Posterior circulation arterial ischemic stroke (PCAIS) is less common than anterior circulation stroke, accounting for 20% to 44% of cases in adult population-based stroke registries. Atherosclerosis-related large artery occlusive disease is the most common stroke subtype in adults. There are limited data on risk factors, stroke subtypes, and recurrence risk for childhood PCAIS. There are few radiological descriptions of vertebrobasilar infarct topography and vascular imaging abnormalities in the pediatric population. In a mixed retrospective/prospective series of 22 children with PCAIS from the United Kingdom, 59% had multiple vertebrobasilar territory infarcts. Vertebral arterial dissection was the most common cause, identified in 45% of cases. One-fifth of cases had recurrent strokes.

The aims of this study were to describe the radiological features of posterior circulation stroke in a prospective series of Australian children aged 1 month to 17 years to determine whether there are differences in infarct topography, vascular imaging abnormalities, risk factors, stroke subtype, and recurrence risk when compared to adults.

Materials and Methods
Definitions
Posterior circulation arterial ischemic stroke was defined as an acute neurological deficit lasting >24 hours and brain imaging confirming parenchymal infarction within the vertebrobasilar territory. The New England Medical Centre Posterior Circulation Topographical Classification System was used to describe infarct location. Infarcts were divided into proximal, middle, and distal segments of the posterior circulation. The proximal segment is supplied by the intracranial vertebral arteries and the posterior inferior cerebellar arteries. The middle segment is supplied by the basilar artery and its penetrating branches up to, but not including, the superior cerebellar arteries. The distal segment is supplied by the superior cerebellar arteries, distal basilar artery, and the posterior cerebral arteries (Figure 1).

Vascular abnormalities on magnetic resonance angiography (MRA) or conventional angiography (CA) were described in terms of location, severity (focal or segmental stenosis, occlusion, and presence of collaterals), and evolution over time. A pediatric modification of the TOAST classification system was used to define stroke subtypes that included sickle cell disease, cardiac embolism, cervical arterial dissection, Moyamoya disease, steno-occlusive cerebral arteriopathy, other determined etiology, multiple probable/possible etiologies, and undetermined etiology.

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Cerebral arteriopathy was defined as a focal or segmental narrowing of the vessel lumen with flow abnormalities on vascular imaging. In cases in which vascular imaging showed an occluded vessel, additional abnormalities were required, such as arterial wall irregularity proximal to the occlusion or persistent vascular abnormalities on follow-up imaging at 3 months (including partial recanalization with persistent irregularity or involvement of a new vessel). Cerebral arteriopathy was subdivided into transient steno-occlusive cerebral arteriopathy or progressive cerebral arteriopathy depending on whether there was progression clinically (with further stroke episodes) or radiologically (with increasing stenosis or involvement of new vessels) beyond 6 months.

Patients
Case ascertainment was through the prospective Royal Children’s Hospital Childhood Ischemic Stroke-Thrombophilia registry. The Royal Children’s Hospital (Melbourne, Australia) is a tertiary pediatric neurology referral center for the states of Victoria and Tasmania, treating 280,000 children annually. Seventy-three consecutive children with arterial ischemic stroke aged between 1 month and 18 years were prospectively recruited to our institutional registry over a 5.5-year period from August 2002 until February 2008. Twenty-seven (37%) of these children with posterior circulation strokes were the subjects of this study. Six children had recurrent posterior circulation events. All children were followed-up for at least 12 months in the stroke clinic. Follow-up MRI to document evolution of vascular abnormalities was performed in all except 2 children in whom stroke mechanism was clearly known.

Forty children with perinatal arterial ischemic stroke were excluded from analysis because previous pediatric studies have shown that risk factors, etiology, and clinical course differ between neonates and older children.

