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Late Consequences of Pediatric Chronic Illness

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Introduction

With the advent of new treatments for chronic pediatric disorders such as cystic fibrosis, juvenile rheumatoid arthritis, and congenital heart disease, more children and adolescents are surviving into adulthood than ever before. Seventy years ago, individuals with cystic fibrosis survived an average of 5 years, while currently the life expectancy for cystic fibrosis is over 30 years [1]. Increased survival has brought new morbidities [2], and may affect psychosocial outcomes of adult life [3]. The prevalence of children suffering from a chronic illness varies widely, but the overall rate is 10% to 20% [4]. Children with chronic illnesses are more likely to have emotional, behavioral and psychiatric symptoms than healthy children [5] and may be psychologically affected or traumatized by medical treatments [6]. On the other hand, resilience is common [7] and chronically ill children do not inevitably develop psychiatric difficulties. This chapter is aimed at helping psychiatric consultants understand how medical, developmental and psychosocial needs are altered in adults who have grown up with chronic pediatric illnesses. Three childhood conditions, congenital heart disease, cystic fibrosis, and rheumatologic disorders, will be discussed in detail, using these conditions as models to illustrate the impact of congenital malformations, genetic disorders, and typically adult disorders occurring in the pediatric age group.

Evaluating Chronically Ill Children and Adolescents

Three aspects of psychiatric consultation in the medically and surgically ill that are specific to working with youth include: 1) an awareness of the cognitive and emotional developmental levels of the patient, 2) the essential role of the family, and 3) a focus on facilitating coping and adjustment to illness in order to follow an optimal developmental trajectory, rather than focus on psychopathology. Clinicians need to be familiar with normal physical, motor,

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language, cognitive, sexual, and emotional development in chronically ill children in order to distinguish normal responses to stress from unhealthy ones [8]. Understanding a child's cognitive ability to process information is essential when communicating about his/her disease. Clinicians cannot assume that chronologic age is equivalent to mental age. Children with medical illness may not develop at the same rate as healthy children because of delayed neurocognitive development, disruptions in education and limited social experiences in addition to the medical condition and treatment influences on intellectual and somatic growth and maturation [9].

The hospital or clinic environment is often distressing or even traumatic for the chronically ill child. Injections, procedures, and surgeries are highly stressful experiences for children. Pain from both medical conditions and treatments can provoke anxiety and affect later pain sensitivities and neurological development [10]. Post-traumatic stress disorder (PTSD) is a risk from traumatic injury or intense hospital experiences such as transplantations. Identifying and easing potentially traumatic situations may decrease the child's stress and improve medical outcomes [11]. Adult clinicians should inquire into a patient's childhood medical disorders and treatments as these early experiences surely influence the patient's trust and utilization of the medical system as they develop into adults.

Impaired growth and development and Impact of Chronic Steroid Use

Physical growth is a dynamic process that starts at conception and ends after full pubertal development [13]. Chronic illness may lead to growth retardation, either because of the illness itself, or because of treatments required for it. Short stature is commonly perceived to be associated with social and psychological disadvantage [14]. Parents often attribute behavioral disorders, anxiety, depression, social and attentional problems to short stature, and are concerned that their children are subjected to height related stressors of being teased or infantilized [14]. However, it is difficult to determine if problems in psychosocial functioning are due to the underlying illness, treatment, or resultant effects such as impaired growth.

Long term administration of systemic corticosteroids (e.g. dexamethasone, prednisone) is a major cause of impaired growth but is often required for children and adolescents with a range of chronic inflammatory, autoimmune, and neoplastic diseases. They are also often used to treat inflammatory bowel disease, asthma, cystic fibrosis, bone marrow and solid organ transplants, nephrotic syndrome and other causes of renal failure, systemic lupus erythematosus, and juvenile arthritis. Children and adolescents with these conditions are at high risk for growth failure, both from their underlying disease and from glucocorticoid therapy.

