INDICE:

Dalle banche dati bibliografiche

ISTITUTO DI RICERCHE FARMACOLOGICHE MARIO NEGRI · IRCCS

GENERALE

Acta Biomed. 2020 Jun;91:48-64.
Transition of care in pediatric oncohematology: a systematic literature review.
Strini V, Daicampi C, Trevisan N, et al.
BACKGROUND: The transition of medical care from a pediatric to an adult environment is a psychological change, a new orientation that requires a self-redefinition of the individual, to understand that changes are taking place in his life. Up to 60 percent of pediatric patients who transition to adult services will experience one or more disease or treatment-related complication as they become adults. A nurse who knows how to recognize potential barriers at an early stage can play a pivotal role in the educational plan for the transition process.
MATERIALS AND METHODS: A literature search was undertaken of PUBMED, CINAHL and The Cochrane Library, with specific inclusion and exclusion criteria, including articles published in the lasts ten years.This literature review has been performed according to the PRISMA statement.
RESULTS: Using the keywords in different combination 38 articles were found in The Cochrane Library, 5877 in PUBMED, 274 in CINAHL. 88 articles were selected after the abstract screening. 31 after removing the duplicates and reading the full text.
DISCUSSION: The main themes surrounding transition of care that emerged from the synthesis are the organization of care within common models of transition, innovative clinical approaches to transition, and the experience of patients and caregivers. The transition from pediatric to adult care of cancer or SCD survivors is an emerging topic in pediatric nursing. The organization of care is affected by the lack of clear and well-structured organizational models. Further research is needed to deepen the understanding of some aspects of the transition

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Per la ricerca degli articoli pubblicati nella letteratura scientifica nel mese in esame sono state consultate le banche dati Medline, Embase e PUBMED utilizzando le seguenti parole chiave (o i loro sinonimi): 'Transitional Care', 'Transitional to Adult Care', 'Children', 'Adolescent', 'Young'. Sono qui riportate le referenze considerate rilevanti e pertinenti.

AIDS Care. 2021 Apr;1-5.

STEPPING UP: RETENTION IN HIV CARE WITHIN AN INTEGRATED HEALTH CARE TRANSITION PROGRAM.

Ryscavage P, Herbert L, Roberts B, et al.

Strategies are needed to optimize HIV health care transition (HCT). We describe HCT outcomes within the University of Maryland STEP Program, which is built upon integration of an adult HIV provider and navigator into the pediatric clinic, and coordinated collaboration between pediatric and adult HIV multi-disciplinary care teams. These outcomes were compared to a historical institutional HCT cohort (Nâ€‰=â€‰50) which attempted transition in an earlier time period (2004-2012). Fifty-eight patients were enrolled during the study period, and 34 attempted HCT. In total, 84 patients underwent attempted HCT. In the STEP cohort, linkage to adult care was 94% and 12 month retention in adult care (95%) was statistically higher compared to the historical cohort. Rates of viral suppression did not differ pre- and post-HCT among STEP Program patients. These results support the concept of an integrated pediatric and adult HIV HCT model though the ability to achieve sustainable HCT success will require further study

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AIDS Behav. 2021 Jan;25:237-48.

DISCLOSURE, SOCIAL SUPPORT, AND MENTAL HEALTH ARE MODIFIABLE FACTORS AFFECTING ENGAGEMENT IN CARE OF PERINATALLY-HIV INFECTED ADOLESCENTS: A QUALITATIVE DYADIC ANALYSIS.

Zanoni BC, Archary M, Subramony T, et al.

Adolescents living with perinatally acquired HIV in South Africa face significant barriers to successful transition from pediatric to adult care. We performed in-depth qualitative interviews with 41 adolescents living with HIV and 18 of their caregivers to investigate modifiable factors to improve engagement in care prior to transition to adult care. Based on dyadic, inductive content analysis, findings suggest that HIV status disclosure, social support, and mental health are targets for improvement in engagement in care. Early disclosure and a sense of belonging facilitated engagement in care, while barriers included delayed or inadequate disclosure, denial, and lack of disclosure to others. Adherence support improved by having a biological mother as a direct supervisor. Barriers to care included changing caregivers, abandonment, undiagnosed mental health problems and learning difficulties. Despite these factors, the majority of adolescents showed resilience and remained engaged in care despite difficult circumstances

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Am J Gastroenterol. 2021 Apr;116:638-46.

ACG CLINICAL REPORT AND RECOMMENDATIONS ON TRANSITION OF CARE IN CHILDREN AND ADOLESCENTS WITH HEREDITARY POLYPOSIS SYNDROMES.

Attard TM, Burke CA, Hyer W, et al.

Transition of care (TOC) in adolescents and young adults (AYAs) with chronic gastrointestinal disorders has received increased attention, especially in those with inflammatory bowel disease. AYAs with hereditary polyposis syndromes are a heterogeneous group of patients with overlapping and complex medical needs. These patients are particularly vulnerable because of the risk of loss of continuity of care and subsequent poor disease outcomes. The Pediatric Committee of the American College of Gastroenterology commissioned a report with recommendations on TOC in AYAs with hereditary polyposis syndromes. This report aims at achieving best practice by both pediatric and adult gastroenterologists despite the paucity of published evidence in this population reflected in the included PRISMA report. Therefore, the group extrapolated findings from the literature related to other chronic gastrointestinal disorders, and a high degree of expert consensus was scored for all recommendations. The report addresses TOC through identifying shared domains followed by specific recommendations in disease management, including models of care, providers and patient and socioeconomic factors relevant to TOC. Areas of strong emphasis include the need for early planning, flexibility in the transition process to maintain continuity during major surgical procedures, patient and family psychological readiness, liaison among team members addressing transition, and changing insurance coverage in this population

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Am J Med Genet C Semin Med Genet. 2021 Mar;187:70-82.

TRANSITION TO VIRTUAL CLINIC: EXPERIENCE IN A MULTIDISCIPLINARY CLINIC FOR DOWN SYNDROME.

Santoro SL, Donelan K, Haugen K, et al.

The COVID-19 pandemic necessitated a rapid transition from in-person office visits to virtual visits in the Down syndrome specialty program at Massachusetts General Hospital (MGH DSP). We describe the clinic transition to virtual visits in April 2020 and reflect on our six-month experience in virtual visits. Clinic metrics were tracked. Electronic survey responses were collected from caregivers attending virtual visits. Input from the MGH DSP team was collected. From April to September 2020, we maintained patient volume (45 visits per month) and overall satisfaction score (6.7 out of 7) following a sudden, unanticipated transition to virtual visits. Survey of

17 caregivers attending virtual visits found that most were equipped with technology, had access to a private location, and most were able to access visit without any limitations. Caregivers appreciated the convenience of virtual visits but sometimes missed the personal connection of an in-person visit. Overall, though, virtual visits were frequently viewed as no different than office visits. Team members identified benefits and challenges of virtual visits, as well as lessons learned from this transition. We were able to maintain multidisciplinary, specialty care with optimal caregiver feedback and sustained number of patient visits

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Arch Dis Child. 2021 May.

MEDICATION DISCREPANCIES IN TRANSITION OF CARE OF HOSPITALISED CHILDREN IN BRAZIL: A MULTICENTRIC STUDY.
Aires-Moreno GT, AlcÃ¢ntara TDS, AraÃºjo DCSA, et al.

OBJECTIVE: To determine the incidence of medication discrepancies in transition points of care of hospitalised children.

DESIGN: A prospective observational multicentre study was carried out between February and August 2019. Data collection consisted of the following steps: sociodemographic data collection, clinical interview with the patient's caregiver, review of patient prescriptions and evaluation of medical records. Medication discrepancies were classified as intentional (documented or undocumented) and unintentional. In addition, discrepancies identified were categorised according to the medication discrepancy taxonomy. Unintentional discrepancies were assessed for potential clinical harm to the patient.

SETTING: Paediatric clinics of four teaching hospitals in Brazil.

PATIENTS: Children aged 1 month-12 years.

FINDINGS: A total of 248 children were included, 77.0% (n=191) patients had at least one intentional discrepancy; 20.2% (n=50) patients had at least one unintended discrepancy and 15.3% (n=38) patients had at least one intentional discrepancy and an unintentional one. The reason for the intentional discrepancy was not documented in 49.6% (n=476) of the cases. The most frequent unintentional discrepancy was medication omission (54.1%; n=66). Low potential to cause discomfort was found in 53 (43.4%) unintentional discrepancies, while 55 (45.1%) had the potential to cause moderate discomfort and 14 (11.5%) could potentially cause severe discomfort.

CONCLUSIONS: Although most medication discrepancies were intentional, the majority of these were not documented by the healthcare professionals. Unintentional discrepancies were often related to medication omission and had a potential risk of causing harm to hospitalised children

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Arch Pediatr. 2021 May;28:257-63.

SMOOTHING THE TRANSITION OF ADOLESCENTS WITH CF FROM PEDIATRIC TO ADULT CARE: PRE-TRANSFER NEEDS.
Genovese VV, Perceval M, Buscarlet-Jardine L, et al.

INTRODUCTION: In France, the cystic fibrosis (CF) care pathway is performed in 45 CF centers, the life expectancy of patients has steadily increased, but to date there are no national recommendations for the transition from pediatric to adult care. The transition to an adult CF center still raises questions about the relevance of its organizational arrangements. The "SAFETIM need" study aimed to identify the organizational needs both of patients and of parents before the transfer to an adult CF center.

METHODS: This was a prospective, observational, multicenter study conducted between July 2017 and December 2018, involving the three CF centers of a regional network in southeastern France. Each adolescent registered with the center and his or her parents were interviewed individually, on the same day, during the 6 months leading up to transfer. They participated in semi-structured interviews during one of their routine consultations at the CF center. The interview manual, based on literature reviews and targeting national recommendations, was tested and validated by the national CF therapeutic education group (GETheM). All interviews were transcribed and checked by two different people, and analyzed by two researchers individually. The results were classified by topic according to content categorization.

RESULTS: Overall, 43 adolescents and 41 parents were interviewed, respectively, who were followed up by CF centers: 14% (n=6) in a mixed CF center (pediatric and adult); 19% (n=8) and 67% (n=29), respectively, in two different pediatric CF centers. Adolescents were between 16 and 19 years old. For adolescents, the average interview time was 5.11min. (standard deviation [SD]: 3.8min; minimum: 2.53min; maximum: 17.14min). For parents, the average interview time was 7.99min (SD: 3.56min, minimum: 3.43min; maximum: 22.50min).

DISCUSSION: Our study enquired only about the preparation and organization of the transfer. We identified three areas of actions matching the needs of adolescents and parents before transfer. The first one is to anticipate team change to prepare follow-up in their future CF center: acquire new skills, consider the future CF center according to the adolescent's curriculum, be involved in the transition process. The second area is to accompany the upcoming change. The care team could help by providing information and support during the start of teenagers' transition toward autonomy. And parents were aware that the CF center change will

reverse roles. They must provide their own knowledge and manage the ambivalence of this as well as letting go. The third one is to announce the transition process and functioning of the future adult CF center, because the transition would require time to find their place (patients and parents) with the new team.
CONCLUSION: The "SAFETIM needs" pre-transfer study results show that we can identify the main criteria to be developed and strengthened, to promote a smooth, high-quality transition from pediatric to adult CF care for patients in France. For most patients, the transition cannot be prepared at the last minute. Caregivers need to develop specific skills in adolescent and young adult care and follow-up. Each team must consider the transition as a normal part of the patient care cycle. While it must be structured, some flexibility must be allowed so as to give everyone the chance to be prepared and to personalize the care

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Arthritis Care Res (Hoboken). 2021 Jan;73:39-47.
CLINICAL TEAM PERSPECTIVES ON THE PSYCHOSOCIAL ASPECTS OF TRANSITION TO ADULT CARE FOR PATIENTS WITH CHILDHOOD-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS.
Bitencourt N, Kramer J, Bermas BL, et al.
OBJECTIVE: The transition from pediatric to adult care for youth with childhood-onset systemic lupus erythematosus (SLE) is a vulnerable period. Adverse outcomes during this transition include gaps in care, unscheduled health care utilization, loss of insurance, and high disease activity. The objective of this study was to examine the clinical care teams' perspective on the psychosocial factors associated with transition outcomes, which are poorly understood in this population.
METHODS: We conducted in-depth interviews with clinical care team members who interact with childhood-onset SLE patients during transfer from pediatric to adult rheumatology. A semistructured interview guide was used to prompt participants' perspectives about the psychosocial factors associated with the transition process for patients with childhood-onset SLE. Audio recordings were transcribed and analyzed using the constant comparative method. We stopped conducting interviews once thematic saturation was achieved.
RESULTS: Thirteen in-depth interviews were conducted. Participants included pediatric rheumatologists (n = 4), adult rheumatologists from both academic and private practice settings (n = 4), nurses (n = 2), a nurse practitioner, a social worker, and a psychologist. We identified several themes deemed by clinical care teams as important during the transition, including the impact of the family, patient resilience and coping mechanisms, the role of mental health and emotional support, and the need for education, peer support, and social connectedness.
CONCLUSION: We identified several psychosocial themes that clinical team members believe impact the transition of patients with childhood-onset SLE into adult care. The role of parental modeling, youth resilience, mental health and emotional care, improved childhood-onset SLE education, and structured peer support and social connectedness are highlighted, which may be amenable to interventions

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Aust J Rural Health. 2021 Feb;29:83-91.
EVALUATION OF A YOUNG ADULT RENAL AND TRANSPLANT TRANSITION CLINIC IN A REGIONAL SETTING: SUPPORTING YOUNG ADULTS AND PARENTS' TRANSITION TO SELF-MANAGEMENT.
Jose K, Le RA, Jeffs L, et al.
OBJECTIVE: This study evaluated the impact of establishing a transition clinic in a regional Australian setting on the lives of young adults living with severe chronic kidney disease and their families.
DESIGN: A qualitative design using the experience-based co-design framework. SETTING: Interviews were held at the Royal Hobart Hospital or the Menzies Institute for Medical Research. The co-design workshop was held at the Royal Hobart Hospital.
PARTICIPANTS: Young people aged 17-29 years living with a kidney transplant or stage 4-5 chronic kidney disease, parents/carers and health professionals.
INTERVENTIONS: Establishment of a young adult renal and transplant clinic.
MAIN OUTCOME MEASURE: Impact of a transition clinic in a regional setting on the lives of young adults living with chronic kidney disease and their families and suggestions for improvement.
RESULTS: Four key themes were identified as follows: The Model of Care; Peer support; Transition towards self-management: Building life skills; Suggestions for improvement and limitations of the service model. The non-institutional, informal clinic setting and social/educational activities facilitated engagement, self-management and peer support for young people and parents. Suggestions for improvement included involvement of older peers, additional life skills sessions and a youth worker.
CONCLUSION: This regional transition clinic is valued by the young people and their parents for generating peer support, building self-management and life skills. Sustainability of the clinic depends upon having the appropriate expertise available, access to a suitable venue and offering a program that meets the needs of young people

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Aust N Z J Public Health. 2021 Feb;45:71-79.

A SOCIOECOLOGICAL DISCOURSE OF CARE OR AN ECONOMISTIC DISCOURSE: WHICH FITS BETTER WITH TRANSITION?

Kay V, Livingstone C.

OBJECTIVE: To analyse a 'socioecological' health promotion discourse and its relationship to orthodox 'economistic' discourse in Australia.

METHOD: In research on health promotion addressing equity and environmental sustainability, we identified a socioecological discourse, based on an ethic of care for people and ecosystems. Using Foucault's concept of discourse as a regime that produces and legitimises certain kinds of knowledge, and ecofeminist historical analysis, we analysed this discourse and its relationship to economism.

RESULTS: The socioecological discourse takes social and ecological wellbeing as primary values, while economism takes production and trade of goods and services, measured by money, as primary. Following British invasion, property-owning white men in Australia had the right to control and profit from land, trade, and the work of women and subordinate peoples. A knowledge regime using money as a primary measure reflects this history. In contrast, a First Nations' primary value expressed in the study was 'look after the land and the children'. Conclusion and implications for public health: Public health often attempts to express value through economism, using monetary measures. However, socioecological discourse, expressed for example through direct measures of social and ecological wellbeing, appears more fit for purpose in promoting a fair and sustainable society

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Autism. 2021 Apr;25:753-66.

DEVELOPMENT AND IMPLEMENTATION OF HEALTH CARE TRANSITION RESOURCES FOR YOUTH WITH AUTISM SPECTRUM DISORDERS WITHIN A PRIMARY CARE MEDICAL HOME.

Harris JF, Gorman LP, Doshi A, et al.

As adolescents become adults, they typically change from seeing a pediatric health care provider to seeing a health care provider who specializes in working with adults. Adolescents with autism spectrum disorder may have more difficulty finding an adult health care provider who is trained to address the varied needs and medical and mental health issues that often are seen with autism spectrum disorder. Without careful planning for transition to adult health care, patients may continue to be seen by pediatric providers who may not be familiar with adult health needs. This quality improvement project focused on improving transition to adult health care by creating varied supports for the patient, family, and the health care team and putting them into action within a pediatric medical practice that serves over 250 adolescent and young adult patients with autism spectrum disorder. Before the supports were put into place, patients and families received limited and inconsistent communication to help them with transition. While the supports helped increase the amount and quality of help patients and families received, medical providers skipped or put off transition discussion in approximately half of well visits for targeted patients. Challenges in implementing the transition process included finding time to discuss transition-related issues with patients/families, preference of medical providers to have social workers discuss transition, and difficulty identifying adult health care providers for patients. This suggests more work is needed to both train and partner with patients, families, and health staff to promote smooth and positive health transitions

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Autism. 2021 Apr;25:731-43.

TRANSITIONING YOUTH WITH AUTISM SPECTRUM DISORDERS AND OTHER SPECIAL HEALTH CARE NEEDS INTO ADULT PRIMARY CARE: A PROVIDER SURVEY.

Ames JL, Massolo ML, Davignon MN, et al.

The transition from pediatric to adult care is a critical inflection point for the long-term health of youth with autism spectrum disorders and other special health care needs. However, for many patients, their caregivers, and providers, the transition lacks coordination. This survey study demonstrates that pediatric and adult providers struggle to implement many components of transition best practices for youth with autism and other chronic conditions, highlighting the urgent need for enhanced medical coordination and additional transition training and resources

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Autism. 2021 Apr;25:705-18.

HEALTHCARE SERVICE UTILIZATION AND COST AMONG TRANSITION-AGE YOUTH WITH AUTISM SPECTRUM DISORDER AND OTHER SPECIAL HEALTHCARE NEEDS.

Ames JL, Massolo ML, Davignon MN, et al.

Youth with autism spectrum disorder often have complex medical needs. Disruptions of healthcare during the transition from pediatric to adult healthcare may put youth with autism spectrum disorder at higher risk of medical emergencies and high medical costs. To understand healthcare utilization during the transition years,

we conducted a study among transition-age youth (14-25years old) receiving healthcare at Kaiser Permanente Northern California during 2014-2015. We examined differences in healthcare utilization and costs among youth with autism spectrum disorder (n=4123), **attention deficit and hyperactivity disorder** (n=20,6015), diabetes mellitus (n=2156), and general population controls (n=20,615). Analyses were also stratified by age and sex. Youth with autism spectrum disorder had the highest utilization of outpatient primary care, mental health, and psychotropic medications and the lowest utilization of obstetrics/gynecology and urgent care. Costs for youth with autism spectrum disorder were higher than those for attention deficit and hyperactivity disorder and general population peers and lower than for diabetes mellitus. Healthcare utilization patterns varied by age. Transition-age youth with autism spectrum disorder generally used healthcare at higher rates relative to attention deficit and hyperactivity disorder and general population peers but at similar or lower rates than diabetes mellitus peers, indicating this group's complex combination of psychiatric and medical healthcare needs. The relatively high utilization of psychiatric services and low utilization of women's health services in transition-age youth with autism spectrum disorder may have implications for long-term health and warrants additional research

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BJGP Open. 2020 Dec;4.

NEW MODELS OF CARE IN GENERAL PRACTICE FOR THE YOUTH MENTAL HEALTH TRANSITION BOUNDARY.

Appleton R, Mughal F, Giacco D, et al.

Blood Purif. 2021 Mar;1-12.

AN INTERDISCIPLINARY APPROACH TO OPTIMIZE THE CARE OF TRANSITIONING ADOLESCENTS AND YOUNG ADULTS WITH CKD.

Diaz-Gonzalez de Ferris ME, D-az-Gonz¡lez de MartÃ-nez ML, D-az-Gonz¡lez de Vel¡zquez AM, et al.

Adolescents and young adults (AYAs) with CKD or end-stage kidney disease (ESKD) have unique medical, dental, psychosocial, neurocognitive, and academic needs and require close interdisciplinary collaboration to optimize their care. The etiology of CKD in AYAs is diverse compared to older adults. With their continuously improved survival, AYAs must start preparation for health-care transition (HCT) from pediatric- to adult-focused health care in the pediatric setting and it must continue at the adult-focused setting, given that their brain maturation and self-management skill acquisition occur until their mid-20s. While the growth and physical maturation of most visible body parts occur before 18 years of age, the prefrontal cortex of the brain, where reasoning, impulse control, and other higher executive functions reside, matures around 25 years of age. The HCT process must be monitored using patient- and caregiver-measuring tools to guide interventions. The HCT process becomes more complex when patients and/or caregivers have a language barrier, different cultural beliefs, or lower literacy levels. In this article, we discuss the unique comorbidities of pediatric-onset CKD/ESKD, provide information for a planned HCT preparation, and suggest interdisciplinary coordination as well as cultural and literacy-appropriate activities to achieve optimal patient outcomes

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BMC Cancer. 2020 Sep;20:898.

IDENTIFYING METRICS OF SUCCESS FOR TRANSITIONAL CARE PRACTICES IN CHILDHOOD CANCER SURVIVORSHIP: A QUALITATIVE INTERVIEW STUDY OF SURVIVORS.

Sadak KT, Gemeda MT, Grafelman M, et al.

Background: Adolescent and young adult (AYA) childhood cancer survivors (CCS) should be empowered to continue their survivor-focused care as they transition into adult medicine. However, the majority of AYA-aged survivors become lost to follow up around the age of typical transition to adulthood. The purpose of this study was to identify, from the patient's perspective, key factors that facilitate successful transitions to adult-centered survivorship care.

Methods: A qualitative study was conducted with AYA CCS (n = 29) from the survivorship clinic of a single institution as key informants. Data were collected through a series of structured phone interviews and subjected to thematic content analysis.

Results: Four major themes with multiple subthemes were identified: (1) transition practices need to be flexible and individually tailored; (2) effective communication is critical to a successful transition; (3) continuity in providers is needed during the transition; and (4) comprehensive care means care that also addresses psycho-social well-being.

Conclusions: From the perspective of AYA CCS, the ideal model of transitional survivorship care could include a patient navigator who promotes provider flexibility, consistent communication, and pro-active comprehensive care that encompasses both medical and psycho-social well-being. Models of care for CCS should be built to provide, or seamlessly facilitate, continuous survivor-focused care across the age continuum.

A longitudinal relationship with a survivor-focused provider can help promote the values that CCS' report as important in transitioning care from pediatric- to adult-centered care

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BMC Fam Pract. 2020 Jul;21:140.

PERSPECTIVES FROM PRIMARY HEALTH CARE PROVIDERS ON THEIR ROLES FOR SUPPORTING ADOLESCENTS AND YOUNG ADULTS TRANSITIONING FROM PEDIATRIC SERVICES.

Schraeder K, Dimitropoulos G, McBrien K, et al.

BACKGROUND: Transitioning from pediatric care to adult-oriented care at age 18 (the age of transfer in most countries and jurisdictions) is a complex process for adolescents and young adults affected by chronic physical health and/or mental health conditions. The role of primary health care (PHC) providers for this population is poorly understood. Perspectives from these providers, such as family physicians and other members of the primary care team, have not been explored in depth.

METHODS: A total of 18 participants (e.g., family physicians, social workers, nurses) were recruited from 6 Primary Care Networks in Calgary, Alberta, Canada. Semi-structured individual interviews were conducted, and transcribed verbatim. A qualitative description approach was used to analyze the data, and included thematic analysis.

RESULTS: Five distinct, yet overlapping, roles of primary health care providers for adolescents and young adults transitioning to adult care resulted from our analysis: (1) being the "common thread" (continuous accessible care); (2) caring for the "whole patient" (comprehensive care); (3) "knowing families" (family-partnered care); (4) "empowering" adolescents and young adults to develop "personal responsibility" (developmentally-appropriate care); and (5) "quarterbacking" care (coordination of specialist and/or community-based care). Participants identified potential benefits of these roles for adolescents and young adults transitioning to adult care, and barriers in practice (e.g., lack of time, having minimal involvement in pediatric care).

CONCLUSIONS: Input from family physicians, who follow their patients across the lifespan and provide the majority of primary care in Canada, are critical for informing and refining recommended transition practices. Our findings provide insights, from PHC providers themselves, to bolster the rationale for primary care involvement during transitions from pediatric specialty and community-based care for AYAs. Solutions to overcome barriers for integrating primary care and specialty care for adolescents and young adults need to be identified, and tested, with input from key stakeholders

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BMC Health Serv Res. 2021 May;21:462.

TRANSITIONING FROM PEDIATRIC TO ADULT CARE AND THE HIV CARE CONTINUUM IN GHANA: A RETROSPECTIVE STUDY.

Abaka P, Nutor JJ.

BACKGROUND: In Sub-Saharan Africa, there are now a significant number of adolescents living with HIV (ALHIV), due to increased access to effective antiretroviral therapy. However, these adolescents are at high risk of dying during the transition to adult care due to various reasons, including lack of preparation for the transition and poor transition arrangements. More knowledge about this issue will lead to a better planned healthcare transition process and preparation for transition from pediatric care to adult care. The aim of this study was to explore the healthcare transitional experiences of ALHIV as they moved from pediatric to adult care.

METHODS: A descriptive exploratory qualitative study was conducted. Purposive sampling method was used to recruit adolescents between 12 and 19 years old. Saturation was realized by the 10th participant. Data were analyzed using thematic content analysis.

RESULTS: Four main themes emerged from the interview data: the transition process, factors facilitating the transition experience, challenges and coping mechanisms of the ALHIV during transition, and suggestions for improvement based on perceptions on the current transitioning approach. A key finding of this study was the sudden preparation for transition, linked to the absence of a structured transition protocol. Even though age was the main reason for transferring the participants from the pediatric to adult clinic, participants' age did not influence whether they attended clinic appointment on their own or accompanied by a care provider; it was dependent on the availability of their parents or caregivers. Participants' parents and adult family caregivers were also integrated into the transition process to some extent. We also found that most of the participants had good patient-provider relationship with their health care providers in both pediatric and adult clinics.

CONCLUSION: Findings support the need to develop a structured healthcare transition policy and age-appropriate transition within the clinic environment. There is also a need for social and community support as ALHIV transition from pediatric to adult care

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BMC Med Ethics. 2021 Mar;22:35.

ETHICAL CHALLENGES OF THE HEALTHCARE TRANSITION TO ADULT ANTIRETROVIRAL THERAPY (ART) CLINICS FOR ADOLESCENTS AND YOUNG PEOPLE WITH HIV IN UGANDA.

Mbalinda SN, Bakeera-Kitaka S, Amooti DL, et al.

BACKGROUND: Whereas many adolescents and young people with HIV require the transfer of care from paediatric/adolescent clinics to adult ART clinics, this transition is beset with a multitude of factors that have the potential to hinder or facilitate the process, thereby raising ethical challenges of the transition process. Decisions made regarding therapy, such as when and how to transition to adult HIV care, should consider ethical benefits and risks. Understanding and addressing ethical challenges in the healthcare transition could ensure a smooth and successful transition. The purpose of this study was to analyze the ethical challenges of transitioning HIV care for adolescents into adult HIV clinics.

METHODS: Data presented were derived from 191 adolescents attending nine different health facilities in Uganda, who constituted 18 focus group discussions. In the discussions, facilitators and barriers regarding adolescents transitioning to adult HIV clinics were explored. Guided by the Silences Framework for data interpretation, thematic data analysis was used to analyze the data. The principles of bioethics and the four-boxes ethics framework for clinical care (patient autonomy, medical indications, the context of care, and quality of life) were used to analyze the ethical issues surrounding the transition from adolescent to adult HIV care.

RESULTS: The key emerging ethical issues were: reduced patient autonomy; increased risk of harm from stigma and loss of privacy and confidentiality; unfriendly adult clinics induce disengagement and disruption of the care continuum; patient preference to transition as a cohort, and contextual factors are critical to a successful transition.

CONCLUSION: The priority outcomes of the healthcare transition for adolescents should address ethical challenges of the healthcare transition such as loss of autonomy, stigma, loss of privacy, and discontinuity of care to ensure retention in HIV care, facilitate long-term self-care, offer ongoing all-inclusive healthcare, promote adolescent health and wellbeing and foster trust in the healthcare system. Identifying and addressing the ethical issues related to what hinders or facilitates successful transitions with targeted interventions for the transition process may ensure adolescents and young people with HIV infection remain healthy across the healthcare transition

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BMC Public Health. 2020 Aug;20:1195.

HEALTHCARE RETENTION AND CLINICAL OUTCOMES AMONG ADOLESCENTS LIVING WITH HIV AFTER TRANSITION FROM PEDIATRIC TO ADULT CARE: A SYSTEMATIC REVIEW.

Ritchwood TD, Malo V, Jones C, et al.

Background: Adolescents living with HIV (ALWH) who transition from pediatric to adult care face several challenges that increase their risk of experiencing treatment interruptions and being lost to HIV care with resultant increased morbidity and mortality. To date, few studies have examined their outcomes post-healthcare transition (HCT), precluding the development and dissemination of evidence-based interventions aimed at retaining ALWH in HIV care both during and after HCT. We conducted a systematic review to synthesize the outcomes of ALWH post-HCT to provide suggestions for future directions.

Methods: We systematically searched several electronic databases through October 2019 using keywords for HIV, HCT and ALWH. We categorized studies by target population, country (i.e., upper-high income and low-middle income), study design (i.e., descriptive, mixed methods, quantitative), outcomes measured, and follow-up period.

Results: A total of 24 studies met inclusion criteria. Studies were categorized according to the following HCT outcomes: retention in HIV care post-HCT (n = 13), changes in CD4+ count and viral load post-HCT (n = 16), and mortality among ALWH post-HCT (n = 7). Most studies (n = 11) examining retention in HIV care indicated that more than 70% of ALWH were retained in care 1-2 years post-HCT while the remaining studies (n = 2) reported retention rates less than 55%. While studies indicated that CD4+ counts and viral loads tended to worsen during the first few years post-HCT, these differences were often not statistically significant. Among all ALWH who transitioned to adult care, a small proportion died within their first seven years post-HCT. Among qualitative studies, common themes included transition readiness (n = 6), provider-patient relationship in the adult clinic setting (n = 6), and concern about the adult clinic setting (n = 4).

Conclusions: Transition outcomes were poorest for ALWH with unsuppressed viremia pre-HCT, suggesting that this subgroup of ALWH may need greater support from their treatment teams and caregivers during and post-HCT to improve clinical outcomes

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BMJ Open. 2020 Jun;10:e033978.

PROGRAMMES TO SUPPORT TRANSITIONS IN CARE FOR CHILDREN AND YOUTH WITH COMPLEX CARE NEEDS AND THEIR FAMILIES: A SCOPING REVIEW PROTOCOL.

Doucet S, Curran JA, Breneol S, et al.

INTRODUCTION: Children and youth with complex care needs (CCNs) and their families experience many care transitions over their lifespan and are consequently vulnerable to the discontinuity or gaps in care that can occur during these transitions. Transitional care programmes, broadly defined as one or more intervention(s) or service(s) that aim to improve continuity of care, are increasingly being developed to address transitions in care for children and youth with CCNs. However, this literature has not yet been systematically examined at a comprehensive level. The purpose of this scoping review is to map the range of programmes that support transitions in care for children and youth with CCNs and their families during two phases of their lifespan: (1) up to the age of 19 years (not including their transition to adult healthcare) and (2) when transitioning from paediatric to adult healthcare.

METHODS AND AND ANALYSIS: The Joanna Briggs Institute methodology for scoping reviews (ScR) will be used for the proposed scoping review. ScR are a type of knowledge synthesis that are useful for addressing exploratory research questions that aim to map key concepts and types of evidence on a topic and can be used to organise what is known about the phenomena. A preliminary search of PubMed was conducted in December 2018.

ETHICS AND DISSEMINATION: Ethical approval is not required where this study is a review of the published and publicly reported literature. The research team's advisory council will develop a research dissemination strategy with goals, target audiences, expertise/leadership, resources and deadlines to maximise project outputs. The end-of-grant activities will be used to raise awareness, promote action and inform future research, policy and practice on this topic

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BMJ Open. 2020 Nov;10:e036573.

DEVELOPMENT OF AN INFORMATIONAL SUPPORT QUESTIONNAIRE OF TRANSITIONAL CARE FOR AGED PATIENTS WITH CHRONIC DISEASE.

Shi X, Geng G, Hua J, et al.

OBJECTIVES: We developed an informational support questionnaire of transitional care (ISQTC) for aged patients with chronic disease and investigated its reliability and validity. SETTING: This study was conducted in three large general hospitals in Nantong, Jiangsu Province, China.

PARTICIPANTS: A total of 130 aged patients with chronic diseases, admitted into outpatient and inpatient departments from three hospitals in China, participated in the study. The inclusion criteria were: (1) patients must provide consent to participate; (2) being 60 years and above; (3) being diagnosed with at least one chronic disease and hospitalised more than two times within the last 1â€‰year; (4) being able to listen, speak, read and write. The exclusion criteria were: (1) refusing to participate; (2) language expression and communication barriers (and having no caregiver to assist in participation); (3) being in intensive care or long-term hospitalisation.

PRIMARY AND SECONDARY OUTCOME MEASURES: The developed questionnaire was validated and tested for reliability. The content validity of the questionnaire was determined through experts' interviews and Delphi expert consultation, and the structure validity of the questionnaire was determined by performing exploratory factor analysis. The coefficient of reliability of the questionnaire was measured using Cronbach's alpha.

RESULTS: Through Delphi expert consultation and exploratory factor analysis, the questionnaire was reduced from four dimensions and 12 items to three dimensions and 11 items. A total of 130 patients responded to the questionnaire. The alpha coefficient was 0.747.

CONCLUSION: The ISQTC is a reliable and valid instrument for evaluating aged patients with chronic disease in transitional care.

TRIAL REGISTRATION DETAILS: ChiCTR1900020923. The trial was registered on 22 January 2019

BMJ Open. 2020 Dec;10:e039972.

ADOLESCENT TRANSITION TO ADULT CARE FOR HIV-INFECTED ADOLESCENTS IN KENYA (ATTACH): STUDY PROTOCOL FOR A HYBRID EFFECTIVENESS-IMPLEMENTATION CLUSTER RANDOMISED TRIAL.

Njuguna IN, Beima-Sofie K, Mburu CW, et al .

Introduction: Successfully transitioning adolescents to adult HIV care is critical for optimising outcomes. Disclosure of HIV status, a prerequisite to transition, remains suboptimal in sub-Saharan Africa. Few interventions have addressed both disclosure and transition. An adolescent transition package (ATP) that combines disclosure and transition tools could support transition and improve outcomes.

Methods and analysis: In this hybrid type 1 effectiveness-implementation cluster randomised controlled trial, 10 HIV clinics with an estimated ≥100 adolescents and young adults age 10-24 living with HIV (ALWHIV) in Kenya will be randomised to implement the ATP and compared with 10 clinics receiving standard of care. The ATP includes provider tools to assist disclosure and transition. Healthcare providers at intervention clinics will receive training on ATP use and support to adapt it through continuous quality improvement cycles over the initial 6 months of the study, with continued implementation for 1 year. The primary outcome is transition readiness among ALWHIV ages 15-24 years, assessed 6 monthly using a 22-item readiness score. Secondary outcomes including retention and viral suppression among ALWHIV at the end of the intervention period (month 18), implementation outcomes (acceptability, feasibility, fidelity, coverage and penetration) and programme costs complement effectiveness outcomes. The primary analysis will be intent to treat, using mixed-effects linear regression models to compare transition readiness scores (overall and by domain (HIV literacy, self-management, communication, support)) over time in control and intervention sites with adjustment for multiple testing, accounting for clustering by clinic and repeated assessments. We will estimate the coefficients and 95% CIs with a two- sided α=0.05.
Ethics and dissemination: The study was approved by the University of Washington Institutional Review Board and the Kenyatta National Hospital Ethics and Research Committee. Study results will be shared with participating facilities, county and national policy-makers.
Trials registration number: NCT03574129; Pre-results

BMJ Open. 2021 Jan;11:e039670.

PATIENT AND PARENT PERSPECTIVES ON TRANSITION FROM PAEDIATRIC TO ADULT HEALTHCARE IN RHEUMATIC DISEASES: AN INTERVIEW STUDY.

Jiang I, Major G, Singh-Grewal D, et al.

OBJECTIVES: To describe the experiences, priorities, and needs of patients with rheumatic disease and their parents during transition from paediatric to adult healthcare. SETTING: Face-to-face and telephone semistructured interviews were conducted from December 2018 to September 2019 recruited from five hospital centres in Australia.
PARTICIPANTS: Fourteen young people and 16 parents were interviewed. Young people were included if they were English speaking, aged 14-25 years, diagnosed with an inflammatory rheumatic disease (eg, juvenile idiopathic arthritis, juvenile dermatomyositis, systemic lupus erythematosus, panniculitis, familial Mediterranean fever) before 18 years of age. Young people were not included if they were diagnosed in the adult setting.
RESULTS: We identified four themes with respective subthemes: avoid repeat of past disruption (maintain disease stability, preserve adjusted personal goals, protect social inclusion); encounter a daunting adult environment (serious and sombre mood, discredited and isolated identity, fear of a rigid system); establish therapeutic alliances with adult rheumatology providers (relinquish a trusting relationship, seek person-focused care, redefine personal-professional boundaries, reassurance of alternative medical supports, transferred trust to adult doctor) and negotiate patient autonomy (confidence in formerly gained independence, alleviate burden on patients, mediate parental anxiety).
CONCLUSIONS: During transition, patients want to maintain disease stability, develop a relationship with their adult provider centralised on personal goals and access support networks. Strategies to comprehensively communicate information between providers, support self-management, and negotiate individualised goals for independence during transition planning may improve satisfaction, and health and treatment outcomes

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BMJ Paediatr Open. 2021;5:e001059.

PAEDIATRIC TO ADULT HEALTHCARE TRANSITION IN RESOURCE-LIMITED SETTINGS: A NARRATIVE REVIEW.

Narla NP, Ratner L, Bastos FV, et al.

BACKGROUND: Ageing into adulthood is challenging at baseline, and doing so with a chronic disease can add increased stress and vulnerability. Worldwide, a substantial care gap exists as children transition from care in a paediatric to adult setting. There is no current consensus on safe and equitable healthcare transition (HCT) for patients with chronic disease in resource-denied settings. Much of the existing literature is specific to HIV care. The objective of this narrative review was to summarise current literature related to adolescent HCT not associated with HIV, in low-income and middle-income countries (LMICs) and other resource-denied settings, in order to inform equitable health policy strategies.
METHODS: A literature search was performed using defined search terms in PubMed and Cumulative Index to Nursing and Allied Health Literature databases to identify all peer-reviewed studies published until January 2020, pertaining to paediatric to adult HCT for adolescents and young adults with chronic disease in resource-denied settings. Following deduplication, 1111 studies were screened and reviewed by two independent

reviewers, of which 10 studies met the inclusion criteria. Resulting studies were included in thematic analysis and narrative synthesis.
RESULTS: Twelve subthemes emerged, leading to recommendations which support equitable and age-appropriate adolescent care. Recommendations include (1) improvement of community health education and resilience tools for puberty, reproductive health and mental health comorbidities; (2) strengthening of health systems to create individualised adolescent-responsive policy; (3) incorporation of social and financial resources in the healthcare setting; and (4) formalisation of institution-wide procedures to address community-identified barriers to successful transition.
CONCLUSION: Limitations of existing evidence relate to the paucity of formal policy for paediatric to adult transition in LMICs for patients with childhood-onset conditions, in the absence of a diagnosis of HIV. With a rise in successful treatments for paediatric-onset chronic disease, adolescent health and transition programmes are needed to guide effective health policy and risk reduction for adolescents in resource-denied settings

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Br J Haematol. 2021 Mar;192:1082-91.
COGNITIVE PERFORMANCE AS A PREDICTOR OF HEALTHCARE TRANSITION IN SICKLE CELL DISEASE.
Saulsberry-Abate AC, Partanen M, Porter JS, et al.
Neurocognitive deficits in sickle cell disease (SCD) may impair adult care engagement. We investigated the relationship between neurocognitive functioning and socio-environmental factors with healthcare transition outcomes. Adolescents aged 15-18 years who had neurocognitive testing and completed a visit with an adult provider were included. Transition outcomes included transfer interval from paediatric to adult care and retention in adult care at 12 and 24 months. Eighty adolescents (59% male, 64% HbSS/HbSβ0 -thalassaemia) were included. Mean age at adult care transfer was 18·0 (±0·3) years and transfer interval was 2·0 (±2·3) months. Higher IQ (P = 0·02; PFDR = 0·05) and higher verbal comprehension (P = 0·008; PFDR = 0·024) were associated with <2 and <6 month transfer intervals respectively. Better performance on measures of attention was associated with higher adult care retention at 12 and 24 months (P = 0·009; PFDR = 0·05 and P = 0·04; PFDR = 0·12 respectively). Transfer intervals <6 months were associated with smaller households (P = 0·02; PFDR = 0·06) and households with fewer children (P = 0·02; PFDR = 0·06). Having a working parent was associated with less retention in adult care at 12 and 24 months (P = 0·01; P = 0·02 respectively). Lower IQ, verbal comprehension, attention difficulties and environmental factors may negatively impact transition outcomes. Neurocognitive function should be considered in transition planning for youth with SCD

Bundesgesundheitsblatt Gesundheitsforschung Gesundheitsschutz. 2020 Jul;63:905-09.
CHALLENGES IN THE CARE OF DISABLED PEOPLE IN THE TRANSITION FROM PEDIATRICS TO ADULT MEDICINE.
Fricke C.
During the transition from adolescence to adulthood, people with disabilities have special problems and needs for adequate medical care and its integration into the overall environment. With the introduction of paragraphs 119c and 43b SGBÂ V, the conditions for medical centers for people with disabilities (MZEB) were created and the special needs of this group of people recognized, while at the same time contributing to the implementation of Art.Â 25 of the United Nations Convention on the Rights of Persons with Disabilities. However, due to access restrictions, the MZEB can only provide adequate care for some of the people with disabilities. Regular conditions also need to be created so that the group of people with disabilities can receive adequate healthcare. The outcome of new care concepts should be confirmed by suitable studies

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Chest. 2021 May.
HOW I DO IT: TRANSITIONING ASTHMA CARE FROM ADOLESCENTS TO ADULTS.
Nanzer AM, Lawton A, D'Ancona G, et al.
Children with asthma grow to become adults with asthma. Adolescents are not simply older children and do not automatically transform into independent adults, nor do they become proficient in self-management of their condition overnight. Adolescence is a high-risk time for many people with asthma, with increased risk of asthma-related morbidity and mortality. Children with high-risk asthma attend hospital-based asthma clinics with their parents until they reach young adulthood and parents usually take on the significant burden of disease management on behalf of their children. Once patients are transferred to adult medical teams, many will continue to have limited knowledge about their asthma, limited understanding of how to manage their symptoms and co-morbidities, and limited comprehension of how and why to take their regular medication. Adolescence is a critical time of change during which young people yearn for autonomy. Effective transition gives young people the skills and knowledge necessary to manage their health independently and provides the substrate for autonomous care, the bed rock of long-term conditions. This review focusses on the

challenges of adolescent health care and provides guidance on how to take a planned, patient centred approach to ensure each transition is effective and safe

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Child Abuse Negl. 2021 May;105104.

A COLLECTIVE IMPACT APPROACH TO SUPPORTING YOUTH TRANSITIONING OUT OF GOVERNMENT CARE.

Smith A, Peled M, Martin S.

BACKGROUND: The issues faced by young people transitioning out of government care are complex, and improving outcomes requires the collaboration of multiple stakeholders (Lopez & Allen, 2007).

OBJECTIVE, PARTICIPANTS, AND SETTING: In Vancouver, Western Canada, 60 agencies and 20 youth from government care are working in partnership using a collective impact approach to address the systemic issues and barriers to healthy development that youth from care experience. Collective impact is an approach to tackling complex social problems which requires collaboration across government, business, funders, charitable organizations, and community members to achieve significant and lasting social change (Hanleybrown et al., 2012). The Vancouver collective operates working groups (co-chaired by youth with care experience) that are addressing challenges in the areas of education, employment, housing, meaningful connections (e.g., sustainable family-type supports), health, wellness and culture.

METHODS: A mixed-method evaluation has included quantitative and qualitative data, collected through outcomes, diaries, surveys, and focus groups, to measure process and outcomes.

RESULTS: Findings help to inform the ongoing development and activities of the collective. Findings indicate the collective is making progress in meeting its goals. Partners continue to be engaged and committed to improving outcomes for youth transitioning out of care, and there are improvements in a number of key areas such as education, collaborative working, and youth engagement.

CONCLUSION: A collective impact approach that includes youth's voices, and collaboration across multiple stakeholders, can increase the likelihood of improving outcomes for young people transitioning out of government care

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Child Care Health Dev. 2021 Apr.

GETTING READY FOR TRANSITION TO ADULT CARE: TOOL VALIDATION AND MULTI-INFORMANT STRATEGY USING THE TRANSITION READINESS ASSESSMENT QUESTIONNAIRE IN PEDIATRICS.

Chapados P, Aramideh J, Lamore K, et al.

Background: Transitioning from pediatric to adult healthcare can be challenging and lead to severe consequences if done suboptimally. The Transition Readiness Assessment Questionnaire (TRAQ) was developed to assess adolescent and young adult (AYA) patients' transition readiness. In this study, we aimed to (1) document the psychometric properties of the French-language version of the TRAQ (TRAQ-FR), (2) assess agreements and discrepancies between AYA patients' and their primary caregivers' TRAQ-FR scores, and (3) identify transition readiness contributors.

Methods: French-speaking AYA patients (n = 175) and primary caregivers (n = 168) were recruited from five clinics in a tertiary Canadian hospital and asked to complete the TRAQ-FR, the Pediatric Quality of Life Inventory™ 4.0 (PedsQL™ 4.0), and a sociodemographic questionnaire. The validity of the TRAQ-FR was assessed using confirmatory factor analyses (CFA). Agreements and discrepancies were evaluated using intraclass correlation coefficients and paired-sample t tests. Contributors of transition readiness were identified using regression analyses.

Results: The five-factor model of the TRAQ was supported, with the TRAQ-FR global scale showing good internal consistency for both AYA patients' and primary caregivers' scores (α = .85-.87). AYA patients and primary caregivers showed good absolute agreement on the TRAQ-FR global scale with AYA patients scoring higher than primary caregivers (ICC = .80; d = .25). AYA patients' age and sex were found to be contributors of transition readiness.

Conclusions: The TRAQ-FR was found to have good psychometric properties when completed by both AYA patients and primary caregivers. Additional research is needed to explore the predictive validity and clinical use of the TRAQ-FR

Clin Infect Dis. 2020 Jun;71:133-41.

HEALTHCARE TRANSITION OUTCOMES AMONG YOUNG ADULTS WITH PERINATALLY ACQUIRED HUMAN IMMUNODEFICIENCY VIRUS INFECTION IN THE UNITED STATES.

Tassiopoulos K, Huo Y, Patel K, et al.

BACKGROUND: Young adults with perinatally acquired HIV (YPHIVs) living in the United States are transitioning to adult clinical care, yet there is little information on factors that affect transition outcomes.

METHODS: YPHIVs aged â‰¥18 years in the Pediatric HIV/AIDS Cohort Study (PHACS) AMP Up cohort approaching or having completed transition from pediatric to adult healthcare were included. Demographic and clinical characteristics and self-reported ability to self-manage healthcare were compared by transition status, and multivariable logistic regression models examined factors associated with satisfaction with, and retention in, adult clinical care (clinic visit within the previous 6 months).
RESULTS: Most of the 455 YPHIVs, regardless of transition status, reported satisfaction with their clinic and care provider, but many reported antiretroviral medication nonadherence. Of the 124 YPHIVs who had transitioned, 56% had periods of unsuppressed HIV-1 RNA in the year before transition. Those who had transitioned were more likely to report high ability to self-manage their healthcare (ability to manage â‰¥7 of 8 skills) than those not transitioned. High self-management was associated with retention after transition (odds ratio, 3.40; 95% confidence interval, 1.33-9.12). Higher perceived emotional social support was also associated with retention. Older age at transition was associated with greater satisfaction with provider and clinic.
CONCLUSIONS: YPHIVs have positive associations with their clinical care around the time of their transition to adult care, but unsuppressed viral load and suboptimal adherence are a concern. Strengthening skills that increase ability to self-manage care and enhance social support may increase retention in care and improve clinical health

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Clin Invest Med. 2020 Jun;43:E14-E23.
OPTIMIZING CHILDHOOD ONCOLOGY CARE TRANSITION FROM PEDIATRIC TO ADULT SETTINGS: A SURVEY OF PRIMARY CARE PHYSICIANS' AND RESIDENTS' PERSPECTIVES.
Marcoux SMMP, LaverdiÃ¨re C.
PURPOSE: The majority of childhood cancer survivors suffer from late adverse effects after the completion of treatment. The prospect of most survivors reaching middle-age is a relatively new phenomenon, and the ways by which current and future primary care physicians (PCPs) will address this novel public health challenge are uncertain.
METHODS: A survey assessing knowledge level and information delivery preferences regarding long-term follow-up guidelines for adult patients having survived a childhood cancer was distributed by e-mail through the Quebec (Canada) national associations of PCPs and residents (n=238).
RESULTS: Participants reported an estimated average of 2.9 ± 1.9 cancer survivors in their yearly caseload, and only 35.3% recalled having provided services to at least one survivor in the last year. Most participants indicated ignoring validated follow-up guidelines for these patients (average score 1.66 on a Likert scale from "1-totally disagreeing" to "5-totally agreeing"). Scarce access to personalized follow-up guidelines and lack of clinical exposure to cancer survivors were identified as main obstacles in providing optimal care to these patients (respective averages of 1.66 and 1.84 on a Likert scale from "1- is a major obstacle" to "5-is not an obstacle at all").
CONCLUSION: The PCPs and residents rarely provide care for childhood cancer adult survivors. On an individual basis, there is a clear need for increased awareness, education and collaboration regarding long-term care of childhood cancer adult survivors during medical training. On a more global basis, structural, organizational and cultural changes are also needed to ensure adequate care transition

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Clin J Oncol Nurs. 2020 Aug;24:451-54.
YOUNG ADULTS WITH SICKLE CELL DISEASE: CHALLENGES WITH TRANSITION TO ADULT HEALTH CARE.
Varty M, Popejoy LL.
Because life expectancy has increased greatly in the past few decades for individuals living with sickle cell disease (SCD), transition to the adult healthcare setting has become a necessity to continue disease management. Transition for young adults with SCD is associated with declining health outcomes, including increased acute care use and mortality. Nurses can assist young adults with SCD who are at risk after transition by assessing the young adult's ability to carry out disease self-management, facilitating the supportive role of the family, and recognizing young adults who may have difficulty accessing healthcare resources and providers

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Curr Gastroenterol Rep. 2021 Jan;23:3.
TRANSITION OF PEDIATRIC LIVER TRANSPLANT PATIENTS TO ADULT CARE: A REVIEW.
Stevens JP, Hall L, Gupta NA.
PURPOSE OF REVIEW: Many pediatric liver transplant patients are surviving to adulthood, and providers have come to recognize the importance of effectively transitioning these patients to an adult hepatologist. The review aims to analyze the most recent literature regarding patient outcomes after transition, barriers to

successful transition, recommendations from clinicians and medical societies regarding transition programs, and to provide personal insights from our experience in transitioning liver transplant recipients.
RECENT FINDINGS: While results were variable between studies, many recent reports show significant morbidity and mortality in patients following transition to adult care. Medical non-adherence is frequently seen in adolescents and young adults both prior to and after transition, and is consistently associated with higher rates of rejection, graft loss, and death. In general, transplant programs with a formal transition process had better patient outcomes though recent findings are mostly-single center and direct comparison between programs is difficult. Societal recommendations for how to create a transition program contain a number of common themes that we have categorized for easier understanding. Successful transition is vital to the continued health of pediatric liver transplant patients. While an effective transition program includes a number of key components, it should be individualized to best function within a given transplant center. Here, we have reviewed a number of recent single-center retrospective studies on transition, but multi-site retrospective or prospective data is lacking, and is a fertile area for future research

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Curr Opin Pediatr. 2020 Jun;32:446-52.
AUTISM: CONSIDERATIONS FOR TRANSITIONS OF CARE INTO ADULTHOOD.
Enner S, Ahmad S, Morse AM, et al.
PURPOSE OF REVIEW: The steady rise in number of youth diagnosed with autism spectrum disorder (ASD) has led to the need to examine transition of care considerations specific to ASD. Improved understanding and guidance addressing these needs will allow pediatric and adult providers to work together to optimize social, medical, and occupational outcomes for these patients.
RECENT FINDINGS: Health-care transition is a delicate time when children with ASD outgrow the services of pediatric programs and enter a fragmented healthcare system that is unfamiliar, insufficiently knowledgeable, and underfunded for their needs.
SUMMARY: Increasing autism prevalence and an aging population with autism lend urgency to improve outcomes in children transitioning to adult-care. Research reveals poor consequences in social support, education, vocational training and employment, housing, and healthcare. Specific considerations to address these issues and ensure successful transition from pediatric to adult care are needed

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Dev Med Child Neurol. 2021 May;63:560-65.
DEVELOPMENT OF THE FEARLESS, TEARLESS TRANSITION MODEL OF CARE FOR ADOLESCENTS WITH AN INTELLECTUAL DISABILITY AND/OR AUTISM SPECTRUM DISORDER WITH MENTAL HEALTH COMORBIDITIES.
Culnane E, Loftus H, Efron D, et al.
AIM: First, to understand the barriers to achieving effective transition and the supports required from the perspective of parents and carers, adolescents with intellectual disability and/or autism spectrum disorder and co-existing mental health disorders (often termed 'dual disability'), and those who provide services to this group. Second, to develop an informed model of shared care to improve the transition of adolescents with dual disabilities.
METHOD: Carers and a young adult with a dual disability were surveyed about their experience of transition care. Other key stakeholders including paediatricians, general practitioners, and policy makers were also interviewed. These data informed the model of care.
RESULTS: Paediatricians and general practitioners reported difficulties establishing working relationships to foster smooth transitions, and carers reported lacking a regular general practitioner with adequate expertise to care for people with dual disabilities. A process of shared care between paediatricians and general practitioners was developed and initiated by a dedicated transition manager, who assisted with care coordination and service linkages. Standardized clinical assessment tools were also introduced to determine patient and carer support needs.
INTERPRETATION: This study highlights the potential to improve transition outcomes for adolescents with dual disabilities and their carers through early transition planning, consistent methods of assessing patient and carer needs, and shared care.
WHAT THIS PAPER ADDS: Adolescents with co-occurring disabilities require a collaborative health and disability service interface. Fearless, Tearless Transition is a new approach to transitioning adolescents with dual disabilities from paediatric to adult care. Carers of adolescents with dual disabilities require support navigating and negotiating services. Engaging general practitioners and paediatricians in shared care early during the transition process is essential

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Digestion. 2021;102:18-24.

TRANSITIONAL CARE FOR PATIENTS WITH INFLAMMATORY BOWEL DISEASE: JAPANESE EXPERIENCE.

Kumagai H, Suzuki Y, Shimizu T.

BACKGROUND: Transition-related healthcare intervention has recently been receiving worldwide attention. Given the increasing incidence of pediatric inflammatory bowel disease (IBD) and its lifelong impact, transitional care has become an important issue. In Japan, guidelines to support the autonomy of IBD patients during transition were recently published.

SUMMARY: Here, we review current issues regarding care for IBD patients during transition from the pediatric to adult period in order to identify the barriers and key elements for successful transition in the context of the Japanese system. Although no single optimal model of transitional care exists, crucial elements identified so far include a joint pediatric/adult clinic or alternating visits between pediatric and adult healthcare providers, a multidisciplinary approach, and good coordination among stakeholders. Self-reliance and independence of patients with childhood-onset IBD are also considered essential for successful transition. Various tools for assessment of transition readiness have been validated and are considered useful. Better outcomes are expected for individually tailored transition, including improvements in medication adherence, perceived health status, quality of life, and self-management. The timing of transfer from a pediatric to an adult gastroenterologist should not be fixed because the issue is not chronological age but rather the degree of individual maturity. We also propose a standardized medical summary with a checklist template for official referral of patients from a pediatric to an adult gastroenterologist. Key Messages: Transition programs require a multidisciplinary approach with a coordinator (IBD nurse) and optimal collaboration and communication. Lack of resources and funding are also pertinent issues

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Epilepsy Behav. 2020 Oct;111:107242.

PILOT DATA AND CASE EXAMPLE OF THE INITIAL VISIT IN A MULTIDISCIPLINARY TRANSITION-AGE PROGRAM (TAP).

Hughes-Scalise A, Reger KL, Gergen MA.

The process of transition from pediatric to adult epilepsy care has received increased attention and emphasis in recent literature, particularly related to the assertion that effective transition is likely to lead to improved medical and psychosocial outcomes. However, the majority of current transition literature focuses on the structure of a transition program, with very little research providing relevant clinical data during the transition period and beyond. The current paper attempts to address this gap in the literature by providing pilot data on participants who engaged in the initial visit of a multidisciplinary transition-focused program housed in a level 4 epilepsy center in the Midwest. Pilot data are presented on 28 participants (36% female) who completed the initial transition appointment. All but one participant presented with a positive history for a neurobehavioral comorbidity, the most common of which included anxiety (61%), attention-deficit/hyperactivity disorder (ADHD; 39%) and depression (36%). Seventy-seven percent of participants further identified a current neurobehavioral comorbidity that was impacting their psychosocial functioning. Recommendations provided most frequently involved increased independence with epilepsy management (64%), increased independence with self-care/independent living (82%), psychological intervention (43%), and increased socialization (43%). A case example is also provided to further highlight program process and outcomes of the initial visit. Pilot results emphasize the value of multidisciplinary care involving psychosocial providers to facilitate a smooth transition between pediatric and adult healthcare

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Epilepsy Behav. 2020 Aug;109:107096.

BARRIERS TO TRANSITION FROM PEDIATRIC TO ADULT CARE FOR PATIENTS WITH DRAVET SYNDROME: A FOCUS GROUP STUDY OF CAREGIVERS.

Boyce DM, Devinsky O, Meskis MA.

Caregivers of individuals with intellectual and developmental disabilities and epilepsy such as Dravet syndrome (DS) must navigate a complex web of state and community services through the transition from child-centered to adult-oriented healthcare. This study examined barriers to successful transition from the caregivers' perspective. Primary caregivers of teenagers or adults with DS who had contemplated or completed transition to adult care were eligible. A three-week, asynchronous, web-based focus group was conducted on Facebook. Data were analyzed in an iterative process based on a Grounded Theory approach. Participants reviewed findings for accuracy. Transition success was defined by how well it ensured adequate care for the child when caregivers became unable to provide it. Existing transition programs were described as "not for our kids." All caregivers reported that transition programs began too late. Challenges to identifying suitable providers were formidable, with 71% of adult patients still being seen by pediatric neurologists. Many adult physicians lacked a general knowledge of DS, yet caregivers perceived that adult physicians were unwilling to listen to caregivers, and few were comfortable accommodating patients with intellectual disabilities and challenging behaviors. Community programs often excluded patients with DS, and rural healthcare disparities created

additional barriers. Analysis produced recommendations for improving the transition process including the creation of a certified Transition Navigator position in the clinical setting. The limitations of this focus group analysis include possible selection bias, but our study identified key issues and pathways to improve the transition process for patients with DS and their caregivers

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Eur J Pediatr. 2021 Jun;180:1817-23.

ADHERENCE TO GLUTEN-FREE DIET AND FOLLOW-UP OF PEDIATRIC CELIAC DISEASE PATIENTS, DURING CHILDHOOD AND AFTER TRANSITION TO ADULT CARE.

Kori M, Goldstein S, Hofi L, et al.

Long-term data on pediatric celiac disease (CD) patients after transition to adult care is scarce. We aimed to evaluate patients' adherence to a gluten-free diet (GFD), the normalization of celiac serology and the frequency of follow-up before age 18, and to study changes in adherence and follow-up frequency after transition to adult care. Presenting symptoms, serology and biopsy results, patients' reported GFD adherence, frequency of follow-up visits, and complications before and after 18 years were collected for CD patients diagnosed between 1998 and 2017. Of 441 CD patients diagnosed and followed in childhood, a quarter (108/441) were over 18 y (years) at data collection. Median age at diagnosis 7.1 y (9 months-18 y), at data collection 23 y (18-38 y), disease duration 11.3 y (2-36 y). Below the age of 18 y, most patients 386/436 (88.5%) reported adherence to GFD, and most 372/425 (85.7%) normalized serology. Of the 441Â patients, only 3 failed to attend any follow-up visit, and 338/441 (76.6%) attended yearly visits. Over the age 18 y, serology testing was done in 78/108 (72.2%), every 1-3 y in 46/78 (59%). Serology normalized in 61/78 (78.2%). Most adult patients 77/108 (71.5%) never attended a gastroenterology clinic. CD-related complications were rare. Younger age at diagnosis, regular follow-up visits in childhood, resolution of symptoms, and normalization of serology before age 18 were identified as predictors of negative serology after the age of 18 y.

Conclusions: Children who have regular follow-up and normalize serology before age 18 years are likely to maintain a GFD and have negative serology as adults. What is Known: â€¢ The rate of adherence to gluten-free diet (GFD) is higher among children compared to adults. Data on long-term follow-up after transition to adult care is scarce. What is New: Patients diagnosed with CD at a younger age (<12 y), who have yearly follow-up visits, resolution of symptoms, and negative serology in childhood are very likely to maintain GFD and have negative serology as adults. â€¢ Even though most patients do not attend GI clinics after transition to adulthood, most adhere to GFD, and complications are rare

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Expert Rev Clin Immunol. 2021 Feb;17:155-61.

THE IMPORTANCE OF TRANSITION FROM PEDIATRIC TO ADULT RHEUMATOLOGY CARE IN JUVENILE IDIOPATHIC ARTHRITIS.

de Oliveira RJ, Kishimoto ST, de Souza DP, et al.

Introduction: Juvenile idiopathic arthritis (JIA) is the most common rheumatic inflammatory condition in childhood. The long-term morbidity, mortality, and quality of life have improved with the earlier use of disease-modifying drugs (DMARDs) and the availability of biology disease-modifying drugs (bDMARDs). Despite the improvement of treatment, around 50% of the patients reach adulthood with articular and/or extra articular disease activity. A careful planned transition from pediatric to adult care is necessary to reduce the loss of follow-up that is associated with stopping medications, flares, and disability due to untreated arthritis or uveitis. Areas covered: This narrative review provides an overview of the importance of transition in JIA Articles were selected from Pubmed searches.

Expert opinion: JIA patients, family, and healthcare workers have to be trained to provide an effective transition plan, based on local and national policies. Important aspects such as expectations, maturation, disease characteristics, disease activity, adherence, disability, and psychological aspects among others have to be considered and addressed during the transition phase to improve self-esteem, self-assurance, and quality of life

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Fam Pract. 2020 Oct.

STRATEGIES FOR IMPROVING PRIMARY CARE FOR ADOLESCENTS AND YOUNG ADULTS TRANSITIONING FROM PEDIATRIC SERVICES: PERSPECTIVES OF CANADIAN PRIMARY HEALTH CARE PROFESSIONALS.

Schraeder K, Dimitropoulos G, Allemang B, et al.

BACKGROUND: Family physicians and other members of the primary health care (PHC) team may be ideally positioned to provide transition care to adolescents and young adults (AYAs; aged 12-25 years) exiting pediatric specialty services. Potential solutions to well-known challenges associated with integrating PHC and specialty care need to be explored.