Twenty-six underwent MRI at our institution on 1.5- or 3.0-Tesla magnets (GE or Siemens Avanto). The presence of a left ventricular assist device prevented us from performing MRI at presentation in 1 child awaiting cardiac transplantation. This patient subsequently underwent follow-up MRI and MRA after transplantation. Sequences performed included axial T1, axial and coronal T2, axial fluid-attenuated inversion recovery, diffusion-weighted imaging, apparent diffusion coefficient images, and 3-dimensional time-of-flight MRA of the intracranial circulation. Contrast enhanced MRA of the neck vessels with axial fat-saturated T1-weighted images was performed in selected cases if there was clinical suspicion of arterial dissection, history of trauma, headache, or neck pain. Conventional angiography was performed in selected cases if the initial MRA was nondiagnostic or other diagnostic investigations failed to identify an underlying cause, if there were multifocal posterior circulation infarcts, if the child had recurrent events despite a previously normal MRA, or if surgical planning was required for cases of Moyamoya or aneurysms. Other diagnostic work-up included prothrombotic studies and echocardiography, except for 2 children (subjects 13 and 23; Table 1) with an obvious cause.

Two pediatric neuroradiologists blinded to the clinical history reviewed the imaging studies. A pediatric neurologist reviewed the clinical notes, hematologic, biochemical, and cardiac investigations to determine risk factors, stroke subtype, and recurrence rates. Radiological findings were correlated with clinical data after initial blinded review of the MRI, MRA, and CA studies. This study was approved by our institutional Medical Ethics Committee.

Results
Demographic Characteristics
Twenty-seven children with posterior circulation strokes were identified; 19 patients (70%) were male. Twenty-three children were white, 2 were of South East Asian origin, and 2 were of East Indian ethnic origin. Mean age at diagnosis of first posterior circulation stroke was 7 years 11 months (range, 1 month to 16 years 6 months). Mean duration of follow-up was 4 years 3 months (range, 17 months to 6 years 11 months; Table 1).

Infarct Topography
Infarct topography is described for 27 initial and 7 recurrent posterior circulation events. Fourteen (41%) posterior circulation infarcts were identified; 9 patients (64%) were male. Twenty-three children were white, 2 were of South East Asian origin, and 2 were of East Indian ethnic origin. Mean age at diagnosis of first posterior circulation stroke was 7 years 11 months (range, 1 month to 16 years 6 months). Mean duration of follow-up was 4 years 3 months (range, 17 months to 6 years 11 months; Table 1).

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Infarcts were confined to the middle segment in 2 events, affecting basilar perforators, and to the proximal posterior inferior cerebellar arteries territory in 1 event. Multiple segments were involved in 6 events. Therefore, 31 events (85%) were distal inclusive infarcts (Figure 1 and Table 2). Nine children also had anterior circulation distribution infarcts; 4 were concurrent with the posterior circulation event and 5 were separate events (Table 1).
<table>
<thead>
<tr>
<th>Patient Event**</th>
<th>Vertebro-Basilar Segment</th>
<th>Infarct Vascular Topography</th>
<th>Single or Multiple Infarcts</th>
<th>Intracranial MRA</th>
<th>Neck MRA</th>
<th>DSA</th>
<th>MRA Correlation With DSA</th>
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<tbody>
<tr>
<td>1 (i) D</td>
<td>L P2 PCA</td>
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<td>S</td>
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<td>R P2 stenosis, L P2 stenosis</td>
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<td>5</td>
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<td>R ICA/trifurcation critical stenosis</td>
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<td>NP</td>
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<td>NP</td>
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<td>S</td>
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<tr>
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<tr>
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<td>S</td>
<td>N</td>
<td>N</td>
<td>N</td>
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<tr>
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<td>S</td>
<td>N</td>
<td>NP</td>
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</tbody>
</table>

AICA indicates anterior inferior cerebellar artery; BA, basilar artery perforators; CA, conventional angiography; D, distal; DSA, digital subtraction angiography; L, left; M, middle; MCA, middle cerebral artery; Mu, multiple; N, normal; NA, not applicable; NP, not performed; P, proximal; PCA, posterior cerebral artery; PCOM, posterior communicating artery; PICA, posterior inferior cerebellar artery; P1, P1 segment of the posterior cerebral artery; P2, P2 segment of the posterior cerebral artery; R, right; S, single; SCA, superior cerebellar artery.