Multiple mechanisms play a role in glucocorticoid impact on bone development and growth. In the short term, bone loss and deterioration depend on the type and dose of glucocorticoid, and occur most prominently in the first six months of treatment. Treatments directed at preventing bone loss during this period are more effective than attempts to compensate for lost growth later on [15]. Glucocorticoids have direct effects on the growth plate and disrupt growth plate vasculature. They have a suppressive effect on osteoblastogenesis, and promote apoptosis of osteoblasts and osteocytes. This may lead to decreased bone formation and osteonecrosis or avascular necrosis of bone. Glucocorticoids may promote calcium loss through the kidneys and gastrointestinal tract, increasing bone remodeling and osteoclastic activity due to secondary hyperparathyroidism. High dose glucocorticoid therapy can attenuate physiologic growth hormone secretion and increase somatostatin tone, and may also impair attainment of peak bone mass and delay growth through direct effects on gonadotrophin and sex steroids [16].

Growth retardation has been reported in children with chronic inflammatory bowel disease (IBD), including ulcerative colitis (UC) and especially in those with Crohn's disease (CD) [17]. Typically children with IBD grow more slowly before diagnosis and when disease is active. Growth retardation has been reported in 15% to 40% of children with IBD [18]. Decreased height velocity may be the earliest indicator preceding the diagnosis of CD. Chronic low nutrition is generally considered an important reason for growth impairment. Treating IBD may restore growth velocity, but ultimately the prolonged use of glucocorticoids may itself lead to growth retardation. Eventual height is usually normal in UC and near normal in CD, and delayed puberty may compensate for the period of poor growth earlier in life [17]. In pediatric patients with IBD treated with glucocorticoids, there is also a higher incidence of osteoporosis, glaucoma, and cataracts compared to adult patients [19]. Steroid sparing agents, such as 6-mercaptopurine may prevent growth retardation associated with chronic steroid use [18]. Mercaptopurine and its prodrug, azathioprine, are effective in maintaining remission in children with CD and may improve growth velocity and final adult height by controlling the disorder and sparing the child long term glucocorticoid treatment [20].

Children with chronic renal disease may also have growth retardation, and are often dependent on glucocorticoid treatment to control their disease. Prednisone is associated with impairment of growth and body height in a dose dependent fashion [21]. Children with severe steroid-dependent nephrotic syndrome are at risk of permanent growth retardation due to prolonged courses of steroid treatment [22]. Suboptimal final height and marked weight gain are common after renal transplant, and may result in significant obesity. After transplant, some children show improved growth, but height remains suboptimal, and steroids needed to maintain the transplant contribute to obesity [23]. Management of growth retardation before transplantation and further reduction in the steroid dose after transplantation may increase final height of children with chronic renal failure [24].

In children with severe rheumatic disorders, treatment with glucocorticoids is frequently needed and is associated with growth retardation and osteopenia [25]. Growth hormone treatment may improve growth and lean body mass, but these benefits disappear when growth hormone therapy is stopped. Long term growth hormone treatment is necessary to maintain a potential positive effect on bone density and metabolism [26]. Children with mild or moderate juvenile idiopathic arthritis disease and lower medication requirements responded better to growth hormone therapy than those with active disease [27]. Using growth hormone earlier may prevent growth deterioration and metabolic complications induced by chronic inflammation and prolonged steroid therapy [28]. Chronic inflammation and prednisone therapy may adversely affect growth, and final height may be closely dependent on both severity of growth retardation during the active phase of the disease and on linear growth after remission [29]. After remission of active disease and discontinuation of prednisone treatment, 70% of children will show catch-up growth, but 30% show persistent loss of height [30]. This has led to the recommendation that early initiation of growth hormone therapy may prevent growth deterioration and other metabolic complications induced by chronic inflammation and long term steroid therapy [30]. Wider use of growth hormone in children and adolescents with rheumatic disorders is not without risk, however, and growth hormone may lead to a flare of previously well controlled systemic lupus erythematosus [31].