OBJECTIVE: To identify strategies to transition care by PHC professionals for AYAs with chronic conditions transitioning from pediatric to adult-oriented care.
METHODS: Participants were recruited from six Primary Care Networks in Calgary, Alberta. A total of 18 semi-structured individual interviews were completed, and transcribed verbatim. Data were analyzed using a qualitative description approach, involving thematic analysis.
RESULTS: Participants offered a range of strategies for supporting AYAs with chronic conditions. Our analysis resulted in three overarching themes: (i) educating AYAs, families, and providers about the critical role of primary care; (ii) adapting existing primary care supports for AYAs and (iii) designing new tools or primary care practices for transition care.
CONCLUSIONS: Ongoing and continuous primary care is important for AYAs involved with specialty pediatric services. Participants highlighted a need to educate AYAs, families and providers about the critical role of PHC. Solutions to improve collaboration between PHC and pediatric specialist providers would benefit from additional perspectives from providers, AYAs and families. These findings will inform the development of a primary care-based intervention to improve transitional care

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Fam Process. 2020 Jun;59:477-91.
THE CULTURAL ADAPTATION OF A TRANSITION PROGRAM FOR LATINO FAMILIES OF YOUTH WITH AUTISM SPECTRUM DISORDER.
Kuhn JL, Vanegas SB, Salgado R, et al.
During the transition to adulthood, effective and culturally relevant supports are critical for families of youth with autism spectrum disorder (ASD). There is a dearth of documented program development and research on supports for Spanish-speaking Latino families during this life stage. The present work describes the cultural adaptation process of an evidence-based transition program for Latino families of youth with ASD. A model of the actions necessary to meaningfully conduct a cultural adaptation in this context is described. After implementing the culturally adapted program titled Juntos en la TransiciÃ³n with five Spanish-speaking families, parents reported high social validity of the program through surveys and interviews. The cultural adaptation process followed in this work is important for the further development of programs that address the transition needs of Latino youth with ASD and their families. Our impressions may also be useful to those who aim to develop culturally sensitive and ecologically valid multifamily group intervention programs for families from cultural and linguistic minority groups

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Front Pediatr. 2020;8:322.
THE TRANSITION OF CARE FROM PEDIATRIC TO ADULT HEALTH-CARE SERVICES OF VERTICALLY HIV-INFECTED ADOLESCENTS: A PILOT STUDY.
Continisio GI, Lo VA, Basile FW, et al.
Objective: Clinical and psychological HIV-related problems peak during adolescence, which coincides with transition of children and adolescents infected from mothers from pediatric to adult reference centers for HIV infection. Transition often is done without specific programs. We wanted to explore transition as an opportunity to increase the efficacy of care and the psychological well-being through a specific program.
Methods: Thirteen vertically infected patients aged 13-20 years were followed up for 24 months by pediatricians, infectious disease specialists, a psychologist, and a nurse. Interventions consisted in joint clinic, simplification of therapy, patient group discussions, HIV infection explanations, and psychological support, lasting 12 months. Efficacy was measured by viro-immunological outcomes and adherence to therapy and psychological tests. Clinical, viro-immunological, and psychological evaluations were performed at 0 (T0) and 12 months (T12) and 6 months after transition to an adult center (T18). Psychological outcomes were assessed using standardized questionnaires for quality of life and self-esteem.
Results: In 11/13 participants, pills administrations/day were significantly reduced. Patients with undetectable viral load and CD4+ >25% increased from 61 to 77% and from 61 to 74%, respectively. Six months after transition, all patients exhibited an undetectable viral load. Adolescents' awareness of the severity of the disease and the risk of sexual transmission was generally poor. Patients classified with "severe" psychological distress according to the quality of life index decreased from 38 to 15% and well-being increased. Similar results were observed 6 months after the transition to adult care. No effect was observed on self-esteem index.
Conclusions: Specific protocols for transition should be developed to optimize resilience and psychological well-being, including routine psychological support for adolescents with HIV infection transiting from pediatric to adult centers for HIV infection

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Front Pediatr. 2020;8:548839.

TRANSITION TO ADULT CARE IN CHILDREN ON LONG-TERM VENTILATION.

Onofri A, Broomfield A, Tan HL.

The number of children on long-term ventilation (LTV) has exponentially increased over the past few decades. Improvements in management of ventilation coupled with improvements in standards of medical care are increasingly allowing young people on LTV to survive into adulthood. The process of transition from the pediatric to the adult healthcare system is challenging and requires special attention. This review aims to provide an overview on transition to adult care for children on LTV. Firstly, examining effective models of transition in other childhood onset chronic conditions as a template, whilst highlighting the unique aspects of transition in LTV patients and secondly, summarizing the main relevant findings in the literature on the topic and emphasizing the importance of a multidisciplinary approach to this process

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Front Public Health. 2020;8:605149.

LOST IN TRANSITION: HEALTH CARE EXPERIENCES OF ADULTS BORN VERY PRETERM-A QUALITATIVE APPROACH.

Perez A, Thiede L, et al.

Introduction: Adults Born Very Preterm (ABP) are an underperceived but steadily increasing patient population. It has been shown that they face multiple physical, mental and emotional health problems as they age. Very little is known about their specific health care needs beyond childhood and adolescence. This article focuses on their personal perspectives: it explores how they feel embedded in established health care structures and points to health care-related barriers they face.

Methods: We conducted 20 individual in-depth interviews with adults born preterm aged 20-54 years with a gestational age (GA) below 33 weeks at birth and birth weights ranging from 870-1,950 g. Qualitative content analysis of the narrative interview data was conducted to identify themes related to self-perceived health, health care satisfaction, and social well-being.

Results: The majority (85%) of the study participants reported that their former prematurity is still of concern in their everyday lives as adults. The prevalence of self-reported physical (65%) and mental (45%) long-term sequelae of prematurity was high. Most participants expressed dissatisfaction with health care services regarding their former prematurity. Lack of consideration for their prematurity status by adult health care providers and the invisibility of the often subtle impairments they face were named as main barriers to receiving adequate health care. Age and burden of disease were important factors influencing participants' perception of their own health and their health care satisfaction. All participants expressed great interest in the provision of specialized, custom-tailored health-care services, taking the individual history of prematurity into account.

Discussion: Adults born preterm are a patient population underperceived by the health care system. Longterm effects of very preterm birth, affecting various domains of life, may become a substantial burden of disease in a subgroup of formerly preterm individuals and should therefore be taken into consideration by adult health care providers

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Growth Horm IGF Res. 2021 Feb;56:101375.

ADULT GROWTH HORMONE DEFICIENCY: OPTIMIZING TRANSITION OF CARE FROM PEDIATRIC TO ADULT SERVICES.

Yuen KCJ, Alter CA, Miller BS, et al.

OBJECTIVE: Most patients with childhood-onset growth hormone deficiency (CO-GHD) receive treatment with exogenous growth hormone (GH) to facilitate the attainment of their full potential adult height. Recent evidence suggests that continuing GH administration during the transition period between the end of linear growth and full adult maturity is necessary for proper body composition and bone and muscle health, and may also have beneficial effects on metabolic parameters, bone mineral density, and quality of life. The timing of this transition period coincides with the transfer of care from a pediatric to an adult endocrinologist, creating the potential for a care gap as a consequence of losing the patient to follow-up.

DESIGN: An advisory board comprising both pediatric and adult endocrinologists was assembled to address current clinical unmet needs and to collaborate on a structured transitional plan for optimal management of patients with CO-GHD.

INSIGHTS/CONCLUSION: The advisors suggest collaborative, multidisciplinary approaches to ensure continuity of care; ongoing testing and monitoring of GHD status into adulthood; and a clearly structured protocol that includes practical guidance for clinicians to establish best practices for transitioning older adolescents with persistent CO-GHD to adult care

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Headache. 2020 Nov;60:2589-91.

THE DEVELOPMENT OF THE MEDICAL TRANSFER PACKET FOR TRANSITION OF CARE OF THE PEDIATRIC PATIENT WITH HEADACHE.

Orr SL, Gelfand AA, Hranilovich J, et al.

Health Policy. 2020 Oct;124:1121-28.

TRANSITIONAL CARE: A NEW MODEL OF CARE FROM YOUNG AGE TO ADULTHOOD.

Bert F, Camussi E, Gili R, et al.

INTRODUCTION: Transition of care represents the transfer from child to adult care. An effective transition maintains continuity of care and presents better clinical outcomes. This process has assumed growing relevance, thanks to improved survivorship of chronic paediatric patients. Actually, there is no a one-size model fitting for all transitions, but each Service organizes its own clinical pathway.
AIM: The study proposes an organizational model for transition, differentiated according to patient complexity.
METHODS: The working group discussed, through regular meetings, the appropriate transitional model for our Hospital. The working group defined a common scheme of transition and elaborated a synthetic document for patients. Then, the common model is adapted, through clinicians' contribution, for different diseases. The complexity assessment includes clinical data, nursing and social information.
RESULTS: The working group defined a common model identifying the main information to be included and detailed in each transition report. The team defined two pathways based on patient's complexity. In case of good compensation and autonomous management, the adolescent is addressed towards standard transition process, a smoother transition from paediatric to adult care with direct connection among healthcare professionals. In case of complex clinical and/or social conditions, an Interdisciplinary Transition Group (ITG) is activated. The group preventively evaluates each patient in periodic meetings and provides a personalized planning of care. In order to define the complexity of a patient, clinical and social determinants are considered. Some diseases are considered complex by default, while others require ITG involvement in case of multiple comorbidities, severe clinical situation, concomitant social criticality and/or cognitive impairment.
DISCUSSION: Transition of care represents an important phase in chronic diseases management. The proposed model assures a multidisciplinary approach, involving all specialists of both paediatric and adult teams. A key determinant of transition is information transmission. Then, the model proposes a common transition report format. Finally, a further perspective study is already in program, in order to assess clinical effectiveness

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Health Qual Life Outcomes. 2021 Jan;19:23.

IMPACT OF A TRANSITION EDUCATION PROGRAM ON HEALTH-RELATED QUALITY OF LIFE IN PEDIATRIC PATIENTS WITH CONGENITAL HEART DISEASE: STUDY DESIGN FOR A RANDOMISED CONTROLLED TRIAL.

Werner O, Bredy C, Lavastre K, et al.

BACKGROUND: Recent advances in the field of congenital heart disease (CHD) led to an improved prognosis of the patients and in consequence the growth of a new population: the grown up with congenital heart disease. Until recently, more than 50% of these patients were lost to follow up because of the lack of specialized structures. The critical moment is the transition between paediatric and adult unit. Therapeutic education is crucial to solve this issue by helping patients to become independent and responsible. The TRANSITION-CHD randomized trial aims to assess the impact of a transition education program on health-related quality of life (HRQoL) of adolescents and young adults with CHD.
METHODS: Multicentre, randomised, controlled, parallel arm study in CHD patients aged from 13 to 25 years old. Patients will be randomised into 2 groups (education program vs. no intervention). The primary outcome is the change in self-reported HRQoL between baseline and 12-month follow-up. A total of 100 patients in each group is required to observe a significant increase of the overall HRQoL score of 7±13.5 points (on 100) with a power of 80% and an alpha risk of 5%. The secondary outcomes are: clinical outcomes, cardiopulmonary exercise test parameters (peak VO2, VAT, VE/VCO2 slope), level of knowledge of the disease using the Leuven knowledge questionnaire for CHD, physical and psychological status.
DISCUSSION: As the current research is opening on patient related outcomes, and as the level of proof in therapeutic education is still low, we sought to assess the efficacy of a therapeutic education program on HRQoL of CHD patients with a randomized trial.
TRIAL REGISTRATION: This study was approved by the National Ethics Committee (South-Mediterranean IV 2016-A01681-50) and was registered on Clinicaltrials.gov (NCT03005626)

HIV Med. 2021 May.

MORTALITY AND AIDS-DEFINING EVENTS AMONG YOUNG PEOPLE FOLLOWING TRANSITION FROM PAEDIATRIC TO ADULT HIV CARE IN THE UK.

Asad H, Collins IJ, Goodall RL, et al.

Objectives: To investigate risk of AIDS and mortality after transition from paediatric to adult care in a UK cohort of young people with perinatally acquired HIV.

Methods: Records of people aged ≥ 13 years on 31 December 2015 in the UK paediatric HIV cohort (Collaborative HIV Paediatric Study) were linked to those of adults in the UK Collaborative HIV Cohort (CHIC) cohort. We calculated time from transition to a new AIDS event/death, with follow-up censored at the last visit or 31 December 2015, whichever was the earliest. Cumulative incidence of and risk factors for AIDS/mortality were assessed using Kaplan-Meier and Cox regression.

Results: At the final paediatric visit, the 474 participants [51% female, 80% black, 60% born outside the UK, median (interquartile range) age at antiretroviral therapy (ART) initiation = 9 (5-13) years] had a median age of 18 (17-19) years and CD4 count of 471 (280-663) cell/μL; 89% were prescribed ART and 60% overall had a viral load ≤ 400 copies/mL. Over median follow-up in adult care of 3 (2-6) years, 35 (8%) experienced a new AIDS event (n = 25) or death (n = 14) (incidence = 1.8/100 person-years). In multivariable analyses, lower CD4 count at the last paediatric visit [adjusted hazard ratio = 0.8 (95% confidence interval: 0.7-1.0)/100 cells/μL increment] and AIDS diagnosis in paediatric care [2.7 (1.4-5.5)] were associated with a new AIDS event/mortality in adult care.

Conclusions: Young people with perinatally acquired HIV transitioning to adult care with markers of disease progression in paediatric care experienced poorer outcomes in adult care. Increased investment in multidisciplinary specialized services is required to support this population at high risk of morbidity and mortality.

HRB Open Res. 2020;3:61.

TRANSITION FROM CHILD-CENTRED TO ADULT-ORIENTED HEALTHCARE SYSTEMS FOR YOUNG PEOPLE WITH NEURODISABILITY: A SCOPING REVIEW PROTOCOL.

Fortune J, Murphy P, Merchant N, et al.

Background: The transition from child-centred to adult-oriented healthcare is a challenging time for young people with neurodisability. As the prevalence of neurodisability increases, greater numbers of young people will eventually transfer to the adult healthcare system. While there is a growing recognition of the importance of providing quality, transitional care, little is known about how to manage and optimise this process for young people with neurodisability. The objective of this scoping review is to examine and map existing literature related to the transition from child-centred to adult-oriented healthcare systems for young people with neurodisability.

Methods: Systematic literature searches of OVID MEDLINE, EMBASE, PsycINFO, CINAHL, Cochrane Library and Web of Science will be conducted from inception to present. A structured iterative search of grey literature will be conducted. This review will consider all study designs examining the transition from child to adult health services in neurodisability. Two reviewers will independently screen each retrieved title and abstract and assess full-text articles against the inclusion criteria to determine eligibility. Data will be extracted and synthesised quantitatively and qualitatively. The process and reporting will follow PRISMA-ScR guidelines.

Conclusion: This review will provide a broad and systematically mapped synthesis of the extent and nature of the available published and unpublished literature on transition from child-centred to adult-oriented healthcare systems in neurodisability. The results will be used to determine gaps in the current evidence base in order to prioritise areas for future research

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Indian J Pediatr. 2020 Jun;87:411-12.

HEALTH CARE TRANSITION: NEED OF THE HOUR.

Basu S.

Int J Cardiol. 2020 Jul;311:35-36.

TRANSITION TO ADULT CARE FOR ADOLESCENTS WITH CONGENITAL HEART DISEASE - IS THERE A LIGHT AT THE END OF THE TUNNEL?

Vonder M, I .

Int J Environ Res Public Health. 2021 Apr;18.

DEVELOPMENTAL LANGUAGE DISORDER (DLD) IN YOUNG PEOPLE LEAVING CARE IN ENGLAND: A STUDY PROFILING THE LANGUAGE, LITERACY AND COMMUNICATION ABILITIES OF YOUNG PEOPLE TRANSITIONING FROM CARE TO INDEPENDENCE.

Clegg J, Crawford E, Spencer S, et al.

Research indicates children and young people in care have a high prevalence of Developmental Language Disorder (DLD) as part of a complex set of vulnerabilities. This study describes the profile of language, literacy and communication abilities of a cohort of care leavers. The language, literacy and communication abilities of 44 young people leaving care between the ages of 16 and 26 years were assessed using standardized measures. Demographic data about the young people was collected along with a survey to key staff to capture their perceptions and experiences of the language and communication abilities of these young people. Ninety percent of the care leavers' language abilities were below average and over 60% met criteria for DLD in combination with literacy difficulties, developmental disorders and social, emotional and mental health difficulties (SEMH). The implications of unidentified DLD on the lives of young people leaving care is discussed. Earlier identification of DLD is advocated to enable services to intervene to facilitate more positive outcomes and life chances for this very vulnerable population

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Ir J Med Sci. 2020 Aug;189:761-69.

PARTICIPATORY DEVELOPMENT OF A PATIENT-CLINICIAN COMMUNICATION TOOL TO ENHANCE HEALTHCARE TRANSITIONS FOR YOUNG PEOPLE WITH 22Q11.2.

Kerin L, Lynch D, McNicholas F.

BACKGROUND: Individuals with the rare genetic disorder, 22q11.2 deletion syndrome (22q11.2ds), face particular challenges with transition from paediatric to adult health services due to complex physical and mental health care needs, often further complicated by intellectual disability (ID). To date, the lived experience of these young people navigating this complex journey has not been well researched.

AIM: The project sought to understand the lived experiences of young women with 22q11.2ds transitioning from child to adult health services and to elicit recommendations for improvement.

METHODS: Following ethical approval, six female participants, aged 19-35Â years, were recruited through the family support organisation 22q11 Ireland. Adhering to participatory action research (PAR) principles, four full day sessions using creative research methodologies were conducted over a 4-month period.

RESULTS: Participants reported significant difficulties navigating transition between and within clinical services, and reported a lack of information transfer between healthcare services which required multiple retelling of their story. They expressed a low sense of confidence in new healthcare providers and reported ambivalence regarding their own agency and ability to manage clinical appointments without family or 'keyworker' support. Participants co-designed a patient-clinician communication tool to assist in information transfer and to capture salient features of any healthcare consultation.

CONCLUSIONS: There is a recognised need to strengthen transition pathways. This is especially true in this at risk group, given the poorer outcomes associated with transitions in youth with ID along with the additive effect of medical and mental health and learning difficulties that often co-occur in 22q11.2ds. A patient-clinician communication tool, designed by participants, offers a pragmatic approach to optimise healthcare transitions, support continuity of healthcare and personal autonomy

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J Adolesc Health. 2021 Mar.

IMPROVING ADOLESCENT AND YOUNG ADULT ENGAGEMENT IN THE PROCESS OF TRANSITIONING TO ADULT CARE.

Lestishock L, Nova S, Disabato J.

PURPOSE: Health care transition (HCT) is the complex process of changing from pediatric to adult-centered care. Comprehensive HCT processes have been associated with improved outcomes in all elements of the Triple Aim. Nationally accepted best practices emphasize Six Core Elements of HCT, including the use of transition readiness assessment tools completed during clinic visits. Specifically, Got Transition's tools include two 0-10 point self-report scales on the validated domains of importance of changing to an adult provider and managing their healthcare, and confidence in their ability to transition. The aim of this quality improvement project (QIP) was to improve the engagement of adolescents and young adults (AYAs), aged 14-20, in the process of transitioning from pediatric to adult care. The sub-aim focused specifically on parent/caregiver engagement in transition, using the same scales in a tool for parents/caregivers. An urban federally qualified health center initiated this QIP.

METHODS: This QIP utilized the Institute for Healthcare Improvement Model for Improvement and plan-do-study-act cycles.

RESULTS: Eighty-five AYAs and 40 parents/caregivers completed readiness assessments twice. Scores improved overall, reaching statistical significance with a small change in AYA mean scores for importance

(.94) and confidence (.75). Provision of a transition policy and completion of readiness assessments by AYAs and parents/caregivers met the 70% goal. Patient portal enrollments increased from 4.2% to 12.5%, although did not meet the 30% goal.
CONCLUSIONS: Engagement of AYAs and parents/caregivers was improved as a result of this QIP. Successful routine implementation of transition process measures demonstrated improved clinic-wide communication

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J Adolesc Health. 2021 May.
EXPLORING HEALTH LITERACY, TRANSITION READINESS, AND HEALTHCARE UTILIZATION IN MEDICAID CHRONICALLY ILL YOUTH.
Chisolm DJ, Keedy HE, Hart LC, et al.
PURPOSE: Youths with special healthcare needs face challenges transitioning from pediatric to adult health care. Understanding possible mechanisms contributing to poor healthcare transition could improve care. This study explores associations between health literacy (HL), transition readiness, and healthcare utilization.
METHODS: Youths with special healthcare needs aged 12-18 years were recruited from a Medicaid accountable care organization (2012-2017). Outcome measures included transition readiness (Transition Readiness Assessment Questionnaire), and healthcare utilization (any well-check, hospitalization, emergency department [ED] visit, or ambulatory sensitive condition ED visit). Multivariate regression analyses examined whether HL (adequate vs. inadequate) predicted outcomes, after adjusting for covariates. Models were then created to examine whether the effect of HL on healthcare utilization was mediated by transition readiness.
RESULTS: Among 417 youths with special healthcare needs, 67.1% reported adequate HL. Relative to inadequate HL, teens with adequate HL had significantly higher average Transition Readiness Assessment Questionnaire-20 scores (Î²Â = .34, p < .001). Controlling for covariates, HL was a significant predictor of having an ambulatory sensitive condition ED visit and having any ED visits neared significance. There was a positive transition readiness mediation effect on having an ED visit, with higher transition readiness being associated with higher odds of having any ED visit in the mediation analysis.
CONCLUSIONS: HL is independently associated with higher transition readiness and lower ambulatory sensitive condition ED use, but pathways of action require further study

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J Adolesc Health. 2021 Mar.
ASSOCIATIONS BETWEEN HEALTH CARE TRANSITION PREPARATION AMONG YOUTH IN THE US AND OTHER COMPONENTS OF A WELL-FUNCTIONING SYSTEM OF SERVICES.
Ilango SM, Lebrun-Harris LA, Jones JR, et al.
PURPOSE: This study examines the relationships between receipt of health care transition (HCT) preparation among US youth and five other components of a well-functioning system of services (family partnership in decision-making, medical home, early/continuous screening for special health care needs [SHCN], continuous/adequate health insurance, access to community-based services).
METHODS: Data came from the combined 2016-2017 National Survey of Children's Health (nÂ = 29,617 youth ages 12-17). Parents/caregivers answered questions about their child's health care experiences, which were combined to measure receipt of HCT preparation and the other five components of a well-functioning system of services. Unadjusted and adjusted analyses were conducted to examine associations, stratified by youth with and without special health care needs (YSHCN/non-YSHCN).
RESULTS: About 16.7% of YSCHN and 13.9% of non-YSHCN received HCT preparation (pÂ = .0040). Additionally, 25.3% of YSHCN and 27.3% of non-YSHCN received all five remaining components of a system of services (pÂ = .1212). HCT preparation was positively associated with receipt of the combined five components among both YSHCN (adjusted prevalence rate ratioÂ = 1.53, 95% confidence interval: 1.20-1.86) and non-YSHCN (adjusted prevalence rate ratioÂ = 1.63, 95% confidence interval: 1.39-1.88). Regarding individual system of services components, early and continuous screening for SHCN was significantly associated with HCT preparation for both populations. For non-YSHCN only, having a medical home was associated with HCT preparation. The remaining three components were not associated with HCT preparation for either population after adjusting for sociodemographic characteristics.
CONCLUSIONS: Among both YSHCN and non-YSHCN, HCT preparation is positively associated with receipt of early and continuous screening for SHCN as well as the five combined components of a well-functioning system of services

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J Adolesc Young Adult Oncol. 2020 Jul.

THE CREATION OF A COMPREHENSIVE ADOLESCENT AND YOUNG ADULT CANCER SURVIVORSHIP PROGRAM: "LOST IN TRANSITION" NO MORE.

Linendoll N, Murphy-Banks R, Barthel E, et al.

Purpose: The Reid R. Sacco AYA Cancer Program set out to improve survivorship care for AYA-aged patients (15-39 years) of pediatric or AYA cancer. This article discusses the steps in establishing the clinic, including the creation of a database on cancer history, exposures, and attendant risks of late effects. Results from the database tell the broader story of AYAs who seek care within a dedicated survivorship clinic.

Methods: The database was created with REDCap® (Research Electronic Data Capture), a secure web-based, HIPAA compliant application for research and clinical study data. Data were abstracted and analyzed by trained members of the program team.

Results: A total of 144 patients were seen for their initial survivorship visit between January 2013 and September 2019. Regarding physical health, two-thirds of the patients presented with an established late effect, one third with an established medical comorbidity, and 11% (n = 16) with secondary cancer related to their oncologic treatment. In assessing mental health, a significant cohort reported a known affective disorder (32%, n = 46) with one quarter already taking a psychotropic medication. Despite the transient nature of AYAs, 85% of patients remained in care within the long-term follow-up clinical model.

Conclusions: Data presented illustrate how multilayered and complex survivorship care needs can be, as patients enter the clinic with complicated pre-existing psychosocial issues, significant late effects, and comorbidities. This study reinforces the value of a clinical database to better understand AYA survivors with the ultimate goal of optimizing and coordinating care.

J Adolesc Young Adult Oncol. 2021 Apr.

A COMPARISON OF YOUNG ADULTS WITH AND WITHOUT CANCER IN CONCURRENT HOSPICE CARE: IMPLICATIONS FOR TRANSITIONING TO ADULT HEALTH CARE.

Mooney-Doyle K, Keim-Malpass J, Svynarenko R, et al.

Purpose: Concurrent hospice care provides important end-of-life care for youth under 21 years. Those nearing 21 years must decide whether to shift to adult hospice or leave hospice for life-prolonging care. This decision may be challenging for young adults with cancer, given the intensity of oncology care. Yet, little is known about their needs. We compared young adults with and without cancer in concurrent hospice care.

Methods: Retrospective comparative design used data from 2011 to 2013 U.S. Medicaid data files. Decedents were included if they were 20 years of age, enrolled in Medicaid hospice care, and used nonhospice medical services on the same day as hospice care based on their Medicaid claims activity dates. Results: Among 226 decedents, 21% had cancer; more than half were female (60.6%), Caucasian (53.5%), non-Hispanic (77.4%), urban dwelling (58%), and had mental/behavioral disorder (53%). Young adults with cancer were more often non-Caucasian (68.7% vs. 40.4%), technology dependent (47.9% vs. 24.2%), had comorbidities (83.3% vs. 30.3%), and lived in rural (58.3% vs. 37.6%), southern (41.7% vs. 20.8%) areas versus peers without cancer. Those with cancer had significantly fewer live discharges from hospice (5.7 vs. 17.3) and sought treatment for symptoms more often from nonhospice providers (35.4% vs. 14.0%).

Conclusions: Young adults in concurrent hospice experience medical complexity, even at end-of-life. Understanding care accessed at 20 years helps providers guide young adults and families considering options in adult-focused care. Clinical and demographic differences among those with and without cancer in concurrent care highlight needs for research exploring racial and geographic equity

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J Adv Nurs. 2020 Jun;76:1293-306.

A SYSTEMATIC REVIEW ON RANDOMIZED CONTROLLED TRIALS: COACHING ELEMENTS OF DIGITAL SERVICES TO SUPPORT CHRONICALLY ILL ADOLESCENTS DURING TRANSITION OF CARE.

Tornivuori A, Tuominen O, SalanterÃ¤ S, et al.

AIMS: To define digital health services that have been studied among chronically ill adolescents and to describe e-health coaching elements that may have an impact on transition outcomes.

DESIGN: Systematic review without meta-analysis.

DATA SOURCES: MEDLINE (Ovid), Pub Med, Scopus and CINAHL on 28 May 2018.

REVIEW METHODS: Peer-reviewed articles published between January 2008-May 2018 were reviewed following the Cochrane Handbook for Systematic Reviews of Interventions and reported according to the Preferred Reporting Items for Systematic Reviews and Meta-analyses statement.

RESULTS: Twelve randomized controlled trials were included. The interventions varied significantly in duration and content. E-coaching that included human and social support showed positive impact on transition outcomes. Digital health services incorporated into usual care provide efficient and accessible care.

CONCLUSION: E-coaching elements enable tailoring and personalization and present a tool for supporting and motivating chronically ill adolescents during transition of care. Future research should evaluate the effectiveness of e-coaching elements.
IMPACT: Digital services are considered a means for increasing adolescents' motivation for self-care and for increasing their accessibility to health care. The coaching elements in digital services consist of a theoretical basis, human support, interactive means and social support. Included interventions varied in terms of duration, dose, content and design. Our results may serve the development of digital health services for adolescents in transition. E-coaching can be used to engage and motivate chronically ill adolescents to improve health behaviour and self-management during transition of care

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J Adv Nurs. 2021 May;77:2340-52.

INFLUENCE OF TRANSITION READINESS FROM PAEDIATRIC TO ADULT HEALTH CARE ON QUALITY OF LIFE IN CHILD-PARENT DYADS WITH LONG-TERM CONDITIONS.

Ma J, Gong G, Zhang T, et al.

AIM: To delineate the impact of perspective of children's transition readiness from paediatric to adult health care on quality of life in child-parent dyads with long-term conditions. We used Actor-Partner Interdependence Model to identify actor effects (effect of one's own transition readiness on one's own quality of life) and partner effects (effect of one's own transition readiness on the partner's quality of life).
DESIGN: A multi-centre cross-sectional survey.
METHODS: The study was conducted in two paediatric hospitals in China from October 2018-August 2019. We used a researcher-designed questionnaire to collect demographic and clinical characteristics. Furthermore, we used four questionnaires assessing transition readiness and quality of life in child and parent respectively to collect data from 370 child-parent dyads. Structural equation modelling was applied to estimate the effect of actor-partner interdependence models.
RESULTS: The total score of transition readiness had two actor effects on total child and parent quality of life controlling for age ($\hat{I}^2$(children) 3.335, p= .032 and $\hat{I}^2$(parents) 8.952, p < .001), while only one actor effect controlling for gender ($\hat{I}^2$(parents) 8.891, p < .001). Specific transition readiness dimensions had different partner effects on different domains of children and parents' quality of life. Moreover, younger children and fathers had a better quality of life.
CONCLUSION: Our study verified inherently interpersonal relationship that transition readiness appeared to influence quality of life in child-parent dyads with long-term conditions.
IMPACT: This study was the first to verify mutual influence of transition readiness and quality of life in child-parent dyad using actor-partner interdependence model. Nurses who design the transition promoting programs should consider the effective communication between healthcare provider and child-parent dyads and support parents' involvement to improve shared understanding about managing child's condition, especially for older children and mothers

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J Allergy Clin Immunol. 2020 Nov;146:967-83.

CONSENSUS OF THE ITALIAN PRIMARY IMMUNODEFICIENCY NETWORK ON TRANSITION MANAGEMENT FROM PEDIATRIC TO ADULT CARE IN PATIENTS AFFECTED WITH CHILDHOOD-ONSET INBORN ERRORS OF IMMUNITY.

Cirillo E, Giardino G, Ricci S, et al.

Medical advances have dramatically improved the long-term prognosis of children and adolescents with inborn errors of immunity (IEIs). Transfer of the medical care of individuals with pediatric IEIs to adult facilities is also a complex task because of the large number of distinct disorders, which requires involvement of patients and both pediatric and adult care providers. To date, there is no consensus on the optimal pathway of the transitional care process and no specific data are available in the literature regarding patients with IEIs. We aimed to develop a consensus statement on the transition process to adult health care services for patients with IEIs. Physicians from major Italian Primary Immunodeficiency Network centers formulated and answered questions after examining the currently published literature on the transition from childhood to adulthood. The authors voted on each recommendation. The most frequent IEIs sharing common main clinical problems requiring full attention during the transitional phase were categorized into different groups of clinically related disorders. For each group of clinically related disorders, physicians from major Italian Primary Immunodeficiency Network institutions focused on selected clinical issues representing the clinical hallmark during early adulthood

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J Am Acad Dermatol. 2020 Jun.

REVIEW OF TRANSITION OF CARE LITERATURE: EPIDERMOLYSIS BULLOSA, A PARADIGM FOR PATIENTS WITH COMPLEX DERMATOLOGIC CONDITIONS.

Perez VA, Morel KD, Garzon MC, et al.

BACKGROUND: Transition from pediatric to adult care is a critical component of healthcare for children with chronic needs. The characteristics of epidermolysis bullosa (EB) demand higher than average levels of provider support. There is consensus among health care professionals regarding the importance of transition, however, there is a scarcity of practical information regarding models for patients with EB.

OBJECTIVE: Review transition of care (TOC) programs in varying specialties. Highlight practical considerations to facilitate the development of programs for patients with EB and other complex dermatologic conditions.

METHODS: Articles identified via MEDLINE and EMBASE health literature database and screened for relevance to TOC.

RESULTS: Various models for transition exist. A well-executed formal transition program, early introduction, interdisciplinary collaboration and psychosocial support were themes associated with successful outcomes.

LIMITATIONS: TOC programs that have not been described in the literature are not reflected in this review.

CONCLUSIONS: Patients with EB have unique needs that affect transition and span expertise across traditional boundaries such as dependency on others for daily skin care, failure to thrive, and risk of squamous cell carcinoma. Given the rarity of the disease, patients with EB will benefit from collaborative efforts to develop programs to optimize successful transition

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J Assoc Nurses AIDS Care. 2021 Mar.

TRANSITION FROM PEDIATRIC AND ADOLESCENT HIV CARE TO ADULT HIV CARE AND THE PATIENT-PROVIDER RELATIONSHIP: A QUALITATIVE METASYNTHESIS.

Barr EA, Raybin JL, Dunlevy H, et al.

Approximately 5 million adolescents (ages 15-24 years) living with HIV will transition to adult care in the next decade. Only half are engaged in care 12 months post-transition. This qualitative metasynthesis aimed to answer: What effect did the patient-provider relationship (PPR) have on adolescent living with HIV transition? What strategies were suggested to develop trusting relationships to promote engagement and retention in care? Primary qualitative studies from PubMed, CINAHL, and EBSCO (January 2008 to December 2019) were identified. Data were analyzed using team-based thematic synthesis techniques and international standards. Fourteen articles with 478 participants from eight countries were included. Four themes emerged: the familial nature of the PPR, stigma as a bond and barrier, the provider knowing the patient and getting to know new providers, and recommendations supporting transition. The PPR is integral. Collaborative strategies used to build new relationships will support autonomy, decrease stigma, and facilitate trust

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J Asthma. 2020 Oct;57:1119-27.

LOST IN THE TRANSITION FROM PEDIATRIC TO ADULT HEALTHCARE? EXPERIENCES OF YOUNG ADULTS WITH SEVERE ASTHMA.

Ã–dling M, Jonsson M, Janson C, et al.

Objective: Asthma is a multifaceted disease, and severe asthma is likely to be persistent. Patients with severe asthma have the greatest burden and require more healthcare resources than those with mild-to-moderate asthma. The majority with asthma can be managed in primary care, while some patients with severe asthma warrant referral for expert advice regarding management. In adolescence, this involves a transition from pediatric to adult healthcare. This study aimed to explore how young adults with severe asthma experienced the transition process.

Methods: Young adults with severe asthma were recruited from an ongoing Swedish population-based cohort. Qualitative data were obtained through individual interviews (n=16, mean age 23.4 years), and the transcribed data were analyzed with systematic text condensation.

Results: Four categories emerged based on the young adults' experiences: "I have to take responsibility", "A need of being involved", "Feeling left out of the system", and "Lack of engagement". The young adults felt they had to take more responsibility, did not know where to turn, and experienced fewer follow-ups in adult healthcare. Further, they wanted healthcare providers to involve them in self-management during adolescence, and in general, they felt that their asthma received insufficient support from healthcare providers.

Conclusions: Based on how the young adults with severe asthma experienced the transition from pediatric to adult healthcare, it is suggested that healthcare providers together with each patient prepare, plan, and communicate in the transition process for continued care in line with transition guidelines

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J Autism Dev Disord. 2020 Jun;50:2174-87.

GROWING UP WITH FRAGILE X SYNDROME: CONCERNS AND CARE NEEDS OF YOUNG ADULT PATIENTS AND THEIR PARENTS.

Van Remmerden MC, Hoogland L, Mous SE, et al.

Little is known about care needs of young adults with Fragile X Syndrome (FXS). Patient-driven information is needed to improve understanding and support of young adults with FXS. A qualitative study was performed in 5 young adult patients (aged 18-30), and 33 parents of young adults. Concerns and care needs were categorized using the International Classification of Functioning, Disability, and Health. Results indicated concerns on 14 domains for males, and 13 domains for females, including physical, psychological and socio-economical issues. In both groups parents reported high stress levels and a lack of knowledge of FXS in adult care providers. This study revealed concerns on various domains, requiring gender-specific, multidisciplinary transitional care and adult follow-up for patients with FXS

J Autism Dev Disord. 2021 Jan;51:298-306.

ANXIETY AND DEPRESSION REDUCTION AS DISTAL OUTCOMES OF A COLLEGE TRANSITION READINESS PROGRAM FOR ADULTS WITH AUTISM.

Capriola-Hall NN, Brewe AM, Golt J, et al.

Young adults with autism spectrum disorder (ASD) experience increased rates of anxiety and depression which can impact academic success. The Stepped Transition in Education Program for Students with ASD (STEPS) applies cognitive-behavioral principles to help young adults with ASD improve their adjustment to postsecondary education. We aimed to determine if STEPS had an effect on anxiety and depression. Treatment-seeking adults with ASD (n = 32; Mage = 19.74) were randomized to STEPS or transition as usual (TAU; i.e., waitlist control group). STEPS participants evinced significantly greater declines in depressive symptoms from pre-treatment to post-treatment compared to the waitlist. Anxiety symptoms did not significantly change. Results suggest that transition support for young people with ASD may improve mental health

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J Cancer Surviv. 2021 Feb;15:151-62.

TRANSITION FROM PEDIATRIC TO ADULT FOLLOW-UP CARE IN CHILDHOOD CANCER SURVIVORS-A SYSTEMATIC REVIEW.

Otth M, Denzler S, Koenig C, et al.

PURPOSE: The successful transition of childhood cancer survivors from pediatric- to adult-focused long-term follow-up care is crucial and can be a critical period. Knowledge of current transition practices, especially regarding barriers and facilitators perceived by survivors and health care professionals, is important to develop sustainable transition processes and implement them into daily clinical practice. We performed a systematic review with the aim of assessing transition practices, readiness tools, and barriers and facilitators.

METHODS: We searched three databases (PubMed, Embase/Ovid, CINAHL) and included studies published between January 2000 and January 2020. We performed this review according to the PRISMA guidelines and registered the study protocol on PROSPERO; two reviewers independently extracted the content of the included studies.

RESULTS: We included 26 studies: six studies described current transition practices, six assessed transition readiness tools, and 15 assessed barriers and facilitators to transition.

CONCLUSION: The current literature describing transition practices is limited and overlooks adherence to follow-up care as a surrogate marker of transition success. However, the literature provides deep insight into barriers and facilitators to transition and theoretical considerations for the assessment of transition readiness. We showed that knowledge and education are key facilitators to transition that should be integrated into transition practices tailored to the individual needs of each survivor and the possibilities and limitations of each country's health care system.

IMPLICATIONS FOR CANCER SURVIVORS: The current knowledge on barriers and facilitators on transition should be implemented in clinical practice to support sustainable transition processes

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J Child Health Care. 2020 Jun;24:246-59.

HEALTH LITERACY, NUTRITION KNOWLEDGE, AND HEALTH CARE TRANSITION READINESS IN YOUTH WITH CHRONIC KIDNEY DISEASE OR HYPERTENSION: A CROSS-SECTIONAL STUDY .

Zhong Y, Patel N, Ferris M, et al.

This study evaluates the associations of nutrition knowledge and health literacy with health care transition (HCT) readiness and self-management in adolescents and young adults (AYAs) with chronic kidney disease (CKD) or hypertension. Chronically ill AYAs with poor HCT or self-management skills are less likely to achieve favorable health outcomes as they enter adulthood. Health literacy and nutrition knowledge, which are

identified as important contributors to health outcomes, may suggest important points of interventions to improve self-management skills. For the study, we enrolled 59 consecutive patients ages 12-29, with a diagnosis of CKD or hypertension at pediatric- and adult-focused clinics in the United States. Participants completed measures of nutrition knowledge, health literacy, and the Self-management and Transition to Adulthood with R(x) = treatment (STAR(x)) questionnaire. Correlation tests and multivariate regressions were employed for data analysis. The findings show that health literacy was positively associated with self-management skills (p = .050), communication with providers (p = .002) and overall HCT (p = .001) after adjusting for key variables. Disease-specific nutrition knowledge positively predicted communication with providers (p = .002) and overall HCT (p < .001) after adjusting for key variables. Therefore, health literacy and nutrition knowledge predicted self-management and transition readiness. Testing for these skills should be considered in clinics and HCT preparation for AYAs with chronic conditions

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J Child Health Care. 2021 Jun;25:305-19.

'A CONFIDENT PARENT BREEDS A CONFIDENT CHILD': UNDERSTANDING THE EXPERIENCE AND NEEDS OF PARENTS WHOSE CHILDREN WILL TRANSITION FROM PAEDIATRIC TO ADULT CARE.

Shaw KL, Baldwin L, Heath G.

Transitional care for young people with long-term conditions emphasizes the importance of supporting parents, particularly in relation to promoting adolescent healthcare autonomy. Yet, little practical guidance is provided, and transitional care remains suboptimal for many families. This study aimed to examine how parents understand and experience their caregiving role during their child's transition to adult services, to identify parents' needs, and to inform service improvements. Focus groups were undertaken with parents of young people with brittle asthma, osteogenesis imperfecta, or epilepsy. Data were analysed using interpretative phenomenological analysis. Participants (n = 13) described how their parenting roles extended beyond what they consider usual in adolescence. These roles were presented as time consuming, stressful, and unrelenting but necessary to protect children from harm in the face of multiple risks and uncertainties. Such protective strategies were also perceived to hinder adolescent development, family functioning, and their own development as midlife adults. Finding a balance between protecting immediate health and long-term well-being was a major theme. Participants called for improved support, including improved service organization. Recommendations are provided for working with parents and young people to manage the risks and uncertainties associated with their condition, as part of routine transitional care

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J Child Neurol. 2020 Jul;35:536-42.

CONTEXTUALIZED AUTONOMY IN TRANSITIONAL CARE FOR YOUTH WITH NEUROLOGIC CONDITIONS: THE ROLE OF THE PEDIATRIC NEUROLOGIST.

Bogossian A, Majnemer A, Racine E.

Youth with neurologic conditions experience multiple life transitions. The transfer from pediatric to adult health care systems exemplifies one such complex and multifaceted transition that occurs in parallel with developmental, legal, and social changes that may influence the roles and responsibilities of youth and their caregivers. As a result, ethical situations, questions, and challenges may surface in transition care to which pediatric neurologists may be confronted. In this article, we focus on the topic of autonomy and situations that may arise in transition care in the context of pediatric neurology. Building from a clinical case, we present the concept of contextualized autonomy to work through the questions that arise in the case and propose ways of thinking through those challenging situations in transition care

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J Clin Med. 2021 Feb;10.

THE COMING-OF-AGE TRANSITION CARE FOR ADOLESCENTS WITH RHEUMATIC DISEASE-WHERE ARE WE AND WHAT HAVE WE DONE IN ASIA?

Teh KL, Hoh SF, Arkachaisri T.

The transition from pediatric to adult health care is a challenging yet important process in rheumatology as most childhood-onset rheumatic diseases persist into adulthood. Numerous reports on unmet needs as well as evidence of negative impact from poor transition have led to increased efforts to improve transition care, including international guidelines and recommendations. In line with these recommendations, transition programs along with transition readiness assessment tools have been established. Despite these efforts, there are still a lot of work to be done for transition care in rheumatology. This review article focuses on how transition care in rheumatology has developed in recent years and highlights the gaps in current practices

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J Deaf Stud Deaf Educ. 2021 Jan;26:21-45.

FAMILY-CENTERED CARE IN THE TRANSITION TO EARLY HEARING INTERVENTION.

Nickbakht M, Meyer C, Scarinci N, et al.

This study aimed to explore and compare families' and professionals' perspectives on the implementation of family-centered care (FCC) (Moeller, Carr, Seaver, Stredler-Brown, & Holzinger, 2013) during the period between diagnosis of hearing loss (HL) and enrollment in early intervention (EI). A convergent mixed-methods study incorporating self-report questionnaires and semistructured in-depth interviews was used. Seventeen family members of children with HL and the 11 professionals who support these families participated in this study. The results suggested that the services engaged during the transition period partially adhered to the principles of FCC, including the provision of timely access to EI services and provision of emotional and social support. However, areas for improvement identified include strengthening family/professional partnerships, shared decision-making processes, collaborative teamwork, program monitoring, and consistency in the provision of information and support. Qualitative and quantitative research findings also indicated a lack of consistency in service provision during the transition period

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J Int AIDS Soc. 2021 Feb;24:e25676.

EXAMINING HEALTHCARE TRANSITION EXPERIENCES AMONG YOUTH LIVING WITH HIV IN ATLANTA, GEORGIA, USA: A LONGITUDINAL QUALITATIVE STUDY.

Halyard AS, Doraivelu K, Camacho-GonzÃ¡lez AF, et al.

INTRODUCTION: Virtually all youth living with HIV in paediatric/adolescent care must eventually transition to adult-oriented HIV care settings. To date, there is limited evidence examining the perspectives of youth living with HIV longitudinally through the healthcare transition process. The objective of our study was to examine attitudes and experiences of youth living with HIV regarding healthcare transition, including potential change in attitudes and experiences over time.

METHODS: We conducted a longitudinal qualitative interview study within a large, comprehensive HIV care centre in Atlanta, Georgia, USA between August 2016 and October 2019.We interviewed 28 youth living with HIV as part of a longitudinal observational cohort study of youth undergoing healthcare transition. We conducted qualitative interviews both immediately prior to, and one year following the transition from paediatric to adult-oriented care.

RESULTS: Six distinct themes emerged from interviews conducted with youth living with HIV pre-transition: (1) reluctance to transition; (2) paediatric spaces as welcoming, and adult spaces as unwelcoming; (3) varying levels of preparation for transition; and (4) expectation of autonomy in the adult clinic. Analysis of post-transition interviews with the same youth demonstrated: (1) inconsistencies in the transition experience; (2) fear and anxiety about transition quelled by experience; (3) varying reactions to newfound autonomy and (4) communication as the most valuable facilitator of successful transition.

CONCLUSIONS: This study's longitudinal perspective on the healthcare transition experience yields insights that can be incorporated into programming targeting this critically important population. Although our study was conducted in a USA-based clinic with co-located paediatric and adult care services, many of our findings are likely to have relevance in other settings as well. Interventions aiming to improve HIV care engagement through transition should seek to enhance patient-provider communication in both paediatric and adult clinics, improve preparation of patients in paediatric clinics and ease patients gradually into autonomous disease management

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J Med Genet. 2020 Oct.

PHENOTYPES IN ADULT PATIENTS WITH RETT SYNDROME: RESULTS OF A 13-YEAR EXPERIENCE AND INSIGHTS INTO HEALTHCARE TRANSITION.

Peron A, Canevini MP, Ghelma F, et al.

Background: Rett syndrome is a complex genetic disorder with age-specific manifestations and over half of the patients surviving into middle age. However, little information about the phenotype of adult individuals with Rett syndrome is available, and mainly relies on questionnaires completed by caregivers. Here, we assess the clinical manifestations and management of adult patients with Rett syndrome and present our experience in transitioning from the paediatric to the adult clinic.

Methods: We analysed the medical records and molecular data of women aged ≥18 years with a diagnosis of classic Rett syndrome and/or pathogenic variants in MECP2, CDKL5 and FOXG1, who were in charge of our clinic.

Results: Of the 50 women with classic Rett syndrome, 94% had epilepsy (26% drug-resistant), 20% showed extrapyramidal signs, 40% sleep problems and 36% behavioural disorders. Eighty-six % patients exhibited gastrointestinal problems; 70% had scoliosis and 90% low bone density. Breathing irregularities were

diagnosed in 60%. None of the patients had cardiac issues. CDKL5 patients experienced fewer breathing abnormalities than women with classic Rett syndrome.
Conclusion: The delineation of an adult phenotype in Rett syndrome demonstrates the importance of a transitional programme and the need of a dedicated multidisciplinary team to optimise the clinical management of these patients

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J Nurs Scholarsh. 2021 Mar;53(2):198-207

THE TRANSITION TO ADULT HEALTH CARE IN YOUTH WITH SPINA BIFIDA: THEORY, MEASUREMENT, AND INTERVENTIONS.

Holmbeck GN, Kritikos TK, Stern A, et al.

PURPOSE: This article focuses on the transition to adult health care in youth with spina bifida (SB) from the perspective of theory, measurement, and interventions.
METHODS: The purpose of this article is to discuss (a) a theory of linkages between the transfer of medical responsibility from parent to child and the transition from pediatric to adult health care, as mediated by transition readiness; (b) measurement issues in the study of self-management and the transition to adult health care; and (c) U.S.-based and international interventions focused on the transition to adult health care in young adults with SB.
FINDINGS: Individuals with SB must adhere to a complex multicomponent treatment regimen while at the same time managing a unique array of cognitive and psychosocial challenges and comorbidities that hinder self-management, medical adherence, and the transition to adult health care. Moreover, such youth endure multiple transitions to adult health care (e.g., in the areas of urology, orthopedics, neurosurgery, and primary care) that may unfold across different time frames. Finally, three transition-related constructs need to be assessed, namely, transition readiness, transition completion, and transition success.
CONCLUSIONS: SB provides an important exemplar that highlights the complexities of conducting research on the transition to adult health care in youth with chronic health conditions. Many transition trajectories are possible, depending on the functioning level of the child and a host of other factors. Also, no single transition pathway is optimal for all patients with SB.
CLINICAL RELEVANCE: The success of the process by which a child with SB transitions from pediatric to adult health care can have life-sustaining implications for the patient

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J Pediatr (Rio J). 2021 Mar.

GROWTH HORMONE DEFICIENCY AND THE TRANSITION FROM PEDIATRIC TO ADULT CARE.

Tavares ABW, Collett-Solberg PF.

OBJECTIVE: To discuss the approach to patients diagnosed with growth hormone deficiency (GHD) in childhood during the transition period from puberty to adulthood, focusing on the following: (1) physiology; (2) effects of recombinant human GH (rhGH) interruption/reinstitution after adult height achievement; (3) re-evaluation of somatrotropic axis; (4) management of rhGH reinstitution, when necessary.
SOURCE OF DATA: Narrative review of the literature published at PubMed/MEDLINE until September 2020 including original and review articles, systematic reviews and meta-analyses.
SYNTHESIS OF DATA: Growth hormone is crucial for the attainment of normal growth and for adequate somatic development, which does not end concomitantly with linear growth. Retesting adolescents who already meet the criteria that predict adult GHD with high specificity is not necessary. Patients with isolated GHD have a high likelihood of normal response to GH testing after puberty. Adolescents with confirmed GHD upon retesting should restart rhGH replacement and be monitored according to IGF-I levels, clinical parameters, and complementary exams.
CONCLUSION: Patients with isolated idiopathic GHD in childhood are a special group who must be reevaluated for GHD as many of them have normal GH provocative tests upon retesting after puberty. Patients who confirm the persistence of GHD in the transition period should maintain rhGH replacement in order to reach an ideal peak bone mass, satisfactory body composition, lipid and glucose profiles, and quality of life

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J Pediatr Adolesc Gynecol. 2020 Jun;33:255-59.

DIFFICULTIES IN TRANSITION OF CARE FROM PEDIATRIC TO ADULT GYNECOLOGY PROVIDERS: SHOULD WE MAINTAIN CARE INTO ADULTHOOD?

Osborne C, Mannerfeldt J, Brain P, et al.

There is evidence that transfer of care for older adolescent patients to adult care is associated with a deterioration in health, especially in those with chronic conditions. Because several specific conditions in pediatric and adolescent gynecology continue into adulthood, it is important that patients have a seamless

healthcare transition. In this commentary, it is argued that instead of arranging transfer, long-term retention of patients by the same physician or physician team may be the more caring, patient-centered approach

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J Pediatr Adolesc Gynecol. 2021 Apr;34:176-89.

YOUNG VOICES: SEXUAL HEALTH AND TRANSITION CARE NEEDS IN ADOLESCENTS WITH INTERSEX/DIFFERENCES OF SEX DEVELOPMENT-A PILOT STUDY.

Callens N, Kreukels BPC, van de Grift TC.

STUDY OBJECTIVE: To determine the sexual health and well-being needs of current generations of youth with intersex/Differences of Sex Development (DSD) during transition from pediatric to adult health care.
DESIGN: Qualitative narrative analyses, quantitative descriptives, and questionnaires.
SETTING: Peer support networks and outpatient clinics.
PARTICIPANTS: Eighteen adolescents aged 16-21 years with intersex/DSD.
INTERVENTIONS: Semi-structured interviews and/or survey.
MAIN OUTCOME MEASURES: Youths learning about bodily differences, their sexual experiences and motives (eg, agency, pleasure), body image, sexual communication inside and outside of health care, and perceived gaps between current and ideal transitional care. Quantitative and qualitative content of the surveys and interviews were analyzed to identify key topics.
RESULTS: We found that (1) there is a need for open-minded perceptions of health care providers about what it means to have a sex variation: (2) there is a need for continued support and information about lived realities relating to the diagnosis and treatments as well as experiential aspects of sexuality: and (3) there are communication obstacles with providers in a multidisciplinary team setting as well as with parents.
CONCLUSION: A user-centered care perspective for adolescents with sex variations includes their stories and feedback toward service improvement. This pilot study shows that adolescents want to be more involved in their sexual health care in ways that connect to their specific questions. Their desire for information increases as they grow older, and underlines the most essential lesson that health care providers can bring their patients about their bodies: how to care for, respect, and enjoy them

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J Pediatr Adolesc Gynecol. 2021 Jun;34:412-14.

TRANSITION CARE FROM ADOLESCENCE TO ADULTHOOD: A 10-YEAR SERVICE REVIEW OF THE GYNECOLOGICAL IMPLICATIONS FOR YOUNG WOMEN AND GIRLS BORN WITH CLOACAL ANOMALIES.

Blyth UEB, Lall A, Jaffray B, et al.

STUDY OBJECTIVE: To establish the gynecological and reproductive outcomes for girls born with a cloacal anomaly, seen in a pediatric specialist cloaca clinic.
DESIGN: Local approval was granted to conduct this review. Outcomes were retrospectively identified using healthcare records.
PARTICIPANTS: Girls with known cloacal anomaly, seen in the cloaca clinic between 2009 and 2019, who had attained menarche or received gynecological input.
RESULTS: Nine females met the inclusion criteria, who were 12-30 years old. The mean age of menarche was 12 years (SD = 1.29). Two developed obstructed menstruation, requiring surgical intervention. Vaginal stenosis affected all women. Three women underwent revision surgery, and 1 is awaiting surgery. None of the women have attempted a pregnancy, to our knowledge.
CONCLUSION: Cloacal anomaly is a rare complex condition. Female individuals with cloacal anomaly require multidisciplinary gynecology specialist care throughout adolescence and adulthood. Provision of a dedicated gynecological service could improve the quality of life of these women

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J Pediatr Endocrinol Metab. 2020 Sep;33:1165-71.

READINESS FOR TRANSITION TO ADULT CARE IN ADOLESCENTS AND YOUNG ADULTS WITH TURNER SYNDROME.

Sheanon NM, Beal SJ, Kichler JC, et al.

Objectives Turner syndrome (TS) is a complex and chronic medical condition that requires lifelong subspecialty care. Effective transition preparation is needed for successful transfer from pediatric to adult care in order to avoid lapses in medical care, explore health issues such as fertility, and prepare caregivers as adolescents take over responsibility for their own care. The objective of this study was to evaluate accuracy of knowledge of personal medical history and screening guidelines in adolescents and young adults (AYA) with TS.
Methods This was a prospective cross-sectional study of 35 AYA with TS of ages 13-22 years recruited from a tertiary care center. AYA completed questionnaires on personal medical history, knowledge of screening guidelines for TS, and the Transition Readiness Assessment Questionnaire (TRAQ).

Results Eighty percent of AYA with TS were 100% accurate in reporting their personal medical history. Only one-third of AYA with TS were accurate about knowing screening guidelines for individuals with TS. Accuracy about knowing screening guidelines was significantly associated with TRAQ sum scores ($r = 0.45$, $p < 0.05$). However, there was no association between knowledge of personal medical history and TRAQ sum scores.
Conclusions Transition readiness skills, TS-specific knowledge, and accurate awareness of health-care recommendations are related, yet distinct, constructs. Understanding of one's personal medical history is not an adequate surrogate for transition readiness. Validated tools for general transition, like the TRAQ, can be used but need to be complemented by TS-specific assessments and content. Providers are encouraged to identify opportunities for clinical and educational interventions well in advance of starting transfer to adult care

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J Pediatr Gastroenterol Nutr. 2020 Dec;71:704-06.
HEALTH CARE TRANSITION: A TIME OF INCREASED VULNERABILITY FOR PEDIATRIC LIVER TRANSPLANT RECIPIENTS.
Vittorio J.
Improvements in pediatric liver transplantation (LT) have led to an increased number of patients reaching young adulthood. Young adult LT recipients transferring from pediatric to adult models of care have increased rates of rejection, graft loss, and medical complications. The goal of a health care transition program is to optimize health and assist youth in reaching their full potential. The means to achieve this goal requires an organized transition process to support youth in acquiring independent health care skills, preparing for an adult model of care, and transferring to new providers without disruption in treatment. This can only be achieved through a multidisciplinary approach to transition planning. This is often a labor and resource-intensive undertaking, which may not receive the necessary support from local institutions. Widespread implementation requires the assistance and endorsement from governing organizations at the national and international level

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J Pediatr Nurs. 2020 Nov;55:e279-e285.
A CROSS-SECTIONAL SURVEY ON THE TRANSITIONAL CARE OF ADOLESCENTS WITH INFLAMMATORY BOWEL DISEASE IN HUNGARY.
ErÅ‘s A, Veres G, TÃ¡rnok A, et al.
PURPOSE: Since little is known about transitional care practices of adolescents with inflammatory bowel diseases (IBD) in Central-Eastern Europe, we aimed to investigate the currently applied transition practices in Hungary.
DESIGN AND METHODS: A nationwide, multicentre survey was conducted with the invitation of 41 pediatric and adult IBD centres in February 2019.We developed a 34-item questionnaire, which included single- and multiple-choice questions related to the current clinical practice of IBD transition.
RESULTS: The overall response rate was 31.7% (13/41); answers came predominantly from tertiary centres. Only 15.4% of the respondent centres followed international IBD guidelines. The majority of the IBD centres provided transition support; however, responses revealed a marked heterogeneity of these services. Joint visits were held only in 54% of the clinics. Gastroenterologists and next of kin are not provided education regarding transition across most centres (85 and 92%). Although adolescents received age-specific education, transition readiness was not measured. More IBD nurses and dietitians were employed in adult centres than in pediatric ones.
CONCLUSIONS: The current survey revealed critical gaps in the Hungarian IBD transition practices. As the beneficial effects of structured IBD transition programmes are recognized in Hungary, there is a growing need for the introduction of new, more effective transition practices.
PRACTICE IMPLICATIONS: Our results can serve as a basis for planning more effective transition strategies

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J Pediatr Nurs. 2020 Jul;53:e41-e48.
"A BRIDGE OVER TROUBLED WATER": NURSES' LEADERSHIP IN ESTABLISHING YOUNG ADULTS' TRUST UPON THE TRANSITION TO ADULT RENAL-CARE - A DUAL-PERSPECTIVE QUALITATIVE STUDY.
Gabay GG, Tarabeih M.
PURPOSE: Patient trust is strongly related to adherence, but has not been tested in transitional care. Low adherence post-transitions of young adults from pediatrics to adult renal care jeopardizes transplanted kidneys and quality of life. We aimed at identifying barriers to trust of young adults in nurses and trust-building elements upon and post transition.
DESIGN AND METHODS: Following IRB approval, we recruited 21 young adults who underwent kidney transplants before the transition to adult renal care and eleven nurses from adult care in two Israeli tertiary hospitals that perform kidney transplants. We conducted 42 in-depth narrative interviews with young adults and one interview with each nurse. We used thematic analysis guided by Meleis's framework of effective transitions.

RESULTS: Most young adults' attributed negative meanings to the transition to adult care which did not enhance their well-being. Young adults were not provided with resources to promote their autonomy and role-sufficiency; despite preparation processes pre-transition, they were not aware of expectations from them. Their own unmet expectations of clinicians made them feel unsafe, objectified, and helpless and resulted in distrust in professionals, low adherence, and in some cases, dropping out of care and lower quality of life.
CONCLUSIONS: Nurses who focused on building a relationship with young adults rather than on operational tasks established trust and led young adults towards role-sufficiency, satisfaction with care, adherence, and optimized quality of life.
PRACTICE IMPLICATIONS: The proposed recommendations for nurses and clinicians structure the trust-building process using elements to improve transitional care

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J Pediatr Nurs. 2020 Dec;58:1-8.
EVALUATION OF THE TRXANSITION INDEX-PARENT VERSION FOR ASSESSMENT OF READINESS TO TRANSITION TO ADULT CARE AMONG YOUTH WITH CHRONIC CONDITIONS.
Hart LC, et al .
Purpose: To develop and evaluate a parent-proxy measure of youth HCT readiness: the TRxANSITION Index-Parent Version.
Design and methods: We recruited parents (77% female) and youth (ages 12 to 25) to complete transition readiness measures during outpatient clinic visits. The TRxANSITION Index-Parent Version contains two domains: the Parent Knowledge Domain assessing a parent's knowledge of their youth's illness, and the Parent Proxy Domain, which provides a parental perspective regarding a youth's transition readiness skills. We evaluated the TRxANSITION Index - Parent Version for differences between parent and youth reports of HCT readiness, associations between parent's score and youth's characteristics, and item-category, item-sub-index, and sub-index category correlations.
Results: Data from 93 parents-youth dyads were analyzed. Parents scored significantly higher than youth in the Parent Knowledge Domain and similarly in the Parent Proxy Domain. Parents of daughters had significantly higher scores in the Parent Knowledge Domain than parents of sons and reported similar scores to Parents of sons in the Parent Proxy Domain. Only the self-management sub-index significantly correlated with youth's age. The sub-index-domain, item-sub-index, and item-domain correlations assessed were generally large in magnitude ($r > 0.5$).
Conclusions: The TRxANSITION Index-Parent Version shows promise as a means of assessing parent knowledge of a youth's illness and may provide an accurate proxy assessment of a youth HCT readiness skills. Practice implications: Obtaining parental perspective on a youth's HCT readiness may provide useful clinical information during the transition process

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J Pediatr Nurs. 2020 Nov;55:201-10.
YOUTHS' EXPERIENCES OF TRANSITION FROM PEDIATRIC TO ADULT CARE: AN UPDATED QUALITATIVE METASYNTHESIS.
Varty M, Speller-Brown B, Phillips L, et al.
PROBLEM: Improvements in chronic disease management has led to increasing numbers of youth transitioning to adult healthcare. Poor transition can lead to high risks of morbidity and mortality. Understanding adolescents and young adults (AYA) perspectives on transition is essential to developing effective transition preparation. The aim of this metasynthesis was to synthesize qualitative studies assessing the experiences and expectations of transition to adult healthcare settings in AYAs with chronic diseases to update work completed in a prior metasynthesis by Fegran, Hall, Uhrenfeldt, Aagaard, and Ludvigsen (2014).
ELIGIBILITY CRITERIA: A search of PubMed, Medline, PsycINFO, and CINAHL was conducted to gather articles published after February 2011 through June 2019.
SAMPLE: Of 889 articles screened, a total of 33 articles were included in the final analysis.
RESULTS: Seven main themes were found: developing transition readiness, conceiving expectations based upon pediatric healthcare, transitioning leads to an evolving parent role, transitioning leads to an evolving youth role, identifying barriers, lacking transition readiness, and recommendations for improvements.
CONCLUSIONS: Findings of this metasynthesis reaffirmed previous findings. AYAs continue to report deficiencies in meeting the Got Transition® Six Core Elements. The findings highlighted the need to create AYA-centered transition preparation which incorporate support for parents.
IMPLICATIONS: Improvements in transition preparation interventions need to address deficiencies in meeting the Got Transition® Six Core Elements. More research is needed to identify and address barriers implementing the transition process

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J Pediatr Nurs. 2021 Apr;59:173-80.

ADULT PROVIDER PERSPECTIVES ON TRANSITION AND TRANSFER TO ADULT CARE: A MULTI-SPECIALTY, MULTI-INSTITUTIONAL EXPLORATION.

Gray W, Dorriz P, Kim H, et al.

PURPOSE: To identify barriers that transcend multiple adult care specialties and identify potential solutions.

DESIGN AND METHODS: Twenty-one adult care providers practicing in the specialty areas of internal medicine, family medicine, gastroenterology, endocrinology, and neurology participated in one of six semi-structured focus group interviews. Data were coded and analyzed according to the Socio-ecological Model of Adolescent/Young Adult Readiness for Transition (SMART).

RESULTS: Three themes and one subtheme emerged from the data. These fell within the beliefs/expectations, knowledge, access/insurance, and relationships (subtheme) domains of the SMART model. Family beliefs/expectations regarding the provider role, difficulty accessing reliable information, and limited access to mental health and behavioral providers reportedly affect providers' ability to provide optimal health care.

CONCLUSIONS: Adult providers identified several barriers affecting their ability to care for newly transferred patients. Increased education of families and improved methods of communication between providers were recommended. Barriers related to access and insurance are common and require larger systems-level collaborations between health care systems and payor sources.

PRACTICAL IMPLICATIONS: Some recommendations (e.g., educating families on the distinct roles of the PCP vs. specialist, highlighting new treatment opportunities in adult care, conveying trust and endorsing the new provider), represent concrete steps pediatric providers can immediately take to improve transfer. Other steps will require forging bridges across the pediatric and adult care world to expand patient access to medical, mental health, and behavioral services

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J Pediatr Nurs. 2021 May;60:164-67.

A PILOT PROJECT: IMPROVING THE TRANSITION CARE PROCESS FOR NEUROSURGICAL ADOLESCENT PATIENTS WITH INDWELLING SHUNTS TO ADULT CARE.

Johnson A, Marks J, Little J.

BACKGROUND: A formal transition program has not been described for neurosurgical adolescent patients with an indwelling shunt device. Transitioning from pediatric neurosurgical care to adult care without transition guidance has caused abrupt transfer of care at this institution. The goal of this pilot transition program was to help patients and caregivers feel informed and prepared for transition.