*Left ventricular assist device prevented acute MRA imaging.
†PICA compression from uncal herniation secondary to a rapidly enlarging intraventricular cyst.
‡Normal intracranial MRA and contrast MRA imaging of neck vessels but conventional angiography identified cervical vertebral artery dissection.
§Dissection with false aneurysm.
¶MRA overestimated stenosis as an occlusion.
∥MRA underestimated degree of stenosis.
**(i), (ii), and (iii) refer to index and recurrent posterior circulation events in individual patients.
††MRA of neck vessels failed to detect cervical vertebral artery dissection.
Vascular Imaging Findings

Intracranial MRA was performed at presentation in 25 of 27 children. MRA was not performed in 1 child with a posterior cerebral artery territory infarct from uncal herniation and vascular compression attributable to a rapidly enlarging interventricular cyst. MRI could not be performed in another child with a left ventricular assist device. Intracranial MRA results were abnormal in 16 children (59%) and normal in 9 children (Table 1). Abnormalities identified at initial diagnosis included unifocal or multifocal stenosis in 8 children and occlusion in 8 children (Figure 2B). One child had a thrombosed giant basilar artery (Figure 3). Contrast MRA of the neck vessels was performed in 9 children. Vertebral artery dissection with a false aneurysm was identified in 1 child (Figure 4B). No abnormality was identified in the remaining 7 children (Table 1).

Follow-up intracranial MRA was performed in 25 children. There was resolution, improvement, or no change in vascular imaging of abnormalities in 8 children with steno-occlusive cerebral vasculopathy within 6 months of diagnosis. There was transient worsening of vascular abnormalities, followed by stabilization within 6 months in another 2 children with steno-occlusive cerebral vasculopathy (Figure 2E, 2F). One of 7 children with normal initial vascular imaging results had a unifocal stenosis develop within 3 months, which remained unchanged at 12 months, but follow-up imaging remained normal in the other 6. Progression beyond 6 months occurred in 5 children; 3 had basal collateralization typical of that described in Moyamoya syndrome. Complete normalization with recanalization of an occluded vessel was only seen in 1 child with cardioembolic stroke (Table 1).
CA was performed in 11 children, 7 of whom had recurrent TIA or strokes. Contrast MRA of the neck vessels missed vertebral artery dissection in 2 children, overestimated the degree of stenosis in 2 children, and underestimated the degree of stenosis in another. There was good correlation between CA and MRA in the remaining 6 cases (Table 2).

**Stroke Risk Factors, Stroke Subtype, and Recurrence Rates**

Fourteen children (52%) had recognizable risk factors before diagnosis of posterior circulation stroke. Stroke subtype could be classified in 23 (85%) children using the Pediatric Stroke Classification system (Table 1). Nine children had nonprogressive steno-occlusive cerebral arteriopathy. Three children had vertebral artery dissection; 2 had a history of trauma and 1 had an undisplaced fracture of the second cervical vertebra with resolution of a false aneurysm after 12 months. Three children had Moyamoya disease and 4 had cardioembolic stroke. Four had other determined etiologies and 4 had possible or undetermined etiologies.

Three children required emergency posterior fossa decompressive craniotomies for acute obstructive hydrocephalus or cerebellar herniation caused by edema or hemorrhagic conversion of cerebellar infarcts. Fourteen children (52%) had recurrent strokes affecting the posterior circulation in 6 or anterior circulation in 8 children; 6 children had arteriopathies and 7 had congenital heart disease. One child died from complications of the underlying cardiac condition.