Children with bronchial asthma, allergic rhinitis and atopic dermatitis have a 2–5 times higher incidence of short stature, skeletal retardation and delayed puberty. This is likely secondary to the severity and underlying mechanisms of their disorder. Local growth factor prostaglandin E(2) (PGE(2)), which is important in bone mineralization, is a messenger substance for both the immediate and late allergic reaction. The platelet-activating factor (PAF) is one of the strongest mediators in the pathogenesis of allergic disorders and it influences the PGE(2)

synthesis in osteoblasts [32]. Inhaled and nasal glucocorticoids rarely suppress adrenal function, though they may decrease prepubertal growth [33].

Children with CF have reduced growth velocity and delayed adolescent growth spurt [35]. This may result from a combination of poor nutrition, pancreatic insufficiency, chronic inflammatory lung disease, and intestinal disease. Even with vigorous treatment of these problems, including dietary interventions to provide calories and fat soluble vitamins in excess of usual recommended amounts [35], severe CF is associated with poor weight gain and slower growth [36]. Relative insulin deficiency rather than nutritional deprivation or poor clinical status may be implicated in the poor linear growth of children with relatively stable lung disease [36].

Abnormal growth in CF may also be related to the primary dysfunction of a cyclic-AMP-regulated chloride channel. The gene for this chloride channel, CFTR, is found in the thalamus, hypothalamus, and amygdala of the brain, sites related to regulation of appetite, energy expenditure, and sexual maturation [35]. Inhibition of CFTR inhibits secretion of gonadotropin-releasing hormone in cell lines [34]. Failure of CFTR function may be related to inhibition of pubertal maturation as well as growth [34].

Twenty percent of all children in the 1993 National CF Patient Registry were <5th percentile, for height or weight for age [37], 28% were below the 10th percentile for height and 34% were below the 10th percentile for weight [38]. Growth hormone treatment enhances nutrition, linear growth and weight in children with CF [38], in prepubertal children may improve height, weight, bone mineral content and lean body mass [39], and may reduce the number of hospitalizations with or without significantly changing pulmonary function, even in children receiving enteral nutritional supplementation [39,40]. Children homozygous for the Delta F508 mutation, which is associated with the most severe disease, fail to normalize growth despite improved care [41].

Compounding these growth problems, children with CF are often treated with glucocorticoids. Attempts to mitigate the effects of steroids with alternate therapy have yielded equivocal results, and while girls may regain their growth, boys have persistent growth impairment, even when the treatment is discontinued [34]. Focusing on reduced height alone as an important adverse effect of glucocorticoids may reflect cultural rather than medical imperatives [35]. Glucocorticoids may also compound the risk of diabetes, cataracts, and osteoporosis, [35] which are not uncommon in CF itself.

The general consequences of malnutrition in CF may include growth failure, increased mortality, delayed puberty, decreased physical well-being, and psychological disorders [42]. Short stature and pubertal delay emphasize the differences between patients with CF and their peers, and may result in a greater impact on their quality of life than the longer term issues of compromised survival [42,43]. Adjustment and self esteem in patients with CF are less than peers, especially in girls [43]. With intensive, coordinated care in a cystic fibrosis center, outcome and growth may improve independent of improved pulmonary function and normalize in most children with CF [41].

Growth itself is an important prognostic factor in survival of patients with CF. Analysis of data from 19,000 patient records of the National Cystic Fibrosis Patient Registry, showed that shorter patients are much more likely to die before taller patients [44]. Short stature in CF may be a marker of more severe disease [44]. The problem of worsening growth failure in children and adolescents with CF ironically is compounded by increased survival [42]. As patients live longer and their clinical symptoms become more severe, nutritional status may worsen, and growth remains impaired [42]. It is important that linear growth be maximized through medical and nutritional intervention [44]. Bone mineralization is decreased and rates of osteoporosis

are significantly increased in patients with CF as they age; adults with late stage CF are at high risk for fractures and severe kyphosis [45].

Psychosocial Considerations

Normative developmental tasks throughout childhood center on developing a sense of self and acquiring autonomy in all areas of life. However, chronically ill young adults with a wide variety of disorders, when compared to their cohort in the general population, have lesser academic and employment achievement, less vocational education and less permanent employment [45], are more likely to be single [47] and have delayed independence [3]. Cohorts have not been followed long enough to establish whether this group “catches up” developmentally at a later point in adulthood, which might allow for survivors to make the transition from dependence to independence in their relationships and work.