METHODS: The Got Transition®, Six Core Elements of Transition, were used to create this program. Both a policy and a registry to track and monitor patients were created. A validated questionnaire for transition readiness was measured. Education was provided based on the results of the questionnaire to prepare the adolescent for transfer of care. A smartphone application was used to promote health care independence. Transfer to adult neurosurgical care included hand-off between the pediatric and adult teams, child life and social work involvement, and scheduled follow up with an adult neurosurgical provider.

FINDINGS: All patients 14 to 18Â years with indwelling shunts were enrolled in the pilot program. Eight patients completed a baseline transition readiness assessment, received education and anticipatory guidance, and downloaded the smartphone application. At the end of the six month pilot, three patients were successfully transferred to adult care.

DISCUSSION: The integration of a transition readiness questionnaire and smart phone application during this pilot program was feasible and continues to be used at this institution. Adolescent patients with shunts require gradual and carefully planned transition services

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J Pediatr Nurs. 2021 Mar;57:e79-e84.

THE EFFECT OF SELF-EFFICACY, SOCIAL SUPPORT AND QUALITY OF LIFE ON READINESS FOR TRANSITION TO ADULT CARE AMONG ADOLESCENTS WITH CYSTIC FIBROSIS IN TURKEY.

Torun T, et al.

PURPOSE: The aim of this study is to examine the effects of self-efficacy, social support and quality of life on readiness for transition to adult care in adolescents with cystic fibrosis.

DESIGN AND METHODS: A descriptive and cross-sectional study design was used. Data were collected from 50 adolescent between 14 and 17Â years old with cystic fibrosis, by using The Transition Readiness Assessment Questionnaire, Social Support Appraisals Scale for Children, Self-Efficacy Questionnaire for Children and health-related quality-of-life instrument, the KIDSCREEN-10.

RESULTS: A positive correlation was found between the readiness levels of adolescents for transition to adult care and self-efficacy levels. In path analysis, self-efficacy was found to have a significant effect on the level of readiness for transition to adult care. There was not statistically significant relationship between the level of readiness for transition to adult care and health-related quality of life and perceived social support. Path

analysis revealed that health-related quality of life and perceived social support had significant effects on the self-efficacy levels of adolescents.
CONCLUSIONS: Self-efficacy were associated with readiness for the transition to adult care. Although perceived social support and quality of life were not related with transition readiness these variables had significant effects on perceived self-efficacy, which was determined as a factor affecting the readiness for the transition to adult care.
PRACTICE IMPLICATIONS: In adolescents with cystic fibrosis, self-efficacy, social support and quality of life levels should be taken into account when planning preparation programs for transition to adult care

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J Pediatr Nurs. 2021 Mar;61:7-14.
CARE COORDINATION FOR CHILDREN WITH SPECIAL HEALTHCARE NEEDS ANTICIPATING TRANSITION: A PROGRAM EVALUATION.
Morton B, Damato EG, Ciccarelli MR, et al.
PURPOSE: Nearly 20% of U.S. children have special healthcare needs (CSHCN). Difficulties experienced with navigating the array of services for these children has highlighted the value of care coordination to improve care, reduce costs and increase satisfaction. This study evaluated the services delivered within a care coordination program at a transition consultation center for CSHCN. It also compared the advancement of youth by age group toward graduation criteria.
DESIGN AND METHODS: Using a program evaluation method, data were collected via a retrospective chart review. The convenience sample included clinical records from 100 patients aged 11-22 who had a chronic disease or disability.
RESULTS: The comparison of services for those with diagnoses of autism spectrum disorder, cerebral palsy and Down syndrome were uniformly high in supporting primary care and health care financing. Medicaid waiver assistance was provided more frequently to younger adolescents while older adolescents more commonly received support in all other graduation criteria, including primary and specialty care, healthcare financing and decision-making supports.
CONCLUSIONS: Youth served in a transition care coordination program receive a high volume and broad array of services. There are some variations in the types of services by diagnosis and level of support need. Older youth show greater advancement toward graduation criteria.
PRACTICE IMPLICATIONS: This in-depth chart review provides a valuable description of the activities of care coordinators serving CSHCN enduring transition. It enables development of targeted strategies for building care coordination programming and sets an example for the design of future research studies on this topic

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J Pediatr Psychol. 2021 Feb;46:197-207.
HEALTH CARE UTILIZATION, TRANSITION READINESS, AND QUALITY OF LIFE: A LATENT CLASS ANALYSIS.
Traino KA, Sharkey CM, Perez MN, et al.
OBJECTIVE: To identify possible subgroups of health care utilization (HCU) patterns among adolescents and young adults (AYAs) with a chronic medical condition (CMC), and examine how these patterns relate to transition readiness and health-related quality of life (HRQoL).
METHODS: Undergraduates (N = 359; Mage=19.51 years, SD = 1.31) with a self-reported CMC (e.g., asthma, allergies, irritable bowel syndrome) completed measures of demographics, HCU (e.g., presence of specialty or adult providers, recent medical visits), transition readiness, and mental HRQoL (MHC) and physical HRQoL (PHC). Latent class analysis identified four distinct patterns of HCU. The BCH procedure evaluated how these patterns related to transition readiness and HRQoL outcomes.
RESULTS: Based on seven indicators of HCU, a four-class model was found to have optimal fit. Classes were termed High Utilization (n = 95), Adult Primary Care Physician (PCP)-Moderate Utilization (n = 107), Family PCP-Moderate Utilization (n = 81), and Low Utilization (n = 76). Age, family income, and illness controllability predicted class membership. Class membership predicted transition readiness and PHC, but not MHC. The High Utilization group reported the highest transition readiness and the lowest HRQoL, while the Low Utilization group reported the lowest transition readiness and highest HRQoL.
CONCLUSIONS: The present study characterizes the varying degrees to which AYAs with CMCs utilize health care. Our findings suggest poorer PHC may result in higher HCU, and that greater skills and health care engagement may not be sufficient for optimizing HRQoL. Future research should examine the High Utilization subgroup and their risk for poorer HRQoL

J Pediatr Psychol. 2021 Jan;46:1-11.

TRANSITION TO ADULT IBD CARE: A PILOT MULTI-SITE, TELEHEALTH HYBRID INTERVENTION.

Gray WN, Wagoner ST, Schaefer MR, et al.

Objective: Transition to adult IBD care continues to be a challenge. Efficacious models of improving transition to adult care in the United States are lacking. We present data from a pilot, prospective, non-randomized, intervention implemented at IBD centers in the Midwest and Southeast United States.

Design and methods: Adolescents and young adults (AYAs; 16-20 years) with IBD and their parents completed a 4- to 5-month transition program (1 in-person group session; 4 individual telehealth sessions). Primary outcomes were feasibility (i.e., recruitment, retention, fidelity) and acceptability (i.e., program satisfaction). Secondary outcomes were changes in transition readiness, self-management skill acquisition, perceived readiness to transfer to adult care, and disease knowledge.

Results: The study exceeded goals for recruitment (target N = 20; actual: 36) and retention (target: 80%; actual: 86.11%). On average, it took participants 20.91 ± 3.15 weeks to complete our 4- to 5-month intervention and there were no deviations from the study protocol. Participant ratings for overall program satisfaction, perceived helpfulness, and program length and format were positive. Increases in transition readiness, $t(30) = 8.30$, $d = 1.49$, $p < .001$, self-management skill acquisition, $t(30) = 3.93$, $d = 0.70$, $p < .001$, and disease knowledge, $t(30) = 8.20$, $d = 1.58$, $p < .001$ were noted. AYA- and parent-perceived transfer readiness also improved (p's < .05; d's = 0.76-1.68).

Conclusions: This article presents feasibility and acceptability data for a 4- to 5-month transition intervention. Improvements in AYA transition readiness, self-management skill acquisition, IBD knowledge, and AYA/parent perceived transfer readiness were also observed

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J Pediatr Rehabil Med. 2021;14:103-12.

OPINIONS ON REHABILITATION CARE OF YOUNG ADULTS WITH TRANSVERSAL UPPER LIMB REDUCTION DEFICIENCY IN THEIR TRANSITION TO ADULTHOOD.

Huurneman KAM, Lankhorst IMF, Baars ECT, et al.

PURPOSE: Young adults with transversal upper limb reduction deficiency experience limitations regarding education, employment and obtaining a driver's license. Contribution of rehabilitation care within these domains has been reported to be inadequate. This study evaluates the needs and suggestions of participants in rehabilitation care.

METHODS: Two online focus groups with young adults and parents met during 4 consecutive days. Health care professionals joined a face-to-face focus group. Data analysis was based on framework analysis.

RESULTS: The rehabilitation team was mainly consulted for problems with residual limb or for prostheses. Young adults and their parents were mostly unaware of resources regarding education, job selection or obtaining a driver's license. Professionals stated that these subjects were addressed during periodic appointments. Young adults didn't always attend these appointments due to limited perceived benefit. To improve rehabilitation care, participants suggested methods for providing relevant information, facilitating peer contact and offering dedicated training programs to practice work-related tasks, prepare for job interviews or enhance self-confidence.

CONCLUSION: Periodic appointments do not fulfil needs of young adults with transversal upper limb reduction deficiency. To improve care, rehabilitation teams should offer age-relevant information, share peer stories, and create dedicated training programs

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J Pediatr Surg. 2021 Feb;56:257-62.

REACHING ADULTHOOD WITH HIRSCHSPRUNG'S DISEASE: PATIENT EXPERIENCES AND RECOMMENDATIONS FOR TRANSITIONAL CARE.

Hoel AT, Tofft L, BjÃ¸rnland K, et al.

BACKGROUND/PURPOSE: The need for transitional care has gained increased focus in the treatment of patients with congenital colorectal disorders. We aimed to acquire in-depth knowledge about the experiences of adult patients with Hirschsprung's disease (HD) and their suggestions for transitional care.

METHODS: Binational study applying gender equal focus group interviews (FGI).

RESULTS: Seventeen (9 men) of 52 invited patients with median age 29 (19-43) years participated. Three themes evolved from the FGI. "Scarred body and soul" describes the somatic and psychosocial challenges the patients experienced and "limited health literacy on HD" refers to the patients' lack of HD knowledge. "Absent transition" depicts missing transitional care and the patients' inability to find adult HD specialists. The adult HD patients strongly recommended transitional care from early teens with focus on information about HD and establishment of a peer-to-peer program. They also emphasized the possibility of being referred to a pelvic floor center.

CONCLUSIONS: HD negatively influences patients' somatic and psychosocial health in childhood, adolescence and adulthood. Adult HD patients strongly recommend transitional care from early teens and the possibility for referral to a center working with pelvic floor dysfunctions.
LEVEL OF EVIDENCE: IV TYPE OF RESEARCH: Clinical

J Pediatr Urol. 2021 Apr;17:155.
Response to Commentary re 'Barriers in transitioning urologic patients from pediatric to adult care'.
Roth JD, Claeys W, Hoebeke P.

J Pediatr Urol. 2021 Apr;17:144-52.
BARRIERS IN TRANSITIONING UROLOGIC PATIENTS FROM PEDIATRIC TO ADULT CARE.
Claeys W, Roth JD, Hoebeke P.
As the advances in medicine continue to emerge, more children with congenital or pediatric-onset chronic urologic conditions are surviving well into adulthood. This imposes an ever rising there is a need for adequate transition of these patients from pediatric to adult care. Despite position statements from multiple heath care organizations and several models proposed in literature, different issues and gaps in urologic transition continue to exist. Major barriers in this transition are adolescence, a challenging time that is characterized by impulsive behavior and risk taking, and the longstanding relation between both patients and paediatric providers. Both pediatric and adult care providers need to be aware of the special needs of maturing youth with chronic care problems related to education, self-management, legal issues and psychological support during care transition. Furthermore, they need to understand and address the currently existing obstacles for adequate transition. There is need for active communication with each other and the patient to develop sustainable relationships that can support the transitioning process. It is therefore in the greatest interest of the care provider to make this transition as smooth as possible. This paper aims to point out the currently perceived barriers in care transition within the urological context, reflect on previous implemented models for care transition and present proposals for improvement

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J Pediatr Urol. 2021 Apr;17:156-57.
Letter to Editor regarding 'Barriers in transitioning urologic patients from pediatric to adult care'.
Sforza S, Bortot G, Cini C, et al.

J Rheumatol. 2021 Feb.
DIFFERENCES IN HEALTH CARE TRANSITION VIEWS, PRACTICES, AND BARRIERS AMONGST NORTH AMERICAN PEDIATRIC RHEUMATOLOGY CLINICIANS FROM 2010 TO 2018.
Johnson K, Edens C, Sadun RE, et al.
OBJECTIVE: Since 2010, the rheumatology community has developed guidelines and tools to improve healthcare transition. In this study we aimed to compare current transition practices and beliefs among Childhood Arthritis and Rheumatology Research Alliance (CARRA) rheumatology providers with transition practices from a 2010 provider survey published by Chira et al.
METHODS: In 2018, CARRA members completed a 25-item online survey about healthcare transition. Got Transition's Current Assessment of Health Care Transition Activities was used to measure clinical transition processes on a scale of 1 (basic) to 4 (comprehensive). Bivariate analyses were used to compare 2010 and 2018 survey findings.
RESULTS: Over half of CARRA members completed the survey (217/396), including pediatric rheumatologists, adult- and pediatric-trained rheumatologists, pediatric rheumatology fellows, and advanced practice providers. The most common target age to begin transition planning was 15-17 (49%). Most providers transferred patients prior to age 21 or older (75%). Few providers used the American College of Rheumatology transition tools (31%) or have a dedicated transition clinic (23%). Only 17% had a transition policy in place, and 63% did not consistently address healthcare transition with patients. When compared to the 2010 survey, improvement was noted in three of twelve transition barriers: availability of adult primary care providers, availability of adult rheumatologists, and pediatric staff transition knowledge and skills ($p<0.001$ for each). Nevertheless, the mean Current Assessment score was less than 2 for each measurement.
CONCLUSION: This study demonstrates improvement in certain transition barriers and practices since 2010, although implementation of structured transition processes remains inconsistent

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J Sex Med. 2021 Mar;18:632-45.

INDIVIDUAL TREATMENT PROGRESS PREDICTS SATISFACTION WITH TRANSITION-RELATED CARE FOR YOUTH WITH GENDER DYSPHORIA: A PROSPECTIVE CLINICAL COHORT STUDY.

Nieder TO, Mayer TK, Hinz S, et al.

BACKGROUND: The number of adolescents presenting with gender dysphoria (GD) in healthcare services has increased significantly, yet specialized services offering transition-related care (TRC) for trans youth is lacking.

AIM: To investigate satisfaction with TRC, regret, and reasons for (dis)satisfaction with transition-related medical interventions (TRMIs) in trans adolescents who had presented to the Hamburg Gender Identity Service for children and adolescents (Hamburg GIS).

METHODS: Data were collected from a clinical cohort sample of 75 adolescents and young adults diagnosed with GD (81% assigned female at birth) aged 11 to 21Â years (MÂ =Â 17.4) at baseline and follow-up (on a spectrum of ongoing care, on average 2Â years after initial consultation). To determine progress of the youth's medical transitions, an individual treatment progress score (ITPS) was calculated based on number of desired vs received TRMIs.

OUTCOMES: Main outcome measures were satisfaction with TRC at the time of follow-up, ITPS, social support, reasons for regret and termination of TRC, and (dis)satisfaction with TRMIs.

RESULTS: Participants underwent different stages of TRMIs, such as gender-affirming hormone treatment or surgeries, and showed overall high satisfaction with TRC received at the Hamburg GIS. Regression analysis indicated that a higher ITPS (an advanced transition treatment stage) was predictive of higher satisfaction with TRC. Sex assigned at birth, age, and time since initial consultation at the clinic showed no significant effects for satisfaction with TRC, while degree of social support showed a trend. No adolescents regretted undergoing treatment at follow-up. Additional analysis of free-text answers highlighted satisfaction mostly with the physical results of TRMI.

CLINICAL IMPLICATIONS: Because youth were more satisfied with TRC when their individual transition (ITPS) was more progressed, treatment should start in a timely manner to avoid distress from puberty or long waiting lists.

STRENGTHS AND LIMITATIONS: This study is one of the first to report on treatment satisfaction among youth with GD from Europe. The ITPS allowed for a more detailed evaluation of TRMI wishes and experiences in relation to satisfaction with TRC and may close a gap in research on these treatments in adolescent populations. However, all participants were from the same clinic, and strict treatment eligibility criteria may have excluded certain trans adolescents from the study. Low identification rates with non-binary identities prevented comparisons between non-binary and binary genders.

CONCLUSION: The study highlights the role of TRMI and individual treatment or transition progress for youth's overall high satisfaction with TRC received at the Hamburg GIS. Nieder TO, Mayer TK, Hinz S, et al. Individual Treatment Progress Predicts Satisfaction With Transition-Related Care for Youth With Gender Dysphoria: A Prospective Clinical Cohort Study. J Sex Med 2021;18:632-645

J Transcult Nurs. 2021 Jan;32:21-29.

SOCIAL AND PSYCHOLOGICAL FACTORS ASSOCIATED WITH HEALTH CARE TRANSITION FOR YOUNG ADULTS LIVING WITH SICKLE CELL DISEASE.

Clayton-Jones D, Matthie N, Treadwell M, et al.

Introduction: Due to advances in disease management, mortality rates in children with sickle cell disease (SCD) have decreased. However, mortality rates for young adults (YA) increased, and understanding of social and psychological factors is critical. The aim of this study was to explore factors associated with health care transition experiences for YA with SCD.

Method: This was a qualitative descriptive study. A 45-minute semistructured interview was conducted with 13 YA (M = 21.5 years, SD = 1.73).

Results: Results suggest that social and psychological factors and self-management experiences influence health care transition. Eight themes emerged: "need for accessible support"; "early assistance with goal setting"; "incongruence among expectations, experiences, and preparation"; "spiritual distress"; "stigma"; "need for collaboration"; "appreciation for caring providers"; and "feeling isolated."

Discussion: Consideration of cultural contexts will guide nurses in supporting health care transition. Designing culturally relevant interventions that address unique needs for YA living with SCD is warranted

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JBI Evid Synth. 2021 Mar.

THE EXTENT SELF-MANAGEMENT FOR YOUTH AND YOUNG ADULTS WITH SPECIAL HEALTH CARE NEEDS IS ADDRESSED IN HEALTH CARE TRANSITION PLANNING LITERATURE: A SCOPING REVIEW PROTOCOL.

Betz CL, Mannino JE, Cleverley K, et al.

OBJECTIVE: The purpose of this scoping review is to explore the extent to which self-management of youth and young adults with special health care needs is reported in the health care transition literature.

INTRODUCTION: It is essential for youth and young adults with special health care needs to learn the self-management skills, to the extent possible, that are essential in maintaining the stability of their chronic condition to seamlessly transfer to adult care and live independently. Acquisition of self-management competencies for chronic care management is an essential component of health care transition preparation.

INCLUSION CRITERIA: The inclusion criteria will be based upon age and condition designation. The age range of participants will include youth and young adults, aged nine to 35â€Šyears, who have a special health care needs. Inclusion criteria consists of both non-categorical and diagnostic specific terminology for youth and young adults with a childhood acquired chronic condition. Non-categorical terms used include "long-term chronic condition," "special health care needs," "medical complex condition," "complex care needs," "developmental disability," "intellectual disability," "mental health condition," "emotional disabilities," "physical disabilities," "chronic illness," and "chronic condition."

METHODS: The following databases will be accessed for this health care transition scoping review: CINAHL, Cochrane CENTRAL, Embase, Ovid MEDLINE, PsycINFO, and Web of Science. Relevant gray literature will be accessed as well. The Covidence software platform will be used to review citations and full text articles. Two reviewers will independently review abstracts and full texts of studies, and extract data using the data extraction tool. Any conflicts will be resolved with a third reviewer. Review findings will be presented in tabular format and narrative synthesis based upon the scoping review objective

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J Child Health Care. 2020 Sep;1367493520953649.

SURVEY OF US PEDIATRIC NURSES' ROLE IN HEALTH CARE TRANSITION PLANNING: FOCUS ON ASSESSMENT OF SELF-MANAGEMENT ABILITIES OF YOUTH AND YOUNG ADULTS WITH LONG-TERM CONDITIONS.

Betz CL, Mannino JE, Disabato JA.

The survival rates of youth and young adults (YYAs) diagnosed with long-term conditions have improved considerably as 90% now enter adulthood; health care transition planning (HCTP) has emerged as a nursing practice priority. The aim of this national online survey was to investigate the extent to which nurses, recruited from two major United States pediatric nursing organizations are involved with HCTP including assessing YYA self-management abilities (SMA). Findings of a 9-item assessment of self-management abilities subscale of the nurses' role in HTCP tool are reported. The nurse respondents (n = 1269), identified the most frequently assessed SMA was the YYAs' ability to understand and speak about their condition and its treatment (M = 2.3, SD = .89). The least frequently assessed was the YYAs' ability to identify community advocates to help them become more independent (M =1.5, SD = .90). Regression analysis identified significant predictors of the frequency nurses assess YYA for SMA included nurses' level of knowledge, perceived level of importance, HCTP and skills identified in job description, and caring for YYA. Findings indicate HCTP care advancements will necessitate HCTP training and development of nurse-led service efforts to facilitate optimal outcomes for YYA

JMIR Form Res. 2021 May;5:e22915.

A PROVIDER-FACING EHEALTH TOOL FOR TRANSITIONING YOUTH WITH SPECIAL HEALTH CARE NEEDS FROM PEDIATRIC TO ADULT CARE: MIXED METHODS, USER-ENGAGED USABILITY STUDY .

McMaughan DJ, Lin S, Ozmetin J, et al.

BACKGROUND: There is a need for medical education on health care transitions for youth with special health care needs. The Texas Transition Toolkit (the tool) supports providers through a one-stop shop for researching literature on care transitions, a catalog of care transition tools, and guides for developing care transition programs.

OBJECTIVE: This study aims to assess the functionality and usability of the tool with providers working with transition-aged children and youth with special health care needs (representative users).

METHODS: The tool was evaluated using a triangulated mixed methods case study approach consisting of a concurrent think-aloud phase, a satisfaction survey, and a survey of problem relevance and task performance to operationalize and capture functionality and usability. Our mixed methods deep dive into the functionality and usability of the tool focused on 10 representative users from one medical home in Texas and 5 website design experts.

RESULTS: Representative users found the tool to be highly relevant, as demonstrated by the satisfaction score for relevance (138/150, 92%). According to the users, the tool provided comprehensive information

related to health care transitions for youth with special health care needs, with a satisfaction score of 87.3% (131/150) for comprehensive. Overall satisfaction with the tool was high at 81.92% (1065/1300) with a cutoff score of 73.33% (953.4/1300) indicating high satisfaction, but users reported relatively lower satisfaction with search (114/150, 76%) and navigation (ease of use: 114/150, 76%; hyperlinks: 163/200, 81.5%; structure: 159/200, 79.5%). They experienced search- and navigation-related problems (total problems detected: 21/31, 68%) and, based on quality checks, had a relatively low task completion rate for tasks involving finding information (60/80, 75%), which required searching and navigation. The problems identified around search and navigation functionality were relevant (relevance scores ranging from 14.5 to 22, with a cutoff score of 11.7 indicating relevance).
CONCLUSIONS: The tool may help bridge the gaps in training on health care transitions for youth with special health care needs in US medical education. The tool can be used to create structured protocols to help improve provider knowledge, collaboration across pediatric and adult care providers, and the continuity of care as youth with special health care needs transition from pediatric to adult care. The results provided a road map for optimizing the tool and highlighted the importance of evaluating eHealth technologies with representative users

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JMIR Res Protoc. 2021 Apr;10:e24565.
AN INTERVENTION FOR THE TRANSITION FROM PEDIATRIC OR ADOLESCENT TO ADULT-ORIENTED HIV CARE: PROTOCOL FOR THE DEVELOPMENT AND PILOT IMPLEMENTATION OF ITRANSITION.
Tanner AE, Dowshen N, Philbin MM, et al.
BACKGROUND: In the United States, adolescents and young adults are disproportionately affected by HIV and have poorer HIV-related health outcomes than adults. Health care transition (HCT) from pediatric or adolescent to adult-oriented HIV care is associated with disruptions to youths' care retention, medication adherence, and viral suppression. However, no evidence-based interventions exist to improve HCT outcomes for youth living with HIV.
OBJECTIVE: There are 2 phases of this project. Phase 1 involves the iterative development and usability testing of a Social Cognitive Theory-based mobile health (mHealth) HIV HCT intervention (iTransition). In phase 2, we will conduct a pilot implementation trial to assess iTransition's feasibility and acceptability and to establish preliminary efficacy among youth and provider participants.
METHODS: The iterative phase 1 development process will involve in-person and virtual meetings and a design team comprising youth living with HIV and health care providers. The design team will both inform the content and provide feedback on the look, feel, and process of the iTransition intervention. In phase 2, we will recruit 100 transition-eligible youth across two clinical sites in Atlanta, Georgia, and Philadelphia, Pennsylvania, to participate in the historical control group (n=50; data collection only) or the intervention group (n=50) in a pilot implementation trial. We will also recruit 28 provider participants across the pediatric or adolescent and adult clinics at the two sites. Data collection will include electronic medical chart abstraction for clinical outcomes as well as surveys and interviews related to demographic and behavioral characteristics; Social Cognitive Theory constructs; and intervention feasibility, acceptability, and use. Analyses will compare historical control and intervention groups in terms of HCT outcomes, including adult care linkage (primary), care retention, and viral suppression (secondary). Interview data will be analyzed using content analysis to understand the experience with use and acceptability.
RESULTS: Phase 1 (development) of iTransition research activities began in November 2019 and is ongoing. The data collection for the phase 2 pilot implementation trial is expected to be completed in January 2023. Final results are anticipated in summer 2023.
CONCLUSIONS: The development and pilot implementation trial of the iTransition intervention will fill an important gap in understanding the role of mHealth interventions to support HCT outcomes for youth living with HIV.
INTERNATIONAL REGISTERED REPORT IDENTIFIER (IRRID): DERR1-10.2196/24565

Joint Bone Spine. 2021 Jan;88:105047.
COMPARISON OF PAEDIATRIC AND ADULT CLASSIFICATION CRITERIA IN JUVENILE IDIOPATHIC ARTHRITIS DURING THE TRANSITION FROM PAEDIATRIC TO ADULT CARE.
Debrach AC, Rougelot A, Beaumel A, et al.
OBJECTIVES: To determine the characteristics of juvenile idiopathic arthritis (JIA) patients seen during the transition period in order to compare paediatric classification criteria with those for adults.
METHODS: Patients with JIA according to the ILAR classification and who had a consultation at transition between 2010 and 2017 were included in a retrospective bi-centre (Lyon, Lausanne) study. JIA classification criteria were compared to ACR/EULAR 2010 criteria for rheumatoid arthritis (RA), Yamaguchi criteria for adult-onset Still's disease (AOSD), ASAS criteria for spondyloarthritis and CASPAR criteria for psoriatic arthritis.

RESULTS: One hundred and thirty patients were included: 13.9% with systemic JIA, 22.3% with polyarticular JIA, 22.3% with oligoarticular JIA, 34.6% with enthesitis-related arthritis (ERA) and 6.9% with psoriatic arthritis; 13.1% had suffered from uveitis; 14.5% of patients had erosions or carpitis, mainly those with psoriatic arthritis, polyarticular or systemic JIA; 37.5% of patients with ERA displayed radiological sacroiliitis. When comparing paediatric JIA criteria with adult classifications, we found that: 66.6% of patients with systemic JIA fulfilled the criteria for AOSD, 87.5% of rheumatoid factor-positive polyarticular JIA and 9.5% of rheumatoid factor-negative polyarticular JIA met the criteria for RA, and 34.5% of oligoarticular JIA fulfilled the criteria for spondyloarthritis. Finally, 77.7% of patients with ERA met the criteria for spondyloarthritis, and 100% of patients with psoriatic arthritis JIA met the criteria for psoriatic arthritis.
CONCLUSION: Oligoarticular JIA and rheumatoid factor-negative polyarticular JIA seem to be paediatric entities, whereas the other types of JIA tended to meet the respective adult classification criteria

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Jpn J Nurs Sci. 2020 Jul;17:e12323.

NURSES' PERCEPTIONS REGARDING TRANSITIONAL CARE FOR ADOLESCENTS AND YOUNG ADULTS WITH CHILDHOOD-ONSET CHRONIC DISEASES.

Suzuki S, Kita S, Morisaki M, et al.

AIM: Nurses are expected to have a role in the transition of care from pediatric to adult medical practices for adolescents and young adults with childhood-onset chronic diseases. This study compares the experience, knowledge, and perceptions regarding the ideal care among adult unit and pediatric nurses regarding the transition to adult care for those with childhood-onset chronic diseases.
METHODS: A cross-sectional study using self-report questionnaires was conducted with nurses in a tertiary hospital in Tokyo. Questions were generated based on a literature review and expert discussion. Data from 1,064 participants were analyzed (adult unit nurses: n = 959, 90.1%; pediatric nurses: n = 105, 9.9%).
RESULTS: Among 623 adult unit nurses who had care experience for adult patients with a childhood-onset chronic disease, 458 nurses (73.6%) were unaware of the concept of transitional care. As the obstructive factors for transition, pediatric nurses recognized problems in healthcare providers' attitudes and lack of transitional care coordinators, while the adult unit nurses emphasized the patients' wishes to continue to receive pediatric healthcare. Most adult unit nurses expected pediatric nurses to function as transitional care coordinators.
CONCLUSION: Adult unit and pediatric nurses had different perceptions of the barriers in transitioning children with chronic diseases to adult care. It is important to have educational programs focusing on transitional care for all nurses, both to enable pediatric nurses to improve transition readiness of children with chronic diseases and to offer adult patients with a childhood-onset chronic disease continuing support through adult unit nurses

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Lancet Child Adolesc Health. 2021 Jan;5:9-11.

TRANSITION OF CARE FOR ADOLESCENTS WITH CHRONIC PAIN.

Kashikar-Zuck S.

LGBT Health. 2021 Apr.

TRANSITION FROM PEDIATRIC TO ADULT CARE FOR TRANSGENDER YOUTH: A QUALITATIVE STUDY OF PATIENT, PARENT, AND PROVIDER PERSPECTIVES.

Pham T, GarcÃ-a A, Tsai M, et al.

Purpose: No information exists on the needs of transgender youth transitioning their gender-affirming health care from pediatric to adult settings. We obtained perspectives of transgender youth, their parents, and providers, and aimed to identify barriers and unmet needs during the transition of care.
Methods: Five online focus groups were conducted between February and March 2019 with separate groups for transgender youth 13-17 and 18-21 years old; parents of transgender youth 13-17 and 18-21 years old; and gender-affirming health care providers. Thematic analysis of transcripts was conducted by two researchers. Pooled Cohen's Îº was 0.83, indicating excellent inter-rater reliability.
Results: Sixty-six participants (29 youth, 27 parents, and 10 providers) identified 10 themes. Themes related to barriers to transition included access and insurance challenges, patient readiness and hesitancy to transfer care, and multidisciplinary-system inefficiencies. Themes related to improving transition focused on prioritizing referrals from trusted sources, establishing gradual patient independence, aligning gender transition goals, and setting impetus for transferring care.
Conclusion: Successful health care transition for transgender youth must consider the intricacies of a complex medical system and challenges that they pose to adolescents' perceived abilities to independently manage health care and willingness to prepare transfer of care. Given that patients, parents, and providers assume important roles during the process, each can uniquely contribute toward ensuring a smooth transition. Efforts

to improve this process should focus on enhancing collaboration between clinics and families through crowdsourcing resources, continued verification of health goals, supporting greater patient autonomy, and delineating an explicit timeline for transition

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Liver Transpl. 2021 Feb.

Reply to: Heldman's National survey of adult transplant hepatologists on the pediatric-to-adult care transition after liver transplantation; What about MedPeds?

El-Baba F, Roxas R.

I decided to write this letter to the editors after a personal patient encounter. My patient is a 22-year-old with a past medical history of autoimmune hepatitis that led to cirrhosis, and he was admitted to my general pediatrics floor team for acalculous cholecystitis. He completed a course of antibiotics, his pain improved and he was discharged. During that hospital visit, his Childs-Pugh was class C and his Model for End-Stage Liver Disease-Sodium was 32. Due to the severity of his illness, I assessed his knowledge about his disorder. He knew his medications well including doses and frequency. However, it was clear that he did not understand the severity of his illness when he told me, "I would like to have a kid in the next year or two."

Lupus. 2020 Sep;29:1206-15.

DISEASE ACTIVITY AND HEALTH-CARE UTILIZATION AMONG YOUNG ADULTS WITH CHILDHOOD-ONSET LUPUS TRANSITIONING TO ADULT CARE: DATA FROM THE PEDIATRIC LUPUS OUTCOMES STUDY.

Haro SL, Lawson EF, Hersh AO.

BACKGROUND: Individuals with childhood-onset systemic lupus erythematosus (cSLE) must transfer from pediatric to adult care. The goal of this study was to examine disease activity and health-care utilization among young adults with cSLE who are undergoing or have recently completed the transfer to adult care.

METHODS: The Pediatric Lupus Outcomes Study (PLOS) is a prospective longitudinal cohort study of young adults aged 18-30 diagnosed with cSLE. We conducted a cross-sectional analysis comparing 47 participants under the care of pediatric rheumatologists to 38 who had completed transfer to adult care. Demographics, disease manifestations, health- care utilization and transition readiness were compared between groups.

RESULTS: Those in the post-transfer group had significantly lower medication usage and were less likely to have seen a rheumatologist in the past year. Disease manifestations, flare rates, and hospitalizations were similar between groups. Nearly a quarter of patients who had transferred to adult care reported difficulties with the process.

CONCLUSION: Post-transfer patients had lower health-care utilization as evidenced by less medication usage and lack of rheumatology follow-up, in spite of the fact that disease activity was similar in both groups. Future studies will assess longitudinal changes in disease activity and damage in this population

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Matern Child Health J. 2020 Jun;24:805.

CORRECTION TO: HEALTH CARE TRANSITION PLANNING AMONG YOUTH WITH ASD AND OTHER MENTAL, BEHAVIORAL, AND DEVELOPMENTAL DISORDERS.

Zablotsky B, Rast J, Bramlett MD, et al.

In the original publication of the article, Figure 1 included footnotes which duplicated information appearing in the figure caption. Therefore the notes of "NOTES: ASD = autism spectrum disorder; MBDD = mental, behavioral, or developmental disorder. Indicators presented are unadjusted estimates. (x) Significantly different than youth with autism spectrum disorder based on adjusted odds ratio ($p < .05$). (y) Significantly different than youth with other mental, behavioral, or developmental disorders based on adjusted odds ratio ($p < .05$)." have been removed. The figure 1 appearing in the original version of the article has been corrected

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Matern Child Health J. 2020 Jun;24:796-804.

HEALTH CARE TRANSITION PLANNING AMONG YOUTH WITH ASD AND OTHER MENTAL, BEHAVIORAL, AND DEVELOPMENTAL DISORDERS.

Zablotsky B, Rast J, Bramlett MD, et al.

OBJECTIVE: To estimate the prevalence of health care transition components among youth with autism spectrum disorder (ASD) aged 12-17 using the 2016 National Survey of Children's Health (NSCH), compared to youth with other mental, behavioral, or developmental disorders (MBDDs) or youth without MBDDs.

METHODS: The 2016 NSCH is a nationally and state representative survey that explores issues of health and well-being among children ages 0-17. Within the NSCH, parents of a subset of youth, ages 12-17, are asked a series of questions about their youth's eventual transition into the adult health care system. The current study

explores components of this transition, comparing youth diagnosed with ASD, youth with other mental, behavioral, or developmental disorders (MBDDs), and youth without MBDDs.
RESULTS: Approximately 1-in-4 youth with ASD had actively worked with their doctor to understand future changes to their health care, significantly less than youth with other MBDDs and youth without MBDDs. Fewer than 2-in-5 youth with ASD had met with their doctor privately or had a parent who knew how their youth would be insured when they reached adulthood.
CONCLUSIONS: The current analysis of a nationally representative sample of youth reveals discrepancies in the proportion of youth with ASD receiving appropriate health care transition planning compared to youth with other MBDDs and youth without MBDDs. These findings suggest the potential for barriers among youth with ASD to effectively transitioning into the adult health care system

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Medicine (Baltimore). 2020 Jul;99:e20899.
HEALTHCARE ACCESS FOR AUTISTIC ADULTS: A SYSTEMATIC REVIEW.
Calleja S, Islam FMA, Kingsley J, et al.
BACKGROUND: People with autism spectrum disorder (ASD) have an increased susceptibility for many chronic health conditions compared with their peers. An increasing number of adolescents are transitioning from pediatric to adult healthcare services. Thus, being able to access appropriate healthcare services that can not only address specific needs of the person but enable them to better manage healthcare conditions and decrease the development of preventable disease is necessary. A systematic review was conducted to identify barriers and enablers of healthcare access for autistic adults.
METHODS: The studies included in the review were quantitative and qualitative and were published between 2003 and 2019. The participants for the review are considered to be adults (over 18 years of age) with a primary diagnosis of ASD.
RESULTS: In total, 1290 studies were initially identified and 13 studies were included based on the inclusion and exclusion criteria outlined in a previous protocol paper. The analysis of these studies identified areas of concern to access appropriate healthcare, such as clinician knowledge, the environment, and life events.
CONCLUSION: Identifying the barriers to healthcare, highlights ways healthcare services can regulate scope of practice, the physical environment, and the process of managing health conditions, thus, autistic adults can strive for optimal health. This review contributes to peer-reviewed evidence for future research and up-to-date information when developing and piloting health interventions for autistic adults.
ETHICS AND DISSEMINATION: There are no human participants, data, or tissue being directly studied for the purposes of the review; therefore, ethics approval and consent to participate is not applicable.
REGISTRATION AND STATUS: PROSPERO 2018 CRD42018116093

MMWR Morb Mortal Wkly Rep. 2020 Aug;69:1180-81.
NOTES FROM THE FIELD: CANDIDA AURIS AND CARBAPENEMASE-PRODUCING ORGANISM PREVALENCE IN A PEDIATRIC HOSPITAL PROVIDING LONG-TERM TRANSITIONAL CARE - CHICAGO, ILLINOIS, 2019.
McPherson TD, Walblay KA, Roop E, et al.

MMWR Morb Mortal Wkly Rep. 2020 Aug;69:1156-60.
SUPPORT FOR TRANSITION FROM ADOLESCENT TO ADULT HEALTH CARE AMONG ADOLESCENTS WITH AND WITHOUT MENTAL, BEHAVIORAL, AND DEVELOPMENTAL DISORDERS - UNITED STATES, 2016-2017.
Leeb RT, Danielson ML, Bitsko RH, et al.
Clinical guidelines recommend that primary care providers (PCPs) provide guidance and support to ensure a planned transition from pediatric to adult health care for adolescents, beginning at age 12 years (1). However, most adolescents do not receive the recommended health care transition planning (2). This is particularly concerning for adolescents with diagnosed mental, behavioral, and developmental disorders (MBDDs) (3), who account for approximately 20% of U.S. adolescents (4). Childhood MBDDs are linked to increased long-term morbidity and mortality; timely health care transition planning might mitigate adverse outcomes (5,6). CDC analyzed pooled, parent-reported data from the 2016 and 2017 National Survey of Children's Health (NSCH), comparing adolescents, aged 12-17 years, with and without MBDDs on a composite measure and specific indicators of recommended health care transition planning by PCPs. Overall, approximately 15% of adolescents received recommended health care transition planning: 15.8% (95% confidence interval [CI] = 14.1%-17.5%) of adolescents with MBDDs, compared with 14.2% (95% CI = 13.2%-15.3%) of adolescents without MBDDs. Relative to peers without MBDDs and after adjusting for age, adolescents with anxiety were 36% more likely to receive recommended health care transition planning, and those with depression were 69% more likely; adolescents with autism spectrum disorder (ASD) were 35% less likely to receive such transition planning, and those with developmental delay* were 25% less likely. Fewer than 20%

of adolescents with MBDDs receiving current treatment met the transition measure. These findings suggest that a minority of adolescents with MBDDs receive recommended transition planning, indicating a potential missed public health opportunity to prevent morbidity and mortality in a population at high risk for health care disengagement (1). Improving access to comprehensive and coordinated programs and services, as well as increasing provider training concerning adolescents' unique mental and physical health care needs (7), could help increase the number of adolescents benefiting from successful health care transitions (4)

Mod Rheumatol. 2021 Apr;1-12.

CHECKLIST FOR RAPID ASSESSMENT OF INDEPENDENCE IN CHILDREN WITH PEDIATRIC RHEUMATIC DISEASES IN TRANSITION TO ADULT MEDICAL CARE.

Miyamae T, Inoue Y, Yamaguchi K.

OBJECTIVES: The Committee for Support of Transition to Adult Medical Care, Pediatric Rheumatology Association of Japan, has developed a checklist for patients with pediatric rheumatologic diseases to evaluate readiness for transition to adult medical care.
METHODS: Using checklists for general pediatric chronic diseases developed by researchers at the Ministry of Health, Labour and Welfare, committee members discussed points for modification or addition specific to pediatric rheumatic diseases and pediatric rheumatism clinical practice.
RESULTS: Three patient-assessment checklists based on patient age and understanding level and a parent-assessment checklist were developed. The checklist for junior high school students and above included a 'Health Education in Adolescence and Young Adulthood' section with items related to sexual knowledge and concerns. Also, items on oral medications and subcutaneous injections management in the 'Self-management of Daily Medical Care,' domain and next medical visits management were added. The checklist for junior high school students included 30 items in seven domains and can be completed within 10 minutes. The checklist was given to 28 children with pediatric rheumatic diseases aged 10 years and older and their mothers.
CONCLUSION: The checklist was developed to share the challenges of independence during transition with patients and parents

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Neurol Clin. 2021 Feb;39:243-56.

THERAPEUTIC PITFALLS IN THE TRANSITION OF NEUROLOGIC PATIENTS FROM PEDIATRIC TO ADULT HEALTH CARE PROVIDERS.

Schnitzler ER, Schneck MJ.

Transitions of care from the pediatric to adult setting are fraught with difficulty. For patients whose neurologic problems began in childhood, there is often a lack of organized multidisciplinary care with the desired neurologic expertise in the adult setting. This monograph highlights those difficulties, reviewing disease-specific instances of the problems with transition from pediatric to adult neurologic care. The use of an arbitrary chronologic age cutoff for transition from pediatric to adult expertise in specific disease state may be a disservice in provision of care, and the disease-specific expertise of providers may outweigh the benefit of an age-related provider focus

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Nurs Open. 2021 https://doi.org/10.1002/nop2.899.

TRANSITIONAL CARE EXPERIENCES OF CAREGIVERS OF PRETERM INFANTS HOSPITALIZED IN A NEONATAL INTENSIVE CARE UNIT: A QUALITATIVE DESCRIPTIVE STUDY.

Ma RH, Zhang Q, Ni ZH, et al.

AIM: To describe the transitional care experiences and nursing needs of caregivers of preterm infants hospitalized in neonatal intensive care units (NICUs).
DESIGN: A qualitative descriptive study.
METHODS: We conducted semi-structured interviews with the 24 caregivers of preterm infants admitted to Children's Hospital, Soochow University. All data were collected by a trained and experienced interviewer. The caregivers' experiences were described using qualitative content analysis.
RESULTS: Six Five themes emerged from the analysis: (a) uncertainty about the disease; (b) anxiety due to restricted visitation; (c) exhaustion from overwork; (d) emotional depression; (e) low care ability; (f) a variety of channels for help and a positive response. This study provides a basis for understanding the needs of their caregivers so that effective coping strategies can be implemented. Nurses' education and practice should focus on understanding the real experiences of the parents of preterm infants during transitional nursing

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Nutrients. 2021 Feb;13.

IMPLEMENTING A TRANSITION PROGRAM FROM PAEDIATRIC TO ADULT SERVICES IN PHENYLKETONURIA: RESULTS AFTER TWO YEARS OF FOLLOW-UP WITH AN ADULT TEAM.

Peres M, Almeida MF, Pinto ÃJ, et al.

We aimed to report the implementation of a phenylketonuria (PKU) transition program and study the effects of follow-up with an adult team on metabolic control, adherence, and loss of follow-up. Fifty-five PKU patients were analysed in the study periods (SP): 2 years before (SP1) and after the beginning of adult care (SP2). Retrospective data on metabolic control and number of clinic appointments were collected for each SP, and protein intakes were analysed. In SP2, three patients (6%) were lost to follow-up. There was a small but statistically significant increase in median number of annual blood spots from SP1 to SP2: 11 (7-15) vs. 14 (7-20); p = 0.002. Mean ± SD of median blood Phe remained stable (525 ± 248 µmol/L vs. 552 Â± 225 µmol/L; p = 0.100); median % of blood Phe < 480 µmol/L decreased (51 (4-96)% vs. 37 (5-85)%; p = 0.041) and median number of clinic appointments increased from SP1 to SP2: (5 (4-6) vs. 11 (8-13); p < 0.001). No significant differences were found regarding any parameter of protein intake. Our results suggest that the implementation of an adult service was successful as impact on metabolic control was limited and attendance remained high. Continuous dietetic care likely contributed to these results by keeping patients in follow-up and committed to treatment

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Open Access Rheumatol. 2020;12:309-21.

YOUNG MINDS: MENTAL HEALTH AND TRANSITIONAL CARE IN ADOLESCENT AND YOUNG ADULT RHEUMATOLOGY.

Palman J, McDonagh JE.

Consideration of the mental health and emotional wellbeing is an important component of health care for all young people, irrespective of setting. Mental health disorders are common during adolescence and young adulthood and young people with rheumatic musculoskeletal diseases (RMD) are not exempt. For such young people, risks of poor outcomes are related to both mental health as well as their RMD. Times of change during adolescence and young adulthood-transitions-are potentially vulnerable life stages for young people with RMD and warrant specific attention in health care provision. Such transitions include those occurring at puberty, during education, training, and employment, socially with moves away from the parental home, as well as from child to adult-centered health services. There is great potential for rheumatology professionals to support young people with RMD at these transitions in view of their frequent encounters and ongoing therapeutic relationships. In this review, we aim to assess the impact of mental health on RMD during adolescence and young adulthood with particular reference to transitional care provision and how rheumatology professionals can be involved in addressing mental health issues during this time of change

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Pan Afr Med J. 2020;37:13.

TRANSITIONING FROM CHILD TO ADULT-ORIENTED HIV CLINICAL CARE FOR ADOLESCENTS LIVING WITH HIV IN ETHIOPIA: RESULTS FROM A RETROSPECTIVE COHORT STUDY.

Broström S, Andersson A, Hallström IK, et al.

INTRODUCTION: Ethiopia has one of the largest number of adolescents living with HIV (ALHIV). As these adolescents reach adulthood they need to transfer from pediatric to adult-oriented clinics. Clear implementation guidelines for transition are lacking and factors associated with successful transition are inadequately investigated. Our objective was to describe the rate and age of transition from child- to adult-oriented care and the factors associated with transition success among ALHIV in selected health facilities in Ethiopia.

METHODS: a retrospective cohort study of adolescents was conducted in eight health facilities in two regions of Ethiopia: Addis Ababa and the Southern Nations, Nationalities and PeoplesÂ´ Region (SNNPR). The study was embedded within a broader study originally aimed at studying clinical outcomes of adolescents. The proportion of adolescents who transitioned was calculated and the association between baseline characteristics and transition was assessed by bivariate and multivariate analysis.

RESULTS: of 1072 adolescents, 8.7% transitioned to adult care. The most frequent age of transition was 15 (range: 10-22). Multivariate analysis generated two significant findings: adolescents from Addis Ababa were more to likely transitioned than adolescents from SNNPR (aOR: 2.18; 95% CI=1.17-4.06; p<0.01), as well as disclosed adolescents compared to those not disclosed of their HIV-status (aOR: 4.19; 95% CI=1.57-11.98; p<0.01).

CONCLUSION: transition occurred in less than 10% of participants, in a wide range of age, indicating a lack of implementation policies regarding the transition process. Thereto, we found that adolescents from Addis Ababa and those disclosed of their disease, were more likely to transition. Further studies are needed to better understand factors associated with transition success

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Patient Educ Couns. 2021 Jan.

SPECIFIC HEALTHCARE RESPONSIBILITIES AND PERCEIVED TRANSITION READINESS AMONG ADOLESCENT SOLID ORGAN TRANSPLANT RECIPIENTS: ADOLESCENT AND CAREGIVER PERSPECTIVES.

Rea KE, Cushman GK, Quast LF, et al.

OBJECTIVE: Adolescents and young adults (AYAs) with solid organ transplants must attain responsibility for healthcare tasks during transition to adult healthcare. However, healthcare systems often initiate transfer based on age and not independence in care. This study examines specific responsibilities distinguishing AYA organ transplant recipients reporting readiness to transfer.

METHODS: 65 AYAs (ages 12-21) with heart, kidney, or liver transplants and 63 caregivers completed questionnaires assessing AYA's transition readiness, healthcare responsibility, and executive functioning. Categorizations included mostly/completely ready versus not at all/somewhat ready to transition; responsibility was compared between groups.

RESULTS: 42% of AYAs and 24% of caregivers reported AYAs as mostly/completely ready to transition. AYAs mostly/completely ready reported similar routine healthcare responsibility (e.g., medication taking, appointment attendance), but greater managerial healthcare responsibility (e.g., knowing insurance details, appointment scheduling), compared to AYAs not at all/somewhat ready to transition.

CONCLUSIONS: All AYAs should be competent in routine healthcare skills foundational for positive health outcomes. However, the managerial tasks distinguish AYAs perceived as ready to transfer to adult healthcare.

PRACTICE IMPLICATIONS: Emphasis on developing responsibility for managerial tasks is warranted. The Hierarchy of Healthcare Transition Readiness Skills is a framework by which AYA responsibility can be gradually increased in preparation for transfer

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Pediatr Cardiol. 2020 Aug;41:1220-30.

HEALTH CARE TRANSITION PERCEPTIONS AMONG PARENTS OF ADOLESCENTS WITH CONGENITAL HEART DEFECTS IN GEORGIA AND NEW YORK.

Gaydos LM, Sommerhalter K, Raskind-Hood C, et al.

With increasing survival trends for children and adolescents with congenital heart defects (CHD), there is a growing need to focus on transition from pediatric to adult specialty cardiac care. To better understand parental perspectives on the transition process, a survey was distributed to 451 parents of adolescents with CHD who had recent contact with the healthcare system in Georgia (GA) and New York (NY). Among respondents, 90.7% reported excellent, very good or good health-related quality of life (HRQoL) for their adolescent. While the majority of parents (77.8%) had been told by a provider about their adolescent's need to transition to adult specialty cardiac care, most reported concerns about transitioning to adult care. Parents were most commonly concerned with replacing the strong relationship with pediatric providers (60.7%), locating an appropriate adult provider (48.7%), and accessing adult health insurance coverage (43.6%). These findings may offer insights into transition planning for adolescents with CHD

Pediatr Clin North Am. 2020 Oct;67:963-71.

OUTCOMES IN ADULT CONGENITAL HEART DISEASE: NEUROCOGNITIVE ISSUES AND TRANSITION OF CARE.

Jacobsen RM.

There is a growing population of patients living with congenital heart disease (CHD), now with more adults living with CHD than children. Adults with CHD have unique health care needs, requiring a thoughtful approach to cardiac, neurocognitive, mental, and physical health issues. They have increased risk of anxiety, depression, pragmatic language impairment, limited social cognition, worse educational attainment and unemployment, and delayed progression into independent adulthood. As a result, it is important to establish an individualized approach to obtain successful transition and transfer of care from the pediatric to adult health care world in this patient population

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Pediatr Infect Dis J. 2021 Apr;40:347-50.

MORTALITY IN PERINATALLY HIV-INFECTED ADOLESCENTS AFTER TRANSITION TO ADULT CARE IN SPAIN.

Berzosa SnA, et al.

INTRODUCTION: After the introduction of combination antiretroviral treatment, (ART) mortality in HIV-infected patients has dramatically decreased. However, it is still high in patients at risk, as adolescents transitioning to adult care (AC) without virologic control. The aim of this study was to characterize mortality and comorbidities of perinatally infected HIV (PHIV) patients after transition to AC.

METHODS: A multicenter retrospective study from patients included in the CoRISpe-FARO Spanish cohort was conducted. PHIV patients who died after transition to AC between 2009 and 2019 were included. Clinical, immunovirologic characteristics, treatments received, comorbidities and causes of death were described.

RESULTS: Among 401 PHIV patients, 14 died (3.5%). All were Spanish, 11/14 (78.6%) women. The median age at diagnosis was 1.5 years (interquartile range [IQR] 0.5-3.9), at transfer to AC was 18 years [16.1-19.9] and at death was 25.8 years [23.6-27.1]. In pediatric units [pediatric care (PC)], CD4+ nadir was 85 cells/Î¼L [IQR 9.7-248.5] and 6/14 patients were classified as C-clinical stage. During AC, all patients were on C-clinical stage and CD4+ nadir dropped to 11.5 cells/Î¼L [4.5-43.3]. cART adherence was extremely poor: in PC, 8/14 patients registered voluntary treatment interruptions; only one had undetectable VL at transition. In AC, 12/14 patients stopped treatment 2 or more periods of time. All deaths were related to advanced HIV disease. Mental health disorders were observed in 7/14 (50%). Main complications described: recurrent bacterial infections (57.1%), wasting syndrome (42.9%), esophageal candidiasis (28.6%) and Pneumocystis jirovecii pneumonia (28.6%). Four women had 11 pregnancies; 5 children were born (none infected).
CONCLUSIONS: Young adults PHIV infected who transition to AC without virologic suppression or proven ability to adhere to ART are at high risk of mortality. Mortality was noted as a consequence of advanced HIV disease, frequent mental health problems and poor adherence to ART

Pediatr Int. 2021 Jan;63:65-71.

TRANSITIONAL CARE FOR INFLAMMATORY BOWEL DISEASE: A SURVEY OF JAPANESE PEDIATRIC GASTROENTEROLOGISTS.

Kumagai H, Kudo T, Uchida K, et al.

BACKGROUND: In 2019 we reported the results of a Japanese national survey designed to explore the views of adult gastroenterologists regarding transitional care for patients with childhood-onset inflammatory bowel disease (IBD). For the present study, we conducted a similar survey of pediatric gastroenterologists to compare the views of the two sets of specialists.
METHODS: The survey conducted in 2019 involved 48 representative members of the Japanese Society for Pediatric Gastroenterology, Hepatology and Nutrition. They were contacted by conventional mail and their answers were not anonymized. Respondents who had already referred patients with IBD to adult gastroenterologists were asked in a questionnaire to rank the importance of specific statements on a Likert scale.
RESULTS: The response rate was 79% and 29 (60%) of the respondents had experienced transitional care for patients with IBD. Transfer to adult care was considered by 90% of the respondents to be the ideal form of medical care for adolescents/young adults with IBD. However, 59% of the respondents had experienced some degree of difficulty when making referrals for such care. The majority of pediatric gastroenterologists considered that the ideal age for transfer was 18-22 years. Among the respondents, physicians at municipal hospitals considered that the presence of diseases other than IBD and a shortage of manpower were significantly more important issues than other practice settings.
CONCLUSIONS: The present survey revealed that the general views regarding transitional care for IBD between pediatric and adult gastroenterologists were similar, except for the appropriate time for transfer. The results underline the importance of preparing a transition program appropriate to practice settings

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Pediatr Neonatol. 2021 Mar;62:146-50.

COMPARISON OF FACTORS ASSOCIATED WITH SUCCESSFUL TRANSITION OF CARE IN PATIENTS WITH HIV VERSUS OTHER CHRONIC DISEASES.

In-Iw S, Lapwech B, Manaboriboon B.

BACKGROUND: Transfer of pediatric patients to adult care is an integral part of optimizing care of chronically ill patients and requires advanced infrastructure and availability of a multidisciplinary team. Thus, assessing factors associated with transition to adult care in this and the other chronic disease group will aid in the targeting intervention programs. The aim of the study was to compare factors associated with transitional readiness and health risk behaviors between adolescents with HIV infection and other chronic diseases.
METHODS: Participants ages 14 to 18 were recruited from chronic care clinics at Siriraj hospital between 2015 and 2016. Self-assessment questionnaires composed of health risk behaviors and a 25-item Likert Scale transition readiness questionnaire with possible scores ranging from 25 to 100 were administered. Analysis was done by SPSS 18.
RESULTS: There were 165 adolescents who participated in the study. Median age was 16 years (range, 14-18). The overall transitional readiness average score was 54.15 ± 8.4 which showed no difference between HIV group (HIVG) and other chronic illness group (non-HIVG). The subjects in the HIVG scored significantly higher in self-management skills (13.03 ± 2.1 vs.12.09 ± 2.8, $p < 0.05$) than their non-HIV counterparts. However, they scored lower in their perception of transition readiness than non-HIVG (13.6 ±2.1 vs. 14.85 ± 2.5, $p < 0.05$). Adolescents who were not HIV-infected were more likely to not want to attend school because of their illness (OR = 4.33, 95% CI = 0.97-19.24.) Conversely, HIV-infected adolescents were more likely to used social media (OR=10.2, 95% CI = 3.26-31.98), consume alcohol beverage (OR = 2.83, 95% CI =1.23-

6.49), smoked cigarettes (OR = 4.17, 95% CI =1.31-13.26), and lack STD knowledge (OR = 3.63, 95% CI = 1.49-8.81) rather than non-HIV infected adolescents.
CONCLUSION: HIV-infected adolescents perceived their self-management skills to be higher than adolescent with other chronic diseases. However, HIV-infected adolescents still possess increased health risk behaviors. To effectively formulate transitional care practice in the Thai context, the program should be focused on health risk behaviors

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Pediatr Pulmonol. 2021 Mar.

A FORMALIZED TRANSITION PROGRAM FOR CYSTIC FIBROSIS: A 10-YEAR RETROSPECTIVE ANALYSIS OF 97 PATIENTS IN LYON.

Bourgeois G, Magne F, Nove JR, et al.

INTRODUCTION: The prognosis of people diagnosed with cystic fibrosis (CF) has dramatically improved over the past decade in France, largely due to advances in CF care management, including an emphasis on chronic maintenance medications. Currently, the majority of French CF patients are adults, which means that they went through a transition process from receiving care at a pediatric CF center to receiving care at an adult CF center. To determine the impact of the transfer on clinical evolution, we report the transition procedure of our CF center in Lyon.
MATERIALS AND METHODS: From January 2006 to December 2016, 97 CF patients underwent a standardized process of transitioning from the pediatric to the adult CF center in Lyon. We compared the clinical evolution of these patients during three periods, starting the year before transition and ending the year after transition. Clinical data taken into account were forced expiratory volume in 1 s (FEV1 in liters), body mass index (BMI in kg/m(2)), pulmonary colonization, number of antibiotic courses, number of days of hospitalization per year, and outpatient visits per year.
RESULTS: No significant differences were observed between respiratory and nutritional status, respiratory microbiome, number of antibiotic courses, or number of hospitalizations or visits when comparing the threeperiods of observation around transition (the year before, the first year after, and the second year after transfer).
CONCLUSION: The standardized transition procedure used in Lyon is associated with the clinical stability of our CF patients

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Pediatr Res. 2021 Mar.

TRANSITION OF CHILDREN WITH LIFE-LIMITING CONDITIONS TO ADULT CARE AND HEALTHCARE USE: A SYSTEMATIC REVIEW.

Jarvis SW, Roberts D, Flemming K, et al.

BACKGROUND: Improved survival has led to increasing numbers of children with life-limiting conditions transitioning to adult healthcare services. There are concerns that transition may lead to a reduction in care quality and increases in emergency care. This review explores evidence for differences in health or social care use post- versus pre-transition to adult services.
METHODS: MEDLINE, EMBASE, CINAHL, PsychINFO and Social Science Citation Index were searched. Studies published in English since 1990 including individuals with any life-limiting condition post- and pre-transition and reporting a health or social care use outcome were included. Data were extracted and quality assessed by one reviewer with 30% checked by an independent reviewer.
RESULTS: Nineteen papers (18 studies) met the inclusion criteria. There was evidence for both increases and decreases (post- versus pre-transition) in outpatient attendance, inpatient admissions, inpatient bed days and health service costs; for increases in Emergency Department visits and for decreases in individuals receiving physiotherapy.
CONCLUSIONS: Evidence for changes in healthcare use post- versus pre-transition is mixed and conflicting, although there is evidence for an increase in Emergency Department visits and a reduction in access to physiotherapy. More high-quality research is needed to better link changes in care to the transition.
IMPACT: Evidence for changes in healthcare use associated with transition to adult services is conflicting. Emergency Department visits increase and access to physiotherapy decreases at transition. There are marked differences between care patterns in the United States and Canada

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Pediatr Rheumatol Online J. 2021 May;19:61.

ASSESSING PREPARATION FOR CARE TRANSITION AMONG ADOLESCENTS WITH RHEUMATOLOGIC DISEASE: A SINGLE-CENTER ASSESSMENT WITH PATIENT SURVEY.

Roberts JE, Halyabar O, Petty CR, et al.

Background: Despite the risk for poor outcomes and gaps in care in the transfer from pediatric to adult care, most pediatric rheumatology centers lack formal transition pathways. As a first step in designing a pathway, we evaluated preparation for transition in a single-center cohort of adolescents and young adults (AYA) with rheumatologic conditions using the ADolescent Assessment of Preparation for Transition (ADAPT) survey.

Findings: AYA most frequently endorsed receiving counseling on taking charge of their health and remembering to take medications. Less than half reported receiving specific counseling about transferring to an adult provider. AYA with lower education attainment compared with those who had attended some college or higher had lower scores in self-management (1.51 vs 2.52, p = 0.0002), prescription medication counseling (1.96 vs 2.41, p = 0.029), and transfer planning (0.27 vs 1.62, p < 0.001). AYA with a diagnosis of MCTD, Sjögren's or SLE had higher self-management scores than those with other diagnoses (2.6 vs 1.9; p = 0.048). Non-white youth indicated receiving more thorough medication counseling than white youth (2.71 vs 2.07, p = 0.027). When adjusting for age, educational attainment remained an independent predictor of transfer planning (p = 0.037). AYA with longer duration of seeing their physician had higher transition preparation scores (p = 0.021).

Conclusion: Few AYA endorsed receiving comprehensive transition counseling, including discussion of transfer planning. Those who were younger and with lower levels of education had lower preparation scores. A long-term relationship with providers was associated with higher scores. Further research, including longitudinal assessment of transition preparation, is needed to evaluate effective processes to assist vulnerable populations

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Pediatrics. 2020 Jun;145.

TRANSITIONING PATIENTS WITH COMPLEX HEALTH CARE NEEDS TO ADULT PRACTICES: THEORY VERSUS REALITY.

Berkowitz S, Lang P.

Pediatrics. 2020 Nov;146.

IMPROVING TRANSITION TO ADULT CARE FOR THOSE WITH DEVELOPMENTAL DISABILITIES: AN UNCLEAR PATH.

Hart LC.

PLoS ONE. 2020;15:e0233451.

PSYCHOSOCIAL NEEDS AMONG OLDER PERINATALLY INFECTED ADOLESCENTS LIVING WITH HIV AND TRANSITIONING TO ADULT CARE IN KENYA.

Gitahi N, Camlin C, Mwania V, et al.

BACKGROUND: Little data is available on the long-term psychosocial effects of disclosure of HIV status that may occur in late adolescence, even when disclosure is timely. Moreover, few studies have described the post-disclosure psychosocial needs of older adolescents who experience delayed disclosure. This study sought to address existing knowledge gaps in the post-disclosure experiences and psychosocial needs of older adolescents living with HIV (ALWHIV).

METHODS: We conducted focus group discussions (FGDs) and in-depth interviews (IDIs) among older perinatally infected adolescents aged 16-19 years We collected socio-demographic data and baseline viral load (copies/ml) results for the preceding six months using interviewer-administered questionnaires and clinical notes abstraction. We analysed data inductively and deductively to identify themes related to the experiences and expectations of adolescents with the disclosure and post-disclosure period.

RESULTS: Adolescents who reported having received timely disclosure expressed that as they grew older, they began to comprehend the lifelong repercussions of an HIV diagnosis and experienced a re-emergence of the negative feelings similar to those experienced during the post-disclosure period. Those who received the knowledge of their HIV status during late adolescence experienced prolonged periods of negative self-perception and anger at not receiving their HIV status earlier. They also expressed a need for more information during the disclosure process on the prevention of onward transmission of the virus, safe conception practices resulting in HIV negative children, and information on how to disclose their HIV status to sexual partners or peers. Anticipated stigma was experienced universally by these older adolescents and was a major barrier towards adherence and coping with an HIV status. Caregivers or siblings with a similar HIV status were a source of social support. Adolescents felt that the support of peers (ALWHIV) helped them to accept their HIV status and to learn how to develop a positive outlook on life.

CONCLUSION: Provision of psychosocial care in late adolescence during the transition to adult care is critical in ensuring the resolution of re-emergent negative emotions. Comprehensive information on HIV prevention and sexual reproductive health should be a crucial component of post-disclosure care for older adolescents. HIV Disclosure and adolescent transition guidelines should include these components to optimize psychosocial care for older adolescents

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PLoS ONE. 2020;15:e0240918.

TRANSITION FROM PEDIATRIC TO ADULT CARE FOR ADOLESCENTS LIVING WITH HIV IN SOUTH AFRICA: A NATURAL EXPERIMENT AND SURVIVAL ANALYSIS.

Zanoni BC, Archary M, Sibaya T, et al.

OBJECTIVE: To determine rates of retention and viral suppression among adolescents living with perinatally-acquired HIV who remained in pediatric care compared to those who transitioned to adult care.

