Childhood Posterior Circulation Arterial Ischemic Stroke

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Background and Purpose—Limited data exist on childhood posterior circulation arterial ischemic stroke (PCAIS). We describe clinical and radiological features of childhood PCAIS to determine whether there are differences in infarct topography, vascular abnormalities, risk factors, and stroke subtypes when compared to adults.

Methods—Children with radiologically confirmed PCAIS were prospectively identified from August 2002 to February 2008. Infarcts were divided into proximal, middle, and distal posterior circulation segments utilizing an adult topographical classification system. Vascular abnormalities were described in terms of location, severity, and evolution over time. A pediatric modification of the TOAST classification system was used to define stroke subtypes.

Results—Twenty-seven (37%) of 73 children recruited to our registry had 34 radiologically confirmed PCAIS events. Infarct location was distal (25), middle (2), proximal (1), and multiple segments (6). Fourteen events were associated with single infarcts and 20 were associated with multiple infarcts. Magnetic resonance angiography was abnormal in 16 of 25 children with PCAIS at presentation with stenosis (8) or occlusion (8). Vascular lesions progressed (5), transiently worsened before stabilizing (2), remained stable or improved (8), or normalized (1) over 12 months. Stroke subtypes included nonprogressive steno-occlusive cerebral arteriopathy (9), cardioembolic (4), dissection (3), Moyamoya syndrome (3), other determined (4), probable (1), and undetermined etiology (3). Fifty-two percent of children had recurrent posterior (6) or anterior (8) strokes.

Conclusions—Nonprogressive arteriopathies are the most common cause of childhood PCAIS, usually affecting distal segments. Atherosclerosis-related risk factors do not play an important role in stroke causation. PCAIS is frequently associated with recurrent events. (Stroke. 2010;41:2201-2209.)

Key Words: arteriopathy ◼ cardiac ◼ childhood ◼ etiology ◼ infarct ◼ posterior circulation ◼ risk factors ◼ stroke ◼ vertebrobasilar

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.1–3 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,4 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.4

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.5 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 arteries5 (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.6

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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 events were associated with single infarcts and 20 events were associated with multifocal infarcts. Infarcts were confined to the distal posterior circulation segment in 25 (73%) of 34 events. All but 2 distal segment infarcts were in the posterior cerebral arteries territory, with 8 involving the proximal P1 segment and 15 involving the P2 segment.
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 2. Infarct Topography and Vascular Imaging Findings for 34 Posterior Circulation Events in 27 Children