Cross-sectional data on almost 100,000 children younger than 18 years in the 1992–4 National Health Interview Survey showed that an estimated 6.5 % of all US children experienced some degree of disability (defined as a long-term reduction in ability to conduct social role activities such as school or play) because of a chronic condition [48]. The presence of a childhood disability is associated with elevated use of health care services, and these youth are four times as likely to be hospitalized and have eight times as many total hospital days as the general population. As these children survive longer, physicians have become increasingly aware that the natural progression of pediatric disease and the consequences of treatment can adversely affect brain development, and compromises in cognitive abilities leading to mild impairments may have more permanent consequences in adolescence and adulthood [9]. Thus, childhood disability from chronic conditions significantly affects the educational and health care systems in the long term. Additionally, added caretaking demands and lost income for parents and the eventual detrimental impact of childhood disability on social and economic status in adulthood all take a toll [48].

Stage or severity of illness, including the degree of life threat, do not independently predict a person’s adjustment to chronic illness. Low self-esteem in childhood, poor school attendance, and family factors all play a significant role. Older children, boys, children from poor families, and those from single-parent households have a significantly higher prevalence of disability even after multivariate analysis [48]. Medical illness may have some protective effects. For example, in IDDM, young adults have less frequent drug use, fewer criminal convictions, and less cigarette smoking [49].

The high degree of variation in outcome measures such as restricted activity, school loss days, severity of limitation, and use of medical services for different childhood conditions is notable and suggests that non-categorical approaches for studying chronic illness disability may not be adequate. Consideration of individual underlying conditions may be more relevant when developing health and social policies and improving outcomes of specific chronic conditions. This is additionally supported by a literature review of the efficacy of psychological interventions for pediatric chronic illnesses, which found 19 studies since 1980 that met criteria of external and internal validity for diabetes, CF, cancer and sickle cell disease but, overall, showed a lack of evidence of what sort of intervention is best for whom [51]. Interventions need to be individually targeted and efficacy of different approaches needs to be assessed.

Psychiatric Consequences

Investigations of psychiatric disorders in pediatric conditions have been limited by small and varied demographic samples, lack of consistent testing measurements, frequent sub-threshold DSM-IV diagnoses, and lack of appropriate control groups. The full range of developmental and childhood psychiatric disorders including adjustment disorder, major depression, anxiety,

and delirium are seen in children and adolescents with chronic illness. Psychiatric disorders are most likely to be present when the chronic physical disorder involves the brain. Some aspects of treatment of life-threatening medical illness may be experienced as repeated trauma, which might not impact during or immediately after treatment, but rather appear as long term effects on affect modulation and interpersonal relationships [52].

Congenital Heart Disease

Depression is common in patients with congenital heart disease, and can exacerbate the physical consequences of the illness [53]. Children with congenital heart disease may have more medical fear and general fear of the unknown, physiologic anxiety, depression, and delinquent behaviors than the general population [54]. Those with cyanotic malformations may be at higher risk for these problems, which may be further exacerbated by maternal anxiety [54]. Adults with more complex, cyanotic disorders may have more problems with depression than those with less severe lesions [55]. Slightly over 36% of adults with congenital heart disease meet DSM-IV criteria for depressive episode or generalized anxiety disorder, and an additional 27% meet criteria for problematic emotional functioning [55]. Risk for either depression or anxiety is significantly correlated with medical severity [55]. However, current research demonstrates a wide range of psychiatric disturbances in adult survivors of congenital heart disease, including some studies which demonstrate a lower prevalence of psychopathology [56]. But even when patients are found to have symptoms of psychiatric illness, they rarely receive psychiatric treatment [56,57]. Once surgery and hospitalizations are in the past, adult patients may strive to obtain a normal life, and having survived a life-threatening illness, they may not worry about minor difficulties and ultimately achieve good social and vocational adjustment [56].

