Middle cerebral artery aneurysms in children: case series and review

Clinical article

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Object. Pediatric intracranial aneurysms are rare lesions that differ from their adult counterparts. Aneurysms involving the middle cerebral artery (MCA) are particularly challenging to treat in children, as they are often fusiform and cannot undergo direct clipping alone. The authors recently treated a patient with a heavily calcified, dysplastic, left-sided MCA aneurysm. The present study was performed to evaluate the authors’ previous operative and follow-up experience with these difficult lesions.

Methods. The authors performed a review of a prospectively maintained database of all aneurysms treated at Methodist Hospital in Indianapolis, Indiana, from January 1990 through November 2010. Relevant operative notes, clinical charts, and radiological reports were reviewed for all patients 18 years of age or younger.

Results. A total of 2949 patients with aneurysms were treated over the study period, including 28 children (0.95%). Seven children harbored MCA aneurysms. Five of these 7 aneurysms (71.4%) were fusiform. Two patients were treated with direct clipping, 2 underwent parent vessel occlusion without bypass, and 3 underwent aneurysm trapping with extracranial-intracranial vessel bypass. Long-term follow-up data were available in 6 cases. All 6 patients had a 1-year follow-up Glasgow Outcome Scale score of 5. Long-term radiological follow-up was available in 4 patients. One patient required a reoperation for a recurrent aneurysm 4 years after the initial surgery.

Conclusions. Middle cerebral artery aneurysms in children are often fusiform, giant, and incorporate the origins of proximal artery branches. Direct clipping may not be possible; trapping of the lesion may be required. Children seem to tolerate surgical trapping with or without bypass extremely well. Aggressive therapy of these rare lesions in children is warranted, as even patients presenting with a poor clinical grade may have excellent outcomes. Long-term surveillance imaging is necessary because of the risk of aneurysm recurrence. (DOI: 10.3171/2011.4.PEDS10583)

Key Words • fusiform aneurysm • middle cerebral artery • vascular disorders • aneurysm trapping • extracranial-intracranial bypass

Intracranial aneurysms in children differ from those in adults in frequency and morphology. Middle cerebral artery aneurysms in children are often large, ectatic, and fusiform. Direct clipping or endovascular coiling of these lesions may not be possible. Strategies for treating MCA aneurysms are well described in adults, but there are few reports describing long-term results in children.

Abbreviations used in this paper: ACoA = anterior communicating artery; EC-IC = extracranial to intracranial; ECA = external carotid artery; GCS = Glasgow Coma Scale; GOS = Glasgow Outcome Scale; ICA = internal carotid artery; MCA = middle cerebral artery; SAH = subarachnoid hemorrhage; STA = superficial temporal artery.

A 10-year-old child recently presented to our service with a heavily calcified giant MCA aneurysm. This case highlighted many of the surgical issues inherent in treating these difficult lesions. We present our experience in treating these challenging lesions.

Methods

Goodman Campbell Brain and Spine prospectively maintains a database for all aneurysms treated at Methodist Hospital in Indianapolis, Indiana. A retrospective review of this prospectively collected database was performed for the study period of January 1990 through November 2010. We reviewed all cases of patients ≤ 18
years of age treated for intracranial aneurysms. We briefly review our overall pediatric aneurysm experience for comparison. We specifically highlight our experience in treating aneurysms of the MCA. The study began after approval by the local institutional review board.

The clinical status at admission was recorded using the GCS scores and Hunt and Hess grades. The CT appearance was classified by Fisher grade. Outcomes are reported using the GOS, with a score of 5 indicating a neurologically normal outcome, 4 indicating disability but independence, 3 indicating disability, 2 indicating severe disability or vegetative state, and 1 indicating death. Clinical outcomes were recorded at hospital discharge and at 1 year.

Statistical analysis was performed using SPSS for Windows software version 18. A multivariate linear regression analysis was performed with rank-transformed data to assess the independent contributions of age and presenting GCS scores with the outcomes at discharge and 1 year. A nonparametric Mann-Whitney U-test was performed to assess the comparison of age between groups. Statistical significance was set at p = 0.05.

Results

From January 1990 through November 2010, we treated 2949 patients with intracranial aneurysms. There were 28 patients (0.95%) ≤ 18 years of age. One patient harbored 3 aneurysms; therefore, a total of 30 aneurysms were present in the 28 patients.

The median age of the 28 patients was 14 years (range 5–18 years). Anatomically, the most commonly involved vessels were the ICA (7 [23.3%] of 30 lesions) and MCA (7 [23.3%] of 30 lesions). Fourteen aneurysms (46.7%) were saccular and 11 (36.7%) were fusiform or dissecting.

