Imaging in paediatric stroke

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Introduction
Stroke is defined as a cerebrovascular abnormality that results in a rapid onset of neurological symptoms lasting longer than 24 hours or causing death.1 Stroke in children is an underrecognised condition, despite being a common cause of childhood death.2,3 The neonate is particularly vulnerable, with neonatal stroke being nine times more common than in the general paediatric population.4 The aetiology of stroke in children is markedly different to adults (see table 1 for causes). Stroke in children is due to infarction (eg figure 1) and haemorrhage (eg figure 2) from various causes in equal proportions. This is in comparison to ischaemic stroke from atherosclerosis that accounts for 80-90% of adult strokes.5 Diagnosing stroke is challenging in children as signs and symptoms can be difficult to elicit, or subtle, especially in neonates. For this reason, children often present in a delayed fashion. This makes neuroimaging vitally important in the early diagnosis or exclusion of stroke in the acutely unwell child where their capacity for recovery is greater than in the adult population.6

Imaging protocol
Both imaging and interpreting the paediatric neural axis pose challenges in children with suspected stroke. Unwell children and neonates may require sedation prior to scanning to obtain diagnostic images with either oral sedatives or a general anaesthetic, especially when undertaking a magnetic resonance imaging (MRI) scan, which requires the child to lie still for relatively long periods.

In the acute setting, unenhanced computed tomography (CT) remains appropriate in certain circumstances despite the use of ionising radiation. CT is quick, readily available and able to visualise acute intracranial haemorrhage or mass effect requiring emergency neurosurgery. CT angiography and or venography can provide additional information if required following review. MRI provides superior anatomical as well as further functional information over CT. MR is the imaging modality of choice in paediatrics and should be used when feasible to determine the underlying cause of stroke. Angiographic imaging from the aortic arch to the proximal intracranial arteries should be performed within 48 hours in all children with suspected arterial stroke.7 See table 2 for a protocol used to investigate all children with suspected stroke at our institution.

Image interpretation and differential diagnoses
Moyamoya syndrome
This is hypoplasia or occlusion of the terminal portion of the internal carotid arteries and proximal intracranial arteries. There are many causes and associations including sickle cell disease, Down's syndrome, neurofibromatosis type 1, cranial radiotherapy, hereditary spherocytosis, HIV, glycogen storage disease type 1a, TB meningitis, congenital heart disease, hyperthyroidism and renal artery stenosis. Occlusion or narrowing of the supra-clinoid internal carotid artery causes formation of collateral vessels over time in the perforator territories, which appear as a ‘puff of smoke’ on angiography. Moyamoya syndrome can cause ischaemic stroke from hypoperfusion of the anterior circulation or haemorrhagic stroke from the multiple friable collateral perforator arteries.8 Figure 3 shows a case of Moyamoya-like phenomenon.

Intracranial vasculitides
A wide variety of conditions can result in inflammation of the intracranial circulation.9 These include intracranial infections, systemic vasculitides and granulomatous disorders such as sarcoidosis and granulomatosis with polyangiitis. The inflammatory process can result in arterial occlusion/narrowing and aneurysm formation. This can result in both ischaemic and haemorrhagic stroke. Common viral pathogens are varicella, HIV and hepatitis C. Bacteria include Borrelia burgdorferi (Lyme disease) and mycobacteria. Fungal and parasitic infections have also been described.10

Arteriovenous malformation (AVM)
An AVM is a collection of arteries that drain directly into veins without a capillary bed either from a direct fistulous connection or a nidus. A flow-related aneurysm can sometimes form. These vascular anomalies have an increased propensity to bleed of ~3% per year causing a haemorrhagic stroke.11

Arterial dissection
A tear within the arterial wall results in intramural haematoma, usually following blunt trauma or abrupt hyperextension of the neck, and is increasingly recognised as a cause of stroke in the paediatric population.12 This is thought to cause narrowing of the lumen and embolus resulting in cerebral infarction. Dissection is most commonly seen in the extracranial arteries, with the anterior circulation more commonly involved. Certain conditions including connective tissue disorders such as Ehlers-Danlos and osteogenesis imperfecta increase the risk of an arterial dissection occurring following relatively trivial trauma.

Venous infarction
Thrombus can form within the dural venous sinuses from many causes including coagulopathies, infection, trauma, dehydration, haematological disorders and chemotherapies.13 Acute thrombosis appears hyperdense on an unenhanced CT and fails to opacify on CT venogram. Clot within the venous sinuses has variable signal characteristics on MRI depending on the age of the thrombosis. The region of infarction does not follow a particular arterial territory and depends on the venous sinus occluded. The resulting infarcts are often haemorrhagic and multifocal. This condition is often difficult to diagnose clinically, however mortality has fallen in recent years, possibly due to more timely recognition/imaging diagnosis and treatment.
Meningitis

Infection of the meninges can result in ischaemic stroke from large or small vessel arterial occlusion from arterial wall inflammation and necrosis, venous sinus thrombosis usually due to adjacent subdural empyema, or cardiopulmonary arrest from sepsis.15

Cardiopulmonary arrest

In children, cardiopulmonary arrest commonly results from sepsis, drowning, choking or non-accidental injury. The hypoxic ischaemic injury can be acute or chronic depending on the insult. In the acute or profound pattern, the hypoxic injury is severe affecting the basal ganglia predominantly. In the chronic or partial pattern, there is a peripheral perfusion injury affecting the watershed territories. There is increased overlap between these two patterns with more severe injury. The structures affected can vary depending on the age of the child.16 Figure 4 shows a case of cardiorespiratory arrest.