METHODS: We evaluated a natural experiment involving adolescents living with perinatally-acquired HIV who were attending a government-supported antiretroviral clinic in KwaZulu-Natal, South Africa. Prior to 2011, all adolescents transitioned to adult care at 12 years of age. Due to a policy change, all adolescents were retained in pediatric care after 2011. We analyzed adolescents two years before and two years after this policy change. Outcomes were retention in care and HIV viral suppression one year after transition to adult care or the 13th birthday if remaining in pediatric care.

RESULTS: In the natural experiment, 180 adolescents who turned 12 years old between 2011 and 2014 were evaluated; 35 (20%) transitioned to adult care under the old policy and 145 (80%) remained in pediatric care under the new policy. Adolescents who transitioned to the adult clinic had lower rates of retention in care (49%; 17/35) compared to adolescents remaining in the pediatric clinic (92%; 134/145; p<0.001). Retention in care was lower (ARR 0.59; 95%CI 0.43-0.82; p = 0.001) and viral suppression was similar (ARR = 1.06, 95%CI 0.89-1.26; p = 0.53) for adolescents who transitioned to adult care compared to adolescents remaining in pediatric care.

CONCLUSION: Adolescents living with perinatally-acquired HIV appear to have higher retention in care when cared for in pediatric clinics compared to adult clinics. Longer-term follow-up is needed to fully assess viral suppression

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PLoS ONE. 2021;16:e0249971.

TRANSITION TO ADULT CARE: EXPLORING FACTORS ASSOCIATED WITH TRANSITION READINESS AMONG ADOLESCENTS AND YOUNG PEOPLE IN ADOLESCENT ART CLINICS IN UGANDA.

Mbalinda SN, Bakeera-Kitaka S, Lusota DA, et al.

BACKGROUND: Transition readiness refers to a client who knows about his/her illness and oriented towards future goals and hopes, shows skills needed to negotiate healthcare, and can assume responsibility for his/her treatment, and participate in decision-making that ensures uninterrupted care during and after the care transition to adult HIV care. There is a paucity of research on effective transition strategies. This study explored factors associated with adolescent readiness for the transition into adult care in Uganda.

METHODS: A cross-sectional study was conducted among 786 adolescents, and young people living with HIV randomly selected from 9 antiretroviral therapy clinics, utilizing a structured questionnaire. The readiness level was determined using a pre-existing scale from the Ministry of Health, and adolescents were categorized as ready or not ready for the transition. Bivariate and multivariate analyses were conducted.

RESULTS: A total of 786 adolescents were included in this study. The mean age of participants was 17.48 years (SD = 4). The majority of the participants, 484 (61.6%), were females. Most of the participants, 363 (46.2%), had no education. The majority of the participants, 549 (69.8%), were on first-line treatment. Multivariate logistic regression analysis found that readiness to transition into adult care remained significantly associated with having acquired a tertiary education (AOR 4.535, 95% CI 1.243-16.546, P = 0.022), trusting peer educators for HIV treatment (AOR 16.222, 95% CI 1.835-143.412, P = 0.012), having received counselling on transition to adult services (AOR 2.349, 95% CI 1.004-5.495, P = 0.049), having visited an adult clinic to prepare for transition (AOR 6.616, 95% CI 2.435-17.987, P = < 0.001) and being satisfied with the transition process in general (AOR 0.213, 95% CI 0.069-0.658, P = 0.007).

CONCLUSION: The perceived readiness to transition care among young adults was low. A series of individual, social and health system and services factors may determine successful transition readiness among adolescents in Uganda. Transition readiness may be enhanced by strengthening the implementation of age-appropriate and individualized case management transition at all sites while creating supportive family, peer, and healthcare environments

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Respir Care. 2020 Dec;65:1916-22.

TRANSITION FROM PEDIATRIC TO ADULT CARE FOR YOUNG ADULTS WITH CHRONIC RESPIRATORY DISEASE.

Willis LD.

Advances in medicine and technology have led to improved survival rates of children with chronic respiratory disease such as cystic fibrosis, neuromuscular disease, and ventilator dependence. Survival into adulthood has created the need for adult specialists for conditions originating in childhood. Transition from pediatric to adult health care is a process that requires advanced planning and preparation and is not a one-time transfer event. Transition should be standard practice, but many children with special health care do not experience successful transition outcomes. Barriers to successful transition include lack of a standardized process, inadequate planning, and poor communication. Adverse outcomes have occurred in cases of abrupt or haphazard transfers. A successful transition process includes early introduction and ongoing discussion that engages the adolescent to plan and prepare for the eventual transfer of care. Care responsibilities should be gradually shifted from the parent to the adolescent in a manner appropriate for the adolescent's age and developmental status. Good communication and collaboration between pediatric and adult care teams is crucial to ensure a smooth transfer of care. Incorporating the 6 core elements of transition can be helpful in developing a successful transition program. This narrative review summarizes the literature for health care transition from pediatric to adult care including the rationale, barriers, factors associated with successful transition, and special considerations. The intent of this review is to increase clinician awareness of health care transitions and the components necessary for an effective transfer of young adults with chronic respiratory disease. Understanding the transition process is an important consideration for both pediatric and adult clinicians, including respiratory therapists

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Rev Salud Publica (Bogota). 2020 Nov;20:784-86.

TRANSITIONAL CARE FROM ADOLESCENCE TO ADULTHOOD.

Figueroa-Gutiérrez LM, Soto-Chaquir M.

Transitional care aims to facilitate the effective transfer of children suffering from chronic diseases to the medical staff in charge of adult care, ensuring appropriate long-term management, early identification of possible complications, and reduction of morbidity and costs associated with the provision of health services. In several countries, significant progress in this regard has been made, and even consensus on the aspects necessary for the development of transitional care has been reached, including the general principles from the policy to its implementation, with good results in the patients. Despite these advances, in many countries such as Colombia, where the pediatric population suffering from chronic diseases that reach adolescence and then adulthood Is on the rise, little is known about transitional care. It is necessary to generate research and interdisciplinary works to meet the multiple needs of this emerging population, their families and caregivers

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Rheumatology (Oxford). 2020 Dec;59:3737-50.

PATIENTS' ATTITUDES AND EXPERIENCES OF TRANSITION FROM PAEDIATRIC TO ADULT HEALTHCARE IN RHEUMATOLOGY: A QUALITATIVE SYSTEMATIC REVIEW.

Kelly A, Niddrie F, Tunnicliffe DJ, et al.

OBJECTIVES: We aimed to describe patients' attitudes and experiences of transition from paediatric to adult healthcare in rheumatology to inform patient-centred transitional care programmes.

METHODS: We searched MEDLINE, EMBASE, PsycINFO and CINAHL to August 2019 and used thematic synthesis to analyse the findings.

RESULTS: From 26 studies involving 451 people with juvenile-onset rheumatic conditions we identified six themes: a sense of belonging (comfort in familiarity, connectedness in shared experiences, reassurance in being with others of a similar age, desire for normality and acceptance); preparedness for sudden changes (confidence through guided introductions to the adult environment, rapport from continuity of care, security in a reliable point of contact, minimizing lifestyle disruptions); abandonment and fear of the unknown (abrupt and forced independence, ill-equipped to hand over medical information, shocked by meeting adults with visible damage and disability, vulnerability in the loss of privacy); anonymous and dismissed in adult care (deprived of human focus, sterile and uninviting environment, disregard of debilitating pain and fatigue); quest for autonomy (controlled and patronized in the paediatric environment, liberated from the authority of others, freedom to communicate openly); and tensions in parental involvement (overshadowed by parental presence, guilt of excluding parents, reluctant withdrawal of parental support).

CONCLUSION: Young people feel dismissed, abandoned, ill-prepared and out of control during transition. However, successful transition can be supported by preparing for changes, creating a sense of belonging and negotiating parental involvement and autonomy. Incorporating patient-identified priorities into transitional services may improve satisfaction and outcomes in young people with juvenile-onset rheumatic conditions

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S Afr J Psychiatr. 2020;26:1523.

DEMOGRAPHIC AND CLINICAL PROFILE OF PATIENTS UTILISING A TRANSITIONAL CARE INTERVENTION IN THE WESTERN CAPE, SOUTH AFRICA.

Botha HF, Koen L, Niehaus DJH, et al.

BACKGROUND: The World Health Organization's action plan for 2020 has identified the need for service-based data to motivate for more appropriate community-based services. To date, there is no published data from step-up or step-down facilities in South Africa. AIM: To describe the demographic and clinical profile of all patients admitted to New Beginnings between 01 January 2011 and 31 December 2015.
SETTING: New Beginnings is an intermediary care facility focused on psychosocial rehabilitation and accommodates 40 patients in a step-up or step-down setting.
METHODS: In this retrospective audit, we reviewed the medical records of all patients (N = 730) admitted to New Beginnings between 01 January 2011 and 31 December 2015.
RESULTS: Most admissions were male (n = 600; 82.2%), unmarried (92.1%) and unemployed (92.7%) patients with a mean age of 28 years. Only 20.7% had completed their schooling and 37.9% were receiving a disability grant. Most patients lived in the Cape Town Metro area (89%) with their families (94.7%), and 75.6% had no children. Schizophrenia (53.7%) was the most common primary psychiatric diagnosis, and most patients were on a combination of oral and depot treatment (46.8%). Illicit substances were used by 75.9% of patients with 30% using both cannabis and methamphetamine. Most patients (74.9%) had only one admission to New Beginnings.
CONCLUSIONS: These baseline data could inform improved service delivery. Further research is needed to evaluate the success of New Beginnings and highlight the need for more of these facilities in the Western Cape and across South Africa

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Semin Arthritis Rheum. 2021 Apr;51:353-59.

PREDICTORS OF ADVERSE OUTCOMES IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS TRANSITIONING TO ADULT CARE.

Bitencourt N, Makris UE, Solow EB, et al.

BACKGROUND: The transition from pediatric to adult care is a vulnerable period for individuals with chronic diseases. We sought to identify risk factors associated with poor outcomes in patients with childhood-onset systemic lupus erythematosus (cSLE) who have transitioned to adult care.
METHODS: A retrospective analysis of cSLE patients was performed. Outcomes of interest were development of end-stage renal disease (ESRD) or death and time to first hospitalization following final pediatric rheumatology visit. Multivariable logistic and Cox regression models were used.
RESULTS: Of 190 patients with cSLE, 21 (11%) developed ESRD and 9 (5%) died following the final pediatric rheumatology visit. In logistic regression, public insurance, history of Child Protective Services involvement, and an unscheduled hospitalization during the final year in pediatric care were predictive of ESRD or death (odds ratio (95% confidence intervals (CI)) 6.7 (1.5-30.7), 6.6 (2.3-19.1), and 3.2 (1.3-8.3), respectively). Among 114 patients with healthcare utilization data, 53% had a hospitalization in adult care. In Cox regression analysis, a pediatric outpatient opioid prescription was associated with shorter time to adult hospitalization and White or Asian race was associated with longer time to adult hospitalization (hazard ratio (CI) 3.5 (1.7-7.0) and 0.1 (0.03-0.4), respectively).
CONCLUSIONS: Risks factors associated with poor outcomes in adult care amongst patients with cSLE include public insurance, history of Child Protective Services involvement, unscheduled care utilization in pediatric care, pediatric outpatient opioid prescription, Black race and Hispanic ethnicity. Efforts to improve long-term outcomes among patients with cSLE should focus on these populations

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Semin Pediatr Surg. 2020 Aug;29:150948.

TRANSITIONAL CARE OF BILIARY ATRESIA.

Samyn M.

Worldwide native liver survival (NLS) for young adults (>20 years) with biliary atresia varies between 14% and 44% with the majority of patients developing complications in adulthood. Cholangitis and portal hypertension with variceal bleeding are the most common complications and development of these during adolescence associated with the need for liver transplantation during adulthood. Adult listing criteria, typically developed on the background of adult liver disease might not be applicable to this patient population and leads to longer waiting time and risk of deterioration of their medical condition. Current data on growth and puberty in young people with biliary atresia surviving with native liver are rare. Pregnancy has been associated with serious complications in particular for those patients with advanced liver disease and, close follow up by specialist teams recommended. The long-term effect of having a chronic liver disease such as biliary atresia on neuro-cognitive and pubertal development has not been sufficiently explored to date despite reports of a high

prevalence of additional educational needs in this cohort. In addition, patients and parents report inferior health related quality of life compared to healthy peers and similar to that of children post liver transplantation. Moving on from paediatric to adult services is challenging for young people and their parents and adult health professionals might not be familiar with the condition and complications. Young people deserve to be looked after by specialist, multidisciplinary services who provide holistic care and address their psychosocial needs in addition to the medical needs

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Southampton (UK): NIHR Journals Library; 2020 Nov. Health Services and Delivery Research.

UNDERSTANDING THE HEALTH-CARE EXPERIENCES OF PEOPLE WITH SICKLE CELL DISORDER TRANSITIONING FROM PAEDIATRIC TO ADULT SERVICES: THIS SICKLE CELL LIFE, A LONGITUDINAL QUALITATIVE STUDY

Renedo A, Miles S, Chakravorty S, et al.

BACKGROUND: Transitions from paediatric to adult health-care services cause problems worldwide, particularly for young people with long-term conditions. Sickle cell disorder brings particular challenges needing urgent action.

OBJECTIVES: Understand health-care transitions of young people with sickle cell disorder and how these interact with broader transitions to adulthood to improve services and support.

METHODS: We used a longitudinal design in two English cities. Data collection included 80 qualitative interviews with young people (aged 13-21 years) with sickle cell disorder. We conducted 27 one-off interviews and 53 repeat interviews (i.e. interviews conducted two or three times over 18 months) with 48 participants (30 females and 18 males). We additionally interviewed 10 sickle cell disease specialist health-care providers. We used an inductive approach to analysis and co-produced the study with patients and carers.

RESULTS: Key challenges relate to young people's voices being ignored. Participants reported that their knowledge of sickle cell disorder and their own needs are disregarded in hospital settings, in school and by peers. Outside specialist services, health-care staff refuse to recognise patient expertise, reducing patients say in decisions about their own care, particularly during unplanned care in accident and emergency departments and on general hospital wards. Participants told us that in transitioning to adult care they came to realise that sickle cell disorder is poorly understood by non-specialist health-care providers. As a result, participants said that they lack trust in staff's ability to treat them correctly and that they try to avoid hospital. Participants reported that they try to manage painful episodes at home, knowing that this is risky. Participants described engaging in social silencing (i.e. reluctance to talk about and disclose their condition for fear that others will not listen or will not understand) outside hospital; for instance, they would avoid mentioning cell sickle disorder to explain fatigue. Their self-management tactics include internalising their illness experiences, for instance by concealing pain to protect others from worrying. Participants find that working to stay healthy is difficult to reconcile with developing identities to meet adult life goals. Participants have to engage in relentless self-disciplining when trying to achieve educational goals, yet working hard is incompatible with being a good adult patient because it can be risky for health. Participants reported that they struggle to reconcile these conflicting demands.

LIMITATIONS: Our findings are derived from interviews with a group of young people in England and reflect what they told us (influenced by how they perceived us). We do not claim to represent all young people with sickle cell disorder.

CONCLUSIONS: Our findings reveal poor care for young people with sickle cell disorder outside specialist services. To improve this, it is vital to engage with young people as experts in their own condition, recognise the legitimacy of their voices and train non-specialist hospital staff in sickle cell disorder care. Young people must be supported both in and outside health-care settings to develop identities that can help them to achieve life goals.

FUTURE WORK: Future work should include research into the understanding and perceptions of sickle cell disease among non-specialist health-care staff to inform future training. Whole-school interventions should be developed and evaluated to increase sickle cell disorder awareness.

FUNDING: This project was funded by the National Institute for Health Research (NIHR) Health Services and Delivery Research programme and will be published in full in Health Services and Delivery Research; Vol. 8, No. 44. See the NIHR Journals Library website for further project information

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Syst Rev. 2021 Feb;10:46.

PRIMARY CARE DURING THE TRANSITION TO ADULT CARE FOR ADOLESCENTS INVOLVED WITH PEDIATRIC SPECIALTY SERVICES: A SCOPING REVIEW PROTOCOL.

Schraeder K, Allemang B, Scott C, et al.

BACKGROUND: Of the 15-20% of youth in North America affected by a chronic health condition (e.g., type 1 diabetes, cystic fibrosis) and/or mental health or neurodevelopmental disorder (e.g., depression, eating disorder, Attention Deficit-Hyperactivity Disorder), many often require lifelong specialist healthcare services.

Ongoing primary care during childhood and into young adulthood is recommended by best practice guidelines. To date, it is largely unknown if, how, and when primary care physicians (PCPs; such as family physicians) collaborate with specialists as AYAs leave pediatric-oriented services. The proposed scoping review will synthesize the available literature on the roles of PCPs for AYAs with chronic conditions leaving pediatric specialty care and identify potential benefits and challenges of maintaining PCP involvement during transition.
METHODS: Arksey and O'Malley's original scoping review framework will be utilized with guidance from Levac and colleagues and the Joanna Briggs Institute. A search of databases including MEDLINE (OVID), EMBASE, PsycINFO, and CINAHL will be conducted following the development of a strategic search strategy. Eligible studies will (i) be published in English from January 2004 onwards, (ii) focus on AYAs (ages 12-25) with a chronic condition(s) who have received specialist services during childhood, and (iii) include relevant findings about the roles of PCPs during transition to adult services. A data extraction tool will be developed and piloted on a subset of studies. Both quantitative and qualitative data will be synthesized.
DISCUSSION: Key themes about the roles of PCPs for AYAs involved with specialist services will be identified through this review. Findings will inform the development and evaluation of a primary-care based intervention to improve transition care for AYAs with chronic conditions

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Thromb Res. 2021 Apr;200:23-29.
TRANSITION OF CARE FOR PEDIATRIC AND ADULT PATIENTS WITH VENOUS THROMBOEMBOLISM: A NATIONAL QUALITY IMPROVEMENT PROJECT FROM THE AMERICAN THROMBOSIS AND HEMOSTASIS NETWORK (ATHN).
DeSancho M, Munn JE, Billett HH, et al.
BACKGROUND: Transition of care (TOC) for management of anticoagulation from inpatient to outpatient setting for patients with acute venous thromboembolism (VTE) poses serious safety concerns. We implemented a national quality improvement educational initiative to address this issue.
METHODS: Pediatric and adult patients admitted for their first VTE were prospectively enrolled at 16 centers from January 2016 to December 2018. Patient demographics, VTE diagnosis, risk factors, and treatment characteristics were collected. There were two phases: pre-intervention (PI) and quality intervention (QI). The PI phase assessed the quality and patient understanding and satisfaction of anticoagulation instructions given at hospital discharge and adherence to these instructions via a patient and/or caregiver feedback questionnaire (PFQ) and a patient knowledge questionnaire (PKQ) at 30 days. The QI phase provided patient and/or caregiver enhanced education regarding anticoagulation therapy and VTE at hospital discharge using a comprehensive discharge instruction module and a phone call follow-up at one week. Patient and/or caregiver knowledge at 7 and 30Â days was assessed with the same PFQ and PKQ and compared to the PI baseline measures.
RESULTS: Of the 409 study patients, 210 (51%) were adults, 218 (53%) females, and 316 (77%) White. Deep vein thrombosis (62.8%) and pulmonary embolism (47.9%) were the most common VTE in children and adults, respectively. Day 30 PFQ scores were significantly higher in the QI phase compared to the PI phase by 11% ($p < 0.01$). Day 30 PKQ demonstrated enhanced teaching (93.7% vs. 83.5%, p-value 0.004) and disease recognition (89.6% vs. 84.6% $p = 0.03$) in the QI phase than the PI phase.
CONCLUSION: Comprehensive VTE discharge instructions followed by a 1-week post-discharge phone call strengthen patient and caregiver knowledge, satisfaction of education given and care provided, and disease recognition

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Transplant Proc. 2020 Jun;52:1496-99.
TRANSITION TO ADULT CARE FOR PEDIATRIC LIVER TRANSPLANT RECIPIENTS.
Quintero J, et al.
In recent years a growing number of pediatric liver transplant recipients are reaching adulthood and are transferred to an adult team. Because pediatric to adult transition has become a common event with many particularities, specific clinical protocols are needed to guide professionals in this process. Transition must be seen as a complex process of high vulnerability for the patient. The incorrect assumption that the transition process is only a bureaucratic transfer of information leads to inappropriate transition procedures that result in young patients not ready to move to adult units with guaranteed success. To ensure this success, a correct coordination and transmission of the information, accompaniment by the health professional during the whole process, and the empowerment of the patient are required. To have a successful transition, a person within the pediatric team must be in charge of the process (named worker)

Urology. 2021 Feb;148:306-13.
VARIABILITY IN KIDNEY FUNCTION ESTIMATES IN EMERGING ADULTS WITH SPINA BIFIDA: IMPLICATIONS FOR TRANSITIONING FROM PEDIATRIC TO ADULT CARE.
Bowen DK, Balmert LC, Meyer T, et al.
OBJECTIVE: To examine the variability of estimated glomerular filtration rate (eGFR) in emerging adults with spina bifida (SB) by comparing multiple equations across the transitional age period, hypothesizing that creatinine (Cr)-based equations show greater variability than cystatin-C (CysC)- or combination-based equations.
METHODS: A retrospective cohort study was performed from 2012 to 2017 at a multidisciplinary SB clinic. Emerging adults were defined as patients ages 18-28 years old. Four pediatric, 3 adult, and 3 averaged eGFR equations were considered. Cross-sectional variability in eGFR data was assessed using coefficients of variation, chronic kidney disease (CKD) stage classification, and pairwise percent relative difference in eGFR between analogous pediatric and adult equations based on included lab values. Longitudinal changes in eGFR over time were compared across equations using a covariance pattern model accounting for repeated measures.
RESULTS: Seventy-five emerging adults with SB (median age 21.8 years; 55% female; 83% with myelomeningocele) were included in cross-sectional analyses. Adult equations gave higher median eGFRs by 22%-27% and generally milder CKD stage classification than analogous pediatric equations. In longitudinal analyses (median follow-up of 22 months), all equations conferred negative eGFR changes over time (range -1.9 to -4.3 mL/min/1.73m(2) per year) that were not significantly different.
CONCLUSION: In emerging adults with SB, adult equations demonstrated higher median eGFRs by 22%-27% compared to analogous pediatric equations, even with Cystatin-C, and generally downstaged CKD stage classification. The same eGFR equation should be used for serial kidney function monitoring in emerging adults with SB who transition care from pediatric to adult services

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West J Nurs Res. 2020 Jul;42:483-84.
FOCUSING ON ADULT HEALTH CARE TO IMPROVE TRANSITION FOR YOUTH WITH CHRONIC DISEASE.
Varty M, Popejoy LL.

World J Urol. 2021 Apr;39:1003-11.
CHRONIC KIDNEY DISEASE AND UPPER TRACT CONCERNS AFTER CONGENITAL AND ACQUIRED URINARY TRACT ABNORMALITIES: CONSIDERATIONS FOR TRANSITION OF CARE IN TEENS AND YOUNG ADULTS.
Yerkes EB, Baum M, Chu DI.
PURPOSE: To emphasize the burden that chronic kidney disease (CKD) and its complications place on overall health and well-being over the lifetime in individuals with congenital and acquired urinary tract abnormalities.
METHODS: Topic-based literature review was performed and professional opinion was obtained to describe the scope of medical challenges faced by both teens and adults and their health care providers in the context of congenital and acquired urinary tract abnormalities.
RESULTS: Challenges include accurate assessment of glomerular filtration rate; engaging for consistent surveillance of blood pressure, proteinuria, and medical complications of CKD that increase the risk of progression to end-stage renal disease and affect general health; achieving early referral to nephrology for better outcomes; managing renal complications within the unique limitations of lower urinary tract function; treating upper tract urolithiasis in the atypical urinary tract; and preparing for successful renal transplant.
CONCLUSION: In individuals with congenital or acquired abnormalities of the urinary tract, there is an inherent risk of CKD with its associated morbidity and increased mortality risk. Interplay between the upper and lower urinary tract impacts CKD progression. Collaborative management between urology and nephrology is highly recommended to address the unique challenges for each individual over the lifetime

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Z Kinder Jugendpsychiatr Psychother. 2020 Nov;48:429-33.
ANXIETY AND DEPRESSION IN TRANSITION - DESIDERATA FOR IMPROVED CARE AND RESEARCH: RESULTS OF THE JOINT TASK FORCE TRANSITION OF DGPPN AND DGKJP.
Kölch MG, Romanos M, Roth-Sackenheim C, et al.
Anxiety and Depression in Transition - Desiderata for Improved Care and Research: Results of the Joint Task Force Transition of DGPPN and DGKJP Abstract. Affective disorders (e. g., anxiety, depression) frequently begin during adolescence. Yet therapeutic approaches during adolescence differ in some respects from those employed during adulthood. During the transition from adolescence to adulthood, there is a high risk of discontinuation of therapeutic treatment, which may consequently affect integration in employment. There is a

need for age-specific therapeutic strategies that address the relevant issues of adolescents as well as the presently unmet needs in research and treatment for this specific population

EPILESSIA

Epilepsia. 2020 Sep;61:1999-2009.

INTEGRATING QUALITY IMPROVEMENT INTO THE ECHO MODEL TO IMPROVE CARE FOR CHILDREN AND YOUTH WITH EPILEPSY.

Joshi S, Gali K, Radecki L, et al.

Objective: Project ECHO (Extension for Community Healthcare Outcomes), a telementoring program, utilizes lectures, case-based learning, and an "all teach-all learn" approach to increase primary care provider (PCP) knowledge/confidence in managing chronic health conditions. The American Academy of Pediatrics (AAP) Epilepsy and Comorbidities ECHO incorporated quality improvement (QI) methodology to create meaningful practice change, while increasing PCP knowledge/self-efficacy in epilepsy management using the ECHO model.

Methods: Monthly ECHO sessions (May 2018 to December 2018) included lectures, case presentations/discussion, and QI review. Pediatric practices were recruited through the AAP. Practices engaged in ECHO sessions and improvement activities including monthly Plan-Do-Study-Act cycles, team huddles, chart reviews, and QI coaching calls to facilitate practice change. They were provided resource toolkits with documentation templates, safety handouts, and medication side effects sheets. QI measures were selected from the American Academy of Neurology Measurement Set for Epilepsy. The AAP Quality Improvement Data Aggregator was used for data entry, run chart development, and tracking outcomes. Participants completed retrospective surveys to assess changes in knowledge and self-efficacy.

Results: Seven practices participated across five states. Average session attendance was 14 health professionals (range = 13-17). A total of 479 chart reviews demonstrated improvement in six of seven measures: health care transition (45.3%, P = .005), safety education (41.6%, P = .036), mental/behavioral health screening (32.2% P = .027), tertiary center referral (26.7%, not significant [n.s.]), antiseizure therapy side effects (23%, n.s.), and documenting seizure frequency (7.1%, n.s.); counseling for women of childbearing age decreased by 7.8%.

Significance: This project demonstrated that integrating QI into an ECHO model results in practice change and increases PCP knowledge/confidence/self-efficacy in managing epilepsy

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Epilepsy Behav. 2020 Dec;113:107576.

PARTICIPATION IN HEALTHCARE BEHAVIOR BY ADOLESCENTS WITH EPILEPSY AND FACTORS THAT INFLUENCE IT DURING THE TRANSITION PERIOD: A CROSS-SECTIONAL STUDY IN CHINA.

Cui C, Li SZ, Zheng XL, et al.

Purpose: Given the importance of adolescents' participation in the care of their chronic diseases during their transitional period in the healthcare system, the present study investigated the degree of participation in

healthcare behavior by Chinese adolescents with chronic epilepsy and identified factors that should be addressed by health interventions.
Methods: The study used a convenience sample of 1238 adolescent patients with epilepsy, who were hospitalized in 17 tertiary A-level children or maternal and child specialty hospitals in China between January 2017 and March 2020. Several scales were used to measure their degree of participation in healthcare behavior and the factors that influence it. Data collection was conducted after uniform training of the investigators. The adolescents who met the inclusion and exclusion criteria could scan the QR code of the questionnaire via a mobile phone.
Results: The age of the participants ranged from 12.2 to 17.8 years (mean 14.2 years), and the sample had a male-to-female ratio of 1.25:1. The patients' average total score of participation in healthcare behavior was 125.58 (SD = 12.25), which was lower than the norm for China. Their scores on the six dimensions of participation were highest for information interaction, followed in descending order by medical decision-making, treatment and care, appeal, diagnosis and treatment decision-making, and questioning supervision. Multiple linear regression found significant associations between health-care participation and five personal and disease variables (gender, age, course of disease, number of comorbid diseases, type of family structure), self-efficacy, and coping styles (cognitive-palliative and acceptance), which explained 52.1% of the variance in patients' total scores on participating in healthcare behavior.
Conclusions: The participation of young Chinese patients with epilepsy in transitional healthcare behavior needs to be improved. Participation was positively associated with being female, a longer course of disease, fewer comorbidities, and living in a nuclear family. Patients who used cognitive-palliative and acceptance coping styles and those who had higher self-efficacy also had significantly higher levels of participation in healthcare behavior. The study provides useful reference points for adolescents with chronic disease to participate in healthcare programs, in order to achieve a smooth transition from childhood to adulthood

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Epilepsy Behav. 2020 Oct;111:107282.
TELEHEALTH IN PEDIATRIC EPILEPSY CARE: A RAPID TRANSITION DURING THE COVID-19 PANDEMIC.
Sattar S, Kuperman R.
Telehealth's first literature reference is an article in 1879 in the Lancet about using the telephone to reduce unnecessary office visits (Institute of Medicine & Board on Health Care Services, 2012). However, providers have been slow to adopt telehealth into their clinical practice secondary to barriers such as cost and reimbursement (Kane and Gillis, 2018) [2]. The advent of shelter in place orders combined with the ongoing need defined by the Centers for Medicare & Medicaid Services (CMS) Administrator Seema Verma "for all Americans, and particularly vulnerable populations who are at heightened risk, to be able to access their providers" has resulted in the rapid implementation of telehealth across multiple specialties. The goal of this paper is to provide a practical framework for translating quality care in epilepsy as defined by the American Academy of Neurology (AAN) guidelines into a virtual care environment. We will also discuss the use and limitations of point of care testing in epilepsy management

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Epilepsy Behav. 2021 May;120:107981.
PEDIATRIC AND ADULT EPILEPTOLOGIST PERSPECTIVES AND EXPERIENCES OF PEDIATRIC TO ADULT EPILEPSY CARE TRANSITION: "SAYING GOODBYE AND OPENING A DOOR.".
Pieters H, Watson M, Baca CM.
We aimed to describe perspectives of transition and transfer of adolescents and young adults with childhood-onset epilepsy from pediatric to adult care from the viewpoints of both pediatric and adult epileptologists. Telephone semi-structured interviews with pediatric (n=15) and adult (n=11) epileptologists at leading U.S. epilepsy centers were used to collect data about the transition process. Interviews were audio-recorded, transcribed, systematically coded using thematic analysis by two independent researchers, and subsequently checked for agreement during regular meetings. Participants were on average 46â€¯years old (SD=7.4), 50% male, 91% Non-Hispanic and 85% Caucasian; all had completed a formal epilepsy or clinical neurophysiology fellowship (mean 11 years since terminal training) and were employed at a comprehensive epilepsy center. Three interrelated themes regarding epileptologists' perspectives of epilepsy transition and transfer of care were evident: (1) the process is unnatural and disruptive, (2) clinicians make the best of challenges, and (3) the epilepsy transition process includes a spectrum of broad needs some of which are unique to epilepsy care while others are common to other chronic diseases. Despite challenges, epilepsy clinicians spontaneously expressed stress, empathy, and commitment to providing the best possible care

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Epilepsy Behav. 2021 Apr;117:107805.

ESTABLISHING A LEARNING HEALTHCARE SYSTEM TO IMPROVE HEALTH OUTCOMES FOR PEOPLE WITH EPILEPSY.

Donahue MA, Herman ST, Dass D, et al.

OBJECTIVE: To describe the organization of the Epilepsy Learning Healthcare System (ELHS), a network that aims to improve care outcomes for people with epilepsy (PWE).

MATERIALS AND METHODS: Patients and family partners, providers, researchers, epidemiologists, and other leaders collaborated to recruit epilepsy centers and community services organizations into a novel learning network. A multidisciplinary Coordinating Committee developed ELHS governance and organizational structure, including four key planning Cores (Community, Clinical, Quality Improvement, and Data). Through Quality Improvement (QI) methodology grounded in the Institute for Healthcare Improvement (IHI) model, including iterative Plan-Do-Study-Act (PDSA) rapid learning cycles and other learning and sharing sessions, ELHS equipped epilepsy centers and community organizations with tools to standardize, measure, share, and improve key aspects of epilepsy care. The initial learning cycles addressed provider documentation of seizure frequency and type, and also screening for medication adherence barriers. Rapid learning cycles have been carried out on these initial measures in both clinical centers and community-based settings. Additional key measures have been defined for quality of life, screening, and treatment for mental health and behavioral comorbidities, transition from pediatric to adult care, counseling for women and girls living with epilepsy, referral for specialty care, and prevention and treatment of seizure clusters and status epilepticus.

RESULTS: It is feasible to adopt a learning healthcare system framework in epilepsy centers and community services organizations. Through structured collaboration between epilepsy care providers, community support organizations, PWE, and their families/caregivers we have identified new opportunities to improve outcomes that are not available in traditional care models

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J Child Neurol. 2021 Jan;36:60-64.

RESIDENT TRAINING IN TRANSITIONING YOUTH WITH EPILEPSY INTO ADULT CARE.

Tirol FG, Kumar A.

OBJECTIVE: To appraise the current training of Neurology (N), Pediatric (P), and Med-Peds (MP) residents at MedStar Georgetown University Hospital (MGUH) in providing care to patients with epilepsy who are transitioning from pediatric to adult care.

METHODS: Through an online questionnaire, we surveyed Neurology, Pediatric, and Med-Peds residents to assess their knowledge, confidence, and experience at transitioning youth with epilepsy to adult-oriented health care.

RESULTS: N, P, and MP residents generally rated their knowledge and confidence at providing transition care to youth with epilepsy to be poor; however, P and MP residents rated higher in limited measures of knowledge and experience.

CONCLUSION: Our appraisal of resident training in transitions care for youth with epilepsy has highlighted training elements in our institution that require attention for both adult and pediatric providers, leading to the formulation of an educational intervention that will promote experiential and multimodal approaches in this area

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Seizure. 2021 May;88:1-6.

TRANSITION FROM PEDIATRIC TO ADULT CARE IN A JAPANESE COHORT OF CHILDHOOD-ONSET EPILEPSY: PREVALENCE OF EPILEPTIC SYNDROMES AND COMPLEXITY IN THE TRANSITION.

Oguni H, Ito S, Nishikawa A, et al.

AIM: We retrospectively examined patients with childhood-onset epilepsy who transitioned from pediatric to adult care to reveal the clinical characteristics and evaluate the complexity of transitioning.

METHODS: The subjects were 220 patients (89 males, 131 females) who had been treated at our pediatric epilepsy clinic and had transferred to adult care between 2014 and 2018 without attending a transition clinic or program. The demographic data of the patients were retrospectively analyzed.

RESULTS: The ages at transition ranged from 15 to 54 years (median: 27 years old). There were 91 patients with focal epilepsies (FEs) and 129 patients with generalized epilepsies [genetic generalized epilepsy (GGE) n = 30, generalized epilepsy of various etiologies (GEv) n = 99]. A most frequent epileptic syndrome was temporal lobe epilepsy followed by frontal lobe epilepsy in FEs, GTCS only followed by juvenile myoclonic epilepsy in GGE and Lennox-Gastaut syndrome followed by Dravet syndrome in GEv. At the age of transition, a total of 77 of the 96 patients with developmental and epileptic encephalopathies (DEE) had pharmacoresistant seizures, which was positively correlated with a late transition age (P0.05). More than monthly seizures and greater than moderate disabilities were noted in 45% and 55% of the patients, respectively.

CONCLUSION: The patients with childhood-onset epilepsy transitioned to adult care from the hospital-based pediatric epilepsy clinic were characterized by generalized>focal epilepsy, a frequent complication of DEE,

more than monthly seizures, and worse than moderate intellectual disabilities. The complication of DEE made a smooth transition difficult and delayed the transition age

DIABETE

Acta Paediatr. 2020 Oct;109:2057-66.

DEVELOPMENT AND VALIDATION OF A QUESTIONNAIRE TO ASSESS YOUNG PATIENTS' EXPERIENCES WITH DIABETES CARE AND TRANSITION.

Hodnekvam K, Iversen HH, Brunborg C, et al.

AIM: To describe the development and validation of a questionnaire in a national Norwegian population-based cohort study designed to assess the experiences of young people with type 1 diabetes who had made the transition from paediatric to adult diabetes care.

METHODS: The questionnaire was developed by the authors based on literature searches, focus group interviews, discussions with experts and cognitive interviews. We included 776 individuals with type 1 diabetes who were last registered in the Norwegian Childhood Diabetes Registry between 2009 and 2012 and had been receiving adult health care for at least 2 years. The data quality was analysed, factor analysis was performed, and the internal reliability, test-retest reliability and construct validity were determined.

RESULTS: The response rate was 321 patients (41.4%); 57.6% were female, and the average age at recruitment was 22.9± 1.2 years. Seven factors were identified. Satisfactory evidence was provided for the internal consistency, reliability and construct validity of the questionnaire. All scales met the criterion of Cronbach's alpha above 0.4. The test-retest correlations ranged from 0.64 to 0.92.

CONCLUSION: The thorough validation of the questionnaire proved satisfactory and indicated that it may be of value for further studies measuring patients' experiences with diabetes care and transition

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Ann Pediatr Endocrinol Metab. 2021 Feb.

A NATIONAL SURVEY OF TRANSITION FROM PEDIATRIC TO ADULT HEALTHCARE PROVIDERS FOR ADOLESCENTS AND YOUNG ADULTS WITH TYPE 1 DIABETES: PERSPECTIVES OF PEDIATRIC ENDOCRINOLOGISTS IN KOREA.

Kim JH, Yoo JH.

PURPOSE: Transition from pediatric to adult healthcare for adolescents and young adults with type 1 diabetes (T1D) increases the risk for poor outcomes. This study aimed to describe the circumstances and clinical practice patterns associated with this transition care from a nationwide survey of pediatric endocrinologists in Korea.

METHODS: An electronic survey regarding healthcare transition of T1D patients was administered to 143 pediatric endocrinologists registered in the Korean Society of Pediatric Endocrinology.

RESULTS: Response rate was 50.2% (n = 72). Among responders, 58.3% (n = 42) were females and 70.8% (n = 51) worked in academic medical centers. Main reasons for transfer to adult care were request from a patient or family (69.6%) and age 18 years (42.0%). Impediments to transition were long-lasting therapeutic relationship (72.9%) and lack of adult specialists in T1D care (62.9%). Communication between pediatric and adult endocrinologists was via non-structured patient summary (68.6%) and telephone or e-mail (27.1%).

Responders reported that successful transition requires development of transition protocols (79.2%) and multidisciplinary team approach for transition care (52.8%).
CONCLUSION: Transition care of T1D patients was a challenge to pediatric endocrinologists in Korea. Development of protocols for transition care for healthcare providers and improvement of diabetes self-management skills for patients are needed

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Aust Health Rev. 2020 Aug;44:601-08.

SERVICE USE OF YOUNG PEOPLE WITH TYPE 1 DIABETES AFTER TRANSITION FROM PAEDIATRIC TO ADULT-BASED DIABETES HEALTH CARE.

Perry L, Dunbabin J, Xu X, et al.

Objective The aim of this study was to determine, in the first 2 years after the last planned appointment with paediatric diabetes services for young people with Type 1 diabetes (T1D): (1) the number of planned and unplanned healthcare contacts and HbA1c measurements made; (2) factors linked to diabetes-related service use; and (3) factors predictive of the number of planned and unplanned service contacts, and of meeting the minimum number of planned service contacts.
Methods Healthcare records of a major public healthcare provider in Australia were audited for preventive and acute service use by young people with T1D transferring from paediatric to adult public healthcare services. Statistical analyses included use of t-tests and logistic regression modelling.
Results Of 172 young people with T1D, 21% had no planned specialist care and 49% accessed acute services for diabetes-related matters. Residents of metropolitan areas and users of continuous subcutaneous insulin infusion therapy were more likely to access specialist care and were less likely to use acute services for unplanned care. Those achieving a minimum of nine planned care contacts in 2 years had a shorter duration between the last paediatric and first adult healthcare contact.
Conclusions Lack of specialist care in early adult years and non-metropolitan relative disadvantage compromise the present and future health of young people with diabetes. What is known about the topic? Well-managed transition is thought to offer the best chance of achieving cost-effective continuing engagement with specialist services for planned preventive care, effective T1D self-management and deferral or early attention to diabetes-related vascular complications. However, transition is commonly reported as problematic. What does this paper add? The findings of this study indicate a positive trend but continuing need to improve transition care for young people with T1D, especially those living in non-metropolitan areas and those not using continuous subcutaneous insulin infusion therapy. What are the implications for practitioners? Without service innovation, suboptimal and delayed access to planned care, high use of acute services for unplanned care and poor glycaemic control will continue to threaten the future health and well-being of young people with T1D

Can J Diabetes. 2021 Mar.

TRANSITION OF YOUTH WITH TYPE 2 DIABETES: PREDICTORS OF HEALTH-CARE UTILIZATION AFTER TRANSITION TO ADULT CARE FROM POPULATION-BASED ADMINISTRATIVE DATA.

Pundyk KJ, Sellers EAC, Kroeker K, et al.

OBJECTIVES: In this study, we aimed to compare health-care visits pre- and posttransition from pediatric to adult care between youth with type 2 and type 1 diabetes.
METHODS: We linked a clinical database with the Manitoba Population Research Data Repository to compare health-care visits 2 years before and after transition, and investigated baseline factors influencing health-care engagement.
RESULTS: Youth with type 2 diabetes (n=196) vs type 1 diabetes (n=456) were more likely to be female (61% vs 44%), older at diagnosis (13.6 vs 10.6 years), live in northern regions and to be in the lowest socioeconomic status quartile (53% vs 5.4%). Seventy-six percent of youth with type 2 diabetes attended a follow-up visit within 2 years of transition compared to 97% of youth with type 1 diabetes. Youth with type 2 diabetes had higher rates of hospitalization pretransition (19.6 vs 11.6 admissions/100 patient years) and posttransition (24.7 vs 11.7 admissions/100 patient years) and fewer medical visits (pretransition: 2.4 vs 3.0 visits/person year [p<0.01]; posttransition: 1.6 vs 2.1 visits/person year [p<0.01]). Accounting for sex, geography, age, education, socioeconomic status and diabetes type, achieving 4 visits in 2 years posttransition was predicted by the number of visits pretransition (odds ratio, 1.35; 95% confidence interval, 1.23 to 1.49) and diabetes type (type 2 diabetes: odds ratio, 0.57; 95% confidence interval, 0.34 to 0.98).
CONCLUSIONS: Youth with type 2 diabetes attend fewer medical follow-up visits pre- and posttransition to adult care compared to youth with type 1 diabetes. Focussed, informed, specific transition planning is needed that addresses the unique characteristics of this population

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Child Care Health Dev. 2020 Nov;46:692-702.

THE TRANSITION EXPERIENCES OF ADOLESCENTS WITH TYPE 1 DIABETES FROM PAEDIATRIC TO ADULT CARE PROVIDERS.

McDowell ME, Litchman ML, Guo JW.

PURPOSE: Emerging adults with type 1 diabetes (T1D) are at increased risk for poor health outcomes as they transition from paediatric to adult healthcare providers. This is in part due to the complexities of young adult life as individuals with T1D enter the workforce, leave home or start college while learning to manage the disease on their own. We sought to identify the barriers and facilitators adolescents face during their emerging adult years with T1D.

METHODS: Young adults, aged 24-35, who lived with T1D during their adolescent years were recruited online to complete a survey regarding their experience with care transition. Categorical data were analysed using descriptive statistics. A thematic analysis, guided by the Framework for Emerging Adults with T1D, was used to explore the free-text data.

RESULTS: In total, 25 adults (84% female) with mean age of 28 Â± 3.2 years participated. Themes that arose from the analysis of the paediatric to adult care transition experiences included (1) importance of support from key players, (2) challenges navigating the healthcare system, (3) mental health needs of emerging adults with T1D, (4) managing day-to-day life with T1D and (5) early independence to ease transition.

CONCLUSION: Individuals with T1D face a variety of challenges as they transition from paediatric to adult care providers. A proactive approach in educating adolescents is needed

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Clin Diabetes Endocrinol. 2020;6:11.

IMPROVING THE TRANSITIONING OF PEDIATRIC PATIENTS WITH TYPE 1 DIABETES INTO ADULT CARE BY INITIATING A DEDICATED SINGLE SESSION TRANSFER CLINIC.

Williams S, Newhook LAA, Power H, et al.

BACKGROUND: Young adults with type 1 diabetes face potential health problems and disruptions in accessing care related to their move from pediatrics into adult care. At a medium-sized pediatric hospital with no formal transition support program, we developed and evaluated the use of a single-session transfer clinic as an initial quality improvement intervention to improve patient satisfaction, clinic attendance, and knowledge of transition related issues.

METHODS: Following a jurisdictional scan of other diabetes programs, the pediatric diabetes program developed a half-day transfer clinic. After the first transfer clinic was held, evaluation surveys were completed by patients, parents, and healthcare providers. Based on the feedback received, we altered the structure and evaluated the revised clinic by surveying patients and parents.

RESULTS: All patients and parents who attended reported high levels of satisfaction with the clinic. Providers were also mostly positive regarding their participation. Feedback from the first clinic was used to modify the structure of the second clinic to better meet the needs of participants and to allow the clinic to run more efficiently. The use of group sessions and adapting resources developed by other diabetes programs were viewed favourably by participants and lessened the burden on staff who delivered the clinic.

CONCLUSIONS: A half-day transfer clinic is a viable step towards improving patient and parent satisfaction during the transition into adult care without requiring additional staff or significant expenditures of new resources. This type of clinic can also be incorporated into a larger program of transition supports or be adopted by programs serving young adults with other chronic diseases

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Diabet Med. 2020 Aug;37:1407-09.

VARIATIONS IN DIABETES TRANSITION CARE FOR CHILDREN AND YOUNG PEOPLE: A NATIONAL SURVEY.

Ng SM, Lay JT, Regan F, et al.

Diabet Med. 2021 Jun;38:e14541.

PSYCHIATRIC DISORDERS IN EMERGING ADULTS WITH DIABETES TRANSITIONING TO ADULT CARE: A RETROSPECTIVE COHORT STUDY.

Robinson ME, Simard M, Larocque I, et al.

AIMS: During transition from paediatric to adult diabetes care, adolescents with diabetes are at increased risk of psychiatric disorders compared with those without diabetes. Prolonged gaps between the last paediatric and first adult diabetes care visit are associated with higher perceived stress and lower life satisfaction. We assessed the effect of a gap (>180 days) in establishing adult diabetes care on the risk of psychiatric disorders and determined other risk factors associated with psychiatric disorders during the transfer to adult care.

METHODS: Using provincial health administrative databases, we conducted a retrospective cohort study of individuals from Quebec, Canada, diagnosed with diabetes between ages 1 and 15 years in 1997-2015. These

individuals were followed from 6Â months after their last paediatric visit until age 25 years. We used multivariable Cox proportional hazard models to determine the association of gap in care with psychiatric disorders risk.
RESULTS: Among 1772 youth with diabetes, 740 (42%) had a gap in care. There was a non-statistically significant association between gap in care and mood disorders diagnosed in the emergency department or hospital (hazard ratio [HR] 1.38, 95% confidence interval [CI]: [0.92, 2.07]). Older age at transfer, recent birth year and higher number of all-cause emergency department visits in the year before transfer increased the risks of psychiatric disorders.
CONCLUSIONS: Prolonged gaps in care during transfer to adult care are common and may be associated with increased psychiatric disorder risk. Developmental factors associated with adolescence and emerging adulthood may further amplify this risk

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Diabetes Care. 2021 Apr.
THE RAPID TRANSITION TO TELEMEDICINE AND ITS EFFECT ON ACCESS TO CARE FOR PATIENTS WITH TYPE 1 DIABETES DURING THE COVID-19 PANDEMIC.
Tilden DR, Datye KA, Moore DJ, et al.
OBJECTIVE: We compared the uptake of telemedicine for diabetes care across multiple demographic groups during the coronavirus disease 2019 pandemic to understand the impact of telemedicine adoption on access to care.
RESEARCH DESIGN AND METHODS: The study analyzed demographic information of patients with type 1 diabetes seen between 1 January 2018 and 30 June 2020 at a single center. We compared the odds of completing a visit via telemedicine across multiple demographic characteristics.
RESULTS: Among 28,977 patient visits, the odds of completing a visit via telemedicine were lower among non-English-speaking (1.7% vs. 2.7%; adjusted odds ratio [OR] 0.45, 95% CI 0.26-0.79) and Medicaid-insured (32.0% vs. 35.9%; OR 0.83, 95% CI 0.72-0.95) pediatric patients. No clinically significant differences were observed for other demographic factors.
CONCLUSIONS: Rapid transition to telemedicine did not significantly impact access to diabetes care for most demographic groups. However, disparities in access to care for historically marginalized groups merit close attention to ensure that use of telemedicine does not exacerbate these inequities

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Diabetes Educ. 2020 Jun;46:252-60.
HEALTH CARE TRANSITION IN TYPE 1 DIABETES: PERSPECTIVES OF DIABETES CARE AND EDUCATION SPECIALISTS CARING FOR YOUNG ADULTS.
Goethals ER, La Banca RO, Forbes PW, et al.
PURPOSE: The purpose of the study was to describe experiences reported by diabetes care and education specialists caring for young adults with type 1 diabetes and to assess perceived deficiencies in clinical resources and barriers to care delivery.
METHODS: A 60-item electronic survey was fielded through email to members of the Association of Diabetes Care and Education Specialists (ADCES). Respondents completed a survey consisting of: (1) clinical practice characteristics and respondents' demographics; (2) health care transition components (eg, referrals) and their perceived importance; (3) framework of current clinical diabetes care delivery and perceived need for additional support; and (4) perceived barriers regarding clinical care delivery. Statistical analyses included descriptive statistics, chi-square tests, and logistic regression.
RESULTS: Respondents (N = 531, 96% female, median years in practice = 13; interquartile range = 7-20) represented 49 states plus the District of Columbia. Although 88% of respondents reported reviewing pediatric records as important/very important, only 22% often/always reviewed them. Although 58% of respondents noted easy access to mental health care providers for young adults, 50% stated a need for additional resources. Furthermore, diabetes care and education specialists without easy access to mental health professionals were significantly more likely to report barriers to diabetes management for young adults with depression, substance abuse, eating disorders, and developmental disabilities.
CONCLUSION: Study findings highlight modifiable factors that may improve diabetes care coordination for transitioning young adults. Uniform approaches and increased access to trained mental health professionals may help support diabetes care and education specialists in their care of young adults with type 1 diabetes

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Diabetes Spectr. 2020 Nov;33:331-38.

HEALTH CARE TRANSITION IN YOUTH WITH TYPE 1 DIABETES AND AN A1C >9%: QUALITATIVE ANALYSIS OF PRE-TRANSITION PERSPECTIVES.

Tremblay ES, Ruiz J, Buccigrosso T, et al.

OBJECTIVE | To explore expectations for transition to adult care and experiences with transition planning among adolescents and young adults with type 1 diabetes and an A1C >9% at a tertiary care U.S. pediatric center.

METHODS | We conducted semi-structured interviews in a purposive sample of patients 14-23 years of age who had had type 1 diabetes for at least 1 year and had an A1C >9%. A multidisciplinary team conducted iterative thematic analysis with deductive and inductive coding aided by NVivo software.

RESULTS | Fourteen subjects participated (nine adolescents and five young adults, mean age 17.1 ± 3.2 years, 57% male, 79% Caucasian, 14% Hispanic, diabetes duration 8.2 ± 4.6 years, mean A1C 10.0 ± 0.8% for adolescents and 10.1 ± 0.7% for young adults). Qualitative analysis yielded four key themes. The first was lack of formal preparation; participants of all ages demonstrated a lack of preparation for transition and ignorance about the process, describing it as coming "out of the blue." The second was a desire for delayed and gradual transition; participants wanted to defer being "serious" about transition to a later/uncertain date, with a preference to "wait until I'm older" among all ages. Participants described ideal transition as a gradual process, taking place "a little at a time." The third was attachment to pediatric providers; participants demonstrated a nearly universal attachment to and "familiarity" with their pediatric diabetes care providers and expressed worries about an "uncomfortable" transition to adult providers. The fourth was concern about an impersonal adult care setting: participants perceived adult care as "formal," "scarier," and "tougher," with increased criticism about poor control; participants expressed fear that adult providers would not "know me" or appreciate "my diabetes journey."

CONCLUSION | We demonstrated a lack of transition preparation and anxiety about transition and adult care among youth with type 1 diabetes and elevated A1C. Our results may help guide early, iterative pediatric transition counseling, with a special focus on addressing attachment and fears about adult diabetes care

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Diabetes Spectr. 2020 Aug;33:255-63.

YOUNG ADULTS WITH TYPE 1 DIABETES PREPARING TO TRANSITION TO ADULT CARE: PSYCHOSOCIAL FUNCTIONING AND ASSOCIATIONS WITH SELF-MANAGEMENT AND HEALTH OUTCOMES.

Gutierrez-Colina AM, Corathers S, Beal S, et al.

BACKGROUND | Young adulthood is a vulnerable developmental period associated with increased risk for suboptimal health outcomes in youth with type 1 diabetes. Psychosocial factors have been associated with self-management and glycemic control in younger populations, but the extent to which these associations exist among young adults is poorly understood. This study aimed to examine the psychosocial functioning of young adults with type 1 diabetes and associated clinical outcomes.

METHODS | Participants included young adults (n = 44) between the ages of 18 and 23 years in a pediatric setting who were preparing to transition to adult care. All participants completed self-report measures of psychosocial functioning at baseline as part of this longitudinal observational study. Outcome data included glycemic control, frequency of blood glucose monitoring, and self-management ratings at baseline and 1-year follow-up.

RESULTS | Young adults with type 1 diabetes reported higher levels of depressive symptoms, lower self-efficacy, and more risk behaviors compared with previously published scores for adolescents. Young adults also reported greater resilience and transition readiness than their younger counterparts. Psychosocial variables were differentially related to glycemic control and frequency of blood glucose monitoring both cross-sectionally and longitudinally.

CONCLUSION | This study provides key information about the psychosocial functioning of young adults with type 1 diabetes. It identifies relevant psychosocial factors that are associated with meaningful health outcomes during the transition preparation period. These findings may inform the development of clinical programs aimed at promoting transition preparation and health outcomes in young adults with type 1 diabetes

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Diabetologia. 2021 Apr;64:758-66.

IMPROVED TRANSITION TO ADULT CARE IN YOUTH WITH TYPE 1 DIABETES: A PRAGMATIC CLINICAL TRIAL.

Butalia S, Crawford SG, McGuire KA, et al.

AIMS/HYPOTHESIS: Youth with type 1 diabetes are at high risk for loss to follow-up during the transition from paediatric to adult diabetes care. Our aim was to assess the effect of a communication technology enhanced transition coordinator intervention compared with usual care on clinic attendance among transitioning youth with type 1 diabetes.

METHODS: In this open label, pragmatic clinical trial of youth with type 1 diabetes, aged 17-18 years, transitioning from paediatric to adult diabetes care, the intervention group received support from a transition coordinator who used communication technology and the control group received usual care. The primary outcome was the proportion of individuals that did not attend at least one routine clinic visit in adult diabetes care within 1 year after transfer. Secondary outcomes included diabetes-related clinical outcomes and quality of life measures.
RESULTS: There were no baseline differences in age, sex, HbA(1c) and number of follow-up visits, emergency department visits and diabetic ketoacidosis admissions in the 1 year prior to transition between the usual care (n =101) and intervention (n =102) groups. In the year following transfer, 47.1% in the usual care group vs 11.9% in the intervention group did not attend any outpatient diabetes appointments (p <0.01). There were no differences in glycaemic control or diabetic ketoacidosis post transfer.
CONCLUSIONS/INTERPRETATION: Our intervention was successful in improving clinic attendance among transitioning youth with type 1 diabetes. Importantly, this programme used simple, readily accessible communication technologies, which increases the sustainability and transferability of this strategy.
TRIAL REGISTRATION: isrctn.org ISRCTN13459962

Endocrinol Diabetes Nutr. 2021 Feb;68:82-91.

RESULTS OF A SPECIFIC AND STRUCTURED PROGRAM IN THE TRANSITION OF YOUNG PATIENTS WITH TYPE 1 DIABETES FROM THE PAEDIATRIC CENTER TO AN ADULT HOSPITAL. THE EXPERIENCE OF A DECADE.

Vidal FM, JansÃ IM, Roca ED, et al.

Objective: Evaluate the results of a healthcare and therapeutic education programme (TEP) aimed at young patients with type 1 diabetes (T1D) transferred from a paediatric centre.
Methodology: This was a prospective, pre-postest in young T1D patients transferred from 2005-2015. The programme has four phases: coordinated transfer, evaluation and objective pacting, knowledge (DKQ2) adherence (SCI-R.es) and quality of life (DQoL and SF12). Results were compared according to Multiple Daily Injections (MDI) vs. Continuous Subcutaneous Insulin Infusión (CSII) and adherence (SCI-R.es < 65 vs. > 65%).
Results: A total of 330 patients were transferred (age 18.19 ± 0.82 years, 49% females, glycated haemoglobin [HbA1c] 8.6 ± 1.4%). The programme was completed by 68%, and 61% did a group course. While no changes in HbA1c were observed at one year (8.3 ± 1.4 vs. 8.2 ± 1.4%), there were changes in severe hypoglycaemias/patient/year (0.23 ± 0.64 to 0.05 ± 0.34 p < 0.001) and mild > 5 hypoglycaemias/patient/week (6.9% vs. 3.9% p = 0.09). DQK2 knowledge increased (25.7 ± 3.6 vs. 27.8 ± 3.8 p < 0.001), with no changes in quality of life or grade of adherence. Patients with CSII (n = 21) performed more blood glucose controls and showed greater programme adherence with no changes in metabolic control. Patients with the best initial adherence presented the best control (p < 0.0001). A lower initial HbA1c and receiving the group course were associated with better clinical HbA1c results ≥ 0.5% (p < 0.05)
CONCLUSIONS: The TEP improved some parameters of metabolic control without modifying the quality of life in young T1D patients. When comparing patients on MDI vs. CSII, there were no differences in metabolic control but there were when differences were evaluated considering treatment adherence

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Front Med (Lausanne). 2021;8:652358.

CHALLENGES IN TRANSITION FROM CHILDHOOD TO ADULTHOOD CARE IN RARE METABOLIC DISEASES: RESULTS FROM THE FIRST MULTI-CENTER EUROPEAN SURVEY.

Stepien KM, KieÄ‡-Wilk B, Lampe C, et al.

Inherited Metabolic Diseases (IMDs) are rare diseases caused by genetic defects in biochemical pathways. Earlier diagnosis and advances in treatment have improved the life expectancy of IMD patients over the last decades, with the majority of patients now surviving beyond the age of 20. This has created a new challenge: as they grow up, the care of IMD patients' needs to be transferred from metabolic pediatricians to metabolic physicians specialized in treating adults, through a process called "transition." The purpose of this study was to assess how this transition is managed in Europe: a survey was sent to all 77 centers of the European Reference Network for Hereditary Metabolic Disorders (MetabERN) to collect information and to identify unmet needs regarding the transition process. Data was collected from 63/77 (81%) healthcare providers (HCPs) from 20 EU countries. Responders were mostly metabolic pediatricians; of these, only ~40% have received appropriate training in health issues of adolescent metabolic patients. In most centers (~67%) there is no designated transition coordinator. About 50% of centers provide a written individualized transition protocol, which is standardized in just ~20% of cases. In 77% of centers, pediatricians share a medical summary, transition letter and emergency plan with the adult team and the patient. According to our responders, 11% of patients remain under pediatric care throughout their life. The main challenges identified by HCPs in managing transition are lack of time and shortage of adult metabolic physician positions, while the implementations that

are most required for a successful transition include: medical staff dedicated to transition, a transition coordinator, and specific metabolic training for adult physicians. Our study shows that the transition process of IMD patients in Europe is far from standardized and in most cases is inadequate or non-existent. A transition coordinator to facilitate collaboration between the pediatric and adult healthcare teams should be central to any transition program. Standardized operating procedures, together with adequate financial resources and specific training for adult physicians focused on IMDs are the key aspects that must be improved in the rare metabolic field to establish successful transition processes in Europe

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Health Sci Rep. 2020 Sep;3:e181.

YOUTH WITH DIABETES AND THEIR PARENTS' PERSPECTIVES ON TRANSITION CARE FROM PEDIATRIC TO ADULT DIABETES CARE SERVICES: A QUALITATIVE STUDY.

Butalia S, McGuire KA, Dyjur D, et al.

BACKGROUND AND AIMS: When youth with diabetes transition from pediatric to adult diabetes care, they are at high risk for loss to follow up and worsening glucose control. We aimed to gain insight on how to improve the transition of youth with type 1 diabetes from pediatric to adult diabetes care from the patients' and parents' perspective.

METHODS: We conducted focus groups in youth with type 1 diabetes in transition from pediatric to adult diabetes care and their parents, in Calgary, Alberta, between June and August 2014. Eligibility criteria included: (a) type 1 diabetes; (b) aged 15 to 25 years; (c) have or had received care at the pediatric hospital; and, (d) either pre or post-transfer; or, (e) parents of recently transferred youth. Purposive sampling was used, and the theoretical framework used was the Integrated Behaviour Model. Participants were asked about positive, negative, or challenging experiences related to diabetes and transition, solutions to challenges, and tools and strategies to improve and better support transition. Thematic analysis was conducted after focus groups were recorded and transcribed.

RESULTS: Three focus groups were conducted: pre-transfer youth with diabetes (4 females and 3 males; median age 17.5 years, IQR 1.3 years); post-transfer young adults with diabetes (2 females and 2 males; median age 23.5 years, IQR 1.2 years); and parents of recently transferred young adults with diabetes (n = 3). Main themes were: (a) communication technology; (b) the need for more transition and diabetes education and preparation during transition; and, (c) the importance and need for social and peer support.

CONCLUSION: This study describes specific areas that may improve diabetes transfer and transition from pediatric to adult diabetes care. This information can help inform clinical care delivery for transition and the development of programs, strategies, and interventions to improve transition care

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Indian J Pediatr. 2020 Jun;87:421-26.

NEED AND FEASIBILITY OF A TRANSITION CLINIC FOR ADOLESCENTS WITH CHRONIC ILLNESS: A QUALITATIVE STUDY.

Menon J, Peter AM, Nayar L, et al.

OBJECTIVE: To assess the need and feasibility of a Transition Clinic in the care of adolescents with chronic illness.

METHODS: A qualitative study, piloted by a questionnaire-based survey, followed by Focus Group Discussions was conducted in the Department of Pediatrics in a government teaching hospital in Kerala. The participants were adolescents with chronic disease, viz., renal disease, diabetes, HIV/AIDS, cancer survivors with Hepatitis B, parents of renal patients, pediatricians and adult specialists. Twenty patients were administered a structured, rated, 18-item questionnaire regarding their attitude and preparedness for transfer to adult departments. The need and feasibility of a Transition Clinic was discussed by 7 focus groups viz., pediatricians, adult physicians, 4 patient groups and 1 parent group. Thematic analysis of data was done.

RESULTS: Except among pediatricians, awareness of the concept of Transition Clinic was low. All participants agreed that abrupt transfer of care hinders treatment and that there is a definite need for a Transition Clinic. Pediatricians discussed the complex needs of adolescents and limitations of their care. Adult specialist physicians described their inadequacy to handle adolescent problems and difficulties posed by large volumes of patients. The adolescents and young adults with chronic illness discussed their reluctance to shift to the adult departments. Parents voiced their concerns about the future of their children. All groups gave constructive suggestions for conduct of the clinic and smooth transition.

CONCLUSIONS: There is a definite need for a Transition Clinic in the management of adolescents with chronic illness to ensure smooth transfer of care. Introducing such clinics in the existing health framework is feasible using a multidisciplinary approach

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J Pediatr Psychol. 2020 Aug;45:767-79.

VALIDATION OF THE HEALTHCARE TRANSITION OUTCOMES INVENTORY FOR YOUNG ADULTS WITH TYPE 1 DIABETES.

Pierce J, Hossain J, Gannon A.

OBJECTIVE: We recently developed and content validated the Healthcare Transition Outcomes Inventory (HCTOI), a stakeholder vetted, multidimensional measure of the outcomes of the transition from pediatric to adult healthcare for young adults (YA) with type 1 diabetes (T1D). In this study, we aimed to evaluate the psychometric properties of the HCTOI.

METHODS: We collected and analyzed cross-sectional data from 128 YA (18-25 years old) with T1D to evaluate the psychometric properties of the HCTOI. We conducted confirmatory factor analysis (CFA), item analysis, and examined reliability and validity in relation to measures of quality of life, diabetes distress, regimen adherence, and glycemic control.

RESULTS: CFA supported a five-factor solution: integration of T1D into emerging adult roles, balance of parental support with T1D autonomy, establishing and maintaining continuity of care, forming a collaborative patient-provider relationship, and ownership of T1D. We reduced the HCTOI from 54 to 34 items. The HCTOI demonstrated adequate internal consistency (Î±'s = 0.62-0.87) and significant correlations demonstrated construct (quality of life, diabetes distress) and criterion validity (adherence, glycemic control).

