

Chapter 26

Transitioning from Pediatric to Adult Medical Care

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“It takes courage to grow up and become who you really are.” – E.E. Cummings

“To exist is to change, to change is to mature, to mature is to go on creating oneself endlessly.” – Henri Bergson

Introduction

Children and adolescents with chronic medical conditions face an uphill battle as they navigate the challenges of becoming an adult. Chronic medical conditions lead to reduced school participation that can impair school performance and ultimately limit employment attainment. Decreased community participation due to medical care needs and concerns about body image may lead to decreased

practical knowledge regarding independent living and increased rates of depression, anxiety, and social/legal problems [1, 2].

Individuals with telomere biology disorders (TBD) come to the challenge of adulthood from differing crossroads depending on the history of their disease. For some patients with TBD diagnosed in early childhood, the major challenge may be navigating a transition from pediatric to adult medical providers across a wide swath of specialties while trying to maximize educational attainment or entering the workforce. For others, the diagnosis of a TBD may not be made until late adolescence, forcing teenagers to cope with a new diagnosis that has the potential to greatly disrupt preset plans for college and beyond. Finally, other individuals with TBD may not yet be aware of their diagnosis but may be experiencing fear and stress of symptoms that have yet to be explained.

While the burdens of having a chronic medical condition such as TBD are immense, there are some benefits when contemplating a successful transition to adulthood. Taking ownership of complex medical care leads to maturity that may serve patients well in college, initial employment, and beyond. In addition, resilience learned through years of overcoming medical challenges can make navigating the highs and lows of adjusting to the workforce and other aspects of the adult world easier. The purpose of this chapter is to discuss how TBD individuals, families, and their providers can facilitate the process of transition from pediatric to adult medical care to make this potentially stressful transition go smoothly during what is already a challenging time for young adults and their families.

What is Pediatric to Adult Transition?

The Society for Adolescent Medicine recognized the need for formalized pediatric to adult transition for patients with chronic conditions in the early 1990s, stating that these patients would benefit from “the purposeful, planned movement of youth with special health care needs from child-centered to adult-oriented care” [3]. Nearly three decades later, despite extensive research and clinical initiatives, only 17% of youth with special health care needs meet benchmarks for having sufficient transition planning [4]. For TBD patients, the pediatric to adult transition is made particularly challenging due to the rarity of TBD and the lack of familiarity of its management by many pediatric and adult providers. Additionally, diverse disease presentations involving multiple organ systems require multispecialty care coordination. Patients may have difficulties maintaining adequate healthcare insurance coverage and identifying a suitable adult “medical home” to both facilitate the transition to adult care and provide long-term care continuity once the transition to adult care has been completed.

You Are Not Alone

The challenges associated with the transition from pediatric to adult health care are not unique to individuals with TBD. Approximately 10 million children (~20%) in the United States have chronic diseases that qualify as special health care needs, and at least 500,000 individuals with these childhood-acquired conditions requiring transition planning each year [5]. Recognizing the need for a systematic approach to optimize care for this young adult population, the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians-American Society of Internal Medicine issued a consensus statement in 2002 that included several specific recommendations for transition [6]. These recommendations have recently been updated [7], focusing on six core elements of preparation for both the pediatric and adult practices involved in patient care transition (Table 1).

Table 1. Six core elements of pediatric to adult transition.

Responsibilities	Pediatric Practice	Adult Practice	Tips for Parents/ Families to Ensure Successful Transition
Transition care policies/ procedures	<ul style="list-style-type: none"> • End date of care provision • Services/resources provided 	<ul style="list-style-type: none"> • Age-based care start date • Patient acceptance policy 	Ask your team about their policies
Tracking and monitoring	<ul style="list-style-type: none"> • Youth/family preparation 	<ul style="list-style-type: none"> • Review of relevant records 	Ask how tracking is performed in your center
Readiness/ orientation	<ul style="list-style-type: none"> • Formal readiness assessments 	<ul style="list-style-type: none"> • Welcome and FAQ documents to orient young adults to practice 	Start discussing readiness approach by age 14
Planning and integration	<ul style="list-style-type: none"> • Develop individualized transition plan • Medical summary • Communication 	<ul style="list-style-type: none"> • Communication with pediatric clinician • Receipt of transition package 	Basic plan should be in place by age 18 even if transition occurs several years later
Transfer	<ul style="list-style-type: none"> • Final visit • Define pediatric responsibilities to be retained during transition 	<ul style="list-style-type: none"> • Initial visit • Self-management skills assessment 	Ensure that the first adult practice appointment is scheduled by the date of last pediatric appointment
Transition completion	<ul style="list-style-type: none"> • Seek feedback on transition • Confirmation of ongoing adult practice appointments 	<ul style="list-style-type: none"> • Ongoing care • Self care skill building • Communication with prior providers when necessary 	Ensure that the pediatric and adult practices are communicating over at least the first 6-12 months of the transition