Discussion

Paediatric stroke remains a poorly recognised and researched condition compared to the ischaemic stroke from atherosclerosis seen in adults, despite the associated significant morbidity and mortality. The causes of stroke in children are very varied with resulting haemorrhage and infarction contributing in equal proportions. Imaging remains at the cornerstone of diagnosis, with MRI often identifying the underlying cause as demonstrated in figures 1-4. Increasing availability and subsequent use of MRI to delineate the aetiology of stroke in children, who are often difficult to accurately assess clinically, will hopefully continue to improve outcomes through more timely diagnosis and management.

References


Figure 1

MRI head from a 16-year-old admitted with sudden onset left-sided weakness, this shows high T2 signal with DWI restriction along the watershed territory of the right anterior and posterior cerebral arteries, in keeping with watershed infarction. The right internal carotid is occluded on the MRA. All figures are reproduced from ‘A pictorial review of imaging in paediatric stroke’ with permission from the BMJ Publishing Group Ltd.17

Figure 2

Cerebral imaging from a 13-year-old girl, admitted with collapse. On admission her pupils were fixed and dilated. The patient required intubation following a respiratory arrest. The non-contrast CT performed following resuscitation showed acute blood (high attenuation material), in the ventricular system effacing the fourth ventricle. The CT angiogram shows the large draining vessel of an AVM leading away from the site of haemorrhage.
**Figure 3**
A 12-year-old girl with sickle cell disease was admitted after developing a sudden onset of right-sided neurological symptoms. T2 weighted image (T2WI) and MRA show loss of both middle cerebral arteries along with multiple lenticulostriatal collaterals in keeping with Moyamoya. On the T2WI there is high signal in the left occipital lobe, which is high signal on diffusion weighted imaging, suggesting acute infarction.

**Figure 4**
Cerebral imaging from a 13-year-old boy after being found hanging. There is subtly increased T2 signal throughout the grey matter, with corresponding restricted diffusion on DWI. These findings fit with diffuse cerebral hypoxia.

### Table 1
**Causes of paediatric stroke.**

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Causes</th>
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<tbody>
<tr>
<td>Arteriopathy</td>
<td>Moyamoya</td>
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<tr>
<td></td>
<td>Syndromic</td>
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<tr>
<td></td>
<td>Acquired</td>
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<td>Cardio-embolic disease</td>
<td>Congenital heart disease</td>
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<td>Arrhythmia</td>
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<td>Endocarditis</td>
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<td>Cardiopulmonary arrest</td>
<td>Sepsis</td>
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<td>Drowning</td>
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<td>Non-accidental injury</td>
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<td>Metabolic encephalopathy</td>
<td>Mitochondrial encephalopathy</td>
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<td></td>
<td>Organic aciduria</td>
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<td>Vascular lesions</td>
<td>Aneurysm</td>
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<td></td>
<td>Vascular malformations, eg arteriovenous</td>
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<td></td>
<td>malformations</td>
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<td></td>
<td>Dissection</td>
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<td>Venous infarction</td>
<td>Trauma</td>
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<td>Infection</td>
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<td>Dehydration</td>
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<td>Chemotherapy agents</td>
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<td></td>
<td>Leukaemia</td>
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<td>Haematological/oncological</td>
<td>Coagulopathy – factor deficiencies, lupus</td>
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<td></td>
<td>anticoagulant, protein C/S deficiency</td>
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<td></td>
<td>Thrombophilia</td>
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<td>Tuberous sclerosis</td>
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<td>Neonatal</td>
<td>Perinatal arterial ischaemic stroke</td>
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<td>Venous infarction</td>
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<td>Chronic partial asphyxia at term</td>
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**Table 2**
**MRI protocol.**

<table>
<thead>
<tr>
<th>Head</th>
<th>Axial T2</th>
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<tbody>
<tr>
<td></td>
<td>Diffusion weighted imaging</td>
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<tr>
<td></td>
<td>Coronal T1</td>
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<td>Post contrast T1</td>
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<td>Neck</td>
<td>Axial T1 fat saturation</td>
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<td></td>
<td>Axial T2</td>
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<tr>
<td>Angiography</td>
<td>Circle of Willis MRA</td>
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<td>Time of flight MRA of the neck</td>
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<td>Contrast enhanced MRA arch and neck</td>
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Causes of paediatric stroke.