Progress in Pediatric Cardiology 39 (2015) 113–118

Contents lists available at ScienceDirect

Progress in Pediatric Cardiology

journal homepage: www.elsevier.com/locate/ppedcard

Review

Neurodevelopmental and psychosocial outcomes across the congenital heart disease lifespan

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ARTICLE INFO

Available online 21 October 2015

Keywords:
Congenital heart disease
Neurodevelopmental
Neurocognitive
Psychosocial
Quality of life

ABSTRACT

Infants, children, and adolescents with congenital heart disease are known to be at elevated risk of neurodevelopmental deficits compared to healthy children, with the risk and severity of impairment increasing with greater complexity of the cardiac lesion. These deficits may intensify with time, becoming more pronounced as children attempt to master more complex tasks requiring integration of multiple cognitive domains. These deficits do not disappear upon transition to adulthood and there are increasing numbers of adults with congenital heart disease who are now reaching middle-adulthood and older-adulthood. The neurocognitive functioning of adults with congenital heart disease, however, has not received the attention of pediatric neurodevelopment outcomes and remains largely unknown. Challenges that may present in adulthood include poor performance in higher education as well as difficulties maintaining employment or interpersonal relationships. In addition to potential neurodevelopmental deficits, children and adults with congenital heart disease also face psychological and social challenges. In this review, we consider neurodevelopmental and psychosocial outcomes of individuals with congenital heart disease across the lifespan.

1. Introduction

As a result of significant advances in pediatric and surgical cardiology care, the majority of infants with congenital heart disease (CHD) now reach adulthood [1,2]. However, improved survival has been shadowed by significant neurodevelopmental morbidity with numerous studies demonstrating increased risk for both neurodevelopmental and behavioral impairment [3–8]. Such deficits do not disappear upon transition to adulthood and impact numerous patient-centered outcomes including functional status, psychosocial well-being and health-related quality of life (QoL). In this review, we offer a lifespan approach to review the neurodevelopmental and psychosocial outcomes of pediatric and adult patients with CHD.

2. Pediatric Neurodevelopmental Outcomes

2.1. What Is Known About Pediatric Neurodevelopmental Outcomes?

CHD survivors are at greater risk for neurodevelopmental deficits compared to healthy children with the risk and severity of developmental impairment increasing with greater complexity of the cardiac lesion [8]. Numerous risk factors have been implicated including underlying syndromes or genetic disorders, the circulatory abnormalities specific to the heart defect, required medical and surgical therapies, and the psychosocial stress of chronic illness [5,8]. However, known risk factors only explain approximately 30% of observed variation in neurodevelopmental outcomes, highlighting the complexity of this issue as adverse outcomes are multi-factorial, inter-related, and likely cumulative [5].

In infancy, the most pronounced neurodevelopmental deficit occurs in the motor domain [9]. However, by early childhood, multiple deficits begin to emerge as a distinctive pattern of neurodevelopmental impairment characterized by mild cognitive deficits, impairments in language (development, expressive, and receptive), impaired executive function, gross and fine motor deficiencies, difficulty with visual construction and perception, and inattention and impulsivity [8,10,11] These deficits may intensify with time, becoming more pronounced as children attempt to master more complex tasks requiring integration of multiple cognitive domains [6,8].

Although the majority of children and adolescents score in the low normal range for general intelligence as expressed by the intelligence quotient (IQ) score, significant variability exists, with a range larger than expected for the normative population [12]. Furthermore, children with CHD exhibit more pronounced deficits in domains requiring integration such as visual-spatial processing, developing complex narratives, and symbolic play [12]. Children with CHD are also at risk for

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http://dx.doi.org/10.1016/j.ppedcard.2015.10.011
1058-9813/

lower academic achievement as reflected in standardized testing in reading and mathematics [4,7].

Children and adolescents with CHD also have a much higher prevalence of attention deficit and hyperactivity disorder (ADHD), with one study reporting a rate 3–4 times higher than the general population [10]. Forty-six percent of parents with children who underwent Fontan palliation report issues with attention [13]. Children and adolescents with complex CHD are also at higher risk for impaired executive function, further exacerbating attention issues and also leading to worse working memory, deficits in organizational and planning abilities, and behavioral dysregulation [14,15].

This broad spectrum of neurodevelopmental impairment leads to a higher likelihood of learning disabilities, abnormalities of speech, language, and behavior and use of special services [8,10]. Nearly half of children 5 to 10 years of age with complex CHD receive at least one remedial service in the form of tutoring, early intervention, occupational therapy, or special education [7]. By adolescence, this number increases to 65% for children with D-transposition of the great arteries (TGA) and 82% for children with tetralogy of Fallot (TOF) [10,11].

