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Neurosurgery in paediatric strokes

S. Ganapathy* and P. Pandey

Abstract

Paediatric strokes are a different entity owing to the difference in pathological entity causing the stroke as well as difficulty in treatment and management due to the presence of a growing brain and small vascular volume making surgery and endovascular intervention dangerous. Yet, the high neuronal plasticity coupled with unique surgical and endovascular procedures makes surgery in these conditions rewarding with improving morbidity and mortality statistics. The field is young and dynamic leading to constant change and updating. We attempt to review the current recommendations with our own experience in paediatric neurosurgery for paediatric strokes and present an overview of common conditions causing paediatric strokes. A brief review of the literature is also supplied for reference.

Keywords: Paediatric strokes, Moyamoya disease, Paediatric aneurysms, Paediatric arteriovenous malformations, Revascularization surgery

Introduction

Paediatric strokes form a very unique subset of strokes [1–4]. They are usually secondary to cardiac disorders (almost one third); haematologic disorders such as sickle cell disease (SCD); prothrombotic disorders; infections such as varicella, human immunodeficiency virus (HIV) infection, meningitis, and tuberculous (TB) meningitis; vascular syndromes; metabolic disorders; vasculitis, due to paraneoplastic syndromes, as a consequence of trauma, and finally even as a side effect of certain drugs [1–7]. (Fig. 1).

Paediatric strokes present in 2 main ways, as in adults: haemorrhagic or ischaemic [2–8].. The causes vary. Haemorrhagic strokes include diseases such as arteriovenous malformations (AVMs) and aneurysms. Ischaemic strokes are predominantly caused by diseases such as moyamoya disease, and moyamoya syndrome [3–10] (Fig. 2).

We attempt to present certain distinct surgical syndromes and their surgical treatment below.

Moyamoya disease

This is a chronic cerebrovascular disease characterized by stenosis and eventual occlusion of the bilateral terminal internal carotid arteries (ICA) with simultaneous collaterals being produced at the base of the brain [3–6, 9]. Moyamoya disease has a bimodal presentation and occurs either between 5 and 9 years or in adults between 45 and 49 years [4, 5, 7, 8]. Moyamoya disease represents approximately 6% of childhood strokes with a female to male ratio of 2.18. Fifteen per cent of cases are familial [6–12]. Lastly, the disease can be unilateral or bilateral. Approximately 40% of unilateral cases progress to bilateral forms [12].

Moyamoya disease presents in different ways. The commonest symptoms are that of ischaemia, or haemorrhage, along with seizures and headache [12–14]. The disease is associated with a variety of syndromes making its existence complex and difficult to diagnose [15, 16]. Some of those diseases include Down's syndrome, neurofibromatosis, or even sickle cell disease (SCD) [16–19]. Most children present with transient ischaemic attacks (TIAs) or established strokes. Alternating symptoms such as drop attacks, seizures, and headaches also occur frequently [18].

Symptomatic patients treated medically have a poor prognosis (Fig. 3). Around 40 to 82% of medically managed patients sustain a new stroke within 5 years (13–15% per year). Surgically treated patients fare better, compared with the natural history of the disease [18–22].

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CAUSES OF PEDIATRIC STROKES:

1. Cardiac disease (approximates 30% of all strokes)
2. Hematologic disorders (e.g. Sickle Cell Disease)
3. Prothrombotic states
4. Infections (e.g. Varicella, HIV, TB meningitis)
5. Vascular lesions (AVMS, Aneurysms)
6. Metabolic disorders
7. Vasculitis
8. Paraneoplastic syndromes
9. Traumatic
10. Drug induced

Fig. 1 Causes of paediatric strokes

SURGICAL DISEASES CAUSING STROKES:

1. Hemorrhagic strokes
   a. Arteriovenous Malformations (AVMs)
   b. Aneurysms
2. Ischemic
   a. Moyamoya disease
   b. Moyamoya syndrome

Fig. 2 Surgical diseases causing strokes

Moyamoya disease: medical treatment

Author	Stroke risk/year
Hallmeier	12%
Gross	13.3%
Cho	4.3%
Meta-analysis	
Jeon JP, et al, J Neurosurg 2017	

Fig. 3 Moyamoya disease: medical treatment

Bypass surgery significantly decreased stroke events ($p < 0.001$). Direct surgery is significantly better than indirect revascularization ($p = 0.028$). There is no difference in complications ($p = 0.176$) between approaches [21].