Pediatric and Adult Care in TBD:

Different Styles but Same Need for Medical Home

Pediatric and adult care models are broadly different (Table 2). Young adults transitioning to the adult care model need to be aware of these differences to avoid lapses in quality of care. The task of establishing a “medical home” in the adult care system is an additional aspect that can be particularly challenging for TBD individuals who frequently have multispecialty care needs. The medical home model is a way of delivering health care that is coordinated, patient and family centered, and culturally appropriate [7]. In this model, the medical home is the provider who serves as the hub of the medical wheel, keeping track of the current status and screening needs across the spectrum of a patient’s health care needs. Proven benefits of utilizing a medical home care model for patients with complex medical conditions like TBD include:

- Easier access to services
- Consistent and coordinated care
- More efficient and effective use of resources
- Better support to individuals and families
- Improved health, developmental, educational, vocational, psychosocial, and functional outcomes

The adoption of this model is sometimes more challenging for rare complex diseases, such as TBD, because the medical home provider needs to have a good understanding of all aspects of TBD. For children with TBD, subspecialists at large academic centers such as pediatric hematologists, immunologists, geneticists or stem cell transplant physicians frequently provide this medical home. In the United States, due to special provisions by Medicaid and private insurance policies for pediatric patients, pediatric academic practices can overcome insurance barriers to seeing patients who may be “out of network” more readily than can many adult practices. Tertiary care pediatric

practices are also frequently equipped to handle patient complexity and have established connections with multiple subspecialists required for TBD patient care.

Table 2. A comparison of pediatric vs. adult care.

Pediatric Care	Adult Care
Parents and medical providers oversee care and monitor symptoms	Care is self-directed and self-monitored by patients
Parents make treatment decisions and may shield patients from details regarding prognosis	Patients have final say in treatment decisions and must be able to handle discussing prognosis
Parents schedule appointments and find new providers as needed	Patients must schedule and keep appointments and find new providers on their own
Late appointments and last-minute rescheduling often tolerated	Being late or rescheduling appointments at the last minute may result in significant delays in care
Transportation provided by parents	Patient must provide own transportation
Social work and behavioral health services are integrated into pediatric subspecialty clinics	Patient must seek outside support services for financial and emotional issues
Parents are responsible for maintaining insurance coverage and healthcare payments	Patient is responsible for maintaining insurance coverage and healthcare payments
Parents manage home medications and home care treatments	Patient must obtain prescriptions and refills and manage home care needs

In the United States, at the time of transition to adult care, some TBD patients may encounter restrictions on adult subspecialty practices that are “in-network” with their insurance. While there are notable exceptions, adult subspecialty practices that are “in-network” may not be equipped with the same resources and connections to efficiently provide this same medical home after patients have transitioned to adult care. Another important distinction is that adult hematology and immunology physicians see fewer patients with TBD and are more familiar with later onset TBD presentations. This is in part because many patients with TBD have undergone

hematopoietic cell transplantation (HCT) by the time of adult care transition.

Consequently, adult hematology or immunology practices are not as central to the care of patients who have already undergone stem cell transplant, and are less familiar with complications seen in patients with more severe, pediatric-onset disease.

