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Management strategy in pediatric moyamoya angiopathy: the Zurich Moyamoya Center experience

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Introduction and Objectives: Moyamoya is a dynamic and progressive steno-occlusive angiopathy of the circle of Willis presenting with repetitive ischemic strokes in the pediatric age group. The management strategy of newly diagnosed children referred to our moyamoya center is presented.

Method / Patient Selection: Twenty-six newly diagnosed moyamoya children, referred from within Europe and internationally, were managed at our moyamoya center between March 2011 - December 2013. Mean age was 8 years (range 1-17 years). Female to male ratio was 15:11.

Results: Diagnostic workup consisted of clinical-neurological evaluation including child development/cognition testing, diagnostic 6-vessel cerebral angiography, MRI and H215O-PET scan with Diamox challenge.

Headache and transient ischemic attacks were common presenting symptoms. Moyamoya syndrome was seen in half of the children (13/26). Two of the syndromic moyamoya children were asymptomatic clinically. Bilateral cortical/subcortical infarcts in distribution territories of anterior cerebral artery (ACA), middle cerebral artery (MCA) and/or posterior cerebral artery (PCA) were seen mainly in children of age < 5 years. Unilateral angiopathy was seen in 4 children. Twenty-four children showed bilaterally decreased perfusion reserves on Diamox-H215O-PET in ACA, MCA and /or PCA territories. Depending on symptomatology, extent of angiopathy and territorial perfusion reserve deficits, multiple 1 or 2 staged cerebral revascularization surgeries were performed. Clinical-neuroradiological follow up at 6 months and 1-3 years after last surgery is ongoing and to be continued into adulthood. The first 15 children at average post-op follow up of 9 months (6 months-1 year) remain stroke free.

Conclusions: Moyamoya in Europe and internationally is highlighted. It is not only limited to the Asian population. Accurate diagnostic work-up and thorough pre surgical planning is necessary for a good clinical outcome. A dedicated center i.e. an interdisciplinary medical and neurosurgical team with an infrastructure appropriate for the pediatric age group is pivotal for optimal management.