Optimizing the Developmental Follow-Up of Children and Adolescents Born with a Congenital Heart Defect

by

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“All things are difficult before they are easy.”

- Thomas Fuller

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ABSTRACT

Background and Objective: Children and adolescents with congenital heart defects (CHD) are at high risk for developmental delays. However, current developmental follow-up practices in Canada may not successfully identify their challenges in a timely manner. The objective of this project was to assemble comprehensive data sources needed to support the development of recommendations to improve the early identification of developmental challenges in children and adolescents with CHD.

Methods: This project was composed of three studies: 1) a systematic review to examine the prevalence and extent of motor difficulties in children (0-18 years) with CHD, 2) a mixed-methods study conducted with health professionals involved with the developmental follow-up of children with CHD in pediatric tertiary care centers to describe current Canadian developmental follow-up practices and to explore barriers to optimal follow-up, and 3) a qualitative study using interpretive description to explore the perspectives of youths with CHD and parents of children with CHD with respect to developmental follow-up.

Results: Although several systematic reviews have been published focusing on multiple developmental domains in children with CHD, a comprehensive review of motor outcomes across all age groups was lacking to fully appreciate the outcomes of interest for follow-up. The results of the first study (manuscript 1) showed that the overall prevalence of motor impairments (i.e., more than one standard deviation (SD) below the normative mean) ranged from 12.3% to 68.6% across childhood. While our results suggest that the overall prevalence of motor impairments remains stable across childhood and adolescence, severe motor impairments (below SD) appear to be more prevalent in younger children. In the second study (manuscript 2), we reported that four of the eight tertiary care centers providing open-heart surgery in Canada had a systematic developmental follow-up program that included screening and formal evaluation. These programs were only accessible to a subset of children with CHD identified to be at higher risk and focused on younger children. Participating clinicians described current practices as suboptimal and would like to develop a more systematic developmental follow-up program or expand their existing program. Participants emphasized the lack of human resources, financial support and

limited dedicated time as important barriers. In the third study (manuscript 3), the parents of children with CHD expressed that the limitations of current developmental follow-up practices in Canada put an undue burden on the families as they needed to assume new roles such as case managers and/or advocates to address their child's developmental limitations. This additional burden resulted in a high level of parental stress, which, in turn, affected the entire family system. The participants highlighted that easier access to both assessments and intervention services within the healthcare and school systems, at key transition points, as well as better communication between professionals would alleviate some of the parental burden. In addition, youth with CHD (manuscript 4) reported that with greater social and academic demands as well as an increased level of autonomy associated with older age, some faced new challenges that they had not encountered as children. Youth with CHD identified four aspects of the continuum of care that required attention to better respond to their needs. First, the format of the developmental follow-up should be adapted to their unique challenges. Second, resources could be more easily accessible throughout childhood and adolescence. Third, planning for transition to adult healthcare is essential to ensure continuity of services. Finally, they identified the school system as an essential component of the continuum of care that should have appropriate resource support.

Conclusion: Early identification of challenges for children and adolescents with CHD is not optimal across developmental domains at key transition points, from the perspectives of service users and service providers. We identified several possible approaches that could be considered to enhance earlier identification and timely interventions to address the needs of children and adolescents with CHD and their families. It is now essential to develop policy recommendations to optimize the developmental follow-up practices in Canada for children with CHD. This process should involve all relevant stakeholders and include a consultation process in the form of a nominal group process, consensus development panel and/or Delphi procedure.

ABRÉGÉ

Contexte et Objectif: Les enfants et les adolescents nés avec une cardiopathie congénitale (CC) présentent un risque élevé de retards développementaux. Cependant, il demeure incertain si les pratiques actuelles de suivi développemental au Canada identifient adéquatement et rapidement ces problématiques. L'objectif de ce projet était d'assembler les données nécessaires à l'élaboration de recommandations visant à améliorer l'identification précoce des problèmes développementaux chez les enfants et les adolescents atteints de CC au Canada.

Méthodes: Ce projet était composé de trois études : 1) une revue systématique ayant pour but d'examiner la prévalence et l'étendue des difficultés motrices chez les enfants (0-18 ans) nés avec une CC, 2) une étude à méthodes mixtes effectuée auprès de professionnels de la santé impliqués dans le suivi développemental des enfants nés avec une CC dans les centres de soins tertiaires pédiatriques et ayant pour but de décrire les pratiques canadiennes actuelles de suivi développemental et d'explorer les obstacles à un suivi optimal, et 3) une étude qualitative utilisant la description interprétative afin d'explorer le point de vue des jeunes nés avec une CC et des parents d'enfants vivant avec une CC en ce qui concerne le suivi développemental.

Résultats: Bien que plusieurs revues systématiques portant sur de multiples domaines développementaux chez les enfants nés avec une CC aient été publiées, aucune revue des aptitudes motrices n'avait été effectuée par groupes d'âge. Les résultats de notre revue systématique (manuscrit 1) ont démontré que la prévalence de déficiences motrices chez les enfants nés avec une CC (c'est-à-dire à plus d'un écart-type (ET) sous la moyenne normative) variait de 12,3 % à 68,6 % au cours de l'enfance et de l'adolescence. Bien que nos résultats suggèrent que la prévalence des déficiences motrices (< -1 ET) reste stable au cours de l'enfance et de l'adolescence, les déficiences motrices sévères (< -2 ET) semblent être plus fréquentes durant la petite enfance. Dans la deuxième étude (manuscrit 2), nous avons identifié que quatre des huit centres de soins tertiaires offrant des chirurgies pédiatriques à cœur ouvert au Canada avaient un programme de suivi systématique du développement comprenant un dépistage et une évaluation formelle. Ces programmes n'étaient, toutefois, accessibles qu'à un sous-groupe d'enfants avec une CC identifiés comme présentant un risque très élevé de délais

développementaux et étaient axés sur les très jeunes enfants. Les cliniciens participants ont décrit les pratiques actuelles comme étant sous-optimales et ont exprimé vouloir développer un programme de suivi développemental plus systématique ou étendre les programmes existants. Les participants ont souligné le manque de ressources humaines, de soutien financier et de temps comme étant des obstacles importants. Dans la troisième étude (manuscrit 3), les parents d'enfants atteints d'une CC ont indiqué que les lacunes dans les pratiques actuelles de suivi développemental au Canada imposaient un fardeau indu aux parents, car ceux-ci devaient assumer de nouveaux rôles tels que gérer les multiples rendez-vous ou revendiquer les services nécessaires au développement de leur enfant. Ce fardeau supplémentaire a entraîné un niveau de stress parental élevé, qui, à son tour, a affecté l'ensemble du système familial. Les participants ont souligné qu'un accès plus facile aux services d'évaluation et d'interventions dans les systèmes de santé et scolaire à certains moments clés du développement, ainsi qu'une meilleure communication entre les professionnels, allégeraient, en partie, cette charge parentale. De plus, les jeunes atteints d'une CC (manuscrit 4) ont signalé qu'avec des exigences sociales et scolaires plus élevées ainsi qu'un niveau d'autonomie accrue à l'adolescence, certains faisaient face à de nouveaux défis auxquels ils n'avaient pas été confrontés pendant leur enfance. Les jeunes atteints d'une CC ont identifié quatre aspects du continuum de soins qui nécessitent des changements afin de mieux répondre à leurs besoins. Premièrement, le format du suivi développemental doit être mieux adapté à leurs besoins et défis. Deuxièmement, les ressources devraient être plus facilement accessibles tout au long de l'enfance et de l'adolescence. Troisièmement, planifier la transition vers le système de santé des adultes est essentiel afin d'assurer la continuité des services. Enfin, ils ont identifié que le système scolaire est une composante essentielle du continuum de soins et devrait disposer de ressources de soutien appropriées.

Conclusion: Que ce soit du point de vue des utilisateurs ou des prestataires de services, l'identification précoce des problématiques développementales chez les enfants et les adolescents nés avec une CC n'est pas optimale pour tous les domaines de développement et lors de transition clés. Nous avons identifié plusieurs approches pouvant envisagées pour améliorer l'identification des problématiques développementales et les interventions précoces

afin de mieux répondre aux besoins des enfants et adolescents nés avec une CC et à ceux de leur famille. Il est maintenant essentiel d'élaborer des recommandations pour optimiser les pratiques de suivi développemental au Canada pour les enfants atteints d’une CC. Ce processus devrait impliquer toutes les parties prenantes et inclure un processus de consultation sous la forme d'un processus de groupe nominal, d'un panel d'élaboration de consensus et/ou d'une procédure Delphi.

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This achievement would not have been possible without the support from all of you.

PREFACE

CONTRIBUTION TO ORIGINAL KNOWLEDGE

The studies presented in this thesis present original scholarship. Novel contributions to the field of congenital heart defects in children are:

(i) Synthesizing the published evidence on the prevalence and extent of motor difficulties across childhood and adolescence in individuals with CHD.

(ii) Determining current practices for the developmental follow-up of children with CHD in all Canadian pediatric health centres that conduct open-heart surgery.

(iii) Identifying perceived barriers to the implementation of optimal developmental follow-up in Canada, from the perspective of health professionals.

(iv) Identifying limitations in current developmental follow-up practices from the point of view of youth with CHD and parents of children with CHD.

(v) Describing optimal developmental practices from the perspective of youth with CHD, parents of children with CHD, and healthcare professionals involved in the developmental follow-up of children with CHD.

CONTRIBUTION OF AUTHORS

All four manuscripts included in this thesis are the work of Marie-Eve Bolduc with the guidance of her supervisors Dr. Marie Brossard-Racine and Dr. Annette Majnemer. The doctoral candidate designed the studies, developed the consent forms, questionnaire, and interview guide, coordinated ethics approval, performed the interviews and analyses, drafted the manuscripts, and reviewed the final version of the manuscript.

Dr. Marie-Brossard-Racine and Dr. Annette Majnemer conceptualized and designed the studies along with the PhD candidate. They contributed to the development of the questionnaires and interview guides and participated in the analysis. They provided ongoing support, offering feedback on the manuscripts and throughout the implementation of the studies and thesis writing process.

Dr. Janet Rennick and Dr. Isabelle Gagnon contributed to the conceptualization of the studies, critically reviewed the study protocol, and carefully reviewed and provided feedback on the manuscripts.

For manuscript 1, Eliane Dionne acted as a second reviewer for the selection of the studies, extraction of the data, and quality appraisal of the selected studies for the first manuscript.

For manuscripts 3 and 4, Eva Sokol, a parent partner, contributed to the study design, critically reviewed the study protocol, contributed to the development of the questionnaire and interview guide, and reviewed manuscripts 3 and 4.

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CHAPTER 1: CONGENITAL HEART DEFECTS: A DISEASE OF THE HEART AND BRAIN

Congenital Heart Defects (CHDs), also known as congenital heart disease, can be defined as defects affecting the structure of the heart and great vessels (Casey, 2016; Nakamura, 2009). They are Canada's leading birth defects, with an incidence of 1.2% of total births per year (Go et al., 2013). Twenty-four to 50 percent of these lesions are complex and require open-heart surgery during infancy (Go et al., 2013; Hoffman & Kaplan, 2002). As a result of recent advances in medical and surgical management, survival rates of infants with CHDs have now reached 97% (Mandalenakis et al., 2020). However, as these cohorts of children with complex CHDs are getting older, there is now clear evidence that they constitute a unique population at high risk for developmental delays (Sherlock et al., 2009; Soul et al., 2009; Stegeman et al., 2022). Hence, with the increase in survival rates, the focus of the clinical and scientific communities has now shifted to improving developmental outcomes.

1.1 THE IMPACT OF CHDS ON BRAIN DEVELOPMENT

The mechanisms through which CHDs affect child and brain development are multifactorial, sequential, and cumulative. They result from a complex interplay between personal and environmental factors, beginning in the fetal period and extending throughout childhood (Marelli, 2020; McQuillen et al., 2012; McQuillen & Miller, 2010). Early research focused primarily on the perioperative factors associated with brain injuries. However, there is now a growing body of evidence that CHDs also have a deleterious effect on brain development in the fetal period (Clouchoux et al., 2013; Ortinau et al., 2013; Schellen et al., 2015). In this section, we will explore the mechanisms through which CHDs can affect brain integrity and development.

Fetal circulation differs from adult circulation, mainly due to the presence of two fetal shunts, the ductus arteriosus, located between the pulmonary artery and the aorta, and the foramen ovale, connecting the two atria (Claessens et al., 2017; Rudolph, 2007; Snarr et al., 2017). These shunts, coupled with increased fetal lung resistance, allow the blood to bypass the lungs (McQuillen et al., 2010; Murphy, 2005). More specifically, in fetal circulation, the oxygen-rich blood comes from the placenta and is directed to the left atrium where it mixes with a limited amount of pulmonary venous blood (McQuillen et al., 2010; Murphy, 2005; Rudolph, 2007). The

blood is subsequently sent to the left ventricle where oxygen saturation is approximately 65% (Rudolph, 2007). From the left ventricle, there is preferential streaming of the oxygen and nutrient-rich blood toward the brain and upper extremities through the aorta (Bhattacharya & Stubblefield, 2016; Donofrio et al., 2011; Wernovsky & Licht, 2016). In fetuses with transposition of the great arteries (TGA), the pulmonary artery is connected to the left ventricle and the aorta to the right ventricle (McQuillen et al., 2010; Murphy, 2005). Therefore, the brain receives blood with lower oxygen saturation and blood with high oxygen levels is directed toward the lungs and lower body (Barkhuizen et al., 2021; Claessens et al., 2017; Peyvandi et al., 2019). In hypoplastic left heart syndrome (HLHS), the increased pressure in the left atrium causes a reversal of the blood flow in the foramen ovale, resulting in a highly reduced ventricular output, with very limited or nonexistent flow to the ascending aorta (Clouchoux et al., 2013; Limperopoulos et al., 2010; McQuillen et al., 2010; Peyvandi et al., 2019). Therefore, cerebral blood flow occurs in a retrograde manner and is dependent on the ductus arteriosus (McQuillen et al., 2010; Peyvandi et al., 2019). Furthermore, in the right ventricle, the oxygenated blood mixes with the blood originating from the venous returns, lowering the saturation level to approximately 60% (McQuillen et al., 2010). Other types of CHDs may cause mixing of the blood with high oxygen saturation levels and blood with low oxygen saturation via a septal defect (Claessens et al., 2017). Hence, in fetuses with CHDs, decreased oxygen saturation and/or brain perfusion during brain development may cause significant delays in cortical maturation, decreased brain volumes and abnormal microstructural development (Brossard-Racine et al., 2014; Clouchoux et al., 2013; Kelly et al., 2019; Lim et al., 2016; Limperopoulos et al., 2010; Masoller et al., 2016; Schellen et al., 2015; Sun et al., 2015).

In addition to the developmental abnormalities described above, acquired brain injuries have also been observed on the brain magnetic resonance imaging (MRI) of individuals with CHDs. Although it was once thought that acquired brain injuries happened mainly during the intraoperative period, these abnormalities, most frequently white matter focal lesions in the periventricular region, have also been detected prior to surgery. The presence of brain immaturity, as described above, has, in fact, been associated with a higher risk of preoperative white matter injury (Andropoulos et al., 2010; Dimitropoulos et al., 2013; Goff et al., 2014;

McQuillen et al., 2010; McQuillen et al., 2003). In preterm infants, it has been demonstrated that the susceptibility of the immature brain to injury is associated with the increased vulnerability of oligodendrocyte progenitor cells and subplate neurons (McQuillen & Miller, 2010; Volpe, 2014). In the immature brain, the population of immature oligodendrocytes is comparatively much larger than in the more mature brain (Back et al., 2001). This creates a period of increased susceptibility to injury since these immature neuroglia are more vulnerable to hypoxia-ischemia than their mature counterparts (Back et al., 2001). The high prevalence of periventricular leukomalacia in individuals with CHDs, indicating impaired myelination process, suggests the involvement of oligodendrocytes (Goff et al., 2014). Furthermore, the vulnerability of subplate neurons reported in the fetal period, may continue to play an important role in the high incidence of brain injuries throughout neonatal life (McQuillen et al., 2003). Several risk factors for intraoperative brain injuries have also been investigated such as the type of bypass and blood pH but the results are inconclusive (Peyvandi et al., 2019). Nevertheless, in the early postoperative period, a predictable drop in cardiac output, referred to as low cardiac output syndrome has been documented in 10-25% of infants (Ballweg et al., 2007; Chandler & Kirsch, 2016; Hoffman et al., 2003). The hypotension and hypoxemia associated with this syndrome have been associated with an increased prevalence of acquired brain injury (Peyvandi et al., 2019). Overall, acquired abnormalities, including white matter injuries and strokes, have been frequently reported in infants with a complex CHD, with an overall prevalence ranging between 71 and 83% post-surgery (Dent et al., 2006; Mahle et al., 2002). Furthermore, a recent meta-analysis showed that adolescents with CHDs are 15.6 times more likely to present with brain abnormalities, including structural and microstructural abnormalities when compared to typically developing peers (Bolduc et al., 2018). In summary, children with CHDs are at high risk for brain immaturity and injury, which may occur as a result of multiple exposures to hypoxic and/or ischemic insults and are long-lasting. These sustained abnormalities in brain development are believed to be at the origin of a range of developmental delays and deficits manifesting throughout childhood, as described below.

Brain development results from a complex interplay between biological and environmental factors (Walker et al., 2011). Beyond the medical factors, early life experiences will also have an

impact on the developing brain (Belsky & de Haan, 2011). Children who are hospitalized in infancy are exposed to painful procedures, loud sounds, continuous lighting and limited social interactions that can have long-lasting effects on their development (Bisogni et al., 2015; Santos et al., 2015). Furthermore, early in the child's life, the parents of children with CHDs are confronted with the stress of the diagnosis and prognosis, intermittent separation from their child as well as witnessing medical procedures (Wernovsky & Licht, 2016). Once discharged, children may experience feeding difficulties, or require frequent rehospitalization, which can further increase parental stress (Golfenshtein et al., 2015). A recent literature review examined the psychological health of parents of children with CHDs and found that most studies report an increased level of stress, anxiety or depression when compared to parents of healthy children (Wei et al., 2015). In fact, one study reported that parents of children with CHDs were at risk of persisting posttraumatic stress disorder (PTSD), where 15% of mothers and 10% of fathers met the criteria for PTSD and approximately another 20% had symptoms of partial PTSD (Helfricht et al., 2008). These early experiences can have a negative impact on the child-parent relationship (Goldberg et al., 1991b; Ljubica et al., 2013), which can contribute to the neurodevelopmental deficits observed in children and adolescents with CHDs (Bellinger et al., 2015; Bellinger et al., 2011; Latal et al., 2016; von Rhein et al., 2014). Having a child with a CHD can also bring about a change in the parenting style (Wernovsky & Licht, 2016). In fact, studies have shown that parents of children with severe illnesses may be overprotective and set lower expectations for their child (Brosig et al., 2007; Carey et al., 2002). Since decreased early learning opportunities can translate into impaired dendritic arborization, dendritic spine density and decreased number of synapses leading to decreased cortical thickness (Belsky & de Haan, 2011; Ljubica et al., 2013; Stiles & Jernigan, 2010; Walker et al., 2011), it can be hypothesized that a more protective parenting style could be negatively associated with brain development. In a review of current literature, Belsky and de Hann (2011) highlighted that parenting is associated with brain development. However, further studies are needed to examine the extent to which this association is present in children with CHDs.

1.2 Developmental Outcomes and Everyday Functioning

Recent studies revealed an association between the presence of brain abnormalities and subsequent developmental impairments in children and adolescents with CHDs (Bolduc et al., 2018; Hiraiwa et al., 2020; Meuwly et al., 2019). This could explain, in part, the high prevalence of developmental impairments in individuals with CHDs. Indeed, studies have shown that 25%-50% of children with complex CHDs without genetic abnormalities, such as transposition of the great arteries and single functional ventricle, exhibit such difficulties (Bellinger et al., 2011; Gaynor et al., 2015; Majnemer et al., 2009; Marino et al., 2012; McCusker, 2016; Mussatto et al., 2014). These impairments may affect one or more developmental domains and their severity ranges from mild to moderate for most children with important impacts on daily life functioning in the home, school and community (Liamlahi & Latal, 2019). While some delays may be identified early in the child's life, other difficulties such as attention span or executive functioning may only be apparent later in childhood or adolescence (Gaudet et al., 2021; Ilardi et al., 2020). Furthermore, the nature of deficits in a particular developmental domain (e.g., motor) may become more complex (e.g., coordination, balance, motor planning) with greater impacts on daily living skills, academic performance and inclusion in community-based activities. In the following sections, we will synthesize the key findings of published evidence on developmental outcomes in children and adolescents with CHDs by domain.

1.2.1 Cognition

Although mean intelligence quotient (IQ) scores remained generally within normal limits, the literature has reported lower cognitive abilities in children and adolescents with complex CHDs when compared to normative data or typically developing controls (Feldmann et al., 2021; Griffin et al., 2003; Huisenga et al., 2021; Karsdorp et al., 2007; Miatton et al., 2006; Siciliano et al., 2019). In terms of executive function, a construct that has received greater attention over the past ten years, a recent meta-analysis suggested a medium effect size (0.5) for executive functioning scores of adolescents and young adults with CHDs when compared to those of typically developing children, which corresponds to a score that is approximately 0.5 standard deviations below those of typically developing children (Mills et al., 2018). The literature also suggests a greater risk for impairment in attention and memory skills (components of executive

functioning) (Mills et al., 2018; Sterken et al., 2015). Finally, several studies have also demonstrated the presence of visuomotor and visual-spatial difficulties in children with CHDs (Kirshbom et al., 2005; Miatton et al., 2007; Wotherspoon et al., 2020).

1.2.2 Psycho-Emotional

The presence of behavioural difficulties in children and adolescents with more complex forms of CHDs was demonstrated in four systematic reviews (Abda et al., 2018; Clancy et al., 2020; Huisenga et al., 2021; Karsdorp et al., 2007). Overall, behavioural problems were present in 25% of children and adolescents with CHDs, with internalizing behaviours (e.g., anxious, withdrawn, depressed behaviour or somatic complaints) being more frequent than externalizing behaviours (e.g., rule-breaking, aggressive-behaviour) (Abda et al., 2018). Behavioural difficulties were severe enough to significantly impact activities and/or relationships in 57.8% of children with complex CHDs. There is also evidence that children and adolescents with CHD have a decreased social cognition, an essential skill for social interactions (Abda et al., 2018). In addition, social skill impairments were observed in infants and children with CHDs (Clancy et al., 2020). Finally, the heterogeneity of the results with regard to emotional functioning limits our ability to draw clear conclusions for these domains and raises the possibility that important moderators exist for this construct (Jackson et al., 2015).

1.2.3. Language

Reviews suggest the presence of language impairments in children with complex CHDs (Huisenga et al., 2021; Miatton et al., 2006). Phonetic awareness, speech production and comprehension have been found to be impaired in this population (Bellinger et al., 2003). A more recent longitudinal study by Fourdain et al. (2019) found that both expressive and receptive language skills were affected at 12 months when compared to normative data. However, a significant difference was no longer evident for receptive language at 24 months (Fourdain et al., 2019).

1.2.4. Motor

Until recently, the presence of motor impairments, especially in older children and adolescents with CHDs, had not received as much attention as the psychosocial and cognitive domains. In a systematic review of randomized control trials and prospective observational cohort studies

published in 2010, Snookes and colleagues (Snookes et al., 2010) examined the impact of CHDs following cardiac surgery performed during the first six months of life, on motor and cognitive outcomes. The average motor score in the reviewed studies was approximately two standard deviations below the normative mean for children aged one to three years. However, in this review, the extent of motor limitations in the older age groups could not be ascertained due to the limited number of included studies that examined preschool and school-aged children. Other reviews yielded similar results (Khalil et al., 2014; Miatton et al., 2006; Mills et al., 2018), but they grouped infants, children and adolescents together, making it difficult to ascertain whether the motor difficulties persist beyond early childhood. A recent review by Huisenga and colleagues (2021) synthesized developmental outcomes across age groups. However, since they reported data for various domains, detailed information on motor skills for children beyond two years of age and data for adolescents were not presented. To address this gap, we conducted a systematic review that examined the prevalence and extent of motor impairments for children across age groups from infancy to adolescence (chapter 4) (Bolduc et al., 2020).

1.2.5 Participation in Everyday Life Roles

The relationship between developmental deficits (e.g., poor executive function, attention difficulties, decreased motor coordination) and participation in daily activities and life roles (e.g., daily living skills, socialization, school performance, leisure) is mediated by social, environmental and personal factors that can either enhance or hinder participation for children with disabilities (Law et al., 2006). As a result, it is essential to independently examine children and adolescents' performance in daily activities and life roles, to appreciate the impacts of developmental delays. A recent study examining leisure participation in adolescents with CHDs found that they mostly participated in social and recreational leisure activities but did not participate as much in active physical activities or skill-based leisure activities (Majnemer et al., 2020). Decreased participation in sports in children and adolescents with CHDs has also been reported by others (Arvidsson et al., 2009; Binkhorst et al., 2008). On the other hand, two recent reviews examined physical activity in children with CHDs (Acosta-Dighero et al., 2020; Skovdahl et al., 2021). Although evidence suggests lower levels of physical activity for children with CHDs when compared to controls, the variation in methodologies used across studies limits our ability to draw clear

conclusions. Academic achievement and educational attainment in children with CHDs were summarized in five reviews, which together suggest decreased performance when compared to peers (Cocomello et al., 2021; Glinianaia et al., 2021; Griffin et al., 2003; Miatton et al., 2006; Mills et al., 2018). Decreased ability to perform activities of daily living has also been reported (Easson et al., 2019; Granberg et al., 2008; Limperopoulos et al., 2001). Limperopoulos and colleagues (2001) reported that functional difficulties were present in 37% of young children with CHDs in the self-care, mobility and cognition domains while Granberg et al. identified decreased overall performance in activities of daily living (Granberg et al., 2008). Finally, challenges in daily living skills were found in 14.7% of adolescents with a complex CHD (Easson et al., 2019).

1.3 Early Identification of Developmental Challenges

1.3.1 Current Practices for the Identification of Developmental Impairments

Timely identification of developmental impairments is essential to limit their impact on participation in daily functions and roles as well as to facilitate access to intervention services and to provide caregiver support, education and coaching (Albaghli et al., 2019; Bailey et al., 2017; Blanche et al., 2016; Council on Children With Disabilities et al., 2006). The effectiveness of early interventions is well supported in other populations such as premature children who share many similarities with children with CHDs with regard to developmental outcomes (Cioni et al., 2016; Easson et al., 2019; Majnemer et al., 2017; Spittle et al., 2015; Wehrle et al., 2022). There is also preliminary evidence of their effectiveness on motor and language skills in children with CHDs (Fourdain et al., 2020). For older children, the use of compensatory strategies such as individualized education plans, psychological support and peer support can be used to optimize functioning (Majnemer, 1998).

Developmental follow-up can take the form of surveillance, screening and/or evaluation. Developmental surveillance includes documenting risk factors as well as parents' concerns over time (Council on Children With Disabilities et al., 2006). Developmental screening tools are short validated assessments, often completed by the parents, that can be used to further assess areas of concern raised during the developmental surveillance (Council on Children With Disabilities et al., 2006). Finally, developmental assessments are formal evaluations used by healthcare

professionals to identify specific developmental disorder(s) and are used to plan therapeutic interventions (Council on Children With Disabilities et al., 2006).

In 2012, the American Heart Association (AHA) published the first scientific statement focused on care practices to optimize the developmental outcomes of children with CHDs (Marino et al., 2012). The algorithm proposed in this statement is based on the presence of risk factors for developmental delays. These include open-heart surgery, cyanotic CHDs, comorbidities such as prematurity or genetic abnormality, mechanical support, heart transplant, cardiopulmonary resuscitation, postoperative hospitalization longer than two weeks, perioperative seizures, or brain abnormalities. In this statement, the recommendations included regular developmental surveillance of children with CHDs by the primary care provider (Marino et al., 2012). In addition, it was recommended that all high-risk infants be referred for formal developmental evaluations and early interventions and that monitoring should continue throughout childhood and adolescence, given that different problems may emerge at different ages. More specifically, it was recommended that these formal evaluations take place between 12 and 24 months, 3 to 5 years and 11-12 years, and assess the following domains: cognition, behaviour and psychosocial adjustment, as well as fine and gross motor skills. Screening for autism spectrum disorder was also recommended between 18 and 24 months. For children who do not present with any risk factors described above, surveillance is recommended, and screening should be performed if surveillance demonstrates a risk of developmental disability. The Cardiac Neurodevelopmental Outcomes Collaborative recently proposed to expand the follow-up strategies recommended by the AHA. The recommendations included new key time points for the re-evaluation of outcomes at 6 months, 18 months, 36 months, 5 years, 8-9 years, 10-11 years, 13-14 years and 18 years (Ilardi et al., 2020; Ware et al., 2020).

1.3.2 Optimal Developmental Follow-up Practices in the Canadian Context

The 2012 AHA statement has been instrumental in raising awareness of the need for closer monitoring of the development of children with CHDs in the clinical community. However, one study suggested that only 21% of pediatric primary care providers in the United States of America are aware of the recommendations included in this statement and that even when aware, formal developmental assessment was not always performed due to difficulty in accessing specialists

and/or resistance by families (Knutson et al., 2016). Different systemic and institutional barriers such as the lack of available specialists trained to perform standardized developmental assessments or the lack of time and resources may also interfere with the implementation of the proposed clinical recommendations (Fischer et al., 2016). However, no study has explored the challenges experienced by health professionals in implementing the developmental follow-up recommendations for children with CHDs in Canada. This information is crucial given that the AHA recommendations were developed for the American healthcare context, where healthcare plans are provided by private companies and may not be applicable to the Canadian healthcare context, which differs from both an accessibility and funding standpoint.

Based on the AGREE-II instrument (AGREE Collaboration, 2003), a tool designed to assess the quality of practice guidelines, the elaboration of quality guidelines for the developmental follow-up of Canadian children and adolescents with CHDs should include the collection of data from various sources. This comprises a systematic search of relevant evidence and consultations with all relevant stakeholders. In fact, recent literature widely recognizes the importance of user involvement (stakeholder engagement) in the development of clinical guidelines (Harding et al., 2011; Nilsen et al., 2006). Involving the user in the development of clinical guidelines can result in a greater ability to address their concerns, and to implement them with consideration of existing barriers, resulting in overall better care (Nilsen et al., 2006).

1.4 SUMMARY

CHD is Canada's most prevalent birth defect. Children and adolescents with CHDs are at high risk for brain injury and maldevelopment, and subsequent developmental impairments in various domains including cognition, psycho-emotional, language and motor domains. Evidence shows that these challenges can have a significant impact on their participation in daily functions and life roles if not addressed in a timely manner. Unfortunately, current developmental follow-up practices across Canada may not be effective in detecting these delays. Information on current practices and barriers as well as consultation with relevant stakeholders (users) are necessary to develop a comprehensive understanding of what optimal follow-up practices should be in the Canadian context.

CHAPTER 2: RATIONALE AND OBJECTIVES OF THE THESIS

2.1 RATIONALE

Congenital heart defects (CHDs) are Canada's most common birth defect and are associated with lifelong challenges in a substantial subset of individuals. By detecting these challenges in a timely manner and providing the families with the services they need, we can limit their impact on academic, social and daily functioning (Bailey et al., 2017; Blanche et al., 2016). Although practice recommendations for the developmental follow-up of children with CHDs requiring open-heart surgery were developed in 2012 in the United States of America (Marino et al., 2012), their uptake and applicability to the Canadian context have not been investigated. Unfortunately, there is no standardized set of developmental and domain-specific assessments currently recommended across Canada for children and adolescents with CHDs, in contrast with other similar high-risk populations (Albaghli et al., 2019). Hence, current developmental follow-up practices may not allow for the timely detection of developmental challenges, and important opportunities for interventions may be lost (Tierney & Nelson, 2009).

The development of practice recommendations for the developmental follow-up of children and adolescents with CHDs requiring open-heart surgery necessitates the collection of data from several sources, including a comprehensive synthesis of the literature of developmental outcomes (Brouwers et al., 2010). Although most domains had been well synthesized through systematic reviews and meta-analyses, the prevalence and extent of motor impairments across age groups were not well defined before this project. This information is essential to identify the type of assessments that need to be administered to each age group. In addition, a description of current barriers and facilitators is also a key component of guideline development (Brouwers et al., 2010). However, no data existed on current developmental surveillance practices for children with a complex CHD or barriers to implementation of optimal practices across Canada. This information is important so that gaps in current practices can be identified and context-appropriate recommendations can be developed. Finally, it is now well recognized that the development of relevant guidelines needs to include consultations with all stakeholders (AGREE Collaboration, 2003; Brouwers et al., 2010). In the case of developmental follow-up of children

with CHDs, this includes the youth themselves, parents of children with CHDs and healthcare professionals.

