

Research Explained

Key Findings: Factors Associated with Neurodevelopment for Children with Single Ventricle Lesions

The *Journal of Pediatrics* published a study in September, 2014: "[Factors Associated with Neurodevelopment for Children with Single Ventricle Lesions](#)".¹

The NPC-QIC Research and Publication Committee reviewed this article and a summary of the findings can be found below.

Main Finding from this Study:

While there has been great improvement in survival for patients with hypoplastic left heart syndrome (HLHS) and other similar single ventricle lesions that require the Norwood procedure, these patients have a higher prevalence of developmental and behavioral abnormalities. Many studies on neurodevelopmental outcomes in this high-risk group of children have been from a single center and/or describe outcomes in infants or school-aged children. Little is known about the assessment of development in children with single ventricle at preschool age.

This study describes neurodevelopmental outcomes at 3 years of age in children with HLHS and other forms of single ventricle who previously participated in the multicenter Single Ventricle Reconstruction Trial, a comparison of two groups who were randomly assigned to get one shunt type or the other at the time of their Norwood surgery. The authors of this study concluded that on average, children with HLHS or single right-ventricle have impaired neurodevelopment at 3 years of age compared to the general population. Lower scores are unrelated to their shunt type. Delay was detected in at least one area of development for 51% of the children.

About this study:

- **Why is this study important?**

This is a very important study because this is a very large group of patients with this heart defect (more than 200) from many different medical centers about whom we have a lot of very good baseline information from the initial Single Ventricle Reconstruction Trial (SVR-I) done when these patients were infants. While infants with low scores on earlier developmental testing at 14 months of age were more likely to have low scores at 3 years of age, their analysis suggests that even when 14-month test scores were in the normal range, children with single ventricle may be at risk for developmental impairment at preschool age and beyond. It will be very important to continue to follow them over time, even for their entire lives.

- **How was this study performed?**

This study invited parents/guardians of all patients who had been previously enrolled in the Single Ventricle Reconstruction trial who survived without heart transplantation to complete annual questionnaires on the child's development, behavior, quality of life, and functional status. Information about interim medical history was also collected. Development at 3 years of age was measured with the parent-completed Ages and Stages Questionnaire (ASQ). Behavior was measured with the Parent-Report Behavior Assessment System for Children (BASC). Parents also completed the Pediatric Quality of Life Inventory and the Functional Status questionnaires. The main comparison was between scores in the single ventricle group and the general population scores. In addition, they looked at differences in scores between the two types of surgical shunts and tried to identify any factors that were more common in patients with the greatest developmental delays.

- **What were the results of the research?**

- 203 completed the Ages and Stages Questionnaire (ASQ)
- At 3 years of age, average scores for each area of development measured on the ASQ questionnaire were significantly lower than those for the general population at the same age.
 - The percentage of patients with single ventricle with delayed scores was 20% for the Communication scale, 30% for the Gross Motor scale, 35% for the Fine Motor scale, 24% for the Problem Solving scale, and 17% for the Personal-Social scale.
 - Delay was detected in at least one area for 51% of the group.
 - Scores did not differ by type of shunt.
- Infants with low scores on earlier developmental testing at 14 months of age were more likely to have low scores at 3 years of age, however, even when 14 month scores were within the normal range, some children were still at risk for developmental delay.
- Children with more complications and longer hospitalization after Norwood surgery, hearing or vision problems, and those with persistent feeding problems at 2 years of age were more likely to have lower developmental scores.

- **What are the limitations of this study?**

Developmental and behavioral questionnaires were not completed for all eligible children in the study. The study also relied largely on parental report.

- **What are the takeaway messages considering the results and limitations of this study?**

Children with HLHS or other single right-ventricle anomalies are at increased risk for developmental impairments at 3 years of age. While there are some factors associated with increased risk for impairment, many children without these risk factors develop impairments, and thus monitoring is important for everyone. Development in these children should continue to be monitored throughout infancy and childhood to improve recognition of delays and potential for services to improve outcomes.

¹Goldberg CS, Lu M, Sleeper LA, Mahle WT, Gaynor JW, Williams IA, Mussatto KA, Ohye RG, Graham EM, Frank DU, Jacobs JP, Krawczeski C, Lambert L, Lewis A, Pemberton VL, Sananes R, Sood E, Wechsler SB, Bellinger DC, Newburger JW; Pediatric Heart Network Investigators. Factors associated with neurodevelopment for children with single ventricle lesions. *J Pediatr.* 2014;165(3):490-496.