**Discussion**

Thirty-seven percent of children with arterial ischemic stroke at our institution had involvement of the posterior circulation. This is much higher than that of a previous mixed retrospective/prospective study from the United Kingdom, where only 22 cases of PCAIS were identified over a 22-year period from a total population of >200 children with arterial stroke. These differences may be attributable to better detection of PCAIS with the advent of MRI. Our findings are more consistent with large adult registries in white and Asian populations in whom posterior circulation stroke accounts for 38% to 40% of all cases.

Boys were over-represented in our series, consistent with recently published data from the International Pediatric Stroke Study. It has been suggested that this may be because of behavioral differences that predispose boys to trauma and arterial dissection. However, cervical arterial dissection was identified on vascular imaging in 10% of children in our series, in contrast to the United Kingdom study in which trauma-related vertebral artery dissection was identified in <10% of children in our series, in contrast to the United Kingdom study in which trauma-related vertebral artery dissection was identified in 45% of cases.

Infarcts most often involved the distal segment of the posterior circulation territory in adult United States and Swiss registries, but the middle segment of the posterior circulation was most commonly involved in another large adult Korean stroke registry, suggesting infarct topography may be influenced by ethnicity. Almost all the infarcts in our pediatric series involved the distal segments of the vertebro-basilar circulation. Fifty-nine percent of children had multiple infarcts, which is similar to the United Kingdom pediatric series.

Sensitivity of noninvasive vascular imaging is an important issue in childhood arterial ischemic stroke because vascular abnormalities are associated with an increased recurrence risk of up to 66%. Intracranial vascular imaging abnormalities including stenoses or occlusion were identified by MRA in...
more than half of our cases at initial diagnosis. MRA overestimated the degree of stenosis in 1 child and underestimated the degree of stenosis in 2 children when compared to CA. Contrast MRA of the neck vessels failed to identify cervical arterial dissection in 2 children in our series. Both proceeded to CA because of recurrent posterior circulation events.

In a pediatric series of 36 children comparing MRA to CA, MRA was diagnostic in most children with large-vessel occlusions, stenoses, or Moyamoya, but it failed to detect collateral vessels in some patients. CA results were abnormal in 4 of 9 patients with normal MRA results, and it identified additional abnormalities not detected on MRA in another 13 children. The CA findings altered clinical management in 11 children. In another pediatric series of 24 children there was good correlation between MRA and CA for presence or absence of arterial lesions, but there was discordance between the 2 modalities in 25% of children, with MRA overestimating the degree of stenosis or suggesting occlusion when there was still flow on CA. Furthermore, CA detected distal abnormalities of small arteries that were not evident on MRA.

Better imaging protocols such as contrast-enhanced 3-dimensional time of flight MRA through the superior mediastinum, neck, and skull base, 3-dimensional multiple overlapping thin section acquisition MRI of the skull base and circle of Willis, axial noncontrast, non-fat-suppressed, and fat-suppressed T1-weighted images, and T2-weighted spin-echo MRI from the aortic arch through to the circle of Willis have improved detection of vascular abnormalities, but sensitivity is still not equal to that of CA because of artifacts arising from flow voids. Therefore, there remains a strong argument for performing CA in children with normal MRA, particularly if there is a continued suspicion of dissection or a small-vessel cerebral vasculitis. MRA can miss intimal flaps or double lumen of dissection. This can be attributable to segmental blurring or signal intensity loss within the vertebral arteries. In the early and chronic stages, hematoma is usually isointense to surrounding structures, whereas it is almost invariably bright on T1-weighted images between 7 days and 2 months, with a characteristic crescent-shape hyperintense area around an eccentric flow void.