The implementation of standardized evidence-based practices for the developmental follow-up of Canadian children would ensure that delays are identified in a timely manner and that the resources can be put in place to support the development of each child with a CHD requiring open-heart surgery. Collecting the necessary information to develop these guidelines is the first step of this important process.

2.2 Objectives

2.2.1 Overarching Objective

The overarching objective of this thesis was to assemble comprehensive data sources needed to support the development of recommendations to improve the early identification of developmental challenges in children and adolescents with CHDs requiring open-heart surgery.

2.2.2 Specific Objectives

The specific objectives of this doctoral project were threefold:

1) To examine the prevalence and extent of motor difficulties in infants, children and adolescents with CHDs requiring open-heart surgery.
2) To describe current Canadian developmental follow-up practices and to explore barriers to optimal follow-up.
3) To explore the perspectives of parents of children and adolescents with CHDs and those of adolescents and young adults with CHDs with respect to the developmental follow-up.

2.3 Overall Study Design and Methods

This is a manuscript-based thesis constructed around four manuscripts. The texts are reproduced exactly as published or submitted. This project is composed of three studies presented in four distinct manuscripts.

Study I (manuscript 1) is a systematic review of motor outcomes in children with CHDs. This review complements existing reviews on other developmental domains in this population. It included original studies published in both French and English that reported motor outcomes in

children and/or adolescents (0-18 years) who were born with a CHD requiring open-heart surgery in the first two years of life.

Manuscript 1 : Bolduc, M.-E., Dionne, E., Gagnon, I., Rennick, J. E., Majnemer, A., & Brossard-Racine, M. (2020). Motor impairment in children with congenital heart defects: A systematic review. *Pediatrics*, *146*(6). https://doi.org/10.1542/peds.2020-0083

Study II (manuscript 2) is an exploratory sequential mixed-methods study of current practices in the developmental follow-up of infants, children and adolescents with CHDs requiring open-heart surgery in Canada. All centers performing pediatric open-heart surgery in Canada were approached to participate in this study. Participants were first sent a questionnaire that included descriptive information and current follow-up practices. Telephone surveys were then conducted to explore the feasibility and acceptability of current recommendations, and barriers to optimal developmental follow-up.

Manuscript 2: Bolduc, M.-E., Rennick, J. E., Gagnon, I., Majnemer, A., & Brossard-Racine, M. (2022). Canadian developmental follow-up practices in children with congenital heart defects: A national environmental scan. *CJC Pediatric and Congenital Heart Disease*, *1*(1), 3-10. https://doi.org/10.1016/j.cjcpc.2021.11.002

Study III (manuscripts 3 and 4) is a qualitative study using an interpretive description methodology. Through interviews with youth with CHDs (manuscript 3) and parents of children with CHDs (manuscript 4), we developed comprehensive and contextualized interpretations of their perspectives on developmental follow-up. Comparative analysis was used to find commonalities and differences in participant experiences and preferences. The final step of the analysis consisted of developing new conceptualizations of the phenomenon that will contribute to improving the identification of developmental challenges in children and adolescents with CHDs.

Manuscript 3 (parents): Bolduc, M.-E., Rennick, J. E., Gagnon, I., Majnemer, A., & Brossard-Racine, M. (manuscript submitted for publication). Navigating the healthcare system with

my child with CHD: A parental perspective on Canadian developmental follow-up practices.

Manuscript 4 (youth): Bolduc, M.-E., Rennick, J. E., Gagnon, I., Brossard-Racine, M., & Majnemer, A. (manuscript submitted for publication). Identifying developmental challenges of youth with congenital heart defects: A patient-oriented perspective.

CHAPTER 3: INTRODUCTION TO THE SYSTEMATIC REVIEW ON MOTOR OUTCOME

Although several studies had described the extent and nature of cognitive and behavioral sequelae in children and adolescents with complex CHDs, the literature on motor outcomes had not been systematically synthesized for each age group before this study. As a result, it remained difficult to clearly articulate the overall prevalence and extent of motor impairments across childhood and adolescence.

This evidence is necessary for determining optimal developmental follow-up practices that include the selection of appropriate assessment tools at each key time point and for all domains at risk of delays (Ilardi et al., 2020; Marino et al., 2012). This information is also essential to implement intervention services that address all affected developmental domains (Ware et al., 2020). If not identified and addressed, motor difficulties may negatively affect anxiety levels, self-esteem, and social-emotional well-being (Piek et al., 2006; Poole et al., 2018).

The manuscript that follows addresses our first specific objective to examine the prevalence and extent of motor difficulties in infants, children and adolescents with CHDs requiring open-heart surgery.

This manuscript has been published in *Pediatrics.*

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CHAPTER 4: MOTOR IMPAIRMENT IN CHILDREN WITH CONGENITAL HEART DEFECTS: A SYSTEMATIC REVIEW

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Abbreviations: CHD: Congenital Heart Defect; IQR: Interquartile range; SD: Standard Deviation

Table of content summary

This review highlights the presence of significant motor skill impairments in children with CHDs and suggests that these difficulties are present throughout childhood and adolescence.

CONTRIBUTOR'S STATEMENT PAGE

Ms. Bolduc conceptualized and designed the review, extracted the data, performed the quality appraisal, drafted the first version of the manuscript, reviewed and approved the final version of the manuscript.

Ms. Dionne extracted the data, performed the quality appraisal, contributed to drafting the manuscript and reviewed and approved the final version of the manuscript.

Dr. Majnemer designed the study approach, contributed to data analysis and drafting of the manuscript, critically reviewed and approved the final version of the manuscript.

Dr. Brossard-Racine conceptualized and designed of the study, contributed to data analysis and drafting of the manuscript, critically reviewed and approved the manuscript.

Dr. Gagnon contributed to the conceptualization of the study, critically reviewed the study protocol and draft of the manuscript, critically reviewed and approved the manuscript.

Dr. Rennick contributed to the conceptualization of the study, critically reviewed the study protocol and draft of the manuscript, critically reviewed and approved the manuscript.

All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

ABSTRACT

Context: With improvements in survival rates in newborns with congenital heart defects (CHD), focus has now shifted toward enhancing neurodevelopmental outcomes across their lifespan.

Objective: To systematically review the prevalence and extent of motor difficulties in infants, children and adolescents with CHD requiring open-heart surgery.

Data Sources: Embase, Medline and CINAHL.

Study Selection: Original studies published between 1997 and 2019 examining gross and/or fine motor skills in children (0-18 years) born with a CHD requiring open-heart surgery were selected by two independent reviewers.

Data Extraction: The prevalence of motor impairments and mean scores on standardized motor assessments were extracted. Findings were grouped in five categories based on the age of the children.

Results: Forty-six original studies were included in this systematic review. The prevalence of mild to severe motor impairments (scores < -1 SD below normative data or controls) across childhood ranged from 12.3 % to 68.6% and from 0% to 60.0% for severe motor impairments (< -2 SD). While our results suggest that the overall prevalence of motor impairments < -1 SD remains rather constant across childhood and adolescence, severe motor impairments (< -2 SD) appear to be more prevalent in younger children.

Limitations: Variability in sampling and methodology between the reviewed studies is the most important limitation of this review.

Conclusions: The results of this review highlight that infants with CHD have an increased risk of motor skill impairments across infancy, childhood and adolescence. These findings stress the importance of systematic standardized screening or evaluation of motor skills across childhood and adolescence in children with CHD requiring open-heart surgery.

Introduction

Congenital heart defects (CHDs) are the most common congenital anomalies with an incidence of 1.2% of total births[1]. As a result of improvements in neonatal surgery and perioperative management[2-4], mortality attributable to a cardiac defect has decreased significantly in the past decades[1, 5], and most infants born with a CHD are now expected to reach adulthood. Therefore, focus has now shifted toward better defining and improving neurodevelopmental outcomes of these individuals across their lifespan.

Children with CHD requiring open-heart surgery have an overall estimated prevalence of developmental impairments ranging from 25% to 50%[3]. Neurocognitive outcomes in children with CHD have been well documented and synthesized using systematic reviews and meta-analyses. These reviews reported a higher prevalence of lower intellectual abilities[6, 7], impairments in executive function[8], attention and memory difficulties[8], and behavioral difficulties[7, 9]. Conversely, the presence of motor impairments in children with CHD has not been comprehensively synthesized. A clear understanding of motor impairments across childhood is essential in order to develop more targeted and comprehensive guidelines for the developmental surveillance of these individuals. Therefore, the objective of this systematic review is to examine the prevalence and extent of motor difficulties in infants, children and adolescents with CHD.

Methods

Eligibility Criteria

Original studies published in English or French and examining motor outcomes in infants, children and/or adolescents (0-18 years) who were born with a CHD requiring open-heart surgery (with or without cardiopulmonary bypass) in the first two years of life were included in this review. Published randomized controlled trials, observational cohort or cross-sectional studies, and case-control studies that used standardized assessments to assess fine and/or gross motor skills were selected. Conference abstracts were not included due to our limited ability to assess the quality of the studies and risk of bias for this type of publication. Due to the fast-evolving surgical and medical care, only studies published since 1997 were included. Studies that included individuals born preterm (i.e. <35 weeks of gestation), with genetic anomalies or requiring heart transplant

were excluded unless the results of the subgroup without these confounders were presented separately. We excluded these specific conditions in an attempt to enhance uniformity of the participants, as these conditions have been independently associated with an increased risk of developmental delays. For similar reasons, reporting of motor assessments was limited to outcomes after open-heart surgery. In the case of longitudinal studies that presented the results of motor skills assessment at two timepoints within the same age category (e.g., twice during infancy), only the data for the larger sample were analyzed.

Search Strategy

A search of studies examining gross and fine motor skills in children, aged 0-18 years, born with a CHD was performed in Embase, Medline and CINAHL in September 2017 and updated in September 2019. Medical subject headings (MESH) were used and exploded where appropriate. Boolean operators were used to combine *congenital heart defect/disease* and *psychomotor performance/motor skills/motor performance/child development* (see Supplemental Table 1 for detailed search). References of the selected articles were screened to identify additional studies. A librarian assisted the authors in the development of the search strategy.

The study selection consisted of a two-step process. First, titles and abstracts were screened to determine if the studies met the inclusion criteria. Then, full texts of the selected articles were reviewed. Data extraction and study appraisal were independently performed by two reviewers (M.B. & E.D.). A data extraction form was developed based on the Institute of Medicine's Standards for Systematic Reviews[10], the University of York's Centre for Reviews and Dissemination's guidelines for reviews[11], and the STROBE statement checklist[12]. The Cochrane Risk of Bias Tool[13] was used for the quality appraisal of the selected studies. This tool examines the risk of selection bias, performance bias, detection bias, attrition bias, and reporting bias which are subsequently categorized as low risk, high risk or unclear risk based on the judgement of the reviewers. In case of disagreement, the decisions were discussed until a consensus was reached. Authors of the selected studies were contacted if clarification was needed to confirm that the article met the inclusion/exclusion criteria or if additional information was required to be included in our analyses. Some of these authors generously provided unpublished data or new calculations that excluded children who were born prematurely or had genetic syndromes.

Analysis

To better appreciate the difference in motor skills across a child's developmental trajectory, we clustered the findings in the following categories based on the mean age of the children at the time of assessment: 1) Infancy (<1 year of age) 2) Toddler years (1 to <3 years); 3) Preschool age (3 to <6 years) 3) School age (6 to <13 years) 4) Adolescence (13 to <19 years). Mild to severe motor skill impairment was defined as a motor score lower than one standard deviation (SD) from the normative mean or below the 15th percentile, while severe motor impairment was defined as a motor score lower than two SD from the normative mean or below the 5th percentile. In order to account for the range of severity in motor impairments and to be consistent with the definition of motor impairments reported across studies, we report the prevalence of all motor impairments as defined by -1 SD below the mean (i.e. mild to severe) and the presence severe motor impairment only (< -2 SD) separately using descriptive statistics.

In addition, effect size (Hedge's g statistic) for the difference in motor skills between children with CHD and typically developing children for each study that presented means and standard deviations was calculated independently for each study. The magnitude of the effect sizes were interpreted as follows: 0.2–0.5 small, 0.5–0.8 moderate, and > 0.8 large effect size based on Cohen's convention[14]. Range and interquartile range (IQR) were used to summarize prevalence and effect sizes. Finally, we synthesized the results of the studies that compared subgroups of children with different cardiac physiology and those that reported fine and gross motor skills separately.

Results

Study Selection

A total of 1192 studies were identified through the literature search. Of the 1002 studies remaining after the removal of duplicates, 805 were eliminated in the first stage of screening which involved screening titles and abstracts, and 156 were subsequently eliminated after reading the full text. Five additional studies were identified through a manual search of selected articles' reference lists, leaving 46 original studies to undergo data extraction and analysis in this review. The detailed study selection process is presented in Supplemental Figure 1.

Study Characteristics

Studies included in this review originated from five continents. Sixteen (35%) studies were conducted in the United States of America (USA)[15-30], seven (15%) in Canada[31-37], six (13%) in Germany[38-43], four (9%) in Australia[44-47], two (4%) in Netherlands[48, 49], two (4%) in Switzerland[50, 51], two (4%) in Finland[52, 53], two (4%) in Japan[54, 55], one (2%) in Brazil[56], one (2%) in India[57] and one (2%) in Norway[58]. In addition, two (4%) studies were conducted in centers located both in Canada and the USA[59, 60]. Twenty-eight (61%) studies used a cohort study design[15, 20, 23-25, 29-35, 37, 38, 41, 43-47, 49-52, 54, 56, 58, 60], nine (20%) a randomized control trial design[16-19, 22, 27, 28, 48, 59], six (13%) a cross-sectional design[21, 26, 36, 39, 53, 57], two (4%) a case-control design [42, 55], and one (2%) a case series design[40]. Studies included in this review examined samples composed of children with different types of CHD, including hypoplastic left heart syndrome or other types of single ventricle defects exclusively (eight studies, 17%)[20, 22, 23, 26, 32, 52, 53, 59], transposition of the great arteries (seven studies, 15%)[16, 18, 19, 33, 40, 41, 49], ventricular septal defect (two studies, 4%)[28, 55], total anomalous pulmonary venous connection (one study, 2%)[31], and aortic arch obstruction (one study, 2%)[48]. Twenty-seven studies (59%) included samples composed of children with mixed CHD diagnoses[15, 17, 21, 24, 25, 27, 29, 30, 34-39, 42-47, 50, 51, 54, 56-58, 60]. Study participants' ages ranged from a few days of age to late adolescence (i.e. 19 years).

Quality of the Studies

Overall, fourteen (30%) studies presented a low risk of bias on all assessed criteria, four (9%) studies presented a high risk of bias in one category and only one (2%) study presented a high risk of bias in two categories. With regards to possible performance bias, the assessors were blind to medical history, diagnosis, other assessment results, or treatment assignment in 17 of the 46 (37%) studies. In terms of possible attrition bias, two (4%) studies lost a large number of participants and did not consider attrition in their analysis, therefore presenting a high risk of bias for this criterion, while four (9%) studies did not report attrition numbers. Finally, baseline imbalance in factors related to outcomes such as age, sex, maternal education was present in four (9%) studies. Detailed information on the quality of the studies is presented in supplemental Table 2.

Motor Skills

Across ages

Prevalence: The prevalence of mild to severe motor impairments (< -1 SD) ranged from 12.3% to 68.6% (IQR: 23.4%-52.2%) in children and adolescents with CHD based on the results from thirteen studies that presented this outcome. Severe motor impairments (< -2 SD) were reported in 22 studies and ranged from 0.0% to 60.0% (IQR: 7.2%-29.8%) in children and adolescents with CHD (Figure 1).

Extent of impairment: The effect size for motor skill differences between children with CHD and typically developing children across ages ranged from -0.07 to 2.19 (IQR: 0.52 to 1.34) across 38 studies.

Infancy (Birth to <12-month-old)

Nine studies examined motor skills in infants younger than 12 months following open-heart surgery for CHD[15, 20, 21, 24, 25, 28, 46, 56, 57]. Study results and characteristics are presented in Table 1.

Prevalence: In this age group, only one study described the prevalence of motor impairments < -1 SD. This study reported a 20.0% prevalence of motor skill difficulties[21]. Additionally, Rocha et al.[56] reported suspected delays in 55.0% of infants with CHD based on the results of a screening test. The prevalence of severe motor impairments (< -2 SD) in infants with CHD ranged from 0.0% to 54.6% (IQR: 8.4%-50.9%)[20, 21, 24, 57]. The results are represented in Figure 2.

Extent of impairment: Among the eight studies that compared the mean motor scores of infants with CHD to normative data or controls[15, 20, 21, 24, 25, 28, 46, 57], six found that infants with CHD obtained significantly poorer scores than their comparison. The other two also found lower mean scores but the results were not statistically significant. The effect size ranged from 0.12 to 1.61 (IQR: 0.53 to 1.43).

Toddler Years (1-year-old to <3-year-old)

Thirty studies examined motor development in toddlers[15-17, 22, 24-27, 29-34, 37, 38, 42, 44, 46-49, 51-55, 57, 59, 60]. Study characteristics and results are presented in Table 2.

Prevalence: The prevalence of mild to severe motor impairments (< -1 SD) ranged from 12.3% to 59.0% in toddlers with CHD (IQR: 14.5%-58.6%)[30, 44, 49, 55, 59], while the prevalence of severe motor impairments (< -2 SD) ranged from 1.5% to 60.0% (IQR: 6.0%-26.1%)[16, 17, 24, 30-33, 44, 47, 49, 51, 57, 59]. One study that included toddlers with single ventricle pathologies identified a significantly higher prevalence of severe motor impairment (60%) using the Bayley Scales of Infant and Toddler Development II when compared to the other studies in this category[51]. Another study defined motor impairment as scores < -1.5 SD below the mean, and therefore could not be synthesized with the other studies, reported 42% of motor impairment in their sample[34]. The results are represented in Figure 2.

Extent of impairment: Twenty-four of the 28 studies (85.7%) that presented this outcome reported that at least one subgroup of children with CHD performed significantly worse on motor assessments than typically developing children. Overall, the effect size for motor skill differences between toddlers with CHD requiring open-heart surgery and typically developing children ranged from -0.07 to 2.19 (IQR= 0.47 to 1.49) [15-17, 22, 25-27, 29-34, 37, 38, 42, 44, 46-49, 52-55, 57, 59, 60].

Preschool Age (3-year-old to < 6-year-old)

Ten studies[19, 23, 35, 40, 42, 45, 48, 50, 52, 54] reported motor outcomes in preschool children. The characteristics and results of these studies are presented in Table 3.

Prevalence: Only two studies reported the prevalence of children with motor scores < -1 SD in this age group. The prevalence of preschool children with mild to severe motor impairments ranged from 32.4% to 68.6%[35, 40], while the prevalence of children with severe motor impairments (< -2 SD) ranged from 7.7%-to 28.6% [23, 35, 48]. The results are represented in Figure 2.

Extent of impairment: All studies examining motor scores in preschoolers found a significant difference when compared to typically developing children. Overall, preschool children with CHD were found to have a difference in motor scores when compared to normative data or controls that ranged between 0.18 and 1.38 standard deviation (IQR: 0.66 to 1.19) [23, 35, 40, 42, 45, 52, 54]. In addition, the results of two studies could not be synthesized above because they did not provide a prevalence nor mean score and SD. One of these studies reported that preschool age children perform worse on norm-referenced motor assessments, with an average score at the 9th

percentile for gross motor skills and 4th percentile in fine motor skills[19]. The second study reported significantly lower balance and aiming and catching skills in preschool children with CHD when compared to the assessment's normative data but did not find a significant difference on the assessment's total score nor on the manual dexterity index[50].

School-age (6-year-old to <13-year-old)

Motor performance in school-age children was presented in five studies[18, 39, 41, 43, 58]. Their characteristics and results are summarized in Table 4.

Prevalence: The prevalence of motor skills impairments (< -1 SD) in school-age children ranged from 26.7% to 46.1% (IQR: 30.6%-45.2%)[18, 39, 41, 58], and from 7.5% to 25.8% for severe motor impairments (< -2 SD) [39, 41, 58].

Extent of impairment: Mean scores in school age children with CHD were lower than those of typically developing children in all reviewed studies. The effect size ranged from 0.16 to 1.21 across studies, IQR: 0.24 to 1.14)[39, 41, 43, 58]. The results are represented in Figure 2.

Adolescence (13-year-old to < 19-year-old)

Only one study examined the extent and prevalence of motor impairment in adolescents with CHD (Table 4).

Prevalence: Easson et al.[36] found that 42.4% of adolescents with CHD presented with mild to severe motor impairments (scores < -1 SD) and that 18.2% had severe motor impairments (< -2 SD). The results are presented in Figure 2.

Extent of impairment: The average total score of adolescents in that study corresponds to the 5th to 15th percentile when compared to norms.

Comparison of Outcome between Children with Different Cardiac Physiology

Seven studies compared children with a single ventricle to those with two ventricle physiologies[15, 24, 25, 30, 37, 54, 60]. Five of these reported significant differences in at least one age group, with the single ventricle group performing worse[15, 24, 25, 54, 60]. The other two studies similarly found lower scores in infants and toddlers with single ventricle physiology, but the differences were not statistically significant[30, 37]. In addition, one study compared children with

hypoplastic left heart syndrome to other single ventricle physiology and found that they performed significantly worse[53]. Finally, two studies reported better motor outcome in toddlers with transposition of the great arteries when compared to children with other two ventricle defects.

Fine and Gross Motor Function

We compared the prevalence of fine and gross motor impairments (< -1 SD) in children with CHD. Three studies reported gross and fine motor scores separately[35, 36, 40]. The results of these three studies suggested no significant differences between the two motor skill domains. Overall, gross motor difficulties were present in 18.2% to 68.8% of participants with CHD, while fine motor impairments ranged from 23.2% to 55.8%.

DISCUSSION

This review highlights the high prevalence of motor impairments throughout childhood and adolescence in children with CHD requiring open-heart surgery. Indeed, the results of our systematic review have shown that approximately one third of children with CHD have delayed motor skills and that these impairments are present in comparable proportions throughout development. Conversely, we found that the prevalence of severe motor impairments (< -2 SD) seems to be higher in younger children, which is in line with preliminary longitudinal observational studies suggesting that motor skills may improve over time in children with CHD[52, 61]. This may be explained in part by the prolonged hospitalization that some infants may experience perioperatively and the recommended movement restrictions that follow surgery which together could delay the acquisition of age-appropriate motor skills. Moreover, the severity of the motor impairments may be decreased with development and after receiving rehabilitation intervention.

The extent of the differences in motor skills detected in the current review indicates that motor scores in children and adolescents with CHD requiring open-heart surgery were approximately one SD below those of typically developing children. In daily life, these children may seem clumsy, experience difficulty scribbling, cutting, doing buttons or zippers, jumping or avoid participation in motor-based activities[62, 63]. Although these motor impairments may seem subtle or “mild”,

they can have long-lasting impacts on the psychological and physical well-being of individuals. For instance, children and adolescents with developmental coordination disorder, a disorder affecting motor coordination, have lower self-esteem and higher anxiety when compared to age-matched controls[64]. In typically developing children, poorer motor coordination is associated with impaired emotion recognition and social behavior[65]. Furthermore, motor impairments are associated with reduced participation in physical activity, which can have an important impact on health[66, 67]. In a recent paper by Majnemer et al., poorer motor performance in adolescents with CHD has also been associated with lower participation in active-physical and social activities[68]. It is therefore expected that the presence of motor impairments during childhood will have long-lasting consequences on daily function, self-determination and well-being unless effective interventions to overcome these difficulties are provided. For infants and younger children, investments in the development and implementation of early intervention programs by children who are at high risk of developmental delays has been supported by various organizations and governmental agencies[69, 70]. They have proven to be effective in other high-risk populations[71] and there is preliminary evidence supporting their effectiveness in children with CHD[72]. For children and adolescents, other approaches such as the Cognitive Orientation to Occupational Performance (CO-OP) intervention as well as occupational and physical therapy have also been shown to be effective in children with developmental coordination disorder[73], who present a motor profile similar to that of children with CHD[36].

Early identification of motor impairments and timely referral to rehabilitation services is essential to optimize daily function and quality of life. However, the subtleness of the impairments identified in this review may pose a challenge to early identification. In 2012, the American Heart Association published guidelines for the developmental follow-up of children with CHD[3]. In these guidelines, it was recommended that all children who underwent open-heart surgery be referred for formal developmental evaluation and early intervention by the primary care provider or subspecialist. Currently in North America, many tertiary care centers have implemented developmental follow-up clinics for children with CHD. However, other centers rely on the assessment of the cardiologist, neonatologist or primary care providers during routine follow-up. The implementation of the guidelines has not been investigated outside the United States or in

American tertiary care settings, however, one study suggests that awareness of the recommendations is limited among American primary care providers[74]. Current clinical practices often rely on general observations of motor milestones and parental reports and may not be sensitive enough to promptly detect more subtle difficulties, especially in older children who have learned to cope and compensate for these difficulties. The results of this review demonstrate that standardized screening and evaluation of developmental delays, including motor delays, throughout childhood and adolescence are essential. Timely referral to rehabilitation services is required when a delay is identified in order to provide these children with the best opportunities to optimize their motor skills, particularly as this review suggests that these difficulties will not be outgrown.

This review also suggests that while all children with CHD who underwent open-heart surgery have a higher prevalence of motor impairments, their severity may differ based on the complexity of the defect. Infants and toddlers with single ventricle physiology were found to have significantly poorer motor abilities than children with two ventricles. These findings are in line with a number of studies that have suggested that specific diagnoses may be associated with severity of motor impairments[17, 39, 52, 53]. The poorer developmental outcomes in children with single ventricle physiology may be attributed to various risk factors including reduced substrate delivery starting in the fetal period[75-77], the need for multiple surgeries[38, 44, 59, 78] and the higher prevalence of brain abnormalities[48, 52, 79]. Although acquired brain injury and brain developmental malformation are frequent in neonates with CHD[80], they, along with the other clinical factors found to be associated with motor skills, have not been consistently reported or evaluated in the reviewed studies. More studies examining risk factors associated with motor delays are needed to identify possible avenues to optimize outcomes. However, considering that all children with CHD who underwent open-heart surgery are at high risk of motor impairments across childhood and adolescence, each would benefit from close follow-up with standardized screening or evaluation of motor skills notwithstanding the presence of additional risk factors.

LIMITATIONS

This review must be interpreted in the context of its limitations. Variability in sampling and methodology between reviewed studies is the most important limitation in this review. The reviewed studies included children with different cardiac physiology, who may have different motor outcome. In terms of methodology, the different motor evaluation tools used may have resulted in heterogeneous results across the different studies since assessments may vary in sensitivity or types of motor skills assessed. Furthermore, different versions of the same test could yield different results. A recent study concluded that the third edition of the Bayley Scales of Infant and Toddler Development may underestimate motor impairments[81]. In this review, 17 studies used the second edition and four used the third edition of that test. Finally, several studies have identified a limited ability for the Alberta Infant Motor Scales to discriminate between infants above 15 months of age[46, 82, 83]. This ceiling effect could have confounded results of three of the reviewed studies examining motor skills in toddlers and may have contributed to the variability in that age group. Nonetheless, these different tools are valid measures of motor performance, and identify children with impairment at different ages.

In addition, although we included only studies that excluded children with genetic findings or syndromes, not all children were systematically tested and, therefore, it is possible that some studies included a larger proportion of children with genetic variants, a subgroup which may have poorer motor outcome[84, 85]. In addition, our review excluded studies that included children who had undergone heart transplant and who may present with more severe developmental difficulties. Therefore, our results cannot be generalized to that subgroup. Nevertheless, our results may include a small subset of children who required a heart transplant at a later timepoint in their life; however this remains purely speculative." While we could identify a risk of bias in some of the reviewed studies, the evaluation of the risk of bias across reviewed studies remained limited due to the few studies that were assessed as having a high risk of bias in each domain. Nevertheless, the lack of explicit blinding found in several studies constitutes an important limitation.

CONCLUSION

The results of this review highlight that infants with CHD have an increased risk of motor skill impairments across infancy, childhood and adolescence. These findings stress the importance of the latest American Heart Association guidelines that recommend the implementation of systematic standardized screening or evaluation of motor skills across childhood and adolescence in children with CHD requiring open-heart surgery to allow for timely detection of motor impairments throughout childhood and adolescence.