Options for maintaining a medical home for patients with TBD transitioning to adult care models thus include the following:

- Identify a regional expert in the care of TBD who practices in the adult care space that is willing to serve as a medical home. While historically such providers were rare, there are fortunately increasing numbers of internal medicine-trained subspecialists who are developing familiarity and expertise in caring for patients with TBD. At the time of transition, your pediatric provider can help provide a list of potential adult care providers in your area. Patients and families can work proactively with a social worker or insurance specialist to identify adult tertiary care practices or consider alternative insurance options to ensure access to a tertiary care center with TBD expertise. However, unfortunately, in the United States, “in-network” insurance restrictions may still create a barrier to accessing a specific regional center.
- Establish care in a dedicated primary care physician (PCP) office with providers trained in internal or family medicine who have experience in managing patients with chronic medical conditions. While this model can be quite successful, it does require establishing communication between this PCP and regional experts in TBD management to ensure an adequate level of TBD expertise.
- Maintain a medical home provider based in a pediatric practice, while transitioning subspecialty care to adult care providers. This option provides optimal continuity by including a provider with first-hand knowledge of a patient’s medical history in their adult care. However, this option requires that the pediatric provider’s hospital system, subspecialty practice, and the patient’s insurance plan allow ongoing medical care of adult patients within a pediatric practice.

Timing of Transition

At age 18, a person has the right to vote, make medical decisions, provide written informed consent to research studies or treatments, control who has access to their medical information, designate a health care power of attorney/health care agent, and create an advanced directive/living will. Whereas this legal transition from childhood to adulthood is abrupt, the real-world transition to adulthood is a gradual process that differs widely among individuals. Moreover, the transition from pediatric- to adult-oriented health care coincides with emerging adulthood in other life domains such as education, employment, social relationships, and independent living.

Ideally, the knowledge and skills needed for transition are learned over time and tailored to the developmental stage of the individual (Table 3). Young children are primarily recipients of care, with management provided by parents and medical providers. Grade School-age children should be provided with developmentally appropriate information and can often begin to participate in aspects of their care. As patients mature through adolescence, parents and providers should make efforts to engage adolescents in a shared management model, thereby beginning the transition process by having teens play an active participatory role in their care management [8]. By the end of the transition process, patients should have the skills to be the primary supervisors of their care, with parents and providers primarily serving as resources and consultants.

Table 3. Role transition in care management from early childhood to adulthood.

Adapted from Kieckhefer & Trahms, *Pediatric Nursing*, 2000.

Age	Provider	Parent/Family	Patient
Early Childhood	Major responsibility	Provides care	Receives care
Grade School Age	Support to family and patient	Manager	Participates
Secondary School Age	Consultant	Supervisor	Manager
Adulthood	Resource	Consultant	Supervisor

Determining Readiness: One Size Does Not Fit All

A number of factors influence transition readiness for individuals with chronic health care needs. In a 2013 study of adolescent and young adult survivors of childhood cancer, a stakeholder validation analysis identified several factors critical to successful transition [9], of which some pre-existing factors can be challenging to change, while others can be modified through a comprehensive transition program (Table 4). For example, while sociodemographic factors, health care access, and global cognitive ability are often static factors, at least in the short term, efforts to promote medical literacy, self-sufficiency, and even anger management/maturity skills through training can greatly facilitate the success of pediatric to adult transition.

Table 4. Patient-specific factors influencing transition timing. Adapted from Schwartz et al, 2013.

Pre Existing Factors	Modifiable Factors
<ul style="list-style-type: none"> ● Social and demographic factors ● Health care access/insurance ● Medical status ● Cognition 	<ul style="list-style-type: none"> ● Knowledge ● Self-sufficiency ● Expectations ● Maturity ● Motivation ● Communication ● Emotions

In practice, for most youth, transition to adult care takes place around the age of 22. However, because the pediatric–adult transition process is influenced by a variety of personal, condition-specific, and sociocultural factors, there is no one set of recommendations and milestones applicable to all individuals, particularly in a heterogeneous disease such as TBD where the care needs are widely variable. There are, however, clues and milestones that suggest a young adult is ready for transition to adult care. These include:

- Lack of comfort in addressing important areas of health with a pediatric provider
- Feeling out of place in the overall environment of a pediatric clinic
- The preferred family support at clinic visits is a boyfriend, girlfriend, or spouse as opposed to a parent, or if a patient is themselves a parent.
- Subspecialists involved in the care team begin to require transition per hospital policies
- Completion of formal education

