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Outcomes regarding the central nervous system in children with complex congenital cardiac malformations

Gil Wernovsky

Division of Pediatric Cardiology, The Cardiac Center at The Children's Hospital of Philadelphia, and Department of Pediatrics, University of Pennsylvania School of Medicine, Pennsylvania, United States of America

THE NUMBER OF CHILDREN NOW SURVIVING surgical interventions because of congenital cardiac disease continues to increase. Indeed, it is estimated that, by the end of 2005, there will be more adults than children with congenital cardiac malformations. The complex challenges facing this growing population are only now becoming apparent, including the continued hazard for mortality, the risk of reoperation, and newly-emerging late sequels of surgery. In particular, children with severe lesions requiring intervention in the neonatal period are surviving at levels thought to be highly improbable no more than two decades ago. Heralded by the availability of prostaglandin E_1 in the late 1970s, improvements in technology, such as echocardiography, and color Doppler in particular, intensive care and intraoperative support, have recently allowed surgeons to reconstruct the hearts of many children with otherwise lethal defects. Much of the published literature has focused on the cardiac sequels in this population. Increasing attention has recently been given, nonetheless, to the neurodevelopmental outcomes in this population of patients.

Until recently, clinical reports focused on early postoperative problems, such as stroke and seizures. It is now increasingly recognized that longer term concerns, such as abnormal performance at school, learning disabilities, and behavioral issues, are considerably more prevalent. While perioperative stroke is rare, and postoperative seizures are considerably less prevalent than a decade ago, it is estimated that at least half of the children surviving surgery as neonates and infants have problems when they reach school-age with learning, visual motor integration, and behaviour. Applicability of many of the reports in the literature has been limited by several factors, including the era in which the surgery was performed, the age of the patient at the time of collection of the data, the heterogeneity of the particular congenital cardiac malformation, and variable designs of the studies and the tools used for assessment. Given these limitations, certain consistent themes have arisen across surgical eras and diagnoses. Most studies have reported that while cognitive abilities, or the "intelligence", of these children are generally within the normal range in the absence of confounding cerebral abnormalities or syndromes, reports consistently demonstrate mean values for the group that are significantly less than expected. Importantly, more sophisticated standardized testing has revealed problems in attention, hyperactivity, fine and gross motor control, visual-motor integration and executive functioning, resulting in less than optimal performance at school, and low self-esteem, for these children.

As my colleague Kathy Mussatto and I describe in Chapter 17 of this supplement, abnormal neurodevelopmental outcomes have become the single most prevalent long-term adverse outcome in our patients, more common in many groups of patients than late mortality, reoperation, arrhythmias, or limited capabilities for exercise. While many of the responsible factors are beyond the control of the medical and surgical practitioners, for example, socioeconomic state, and genetics, it is important, if we are further to improve the outcomes for our patients and their families, that we obtain a better understanding of how to protect the brain in the operating room and intensive care unit. The chapters which follow in this section of our supplement focus on the current investigations

Correspondence to: Gil Wernovsky MD, Division of Pediatric Cardiology, Children's Hospital of Philadelphia, 34th Street and Civic Center Boulevard, Philadelphia, PA 19104, USA. Tel: +1 215 590 6067; Fax: +1 215 590 5825; E-mail: Wernovsky@email.chop.edu

in this arena, including work both in the animal laboratory and clinical arena.

In Chapter 21, Bill DeCampli presents a thoughtful review of the literature, along with his recently completed studies in piglets, to assess neuronal damage and behavioural outcomes following various techniques used in cardiopulmonary bypass, including deep hypothermic circulatory arrest, and the "low-flow" and "high-flow" variants of hypothermic bypass. Additional studies in piglets are presented which compare alpha-stat and pH stat management during core cooling, which correlate well with studies in humans. In Chapter 22, Ross Ungerleider reviews current techniques to minimize morbidity in the neonate and young infant undergoing cardiopulmonary bypass. Ross provides clinical correlates to the principles reviewed in Chapter 21 by Bill DeCampli regarding the use of deep hypothermic circulatory arrest, as well as the rationale for the routine use of ultrafiltration and occasional delayed sternal closure. It is increasingly recognized that the immediate postoperative period is one of increased vulnerability to neurological injury, and in Chapter 23, George Hoffman reviews the growing information obtained by his excellent group of investigators from the Children's Hospital of Wisconsin on postoperative monitoring of regional saturations of oxygen in the brain and other organs. Finally, in Chapter 24, Carmen Giacomuzzi and her colleagues at Doernbecher Children's Hospital in Portland, Oregon, describe a similar methodology but applied to a the subset of patients considered at high risk during the first stage of the Norwood sequence of operations for hypoplastic left heart syndrome followed by routine ventricular assist.

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