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Study	Country	Design Cohort	Population (Dx)		Surgery (Type, age, year)	Age	n (controls)	Outcome	Results All CHD	Results Controls and/or Subgroups	
Aly et al. 2017	USA	Cohort CNHI	TGA 24% CoA 5% TOF 4% Other 2V 12%	HLHS 27% HLHS variants 20% Other SV 7%	< 1 month	6 months[a]	54	BSID II (PDI)	76 ± 15	*SV* 68 ± 15[†]	*2V* 83 ± 9[†]
Bhutta et al. 2012	USA	RCT	VSD		2004-2007	5 months	24	BSID II (PDI)	Unpublished data provided by authors used for descriptive statistics.	Published results: No significant differences in pre-operative and post-operative data.	
Cheatham et al. 2015	USA	Cohort NCH	HLHS		Hybrid stage 1 palliation 2010-2012	62 days (mean)	14 (6)	TIMP	63.9 ± 18.1[*] 54.6% < -2SD data for control not reported		
Chen et al. 2015	USA	Cross-sectional	SV 40% TGA 40% Interrupted Aortic Arch 10% TOF 10%		< 1 month	105 days (mean)	10 (14)	BSID III (PDI)	90.10 ± 7.36 2/10 (20%) < -1SD 0/10 < -2SD	*Controls* 95.86 ±10.26 1/14 (7.1%) < -1SD 0/10 < -2SD	
Hoskoppal et al. 2010	USA	Cohort Omaha	SV 26% 2V 74%		< 8 months 1999-2006	8.8 months (mean)	100	BSID II (PDI)		*SV* BSID 75.8 ± 14[†]	*2V* BSID 91.3 ± 13.2[†]
								AIMS		AIMS 17/23 (74%) < 5th percentile[†]	AIMS 20/70 (29%) < 5th percentile[†]
								RGDI		RGDI 47.6 ± 18.5[†]	RGDI 60.8 ± 22.5[†]
Long et al. 2012	Australia	Cohort RCH	SV 32% 2V 68%		< 8 weeks 2006-2008	4.2 months (mean)	48	AIMS	Unpublished data provided by authors used for descriptive statistics.	Published results: 11.76 ± 3.03	
Medoff-Cooper et al. 2016	USA	Cohort CHP	HLHS 24% TGA 24% CoA 10% DILV 7% TOF 7% DORV 6% Tricuspid atresia 6%		< 30 days	6 months[a]	51	BSID II (PDI)	81 ± 14	*SV* 76.58 ± 14.77	*2V* 83.97 ±13.47

Rocha et al. 2009	Brazil	Cohort HCSA	VSD 25% AVSD 25% Shone's complex 5% Aortic Stenosis with VSD 5% TOF10% TAPVC 10% PA 10% ASD 5% Truncus arteriosus 5%	2001-2002	6.7 months (mean)	20	DDST II	55% suspicion of delay
Solomon et al. 2018	India	Cross-Sectional	Acyanotic (49%) Cyanotic (51%)	2013-2014	5 months (median)	158	DASII	81.2 ± 33.02 53/158 (33.5 %) < -2SD

Legend: 2V, Two Ventricles; AIMS, Alberta Infant Motor Scale; ASD, Atrial Septal Defect; AVSD: Atrioventricular Septal Defect; BSID: Bailey Scales of Infant Development; CHD: Congenital Heart Defect; CHP: Children's Hospital of Philadelphia; CoA: Coarctation of the Aorta; CNHI: Children's National Heart Institute; DASII: Developmental Assessment Scale for Indian Infants; DDST: Denver Developmental Screening Test ; DILV; Double Inlet Left Ventricle; DORV: Double Outlet Right Ventricle; Dx: Diagnosis; FM: Fine Motor ; GM: Gross Motor; HCSA: Hospital da Criança Santo Antônio; HLHS: Hypoplastic Left Heart Syndrome; NCH: Nationwide Children's Hospital; PA: Pulmonary Atresia; PDI: Psychomotor development index; RCH: Royal Children's Hospital; RCT: Randomized controlled trial; RGDI: Revised Gesell Developmental Index; SD: Standard Deviation; SV: Single Ventricle; TAPVC: Total anomalous pulmonary venous connection ; TGA: Transposition of the great arteries; TIMP: Test of Infant Motor Performance; TOF: Tetralogy of Fallot; USA: United States of America; VSD: Ventricular Septal Defect

* Significant difference between cases and norms

† Significant difference between subgroups

[a] based on methodology

Table 1. Characteristics and results of studies evaluating motor outcomes in infants

Study	Country	Design Cohort	Population (Dx)	Surgery (Type, age, year)	Age	n (controls)	Outcome	Results All CHD	Results Controls and/or Subgroups		
Alton et al. 2007	Canada	Cohort SCH	TAPVC	< 6 weeks 1996-2004	21 months (mean)	34	BSID II (PDI)	89 ± 13 2/34 (6%) < -2 SD			
Aly et al. 2017	USA	Cohort CNHI	TGA 24% HLHS 27% CoA 5% HLHS TOF 4% variants 20% Other 2V Other SV 7% 12%	< 1 month	21 months [a]	54	BSID II (PDI)	74 ± 17	*SV* 68 ± 17 †	*2V* 82 ± 15 †	
Andropoulos et al. 2014	USA	Retrospective Cohort BCM	SV 47% TGA 33% Other 2V 20%	CPB > 60 min	12 months [a]	59	BSID III (PDI)	89.6 ± 14.1			
Atallah et al. 2008	Canada	Cohort SCH	HLHS or its variants	Norwood 1996-2005 MBTS 1996-2002 RVPA 2002-2005	21 months (mean)	56	BSID II (PDI)		*MBTS* 67 ± 19 † 18/30 (60%) ≤ -2SD†	*RVPA* 78 ± 18 † 8/26 (31 %) < -2SD	
Bartlett et al. 2004	USA	RCT CHB	TGA	ASO 1988-2000	12 months [a]	272	BSID BSID II (PDI)		*Prenatal diagnosis* z score: -0.92 ± 0.93 6% ≤ -2 SD	*Postnatal diagnosis* z score: -0.88 ± 1.05 17% ≤ -2 SD	
Bellinger et al. 2001	USA	RCT CHB	TGA 52.3 % TOF/Other 34.6% VSD/CAVC 13.1%	< 9 months 1992-1996	13.2 months (mean)	111	BSID (PDI)		*TGA* 99.0 ± 16.8† 14% < -2SD	*VSD/CAVC* 90.3 ± 18.3 31% < -2SD	*TOF/Other* 89.7 ± 19.8 25% < -2SD
Claessens et al. 2018	Netherlands	RCT WCH	Aortic arch obstruction		24 months	32	Dutch BSID III (PDI)	101 ± 11	*No/Mild WMI* GM 9 (8-10) FM 12 (10-15)	*WMI* GM 8 (6–9) FM 12 (11–14)	
Dittrich et al. 2003	Germany	Cohort German Heart Institute	TGA 24% APVC 4% CoA 15% ASD/VSD 28% TOF 15% PDA 3% Other 7%	<11 months 1998-1999	12.2 months (median)	90 (20)	Griffiths Locomotor skills		*Corrective surgery* 99.4 ± 20.1†◊	*Palliative surgery* 73.5 ± 16.4†◊	*Controls* 108.2 ± 19.7
Freed et al. 2006	Canada	Cohort SCH	TGA	1996-2004	18-24 months[a]	82	BSID II (PDI)	92 ± 15 5/82 (6.1%) < -2SD			

Goldberg et al. 2007	USA	RCT UMCHC	HLHS 87% Other SV 13%		Norwood < 30 days 2001-2005	12 months (median)	50	BSID-II (PDI)	77.1 ± 21	*RCP* 74.0± 20.3 *DHCA* 79.6±20.9
Gunn et al. 2016	Australia	Cohort	TGA 29% CoA 8% Other 2V 17%	PA 15% HLHS 21% Other SV 10%	< 2 months 2005-2008	24 months (mean)	130	BSID-III (PDI)	96.8 ± 12.5* 16/130 (12%) < -1SD 2/130 (2%) <70	SV performed significantly worse than 2V
Hoskoppal et al. 2010	USA	Cohort Omaha	SV 26% 2V 74%		< 8 months 1999-2006	17 months (mean)	47	AIMS		*SV* AIMS 3/10 (30%) <5th percentile *2V* AIMS 9/40 (23%) <5th percentile
								RGDI		Gesell 74.5 ± 16.1 Gesell 78.7 ± 23.6
Hülser et al. 2007	Germany	Case-Control	TGA 57% VSD 43%		after 1996	2 years [a]	17 (5)	ET6-6		*TGA* *VSD* *Controls* 0.29± 0.80◊ 0.08± 1.38◊ 0.34± 0.84
Ibuki et al, 2012	Japan	Cohort Toyama University Hospital	TGA 30% SV 70%		2003-2009	15.4 months (mean)	33 (46)	BSID II (PDI)		*TGA* 94.6 ±13.9 † *SV* 75.9 ± 18.5 *†
Knirsch et al. 2012	Switzerland	Cohort UCHZ	HLSH 75% DILV 20% DROV 5%		Norwood or Hybrid < 2 months	12 months[a]	20	BSID II (PDI)		Median 57 (49-99)* 12/20 (60%) < -2SD
Limperopoulos et al. 2002	Canada	Cohort MCH	Mixed		<2 years	20.7 months (mean)	81	PDMS	GM 84.1 ± 14.7 42% < 1.5 SD FM 83.1 ± 13.9 42% < 1.5 SD	
								Griffiths	102.8 ± 17.5 26% < 1.5 SD	
Long et al. 2012	Australia	Cohort RCH	2V 68% SV 32%		< 8 weeks 2006-2008	16 months (mean)	45 [d]	AIMS	Unpublished data provided by author for meta-analysis purpose only.	Published results: 52.79 ± 8.19
Mackie et al., 2013	Canada	Cohort Stollery	HLSH 26% TGA 36% TAPVC 16% Other 21%		< 6 weeks 2002-2006	18-24 months	47	BSID II (PDI)		Norwood 77.5 ± 18.6 Other 82.5 ± 17.1
										Other diagnostic group were not included because they overlap with samples presented in Freed et al. (2006) and Alton et al. (2007).

Maenpaa et al. 2016	Finland	Cohort HUCH	HLHS 60% SV 40%	2002-2005	12.3 months (mean)	30 (42)	AIMS		*HLHS* 70 ± 21	*SV* 72 ± 23	*Controls* 92 ± 8
Matsuzaki et al. 2010	Japan	Case-control	VSD	2004-2008	12 months[a]	39 (108)	BSID II (PDI)	79.6 ± 16.0 * 23/39 59% < -1SD below control	*Controls* 93.6 ±14.5		
Medoff-Cooper et al. 2017	USA	Cohort CHP	HLHS 24% TGA 24% CoA 10% DILV 7% TOF 7% DORV 6% Tricuspid atresia 6%	< 30 days	12 months[a]	72	BSID II (PDI)	80 ± 16	*SV* 73.94 ± 15.78 †	*2V* 84.58 ±15.33†	
Peyvandi et al. 2018	USA Canada	Cohort UCSF-UBC	SV TGA		12 months[a]	104	BSID II (PDI)		*SV* 70.7 ± 28.3	*TGA* 83.2 ± 21.23	
Ravishankar et al. 2013	USA Canada	RCT ISV Trial	HLSH 59% Other SV 41%	SCPC 2003-2007	14 months (mean)	170	BSID II (PDI)	80.3 ± 18.1 58% < -1SD 28% < -2SD			
Robertson et al. 2004	Australia	Cohort Prince Charles Hospital	TGA 34% TOF 26% VSD 23 % TAPVC 6% CAT 6% SV 6%	1999-2001	1 year [a]	35	BSID II (PDI)	89 ± 20 6/35 (17.1%) < -2 SD Significantly lower than pre-surgery			
Sarajuuri et al. 2010	Finland	Cross-sectional	SV 61% HLHS 39%	Norwood 2002-2005	30.2 months (median)	34 (41)	BSID II (PDI)		*HLHS* 80.7 ± 27.1◊† 91.0 (27-118)	*SV* 94.5 ± 10.8◊† 92.5 (79-118)	*Controls* 105.3 ± 9.1 106 (86-121)
Solomon et al. 2018	India	Cross-Sectional	Acyanotic (49%) Cyanotic (51%)	2013-2014	14 months (median)	152	DASII	92.4 ± 26.02 22/152 14.5% < -2SD			
Toet et al. 2005	Netherlands	Cohort WCH	TGA (no brain abnormalities on preoperative ultrasound)	ASO 1998-2000	30-36 months (range)	17	BSID (PDI)	101.1 ± 17.5[c] 3/18 17% < -1SD 1/18 6% < -2SD			
Visconti et al. 2006	USA	Cross-sectional CHB	HLHS 79% SV with aortic arch obstruction 21%	Norwood 1999-2004	1 year [a]	29	BSID (PDI)	75.2 ± 14.5			
Williams et al. 2012	USA	Cohort MSCH	TGA 31% TOF 38% HLHS 31%	2008-2009	19 months (mean)	13	BSID III (PDI)	Scores below average			

Wypij et al. 2008	USA	RCT Hematocrit trial	TGA 40% TOF 34% VSD 27%	1996-2004	1 year [a]	215	BSID II	86.2 ± 15.7

2V: Two Ventricles; AIMS: Alberta Infant Motor Scale; APVC: Anomalous Pulmonary Venous Connection; ASD: Atrial Septal Defect; ASO: Arterial Switch Operation; BCM: Baylor College of Medicine; BSID: Bailey Scales of Infant Development; CAT: Complex Arterial Trunk; CAVC: Complete Atrioventricular Canal Defect; CHB: Children's Hospital Boston; CHD: Congenital Heart Defect; CHP: Children's Hospital of Philadelphia; CHW: Children's Hospital of Wisconsin; CNHI: Children's National Heart Institute; CPB: Cardiopulmonary Bypass; CoA: Coarctation of the Aorta; DASII: Developmental Assessment Scale for Indian Infants; DHCA: Deep hypothermic circulatory arrest; DILV; Double Inlet Left Ventricle; DORV: Double Outlet Right Ventricle; Dx: Diagnosis; FM: Fine Motor; GM: Gross Motor; Griffiths: Griffith Developmental Scales; HLHS: Hypoplastic Left Heart Syndrome; HUCH: Helsinki University Central Hospital; ISV: Infant Single Ventricle; MBTS: Modified Blalock-Taussig Shunt; MCH: Montreal Children's Hospital; MSCH: Morgan Stanley Children's Hospital; PA: Pulmonary Atresia; PDA: Patent Ductus Arteriosus; PDI: Psychomotor development index; PDMS: Peabody Developmental Motor Scale; RCH: Royal Children's Hospital; RCP: Regional Cerebral Perfusion; RCT: Randomized Controlled Trial; RGDI: Revised Gesell Developmental Index; RVPA: Right Ventricle-to Pulmonary Artery; SCH: Strollery Children's Hospital; SCPC: Superior Cavopulmonary Connection; SD: Standard Deviation; SV: Single Ventricle; TAPVC: Total Anomalous Pulmonary Venous Connection ; TGA: Transposition of the Great Arteries; TOF: Tetralogy of Fallot; UCHZ: University Children's Hospital Zurich; UCSF-UBC: University of California-San Francisco Benioff Children's Hospital (UCSF) and University of British Columbia (UBC); UMCHC: University of Michigan Congenital heart center; USA: United States of America; VSD: Ventricular Septal Defect; WCH: Wilhelmina Children's Hospital; WMI: White Matter Injury

* Significant difference between cases and norms
◊ Significant difference between cases and controls
† Significant difference between subgroups
[a] Based on methodology

Table 2. Characteristics and results of studies evaluating motor outcomes in toddlers

Study	Country	Design Cohort	Population (Dx)		Surgery (Type, age, year)	Age	n (controls)	Outcome	Results All CHD	Results Controls and/or Subgroups			
Bellinger et al. 1999	USA	RCT	TGA		ASO < 3 months 1988-1992	49 months (mean)	158	PDMS	GM 9th percentile FM 4th percentile	*IVS CA* GM 257 ± 18 FM 190 ± 10	*IVS LFB* GM 262 ±-14 FM 193 ± 10	*VSD CA* GM 246 ± 15 FM 186 ± 12	*VSD LFB* GM 259 ± 16.1 FM 192 ± 8
Claessens et al. 2018	Netherlands	RCT WCH	Aortic arch obstruction			5.9 years (mean)	34	Dutch M-ABC II	20th centile (SD 21st) 9/34 (26%)< -2 SD	*No WMI* Total 8 (5-9)		*WMI* Total 6 (5-7)	
Hoffman et al. 2005	USA	Cohort CHW	HLHS 1996-1999		Fontan Procedure	4.5 years (mean)	13	MSCA-M	42 ± 10* 1/13 (8%) < 2SD				
Hövels-Gürich et al. 1997	Germany	Case series	TGA		ASO 1986-1992	5.4 years (mean)	56	KSBCT (GM)	88.7 ± 14.4* 32.4% * < -1SD	*No neurologic damage* 90.1 ± 13.5 25.6% < -1SD			
								DDST	FM 22.1% impairment GM 23.4% impairment				
Hülser et al. 2007	Germany	Case-Control	TGA 57% VSD 43%		after 1996	5-6 years[a]	14 (10)	ET6-6		*TGA* 0.30 ±0.98◊ (distance score)	*VSD* 0.10 ± 0.69◊	*Controls* 0.88 ± 0.41	
Ibuki et al, 2012	Japan	Cohort Toyama	TGA 30% SV 0%		2003-2009	38.8 months (mean)	33	BSID II (PDI)		*TGA* 97.3 ±13.4 †		*SV* 79.3± 18.5 †*	
Krueger et al. 2015	Switzerland	Cohort UCHZ	Acyanotic VSD 18% AVSD 5% CoA 4% Other Acyanotic 6%	Cyanotic TGA 29% TOF 9% PA 5% DORV 3% TAC 4% TAPVC 3% Other cyanotic 2V 2% SV 11%	< 1 year 2004-2008	4.3 years (mean)	141	M-ABC-2	Mean percentile (range) Total score 47.8 (2-98) Balance 44.9 (1-99) * Aiming and catching 44.0 (1-100) * Manual dexterity 46.8 (1-98)				
Long et al. 2016	Australia	Cohort RCH	Mixed Biventricular 70% SV 30%		< 2 months 2006-2008	5 years[a]	32 [d]	BOT-2	Unpublished data provided by author for meta-analysis purpose only.	Published results: Brief Standard Score 44 (39–48) 11/33 (32%) < -1SD			

Maenpaa et al. 2016	Finland	Cohort HUCH	HLHS 60% SV 40%	2002-2005	5.1 years (mean)	30 (42)	M-ABC		*HLHS* 69 ± 26◊	*SV* 70 ± 20◊	*Controls* 88 ± 12
Majnemer et al. 2006	Canada	Cohort MCH	TGA 27% TOF 26% VSD 11% SV variants 10% DORV 6% Others 20%	< 2 years	64.2 months (mean)	77 (42)	PDMS	GM 82.7 ± 12.3 68.6 % < 1 SD 29% < 2SD FM 86.2 ± 16.3 55.8 % < 1 SD 20% < 2SD			

ASO: Arterial Switch Operation; AVSD: Atrioventricular Septal Defect; BOT-2: Bruininks-Oseretsky Test of Motor Proficiency Brief Form, Second Edition; BSID: Bailey Scales of Infant Development; CA: Total Circulatory Arrest; CHD: Congenital Heart Defect; CHW: Children's Hospital of Wisconsin; CoA: Coarctation of the Aorta; DDST: Denver Developmental Screening Test ; DORV: Double Outlet Right Ventricle; Dx: Diagnosis; FM: Fine Motor; GM: Gross Motor; HLHS: Hypoplastic Left Heart Syndrome; HUCH: Helsinki University Central Hospital; IQR: Interquartile range; IVS: Intact Ventricular Septum; KSBCT: Kiphard Shilling Body Coordination Test; LFB: Low-Flow Cardiopulmunary Bypass; M-ABC: Movement ABC; MCH: Montreal Children's Hospital; MSCA-M: McCarthy Scale of Children's abilities -Motor; PA: Pulmonary Atresia; PDI: Psychomotor development index; RCH: Royal Children's Hospital; RCT: Randomized Controlled Trial; PDMS: Peadbody Developmental Motor Scale; SD: Standard Deviation; SV: Single Ventricle; TAC: Truncus Arteriosus Communis; TAPVC: Total Anomalous Pulmonary Venous Connection ; Toyama: Toyama University Hospital; TGA: Transposition of the Great Arteries; TOF: Tetralogy of Fallot; UCHZ: University Children's Hospital Zurich; USA: United States of America; Zurich VSD: Ventricular Septal Defect; WCH: Wilhelmina Children's Hospital; WMI: White Matter Injury

* Significant difference between cases and norms

◊ Significant difference between cases and controls

† Significant difference between subgroups

[a] Based on methodology

Table 3. Characteristics and results of studies evaluating motor outcomes in preschool-aged children

Age Category	Study	Country	Design (Cohort)	Population (Dx)	Surgery (Type, age, year)	Age	N (controls)	Outcome	Results All CHD	Results Controls and/or Subgroups	
School Age	Bellinger et al. 2003	USA	RCT BCAS	TGA	ASO <3 months 1988-1992	8.1 (mean)	154	Grooved Pegboard		*TCA* n=79 60% < -1SD†	*LFCPB* n=75 32% < -1SD†
	Holm et al. 2007	Norway	Cohort RRMC	TOF 24% TGA 27% Hypoplastic right or left ventricle 15% Tricuspid atresia 2.5% Others 32%	< 12 months	10.3 (mean)	120	M-ABC	Total score 10.0 ± 7.7◊ 51/120 (42.5 %) <15th percentile◊ 31/120 (25.8%) <5th percentile◊ Dexterity 4.3 ± 4.0◊ Ball skills 2.4 ± 2.4◊ Balance 3.3 ±3.6◊	*Controls* Total motor score 4.0 ± 3.7 28/385 (7.3%) <15th percentile 9/385 (2.3%) <5th percentile Dexterity 2.1 ±2.6 Ball skills 0.9 ± 1.5 Balance 1.0 ± 1.7	
	Hövels-Gürich et al. 2002	Germany	Cohort Aachen	TGA	ASO 1986-1992	10.5 years (mean)	60	KSBCT	16/60 (26.7%) < -1SD 9/60 (15%) < -2SD		
	Hövels-Gürich et al. 2006	Germany	Cross-sectional	TOF 50% VSD 50%	1993-1998	7.4 years (mean)	40	KSBCT	86.2 ± 12.8 * 17/40 (42.5%) < -1SD 3/40 (7.5%) < -2SD	*TOF* 80.4 ± 9.5 † 13/20 (65%) <-1SD 1/20 (5%) < -2SD	*VSD* 91.6 ± 13.3 † 4/20 (20%) < -1SD 2/20 (10%) <-2SD
	Mittnacht et al. 2015	Germany	Cohort Heidelberg	TGA 25% AVSD 11% UAVC 19% VSD 14% TOF/DORV Other & PS 11% acyanotic Other 7% 14%	1994-1995	10.7 years (median)	28	LOS KF 18		*T3* Median (range) 47.5 (31-61) [a]	*Placebo* Median (range) 48 (35-73) [a]
Adolescence	Easson et al. 2018	Canada	Cross sectional	SV 16% Biventricular 84%	< 2 years	15.7 years (mean)	66	M-ABC	Total score[b] 68.3 ± 19.4 28/66 (42.4.%) < -1SD 12/66 (18.2%) < -2SD Manual dexterity[b] 7.7 ± 3.2 22/66 (33.3%) < -1SD 17/66 (25.8%) < -2SD Aiming & catching[b] 8.3 ± 3.9 20/66 (30.3%) < -1SD 15/66 (21.2%) < -2SD		

											Balance[b] 9.6 ± 3.9 12/66 (18.2%) < -1SD 9/66 (13.6%) < -2SD

ASO: Arterial Switch Operation; AVSD: atrioventricular septal defect; BCAS: Boston Circulatory Arrest Study; CHD: Congenital Heart Disease; DORV: Double Outlet Right Ventricle; Dx: Diagnosis; KSBCT: Kiphard Shilling Body Coordination Test; LFCPB: Low-Flow Cardiopulmonary bypass; LOS KF 18: Lincoln-Oseretsky Motor Development Scale; M-ABC: Movement ABC; PS: Pulmunary Stenosis; RCT: Randomized Controlled Trial; RRMC: Rikshospitalet-Radiumhospitalet Medical Centre; SD: Standard Deviation; SV: Single Ventricle; T3: tri-iodothyronine; TCA: Total Circulatory Arrest ; TGA: Transposition of the Great Arteries; TOF: Tetralogy of Fallot; UAVC: Univentricular Atrioventricular Connection; USA: United States of America; VSD: Ventricular Septal Defect

* Significant difference between cases and norms

◊ Significant difference between cases and controls

† Significant difference between subgroups

[a] Within normal range

[b] Unpublished data provided by author

Table 4. Characteristics and results of studies evaluating motor outcomes in school-aged children and adolescents

Database	Population	Outcome
Medline	exp Heart Defects, Congenital/ (congenital heart adj (disease or defect*)).ti,ab,kf.	psychomotor performance/ motor skills/ ((motor skills* or motor performance or psychomotor performance).ti,ab,kf. exp Motor Skills Disorders/ "Child Development"/
Embase	Exp congenital heart disease (congenital heart adj (disease or defect*)).ti,ab,kw	Exp motor performance Exp psychomotor performance/ (motor skill* or motor performance or psychomotor performance). ti,ab,kw exp child development/
CINAHL	(MH "Heart Defects, Congenital+") TX congenital heart defect* TX congenital heart disease	(MH "Motor Skills+") OR (MH "Motor Skills Disorders") (MH "Child Development")

Supplemental Table 1. Search strategy

Authors (years)	Blinding	Attrition bias	Baseline Imbalance
Alton et al. 2007	Unclear risk	Low risk - Low attrition	Low risk
Aly et al. 2017	Unclear risk	Low risk - Low attrition	Low risk
Andropoulos et al. 2014	Low risk - Blind to diagnosis or surgery	Low risk - Low attrition	Low risk
Atallah et al. 2008	Unclear risk	Low risk - Low attrition	Low risk
Bartlett et al. 2004	Unclear risk	Low risk - Low attrition	Low risk
Bellinger et al. 1999	Low risk - Blind to treatment assignment and clinical course	Low risk - Low attrition	Low risk
Bellinger et al. 2001	Low risk - Blind to treatment assignment	Low risk	Low risk
Bellinger et al. 2003	Unclear risk	Low risk - Low attrition	Low risk
Bhutta et al. 2012	Low risk - Blind to treatment group	Low risk - Low attrition	Low risk
Cheatham et al. 2015	Unclear risk	Low risk - Low attrition	Low risk
Chen et al. 2015	Unclear risk	Low risk - Low attrition	High risk - 70% male in CHD compared to 28.5% in controls
Claessens et al. 2018	Unclear risk	Low risk - Low attrition	Low risk
Dittrich et al. 2003	Low risk - Blind to type of cardiac defect and medical history	Low risk - Low attrition	Low risk
Easson et al. 2018	Low risk - Blind to medical history and other tests results	Low risk - Low attrition	Low risk
Freed et al. 2006	Unclear risk	Low risk - Low attrition	Low risk
Goldberg et al. 2007	Low risk - Blind to surgical technique	Low risk	Low risk
Gunn et al. 2016	Unclear risk	Low risk	Low risk
Hoffman et al. 2005	Unclear risk	Low risk	Low risk
Holm et al. 2007	Unclear risk	Low risk	Low risk
Hoskoppal et al. 2010	Unclear risk	High risk 58% attrition	Low risk
Hövels-Gürich et al. 1997	Unclear risk	Low risk	Low risk
Hövels-Gürich et al. 2002	Unclear risk	Low risk	Low risk
Hövels-Gürich et al. 2006	Unclear risk	Unclear risk	Low risk
Hülser et al. 2007	Unclear risk	High risk - 66% and 72% attrition for 2-year-old and 5-6 years outcomes respectively	High risk - Significant difference in maternal education between cases and controls.

Ibuki et al, 2012	Unclear risk	Unclear risk	Low risk
Knirsch et. al 2012	Unclear risk	Low risk	Low risk
Krueger et al. 2015	Unclear risk	Low risk	Low risk
Limperopoulos et al. 2002	Low risk - Blind to type of cardiac defect, operative procedures and other pertinent medical history.	Low risk	Low risk
Long, Harris et al. 2012	Low risk - Blind to infant's medical history	Low risk	Low risk
Long et al. 2016	Low risk - Blind to medical and surgical history	Low risk	Low risk
Mackie et al. 2013	Unclear risk	Low risk	Low risk
Maenpaa et al. 2016	Unclear risk	Low risk	Low risk
Majnemer et al. 2006	Low risk - Blind to medical history and findings of previous developmental assessments	Low risk	Low risk
Matsuzaki et al. 2010	Unclear risk	Unclear risk	Low risk
Medoff-Cooper et al. 2016	Unclear risk	Low risk	Low risk
Mittnacht et al. 2015	Low risk - Blind to treatment assignment	Unclear risk	Low risk - CHD were older than controls
Peyvandi et al. 2018	Low risk - Blind to diagnosis, clinical factors, and brain imaging findings	Low risk	High risk - Significant difference in male/female ratio between the 2 groups.
Ravishankar et al. 2013	Low risk - Double blind	Low risk	High risk - Significant between site differences for motor outcome
Robertson et al. 2004	Unclear risk	Low risk	Low risk
Rocha et al. 2009	Low risk - Blind	Low risk	Low risk
Sarajuuri et al. 2010	Unclear risk	Low risk	Low risk
Solomon et al. 2018	Unclear risk	Low risk	Low risk
Toet et al. 2005	Unclear risk	Low risk	Low risk
Visconti et al. 2006	Unclear risk	Low risk	Low risk
Williams et al. 2012	Low risk - Blind to diagnosis	Low risk	Low risk
Wypij et al. 2008	Low risk - Blind to treatment assignment	Low risk	Low risk

Supplemental Table 2. Quality assessment of reviewed studies

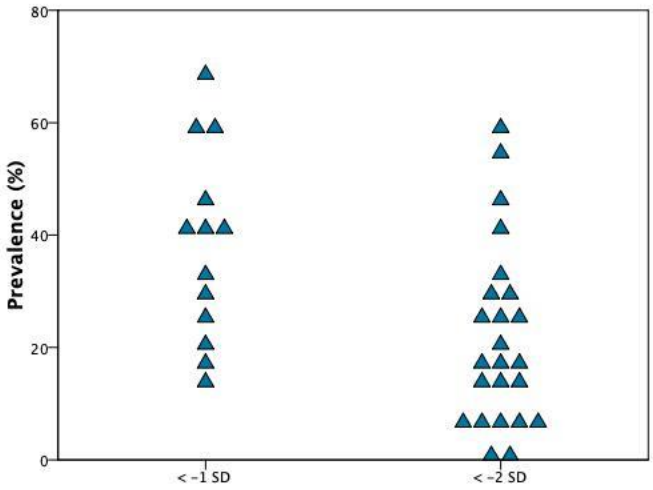


Figure 1. Prevalence of Motor Impairments Across Age Groups. In this figure, each triangle represents a study

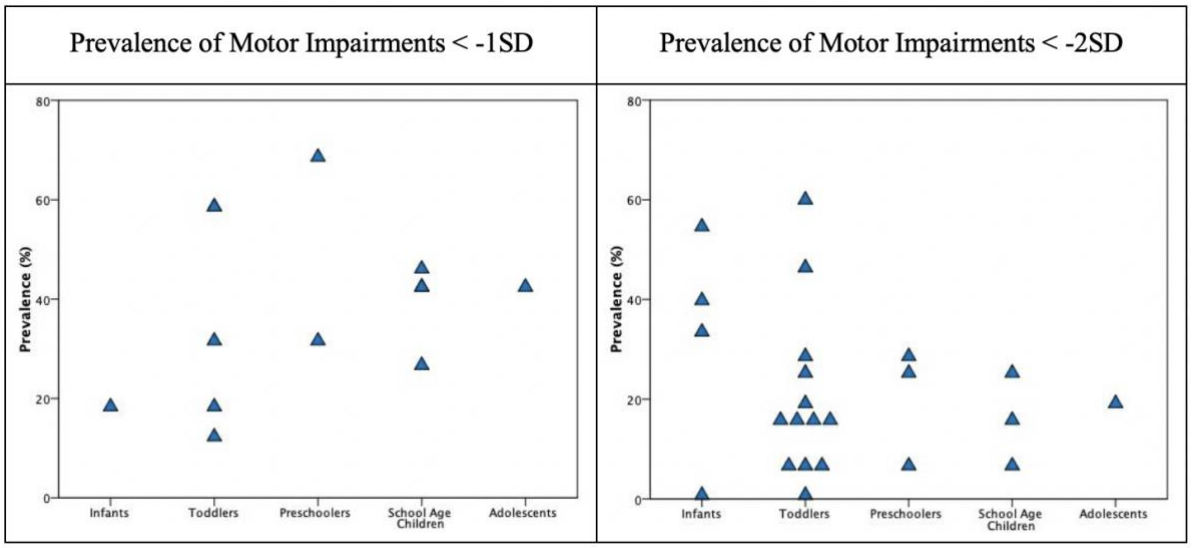
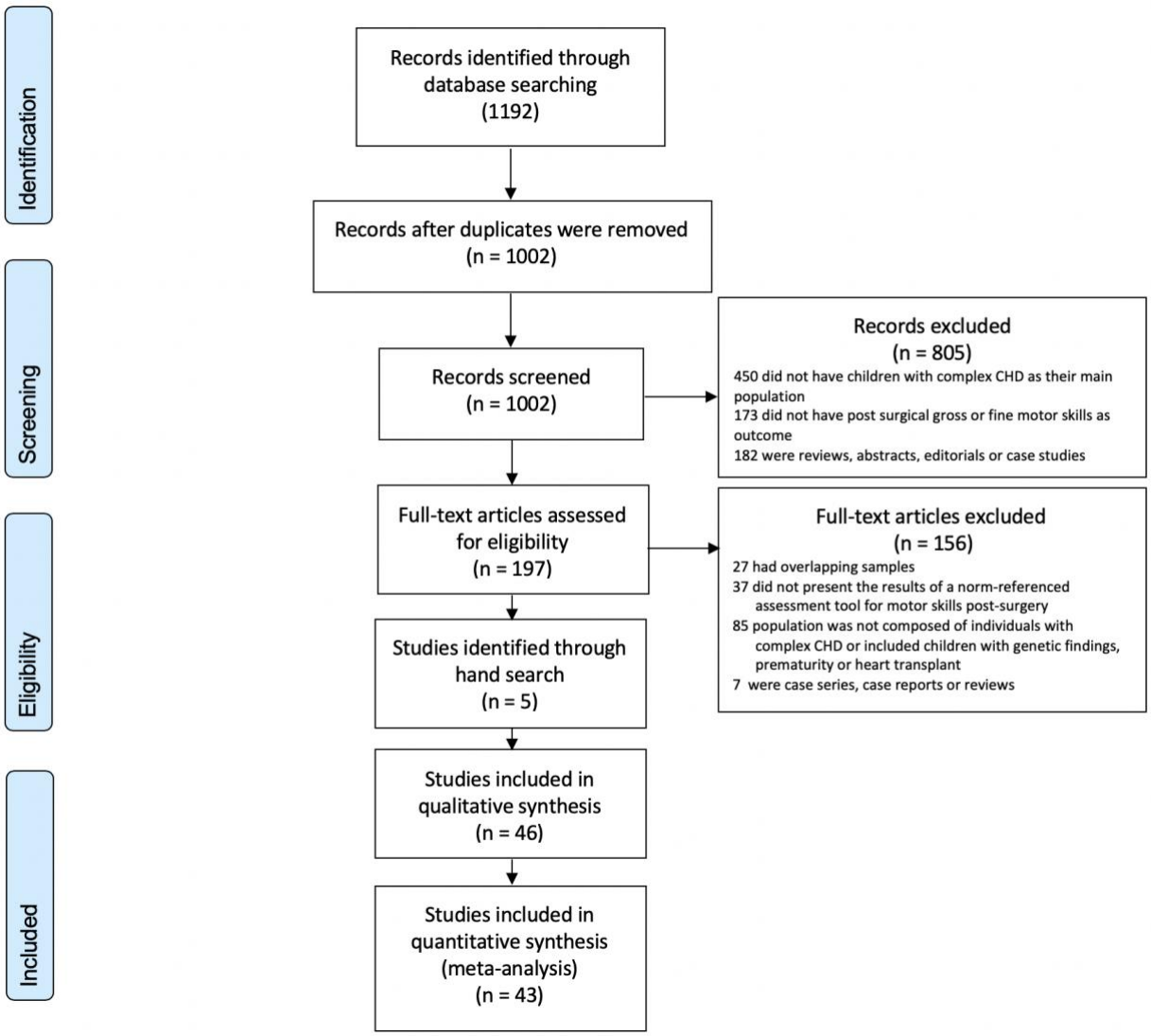


Figure 2: Prevalence of motor impairments for each age group. In this figure, each triangle represents a study



Supplemental Figure 3. Study selection

CHAPTER 5: INTRODUCTION TO THE CANADIAN ENVIRONMENTAL SCAN

In the previous chapters, we examined the frequency and extent of developmental impairments in children and adolescents with a congenital heart defect (CHD). Considering the high prevalence of challenges reported and their possible impact on daily functioning, it is essential to have systematic developmental follow-up practices in place to identify delays in a timely manner and ensure that appropriate resources and supports are provided to optimize outcomes.