Embolism from cardioembolic and proximal arterial sources were the most common stroke mechanism identified in adults in the New England Medical Centre posterior circulation stroke registry, accounting for 40% to 54% of all cases. Large-artery occlusive disease causing hemodynamic ischemia was seen in 32% to 35% of cases, and branch artery occlusion was seen in only 14% to 17% of cases. In contrast, hemodynamic ischemia secondary to large-artery occlusive disease was the most common mechanism in a Korean stroke registry, accounting for 50% of cases, followed by small-vessel disease in 33% of cases. Cardioembolic stroke was only identified in 10% of cases, but this may be an under-

Figure 4. A 16-year-old boy with a history of neck trauma 18 months before presentation and a 6-week history of vertiginous episodes. T2-weighted MRI showing an acute right cerebellar hemisphere infarct and established infarcts in both thalami (A). Contrast MRA of the neck vessels showing a right vertebral dissection with false aneurysm (B), confirmed on conventional angiography (CA) (C). Repeat CA 12 months later showing spontaneous resolution of the aneurysm (D).
representation because cardiac imaging was only performed in selected cases with a high clinical index of suspicion of a cardioembolic source. Embolism from cardiac or extracranial arterial sources only accounted for one-quarter of cases in our pediatric series, suggesting that PCAIS is more often caused by intracranial arteriopathies.

Adult etiologic classification systems are difficult to apply to the pediatric population because children do not have atherosclerosis-related risk factors causing large-artery occlusive disease or small-vessel lacunar occlusive disease. Risk factors for pediatric stroke in children are more variable and include nonatherosclerotic arteriopathies, cardiac disorders, congenital or acquired thrombophilies, infection, and rare genetic or metabolic disorders. We were able to classify stroke subtype in 85% of cases using a modified pediatric version of TOAST. Nonatherosclerotic arteriopathies collectively accounted for more than half of the cases seen at our institution, in contrast to adults in whom atherosclerotic-related large-artery occlusive disease is the most common cause of posterior circulation stroke. There is increasing evidence that arteriopathies play a major role in pediatric stroke, accounting for >50% of strokes and up to 80% of cases once cardiac causes are excluded.

Steno-occlusive cerebral arteriopathy was the most common etiologic subgroup in our study. The term transient cerebral arteriopathy is synonymous with steno-occlusive cerebral arteriopathy and refers to lack of radiological progression of vascular disease beyond 6 months. Transient cerebral arteriopathy typically affects the anterior circulation, but 3 of 9 children originally described with transient cerebral arteriopathy had posterior circulation involvement.

Preceding varicella infection has been described in 33% to 64% of children with transient cerebral arteriopathy. The pathogenesis of post-varicella arteriopathy is poorly understood, but retrograde viral transmission along trigeminal nerve afferents may trigger an inflammatory response in the vessel wall. Posterior circulation steno-occlusive cerebral arteriopathy may be attributable to a different mechanism, because only 1 child in our series had a history of recent varicella infection, and the vertebrobasilar circulation is mainly innervated from branches of the C2 dorsal root.

Serial imaging is important because it may not be clear at initial diagnosis whether the child has a transient or progressive arteriopathy because of the variable angiographic patterns of evolution over time. Three children in our series with steno-occlusive cerebral arteriopathy had initial angiographic progression in the first 6 months before stabilization. This pattern of clinical and radiological progression has been described in other series.

Half of the children in our study had recurrent events, compared to only 20% of children in the United Kingdom study. Recurrence is dependent on etiology, with the risk being highest in patients with arteriopathies and cardiac disease. Life-threatening increased intracranial pressure is a recognized complication of posterior circulation stroke, particularly in young adults, because of the limited capacity of the posterior fossa to accommodate cerebral edema. Three children in our series required emergency decompressive craniotomies, highlighting the importance of close clinical and radiological surveillance after PCAIS diagnosis in children.