Cystic Fibrosis

As a group, adults with cystic fibrosis do not demonstrate significant levels of depression, anxiety, or other psychopathology [58]. Psychological and psychosocial functioning in patients with cystic fibrosis are similar to that of healthy peers, at least until their disease becomes severe and their physical and social activities become increasingly limited [59]. They may then have an increased risk for psychiatric problems, such as depression, and typically score poorly on physical functioning measures in quality of life assessments [59]. Coping styles have a large effect on quality of life, and compliance is a complicated problem for many patients [59]. Men may be at higher risk for depression and anxiety, and better lung function predicts less anxiety. Higher level of psychosocial support is a strong predictor of better psychological functioning [58]. Major psychiatric illness may also occur in adults with cystic fibrosis, and both bipolar disorder [60] and paranoia [61] have been reported.

Rheumatic Disorders

Pediatric rheumatic disorders are more common in girls than boys, and affect about 200,000 children in the United States. Juvenile rheumatoid arthritis accounts for 75–85% of rheumatic diseases affecting children. These disorders are not curable, and the main goal of treatment is disease management, which includes reducing pain, controlling inflammation, maintaining function, and preventing deformities or persistent organ failure.[62] Rheumatologic disorders in pediatric patients are typically more severe clinically than the same disorders in adults, and may have significant psychiatric and medical consequences. These conditions are treated with potent immunosuppressive medications, steroids, and immune modulators that also may be associated with psychiatric side effects. Psychiatric symptoms may be a reflection of an autoimmune process or related to the underlying vasculitis, or the psychiatric disorder may be unrelated and coincidental. Children with rheumatologic disorders are at an increased risk for adjustment problems, particularly internalizing problems such as anxiety and depression, often

seen in association with higher levels of family stress. Higher social support reduces the risk for these adjustment disorders [62]. Classmate and parental support appear to be the best predictor of adjustment [62], and in contrast, poor body image and lack of satisfaction with social support are most predictive of poor psychosocial adjustment and function [63]. Parental functioning appears to play a role in determining the long term effects of psychiatric complications of pediatric rheumatologic disorders, and parental distress and maternal depression are associated with more behavior problems in the ill child [64].

Juvenile Rheumatoid [Idiopathic] Arthritis (JIA) is the most common rheumatologic disorder in childhood. Central nervous system involvement is rare in JIA, but depressive and anxiety disorders are common and are attributed to social isolation, chronic pain, and deformity. In adults with long standing JIA, who on average had 28 years of illness, 31.6% were anxious, 5.2% were depressed, and 21.1% had previously suffered from depression [63]. Pain intensity and level of anxiety were higher in this group of adults than in children and adolescents with JIA [63]. Both physical and psychologic factors influence pain, and psychologic variables explain the majority of variance in depression and anxiety in adults with JIA [63].

As a patient enters adulthood, pain coping strategies become an important predictor of pain, and higher levels of denial and dependence are detrimental to pain management [63]. The age of onset of disease may have an effect later in life on the effectiveness of learned coping strategies to avoid anxiety of depression, and a later age of onset is associated with impaired self identity and self confidence and a greater sense of loss [63].

Systemic lupus erythematosus (SLE) is the prototypical generalized autoimmune disease, with variable clinical features affecting all organ systems. Central nervous system involvement occurs in more than half of children and adolescents with SLE, usually early in the course of the disease, and typically includes both neurologic and psychiatric symptoms [65,66,67]. Cognitive, mood, and psychotic symptoms in CNS lupus may be related to an underlying vasculitis, or may reflect the impact of anti-phospholipid, anti-neuronal, anti-receptor antibodies. While there are numerous studies of psychiatric problems in adult patients with SLE, and some describing mood, cognitive, and psychotic problems in pediatric SLE patients [68], there are no outcome studies of the long term consequences of pediatric psychosocial dysfunction.

The presence of delirium, psychosis, confusion, depression, or mania in a patient with SLE suggests primary CNS involvement, and the diagnosis of CNS-SLE should be suspected when neuropsychiatric symptoms occur in patients known to have SLE, and investigated in children and adolescents with the acute onset of delirium or profound psychiatric symptoms with or without neurologic symptoms [68].