Recommendations for best practices with regard to the developmental follow-up of children with a CHD have been published by the American Heart Association (Marino et al., 2012). The implementation of the proposed algorithm, which is based on the American healthcare system, has faced some challenges within the United States of America itself (Knutson et al., 2016). In fact, difficulty accessing specialists and the resistance of families have been documented as important barriers (Knutson et al., 2016).

There is currently no available data on developmental follow-up practices or barriers to implementing optimal practices in Canada. Barriers to guideline implementation can be multiple. They exist at individual, organizational (structure) and attitudinal levels (Forsner et al., 2010). The Canadian healthcare system differs from the American system both from an accessibility and funding perspective, and it is therefore expected that the barriers faced during the implementation of the recommendations would also differ. In fact, it is important to question whether their implementation is even feasible in the Canadian context.

The next chapter presents the results of a national environmental scan that was conducted with all Canadian institutions that perform pediatric open-heart surgery. The primary aim of this study was to describe current developmental follow-up practices in tertiary care centers that perform pediatric open-heart surgery in Canada. The secondary aim was to explore perceptions of structural barriers to optimal developmental follow-up of children with a CHD post open-heart surgery in Canada.

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CHAPTER 6: CANADIAN DEVELOPMENTAL FOLLOW-UP PRACTICES IN CHILDREN WITH CONGENITAL HEART DEFECTS: A NATIONAL ENVIRONMENTAL SCAN

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ABSTRACT

Background: Developmental follow-up is central to the timely identification of delays in at-risk children. Data regarding the follow-up of children with congenital heart defects (CHD) post open-heart surgery across Canada is currently lacking. The objective of this study was to describe current Canadian developmental follow-up practices and to explore barriers to optimal follow-up.

Methods: A cross-sectional study was implemented with health professionals involved with the developmental follow-up of children with CHD in the eight specialized hospitals that perform pediatric open-heart surgery in Canada. A questionnaire collected descriptive information about the setting and current follow-up practices. Additionally, an interview was conducted to explore what would be considered optimal developmental follow-up in Canada and identify current barriers.

Results: Four of the eight tertiary care centers had a systematic developmental follow-up program that included screening and formal evaluation. These programs were only accessible to a subset of children with CHD identified to be at higher risk. Participants described current practices as suboptimal and would like to develop a more systematic developmental follow-up program or expand an existing one. Participants emphasized the lack of human resources, financial supports and limited dedicated time as important barriers to offering optimal follow-up care.

Conclusion: Current follow-up practices in Canada are considered suboptimal by healthcare specialists involved with children with CHD. These practices may fail to promptly identify children and adolescents with CHD who experience developmental challenges. It is essential that we develop national recommendations to optimize the developmental follow-up practices in Canada for this high-risk population.

RÉSUMÉ

Contexte: Le suivi développemental est essentiel afin d'identifier rapidement les retards chez les enfants à risque. Il n'existe actuellement pas de données sur le suivi des enfants atteints d'une cardiopathie congénitale ayant nécessité une intervention chirurgicale à cœur ouvert. L'objectif de cette étude consistait à décrire les pratiques actuelles de suivi développemental et à explorer les obstacles à un suivi optimal au Canada.

Méthodologie: Une étude transversale a été menée auprès de professionnels de la santé assurant le suivi du développement d'enfants atteints de cardiopathie congénitale dans huit hôpitaux spécialisés qui pratiquent des chirurgies à cœur ouvert au Canada. Un questionnaire a permis de recueillir des renseignements descriptifs sur les établissements et les pratiques actuelles de suivi. De plus, une entrevue a été menée pour explorer ce qui pourrait être considéré comme un suivi optimal du développement au Canada et cerner les obstacles actuels.

Résultats: Quatre des huit centres de soins tertiaires disposaient d'un programme de suivi systématique du développement qui comprenait un dépistage et une évaluation formelle. Ces programmes n'étaient accessibles qu'à un sous-groupe d'enfants atteints d'une cardiopathie congénitale, identifiés comme étant à risque élevé de retard de développement. Les participants ont décrit les pratiques actuelles comme sous-optimales et souhaitaient mettre en place un programme de suivi développemental plus systématique ou à élargir un pro- gramme existant. Les participants ont souligné le manque de ressources humaines et financières ainsi que le peu de temps qui peut être consacré au suivi comme étant les principaux obstacles pour offrir un suivi optimal.

Conclusions: Les professionnels de la santé œuvrant dans le traitement des enfants atteints de cardiopathie congénitale considèrent que les pratiques actuelles de suivi au Canada sont sous-optimales. Ces pratiques peuvent ne pas permettre d'identifier rapidement les enfants et les adolescents atteints de cardiopathie congénitale qui présentent des retards de développement. Il est essentiel que nous élaborions des recommandations nationales pour optimiser les pratiques de suivi développemental au Canada pour cette population à risque élevé.

Keywords

Congenital heart defect, surveillance, evaluation, follow-up, neurodevelopment, health services

INTRODUCTION

Children and adolescents with congenital heart disease (CHD) are at high risk for developmental delays.[1-3] These delays can affect various developmental domains such as cognitive, language, psychosocial and motor abilities to different degrees based, in part, on the complexity of the CHD.[1-4] While approximately 10% of children with less complex forms of CHD (e.g., atrial septal defect and ventricular septal defect) experience persisting developmental impairments, delays are documented in up to 50% of the children with more complex CHD (e.g., tetralogy of Fallot, coarctation of the aorta, transposition of the great arteries, and single functional ventricle).[4] Some of these developmental challenges will become evident in early childhood but others, such as impaired executive functions and learning, become apparent only as the child gets older or during adolescences.[1, 4, 5]

Developmental follow-up practices for at-risk populations include surveillance, screening and/or the formal evaluation of developmental domains and activity limitations.[6] Developmental surveillance is a flexible and longitudinal process that involves identifying and documenting the presence of risk factors, as well as the collecting of parents' concerns over time.[6] Developmental screening typically relies on short validated parental questionnaires, which can be used on their own or to further assess areas of concerns identified during the developmental surveillance.[6] Finally, formal developmental evaluation refers to the use of standardized evaluations of developmental domains performed by healthcare professionals to assess the presence of developmental disorders or impairments.[6] Appropriate follow-up is essential to the early identification of impairments and subsequent referral to timely interventions. This is especially important during the first years of life to capitalize on the potential of early intervention to optimize child development.[7] However, given that developmental challenges evolve across childhood and adolescence in survivors of CHD, developmental follow-up should continue throughout childhood and adolescence.

In 2012, the American Heart Association (AHA) published the first scientific statement that recommended systematic follow-up practices to optimize the developmental outcome of children with CHD.[4] They proposed an algorithm for the surveillance, screening, and evaluation of children with CHD based on their risk of developmental disorders. In this statement, the primary care provider is recommended to perform developmental surveillance during routine follow-up visits for all children with CHD. In addition, children identified at higher risk of delays, such as those who underwent open-heart surgery, should be directly referred for formal developmental evaluation at specific timepoints during

childhood.[4] In addition to the administration of child-specific measures, the AHA statement also recommends parent-child observation to assess, social skills, language, and parental stress.

The AHA 2012 statement has been instrumental in raising awareness of the complex structure and process that need to be put in place for the optimal monitoring of children with CHD and is regarded as the gold standard in various countries.[4, 8, 9] However, it remains unclear to what extent these recommendations were implemented outside the United States (USA) and whether their implementation is feasible in other contexts. In countries with publicly funded healthcare systems such as Canada, the absence of the medical home structure, on which the recommendations were based, may present a barrier to the implementation of these recommendations. In order to implement optimal practices for Canadian children with CHD, it is essential to better understand the structures of follow-up care already in place. Moreover, identifying the barriers for the implementation of systematic follow-up programs is essential to develop effective strategies to translate evidence into daily practices. Therefore, the primary objective of this study was to describe current developmental follow-up practices in tertiary care centers that perform pediatric open-heart surgery in Canada. As a secondary objective, we aimed to explore structural barriers to optimal developmental follow-up of children with CHD post open-heart surgery in Canada.

Materials and Methods

This cross-sectional study used an exploratory sequential mixed-methods approach. Quantitative data were collected from healthcare professionals at eligible institutions using a single questionnaire aimed to capture institutions' characteristics and surveillance practices. Qualitative data were collected via telephone interviews to explore respondents' perspectives regarding optimal developmental follow-up care and barriers to achieving it.

Recruitment

All institutions that performed pediatric open-heart surgery in Canada listed on the Canadian Cardiovascular Society website[10] were eligible for this study. We contacted the head of cardiology and/or developmental follow-up in each institution. We first confirmed the institution's eligibility and asked them to identify a healthcare professional with excellent knowledge of their institution's current follow-up practices to participate in the study. Nurses, cardiologists, pediatricians, psychologists, and occupational therapists involved in the developmental surveillance of children and adolescents with CHD

were all eligible professionals to participate in this study. Once the healthcare professional was identified, they were contacted to confirm eligibility and interest in participating in this study. The participant was then sent the consent form and questionnaire via email and was encouraged to gather information from other professionals/units when necessary. Additional departments were contacted if the developmental follow-up of children with CHD was overseen by more than one department or unit.

Data Collection

The questionnaire was divided in two sections: 1) descriptive information, and 2) current surveillance practices. It included both categorical and short answer questions to identify unit characteristics and current surveillance practices for children with CHD post open-heart surgery. During the subsequent telephone interview, answers to the questionnaire were reviewed if clarification was needed. The interview primarily addressed the feasibility and acceptability of AHA recommendations and explored what would be considered optimal developmental follow-up in Canada and identify current barriers. Interviews were conducted in either French or English and were audio recorded.

Analysis

Demographic and other descriptive data obtained from the questionnaires and interviews were synthesized using descriptive statistics. Current practices were summarized using schematic representations of the children's developmental follow-up trajectories. Qualitative data were transcribed verbatim and analyzed using NVivo 11 software (QSR International) to categorize and analyze data, as well as provide illustrative quotes. Thematic analysis was used to identify common patterns or trends in responses using inductive and deductive coding. A mixed-method matrix was used to integrate and interpret the qualitative and quantitative data during the analysis. French quotes were translated to English to maintain anonymity of the source. All analyses were performed by the first author (MEB) and discussed with the research team.

Results

All eight tertiary centers that performed pediatric open-heart surgeries in Canada participated in our study. A total of ten practicing health professionals were interviewed for this study, and included cardiologists (3/10), nurses (3/10), pediatricians (2/10), one neonatologist and one psychologist. They worked either in the Division of Cardiology (4/10), Neonatal Follow-up (3/10), Child Psychology (1/10), or in a specialized developmental follow-up program for children with CHD (2/10). All participants had

been working in their current unit for a minimum of 5 years except for one who had been there for 1.5 years. The participants were selected based on their knowledge of the follow-up practices of the children who undergo surgical repair of their heart defects. The institutions were either pediatric hospitals or a pediatric pavilion of a larger general hospital or university health center and were located in five different provinces (Alberta, British Columbia, Nova Scotia, Ontario, Quebec. The number of pediatric open-heart surgeries performed yearly from 2017 to 2019 in these centers ranged from 68 to 600 (median 108/year). At most centers (6/8), only one unit or clinic was overseeing the developmental follow-up of children with CHD. Two institutions had follow-up services that could involve more than one unit or clinic.

A. Current Canadian Developmental Follow-Up Practices

Developmental follow-up care practices

We identified four different follow-up pathways across the participating Canadian institutions. Half of the institutions (4/8) had a structured developmental follow-up program for children with CHD that included formal surveillance, screening, and evaluation. Two of these programs were set up as independent developmental follow-up clinics while the other two were housed within the center's Neonatal Follow-up Program. The two centers that performed the highest number of surgeries had a structured developmental follow-up program. Of the four remaining centers, one offered a subgroup of infants who were discharged from the Neonatal Intensive Care Unit with a formalized surveillance program by developmental pediatricians through the Neonatal Follow-up Program and another center systematically referred children with single ventricle physiologies for psychological evaluation. Finally, the developmental follow-up services for children with CHD in two institutions was limited to informal surveillance during cardiology appointments. The different structures are illustrated in Figure 1.

Eligibility for developmental follow-up care

Only a subset of children with CHD requiring open-heart surgery were eligible to be followed by the four structured developmental follow-up programs. The eligibility criteria for these programs varied from one center to another and were based on potential increased risk (e.g., age at surgery, diagnosis, and presence of additional risk factors). The main criteria are summarized in Table 1.

Duration and frequency of developmental follow-up

For the four centers that offered a structured developmental follow-up program that included systematic screening and evaluation throughout infancy and preschool age, formal evaluations were typically performed twice during the first year of life, then once a year or once every two years in children between one and five years of age. During school age and adolescence, developmental follow-ups were offered only if there was a previously identified concern. The frequency of follow-up for these programs are presented in Table 2.

Children who benefited from periodic surveillance through the Neonatal Follow-up Program were evaluated at five timepoints from the time of their surgery to five years of age (4 months, 9 months, 18 months, 36 months, and preschool years). The children followed by Child Psychology underwent a standardized evaluation between 18-24 months and at 5 years of age. Finally, the frequency of surveillance through the Cardiology unit depended on the cardiac diagnosis and other factors related to the heart condition. Children were typically followed through cardiology until they were 18 years of age except in one institution that continued following their patients throughout their entire life.

Format and tools used for surveillance, screening, and evaluation

The four developmental follow-up programs relied on different approaches to identify developmental delays. While one program used a combination of phone surveillance, web-based screening questionnaires and in-person evaluation, the three others relied on in-person screening and evaluation tools. Chosen screening and evaluation tools focused on different developmental domains (e.g., motor, cognition, language) or were disorder specific (i.e., autism). The tools used by the four developmental programs at the time of this study are summarized in supplemental Table S1.

Referral to intervention services

The study participants referred children with CHD to a variety of services for different reasons ranging from general concerns to hearing loss. Please refer to table 3 for detailed information on these services and reasons for referrals. The process to refer children to intervention services differed widely from one institution to the other and between provinces. For example, one province had a developmental program to which parents can self-refer, while in another province, only children with a score two standard deviations below the normative mean on a formal evaluation could access rehabilitation services within the public healthcare system and was per physicians' referral only. Referral to

intervention services was considered frequent (36-70%) at three institutions and rare (<10%) or occasional (11-35%) at five of them. As illustrated in Figure 1, referrals to rehabilitation services were more frequent in centers with a structured developmental follow-up program that included systematic screening and evaluations compared to those where children's development was followed through surveillance only.

Parental mental health

Parents' mental health was formally screened in two of the eight centers. In these institutions, the parents were given a questionnaire to complete, which was then reviewed by a nurse or psychologist. The tools used for assessing parental stress are described in Supplemental Table S1. One centre reported that family coping was addressed informally but systematically during history taking for cardiac follow-up. Finally, at one center with a formal follow-up program a social worker was involved in supporting families at both an in-patient and out-patient level; however, it was not clear whether any formal screening or evaluation took place.

B. Optimal Developmental Follow-Up in Canada

Eight out of the ten participants interviewed reported being familiar or somewhat familiar with the AHA 2012 statement regarding recommended developmental follow-up care practices.[4] These recommendations for the developmental follow-up of infants at high risk for developmental delays are summarized in Figure 2. Of those who were familiar with it, two participants mentioned that the recommendations could not realistically be applied in Canada given the differences in our healthcare system and limited resources. With respect to the feasibility of using the AHA statement [4] in Canada, one participant stated:

"I do remember I looked at them thinking that's not going to fly. […], that's how a lot of the guidelines in the States apply to us… right? From a resource potential, we don't have that." (Participant 8)

Conversely, one participant expressed that it might be more realistic to implement this type of follow-up in Canada since the families would not have to pay out of pocket for the services. The other five participants expressed that the AHA statement was useful but would have to be adapted to the Canadian context. One participant stated:

"From my perspective, I think a lot of the concepts can be applied in Canada, where healthcare is provincial, I would say […] with the ability to tweak some of it to fit with the way we provide care in [the

provinces] and Canada. Because in the States, they use very much the medical home and we are not quite the same, but we do have things like [developmental] program[s] available. So, I feel that with tweaking, it's a mold that can be used in Canada and I would support developing something like that for Canadian and provincial guidelines." (Participant 9)

Some participants proposed making changes to the timing of evaluations while others suggested the implementation of a screening process to identify children who would benefit from standardized evaluation. In addition, two participants noted the expertise of developmental specialists and questioned whether informal surveillance from these professionals might simply replace some of the more formal evaluations. One participant expressed: *"Probably, in the United States, it [a standardized evaluation] may be necessary. But in here, I mean it's more or less done. The pediatricians are very good, they have a lot of developmental training, and you know, our public health system is good…"* (Participant 9)

Overall, the majority of healthcare professionals (9/10) concurred that children with CHD requiring open-heart surgery at their institution would benefit from a more systematic approach as proposed in the AHA statement. Ideally, this would take the form of new developmental follow-up programs or expanding existing programs where a larger group of children with CHD would be eligible and periodically followed for a longer period. The importance of a multidisciplinary and collaborative approach was also identified by participants when exploring the feasibility of implementing a more structured follow-up program.

"Optimal would be this is child x who has this disease, they are screening at like you said one year, three years, at 10 years, automatically comes up. That formalized screening automatically is there, this is the path you are on, I don't have to worry about referring and are you going to get seen, etc. And then in between we do our informal screening to make sure that nothing has come up in between, obviously. But if we are talking about formalized stuff, it would be fantastic to have one stop shop." (Participant 8)

C. Barriers to Developmental Follow-Up

When asked about the barriers to implementing new or expanding follow-up programs, nine of the ten participants identified the lack of human or financial resources as the primary barrier of importance. One participant said: *"When you are doing outcomes research, it's really, really hard to get research money because they say the hospital should pay. And it's the opposite, right? The hospital and the research funders both say somebody else should pay for outcomes research."* (Participant 7)

Participants indicated that human and financial resources are closely related because of the financial implications of hiring and retaining healthcare professionals who can administer the formal evaluations, as expressed in the following quote: *"It goes together. It's the budget that brings the staff. There may be staff but there are no positions… there is no money […] to have additional positions to do that"* (Participant 6)

Some participants also identified that the funding for the follow-up programs currently in place in the four centers often comes from research grants or donations. Therefore, new and sustainable funds were required to develop new programs or expand existing ones. The amount of time and training required to perform formal evaluations and the presence of waitlists for evaluation were identified as additional barriers to implementing more frequent developmental evaluations.

"We don't really have the resources and worse the training to do a Bayley or it's not [possible] in our visit that's relatively short." (Participant 1)

Finally, one participant highlighted that it is sometimes difficult for parents to come back to the hospital for follow-up. *"I think that sometimes people are travelling, I think what's probably different from the States, I can't say for sure, but our catchment area is so huge."* (Participant 3)

The participants identified further challenges specific to the referral of children with identified developmental delays to intervention services. Once again, the challenges varied from one province to another. The absence of a centralized service point to access community resources was identified as a barrier in some institutions. Health care professionals exclusively working in hospitals expressed that they may not be optimally positioned to identify all potential resources offered in the community and that a centralized navigator with established relationships with community resources may be better suited to direct the patients to the appropriate services once a developmental delay is identified. Some participants also reported the presence of long waitlists as important barriers to timely intervention.

"[…] the biggest issue is obviously access and waiting and when it comes to some of those services we talk with families directly about what they have with regards to their own private insurance, right? And whether or not they have access or the wherewithal to have private testing." (Participant 8)

"You know the hospital does short, not very intensive interventions; the physiotherapist cannot see the child more than every month. In rehabilitation, if it is very severe, they […] will have more [interventions],

but I will refer them at six months, and they will wait [another] 6 to 12 months. It is not like [in the United States]." (Participant 2)

DISCUSSION

It is well established that children with CHD, who present a high risk for developmental delays, can benefit from a systematic follow-up program.[6] In the first years of life, timely identification of delays provides access to a critical window of enhanced brain plasticity and reorganization as a result of early interventions.[11] Periodic interventions during childhood and adolescence ensure that new challenges can be identified promptly, thus minimizing or preventing deleterious consequences on academic performance, age-appropriate daily life skills and psychological well-being. Our results show that only half of the tertiary care centers that perform infant open-heart surgery in Canada have a systematic process in place for developmental follow-up that includes formal screening and evaluation at specific timepoints during infancy and childhood. Moreover, these follow-up programs were only accessible to a subset of children with CHD requiring open-heart surgery who met very specific criteria, whereas others also known to be at high risk were not followed. Although most children benefited from informal developmental surveillance at every cardiology visit, as recommended by the Council of Children with Disabilities,[6] a large portion of the children who had open-heart surgery, a known risk factor for developmental disability, failed to have the formal evaluations in infancy (12-24 months), preschool years (3-5 years) and childhood (11-12 years) that are recommended by the AHA 2012 Statement.[6]

Many participants highlighted the importance of having a systematic process in place from diagnosis to intervention. In addition, if a developmental delay is identified during the evaluation, a centralized referral process was recognized as being important in order to refer the children to the appropriate intervention services. Our results demonstrate that children who undergo standardized evaluations were more likely to be referred to early intervention or rehabilitation services. Hence, a more systematic approach would provide a more consistent access to intervention services.

The importance of a family-centered approach in order to best meet the developmental needs of children with CHD is well substantiated in the literature.[12] This approach recognizes the role that the child's family plays with regard to their development and overall well-being.[12] High parental stress, which has been well documented for parents of children with CHD, can have negative impacts on a child's

development and has been associated with development and behavioural challenges.[13-15] As a result, it is important that parents' psychological well-being be evaluated as part of the child's follow-up. Among the eight centers that perform pediatric open-heart surgery in Canada, only three systematically assessed parental stress through surveillance or screening. As a result, many Canadian parents of children with CHD may not receive the care they need to cope with the added stress and anxiety generated by their child's condition.

Health care professionals involved in the developmental follow-up of children with CHD raised concerns with the feasibility of adhering to the recommendations of the AHA 2012 Statement in Canada. They highlighted that the process recommended in this statement may not be fully applicable to the Canadian health are context. The funding models are very different in the Canadian and American healthcare systems. Both countries rely on health insurance to cover the majority of healthcare fees. However, the difference lies in the organization that provides this coverage. In the USA, the health insurance can be acquired through private companies or can be provided by the government for some specific groups.[16] In Canada, healthcare is universal, provided by the government and funded through taxes.[16] Some private services are available and partially covered for those who benefit from additional private insurance. Moreover, the AHA 2012 Statement is based on the medical home model, where the primary care provider offers comprehensive care and manages the need for consultations and referrals.[17] The concept of medical home does not exist in Canada. Nevertheless, the coordination of care is often the responsibility of the family physician or pediatrician. The Canadian healthcare system also varies from one province to another since healthcare is under provincial jurisdiction. Therefore, access to resources differs at the provincial level as well. Hence, it is essential that the AHA 2012 Statement be adapted to the Canadian context.

Participants in this study identified different barriers to implementing optimal developmental follow-up care for children with CHD. The lack of financial resources and skilled healthcare professionals to administer the evaluations was raised by the majority of the participants. The time required to complete numerous standardized evaluations at regular intervals was also recognized as an important barrier to implementing the AHA 2012 Statement.[4] A study by Donnellan et al. (2013) on the barriers to the implementation of stroke guidelines in Ireland has found similar results in terms of the need for additional resources.[18] They concluded that successful implementation of practice guidelines involves a

complex interplay between structural factors, processes and attitudes of the healthcare professionals involved. Nevertheless, even with limited resources, some institutions have successfully implemented developmental follow-up for children with CHD by pulling resources from different departments/units together.[19] Therefore, it is important to carefully assess barriers and facilitators to find cost-effective solutions to implement this complex structure.

Gagliardi et al., conducted a scoping review that examined implementations of guidelines and identified strategies that can be used to address different barriers.[20] They found that education is the most commonly used approached to enhance awareness of guidelines but has little effect on changing practice.[20] Conversely, financial investments, structural changes or identification of strategies to overcome barriers were only rarely used or completely absent.[20] Considering that a large majority of the interviewed healthcare professionals were aware of the AHA Statement but face various barriers in its implementation, the use of a local champion may prove to be helpful to optimize current practices. Before the COVID-19 pandemic, the Canadian follow-up programs that offered screening and evaluations predominantly relied on face-to-face screening and evaluation. The past year has forced healthcare professionals to explore new ways in which services could be offered remotely. There have been preliminary successful reports of effective administration of some pediatric evaluations through telehealth.[21, 22] These recent reports, coupled with use of self-report screening tools and measures, suggest that novel modes of service delivery may provide solutions to address some of the barriers in optimizing follow-up in Canada.

The results of this study should be interpreted in the context of their limitations. Despite efforts to ensure that questionnaires were administered to participants with excellent knowledge of their institution's follow-up practices, the information was typically gathered from a single source. This practice may have introduced a bias as some information could have been omitted. It is also important to note that this study did not include community pediatricians and family doctors. Therefore, we cannot state with certainty that developmental evaluation and screening does not take place in the community. However, based on our knowledge of the Canadian organizational structure, formal community screening and evaluation for CHD is most likely limited or absent. This would need to be confirmed in future studies. Consistent with these assumptions, a recent study suggests that only 21% of pediatric primary care providers in the USA are aware of the AHA 2012 Statement and that even when aware,

formal developmental evaluation was often not performed due to difficulty in accessing specialists and/or resistance by families.[23] Future studies will be necessary to develop expert-based recommendations to adapt the AHA statement to the Canadian context.

CONCLUSION

This study focuses specifically on the Canadian hospitals that provide infant open-heart surgery to children with complex CHD. This study demonstrates that only a subset of Canadian children with the most complex CHD requiring infant open-heart surgery benefited from a systematic developmental follow-up program that included systematic screening and evaluation throughout childhood. Current follow-up practices in healthcare institutions across Canada are considered suboptimal from the point of view of most health professionals in this study. Current practices may fail to promptly identify children and adolescents with CHD who experience developmental challenges. Therefore, it is essential to develop policy recommendations to optimize the developmental follow-up practices in Canada for children with CHD. The results of this study lay the foundations to better understanding current barriers and opportunities within the Canadian healthcare system. Future studies should explore the use of follow-up services in other regional and community-based centres that do not conduct open-heart surgery but may follow these children following surgery.

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	Institutions (n)
Children with CHD and additional risk factors (e.g., prematurity, extracorporeal membrane oxygenation, heart transplant, cardiac arrest, etc.)	3
CHD requiring surgery with bypass < 6 weeks	2
Complex CHD requiring surgery before 1 year of age	2
Children with a single ventricle pathology	1
It is important to note that some programs used more than one of the listed criteria to determine eligibility to their program.	

Table 1. Eligibility criteria for structured developmental follow-up programs

Age	**Frequency of screening and evaluation**
Infancy: 0-12 months	Twice (n=3) Once (n=1) No formal screening or evaluation (n=1)
Toddlerhood: 1-3 years	Twice (n=3) Once (n=2)
Preschool age: 3-5 years	Twice (n=1) Once (n=4) No formal follow-up (n=1)
School age: 5-12 years	Once (n=2) If concerned (n=3)
Adolescence: 13-18 years	If concerned (n=3) No formal follow-up (n=2)

Table 2. Frequency of screening and evaluation within the four structured developmental follow-up programs

Service referrals	Psychologists/neuropsychologists (9/10) Speech-language pathologists (9/10) Occupational therapists (7/10) Physical therapists (5/10) Developmental pediatricians (1/10) Special educators (1/10) Existing developmental programs (2/10)
Reasons for referral	General concerns identified during developmental follow-up (9/10) Feeding difficulties (6/10) Behavioral difficulties (7/10) Motor difficulties (5/10) Academic difficulties (5/10) Language difficulties (2/10) Autism (1/10) Hearing loss (1/10)

Table 3. Service referrals and the reason for referral

Domain	Screening & Evaluation Tool
Global Development	• Ages and Stages Questionnaires 3rd edition • Ages and Stages Questionnaires: Social-Emotional • Bayley Scales of Infant and Toddler Development 3rd edition
Motor skills	• Abnormal Involuntary Movement Scale • Bruininks-Oseretsky Test of Motor Proficiency 2nd edition • General Movements Assessment • Grooved Pegboard • Hammersmith Infant Neurological Examination • Movement Assessment Battery for Children 2nd edition • Movement Assessment of Infants • Peabody Developmental Motor Scales 2nd edition • Posture and Fine Motor Assessment of Infants
Behavior	• Behavior Rating Index of Executive Function 2nd edition • Behavior Rating Index of Executive Function- Preschool 2nd edition • Adaptive Behavior Assessment System 3rd edition • Behavior Assessment System for Children 3rd edition • Child Behavior Checklist • Child Teacher Report Form
Cognitive skills (intelligence, memory, and attention)	• California Verbal Learning Test – Children's Version • The Differential Ability Scales 2nd edition • Delis-Kaplan Executive Function System • Rey-Osterrieth complex Figure • Test of Everyday Attention for Children • Wide Range Assessment of Memory and Learning (Story & Design Memory) • Wechsler Intelligence Scale for Children 4th edition • Wechsler Preschool and Primary Scale of Intelligence 5th edition
Language and Communication	• Communication and Symbolic Behavior Scale Developmental Profile Infant- Toddler Checklist • MacArthur-Bates Communicative Development Inventories • Expressive Vocabulary Test 3rd edition • Peabody Picture Vocabulary Test • Preschool Language Scales 5th edition

	• Comprehensive Test of Phonological Processing-Second Edition
Social Skills/Autism	• Modified Checklist for Autism in Toddlers • Social Responsiveness Scale 2nd edition
Sensory	• Sensory Profile-Sensory Profile- Child Sensory Profile 2nd edition
Visual Motor Integration	• VMI-Beery-Buktenica Developmental Test of Visual Motor Integration
Academic Achievement	• Bracken: School Readiness Scale • Wechsler Individual Achievement Test 3rd edition
Psychosocial	• Depression, Anxiety and Stress Scale (DASS-21) • Nepsy-II
Audiology	• Behavioral audiometry • Ear specific audiometry
Parents' mental health	• Modified Perinatal Post Traumatic Stress Disorder Questionnaire • Parenting Stress Index – Short Form

Supplemental Table S1. Tools used for screening and evaluation

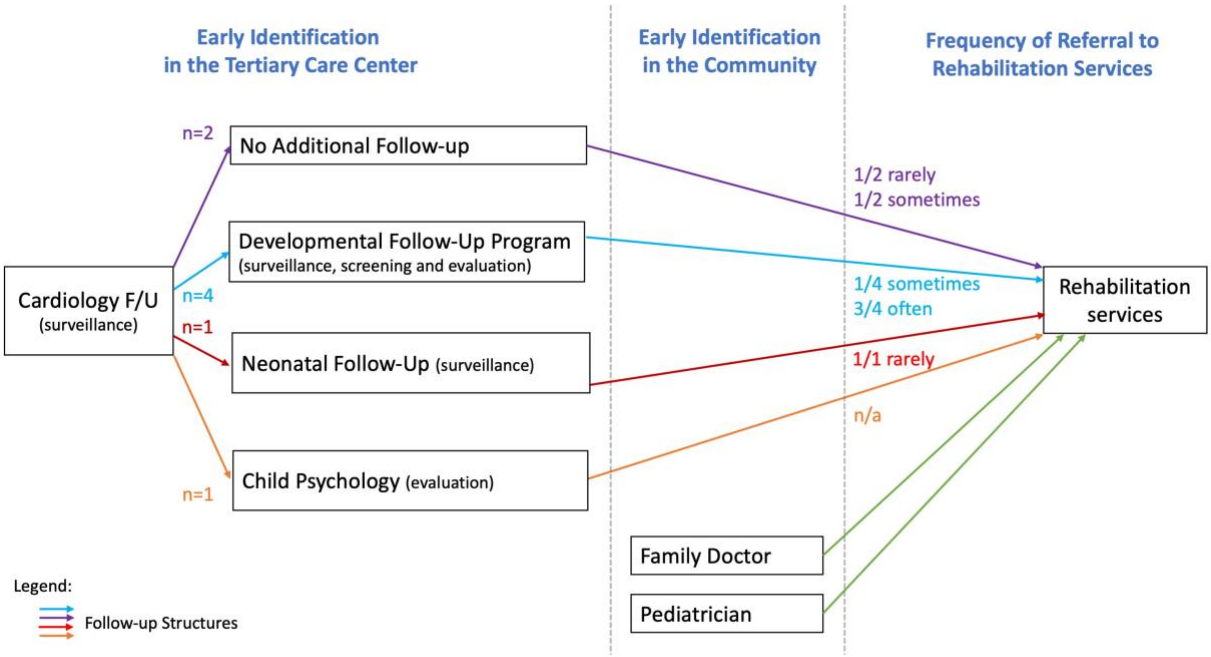

Figure 1: Developmental follow-up care practices in Canadian tertiary care centers

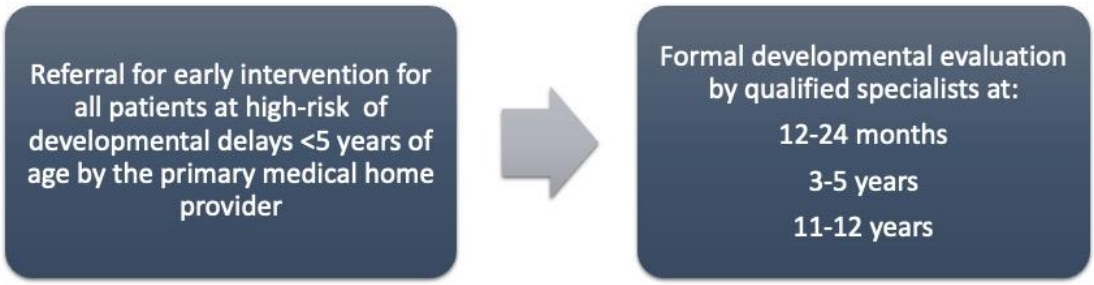

Figure 2. Summary of the AHA recommendations for the developmental follow-up of high-risk children with CHD[4]

CHAPTER 7: INTRODUCTION TO THE PARENTAL PERSPECTIVES

We have previously identified that current developmental follow-up practices in Canada are suboptimal from the point of view of healthcare professionals involved in the developmental follow-up at the institutions that perform pediatric open-heart surgery in Canada. Furthermore, financial and human resources appeared to be limiting the implementation of best practices (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). However, service users (i.e., consumers, parents, or patients) are another important group whose perspectives need to be taken into consideration when appraising current practices. Exploring the perspectives of parents of children with a congenital heart defect (CHD) as well as adolescents and young adults with a CHD is central to developing high-quality practice recommendations for the timely detection of developmental challenges in children and adolescents with a CHD.