2.2. Opportunities for Intervention

Research is still required to elucidate the neurodevelopmental, psychosocial and physical morbidity factors that may be prevented or attenuated through intervention. In the meantime, incorporation of new clinical evaluation and management algorithms have been proposed for early identification of developmental impairment and prompt initiation of intervention [8]. In 2012, the American Heart Association published a management algorithm that stratified children with CHD on the basis of established risk factors. Given the potential for risk to change over time, routine surveillance and screening were recommended for all children with CHD at specified time intervals. For those deemed high-risk for developmental impairment, formal periodic developmental and medical evaluations were also endorsed [8]. Neurodevelopmental follow-up clinics have been established at numerous pediatric cardiac centers to provide the necessary expertise to identify and manage developmental impairment through multidisciplinary teams which may include a developmental pediatrician, psychologist, neurologist, special education or school interventionalist, speech and language pathologist, physical therapist and/or occupational therapist [8]. Early identification of genetic conditions is also important as children with both CHD and a genetic defect or syndrome are much more likely to have developmental deficits [8]. In the child or adolescent struggling with ADHD, optimal diagnosis and management can be achieved with multimodal interventions including pharmacotherapy, behavioral therapy and psychoeducational interventions [8].

3. Pediatric Psychosocial Outcomes

3.1. What Is Known About Pediatric Psychosocial Outcomes?

Children and adolescents with CHD are also at increased risk for impaired social cognition. The prevalence of "internalizing" problems (such as anxiety, depression, withdrawal, and somatization) and "externalizing" problems (attention, aggression) range from approximately 15% to 25% by parent report [10,16]. While studies on psychiatric disorders in children with CHD are limited, one study reports psychiatric illness in more than 20% of adolescents and young adults with CHD [17]. Another study comparing adolescents with TGA with a control group showed worse clinician-rated symptoms of depression, anxiety, and disruptive behaviors [18]. Worse global psychosocial functioning was associated with greater parental stress and lower cognitive function [18].

The cumulative effect of neurodevelopmental and psychosocial impairment significantly impacts overall QoL. For example, lower full-scale IQ and worse performance in academic achievement were associated with lower parent-reported psychosocial QoL scores at 8 years of age [19]. In a study investigating QoL in children following Fontan palliation, parent-reported psychosocial QoL scores were significantly lower [20]. Impaired executive functioning, gross motor ability, and the presence of internalizing disorders such as anxiety or depression have been associated with significantly lower QoL scores after controlling for patient demographics and other important clinical covariates [14]. Executive functioning, gross motor ability, and internalizing disorders have been shown to account for 42–50% of the variance in both patient and parent proxy-reported Psychosocial Impact QoL scores. In addition, executive dysfunction accounted for 37–54% of the variation noted in a patient and parent-reported school functioning QoL score [14].

3.2. Opportunities for Intervention

The best approach to psychosocial adjustment, behavioral problems, and social issues is enhanced prevention through early childhood surveillance, screening, detection, counseling, and management strategies that target normalization, social skills development, and healthy self-perception. Behavior should be monitored at every medical visit from infancy through adolescence through routine measurement of health-related QoL, psychosocial and behavioral functioning. Behavior screening tests that are most useful in the pediatric settings are parent-completed questionnaires as outlined in the 2010 American Academy of Pediatrics Task Force on Mental Health ("Enhancing Pediatric Mental Health Care") [21]. For adolescents, patient input combined with parent and teacher questionnaires provides the most comprehensive evaluation of psychosocial and mental health [8].

3.3. Transition

The neurodevelopmental and psychosocial issues noted in adolescence in CHD survivors typically persist as patients transition from pediatric to adult cardiovascular care. To further complicate this issue, adolescents with CHD must not only navigate the normative transitions of adolescence, but also develop an appropriate sense of identity in the context of chronic illness [22,23]. In fact, these issues likely have a tremendous impact on the life success of adults with CHD. There are no data on the impact of pediatric neurodevelopmental and psychosocial issues on the QoL of adults with CHD. However, there are data on the impact of adolescent neurodevelopmental issues on adult QoL in the Attention Deficit Hyperactivity Disorder (ADHD) population. In two recent cohort studies on adolescents with ADHD with more than 20 years follow-up, adolescents with ADHD were noted to have worse impaired physical and mental health, greater external stress with impaired work performance and higher financial stress, an increased risk of mortality from suicide, and a greater risk of psychiatric disorders including substance dependence, anxiety, and depression [24,25]. These data are particularly troubling since CHD survivors often have ADHD in addition to other complex significant neurodevelopmental and psychosocial issues.

A 2011 scientific statement from the American Heart Association outlines best practices in managing transition for adolescents with CHD [23]. This document emphasizes that neurodevelopmental and psychosocial factors may significantly impact adolescents' abilities to gain knowledge and develop health care self-management strategies. The adolescent neurodevelopmental and psychosocial phenotype has significant implications on care transition and the life success of the adult with CHD [26]. Adults with moderate to severe complexity CHD have an increased risk of anxiety, depression, pragmatic language impairment and social cognition issues, worse educational attainment, underemployment, and delayed progression into independent adulthood [8,27–30].