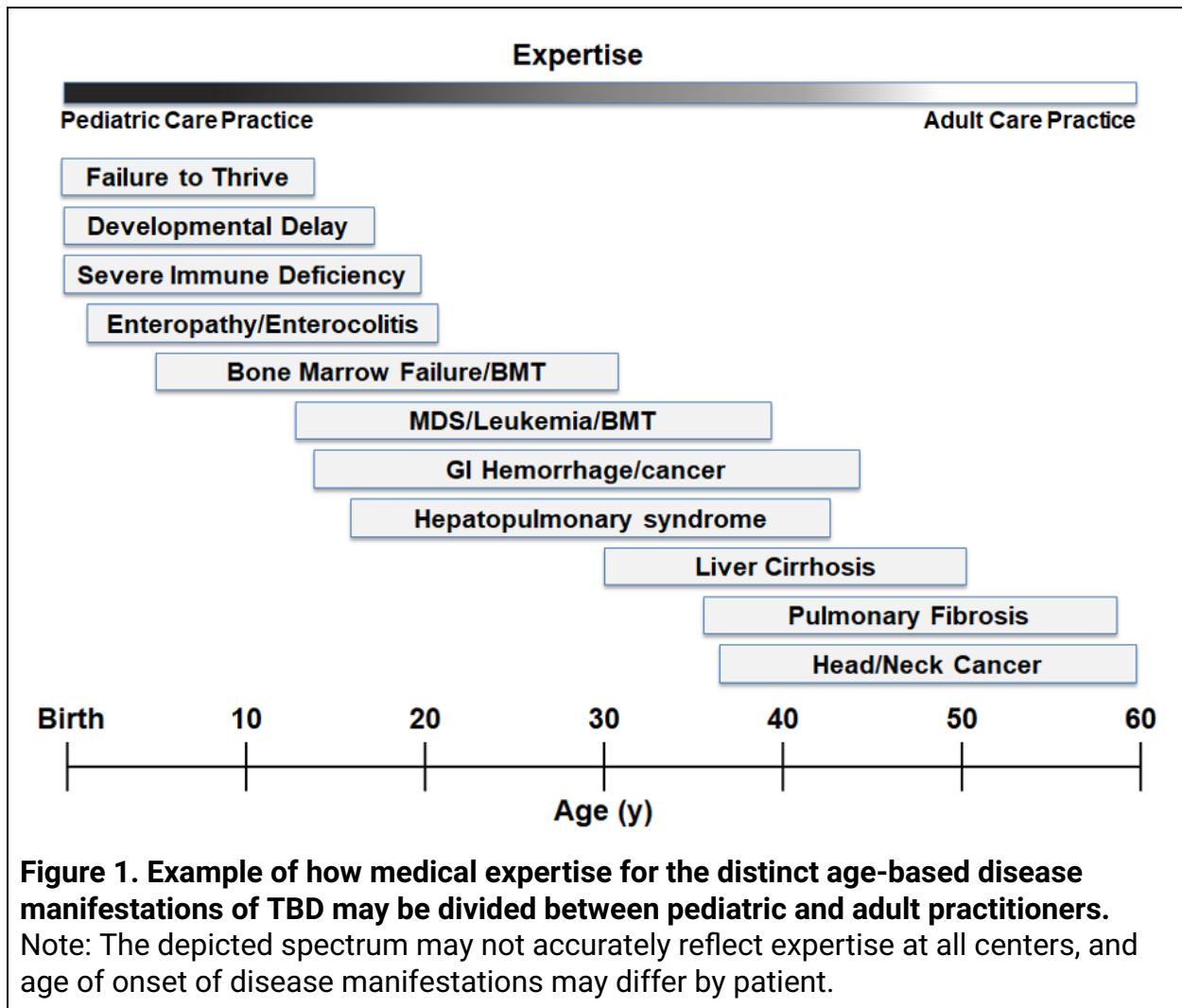
Disease-Specific Factors Influencing the Timing of Transition in TBD

Patient-specific, pre-existing factors heavily influence the timing and nature of the transition to adult care for patients with TBD. For those with severe forms of TBD in whom the diagnosis has been made in early childhood, the focus of pediatric-adult transition is often transitioning multiple previously established subspecialty services to adult care. For these patients, this transition may be occurring amid an overall adjustment to the myriad of challenges of young adulthood while they continue to have multiple long-standing medical limitations. In other cases, TBD may be diagnosed in late adolescence and may present with bone marrow failure or perhaps liver or hepatopulmonary concerns. These patients may need to decide whether to establish new subspecialty care in pediatric versus adult centers at a time where they are still learning about their diagnosis and what medical services they may need.