Developmental follow-up for children with a CHD starts during infancy and extends until the adolescent transitions to adult care. Consequently, it is important to explore the perspectives of service users along their entire developmental trajectory. As a result, we recruited parents of children aged 5 to 14 years, to discuss their viewpoints with regard to the developmental follow-up of their children during infancy and childhood. In addition, youth 13 to 22 years of age were approached to discuss their experiences with the developmental follow-up received during childhood, adolescence and young adulthood. The overall objective was common to both groups and the data were collected simultaneously using an iterative process. The aim of this study was to explore the perspectives of parents of children and adolescents with CHDs and those of adolescents and young adults with CHDs with respect to developmental follow-up. The results are presented in two separate manuscripts to best describe the preoccupations of each group of service users.

This third manuscript reflects the perspectives of parents with children with CHDs and has been submitted for publication on April 19th, 2022.

Bolduc, M.-E., Rennick, J. E., Gagnon, I., Majnemer, A., & Brossard-Racine, M. (2022). Navigating the Health Care System with my Child with CHD: Parental Perspectives on the Canadian Developmental Follow-Up Practices. *Manuscript submitted for publication*.

The youth's perspective will be presented in Chapter 10.

CHAPTER 8: NAVIGATING THE HEALTHCARE SYSTEM WITH MY CHILD WITH CHD: PARENTAL PERSPECTIVES ON CANADIAN DEVELOPMENTAL FOLLOW-UP PRACTICES

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Short Title**:** Parental Perspectives on Canadian Developmental Follow-Up for CHD

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Prior presentation of study data**:**

Data has been presented as a poster at the 10th Annual Scientific Sessions of Cardiac Neurodevelopment Outcome Collaborative in November 2021.

List of Abbreviations

CHD: Congenital Heart Defect

ABSTRACT

Objective

Parents of children with a congenital heart defect (CHD) face several barriers when trying to access the services needed to promote their child's development. In fact, current developmental follow-up practices in Canada may not optimally identify developmental challenges in a timely manner and important opportunities for interventions may be lost. This study aimed to explore the perspectives of parents of children and adolescents with a CHD with respect to developmental follow-up.

Study Design

Interpretive description was used as a methodological approach for this qualitative study. Parents of children aged 5-15 years with a complex CHD living in Canada were recruited. Semi-structured interviews that aimed to explore their perspectives regarding their child's developmental follow-up were conducted.

Results

Fifteen parents of Canadian children with a CHD were recruited for this study. They expressed that the lack of systematic and responsive developmental follow-up services and limited access to resources to support their child's development placed an undue burden on their families, and, as a result, they needed to assume new roles as case managers or advocates to address these limitations. This additional burden resulted in a high level of parental stress, which, in turn, affected the parent-child relationship and siblings.

Conclusion

The limitations of the current developmental follow-up practices put undue pressure on the parents of children with complex CHD. From the perspective of these parents, a more systematic approach to developmental follow-up would allow for timely identification of challenges and would promote a more positive parent-child relationship.

BACKGROUND

Children with a complex congenital heart defect (CHD) requiring open-heart surgery in infancy are at high risk of developmental delays that may affect multiple developmental domains including motor skills, language, cognition, behavior and academic skills, which can arise at different time points during childhood and adolescence (1-3). Although developmental difficulties are often mild to moderate, their frequency remains high and they are associated with activity limitations at school, in self-care and in community participation (4). Thus, in recent years, the improvement of developmental outcomes has become a priority for both the clinical and research communities.

Improvement of developmental outcomes is dependent on a system that provides timely identification of developmental delays and subsequent referral for interventions. This is well supported in the literature on other high-risk infant populations, especially when implemented in a timely manner (5). There is also growing evidence of the effectiveness of early interventions in the CHD population (6, 7). Moreover, children and families can be offered support, resources and strategies to functionally adapt to some challenges that cannot be remediated (8). Conversely, developmental challenges that are left unattended may have long-lasting consequences on school performance, self-esteem, anxiety and social relationships (8, 9).

In an effort to optimize the developmental trajectories of children with a CHD, the American Heart Association (AHA) released a statement emphasizing the importance of systematic follow-up services for all children with a CHD (10). This statement indicates that follow-up care for high-risk children with a CHD should include surveillance (monitoring of parents' concerns over time), screening (questionnaires) and formal evaluations. Formal evaluations are recommended at 12-24 months, 3-5 years and 11-12 years for this group. However, we recently demonstrated that current developmental follow-up practices in Canada vary greatly across centers and may not optimally identify challenges in a timely manner (11). As a consequence, important opportunities for interventions to enhance outcomes may be lost.

Parents of young children with a CHD highlighted the importance of regular monitoring in the context of a mixed-methods study that explored their overall experiences and needs related to healthcare delivery (12). They also identified several barriers to accessing interventions such as a lack of awareness of available resources, and the distance and cost associated with attending appointments (12). Although

the participants mentioned the importance of monitoring, no question specifically explored developmental follow-up experiences and preferences. Therefore, it is impossible to determine if the preferences, barriers and facilitators are similar for developmental follow-up and interventions. Hence, the aim of this study is to explore the perspectives of parents of children and adolescents with a CHD with respect to the developmental follow-up of their child.

METHODS

DESIGN

This qualitative study used interpretive description as a methodological approach. Interpretive description is a flexible, non-categorical qualitative research methodology designed to develop an understanding of human experiences that recognizes each experience as constructed and contextual and is applicable to clinical contexts (13, 14).

SAMPLE AND PROCEDURE

Parents of children aged 5 to 15 years with a CHD requiring open-heart surgery before two years of age and who had received health services in Canada were approached to participate in this study. Parents whose children were born prematurely, or with genetic conditions were not eligible because these factors presumably affected the extent and type of developmental follow-up received. Participants were recruited through support groups and associations for families of children with a CHD. In addition, flyers inviting parents to participate in our study were sent to the Cardiology Divisions of selected children's hospitals across Canada. Parents interested in participating in the study contacted us by phone or email. We first confirmed their eligibility and then sent the consent form and questionnaire. Participants were purposively recruited, from the pool of interested participants, to capture users' experiences across infancy, preschool and elementary school-age years and early adolescence, to represent different geographical locations (urban and rural), and representations of children from both sexes. These criteria were selected to provide the greatest variations in our sample. Recruitment ended when no new themes were identified in two subsequent interviews.

We emailed each participating parent a questionnaire that included demographic questions and information on their child's developmental profile. Questions included the age and sex of the child with a CHD, geographical location, socioeconomic information, and developmental challenges encountered. Parents were asked to return the completed questionnaire prior to their interview. Semi-structured

interviews were conducted by videoconference or telephone (see Interview Guide, Appendix A). Interviews were conducted in French or English. Informed consent was obtained from each study participant. Ethical approval for this study was obtained from the McGill University Health Centre Research Ethics Board #2020-5921.

Analyses

Interviews were transcribed verbatim and the transcripts were analyzed using NVivo 11 software (QSR International). Data were coded using both inductive and deductive coding. The deductive codes were based on the interview guide questions. Comparative analysis was then used to find commonalities and differences in parents' experiences (15). This method allowed us to discern commonalities between participants' experiences and preferences while acknowledging individual care experiences and their complexity. New conceptualizations of the parents' perspectives on the developmental follow-up of children and adolescents with a CHD were then developed. Finally, quotes that best represented the data collected were selected and the interpretations were developed. French quotes were translated by the first author. Data collection and analysis took place concurrently in an iterative manner. This allowed for tentative interpretations to be discussed in subsequent interviews, to allow participants to expand, clarify and/or elaborate on the proposed interpretations.

Trustworthiness

Four criteria were used to ensure the trustworthiness of this study: epistemological integrity, representative credibility, analytic logic and interpretive authority (13). Various strategies were used from the design of the study to the analysis to support these criteria. First, to ensure epistemological integrity, we used a research question, data collection techniques and analysis methods that are consistent with the theoretical and philosophical underpinnings of interpretive description. Secondly, we used purposive sampling to have the highest level of variation in our sample to ensure that the selected parents were representative of the population. Thirdly, the same analytical process was used for each interview and the reasoning was made explicit throughout the manuscript. Finally, interpretations were discussed during subsequent interviews and among the research team and reviewed by a parent partner to enhance interpretive authority.

RESULTS

A total of 38 parents contacted us to participate in the study. Six were not eligible because they did not meet the inclusion criteria. Purposive sampling was performed among the remaining 32 parents. A total of fifteen parents of children with a CHD now aged 5-14 years (mean 9.4 years) participated in the study. All interviews were conducted with parents, who self-identified as mothers of children with a CHD and, in three cases, fathers were also present for part of the interview. Interviews lasted an average length of 42 minutes (range: 22-95 minutes). Participants' children were born with a wide variety of a CHD diagnoses requiring open-heart surgery and families lived in different regions and had variable socioeconomic characteristics (Table 1). All but one child with a CHD experienced challenges in one or more of the following surveyed domains: academic performance (n=10), behavior (n=9), gross motor (n=9), fine motor (n=9), cognitive (n=7) and language (n=6) skills. Although the questionnaire did not specifically ask about these difficulties, four parents reported that their child with a CHD had difficulty falling asleep or sleeping through the night and four reported sensory disturbances. These disturbances were described as oral hypersensitivity, need for sensory stimulation, or hypersensitivity affecting multiple senses. In terms of developmental follow-up, twelve of the fifteen parents had experienced screening and/or formal evaluation for their child at some point, and three had access to surveillance only.

Although parents of children with a CHD expressed gratitude and were very satisfied overall with the care their child had received for their cardiac condition, most of them voiced concerns regarding the developmental follow-up they received. Three main themes provided an overview of the perspectives of parents on their child's developmental care:

A. Perspectives on current developmental follow-up care: Limited accessibility from identification to intervention;

B. Increased parental burden: Struggling to fill the gaps in developmental follow-up;

C. Impact on the family: Seeking to establish a sense of normalcy (figure 1).

A. Perspectives on Current Developmental Care: Limited Accessibility From Identification to Intervention

1) Lack of systematic and responsive developmental follow-up services across childhood

Even if parents mentioned having access to a formal evaluation through public or private resources for their child, some participants reported that they had to request (and sometimes even insist) on being referred to those during routine appointments with their cardiologist, pediatrician or family doctor. Three parents expressed that they gained access to formal evaluation through participation in research studies. Many parents expressed concerns with the absence of systematic follow-up services for all children with a CHD at risk of developmental delays: *"Especially kids who have open-heart on bypass… I wish it was more just universal that all kids get referred into whatever the program is in their community, at least for a primary assessment. In my ideal world, that is just more standard of care as opposed to something that you have to wait for a problem to be discovered and know how to navigate the system to get there…."* Parents also voiced concerns with regard to the responsiveness of current practices to formally identify the developmental delays or challenges their child was experiencing. They expressed that sometimes they brought up concerns with healthcare professionals who preferred to wait and see if the problems would resolve by themselves before referring to another healthcare professional for further assessment or intervention: *"If you wait too long or if you wait until four years old, then it does not give much time to do therapy before your child goes to school."*.

Several parents insisted on the importance of having formal evaluations in addition to questionnaires as part of the developmental follow-up: *"I think there should be batteries of tests too, because questionnaires are one thing. It's good, but it doesn't meet all the needs. There are perhaps things that the therapist can notice that the parent does not see."* Most parents expressed that it was reassuring to know how their child was developing. Conversely, two parents, including one whose child did not experience challenges, expressed that those formal evaluations can sometimes be stressful for both the child and the parents and that adding additional hospital visits can also be overwhelming, especially in the first years of life: *"Maybe as a first-time parent, I may have opted for some of the screening, or some of these questionnaires, or the developmental tests, but knowing what I know now about development and how the range of normal is so vast… I think maybe if my child was not exactly hitting the milestones that they quote-unquote 'should be', it would again be stressful, and I don't know if that would be totally necessary."* Parents were aware and sensitive to the lack of resources in the healthcare system; however,

they believed that investing in early detection is worthwhile: *"I imagine, like anything, if you can pick up on issues early, even though there's a cost to that identification, maybe there's an easier fix at that early stage whereas if it continues to go on through life and now what burden are they putting on the healthcare system…".* In response to the limited resources and stress of evaluations, some parents suggested a tiered approach where they could start with screening questionnaires and follow-up with a formal evaluation only if the questionnaire raised concerns. One parent who was part of another research project, however, found using multiple questionnaires that often overlapped was both frustrating and time-consuming for parents.

The timing of these evaluations was also considered important. Overall, parents agreed that the frequency of administration should be higher in early childhood, with some parents suggesting developmental follow-up every 6 months, and others every year, or every two years. Although opinions varied on the exact timing and frequency of the assessments, most parents agreed that a comprehensive evaluation was needed before children start school so that the required support could be put in place: *"Once the school machine starts rolling, he's on this trajectory and so it's hard now. […] We don't really have the assessment or much support yet on how we can support him, and the more years we get down the harder it's going to be."* Some parents reflected that because their child did not go to preschool due to their higher health risks, an important safety net for early identification of challenges might have been missed. Parents also highlighted the importance of systematic developmental follow-up that continues beyond school entry.

Some participants also identified the importance of a centralized follow-up process to facilitate the circulation of information between professionals: *"Every time you have to repeat that, firstly it takes a lot of time, it takes your energy, it reminds you of bad memories."* A more centralized process could also prevent gaps in service delivery, especially at key transition points: *"They totally just give the information, they said the school should take over from there, but the school system never did any follow-up."* Furthermore, parents highlighted the importance of having a point of contact in case a new concern arose in between assessments: *"Even (sigh) someone to contact. If anything comes up over the next three years, you can contact this person, or this department and we can help you with any questions you might have or be able to direct you in the right direction. That might have been helpful as well because there's lots of times where I'd grab my heart book that I got when she was born, and I sort of flipped through for information or people to contact and there was really nobody I could contact."*

2) Supporting developmental needs: From community resources to education for parents and educators

Parents referred to intervention services were often confronted by the scarcity of resources both in the healthcare and educational systems. Some identified that receiving a formal diagnosis was an advantage to accessing resources: *"You can say, [for] better or worse, she got diagnosed with the intellectual disability, so that's why she qualified for some programs, like say Special Olympics, designed for kids or adults, or people with mental disability. […] There's no program at all for kids with normal intellectual ability but [who have] some physical delays to participate [in]."* Unfortunately, receiving a diagnosis was not helpful to all parents: *"Even after finding all the resources and having her diagnosis [of attention deficit disorder] [it] didn't get her anywhere anyway."* Participants reported that the subtleness of challenges associated with their child's CHD may contribute to the difficulties in accessing the resources: *"We squeaked until we were able to get something [resources] because they often overlook kids that [have moderate difficulties], because they aren't as bad as other kids, or they feel that other kids are in more need of, and it's the kids that are in need but just kind of on the borderline that fall through the cracks".*

The presence of waitlists for resources was also problematic, especially when referrals were not made in a timely manner: *"The thing is everything is waitlist and no services that you get right away. […] So, it either depends on [if] you wait, […] or you just simply go private."* For families who lived further from large urban centers, the limited accessibility to professionals was even more striking. One parent who lived in a rural area suggested online consultations as a solution to this problem.

Within the school system, the supports children had access to were also limited and varied from year to year depending on the teacher's background: *"We are really lucky we had an amazing grade four teacher who was very understanding, and he did a psych major within his teaching, so he had a really good outlook on things, [a] unique way and used tools that he had in his toolbox that I had never seen a teacher use before which was awesome."* A mother suggested that to address the limited supports available within the school system, parents could provide the teachers with educative materials on CHDs, its impact on development and how to best support their child's needs: *"If there was [a] little tool kit ready when you start school […] and even just hand out to the teacher like 'these are some of the strategies that we find might help'."*

Some parents had access to resources they appreciated. Parents from two provinces (British Columbia and New Brunswick) mentioned they could self-refer to a developmental follow-up program or rehabilitation services for the first 3 to 5 years of life depending on the province. Other parents had access to community services to support their child development. For example, one parent had a weekly inclusive physical literacy class offered by students from various backgrounds such as kinesiology and physiotherapy, through a program affiliated with their local college. Community services such as swim therapy were also found to be beneficial by parents of children with a CHD.

Finally, parents suggested that providing them with the tools to best support their child's development would be valuable: *"Even if it's just sometimes […] to give suggestions to parents. […] There are parents who don't know as much how to stimulate development. It's [the evaluation] an opportunity to give parents strategies on how to stimulate development. […] It could be beneficial to parents too."*

B. Increased Parental Burden: Struggling to Fill the Gaps in Developmental Follow-Up

Many participants expressed that the limitations in current developmental follow-up practices resulted in an increased parental burden as they had to fill these gaps if they wanted their child to receive the care they needed. Since developmental follow-up mostly took the form of surveillance, parents felt that the responsibility of identifying delays rested on their shoulders. Some parents did not feel competent to identify these difficulties accurately: *"So it's me who sees it [what needs to be worked on] and then I see him developing. I see his problems. I know all the little things he has to work on but it's not easy either because I'm a carpenter, so I do my best."* Identifying delays seemed especially difficult for first-time parents who did not have a point of comparison. Some parents also found it difficult to identify delays in their child with a CHD: *"When really you are not holding the bar as high as you would hold it for someone who hadn't gone through so much."* Conversely, other parents felt that they were constantly keeping a critical eye on their child's development to ensure they could report any observed concerns to their healthcare practitioner.

Given the limited access to resources, parents often became advocates for their child to acquire the services they needed: *"I've definitely been her number one advocate her entire life. […] With the whole school system, to be honest, if I had not been the squeaky wheel, we would not have had the assistance we have now which is vital to her education."* Some parents hired professionals to provide additional support for their child when public resources were not available, but this option was not available to all

parents because of the associated costs: *"I found someone in the private sector at the beginning. She came to our house a few times. Then, of course, the insurance does not cover much, so she just came a few times, and she gave some suggestions…"* Parents also expressed that they are often worried that they perhaps could do more to support the needs of their child and they didn't always feel equipped to provide the required support: *"We get the email from the school about how he's not doing his work. I'm like 'what am I supposed to do?'… I'm not in education. […]. The school is struggling, we are struggling, and again it just keeps rolling on."*

Parents raised ethical concerns related to the need to advocate for their child's care. One parent identified that when care depended on parents advocating for their child, it can lead to inequalities: *"[We] shouldn't only be getting services when parents advocate. Because lots of people are busy. I think the children deserve better follow-up even if they don't have parents that have them under magnifying glasses. It should be equal for everybody."*

The lack of systematic and centralized follow-up systems meant that parents had to navigate the healthcare system to find the resources they needed to support their child. Parents felt they had to take on the roles of case manager to coordinate their child's appointments and ensure information was communicated to all professionals: *"We're lucky but I still find there is a lot that I had to do... it was like a full-time job, all the time. Being on the phone and going to appointments. […] I felt that a lot depended on me. I was the one who had to go and look for all the things [resources]."* Some mothers had to stop working given the amount of time required for these additional responsibilities: *"That's why I choose not to work […], so I can have a little bit more time to be a case manager for my little one. I know, in a way, I'm privileged to do that. […] It's a lot for families. It's really a lot. It sucks your energy, I tell you. Emotionally, it drains you."*

C. Impact on the Family: Seeking to Establish a Sense of Normalcy

Parents of children with a CHD expressed that they experienced a high level of stress. The stress was initially caused by their child's heart condition, the diagnosis, perinatal circumstances and surgery: *"I'm already on alarm myself since the surgery, his heart condition was totally unexpected and we weren't aware of it, so like I'm always a mom on edge."* However, after this period, most parents continued to feel anxious about their child's development: *"Still today, as school is coming up, I have anxiety… how*

will third grade be? […] At every stage, every year of life, it's all the time-, you think about it whether you want it or not."

The interviewees also reported an overall sense of being overwhelmed by the multiple roles and the added responsibilities that they must take on: *"The time and energy that it takes... If someone could just decrease the stress for the parents."* More specifically, some parents described the role of identifying observed delays and reporting them to the healthcare professionals as an important source of stress: *"He was my first child, he had a major heart condition, I felt like such a first-time mom… I'm going to cry… who didn't have a lot of other moms going through the same thing. So it's really hard to know what is normal, and what is not normal."*

Some parents revealed how the diagnosis of CHD affected the entire family. They expressed that the parent-child relationship was affected because they did not have the same opportunity to spend quality time with their child as a baby: *"You just don't have a normal time with your baby so that is important for the parent too, for the mental health of the parent and the baby. […] You want to cherish the moments with your child and sometimes the anxiety takes over on it… it's something that I said to myself 'I want to enjoy every second' because we almost lost our child more than once. You want to enjoy every minute you have with him."* A few parents expressed that their role in the identification of delays conflicted with the role they should play as a parent: *"That's also your role as a mom, right? To just be so proud, and happy about where they've been and where they are now."*

One parent expressed how having a child who needs critical care at birth negatively impacted the sense of empowerment as a parent: *"You feel completely powerless, no control and you want to do something for your child."* Some events also affected the parents' sense of self-efficacy: *"She failed to thrive at the beginning. She wasn't putting weight on and I was getting made to feel like I was a bad mom because she wasn't putting weight on."* One parent expressed that it would be important to treat them as normal parents despite the circumstances: *"My son was our first child, so they forgot the normal things you need to do with a baby and the normal steps. Your child is so sick that at first it is like survival, but they must not forget that you are going home anyway with a baby, you have to take care of this baby. I need the basics. The other parents, they showed them how to wash their babies before they go home. We haven't been shown anything I knew how to give her injections and gavage, but all the normal things were forgotten. […] I find that there is this side there that they should not forget also, the human being."*

One parent expressed that not having the adequate resources in place can impact the child's mental health. When referring to the feedback received at school they said: *"Everyday, grade one, sad faces, sad faces stamped on her work. Sad faces. Good self-esteem there people. […] Don't tell me she's a bad student. She's having a hard time learning."* The impact of the CHD was not only felt by parents and children with a CHD. A parent also described how the disruptions can also have an impact on siblings: *"[It] really takes a toll on my other kids. It's really difficult. That's why my older daughter developed depression. The one kid can affect the whole family."*

A number of parents expressed how important social support is to address some of these needs: *"We were very fortunate that as soon as we got the diagnosis that [Child] had his heart issue, they connected us to [another family]. Obviously, everybody's experience is unique, but I do feel that's […] important, it's almost like having a pillow for landing. Just to know you can talk to some people who had similar experiences…"*

DISCUSSION

Participants in this study identified the importance of systematic, accessible, and responsive developmental follow-up services, where information on the child and available resources is centralized. Without such a system, the burden of identifying delays and adequately supporting the needs of children with a CHD falls on the shoulders of the parents, increasing their stress and ultimately impacting the entire family. Indeed, as the healthcare focus for children with a CHD shifts from survival to improving developmental outcomes, it is important to ensure that the family is considered when defining best practices. Family-centered care is a complex model that positions families as the experts of their needs. It includes sharing information, collaborating in care, and empowering parents (16, 17). The importance of this approach has been recognized by various institutions and shown to result in improved patient safety and health outcomes (18).

Systematic developmental follow-up is required to identify challenges in a timely manner so that the resources can be put in place to support development, thus avoiding gaps in continuity of care. Although the AHA (10) recommends formal evaluation at three timepoints during childhood (12-24 months, 3-5 years and 11-12 years) and early intervention for all children with a CHD requiring open-heart surgery, some parents suggested a tiered approach in which formal evaluation would be used only when screening results indicate potential developmental delays rather than systematic formal evaluations at

all key time points. The Ages & Stages Questionnaires (ASQ), Third Edition (19) could be a clinically useful tool to screen children with a CHD up to 5.5 years of age for developmental delays given its sensitivity, specificity and predictive value (20). In fact, this approach has been successfully implemented in Australia (21). This could present a cost-effective manner to ensure that challenges are identified early before they have long-term consequences on the child. This approach may also be less stressful and burdensome for the parents of children who experience fewer challenges and for youth with a CHD (22). To our knowledge, there is no equivalent for the ASQ for older children. However, the Vineland Adaptive Behavior Scales, third edition (23) have frequently been used in research studies that included children with a CHD and can be used up to 21 years of age (24, 25). However, this tool uses an interview format to collect information from the parents and thus is more resource intensive.

Offering comprehensive developmental follow-up that includes surveillance of all domains at risk of being affected is also essential for the timely identification of delays. Some parents in this study identified sleep and sensory regulations as challenging for the child and reported that they did not receive adequate follow-up for these difficulties. These two domains are not included in the AHA (10) recommendations and have not been well studied. Nevertheless, recent recommendations by the Cardiac Neurodevelopmental Outcome Collaborative suggest using the BEARS screening tool (26) to assess sleep, but no assessment tool has been recommended for sensory function (27). Future studies are needed to better identify the prevalence of these difficulties in children with a CHD and to determine how they can best be assessed. Finally, parents were also concerned with the limited availability of resources to stimulate their child's development or interventions for identified difficulties, both in the healthcare and school systems. This need for enhanced support throughout childhood and adolescence has also been reported by youth with a CHD (22). Enhancing the accessibility of resources within the school system needs to be formally examined in future studies. Nevertheless, the parents have identified various strategies that rely on education, community resources, telehealth and other technologies that could be put in place to support their child's needs.

The participants in this study reported a heavy burden related to managing the care of their child with a CHD. Canadian parents expressed that they had to assume new roles such as case managers, administrators, or advocates, as a direct consequence of the current gaps in developmental follow-up practices. This is in line with a previous Swiss study in which parents reported feeling exhausted from additional responsibilities with regard to the neuromotor development of their infant with a CHD (28).

The increased burden resulted in increased levels of stress for participants in our study. This could explain, in part, the increased level of parental stress, anxiety and depression reported in previous studies (29, 30). Studies have shown that parental stress is associated with the child's cognitive ability and behavior (31-33). Hence, it is essential that screening for parental mental health and access to psychological and social supports for their child be included as part of family-centred care. Those supports should include education on parental self-care and promotion of child development and should facilitate social supports (34). Furthermore, the lack of quality time with their child and a decreased sense empowerment and self-efficacy was described as having an impact on the parent-child relationship by study participants. This could relate to changes in parenting styles and decreased attachment between the mother and child (35-37). Finally, welcoming a child with a CHD may be associated with changes in family functioning (38, 39). A recent systematic review reported psychosocial well-being to be negatively impacted in 40% of siblings of children with a CHD (40). Thus, assessment and support for siblings also need to be considered.

The findings of this study support the post-intensive care syndrome – Pediatrics (PICS-p) conceptual model (41). This model captures the relationship between the child's developmental outcome and the family's psychosocial well-being described in this study. Hence, this framework may prove to be helpful in developing a comprehensive approach to developmental care for children with CHD and their families in the future.

This study presents some limitations. Our sample consisted almost exclusively of mothers and may not represent the views of fathers of children with a CHD. Nevertheless, the mothers often described the experience they had as parents or as a family and some fathers were present during a portion of the interviews. In addition, despite our far-reaching recruitment strategy, we could not enroll participants from all ten Canadian provinces; therefore, potential gaps and strategies that exist in provinces from which we did not have participants may have been missed.

CONCLUSION

The limitations of current developmental follow-up practices put undue stress and burden on the parents of children with a complex CHD. From the perspective of parents, a universal and systematic approach to developmental follow-up would allow for timely identification of challenges, enable initiation of interventions and supports to promote the child's development and promote more positive

parent-child relationships. Thus, a family-centered approach to follow-up care is warranted for children with CHD.

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Child's primary congenital heart diagnosis*	5 Single ventricle physiologies • hypoplastic left heart syndrome • double outlet right ventricle 10 Two ventricle physiologies • bicuspid aortic valve • ventricular septal defect • transposition of the great arteries • tetralogy of Fallot • arch interruption type A • severely dysplastic mitral valve • truncus arteriosus
Province of residence	2 Alberta 7 British Columbia 1 New Brunswick 5 Quebec
Geographical regions	8 Rural (≥50 km from urban centre) 4 Suburban (<50 km from urban centre) 3 Urban (population > 100,000)
Mother's highest level of education completed	0 High school completed 8 CEGEP, College certification, or technical program 2 University graduation or standard 4-year college 5 Graduate school (graduate degree)
Father's highest level of education completed	2 High school completed 9 CEGEP, College certification, or technical program 1 University graduation or standard 4-year college 3 Graduate school (graduate degree)
Family income	1 $20 000$ to $39 999 2 $40 000$ to $59 999 1 $80 000$ to $99 999 8 Above $100 000 3 Prefer not to answer

Table 1: Cardiac diagnoses and family demographics

*as reported by parents

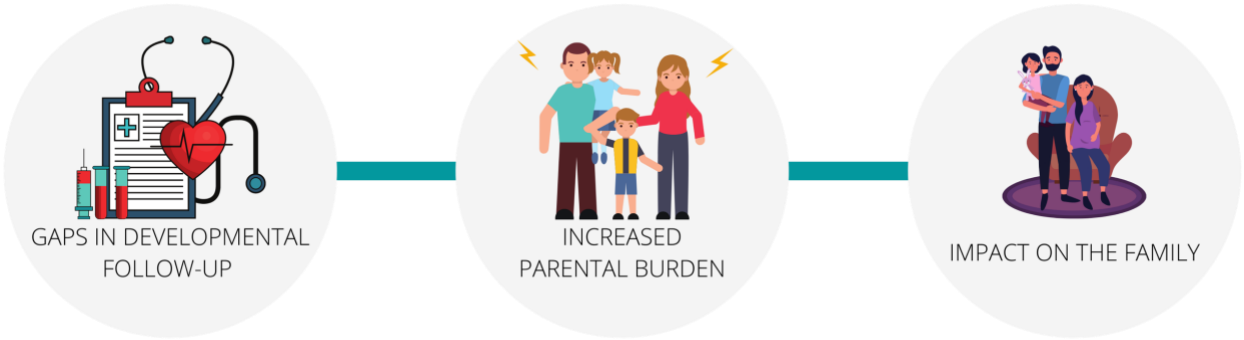
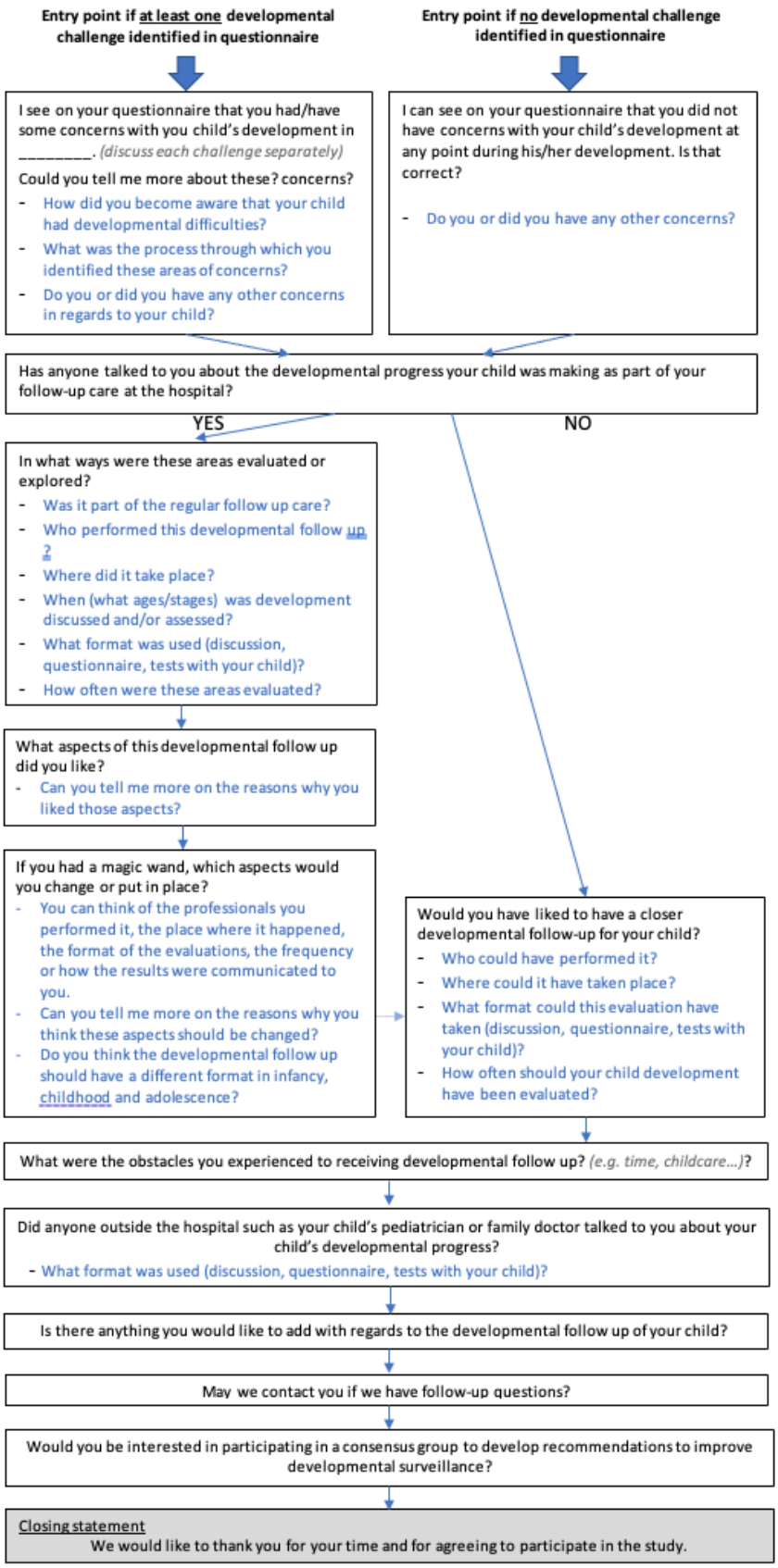


Figure 1. Impact of the current limitations in developmental follow-up practices on families of children with CHD

APPENDIX A: INTERVIEW GUIDE



CHAPTER 9: INTRODUCTION TO THE YOUTHS' PERSPECTIVES

In the previous manuscript, we explored the perspectives of parents of children with a congenital heart defect (CHD) with regard to the developmental follow-up of their child from birth to 14 years of age. Parents clearly identified gaps in current practices. Those gaps resulted in a limited ability of the current Canadian healthcare and school systems to identify developmental delays and provide the resources to optimize the child's development and functioning.