CONCLUSIONS: The HCTOI demonstrated promising initial psychometric properties. As the first measure of the multiple dimensions of healthcare transition outcomes, the HCTOI provides a means to examine longitudinal relations between transition readiness and outcomes and to assess the efficacy or effectiveness of interventions and programs designed to improve the transition process for YA with T1D

OTJR (Thorofare N J). 2021 Jan;41:6-14.

ENGAGEMENT IN HOUSEHOLD CHORES IN YOUTH WITH CHRONIC CONDITIONS: HEALTH CARE TRANSITION IMPLICATIONS.

Richards J, Nazareth M, van Tilburg MAL, et al.

This study examined associations between chores engagement, self-management, and transition readiness in youth with chronic conditions. Youths with various chronic conditions attending a therapeutic camp, and their parents participated. Responses of 165 campers and their parents were analyzed (mean camper age 12.3 ± 2.6 years, 47.9% males, 79.4% White). The most common diagnoses were diabetes, spina bifida, cerebral palsy, and sickle cell anemia. Youth who completed chores manifested higher overall health care transition readiness (Î²^ = 5.17, p = .026) and better communication with providers (Î²^ = 2.98, p = .006) than youth who completed no chores. Higher chores frequency was not more predictive of higher health care transition readiness scores above and beyond the effects of having chores at all. These results suggest that responsible health care behaviors are related to similar actions in other areas of life. Assignment of chores may promote self-management and health care transition readiness in youth with chronic conditions

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Z Gastroenterol. 2021 Mar;59:250-54.

TRANSITION OF CARE IN A CASE OF OBESITY WITH METABOLIC SURGERY.

Stroh C, Luderer D, Meyer F.

The World Health Summit 2011 confirmed the epidemic-like occurrence of diabetes mellitus and obesity. In Germany, 62.7 % and 21.9 % of the population have a BMI of more than 25 kg/m² and more than 30 kg/m2, respectively. Currently, 10.5 obese people per 100 000 German inhabitants undergo bariatric surgery, while 86 and 114.8 per 100 000 in France and in Sweden, respectively, favor bariatric surgical interventions.

Aim: By means of a scientific case report, the instructive case of a young patient with morbid obesity is illustrated based on 1) selective references from the medical literature and 2) insights from the daily clinical practice in the case-specific medical and perioperative management after successful surgery for malformation in his childhood and, thus, the limited therapeutic options of metabolic surgery.

Case report (case-, diagnostic-, and treatment-specific aspects): 35-year-old patient with morbid obesity. **Medical history**: Status after surgical intervention for gastroschisis as a newborn (surgery report not available).

Clinical findings: Super obesity characterized by 234 kg and 174 cm (→ BMI: 77.3 kg/m²), hypogonadotrophic hypogonadism.

Approach & course: · Initial treatment with gastric balloon followed by a weight reduction of 46 kg within the first 6 months; however, despite weight reduction, development of an insulin-dependent diabetes with insulin resistance from a diet-based diabetes. · Repeat gastric balloon therapy for "bridging" but with no further weight reduction despite additional administration of GLP-1 analogues. ·

Surgical intervention: Removal of the balloon - termination because of excessive adhesions to the liver and spleen as well as filiforme hepatic lesions (histopathology: liver hamartoma).

Open surgery: extensive adhesiolysis because of previous pediatric surgery for gastroschisis, including associated non-rotation of the intestine with complete right-sided position of the intestine (left side: colon; right flexure: at infralienal position) prompting single-anastomosis duodeno-ileostomy (SADI)-procedure, leaving the stomach in situ with simultaneous cholecystectomy and herniotomy in sublay technique.
Outcome (early postoperative and mid- to long-term): The patient tolerated the intervention well. Postoperative course was uneventful with regard to mobilization, beginning of oral nutrition, and wound healing; there was a subsequent weight reduction due to a "common channel" of 250 cm.
Conclusion: While the increase of obesity prevalence in adults has currently stopped, incidence in children and teenagers is rapidly rising. The consequence might be that children and young adults who have undergone bariatric surgery in childhood and adolescence can develop complications from these former interventions as adults. Therefore, it is reasonable to recommend follow-up investigations within specialized centers according to well-established standards. On the other hand, the increasing prevalence of obesity in childhood leads to the possibility that adults who underwent pediatric surgery because of embryonal malformations may require an appointment with a bariatric surgeon at some point. For these patients (as a representative example of the transition of care phenomenon), the risk of metabolic surgical intervention is increased; such operations require the appropriate knowledge and expertise of the bariatric surgeon on embryonal malformations and their approach by pediatric surgery

ADHD

Bundesgesundheitsblatt Gesundheitsforschung Gesundheitsschutz. 2020 Jul;63:910-15.

ADHD IN THE TRANSITION TO ADULTHOOD: PREVALENCE, SYMPTOMS, RISKS, AND CARE.

Philipsen A, Döpfner M.

Attention-deficit/hyperactivity disorder (ADHD) is a common neurodevelopmental disorder. In contrast to earlier assumptions, ADHD at least partially persists into adulthood in 50-80% of the patients.This narrative review article highlights the risks, treatment options, and care requirements associated with the transition to adulthood. Available epidemiological and routine care data and guidelines are reviewed and screened for indications and recommendations to improve the health-care of adolescents with ADHD.Epidemiological and routine care data point to a care gap for adolescents with ADHD in the sensitive phase of transition from adolescence to adulthood. Specific transition concepts should be expanded and their effectiveness scientifically investigated

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Health Soc Care Community. 2020 Oct.

EXPERIENCES OF TRANSITION FROM CHILDREN'S TO ADULT'S HEALTHCARE SERVICES FOR YOUNG PEOPLE WITH A NEURODEVELOPMENTAL CONDITION.

Shanahan P, Ollis L, Balla K, et al.

Previous research has highlighted a lack of continuity of care when young people with a neurodevelopmental condition make the transition from children's to adult specialist healthcare services. A lack of planning, consistency, and availability of adult services has been found to lead to; increased anxiety, poor health outcomes, reduced support and some young people not receiving healthcare. The majority of transition research has focused on what health professionals consider important in the transition process, rather than focusing on the experiences of the young people and those closest to them. Our objective was to gather evidence from young people (and their families) who had experienced transition from children's to adult specialist healthcare services through semi-structured interviews. Volunteers were recruited from two London boroughs. All young people were aged between 18 and 25Â years with a neurodevelopmental condition (Attention Deficit Hyperactivity Disorder, Autism Spectrum Disorder and/or an Intellectual Disability). Overall, we interviewed six young people with support from a family member. Five further family members were interviewed on behalf of the young person. In total, ten semi-structured interviews were transcribed verbatim and analysed using Interpretative Phenomenological Analysis. Four themes emerged from the analysis: (a) Parents as advocates, (b) Availability of adult's specialist health and social care services, (c) Lack of information sharing and (d) Transition as a binary, abrupt change. Our findings suggest the transition experience could be improved by changing service specifications to incorporate assessment and handover across the age range of 16-20Â years. Additionally, statutory services should understand and provide the coordination role now offered by parents in transition. We suggest future research could evaluate the feasibility

of a patient-owned online information sharing tool with information about relevant services for young people and their families

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Scand J Occup Ther. 2021 Feb;28(2):78-90

PROOF OF CONCEPT: THE TRANSITION PROGRAM FOR YOUNG ADULTS WITH AUTISM SPECTRUM DISORDER AND/OR ATTENTION DEFICIT HYPERACTIVITY DISORDER.

Jonsson U, Coco C, Fridell A, et al.

BACKGROUND: The support needs of people with neurodevelopmental disorders are not sufficiently met during the initial years of adulthood.

AIM: To evaluate feasibility and preliminary effects of a novel programme designed to empower young adults with autism spectrum disorder (ASD) and/or attention deficit hyperactivity disorder (ADHD) to make progress within significant life domains (i.e. work, education, finance, housing/household management, health, leisure/participation in society, and relationships/social network).

MATERIAL AND METHOD: TRANSITION is a 24-week programme that combines group-based workshops with personalised support based on goal attainment scaling. The study enrolled 26 young adults (50% females; age 17-24years) in the normative intellectual range, diagnosed with ASD (n=8), ADHD (n=4), or both (n=14). The intervention was delivered by the regular staff of publicly funded psychiatric services in Stockholm, Sweden.

RESULTS: The programme was possible to implement with minor deviations from the manual. Participants and staff generally viewed the intervention positively, but also provided feedback to guide further improvement. There was a high degree of attendance throughout, with 21 participants (81%) completing the programme. All completers exceeded their predefined goal expectations within at least one domain.

CONCLUSIONS: The TRANSITION-programme is a promising concept that deserves further evaluation

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A transition clinic model for inflammatory bowel disease between two tertiary care centers: outcomes and predictive factors

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Abstract. – OBJECTIVE: Few models of transition have been proposed for inflammatory bowel disease (IBD). The aim of the present study is to evaluate the feasibility of a transition model and the predictive factors for success/failure.

PATIENTS AND METHODS: Patients with low activity or remission IBD were enrolled. Proposed model: three meetings every four-six weeks: the first one in the pediatric center (Bambino Gesù Children's Hospital); the second one, in the adult center (Foundation Polyclinic University A. Gemelli), with pediatric gastroenterologists; the last one, in the adult center, with adult gastroenterologists only. Questionnaires included anxiety and depression clinical scale, self-efficacy, quality of life, visual-analogic scale (VAS). Transition was considered successful if the three steps were completed.

RESULTS: Twenty patients were enrolled (range 18-25 years; M/F: 12/8; Ulcerative Colitis/ Crohn's Disease 10/10); eight accepted the transition program, four delayed the process and eight refused. Patients who completed transition generated higher scores on the resilience scale, better scores on well-being perception, and had lower anxiety scores. Patients who failed transition were mostly women. The perceived utility of the transition program was scored 7.3 on a VAS scale.

CONCLUSIONS: The proposed transition program seems to be feasible. Psychological scores may help in selecting patients and predicting outcomes.

Key Words:
Ulcerative colitis, Crohn's disease, Self-efficacy, Quality of life, Children.

Introduction

Crohn's Disease (CD) and Ulcerative Colitis (UC) are chronic diseases affecting children and adolescents in up to 25% of cases[1]. The early age incidence is increasing, typically with more extensive and severe forms when compared to adulthood[2]. Reaching the adulthood, this growing cohort of patients needs to undergo a very "special moment", the transfer from the pediatric center to the adult one. They have to move from a center where the care management refers to parents, to another where the care management is referred to the patients themselves. The chronic nature of these diseases, characterized by an alternation of exacerbation and remission, and the high associat-

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ed morbidity, makes the transition to the adult clinic an obligated step. This step is a delicate moment, and no standardized protocols exist up to now[3].

Research in other disciplines (rheumatic diseases, cystic fibrosis, diabetes mellitus type 1) shows that a structured program correlates with a better compliance, a better control of patient's disease and higher satisfaction[4]. In inflammatory bowel disease (IBD) this process should start at the pediatric center and should provide the young people with the necessary tools and the appropriate knowledge to make them independent in managing their disease[5].

Only a few models of transition clinics have been described for IBD, and almost none arising from the Italian cohort. A model of transition is proposed in this study, involving two tertiary centers for pediatric and adult IBD: Bambino Gesù Children's Hospital and Fondazione Policlinico Gemelli IRCCS. The aim of the study is to assess the feasibility and effectiveness of the proposed transition model. Furthermore, as secondary objective, the possible predictors for success/failure of the transition are analyzed.

Patients and Methods

Patients

Patients were enrolled based on the following inclusion criteria: diagnosis of CD or UC, according to current guidelines from European Crohn's and Colitis Organization (ECCO) and European Society for Paediatric Gastroenterology Hepatology and Nutrition (ESPGHAN), remission or low activity disease, age ≥18 years old, and follow up of at least 2 years in the pediatric center. The disease was considered stable when no significant clinical variations were found in the last 4 weeks before the T1 visit (variation greater than 3 points at clinical Mayo score for UC patients or higher than 3 at Harvey Bradshaw index for CD patients). Exclusion criteria were age <18 years and patients refusing to fill out the questionnaire or rejecting the transition process. The clinical characteristics of the study population and the enrolled patients are summarized in Table I and II, respectively. Ethical approval was obtained from local institutional review board (protocol number P/491/CE/2011). The transition was proposed to candidate patients between January and June 2015.

The Transition Clinic Model

The proposed transition model consists of three meetings/visits, fixed about 1 month apart, involving patients and pediatric/adult caregivers.

Table I. Characteristics of the studied population.

		Patient at T0 (% of total)
Male/Female		12/8
Mean age		20.2 (± 1.76)
Type of disease CD/UC		10/10
Time at OPBG		5 (± 2,23)
Age at diagnosis		15.2 (± 3.44)
Number of IBD centers changed following the diagnosis		1.3 (0.57)
surgery		10 (50%)
IBDQ		171.36 (± 35.59)
Previous treatment		
1. biologics		4 (20%)
2. immuesuppressants		12 (60%)
3. steroids		20 (100%)
4. antibiotics		5 (25%)
5. mesalamine		19 (95%)
Current treatment		
1. biologics		4 (20%)
2. immunesuppressants		5 (25%)
3. steroids		3 (15%)
4. antibiotics		0
5. mesalamine		12 (60%)
Montreal classification		
CD	A1	6 (30%)
	A2	4 (20%)
	A3	0
	L1	0
	L2	3 (15%)
	L3	6 (%30%)
	Upper disease	1 (5%)
	B1	5 (25%)
	B2	4 (20%)
	B3	1 (5%)
	P	3 (15%)
UC	E1	1 (5%)
	E2	2 (10%)
	E3	7 (35%)
	S0	0
	S1	2 (10%)
	S2	7 (355)
	S3	1 (5%)

The first visit (T1) is performed at the children's hospital, when the transition is "officially" proposed and explained. The second meeting (T2) is performed in the adult center, in the presence of both adult and pediatric gastroenterologists. The last meeting (T3) takes place at the adult unit with the adult gastroenterologist. The third meeting is still a dedicated visit, but carried out in complete independence and autonomy, similarly to the setting of the adult IBD clinic. Questionnaires are administered during the three visits. Physicians need to complete an independent questionnaire. The proposed model is described in Figure 1.

Table II. Characteristics of the enrolled patients.

	Success at T2	Failure at T2	*p*-value	Success at T3	Failure at T3	*p*-value
M/F	8/3	1/4	0.106	8/0	0/3	0.001
Mean age at transition	20.45 (± 1.63)	19 (± 1)	0.09	20.62 (± 1.84)	19.14 (± 0.89)	0.07
CD/UC	6/5	1/4	0.308	6/2	1/6	0.041
Follow up at OPBG	5 (± 1,94)	5,8 (± 2.16)	0.337	5,57 (± 1.98)	5.28 (± 2.05)	0.778
Mean number of other hospitals previuos to OPBG	1.36 (± 0.67)	1.2 (± 0.44)	0.631	1.37 (± 0.74)	1.14 (± 0.37)	0.470

Definition of Transition Outcomes

The transition was considered successful when the three meetings were completed. Failure could occur at each visit, and it was defined as the patient's unwillingness to go through the process or as not showing up at the appointment.

Questionnaires for patients:

During the first visit (T1), the patient was asked to fill the following questionnaires:

- HADS (anxiety and depression clinical scale)
- GSES (Generalized Self-Efficacy scale)
- CD-RISC (Connor-Davidson scale)
- IBDQ (IBD quality of life Questionnaire)
- VAS (visual-analogic scale) to evaluate respectively:
  - the current state of patient's health
  - disease activity in the last week
  - patient's personal perception about the independence in the disease management
  - confidence in the pediatric physician/team
  - grade of comprehension perceived about the adult physician/team
  - confidence in the adult physician/team

During the visits T2 and T3, the patient was asked to fill in only a few of the VAS.

HADS (The Hospital Anxiety and Depression Scale)

This test consists of l4 items exploring depression and anxiety. The timeframe analyzed is that of the previous two weeks, and for each answer there is a numerical score, expressed on a 4-point scale (0-3). The total score is obtained by

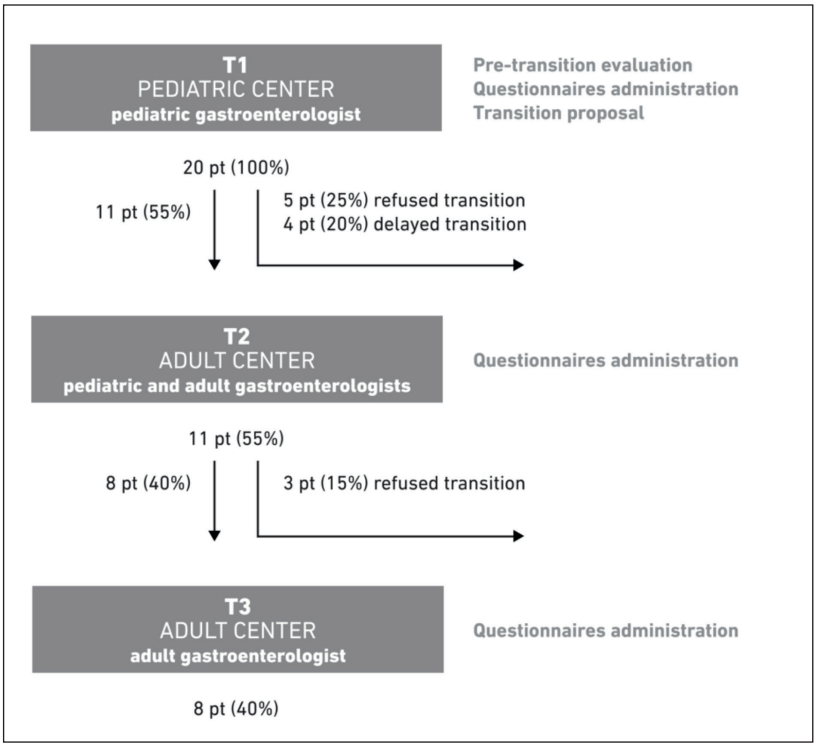

Figure 1. Chart of the proposed model and main outcomes.

summing up each item, and it ranges from 0 to 21 for anxiety or depression each. A score between 0 and 7 for each subscale can be considered normal. A score equal to or greater than 11 indicates the likely presence of a mood disorder. A score between 8 and 10 is suggestive of the presence of the state explored[6].

Generalized Self-Efficacy

The GSES was created to measure the perceived self-efficacy. Self-efficacy is defined as one's belief in one's ability to succeed in specific situations or accomplish a task[7]. The first version of the scale was created in Germany by Jerusalem and Schwarzer (1986) and consisted of 20 items, later reduced to 10 items (Jerusalem, Schwarzer, 1986; Schwarzer and Jerusalem, 1989, 1995). The GSES is a one-dimensional scale and uses a Likert scale of four steps (1 = "not true" to 4 = "completely true"). Individual differences are explored in terms of motivations, attitudes, learning and task performance. There are 10 items in total, and the maximum score is therefore 40. The higher the score, the greater the self-efficacy. The Italian version has been translated and validated by Sibilia, Schwarzer, Jerusalem (1995) [8].

CD-RISC

This scale is used to assess the resilience. The authors Connor and Davidson define resilience as "the ability to thrive also in difficult moments"[9]. According to the authors, it can be considered as a measure of stress-management capability. The CD-RISK, in the proposed version, is composed of 25 items, each based on a 5-point scale: (0) almost never true, (1) rarely true, (2) is true sometimes, (3) often true, (4) true in almost all cases. The score can thus vary between 0 and 100. The higher the score, the greater the level of resilience[9].

IBDQ

The questionnaire aims to evaluate the quality of life of patients with IBD, in reference to the last two weeks prior to completing the questionnaire. The quality of life is indeed a subjective index of perceived health. This questionnaire has proved to be a valuable tool that reflects important changes in health status and can also be used in clinical trials to measure the effectiveness of therapy. The questionnaire consists of 32 items that explore four dimensions: a) intestinal symptoms (10 items); b) State of emotional health (12 items); c) systemic symptoms (5 items); d) social functions (5 items). For each item, the patient is asked to express their opinion using a 7-point Likert scale (1 - worst function to 7 - best function). The higher the score, the better the quality of life of the patient. The minimum possible score is 32, the maximum 224[10,11].

Statistical Analysis

Database was imported in the IC STATA12 statistical software for MAC. The descriptive analysis was conducted with the support of the MICROSOFT EXCEL software for the creation of graphs and charts. The inferential analysis was performed using non-parametric tests for continuous variables: Spearman rank correlation test, and Mann-Whitney test. The hypothesis was rejected for alpha $p<0.05$.

Results

Feasibility of the Model

The present model was proposed to 20 patients, as potential candidates for transition. At the end of the first meeting, 5 patients refused the transition, while 15 patients were favorable (Figure 1). Four patients from the latter group were qualified as unstable during the T1 visit, according to a clinical evaluation, and pediatricians postponed their transition. These four patients were not considered in further analysis. The transition was continued with 11 patients (55% of the enrolled patients). Three patients refused to continue with the third visit (T3). A total of 8 patients (40%) completed the proposed model of transition.

The visits were organized properly, and no major problems were encountered during the process. For these reasons, the proposed model appeared to be feasible.

All patients enrolled in the program and called back for a delayed questionnaire appointed 7.3 on a 0-10 VAS scale to the utility of the transition program. Ninety percent of the contacted patients were glad about this experience and would repeat it again or suggest it to other patients (data not shown).

Disease Awareness and Knowledge of the Transition Process in Candidates of the Transition Clinic

Overall, the patients displayed a high perception of their independence in managing the disease and about the transition process (Figure 2A and 2B), in particular, by an average score of 77 on a 1 to 100 VAS of the perception of independence, and 2.42 of the readiness to transfer (tak-

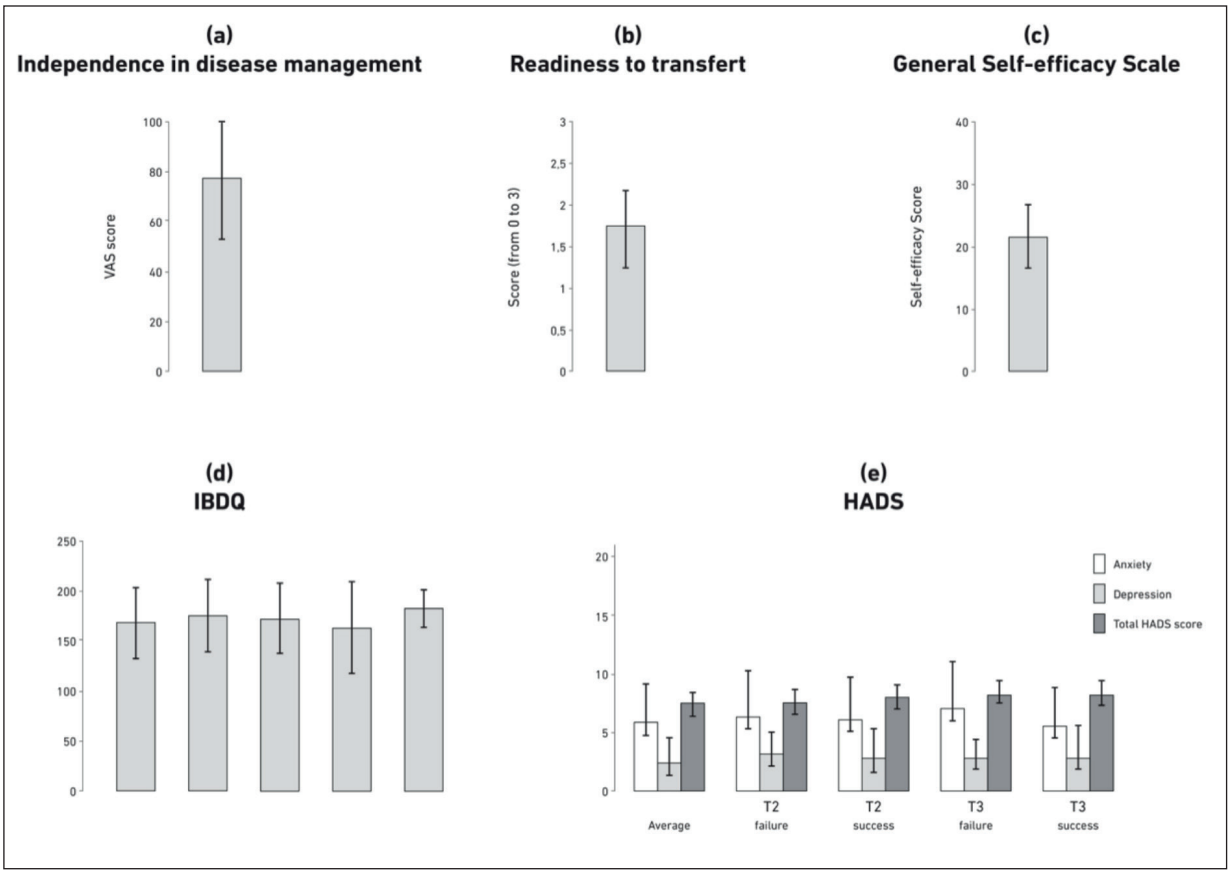

Figure 2. Disease awareness and knowledge of the transition process in candidates of the transition clinic (**A**-**C**). Multiparametric evaluation of patients and predictor factors of success for the proposed transition clinic process (**D**-**E**).

en from a Likert scale of four steps from 0 "disagree" to 3 "agree"). They displayed high trust in the physician, both pediatrician and the adult gastroenterologist with a mean value 90 (scores expressed on a 1 to 100 VAS). There are no differences in confidence between the pediatrician and the adult gastroenterologist in either T2 or T3 (data not shown). The average score, based on the total number of patients in the questionnaire designed to measure the generalized self-efficacy, is of 27.75/40 points (Figure 2C).

Multiparametric Evaluation of Patients and Predictor Factors of Success for the Proposed Transition Clinic Process

Classifying patients into failure and success of the transition process, a higher average score for trust in physician was found in success compared to failure ($p<0.05$).

The quality of life, assessed by IBDQ, was higher in success at T3 (185.37 points) compared to the total average (171.36 points) and failure at T3 (165.14 points) ($p<0.05$) (Figure 2D). These results paralleled with the perceived well-being measured by VAS scale (data not shown).

None of the patients generated any significant scores for anxiety and/or depression. When assigning patients to either failure or success, higher scores on the anxiety scale were registered in the failure group (6.25) compared to the success group (5.5) (Figure 2E). On the depression scale, although the scores relating to failures were slightly higher, the results did not show any statistical significance.

Self-efficacy assessment showed that higher scores were observed in patients failing the transition at T2 and at T3 compared to success ($p<0.05$).

More consistent results emerged from the analysis of the CD-RISC. In particular, success at T2 and at T3 showed a higher total score (70 points at T2 and 69.62 points at T3) compared to groups failing the transition at T2 and at T3 (60.2 points and 62.85 points, respectively).

This is more evident in the domains of personal responsibility - tenacity (23.72 points in T2 success group and 23.12 points in the T3 success group, 18.6 points in T2 failure group and 19.85 points in the group of failures to T3), in self-confidence (20.36 and 20.75 points in successful groups at T2 and at T3, 17 points in failure at T2 and 28 points of failure groups at T3) and that relating to the acceptance of the positive changes (14.45 and 14.85 for successful groups at T2 and at T3; 13.6 and 13.57 points for the groups who rejected the transition at T2 and at T3, respectively). Less significant are the differences observed in the domains related to the spiritual influences and control.

Discussion

The present study shows the real situation of two tertiary care centers in Italy. The proposed transition model has been designed taking into account the recommendations by the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN)[12] and considering the most recent major European and American pieces of evidence[13]. The structure of the program and the results were also compared with the recommendations by the Joint Expert Panel attended by the Italian Society of Paediatric Gastroenterology, Hepatology and Nutrition (SIGENP), the Italian Association of Hospital Gastroenterologists and Endoscopists (AIGO), the Italian Society of Endoscopy (SIED), and the Italian Society of Gastroenterology (SIGE).

The patients enrolled were homogeneous in age and clinical features. The proposal was accepted by 75% of patients, and we have completed the transfer of 40% of them. The sample size is small and the analyzed timeframe is short. This could influence the outcomes, considering the remitting-relapsing nature of IBD. The literature sources have underlined the importance for this process to occur in a stable phase of the disease, and therefore 5 amenable patients were postponed in this series.

At the beginning of the transition program, patients with CD and UC were equally represented, while patients who completed the transition program had CD (statistically definitive). Similarly, patients who completed the program were males, while at the beginning of the program there were 12 males and 8 females (statistically definitive). Patients who completed the program were older than those that failed in the process (difference about one year).

The study analyzed the perception of patients about the necessity of the transfer and their knowledge of the disease. Patients expressed a positive feedback when asked to judge their own perception of the transfer readiness. The independence perception was also positive. The trust placed by the young adult patients in the doctor was an important element in determining the success or failure of the transition.

To analyze predictive factors for success or failure, the success/failure at T2 and T3 were compared. The quality of life, assessed by IBDQ, was higher in successfully transferred patients at T2 and T3 compared to patients not responding to the proposal. The difference is significant considering the quality of life to be a subjective index of perceived health. It might be useful to propose the transfer to a stage where this perception is high. Similar results were obtained in evaluating the perception of the patients' well-being, which was greater in the success groups.

When analyzing the psychological characteristics of patients through the specific questionnaire (HADS), none of the patients had a significant score for anxiety and depression. In the group of transfer failure at T2 and T3 the scores relating to anxiety scale were slightly higher.

Another potential predicting factor for transition's effectiveness was the perceived self-efficacy, as measured on the scale of generalized self-efficacy. This enables to assess the individual differences in terms of motivation, attitudes, learning and task performance. In the present study, a higher score was observed in patients who had rejected the transition at T2 or at T3. This appears to be the only aspect mostly present in failure groups. This result may suggest that the patients who have not completed the transition are considered to be more independent and therefore feel less need to resort to a structured path.

Some interesting results were obtained using the CD-RISC. This scale is used to assess the resilience, as "personal ability to thrive in face of difficulty". Patients that passed the transition at T2 and at T3 generated higher average scores than groups that rejected the transition. Furthermore, patients respondent to the transition were more tenacious, had greater self-confidence, and therefore they could better manage negative emotions and have a greater positive acceptance to changes.

Regarding the "medical evaluation questionnaire", some interesting data have emerged about

the different assessment expressed by pediatrician and gastroenterologist: the pediatrician is more generous in the evaluation of their patients, and tends to assign higher scores. Only for the questions on the readiness to transfer, the scores given by the adult gastroenterologist are similar but slightly higher than the pediatrician's. The most important difference is related to the doctor-patient relationship's quality. As expected, the pediatrician's score differs from the adult gastroenterologist's, generating a higher value, since it is based on a mutual trust and understanding, built over time. In addition, the initial judgment by the pediatrician regarding the transfer readiness is consistent with what has been observed. Patients who complete the first and the second meeting at the adult hospital are considered more ready, as well as more prepared with an understanding of the transition process. As a result, this can highlight an important role of the transition program in increasing disease knowledge, and in the perceived transfer readiness, expressed by the upward trend of the scores assigned by the patient to the specific questions.

Conclusions

The proposed transition program seems to be feasible and effective. However, it is necessary to expand the sample size and apply a long-term follow up. The most difficult patients to be transferred are female with UC, who feel autonomous and independent and place less faith in the adult doctor. Their health and well-being at the time of the transfer is lower than that of the transferred patients.

The patients who complete the transition have different psychological characteristics: they appear to be less anxious, more tenacious, and more responsive to changes. These characteristics may be useful in discriminating the positive or negative response towards the transition and could be considered to better prepare patients for transfer.

The goal of the transitional path is to ensure the continuity of care, taking into account the physical, social and emotional development of the patient. A successful transitional program should promote adherence to treatment, expand the knowledge of the disease, and encourage the patient's autonomy in managing it independently, with the aim of improving or maintaining a stable control of the disease.

Author Contributions

Scaldaferri, Romeo, De Angelis and Ricca designed the work, performed the follow up visits and drafted the manuscript. Angelino revised the final draft of the manuscript and edited figures and tables. Ricca, Filoni, Ferrarese, Borrelli and Camardese administered and interpreted the psychological questionnaires. Scaldaferri, Filoni, Torroni, Faraci, Rea, Giorgio performed the follow up visits and acquisition and interpretation of clinical data. Lopetuso, Pizzoferrato, and Gaetani contributed to the acquisition and interpretation of clinical data. Poscia performed statistical analysis. Gasbarrini and Dall'Oglio contributed to design of the work and supervised the study. All Authors critically revised the manuscript for important intellectual content. All Authors approved the final version of the manuscript.

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Conflict of Interests

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Transitional care: A new model of care from young age to adulthood

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ABSTRACT

Introduction: Transition of care represents the transfer from child to adult care. An effective transition maintains continuity of care and presents better clinical outcomes. This process has assumed growing relevance, thanks to improved survivorship of chronic paediatric patients. Actually, there is no a one-size model fitting for all transitions, but each Service organizes its own clinical pathway.
Aim: The study proposes an organizational model for transition, differentiated according to patient complexity.
Methods: The working group discussed, through regular meetings, the appropriate transitional model for our Hospital. The working group defined a common scheme of transition and elaborated a synthetic document for patients. Then, the common model is adapted, through clinicians' contribution, for different diseases. The complexity assessment includes clinical data, nursing and social information.
Results: The working group defined a common model identifying the main information to be included and detailed in each transition report. The team defined two pathways based on patient's complexity. In case of good compensation and autonomous management, the adolescent is addressed towards standard transition process, a smoother transition from paediatric to adult care with direct connection among healthcare professionals. In case of complex clinical and/or social conditions, an Interdisciplinary Transition Group (ITG) is activated. The group preventively evaluates each patient in periodic meetings and provides a personalized planning of care. In order to define the complexity of a patient, clinical and social determinants are considered. Some diseases are considered complex by default, while others require ITG involvement in case of multiple comorbidities, severe clinical situation, concomitant social criticality and/or cognitive impairment.
Discussion: Transition of care represents an important phase in chronic diseases management. The proposed model assures a multidisciplinary approach, involving all specialists of both paediatric and adult teams. A key determinant of transition is information transmission. Then, the model proposes a common transition report format. Finally, a further perspective study is already in program, in order to assess clinical effectiveness.

1. Introduction

Transition of care is the transfer, for pediatric patients with chronic diseases, from child to adult care [1]. This healthcare transition is meaningful as corresponds to the passage from a family-centered and parents reliant model of care (pediatric care) to adult care, mainly focused on individual patient and requiring more autonomy in disease management [2]. Three main goals in transition are unanimously recognized: the achievement of sufficient skills and knowledge before the transfer, the readiness of patient and parents, and the correct information of adult care professionals regarding patient clinical history [3]. An effective transition should consent the maintenance of the continuity of care [4], and must be preceded by a transition readiness assessment and an education process [5]. Nevertheless, actually, a well-validated questionnaire measuring readiness for transfer is not yet universally recognized [6]. Transition is not a single moment, but an organized process precociously initiated, finishing with the effective transition of care [7,8]. For example, considering type 1 diabetes, experts recommend a transition of at least 12 months

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[9]. The importance of a multidisciplinary approach in transition is highlighted [1]. Indeed, transitional care is a holistic active process addressing medical, psychological and social needs of young patients [10]. Therefore, different healthcare professionals, as well as patients and families take part to transition [8]. In this context, increasing evidence indicates the important role of nurses as case managers, coordinating the multidisciplinary group, acting as a junction among specialists, patients and families [11]. Indeed, the transition process can be critical not only for patients, but also for parents. However, they can act as important key facilitators for children, supporting them in becoming aware of their diseases [12]. Actually, thanks to advances in medical care, children with different chronic diseases successfully reach adulthood, improving survival and quality of life. As a consequence, increasing numbers of adolescents have to be transited to adult care [13–15]. In accordance with this epidemiological framework, in recent years, international literature focuses on transitional care, especially considering some diseases as sickle cell disease [16], inflammatory bowel disease [5], type 1 diabetes [7], or rheumatic diseases [8]. Main critical issues in transition are insufficient patient preparation, subsistence of pediatric care in adulthood and dropouts in medical supervision [17]. Other emerging difficulties are: the need to integrate psychological and clinical care, excessive parental involvement, and multidisciplinary coordination [18,19]. Unfortunately, especially considering rare diseases, many patients do not receive age-appropriate medical care throughout adulthood. Indeed, for some typical pediatric diseases, clinical experience in adult settings is still limited [20]. An ineffective transition can determine negative outcomes for young patients [1]. Then, transition represents a critical phase, since negative outcomes in this period could have acute and long-term consequences [1]. For example, ineffective transition in type 1 diabetes potentially translates in a decrease in control visits, an increase in glycated hemoglobin (HbA1c) and a rise in hospitalization and emergency visits [7]. The clinical outcomes, for monitoring transition, vary in different diseases and can be represented by specific biochemical indicators or by organizational parameters as missed follow-up visits [21]. Moreover, other indicators must be considered as quality of life, patient and family satisfaction, and adherence to treatment [22].

The growing relevance of transitional care is highlighted by the development of international guidelines [17]. In Italy, the National Government, with the recent "National Plan in Chronic Care", identified transitional care as a priority area for action [23]. Despite the increasing interest regarding this theme, there are still many topics debated, such as the correct age for transition [1]. About this, some studies refer to 18 years old, while others suggest 16 years or even precocious transitions [1,19]. In any case, transition corresponds with adolescence, a potentially critical phase of life, and some degree of flexibility is suggested [1,5,19]. Indeed, transition is not exclusively related to chronological age, but mainly refers to patient maturity [8]. Actually, there is no a one-size model fitting for all transitions, but each Service or Hospital organizes its own clinical pathway [8,24]. Scientific Societies identified some core steps in providing transitional care: communication of transition, patient empowerment, providing to adult care all medical records and summaries, and flexibility [25]. Moreover, evidences regarding long-term outcomes and cost-effectiveness of transitional care often are lacking [5,10]. A recent systematic review regarding transitional care, considering a total of 238 participants, advocates the necessity of further studies with longer follow-ups [26]. Despite the absence of a single model for transitional care, emerging evidences identified the importance of differentiated pathways according to clinical complexity [27–30]. About this, a recent English study estimates that approximately 30 % of the population requiring transition presents a complex chronic condition or other complex nursing needs, requiring a more tailored program, while the remaining 70 % presents a good clinical control. For stable patients a smoother transition process is possible as well as desirable [30,31]. The definition of clinical complexity in children, despite several tools proposed, is still subjective, considering clinical conditions, comorbidities, psychological, and social needs [32]. The recognition of medical complexity in children is vital, because this population presents extensive healthcare needs, often experiencing important functional limitations and representing a group of high resource utilizers [33]. Moreover, in defining complexity of children is important to consider, also, social determinants [34,35]. Indeed, social criticalities can significantly affect clinical outcomes [34]. The presence of social risk factors increases the family need of care coordination and often requires the involvement of other services (e.g. Social Services), even for those patients not considered clinically complex [36].

2. Objectives

The study describes the organizational model for transitional care developed in our Hospital considering transition from pediatric to adult care for different chronic diseases. The authors described two processes of transition differentiated according to patient complexity. Moreover, preliminary data regarding endocrinology patients transited are reported. The model proposed aimed to provide valuable solutions to the criticalities described throughout the introduction, in particular the lack of care integration, the need of multidisciplinary assistance, the improvement of patient autonomy, and the need of more in-depth data collection and transmission between different healthcare teams.

3. Methods

3.1. Context

"*A.O.U. Città della Salute e della Scienza di Torino*" Hospital (City of Science and Health - AOU), located in Turin, represents one of the biggest University Hospitals in Europe, with 2400 beds, nearly 100,000 admissions/year and 200,000 visits in Emergency Department (ED). Following a regional act in 2012, four important Regional Hospitals were unified, forming the City of Science and Health. This facility is composed by Molinette Hospital, S. Anna Hospital, Regina Margherita Hospital and CTO Hospital. Molinette is the biggest hospital in Piedmont Region, and is an important landmark for adult care. Regina Margherita Hospital is a pediatric hospital, while S. Anna Hospital is specialized in taking care of women's diseases and pregnancy. Finally, CTO is devoted to orthopedic surgery, traumatology, plastic surgery and neurosurgery. All together these hospitals guarantee highly specialized diagnosis, care and health assistance for adults, women and children. In particular, Regina Margherita Hospital represents a regional and national referral center for several pediatric diseases, especially considering rare diseases. Therefore, a growing number of adolescents with chronic diseases need to be effectively transferred from pediatric care (Regina Margherita) to adult services, preferably adult hospitals of the AOU or other reference centers [37]. Considering the specificity of our setting, assuring care for the entire life-span of chronic patients, as well as the role of regional or national referral for several clinical branches, transition of care represent an important topic of discussion. The main challenges individuated in our setting are:

- Improving patient and family autonomy in diseases management, prior to proceed with transition;
- Assuring the continuity of care to patients in different Hospital settings, often characterized by different approaches to subjects;

- Assuring a correct and complete informational transition among the healthcare professionals involved in the process;
- Limiting, as far as possible, the losses of patients to follow-ups;
- Consenting an effective transition within the AOU or to other Hospitals, according to patient and family preferences.

3.2. Model definition

During November 2015, with a specific resolution, a Hospital Commission was established, in order to define the organization and the activities related to transitional care. The Commission started its work with the activities aiming to:

- Define and describe the transition pathways already active in the Hospital;
- Identify, trough specific audits with involved healthcare professionals, the critical points emerging in the transition from pediatric to adult care;
- Collect operational proposals to give full and complete response to the transition needs identified.

The works of the Commission lasted 2 years, consisting of regular meetings, with the preparation of specific documents for patients and healthcare professionals, as well as analyses and comparisons with the existing literature.

A multidisciplinary group for transition, composed by: medical doctors (medicals specialized in Public Health, pediatricians, child endocrinologists, pulmonologists, child neuro-psychiatrists, pediatric surgeons, doctors in internal medicine, endocrinologists, diabetes specialists and neurologists), pediatric and adult nurses, psychologists and social workers, was specifically composed during 2017. This group (acting as a Delphi group) discussed, through regular meetings (about 20), the appropriate transitional model to be adopted in our Hospital, contextualizing it, through clinicians' contribution, for different diseases. The working group defined a common scheme of transition, explicated throughout transition programs shared with all clinicians involved. Then, the general scheme was specifically applied in different contexts. Moreover, a synthetic explanatory document for patients and families was prepared and published on the Hospital website. The group defined two general pathways based on clinical and social complexity. The group collected the quantitative data available regarding transition, using them as a fundamental background in the model definition, in order to better frame the real dimensions of the problem, providing some important hints to the model definition. These quantitative records were completed by some random qualitative interviews to patients to be transited or already transited, and to their families. These interviews were conducted by healthcare professionals specifically trained, and were employed to integrate the point of view of patients and families within the proposed model.

The Commission concluded its work during 2018, and during the same years, the Interdisciplinary Groups were activated, with a composition and a periodicity of meetings strictly depending from the diseases treated.

The present work describes the model characteristics and some preliminary results. However, a supplementary study is already implemented, in order to investigate the effectiveness of the proposed model with more updated and expansive data.

3.3. Complexity assessment

The working team elaborated two different pathways, based on complexity assessment. For an adequate assessment, a tool for complexity identification was discussed and elaborated, integrating clinical data, nursing and social information.

3.4. Statistical analysis

Preliminary results of the Endocrinology Unit were assessed, in order to define the transitional pathway. All endocrinology pediatric patients transited to adult care from January 2016 to January 2019 were considered. Since a well-structured plan of transition for patients with type 1 Diabetes already existed in our Hospitals, these patients were excluded from preliminary data reported in the article. For each patient, the following data were collected: diagnosis, age at diagnosis, age at transition, number of jointed visits in the adult center, drop-outs to transition and then during the first year of adult care, time between first adult visit and last pediatric assessment, frequency of visits pre- and post- transition and ED visits and unexpected hospitalization post-transition. Numbers and percentages of patients at different steps were computed. Descriptive statistics were applied in order to define sample characteristics, as mean, standard deviation (SD), and range for continuous variables and frequencies and percentage for categorical variables. All statistical analyses were performed using STATA 15. '

4. Results

4.1. Model description

The working group elaborated a general transition scheme to be applied for all diseases (see Fig. 1). The general pathway is described below. Since all considered diseases are chronic conditions, the communication with parents and patients regarding transition is generally implemented precociously, if possible since diagnosis. The progressive preparation of patients, including patient and family empowerment, took place during adolescence with individual and small-group meetings with the healthcare team (nurses, doctors, psychologist and social workers). Finally, the effective transfer of setting is planned at 18 years old, or as soon as the patient showed adequate autonomy in disease management. This model assures flexibility. Indeed, the age at transition could be postponed in case of concomitant cognitive or behavioral impairments, delays in anthropometric development, variations in therapy regimen, completion of surgical plans, and/or acute diseases. The preparation process included patient and caregiver empowerment regarding disease knowledge, therapy and potential urgent situations management. An effective transition presents as a requisite the achievement of adequate autonomy in disease management. Indeed, insufficient self-efficacy can determine variations or deferments in transition and/or the activation of a multidisciplinary evaluation. Several pediatric chronic diseases can occur in different ethnic groups. Naturally, any linguistic and/or cultural barrier can complicate the communication process. Consequently, the team of care has to evaluate the need of a cultural mediation support. This Service is available in our Hospital, and can be requested in any phase of care. Moreover, since chronic diseases can determine disabilities and/or cognitive impairment and/or social burden the need of psychological support for patients and families has to be assessed, also during transition. Moreover, during transition, any concomitant social problem must be adequately assessed. In particular, the working team recognizes as particular conditions of fragility: institutionalization, foster care, precarious family or economic situations. A specific document, integral part of the transition report, including nursing and social information, has specifically been prepared. During transition from pediatric to adult care, the transmission of clinical and social information between the two healthcare teams is necessary. This communication takes place, mainly, through the drafting of a detailed transitional report (medical, nursing, social, psychological), elaborated by the pediatric team. This report is prepared before the last visit in the pediatric setting

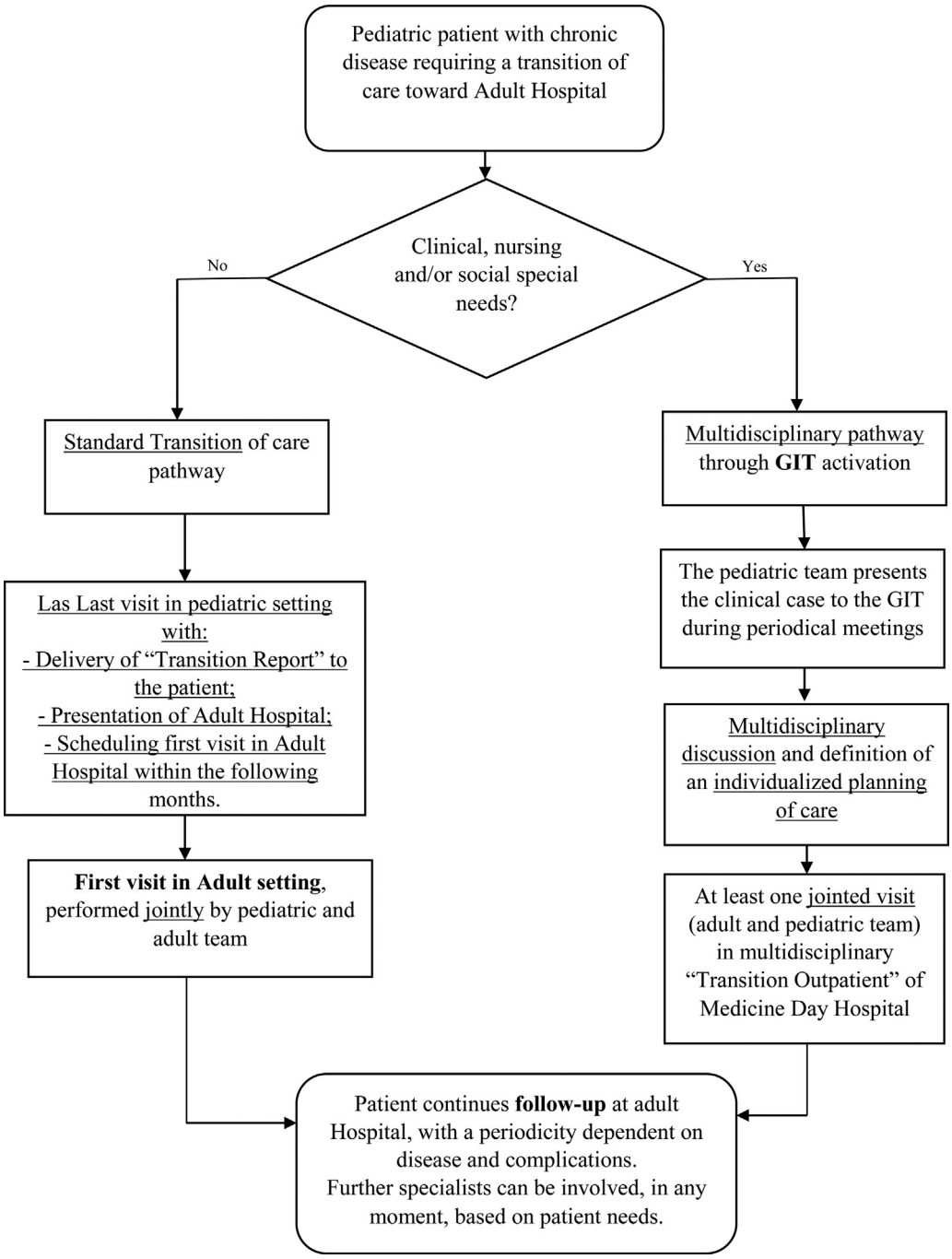

Fig. 1. The following figure synthetizes the Transitional Care model of our Hospital.

and delivered to the patient in this occasion, and then transmitted to the adult team of care. The working group defined a common model for this report, identifying the main information to be included and detailed as:

- Active clinical problems and treatment plans;
- Concomitant diseases and complications;
- Psychological condition of patient and family, and if necessary activation of psychological support;
- Fertility aspects (including if necessary a genetic consult);
- Potential urgent conditions;
- Follow-up characteristics: e.g. modification in standard follow-up;
- Use of aids and/or therapeutic plans;
- Vaccination status;
- Rehabilitation needs;
- Specialized nursing care needs;
- Social determinants.

This report is essential in order to assure the continuity of care between pediatric and adult teams, providing to adult healthcare professionals the main information required to guarantee the correct take in care of each patient. Indeed, the report clearly summarized the clinical history of individual patient during pediatric age. Finally, the report is an important multidisciplinary tool, in order to give a complete view of the patient situation, including clinical information, nursing and social details. Sharing in advance

this report among the teams consent to jointly discuss the case, prior to proceeding with the effective transition.

Many chronic diseases can determine potential urgent conditions. The patient/caregiver during transition is educated regarding the management of these situations. Moreover, patient and caregiver are informed of the change in ED referral at the age of 18 (from pediatric to adult ED). For some diseases, during transition, an emergency card is provided to patients, containing first aid information. During transition the healthcare team presents to patients the available tools supporting self-management e.g. smart-phone applications. Rehabilitation needs are multidisciplinary assessed and the individualized rehabilitative program is generally completed by Territory Services. The plan is defined considering the autonomy degree of patient/caregiver. The duration and frequency of the intervention strictly depends on patient needs and disease stage. Specialized nursing needs are assessed considering patient autonomy and self-efficacy. Transition acts as an important educative opportunity, in order to improve patient knowledge regarding the disease and therapy as well as to reduce the dependence from pediatric care. The healthcare professionals involved vary considering different diseases, with the identification of a case manager, usually a nurse, with coordinating functions. In any case, on the basis of patient needs, nurses of continuity of care, social workers or other professionals can be involved. Moreover, during transition a direct connection with Primary Care is expected. In particular, General Practitioners (GP's) need to be involved through direct participation to transition visits or through the transmission of clinical documentation. Other territorial health and social services are specifically involved depending on patient needs. Moreover, the collaboration with voluntary associations, scientific societies, schools and sport associations can provide an added value to the process. Generally, the Adult Center of reference is Molinette Hospital. However, each patient can request a transition toward other regional or extra-regional Hospitals, for example in case of logistic or organizational problems. The choice of the adult setting of care should be shared between patient, family and pediatric team. Indeed, the Adult Hospital has to respond to some professional and structural requirements. In particular, the hospital must guarantee adequate multidisciplinary experience as well as sufficient technological and diagnostic supports. Then, the pediatric team prepares a detailed report for the adult care team. Finally, specific tools for monitoring the entire process are identified, considering quantitative indicators and questionnaires, investigating patient quality of life, satisfaction and adherence to therapy/visits.

4.2. Complexity definition

The working team defined two pathways of transition based on patient's complexity. In case of chronic patients with good compensation and satisfactory autonomous management, the adolescent is addressed towards a standard transition process. In these cases, a smooth transition from pediatric to adult care is possible, with a direct connection among the involved healthcare professionals. The transition required a last visit in the pediatric setting. During this visit the adult setting of care is described to patient and family and the first visit in the new setting is programmed within the following months. In this occasion, the pediatric team gives to the patient the "transition report", containing the information described above. The patient transmits this report to his GPs and takes it with him at the first visit in the new setting. Moreover, during transition a presentation brochure is provided to patients, presenting adult services as well as the contacts of the new team of care. Then, the first visit in the adult setting is carried out jointly between pediatric and adult specialists. Afterwards, the follow-up continues in the new setting with a periodicity dependent on the pathology. After effective transition, adult specialists can contact the pediatric team for further consultations or in case of emerging issues requiring a jointly definition. In each moment of transition, further professionals could be involved, according to patient needs. If necessary, more jointly visits can be organized. In case of complex clinical and/or social conditions, such as multiple comorbidities or particularly severe clinical conditions, an alternative transition pathway is initiated. In these cases, an Interdisciplinary Transition Group (ITG) is activated. The ITG is composed by all the professionals involved in pediatric and adult care. A Chief Medical Officer, expert in continuity of care, acts as coordinator of the group. In this team, also, nurses, social workers and psychologists are involved (for both the pediatric and the adult team). The group preventively evaluates each patient and the available medical documentations. Then, the pediatric team presents to ITG in periodic meetings the clinical cases considering all aspects (clinical, nursing, social and psychology). Therefore, the group proceeds with an individual and personalized planning of the following pathway of care. In these cases, adult care can benefit of the contribution of the Unified Day Hospital of Medicines, located in Molinette Hospital, with a dedicated multidisciplinary outpatient service for transition of care. At least one of the visits taking place in the adult setting must be carried out jointly with the pediatric team, in order to assure the continuity of care. To define the complexity of a patient and, therefore, identify the necessity of the ITG activation, clinical and social determinants are considered. Some diseases are considered complex by default as associated with multiple comorbidities, severe clinical situations or extremely rare incidence as some genetic syndromes, for example McCune Albright Syndrome. Conversely, other diseases generally follow standard transition (e.g. type 1 diabetes, coagulopathies, etc.), requiring the ITG involvement exclusively in case of multiple comorbidities, severe clinical situation, concomitant social criticalities, and/or cognitive impairment. Social determinants are considered as integral part of the patient complexity evaluation, in accordance with the international evidences in the field [34,36]. In this context, specific potential determinants of social frailty are identified in: foster care, caregiver necessity, critical socio-economic status, family structure and critical housing conditions.

In our specific context, it is estimated that around 15–20 % of children to be transited each year need the ITG activation. This process is extremely variable depending from the underlying diseases of the subject as well as the social condition. In particular, some rare diseases are considered complex by definition e.g. some severe neurological condition and genetic diseases associated to mental impairment. Indeed, in these cases, the multidisciplinary team seems essential in order to effectively take in charge the entire household, assuring the active and precocious involvement of social workers and nurses (in the Hospital and on the territory).

5. Preliminary results

The data of pediatric endocrinology patients transited to adult care from January 2016 to January 2019 were collected. Since a well-structured pathway for diabetic patients already existed in our facility, these patients were excluded from these preliminary analyses. During the considered period, 61 patients were successfully transited to adult care. Table 1 describes the main characteristics of transited patients, grouped by main baseline disease. Thirteen patients (21.3 %) presented a pituitary or hypothalamic disease as principal diagnosis (6 boys and 7 girls). The mean age of transition of these patients was 19.6 years old (range: 16–27 years), and all patients were addressed to the AOU Endocrinology Unit for their follow-up. No patients have concomitant motor disabilities, while one patient suffers of epilepsy and blindness and one presented concomitant social criticalities. Fifteen patients (24.6 %) presented

Table 1
Main characteristics of endocrinology-transited patients. The table described the main characteristics of endocrinology-transited patients, grouped by the main baseline diseases.

Pathology treated	Sex	Mean age at diagnosis	Mean age at transition	Hospital of transition	Additional disabilities	Contacts with GP's
Hypothalamus and pituitary diseases (n = 13)	Males: 6 (46.1) Females: 7 (53.8)	9.4 years (range: 1–14 years)	19.6 years (range: 16–27 years)	Molinette endocrinology: 13 (100)	2 (15.4)	0
Adrenogenital syndrome (n = 15)	Males: 5 (33.3) Females: 10 (66.7)	1.7 years (range: 0–14 years)	20.9 years (range: 18–38 years)	Molinette endocrinology: 15 (100)	2 (13.3)	0
Neurofibromatosis type 1 (n = 18)	Males: 10 (55.6) Females: 8 (44.4)	3.8 years (range: 4 month-13 years)	18.8 years (range: 16–23 years)	Dermatology: 14 (77.8) Rare disease Day Hospital: 3 (16.7) Other regional centres: 1 (5.5)	2 (11.1)	0
Down syndrome (n = 13)	Males: 7 (53.8) Females: 6 (46.2)	From birth	17.9 years (range: 15–19 years)	Molinette endocrinology: 13 (100)	7 (53.0)	0
Other pathologies (n = 3)	Males: 1 (33.3) Females: 2 (66.7)	–	–	–	–	–

as primary diagnosis a congenital adrenal syndrome (5 boys and 10 girls), with a mean age of transition of 20.9 years old (age: 18–38 years). All patients were addressed to Endocrinology Unit of Molinette Hospital for adult care. No patients presented motor disabilities, while two have concomitant social criticalities needing to be further assessed during transition. Eighteen patients (29.5 %) presented an initial diagnosis of neurofibromatosis 1 (10 boys and 8 girls), with a mean age of transition of 18.8 years (range: 16–23 years). Seventeen patients (94.4 %) were addressed to Molinette Hospital for adult care (to Dermatology Unit or to Rare Disease Day Hospital), while one patient continued his follow-up at Biella Hospital (another regional referral center). Two patients presented concomitant motor disabilities, while none suffers of social issues. Thirteen of the transited patients (21.3 %) presented Down syndrome (7 boys and 6 girls), with a mean age of transition of 17.9 years old (range: 15–19 years). Finally three patients (4.9 %) with other endocrinologic diagnosis were transited during the period of interest.

Table 2 reports the main outcomes of transition for the 61 patients transited during the period of interest. Out of 61 patients, 48 patients attended at least one jointed transitional visit (78.7 %), for two patients there were not sufficient data (3.3 %), while 11 patients did not attend any scheduled transition visits with a drop-out of 18.0 % to transition. The time elapsed from the last visit in pediatric setting and the first in the adult Hospital ranged from 7 days to 21 months. Finally, 57 patients (93.4 %) did not present any ED visit or hospitalization after transition, 2 patients (3.3 %) presented at least one ED visit, while for other 2 patients (3.3 %) there were not further data regarding ED visits after transition.

6. Discussion

Transition of care from pediatric to adult hospitals represents an important phase in chronic diseases management, corresponding to adolescence a generally critical period [1]. The effectiveness of clinical transition is important considering patient short- and long-term clinical outcomes [1,7]. Thanks to clinical and technological innovations, survival and quality of life of pediatric patients with chronic diseases is considerably improved [31]. Therefore, increasing numbers of adolescents and young patients have to be transited to adult care [13–15]. The growing impact of this process was associated to an increased attention towards this field [8,17,23]. Actually, there is no a one-size model fitting for all transitions, but each Service organizes its own clinical pathway [8,24]. Therefore, the aim of the present study was to define and describe an organizational model to be applied in our specific context, and potentially adapted to similar settings. According to recent international literature [30,31], we identify two different models of transition considering patient clinical and social complexity. In particular, in case of good clinical control a smoother process was defined, with a direct connection between all specialists involved (pediatric and adult care). On the other hand, in case of complex diseases (e.g. multiple comorbidity, severe clinical situation, social fragility, cognitive impairment, growth delays. . .) a dedicated multidisciplinary group is activated, in order to plan the following care in adult setting. The importance of coordination during transition is largely proven [1,10]. Our model assures a multidisciplinary approach, involving all specialists taking care of patients, nurses, social workers, continuity of care professionals and psychologists, of both pediatric and adult teams. The nurses of the two teams of care engage the role of case managers, acting as coordinators among specialists, and connection with patient and families. Considering the model proposed, the family is continuously involved in the process. In particular, considering chronic diseases, this awareness process is precociously initiated. On the other hand, a more in-depth patient empowerment process is started as soon as possible, with a specific focus on adolescence, favoring patient autonomy also trough small-group activities. In case of ITG activation, a Chief Medical Officer, expert in continuity of care, acts as coordinator of the group. The definition of complexity in pediatric age is still debated [32,33]. However, in our model to identify complex patients, we combined clinical and social evaluation, supporting the pediatric team in complexity identification. A further study is programmed in order to elaborate a specific "complexity scale", including information on comorbidities, motor difficulties, need of a caregiver, cognitive impairment and social criticalities. Preliminary results on Endocrinology Units described the spread of these characteristics in our population. The proposed scale, defined multidisciplinary with doctors, nurses, social assistants and psychologists, will be tested and further assessed in future quantitative and qualitative studies, in order to identify the items more predictive of clinical complexity.

A key determinant in transition success is information transmission [18,19]. In order to deal with this potential obstacle, our model proposes a transition report format. Then, the format is specifically completed by pediatric team and shared with the adult team of care and GPs. The report contains clinical, nursing, social-care and

Table 2
Main registered outcomes of transition. The Table reports the main registered outcomes of transition for the 61-endocrinology patients transited to adult care during the period of interest. The table reports the visits attended by patients in the two-year period after effective transition. Transition included 1-2 joined visits, then the FU is continued in the adult setting accordingly to a clinical schedule related to the clinical conditions of patients.

Number of transitional visits	Compliance to visits at 1-year	Time between last paediatric visit and first adult visit	Frequency of pre-transition visits	Frequency of post-transition visits	Not-programmed hospitalization or Emergency Department access
2 visits: 46 (75.4) 1 visit: 2 (3.3) 0: 11 (18.0) No collected data: 2 (3.3)	Drop-outs: 4 (6.6)	7.3 months (range: 7 days – 21 months)	8.6 months (range: 2–12 months)	8.4 months (range: 3–18 months)	2 (3.3)

psychological information. This transition report can act as a key facilitator in continuity of care, one on the main goal of effective transition [3].

According to literature evidences [7,8], transition is not a single moment, but a process beginning in pre-adolescence with progressive patient empowerment and ending with effective transition. In our model, the importance of patient and family education is carefully considered, and self-efficacy and autonomy in disease management are defined as indispensable pre-requisites for starting transition. Moreover, flexibility in the process is guaranteed, as required by international guidelines [17,25]. Indeed, transition is precociously initiated and can be delayed considering clinical issues and insufficient autonomy in disease management. The flexibility of the proposed model is confirmed assessing preliminary data of Endocrinology Unit regarding the effective age of transition, presenting a variable range of ages strictly depending on patient characteristics, social criticalities and comorbidities. This decision is in accordance with the scientific evidences suggesting that transition is not related only to chronological age, but mainly to patient maturity [8]. Transition involves not only patients, but their entire families [12]. Therefore, the whole family is considered when assessing disease burden, especially concerning the need for psychological support.

The model plans to measure some common indicators in order to assess the effectiveness of the process. In particular, dropouts to scheduled transitional visits will be carefully measured consenting, when possible, to patients to re-schedule appointments. The analyzed period, presented in preliminary results, included the first patients transited following the new proposed model, outlining promising results as nearly 80 % of patients attended at least one jointed transitional visit.

Considering the preliminary results on dropout indicator (18.0 % of endocrinology transited patients), attrition should be carefully considered defining target approaches in order to decrease this concerning phenomenon. Moreover, the resort to emergency care after transition will be considered. As presented in the preliminary results this indicator was favorable in our study (lower than 4%). Also the time between the last visit in pediatric setting and adult care seemed reasonable (see Table 2) considering the involved diseases. Concerning the frequency of visits in the two settings, prior and after transition, only slight differences were seen. A criticality potentially emerging from preliminary data is the scarce connection with GP's, and for certain this step need to be further investigated in subsequent analyses. As expected, nearly all transited patients were addressed to the adult departments of the AOU, with a little resort to other regional referral centers. This aspect requires to be further considered, since it is important to satisfy patients and family preferences, also considering logistic necessities. At this regard, a more close cooperation with other local centers needs to be established. Other specific clinical indicators will be defined considering the specific diseases in study e.g. Hb1ac for diabetes.

The described model is formally adopted in our Hospital from 2018, with previous pilot experiences in some Units as Pediatric Endocrinology. The following programmed steps are to monitor and evaluate the effectiveness of transition. Therefore, a further perspective study is already in program from 2019, in order to assess clinical effectiveness and continuity of care for young patients transited according to the adopted model. Moreover, this study will assess also qualitatively patient, family and healthcare professionals experiences regarding transition using questionnaires and interviews.

