Caring for the adolescent with surgically repaired D-transposition of the great arteries can be one of the most satisfying experiences in congenital cardiology. The operative mortality for the arterial switch procedure is now <3%. Few of these children require cardiac medications; exercise limitations are minimal; and very rarely do the patients require hospitalization. In many respects, advances in pediatric cardiology and cardiac surgery have succeeded in curing this once lethal condition.

Interestingly, the original hypothesis that a strategy of predominantly low-flow cardiopulmonary bypass compared with predominant hypothermic circulatory arrest might improve neurocognitive outcomes nowadays seems less intriguing. Recent data from the Society of Thoracic Surgeons suggest that the majority of centers have moved away from predominant circulatory arrest support strategies for the arterial switch procedure. In an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database of 548 patients undergoing arterial switch operations with or without ventricular septal defect repair (but without arch reconstruction) in 2010, only 88 of 548 operations (16.1%) used deep hypothermic circulatory arrest. In these cases, the duration of deep hypothermic circulatory arrest was quite short. The median duration of deep hypothermic circulatory arrest was 9.0 minutes (interquartile range, 6.0–18.5 minutes).

Even the analysis of patient- and procedure-related risk factors seems to provide few clues as to how one might change perioperative management to provide neuroprotection in the current era. As noted, extended periods of hypothermic arrest are uncommon in the current surgical management of D-transposition of the great arteries. Thus, postoperative seizures, the strongest predictor of adverse outcome from the BCAS, are also less common. Contemporary data from centers using predominantly deep hypothermic circulatory arrest or predominantly high-flow cardiopulmonary bypass have reported the incidence electroencephalographic seizures to be significantly less than the 19.9% incidence reported in the BCAS.3–5

Rather than the risk factor analysis, the most compelling findings from this study relate to impact of neurocognitive and social impairments on the daily lives of these teenagers. Almost 40% of the subjects in this follow-up study received tutoring for schoolwork, and 12% were taking at least 1 medication for psychiatric disorder. The need for such additional services and therapy relates in large part to the neurodevelopmental signature of adolescents with surgically repaired D-transposition of the great arteries. These children’s performance is characterized by impairment in executive function, cognitive flexibility, visual perceptual skills, and social cognition. There is growing evidence that executive functions continue to develop through late adolescence and into adulthood and are related to important outcomes such as academic achievement.6 Along with information-processing speed, executive improvements facilitate the development of more abstract and efficient processing of language. These skills are particularly critical to debate and persuasion, which themselves are essential for success in higher education and beyond.

It is likely that the very neuropsychological domains that are problematic for adolescents with D-transposition of the great arteries are also essential to successful transition to adulthood, especially to adult self-care. This may explain in part why so many young adults with congenital heart disease fail to navigate the transition to appropriate, regular medical care.7 Identifying and remediating these deficits has been a major focus of the adult transition programs of other chronic childhood diseases.8

Much of our focus in pediatric cardiac care to this point has been centered on strategies to mitigate procedure-related brain injury. The seminal work by the investigators in Boston regarding the optimal hematocrit during cardiopulmonary bypass is such an example.9 However, our understanding of neurodevelopmental impairment has evolved considerably since the inception of the BCAS. It is increasingly recognized that many of the factors that lead to later deficits are outside the control of cardiologists, cardiac surgeons, and perfusionists. These deficits may be hard-wired.10 Therefore, it may be worthwhile to turn our attention to strategies to provide cognitive rehabilitation. Other fields such as pediatric oncology and neurotrauma have begun to explore strategies to enhance the performance of teenage survivors.11,12

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Going forward, the investigators will analyze the impairments described above in the context of diffusion tensor, volumetric, and functional magnetic resonance imaging data. We know that the neural circuitry that joins the prefrontal cortex to posterior cortical and subcortical areas is critically important in executive function. Importantly, data from the preterm population have demonstrated that differential decreases in white matter volumes correlate with impairment in executive function. The impairments reported in the adolescents enrolled in the BCAS represent the logical manifestation of the pattern of early injury that has emerged from neuroimaging in contemporary cohorts of neonates with D-transposition of the great arteries. Our hope is that these neuroanatomical correlates will allow us to work backward and forward to better understand the origin of these deficits and to explore possible therapeutic avenues.

The findings of this study represent a clarion call for ongoing surveillance for neurological impairment in the congenital heart population. For school-aged children and adolescents, measuring IQ alone is not sufficient to provide an accurate and comprehensive understanding of a patient’s functioning in these areas. Acquisition of higher-order planning and organizational skills is needed for success as children enter and progress through middle school and high school. Although it may not be practical to replicate the comprehensive assessment of the BCAS investigators in our routine clinical care, we should supplement the screening performed by schools, paying particular attention to working memory, executive function, and social cognition, to build on the surgical success of procedures such as the arterial switch operation.

Disclosures

None.

References


Boston Circulatory Arrest Study at 16 Years: Handing Over the Keys
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