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EDITORIAL COMMENTARY

Leaving the nest: Facing adulthood after the arterial switch operation

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See related article on pages 2190-9.

The report in this issue of the *Journal* by Heinrichs and colleagues¹ from Aachen is an important companion study

to the Boston Circulatory Arrest Study (BCAS). A deeper understanding of each is enabled by reviewing the similarities and differences between them.

Let's start with the similarities. Both groups studied neonates undergoing the arterial switch during a similar era (BCAS, 1988-1992; Aachen, 1986-1992), and both groups have performed detailed neurodevelopmental follow-up testing in addition to neuroanatomic imaging with magnetic resonance imaging (MRI).^{1,2} Both groups have reevaluated these patients periodically (ages 4-5, 8-10, and 16-17 years) and have reported significant neurologic impairment across similar domains, primarily in motor skills, language function, and sensory processing skills.

There are, however, some important methodologic differences to note. The BCAS examined the effects of low-flow cardiopulmonary bypass compared to deep

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hypothermic circulatory arrest (DHCA),² whereas the Aachen group performed all operations under DHCA, averaging 60 minutes very consistently (± 3.3 minutes).¹

Because all Aachen patients underwent a relatively long and uniform period of DHCA (60.2 ± 3.3 minutes), the contribution of DHCA duration to outcomes could not be assessed.¹ In the BCAS, this comparison was central to the study, and patients exposed to significantly shorter duration of DHCA (the low-flow cardiopulmonary bypass group) scored significantly better on tests of motor function, speech, visual motor tracking, and auditory analysis than did children randomly assigned to undergo DHCA, at least until the age of 8 years.² In both the Aachen and the BCAS data, these discrete domain differences were less distinct at the age of 16 years, although problems remained.^{1,2}

At adolescence, most of the patients in the BCAS group (65%) had received remedial academic or behavioral services, and scores on a battery of neurocognitive tests tended to be lower than those of the normative population.¹ Likewise, in the current report from Aachen of Heinrichs and colleagues,¹ the average results of full-scale IQ testing were not different from population norms; however, 5 times as many patients scored 2 SD below the norm (11% vs 2%). Furthermore, although Heinrichs and colleagues¹ reported that 90% of their patients were neurologically “normal,” 10% were not, and some of these demonstrated devastating neurologic injury.

These data, and the changes seen with time, are open to interpretation. It may be that by the time these children reach adolescence, they may have acquired (or been taught) a repertoire of coping and behavioral skills that allow them to compensate partially for the underlying deficits that were identified earlier. Alternatively, it may simply be a matter of time needed for biologic recovery from the injury.

In addition to neurodevelopmental testing, both groups have also used MRI to evaluate neuroanatomic correlates with functional outcomes. In this case, the methodologic differences in MRI image acquisition between the 2 studies may provide some additional insight into these patients.

In the study of Heinrichs and colleagues,¹ the Aachen group used a more powerful magnet (3 T), which may provide greater anatomic detail than the 1.5-T MRI used in the BCAS. Whereas the BCAS identified MRI abnormalities in 33% of adolescents,² the Aachen group reported that 66% of patients demonstrated focal white matter injury, 25% reduced brain volume, and 5% “marked” encephalomalacia.¹ They were also able to identify a correlation between the MRI findings and neurocognitive deficits, something the BCAS did not do.

Although it used a weaker 1.5-T magnet, the BCAS used the technique of diffusion tensor imaging, which revealed unsuspected and wide spread microstructural white matter injury not apparent on conventional MRI sequences reported by the Aachen group, suggesting that this brain injury may be related to the deficits that these patients exhibit.³

Finally, consistent with a large body of literature, both groups did report that socioeconomic status did correlate with neurocognitive outcomes. Although socioeconomic status did not cause MRI abnormalities, and genetic factors are almost certainly involved, one should not discount the possibility that the availability and application of early interventions designed to improve function across the affected domains might be very helpful.

In summary, the methodologic differences between the 2 studies provide complementary information, the sum of which is a more powerful than that obtained from either alone. These similar findings between 2 unrelated studies suggest the occurrence of a fundamental biologic phenomenon, one we are just beginning to characterize. Unfortunately, even sophisticated characterizations do not provide an etiology. A panoply of possibilities have been proposed, including exposure of the fetal brain to hypoxemic conditions, altered fetal blood flow patterns, and inherent brain immaturity associated with congenital heart disease. What has become clearer with increasing clinical experience is that our attempts to adopt surgical, anesthetic, and critical care techniques designed to reduce brain injury have met with little success.

Although these efforts will and should continue, a shift in our approach may be warranted. It may be time to consider a strategy than coemphasizes *prevention* and *treatment* of the neurocognitive impairment that has been documented in these studies. This is especially critical as we try to help prepare these young patients to cope with the challenges of adult independence.

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