In indirect revascularization, the EDAMS technique involving an encephalo-dural-angio-myo synangiosis has both benefits and disadvantages [22–24]. The benefit is of generally shorter operative times and fewer technical challenges [25, 26]. The disadvantages however are many. The main problem is a delay of weeks to months for new vessel formation. Collateral vessels required 3 to 4 months to develop [26]. Some series demonstrate an increased risk of stroke in this period. Therefore, our preference is for a combined revascularization, where one employs a superficial temporal artery (STA) to middle cerebral artery (MCA) STA-MCA bypass combined with indirect methods like dural and muscle inversion [27] (Fig. 4). This gives the patient the benefits of both methods. There is an immediate reduction of ischaemic risk, with the gradual development of collaterals from the indirect method for robust and necessary revascularization [25, 27].

The rationale to do both procedures together is that immediate risk reduction is important, especially in patients having repeated symptoms. It is recognized that direct graft may not give good revascularization to the entire hemisphere [18–20]. Fifty per cent of revascularization in direct bypass surgeries ultimately comes from indirect sources. Therefore, a combined procedure offers the benefit of both procedures with immediate and long-term revascularization issues attended to. A direct extracranial-intracranial arterial bypass is preferred where end-to-side anastomosis of parietal/frontal branch of the superficial temporal artery (STA) with cortical branch of the middle cerebral artery (MCA) is done using 10.0 interrupted sutures. The EDAMS indirect revascularization involves using the temporalis muscle with the deep temporal artery, the STA, and the middle meningeal artery as adjuncts to facilitate neovascularization.

Complications include the risk of perioperative stroke which is around 1–2%, hyperperfusion to the brain leading to bleeds, and inadequate revascularization which leads to a persistence of the clinical syndrome of strokes or bleeds despite surgery [26]. Therefore, combined revascularization is an effective technique for symptomatic moyamoya disease (MMD) with reasonable morbidity and mortality. Long-term results are excellent in both ischaemic and haemorrhagic forms of the disease with combined revascularization [27].

Paediatric aneurysms

Paediatric aneurysms are rare, unlike the adults. Most of the institutional series are small, thereby making data unreliable and difficult to predict. Some are related to arterial dissections or head trauma (5–10%). Other causes include genetic diseases like Ehrler-Danlos syndrome (EDS), Marfan's syndrome, polycystic kidney disease (ADPKD), hereditary haemorrhagic telangiectasia (HHT), and mycotic aneurysms caused by infections due to *Streptococcus viridans* or *Staphylococcus aureus* account for around 8–10% as well.

The incidence is approximately 0.05–0.09 per 100,000 persons per year in children less than 15 years but 0.5 per 100,000 in children greater than 15. Unlike in adults, paediatric aneurysms are commoner in males as compared to females. They present as in adults with SAH

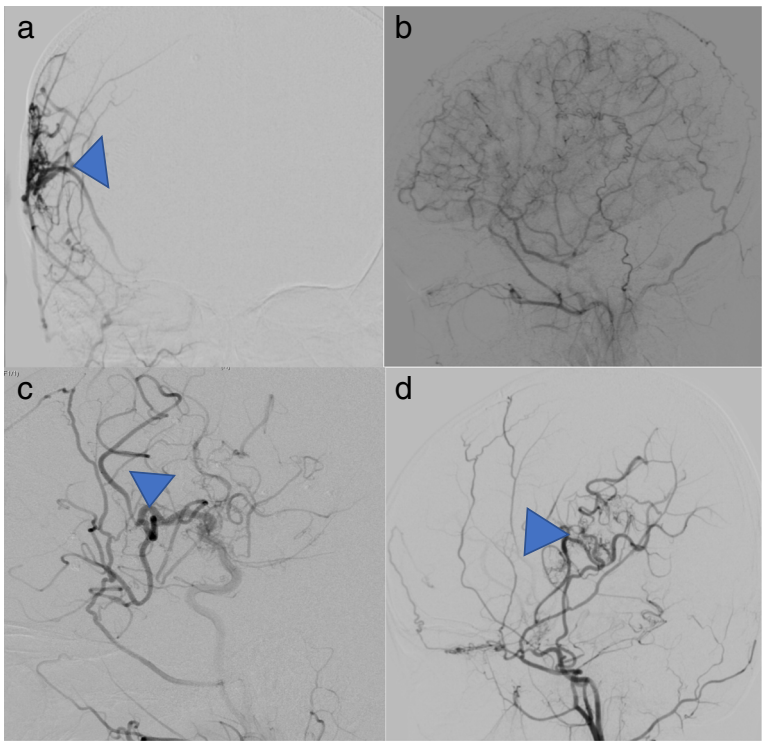

Fig. 4 **a** MCA filling from the anastomosis. **b** MCA filling from an ECA injection. **c** Filling of the external carotid artery (ECA) circulation with the bypass section seen along with **d** ECA circulation with MCA filling as shown

and mass effect. Many are incidentally discovered as well.