The main limitations of our study are that we considered only our Hospital organization and a limited spectrum of diseases (in particular endocrinology diseases). These constraints could lead to some issues of generalizability of the model in other contexts. However, the progressive extension and contextual application to other diseases is already expected, for example considering neurological diseases and complex genetic syndromes. Moreover, it could be interesting to expand the proposed model to other hospitals, adapting it to the peculiarity of the chosen settings.

7. Conclusions

In conclusion, transition of care is a theme of growing interest [2,17,23], however a unique transition model did not exist [8,24]. Therefore, considering international guidelines and evidences [1,10,17], we proposed a transition model adapted to our clinical context, potentially applicable to other clinical settings or diseases.

The proposed model assures holistic care to patient and family, aiming to ensure therapeutic continuity, maintenance of good clinical outcomes and promoting young patients wellbeing. In order to achieve these goals, the multidisciplinary approach is extremely important assuring the involvement and coordination among nurses, clinicians, social workers, psychologists, and all other professionals involved, in both pediatric and adult care setting. In conclusion, the proposed model consented to effectively face in our site the issues related to the preservation of the continuity of care for transited patients, assuring a multidisciplinary take in charge. Moreover, in case of complex patients, the activation of ITG guarantees the multidisciplinary approach to patient needs, effectively fixing this important matter. In this context, the involvement of GPs results extremely important. Indeed, GPs have the key role of patients' management on Territory. The active involvement will be obtained through the direct participation of GPs to the transition visits. In order to collect also their opinion, potentially improving this important part of the model, interviews will be planned and conducted also at Territory level to collect emerging criticalities at this step. Finally, further studies regarding clinical and organizational outcomes are desirable, in order to implement potential improvement or corrective actions. About this, a prospective cohort study is already planned in our hospital from 2019. This study will investigate outcomes and process indica-

tors, as well as qualitative experiences through questionnaires and semi-structured interviews with patients, parents and healthcare professionals. Considering the importance of integration at different levels, also beyond the hospital, it seems particularly important to share the proposed model at a Regional level, in order to start a productive discussion. The aim of a similar thread is to develop a shared policy to be applied on the entire regional territory. In this way, the proposed model would be tested also in different contexts, solving the abovementioned issue of generalizability. About this, the working group already has disseminated the proposed model at different levels, in order to start a profitable collaboration.

In summary, the main limitations of the present study are related to the model implementation in a single center and to a limited number of diseases, namely endocrinology illnesses. However, as previously mentioned, these constraints will be addressed in the following months, trough the implementation of a prospective study and the extension of the model to other settings and diseases.

In conclusion, the model extensively described throughout the article is intended to solve the main issue regarding transition mentioned within the introduction. In particular, the new model seek to solve the problems related to the fragmentation of care for transited patients and the organizational issues related to the communication necessities between two different teams of care. Furthermore, the suggested pattern of transition allows to progressively preparing adolescents to the transition of care, effectively dealing with issues of dependence, gradually increasing patients' autonomy, simultaneously preparing the entire family. Eventually, the establishment of ITGs in our center assessed the criticalities related to patients' complexity during childhood.

Declaration of Competing Interest

The authors report no declarations of interest.

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Consensus of the Italian Primary Immunodeficiency Network on transition management from pediatric to adult care in patients affected with childhood-onset inborn errors of immunity

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Medical advances have dramatically improved the long-term prognosis of children and adolescents with inborn errors of immunity (IEIs). Transfer of the medical care of individuals with pediatric IEIs to adult facilities is also a complex task because of the large number of distinct disorders, which requires involvement of patients and both pediatric and adult care providers. To date, there is no consensus on the optimal pathway of the transitional care process and no specific data are available in the literature regarding patients with IEIs. We aimed to develop a consensus statement on the transition process to adult health care services for patients with IEIs. Physicians from major Italian Primary Immunodeficiency Network centers formulated and answered questions after examining the currently published literature on the transition from childhood to adulthood. The authors voted on each recommendation. The most frequent IEIs sharing common main clinical problems requiring full attention during the transitional phase were categorized into different groups of clinically related disorders. For each group of clinically related disorders, physicians from major Italian Primary Immunodeficiency Network institutions focused on selected clinical issues representing the clinical hallmark during early adulthood. (J Allergy Clin Immunol 2020;146:967-83.)

Key words: *Transitional care, inborn errors of immunity, primary immunodeficiency, humoral immune defects, DiGeorge syndrome, combined immunodeficiency, innate immune defects, DNA repair syndromes, Italian Network of Primary Immunodeficiencies*

Inborn errors of immunity (IEIs) are a group of more than 400 rare inherited disorders due to immune system impairment, the overall estimated prevalence of which is approximately 1 in 1000 to 1 in 5000.[1] The development of recommendations for good clinical practice in management of these disorders is hampered by the low number of patients affected with each single disorder. The spectrum of clinical features may be very wide. The issue of how to manage and treat young adults with severe chronic conditions that were often lethal in childhood until a few decades ago is being raised in many clinical settings. IEIs are usually diagnosed in pediatric age. However, thanks to the increased scientific knowledge of these disorders and the recent advances in

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Abbreviations used
AML: Acute myeloid leukemia
A-T: Ataxia-telangiectasia
CGD: Chronic granulomatous disease
CID: Combined immunodeficiency
CMC: Chronic mucocutaneous candidiasis
CN: Congenital neutropenia
CRD: Clinically related disorder
CVID: Common variable immunodeficiency
DGS: DiGeorge syndrome
DKC: Dyskeratosis congenita
DRS: DNA repair syndrome
G-CSF: Granulocyte–colony-stimulating factor
GOF: Gain-of-function
GT: Gene therapy
HIES: Hyper-IgE syndrome
HSC: Hematopoietic stem cell
HSCT: Hematopoietic stem cell transplantation
ID: Intellectual disability
IEI: Inborn error of immunity
IPINet: Italian Network of Primary Immunodeficiencies
IRT: Immunoglobulin replacement therapy
LAD: Leukocyte adhesion defect
LTFU: Long-term follow-up
MRI: Magnetic resonance imaging
NASH: Nonalcoholic steatohepatitis
QoL: Quality of life
SCID: Severe combined immunodeficiency
STAT1: Signal transducer and activator of transcription 1
XLA: X-linked agammaglobulinemia

innovative therapeutic options and medical care, the cohort of patients entering adulthood is growing year by year.[2]

According to Blum et al, transition is defined as "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems."[3] To date, specific literature on IEIs transition of care is not available. Peculiarities of the adolescent affected with IEIs, such as worsening of chronic lung disease, progressive increase in tumor risk, immune dysregulation and related clinical issues, recently identified phenotypes, and psychological issues, make the transitional care for these patients a unique task.

The transitional process to a dedicated adult team is critical to maintaining a high quality of care during long-term follow-up (LTFU). Synergy between the pediatric and adult teams should be ensured during the period of transition to make the process less stressful for the patient and the family.[4,5] A key element in the transition is the training of adult specialists from different medical branches (gastroenterologist, hematologist, pulmonologist, etc) in the peculiarities of patients with IEIs. To ensure the best management, their medical approach should be tailored to the patient's comorbidities, either related to the preexisting IEIs or secondary to therapies. This also requires an in-depth knowledge of the pathogenic mechanism underlying the specific IEI.

The aim of this work is to propose recommendations on transition of care for the main phenotypes of IEIs categorized into different groups of clinically related disorders (CRDs). Each group of CRDs consists of IEIs sharing common main clinical problems that require full attention during the transitional phase. We have focused on the individual peculiarities of each group of CRDs in an attempt to identify the optimal multidisciplinary teams, by analogy with similar disease models, showing partial clinical overlap with IEIs.

METHODS

The participants were physicians of the main clinical Centers of the Italian Network of Primary Immunodeficiencies (IPINet) who are experts in the management of IEIs. IEIs that share common clinical hallmarks requiring careful attention during the transition phase from adolescence to adulthood were grouped as distinct sets of CRDs. Currently, IPINet centers have approximately 3100 patients with IEIs in follow-up. A steering committee was set up to develop a general questionnaire for each group of CRDs, eventually resulting in a variable number of statements about transitional care key points, which have been rated by all the authors. Each statement is based on evidence drawn from (1) studies involving cases and case series of patients with IEIs, (2) management of other rare diseases with clinical similarities to each group of CRDs, (3) rules of clinical good practice derived from expert-based opinions, and (4) review of the literature from databases such as PubMed and Google Scholar. The literature review was carried out by using the following key words: *transition of care* or *transition of management* or *continuity of care* AND *adolescence* or *pediatrics* or *young adults* or *pediatric patient* AND *primary immunodeficiency* or *inborn errors of immunity*. The literature regarding transitional care in various other pediatric disorders sharing similarities with the clinical hallmarks of each group of CRDs was reviewed as well. Recommendations were rated from –1 (total disagreement) to +1 (total agreement). Agreement was defined as the sum of percentages of the ratings of strongly agree (rated +1) and agree (rated +0.5). Disagreement was defined as the sum of the ratings of strongly disagree (rated –1) and agree (rated –0.5). If at least 75% of the raters agreed with the statement and a mean score of 0.75 or higher was reached, that specific recommendation was assumed to have reached consensus. Furthermore, statements that reached a 75% level of agreement and a mean score between 0.65 and 0.74 were assumed to have reached only partial consensus. No consensus was defined as a mean score lower than 0.64 and agreement by less than 75% of the raters. According to Delphi methodology, after the third round of opinions, the consensus was approved.

RESULTS

General principles for transitional care for IEIs have been summarized in Table I. The majority of raters agreed with the statement that transition of care is critical to ensure appropriate LTFU for patients with IEIs (rate of agreement 96%), for the best management of major clinical issues persisting since childhood and those emerging over time (rate of agreement 100%). Regardless of the specific CRD group, the process should include shared patient-centred and family-centred decision making with the multidisciplinary team (rate of agreement 100%). Patients must learn how to cope with the long-term psychological effects of having a chronic disease. Thus, psychological support or stress management should be offered to the patients to help them deal with their condition in different phases of disease and, in particular, during transition (rate of agreement 100%). To facilitate the transitional process, an adolescent with an undiagnosed condition should not leave the pediatric facility without a diagnostic reevaluation (rate of agreement 86%). Transition should start at the age of 14 years and end by the age of 25 years, but no time limit should be mandated (rate of agreement 96%). The accompaniment, defined as a period when the patient is followed-up simultaneously by pediatricians and adult specialists, should last at least 3 years before the final transfer (rate of agreement 96%). Preliminarily (as shown in Table II), the committee has identified 6 groups of CRDs consisting of IEIs sharing common clinical hallmarks that deserve particular attention during transition. A few general statements were common to all CRDs. In Table III, these statements are indicated

TABLE I. Overarching principles for transitional care for patients with IEIs

Overarching principles	Level	Strength	Level of agreement (%)
Transition of care is critical to ensure appropriate long-term follow-up for patients with IEIs	4	D	96
Transition of care is warranted to ensure best management of clinical issues persisting from childhood and those emerging over time	4	D	100
The care of patients with IEIs should include shared patient-centered and family-centered decision making with the multidisciplinary team	4	D	100
In patients with IEIs, psychosocial support is recommended during the transition phase	4	D	100
Adolescents with an undiagnosed condition should not leave the pediatric facility without a diagnostic reevaluation	4	D	86
Transitional care should be planned during adolescence, preferably starting at the age of 14 years and should end by the age of 25 years; however, no time limit should be mandated	4	D	96
The accompaniment should last at least 3 years before the final transfer	4	D	96
Key documentation and records should be included in the transition package	4	D	100

Level of evidence for chronic pediatric diseases sharing similarities with IEIs. The numeral *4* indicates expert opinion; the uppercase *D* indicates maximal strength based on level 4 evidence. Level of agreement is the percentage of experts who agreed on the recommendation during the final voting round of the consensus.

along with the level of consensus for each CRD group. It should be noted that all the experts agreed that the transitional team should be multidisciplinary even though the composition of the team relied on the clinical peculiarities of the CRD.

Common clinical hallmarks of the humoral immune defects group of CRDs during transition

More than half of all patients with an IEI have disorders characterized by a predominant impairment in antibody production. The International Union of Immunological Societies (IUIS) classification of IEIs provided a current overview of this group of diseases.[1] In this consensus article, we have focused on common variable immunodeficiency (CVID) and other B-cell deficiencies impairing 2 or more immunoglobulin classes; we have done so because selective antibody deficiencies, even though more frequent, are usually less severe and only rarely share clinical manifestations with more severe B-cell disorders.

The increased susceptibility to infections, usually from encapsulated bacteria mainly involves respiratory tract, but all organs may potentially be affected (Table II). X-linked agammaglobulinemia (XLA) has been associated with life-threatening infections caused by viruses of the genus *Enterovirus*, thus resulting in neurologic sequelae. A reduced risk of severe infections and an improved long-term quality of life (QoL) is achieved under adequate immunoglobulin replacement therapy (IRT) through intravenous IgG or subcutaneous IgG and antibiotic prophylaxis, even though less severe infections can persist during the follow-up.[6-11] Antibiotic therapy requires an appropriate stewardship, in particular, during adulthood. Live attenuated vaccines are not recommended in patients receiving IRT, whereas inactivated vaccines may be administered because they could elicit T-cell–mediated responses. Yearly influenza vaccination is recommended for patients with B-cell defects and household contacts.[12-15]

Respiratory complications are the lifelong hallmark of this group of CRDs.[10,11] Chronic lung disease has a prevalence higher than 50% in almost all adult age groups with CVID, and it may affect almost half of patients with XLA by their 40s to 50s despite adequate levels of IRT.[8-10,16-18] Granulomatous lymphocytic interstitial lymphocytic disease (GLILD) has a severe impact on the outcome. The extent of lung damage should be evaluated at baseline and during the follow-up through extensive lung functional tests, imaging techniques (such as high-resolution computed tomography and magnetic resonance imaging [MRI]), and other tests, depending on clinical features. Prevention, through early diagnosis and regular monitoring, can reduce the morbidity associated with chronic lung disease. A personalized respiratory physiotherapy program is often required. Gastrointestinal manifestations are among the most frequent clinical problems during LTFU. Adult patients usually report abdominal pain, bloating, nausea, vomiting, diarrhea and weight loss. Affected individuals may also experience impaired ability to absorb nutrients.[19] Moreover, chronic gastrointestinal inflammatory disease is frequent, particularly in CVID in early adulthood. Along with chronic enteropathy, manifestations can also include gastritis and pernicious anemia. A risk of *Helicobacter pylori* infection has also been reported.[20,21] Liver nodular regenerative hyperplasia has been documented in up to 5% of adult patients with CVID,[19,22,23] and it may quickly progress to hepatitis, portal hypertension, and hepatic failure. Thus, it should be managed aggressively: biopsy is often required to assess the inflammatory infiltrate, which can benefit of immunosuppressive treatment.[22]

The prevalence of autoimmunity in patients with CVID varies from 7% to 33% in different studies.[9,24-29] Fischer et al showed that autoimmune manifestations occur throughout the patient's lifetime and that the overall survival time was significantly reduced in patients with autoimmune manifestations.[30] Treatment of noninfectious complications benefits from tailored approaches, including immunosuppressive treatment.

Malignancy is a major cause of early mortality in cohorts of adults with CVID. Up to 20% of adult patients with CVID develop cancer,[21,31-34] including malignancies of lymphatic tissue, such as non-Hodgkin lymphoma and gastrointestinal cancer, which is almost 50 times more frequent than in the healthy population.[21,35-37] Gastrointestinal tract malignancies (eg, gastric adenocarcinoma, liver carcinoma, colon adenocarcinoma) have been reported in patients with XLA.[11,38,39]

Group–specific consensus. As reported in Table IV, an agreement for 6 group-specific statements was reached and assumed as IPINet consensus. Thus, lifelong care of patients with confirmed B-cell defects should be led by a clinical immunologist as the case manager (rate of agreement 96%; score 0.76 ± 0.25). The continuity of IRT adherence and periodic

TABLE II. Common hallmarks of the clinical entities within each group CRDs during adolescence and transitional age

CRD	Main disorders included	Hallmarks	Main issues
Humoral defects	XLA or AR-agammaglobulinemia, CVID	Susceptibility to infections	Encapsulated bacteria
		Chronic lung disease	Bronchiectasis, GLILD, otitis, sinusopathy
		Chronic gastrointestinal involvement	Chronic enteropathy, malnutrition, primary biliary cirrhosis, hepatitis, and nodular regenerative hyperplasia, *Giardia lamblia* and/or bacterial or viral recurrent infections
		Long-lasting autoimmunity and autoinflammation	Thrombocytopenia, hemolytic anemia, arthritis, IBD, granuloma
		Lymphoproliferative disorders and malignancy	Lymphadenopathy, splenomegaly, lymphoma, solid cancer
		Susceptibility to infections	Osteomyelitis, meningitis
DGS spectrum	22q11.2 deletion syndrome, DiGeorge-like syndromes	Congenital and acquired cardiovascular anomalies	Postsurgery lifelong FUP, bacterial endocarditis, arrhythmia, ventricular dysfunction, and aortic root dilatation
		Neuropsychiatric disorders	Neurodevelopment decline, psychotic disorder, schizophrenia, anxiety, autism spectrum disorder, ADHD, depressive and mood disorders, brain abnormalities
		Immunologic features	Susceptibility to infections
			Autoimmune cytopenia and cancer susceptibility
		Endocrine aspects	Hypoparathyroidism/hypocalcemia, hypothyroidism, hyperthyroidism, obesity
		Orthopedic alterations	Cervical spine anomalies, severe scoliosis
		Genetic counseling	Reproductive genetic counseling and contraception
		Genitourinary anomalies	Unilateral renal agenesis, multicystic dysplastic kidney
CID	CID, late-onset CID and untreated SCID, syndromic CID, *CD40*, *CD40L* deficiency	Immunologic features	Progressive lymphopenia, severe/ atypical infections
		Immune dysregulation and autoimmunity	Cytopenia, vasculitis, HLH, granulomata, IBD, diabetes, thyroiditis, neuropathy
		Dermatologic issues	Severe eczema, CMC, warts
		Lymphoproliferative disorders and malignancy	Lymphoma, EBV-related lymphoproliferation, solid cancer
		Extraimmunologic manifestations	Involvement of several organs and systems
		Drug-related side effects	Drug toxicity, antibiotic resistance
Innate immunity defects	CGD, SCN, LAD, CMC, HIES	Susceptibility to infection	Fungi, *Staphylococcus aureus, Serratia marcescens, Nocardia* spp*, Salmonella*, and bacillus Calmette-Guérin infections
		Inflammation and autoimmunity	CGD colitis, systemic autoimmune disorders
		Malignancy	Hematopoietic and solid cancer
		Extraimmunologic manifestations	ID, growth retardation, skeletal and skin defects, albinism, metabolic diseases, vascular abnormalities
DRSs, Inborn errors with malignancies	A-T, A-T–like disorders, NBS, BS	Neurologic features	Progressive neurologic degeneration, extrapyramidal involvement
	DKC, telomeropathies	Immunologic features	T- and B-lymphocyte deficiency, low Ig levels, susceptibility to infections
		Respiratory manifestations	Chronic bronchopneumopathy, ILD, pharyngeal incoordination, and respiratory muscles insufficiency
		Nutritional problems	Malnutrition, metabolic disorder
		Endocrine system and metabolic status	IGF-1 deficiency; pubertal delay; gonadal, thyroid, and adrenal gland dysfunction; insulin resistance; NASH; cardiovascular risk
		Immune dysregulation and cancer	Granuloma, lymphoid and solid tumors
		Hematologic features and malignancy	Bone marrow failure and cancer susceptibility
		Skin and annexa features	Oral leukoplakia, dystrophic nails, and reticular skin pigmentation
		Premature aging	Graying hair, liver fibrosis, portal hypertension, osteopenia

(Continued)

TABLE II. *(Continued)*

CRD	Main disorders included	Hallmarks	Main issues
HSCT or GT	Severe forms of IEIs (SCID, IPEX, CGD, HLH, WAS)	End-organ damage Long-term side effects of conditioning regimen Uncorrected disease manifestations Incomplete immune reconstitution Secondary cancer Chronic graft-versus-host disease Genetic counseling and sterility treatment	Infectious and noninfectious sequelae Cataract, endocrine dysfunction, liver and lung disease, metabolic syndrome Extraimmunologic manifestations Recurrent infections, autoimmune manifestations, immunoglobulin substitution, antimicrobial prophylaxis, immunosuppression Intrafamilial recurrence, drug-related sterility

AR, Autosomal recessive; *BS*, Bloom syndrome; *CMC,* chronic mucocutaneous candidiasis; *DKC*, dyskeratosis congenita; *FUP,* follow-up; *GLILD*, granulomatous lymphocytic interstitial lymphocytic disease; *HLH*, hemophagocytic lymphohistiocytosis; *IBD*, inflammatory bowel disease; IGF-1, insulin-like growth factor-1; *ILD*, interstitial lung disease; *IPEX*, immunodysregulation polyendocrinopathy enteropathy X-linked; *LAD,* leukocyte adhesion defect; *NASH*, nonalcoholic steatohepatitis; *NB*S, Nijmegen breakage syndrome; *SCN*, severe congenital neutropenia; *WAS*, Wiskott-Aldrich syndrome.

TABLE III. General statements for all CRDs with or without consensus

	Level of agreement (%) (mean score ± SD)					
Statement	Humoral immune defects	DGS	CID or untreated SCID	Innate immune disorders	DRSs and IEIs with malignancies	HSCT or GT
How should the transitional team be composed?						
A pediatric immunologist and an adult immunologist in a multidisciplinary setting, as required	92* (0.76 ± 0.35)	100† (0.81 ± 0.24)	100‡ (0.87 ± 0.22)	100§ (0.87 ± 0.22)	100‖ (0.83 ± 0.24)	96¶ (0.73 ± 0.25)
What are the process outcomes?						
Patient and family satisfaction	92 (0.75 ± 0.35)	92 (0.75 ± 0.35)	88 (0.68 ± 0.35)	74 (0.69 ± 0.37)	92 (0.76 ± 0.25)	96 (0.58 ± 0.30)
Adherence to therapies	92 (0.88 ± 0.21)	96 (0.73 ± 0.25)	92 (0.70 ± 0.25)	77 (0.69 ± 0.37)	76 (0.56 ± 0.47)	NA
Adherence to follow-up	92 (0.90 ± 0.20)	96 (0.73 ± 0.25)	92 (0.57 ± 0.38)	81 (0.67 ± 0.35)	89 (0.68 ± 0.35)	100 (0.73 ± 0.25)
Patient and family empowerment	96 (0.66 ± 0.24)	96 (0.84 ± 0.23)	NA	77 (0.78 ± 0.25)	89 (0.77 ± 0.25)	96 (0.68 ± 0.24)
Disease activity status#	NA	96 (0.80 ± 0.24)	96 (0.80 ± 0.24)	78 (0.68 ± 0.36)	92 (0.76 ± 0.25)	100 (0.69 ± 0.24)

NA, Not applicable.

Level of agreement is the sum of the percentage of strong agreement (rated +1) and level of agreement (rated +0.5); consensus is defined as a 75% or higher rate of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher rate of agreement and a mean ranging from 0.65 to 0.74.

*Pediatric and adult nurse, family pediatrician, general practitioner, gastroenterologist, hematologist, pulmonologist, and psychologist.

†Psychiatrist, neurologist, endocrinologist, cardiologist, hematologist, and psychologist.

‡Infectiologist, endocrinologist, gastroenterologist, nutritionist, pulmonologist, gynecologist, dermatologist, and psychologist.

§Pulmonologist, gastroenterologist, cardiologist, dermatologist, oncologist, and orthopedist.

‖Neurologist, pulmonologist, gastroenterologist, cardiologist, dermatologist, oncologist, orthopedist, and psychologist.

¶Medical professional experienced in HSCT for adult patients with IEIs.

#Infection rate, admissions in emergency unit, extraimmunologic. complications, including neurologic, endocrine, malnutrition, abnormal liver and heart function, and joint deformities.

monitoring of serum IgG levels should be assessed (rate of agreement 92%; score 0.84 ± 0.33), making the patient aware of the potential complications of not adhering to IRT and for better self-regulation. The adherence to lung physiotherapy (rate of agreement 92%; score 0.88 ± 0.21) should be periodically evaluated, and monitoring of hematologic (eg, malignancy and cytopenia), gastrointestinal (eg, granulomatous diseases), and pulmonary complications must be ensured (rate of agreement 100%; score 0.94 ± 0.16). An adapted vaccination course should also be planned during adulthood in consideration of IRT (partial consensus; rate of agreement 96%; score 0.69 ± 0.34), and an annual complete follow-up should be guaranteed (partial consensus; score 0.72 ± 0.25). There was no consensus on the contraindications to starting the transition.

Common clinical hallmarks of the DGS group of CRDs during transition

22q11.2 deletion syndrome is the most frequent chromosomal microdeletion disorder underlying DiGeorge syndrome (DGS), occurring in up to 1 in 4000 live births.[40] In a few cases, cytogenetic abnormalities other than 22q11.2 or *TBX1* haploinsufficiency have been found in patients with a DGS clinical phenotype, including velocardiofacial syndrome or conotruncal anomaly face syndrome. Clinical features deserving maximal attention during adolescence and adulthood may be variable among different subjects.[40] The multitude of clinical manifestations associated with the syndrome and the high variability of the clinical phenotype complicate the transition of care in this syndrome.[40] It should be noted that in addition to the predominant

TABLE IV. Recommendation on transitional care for patients with humoral immune defects

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
No consensus			
Unstable clinical condition (eg, autoimmune flares, acute infection)	74	26	0.46 ± 0.66
Who should be the case manager within the adult team?			
Consensus			
Clinical immunologist	96	4	0.76 ± 0.25
No consensus			
Pulmonologist	29	71	–0.06 ± 0.61
Internist	89	11	0.62 ± 0.41
Any health care professional who assumes full responsibility for care coordination and planning	44	56	–0.01 ± 0.61
What are the peculiarities to focus on in this specific CRD?			
Consensus			
To ensure continuity of IRT adherence and monitoring	92	8	0.84 ± 0.33
To evaluate periodically adherence to lung physiotherapy	92	8	0.88 ± 0.21
To focus on hematologic, gastrointestinal, and lung complications	100	0	0.94 ± 0.16
Partial consensus			
To sensitize and monitor an adapted vaccination course	96	4	0.69 ± 0.34
To guarantee annual complete follow-up	89	11	0.72 ± 0.25

Level of agreement is the sum of the percentage of strong agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement is the sum of percentage of strong disagreement (rated –1) and percentage of disagreement (rated –0.5). Consensus is defined as a 75% or higher level of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher level of agreement and a mean score ranging from 0.65 to 0.74.

features summarized in Table II, the clinical phenotype may also encompass other manifestations, including dysmorphic features, which profoundly affect the self-confidence and social life, thus requiring special attention during adolescence.[27,41] A successful transfer to adult services is a very complicated process, especially in those patients with various organ and intellectual or developmental disabilities. Practical guidelines for managing pediatric and adult patients with 22q11.2 deletion syndrome have been proposed.

The specificity and critical issues of transition of care in this condition are summarized in the Table II.

Of note, several congenital heart disorders require lesion-specific management and lifetime surveillance, even after corrective surgery.[27,42] Furthermore, patients with DGS may develop cardiac manifestations during the follow-up as a result of comorbidities such as chronic kidney disease, leading to hypertension, endocrine dysfunction (parathyroid or thyroid disorders) causing arrhythmias, low physical activity, and side effects of antipsychotic therapies resulting in metabolic syndrome.[42]

As for neuropsychiatric disorders, intellectual disability (ID) and subsequent impaired social processing are common in DGS, with variable degrees of severity. A gradual decline in neurodevelopment throughout the lifespan has been reported, and a link between ID and psychiatric disease has been hypothesized.[27,43-45] Overall, the neuropsychiatric phenotype may evolve over time, with earlier symptoms persisting or being replaced by others with the transition from childhood to adulthood, thus entailing a strong need for continuous follow-up. Autism spectrum disorder and attention-deficit/hyperactivity disorder emerge during childhood and can have an impact on adult life as an independent issue. There is no apparent correlation between these disorders and the subsequent development of schizophrenia.[46] Interestingly, risk factors for the development of schizophrenia include the aforementioned early cognitive decline, social and executive dysfunction, and depressive and anxiety disorders,[47] which are common in adult patients. Among the other factors that may affect neuropsychiatric outcome and its interindividual variability are lower parental socioeconomic status and intrusive parenting style, which have been shown to be related to worse social functioning.[48] Special attention should also be placed on environmental demands: tracing a thorough outline of the individual's intellectual and psychiatric phenotype is mandatory to plan the required social adaptations to avoid the stress deriving from a mismatch.[27,47] Ear and palatal anomalies may be associated with hearing loss (found in 30%-40% of patients) and speech disorders,[40] which may often interfere with social functioning.[27,49]

Immune alterations may be very variable, ranging from partial DGS (characterized by normal or mild reduction of T-cell number and function) to complete DGS, in which the T-cell defect is more profound, resembling a severe combined immunodeficiency (SCID)-like phenotype associated with atypical infections.[50-52] Velopharyngeal insufficiency, gastroesophageal reflux, and asthma/rhinitis also may contribute to susceptibility to infection.[53] The incidence of infections tends to decrease with age. However, a few patients may require that prophylactic treatment with broad-spectrum antibiotics and, rarely, intravenous IgG or subcutaneous IgG also be continued in adulthood.[50] In this scenario, immunoglobulin levels, T-cell counts, and vaccine-specific antibody titers should be controlled over time, along with the frequency of infections. Counts of T cells, which show a senescent and memory phenotype, tend to increase with age.[54,55] Patients with DGS may also develop autoimmunity and immune dysregulation later in life, especially autoimmune cytopenias.[56] Notably, peculiar immunophenotypic alterations, such as decreased numbers of naive $CD4^+$ cells and class-switched B cells, have been identified early after diagnosis of DGS in patients who developed such complications; therefore, closer monitoring is warranted for these subgroups of patients.[56]

A higher overall rate of malignancies (eg, thyroid cancer, leukemia, lymphoma) has also been reported.[57]

Genetic counseling. Reproductive genetic counseling should be offered to patients, starting from adolescence and continuing throughout adulthood. Future parents should be informed about recurrence risk, even in the absence of causative deletion in their genotype, because of the potential germline mosaicism, and they should also be informed about broad interfamilial and intrafamilial variability in clinical phenotype.[40,49,50] They should be offered diagnostic testing by chorionic villus sampling or amniocentesis.[53] Gynecologic counseling during adolescence should be offered to prevent unplanned pregnancies.

Group-specific consensus. As reported in Table V, agreement regarding 6 group-specific statements was reached and assumed as IPINet consensus. Because of the wide spectrum of manifestations of this group of diseases, the case manager within the adult team should be identified with consideration for the prominent phenotypic feature of each patient (rate of agreement 96%; score 0.78 ± 0.25). Particular attention should be paid to cardiovascular (rate of agreement 100%; score 0.76 ± 0.25), neuropsychiatric (rate of agreement 96%; score 0.81 ± 0.34), and orthopedic (rate of agreement 96%; score 0.75 ± 0.25) issues, and genetic counseling (rate of agreement 96; score 0.78 ± 0.25) should be planned. Even though immunodeficiency may be more attenuated in adult patients, the raters agreed that attention should be paid to immune-related manifestations (partial consensus; rate of agreement 96%; score 0.70 ± 0.34).

In contrast, there was no consensus on conditions that contraindicate the transition. In all, 89% of raters agreed on the need to identify subspecialists who must be trained on the specificity of the peculiar features in the DiGeorge spectrum syndrome; however, a definitive consensus on this issue was not reached (score 0.62 ± 0.33). Furthermore, the majority of raters (37% and 45%, respectively) believed that a diagnosis close to the age of transition or the presence of smaller affected siblings may be not a significant obstacle to transition.

Common clinical hallmarks of the CID and untreated SCID group of CRDs during transition

SCIDs and late-onset combined immunodeficiencies are a group of heterogeneous genetic disorders characterized by severe recurrent infections, and impaired cellular and humoral functionality.[58,59] Because most patients with SCIDs are successfully treated with hematopoietic stem cell transplantation (HSCT), the transition of care is guided mainly by hematologists with expertise in HSCT who ensure LTFU of HSCT survivors. On the other hand, patients with milder phenotypes characterized by a late onset of the disease need to be closely evaluated for progression of the disease, including lymphopenia, and possible complications. Within this group, patients with profound combined immunodeficiency (CID) associated with severe infections and/or immune dysregulation often have an uncertain indication for HSCT, and its timing is debated on account of transplantation risks.[60] Moreover, patients who carry hypomorphic mutations of known SCID-causing genes can be diagnosed in adolescence or adulthood when they have a milder clinical presentation or are even clinically asymptomatic.[61] In such cases a potential progressive immunologic deterioration may be expected.[62-64] The specificity and critical issues of transition of care of patients with this condition are summarized in Table II.

Increased susceptibility to bacterial, fungal, or viral infections frequently results in chronic obstructive lung disease and/or bronchiectasis and chronic organs damage.[58,59] An atypical course of some infections (such as *Helicobacter* bacteremia, adenovirus/cytomegalovirus, EBV infections and, rarely, mycobacteriosis), chronic candidiasis, warts caused by human papilloma virus, or recurrent molluscum contagiosum, is also frequent in these patients. Moreover, patients undergoing lifelong prophylactic treatments should be regularly checked for the occurrence of drug-related side effects.

In some patients, the phenotype is predominated by immune dysregulation and autoimmunity, which can be triggered by infections. The prototype of the late-onset subgroup is recombination-activating gene hypomorphic deficiency, in which autoimmunity and hyperinflammation are common.[60,65] These patients should be regularly checked for the occurrence of EBV-related lymphoma or Hodgkin-like features. Uncontrolled hyperinflammation, as life-threatening hemophagocytic lymphohistiocytosis, may occur in all forms when proper clearance of an infectious agent fails to be achieved. Attention should also be paid to the transitional care of patients affected with radiosensitive disorders due to defects of nonhomologous end joining factors such as DNA ligase IV, which are associated with increased risk of developing leukemia and other lymphoproliferative disorders.[66]

CIDs with extraimmunologic manifestations (CIDs with associated or syndromic features), according to the phenotypic classification by IUIS, were also included within this group.[1] Several organs and systems may be involved, displaying microcephaly, dysmorphic facies, and developmental delay or skeletal, endocrine, and hematologic (eg, microthrombocytopenia) abnormalities. Malabsorption is a frequent issue.

CRD group–specific consensus. After a literature review and scoring, IPINet consensus was reached concerning a number of critical issues regarding transitional care for patients with CIDs and untreated patients with the clinical spectrum of SCIDs (Table VI). For patients with very rare untreated SCIDs, transition should be avoided if the adult center is not fully equipped to handle the potential severe progression and frequent clinical instability of these patients (rate of agreement 96%; score 0.75 ± 0.35), and transition should be postponed if the patient is critically ill (partial consensus; rate of agreement 100%; score 0.66 ± 0.33). The case manager within the adult team should preferentially be the clinical immunologist (rate of agreement 92%; score 0.78 ± 0.35). Attention should be paid to management of chronic end-organ damage and several extraimmunologic manifestations (rate of agreement 100%; score 0.83 ± 0.24), and new disease therapeutic strategies (rate of agreement 100%; score 0.86 ± 0.22) and indications for HSCT (rate of agreement 100%; score 0.81 ± 0.24) should be periodically discussed. Multidisciplinary discussion of diagnosis and further tests to improve the work-up of undiagnosed patients should be encouraged (rate of agreement 100%; score 0.81 ± 0.24), and drug-related side effects should be evaluated (rate of agreement 100%; score 0.79 ± 0.35). Even though high-efficiency particle air filtration rooms or laminar flow units are in general not mandatory, the adult center should ensure immediate medical care when necessary and the patient should be admitted to a single room (rate of agreement 96%; score 0.79 ± 0.34). In addition, monitoring of the progression of immunologic deterioration (partial consensus; rate of agreement 96%; score 0.72 ± 0.36), end-stage disease (partial consensus; rate of agreement 96%; score 0.68 ± 0.34), endocrine complications

TABLE V. Recommendation on transitional care for patients with DiGeorge spectrum syndrome

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
No consensus			
Lack of subspecialist experts in the specific aspects of the disease	89	11	0.62 ± 0.33
Recent diagnosis close to the age of transition	63	37	0.28 ± 0.61
Younger siblings affected	55	45	0.13 ± 0.62
Who should be the case manager within the adult team?			
Consensus			
The choice should take into account the prominent phenotypic feature	96	4	0.78 ± 0.25
No consensus			
Clinical immunologist	81	19	0.62 ± 0.48
Geneticist	51	49	0.09 ± 0.63
Internist	89	11	0.58 ± 0.40
Any health care professional who assumes full responsibility for care coordination and planning	74	26	0.35 ± 0.64
What are the peculiarities to focus on in this specific CRD?			
Consensus			
To manage cardiovascular anomalies, including congenital and acquired conditions (eg, congenital heart disorders, bacterial endocarditis, rhythm disturbances, ventricular dysfunction and aortic root dilatation, metabolic syndrome)	100	0	0.76 ± 0.25
To monitor the evolution of neuropsychiatric disorders	96	4	0.81 ± 0.34
To evaluate the progression of orthopedic impairment (ie, scoliosis, club foot)	96	4	0.75 ± 0.25
Genetic counseling	96	4	0.78 ± 0.25
Partial consensus			
To evaluate immune functionality and immune-related manifestations	96	4	0.70 ± 0.34
No consensus			
To guarantee complete follow-up biannually	76	24	0.45 ± 0.55

Level of agreement is the sum of the percentage of strong agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement is the sum of the percentage of strong disagreement (rated –1) and the percentage of disagreement (rated –0.5). Consensus is defined as a 75% of higher level of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher level of agreement and a mean score ranging from 0.65 to 0.74.

(partial consensus; rate of agreement 100%; score 0.72 ± 0.25), and QoL (partial consensus; rate of agreement 96%; score 0.66 ± 0.33) should be performed.

No consensus was reached regarding the follow-up intervals that should be adapted for each subject or regarding nutritional issues.

Common clinical hallmarks of the innate and intrinsic immunity defects group of CRDs during transition

Defects in innate and intrinsic immunity encompass a heterogeneous group of inherited diseases associated with invasive, life-threatening infections characterized mainly by increased susceptibility to a single or narrow group of microorganisms and immune dysregulation leading to autoimmune and autoinflammatory disorders**.** Within this CRD group, particular attention in the transitional care process must be paid to congenital defects of phagocyte function and number, such as chronic granulomatous disease (CGD), leukocyte adhesion deficiency (LAD), congenital neutropenias (CNs), chronic mucocutaneous candidiasis (CMC) due to signal transducer and activator of transcription 1 (*STAT1*) gain-of-function (GOF) mutations, and hyper-IgE syndrome (HIES).[1,67] All these disorders, although profoundly different in their pathogenic mechanism, share similarities in the major clinical problems that may characterize patients during adolescence and early adulthood.

The main common clinical hallmark of patients with innate and intrinsic immunity includes an increased risk of bacterial and fungal infections. In particular, patients affected with CGD, or with CNs, may develop severe recurrent infections mainly due to *Aspergillus* spp and *Staphylococcus aureus*.[68-70] A recent study documented that one-third of all infectious events occurred in patients with CGD after the age of 16 years. Similarly, noninfectious granuloma or inflammatory granulomatous bowel disease in patients with CGD occurred during the same period in 46% of patients. The prognosis of CGD has greatly improved since it was first described thanks to earlier diagnosis, better management of infectious and inflammatory complications, antibacterial and antifungal prophylaxis, and good outcome after HSCT.[71,72] The aging process of the population of individuals with CGD, as a paradigm of the whole CRD group, poses new challenging problems because of the lifetime recurrence of pulmonary manifestations and sequelae; growth failure; and noninfectious liver disease, including toxic drug–induced hepatitis, nodular regenerative hyperplasia, noncirrhotic portal hypertension, and several autoimmune manifestations. Although the frequency of inflammatory episodes tends to increase slightly after the age of 16 years, infectious events tend to decrease over time.[73] Thus, although infections remain the first cause of death, inflammatory complications, mainly pulmonary or digestive, seem to predominate during adulthood. For patients with CN, the availability of granulocyte–colony-stimulating factor (G-CSF) therapy drastically changed the QoL and their overall survival is now estimated at approximately 80%. However, about 10% of patients (mainly G-CSF nonresponders) still die from severe bacterial infections.[74] *STAT1* GOF mutation accounts for half of the cases of CMC. Affected patients are at risk of invasive candidiasis (sepsis,

TABLE VI. Recommendation on transitional care for patients with CID or untreated SCID

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
Consensus			
For untreated patients with SCID, transition may be not feasible if the adult center is not fully equipped	96	4	0.75 ± 0.35
Partial consensus			
Critical illness (eg, active opportunistic infections or severe invasive/systemic infections or uncontrolled immune dysregulation manifestations)	100	0	0.66 ± 0.33
Who should be the case manager within the adult team?			
Consensus			
Clinical immunologist	92	8	0.78 ± 0.35
No consensus			
Hematologist	63	37	0.23 ± 0.57
Pulmonologist	19	81	–0.38 ± 0.41
Internist	78	22	0.42 ± 0.51
Any health care professional who assumes full responsibility for care coordination and planning	41	59	–0.12 ± 0.54
What are the peculiarities to focus on in this specific CRD?			
Consensus			
To manage each chronic end-organ damage	100	0	0.83 ± 0.24
To manage several extraimmunologic features	100	0	0.83 ± 0.24
To periodically discuss new disease therapeutic strategies	100	0	0.86 ± 0.22
Periodic multidisciplinary discussion of diagnosis and further tests to improve the work-up in undiagnosed patients	100	0	0.81 ± 0.24
Periodic discussion of indication to HSCT (especially in late-onset and evolving diseases)	100	0	0.81 ± 0.24
To monitor drug-related side effects	100	0	0.79 ± 0.35
The adult center should ensure immediate medical care when necessary and isolation of the patient in a single room	96	4	0.79 ± 0.34
Partial consensus			
To monitor the progression of functional immunologic deterioration	96	4	0.72 ± 0.36
To manage the progression to end-stage disease	96	4	0.68 ± 0.34
To monitor endocrine complications (primary or secondary to therapies)	100	0	0.72 ± 0.25
To evaluate QoL index	96	4	0.66 ± 0.33
No consensus			
To guarantee complete follow-up biannually	92	8	0.57 ± 0.38
To provide exclusive or partial enteral nutrition	92	8	0.62 ± 0.40

Level of agreement is the sum of the percentage of strong agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement is the sum of the percentage of strong disagreement (rated –1) and the percentage of disagreement (rated –0.5). Consensus is defined as a 75% or higher rate of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher rate of agreement and a mean ranging from 0.65 to 0.74.

nephritis), or other fungal infections, which may manifest in adulthood also.[75,76] As CMC may be the cause of impairment in activities of daily living or social barriers, long-term prophylaxis should be continued for these patients. Autoimmune manifestations, including thyroiditis, type 1 diabetes, cytopenia, and enteropathy, may occur in the whole CRD group during the second decade of life.[77]

The risk of malignancies in CMC, HIES, and CN is high. Most patients with CN are at increased risk of progression to myelodysplasia and acute myeloid leukemia (AML). The cumulative incidence of myelodysplasia and/or AML is estimated at 11% at 20 years of age and at 22% after 15 years of G-CSF therapy. Although it has been clearly demonstrated that G-CSF dramatically improves survival of patients with CN, it is also likely that G-CSF itself affects clonal evolution. The risk of developing AML or myelodysplasia varies considerably across the spectrum of genetic etiologies: by 30 years of age, this rate is estimated to be roughly 60% in patients with *GATA2* mutations, 30% in patients with *SBDS* mutations, and 15% in patients with *ELANE* mutations. Chromosomal abnormalities, including monosomy 7 and gain of chromosome 21, as well as additional acquired somatic mutations in *CSF3R* and *RUNX1* genes, are frequently detected before AML transformation and should be checked.[61] Oral squamous cell carcinoma and esophageal cancer constitute the most common cause of death due to malignancy in patients with *STAT1* GOF mutations. Other cancers have also been reported in affected patients: cutaneous, gastrointestinal, or laryngeal carcinoma; melanoma; or even leukemia. About 7% of patients with HIES also develop malignancies, especially various types of lymphoma.[78]

As for extraimmunologic manifestations, severe mental and growth retardation in late childhood as a consequence of defect in fucose metabolism have been reported in patients with LAD-2. The risk of bleeding is also an important issue in LAD-3 on account of mutation in the kindlin-3 gene, affecting the integrin activation cascade. CNs are sometimes associated with a multiplicity of syndromic features that may include oculocutaneous albinism, metabolic diseases, and bone marrow failure syndromes. When compared with children without CGD, children with CGD exhibit higher rates of difficulty acquiring social and/or school skills, difficulty establishing peer relationships, and conduct and/or emotional problems.[79] In contrast, adults with CGD report greater

TABLE VII. Recommendation on transitional care for patients with innate immune disorders

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
Consensus			
Critical illness (eg, active or severe invasive/systemic infections, uncontrolled inflammatory/autoimmune manifestations)	92	8	0.76 ± 0.42
Recent onset or treatment of myelodysplasia or leukemia or HSCT	92	8	0.76 ± 0.42
Who should be the case manager within the adult team?			
Consensus			
Clinical immunologist	96	4	0.82 ± 0.24
Partial consensus			
Internist	92	8	0.70 ± 0.42
No consensus			
Pulmonologist	34	66	–0.23 ± 0.45
Infectiologist	70	30	0.23 ± 0.51
What are the peculiarities to focus on in this specific CRD?			
Consensus			
To monitor severe and recurrent infections	100	0	0.82 ± 0.24
To monitor recurrence of infections due to a single group of microorganisms	89	11	0.85 ± 0.23
To manage accelerated progression of common infections	89	11	0.87 ± 0.22
To manage inflammatory and autoimmune diseases	96	4	0.84 ± 0.23
To manage progressive and chronic end-organ damage	100	0	0.87 ± 0.22
To manage extraimmunologic condition	92	8	0.76 ± 0.25
Malignancy surveillance	96	4	0.78 ± 0.25
To monitor drug-related side effects/multidrug resistance	81	19	0.84 ± 0.23
To evaluate new disease therapeutic strategies	81	19	0.81 ± 0.24
To manage pregnancy and genetic counseling	77	23	0.78 ± 0.25
To evaluate QoL index	81	19	0.81 ± 0.24
Partial consensus			
To manage malnutrition and growth failure and/or pubertal delay	85	15	0.72 ± 0.36

Level of agreement is the sum of the percentage of strong agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement in the sum of the percentage of strong disagreement (rated –1) and the percentage of disagreement (rated –0.5). Consensus is defined as a 75% or higher level of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher level of agreement and a mean score ranging from 0.65 to 0.74.

difficulties in either mental and physical area than adults without CGD do. Clinical status may affect psychological and school dimensions and social networking.[59] Inflammatory symptomatic cerebral and extracerebral aneurysms and aortic calcification have been detected at a higher rate than in the general population and at a younger age (23 vs 50 years) in patients with *STAT1* GOF mutation syndrome than in those without such mutation.[80,81] Cumulative survival rate was significantly lower in patients who developed invasive infections, cancer, and/or symptomatic aneurysms.[77] In patients with HIES, facial abnormalities, retention of the primary teeth, skeletal abnormalities such as osteopenia resulting in multiple fractures for minor traumas and scoliosis, joint hyperextensibility, various vascular malformations, and chronic severe eczema have been described.[52,78,82]

Several genetic defects associated with susceptibility to specific pathogens or a narrow group of pathogens (mycobacteria, pyogenes, human papilloma virus, herpes simplex virus, and other viruses; *Candida* spp, and other fungi) have been recently described, thus requiring a personalized approach.

CRD group–specific consensus. After a literature review and scoring, IPINet consensus was reached concerning a number of critical issues on transitional care for innate and intrinsic immunity defects (Table VII). Active or severe invasive and/or systemic infections, uncontrolled inflammatory or autoimmune manifestations, recent onset or treatment of myelodysplasia or leukemia, or HSCT might contraindicate the transition process (rate of agreement for both 92%; score 0.76 ± 0.42). The case manager within the adult team should be the clinical immunologist (rate of agreement 96%; score 0.82 ± 0.24); alternatively, the internist should be considered (partial consensus; rate of agreement 92%; score; 0.70 ± 0.42). To manage inflammatory, autoimmune (rate of agreement 96%; score 0.84 ± 0.23), chronic end-organ damage (rate of agreement 100%; score 0.87 ± 0.22), and extraimmunologic manifestations (rate of agreement 92%; score 0.76 ± 0.25), including the risk of aneurysm in *STAT1* GOF mutations; special attention should be paid to monitoring recurrence and severity of infections (rate of agreement 100%; score 0.82 ± 0.24), especially those due to a single group of microorganisms (rate of agreement 89%; score 0.85 ± 0.23) or with accelerated progression (rate of agreement 89%; score 0.87 ± 0.22). Malignancy surveillance, including cytogenetic studies, is mandatory in several forms (rate of agreement 96%; score 0.78 ± 0.25). Monitoring of drug-related side effects, multidrug resistance (rate of agreement 81%; score 0.84 ± 0.23), and QoL should be carried out (rate of agreement 81%; score 0.81 ± 0.24). New disease therapeutic strategies should be discussed periodically (rate of agreement 81%; score 0.81 ± 0.24), and pregnancy management and counseling should be offered (rate of agreement 77%; score 0.78 ± 0.25). Attention should be paid to malnutrition risk and to the growth failure and/or pubertal

delay (partial consensus; rate of agreement 85%; score 0.72 ± 0.36).

Common clinical hallmarks of the DNA repair syndromes and IEIs with malignancy susceptibility group of CRDs

DNA repair syndromes (DRSs) are a heterogeneous group of conditions characterized by defects in the process of DNA damage repair and, clinically, mainly by neurologic, immunologic, and systemic involvement. The CRD group includes ataxia-telangiectasia (A-T) and AT-like disorders, Nijmegen breakage syndrome, Bloom syndrome, and other rarer disorders.[24] Other IEIs associated with cancer susceptibility include dyskeratosis congenita, an inherited bone marrow failure syndrome that is typically characterized by skin and annexa features, as illustrated in Table II. To date, several genetic alterations may modify telomere maintenance and function.[83] Bone marrow failure is the primary cause of death in these patients, and therefore, HSCT is the only curative treatment.[84] However, HSCT cannot correct other systemic manifestations of defects of telomere maintenance.[85] In rarer telomeropathies, premature aging, liver fibrosis, and predisposition to myelodysplastic syndrome and myeloid leukemia are reported. All these conditions share a poor prognosis as a result of development of serious complications such as malignancies and bone marrow failure.

A-T (Online Mendelian Inheritance in Man no. 208900), although rare, is the most frequent DRS and will be used as a model for management and surveillance for the whole group; the remaining DRSs, share similar management protocols with A-T.[86] The main causes of death in A-T are respiratory infections, leading to progressive respiratory failure, and cancer.[87] The multiple and different functions played by Ataxia telangiectasia mutated (ATM) protein explain well the wide variety of clinical manifestations in individuals with A-T and the severity of their clinical phenotype (classic vs mild).[86] Affected individuals with classic A-T usually survive until the second or third decade of life, whereas individuals with milder cases can reach their 60s.[24,88]

The improvement in the quality of care has changed the natural history of A-T by lengthening life expectancy and revealing new clinical manifestations typically associated with young adulthood.[89,90] Given the complexity of these heterogeneous conditions, transition of care must be tailored to the individual patient.

A-T is characterized by progressive cerebellar degeneration, extrapyramidal disorders, and oculomotor apraxia.[91,92] Progressive motor dysfunction is responsible for postural anomalies, dysarthria, respiratory failure, and joint contractures that worsen the nutritional aspect. These manifestations are variably represented in the young adult with A-T, who invariably also experiences loss of gait and significant movement disorders.[93] Regarding the neurologic manifestations, which are only transiently controlled by therapies,[12,94,95] adult neurologists (mainly experts in movement disorders), physiatrists, psychologists, and therapists with expertise in motor disability, respiratory, speech, and occupational rehabilitation are required.[24,89]

About two-thirds of patients with A-T have an immune defect with a reduction in T- and B-lymphocyte counts that is often associated with deficiency of 1 or more immunoglobulin classes. About 10% of patients show hyper-IgM syndrome, which together with the IgG2 deficiency, is associated with a worse clinical course. Immunodeficiency leads to increased susceptibility to infections, which affect the lung almost exclusively.[90] Under certain circumstances, antibiotic prophylaxis, IRT, and vaccinations have proved effective in the prevention of infectious diseases.[12,52] HSCT has been used in very few patients, but the results are not encouraging.[2]

Respiratory disease progresses with age and includes (1) respiratory chronic disease due to recurrent infections, (2) interstitial lung diseases, and (3) respiratory manifestations due to the neurologic dysfunction.[87,96] Optimal multispecialist respiratory management of young adult patients is necessary owing to the recurrence of respiratory manifestations, pharyngeal incoordination, and insufficiency of the respiratory muscles. Specific guidelines for antibiotic prophylaxis in A-T are not available; however, indirect indications may be obtained from the experience of cystic fibrosis, or as suggested by expert opinions. IRT is generally considered only in the case of frequent respiratory infections and poor antibody response to specific immunizations. IL-6 and IL-8 may serve as potential biomarkers to identify subjects at higher risk of lung failure.[41,89,97] Interstitial lung disease occurs in about 25% of patients with A-T and usually develops during adolescence. Regular functional monitoring of lung airways by means of spirometry and respiratory physiotherapy is required. Chest radiographs and high-resolution computed tomography scan may show different degrees of inflammation and pulmonary fibrosis, but they are not indicated in these patients; MRI should be preferred given the radio sensitivity.[98]

The nutritional status of adolescents with A-T is often compromised, which is, at least in part, explained by swallowing disorders, recurrent infections, respiratory failure, and reduced motor activity. The involvement of the endocrine and skeletal systems, along with frequent metabolic alterations, contributes to malnutrition.[99,100]

The treatment of endocrine abnormalities is mainly based on use of replacement therapies and on control of the nutritional intake.[101] Vitamin D and lipid metabolism may be impaired in a number of cases. Nonalcoholic steatohepatitis (NASH) is a fairly common condition in adults with A-T, and it parallels metabolic syndrome. NASH leads to liver failure with progressive and irreversible liver fibrosis due to accumulation of fats and to the subsequent inflammatory process. FibroScan, MRI, and MRI with elastography are also useful in NASH monitoring. In the absence of a specific therapeutic treatment, adjustments of the nutritional plan on the basis of liver conditions remain the main supportive treatment.[102,103]

Glucose intolerance, alteration of lipid metabolism, metabolic syndrome, and low levels of vitamin E contribute to the risk of acute (myocardial infarction and stroke) and chronic cardiovascular diseases (eg, arterial hypertension). The use of antioxidant drugs, which are also indicated for the treatment of NASH, is supported by evidence of efficacy in small groups of cases so far reported.

Granuloma, a predominantly inflammatory lesion, frequently occurs in A-T; it mainly affects the skin and, less frequently, the bones, joints, or internal organs. It is likely related to previous administration of the rubella vaccine and has a chronic progressive course. Therapeutic options include topical corticosteroids associated with immunoglobulins, TNF inhibitors; more recently, allogenic HSCT has been proposed.[65,104]

Malignancies mainly affect blood and lymphoid organs, with an increase of solid tumors in young adulthood. Surveillance protocols and treatments require strong attention and careful

monitoring, as patients are hypersensitive to radiography, radiomimetic drugs, and chemotherapy, which are often fatal.

CRD group–specific consensus. After a literature review and scoring, IPINet consensus was reached concerning the following critical issues on transitional care for DRSs and IEIs with susceptibility to malignancy (Table VIII): severe respiratory failure, profound malnutrition or cachexia, advanced neurodegeneration, and end-stage cancers in extremely compromised patients might contraindicate the transition process (rate of agreement 96%; score 0.76 ± 0.35). A partial consensus was reached concerning the case manager within the adult team, who in the opinion of many raters should be the clinical immunologist (rate of agreement 92%; score 0.67 ± 0.34). Special attention should be paid to monitoring multisystemic disease, neurologic deterioration, respiratory failure, and endocrine imbalance, as well as to conducting cancer surveillance (rate of agreement 89%; score 0.81 ± 0.24). Close cooperation between pediatric and adult care physicians is mandatory (rate of agreement 89%; score 0.77 ± 0.25); relevant care professionals such as physical, speech, respiratory, and occupational therapists and nutritionists are highly recommended (rate of agreement 100%; score 0.83 ± 0.24), and the support of a psychologist is highly suggested for patients and their families (rate of agreement 100%; score 0.81 ± 0.24). Younger adults may take advantage of assistance from special education teachers in the transition process (rate of agreement 92%; score 0.78 ± 0.25), and periodic discussions aimed at the mitigation of disease-specific barriers, including aspects related to incomplete understanding of disease pathophysiology and at the evaluation of therapeutic opportunities, should be held (partial consensus; rate of agreement 89; score 0.68 ± 0.24).

No consensus was reached regarding the evaluation of QoL because the majority of raters believed that it may be related more to the natural history of the disorder in adolescence rather than to the transition process.

Common clinical hallmarks in the transitional care for patients having undergone HSCT or GT in childhood

Cellular therapy is increasingly being used for patients with IEIs. Treatment approaches include allogeneic HSCT or autologous hematopoietic stem cells (HSCs) with gene therapy (GT).[105] Allogeneic HSCT is a well-established treatment for the most severe forms of IEIs, including SCID, immunodysregulation polyendocrinopathy enteropathy X-linked syndrome, Wiskott-Aldrich syndrome, and CGD.[106-109] The development of less toxic conditioning regimens (eg, use of reduced dose busulfan, treosulfan, and/or fludarabine) and more sophisticated stem cell procurement and *in vitro* manipulation have led to more widespread use of HSCT for patients with IEIs. Thus, it has been proposed as a treatment option for several other IEIs (eg, CID, lypopolysaccharide-responsive beige-like anchor *[LRBA]* protein, or adenosine deaminase 2 deficiency).[60,110,111] The GT approach has been identified as a successful definitive treatment option for several IEIs, with some advantages over allogeneic HSCT, including the use of autologous HSC and a reduced rate of mortality and complications thanks to less intensive chemotherapeutic agents.[112,113] Patients who received HSCT or HSC GT require LTFU, and transition of care to a dedicated adult team is critical to maintain a high quality of care over the long term.

In particular, attention should be paid to the different clinical aspects, as illustrated in Table II. The uncorrected gene defect in nonhematopoietic tissues may be responsible for ongoing extraimmunologic manifestations.[2] Immune function may not be completely restored by cellular therapy on account of incomplete engraftment or rejection in the case of HSCT or insufficient transduction and loss of gene-corrected cells in GT trials. Specific HSCT settings are associated with a high risk of incomplete immune reconstitution (eg, unconditioned HSCT for IL-2 receptor subunit gamma deficiency is associated with absent B-cell engraftment).[114] Patients might require further cell therapy, or they might need lifelong prophylactic measures to control the disease manifestations. In the latter case, monitoring of complications related to the original IEI should be guaranteed over the course of LTFU. Vaccination schedule and the specific antibody response should be carefully monitored over time.[115]

Among patients surviving cellular therapy for IEIs, an increased risk of cancer might exist because of (1) the specific gene defect (eg, DNA instability); (2) long-term effects of the conditioning regimen; or (3) potentially, insertional oncogenesis (GT).[116,117]

Genetic counseling and sterility treatment. Survivors of cellular therapy for IEIs require genetic counseling to assess the risk of transmitting the disease to their offspring. Moreover, drugs used in the conditioning regimen can cause sterility in some patients. Survivors need adequate counseling and treatment for fertility preservation and also regarding the use of cryopreserved ovules, ovarian tissue, or sperm.[118]

CRD group–specific consensus. Only limited data are available to guide the choice of the optimal clinical model to provide such transitional care. As shown in Table IX, agreement was achieved regarding extraimmunologic manifestations, drug toxicity, and risk for the offspring (rate of agreement 92%; score 0.80 ± 0.25), as well as regarding the need for monitoring of uncorrected manifestations and incomplete immune reconstitution (rate of agreement 100%; score 0.78 ± 0.25), severe organ damage, and chronic graft-versus-host disease (rate of agreement 100%; score 0.82 ± 0.24) and evaluation of QoL (partial consensus; rate of agreement 96%; score 0.66 ± 0.24).

No consensus was reached regarding those clinical conditions for which starting the transition is deemed inappropriate or on the evaluation of vaccination status.

The available information is mostly derived from patients receiving HSCT for childhood cancer, and little is known regarding patients undergoing cellular therapy for IEIs.[2,5,119] However, the majority of raters agreed that the adult team needs to incorporate medical personnel specifically trained in HSCT or GT for IEIs. Because only a few centers worldwide perform cellular therapy for adults with IEIs, the capacity of these structures might not suffice to address the need for all patients requiring LTFU. We propose that in a setting in which such professionals are not available, the LTFU might be provided by a hematologist experienced in cellular therapy in close collaboration with an immunologist experienced in IEIs. Second, patients receiving GT are often enrolled in study protocols that require a specific long-term follow-up as required by the competent authorities (US Food and Drug Administration and European Medicines Agency). In such a setting, the transition to another team might cause issues in the compliance with study requirements. The follow-up plan should be clearly stated, and the results of relevant

TABLE VIII. Recommendation on transitional care for patients with DRSs and IEIs with malignancy susceptibility

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
Consensus			
Severe respiratory failure, profound malnutrition/cachexia	96	4	0.76 ± 0.35
Advanced neurodegeneration	96	4	0.76 ± 0.35
End-stage cancers in extremely compromised patients	96	4	0.76 ± 0.35
Who should be the case manager within the adult team?			
Partial consensus			
Clinical immunologist	92	8	0.67 ± 0.34
No consensus			
Neurologist	56	44	0.13 ± 0.67
Pulmonologist	45	55	0 ± 0.56
Internist	85	15	0.56 ± 0.46
Any health care professional who assumes full responsibility for care coordination and planning	56	44	0.13 ± 0.67
What are the peculiarities to focus on in this specific CRD?			
Consensus			
To monitor several aspects of the multisystemic disease	89	11	0.81 ± 0.24
To monitor neurologic deterioration, respiratory failure, endocrine imbalance, and cancer surveillance	89	11	0.81 ± 0.24
To ensure a solid cooperation between pediatric and adult care physicians	89	11	0.77 ± 0.25
To ensure care by physical, speech, respiratory, occupational therapists and nutritionists	100	0	0.83 ± 0.24
To ensure psychologist support	100	0	0.81 ± 0.24
To ensure educator teacher support	92	8	0.78 ± 0.25
Partial consensus			
To mitigate the presence of disease-specific barriers (eg, incomplete understanding of disease pathophysiology, limitations of information)	89	11	0.68 ± 0.24
To discuss current therapeutic strategies and difficulty of experimental trials	89	11	0.68 ± 0.24
No consensus			
To evaluate QoL index	81	19	0.64 ± 0.42

Level of agreement is the sum of the percentage of strong agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement is the percentage of strong disagreement (rated –1) and the percentage of disagreement (rated –0.5). Consensus is defined as a 75% or higher level of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher level of agreement and a mean score ranging from 0.65 to 0.74.

TABLE IX. Recommendation on transitional care for patients with IEIs who underwent HSCT or gene therapy

Statement	Level of agreement (%)	Level of disagreement (%)	Mean score ± SD
Which clinical conditions mean that it is not appropriate to start the transition?			
No consensus			
Enrolment in a study protocol requiring a specific long-term follow-up (mainly after gene therapy)	77	23	0.50 ± 0.54
Unstable chimerism	77	23	0.35 ± 0.49
What are the peculiarities to focus on in this specific DRG?			
Consensus			
To manage several aspects related both to the genetic disease (eg, nonimmunologic disease manifestations, risk for the offspring) and to the treatment (ie, late toxicity of the conditioning regimen and infused products on fertility, endocrinologic, pneumologic, auxologic aspects)	92	8	0.80 ± 0.25
To monitor uncorrected manifestations and incomplete immune reconstitution	100	0	0.78 ± 0.25
To monitor organ damage and cGVHD	100	0	0.82 ± 0.24
Partial consensus			
To evaluate QoL index	96	4	0.66 ± 0.24
No consensus			
To evaluate the response to vaccinations and attendance to the community	92	8	0.59 ± 0.31

cGVHD, Chronic graft versus host disease.
Level of agreement is the sum of percentage of strongly agreement (rated +1) and the percentage of agreement (rated +0.5); level of disagreement is the sum of the percentage of strong disagreement (rated –1) and the percentage of disagreement (rated –0.5). Consensus is defined as a 75% or higher level of agreement and a mean score of 0.75 or higher. Partial consensus is defined as a 75% or higher level of agreement and a mean score ranging from 0.65 to 0.74.

assessments should be transmitted to the original team that treated the patient with GT.

