Assessment and Management of Paediatric Intracranial Aneurysm. A single center experience.

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Authors: A. J. Schmitt¹, F. Robertson¹, S. Brew², S. Bhaté¹, V. Ganesan¹, A. Rennie¹, London/UK, ²Auckland/NZ
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Aims and objectives

Intracranial aneurysms are rare in children, with a prevalence of 0.5% to 4.6% quoted in the literature (1-4).

They differ in many ways from intracranial aneurysms seen in adults. Previous studies have shown, that they have a higher incidence of posterior circulation aneurysms and are more predominant in males (2,5-7). Furthermore, the incidence of giant and fusiform aneurysms is higher and anterior circulation aneurysms are more commonly located at the terminal internal carotid artery (ICA) in comparison to adults (6), where the most common location is at the anterior cerebral artery/ anterior communicating artery (Acom/ACA) junction.

Children are not exposed to the same environmental factors, which are risk factors for the adult population, but often have underlying conditions, predisposing them to aneurysm formation. Common diseases found in children are connective tissue disorders, such as Ehlers-Danlos syndrome, polycystic kidney disease, aortic coarctation, immunocompression or various infections (8,9,10).

To further contribute to the understanding of this rare disease, the authors reviewed their clinical experience with 22 paediatric aneurysms, which occurred in 21 patients under the age of 18 years.
Methods and materials

Between 2005 and 2013, 21 paediatric patients with 22 intracranial aneurysms presented to our neurovascular service. Patients with associated aneurysms seen in Vein of Galen malformations or arteriovenous malformations were not included in the analysis.

The authors retrospectively reviewed all radiological studies. The preoperative computerized tomography (CT) scans or magnetic resonance imaging (MRI) were analysed for signs of subarachnoid haemorrhage (SAH). This diagnosis was confirmed with CT findings of blood within the subarachnoid space or basal cisterns or with blood seen on susceptibility weighted MRI imaging.

The preoperative cross sectional and angiographic imaging was analysed to further characterise the location, size and morphology of the aneurysms.

The aneurysms were classified considering their vessel wall pathology as previously described by Krings et al. (11) and also the classification described by Lasjaunias et al. (12).

The following categories were distinguished: Classic saccular aneurysm (berry-like), segmental ectasia, dissecting aneurysm, mycotic or microbial (fusiform), immunodeficiency related, neoplastic and traumatic.

The follow up imaging, which was obtained at various time points, was assessed for degree of aneurysm occlusion, possible re-perfusion of the aneurysm and other findings, such as complications arising from the treatment and their long term sequelae.

The clinical records (initial clinical presentation, underlying disease, risk factors, outcome), intraoperative reports and all clinical correspondence was were reviewed retrospectively for each patient.

All patients presenting with SAH were graded according to the Hunt and Hess rating scale for subarachnoid haemorrhage (13). As per consensus, Grade 0-3 were considered to indicate a good preoperative status and grade 4-5 were thought to represent a poor status.
The post-operative outcome was assessed using the Glasgow outcome score (GOS), where GOS 1-3 was considered a poor outcome and 4-5 indicated a good performance status (14).
Results

Demographics and aneurysm characteristics:

From 2005 to 2013, 21 patients were referred to the neurovascular service at Great Ormond Street Hospital with a diagnosis of intracranial aneurysm.

This included 8 female and 13 male resulting in a female to male ratio of 1: 1.63. The median age at presentation was 9.4 years (range: 0.1 -16.2 years).

62% of patients presented with evidence of subarachnoid blood on initial imaging. The average Fisher grade was 3.7 (Fisher grade unknown in 1 patient). Most of the patients presented with associated headache (52%) or collapse (33%). Other common presentations were seizure (24%), vomiting (19%) and ptosis (10%) and 14% of patients were diagnosed incidentally.

Three of the 22 aneurysms were due to trauma, 1 was found in a patient with immunodeficiency, 1 was an ectatic section of a vessel with an associated saccular aneurysm, 7 were classic saccular aneurysms, 6 were fusiform and 3 were due to dissection, with a dissection flap seen in 1 of them.

One aneurysm was due to segmental ectasia, a concept introduced by Lasjaunias et al. in 2000 (15).

The location of the aneurysm in relation to their aetiology is demonstrated in Figure 1.

The authors identified relevant underlying conditions in four patients, as further demonstrated in Figure 2.