Unlike in adults, ICA is the commonest site of aneurysms in children. In order of the highest incidence, the locations of paediatric aneurysms include ICA bifurcation (commonest), MCA, and anterior communicating artery. Posterior circulation aneurysms are 2–3 times commoner than in adults with 20% of all paediatric aneurysms occurring in the posterior circulation. Almost one fourth to one third are giant aneurysms, which is rare in adults. Giant aneurysms are also commoner in the vertebrobasilar circulation. There is also a higher incidence of mycotic aneurysms (8%) as well as aneurysms in the distal branches of vessels where 70% present with haemorrhage. Aneurysms are also increasingly associated with vasculitis and MMD. There is also a very high incidence of dissecting and fusiform aneurysms, along with supraclinoid ICA, MCA, and posterior cerebral artery (PCA) aneurysms (P1–P2 aneurysms) [19, 21, 25, 26].

Because of young age and long life expectancy, surgical clipping is preferred over coiling, as it is considered to be more durable with significantly less re-treatment required. There is also a higher fraction of complex aneurysms where coiling becomes impossible necessitating open microsurgery. There is also a higher incidence of complex surgical procedures apart from simple clipping such as external carotid artery to internal carotid artery (ECA-ICA) bypass and parent vessel occlusion coupled sometimes with complex endovascular procedures in a hybrid approach enabling the best prognosis [24, 27, 28]. There is also a higher incidence of spontaneous occlusion and thrombosis of aneurysms making the intervention in all cases unnecessary [26].

MMD aneurysms occupy a special place in paediatric aneurysms where aneurysms are associated with MMD and rupture causing bleeds and concomitant infarcts (Fig. 5). Treatment can be either endovascular or surgical. Vascular access can be a problem, especially in stenosed blood vessels making endovascular procedures difficult. In patients presenting with both ischaemia and bleeds, clipping of aneurysm and simultaneous indirect revascularization can be performed [26, 27].

Paediatric arteriovenous malformations

These are rare in children, constituting approximately 3% of all AVMs. They are much more likely to present with rupture. Long life expectancy and rupture status, along with the presence of the developing brain, make for a very different treatment regimen as compared to adult s[29].. Paediatric AVMs are the commonest cause of spontaneous intracerebral haemorrhage (ICH) in the paediatric population (Fig. 6). The prevalence ranges approximately between 0.06 and 0.11%. Most children present with rupture, a few with seizure/headache. The

