Neurodevelopmental Outcomes of the Child with Congenital Heart Disease

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KEYWORDS

• Congenital heart disease • Development • Impairment • Outcome • School-age
• Genetic disorder • Examination

KEY POINTS

- Noncardiac complications and comorbidities play a critical role for lifelong quality of life and academic achievement in children born with congenital heart disease (CHD).
- A significant proportion of children with CHD manifest neurodevelopmental impairments, affecting cognition, motor and language development, and higher-order cognitive functions.
- Impairments are often of mild to moderate severity; however, they may occur in conjunction and significantly affect school performance, integration, and academic achievement.
- Developmental problems seem not to diminish, but rather become more apparent as demands on the developing child, in particular the school-age child, increase.
- Major risk factors for impaired development are genetic disorders, delayed preoperative brain development, a complicated postoperative course, and a poor socioeconomic environment.

INTRODUCTION

Moderate and severe congenital heart disease (CHD) occurs in about 6 per 1000 live births[1] and constitutes the most frequent congenital malformation in childhood. Around half of all newborns with a CHD require immediate surgical intervention to survive. With dramatic improvements in neonatal, perioperative care, and surgical management, survival rates have increased and reached rates of 80% to 90% even for the most complex forms of CHD.[2] In addition, cardiac outcome is often good or even excellent, allowing these children to survive into adulthood. This has led to the

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situation where more adults are alive now with CHD than children.[3] With this development from a childhood disease to an adult disease, it is critical to address potential cardiac and noncardiac complications. Restrictions in cardiac function, but also impairments in cognitive and academic performance, are associated with lifelong individual psychological and societal financial burdens.[4] The description of noncardiac consequences, such as neuropsychological and psychoemotional problems, becomes even more critical in this view. Only with this information can clinicians sensitize primary care providers and specialists, inform patients and parents, develop programs for early detection, and ultimately develop therapeutic strategies to reduce or prevent these sequelae.

This article addresses the spectrum, severity, and evolution of potential neurodevelopmental and neuropsychological problems associated with CHD. Furthermore, in every chronic childhood disorder including CHD, child and parental quality of life are affected. It is beyond the focus of this article to also cover this important topic, but see referenced review articles.[5,6]

GENETIC COMORBIDITY

When discussing neurodevelopmental impairments in children with CHD, one has to consider that about one-third of all newborns and children operated for CHD have an underlying genetic disorder.[7] The most frequent disorders include trisomy 21 syndrome, 22q11 microdeletion syndrome, and CHARGE syndrome. With more sophisticated and readily available genetic screening options, the proportion of children born with CHD who are diagnosed with a genetic comorbidity will likely increase.[7] Thus, when describing the neurodevelopmental outcome in association with CHD, keep in mind that outcome in children with a genetic comorbidity is even poorer[8,9] and is strongly determined by that specific genetic aberration. There is still a large variability in outcome (eg, intelligence quotient [IQ]) within a given genetic disorder, but the IQ distribution is significantly shifted even more leftward than for CHD alone. Importantly, these children are being operated for their CHD, but often are faced with a variety of noncardiac medical problems, such as leukemia or immunodeficiency, and hearing and visual impairments. Therefore, these children need particular attention and an interdisciplinary approach that cannot be provided by the cardiologist or primary care provider alone. In addition, when following children with CHD, the diagnosis or suspicion of a genetic disorder may only arise during follow-up after children show poorer development progress with lower IQ scores than expected or explained by the CHD or the perioperative course. In combination with subtle facial or other minor dysmorphisms or other organ disorders, evaluation for a genetic disorder must then be initiated. Often a diagnosis of a genetic disorder is then made, which is essential in the view of prognosis, family counseling, and search for other organ malfunctions.

Fig. 1 shows a schematic overview of IQ outcome curves compared with norms in children with CHD and in those with genetic comorbidities. Importantly, the graph illustrates the significant overlap of performance despite differing mean IQs between these groups and despite higher proportions of children in each category performing below clinically relevant cut-off values, such as an IQ of 70 or 85 (-2 standard deviation and -1 standard deviation of the mean, respectively). This also translates into clinical experience: children with CHD may perform well within the norm or even in a high-normal level. In addition, clinical parameters during the neonatal period and surgical treatment variables very rarely help to predict outcome in an individual child. Little is still known about the factors that lead to a poor performance in one child and to