National Pediatric Cardiology
Quality Improvement Collaborative

Research Explained

Six-Year Neurodevelopmental Outcomes for Children with Single-Ventricle Physiology

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ABOUT THIS STUDY

The goal of this study is to describe the patterns of neurodevelopmental problems in children with hypoplastic left heart syndrome (HLHS) and other complex congenital heart defects (cCHDs) over the first years of childhood and to determine which early tests and evaluations can predict behavior problems at 6 years of age

Why is this study important?

- Neurodevelopmental deficits are the most common long-term complication for children with HLHS and other cCHDs.
- Neurodevelopmental deficits can include learning disabilities, attention deficit disorders, and other difficulties with learning, conduct, and behavior.
- Neurodevelopmental deficits can impact a person's ability succeed in school and employment. They can make the quality of life worse for the child and their family.
- This study of young children (6 or younger) with single ventricle defects like HLHS describes patterns in how neurodevelopmental deficits are diagnosed during a child's first years of childhood and how common they are in children with these heart conditions. This study also determines if testing for these deficits at a young age can predict learning, emotional, and behavioral difficulties later in childhood.

How was this study performed?

- This study used information from a large multi-center clinical trial called the Pediatric Heart Network Single Ventricle Reconstruction (SVR) Trial and a later follow-up study (SVR-II).
- The SVR Trial compared outcomes for children who had either of 2 different types of Norwood procedure, and included neurodevelopmental testing at 14 months and 3 years of age.
- The SVR-II study used two developmental skills questionnaires (BASC-2 and ASQ) and conducted interviews of the parents of the children who participated in the original SVR Trial.
- The BASC-2 questionnaire collected information about the child's behavior including anxiety, depression, hyperactivity, problem behaviors, communication skills, and daily living activities. The ASQ parent questionnaire collected information about the child's communication skills, motor skills, problem-solving skills, personal and social skills.

What were the results of the study?

- 291 patients who completed both the SVR Trial and SVR-II were included in this study.
- Parents were more likely to report behavior problems in their children at age 6 years than at younger ages.
- The biggest differences in children in this study compared to other children were skills for communication, and for completion of daily activities (adaptive function skills).
- As children in this study got older, more parents reported that their child had difficulties with communication and completion of daily activities (adaptive function skills).
- At the age of 6 years, one-third of children (29%) in this study had difficulties with communication and completion of daily activities (adaptive function skills).
- As children in this study got older, more parents reported problems with hyperactivity and aggression.
- Parent-reported problems with anxiety and depression in their child were within normal ranges for these age groups.
- In the children in this study, the BSID-II test at 14 months of age did not reliably predict adaptive function skills at age 6.
- Children in this study who had poorer scores on the problem-solving and communication sections of the ASQ parent questionnaire were more likely to have problems with adaptive function skills at age 6.

- As children grew older in this study, the ASQ parent questionnaire's ability to predict difficulties with problem solving and adaptive functioning improved significantly.

What were the limitations of the study?

- The children who completed this study came from wealthier families, were less likely to have a genetic syndrome, and tended to have a higher score on the BSID-II test at 14 months of age.
- Most of the information collected for this study was reported by parents instead of through in-person evaluations.
- This study may be under-reporting the degree of neurodevelopmental deficits in children with single ventricle heart disease.

What it all means

- In this study, early problems in problem-solving and communication were strongly related to poor adaptive function skills at 6 years of age.
- Specifically, in this study, early fine-motor skill development was important in the development of adaptive function skills at an older age.
- Many children who had difficulties with adaptive function skills at 6 years of age were not identified on earlier neurodevelopmental assessments.
- For children with congenital heart disease, it is important to have repeated neurodevelopmental evaluations throughout their lifespan because new problems may be detected as a child ages and when school and social life becomes more complicated.