Overestimating Neuroprotection in Congenital Heart Disease: Problems With Bayley III Outcomes

AUTHOR: Michael E. Msall, MD

Kennedy Research Center on Intellectual and Developmental Disabilities, Section of Developmental and Behavioral Pediatrics, University of Chicago Comer Children's Hospital, Chicago, Illinois

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Address correspondence to Michael E. Msall, MD, Kennedy Research Center on Intellectual and Developmental Disabilities, Section of Developmental and Behavioral Pediatrics, University of Chicago Comer Children's Hospital, 950 E 61st St, SSC Room 207, Chicago, IL 60637. E-mail: mmsall@peds.bsd.uchicago.edu

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Major advances in the care of children with congenital heart disease include prenatal diagnosis, prompt and often prenatal referral to pediatric cardiology centers of excellence, early stabilization of abnormal physiology with single-stage corrective repair before 6 weeks of age, and attention to medical and environmental stressors that affect the vulnerability of a child's immature central nervous system.¹⁻³

In this issue of *Pediatrics*, Acton et al⁴ describe the developmental outcomes of surviving children from a large prospective Canadian cohort after early congenital surgical heart repair at a regional center of excellence. The children had surgery at a mean age of 14 days and underwent assessments at a mean age of 21 months by using 2 versions of the well-standardized Bayley scales (Bayley II and III).^{5,6} These scales were used to assess gross and fine motor skills and verbal and nonverbal cognitive performance during structured evaluations.

WHAT DID THEY FIND?

First, among these critically ill term neonates with malformations including transposition of the great arteries, hypoplastic left heart, total anomalous pulmonary venous return, tetralogy of Fallot, and interrupted aortic arch, 90% survived. All 168 survivors were assessed in follow-up. However, 9% (14 chromosomal, 1 fetal alcohol syndrome) were excluded from this analysis because their genetic disorders were associated with intellectual disability. Eleven children required extracorporeal membrane oxygenation or cardiac transplantation, which reflected prolonged physiologic instability and difficulty with initial primary repair. When using the Bayley III cognitive and motor scales, disability defined as a standard score of <70 occurred in 2.7%. This score is similar to that in the normative term population without congenital heart disease during the standardization of the Bayley III.

ARE THESE RESULTS AN INDICATOR OF SUBSTANTIALLY IMPROVED NEUROPROTECTION BECAUSE OF EARLY DETECTION, STABILIZATION, AND IMPROVED INTERVENTIONS?

Among the entire cohort, the mean cognitive composite score (CS) was 96, the language CS was 91, and the motor CS was 94. These scores are 0.6 z and 0.3 z below the mean for peers assuming a CS of 100 ± 15. These results indicate that on a population level, developmental outcomes are broadly typical. This outcome is indeed a cause for celebration.

HOW DO THESE RESULTS COMPARE WITH THE PREVIOUS GOLD STANDARD, THE BAYLEY II?

Among 25 children given both the Bayley II and the Bayley III, the mean Bayley II Mental Developmental Index was 86 and the Performance Developmental Index was 83. These group means are 10 cognitive

points and 7 motor points lower than comparable scores on the Bayley III ($P < .001$ and $P < .009$). If one assumes that between the 1993 Bayley II norms and the 2006 Bayley III norms, an increase of 0.3 points occurs per year, then their scores of 86 and 84 should be compared with a mean of 104 on the Bayley II.⁷ These mean scores reflect substantially (>1 SD) poorer performance on cognitive and motor tasks compared with term peers in the normative sample. It is important to note that the Bayley III is overestimating the child's developmental outcome so that "all the children are above average" (see http://en.wikipedia.org/wiki/Lake_Wobegone#The_Lake_Wobegone_effect).

WHAT DOES THIS MEAN?

First, these findings suggest that we exercise caution when using new assessments for measuring early child-

hood developmental outcomes. Second, Acton et al have demonstrated the crucial importance of using both the new tool and the historic standard. In addition, it is also critically important to have a subset of the cohort compared with term controls before the new tool is applied on specific at-risk populations from diverse geographic and cultural regions. Last, it allows us to go beyond early childhood outcomes by using a single standardized score (eg, a Bayley II Mental Developmental Index of 75) and examine the preschool developmental tasks of perception, coordination, communication, problem-solving, behavioral regulation, adaptive skills, executive function, and social interaction, all of which are more substantially related to higher cortical processes that ultimately underlie academic and community success than are early childhood Bayley scores.^{8,9}

WHERE DO WE GO NEXT?

In an era of scarce resources we must develop collaborative tools for measuring health and developmental outcomes after critical illness, which will allow us to go beyond the limitations of a summary score on the Bayley scales in early childhood. It will allow us to focus instead on the complexity of risk and resiliency with respect to neurodevelopmental processes that are most relevant to learning and school success.¹⁰

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