SLE is associated with both primary and secondary effects on psychosocial functioning. The prevalence rate of depressive symptoms with SLE is 30% [69]. Depression may be related to the effect of pain and fatigue on mood symptoms [70]. Psychosis, depression, anxiety, cognitive deficits and emotional distress are frequently seen in patients with SLE [71]. Depression and other neuropsychiatric symptoms in patients with SLE are associated with increased risk for suicidal behavior [72]. Fortunately, depression in patients with SLE usually responds to antidepressant therapy [69].

Depression may predict cognitive dysfunction in patients with SLE [73]. Emotional disturbances and problems with social functioning, personal discomfort in social situations, and depressed mood are more frequent in SLE patients with skin and joint abnormalities, confirming that psychosocial dysfunction may not only be a reflection of direct CNS involvement [74].

The pathogenesis of neuropsychiatric SLE has been attributed to autoantibody mediated neural dysfunction, vasculopathy, and coagulopathy. Several autoantibodies have been reported in serum and cerebrospinal fluid, including anti-neuronal, anti-ribosomal P proteins, anti-glial fibrillary acidic proteins, anti-phospholipid, and anti-endothelial antibodies [75]. Anti-Nedd5 antibodies have been found to be associated with psychosis in SLE patients [75]. Anti-N-methyl-D-aspartate receptor antibodies have been associated with neuropsychiatric symptoms in SLE, but their presence alone does not explain cognitive dysfunction, depression, or anxiety in patients with SLE [76]. Anti-NR2a antibodies may be associated with depressed mood but may not be associated with cognitive dysfunction in patients with SLE [77]. Anti-neuronal antibodies and abnormalities seen on SPECT scan appear to be useful in diagnosing CNS-SLE in pediatric patients [68].

Transition to Adult Care

Adolescence is a complex period of transition from childhood to adulthood. Having a chronic illness adds to the complexity (Table 1). Issues of puberty, autonomy, personal identity, sexuality, education and vocational choices become more difficult for an adolescent who is also coping with chronic illness. This period may be further complicated by medical setbacks, impaired physical or mental abilities, forced dependence, and perceived poor prognosis [78]. For adolescents with a chronic illness, perception of their illness severity is directly related to their psychosocial well being [79]. A generation ago, few children with severe chronic illness or disability survived to 21 years, and issues of transition from pediatric to adult health care were rarely considered [78]. But now over 90% of children with chronic or disabling conditions will survive beyond the second decade, and over 30% of young people 10 to 17 years old have a chronic condition [80].

The previously unanticipated problem of transitioning a pediatric patient with congenital heart disease, malignancies, rheumatologic disorders, CF, or transplants to adult care is beginning to be addressed. There are two different approaches to planning for the transition of adolescents with chronic medical problems to adult medical care. One approach is transfer to a specialized center with care focused on pediatric disorders in adults, such as with CF or congenital heart disease [81]. The other involves the transfer to adult specialists of patients with the pediatric onset of what is usually considered an adult disorder, such as IBD or rheumatologic disorders [82]. Either way, the process of transition is usually begun by discussing the planned transfer to adult care long (as much as a year or more) before it is anticipated to occur, and ideally the process should be a guided educational and therapeutic transition rather than an administrative event [83]. Transition can be viewed as a positive step as the patient graduates from a pediatric to adult program. It may be difficult for chronically ill adolescent patients to break ties with their pediatrician, to feel accepted by their peer group, and to plan realistically for the future [84], although typically parents and pediatricians are more concerned about the transition than are the patients and their adult physicians [85].

It is often difficult for pediatricians to let go of their patients, and the age limit of pediatric practice has been steadily increasing. In 1938, the American Academy of Pediatrics defined the limit of pediatric practice at 16 to 18 years. By 1972, this age limit had increased to 21 years. In 1990 the American Academy of Pediatrics stated that the services of a pediatrician may continue to be the optimal source of health care past the age of 21 years, which may have the effect of prolonging entry into adulthood for young individuals with chronic diseases [85]. In 1989 the Surgeon General of the United States convened a multidisciplinary conference to address strategies for the implementation of medical care systems for growing youth with special needs, and a transition manual [86] was published to aid health professionals in the transfer of chronically ill adolescents to adult care systems. The British National Health Service has highlighted the importance of ensuring safe and effective transition from children's services

into adulthood [87]. The American Academy of Pediatrics Committee on Children with Disabilities and Committee on Adolescence issued a policy statement addressing issues of transition in 1996 which remains the current standard [88].