Nineteen of the 28 patients (67.9%) presented with hemorrhage, 2 (10.5%) of whom suffered from rebleeding and died prior to treatment. Both of these patients had anterior circulation aneurysms (Case 20 with an anterior cerebral artery aneurysm and Case 27 with ACoA aneurysm [Table 1]). The patient in Case 13 had an aneurysm incidentally discovered on screening for sickle cell anemia. Two patients (7.1%) presented with cerebral infarction. One patient presented with new cranial nerve palsies. Another 5 patients (17.9%) had pseudoaneurysms associated with trauma. The clinical characteristics and outcomes of all patients are shown in Table 1.

Long-term clinical outcome data were available in 24 of the 26 surviving patients. At 1-year follow-up, 20 patients (83.3%) had a GOS score of 5, 2 (8.3%) had a GOS score of 4, 1 (4.2%) had a GOS score of 3, and 1 (4.2%) had a GOS score of 2. There were the 2 aforementioned deaths. Overall, 22 (91.7%) of the 24 patients had a good or excellent outcome. The patient with a GOS score of 2 suffered a severe head injury in a motor vehicle collision. His deficits were related to the original trauma; no new neurological deficits occurred in relation to the pseudoaneurysm. Overall, 16 (64.2%) of the 25 patients presenting with hemorrhage had a good or excellent outcome.

The discharge GOS score was the only independent predictor of 1-year outcome (partial rho = 0.577, p = 0.012). The presenting GCS score, age, Fisher grade, and presence of angiographic vasospasm were not statistically associated with 1-year outcome.

Middle Cerebral Artery Aneurysms

Seven patients harbored MCA aneurysms. Their median age of 10 years (range 5–13 years) was statistically significantly lower than the overall group (p = 0.01). Two patients were girls and 5 were boys. Six patients (85.7%) presented with hemorrhage; 1 presented with stroke. Two patients had saccular aneurysms, 1 of which was bilobed. Five of the 7 patients (71.4%) had fusiform aneurysms. Table 2 provides a summary of the presentations, Hunt and Hess grades, Fisher grades, and details of surgical treatment for patients with MCA aneurysms.

The 2 patients (Cases 1 and 7) with saccular aneurysms underwent direct surgical clipping. In each there was postoperative angiographic confirmation of aneurysm occlusion. These 2 patients also had imaging follow-up at 4 years. In the patient in Case 7, a recurrent aneurysm was present at the surgery site. The aneurysm was in a different orientation than the original. A second craniotomy, with direct clipping, was performed. Subsequent imaging 6 years after the second surgery showed no residual or recurrent aneurysm.

Five patients had fusiform aneurysms that could not be primarily clipped. Instead, these lesions were treated with surgical trapping. Two patients (Cases 10 and 16) underwent trapping alone, without bypass; both presented in severe clinical distress due to hemorrhage. The patient in Case 16 presented with a large intraparenchymal clot in addition to SAH. The clot obscured the aneurysm on the initial angiogram. Surgical evacuation of the clot was performed; unfortunately, rehemorrhage occurred. A repeated angiogram demonstrated the aneurysm. A second surgery was performed. During this procedure, the aneurysm ruptured and was trapped immediately. The STA was damaged during the first surgery and was considered unsuitable for a bypass.

Three patients (Cases 22, 23, and 28) underwent trapping and EC-IC bypass.

Minimum clinical follow-up of 1 year was available for 6 patients (Table 3). All 6 presented with hemorrhage; 3 presented in poor condition (Hunt and Hess Grade IV). All patients had good or excellent outcomes. The discharge GOS score was 5 in 3 patients, 4 in 1 patient, and 3 in 2 patients. Status in all 6 patients either remained or improved to a GOS score of 5 by 1 year. There was no statistical difference in 1-year clinical outcome between patients suffering from hemorrhage with MCA aneurysms and the overall group (p = 0.865, Mann-Whitney U-test).