Because TBD is a rare condition, there are significant geographical differences in access to disease-specific expertise. Tertiary care options may also be quite limited for patients living in some regions. This limited access may result in patients being justifiably reluctant to leave local pediatric providers with whom they have a strong, well-established relationship. A related problem that sometimes limits access to expert care is that private- and state-sponsored health insurance programs for children are often flexible in allowing children with chronic medical needs to seek out medical expertise, even if this means crossing state or provincial boundaries. In contrast, health insurance programs for adult patients may impose restrictions in accessing the same degree of expertise in adult care centers, particularly if expert providers are not a part of the treatment network.

Another major factor in deciding the optimal time to transition to adult care is the set of organ-specific disease manifestations in individual TBD individuals (Figure 1) and the availability of adult subspecialty providers in their local area that can address these

specific complications (Figure 1). For example, expertise in severe immune deficiency, developmental concerns, and even enterocolitis associated with TBD that often occur in early childhood are most readily found in pediatric health systems. In contrast, complications that are seen more frequently in adults with TBD, such as pulmonary fibrosis, head and neck cancer, and end-stage liver cirrhosis, are more frequently cared for by adult medicine practices. Complications that come to the forefront in adolescence and young adulthood, such as bone marrow failure (BMF), development of myelodysplastic syndrome (MDS) or leukemia, worsening gastrointestinal hemorrhage seen in some TBD patients [10], and the increasingly recognized hepatopulmonary syndrome [11] often present the most challenges when considering transition. While expertise in BMF/MDS diagnostics and treatment with HCT typically exist in both pediatric and adult tertiary care practices, patients with GI hemorrhage or hepatopulmonary syndrome may be reluctant to transition to adult care while the acuity of these complications is increasing, despite the fact that adult liver transplant and interventional gastroenterology practices may have greater expertise in managing these complications. Thus, for young adult individuals who are struggling with these complications, it is critical to ensure a comprehensive transition plan focused on maximizing communication between pediatric and adult care teams across subspecialties.



Unfortunately, many patients with TBD also experience neurocognitive or neuropsychiatric deficits. In severe forms of TBD such as Hoyeraal-Hreidarsson and Revesz syndromes, the severity of neuropsychiatric deficits may make the transition to independent medical management infeasible. Nonetheless, many of these individuals will develop complications that are best managed using expertise from adult providers. These patients particularly benefit from formal identification of a medical home, whether from a pediatric provider or one specializing in caring for adults with disabilities, as long as this medical home can involve all necessary subspecialists to create an individualized, cohesive network of care.

Persons with neurocognitive disabilities may have the capacity to designate a healthcare or medical power of attorney, even if they lack the capacity to make more complicated medical decisions for themselves [12]. In other cases, state or other government oversight institutions may have laws or procedures to automatically designate a family member to act as the surrogate decision-maker for individuals who lack the capacity to make medical decisions and are unable to designate a medical power of attorney. Therefore, when significant neurocognitive concerns exist, transition planning should also include assistance with the process of pursuing legal guardianship for families who wish to do so.

General Tips for Focusing on Modifiable Factors to Improve Transition Outcomes

Despite the challenges encountered during the transition process, families and providers who focus on factors that are most easily modifiable (Table 4) can overcome many of the potential obstacles and ensure optimal care through this critical period and beyond. In particular, teens must gain the necessary skills and the knowledge necessary to maintain high-quality health care.

As a basic guideline, between the ages of 11 and 13, most patients should be developmentally able to understand their medical condition and their basic health care needs. This includes being able to:

- Briefly explain their condition to others
- State which doctors they see and for what reason
- State what medicines they take and for what reason
- Take their medications without reminders
- Know their allergies

Patients in this 11-13 year-old age range, with the help of techniques such as a 3-sentence health summary (Table 5) should be able to easily discuss health information with their regular providers, as well as in emergency situations.

By 14 to 16 years of age, developmentally able patients should be directing most of their communications with health care providers. They should also have the opportunity to discuss aspects of their care privately with providers. They should fully understand emergency health care plans and be engaged in accessing community resources to help them navigate school/work challenges posed by their medical condition. In addition, they should begin to understand the processes of health insurance and making appointments.