In this fourth manuscript, we present the viewpoints of youth (13-22 years of age) with a CHD with regard to the developmental follow-up they received as children and adolescents. The interviews with youth and parents were conducted concurrently, using a similar interview guide. Nevertheless, the topics and concerns raised by youth with a CHD were very different from what was expressed by the parents. They reflected on a different period of their life and could articulate personal challenges that had emerged during adolescence, a critical phase of development, and on their transition to early adulthood. It was, therefore, impossible to do justice to both perspectives within the constraints of a single manuscript.

The specific objective of this fourth manuscript was to explore the perspectives of adolescents and young adults with CHDs with respect to developmental follow-up.

The manuscript on the youth perspectives has been submitted for publication on February 8, 2022 and is currently under review.

Bolduc, M.-E., Rennick, J. E., Gagnon, I., Brossard-Racine, M., & Majnemer, A. (2022). Identifying developmental challenges of youth with congenital heart defects: A patient-oriented perspective. *Manuscript submitted for publication*.

CHAPTER 10: IDENTIFYING DEVELOPMENTAL CHALLENGES OF YOUTH WITH CONGENITAL HEART DEFECTS: A PATIENT-ORIENTED PERSPECTIVE

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Short title:

Patient perspectives on identifying developmental challenges

4971 Words

ABSTRACT

Introduction

Youth with congenital heart disease (CHD) are at high risk for a range of developmental impairments that become evident at different times across childhood and adolescence. This study aimed to explore perspectives of youth with CHD with respect to their developmental follow-up across childhood.

Methods

Interpretive description was used as a methodological approach for this qualitative study. Youth aged 12-22 years with a CHD requiring open-heart surgery before two years of age and who had received health services in Canada since birth were enrolled.

Results

Ten youths with CHD, two males and eight females, aged 13-22 years (mean 19.8) participated in this study. With higher social and academic demands as well as increased level of autonomy associated with older age, some youth faced new challenges that they had not encountered as children. Youth with CHD identified four aspects of the continuum of care as needing to be changed to better respond to their needs. First, the format of developmental follow-up needs to be adapted to their unique challenges. Second, resources must be more easily accessible throughout childhood and adolescence. Third, planning for transition to adult care is essential to ensure continuity of services. Finally, they identified that the school system is an essential component of the continuum of care.

Conclusion

Adolescents and young adults with CHD are at high risk of developing physical, academic and psychosocial challenges, however, timely identification of challenges does not appear to be optimal across domains and transition points, from the perspective of the youth themselves. Youth with CHD reported not having the resources and supports they required to optimize their functioning. Our findings suggest that several approaches could be adopted to enhance identification and outcomes to address the limitations of current Canadian practices.

INTRODUCTION

Congenital heart defects (CHD) are Canada's leading birth defect, with an incidence of 1.2% of total births per year.[1] With advances in medical and surgical management, survival rates of infants with CHD have now surpassed 95%.[2] However, a proliferation of outcome studies in children and adolescents with complex CHD, especially in those who underwent open-heart surgery, demonstrate that they are at high risk of developmental delays. These delays can affect different domains including motor skills, cognition, behavior, language and social skills and psychosocial health and can become evident at different times across childhood and adolescence.[3-5]

In 2012, the American Heart Association (AHA) published the first scientific statement focused on care practices to optimize the developmental outcome of children with CHD.[6] This statement recommends ongoing surveillance and systematic evaluation at key time points for all children with CHD (up to 12 years old) who fall under the high risk for developmental delay category, including those who underwent early open-heart surgery. For children over 12 years of age, the healthcare providers are to decide what follow-up plan should be put in place.

Practice guidelines, such as the AHA statement, play a key role in translating scientific knowledge into changes in practice.[7] However, recent literature and knowledge mobilization approaches recognize the importance of user involvement (stakeholder engagement) in the development of such clinical guidelines or practice recommendations.[8-10] Involving the user in the development of these guidelines can result in a greater ability to address their concerns and needs, and to implement them with consideration of existing barriers, resulting in overall better care.[9] Therefore, studies are needed to better delineate the unmet needs related to the developmental follow-up in this population.

No study to date has explored the experiences and preferences of adolescents and young adults with CHD with regard to their developmental follow-up. This information is essential to developing high quality practice recommendations for the timely detection of developmental challenges in children and adolescents with CHD. Therefore, the aim of this study was to explore the perspectives of youth with CHD with respect to their developmental follow-up across childhood.

METHODS

STUDY DESIGN

Interpretive description was used as a methodological approach for this qualitative study. Interpretive description is a non-categorical qualitative research methodology that was developed to address a need for a qualitative research methodology that can be directly applied to improve clinical practice.[11, 12] It is designed to provide a comprehensive understanding of the inherent complexity of subjective experiences. Interpretive description aligns with a constructivist paradigm, where it is recognized that knowledge is constructed and is context dependent. It recognizes the presence of human commonalities as well as individual variations.

SAMPLING AND RECRUITMENT

Youths aged 12-22 years with CHD that required open-heart surgery before two years of age and who had received health services in Canada since birth were eligible to participate in the study. Youths who were born premature, or with genetic conditions were excluded because their developmental follow-up may have been different due to the associated condition(s). Participants were purposively recruited to represent different age groups, genders, geographical locations (urban and rural), and developmental challenges. These criteria were selected to provide the greatest variation in our sample. As data collection progressed, theoretical sampling was used to recruit more participants from the different categories described above, based on new response patterns that emerged, or a need to further explore particular perspectives.

Participants were recruited through support groups and associations for youths with CHD and their families. We also contacted youths who had participated in previous research studies conducted by our group if they had agreed to be contacted for future research projects. Finally, electronic flyers were distributed to healthcare providers practicing in hospitals that offered pediatric open-heart surgery across Canada. Institutional Research Board approval was received for this study and informed consent were obtained from each participant and from the parents of participants under 18 years of age. In addition, assent was obtained for all participants under 18 years of age.

SOURCES OF DATA

Questionnaire: A questionnaire was used to gather individual and demographic information. Participants <18 years were encouraged to complete the questionnaire with their parents. Data from this

questionnaire was used to assist in theoretical sampling and description of sample characteristics. Questions included age and gender of the patient with CHD, geographical location (first three digits of postal code), and socioeconomic information (parental level of education, family income). In addition, a question asked about the type of developmental challenges encountered at different ages. The questionnaire was sent to each participant via email prior to the interview.

Individual interviews: The interviews were conducted by the first author and were designed to last approximately 45 minutes. The interview guide was developed based on existing literature and with the assistance of a parent partner and was continually reviewed during data collection to address new concepts that emerged from the data and to further clarify any emerging questions. Youth with CHD could be accompanied by their parents for the interview if desired. Participants were given the option to participate in a phone interview or via internet using platforms such as Teams or Zoom. Participants also had the choice to do the interview in either French or English. Interviews were audio-recorded.

Qualitative Analysis

Qualitative data obtained during the interviews was transcribed verbatim and analyzed using NVivo 11 software (QSR International) to categorize and analyze data, as well as to provide illustrative quotes. Analysis, using both inductive and deductive coding, and data collection took place concurrently in an iterative manner. This allowed tentative interpretations to be discussed in subsequent interviews in order to allow participants to expand, clarify and/or elaborate on the proposed interpretations. Comparative analysis, derived from grounded theory, was used to find commonalities and differences in participant experiences[13]. The final step consisted of developing new conceptualizations of the phenomenon. Finally, quotes were selected to best represent the data collected and interpretations.

Rigor

This study was guided by four general principles: epistemological integrity, representative credibility, analytic logic and interpretive authority.[11] Epistemological integrity was ensured by exploring the researcher's orientation and making sure that data collection and analysis were consistent with the theoretical and philosophical underpinnings of interpretive description. Representative credibility was supported by sampling participants that are representative of the population. Thick description was also used to allow others to examine the contextual similarity. Analytic logic, a constant inductive reasoning and decision-making process, was ensured by applying the same analytical process to each interview.

Finally, interpretive authority, that the interpretations are trustworthy, was supported by making sure that the interpretations were as bias-free as possible. This was considered in the design of the interview guides and was confirmed by verifying some of the data collected and interpretations during the interviews through an iterative process. Exploiting theoretical underpinnings are essential in interpretive description. The first author's theoretical orientations are influenced by both the philosophical underpinnings of their profession and personal allegiances. As an occupational therapist, they deeply value rehabilitation and believe that it can make a significant contribution to optimize outcome in various populations and especially children.

RESULTS

Ten youths with CHD, two males and eight females, aged 13-22 (median 19.8) participated in this study. Four youths were recruited through support groups and associations, five were recruited from our database of previous participants using purposive sampling and one participant was recruited through snowball sampling. All interviews were conducted by the first author using an interview guide. Three youths chose to have a parent present during the interview. Eight of 10 participants had at least one parent who completed post-secondary education and the two others had at least one parent who completed high school. The detailed demographic information and primary cardiac diagnoses of interviewed participants are presented in Table 1.

OUTCOME: DEVELOPMENTAL CHALLENGES EXPERIENCED BY YOUTHS WITH CHD

With higher social and academic demands as well as increased level of autonomy associated with older age, some youths faced new challenges that they had not encountered as children. In their questionnaires, three youths with CHD identified that they had experienced or were still experiencing difficulties in one developmental domain while four had or continued to have difficulties affecting multiple domains. These participants experienced a wide spectrum of challenges both in terms of the domains affected and the extent of the difficulties. The other three participants did not report any experience of developmental challenges on their questionnaire. However, two of these participants identified limitations during the interview, one reported difficulties in one domain while the other reported some minor difficulties in various domains.

Motor activities and sports

The physical limitations or motor impairments reported by youth with CHD had an important impact on their daily lives. Six participants indicated having gross and/or fine motor difficulties while growing up. Examples included: riding a bicycle, handwriting, or performing other fine motor tasks such as using buttons and zippers.

« It was mainly my writing skills, like writing by hand, that was difficult. I had some pretty incomprehensible handwriting. »

Limitations in cardiovascular function, described as high levels of exertion during physical efforts, also had a significant impact on the participants' daily lives. Two of the participants who had not identified limitations on the questionnaire reported difficulties in this domain.

« I was doing contemporary dance, ballet, it's not like hip hop, but still requires an effort. […] Often, while dancing, it was really harder in terms of effort, especially cardio. I've never played soccer in my life; I couldn't do that. I stayed in figure skating, dancing, just as little effort as possible. »

Physical education classes were commonly described as being challenging and were a source of anxiety for some participants. The focus on team sports and the competitive atmosphere of traditional physical education classes further contributed to the negative experiences. One participant described how they felt about these classes: *«That's why I used to hate it [baseball]. I used to play it in gym class in middle school and I used to hate it because I can never hit the ball with the bat, and everybody is staring at me.»*

Another participant expressed:

«I saw a lot of really competitive people, they run everywhere ... I can run, but after five minutes of effort, you know, I get out of breath, and I see that I was slowing down, and I was always the last. I would put myself down, I said to myself " I'm not good enough", maybe "it's too hard for me, it's because of my defect", […] it's not my fault that I have this, but I always try to imagine what it would [be] like if I had a normal heart… would I be able to run with my friends? If I would not have to say " 'scuse me I have to sit down, catch my breath," that was hard for me. Sports and all that.»

Conversely, some participants who experienced fewer limitations or impairments expressed frustration with not being allowed to participate in sports as much as they would have liked because of limitations imposed by their cardiologist or by their physical education teacher, who, from their perspective, may

have been overprotective. These participants had not reported difficulties in motor activities on their questionnaire.

«Of course, it was not recommended to make physical efforts, but despite everything, at 10 years old, I liked skipping rope with my friends, so I cheated a little without pushing my luck too much. […] It went well [physical education], I had a teacher that was very accommodating, she knew what the stakes were. […] But I think she was a little scared in that regard. There were things that she didn't want me to do but that, I was like: "Yes, I can do this ".»

Academic performance

Academic difficulties were present for most of the participants in this study. Four participants identified academic difficulties in the questionnaire and one additional participant expressed during the interview that a lot of effort was required as well as the need for school resources and parental support to succeed. A sixth participant identified that the help of a tutor was needed in high school for a specific subject. Overall, three participants in this study mentioned having an individualized education program, indicating that challenges were identified and that measures to support these children or adolescents were required.

« I had, well I still have attention problems but in high school it was really more because math and science were really difficult so at school I couldn't concentrate much, and I needed [accommodations]…»

Behavior and mental health

Seven of the ten participants indicated in the questionnaire that they experienced challenges with social relationships, behavior or mental health. In terms of relationships, some participants described themselves as not very sociable. Stress and anxiety were also common among the interviewed youth. These symptoms sometimes took the form of stress linked to social situations or related to discussions about their cardiac condition. Although their behavior and mental health challenges interfered with their daily functioning, most participants had not formally been followed-up by health professionals with regard to these difficulties: *«I think it started out more around school years 10, 11, 12 and then still today like I just talked to you, and I get really stressed out. Of course, I have anxiety, but I didn't go to a doctor for that, but I can tell I have anxiety. »*

One participant was diagnosed with general anxiety in adolescence, following a series of anxiety attacks and a suicidal attempt. Two participants linked their current psychosocial difficulties to the medical

events they went through as children and introduced the concept of physical and psychological trauma that had life-long consequences on their well-being: *«It really left a mark in my brain, it's there and then it's going to stay there. »*

The impact of these challenges on self-esteem was also brought up by two participants: *«I can run, but after five minutes of effort, I get out of breath, and I see that I was slowing down, and I was always the last. I would put myself down, I said to myself " I'm not good enough", maybe "it's too hard for me, it's because of my defect."»*

Continuum of Care: From Healthcare to Education

Need for a format of developmental follow-up that addresses the youth's unique needs

Most of the ten study participants experienced developmental surveillance during routine cardiology appointments or visits to the pediatrician/family doctor. This monitoring process took the form of general questions about different parts of their life such as school and social relationships. In addition, some participants remembered completing standardized evaluations with a psychologist at the hospital. For most of these participants, access to more formal evaluations was initiated by their parents based on parental concern of difficulties that were significantly affecting their child's daily function. Participants enumerated several benefits of formal developmental evaluations. They stated that it helped them better understand their limitations and identified areas of difficulty so that adequate supports could be put in place.

«It [the formal evaluations] really helped me realize that I had a lot of gaps in a lot of areas. […] Even today, it's really the mathematical processes and numbers [that are difficult] for me. I can't easily count money and stuff like that. »

Another participant explained: *«The psychological analysis helped me to better understand how my brain works. »*

With regards to the disadvantages of developmental surveillance, some participants expressed that they may not have felt comfortable bringing up some of their difficulties when speaking with their doctor and that some challenges may have been better captured through self-report questionnaires. In addition, the majority of participants did not consider their academic difficulties to be related to their cardiac condition. Therefore, they would not have identified their academic, social or mental health difficulties to their cardiologist. When asked about whether they had discussed their anxiety with healthcare

professionals at the hospital, one participant answered: *«I never really thought about it because you know, anxiety is more, it's mental health […]. Cardiologist, mental health… in my head they don't go together. »*

Similarly, another participant expressed: *«I might not have seen the connection ... you know, I think I wouldn't mind talking about it with someone, but it might not have been the first person I would have thought to talk about, either to my cardiologist or my family doctor. These are not really people that I would associate with the academic or social difficulties. »*

Nevertheless, formal evaluations were also perceived to have limitations because they are sometimes stressful and require taking time off school/work. As a result, some participants, including the two participants who did not experience limitations, suggested starting with screening self-report questionnaires and follow up with formal evaluations only if the questionnaire raised concerns: *«I think that starting with a questionnaire as a first step and, then maybe a test if there is something special on the questionnaire. »*

Some participants also suggested key timepoints when systematic follow-up would be most important. Academic transitions such as the beginning of kindergarten, high school and college were identified as being periods of change where new difficulties may arise. Others suggested more frequent follow-up such as evaluations every two years, or along with cardiology follow-up visits between five and 12 years of age. Yearly questionnaires to identify academic difficulties were also proposed so that the appropriate supports could be provided in a timely manner. There was no difference in the suggested timing for follow-up when comparing the youth who experienced challenges and those who did not.

Need for intervention services, education, and ongoing support

For many participants, even when developmental concerns were identified, the intervention services were not readily available within the healthcare system. One participant, who had not received services for anxiety expressed that the process to find a provider for intervention services was complicated and it would have been helpful to have a centralized service point to access services through the hospital.

In addition, some participants highlighted the need for education about the impact of their cardiac condition on their daily life. One participant described a difficult situation:

«I remember when I was younger, I was doing gymnastics and I think I was 10, and I was going to do the competition but I wasn't allowed, my doctor decided against, and I didn't understand and it was really

upsetting, it was a really upsetting time in my life, and it was just hard for me to understand why and how come, and I wish that he could have sat me down and explained through my tests or through other people's tests or whatever it was to just help me understand why I had to take these sorts of precautions for myself, because it was just so confusing.»

In addition to receiving services to address their developmental and activity limitations, participants also expressed the need to access healthcare professionals who were knowledgeable of their cardiac condition, to respond to questions about health issues that may arise outside their developmental follow-up. One participant gave the example of not knowing if they could work in a camp with children during the COVID-19 pandemic and having difficulty getting an answer to this question.

Transfer of care: A break in the trusting relationship

Six participants were over the age of 18 and had transitioned to adult care. Transitioning from pediatric to adult care was a period of great concern for many of the older participants. In some instances, both the pediatrician and the pediatric cardiologist who had been following them since birth changed. This change impacted their level of trust in their treating healthcare professionals, which was further amplified by receiving contradictory information at times.

«…the doctor [PEDIATRIC CARDIOLOGIST] told me all the time "even if you have heart disease, it's good that you do physical exercise, you have to play sports". It is true that I am not very sporty by nature so that I did not do that much either but I know that it is good… but then returned to the [ADULT HOSPITAL], my cardiologist told me that I should not exert myself when I was training for my physical education class at [school], I must not be training, it's difficult for my heart. Who do I believe? Both are qualified, both are cardiologists… you know? I'd be more inclined to believe [my pediatric cardiologist] because we've been 18 years together… but right now, she's my new cardiologist, so I have to believe her too ...»

Some participants expressed that the continuity in the care they received was reassuring: *«It's not yet sure how it's going to be when I transition to adult care, compared to pediatric care, but our […] pediatrician will continue to be my doctor even though I'm 18, so that should help.»* Conversely, some participants described the transfer of care to be chaotic at the organizational level. The lack of communication between the institutions delayed the transfer of documents, for which the patient /family needed to keep calling, causing undue stress on the participant and their family.

The School System: An Essential Component of the Continuum of Care

Youth with CHD described the education system as an integral part of the continuum of care. In the first few years of their academic journey, teachers were sometimes the first to identify fine motor delays such as handwriting difficulties. Furthermore, many of the challenges that arose later in childhood and adolescence, such as limited attention span or organizational skills, anxiety or problems with socialization impacted their school performance.

Two of the three participants with an individualized education program felt that the support provided through the school was not adequate. One participant felt that the school failed to provide all the supports required in her individualized education program, while another expressed that their teachers were very understanding and supportive but that they sometimes lacked the resources to optimally support their learning. *«Because they were like, "I can't help you with everything… I can take some homework away and, yes, I can give you more time in your exams" but that wasn't enough. »* Another participant expressed the significant impact of this lack of support on her parent's perception of her efforts at school: *«They made daddy think I was not even trying. »*

In addition, one participant who was struggling during physical education classes expressed that the solution found by their teacher did not have the expected effect: *«They made it easier for me, so you know, they said like "your grades at school, I'm going to grade you easy", but you know, I thought it was like, you know, if you want to give me a grade that I deserve, give it to me, I don't want to make it easier to get a better grade because of my heart condition. So, you thought it was a little …, they put me down a little bit, so I found it a little harder but I'm still trying my best...»*

Some participants suggested that teachers should be provided with training on how to better support them and on the long-lasting impact of physical and emotional trauma of their childhood experiences.

DISCUSSION

Systematic developmental follow-up for high-risk populations is essential to optimize the outcomes of children and adolescents with CHD. It allows for the identification of evolving challenges and helps determine the resources and supports required. The results of this study demonstrate that with increasing social and academic demands over the course of childhood, along with greater expectations for autonomy, many adolescents and young adults with CHD face challenges they may not have

encountered as children. In addition, current follow-up practices may fail to identify these new challenges they experience in a timely manner.

Several findings are important to highlight as they have direct implications for future practice. Many youths we interviewed were not aware of the link between their cardiac condition and some of the developmental challenges that they experienced, particularly those related to their mental health and academic difficulties. This is highly problematic in terms of proactive anticipatory guidance, given the high prevalence of academic difficulties and anxiety in adolescents and adults with CHD.[14, 15] Other participants did not realize that daily struggles, even those perceived as not severe, could benefit from a formal evaluation and intervention services and supports. Similarly, Veldtman et al.[16] found that one third of children with CHD had a poor understanding of their cardiac condition and its long-term impacts on daily functioning.

A recent study highlighted that only half of the tertiary care centers that perform pediatric open-heart surgery in Canada have a formal developmental follow-up program for children with CHD, and none systematically administered standardized screenings or evaluations to adolescents with CHD.[17] Developmental follow-up during adolescence was found to be limited to surveillance, a process during which the medical team would ask questions about different aspects of the development.[17] In the current study, the participants expressed that when their cardiology team or the primary healthcare provider asked questions about possible challenges, they did not always consider mentioning developmental challenges unless the limitations were physical and a direct link with the heart defect was evident to them. Therefore, early identification of challenges may not be optimal when surveillance is used embedded within their general medical examinations. Youths with CHD may have waited until there were substantive and persisting consequences on their daily functioning before reporting these difficulties. The youth would often turn to their parents, who initiated the process of undergoing formal evaluation. The importance of awareness as a first step to seeking help is well reflected in the model of help-seeking for mental health problems developed by Rickwood and colleagues.[18] The use of patient-report measures on developmental progress, academic performance, and psychosocial health at specific critical time points during older childhood and adolescence may be helpful for this high-risk population.

The youth interviewed in this study considered the education system a part of the continuum of care. One of the most important perceived benefits of identifying their academic challenges was to offer

appropriate supports. The high prevalence of academic difficulties in this population is well documented. In a large cohort study, Oster and colleagues found that children with CHD were 1.24 times more likely to have difficulty in math or reading, with 44.6% not meeting the minimum standards, and 20.5% requiring individualized education plans.[15] In another study of approximately 100 children with CHD aged 5 to 10 years of age, close to 50% required support at school and 15% were in a special education class.[19] Participants for whom academic difficulties were formally identified but for whom insufficient resources were provided expressed dissatisfaction and frustration with the education system. Hence, it is important that the school system can support their academic, social and mental health needs once challenges are identified, and that adequate resources can be provided. This is in line with the recommendations from the Early Childhood Systems Working Group for policies integrating learning, health and family to optimize outcomes for all children.[20]

The impact of the cardiac defect on their ability to participate in physical education is another area of concern that youth in this study identified within the education system. The distressing and resentful feelings related to the imposed restrictions for participation into sports have been reported in previous studies and can be associated with a loss of identity for more athletic participants.[21] There are clear guidelines to indicate that sports and physical activities should not be restricted for the majority of youth with CHD,[22, 23] but Reybrouck et al. found that overprotection from parents and others may interfere with these recommendations.[24] Therefore, the reasons for imposing activity limitations need to be further investigated to ensure that youth with CHD have the opportunity to engage in physical activity in the absence of documented contra-indications.

The young adults in this study reported that their transition to adult healthcare was often poorly coordinated and led to a decreased sense of trust in their treating healthcare team. Current evidence suggests that a resultant lack of trust in healthcare professionals could have a deleterious impact on help-seeking behaviours.[25] To address these concerns, the Canadian Pediatric Society brought forward the need for formal transition planning. This included ensuring that adolescents understood their health condition, were involved in managing their own health and were confident in their healthcare.[26] They also highlighted the importance of providing uninterrupted, coordinated and developmentally appropriate care during this transition period.[26] The Committee on Improving the Health, Safety, and Well-Being of Young Adults has also suggested treating young adults as a distinct group to address their

specific context.[27] The beginning of adulthood is a time of life when they face new challenges and added responsibilities in the self-management of their health.[27]

This study must be interpreted within the context of its limitations. It was conducted with a small sample of youth with CHD and there was an over-representation of participants living in the province of Quebec. Therefore, the perspectives expressed by the participants may not be generalizable to some provinces where participants could not be recruited.

CONCLUSION

Children with CHD are at high risk of developing physical, academic and psychosocial challenges, however, early identification does not appear to be optimal across domains and transition points across childhood, from the perspective of the youth themselves. In addition, youth with CHD do not have the resources and supports they require to optimize their functional outcomes. Our findings suggest systematic use of self-report measures, transition planning for adult care and integration of the services between the healthcare and school systems as possible strategies to optimize developmental outcomes of youth with CHD in the future.

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	Age in years	**Gender**	**Main cardiac diagnosis** (as described by the participants)	**Province**	**Geographical Area**	**Presence of identified developmental challenge(s) on questionnaire**
Participant 1	17	Female	Atrioventricular septal defect & mitral valve repair	Ontario	Rural	Yes
Participant 2	17	Male	Tetralogy of Fallot	Alberta	Urban	Yes
Participant 3	20	Female	Tetralogy of Fallot	Quebec	Suburban	Yes
Participant 4	13	Female	Double outlet right ventricle	British Columbia	Rural	No
Participant 5	21	Male	Single ventricle defect	Quebec	Suburban	No
Participant 6	14	Female	Truncus arteriosus	British Columbia	Rural	Yes
Participant 7	19	Female	Tetralogy of Fallot	Quebec	Suburban	Yes
Participant 8	21	Female	Aortic stenosis & septal defect	Quebec	Suburban	No
Participant 9	22	Female	Tetralogy of Fallot	Quebec	Rural	Yes
Participant 10	22	Female	Tetralogy of Fallot	Quebec	Urban	Yes

Table 1. Characteristics of participants

CHAPTER 11: DISCUSSION

Despite advances in the surgical and medical management of congenital heart defects (CHDs), the risk of developmental impairments for children with a CHD requiring surgery remains high. Developmental impairments can affect one or more domains and can arise at various ages and persist throughout childhood and adolescence (Bolduc et al., 2020; Gaudet et al., 2021; Ilardi et al., 2020; Liamlahi & Latal, 2019). Notwithstanding this high risk of delays, our results demonstrate that only half of the Canadian tertiary care centers that perform open-heart surgery had systematic follow-up programs that include screening and evaluation of developmental delays (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). These practices are considered suboptimal from the point of view of many healthcare professionals involved in the developmental follow-up of children with CHDs (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). Parents of children with CHDs and youth with CHDs have also identified important gaps in the services received (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). As a result of these gaps, parents are overwhelmed with the responsibilities they must take on to promote their child's development and sufficient resources to support youth's challenges are not accessible (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Timely identification and interventions may hold the key to successfully mitigate the challenges experienced by children and adolescents with CHDs. Strategies to optimize developmental outcome starts in neonatal life and extend into young adulthood. These strategies can take various forms: 1) prevention of developmental delays, 2) timely identification of delays, and 3) resources to support child development.

11.1 PREVENTION OF DEVELOPMENTAL DELAYS : A FAMILY-CENTERED APPROACH

11.1.1 NEWBORN DEVELOPMENTAL CARE

Prevention of developmental delays from a developmental care perspective starts as early as the first hospitalization. Developmental care within the neonatal or cardiac intensive care unit, such as the newborn individualized developmental care and assessment program (NIDCAP), aims to minimize the impacts of intensive care and maintain the connection between the child and their parents (Als & McAnulty, 2011). Although this approach presents a unique challenge for infants with CHDs in the post-operative period, it is showing promising results for different populations of children at high risk of developmental delays including those with CHDs (Anderson et al., 2020; Butler et al., 2017; Lisanti et al.,

2019). In fact, it has been shown to have a positive impact on brain and child development in children born preterm when measured at school-age (McAnulty et al., 2012). Consistent use of this approach across the tertiary care hospitals that perform open-heart surgery in Canada could address some of the concerns voiced by parents in this study with regard to not having the same opportunities to bond or to spend quality time with their child with a CHD. In addition to providing developmental care within the hospital, a resource guide on developmental stimulation can be shared with parents during the hospital stay or at subsequent follow-up as an additional tool to contribute to the prevention of developmental delays, as suggested by one of the study participants (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022).

11.1.2 Supporting Parental Mental Health

A large proportion of parents of children with CHDs experience increased levels of stress, anxiety or depression when compared to parents of healthy children (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022; Wei et al., 2015). Heightened levels of stress around the time of diagnosis and hospitalization have been well described in previous studies (Golfenshtein et al., 2019; Rempel et al., 2013). The parents in our study also expressed the presence of stress associated with the increased responsibilities they must take on to ensure that their child receives the support they need (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Long-term psychological distress in parents can lead to changes in parenting styles and decreased parent-child attachment (Goldberg et al., 1991a; Schmitz, 2019). Parental mental health has been also shown to mediate self-concept, emotional well-being, cognitive skills and behavioral outcomes in children with CHDs (Berant et al., 2008; Brosig et al., 2007; Golfenshtein et al., 2020; Majnemer et al., 2009; McCusker et al., 2013; McCusker et al., 2007). Consequently, assessment of parental mental health has been included in the most recent recommendations by the American Heart Association and the Cardiac Neurodevelopmental Outcome Collaborative (Ilardi et al., 2020; Marino et al., 2012; Ware et al., 2020). Although parental mental health was listed as a high priority in five cardiac intensive care units across the United States of America, few centers implemented any systematic screening (Miller et al., 2020). In our study, only two of the eight centers that perform pediatric open-heart surgery in Canada systematically screened parental mental health. In a previous study, parents of children with CHDs had identified barriers to discussing their mental health with the healthcare providers (Franklin et al., 2021). These included thinking that caring for their mental health was outside the scope of their child's healthcare providers, fear of a negative perception of their parental ability by the

healthcare team and negative reactions by the healthcare professionals in similar situations in the past (Franklin et al., 2021). Conversely, confidence in the medical team's ability to support their mental health and overt efforts by those healthcare professionals to provide support were viewed as facilitators to having discussions on their mental health (Franklin et al., 2021). Parents in our study have voiced that a more systematic approach to developmental follow-up would decrease their burden and help alleviate their stress (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). In a different study, parents had also reported the need for structured psychosocial support services that include both individualized and multidisciplinary approaches (Gramszlo et al., 2020). Importantly, the NIDCAP approach described above can also contribute to alleviating parental stress in their child's first year of life (Khosravan et al., 2020; van der Pal et al., 2008).