DISCUSSION

Adolescents and young adults affected with IEIs represent a very vulnerable population. In 2011, the American Academy of Pediatrics, with the endorsement of the American Academy of Family Physicians and the American College of Physicians, emphasized the need to plan health care transition for chronic diseases.[4,120] Although programs of transitional care are continuously evolving for patients affected with several pediatric-onset chronic conditions (eg, cystic fibrosis, inflammatory bowel disease, diabetes mellitus, rheumatologic diseases), at present none of them focuses on IEIs. The medical problems at the time of transition of young people with IEIs are often complex and characterized by a marked trend toward progression over time. Thus, transition of care is critical to maintaining high quality of care of patients with IEIs during LTFU, and it should be considered an emerging priority.[5] IEIs are newly identified diseases, with new gene defects continuously discovered, thus implying that time is required to allow a proper overall knowledge among adult health care professionals. Most adult immunologists are dealing mainly with allergy or autoimmunity and not with IEIs, and many adult specialists in gastroenterology, endocrinology, and respiratory diseases have little or no exposure to patients with IEIs during their training. Dynamic programs aimed at improving awareness of physicians to recognize complications, and promoting optimal treatment, should be implemented in each center to break down barriers. Without an established transition program, a significant number of patients may be lost to follow-up during transfer, with consequently higher rates of nonadherence and increased morbidity and mortality.[5,101]

Because of the high number of IEIs, which differ profoundly in terms of clinical phenotypic expression, the transition process may not be a one-size-fits-all undertaking. The IPINet Committee categorized IEIs in different groups of CRDs that share common major clinical hallmarks deserving special attention during the transition. The committee formulated a number of statements for each CRD group to help physicians during the process. Both pediatricians and adult care providers need to screen for, prevent, and treat unique medical and psychosocial comorbidities associated with different IEIs in a tailored approach to health.[60] Adaptation of vaccination schedule, monitoring of the effects of advanced therapies and IRT, cancer surveillance, and promotion of preventive health measures and compliance with care (including lung physiotherapy) are critical to achieve successful clinical management in this population. A few patients with IEIs have different degrees of ID, requiring a personalized approach to transition, including decision-making support and neuropsychiatric evaluations by trained providers.

New technologies, such as chromosome microarray and high-throughput next-generation sequencing, were not available when patients with childhood-onset IEIs were first investigated.[60] Therefore, adolescents without a definite molecular diagnosis should be reevaluated before transition.

One of the main issues that emerged is that the transition may be a very difficult task for those patients with more complex syndromes, mainly when a specialized, fully trained, and well-equipped adult center is not available. Adult specialist physicians should have dedicated expertise in the specific clinical problem within the context of the IEI pathogenic mechanism.

Critical health conditions have been considered by the committee a contraindication to the transition for most IEIs. Nevertheless, to avoid sudden decisions that are forced or imposed under unexpected and rapidly progressive conditions in the context of an intensive care setting health care, providers should carefully plan in advance the transition for patients with severe chronic organ damage. Young adults with IEIs experience psychosocial issues, such as depression, anxiety, suicidal ideation, relational difficulties, and engagement in risky behaviors, which require careful psychological support.

Going forward, improving transitional services will require careful evaluation of several issues through *ad hoc* studies focusing on development of valid measures of quality improvement and implementation and also taking local resources into consideration. Quantification of health-related QoL in IEIs has recently been initiated as an important tool to evaluate patients' health status over time, to assess the effects of therapeutic intervention, and the appropriateness of the transitional process.[59] Unlike for other chronic conditions such as diabetes or cystic fibrosis, for which QoL measures are available, for IEIs, only the CVID-QoL questionnaire has been developed as a disease-specific instrument for adults with CVID.[121] Several variables have been suggested as objective outcome measures for evaluating a successful transition, including reduced attempts to return to child-centered care. It must be emphasized that disease-specific outcome measures are to be preferred. Unfortunately, tools to evaluate the process outcome for IEIs are not available. Instruments validated for other chronic diseases, such as the Karnofsky Performance Status Scale, the Mind the Gap scale or the On Your Own Feet Transfer Experiences Scale, the Hospital Anxiety and Depression Scale, and the Treatment Satisfaction Questionnaire for Medication-9, may be used to evaluate disease activity status, anxiety, depression, or adherence to therapy. Patient and family empowerment should be checked according to the American College of Physicians to evaluate the transition readiness. The frequency of follow-up has not reached universal consensus because data obtained during the transitional age are not available. However, follow-up should be adapted to each IEI, to the peculiarity of each patient, and to center rules. Subspecialty physicians should be involved by the Case Manager on an individual basis.

In Developing Countries with a poor economy, the shortage of health care providers and the difficulty of creating multidisciplinary teams make planning transitional care even more complex, and thus, protocols should be adapted to local environment.

Our scope was aimed at defining a general framework to help the transition process in the group of IEIs, which are extraordinarily complex diseases that invariably challenge physicians, patients, and their families.

Conclusion

We have presented a consensus statement on the transitional care for patients with different IEIs that can serve as a rational basis for future experimental studies and guidance. These guidelines will be updated periodically as more evidence becomes available.

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Clinical implications: A consensus on the transition process to adult health care services for patients with IEIs has been formulated by physicians from the IPINet to improve clinical practice.

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ORIGINAL RESEARCH
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The Transition of Care From Pediatric to Adult Health-Care Services of Vertically HIV-Infected Adolescents: A Pilot Study

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Objective: Clinical and psychological HIV-related problems peak during adolescence, which coincides with transition of children and adolescents infected from mothers from pediatric to adult reference centers for HIV infection. Transition often is done without specific programs. We wanted to explore transition as an opportunity to increase the efficacy of care and the psychological well-being through a specific program.

Methods: Thirteen vertically infected patients aged 13–20 years were followed up for 24 months by pediatricians, infectious disease specialists, a psychologist, and a nurse. Interventions consisted in joint clinic, simplification of therapy, patient group discussions, HIV infection explanations, and psychological support, lasting 12 months. Efficacy was measured by viro-immunological outcomes and adherence to therapy and psychological tests. Clinical, viro-immunological, and psychological evaluations were performed at 0 (T0) and 12 months (T12) and 6 months after transition to an adult center (T18). Psychological outcomes were assessed using standardized questionnaires for quality of life and self-esteem.

Results: In 11/13 participants, pills administrations/day were significantly reduced. Patients with undetectable viral load and CD4+ >25% increased from 61 to 77% and from 61 to 74%, respectively. Six months after transition, all patients exhibited an undetectable viral load. Adolescents' awareness of the severity of the disease and the risk of sexual transmission was generally poor. Patients classified with "severe" psychological distress according to the quality of life index decreased from 38 to 15% and well-being increased. Similar results were observed 6 months after the transition to adult care. No effect was observed on self-esteem index.

Conclusions: Specific protocols for transition should be developed to optimize resilience and psychological well-being, including routine psychological support for adolescents with HIV infection transiting from pediatric to adult centers for HIV infection.

Keywords: HIV, adolescents, transition, biopsychosocial approach, well-being

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INTRODUCTION

An increasing number of vertically HIV-infected adolescents are referred to centers for adult (1). In pediatric HIV infection, as in many other childhood illnesses, the chronicization of the disease has led to new problems including adherence, life-long treatment self-management of the medication side effects, stigma, and—of major importance—awareness of sexual transmission (2, 3). The number of drugs approved for pediatric patients is limited compared with that for adults, and this may hamper adherence with antiretroviral therapy (ART), because non-adherence is often associated with side effects and psychosocial implications in children and adolescents (4–6). Moreover, psychological and social HIV-related problems are amplified in children and adolescents growing to adulthood (7–9). In a previous study, using the International Classification of Functioning, Disability and Health instrument (ICF), we found that psychosocial issues had a major impact on quality of life than had clinical problems in a population of children and adolescents with HIV infection (10). In addition, the stigma associated with HIV infection is a major barrier to social and psychological health (11). Sexual transmission of HIV infection inhibits the development of an emotional life in adolescents, affecting the psychosocial stability and the sexual health (12).

All these factors complicate the management of teenagers with HIV infection and negatively impact adherence to ART, medical visits, and psychosocial functioning. Up to 20% of pediatric patients with HIV infection show poor short-term adherence (within the last month) to ART, and this percentage increases to 50% when adherence is assessed in the long term (within the last 3 months) (13–15). The complexity of therapy is one of the major determinants of low adherence in children with HIV. Current guidelines highlight the importance of tailored strategies to implement ART adherence, particularly in adolescents (https://aidsinfo.nih.gov/guidelines/html/1/adult-and-adolescent-arv-guidelines/30/adherence-to-art). Reduction of pill number and administrations per day to once per day fixed dose combination scheme is approved for adult patients only.

The transition to centers for HIV care of adults represents a bridge between the dependence and protection typical of pediatric patient and the liberty, responsibility, and self-management of adult patients and is characterized by specific problems and emotional–relational behaviors in a dynamic equilibrium. However, transition to adult health-care settings is only one of the aspects of the more general growth to adulthood, during which younger people undergo a change that is both systemic and cultural. Although specific programs for transition to adult care centers have been reported for cystic fibrosis, diabetes mellitus, and other chronic diseases, only limited data are available on the transition of adolescents with HIV (16). Transition is associated with several problems and risks, including loss to follow-up, low adherence, disruption of psychological health, problems in hiding HIV status, stigma, and isolation from peers. Therefore, transition is a delicate period in which management changes, and this begs risks for adolescents. Yet transition is often achieved without a specific program, and adolescents are simply referred to centers for adult patients, where management and approach are different from those of pediatric centers. We wanted to explore transition as an opportunity to increase the efficacy of care and the psychological well-being to ultimately increase resilience, a major resource for HIV-infected patients.

The American Academy of Pediatrics states that health-care providers should develop a formal process for transitioning HIV-infected adolescents to adult health care between the ages of 18 and 25, starting with the patients in their early teens (17). However, in most centers for pediatric HIV infection, the patients are either transferred to adult care without a specific program or continue their follow-up in pediatric infectious disease units.

The Transition of Seropositive Children to Adulthood (TOSCA) project was a pilot study carried out to test the efficacy of an integrated multifaceted intervention aimed at optimizing the transition of care from pediatric to adult health-care services. We hypothesized that a specific medical and psychological intervention applied during transition of care to the adult care center improves the viro-immunological and psychological conditions of adolescents, ultimately increasing their resilience.

METHODS

Study Protocol and Population

The Reference Center for Pediatric HIV/AIDS of the University of Naples Federico II covers a territory of about 7 million inhabitants of Southern Italy and manages about 40–50 HIV-infected children and adolescents with one to three new cases of vertical HIV infection every year.

All patients aged $\geq$13 and $\leq$20 years who already had received the disclosure of HIV infection and for whom informed consent had been obtained were included in the present study. The study protocol included three phases: in the first phase, we created the multidisciplinary team and developed the study materials. All patients underwent full clinical, immunological, and virological evaluations. In addition, information regarding disease knowledge, adherence to therapy, and psychological status (including self-esteem) was obtained. Next, all patients underwent the bundle of clinical and psychological interventions as detailed below. All the parameters were recorded at baseline (T0) and 12 (T12) months after the preparation to transition was achieved. In the third phase, patients were transferred to adult care. All the parameters were recorded again 6 months after the transition (T18).

The intervention consisted in a bundle of initiatives run by a multidisciplinary group including a pediatrician specialist (PID) as well as an infectious disease specialist (AID), a dedicated nurse, a psychologist, and a social assistant. The group met periodically to discuss study protocol, progresses, barriers, and interventions.

All patients enrolled had been followed up since birth or the initial diagnosis of vertical HIV infection. Before this study, PID took care of HIV-infected adolescents up to 20 years of age in specific pediatric reference and then referred them to AID in the reference center for adult HIV-infected patients. Transition only included a brief discussion and written reports of their clinical and immunological state, previous ART regimen, concomitant diseases, and adherence to ART by the PID specialist.

OUTCOME PARAMETERS AND INTERVENTION BUNDLE

Outcome Parameters

According to the study protocol, the clinical and psychological evaluations were performed before starting the intervention (T0), after 12 months (T12) months, which is during transition, and 6 months after transition.

Viro-Immunological Parameters

Every 2 months, each patient with HIV underwent full evaluation of viral load (HIV RNA measured as the number of HIV copies/ml of blood) and of $CD4^+$ cell count (measured as the number of $CD4^+$ cells/mm^3 of blood). Both parameters are markers of infection status and of ART efficacy. The same parameters were recorded at T0, T6, T12, and T18 to have a closer control of the state of infection.

Patient's Knowledge of HIV Infection

To assess the knowledge of HIV infection, a brief questionnaire was developed with six multiple-choice questions, investigating awareness of modality of HIV transmission and importance of ART. The questionnaire was administered at the beginning of the study and at T12. Each correct response was scored 1, and a wrong response was scored 0.

After baseline assessment, each patient was asked to participate to a discussion on HIV infection, transmission, and management. This was repeated during the intervention period at least two times, and at the end of the intervention, the questionnaire was administered again.

Evaluation of Psychological Well-Being

Two standardized questionnaires were used to investigate patients' well-being and self-esteem:

- The *Psychological General Well-Being* (*PGWB*) index is an established patient-reported instrument, which provides self-reported assessment of psychological health. It does not include an evaluation of physical health. This 22-item instrument includes six domains: Anxiety, Depressed Mood, Positive Well-being, Self-Control, General Health, and Vitality. The 22 items generate an overall index or total score for general well-being. The original score for each item ranges from 0 to 5, giving a possible total score 0–110 (18). A total score < 59 indicates severe distress, 60–72 moderate distress, and 73–98 no distress. Higher scores indicate higher levels of well-being as perceived by the patient. PGWB takes 10 min or less to complete and is generally well-accepted.
- The *Multidimensional Self-Esteem Test* (*TMA*) is a validated questionnaire for self-esteem particularly in adolescents in its multiple dimensions. The test includes six areas (personal, school, emotional, skills, family, and body), and consists of six groups of 25 items for each area with each item requiring one of four possible answers: absolutely true, true, not true, and absolutely not true. The average scores for self-esteem in a standard reference sample range between 85 and 115 (19).

INTERVENTION BUNDLE

Joint Pediatric/Adult Clinic

A joint transition outpatient clinic was set up by PID and AID physicians and a study nurse in order to have a joint medical visit every 2 months. The last two medical visits for each patient were held at the Department of Adult Infectious Diseases, with the presence of PID, in order to achieve a smooth transition. In the last visit, PID and AID jointly discussed each patient's treatments and simplification of ART regimen in the presence of the patient and his/her family.

Knowing HIV Infection, Its Transmission Routes, Its Consequences, and Self-Management

A 30- to 60-min face-to-face education session focusing on HIV infection, its clinical manifestations, risks, transmission routes and prevention measures, and principles of ART and adherence was provided to each patient by a PID, and it took place at the Department of Pediatrics. The meeting included time for questions and answers. At the end of session, the physician scored the availability/involvement of each patient using a 5-point Likert scale (1 = strongly disagree to 5 = strongly agree). In order to better understand patients' doubts and requests, overcome barriers linked to anxiety and embarrassment, and drive future meetings, PID specialists and the nurse also answered questions collected from patients, written anonymously and collected in a box in a blinded fashion.

Psychological Support and Group Discussion: Resilience as a Resource

A psychologist experienced in the management of pediatric HIV infection organized individual meetings once a month; in addition, group meetings with all patients and their families were organized twice a year. A study nurse and a gynecologist took part in these meetings to provide information on prevention of HIV transmission and protection during sexual contacts.

A bundle of interventions and their time points are reported in **Table 1**.

STATISTICAL ANALYSIS

Quantitative variables were reported as means ± standard deviation (*SD*), and variables with skewed distributions were presented as medians and interquartile ranges (IQRs). Results were compared by *t*-test or the Mann–Whitney non-parametric test, as appropriate. Categorical variables were summarized and reported as frequencies and percentages and compared by Fisher's exact test or chi-square test, as appropriate.

Two-sided $p < 0.05$ were considered statistically significant. Statistical analysis was performed using the SPSS software (version 20.0 for Windows; SPSS Inc., Chicago, IL, USA).

TABLE 1 | Bundle of interventions and their time points.

Intervention	T0	T6	T12	T18
Phase 1				
Informed consent	X			
Development of multidisciplinary team	X			
Development of study material and database	X			
Patient enrollment	X			
Phase 2				
Clinical evaluation	X	X	X	X
Assessment HIV immunological class	X		X*	X
Assessment CD4+ count and percentage	X	X	X	X
Assessment HIV viral load	X	X	X	X
Educational session	X	X	X	
ART review and simplification		X	X	
Administration of knowledge questionnaire	X		X	
Psychological interview	X		X	
Administration of PGWB questionnaire	X		X	X
Administration of TMA questionnaire	X		X	
Phase 3				
Transition of adolescents to AID unit				X
Psychological follow-up				
Analysis of results				X

ART, antiretroviral therapy; PGWB, Psychological General Well-Being; TMA, Multidimensional Self-Esteem Test.
**Or in case of any acute illness defining severe AIDS.*

DATA MASKING, ETHICS, AND FINANCIAL SUPPORT

Single codes were assigned to each enrolled patient to collect and report data anonymously. All study data were collected through manual chart and reviewed by two persons (ALV and EB) involved in the study and recorded in a Microsoft Excel database. The results of psychological tests were analyzed by a psychologist and a social worker and subsequently recorded in the same database. At enrollment, the study protocol was presented and discussed with patients and caregivers who signed a specific informed consent according to patients' age.

The study was conducted according to the Declaration of Helsinki, and the protocol was approved by the Ethical Committee of the University Federico II of Naples (protocol number 229/15). The TOSCA study was supported by a competitive grant by GILEAD Sciences S.r.l. (Milan, Italy).

RESULTS

Study Population

The eligible population included 13 adolescents with vertical HIV infection on ART followed up to the reference center of Naples. All patients (age range 14–20 years) were enrolled. The general characteristics of the population at enrollment are shown in **Table 2**.

TABLE 2 | General characteristics of the study population and HIV class at study enrollment.

Characteristics	*N*
Total enrolled patients (male/female)	13 (5/8)
Median age at diagnosis, years (range)	2 (1–10)
Median age at enrollment, years (range)	17 (14–20)
Ethnicity	
Caucasian	11/13
African	2/13
Years on ART, mean (SD)	14 (3)
HIV disease and infection class*at diagnosis	***n***
Class (C1/C2/C3)	0/0/1
Class (B1/B2/B3)	1/1/6
Class (A1/A2/A3)	3/1/0
Patients with undetectable HIV viral load§	8

ART, antiretroviral therapy.
**HIV disease staging is defined according to the U.S. Centers for Disease Control and Prevention (CDC) pediatric classification system.*
§Defined as HIV RNA < 40 copies/ml.

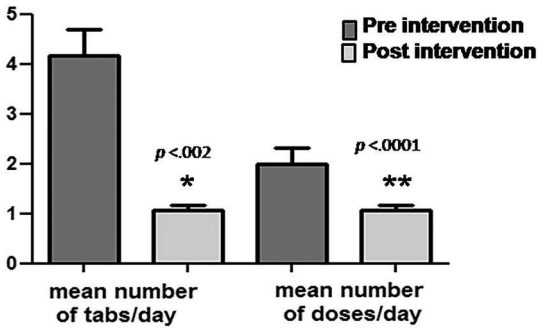

FIGURE 1 | Simplification of therapy by applying the drug protocols available for adult patients. Both the mean number of daily pills and number of daily drug doses were significantly reduced (4.18 ± 1.72 to 2.0 ± 1.1, $p < 0.002$, and 1.82 ± 0.4 to 1.09 ± 0.3, $p < 0.0001$, respectively) during the transition period. *$p < 0.005$; **$p < 0.0001$.

During the study, one patient developed a severe psychiatric disorder and dropped out before the end of transition process (T12). For this patient, no data are available for T18.

Modifications of Antiretroviral Therapy

In 11/13 participants (85% of the total), the pediatric drug regimen was simplified by adopting the adult protocol during the 12 months of transition protocol. Both the number of daily pills and doses were significantly reduced (from 4.18 ± 1.72 to 2.0 ± 1.1, $p < 0.002$, and from 1.82 ± 0.4 to 1.09 ± 0.3, respectively, $p < 0.0001$). For three of the nine patients (33.3%) on a multi-tablet regimen, it was possible to switch to once-daily fixed dose combination scheme (**Figure 1**).

Viro-Immunological Parameters

At 12 months post-intervention, the total number of patients with undetectable viral load in blood increased. An increase in

CD4+ lymphocytes was documented at T12, when 11/13 patients showed a percentage of CD4+ above 25%. At 18 months, all transited patients were viro-suppressed, and an increase of mean CD4+ cell count was observed, although the number of patients with a percentage of CD4+ above 25% slightly decreased. The results of HIV viral load and of the CD4+ lymphocytes count at T0, T6, T12, and T18 are reported in **Table 3**.

Knowledge of HIV

In the pre-intervention evaluation, none of the patients was able to correctly answer all the questions. However, at the end of the project, 3/13 patients gave all the correct answers to the six questions in the questionnaire (0 vs. 23%). On average, the mean number of correct answers, at baseline, was 3.62 ± 1.39; at the end of the study, this number increased to 4.31 ± 1.38 (p.21). Of note, 5/13 (38%) patients answered "I do not know" to the question: "Based on current knowledge, can you recover from HIV infection?" Furthermore, eight patients (61%) reported that they did not consider the therapy useful to protect partners or close contacts from being infected. Surprisingly, all patients showed a high level of willingness to participate in the assessment sessions and the discussion, both in the pre-intervention and post-intervention phases; and the number of missed meetings was limited.

TABLE 3 | Viral and immunological modifications observed in HIV-infected adolescents at baseline, during, and 6 months after the intervention.

Characteristics	T0 (n = 13)	T6 (n = 13)	T12 (n = 13)	T18 (n = 12)
HIV viral load				
HIV RNA < 40 copies/ml (n, % of patients)	8 (61.5)	8 (61.5)	10 (77)	12 (100)
Immunological status				
T CD4+ < 15% (n, %)	1 (7.7)	0 (0)	0 (0)	0 (0)
T CD4+ 15–25% (n, %)	4 (30.8)	4 (30.8)	2 (15.4)	3 (25)
T CD4+ > 25% (n, %)	8 (61.5)	9 (69.2)	11 (84.6)	9 (75)
T CD4+ absolute count (mean ± SD)	711 ± 357	728 ± 338	724 ± 297	799 ± 411

Psychological Interventions

A total of 25 psychological interviews and 25 PGWB and 23 TMA tests were completed. With regard to the psychological interviews, most of the patients showed an adequate level of participation as judged by the timeliness and the adherence to the scheduled meetings. The patients reported their personal experience of living with a chronic infectious disease, their relationships with peers, and the psychological burden of the disease and its therapy. Individual experiences, personal worries, and possible solutions were shared with the group and discussed. The concept of "transition" was extensively discussed as an active passage to adulthood and a necessary step to self-management of HIV infection, but also as an opportunity to effectively address unresolved issues.

All patients answered the PGWB and TMA questionnaires, showing good levels of participation and cooperation. As shown in **Figure 2A**, at baseline assessment, a "severe" level of distress was found in 5 of the 13 patients (38%), with scores lower than 59. Seven other patients with scores between 73 and 98 and 1 > 100 showed no distress or even positive well-being on PGWB. After 12 months, only two patients (15%) were still in the severe distress range, one showed moderate distress, and the remaining nine had no distress. Nine out of twelve patients showed an increase in the global wellness index (PGWB) scores compared with the baseline, but in three cases, the scores were lower

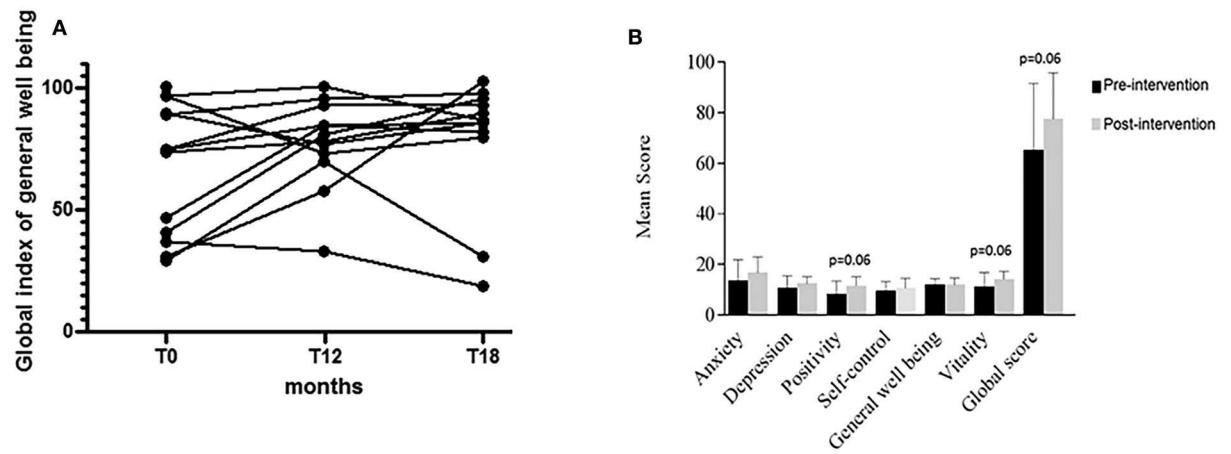

FIGURE 2 | Assessment of general psychological well-being. **(A)** At baseline assessment, a "severe" level of distress was found in 5 of 13 patients (38%), with scores lower than 59. At the end of intervention (T12), nine of the 12 remaining patients showed an increase in the PGWB scores compared with the baseline pattern, but in three cases, the scores were lower compared with baseline. One patient, who showed a condition of severe distress at baseline, improved at the end of the intervention but returned to his/her basal condition as early as 6 months after transition. One patient showed a slight deterioration of the well-being index during the intervention, which further worsened after transition. **(B)** The score increase was more pronounced in the two specific domains of vitality and positivity. The global score of general well-being increased at the end of intervention, although the value did not reach statistical significance (p = 0.06).

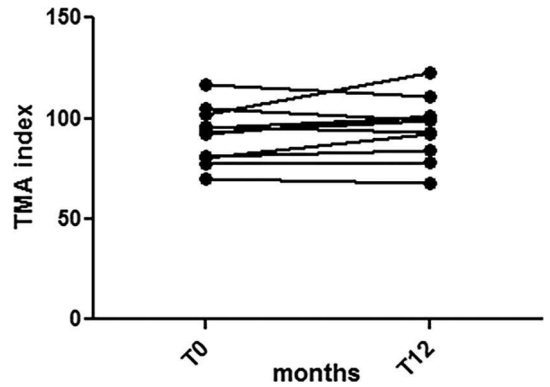

FIGURE 3 | Self-esteem assessment. Seven out eleven patients with HIV infection reached or exceeded a score of 85 at T0. After 12 months of intervention, 8/11 had a score higher than 85. Of the four patients with an initial score below 85, only one patient scored more than 85 at the 12-month follow-up; the other three did not show any improvement in self-esteem. Overall, the results showed that HIV-infected adolescents exhibited a good level of self-esteem.

compared with baseline. Surprisingly, one patient with severe distress at baseline improved at the end of the intervention but returned to his basal condition already 6 months after transition. One patient showed a slight deterioration on the well-being index during the intervention and showed a further slight worsening after the transition (**Figure 2A**). The global score of general well-being increased at the end of intervention from 68 to 77.5, although the difference did not reach statistical significance ($p = 0.06$). The increase was more pronounced in the two specific domains of vitality and positivity (**Figure 2B**). Six months after transition, the mean global score of general well-being was 79, indicating no distress.

Eleven of the 13 patients also completed the two-point evaluation of TMA (T0 and T12). This test showed that the majority of patients (7/11) with HIV infection had a fair level of self-esteem, having reached or exceeded a score of 85 at T0. After 12 months of intervention, 8/11 patients had a score higher than 85. Of the four patients with an initial score below 85, only one patient scored more than 85 at the 12-month follow-up; the other three did not show any improvement of self-esteem (**Figure 3**).

DISCUSSION

Transition provides an opportunity to improve clinical and psychological outcome and increase their resilience, and we observed that viro-immunological parameters as well as psychological health improved or showed an improving trend upon application of multifaceted clinical and psychological intervention.

In children and adolescents, ART is effective in controlling HIV-related symptoms; but the overall quality of life is far from optimal, owing to problems in the psychosocial sphere (10). In a previous study, we observed a major burden of psychosocial issues in a population of HIV-infected compared to a population of non-HIV-infected children (20). Poor socioeconomic conditions, high rates of unemployment, family breakdown, and problems in HIV disclosure are the most difficult obstacles in the management of HIV-infected adolescents (21). This indicates that psychosocial parameters, rather than just the clinical aspects, should be a target of effective care of HIV infection. Transition to adult care centers provides a major opportunity to optimize management, and this should be regarded as a purposeful process in which short- and long-term goals can be constantly reassessed (22).

We designed an integrated multifaceted intervention with the aim of taking advantage of transition to introduce a medical–psychological intervention and tested it in a pilot study. Our data showed that at the end of the intervention, all the assessed outcomes, and immuno-virological psychological parameters improved; and also, knowledge of the disease improved. The number of patients with CD4+ count above 25% and the number of viro-suppressed patients increased, probably as a result of the simplification of therapy upon application of adult patient schemes. We do not know which intervention among those applied was more effective. The sample size was limited, and the individual conditions were rather heterogeneous. The effects were also affected by family settings. However, the improvement was evident, and many, although not all, unresolved problems improved in all patients.

The interviews provided a worrying scenario with most adolescents not being aware of the peculiar features of the infection and its transmission. Many patients were not aware of sex transmission and protection opportunities. This finding suggests that their attention is mainly focused on themselves and not on their partners and that HIV-infected adolescents need more information about the disease.

However, after intervention, the number of patients with severe distress decreased significantly as indicated by an increase of their global wellness index score. It is probable that the transition process and the cooperation between PID and AID specialists positively affected the sense of safety of patients, lowering the degree of anxiety related to the change of the health-care team and place of care. Although at baseline 38% of patients showed a "severe" level of distress, the majority of adolescents showed a fair degree of self-esteem, having reached or exceeded a score higher than 85, and no further increase was observed at the end of the intervention. Recent data show that relational self-esteem is positively associated with psychological well-being and indicate a connection between relational self-esteem and social support among vulnerable children such as HIV-infected children and adolescents (23). The high self-esteem observed in our population before the intervention may derive at least in part from the psychological support, which is routinely provided by the pediatric center team. A progressive ability of self-management associated with increase in awareness makes the transition to an adult care center easier.

The positive effect of the transitioning program on both viro-immunological and psychological outcomes was maintained for at least 6 months after transition, when the patients were followed up by AID. All adolescents maintained good clinical and

immuno-virological control of the disease, and there was no case "lost to follow up."

The major limitations of the study are the small sample size and the absence of comparative group. Despite these limitations, the results of this pilot study demonstrate for the first time the efficacy of a multifaceted transition process in HIV-infected adolescents and provide a conceptual framework for randomized controlled multicenter trials in larger populations.

In conclusion, transition to the adult health-care system is a delicate step that may lead to a clinical improvement of the disease and also decreasing psychological distress, optimizing resilience, which is a major resource in HIV infection. An integrated multifaceted intervention jointly conducted by pediatricians and adult physician and by expert psychologists is effective in increasing the knowledge of HIV infection, controlling the degree of distress experienced, and providing information about sexual precautions. Finally, it improves the quality of life of vertically acquired HIV adolescents, providing relief to the psychological problems that are often neglected by a "pure" medical care.

DATA AVAILABILITY STATEMENT

The datasets generated for this study are available on request to the corresponding author.

ETHICS STATEMENT

The studies involving human participants were reviewed and approved by Ethical Committee of the University Federico II of Naples. Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin.

AUTHOR CONTRIBUTIONS

EB, GC, AL, and AG made substantial contributions to the conception or design of the work or the acquisition, statistical analysis, or interpretation of data for the work. AG drafted the work or revised it critically for important intellectual content. CR, CS, CM, and FB organized the database and acquired and analyzed the data for the work. GP and MC made contributions to the conception or design. All authors contributed to manuscript revision, read, and approved the submitted version.

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Letter to the Editor

Letter to Editor regarding 'Barriers in transitioning urologic patients from pediatric to adult care'

Dear Editor,

We refer to the article: 'Barriers in transitioning urologic patients from pediatric to adult care' published in Journal Of Pediatric Urology (December 2020) https://doi.org/10.1016/j.jpurol.2020.12.020 [1].

The authors nicely summarize the strengths and the critical problems related to the transitional period. Furthermore, they produced a snapshot of the major concerns for the physicians and the healthcare system: longstanding patient relationship, developmental care, lack of training in pediatric-onset conditions, adolescent healthcare, extensive time to establish trust, lack of previous clinical information, inappropriateness of the infrastructures. They reported a large American consensus statement and further updates where it's stressed the importance of a high-quality care transition program. Finally, they provided a list of possible transitional models even if the majority were run with few patients, in an experimental setting or are not yet validated [2].

In this background, with lack of a definite and definitive consensus, expertise, and space within the health system for the transitional care patients, we're trying to answer this unmeet question by presenting our idea/project for an Interhospital Department (ID) named 'Centro Interaziendale di Chirurgia Urologica Pediatrica'. ID consists of Meyer Children Hospital and Careggi Hospital, under the supervision of the University of Florence. These structures are symbolically divided by few meters and started to collaborate for patient's healthcare. ID was created in 2015: it provides a multidisciplinary approach with a sharing of technology and expertise for complex and lifelong healthcare. The link between the two structures allowed us to share technologies and personal resources typical of the adult setting for the children; in the last five years we developed robotic urological surgery and retrograde intrarenal surgery as well as percutaneous nephrolithotomy for pediatric patients [3,4].

Furthermore, we aimed to create a transitional care pathway for the importance of the follow up into adulthood of those patients with urinary tract malformations such as spina bifida, hypospadias, bladder exstrophy, posterior urethral valves.

We proposed to set up a program as follows: between 11 and 14 years old the patients attend an outpatient visit with the pediatric physician of trust, where the following steps are shared and defined. Between 14 and 15 years old, 3—4 outpatient visits together with patients, families and two physicians are scheduled. From 16 years old, or when the patient feels ready, he/she attends outpatient visits with the adult care providers, inside the pediatric hospital. Always available in both hospitals there's a multidisciplinary transitional team made of multiple health figures: urologists, nephrologists, gynecologists, andrologists, endocrinologists, internists, psychiatrists, psychologists.

Unfortunately, due to COVID-19 pandemic, some ordinary activities were stopped and others reduced and some parts of our approved project of a ID is only in the 'starting box' of a correct run for these patients [5,6].

In conclusion, increasing the average life of these patients with urological malformations, a structured transitional program, shared by interhospital specialists, appears to us as well as feasible, moreover almost mandatory in the taking care of these patients. Aim of this project is to preserve renal function, reduce the number of unnecessary visits to the emergency department and make these patients feel as welcome as possible both in their self-determination and self-esteem and in the ordinary adult society.

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RESEARCH Open Access

Transition of inflammatory bowel disease patients from pediatric to adult care: an observational study on a joint-visits approach

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Abstract

Background: Transition from pediatric to adult care of patients affected by Inflammatory Bowel Disease (IBD) is a critical step that needs specific care and multidisciplinary involvement. The aim of our study was to evaluate the outcome of the transition process of a cohort of IBD patients, exploring their readiness and the possible impact on quality of life.

Methods: This observational study followed transitioned IBD patients from pediatric to adult care. Transition was carried-out through combined visits, jointly performed by the pediatrician and the adult gastroenterologist. Clinical data were collected before and after transition. A subgroup of patients was submitted to an anonymous online questionnaire of 38 items based on the validated questionnaires TRAQ and SIBDQ within the first 6 months from the beginning of the transition process.

Results: Eighty-two patients with IBD were enrolled, with a mean age at transition of 20.2±2.7 years. Before transition, 40.2% of patients already had major surgery and 64.6% started biologics. At transition, 24% of patients were in moderate to severe active phase of their disease and 40% of them had already been treated with ≥ 2 biologics. The mean score of the TRAQ questionnaires collected is 3.4±1.5 and the mean score of SIBDQ is 53.9±9.8. A significant association was found between a TRAQ mean score > 3 and a SIBDQ > 50 (p=0.0129). Overall, 75% of patients had a positive opinion of the transition model adopted.

Conclusions: A strong association has been found between TRAQ and SIBDQ questionnaires, showing how transition readiness has a direct impact on the quality of life of the young adult with IBD.

Keywords: Transition, Pediatric IBD, Crohn's disease, Ulcerative colitis, TRAQ, SIBDQ

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Introduction

Up to the 25% of patients affected by Inflammatory Bowel Disease (IBD) receive their diagnosis before the age of 20 years, with a constantly growing incidence of the diagnosis made during childhood [1]. Pediatric IBD shows peculiar features compared to adult one, including a more extensive disease and a more frequent upper gastrointestinal tract involvement. Moreover, patients with an onset diagnosed during childhood are recently more likely to receive immunosuppressive or biological therapies since from the moment of diagnosis [2]. Children with IBD are found to have frequent mood disorders and are considered at higher risk for difficulties in social, family and school functioning [3]. A crucial phase in the management of children with IBD is the transition from the pediatric to the adult IBD care system, requiring a full achievement of maturity and self-management skills, which not occur at the same age for each individual [4]. The "ideal" patient for a transition process should be a young independent adult in a clinical remission phase, with no change of medical therapy or surgical intervention necessarily planned [5].

The guideline of the British Society of Gastroenterology on transition of young adults with chronic digestive disease recommends a shared management between pediatrician and adult center, through an educational and conscious process that involves both the patient and his/her family, verifying the readiness of the young and his/her level of awareness [6]. The Transition Readiness Assessment Questionnaire (TRAQ) is one of the most useful tool (not disease specific) for assessing patient's readiness to fulfill the transition from the pediatric health service to the adult care [7], exploring both the self-management domain (e.g. handling medications, arranging medical follow-up visits, managing finances, health insurance) and the self-advocacy one (e.g. communication with providers and managing activities of daily living and use of school and community resources). As far as IBD are concerned, age seems to be the best predictor of TRAQ score, and lower scores on the medication management section are associated with higher risk of nonadherence [8].

The Short Inflammatory Bowel Disease Questionnaire (SIBDQ) is a validated health-related quality of life (HRQoL) tool, designed to find out IBD symptoms, emotional status and limitation in social activities due to IBD symptoms in the last 2 weeks [9].

The aim of this study is to evaluate the outcomes of the transition process of consecutive patients transferred from two pediatric referral hospitals to an IBD adult hospital unit, in order to explore patients' readiness and its association to the quality of life.

Materials and methods

Data collection

The study was carried out at IBD Unit - Fondazione Policlinico Universitario A. Gemelli IRCCS, Rome Italy. Consecutive patients with a previous diagnosis of pediatric IBD made within the age of 19, who transited from two different pediatric centers in Rome (Italy), the "Bambino Gesù Pediatric Hospital" (OPBG) and the "Policlinico Umberto I" on a period going from January 2014 to June 2019, were included. For each patient transition have been carried out through two joint visits, with the presence of both pediatric and adult gastroenterologist specialists.

The first visit occurred at the pediatric center, where patients, parents and doctors examined the previous medical history and planned the timing of transition, according to the patient's clinical status and needs, evaluating his grade of maturity and awareness of disease.

The second visit occurred instead at the adult center, giving to the patients the possibility to discuss with doctors about future plans and therapies in a more autonomous and conscious way, weather in presence or not of parents according to their will. From that moment, follow-up of patients has been made by the adult gastroenterologist, with a maximum distance of 2 months within two different visits and with an average length of the follow-up period of 18 months before the end of the data collection.

Clinical data that were collected at the moment of enrolment have been gender, age at diagnosis and at time of transition, type and location of IBD according to the Montreal classification [10], previous surgery, previous and current therapies at time of transition, clinical disease activity at diagnosis and at transition visit (measured with Partial Mayo Score (PMS) for Ulcerative Colitis (UC) and with Harvey-Bradshaw Index (HBI) for Crohn's Disease (CD) [11, 12]) and available endoscopic and imaging reports. Relapse was defined as worsening of symptoms, change of treatment or need for surgery.

Patients were then submitted to an anonymous online questionnaire of 38 items based on the validated questionnaires TRAQ and SIBDQ and administered within the first 6 months from the beginning of the transition process. Questions from TRAQ about health insurance covers had been removed, due to the differences of the Italian public healthcare system. Poor transition readiness has been defined as a TRAQ mean score ≤ 3 points out of 5. The SIBDQ consists instead of 10 questions, scored of a seven-point scale with higher scores indicating a better quality of life. Good HRQoL was defined as a score above 50 points out of 70 [13].

Two final questions were added in order to evaluate the quality of transition process: 1) When do you think

the transition process from your pediatrician to your gastroenterologist should have taken place? 2) What do you think are the most important necessary conditions for an ideal transition?

The study protocol conforms to the ethical guidelines and it has been approved by the ethical committee of the centers involved. An informed consent has been obtained from all patients.

Statistical analysis

Categorical variables were synthesized with frequencies and percentages, continuous variables with averages and the measurement of the standard deviation (SD). The statistical association of the events, the graphical representation of the same ones and the evaluation of the significance had been carried out with MATLAB and Statistics Toolbox (The MathWorks, Natick, Massachusetts, US). Associations with a p-value < 0.05 were considered significantly different from zero.

Table 1 Baseline patients' characteristics

Number of Patients	82
CD	49 (59.8%)
UC	33 (40.2%)
Sex	42 M, 40 F
CD	30 M (61.2%)
UC	21 F (63.6%)
Age	
At diagnosis	11.8±3.5 years
At transition	20.1±2.7 years
Localization CD	
Ileal- n (%)	9 (18%)
Colic- n(%)	6 (12%)
Ileo-colic- n(%)	30 (61%)
Upper GI- n (%)	4 (8%) exclusive 8 (16%) in addition
Perianal Disease	15 (30.6%)
Phenotype Pattern CD	
Inflammatory (B1)	17 (34.7%)
Stricturing (B2)	24 (49%)
Penetrating (B3)	8 (16.3%)
Growth Failure	21 (42.9%)
Localization UC	
Ulcerative Proctitis (E1)	1 (3%)
Distal colitis (E2)	6 (18.2%)
Pancolitis (E3)	26 (78.8%)

Results

Patients' characteristics

Before transition

A total of 82 patients were enrolled, of which 57 came from the OPBG and 25 from the Policlinico Umberto I. Forty-two patients were males (51.2%) and 49 (59.8%) were affected by CD. The mean age at diagnosis was 11.8±3.5 years. Among 49 patients with CD, 32 patients (65.3%) had a penetrating or stricturing disease, 21 (42.9%) showed growth failure at the time of diagnosis and 15 (30.6%) had a perianal disease. Of 33 patients with UC, 26 (78.8%) had pancolitis.

Table 1 summarizes baseline patients' characteristics.

With regard to the medications taken before transition, 75% of patients received corticosteroids for more than 3 months during childhood. Fifty-six patients (68.3%) began an immunosuppressive therapy with thiopurines (still ongoing at transition only in 8 patients, 14.3%), and 9 patients received other types of immunosuppressants (cyclosporine, methotrexate, thalidomide).

Fifty-three patients (64.6%) started at least one biological therapy in pediatric age (that is ongoing in 62% of patients at time of transition) and 15 (28%) already tried ≥ 2 different types of biologics.

Up to the 40.2% of patients underwent one or more surgical intervention, more frequently for CD patients compared to UC (58% vs 15%, P=0.0003). Factor that associated with a higher risk of major surgeries occurred during the pediatric age have been a complicated (both stricturing and penetrating) CD (p=0.015), two or more types of biologics employed before transition (p=0.005) and a more severe clinical activity at diagnosis (p=0.001) (Fig. 1).

No significant difference ($p > 0.05$) has been found between patients coming from the two different centers in terms of mean age at diagnosis and transition, number and type of therapies and disease activity.

At time of transition

The average age observed at transition was 20.1±2.7 years, with an average time between the diagnosis and the beginning of transition to the adult center of 8.3±4.6 years. Only 22% of patients who carried out transition between ages 14 and 18 years were in a complete clinical remission phase, compared to the 60% of those who carried it out between ages 19 and 24 years (p=0.002, Fig. 2). No significant association was found between the age at transition and other parameters such as type of disease (p=0.89), previous surgery (p=0.44), clinical activity at diagnosis (p=0.78) and early onset (< 6 years) disease s (p=0.66).

Overall, 24% of patients carried out transition while on a moderate or severe disease activity of which 40% had already been treated with ≥ 2 biologics. 47.5% of patients had to change or start a new biologic therapy within 18

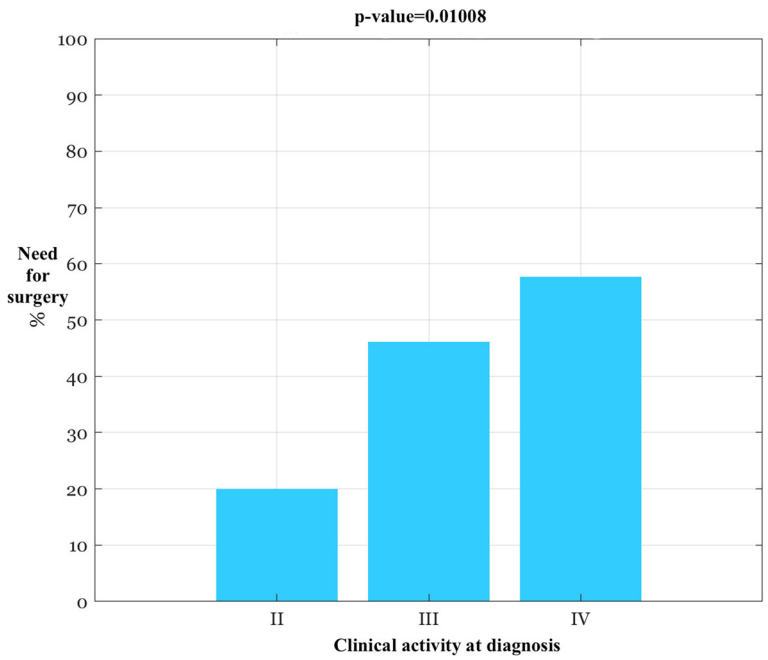

Fig. 1 Association between probability of undergoing surgery before transition and clinical activity at diagnosis (HBI and PMS)

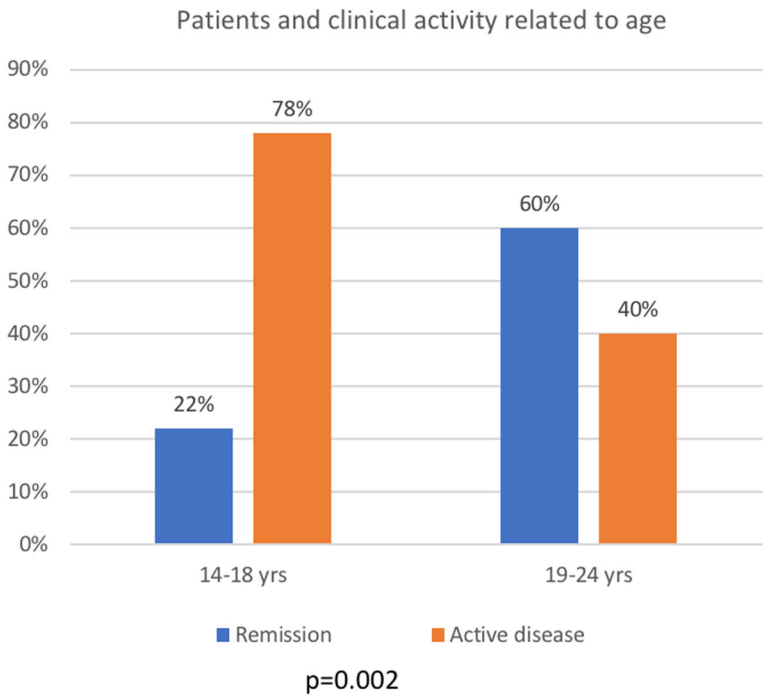

Fig. 2 Transitioned patients divided by age and clinical disease activity

months after transition, and 21% of patients needed IBD-related surgery within the first 2 years after transition.

Transition while on moderate to severe active disease was significantly associated with a worsening of symptoms during the first year of follow-up visits at the adult center (p=0.002).

On the other hand, even if the 76% of patients carried out transition in a clinical remission phase or mild activity of disease (HBI or PMS = I or II), just the 80% of them really confirmed their clinical well-being with a complete endoscopic remission, and the 7.3% of their endoscopies revealed a moderate to severe endoscopic activity. Conversely, a complete endoscopic remission has been found to be related to a clinical remission with a correspondence of the 96%.

Furthermore, 64% of total patients carried out transition on a both clinical and endoscopic remission, and still maintain this condition at follow-up visits.

Assessment patients' readiness and quality of life

A selected group of fifty-three of patients, comparable for age and disease's characteristics to the population of the study, answered the online anonymous questionnaires. Among them, up to 70% still go to follow-up visits at the adult center along with their parents, and 89% of them report that parents still have an important role in the management of therapy.

Table 2 summarizes patients' answers.

The mean value of the TRAQ questionnaires' scores collected is 3.4±1.5 out of 5, and the mean score of SIBDQ is 53.9±9.8 out of 70.

Considering as poor transition readiness a TRAQ score ≤ 3 out of 5, and as a good HRQoL a SIBDQ score > 50, significant association was found between a TRAQ mean score > 3 and a SIBDQ > 50 (p=0.0129). In fact, the 80.5% of patients with TRAQ mean score > 3 reported scores > 50 of SIBDQ, compared to the 41.7% of those with TRAQ mean score ≤ 3 (Fig. 3).

Conversely, considering patients referring a good HRQoL (SIBDQ > 50), the 86.8% reported TRAQ scores > 3, against the 13.2% that reported scores ≤ 3(Fig. 4).

No significant differences were observed among TRAQ mean scores of patients who completed transition within the age of 19 (3.3±0.5) compared to those who passed after 20 years (3.4±0.5), with p=0.36.

Overall, 75% of patients have a positive opinion of the transition model adopted. According to the answers collected, the three most important aspects for an ideal transition for patients are: timing of transition in a phase of stable remission (52% of patients), the access to scheduled joint visits with both pediatricians and adult gastroenterologists (49%) and the presence of psychologist and nutritionist figures in the adult center (34%). Less important, according to the patients' opinion: the short distance between the two centers (24%), the possibility to choose independently the adult gastroenterologist (9%) and the protraction of transition for longer than 1 year (8%).

Table 2 Transition Readiness Assessment Questionnaire (TRAQ) answers

Questions	Answers' Mean Value (out of 5)
1. Do you know what to do if you are having a bad reaction to your medications	2.5 ± 1.2
2. Do you take medications correctly and on your own?	4,1 ± 1.1
3. Do you reorder medications before they run out?	4.6 ± 0.8
4. Do you call the doctor's office to make an appointment?	3.2 ± 1.5
5. Do you follow-up on any referral for tests, check-ups or labs?	4 ± 1.2
6. Do you arrange for your ride to medical appointments?	2.2 ± 1.6
7. Do you call the doctor about unusual changes in your health (For example: Allergic reactions)?	3.9 ± 1.3
8. Do you know what your health insurance covers?	3.3 ± 1.1
9. Do you manage your money & budget household expenses (For example: use checking/debit card)?	2.7 ± 1.4
10. Do you fill out the medical history form, including a list of your allergies?	4.5 ± 0.7
11. Do you keep a calendar or list of medical and other appointments?	1.8 ± 1.2
12. Do you make a list of questions before the doctor's visit?	1.9 ± 0.7
13. Do you tell the doctor or nurse what you are feeling?	4.1 ± 1.2
14. Do you answer questions that are asked by the doctor, nurse, or clinic staff?	4.8 ± 0.7
15. Do you help plan or prepare meals/food?	3.3 ± 1.3
16. Do you keep home/room clean or clean-up after meals?	3.1 ± 1.3
Total mean score	3.4 ± 1.5

Legend: 1 - No, I do not know how. 2 - No, but I want to learn. 3 - No, but I am learning to do this. 4 - Yes, I have started doing this. 5 - Yes, I always do this when I need to

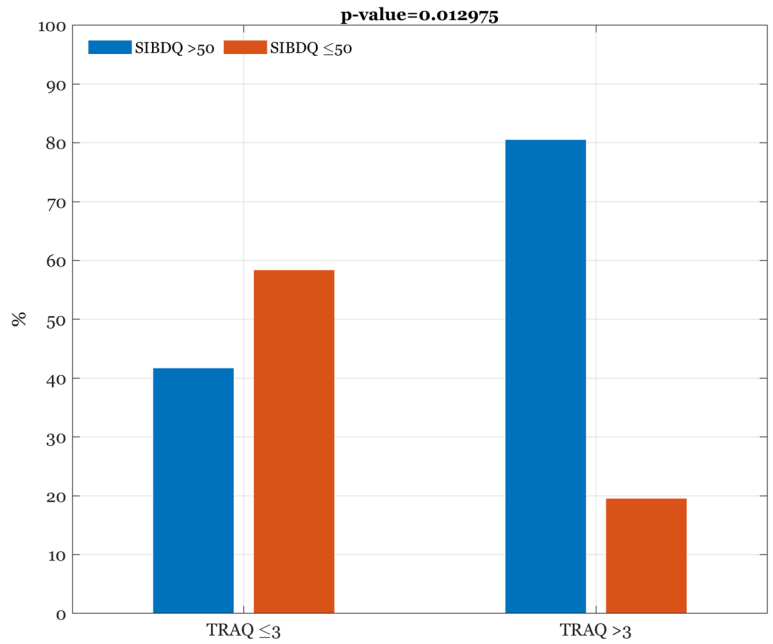

Fig. 3 Association between TRAQ and SIBDQ scores

Forty percent of patients believe that the disease has delayed their educational and professional pathway, and 90% of patients who underwent surgery believe that surgery has overall improved their quality of life. The 88% of patients say they feel better now than they did in pediatric age, and 70% report an actual clinical well-being. The 45% of patients finally reported the presence of other cases of IBD in the family.

No significant difference ($p > 0.05$) has been found between patients coming from the two different centers in terms of questionnaire's answers or short and long-term outcomes during follow-up visits.

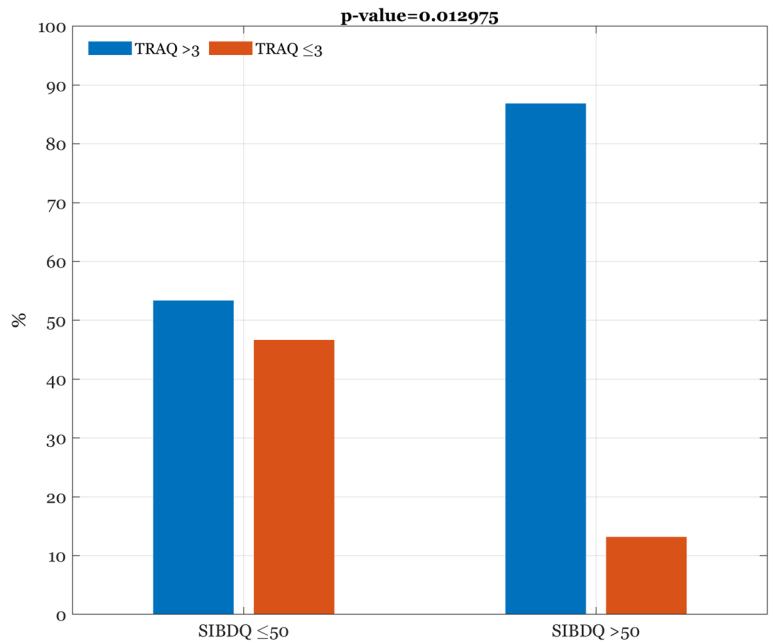

Fig. 4 Association between SIBDQ and TRAQ scores

Discussion

We have explored the outcome of the transition process of 82 consecutive IBD patients coming from two pediatric referral hospitals to one IBD adult university hospital center, their readiness and the consequent impact on quality of life. Our model included two combined pediatric–adult visits, the first one at the children's hospitals, in presence of parents, and the second one at the adult center. Despite there is no evidence that this model of transitional care is superior over others, the "joint-visit" model has been shown to be associated with a higher effectiveness on transmitting clinical data and building confidence in adults' clinicians [14].

A recent study has been conducted on the transition process from pediatric to adult care of 106 young patients affected by IBD, investigating the 1-year success outcomes of this process [15]. This study has shown a significant maintenance or even a reduction of the disease activity scores and number of exacerbations in those patients who were in a remission phase at the time of transition. Our results confirm this data, strengthening the importance of the right timing of transition to the adult center.

For this reason, no strict chronological criteria have been considered in order to plan the timing of transition, and a case-by-case selection, according to IBD pediatricians' judgment, has been used when possible.

The mean age observed at transition in our cohort was 20.1±2.7 years, exceeding the recommended ideal interval of the age of 17 and 19 [6]. In addition to this, patients who carried out transition with a moderate to severe disease activity have been resulted to be younger than those who transited with a mild or absent one (average age of 19.3±2.4 years vs. 20.2±2.5 years). This fact could be attributed to a lower engagement towards transition process made by patients and caregivers in those cases that did not present symptoms.

For this reason it can be also assumed that patients who transit on a younger age, and who often reach the adult hospital in need for hospitalization and surgery, making the experience of the transition even more traumatic, could be considered at higher risk of a further clinical deterioration in the first years of adulthood, and should need greater clinical care in order to obtain the best possible outcome in terms of quality of life, fundamental parameter to guarantee a good compliance to therapy in the future.

Data collected also show that main risk factors linked to a higher rate of surgery within the first year from transition can be considered the CD phenotype (particularly B2 and B3 behavior phenotypes), the number of biologics drugs assumed during the pediatric age, the clinical activity at diagnosis and the absence of an endoscopic or clinical remission.

An active disease at the time of transition has been already associated with an unsuccessful transition [16]. According to this, patients should complete transition to the adult center while in clinical remission or mild disease activity. Conversely, the transition of active patients should be rather delayed until the resolution of the acute phase, in order to avoid psychological traumatisms and to promote a greater confidence in the adult specialist and a good compliance with the therapy management [17].

Furthermore, it has been recently observed in an US cohort the absence of association between TRAQ scores and different measures of health [18]. In our cohort, instead, we found a significative positive association between TRAQ and SIBDQ questionnaires, highlighting the importance to assess a real readiness and demonstrating the association between patients' transition readiness and higher HRQoL.

This fact gets even more important considering that we did not observe significant differences in terms of TRAQ mean scores among patients of different age, ant that we found more or less the same level of independence and awareness of disease within different groups, proving that transitions carried out at older ages do not improve readiness and quality of life score in significant way.

Our patients' cohort expressed generally satisfaction about the transition process adopted, in particular about the opportunity to join combined pediatric-adult visits. Parents seem to still play a central role in patients' lives and in their relationship with disease, especially for managing scheduled appointments, medications (including also keeping in memory of previous side effects or intolerance) and talking with providers.

This confirms the urgent need to structure educational programs during childhood, including also parents, in order to stimulate the acquisition by patients of a greater independence and capability of self-management.

Our study has some limitations, of which main ones can be considered the inclusion of two pediatric centers, with possible different therapy approaches, the single model of transition analyzed and the low percentage of children in remission enrolled between patients who carried out transition within the recommended age of 19 years.

The first limitation could lead to some biases related to the lack of homogeneity in the management and follow up, even no significant differences have been found in terms of short and long-term outcomes. Patients enrolled were similar in terms of age, and the same associations have been found in those who carried out transition in an active phase of disease, even if coming from different centers. On the other hand, our study could be considered a realistic model where patients

with different backgrounds can converge to a single adult center through a standardized path.

In conclusion, our data support that mild activity of disease at transition and a high level of adolescent's autonomy skills are the most important aspects of considering for transition and for assuring a better HRQoL in this difficult phase of care.

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None.

Authors' contributions
AC, DP and AA made substantial contributions to conception and design, acquisition of data and analysis and interpretation of data. AC and DP wrote the manuscript. AA, LG and AG reviewed and revised the manuscript. FB, DK, BP, MA and SC were involved in the recruiting of patients. All authors read and approved the final manuscript.

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Availability of data and materials
The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

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The study was approved by the Ethical Committee of the centers involved. The procedures were in accordance with the ethical standards of the Declaration of Helsinki.

Consent for publication
All authors have read and approved the content and agree to submit for consideration for publication in the journal.

Competing interests
The authors declare that they have no competing interests.

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Publisher's Note

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ORIGINAL RESEARCH

Phenotypes in adult patients with Rett syndrome: results of a 13-year experience and insights into healthcare transition

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ABSTRACT

Background Rett syndrome is a complex genetic disorder with age-specific manifestations and over half of the patients surviving into middle age. However, little information about the phenotype of adult individuals with Rett syndrome is available, and mainly relies on questionnaires completed by caregivers. Here, we assess the clinical manifestations and management of adult patients with Rett syndrome and present our experience in transitioning from the paediatric to the adult clinic.

Methods We analysed the medical records and molecular data of women aged ≥18 years with a diagnosis of classic Rett syndrome and/or pathogenic variants in *MECP2*, *CDKL5* and *FOXG1*, who were in charge of our clinic.

Results Of the 50 women with classic Rett syndrome, 94% had epilepsy (26% drug-resistant), 20% showed extrapyramidal signs, 40% sleep problems and 36% behavioural disorders. Eighty-six % patients exhibited gastrointestinal problems; 70% had scoliosis and 90% low bone density. Breathing irregularities were diagnosed in 60%. None of the patients had cardiac issues. CDKL5 patients experienced fewer breathing abnormalities than women with classic Rett syndrome.

Conclusion The delineation of an adult phenotype in Rett syndrome demonstrates the importance of a transitional programme and the need of a dedicated multidisciplinary team to optimise the clinical management of these patients.

INTRODUCTION

Rett syndrome (RTT, OMIM #312750) is one of the most common causes of intellectual disability (ID) in females, with an incidence of 1 in 10 000 .[1] According to the 2010 diagnostic criteria,[1] typical RTT is defined by the presence of regression of purposeful hand use and spoken language, and development of gait abnormalities and hand stereotypies. In addition to the core neurodevelopmental manifestations, affected girls often show variable degrees of autonomic dysfunction,[2] growth retardation, gastrointestinal (GI) discomfort[3] and epilepsy.[4] The clinical outcome is frequently complicated by variable motor impairment,[5] skeletal abnormalities such as scoliosis,[6] low bone density and increased risk of fractures.[7]

Approximately 60% of individuals with RTT from large cohorts from North America[8] and Australia[9] have been reported to potentially survive into middle age. Clinicians are therefore increasingly faced with managing the complexity of RTT in adulthood. However, little information has been available about these patients' health status, and most studies were conducted using questionnaires completed by caregivers. In 2013, researchers from Maastricht University together with the Dutch Rett Syndrome Association reported a slow and progressive deterioration of gross motor functioning in contrast to better preserved cognitive functioning, less autonomic and epileptic features and overall good general health in adult patients.[10] In 2010, we evaluated the main clinical features and health status of adult Italian patients with RTT, and identified sleep, behavioural issues, autonomic disorders, GI and musculoskeletal problems as major problems, whereas epilepsy tended to improve.[11] The major limitation of our previous study was that data were obtained from the patients' parents in most cases, introducing a bias in the interpretation of some clinical details. Nevertheless, that study taught us that the complex phenotype of individuals with RTT requires more careful and extensive medical care, although guidelines for clinical management of adolescents and adults with RTT are not available. The majority of adults with RTT are treated in paediatric centres because specialised adult clinics are rare.

At our hospital, we have established a transitional programme from paediatric to adult care for girls with RTT. In this paper, we will discuss the functioning of our specialised adult RTT clinic, and present the results of a study that investigated the medical issues in adults 13 years after the clinic was established. Starting from our clinical observations, we will discuss the major healthcare issues described thus far in women with RTT and propose a follow-up protocol for these patients in adulthood.

MATERIALS AND METHODS

Rett clinic

At San Paolo University Hospital in Milan (Italy), a group of physicians has developed experience in caring for patients with RTT, and a multidisciplinary Rett clinic was established in 2006. Overall, we have been caring for about 130 patients, of whom half are paediatric and half adults.

A paediatric neurologist and a paediatrician coordinate clinical care for children up to age 18

years, and a neurologist is in charge of the adult clinic. These physicians also serve as clinical managers and schedule clinical encounters based on individual needs and following the local guidelines on surveillance and management (http://malattierare.marionegri.it/images/downloads/PDTA/PDTA_schede/sclerosi_tuberosa.pdf). In addition to neurologists and paediatricians, the multidisciplinary team consists of cardiologists, internal medicine doctors, a general surgeon, a clinical geneticist, a physical medicine doctor and a gynaecologist, while other specialists are involved only when needed. A medical unit called Disabled Advanced Medical Assistance (DAMA) has dedicated spaces and personnel (internal medicine, general surgery and registered nurses) for individuals with ID and complex disabilities, and a phone line and fast track to the emergency room for these patients. All the patients with ID who are seen at DAMA also receive a yearly dental visit.

Ours is currently the only adult RTT clinic in Northern Italy, and receives referrals from several other paediatric clinics when patients turn 18. We have a formal programme for transition and transfer of care that takes into account external referrals and ensures continuity of care for those paediatric individuals who are already known to our staff.

Based on the model of the tuberous sclerosis complex clinic,[12] individuals with RTT are usually seen as outpatients (children every 6 months, adults yearly), and we try to schedule all specialty visits during 1 or 2 days. The clinical manager performs the last visit, reviewing all notes from other specialties' visits, discussing the results and making a follow-up plan. He/she keeps contact with family doctors, therapists and patient organisations to coordinate care. After each clinical encounter, the results of each visit are transferred into an ad hoc database, which we use for research.

Patients' data

We included in the study all patients aged ≥18 years who were in charge to our Rett clinic (mostly since it was established) and who received the last medical visit between January 2018 and December 2019. The diagnosis of RTT was made according to the diagnostic criteria in use.[1] We recorded demographic and genetic information for each patient, and evaluated major clinical issues and interventions.

RESULTS

We included in this study 56 adult patients with RTT. Median age was 29 years (range 19–49). Forty-seven out of 56 (84%) had a pathogenic variant in *MECP2* (all except four had a diagnosis of classic RTT: one with preserved speech variant, and three with early onset seizure variant). Two patients carried a pathogenic variant in *CDKL5*, and two in *FOXG1*. The characteristics of these patients are summarised in table 1.