By late adolescence and early young adulthood (17-22 years of age), patients should be able to schedule their own appointments and arrange necessary transportation. They should have full knowledge of their health insurance coverage and have a sense of how they will maintain coverage in the future, based on their career plans and goals. They should also play the primary role in maintaining health records by using a “care binder” notebook or another system for organizing health information. They should be able to update and maintain access to written health summaries and emergency care cards. Resources that can help adolescents and young adults acquire these skill sets are listed in Appendix 1.

Table 5. Patient-specific factors influencing transition timing. Adapted from Schwartz et al, *JAMA Pediatr* 2013.

3-Sentence Health Summary
<p>Method:</p> <p>Sentence 1: State your age, diagnosis, and most important points about your medical history.</p> <p>Sentence 2: State any treatment you are currently receiving.</p> <p>Sentence 3: State what your concerns or symptoms are now.</p>
<p>Example:</p> <ol style="list-style-type: none">1. I am 16 and have a Telomere Biology Disorder called dyskeratosis congenita. I have bone marrow failure and problems with my immune system, and I have had several severe lung and skin infections.2. I receive red blood cell and platelet transfusions once in a while when my counts are low and have required admission to the hospital before when I have had an infection.3. I am here today because I have a fever and a cough, and I just don't feel well.

How Can Parents Help?

While turning over autonomy for healthcare to their adolescent and young adult children can be understandably anxiety-provoking, empowering their child with the skills and confidence to manage their own care are the most critical contributions parents can make to ensuring continued high-level care. A few general tips for how parents can facilitate their child's transition are:

- Start early
- Teach developmentally appropriate information about your child's condition
- Encourage your teen to assume responsibilities
- Provide coaching opportunities and practice independence
- Help your teen understand the future health implications of their condition
- Discuss career and educational goals and their impact on health insurance
- Address decision-making and guardianship issues when necessary

Transition as a Challenge That Must Be Met

Becoming a successful young adult is challenging, even for adolescents who do not have chronic medical conditions such as TBD. Transitioning medical care from pediatric into adult systems is unmistakably an added burden for young adults with TBD during this already difficult developmental period. However, youth who experience a successful, systematic transition have better health outcomes and report improved rates of fulfillment, achievement, and higher self-esteem compared to those who do not attain a smooth transition. Pediatric and adult care providers, along with parents and other family members have an obligation to provide these individuals not only with exceptional medical care, but also the knowledge and skills necessary to ensure an optimal transition experience.

“Every new beginning comes from some other beginning’s end.” – Seneca the Younger

Appendix: Tools and Resources

Websites Focused on Pediatric to Adult Transition and Medical Home Resources

- National Health Care Transition Center: <http://www.gottransition.org>
- American Academy of Pediatrics:
<https://medicalhomeinfo.aap.org/Pages/default.aspx>
- Florida Health and Transition Services: <http://www.floridahats.org>
- New York State Institute for Health Transition Training for Youth with Developmental Disabilities: <https://healthytransitionsny.org>
- Children’s Hospital of Philadelphia Transition to Adulthood:
<http://www.chop.edu/centers-programs/transition-adulthood-program>
https://policylab.chop.edu/sites/default/files/pdf/publications/Transitions_Of_Care.pdf

Building a Health Care Summary and Maintaining Medical Records

- 5-page health summary:
 - https://www.acponline.org/system/files/documents/clinical_information/high_value_care/clinician_resources/pediatric_adult_care_transitions/general_hema/general_hematology_clinical_summary.pdf
 - <https://www.hematology.org/education/clinicians/clinical-priorities/pediatric-to-adult-care-transition>
- Care binder:
<https://www.medicalhomeportal.org/living-with-child/after-a-diagnosis-or-problem-is-identified/caring-for-children-with-special-health-care-needs/managing-and-coordinating-care>

Emergency Plans/Cards

- My Health Passport: <http://www.sickkids.ca/MyHealthPassport>
- Emergency Information Card:
https://www.redcross.org/content/dam/redcross/National/m4240194_ECCard.pdf

Tips for College

- <https://www.familyeducation.com/a-parent-guide-to-managing-chronic-illness-in-college>

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