The limitations of this study were that contrast MRA was not systematically performed in all children. Therefore, it is possible that extracranial dissection was missed in 3 children classified as having steno-occlusive cerebral arteriopathy and 2 classified as having undetermined etiologies. All had single infarcts without recurrent events. It has not been our practice to perform contrast MRA imaging of the neck vessels on the initial scan in all children because of practical issues, including the need for general anesthesia, prolongation of scanning time, and use of contrast agents. The study was performed in a tertiary pediatric center; therefore, the findings may not be applicable to the general pediatric population because of referral bias.

**Conclusion**

In summary, most PCAIS are distal in location and nonatherosclerotic arteriopathies are the most common stroke subtype, accounting for two-thirds of all cases. Traumatic extracranial vertebral artery dissection was not commonly identified. Patients with arteriopathies and congenital heart disease are at higher risk for recurrence, highlighting the importance of ongoing radiological surveillance.

**Disclosures**

None.

**References**


Childhood Posterior Circulation Arterial Ischemic Stroke
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Abstract

Childhood Posterior Circulation Arterial Ischemic Stroke

Mark T. Mackay, MBBS, FRACP; Sanjay P. Prabhu, MBBS, FRCR; Lee Coleman, MBChB, BSc, FRANZCR

Background and purpose: Currently, there are limited reports on childhood posterior circulation arterial ischemic stroke (PCAIS). This study analyzes clinical presentations and imaging features of childhood PCAIS to determine differences in infarct location, vascular anomaly, risk factors, and stroke subtypes between children and adults.

Methods: A total of 73 patients with imaging-confirmed childhood PCAIS were recruited prospectively between August 2002 and February 2008 from the Royal Children’s Hospital. Children were divided into proximal, middle, and distal groups based on infarct location. Vascular anomalies were classified based on location, severity, and progression over time. TOAST criteria were used to subtype childhood PCAIS.

Results: Among the 73 recruited children, 27 (37%) had PCAIS; 34 infarct sites were identified, with proximal involvement in 25 cases, middle in 2 cases, distal in 1 case, and multiple segments in 6 cases. Of the 27 PCAIS cases, 25 were detected by MRA, with arterial stenosis in 14 cases and arterial occlusion in 13 cases. Among the 25 children, 5 cases showed progression, 2 cases showed temporary progression followed by stabilization, 8 cases showed stabilization or improvement, and 1 case showed recovery.

Conclusion: Non-progressive arterial disease is the most common cause of childhood PCAIS, often involving the proximal posterior circulation arteries. Arterial hypertension related to the risk factors is not a significant cause of childhood PCAIS, and childhood PCAIS has a high recurrence rate.

Keywords: Arterial disease, cardiac source, childhood, causation, stroke, posterior circulation, risk factors, stroke, vertebrobasilar artery.

Whole Brain and Regional Hyperintense White Matter Volume and Blood Pressure

Overlap of Genetic Loci Produced by Bivariate, Whole-Genome Linkage Analyses

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Background and purpose: Diffusion-weighted imaging (DWI) shows widespread cerebral white matter (WM) damage in hypertension. Previous studies have suggested that some of this white matter damage may be related to blood pressure fluctuations. In this study, we aimed to identify genetic loci that are associated with both brain white matter volumes and blood pressure using bivariate whole-genome linkage analyses.

Methods: In the San Antonio Family Heart Study, we measured blood pressure and obtained high-resolution (1 mm³) magnetic resonance imaging (MRI) scans for 357 family members (182 females; mean age 47.9 ± 13.2 years). We used bivariate whole-genome linkage analyses to identify genetic loci associated with both brain white matter volumes and blood pressure.

Results: We identified significant associations at 1q24, 1q42, 10q24-q26, and 15q26. Post hoc analyses excluded 55 individuals on antihypertensive medication, and the results were consistent with the whole sample.

Conclusion: These findings suggest that genetic factors may contribute to the association between hypertension and brain white matter damage, but further studies are needed to confirm these results.

Keywords: Brain, brain imaging, genetics, hypertension, white matter integrity, white matter disease, magnetic resonance, magnetic resonance imaging.