Two patients were found to have pathogenic variants in genes encoding GABAa receptor subunits (*GABRG2* and *GABRB2* respectively, one with atypical RTT and one with typical RTT), and have been recently described elsewhere.[13] No pathogenic variants were detected in two individuals with a clinical diagnosis of classic RTT, and testing was not available in another one with classic RTT.

For consistency, we focused our analysis on the 50 patients with *MECP2* pathogenic variants or a clinical diagnosis of typical RTT.

The main medical problems of the adult cohort are represented in figure 1. With regard to motor ability, 27/50 girls (54%) had lost the ability to walk independently, 18/50 (36%)

Table 1 Clinical characteristics of adult patients with *CDKL5* and *FOXG1* pathogenic variants

Patients	Mutation type	Age (years)	Independent walking	Stereotypies/Movement disorder	Epilepsy/ Drug resistance	Sleep disturbances	Behavioural disorder	Gastrointestinal problems	Osteoporosis	Scoliosis	Breathing issues	Cardiac problems
CDKL5 (NM_001323289.2)	c.1648C>T; p.(Arg550Ter)	19	–	+/–	+/+	+	–	+	+	+	–	–
CDKL5 (NM_001323289.2)	c.607G>C; p.(Glu203Gln)	47	–	+/–	+/–	–	–	+	+	+	–	–
FOXG1 (NM_005249.3)	c.256delC; p.(Gln86ArgfsX106)	19	–	+/+	+/–	–	–	+	+	+	–	–
FOXG1 (NM_005249.3)	2 Mb 14q12 deletion (28 780 663–30 780 833; hg19), de novo	44	–	+/–	+/–	–	–	+	+	+	–	–

+: present; –: absent.

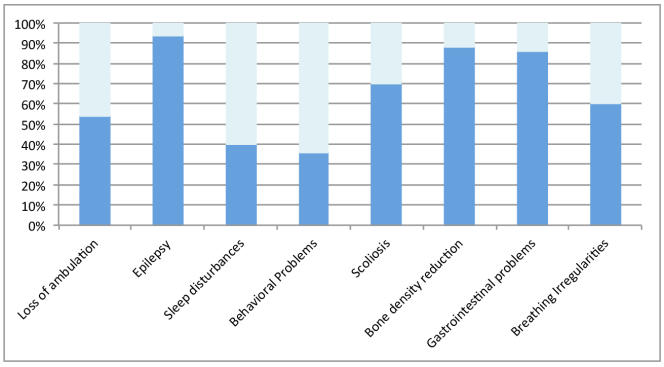

Figure 1 Bar graph showing the medical issues of our cohort of 50 adult patients with classic Rett syndrome. Dark blue: percentage of patients presenting specific problems; light blue: percentage of patients without the specific problems.

were able to walk with support, while 5/50 (10%) were able to walk unattended. As expected in Italy, where individuals with ID are usually taken care of at home, only two women from the present cohort lived in an assisted living facility or nursing home.

Probably due to the bias that our centre is dedicated to epilepsy, the most prevalent disturbances were related to neurological problems: epilepsy was diagnosed in 47/50 (94%), and was drug resistant in 12/47 (26%) (the pathogenic variants found in these patients are listed in online supplemental table S1), with 10 patients (21%) having monthly seizures. All but five patients were on antiepileptic drugs (AEDs). Ten out of 50 (20%) showed extrapyramidal signs, such as dystonia and tremor. Hand stereotypies were evident in all the patients.

Sleep disturbances were diagnosed in 20/50 (40%), and highly interfered with family habits because of frequent nocturnal awakenings in 7/50 (14%). They were usually treated with melatonin. In one patient mirtazapine was used, and in another one gabapentin was prescribed.

Behavioural problems, such as irritability, agitation or crying/screaming outburst, were recorded in 18/50 (36%). Six patients were treated with antipsychotic drugs, mainly risperidone. When suspected, depression was treated with sertraline or quetiapine.

The great majority of our patients have muscular-skeletal problems. Scoliosis was diagnosed in 35/50 (70%). Of these patients, 8 received surgery, 14 needed bracing and the remaining did not show a scoliosis that was severe enough to need treatment. The patients with severe scoliosis were wheel chaired. All but six (88%) had low bone density at densitometry. For these issues, almost all patients received calcium and vitamin D supplementation, but none of them was treated with bisphosphonates.

GI disturbances were seen in 43/50 (86%): constipation in 42/50 (84%), gastro-oesophageal reflux (GERD) in 17/50 (34%); two required Percutaneous Endoscopic Gastrostomy (PEG) and 3/50 (6%) experienced cholelithiasis. Assessment of GI issues was generally clinical, and treatment consisted of a combination of behavioural indications (feeding strategies and posture for GERD; adequate fibres and fluid intake for constipation) and pharmacological therapy (proton-pump inhibitors or domperidone for GERD; psyllium or oral osmotic laxatives for constipation).

Regarding autonomic system disturbances, 30/50 (60%) women exhibited breathing irregularities. Most of them (27/50, 54%) experienced apnoeas, 12/50 (24%) associated with hyperventilation and 7/50 (14%) hyperventilation only.

Heart rate disturbances or other cardiac issues were not identified in any women from this cohort.

Interestingly, 6/50 (12%) women suffered from recurrent urinary tract infections.

Menstrual irregularities were frequently reported, although they did not usually require specific treatments. In two women were ovarian cysts detected, and surgery was indicated in one case.

DISCUSSION

Although RTT is characterised by a recognisable phenotype, the clinical manifestations associated with this syndrome vary with age.[10 11] Recently, an increasing number of studies have started to assess medical morbidities of adult women with RTT,[8 9] but results are not consistent and the Italian population—and a Southern European cohort, more generally speaking—has been rarely evaluated. To better understand the medical problems and to evaluate the efficacy of our transitional model, we conducted a clinical study based on chart reviews of the adult patients with RTT from our clinic (in contrast with previous studies based on surveys completed by caregivers).

Here, we describe the main problems experienced by adults with RTT and potential therapies to address them.

Patients with a clinical or molecular diagnosis of classic RTT (*MECP2*)

Neuromotor features

Motor features are widely compromised in RTT, with variable degrees of impairment related to genotype and age.[14] In our cohort, only 10% of women kept their ability to walk independently. The women who maintained relative good motor performances (eg, walking, climbing and go down the stairs) had presented with a mild neurological phenotype since their childhood: they never had seizures or epilepsy was easily controlled, and they were able to keep objects in their hands for a while. This finding is in line with the report that girls carrying pathogenic variants associated with a milder phenotype achieve better gross motor scores, and some maintain the ability to walk when adults.[9]

Interestingly, neurological impairments such as bradykinesia, dystonia and parkinsonism can be frequently seen in older women, and were also present in our sample in 20% of the women, whereas stereotypic hand movements tend to persist throughout life,[15] and were evident in all the women, with different characteristics. However, no clear correlations were found between dystonia and ambulation. Several patients with movement disorders (dystonia, tremor, parkinsonism) maintained their ability to walk.

Epilepsy

Epilepsy is common in RTT, with prevalence varying between 60% and 90%, although seizure frequency might be overestimated due to the difficulty in differentiating seizures from non-epileptic paroxysmal events.[4] Epilepsy onset usually occurs around 3–5 years of age, but seizures can appear even after 10 years of age. This information should be kept in mind when caring for adolescent and adult patients with RTT.[16]

Since our centre has a long history in treating patients with complex epilepsies, women with RTT followed at San Paolo Hospital probably exhibit a worse neurological phenotype. Indeed, epilepsy was present in 94% of our patients. Seizures were drug resistant in 26%, in line with wider studies involving patients at all ages.[16]

Although previous works demonstrated significant improvement in seizure disorder after the age of 20 years,[10] recent studies showed that the lifetime course of epilepsy is higher than previously reported.[17] Seizure remission is not so common, and only a small percentage of individuals are seizure-free and off AEDs.[11] Moreover, patterns of relapse/remission over the life span are emerging,[4] and modifications of the drug regimens are often required to achieve seizure control. Therefore, clinicians should also consider age-dependency when prescribing appropriate antiepileptic drugs in RTT. We recently reported evidence that valproic acid could be the most effective anti-epileptic drug in younger girls, and carbamazepine in patients aged 15 years or older.[18]

In general, no specific *MECP2* mutation is significantly associated with either seizure prevalence or severity, while higher Clinical Severity Scores and nutritional/growth problems are frequently related to epilepsy.[4]

Sleep disturbances

Sleep problems are present in nearly all patients with RTT.[19] In our previously published cohort, sleep disorders were reported in the majority of adult patients (77%), even though they were considered a mild problem by the parents.[11] Considering the most frequent sleep disorders in relation to ageing, the prevalence of night laughing and screaming tend to decrease with age, while waking remains similar across age groups.[20] In the present cohort, 40% of women exhibited sleep disturbances, and in 14% of them this problem had a remarkable impact on daily habits. This number is consistent with the study by Wong *et al*,[20] who found a decreased prevalence of sleep problems with age.

Regarding therapeutic management of sleep disorders, experience from the Australian group suggests that melatonin is used in <5% of cases and provides only little benefit to the patients.[20] Another Australian study showed the importance of sleep hygiene strategies, such as environment modifications, sleep practices and physiological factors, to reduce sleep disorders in individuals with RTT.[19] If behavioural interventions are inadequate, clinicians should consider the use of medications including chronobiotic regulators such as melatonin, or GABA agonists (eg, gabapentin, mirtazapine).

Behavioural problems

Individuals with RTT have difficulties in communicating symptoms and problems of which caregivers may not be aware. Behavioural problems might therefore be a symptom of a health-related disorder, for which a complete clinical evaluation is mandatory.

In our sample, a third of women with RTT presented behavioural disturbances, which could not be related to health problems or environmental situations despite careful assessment, and were mainly characterised by agitation, crying and irritability. Sudden mood changes or periods of low mood have been described in adults with RTT.[10 11] Cianfaglione *et al*[21] reported significant deterioration in mood as individuals aged. Therefore, depression should be considered as a possible comorbidity in women with RTT that requires identification and appropriate management.

Lastly, also agitation and anxiety have been described in adults with RTT, with the latter improving in women older than 30 years of age.[10]

Scoliosis

As expected and in line with previous studies, which show that up to 75% of girls receive a diagnosis of scoliosis by the age 15 years,[6] 70% of women from our cohort had scoliosis. Protective effects of walking ability[22] and milder genotype (such as the p.R133C, p.R294X and p.R306C pathogenic variants) have been demonstrated.[23]

In order to monitor the progression of scoliosis, 6 monthly plain X-rays are suggested if the Cobb angle is >25° before skeletal maturity and 12 monthly X-rays after skeletal maturity until evidence of no further progression, following a specific protocol.[24]

Although not proven to modify spine curvature in RTT, bracing is recommended in order to reduce the progression of scoliosis when curves reach 25°, while surgery should be considered when the Cobb angle is approximately 40° to 50°.[24] Surgical outcomes may improve if intervention is done before the development of severe scoliosis and before the effects of other comorbidities, such as age-related reduction in mobility. Additional non-surgical approaches, such as intensively structured physical therapy environment, have been suggested to improve the Cobb angle,[25] but need further evaluations.

Bone health

Although altered bone mineral density has been reported in about 50% of individuals with RTT, low bone density was documented on densitometry in around 90% of the women in our cohort. We may speculate that this finding can be explained by the characteristics of the sample, with an over-representation of patients treated with antiepileptic drugs and neurologically impaired. Indeed, anticonvulsant therapy, especially with valproate, is an additional risk factor for low bone density and increases fracture risk by threefold in RTT, compared with no or any other prescribed antiepileptic drugs. Also, non-ambulatory patients have a decreased bone mass.[26]

Patients with lower bone density are exposed to a higher risk of fractures, occurring much more frequently in RTT than in the general population. Fractures often occur spontaneously, after minimal trauma or a fall, and predominantly in the upper/lower limbs long bones.

A panel of experts in RTT has recently developed clinical guidelines for the management of bone health.[7] The guidelines are divided in two sections: clinical and bone density assessment, and non-pharmacological and pharmacological intervention strategies. DEXA scans should be used to assess baseline bone mineral density, and monitoring should be done every 1–2 years depending on clinical presentation, and should consider Z-score results as well as previous occurrence of fractures. The increase of physical activity or at least encouragement of supported standing are recommended as non-pharmacological interventions, and the increase of dietary intake of calcium-rich or calcium-fortified food and vitamin D supplementation are advised.[7] While no clear evidence of benefit from the use of bisphosphonates exists, new potential treatment with zoledronic acid in RTT has been recently suggested as it resulted in an increase in bone density and a reduction in the incidence of fractures.[27]

Gastrointestinal issues

It is widely known that GI and nutritional problems persist throughout life in girls and women with RTT.[3] However, the type of problems seems to differ between children and adults. Affected women are significantly less likely to present with vomiting or regurgitation and GERD than children. On the

other hand, underweight, prolonged feeding time and swallowing difficulty—thus considering the need of gastrostomy placement—are typical of older individuals.[3] As expected, also in our cohort constipation was one of the most frequent complaints, experienced by 84% of women, while GERD only by 34%.

Interestingly, we diagnosed cholelithiasis, or gallbladder disease, in a small number of patients. This finding has been recently proven to be relatively frequent in RTT, and should be suspected as one of the causes of abdominal pain.[28]

An international consensus developed guidelines on assessment and management of GI disorders in RTT,[29] with specific focus on GERD, constipation and abdominal bloating. For these issues, conservative strategies together with pharmacological/surgical interventions are recommended.[29]

Management of growth and nutritional aspects is often challenging in RTT, and needs to consider feeding difficulties and nutritional needs. Although no clear recommendations for affected adults have been established, risk of aspiration and prolonged feeding times are factors that should be considered for gastrostomy indication.

Breathing problems

Caregivers reported breathing disturbances in approximately two-thirds of RTT girls in a worldwide-based cohort[2] and in almost all patients with typical RTT in the USA,[30] with onset usually occurring during early childhood. The prevalence of hyperventilation decreases over time,[10] while breath holding tends to persist, as seen in our cohort. This finding can be explained by the lower survival rate of individuals with more severe respiratory problems.[2] Based on the American natural history study,[30] one of the most striking findings is the strong association between prolonged corrected QT interval and severe breathing dysfunction, which links to increased risk for sudden death.

The majority of patients do not receive any treatment for their breathing abnormalities, although the use of topiramate, acetazolamide, buspirone and fluoxetine has shown some benefits.[2]

Besides autonomic breathing disturbances, individuals with RTT may experience more complex respiratory dysfunctions. Lower airway inflammation and ventilation and perfusion mismatch are thought to be the mechanisms involved, leading to lower respiratory tract infections that often require hospitalisation. Walking seems to have protective effects on respiratory health, beyond the influence of specific *MECP2* variants, suggesting that the maintenance of activity programmes should be supported, especially during adolescence and adulthood.[31]

Cardiac issues

Of note, no cardiac problems were present in our cohort, supporting the findings that heart problems may not be as common as previously thought.[32] Individuals with RTT had been reported to show ECG and rhythm abnormalities including prolonged corrected QT interval (QTc) and reduced heart-rate variability.[33] However, more recent findings demonstrated that the prevalence of QTc prolongation in RTT is lower than previously thought and around 7%.[32] No correlation between QTc and genotype was identified, but a positive trend towards a slight increase of QTc with age was reported. However, the clinical significance of QTc prolongation remains unknown.[33] To date, no specific consensus has been established, and treatment is based on individual needs.

Gynaecological issues

Little is known about gynaecological problems in people with RTT. Longitudinal population-based data showed that, at 14 years of age, the median age of menarche in girls with RTT was slightly older than in the general population. This finding could be probably related to body mass index and to genotype.[34]

Menstrual disturbances in women with RTT from our cohort were similar to those in the general population. Hormonal contraception may be used, in forms of oral pills or depot medroxyprogesterone. Dysmenorrhoea can be treated with non-steroidal anti-inflammatory drugs at appropriate doses, since the communication disability can make it difficult to evaluate the severity of pain experienced by these girls and women.

Patients with *CDKL5* pathogenic variants

Although patients carrying mutations in the X linked cyclin-dependent kinase-like 5 gene (*CDKL5*, chr. Xp22.13, OMIM # 300203) were originally diagnosed as having the early onset seizure variant of RTT,[1] recent evidence suggests that this disorder should be considered a distinct clinical entity.[35] One of the major medical issues in these individuals is epilepsy, which is almost universally present and frequently drug-resistant, also in adulthood, as in our two patients. Sleep difficulties as well as GI problems seem to be very common in the *CDKL5* disorder. Scoliosis tends to develop with increasing age, while our experience confirms that breathing abnormalities seem much less prevalent than in classic RTT.[36]

Patients with *FOXG1* pathogenic variants

Since the identification of *FOXG1* (chr. 14q12, OMIM # 164874) as the gene responsible for the congenital form of RTT,[1] the combination of neurodevelopmental and brain imaging features observed in individuals with *FOXG1* pathogenic variants may be sufficiently distinctive to allow clinical recognition of this disorder, considered as the *FOXG1* syndrome.[37] Less than 10 adults with *FOXG1* mutations have been described so far, mainly within large series of patients.[37–39] Their phenotype is characterised by severe postnatal microcephaly, severe ID, absent language, autistic traits, epilepsy[38] and a typical hyperkinetic-dyskinetic movement disorder.[40] Poor sleep patterns, irritability in infancy, unexplained episodes of crying, recurrent aspiration and GERD have also been reported.[37] To date, no specific studies have evaluated the morbidities of these individuals in adulthood. The two women with *FOXG1* pathogenic variants in our cohort experienced epilepsy, but seizures were controlled by AEDs. GI problems, scoliosis and severe osteopenia with fracture were the main problems in our two patients, in addition to ID.

Strengths and limitations

Although smaller in sample size and retrospective in nature, this is one of the first studies that evaluated medical morbidities in adult women with RTT by medical records review in a specialised clinic instead of by surveys completed by caregivers, thus possibly providing more accurate data. The results of this preliminary assessment confirm the utility of our multidisciplinary clinic, which ensures that all the main comorbidities are addressed. On the other hand, the identification of previously underestimated medical issues allowed us to understand what requires to be implemented, such as the presence of a sleep medicine specialist who has now joined the clinic.

Despite having some pitfalls and facing limitations in economic and staff resources, we try to target patients' needs and family's demands, adjusting international guidelines to the Italian reality.

Table 2 Surveillance and management recommendations for adult patients with RTT

Organ system or specialty area	Recommendation
Genetics	Offer genetic testing and family counselling, if not done previously.
Brain	Assess neurological features at each clinical visit at least annually. Sudden changes in behaviour should prompt medical/clinical evaluation to investigate potential medical causes. Be aware of the high prevalence of non-epileptic manifestations (eg, dystonia, stiffness, behavioural changes that could be associated with abdominal pain). Ascertain the presence of sleep disorders with the caregivers and plan additional exams when needed (eg, polysomnography).
	Routine EEG should be performed in individuals with known or suspected seizure activity. The frequency of routine EEG should be determined by clinical need rather than a specific defined interval. Prolonged video EEG, 24 hours or longer, is appropriate when seizure occurrence is unclear or when unexplained sleep or behavioural changes, or other alterations in neurological function are present.
Bone	Monitor bone mineral density every 1–2 years.
	Perform X-ray of the spine and hips to monitor scoliosis progression and investigate potential hip dislocation, based on clinical evaluation.
Gastrointestinal (GI)	Offer clinical evaluation for GI and nutritional problems annually.
	Perform 24 hours diet recall if nutritional indexes are abnormal.
	Request abdominal ultrasound based on clinical judgement (eg, persistent pain).
Teeth	Perform a detailed clinical dental exam at least every 6–12 months.
Heart	Obtain ECG every year in asymptomatic patients of all ages to monitor for conduction defects. More frequent or advanced diagnostic assessment may be required for symptomatic patients or patients with QT prolongation (>450 ms) or cardiac arrhythmias.
General assessment	Perform annually: complete blood count, CRP, electrolytes, albumin and pre-albumin, 25-hydroxy-vitamin D and parathyroid hormone levels, folic acid, vitamin B_{12}, total proteins and protein electrophoresis, ferritin, transferrin, serum iron, kidney, liver, thyroid function and other hormonal assessment, antiepileptic drug serum levels (if the patient is treated for epilepsy).
Gynaecology	Perform a gynaecology visit every 1–2 years. At least one pelvic ultrasound after puberty is recommended.*
	Consider performing pelvic ultrasound annually to look for ovarian cysts when the patient is taking valproic acid.
	Request specialty evaluation if menstruation irregularities or dysmenorrhoea are reported.

*According to the guidelines of the Italian Ministry of Health.
CRP, C-reactive protein; EEG, electroencephalograph; RTT, Rett syndrome.

This has resulted in the creation of local protocols, as seen in table 2, which we propose to the international community. Finally, women with RTT—like women with ID in general—may face several barriers in accessing preventive cancer screening (eg, breast and cervical cancer). Clinicians generally focus more on acute problems or chronic disorders associated with the specific syndrome than to age-related screening programmes. Indeed, these issues should be always kept in mind, especially for individuals with communication disability.

In conclusion, we have discussed the main clinical issues that should be considered when transitioning patients with RTT to adult care, based on data from our study and from the literature. We underline the importance of establishing a multidisciplinary team and educating the adult team members about RTT. Larger international studies assessing the medical problems of adult women with RTT and experiences from other adult clinics are highly encouraged.

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Ethics approval Patients described in the study have been followed at the Epilepsy Center of the San Paolo Hospital, University of Milan (Italy). Data were obtained from a dedicated database of patients with RTT, and informed consent to participate was obtained from the patients' parents. The Ethics Committee of San Paolo Hospital, Milan approved the study (2019/ST/098).

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Transition of care in pediatric oncohematology: a systematic literature review

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Abstract. *Background:* The transition of medical care from a pediatric to an adult environment is a psychological change, a new orientation that requires a self-redefinition of the individual, to understand that changes are taking place in his life. Up to 60 percent of pediatric patients who transition to adult services will experience one or more disease or treatment-related complication as they become adults. A nurse who knows how to recognize potential barriers at an early stage can play a pivotal role in the educational plan for the transition process. *Materials and methods:* A literature search was undertaken of PUBMED, CINAHL and The Cochrane Library, with specific inclusion and exclusion criteria, including articles published in the lasts ten years.This literature review has been performed according to the PRISMA statement. *Results:* Using the keywords in different combination 38 articles were found in The Cochrane Library, 5877 in PUBMED, 274 in CINAHL. 88 articles were selected after the abstract screening. 31 after removing the duplicates and reading the full text. *Discussion:* The main themes surrounding transition of care that emerged from the synthesis are the organization of care within common models of transition, innovative clinical approaches to transition, and the experience of patients and caregivers. The transition from pediatric to adult care of cancer or SCD survivors is an emerging topic in pediatric nursing. The organization of care is affected by the lack of clear and well-structured organizational models. Further research is needed to deepen the understanding of some aspects of the transition. (www.actabiomedica.it)

Key words: oncology, sickle cell disease, pediatric, nurse, transition, review, models, organization, care, cancer

Background

The transition of medical care from a pediatric to an adult environment is a psychological change, a new orientation that requires a self-redefinition of the individual, to understand that changes are taking place in his life (1). Transition is defined as a "purposeful, planned process with a goal of providing continuity of care and preparing young adults for greater independence" (2).

Transfer of care from pediatric to adult services may occur between 18 and 21 years of age (3,4). Young adults who make this transition from pediatric oncology are usually long-term survivors, children who have survived cancer and need a follow-up period.

About 90% of young people with chronic health conditions survive and reach adulthood (5), and cancer survivors are a growing population (6). About 75% of survivors say they experience a chronic health condition, more than 40% still have serious health problems and 33% have multiple health problems (7). Many centers don't have a plan to prepare the survivors and their families for a successful transition (6), although a good plan can help young adults maintain optimal health outcomes, promote independence and empower them to manage their own health conditions (1,8).

Up to 60% of pediatric patients who transition to adult services will experience one or more disease or treatment-related complication as they become adults, including endocrine, cardiac, reproductive or psychological side effects, difficulty coping with adverse results of treatment, anxiety about the future, or an altered body image (9).

The same problems are experienced by patients with Sickle Cell Disease (SCD) (10). Almost 95% of patients with SCD live past the age of 18 and therefore require transition to an adult healthcare setting. For these patients, the period between 18 and 22 years of age is associated with an increased risk of mortality and morbidity due to poor adherence to therapy (11,12). As a consequence, pre-transition process measures are an important component of quality care in SCD (10).

Different transition models have been proposed to adapt to this difficult phase: generic models fit the traditional medical training models of pediatric, adolescent, and adult health care providers. In primary care models a family physician, or a primary care physician, is viewed as the care coordinator, and subspecialty consultants are used as needed. Single-site models are similar to generic models; the site of care remains constant as transition occurs from pediatric to adolescent to adult health care (3). Many patients have difficulty coping with this initial phase of the transition process, proved by an increase in access to the first aid, to emergency visits and re-hospitalizations.

Barriers to transition of care are most often classified into one of four groups: patient centered barriers, family centered barriers, pediatric caregiver barriers, and adult caregiver barriers (13,14).

A nurse who knows how to recognize potential barriers at an early stage can play a pivotal role in the educational plan for the transition process: this professional can identify the needs and limits of each young person, and must have a strong cultural background on this aspect (15).

Aim

The present study aims at exploring, through a systematic literature review, the main topics of transition care in the pediatric hemo-oncological and oncological setting, at identifying the strengths and weaknesses of this process, the different organizational phases, the models already tested, and at addressing the experiences lived by the main actors (patient and caregiver).

Materials and Methods

A literature search was undertaken of PUBMED, CINAHL and The Cochrane Library, from March to April 2018.

The following keywords were used in combination to identify relevant publications: transition of care, oncology, cancer. The terms were combined using the Boolean operator AND. Limits for the search were: full text, publications in the last 10 years, articles in English or Italian, and any type of study design.

Inclusion criteria: articles about the transition process between pediatric and adult providers; articles about nursing topics related to the transition process, patient experiences of transition, caregivers experience of transition, facilitators, difficulties and barriers to transition; articles illustrating how handover between healthcare professionals is organized. Both articles exploring the transition process for patients with oncohematological, oncological disorders and sickle cell disease were all included as they share the same clinical environment and healthcare team.

Exclusion criteria: articles about the transition process between adult providers, articles about medical topics such as drug dosage, diagnosis, and costs related to the process, articles about transition processes for patients with a diagnosis other than cancer or sickle cell disease.

This literature review has been performed according to the PRISMA statement (16).

Using the keywords mentioned above, 38 articles were found in The Cochrane Library, but excluded because of lack of relevance. 5877 articles were found in PUBMED and 71 of these were selected, 274 articles were found in CINAHL and 17 of these were selected. A total of 88 articles were selected after the abstract screening. After removing the duplicates, 44 articles were considered.

After the reading of Full Text, 31 articles were selected, because the other fall in the exclusion criteria.

Table 1. Search strings and keywords.

Search number	Found articles	Selected articles	Keywords	Limits	Databases
1	4752	65	Transition of care AND oncology AND cancer	2008-2018, Full text, English and Italian	Pubmed, Cochrane Library
2	3351	42	Transition of care AND oncology	2008-2018, Full text, English and Italian	Pubmed, Cochrane Library
3	4053	46	Transition of care AND cancer	2008-2018, Full text, English and Italian	Pubmed, Cochrane Library
4	3	0	Transition AND cancer AND oncology	2008-2018, Full text, English and Italian	Cochrane Library
5	35	0	Transition AND oncology	2008-2018, Full text, English and Italian	Cochrane Library
6	15	0	Transition AND cancer	2008-2018, Full text, English and Italian	Cochrane Library
7	248	12	Transition of care AND cancer AND oncology	2008-2018, Full text, English and Italian	CINAHL
8	156	4	Transition of care AND cancer	2008-2018, Full text, English and Italian	CINAHL
9	162	8	Transition of care AND oncology	2008-2018, Full text, English and Italian	CINAHL

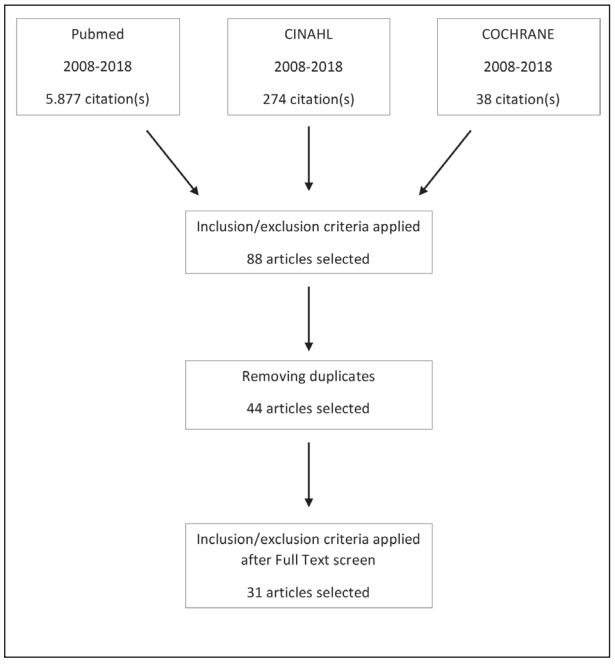

Figure 1. PRISMA Flow Diagram

Of these 6 were reviews of the literature, 1 policy statement, 14 descriptive studies in prospective/retrospective or cross-sectional design, 9 qualitative studies with focus-groups, semi-structured interviews or Delphi studies, 1 qualitative socio-ecological study.

Results

The main themes surrounding transition of care that emerged from the synthesis are the organization of care within common models of transition, innovative clinical approaches to transition, and the lived experience of patients and caregivers.

Organization of care in transition

The definition of transition means the transfer experienced by adolescents and young adults from pediatric to adult care. This transition can be a period of major stress and unfavorable consequences especially for adolescents who have undergone treatment for a serious illness and must move from a sheltered pediatric environment to an independent adult-medicine environment. Adults who transitioned without a concrete plan reported feeling ill-prepared and that their

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
Freyer et al., 2008	Literature review	To identify specific goals and action items in the following key areas: Models of Transitional Care, Survivor/ Family Education, Post-Transitional Care Outcomes, Education of Health Care Professionals, and Health Care Policy and Advocacy.	Not applicable	Deficit in primary care assistance and long-term planning. Some centers use a team with a general adult physician and pediatrician. Pediatric oncologists should develop a plan for patients that accounts for the possible long-term effects that they may experiences as adults. There is a need for increased family education, and additional education for care teams on the transition process.
McPherson et al., 2009	Descriptive cross-sectional study	Primary aim: to describe the preparation and knowledge of adolescents with Sickle Cell Disease during the transition process based on age, sex, degree of severity of the disease. Secondary aim: to identify adolescents' concerns about interfacing with transition process.	69 adolescents with Sickle Cell Disease, 30 females and 39 males, USA- Washington	Older children feel more prepared and have greater levels of knowledge about the process of transition of care. A positive attitude towards this process increases over the years, the difficulty of the process is inversely related to the severity of the disease. An insufficient knowledge and a late introduction to it emerges as the main barriers to the transition process.
Henderson et al., 2010	Literature review	To describe problems and obstacles to the success of transition programs dedicated to child cancer survivors.	Not applicable	Transition programs for child cancer survivors require the input from experts who can act as a bridge between pediatric oncology services and adult primary care services, in order to reduce risks associated with transition. The transition process must take place gradually and can be carried out optimally only by overcoming the concrete problems. The obstacles to the success of transition are put in place by those who should facilitate this process: the health system, the patients and those involved in providing care in the pediatric and adult fields.
Freyer, 2010	Literature review	To explore how the formal transition process can contribute to meeting the medical and psychosocial needs of child cancer survivors who usually have a lack of knowledge on health and health promotion.	Not applicable	Patients who survive childhood cancer are not compliant with the recommended follow-up in adulthood. The systematic transition process is the gold standard, even if there is no model that is ideal or better than others.

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
Nathan et al., 2011	Literature review	To identify a systematic transition plan that considers diagnosis, initiation of therapy, completion of therapy, entry into long-term follow-up care, transfer from pediatric to adult medical providers, and exit from oncology care providers to primary health care providers.	Not applicable.	An appropriate care plan is essential to transfer the patient from a cancer clinic to the primary care setting. Many patients do not have a primary care provider, so the cancer clinic should help them find one. Some clinics accompany the patient during this phase, others discharge them at the end of the therapy without planning for the transition to the adult clinic.
Sobota et al., 2011	Survey, Descriptive cross-sectional study	To describe how the transition process takes place in pediatric hospitals with Sickle Cell Disease centers (logistic mode, identification of a physician in the adult area, patient preparation, program and transition assessment, demographic aspects).	Directors, or delegates, of 45 pediatric hospitals with Sickle Cell Disease centers. USA- Boston	The transition process is initially discussed when the patient is about 15 years old, and is initiated at around 19. 97% of the centers identify a referring physician in the adult area. Most professionals discuss it with the patient and the family, and prepare a plan that identifies needs. About half of the centers review the program annually, 39% measure patient satisfaction. The main obstacle is finding a referring physician in the adult service.
Schwartz et al., 2011	Qualitative study. Social-ecological model	To create a social-ecological model that describes the patient's preparation for the transition phenomenon.	Adolescent and young adult (AYA) with chronic health conditions, including patients with cancer and survivors.	Model divided into 3 parts (patient, parent, physician) that assesses the degree of preparation for transition by age, knowledge of the disease, and cognitive ability. This model considers the influence of health, culture, sociodemographic factors and health system on the style of coping that the patient and the family may develop. The model aims to be universal, therefore there is a need to validate it in specific populations.
Granek et al., 2012	Qualitative study. Grounded theory.	To identify psychological factors involved in the transition process.	Total: 38 patients. 10 patients still under the care of pediatric services. 28 patients who had undergone transition to adult services: 11 successfully transitioned, 17 failed the transition.	It is very important to take into account the psychological factors involved in the preparation of child cancer survivors who are transitioning to adult services. Identifying and addressing the individual psychological needs of these patients can contribute to a successful transition. Moreover, since the attitude towards one's own health is not always regulated by rationality, but is influenced by emotional drives, focusing on the psychological aspects, can help patients to address themselves in a positive way towards the treatment.

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
McInally et al., 2012	Literature review	To explore the meaning of effective transition, highlight some of the challenges faced by young people with cancer, identify gaps in the research literature.	Not applicable	The care provided should be appropriate for the young adult; the patient's concerns must be heard by specialists; the transition of care should promote autonomy, independence and responsibility of the young person; the process must be flexible and planned with the family. There is no shared or emerging model to guide the process.
Sadak et al., 2013	Descriptive cross-sectional study	To generate hypotheses of facilitators of the transition process.	129 young adult (> 16 years old) cancer survivors that have not yet "passed" into the adult setting.	Young patients prefer a clinical team with a pediatric specialist and a clinical setting where there is good flexibility in planning the transition process. The possibility of using network to help the process is poorly considered. There is the necessity to clarify meaning of network, if included as a social network or network created by social media.
Schwartz et al., 2013	Qualitative forms: focus group e semi-structured interviews.	Further validation of the Socialecological Model of Adolescent and Young Adult Readiness to transition (SMART) through feedback from stakeholders: child cancer survivors, their parents and caregiver teams.	14 patients who survived childhood cancer. 18 parents. 10 health professionals specialized in the pediatric field.	Progress in the transition process is hampered by the lack of measurement instruments that could identify and improve current practices. SMART is a theoretical model, a comprehensive and empirically appropriate tool for assessing whether a child cancer survivor is ready for the transition process.
Klassen et al., 2014	Interview. Evaluation scales.	To develop and validate instruments that evaluate when a child cancer survivor is ready to transition from pediatric to adult care.	38 child cancer survivors: 10 still managed by pediatric care, 11 successfully transitioned, 17 failed transition process. 331 child cancer survivors, of these 250 completed the questionnaires.	There is limited knowledge about the experience of the transition process for child cancer survivors. Validated assessment tools can be used to investigate obstacles and / or facilitators to the transition process from pediatric to adult care. Creation of three evaluation scales: 1. Cancer Worry Scale 2. Self-management skills scale 3. Expectation scale
Fernandes et al., 2014	Descriptive study.	To determine patient and parent attitudes and perceptions of the education provided during the transition process, and obstacles to transition .	155 patients with various chronic childhood illnesses, aged between 16 and 25.	Most patients and parents say they have received information and training on the health condition. There are significant gaps in the educational process that takes place during the transition.
	Self-assessment survey: 30 multiple choice questions and 1 open-ended question.		104 parents or caregivers.	For example: lack of education regarding unprotected sex, birth control, pregnancy, drug abuse, and lack of job counseling. Some barriers to the transition process have been identified: emotional attachment to the pediatric team, and gaps in the provision of adult care. Most patients feel ready to complete the transition process at the age of 25.

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
Andemariam et al., 2014	Descriptive retrospective study.	To describe risk factors for negative outcomes of the transition process.	47 patients with Sickle Cell Disease between the ages of 16 and 24 who experienced the transition process between 2007 and 2012.	The study shows that a transition with a negative outcome is not related to sex, race, episodes of "acute chest syndrome" or hospitalizations for episodes of vasoclusives. There is a correlation with the starting age of the transition (the most favorable outcome for those who start before the age of 21) and with the distance of the adult clinic compared to the pediatric setting.
Bryant et al., 2015	Policy statement	To define the process of preparing pediatric patients with Sickle Cell Disease for the transition of care	Not applicable	It appears necessary to start discussing transition at 12 years old, and start written planning from 14; to get help from organizations in the sector; to include in the plan a multi-professional team, and the family / caregiver; to make sure that parents leave the child alone only a part of the visit from the age of 13, and let completely alone visits from 18 years old. The process ends not in the transition to the adult setting, but when the whole team and the family are sure of the successful outcome of the transition.
Frederick et al., 2016	Qualitative study Focus group	To describe the commonalities and differences between experiences of patients with cancer.	16 patients recruited from a pediatric oncohaematological clinic, aged 21 to 39, who have completed therapy for at least 1 year.	Main themes emerged: education on "self-advocacy", the worry about the future, the role of the family as an obstacle to autonomy, the dependence on parents to book visits and to make health decisions, the expectation of having a close relationship with the doctor, the problem of who to ask for support, the necessity of an individualized plan for the process, different expectations on primary care medical role.
Ganju et al., 2016	Descriptive cross-sectional study.	To evaluate the impact of the previous care, before the transition process, on patient knowledge and awareness of the disease. Identify any demographic or neurocognitive barriers to education.	110 patients enrolled. 93 of these completed the questionnaire.	Participation in patient care program plays an important role in the transmission of information regarding their pathological history and the perception of the risks of future health problems. Care programs for child cancer suvivors must be developed and implemented to fill any gaps in the patients' knowledge of self-management of health.
Svedberg et al., 2016	Cohort observational study. Mixed method.	To explore young adult cancer survivors experiences of support from health services during the transition process.	416 patients diagnosed with acute lymphoblastic leukemia between 1985 and 1997 enrolled in the Swedish Children's Cancer registry. Of these, 144 completed the questionnaire.	Most participants received insufficient physical, mental and social support from health services. During the transition process it is necessary that health services adopt a personalized assistance plan. The approach used must be holistic and must support the patient in managing their life in the best possible way.

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
Szalda et al., 2016	Descriptive cross-sectional study. Questionnaire.	To describe the patient perceptions of the involvement of adult services during follow up.	80 patients transferred from the Survivorship Cancer program at the Children's Hospital in Philadelphia to the adult-focused follow-up. 99 of these decided to participate in the study; 80 completed the questionnaire.	Young adults cancer survivors report a non-optimal involvement and communication during follow-up meetings for adults with cancer. Patients demonstrate a lack of understanding in the importance of follow-up.
Kenney et al., 2016	Descriptive cross-sectional study. Questionnaire.	To describe the current practices and models of transition process; to describe the perceived obstacles during the transition phase.	1586 medical specialists in pediatric oncohematology, members of the Children's Oncology Group. Of these, 507 replied to the electronic questionnaire. Of these, 347 possessed the eligibility criteria.	Systematic transposition practices do not seem to be widely used by pediatric oncologists. Specialists experience many barriers to the transition of patients to adult care. Medical specialists share the goal of providing patients with a systematic transition education to prepare them to manage their health needs independently.
Bashore et al., 2016	Pilot study	To examine the use of an interactive workbook as an educational method for patients facing the transition.	20 child cancer survivors, between 16 and 21 years old, who have completed therapy two years ago.	Those who are less ready to leave pediatric services are less likely to start the transition process. Patients experienced more anxiety at the start of the study than at the end Those who finished the workbook reported they felt more ready for the transition. The workbook is recognized as an instrument, but more education and knowledge is needed on the process.
Margolis et al, 2017	Descriptive retrospective and cross-sectional staudy	To identify strengths and weaknesses in the management of transition from a pediatric to an adult clinical setting for patients with Chronic Granulomatous Disease	33 patients enrolled from 1 January 2011 to 28 February 2014, aged between 18 and 24.	The authors identified that introducing patients to the adult clinical setting before admission was a facilitator to transition Main barriers identified included a lack of full understanding of the patient's disease and treatment regimen, lack of preparation and planning for the transition process, and missed opportunity for Advance Care Planning.
DiNofia et al, 2017	Descriptive cross-sectional study	To describe the wishes of parents of child cancer survivors in the transition process towards an adult setting.	138 enlisted parents, 123 enrolled, 41 responses collected. Parents of patients >16 years of age who participated in the 3 years preceding the "LTFU Program at Children's National Medical Center"	Parents want complete involvement in the transition process. They consider it important to promote the independence and responsibility of their children, to be prepared for the transition process, and to maintain a point of contact at pediatric services.

Table 2. Summary of results

Study Author, Year	Design	Study Aim	Sample, setting	Findings
Sadak et al., 2017	Phenomenological qualitative study	To define the characteristics of a positive transition of care from the point of view of the patient's medical team, patient and parents, with semi-structured telephone interview.	29 professionals (10 doctors, 8 experienced nurses, 6 nurses, 2 psychologists, 1 social worker, 1 dietician, 1 administrative) of 3 institutions.	The study identified the following facilitators of transition: good communication between the pediatric and adult teams, multidisciplinary network of specialists, presence of several services within a structure (as happens mostly in Pediatrics), creating the figure of the "Patient navigator" (a bridge between the two settings), hold regular meetings between the pediatric and adult teams. The identified barrier is the lack of a home care team or primary care physician helping this process.
Quillen et al., 2017	Descriptive pilot study.	To identify and describe barriers that young adults encounter during the transition process within 5 years from the end of the pediatric therapeutic path.	48 young adults, aged between 20 and 25, who completed treatment in pediatrics and transitioned to adult services.	Barriers included a knowledge deficit in the transition process among young patients; lack of physicians' knowledge of the long-term effects; poor education on long-term follow-up. It could be useful to have a contact list of adult hospitals to create a transition plan.
Mouw et al., 2017	Qualitative approach: grounded theory. Interview.	To examine existing models of the transition process, emphasizing strengths and weaknesses. To optimize these models in order to maintain a connection with child cancer survivors who go through the transition process.	20 LTF experts (Long term Follow up): doctors, nurses, social workers, educators, psychologists from 10 institutions affiliated to the Pediatric Oncology group.	Most patients who survive childhood cancer develop physical and / or psychosocial sequelae; however, many subjects do not receive adequate long-term follow-up for screening, prevention and treatment of later complications. Patients benefit from transition models in which there is a greater and better connection between patient and specialist.
Nandakumar et al., 2018	Descriptive study: semi-structured telephone interviews	Describe the attitudes and experiences of child cancer survivors and their parents regarding barriers and facilitators to the transition process.	33 subjects interviewed: 18 patients who survived childhood cancer 15 parents of patients who survived childhood cancer	The obstacles to the transition process include: dependence on pediatric health services, low trust in general practitioners, inadequate communication and cognitive difficulties. Facilitators include trust of physicians, good communication, patient independence, and patient age when transition process is commenced.

transition was based on age rather than readiness or needs (17). These adult patients also reported that their follow-up care had declined since the transfer.

In addition, failure of transition and hence of appropriate surveillance for late effects may have potentially important medical consequences (6). Hence, a well-planned transition to adult care allows AYAs (adolescents and young adults) to optimize their health and ability to independently manage their disease and assume adult roles and functioning (5). Thus, transition programs that prepare pediatric patients with SCD for the adult healthcare environment promote self-advocacy and self-management. Model transition programs use interdisciplinary teams to help adolescents develop this independence and knowledge.

While there is a body of literature on Advance Care Planning with AYA, this topic is often overlooked in the literature on transition (18). This is probably

due to the vast heterogeneity of situations that may be faced by caregivers dealing with this transition, so that the argument may have been considered as too broad. For instance, the pivotal focus in the transition programs for cancer and sickle cell disease (SCD) – one of the cases studied in this review – has so far been mostly focused on a very specific topic, the optimal age to deal with transition.

Care Transition Models

Several models of care for adult survivors of childhood cancer were identified. According to Freyer et al., some institution-based programs transfer young adult survivors from the pediatric oncology clinic to an adult-oriented Long-Term Follow-Up (LTFU) team within the same medical campus, comprising both primary care physicians (e.g. family medicine or internal medicine) and pediatric oncology clinicians (e.g. physician or mid-level provider) (18). At the time of survivor transition (typically between 18–25 years old), the pediatric oncology team needs to prepare a detailed, comprehensive treatment summary to aid the new physician. This document should include a summary of the cancer diagnosis; prior treatment including significant clinical events; an assessment of current health status including a complete physical examination and list of active health problems and psychosocial issues; and potential late effects (with approximate risk estimate, if possible) that may result from the cancer, its treatment, genetic predispositions and any co-morbid conditions.

Freyer (19) classifies transitional care models under three headings. In the cancer center–based model transitional care is delivered within the same system as treatment was given and involves direct, on-site collaboration of the pediatric oncology team and adult care providers. In the community-based model, transition is located in the office or clinic of the care provider, typically a primary care clinician. With the hybrid model, care is also transferred to the office or clinic of the primary care provider but relies on an ongoing interaction with the cancer treatment center that includes bidirectional updates on patient status, assistance with clinical management, and provision of current survivorship care guidelines. For all three models, the pre-transition phase relies on the pediatric long-term follow-up team (typically a pediatric oncologist, an advanced practice nurse, and a medical social worker).

A similar classification is proposed by Granek (20). In their work, some programs transition to a primary care practitioner, while others offer life-long care in specialized survivor clinics. An intermediate model distinguishes between survivors with high and low levels of morbidity. The former shall be included in specialized survivor programs, while the latter can be addressed to primary care practitioners.

The majority of pediatric cancer centers have a formal survivor program or clinic. The remaining centers do not have a specialized Long-Term Follow-Up (LTFU) program or clinic and provide follow-up to survivors in their acute care oncology clinics. Nathan (21) describes transition models for AYA cancer survivors in Canada. Once survivors reach adulthood, few centers have access to a formal program for adult survivors of childhood cancer, whereas the remaining centers discharge survivors to their primary care physician at some point after the completion of therapy. There are no formal survivor programs for adolescents/young adults who receive their acute cancer care in an adult hospital.

There are five core principles for transition arrangements of childhood cancer survivors to be successful (9):

1. The healthcare setting should be appropriate for the client's age and stage of development.
2. Common concerns associated with young adulthood should be addressed in addition to specialty care.
3. Transition should promote autonomy, personal responsibility and self-reliance in young adults.
4. Transition programmes should be flexible to meet the changing needs of young adults.
5. The designated process should be planned with the young adult and their family.

To describe transition practices and barriers to transfer, Kenney electronically surveyed 374 U.S. Children's Oncology Group members. Personal provision of transition education is delivered by the majority of pediatric oncologists, often with the help of other clinical staff. The majority of pediatric oncologists do not use a formal transition assessment tool such as

questionnaire, survey, or checklist to assess their patient's transition readiness (12).

Transition has been studied in depth also for SCD survivors. Andemariam defines the transition period for SCD patients as having three phases: preparatory, transitional, and completion. The preparatory phase is focused on patient education regarding SCD and patient-specific health issues and management (22). It lasts for 6–10 visits done every 4–6 months over a 3-year period. The transitional phase is dedicated to review health summaries, problem lists, and treatment plans with the family and the medical staff. It is focused on empowering the patient and promoting autonomous health management. The completion phase is focused on establishing effective patterns of health care in the adult setting.

Sobota carried out a survey of US pediatric providers and describes transition of SCD patients. Most clinics report having a transition program, although half have been in place for under 2 years (23). There is wide variation in specific transition practices. Close to all centers have an identified accepting adult provider, however, only slightly more than half routinely transfer their patients to an adult hematologist specializing in SCD. Although there has been a recent effort to establish transition programs in pediatric SCD clinics specific practices vary widely. Lack of an accepting adult hematologist with an interest in SCD emerged as a common barrier to transition. One-third of centers allow patients to remain in pediatric care past the cut-off age in cases of cognitive or developmental delay, or needing time to complete a transition program, graduate high school, or find an adult provider.

The majority of the aforementioned studies highlight that the transition age is a main determinant of success or failure of the entire process.

Bashore et al. (1) underlines that the America Academy of Pediatrics (AAP) has established guidelines for clinicians to begin transition as early as 12 years of age, to allow for acquisition of skills necessary for the independence required in adulthood. Not only should the chronological age of the adolescent be considered but also the developmental age of the adolescent.

Andemariam stresses that, in their study, older age at the time of initiation of the modified transition process was associated with poor transition success. The preparatory and transition phases were changed such that both begin at age 16, and the definitive transfer to the adult SCD center is at the age 21. Ideally, patients schedule their first visit to the adult SCD center prior to reaching their 21st birthday, and prior to their last appointment at the pediatric SCD clinic (22).

In all three models proposed in Freyer's classification, the actual transition of care ordinarily takes place when the survivor reaches approximately 18 to 25 years of age and demonstrates transition readiness (19). The transition process needs to be initiated early—it is not too early to begin mentioning transition when the child is initially diagnosed with cancer—beginning at 18 years of age is almost certainly too late.

In another study the majority of respondents transferred childhood cancer survivors to adult care by age 25 years (12). The timing of transfer was most often determined by patients' chronologic age, diagnosis of adult comorbidities, and pregnancy.

According to Bryant et al. (10), a formal discussion about transition and the policy of the practice/institution should begin at age 12 (or when developmentally ready) with both parent and child. All patients should have a written transition plan by age 14. This plan should be developed together with the patient and their family and updated annually.

In a survey of transitioned patients, it has been demonstrated that most participants agreed that the transition should begin in early to mid-adolescence (24). This is needed to optimize education of disease history, current and future survivorship care needs, and medical risk.

In a survey of US pediatric providers, Sobota et al. (23) describes that just over half of the centers are in a system with a required age for transition due to "hospital policy," which ranges from 18–22 years. Transition is first discussed at an mean age of 15.7 years (range 13–18) and transfer occurs at a mean age of 19.6 years (range 18– 25). Age and pregnancy are still the primary factors that determine time of transfer. Pregnancy in adolescents and young adults may not be planned, and therefore, using it as a trigger for transfer is unlikely to allow adequate time for preparation. Using age as a proxy for maturity may be particularly problematic for patients with SCD, who may have neurocognitive delay due to cerebrovascular injury.

According to Quillen (15) pediatric cancer survivors could start transition to adult health care at 21 years of age.

Innovative Approaches to Transitional Care

As underlined by Ganju (25) a transitional clinic for young adult survivors (YAS) of childhood cancer is an evolving model and at present, there is little research evaluating the benefits, implementation, and efficacy of these clinics for pediatric cancer survivors.

Granek stresses that an important implication for practice is to empower teens' sense of identity as a cancer survivor by engaging with their peers and encouraging them to attend cancer-related groups and organizations that provide information and social support for survivors (20). This could be achieved through camps or organizations that involve peers as well as through peer mentor or 'buddy systems' within the health care context.

Freyer (19) propose an interactive online program called Passport for Care, which provides survivors and clinicians with a virtual resource center, where they can enter patient-specific history and receive individualized monitoring recommendations.

A pilot study examined the use of an interactive transition workbook as a method of educating survivors about their medical history, providing necessary information about the transition to adult care, and working with them to establish goals and plans for education and vocational success (1). Having the time to collate this information in an organized manner may have assisted them in processing the transition from pediatrics to adult care in the future.

Klassen et al. (26) developed three scales for childhood cancer survivors that measure concepts identified as barriers and/or facilitators to transitioning successfully to adult-orientated health care. They measure Cancer Worry (about cancer-related issues such relapsing or getting a new type of cancer), self-management skills (investigating skills that adolescents need to acquire to be able to care for their health as adults, such as booking doctor's appointments and filling prescriptions) and expectations (delving into the nature of adult Long Term Follow Upcare, such as expecting to get a reminder call before an appointment).

Sobota, Shah and Mack (27) propose that sending a transfer summary ahead of the first visit in adult care should be part of best practice in transition. According to the results of an expert panel, adult SCD providers would also appreciate direct communication from the pediatric hematologist. Lack of time and reimbursement are often cited as barriers to providing comprehensive transition. To solve this problem, transition advocates have identified billing codes that allow reimbursement for transition activities such as updating a transfer summary (e.g. by billing for "care plan oversight").

A transition model called SMART – Socio-ecological Model of AYA Readiness for Transition was proposed by Schwartz et al. (5). This model of transition extends beyond patient age and patient knowledge and skills by identifying measureable social-ecological components of the transition process and highlighting the potential role of culture and socio-demographics in the transition process, a neglected issue.

Innovative approaches such as peer mentoring programs or web-based interventions may reach more patients compared to the traditional clinic approach (28). Important next steps include further education for patients and adult providers, ensuring adequate transition planning for youth and their families, and research to determine what factors have the most significant impact on transition quality.

The Patient experience

The point of view of YAS is necessary to understand how to achieve a successful transition. To elicit this, patients aged from 16 to 39 years are often asked to complete a questionnaire by e-mail or during their annual survivor clinic visit. The most common questions concern barriers and facilitators to transition, how they received the relevant information, the relationship with their parents, and recommendations for a successful transition. Some studies have investigated gender differences (25,29); the psychological and social aspects of transition (20,21,30,31) and age differences (22,29).

Regarding gender differences, 30 females and 39 males (Median age was 16.7 years) responded to a survey on transition to adult services (29). Female patients reported a higher level of anticipated difficulty

than male patients. No significant differences were found between knowledge, thought, interest, and importance of transition. In the study by Ganju et al. (25) men were less likely than women to expect future health risks from their cancer treatment.

Regarding the psychological aspect, severity of disease and a high rate of hospitalization had a negative impact on patients' interest in learning about transition (18,22,29). Using the Childhood Cancer Survivor Study Neurocognitive Questionnaire (CCSS-NCQ), Ganju et al. (25) found that the patients are more likely to assess their health risks based on their current health states, as opposed to the intensity or duration of their treatment. The transitional period often takes place at a critical time during survivors' development into independent young adults. Many survivors view themselves as completely healthy or invincible and they do not recognize their risk of serious cancer-related health problems and do not adhere to recommended cancer-related follow-up care (21). Furthermore, childhood cancer survivors may experience psychological symptoms of depression, anticipatory anxiety prior to the transfer and posttraumatic stress (PTS) which can hinder their engagement with medical care and make the transfer of care to the adult system a difficult emotional process (30,31). In the study by Svedberg et al. on 213 YAS of pediatric cancer, they found that survivors would have appreciated more follow-up information based on their needs and on their psychosocial health (31). The participants reported they had not received the annual follow-up visit to control the risk of late effects of treatment, did not experience sufficient support for: depression, panic disorders, eating disorders, obsessions, hypochondria and did not receive treatment strategies for physical changes.

Patient age was significantly associated with interest in the transition. Older age at the time of initiation of the modified transition process was associated with poor transition success (22) but patients aged 17–20 years demonstrated significantly greater knowledge and interest in transition, greater self-management skills to make their own appointments and call for medication refills than 14–16 years old (10,29). A slightly older age at transfer may improve readiness for transfer but conversations about high-risk behaviors such as alcohol, tobacco, illicit drug use, sexual intercourse, need to begin in early adolescence (32).

In some studies, the relationship and attachment to the family and to the pediatric healthcare providers has been identified as a barrier. Patients recognize the importance of care received from parents and pediatric health care providers but report this has made them less prepared for autonomy in adult life. Therefore, parents still play a significant role in communication with the healthcare services even when the patients become adults (31,32). Dependence on parents or doctors could be necessary to compensate for cognitive difficulties that result from chemotherapy treatments, as these can negatively affect self-management ability (14). Several patients develop their self-management abilities and understand their disease only after transition experience (11,14). Also, pediatric oncologists find it difficult to transfer long-term patients into adult care because of their long-standing relationship with them (14,32).

Patients suggest that awareness of the differences in care between the pediatric world and the adult world could facilitate transition (28). About 63% of adolescents wanted their pediatric doctor to supply specific information about adult hospitals in the area, 59% requested written information about the transition process, 39% requested help in making the first appointment with an adult provider, 33% asked to be connected to someone who had already gone through the transition process (28), 23% requested help in visiting different adult hospitals, and 17% requested group meetings with other patients to discuss transition (29). Other information requests concern specific names of doctors, information on insurance coverage, ease of appointment scheduling and parking/ transportation (29). Good communication was perceived to enable successful transition and was associated with positive transition attitudes. Communication also provided comfort during transition (14). The most important information for patients with SCD, was concern about the modalities of pain management and planning re-entry for transfusions (22).

Key barriers to transition included dependence on pediatric healthcare providers, less confidence in primary care physicians (PCPs), inadequate communication, and cognitive difficulty (14). Less than half of patients (N = 155) and parents (N = 104) reported receiving any education regarding reproductive health (specifically, unprotected intercourse), impact of

disease on future offspring, birth control, risk of pregnancy, illicit drug use, and future career (32). Lack of knowledge about the disease and anticipatory guidance about the process were the major barriers in transition (29,33). Nearly a quarter of participants in Margolis' study (33 young adult participants, 19 - 27 years of age), reported that they did not feel included in planning the transfer of care or in the actual transition itself. One AYA said, "*There was no transition. It just kind of happened*" (33). Only in one study, a lower income was a demographic factor that correlates with less knowledge (25). Patients who have experienced a greater number of complications, like acute chest syndrome (ACS) episodes and hospitalization for vaso-occlusive crisis (VOC), are less likely to experience a positive transition (22).

In some cases participants expressed dissatisfaction about losing contact with healthcare services after the age of 18 or after being discharged from the pediatric oncology ward at the end of treatment (31), but the real problem is the perceived negative attitude and lack of trust in new adult care provider (18). Specifically in the Emergency Department and inpatient units, the staff were not well informed about SCD (28) or the PCPs did not demonstrate sufficient cancer-specific knowledge to provide the level of care that pediatric HCPs could (14). In particular, young adults with SCD have the perception that health care professionals in the adult world underestimate the degree of pain experienced (28). Some Adult Patients with SDC have reported "*these physicians did not have enough medical knowledge about sickle cell disease*" (11).

Non-clinical risk factors for unsuccessful transitioning were greater travel distance from the patient's home to the adult SCD center (22), or the inaccessibility of care due to distance (14) transfer to another city (11), and insufficient medical insurance (11,14).

According to the results of this literature review, patients demonstrated high levels of awareness about the importance of transition and showed interest in learning about the process. The transition is a time-consuming process and the goal is to become responsible for oneself (28). Svedberg et al. (31) and Frederick et al. (32) underline the need for a personalized, holistic care plan. Survivors desire a multidisciplinary care team that offers care across multiple specialties and subspecialties but it is unclear which disciplines would be central to the clinical team (34,35). The major barriers in transition included dependence on pediatric HCPs, less confidence in PCPs, inadequate communication, and cognitive difficulty (14). The emotional components such as fear, anxiety, gratitude and gaining perspective acted as both facilitators and barriers to transition in different childhood cancer survivors (20). Improvements in the transition process could be made with more written information about local adult providers and the overall transition process through an appropriate medium "like a website or a booklet" (14,29). In some studies, group meetings and visits to adult hospitals were not highly rated (29), while in the qualitative study of Sobota et al. (28) the young adults with SCD suggested meeting the adult provider prior to transfer. During the transition process, patients also need comfort and support for coping with difficult thoughts and memories of traumatic experiences (31). To increase coping, the figures to be involved are: family, survivors, and adult health staff (1).

The Caregiver Experience

The transition phase should assist parents or caregivers in accepting a new role, as they may no longer be directly responsible for the patients' care. Transition means not only a change in the place of care or a change in the referring physician, but also an increased responsibility for the young adult, who must learn to interface directly with the medical specialist and health services (34). Survivorship care plans are an important method for addressing the challenge of safe and effective transfer of care from cancer center to primary care. These documents should be created by the cancer team and shared with patients, families and primary care providers at the end of treatment (21).

While being the only point of reference in the care path creates a unique bond, at the same time it also creates an important barrier. Young patients identify the longstanding dependence on parents for healthcare management as an important barrier, including their reliance on parents to retain critical health information, coordinate appointments, and engage in critical health decisions (24).

A notable difference between survivor and parent point of view on transition emerges from the study of Frederick et al., where only 43% of survivors reported parental inclusion as "very important" in their decision to transition care compared to 83% of parents (24).

This discrepancy could mean that young adults experience a period of increasing independence.

There is also a subset of childhood cancer survivors that develop an unhealthy dependence on their parents for coordination of their health care.

On the other hand, while there are many possible reasons for this discrepancy between desired parental involvement, this difference in scores emphasizes the fact that all models of transitional care for child cancer survivors must allow adaptation to the new care setting to meet the individual needs of each survivor and his family, including parents

Sadak et al. report that parents feel "*worried because they feel like everyone in pediatrics knows exactly what happened [to their children]*" and may wonder if the team of adult-centered survivors "*really know*" and "*understand consequences of having received the treatments*". It is important for parents to know that "*their child is moving to a team of specialists who focuses specifically on caring for the adult survivor [of child cancer]*" (34). The medical team in the adult area should make caregivers more involved in the care path by showing them the informative and teaching materials that they will give their children.

Fernandes et al. shows that 73% of parents support the allocation of resources and materials for more education and assessment prior to transitioning, and 95% of parents supported the allocation of resources to improve the transfer process of patients from pediatric- to adult- oriented care (32).

DiNofia, Shafer, Steacy, & Sadak showed that 100% of parents believed it was important to promote the independence of survivors. Sometimes, these desires can be conflicted. The achievement of autonomy for adolescents with chronic health conditions is often delayed compared to peers without these conditions (36).

However parents also have the opportunity to be proactively involved in supporting the path of their child towards the independence of healthcare (36).

Some parents suggested strategies that could help alleviate their fears, for example by using the resources of a pediatric provider that could help / mediate part of the transition by working together with adult providers and building patients / parents support groups (32).

These two studies agree that additional studies are needed to determine if parental attitudes about transition of care to adult care settings and their inclusion in the transition process are determinants of successful transfers.

The perceived barriers to transition to adult care included deficits in: disease understanding, medication regimen understanding, knowledge about advance directives, and preparation, planning, and practice related to the transfer in care. In this sense, the participation of caregivers is intended as a facilitator and not a barrier (36).

The good preparation and skill of the adult team to succeed at this stage is fundamental: *"educating the families about the idea of transition and that is beneficial is critical*" (34). This literature review has illustrated that sufficient planning of transition is the main contributor to successful outcomes for survivors.

Discussion

The organization of care is affected by the lack of clear and well-structured organizational models. The first problem that arises is the age of the patient when they start this process. The second is whether the care team is adequate for dealing with the transition and what are the professional figures that shall be involved in the process. The third is the active role given to the family. New innovative models to increase the patient's awareness of this transition have been studied. However, emotional factors such as anxiety and fear are still considered as crucial from the perspective of patients, families, or caregivers. Active involvement of the whole family member is necessary to promote survivor autonomy.

This process has to start at an early age and reach the definitive phase with the full maturity of the young adult.

This review highlights how some non-organizational aspects, mainly related to the sphere of the subject's experience, are essential for the success of the process. In fact, unlike studies that deal only with the issues of therapy, the ultimate goal of the transition is

achieved when the patient positively accepts the move into the adult environment and becomes independent and autonomous in dealing with the disease.

The phenomenon of transition faces multiple aspects and includes multiple actors. By dividing the review into these paragraphs, we tried to touch all those fundamental aspects to create a winning treatment plan in the near future.

Conclusion

The transition from pediatric to adult care of cancer or SCD survivors is an emerging topic in pediatric nursing.

This systematic review is the first that includes a review of the transition of care for pediatric patients with cancer or SCD, in all aspects of care.

The review has some limitations: first, it was decided to take into consideration both patients, with oncohematological pathologies and with sickle cell disease, since in most international situations these two groups have in common the same care environment (same hospital unit). However, the two types of patients face different treatments, with treatment paths that can be differently structured, even at the age of taking charge.

Another limitation regards the different types of studies considered to carry out the review: the studies, both quantitative and qualitative, approach the different experiences from different points of view that are sometimes difficult to reconcile. As a result, conflicting results may sometimes emerge.

The strengths concern the originality of the study as the issue of transition, both in the medical and nursing fields, is increasingly prevalent. A review that builds bridges between the different realities can help to create a common care pathway between the two environments.

The review also sought to explore the experiences of both patients and caregivers, considering them both as important and fundamental actors, together with healthcare professionals.

Further research is needed to deepen the understanding of some aspects of the transition care, such as the training provided at university level regarding this process and the possibility of creating an instrument that allows to act as a mediator in the transition process, “like a website or a booklet”.

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