All 6 underwent angiography in the immediate postoperative period, and 4 had long-term radiological follow-up ranging from 4 to 15 years (Table 3). One patient (Case 16) who had undergone surgical trapping without bypass had radiological follow-up at 15 years. Computed tomography angiography showed no recurrence or new aneurysm. In 1 patient who had undergone aneurysm trapping and EC-IC bypass, radiological follow-up at 9 years was obtained. This CT angiogram showed a patent bypass with no residual or recurrent aneurysm.
TABLE 1: Clinical characteristics and outcomes in 28 pediatric patients with aneurysms treated between 1990 and 2010*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Anatomical Location</th>
<th>Aneurysm Type</th>
<th>Presentation/Comorbidity</th>
<th>GCS Score at Presentation†</th>
<th>Treatment</th>
<th>Angiographic Vasospasm at Discharge</th>
<th>Angiographic Vasospasm at 1 Yr</th>
<th>GOS Score</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>6, F</td>
<td>rt MCA fusiform</td>
<td>SAH</td>
<td>10T clip</td>
<td>yes</td>
<td>5</td>
<td>5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>18, F</td>
<td>ACoA, ophthalmic, lt cavernous</td>
<td>saccular, saccular, saccular</td>
<td>SAH; carbamoyl phosphate deficiency</td>
<td>6T endovascular</td>
<td>no</td>
<td>3</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>17, F</td>
<td>feeding vessel for AVM</td>
<td>saccular</td>
<td>ICH</td>
<td>15 resection</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>17, F</td>
<td>PICA</td>
<td>traumatic pseudoaneurysm</td>
<td>IVH, SAH, trauma</td>
<td>4T endovascular</td>
<td>yes</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>16, F</td>
<td>lt ICA</td>
<td>traumatic pseudoaneurysm</td>
<td>trauma, CN palsy</td>
<td>14 endovascular</td>
<td>no</td>
<td>3</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>15, M</td>
<td>lt ICA</td>
<td>traumatic pseudoaneurysm</td>
<td>GSW, CN palsy</td>
<td>15 endovascular</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>12, M</td>
<td>rt MCA</td>
<td>saccular</td>
<td>SAH</td>
<td>14 clip</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>15, F</td>
<td>rt ICA</td>
<td>dissecting</td>
<td>CN palsy</td>
<td>15 balloon occlusion followed by clip ligation</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>17, F</td>
<td>lt VA</td>
<td>dissecting</td>
<td>postcoital SAH</td>
<td>15 endovascular</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>10, F</td>
<td>rt MCA</td>
<td>fusiform</td>
<td>SAH</td>
<td>15 trapping &amp; resection</td>
<td>yes</td>
<td>3</td>
<td>5</td>
<td></td>
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<tr>
<td>11</td>
<td>14, F</td>
<td>lt ICA</td>
<td>saccular</td>
<td>SAH</td>
<td>15 clip</td>
<td>no</td>
<td>5</td>
<td>5</td>
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<tr>
<td>12</td>
<td>17, F</td>
<td>lt pericallosal</td>
<td>saccular</td>
<td>SAH</td>
<td>15 clip</td>
<td>yes</td>
<td>5</td>
<td>5</td>
<td></td>
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<tr>
<td>13</td>
<td>14, M</td>
<td>lt ICA</td>
<td>saccular</td>
<td>sickle cell, incidental finding</td>
<td>15 clip</td>
<td>no</td>
<td>5</td>
<td>5</td>
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<tr>
<td>14</td>
<td>16, F</td>
<td>basilar</td>
<td>fusiform, giant</td>
<td>SAH, hydrocephalus</td>
<td>3 endovascular coiling &amp; vessel ligation</td>
<td>3</td>
<td>5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>14, M</td>
<td>rt ICA</td>
<td>pseudoaneurysm, MVA 6 mos prior to presentation</td>
<td>acute vision loss</td>
<td>15 trapping</td>
<td>no</td>
<td>5</td>
<td>lost to FU</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>13, M</td>
<td>lt MCA</td>
<td>saccular</td>
<td>SAH/ICH</td>
<td>5 rebleed; trapping after intraprop rupture</td>
<td>yes</td>
<td>3</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>13, M</td>
<td>lt ICA</td>
<td>saccular</td>
<td>SAH</td>
<td>7 clip</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>15, F</td>
<td>ACoA</td>
<td>saccular, giant</td>
<td>SAH</td>
<td>15 clip</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>18, M</td>
<td>ACoA</td>
<td>saccular</td>
<td>SAH</td>
<td>13 clip</td>
<td>no</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>9, F</td>
<td>lt ACA</td>
<td>saccular</td>
<td>SAH</td>
<td>3 rebleed; died before treatment</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>7, F</td>
<td>VA</td>
<td>dissecting</td>
<td>infarction</td>
<td>trapping</td>
<td>no</td>
<td>3</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>8, M</td>
<td>lt MCA</td>
<td>fusiform</td>
<td>SAH; familial history of aneurysms</td>
<td>15 trapping w/ EC-IC bypass</td>
<td>no</td>
<td>5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>5, M</td>
<td>rt MCA</td>
<td>fusiform</td>
<td>SAH, seizures</td>
<td>15 trapping w/ EC-IC bypass</td>
<td>no</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>24</td>
<td>6, M</td>
<td>rt pericallosal</td>
<td>traumatic pseudoaneurysm, severe head injury from MVA</td>
<td>incidentally discovered on imaging 9 mos after accident</td>
<td>endovascular</td>
<td>no</td>
<td>2 (deficits from head injury stable after aneurysm treatment)</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>
Case Illustration

Presentation and Examination. This otherwise healthy 10-year-old boy (Case 28, Table 1) began to experience tingling in the right side of his body before going to bed. He woke up the next morning with a dense right hemiplegia in the lower portion of his face and his extremities. He did not experience headache, nausea, or vomiting. He had relatively preserved sensation.