<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>
</tr>
</thead>
<tbody>
<tr>
<td>1 (i)</td>
<td>D</td>
<td>L P2 PCA</td>
<td>S</td>
<td>L P2 occlusion</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>1 (ii)</td>
<td>D</td>
<td>R P2 PCA</td>
<td>S</td>
<td>R P2 occlusion, L P2 stenosis</td>
<td>N</td>
<td>R P2 stenosis, L P2 stenosis</td>
<td>No</td>
</tr>
<tr>
<td>2</td>
<td>D</td>
<td>L P1 PCA</td>
<td>S</td>
<td>L P1 stenosis</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>3 (i)</td>
<td>P, M, D</td>
<td>Bilateral PICA, AICA, SCA, P2 PCA</td>
<td>Mu</td>
<td>BA stenosis with distal occlusion, R AICA, bilateral PCA stenosis</td>
<td>NP</td>
<td>BA, L PCA stenosis</td>
<td>No</td>
</tr>
<tr>
<td>3 (ii)</td>
<td>D</td>
<td>L P2 PCA</td>
<td>S</td>
<td>L P1 stenosis</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>4</td>
<td>D</td>
<td>L P2 PCA</td>
<td>S</td>
<td>L P1 stenosis</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>D</td>
<td>R PCOM</td>
<td>S</td>
<td>R ICA occlusion at trifurcation to ACA, MCA and PCOM</td>
<td>NP</td>
<td>R ICA/trifurcation critical stenosis</td>
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</tr>
<tr>
<td>6 (i)</td>
<td>P, M, D</td>
<td>L PICA, AICA, SCA</td>
<td>Mu</td>
<td>Normal</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>6 (ii)</td>
<td>M, D</td>
<td>BA, R AICA, SCA, R PCA</td>
<td>Mu</td>
<td>BA stenosis, PICa, AICA, SCA origins not visible, P1 occlusion</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>7</td>
<td>D</td>
<td>L P1 PCA</td>
<td>Mu</td>
<td>L P1 occlusion</td>
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<td>L P2 stenosis</td>
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<td>Yes</td>
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<tr>
<td>9</td>
<td>D</td>
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<td>S</td>
<td>L P2 occlusion</td>
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<td>NP</td>
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<tr>
<td>10 (i)</td>
<td>M, D</td>
<td>BA, R AICA, R SCA, bilateral P2 PCA</td>
<td>Mu</td>
<td>Normal</td>
<td>N</td>
<td>Dissection</td>
<td>R vertebral dissection§</td>
</tr>
<tr>
<td>10 (ii)</td>
<td>D</td>
<td>Bilateral P1 PCA</td>
<td>Mu</td>
<td>N</td>
<td>Dissection</td>
<td>R vertebral dissection§</td>
<td>Yes</td>
</tr>
<tr>
<td>11</td>
<td>M, D</td>
<td>L AICA, SCA, R AICA, bilateral P1 PCA</td>
<td>Mu</td>
<td>L P1 occlusion, R PCA stenosis</td>
<td>N‡</td>
<td>L PCA stenosis</td>
<td>L vertebral dissection</td>
</tr>
<tr>
<td>12 (i)</td>
<td>P</td>
<td>R PICA</td>
<td>Mu</td>
<td>N</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>12 (ii)</td>
<td>P, D</td>
<td>R PICA, L PCA</td>
<td>Mu</td>
<td>N</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>12 (iii)</td>
<td>D</td>
<td>L P2 PCA</td>
<td>Mu</td>
<td>N</td>
<td>N‡</td>
<td>R vertebral dissection</td>
<td>No††</td>
</tr>
<tr>
<td>13</td>
<td>D</td>
<td>R P1 PCA</td>
<td>S</td>
<td>Thrombosed BA aneurysm, R P1 occlusion</td>
<td>NP</td>
<td>Thrombosed BA aneurysm, P1 occlusion</td>
<td>Yes</td>
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<tr>
<td>14</td>
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<td>L P2 PCA</td>
<td>S</td>
<td>Bilateral MCA occlusions, P1 stenoses, collaterals</td>
<td>NP</td>
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<tr>
<td>15</td>
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<td>Mu</td>
<td>Bilateral ICA, MCA, ACA, PCA stenoses with collaterals</td>
<td>NP</td>
<td>ICA, MCA, ACA, PCA stenoses with collaterals</td>
<td>Yes</td>
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<tr>
<td>16 (i)</td>
<td>D</td>
<td>R P2 PCA</td>
<td>Mu</td>
<td>R PCA occlusion with collaterals</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
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<tr>
<td>16 (ii)</td>
<td>D</td>
<td>L P2 PCA</td>
<td>Mu</td>
<td>L PCA occlusion with collaterals</td>
<td>N</td>
<td>NP</td>
<td>NA</td>
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<tr>
<td>17</td>
<td>D</td>
<td>Bilateral P2 PCA</td>
<td>Mu</td>
<td>Bilat ICA, MCA, ACA, PCA stenoses with collaterals</td>
<td>NP</td>
<td>ICA, MCA, ACA, PCA stenoses with collaterals</td>
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</tr>
<tr>
<td>18</td>
<td>D</td>
<td>R P2 PCA</td>
<td>S</td>
<td>R P2 PCA stenosis FMD changes in anterior circ</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
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<tr>
<td>19</td>
<td>D</td>
<td>R P1 PCA</td>
<td>S</td>
<td>NP*</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
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<tr>
<td>20</td>
<td>D</td>
<td>L P1 PCA</td>
<td>S</td>
<td>L P1 occlusion</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>21</td>
<td>D</td>
<td>L P2 PCA, L MCA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>22</td>
<td>D</td>
<td>L SCA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>23</td>
<td>D</td>
<td>L P1 PCA</td>
<td>S</td>
<td>NP‡</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>24</td>
<td>D</td>
<td>R P1 PCA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>25 (i)</td>
<td>M</td>
<td>L BA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
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<td>NA</td>
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<tr>
<td>25 (ii)</td>
<td>M</td>
<td>R BA</td>
<td>S</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Yes</td>
</tr>
<tr>
<td>26</td>
<td>D</td>
<td>L P2 PCA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
<tr>
<td>27</td>
<td>D</td>
<td>R P2 PCA</td>
<td>S</td>
<td>N</td>
<td>NP</td>
<td>NP</td>
<td>NA</td>
</tr>
</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 cerebellar artery; R, right; S, single; SCA, superior cerebellar artery.