11.2 Timely Identification of Developmental Delays

11.2.1 Universal, Systematic, and Responsive Follow-up Services for All Children with a CHD at High-Risk for Delays

Timely identification of developmental impairments is essential in order to put in place the resources needed to remediate or compensate for these difficulties. In fact, the children who benefited from a structured developmental follow-up were more likely to be referred for further evaluation and/or interventions (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). Hence, in the past decade, projects and recommendations were developed worldwide to promote systematic developmental follow-up for children with CHDs (Eagleson et al., 2020; Fourdain et al., 2020; Marino et al., 2012). The recommendations put forward by the American Heart Association in 2012 have been widely recognized (Marino et al., 2012). However, different health service delivery models may require these guidelines to be adjusted to their specific contexts (Eagleson et al., 2020; Fourdain et al., 2020). Most of the healthcare professionals interviewed as part of our study recognized that these guidelines need to be adapted before being successfully implemented in Canada (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). The results of our study also demonstrate that, although Canadian healthcare professionals recognized the importance of systematic developmental follow-up, current practices varied greatly from one province to another. Even when a structured developmental follow-up program was in place, only a subset of high-risk children with CHDs were eligible. In addition, no systematic follow-up was offered after five years of age. In fact, healthcare professionals, parents of children with CHDs and youth with CHDs alike identified important gaps in the current developmental follow-up practices in Canada

(Bolduc, Rennick, Gagnon, Majnemer, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Barriers to implementing universal, systematic and responsive follow-up services have been identified as the lack of sufficient and sustainable funding and the limited number of healthcare professionals available to perform formal evaluations (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022).

A number of parents and youths with CHDs raised the possibility of using self-report measures (i.e., patient-reported outcome measures) in lieu of formal evaluations for some follow-up time points (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). The use of self-report measures was also proposed to replace surveillance, a format that may not be effective in identifying challenges that are not physical and for which the link to the cardiac defect is not as obvious (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). This could prove to be a cost-effective approach to the identification of delays. In line with this approach, one of the surveyed structured developmental follow-up programs in Canada used a web-based platform to collect developmental information through questionnaires (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). This platform, adapted from the American data collection system CHADIS (Child Health and Development Interactive System) (Howard & Sturner, 2017) is now available to healthcare professionals across Quebec for the developmental follow-up of all children (Gouvernement du Quebec, 2022). It could be a cost and time-effective alternative to consider for the collection of developmental information necessary to identify challenges in a timely manner. If screening questionnaires are used to identify impairments in children with CHDs, it is important to implement an effective and systematic approach to referring children whose screening questionnaire raises concerns for further evaluation.

The possibility of having a provincial center to centralize developmental follow-up, minimize costs and optimize expertise was also proposed by one of the healthcare professionals we interviewed (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). However, the option of centralizing resources should be carefully considered. In Australia, similar to Canada where the healthcare system is both universal and funded through taxes, different follow-up service models have been examined. In a pilot project, they first implemented a state-wide centralized follow-up program with formal evaluations at specific time points as recommended by the American Heart Association and as suggested by our study participant (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022; Eagleson et al., 2020; Marino et al., 2012). Unfortunately, the financial costs and human resources required to maintain this centralized program

were not sustainable (Eagleson et al., 2020). A second centralized model using screening tools in lieu of evaluations was also found to be unsustainable due to costs and resource needs (Eagleson et al., 2020). They found that a decentralized approach where developmental care was provided across primary, secondary and tertiary centers was the most sustainable model (Eagleson et al., 2020).

11.2.3 Continuity of Services During Transition to Adult Care

Gaps in follow-up during the transition to adult care were also identified by the young adults with CHDs who participated in our study (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). They articulated the difficulties they faced when the healthcare providers they had known and trusted for years changed to new service providers (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). This finding is supported by previous studies on children with long-term conditions including CHDs (Coyne et al., 2019; de Hosson et al., 2021; Gaydos et al., 2020; Mackie et al., 2019). These studies highlighted the importance of gaining trust in the new healthcare provider. This could be promoted by the implementation of joint consultations prior to the transfer (Coyne et al., 2019; de Hosson et al., 2021). Participants in our study also emphasized the importance of a coordinated process where their medical information would be accessible to the new medical team (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). They identified the importance of a contact person to obtain information between follow-up visits if new challenges arise (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). The potential effectiveness of transition interventions in adolescents aged 16-17 years of age to optimize transition readiness for adolescents with CHDs was recently demonstrated (Charles et al., 2021; Moons et al., 2021). The most highly valued components of these sessions included using a health passport, participating in role plays to simulate the clinical environment and goal setting. Organizing an annual day during which information sessions and leisure activities are offered with the goal of improving knowledge of the condition and providing peer support have also been suggested (de Hosson et al., 2020). Fortunately, a growing number of young adults with CHDs are being referred to specialized adult congenital heart defects centres which have been associated with reduced mortality rates (Mackie et al., 2019).

11.2.4 Selection of Assessment Tools and Timing of Assessments

Other important considerations for the effective identification of developmental impairments include assessing all developmental domains that are at-risk of being affected (Ilardi et al., 2020). The domains to be assessed in this population and suggested evaluation tools have been comprehensively synthesized in two articles recently published by the Cardiac Neurodevelopmental Outcome Collaborative (Ilardi et al., 2020; Ware et al., 2020). Overall, the outcomes to be evaluated were grouped under the following domains: cognition/intelligence/memory, language, motor (gross and fine), academic/school readiness, adaptive skills, attention/behavior, autism/social communication, executive functions, social-emotional functioning. In addition, a significant number of parents also reported sensory dysregulation such as sleep disturbances and hypersensitivity (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022; Rempel et al., 2014). Their incidence needs to be further investigated to determine whether systematic screening for these impairments needs to be included in future guidelines.

The Cardiac Neurodevelopmental Outcome Collaborative provided an extensive list of domain-specific assessment tools for each of these domains (Ilardi et al., 2020; Ware et al., 2020). However, there is no recommendation of an assessment tool for gross motor skills beyond five years of age. Based on the results of our systematic review of motor outcomes, we suggest adding an assessment of gross motor function for children aged 6 to 18 years, since mild to moderate motor impairments were consistently present in similar proportions across childhood and adolescence. The Bruininks-Oseretsky test of motor proficiency, second edition (Bruininks & Bruininks, 2005) and the Movement ABC second edition (Henderson et al., 2007) are examples of tests with good psychometric properties that could be used to assess gross and fine motor skills in older children and adolescents with CHDs (Griffiths et al., 2018). Recommendations for assessing the presence and extent of sensory dysregulation are also absent in this list. The Sensory Integration and Praxis tests (Ayres, 1989) and the Sensory Profile 2 (Dunn, 2014) are the most commonly used tools to evaluate sensory processing in children and could be used for the assessment of this domain (Jorquera-Cabrera et al., 2017). The former is an assessment battery comprising 17 subtests while the latter comprises questionnaires (Jorquera-Cabrera et al., 2017).

If implementing an approach to developmental follow-up that uses screening questionnaires as a first step in identifying developmental delays, the Ages & Stages Questionnaires - Third Edition (ASQ-3) (Squires & Bricker, 2009) has been shown to have adequate psychometric properties to detect developmental delays in children with CHDs for children up to school age (Lepine et al., 2022; Noeder et

al., 2017). Nevertheless, Noeder et al. (2017) recommended using a one standard deviation below the mean cut-off to improve sensitivity. Similarly, Eagleson and colleagues (2020) recommend using a low threshold on the ASQ-3 since mild challenges are frequent and often present across multiple domains in children with CHDs. Telephone surveillance using measures such as the Vineland Adaptive Behavior Scales (Sparrow et al., 2016) to identify activity limitations across domains is another potential option for developmental monitoring at key transition points for older children and adolescents with CHDs (Limperopoulos et al., 2006). For youth, with CHDs, the use of a communication paper, a non-standardized digital short questionnaire to identify the topics that adolescents with CHDs would like to discuss, could also be used (de Hosson et al., 2020).

In terms of timing for evaluation, the Cardiac Neurodevelopmental Outcome Collaborative recently proposed new time points for the re-evaluation of developmental and functional outcomes (Ilardi et al., 2020; Ware et al., 2020). Evaluations at 6 months, 18 months, 36 months, 5 years, 8-9 years, 10-11 years, 13-14 years and 18 years were recommended and are well aligned with key moments in the developmental trajectory or important transition time points (Ilardi et al., 2020; Ware et al., 2020). Eagleson and colleagues (2020) recommended similar time points to assess children using the ASQ-3. Although ideal, it may not be possible, considering the barriers identified in the Canadian healthcare system, to implement formal evaluations at eight timepoints across development (Bolduc, Rennick, Gagnon, Majnemer, et al., 2022). Across the interviewed parents in our study, the preferred frequency was variable but the most important transition period identified was the beginning of kindergarten (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Many parents identified the need for a comprehensive evaluation around the age of 4 or 5. Similarly, the youth in our study recommended assessments every year or every two years between 5 and 12 years old with the most important assessment taking place during academic transitions such as the beginning of kindergarten, high school and college (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022). An assessment in the first years of life would also allow access to early intervention during a critical window of enhanced brain plasticity (Cioni et al., 2016).

11.3 Resources to Support Child Development

The final step to providing comprehensive developmental care to children with CHDs and their families is ensuring access to resources that support the development and functioning of children with CHDs

once challenges have been identified. These interventions can include remediation and/or compensation strategies depending on the needs of the child (Majnemer, 1998). Participants in our studies have identified the lack of resources and the presence of long waitlists as critical barriers to accessing the required support (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022) . In Canada, in-home or community-based interventions often focus on the 0-3 year window (Vyas et al., 2021). However, a study of service uses in children with hypoplastic left heart syndrome conducted in Australia showed that, while children visited their primary care practitioner more frequently in their early years, consultations with allied healthcare professionals were most frequent for children aged 6-9 years (Huang et al., 2020). Keeping in mind that resources and funding are limited, parents have proposed that phone consultation or telehealth may be used (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Some parents also mentioned the observed benefits of participating in community services (Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). It is therefore essential that a list of these services and other community resources be provided to the parents and children at various time points during the child's development. Applications such as Jooay© (an application of adapted and inclusive leisure services in communities across Canada) could also become an invaluable source of information for parents, children, and youth alike (Shikako et al., 2021). Finally, youths in our study clearly expressed the need for ongoing support during adolescence since new challenges could arise because of the higher social and academic demands as well as increased level of autonomy expected at this age. They identified the need for better integration of the services offered through the healthcare and school systems in order to prevent any interruption of services. This need for more targeted school-based supports was also highlighted in a recent Canadian study on the use of interventions for children with CHDs and barriers to accessing early intervention (Vyas et al., 2021). In fact, an earlier study has found that more than a third of children with CHDs experiencing difficulties in the motor, cognitive or behavioral domains needed educational supports (Majnemer et al., 2017).

11.4 LIMITATIONS

While this project successfully synthesized motor outcomes in children and adolescents with CHDs, gathered information on current developmental follow-up practices in Canada and explored perspectives on optimal practices from three groups of stakeholders, it also had some limitations. Variations in sampling strategies and the use of different motor evaluation tools may have resulted in heterogeneity across the studies included in our systematic review (study I). In the environmental scan

(study II), information on current practices was obtained from a single source in most institutions. Although we ensured that the healthcare professionals interviewed were knowledgeable of the practices in place, some information may not have been captured. In addition, our study did not examine the role that primary care providers play in the developmental follow-up of children and adolescents with CHDs in Canada. However, based on the interviews we conducted with parents and youth, most family doctors performed informal developmental follow-up in the form of surveillance during routine follow-up appointments (Bolduc, Rennick, Gagnon, Sokol, Brossard-Racine, et al., 2022; Bolduc, Rennick, Gagnon, Sokol, Majnemer, et al., 2022). Notwithstanding our ambitious recruitment strategy, we were not able to recruit parents and youth from all provinces, and youth from the province of Québec were overrepresented (study III). Therefore, the results of our study may not be generalizable to all provinces, and we may not have captured all gaps and potential strategies to optimize developmental follow-up. Additionally, as the primary interviewees in all parent interviews were mothers, we may not have captured how fathers' perspectives differed. Finally, we did not actively recruit children younger than 12 years of age to participate in our study. Therefore, we were unable to explore if their perception of developmental follow-up was different from those of parents and youth.

11.5 Future Directions

This study gathered information from various sources that will be useful in the elaboration of developmental follow-up guidelines that are adapted to the Canadian context. Based on the interviews we conducted with parents, the prevalence of sensory dysregulation seems to be higher in children with CHDs compared to the general population. Studies are needed to specifically examine the prevalence of these issues, investigate their underlying causes, and determine the resources needed by parents and their children with regard to these challenges. In addition, future studies need to examine current developmental follow-up practices offered by community providers and investigate the complementary role they could play in the implementation of systematic developmental services. Finally, the next step is to assemble a committee of experts to develop recommendations for optimal practices across Canada. The elaboration of practice guidelines for the developmental follow-up of children and adolescents with CHDs should involve all relevant stakeholders and include a consultation process in the form of a nominal group process, consensus development panel and/or Delphi procedure.

CHAPTER 12: CONCLUSION

Infants born with a congenital heart defect (CHD) now benefit from lifesaving surgical repairs thanks to recent medical advances. However, with the vast majority of children surviving open-heart surgery, it has become clear that CHDs are associated with a high prevalence of developmental delays that can affect multiple domains and disrupt daily function.

The results of this project have highlighted the need for systematic developmental services that include all developmental domains and span from infancy to adolescence. These services must be available to all children at high risk of developmental delay. Unfortunately, current developmental follow-up practices are not optimal in Canada from the point of view of healthcare professionals, parents and youth with CHDs. These limitations put an undue burden on families who must assume additional roles to compensate for these limitations. This results in an increased level of stress for the parents and can affect the entire family. Gaps in current follow-up practices and limited access to resources also result in unsupported needs for youth with CHDs and lost opportunities to enhance developmental outcomes.

Considering the scarce sources of financing for these services and the limited availability of human resources to perform formal evaluations, it is clear that the guidelines developed by the American Heart Association in the United States of America cannot be implemented in the Canadian context without modification to work around existing barriers. The participants in our study have proposed ideas and strategies to improve developmental care in Canada. This includes the implementation of systematic time points for assessment at key transitions, the use of screening questionnaires and self-report measures (patient-reported outcome measures), the implementation of preventive strategies, better transition planning for transfer to adult care, more resources available within the school system and information on readily available community resources. Telehealth may provide cost-effective opportunities for surveillance and systems navigation.

Implementing more systematic follow-up practices and improving access to resources could promote a more balanced and positive parent-child relationship. Moreover, the provision of timely interventions would optimize the child's physical and mental health and developmental outcomes, so that each child can attain their potential and participate fully at school and in their community.

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APPENDIX I: CONSENT AND ASSENT FORMS

INFORMATION AND CONSENT FORM FOR HEALTHCARE PROFESSIONALS (STUDY II)

INFORMATION AND CONSENT FORM

Healthcare Professionals

Phase II

Research Study Title:	Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect
Protocol number:	2020-5921
Researcher responsible for the research study:	Dr. Marie Brossard-Racine
Co-Investigator(s)/sites:	Marie-Eve Bolduc and Dr. Annette Majnemer
Sponsor:	n/a

INTRODUCTION

We are inviting you to take part in this research study because you are involved in the developmental surveillance of children and adolescents with a complex congenital heart defect (CHD).

However, before you accept to participate in this study and sign this information and consent form, please take the time to read, understand and carefully examine the following information.

We invite you to speak to the researcher responsible for this study (i.e. “the researcher”) or to other members of the research team and ask them any questions you may have about this study. Please also ask a member of the research team about any parts of this consent form you do not understand.

BACKGROUND

Children and adolescents with congenital heart disease (CHD) are at high risk for developmental delays. Early identification through developmental surveillance is essential in order to provide timely interventions. Current developmental follow-up practices in Canada need to be identified to assess if they are optimal.

PURPOSE OF THE RESEARCH STUDY

The purpose of this study is to describe current developmental surveillance practices across Canada and to identify current structural barriers to the developmental surveillance of children with CHD post open-heart surgery.

For this research study, we will recruit approximately 32 participants. One health professional from the cardiology and neonatal follow up units of each pediatric academic health centers across Canada that performs open-heart surgery will be interviewed as part of this study. Health professionals from other departments may also be recruited if involved in the developmental follow up of children and adolescents with CHD

DESCRIPTION OF THE RESEARCH PROCEDURES

This research study will take place at the by phone or internet call (e.g., Skype or Facetime) at the time most convenient to the participant.

1. Duration and number of visits

Your participation in this research study involves in a single phone interview that will last 30-45 minutes.

2. Study Procedures

During your participation in this research study, you will participate in one phone interview. An appointment will be made in advance for this interview. A copy of the questions about current practices will be sent in advance, so that you can review the content, and gather information as needed. The first part of the interview will consist of a survey that will include questions to identify current surveillance practices for children with CHD post open-heart surgery. The second part will consist of questions to gather your opinion on current practices and identify structural barriers to developmental surveillance.

PARTICIPANT'S RESPONSIBILITIES

- Gather all necessary information with regards to the developmental surveillance of children and adolescents with CHD in their department prior to the interview.

BENEFITS ASSOCIATED WITH THE RESEARCH STUDY

There is no direct benefit to you for participating in this research. However, we hope that the study results will contribute to optimizing neurodevelopmental outcomes in children and adolescents with congenital heart defects.

RISKS ASSOCIATED WITH THE RESEARCH STUDY

A possible risk associated with this study is a breach of confidentiality or use of your personal information by a third party. To limit this risk, we will take the steps to protect your confidentiality described in the Confidentiality section, below.

We do not foresee any other risks associated with this study.

VOLUNTARY PARTICIPATION AND THE RIGHT TO WITHDRAW

Your participation in this study is voluntary. Therefore, you may refuse to participate. You may also withdraw from the *ongoing* project at any time, without giving any reason, by informing a member of the study team. Your decision not to participate in the study, or to withdraw from it, will have no bearing on your job or on any work-related evaluations or reports. You will be informed in a timely manner if any information becomes available that may impact your willingness to continue participating in this study.

The researcher or the Research Ethics Board may put an end to your participation without your consent. This may happen if new findings or information indicate that participation is no longer in your interest, if you do not follow study instructions, or if there are administrative reasons to terminate the project.

If you withdraw or are withdrawn from the study, you may also request that the data already collected about you be removed from the study. If you request that your data be removed and the information already collected about you can be identified as yours it will be destroyed. If the data has been anonymized or was always anonymous (i.e. does not contain any information that can be used to identify you), the data will continue to be used in the analysis of the study.

CONFIDENTIALITY

No information that can identify you personally will be collected and it will not be possible to link your study data to you. All the information collected during the research project will remain confidential to the extent provided by law. Your department and institution will only be identified by a code number. The key to the code linking your institution's name to the institution' study number will be kept by the researcher.

All audio-recordings will be transcribed (your words will be written down) in a de-identified fashion (i.e. your name will not appear in the transcripts). The audio-recordings will then be destroyed. It is possible that direct quotes of what you said will be presented in publications and/or conferences. However, precautions will be taken to ensure that it will not be possible to identify you. Your employer will not have access to your answers to the survey.

The study data will be stored for 7 years by the researcher responsible for the study.

The data may be published or shared during scientific meetings; however, precautions will be taken to ensure that it will not be possible to identify you or your institution.

For auditing purposes, the research study files which could include documents that may identify you may be examined by a person mandated by the institution, or the Research Ethics Board. All these individuals and organizations adhere to policies on confidentiality.

You have the right to consult your study file in order to verify the information gathered, and to have it corrected if necessary.

FUNDING OF THE RESEARCH PROJECT

The researcher has received funding from the Richard and Edith Strauss Foundation to conduct this research project.

CONFLICT OF INTERESTS

The researchers have no conflict of interest to declare.

COMPENSATION

You will not receive financial compensation for participating in this research study.

SHARING STUDY RESULTS

If you wish, you will receive a summary of research results by email if you provide your contact information on the form below. Results from this study will be presented at least one scientific conference and published in one peer-reviewed journal.

SHOULD YOU SUFFER ANY HARM

By agreeing to participate in this research project, you are not waiving any of your legal rights nor discharging the researcher, the sponsor or the institution, of their civil and professional responsibilities.

CONTACT INFORMATION

If you have questions or if you have a problem you think may be related to your participation in this research study, or if you would like to withdraw, you may communicate with the researcher or with someone on the research team at the following email address: marie-eve.bolduc@mcgill.ca.

For any question concerning your rights as a research participant taking part in this study, or if you have comments, or wish to file a complaint, you may communicate with:

The Ombudsman of the McGill University Health Centre at the following phone number: 514-934-1934 ext 48306.

OVERVIEW OF ETHICAL ASPECTS OF THE RESEARCH

The McGill University Health Centre Research Ethics Board reviewed this study and is responsible for monitoring it at all participating institutions in the health and social services network in Quebec.

Research Study Title: Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect

SIGNATURES

Signature of the participant

I have reviewed the information and consent form. Both the research study and the information and consent form were explained to me. My questions were answered, and I was given sufficient time to make a decision. After reflection, I consent to participate in this research study in accordance with the conditions stated above.

1) I accept that my participation in the study be audio-recorded.
   Yes ☐ No ☐
2) I authorize a member of the research study to contact me to check the transcript of what I said.
   Yes ☐ No ☐
3) I wish to receive a copy of the study results by email.
   Yes ☐ No ☐ If yes, please provide contact information: _____
4) I authorize a member of the research study to contact me in the future to ask if I am interested in participating in other research.
   Yes ☐ No ☐ If yes, please provide contact information: _____

Name of participant Signature Date

Signature of the person obtaining consent

I have explained the research study and the terms of this information and consent form to the research participant, and I answered all his/her questions.

Name of the person obtaining consent Signature Date

INFORMATION AND CONSENT FORM FOR THE PARENTS OF A CHILD BORN WITH A CONGENITAL HEART DEFECT (STUDY III)

INFORMATION AND CONSENT FORM

Parent of a child born with a congenital heart defect

Phase III

Research Study Title:	Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect
Protocol number:	2020-5921
Researcher responsible for the research study:	Dr. Marie Brossard-Racine
Co-Investigator(s)/sites:	Marie-Eve Bolduc and Dr. Annette Majnemer
Sponsor:	n/a

INTRODUCTION

We are inviting you to take part in this research study because you are the parent of a child or adolescent born with a congenital heart defect that required open-heart surgery before the age of 2 years old.

However, before you accept to take part in this study and sign this information and consent form, please take the time to read, understand and carefully examine the following information. You may also want to discuss this study with your family doctor, a family member or a close friend.

This form may contain words that you do not understand. We invite you to speak to the researcher responsible for this study ("the researcher") or to the co-investigators and ask them any questions you may have about this study. Please also ask a member of the research team about any parts of this consent form you do not understand.

BACKGROUND

Children with a congenital heart defect (CHD) now benefit from early surgical repair and can live a full life. However, some children may experience developmental delays. These difficulties can be better addressed if identified as early as possible. Effective developmental surveillance needs to take into consideration the potential challenges experienced by children and adolescents with CHD, the barriers and the preferences of patients and families. However, the needs and preferences of children and youth with a congenital heart defect as well as their families with regards to the best ways to monitor their development have not been investigated to this day.

PURPOSE OF THE RESEARCH STUDY

The purpose of this study is to define what families would most prefer in terms of developmental surveillance of children and adolescents with congenital heart defects.

For this research study, we will recruit approximately 15 parents of children or adolescents with a congenital heart defect and 10 youth (13-19 years old) born with a congenital heart defect who had open-heart surgery before the age of 2 years old.

DESCRIPTION OF THE RESEARCH PROCEDURES

This research study will take place at the McGill University Health Centre (for participants in the Montreal region), by phone or internet call (e.g., Skype or Facetime) at the time most convenient to the participant.

3. Duration and number of visits

Your participation in this research study involves a single interview that will last approximately 60 minutes.

4. Study Procedures

If you agree to participate in this project, you will first be asked to complete a questionnaire about your child's developmental history and your family's demographics. We will also schedule a one-hour interview about the developmental follow up your child has received. You will have the option to do the interview in person at the McGill University Health Centre (if you are from the Montreal region), at home or another convenient location. If it is not possible to meet for a face-to face interview, it could also be completed by phone or via Internet by using Skype, Facetime, Zoom, or Snapchat.

BENEFITS ASSOCIATED WITH THE RESEARCH STUDY

There is no direct benefit to you for participating in this research. However, we hope that the study results will contribute to improving the developmental follow up for children and adolescents born with a heart defect.

RISKS ASSOCIATED WITH THE RESEARCH STUDY

A possible risk associated with this study is a breach of confidentiality or use of your personal information by a third party. To limit this risk, we will take the steps to protect your confidentiality described in the Confidentiality section, below.

We do not foresee any other risks associated with this study.

VOLUNTARY PARTICIPATION AND THE RIGHT TO WITHDRAW

Your participation in this study is voluntary. Therefore, you may refuse to participate. You may also withdraw from the *ongoing* project at any time, without giving any reason, by informing a member of the study team. Your decision not to participate in the study, or to withdraw from it, will have no impact on the quality of care and services to which your child is otherwise entitled. You will be informed in a timely manner if any information becomes available that may impact your willingness to continue participating in this study.

The researcher or the Research Ethics Board may put an end to your participation without your consent. This may happen if new findings or information indicate that participation is no longer in your interest, if you do not follow study instructions, or if there are administrative reasons to terminate the project.

If you withdraw or are withdrawn from the study, you may also request that the data already collected about you be removed from the study. If you request that your data be removed and the information already collected about you can be identified as yours it will be destroyed. If the data has been anonymized or was always anonymous (i.e. does not contain any information that can be used to identify you), the data will continue to be used in the analysis of the study.

CONFIDENTIALITY

During your participation in this study, the researcher and his/her team will collect and record information about you and your child. They will only collect information necessary for the study.

The following information will be collected from the questionnaire: age, sex, challenges experienced by your child, parental level of education and family income.

All the information collected during the research project will remain confidential to the extent provided by law. You will only be identified by a code number. The key to the code linking your name to your study participant number will be kept by the researcher.

All audio-recordings will be transcribed (your words will be written down) in a de-identified fashion (i.e. your name will not appear in the transcripts). The audio-recordings will then be destroyed. It is possible that direct quotes of what you said will be presented in publications and/or conferences. However, precautions will be taken to ensure that it will not be possible to identify you.

The study data will be stored for 7 years by the researcher responsible for the study.

The data may be published or shared during scientific meetings; however, precautions will be taken to ensure that it will not be possible to identify you.

For auditing purposes, the research study files which could include documents that may identify you may be examined by a person mandated by the institution, or the Research Ethics Board. All these individuals and organizations adhere to policies on confidentiality.

You have the right to consult your study file in order to verify the information gathered, and to have it corrected if necessary.

FUNDING OF THE RESEARCH PROJECT

The researcher has received funding from the Richard and Edith Strauss Foundation to conduct this research project.

CONFLICT OF INTERESTS

The researchers have no conflict of interest to declare.

COMPENSATION

For costs and inconveniences you experienced during this study, you will receive a total compensation in the amount of $45. If you withdraw before study completion, you will receive an amount proportional to your participation.

SHARING STUDY RESULTS

If you wish, you will receive a summary of research results by email if you provide your contact information on the form below. Results from this study will be presented at least one scientific conference and published in one peer-reviewed journal.

SHOULD YOU SUFFER ANY HARM

By agreeing to participate in this research project, you are not waiving any of your legal rights nor discharging the researcher, the sponsor or the institution, of their civil and professional responsibilities.

CONTACT INFORMATION

If you have questions or if you have a problem you think may be related to your participation in this research study, or if you would like to withdraw, you may communicate with the researcher or with someone on the research team at the following email address: marie-eve.bolduc@mcgill.ca.

For any question concerning your rights as a research participant taking part in this study, or if you have comments, or wish to file a complaint, you may communicate with:

The Patient Ombudsman of the McGill University Health Centre at the following phone number: 514-934-1934 ext. 48306.

OVERVIEW OF ETHICAL ASPECTS OF THE RESEARCH

The McGill University Health Centre Research Ethics Board reviewed this study and is responsible for monitoring it at all participating institutions in the health and social services network in Quebec.

Research Study Title: Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect

SIGNATURES

Signature of the participant

I have reviewed the information and consent form. Both the research study and the information and consent form were explained to me. My questions were answered, and I was given sufficient time to make a decision. After reflection, I consent to participate in this research study in accordance with the conditions stated above.

I authorize the study team to have access to my medical record for the purposes of this study.

1) I accept that my participation in the study be audio-recorded.
   Yes ☐ No ☐

2) I authorize a member of the research study to contact me to check the transcript of what I said.
   Yes ☐ No ☐

3) I wish to receive a copy of the study results by email.
   Yes ☐ No ☐ If yes, please provide contact information: _____

4) I authorize a member of the research study to contact me in the future to ask if I am interested in participating in other research.
   Yes ☐ No ☐ If yes, please provide contact information: _____

Name of participant Signature Date

Signature of the person obtaining consent

I have explained the research study and the terms of this information and consent form to the research participant, and I answered all his/her questions.

Name of the person obtaining consent Signature Date

Centre universitaire de santé McGill — McGill University Health Centre

INFORMATION AND CONSENT FORM

Patients with a congenital heart defect

Phase III

Research Study Title:	Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect
Protocol number:	2020-5921
Researcher responsible for the research study:	Marie-Eve Bolduc
Co-Investigator(s)/sites:	Dr. Marie-Brossard-Racine and Dr. Annette Majnemer
Sponsor:	n/a

INTRODUCTION

We are inviting you to take part in this research study because you were born with a congenital heart defect that required open-heart surgery before the age of 2 years old.

However, before you accept to take part in this study and sign this information and consent form, please take the time to read, understand and carefully examine the following information. You may also want to discuss this study with your family doctor, a family member or a close friend.

This form may contain words that you do not understand. We invite you to speak to the researcher responsible for this study ("the researcher") or to the co-investigators and ask them any questions you may have about this study. Please also ask a member of the research team about any parts of this consent form you do not understand.

BACKGROUND

Individuals with a congenital heart defect (CHD) now benefit from early surgical repair and can live a full life. However, some people may experience developmental delays. These difficulties can be better addressed if identified as early as possible. Effective developmental surveillance needs to take into consideration the potential challenges experienced by children and adolescents with CHD, the barriers and the preferences of patients and families. However, the needs and preferences of children and youth with a congenital heart defect as well as their families with regards to the best ways to monitor their development have not been investigated to this day.

PURPOSE OF THE RESEARCH STUDY

The purpose of this study is to define what youth born with a congenital heart defect would most prefer in terms of their developmental surveillance of children and adolescents with congenital heart defects.

For this research study, we will recruit approximately 10 youth (13-19 years old) born with a congenital heart defect who had open-heart surgery before the age of 2 years old and 15 parents of children or adolescents with a congenital heart defect.

DESCRIPTION OF THE RESEARCH PROCEDURES

This research study will take place at the McGill University Health Centre (for participants in the Montreal region), by phone or internet call (e.g., Skype or Facetime).

5. Duration and number of visits

Your participation in this research study involves a single interview that will last approximately 60 minutes.

6. Study Procedures

If you agree to participate in this project, you will first be asked to complete a questionnaire about your developmental history and your family's demographics. We will also schedule a one-hour interview about the developmental follow up you have received. You will have the option to do the interview in person at the McGill University Health Centre (if you are from the Montreal region), at home or another convenient location. If it is not possible to meet for a face-to face interview, it could also be completed by phone or via Internet by using Skype, Facetime, Zoom, or Snapchat. You may accompanied by a parent for this interview if you prefer.

BENEFITS ASSOCIATED WITH THE RESEARCH STUDY

There is no direct benefit to you for participating in this research. However, we hope that the study results will contribute to improving the developmental follow up for children and adolescents born with a heart defect.

RISKS ASSOCIATED WITH THE RESEARCH STUDY

A possible risk associated with this study is a breach of confidentiality or use of your personal information by a third party. To limit this risk, we will take the steps to protect your confidentiality described in the Confidentiality section, below.

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The following information will be collected from the questionnaire: age, sex, challenges experienced, parental level of education and family income.

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All audio-recordings will be transcribed (your words will be written down) in a de-identified fashion (i.e. your name will not appear in the transcripts). The audio-recordings will then be destroyed. It is possible that direct quotes of what you said will be presented in publications and/or conferences. However, precautions will be taken to ensure that it will not be possible to identify you.