The patient’s medical history was unremarkable. There was no family history of intracranial pathology or aneurysms.

The initial CT scan showed a calcified mass in the left sylvian fissure without evidence of hemorrhage (Fig. 1A). A 21 × 14 × 18–mm aneurysm and a capsular stroke were demonstrated on MR imaging/MR angiography (Fig. 1B and C). Angiography showed that the aneurysm involved a segment of the proximal frontal trunk of the left MCA with mural thrombus (Fig. 1D). An injection of the ECA showed a robust frontal branch of the STA with a relatively small parietal branch (Fig. 1E).

We allowed the patient time to recover from the acute period after his stroke. His leg strength and facial movement improved with physical therapy.

Operation. Surgery was performed 3 weeks after the patient’s initial presentation. A standard pterional craniotomy was performed, with care to dissect both the frontal and parietal branch of the STA. The aneurysm calcification extended into the MCA parent artery (Fig. 2A), precluding primary clipping. Given the robust STA frontal branch, we planned an EC-IC bypass. A recipient temporal cortical arterial branch was selected (Fig. 2B). After examining the parietal branch of the STA, we did not believe it had sufficient size or flow to create a viable bypass.

After the bypass was performed, we clip occluded the M1 branch proximal to the aneurysm. Intraoperative angiography showed robust collateral flow filling the frontal lobe despite M1 occlusion (Fig. 2C). However, selective injection of the ECA showed that the aneurysm still filled from the bypass (Fig. 2D). Intraoperative indocyanine green angiography demonstrated both the continued filling of the aneurysm and the collateral filling of the cortical vessels (Fig. 2E). A second clip was placed distal to the aneurysm, completing the trapping (Fig. 2F). A second intraoperative angiogram was obtained with selective injection of both the ICA (Fig. 2G and H) and ECA (Fig. 2I). This showed complete exclusion of the aneurysm with good flow through the bypass. There was also significant collateral flow to both the frontal and temporal lobes.

Postoperative Course. The patient did not have any new postoperative neurological deficits. He was transferred to a rehabilitation center on postoperative Day 3. By the time of discharge to home, he had improved facial function, antigravity strength in the proximal upper extremities, and was ambulatory with a walker. By 3 months, he had Grade 4/5 strength in the leg and was able to walk without a walker. He had continued weakness of Grade 4/5 in the upper extremity and difficulty

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Anatomical Location</th>
<th>Aneurysm Type</th>
<th>Presentation/Comorbidity</th>
<th>GCS at Presentation†</th>
<th>Treatment</th>
<th>Angiographic Vasospasm at Discharge</th>
<th>Angiographic Vasospasm at 1 Yr</th>
<th>Acute Neurological Deficit at Discharge</th>
<th>Acute Neurological Deficit at 1 Yr</th>
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<tr>
<td>25</td>
<td>5, F</td>
<td>rt PCA</td>
<td>dissecting</td>
<td>SAH, SDH, hemiparetic, CN palsy</td>
<td>8T</td>
<td>endovascular vessel occlusion</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>ongoing‡</td>
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<tr>
<td>26</td>
<td>12, F</td>
<td>rt AICA</td>
<td>dissecting</td>
<td>SAH, hydrocephalus</td>
<td>8T</td>
<td>endovascular vessel occlusion</td>
<td>yes</td>
<td>yes</td>
<td>no</td>
<td>ongoing‡</td>
</tr>
<tr>
<td>27</td>
<td>16, M</td>
<td>ACoA</td>
<td>saccular</td>
<td>SAH</td>
<td>14</td>
<td>clipped, died prior to treat ment</td>
<td>yes</td>
<td>no</td>
<td>1</td>
<td></td>
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<tr>
<td>28</td>
<td>10, M</td>
<td>MCA</td>
<td>fusiform</td>
<td>SAH</td>
<td>15</td>
<td>trapping w/ EC-IC bypass</td>
<td>no</td>
<td>ongoing‡</td>
<td>4</td>
<td>ongoing‡</td>
</tr>
</tbody>
</table>

* AICA = anterior inferior cerebellar artery; AVM = arteriovenous malformation; CN = cranial nerve; FU = follow-up; GSW = gunshot wound; IVH = intraventricular hemorrhage; ICH = intracerebral hemorrhage; MVA = motor vehicle accident; PCA = posterior inferior cerebellar artery; SAH = subarachnoid hemorrhage; SDH = subdural hematoma; VA = vertebral artery.† Scores accompanied by a T indicate the patient was intubated.‡ Less than 1 year of follow-up.
with fine motor motion of the hand. He could completely close the right eye and had an emotional smile, although he had difficulty with volitional lower-extremity facial movement.