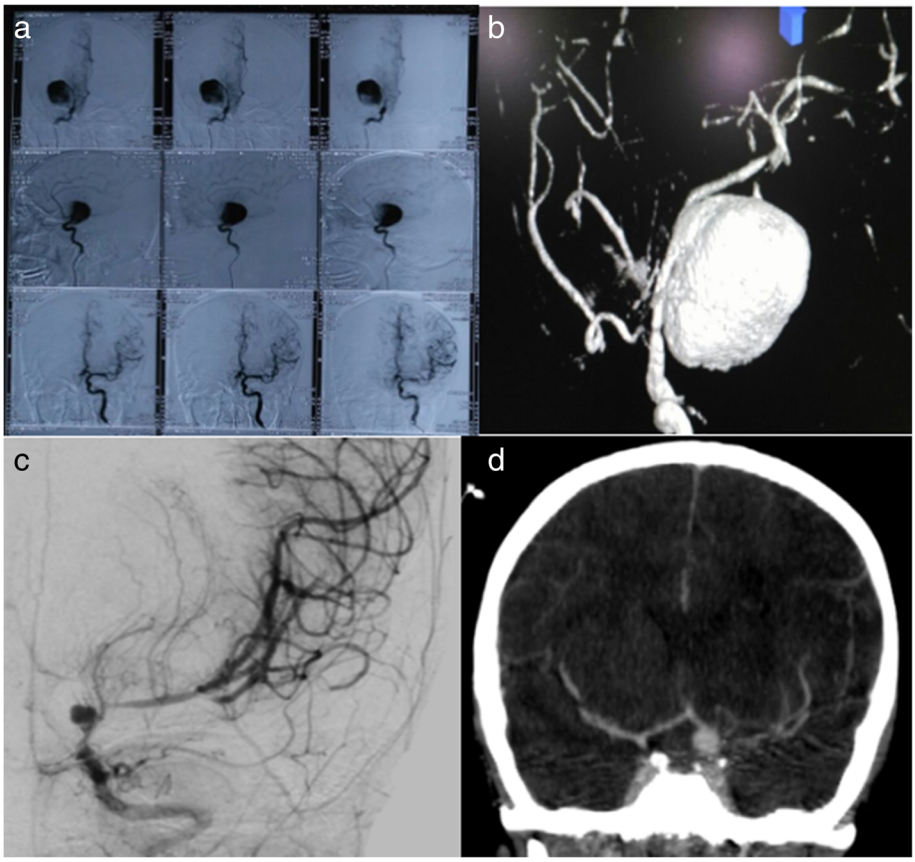

Fig. 5 **a** Angiographic evidence of giant aneurysms. **b** 3D reconstruction of the same aneurysm with a complex architecture. **c** A moyamoya aneurysm where the aneurysm is seen in a background of ICA stenosis and multiple small feeble collaterals. **d** A CT angiogram showing a moyamoya aneurysm in the left ICA

annual haemorrhage rate is around 2–4% per year. Re-haemorrhage rates are 4–6% per year, with mortality rate up to 25% per each event. The risk of haemorrhage depends upon the presence of previous rupture, deep-seated AVMs, infratentorial AVMs, female sex, AVMs associated with aneurysms, or a diffuse morphology [29, 30].

Diagnosis after the clinical presentation of a spontaneous ICH is by MRI/MR angiography (MRA) for AVM localization and treatment planning. Definitive analysis is through a digital subtraction angiography (DSA) where size, location, venous drainage, associated vascular conditions, and dynamic blood flow are noted to decide upon treatment. Multimodality management is often used to treat these conditions [23, 24, 29, 30]. Options include surgery embolization and radiosurgery. Deciding upon the mode of therapy depends upon the Spetzler-Martin grading. Complete surgical resection is the gold

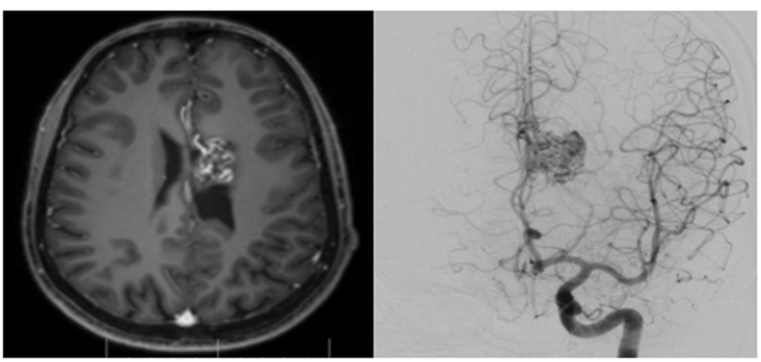

Fig. 6 A parasagittal AVM being fed from the ACAs

standard for AVM treatment. It can be done in lower-grade AVMs with acceptable morbidity. Surgery offers an immediate cure along with haematoma removal. Most series suggest a 90–95% cure rates, with 5% combined morbidity and mortality. Better long-term results in the paediatric population are seen as compared to adults [29, 30].

Radiosurgery was first used by Altschuler in 1989. There were concerns however regarding exposure of developing brains to radiation therapy. Radiation is indicated in deep AVMs, not easily accessed by microsurgery, or in AVMs of the eloquent cortex [30]. Radiation offers 60–70% obliteration rates with 3–6% complication rates. The common complications include radiation oedema along with non-haemorrhagic neurological deficits. Radiosurgery is now being used in certain complex cases. The most important drawback is delayed obliteration while the risk of haemorrhage remains till obliteration [29, 30]. The annual haemorrhage rate after radiosurgery is 1.8–2.8% per year. Embolization is mostly used as an adjunctive treatment despite the rapid evolution of catheter technology, imaging, and embolic materials. Cure rates are 5–20% with complication rates of 7–26%. Embolization is preferred as a pre-surgical adjunct. It is judicious to use a single modality whenever feasible to effect the best results [30].

Conclusion

Both ischaemic and haemorrhagic diseases are encountered in the paediatric population. Surgery plays an important part in the evaluation and management of these diseases. Other rare diseases such as dural arteriovenous fistulae (DAVFs) are treated as per the situation demands.

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Authors' contributions
Dr. Sibhi Ganapathy analysed and interpreted the patient data. Dr. Paritosh Pandey performed the review of the images and cases and reviewed the manuscript data and references. All authors read and approved the final manuscript.

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Dr. Sibhi Ganapathy is a young and upcoming neurosurgeon with over 50 publications to his credit. His interests include paediatric neurosurgery and vascular neurosurgery. Dr. Paritosh Pandey is a senior neurosurgeon and microvascular and endovascular neurosurgeon with decades of experience in the field. He is a renowned author, speaker, and educator who has illuminated the field of neurosurgery for years in India, and across the world.

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