*Left ventricular assist device prevented acute MRA imaging.
†PCA 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 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 infant topography may be influenced by ethnicity. Almost all the infarcts in our pediatric series involved the distal segments of the vertebral-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.

Embolic stroke 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-
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 thrombophilias, 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 vertebralbasilar 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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儿童后循环缺血性卒中的研究

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背景和目的：目前关于儿童后循环动脉缺血性卒中(PCAIS)的相关研究报道有限，本研究通过对儿童后循环动脉卒中的临床表现、影像学特点进行分析，探讨儿童与成人PCAIS在梗死部位、血管畸形情况、危险因素及卒中亚型等方面的差异。

方法：收集2002年8月至2008年2月在皇家儿童医院73例经影像学证实的儿童PCAIS患者临床资料进行前瞻性研究，应用成人血管定位分类系统，依据后循环梗死部位，将患儿分成近段组、中段组、远段组，同时血管畸形依据畸形部位、严重程度及随时间进展情况分别进行阐述分析。应用针对儿科改良版的TOAST分型方法对儿童PCAIS进行卒中亚型分型。

结果：本组募集到的73例儿童病例，确定PCAIS27名(37%)，影像学上可见34个病灶，其中动脉病变部位近段25例，中段2例，远段1例，多节段6例；血管事件中14个累及单病灶，20个累及多病灶。25名PCAIS患儿中16例MRA发现异常，其中动脉狭窄8例，动脉闭塞8例。该16例患儿经过12个月的随访调查结果显示：血管病变进展者5例，暂时进展后稳定者2例，血管稳定或改善者8例，恢复正常者1例。25名PCAIS患儿按卒中亚型分类，非进展的脑血管狭窄闭塞动脉病9例，心源性栓塞4例，动脉夹层剥离3例，烟雾样血管病3例，其他明确病因者4例，可能病因1例以及3例不明原因者。52%的儿童PCAIS有再发后循环(6例)或前循环梗死(8例)。

结论：非进展动脉病是儿童PCAIS最常见病因，常会累及后循环动脉的远段。动脉粥样硬化相关危险因素并非儿童PCAIS的重要原因，而儿童PCAIS存在高复发率。

关键词：动脉病，心源性，儿童，病因，梗死，后循环，危险因素，卒中，椎基底动脉

全脑及局部性脑白质高信号体积与血压：
二变量全基因组连锁分析显示的遗传位点重叠

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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背景和目的：脑白质T2高信号(HWM)体积是大脑完整性的重要的神经影像学标志，且有高度遗传性。病理生理学研究已显示局部、室管膜及皮层下的HWM损害分别与脉压和动脉血压相关。我们通过对健康墨西哥裔美国人的HWM体积和血压进行二变量全基因组连锁分析，以确定其与两者相关的染色体区域。我们的目的是明确这些表型新的基因多向性定量性状位点，并复制之前在全脑HWM体积和血压测量中遗传学的发现。

方法：血压测量以及高分辨率(1 mm³)三维液体衰减反转恢复(FLAIR)成像测得的全脑、皮层下及室管膜的HWM病损体积作为局部定量表型，数据收集于San Antonio Family Heart研究中多个扩大家庭的357位成员(218位女性；平均年龄47.9±13.2岁)。

结果：二变量全基因组连锁分析确定了影响全脑及局部(室管膜)的HWM体积和脉压的有意义的定量性状位点，在染色体位置1q24的标志物D1S196和D1S1619之间，几个其他染色体区域(1q42, 10q24-q26和15q26)显示可能的关联。事后分析结果时排除了55个服用降压药者后再得出的结果，与整个队列研究的结果无本质区别。

结论：这一研究证实了几个预先观察的影响血压和大脑完整性的定量性状位点，并识别出位于染色体1q24的新的有意义的定量性状位点。这一遗传学结果强烈支持基因多向性的作用共同影响血压及脑白质的完整性。

关键词：脑，脑部成像，遗传学，高血压，脑白质疏松症，脑白质病，磁共振，核磁共振成像

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