Discussion

Intracranial aneurysms in children differ from those in adults. Aneurysms are relatively rare in the pediatric population. In a cooperative study examining 6368 aneurysms, only 0.69% occurred in children. Pediatric aneurysms may arise in the context of systemic disease such as connective-tissue disorders, autosomal dominant polycystic kidney disease, hereditary hemorrhagic telangiectasia, sickle cell disease, PHACES syndrome, syphilis, neurofibromatosis Type 1, Klippel-Trenaunay syndrome, tuberous sclerosis, fibromuscular hyperplasia, and Kawasaki disease.

Children have a relatively higher rate of traumatic aneurysms than adults. Five (17.9%) of the 28 patients in our series had traumatic pseudoaneurysms. Our experience with endovascular treatment of proximal traumatic aneurysms has been previously published.

Anatomically, a higher percentage of pediatric aneurysms occur at the ICA bifurcation, MCA, or in the posterior circulation. There is a greater percentage of large or giant aneurysms in children. Patients with giant aneurysms may present with seizures, mass effect, headaches, or cranial nerve palsies. Some series show a higher rate of unruptured aneurysms in children. In our series, however, 19 (67.9%) of 28 patients presented with nontraumatic hemorrhage. Other series have also shown a high rate of hemorrhagic presentation. Many reports have shown a sex prelidence, but ours did not show a significant difference.

Clinically, children tend to have a better outcome after aneurysmal SAH than adults. In previous series, good outcomes varied between 63% and 95% in children who presented with a good clinical grade. In our series, 16 (84.2%) of 19 hemorrhage patients had good or excellent outcomes at 1 year. Two patients died; both suffered from a rehemorrhage prior to treatment. Rebleeding is perhaps more common in the pediatric aneurysm population than in adult aneurysms. In a series of 21 children with SAH, Proust et al. reported a rebleeding rate of 52.4% in a time frame of 6 hours–15 days after the initial hemorrhage.

There are multiple potential reasons why children may experience a better clinical outcome than adults, despite presenting with hemorrhage. Children typically do not have the same comorbidities as adults; they generally do not have a smoking history and are free of cardiac and atherosclerotic diseases. Children often have a more robust collateral circulation than adults. Although children may have angiographically demonstrated vasospasm, the aforementioned factors make them less susceptible to permanent deficits. In our series, the presence of angiographically demonstrated vasospasm was not statistically correlated with 1-year clinical outcome.

Whereas fusiform aneurysms are relatively common in children, they are rare in adults, with an incidence of less than 0.1% in autopsy or radiology series. Patients with fusiform aneurysms may present with mass effect, thromboembolic events, or hemor-

### TABLE 2: Presentation, anatomy, and treatment in 7 patients with MCA aneurysms

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Hemorrhage</th>
<th>Aneurysm Size (mm)</th>
<th>Description</th>
<th>Location</th>
<th>Fisher Grade</th>
<th>Anatomical Projection</th>
<th>Surgical Technique</th>
<th>Angiographic Vasospasm</th>
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<tr>
<td>1</td>
<td>yes</td>
<td>&gt;25</td>
<td>M1</td>
<td>2</td>
<td>IV superior</td>
<td>direct clipping</td>
<td>yes</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>yes</td>
<td>7</td>
<td>MCA bifurcation</td>
<td>2</td>
<td>I posterior</td>
<td>1999: direct clipping for ruptured aneurysm</td>
<td>no</td>
<td></td>
</tr>
<tr>
<td>no</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2003: re-do craniotomy for new aneurysm formation (unruptured)</td>
<td>no</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>yes</td>
<td>fusiform</td>
<td>M2</td>
<td>4</td>
<td>IV circumferential dilation</td>
<td>craniotomy for trapping/resection of aneurysm; shunt for hydrocephalus</td>
<td>yes</td>
<td></td>
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<tr>
<td>16</td>
<td>yes</td>
<td>8 (broad-based neck incorporating anterior origin of M2 branch)</td>
<td>M2</td>
<td>4</td>
<td>IV anterior, superior</td>
<td>intraop rupture; trapping w/o bypass</td>
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</tr>
<tr>
<td>22</td>
<td>yes</td>
<td>23, fusiform dilation</td>
<td>M2</td>
<td>4</td>
<td>I circumferential dilation</td>
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<td>fusiform</td>
<td>M3</td>
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<td>no</td>
<td></td>
</tr>
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<td>28</td>
<td>no</td>
<td>fusiform MCA segment w/ 21 × 14 × 18-mm aneurysm</td>
<td>MCA bifurcation</td>
<td>1</td>
<td>superior</td>
<td>trapping w/h EC-IC bypass</td>
<td>no</td>
<td></td>
</tr>
</tbody>
</table>

