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EDITORIAL COMMENT

Congenital heart disease: New challenges

Cardiopatias congénitas: novos desafios

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The prevalence of congenital heart disease (CHD) is about 1 in 100 live births,[1] with a wide spectrum of disease severity ranging from minor septal defects to life-threatening situations like hypoplastic left heart syndrome, transposition of the great arteries and cyanotic forms of tetralogy of Fallot. Mortality from severe forms of CHD in infancy and childhood has declined markedly in recent decades, mostly driven by advances in pre- and postnatal diagnostics, early corrective surgery and improved postoperative care.[2] Gains in survival until adulthood and increased life expectancy represent a new challenge, since inadequate cerebral blood flow and hypoxia during critical phases of fetal brain development may have irreversible consequences, leading to lifelong cognitive impairment. Recent studies have accordingly aimed to characterize neurocognitive performance and related impacts on psychosocial achievements in these patients, in order to identify their relationship with neonatal and clinical variables that could be the target of intervention.

The paper by Areias et al. published in this issue of the *Journal*[3] is noteworthy as the first holistic and interdisciplinary study between psychology and cardiology carried out in this country. The authors compared the cognitive performance of a cohort of 217 Portuguese CHD patients (cyanotic and noncyanotic forms) with a matched control group (n=80), using a comprehensive neuropsychological test battery to explore the major cognitive domains: memory, executive function, processing speed, attention and visuoconstructive ability. The examination was complemented with behavioral and psychological assessment instruments and a semi-structured interview covering areas of social support, family educational style, environment, self-image, functional limitations and educational achievements. The results confirm a profile of poor performance in all the explored cognitive domains, which is similar to that resulting from the encephalopathy first described in premature infants. As in the latter condition, studies using conventional fetal brain magnetic resonance imaging (MRI) have shown findings compatible with cerebral white matter immaturity that correlated with the severity of the underlying CHD. Furthermore, advanced MRI techniques reveal disorganization of neuronal networks that is associated with the attention deficit hyperactivity disorder seen in adolescents with severe congenital CHD.[4] Research in which imaging data is correlated with clinical outcomes for risk stratification is increasingly used in CHD and should also be considered by this Portuguese group.

Another strength of the current paper[3] is the identification of neurocognitive phenotypes (clusters) which were further correlated with sociodemographic, neonatal, clinical and psychological adjustment variables. The authors identified three phenotypes that differed in all cognitive dimensions: non-impaired (NI), moderately impaired (MI) and globally impaired (GI). As anticipated, the MI and GI clusters included more cyanotic forms of disease, including tetralogy of Fallot and transposition of the great arteries. Of particular interest, compared to the NI cluster, the GI cluster showed worse neonatal indicators (smaller head

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circumference and lower birth weight and length and Apgar scores), as well as poorer psychosocial achievements, including fewer years of schooling and more psychological adjustment problems and aggressive behavior.

These findings highlight the impact of antenatal impairment in brain growth and underline the importance of identifying parameters of cardiac and circulatory function that can be used as surrogates of brain development, particularly in these high-risk patients. For instance, prenatal autonomic activity has been investigated as a marker of early childhood development, which was confirmed in a recent study showing that fetal heart rate variability parameters at 34 to 38 weeks gestational age correlated with 18-month cognition and motor scores.[5] The same applies to the cerebral-to-placental resistance ratio, which has been assessed as a measure of cerebral autoregulation (brain sparing).[6]

With regard to treatment, the establishment of clusters of cognitive functioning like those proposed by Areias et al. may be useful, providing guidance for more personalized and tailored interventions. For less serious conditions, a more conservative approach should be recommended focusing on prevention of further brain insults (particularly during delivery and surgical procedures) and early cognitive stimulation as well as educational support. For severe forms of CHD, besides those described above, new neuroprotective approaches, such as maternal hyperoxygenation at 28 to 41 weeks of gestational age to increase cerebral oxygen delivery during pregnancy,[7] may be considered. Furthermore, these new strategies of intervention should be tested using specific clinical outcomes such as those analyzed in the paper by Areias et al., including psychosocial adjustment, educational performance, quality of life, and independence in adulthood.

Conflicts of interest

The author has no conflicts of interest to declare.

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