

Childhood stroke

Diagnostic and management challenges

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Childhood stroke constitutes a significant health problem. During the past 10 years, the reported incidence of childhood ischemic stroke increased to 2-6/100,000 children/year.¹ This is partly due to increased clinical awareness, improved newer neuroimaging techniques and an increased number of children who now survive lethal disorders, such as leukemia and congenital heart disease, which are known to be complicated by stroke. Over 60% of children surviving a stroke develop some residual handicap, including motor deficits, seizures and mental retardation. The identification of causes and risk factors of stroke in children in a given population is important, since non-atherosclerotic risk factors have different features in children compared to adults and are age and population specific. Research on pediatric stroke is still at its infancy and, especially in Saudi Arabia, baseline epidemiologic data and exploration of risk factors is scarce. To address the shortcomings and in the hope of better diagnostic, management and preventive care directed to childhood stroke in Saudi Arabia, a research proposal was submitted to the Prince Salman Center for Disability Research (PSCDR) on "Stroke in Saudi children. Clinical features, etiology, risk factors and prognosis." This was gracefully funded by PSCDR, and the outcome was important intriguing information. It is our hope that the present Supplement will highlight and disseminate this information, and ultimately fulfill our goal of better preventive and curative care for children with stroke.

The presented combined prospective and retrospective study, which spanned 10 years and 7 months, constituted one of the largest cohorts of children with stroke studied at one medical center. During the study periods July 1992 to March 2003,

117 children (61 males and 56 females) were evaluated; the majority (89%) were Saudis. The calculated annual hospital frequency rate of stroke was 27.1/100,000 of the pediatric (1 month – 12 years) population.² Ischemic stroke accounted for most cases, whereas intracranial hemorrhage was less common. A major risk factor was identified in 93 of the 104 (89.4%) Saudi children.²

The present study uncovered the importance of prothrombotic disorders as risk factors for stroke in Saudi children.³ It is noteworthy that previous studies, whether in Saudi Arabia or the Gulf Region, did not explore the role of prothrombotic conditions in the pathogenesis of childhood stroke. Contrary to previous similar studies from Saudi Arabia, sickle cell disease (SCD) accounted for greater than 10% of the ascertained risk factors and those patients presented with severe manifestations, highlighting the severe phenotype of SCD, which is also prevalent in Saudi Arabia, in addition to the mild type. Of the identified risk factors, presumed perinatal ischemic cerebral injury accounted for stroke in 23 children (22.1%),⁴ and infectious and inflammatory disorders of the circulatory system in 18 (17.3%).⁵ Acute bacterial meningitis constituted an important potentially preventable cause. The study⁵ also described the first cases of stroke as a manifestation of neurobrucellosis, and suggested that investigation for neurobrucellosis should be part of the diagnostic work-up for childhood stroke in Saudi Arabia and other similar countries of the Gulf Region. Congenital toxoplasmosis and rubella syndrome were also identified risk factors for childhood stroke and both are potentially preventable.⁵ Congenital and genetic cerebrovascular anomalies were also identified as important risk factors where neurosurgical intervention has saved lives in several

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cases.⁶ Underlying cardiac anomalies should be excluded in cases of childhood stroke, since the condition can be asymptomatic, as has been identified in one patient in this cohort.⁷ Moyamoya syndrome is another significant underlying cause of stroke in Saudi children,⁸ and was found, in the present study, to be associated with another disease in all identified cases. The association of moyamoya syndrome and protein C deficiency was first reported in this cohort, whereas the associations of the syndrome with wrinkly skin syndrome (OMIM 278250) and Adams-Oliver syndrome (OMIM 100300) have not, hitherto, been described. Revascularization surgery (encephaloduroarteriosynangiosis) was a safe intervention procedure even in patients with hypercoagulable disorders (protein C and S deficiencies) and in SCD. Inherited metabolic diseases (all due to mitochondrial disorders in this cohort) caused stroke in 3.8%; but required specialized reference laboratories for diagnostic confirmation.⁹ Hypernatremic dehydration¹⁰ and post-traumatic arterial dissection¹¹ have also contributed to stroke. Both require challenging but life saving, management.

The outcome of stroke in Saudi children constituted a huge burden, with a high rate of motor, cognitive and language deficits. Residual hemiparesis (irrespective of its effect on daily functions), was detected in 81%, whereas epilepsy affected 58%. The socio-economic consequences of these ailments for the victim, family, and the community at large are quite challenging. Programs aimed at the primary prevention of stroke remain the corner stones in handling this situation.

Based on the experience gained from this study and the international guidelines, a diagnostic algorithm outlining the approach to a child with a suspected stroke/cerebrovascular lesion was designed.¹³ The algorithm might also be of use for managing other children from the Arabian Peninsula and Middle East with similar demographic, socio-economic and ethnic backgrounds. Nationwide, handling of childhood stroke can be achieved effectively by the establishment of specialized stroke care services, designing and implementing a childhood stroke registry via the Internet, and efficient use of Telemedicine to help in the initial management of patients at the various district and regional hospitals.¹⁴

Finally, this Supplement comes out following the tragic death of one of the Investigators, Dr. Ahmed A. Al-Jarallah in June 2004. Since his service as Senior Registrar at the Division of Pediatric Neurology (DPN), he was keen to join and give his contribution to this endeavor. Although he started to feel unwell during the prospective study period of the Project (when he was appointed Consultant Pediatric Neurologist), he kept his enthusiasm in discussing individual cases and required investigations. His death is a sad loss for

the DPN, his colleagues, his patients and above all his family. We sincerely hope this Supplement will add to his deeds.

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