* H & H = Hunt and Hess.
TABLE 3: Clinical and radiological follow-up of 7 patients treated for MCA aneurysms*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Postop Imaging/Result</th>
<th>FU Imaging/Result</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>angiogram; completely clipped w/ no residual aneurysm</td>
<td>4-yr CT angiogram showed no residual aneurysm</td>
<td>upper visual quadrant deficit on 1-yr exam, but visual field tests normal; currently an honor roll student in regular classes &amp; involved in sports</td>
</tr>
<tr>
<td>7</td>
<td>1999: angiogram; completely clipped w/ no residual aneurysm; 2003: angiogram showed complete clipping of 2nd aneurysm w/ no residual</td>
<td>4-yr angiogram showed recurrent aneurysm pointing in opposite direction as original; angiogram 6 yrs after 2nd op showed no residual or recurrent aneurysm</td>
<td>able to compete successfully as a wrestler in HS; graduated from college; currently employed, w/o neurological difficulties</td>
</tr>
<tr>
<td>10</td>
<td>angiogram showed complete ligation of parent branch</td>
<td>none</td>
<td>initially lost to FU, patients was contacted 18 yrs after hemorrhage; is attending college</td>
</tr>
<tr>
<td>16</td>
<td>angiographic occlusion of MCA parent branch; no aneurysm filling; retrograde filling of opercular branches</td>
<td>15-yr CT angiogram showed no residual or new aneurysm</td>
<td>required rehab &amp; speech therapy, but able to return to school; graduated college; currently employed, but has residual difficulty w/ headaches &amp; time/money mgmt skills</td>
</tr>
<tr>
<td>22</td>
<td>angiographic occlusion of parent branch w/ no filling of aneurysm, patent bypass</td>
<td>none</td>
<td>moved out of state; no deficits on 1-yr FU exam</td>
</tr>
<tr>
<td>23</td>
<td>no residual or refilling, patent bypass</td>
<td>6-yr angiogram: no residual or recurrent aneurysm, bypass patent; 9-yr CT angiogram: no residual or recurrent aneurysm, bypass patent</td>
<td>presented w/ lt hemiparesis that resolved w/ rehab; is attending high school &amp; doing well academically; involved in recreational sports w/o difficulty</td>
</tr>
<tr>
<td>28</td>
<td>intraprop angiogram showed occlusion of aneurysm w/ good collat &amp; retrograde filling of MCA territory (see Fig. 2)</td>
<td>none</td>
<td>presented w/ cerebral infarction; hemiplegia improving w/ physical therapy; had 1 seizure in postop period</td>
</tr>
</tbody>
</table>

* HS = high school; mgmt = management; rehab = rehabilitation.

There is a predilection for fusiform aneurysms in certain anatomical locations. Some authors have reported a higher incidence of these lesions in the posterior circulation, although other series have demonstrated an even distribution between anterior and posterior circulations. In the anterior circulation, the MCA is consistently the most commonly involved vessel.

Etiology of MCA Aneurysms

The MCA forms relatively early in embryological development. It appears earlier than most other vessels and supports a relatively large blood flow volume. Any congenital weakness may lead to a dilated, ectatic vessel segment early in a child’s life. Indeed, aneurysms in the very young tend to involve the MCA. In a review of aneurysms in children younger than 1 year of age, Buis et al. found that the prevalence of MCA aneurysms was 3 times higher than aneurysms of any other vessel. In our series, the median age of the patients with MCA aneurysms (10 years) was less than that (14 years) of the overall group.

The early high-flow burden, combined with congenital defects in the parent vessel wall, may explain the relatively high incidence of fusiform aneurysms of the MCA. Cedzich et al. described a case of an 11-month-old child with 13 aneurysms in 1 segment of the left MCA. Pathological analysis revealed an incomplete internal elastic lamina and a thinned media. In a review of the literature, Ferrante et al. reported on the pathological examination of 23 aneurysms in children younger than 5 years of age. The aneurysm walls revealed thinned fibrous tissue and no internal elastic and muscular lamina. In a proposed classification system of nonatherosclerotic fusiform aneurysms, Mizutani et al. described segmental arterial ectasia (Type 2) or dolichoectatic dissecting aneurysms (Type 3) based on the internal elastic lamina and the extent of intimal thickening. In our series, 5 (71.4%) of the 7 patients with MCA aneurysms had fusiform lesions. This was higher than the incidence of fusiform aneurysms (36.7%) of our overall pediatric aneurysm group.