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The data may be published or shared during scientific meetings; however, precautions will be taken to ensure that it will not be possible to identify you.

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CONTACT INFORMATION

If you have questions or if you have a problem you think may be related to your participation in this research study, or if you would like to withdraw, you may communicate with the researcher or with someone on the research team at the following email address: marie-eve.bolduc@mcgill.ca.

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Research Study Title: Optimizing the developmental surveillance of children and adolescents born with a congenital heart defect

SIGNATURES

Signature of the participant

I have reviewed the information and consent form. Both the research study and the information and consent form were explained to me. My questions were answered, and I was given sufficient time to make a decision. After reflection, I consent to participate in this research study in accordance with the conditions stated above.

I authorize the study team to have access to my medical record for the purposes of this study.

1) I accept that my participation in the study be audio-recorded.

   Yes ☐ No ☐

2) I authorize a member of the research study to contact me to check the transcript of what I said.

   Yes ☐ No ☐

3) I wish to receive a copy of the study results by email.

   Yes ☐ No ☐ If yes, please provide contact information: _____

4) I authorize a member of the research study to contact me in the future to ask if I am interested in participating in other research.

   Yes ☐ No ☐ If yes, please provide contact information: _____

Name of participant (<18 years old) Signature Date

Assent of minor, capable of understanding the nature of the research (signature) or Verbal assent of minor obtained by:

_____

Name of parent(s) or legal guardian Signature Date

Name of participant (18 years +) Signature

Date

Signature of the person obtaining consent

I have explained the research study and the terms of this information and consent form to the research participant, and I answered all his/her questions.

Name of the person obtaining consent Signature Date

APPENDIX II: QUESTIONNAIRE AND INTERVIEW FORM FOR STUDY II

Introduction

Thank you for taking the time to participate in our study. As described in our earlier email exchange, I will first ask you questions on current developmental follow-up practices at your institution. Later in this interview, I will ask you about your perspectives on these practices.

Do you have any questions?

Section One: Descriptive Information

1. How would you describe your institution?
   - ϒ General Hospital/University Health Centre (i.e. primarily adult patient population)
   - ϒ Pediatric Hospital/Pediatric University Health Centre

2. How many children had open-heart surgery for a CHD at your institution in the past 3 years?

   2017: _____

   2018: _____

   2019: _____

3. What is your professional background?
   - ϒ Cardiologist
   - ϒ Cardiothoracic Surgeon
   - ϒ Neonatologist
   - ϒ Nurse
   - ϒ Occupational Therapist
   - ϒ Pediatrician

□ Physical Therapist

ϒ Psychologist

ϒ Other, please specify _____

4. In which unit/program do you provide developmental follow-up services?
   ϒ Cardiology

   ϒ Child Development

   ϒ Neonatal Follow-up

   ϒ Occupational Therapy_

   ϒ Pediatric Intensive Care

   ϒ Physical Therapy

   ϒ Psychology

   ϒ Other, please specify _____

5. What is your role within that unit/program?

   _____

6. How many years has it been since graduating from your professional program?
   ϒ <5 years

   ϒ 5-15 years

   ϒ > 15 years

7. For how many years have you been working with the CHD population?
   ϒ <5 years

   ϒ 5-15 years

   ϒ > 15 years

Section Two: Current Follow-up Practices

8. Do healthcare professionals in your unit/program perform **developmental surveillance** for children and adolescents with CHD?
   We define developmental surveillance as a flexible process during which the healthcare professional periodically asks parents about their concerns with regards to the development of their child, performs skilled observation of the child and maintains a developmental history of the child.

   ϒ Yes

   ϒ No

   If yes, to which population (e.g., all children who underwent open-heart surgery before 6 months of age)?

   _____

   _____

   _____

   To your knowledge, is developmental surveillance also performed by another unit/program at your institution?

   ϒ Yes, please specify _____

   ϒ No

9. Do healthcare professionals in your unit perform developmental **screening** for children and adolescents with complex CHD?
   We define screening as the use of a short, validated assessment that can be completed by the parent or health professional to further assess areas of concern.

   ϒ Yes

   ϒ No

   If yes, to which population (e.g., all children who underwent open-heart surgery)?

_____

_____

_____

If yes, which developmental screening questionnaire(s) or tool(s) do you use? _____

_____

_____

To your knowledge, is developmental screening also performed by another unit/program at your institution?

ϒ Yes, please specify _____

ϒ No

10. Do healthcare professionals in your unit perform developmental **evaluations** for children and adolescents with CHD?
We define evaluation as a comprehensive standardized assessment that is performed by trained professionals that can lead to a specific diagnosis and intervention plan.

ϒ Yes

ϒ No

If yes, to which population (e.g., all children who underwent open-heart surgery)?

_____

_____

_____

If yes, which developmental assessments(s) does your unit/program uses and who performs each of these assessments (e.g., Bayley – psychologist)?

Assessment	Healthcare Professional

To your knowledge, are <u>developmental evaluations</u> also performed by another unit/program at your institution?

ϒ Yes, please specify _____

ϒ No

11. Please indicate which healthcare professionals in your unit/program perform developmental surveillance, screening and evaluation? Select all that apply.

	Surveillance	Screening	Evaluation
Cardiologist			
Cardiothoracic Surgeon			
Intensivist			
Neonatologist			
Nurse			
Occupational Therapist			
Pediatrician			
Physical Therapist			
Psychologist			
Other, please specify			

12. Please indicate the frequency at which professionals in your unit/program perform developmental surveillance, screening and evaluation, for each of the following ages?

	Surveillance	Screening	Evaluation
0-12 months old	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other
1 to 3 years old	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other
3 to 5 years old	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other
5 to 12 years old	ϒ Monthly ϒ Twice-annually ϒ Annually	□ Monthly ϒ Twice-annually ϒ Annually	□ Monthly ϒ Twice-annually ϒ Annually

	□ If concerned ϒ Never ϒ Other	□ If concerned ϒ Never ϒ Other	□ If concerned ϒ Never ϒ Other
13 to 18 years old	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other	ϒ Monthly ϒ Twice-annually ϒ Annually ϒ If concerned ϒ Never ϒ Other

13. How often do you refer children and adolescents with CHD (those that had open heart surgery) to rehabilitation services?

ϒ Never (0%)

ϒ Rarely <10%

ϒ Sometimes 11-35%

ϒ Often 36-70%

ϒ Most of the time (>70%)

14. To which of the following specialists do you refer children or adolescents with CHD for evaluation or intervention? Select all that apply.

	Evaluation	**Intervention**
Occupational Therapist		
Physical Therapist		
Psychologist		
Speech Language Pathologist		
Special Educator		
Other: _____		

15. What are the reasons for which you refer children and adolescents with CHD for evaluation or intervention services? Select all that apply.

□ Developmental screening raises concerns (acquisition of milestones)

ϒ Feeding difficulties

ϒ Behavioral difficulties

ϒ Academic difficulties

ϒ Motor difficulties

ϒ Other: _____

16. Do health professionals in your unit/program perform surveillance, screening or evaluation of parents' mental health (e.g., stress, depression)

Probes:

- When are these performed? _____

- Which healthcare professional performs these? _____

*- How is the parents' mental health assessed?*_____

Transition

We will now move to questions related to your perspective on the developmental follow-up of children and adolescents with complex CHD. You have not received these questions in the email.

Section Three: Feasibility and acceptability of guidelines and barriers to developmental follow-up

17. Are you familiar with the 2012 American Heart Association developmental follow-up guidelines?
ϒ Yes

➔ Go to question 18

□ No

➔ Go to question 22

18. What is your opinion on these guidelines?

Probes: Strengths/limitations? Do you think they are feasible in Canada? Acceptable?

Why or why not?

19. Do you use the 2012 AHA's developmental guidelines to inform your practice decisions on follow-up and referral of children and adolescents with CHD?

20. Could you tell me more about why you use or do not use these guidelines?

21. Are you familiar with the screening and evaluation tools recommended in these guidelines?
ϒ Yes

ϒ No

22. Do you consider the current developmental follow-up practices at your institution as being optimal?
ϒ Yes

Probes:

- *Could you tell me more about the reasons why you think current practices are optimal?*

ϒ No

Probes:

- *What are some of the current barriers at your institution in terms of developmental follow-up?*
- *Which practices would you want to change to optimize the developmental follow-up and referral practices?*

23. Is there anything you would like to add with regards to the developmental follow-up of children and adolescents with complex CHD?

24. May we contact you if we have follow-up questions?
    ϒ Yes

    ϒ No

    ➔ Go to closing statement below

For participants from the Montreal region only

25. Would you be interested in participating in a consensus group to develop recommendations to improve developmental follow-up?
    ϒ Yes

    ➔ Go to question 26
    ϒ No

    ➔ Go to closing statement

26. May we contact you for this phase of our project in a few months?
    ϒ Yes, how would you like to be contacted? _____

    ϒ No

Closing statement

We would like to thank you for your time and for agreeing to participate in the study. We will send you a summary of the findings once the study is completed.

APPENDIX III: QUESTIONNAIRES AND INTERVIEW GUIDES FOR STUDY III

PARENTS QUESTIONNAIRE

Please answer the following questions about your child who was born with a heart defect	
Date of birth	_____ DD/MM/YYYY
Sex	☐ Female ☐ Male ☐ Other
Have any of these developmental domains been a concern to you while your child was growing up? Please select all that apply and indicate the approximate age at which your child began experiencing difficulties in the domain	☐ Gross motor skills (e.g., running, kicking, jumping, riding a bicycle) ____ years old ☐ Fine motors skills (e.g., manipulating small objects, tying shoes, doing zippers) ____ years old ☐ Behavior (e.g., attention problems, anxiety, aggressive, impulsive behavior) ____ years old ☐ Cognition (e.g., concepts, memory, problem-solving) ____ years old ☐ Language (e.g., speaking, understanding) ____ years old ☐ School performance (e.g., writing, reading, mathematics) ____ years old ☐ Social relationships ____ years old ☐ Other: _____ ____ years old

Please answer the following questions about your family demographics	
First 3 characters of your postal code	__ __ __
Parent 1: highest level of education completed	☐ Elementary school (6th grade) or less ☐ Partial high school ☐ High school completed ☐ CEGEP, College certification, or technical program completed ☐ University graduation or standard 4-year college (undergraduate degree) ☐ Graduate school (graduate degree)

Please specify Parent 1's relationship with the child	□ Mother □ Father □ Other, please specify _____
Parent 2 highest level of education completed	□ Elementary school (6th grade) or less □ Partial high school □ High school completed □ CEGEP, College certification, or technical program completed □ University graduation or standard 4-year college (undergraduate degree) □ Graduate school (graduate degree)
Please specify Parent's 2 relationship with the child	□ Mother □ Father □ Other, please specify _____
Total Family Income	□ Under 20 000$ □ 20 000$ to 39 999$ □ 40 000$ to 59 999$ □ 60 000$ to 79 999$ □ 80 000$ to 99 999$ □ Above 100 000$ □ Prefer not to answer

SEMI-STRUCTURED INTERVIEW GUIDE – PARENT VERSION - ENGLISH

Interview

- The main questions are in black and the optional follow up questions in blue.
- Notes for the interviewer are in grey

Introduction

My name is Marie-Eve Bolduc, I am a PhD student in rehabilitation at McGill University. I am also an occupational therapist.

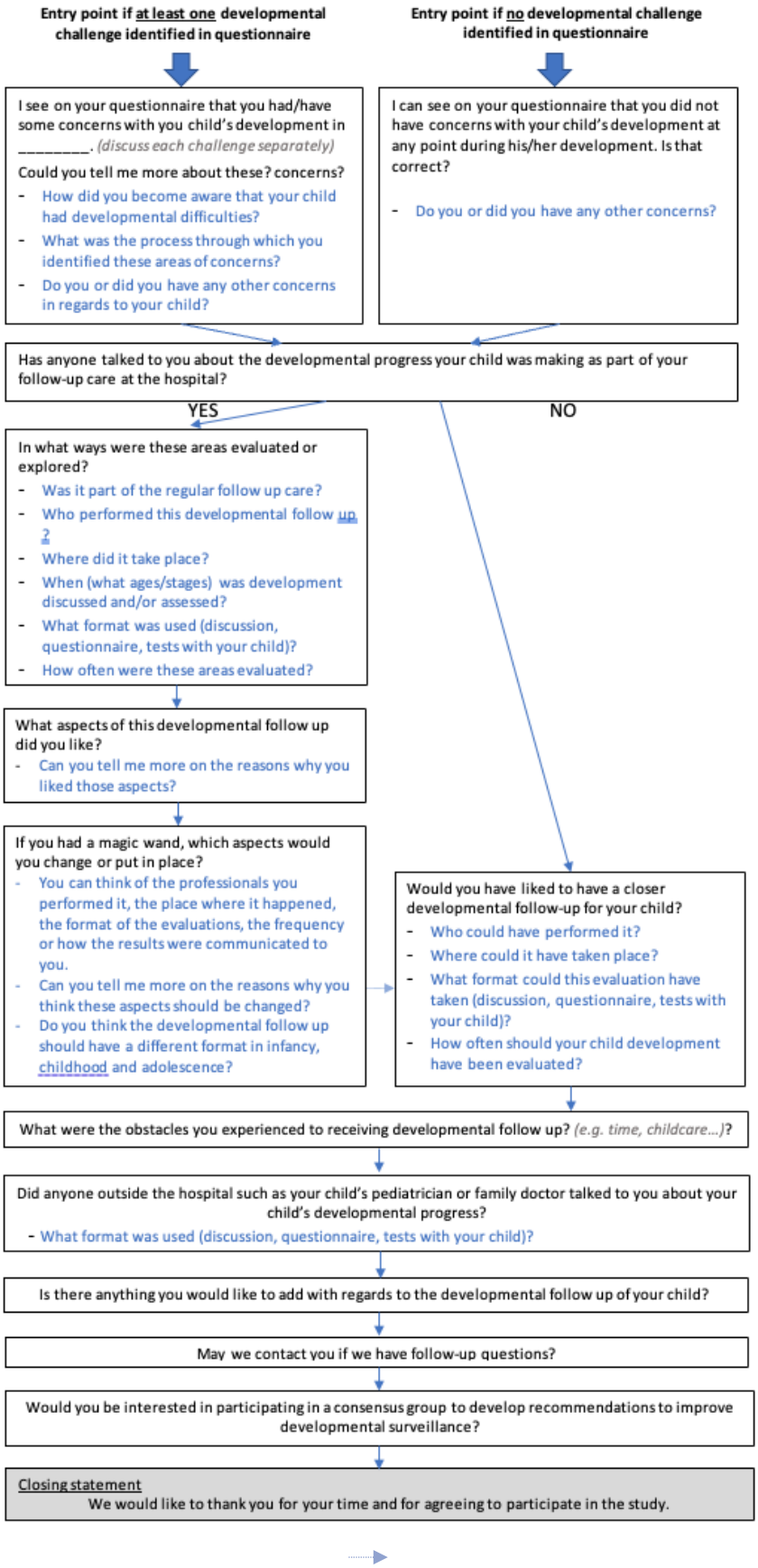
I would first like to thank you for accepting to participate in this interview. We are hoping that the results of this study will improve the developmental follow-up of children and adolescents with CHD across Canada.

During this interview, I will ask you questions about the developmental challenges your child has experienced and about the developmental follow up you have received. The interview will last approximately 45 minutes to one hour.

All your answers will remain confidential. I would like to take notes and record the interview to allow me to go back to your answers when I analyze the data. Is that ok with you?

Do you have any questions before we beginning?

On the questionnaire that you filled up before this interview, we asked you to identify the developmental challenges that your child may have experienced. I would first like to ask you if you were aware that these challenges could be associated with your child's CHD?


Entry point if at least one developmental challenge identified in questionnaire
Entry point if no developmental challenge identified in questionnaire
I see on your questionnaire that you had/have some concerns with you child's development in _____. (discuss each challenge separately)
Could you tell me more about these? concerns?
- How did you become aware that your child had developmental difficulties?
- What was the process through which you identified these areas of concerns?
- Do you or did you have any other concerns in regards to your child?
I can see on your questionnaire that you did not have concerns with your child's development at any point during his/her development. Is that correct?
- Do you or did you have any other concerns?
Has anyone talked to you about the developmental progress your child was making as part of your follow-up care at the hospital?
YES
NO
In what ways were these areas evaluated or explored?
- Was it part of the regular follow up care?
- Who performed this developmental follow up ?
- Where did it take place?
- When (what ages/stages) was development discussed and/or assessed?
- What format was used (discussion, questionnaire, tests with your child)?
- How often were these areas evaluated?
What aspects of this developmental follow up did you like?
- Can you tell me more on the reasons why you liked those aspects?
If you had a magic wand, which aspects would you change or put in place?
- You can think of the professionals you performed it, the place where it happened, the format of the evaluations, the frequency or how the results were communicated to you.
- Can you tell me more on the reasons why you think these aspects should be changed?
- Do you think the developmental follow up should have a different format in infancy, childhood and adolescence?
Would you have liked to have a closer developmental follow-up for your child?
- Who could have performed it?
- Where could it have taken place?
- What format could this evaluation have taken (discussion, questionnaire, tests with your child)?
- How often should your child development have been evaluated?
What were the obstacles you experienced to receiving developmental follow up? (e.g. time, childcare…)?
Did anyone outside the hospital such as your child's pediatrician or family doctor talked to you about your child's developmental progress?
- What format was used (discussion, questionnaire, tests with your child)?
Is there anything you would like to add with regards to the developmental follow up of your child?
May we contact you if we have follow-up questions?
Would you be interested in participating in a consensus group to develop recommendations to improve developmental surveillance?
Closing statement
We would like to thank you for your time and for agreeing to participate in the study.

YOUTH QUESTIONNAIRE

Please answer the following questions about you	
Date of birth	_____ DD/MM/YYYY
Sex	☐ Female ☐ Male ☐ Other
Have you experienced difficulties in any of these developmental domains when growing up? Please select all that apply and indicate the approximate age at which you began to experience these difficulties	☐ Gross motor skills (e.g., running, kicking, jumping, riding a bicycle) ____ years old ☐ Fine motors skills (e.g., manipulating small objects, tying shoes, doing zippers) ____ years old ☐ Behavior (e.g., attention problems, anxiety, aggressive, impulsive behavior) ____ years old ☐ Cognition (e.g., concepts, memory, problem-solving) ____ years old ☐ Language (e.g., speaking, understanding) ____ years old ☐ School performance (e.g., writing, reading, mathematics) ____ years old ☐ Social relationships ____ years old ☐ Other: _____ ____ years old

Please answer the following questions about your family demographics	
First 3 characters of your postal code	_____
Parent 1: highest level of education completed	☐ Elementary school (6th grade) or less ☐ Partial high school ☐ High school completed ☐ CEGEP, College certification, or technical program completed ☐ University graduation or standard 4-year college (undergraduate degree) ☐ Graduate school (graduate degree)

Please specify Parent 1's relationship with you	□ Mother □ Father □ Other, please specify _____
Parent 2 highest level of education completed	□ Elementary school (6th grade) or less □ Partial high school □ High school completed □ CEGEP, College certification, or technical program completed □ University graduation or standard 4-year college (undergraduate degree) □ Graduate school (graduate degree)
Please specify Parent's 2 relationship with you	□ Mother □ Father □ Other, please specify _____
Total Family Income	□ Under 20 000$ □ 20 000$ to 39 999$ □ 40 000$ to 59 999$ □ 60 000$ to 79 999$ □ 80 000$ to 99 999$ □ Above 100 000$ □ Prefer not to answer

STUDY III: SEMI-STRUCTURED INTERVIEW GUIDE – YOUTH VERSION - ENGLISH

Interview

- The main questions are in black and the optional follow up questions in blue.
- Notes for the interviewer are in grey

Introduction

My name is Marie-Eve Bolduc, I am a PhD student in rehabilitation at McGill University. I am also an occupational therapist.

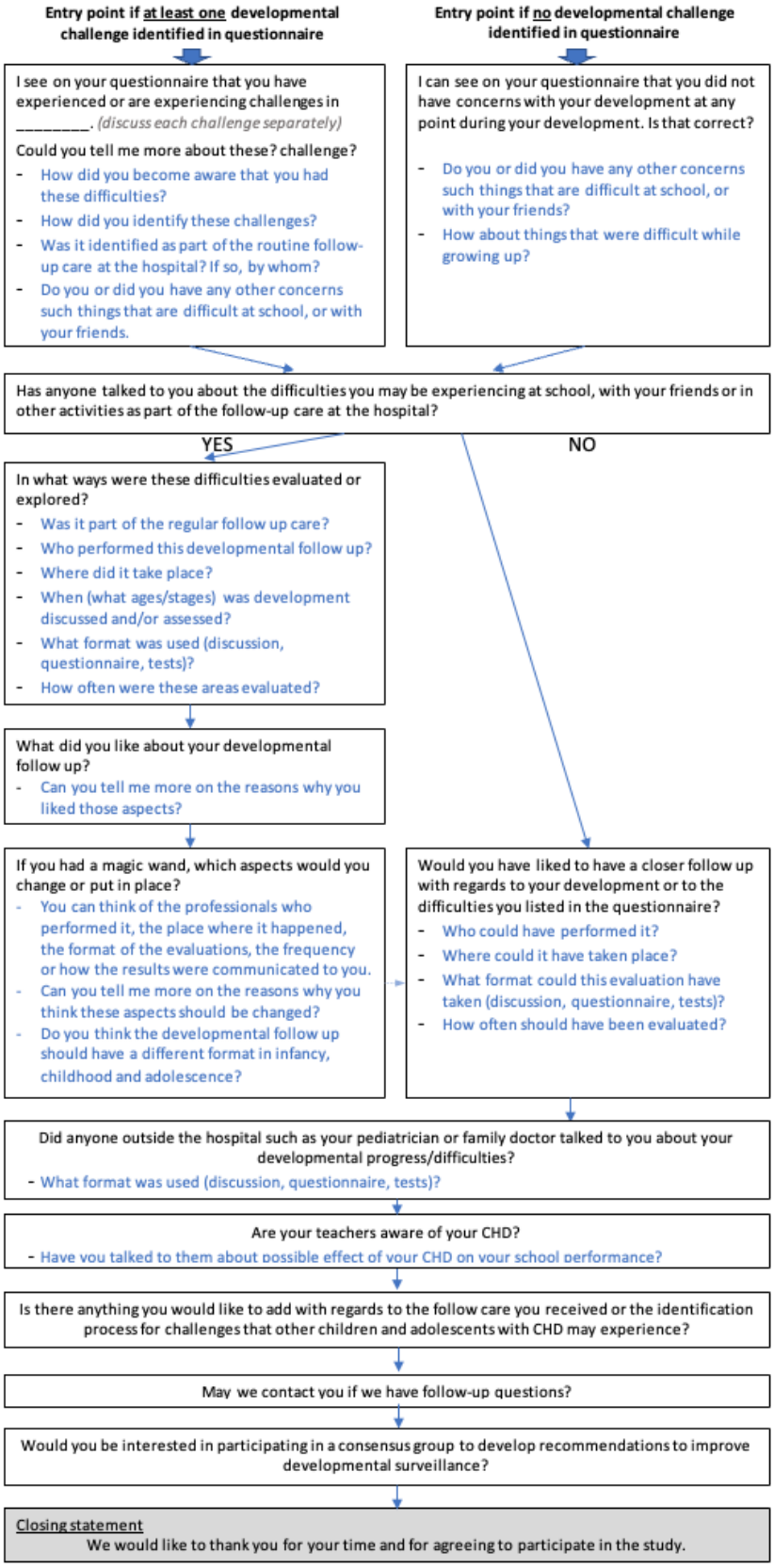
I would first like to thank you for accepting to participate in this interview. We are hoping that the results of this study will improve the developmental follow-up of children and adolescents with CHD across Canada.

During this interview, I will ask you questions about the difficulties you may have experienced growing up or that you experience at home, at school or with your friends and about the developmental follow up you have received with regards to these challenges. The interview will last 45 minutes to one hour.

All your answers will remain confidential. I would like to take notes and record the interview to allow me to go back to your answers when I analyze the data. Is that ok with you?

Do you have any questions before we begin?

On the questionnaire that you filled up before this interview, we asked you to identify challenges that you may have at home, at school or with your friends. I would first like to ask you if you were aware that these challenges could be associated with your CHD?


Entry point if at least one developmental challenge identified in questionnaire
Entry point if no developmental challenge identified in questionnaire
I see on your questionnaire that you have experienced or are experiencing challenges in _____. (discuss each challenge separately)
Could you tell me more about these? challenge?
- How did you become aware that you had these difficulties?
- How did you identify these challenges?
- Was it identified as part of the routine follow-up care at the hospital? If so, by whom?
- Do you or did you have any other concerns such things that are difficult at school, or with your friends.
I can see on your questionnaire that you did not have concerns with your development at any point during your development. Is that correct?
- Do you or did you have any other concerns such things that are difficult at school, or with your friends?
- How about things that were difficult while growing up?
Has anyone talked to you about the difficulties you may be experiencing at school, with your friends or in other activities as part of the follow-up care at the hospital?
YES
NO
In what ways were these difficulties evaluated or explored?
- Was it part of the regular follow up care?
- Who performed this developmental follow up?
- Where did it take place?
- When (what ages/stages) was development discussed and/or assessed?
- What format was used (discussion, questionnaire, tests)?
- How often were these areas evaluated?
What did you like about your developmental follow up?
- Can you tell me more on the reasons why you liked those aspects?
If you had a magic wand, which aspects would you change or put in place?
- You can think of the professionals who performed it, the place where it happened, the format of the evaluations, the frequency or how the results were communicated to you.
- Can you tell me more on the reasons why you think these aspects should be changed?
- Do you think the developmental follow up should have a different format in infancy, childhood and adolescence?
Would you have liked to have a closer follow up with regards to your development or to the difficulties you listed in the questionnaire?
- Who could have performed it?
- Where could it have taken place?
- What format could this evaluation have taken (discussion, questionnaire, tests)?
- How often should have been evaluated?
Did anyone outside the hospital such as your pediatrician or family doctor talked to you about your developmental progress/difficulties?
- What format was used (discussion, questionnaire, tests)?
Are your teachers aware of your CHD?
- Have you talked to them about possible effect of your CHD on your school performance?
Is there anything you would like to add with regards to the follow care you received or the identification process for challenges that other children and adolescents with CHD may experience?
May we contact you if we have follow-up questions?
Would you be interested in participating in a consensus group to develop recommendations to improve developmental surveillance?
Closing statement
We would like to thank you for your time and for agreeing to participate in the study.

APPENDIX IV: RECRUITMENT DOCUMENTS FOR STUDY III

ELECTRONIC FLYER - ENGLISH

IMPROVING THE DEVELOPMENTAL FOLLOW-UP OF CHILDREN AND ADOLESCENTS WITH CONGENITAL HEART DEFECTS

We are currently conducting a research study to define what families would most prefer in terms of developmental follow-up of children and adolescents with heart defects.

WHO CAN PARTICIPATE?

- Youth and young adults (13-25 years old) born with a congenital heart defect who had open-heart surgery before 2 years of age and who received health services in Canada since their surgery.
- Parents of children aged 5-15 years with a congenital heart defect who had open-heart surgery before 2 years of age and who receive health services in Canada since their surgery.

WHAT DOES IT INVOLVE?

- An interview that will last approximately 60 minutes.
- The interview can take place at the hospital, at your home or any other location that may be convenient for you. It could also take place by phone or online call (e.g. Skype).

If you would like to participate or have questions

Contact us at marie-eve.bolduc@mcgill.ca or leave us a message at 514-398-8143.

ELECTRONIC FLYER - FRENCH

AMÉLIORER LE SUIVI DÉVELOPPEMENTALE DES ENFANTS ET ADOLESCENTS NÉS AVEC UNE MALFORMATION CARDIAQUE

Nous menons actuellement une étude pour définir ce que les familles préféreraient en termes de suivi développemental des enfants et des adolescents atteints de malformations cardiaques.

QUI PEUT PARTICIPER?

- Jeunes et jeunes adultes (13-25 ans) nés avec une malformation cardiaque, ayant subi une intervention chirurgicale à cœur ouvert avant l'âge de 2 ans et ayant reçu des services de santé au Canada depuis leur chirurgie.
- Parents d'enfants âgés de 5 à 15 ans présentant une malformation cardiaque, ayant subi une opération à cœur ouvert avant l'âge de 2 ans et ayant reçu des services de santé au Canada depuis leur chirurgie.

CE QUE L'ÉTUDE IMPLIQUE

- Une entrevue d'une durée approximative de 60 minutes.
- L'entretien peut avoir lieu à l'hôpital, chez vous ou à tout autre endroit qui vous convient. Cela pourrait également se faire par téléphone ou par appel en ligne (ex. Skype).

Si vous aimeriez participer ou si vous avez des questions

Contactez-nous à marie-eve.bolduc@mcgill.ca
ou laissez-nous un message au 514-398-8143.

PAPER FLYER - BILINGUAL

LE BUT DE NOTRE ÉTUDE

Les enfants atteints de cardiopathie congénitale bénéficient désormais d'interventions chirurgicales précoces et vivre pleinement. Cependant, certains enfants présentent des retards de développement.

Ces difficultés peuvent être mieux traitées si elles sont identifiées le plus tôt possible. Cependant, les besoins et les préférences des enfants et des jeunes atteints de cardiopathie congénitale ainsi que de leurs familles en ce qui concerne les meilleurs moyens de surveiller leur développement n'ont pas été identifiés.

Ce projet vise à définir ce que les familles préféreraient le plus en termes de surveillance développementale des enfants et des adolescents atteints de malformations cardiaques.

QUI PEUT PARTICIPER ?

- Nous recherchons des jeunes (13-21 ans) nés avec une malformation cardiaque congénitale, ayant subi une intervention chirurgicale à cœur ouvert avant l'âge de 2 ans et qui reçoivent des services de santé au Canada depuis leur naissance.
- Nous recherchons également des parents d'enfants âgés de 5 à 15 ans présentant une cardiopathie congénitale, ayant subi une opération à cœur ouvert avant l'âge de 2 ans et qui reçoivent des services de santé au Canada depuis leur naissance.

EN QUOI CONSISTE MA PARTICIPATION?

- Tous les participants sont invités à un entretien d'une durée approximative de 60 minutes.
- L'entretien peut avoir lieu à l'hôpital, chez vous ou à tout autre endroit qui vous convient. Cela pourrait également se faire par téléphone ou par appel en ligne (ex. Skype).

EST-CE QUE NOUS OFFRONS UNE COMPENSATION?

Oui. Si vous décidez de participer à cette étude, vous recevrez une compensation financière pour votre temps et votre déplacement.

SI VOUS ÊTES INTÉRESSÉ(E) À PARTICIPER OU VOULEZ PLUS D'INFORMATION

Si vous avez des questions ou souhaitez plus d'informations sur cette étude. Vous pouvez nous faire parvenir un courriel au marie-eve.bolduc@mcgill.ca ou nous laisser un message au 514-398-8143.

Improving the Developmental Surveillance of Children and Adolescents with Congenital Heart Defects

Contact us

✉ **marie-eve.bolduc@mcgill.ca**

abcdresearch
ADVANCES IN BRAIN & CHILD DEVELOPMENT RESEARCH LABORATORY

Research Institute of the MUHC

AIM OF OUR STUDY

Children with a congenital heart defect now benefit from early surgical repair and can live a full life. However, some children do experience some developmental delays.

These difficulties can be better addressed if identified as early as possible. However, the needs and preferences of children and youth with a congenital heart defect as well as their families with regards to the best ways to monitor their development have not been identified.

This project aims to define what families would most prefer in terms of developmental surveillance of children and adolescents with heart defects.

WHO CAN PARTICIPATE?

- We are looking for youth (13-21 years old) born with a congenital heart defect who had open-heart surgery before 2 years of age and who have been receiving health services in Canada since birth.
- We are also looking for parents of children aged 5-15 years with a congenital heart defect who had open-heart surgery before 2 years of age and who have been receiving health services in Canada since birth.

WHAT DOES YOUR PARTICIPATION INVOLVE?

- All participants are invited for an interview that will last approximately 60 minutes.
- The interview can take place at the hospital, at your home or any other location that may be convenient for you. It could also take place by phone or online call (e.g. Skype).

DO WE OFFER A COMPENSATION?

Yes. If you choose to participate in this study, you will receive a financial compensation for your time and transportation.

IF YOU ARE INTERESTED IN PARTICIPATING OR NEED MORE INFORMATION

If you have questions or would like more information on this study. You can e-mail us at marie-eve.bolduc@mcgill.ca or leave us a message at 514-398-8143.

Améliorer la Surveillance Développementale des Enfants et Adolescents Nés avec une Malformation Cardiaque

Contactez-nous

✉ **marie-eve.bolduc@mcgill.ca**

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ADVANCES IN BRAIN & CHILD DEVELOPMENT RESEARCH LABORATORY
Institut de Recherche du CUSM