Transfer is not the same as transition. Transfer occurs when information or people move from pediatric to adult care. Transition is a multifaceted process that addresses the psychosocial, educational, and vocational needs of adolescents as they move from child-oriented to adult-oriented care. Ideally, the patient should be provided a coordinated, uninterrupted care plan that is developmentally appropriate and comprehensive [89].

The primary barriers to effective transition are limitations in the health care system itself, related to funding and identification of appropriate resources, rather than family or adolescent resistance [80]. There is little agreement on the best methods or optimal time for transition from pediatric to adult care, but the process of transition can be improved by timely discussion between the patient and family and treatment team members in both the pediatric and adult settings [90]. There are now more than one million adults with congenital heart defects living in the United States who under current guidelines should be seen every 12 to 24 months by a cardiologist with specific CHD expertise to monitor for potential serious complications in this population [81]. Unfortunately, more than a quarter of patients have no cardiac follow-up after they turn 18 years old and pediatric cardiac care is discontinued [81]. Some patients fail to get follow-up for more than ten years and typically demonstrate a poor level of knowledge about their heart condition [91]. Failure to address transition properly during adolescence may result in an adult who cannot effectively be responsible for his/her own care [92].

Noncompliance with treatment, particularly prevalent in adolescents, requires attention to psychological and social issues as well as medical factors [93]. The young person must have sufficient self-management skills to adapt to adult-oriented medical systems, and long term social support may need to be established before the transfer is complete [90].

It is important that pediatric relationships are appropriately terminated as part of the transition process [94]. Pediatricians themselves can become barriers in the process of transitioning to adult care, and may resist the transition process because they lack confidence in their adult colleagues [95]. Pediatric and adult medical practice represent two different medical subcultures [94]. Often there is lack of communication, no common guidelines, and differences in the management of patients by internists compared to pediatricians [96]. For example, adult endocrinologists use different tests to re-evaluate diagnosis and higher doses of medications such as growth hormone to treat adults compared to pediatricians caring for adolescents [96].

A move towards a culture of personal responsibility for health care is crucial for the promotion of the maturing patient's independence [95]. The purpose of transition is to prepare the patient for the transfer to adult medicine by gaining an understanding of the clinical picture of their disease and of the treatment goals and possibilities, and developing a personal responsibility for medication and diet [97]. A carefully planned transition to adult health care should improve self-reliance, enhance autonomy and independence, and support young people in attaining their maximum potential and meaningful adult lives [98].

Summary

There are many challenges in coping with and adapting to life with a chronic disease, and increased survival cannot be assumed to be associated with increased quality of life. A recent systematic review shows there is wide variation in outcomes depending on the definitions and measurements used to estimate the prevalence of chronic health conditions, making the impact of disability on children's health and social functioning difficult to assess, and different authors have called for an international consensus about the conceptual definition of chronic health

conditions in childhood [99]. It is frequently difficult to determine if problems in psychosocial functioning are due to the underlying illness, treatment, or resultant effects of either on physical growth or cognitive development. Mental health assessment and treatment should be an integral component of comprehensive care of chronically ill children and adolescents. Transition of care is an important process that addresses significant changes from child-oriented to adult-oriented care. Adults with chronic health conditions should continue to be evaluated periodically for late consequences of their childhood illness and early medical care, and attention paid to their ongoing psychosocial, psychiatric, educational and vocational needs.

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TABLE 1

Differences between pediatric and adult care [Robertson 2006]

Pediatric care	Adult care
Family oriented	Individual focused
Developmental aspects considered	Specifically focused on health
Coordinates with schools and social services	Less communication with social services or workplace
More help with treatment regimen	More accepting of treatment refusal
More trainee supervision	Less trainee supervision
Paternalistic	Shared treatment decisions