Treatment of MCA Aneurysms

The natural history of an incidentally discovered fusiform MCA aneurysm is unknown. Some authors have argued that fusiform aneurysms are less likely to rupture than saccular aneurysms in children. Therefore, observation may be an appropriate choice in selected patients. Hetts et al. observed 18 patients with long-segment vascular dysplasia for an average of 41 months. There were no hemorrhages during the follow-up period, although in 3 cases the authors documented growth of the aneurysm. The most comprehensive study of fusiform MCA aneurysms in both adults and children was reported by Day et al. In their review of 102 aneurysms from their institution and the literature, the hemorrhage rates of small, large, giant, and serpentine aneurysms were 80%, 62%, 23%, and 14%, respectively. Four of the patients were younger than 1 year of age.
The decision to observe these lesions should be made with caution. In our series, 4 of the 5 patients with fusiform MCA aneurysms presented with hemorrhage. Other authors have described an extremely poor natural history for fusiform aneurysms. Suzuki et al. reviewed 18 cases of serpentine aneurysms of the MCA and concluded that “the outcome of patients were definitely better in the group treated actively than in the group with conservative therapy.” Observation alone has inherent risk. Isono et al. reported a case of an adult patient diagnosed with a dissecting MCA aneurysm without hemorrhage. The patient was observed. He suffered a fatal hemorrhage 4 years after diagnosis. We agree with Day et al. that aggressive treatment should be performed in patients with symptomatic lesions. We also encourage consideration of repair in incidental lesions.

Middle cerebral artery aneurysms may have anatomical characteristics that are unfavorable for endovascular treatment. These include a wide neck, fusiform aneurysm shape, and critical perforators arising from the base. For these reasons, we have preferred open surgery. However, the scope and capabilities of endovascular therapy continue to evolve. Newer techniques such as stenting and remodeling may allow expanded treatment of fusiform aneurysms in the future, although the question of the durability of endovascular treatment in children remains.

Surgical strategies include direct clipping and possible reconstruction of the vessel wall, proximal vessel occlusion, wrapping, bypass alone, or trapping with bypass. Direct clipping was performed in Cases 1 and 7. Despite angiographically “complete” clipping, a recurrence developed in Case 7 four years after the original surgery. The patients in Cases 10, 16, 22, 23, and 28 had fusiform or ectatic dilation of a vessel segment or a broad-based aneurysm neck incorporating part of the parent vessel. In our experience, these aneurysms are difficult or impossible to repair primarily. The defect in the vessel wall is circumferential with abnormal, thinned walls. The entire wall is affected; primary reconstruction must therefore allow flow through the abnormal vessel segment. Other authors have previously cited a high rate of regrowth and rebleeding of these aneurysms after direct clipping, despite excellent initial angiographic results.

Drake and Peerless described their extensive experience in treating 120 giant and/or fusiform aneurysms. In their comprehensive series, 13 of 55 giant MCA aneurysms were fusiform. An overall good or excellent out-

**Fig. 1.** Radiological studies obtained in a 10-year-old child presenting with hemiplegia. **A:** Axial CT scan showing a calcified mass without hemorrhage (arrow). **B** and **C:** Axial MR image MR angiogram demonstrating a $21 \times 14 \times 18$-mm lesion at the MCA bifurcation (circle, B) with stroke (C). **D:** Cerebral angiogram demonstrating a fusiform aneurysm (arrow). Note that much of the bulk of the aneurysm does not fill secondary to mural thrombus and calcification. **E:** External carotid artery angiogram showing a robust frontal branch of the STA (arrow) with a smaller parietal branch.
come was reported in 8 patients (62%). Five patients were 19 years of age or younger. The 2 patients treated with clip-based vessel reconstruction and 2 patients treated with proximal occlusion and bypass grafting fared well. It is noteworthy that the bypass grafts in both patients were found to be occluded on subsequent studies, yet the patients had good clinical outcomes. The authors commented that “neither patient developed any persisting deficit because of a luxuriant leptomeningeal collateral circulation for the anterior and posterior cerebral arteries, which filled the MCA circulation retrogradely, even back in the M2 branches to the region of the aneurysms.” One 16-year-old patient treated with aneurysm wrapping had a poor outcome.

Proximal vessel occlusion may cause thrombosis of the aneurysm and has been used with good success. Lv et al. reported on 16 pediatric patients who underwent proximal vessel occlusion for fusiform aneurysms, including 4 in the MCA distribution. Follow-up angiography demonstrated occlusion of the parent vessel and retrograde collateral perfusion in all cases. All patients experienced a good neurological outcome. With proximal occlusion alone, there may be a chance of retrograde aneurysm filling from collateral flow. In our example case, the aneurysm filled from the bypass after the proximal clip was applied (Fig. 2D and E). It is unknown if this would have filled from collateral flow without the bypass.

Surgical trapping excludes the aneurysm from the circulation both proximally and distally. Trapping has been reported in both adults and children. The safety of proximal vessel occlusion or surgical trapping depends heavily on the extent of collateral flow. The collateral circulation of lenticulostriate perforators is an especially important factor in the treatment of MCA aneurysms. As mentioned previously, children generally lack atherosclerotic vascular disease and tend to have a very robust collateral capability.

Cerebral revascularization with EC-IC bypass may be used in conjunction with vessel occlusion or trapping. Children may have recipient or donor vessels that are too small for bypass, but pioneers in the procedure have shown technical success in vessels as small as
Pediatric middle cerebral artery aneurysms

1 mm. The age at which a bypass becomes technically feasible varies, although the first case of STA-MCA bypass in moyamoya disease was performed by Yükselgil in a 4-year-old child. Suzuki et al. used 5 years of age as a cutoff between direct and indirect revascularization in the treatment of moyamoya disease. Other authors have shown technical and clinical success of EC-IC bypass in children with moyamoya disease. There are few reports of the durability of EC-IC bypass in children. Zhang et al. reported on 2 pediatric patients with giant intracranial aneurysms treated with saphenous vein EC-IC bypass. One patient harbored a fusiform MCA aneurysm. In both cases, the authors observed long-term bypass patency and actual growth of the grafts with growth of the child. Our patients tolerated surgical trapping very well. There were no new neurological deficits or strokes after these procedures. Our preference is to perform an EC-IC bypass whenever technically feasible. However, both patients who underwent aneurysm trapping without bypass had excellent clinical results.

Clinically, all our patients with MCA aneurysms had good to excellent outcomes. All 6 patients with follow-up of 1 year had a GOS score of 5. The follow-up duration ranged from 1 to 18 years (average 9.5 years). These 6 children had excellent clinical outcomes despite the fact that all presented with hemorrhage; on admission, status in 3 patients was Hunt and Hess Grade IV. The clinical outcomes at 1 year did not differ between patients treated for MCA aneurysm rupture–related hemorrhage and the overall pediatric group.

Radiological Follow-Up

The recurrence rate for aneurysms after either clipping or endovascular coiling continues to be a matter of study. Although there is an increasing body of literature on the recurrence rate after coiling, there are remarkably few publications detailing the recurrence rate after clipping. In a widely quoted study, David et al. estimated that the annual recurrence rate after microsurgical clipping was approximately 0.5%. Their study involved primarily adult patients. In their review of 35 pediatric aneurysms with long-term angiographic follow-up, Karakala et al. reported a higher recurrence rate of 8.6% and an estimated annual risk rate of 2.6%. In their series, 1 sacular aneurysm exhibited additional growth in a delayed fashion. We agree with previous authors that long-term surveillance is necessary, even after complete obliteration of the aneurysm by either surgical or endovascular means.

The long-term radiological outcomes in our patients with MCA aneurysms are shown in Table 3. Immediate postoperative angiography revealed complete obliteration of the aneurysm in Case 7. However, the patients suffered a recurrence of the aneurysm 4 years later. One of the 2 patients who underwent surgical trapping without bypass underwent radiological follow-up 15 years after the procedure. The studies showed no residual or new aneurysm. One of the 2 patients with aneurysm trapping and EC-IC bypass underwent follow-up 9 years after the procedure. The CT angiogram demonstrated a patent bypass and no residual or recurrent aneurysm.

Limitations of the Study

Clearly, the rarity of pediatric aneurysm makes definitive conclusions difficult to draw. We did not emphasize the statistical outcomes in this manuscript, as small numbers preclude definite conclusions.

Conclusions

MCA aneurysms present unique challenges in children. The patients are relatively younger and the aneurysms are often fusiform. Surgical clipping with or without distal bypass is well tolerated in children, likely due to robust collateral circulation. Children treated for aneurismal SAH generally fare better than adults. Children with surgically treated MCA aneurysms had good clinical results, despite a high incidence of hemorrhage and fusiform aneurysms in our experience. Long-term radiographic follow-up is warranted in children, as there is the possibility of recurrence even after successful surgical clipping.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Fulkerson. Acquisition of data: Fulkerson, Voorhies, Payner, Leipzig, Redelman. Analysis and interpretation of data: Fulkerson, Payner, Leipzig, Horner, Cohen-Gadol. Drafting the article: Fulkerson, Voorhies, Horner, Cohen-Gadol. Critically revising the article: Fulkerson, Voorhies, Payner, Leipzig, Horner, Cohen-Gadol. Administrative/technical/material support: Redelman. Study supervision: Fulkerson.

Acknowledgments

The authors thank Erin Giesler, R.N., B.S.N., C.N.R.N., for assistance with data collection. Amy A. Fulkerson for editorial support, and Mohammadali M. Shoja, M.D., for statistical assistance.

References


J Neurosurg: Pediatrics / Volume 8 / July 2011


Manuscript submitted December 23, 2010. Accepted April 19, 2011.

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