Treatment:

The majority of the paediatric intracranial aneurysms were treated endovascularly (77.2%), 2 patients with 2 aneurysms (9.1%) underwent surgery and 3 aneurysms (13.6%) were not amenable to treatment.
Eight aneurysms were amenable to treatment with coils only, 2 patients were treated with stent assisted coiling and 3 patients received treatment with flow diverting stents.

One traumatic aneurysm was treated with glue. 3 further aneurysm, one fusiform ICA aneurysm, one dissecting aneurysm of the PCA and one fusiform left vertebral artery aneurysm, were treated with occlusion of the parent vessel.

One patient presenting with a fusiform MCA aneurysm, surgically received a superficial temporal artery to MCA (STA-MCA) bypass and one middle cerebral artery aneurysm was treated by the means of microsurgical clipping.

Three patients did not receive treatment. Out of those, one had a poor pre-operative status and died 3 days post presentation. Another patient presented with fusiform aneurysms due to HIV and as suggested in the literature intracranial aneurysms associated with HIV show a low propensity for progression and often improve on treatment of the underlying condition(16). The patient was treated with antiretroviral therapy and the imaging appearances improved.

The third patient presented with mycotic aneurysms due to an underlying endocarditis, and treatment of his infection lead to improvement of the lesions.

Four patients experienced procedural complications. Of these, 3 had a favourable outcome and one died.

One of the patients with a traumatic, 2 mm, right MCA aneurysm, was treated with glue occlusion. During the procedure a drop of glue embolised into the distal MCA causing a left MCA infarct, leaving the patient with a mild right sided hemiparesis.

Another patient with an enlarging giant fusiform left MCA aneurysm was treated with flow diverting stents. During the procedure the capture coil detached and occluded a small temporal left M2 branch. Fortunately the patient did not show any deficit on follow up.

The third patient presented with a complex, fusiform aneurysm involving the left V4 intradural segment and basilar artery. He was treated with a construct of flow diverting stents and coiling and at the end of the procedure the left posterior inferior cerebellar artery (PICA) was seen to fill through the patent aneurysm neck. The patient however showed a small left PICA infarct on follow up imaging, but they remained intact throughout.
Unfortunately, two of our patients died. One patient was not treated due to a poor pre-operative status and they died shortly after admission. Another patient, who presented with a grade 4 SAH and a small right MCA aneurysm, was treated endovascularly, but a further bleed was demonstrated on the immediate post-operative flat-panel volume CT and they died the next day.

Of the 17 patients that underwent treatment, 15 patients scored 1 on the Raymond-Roy Occlusion Classification scale (17) on their last imaging follow up. Out of those 15 patients 2 had to undergo retreatment to achieve these results. Stable mild rest perfusion of the aneurysm neck (Raymond-Roy 2) was seen in 2 patients.

Figure 4 demonstrates a case of a 14 year old patient diagnosed and treated for a large left vertebral artery aneurysm.

**Recommendations for Clinical assessment:**

In order to gain more information and also to tailor the treatment and estimate the prognosis for each individual patient, thorough preoperative clinical assessment is necessary. Up to date there are no guidelines as how to assess paediatric aneurysm patients but the authors follow a protocol, which aims to capture all possible underlying conditions.

Clinical history taking, including presenting symptoms, known past medical history and family history, is the first step in the preoperative assessment of a newly admitted patient. This is especially important to exclude traumatic aneurysms and have an indication about the general health of the patient and their immediate family.

Thorough haematological and microbiological blood tests are crucial and at the authors’ institution haematological markers for vasculitis (P-ANCA, C-ANCA) are routinely requested in addition to standard blood tests (FBC, WBC, CRP, Coagulation screen).

A physical examination as well as a specialist cardiac review including an echocardiogram and an electrocardiogram as minimal standard in addition to a routine Abdominal ultrasound are part of the standard workup. This step is important to assess for underlying conditions, such as aortic stenosis, coarctation and polycystic kidney disease (PCKD).

The authors routinely refer their paediatric aneurysm patients to the genetics team, which after thorough assessment, decide on necessary investigations.
For all patients, the authors aim to perform a preoperative MRI and MRA next to a noncontrast CT and CT angiogram. Extracranial vascular imaging from the aortic arch upwards is also performed to assess for possible segmental ectasia.

Evaluation of the cross sectional imaging is an important step and often visualised extravascular pathologies (such as underlying trauma, thromboembolic stroke, meningitis, sinusitis, osteomyelitis or syndromic signs) guide the team to an underlying diagnosis.

The protocol varies for emergency cases, where urgent treatment is necessary. For example in a child with a suspected or diagnosed SAH, the initial imaging and clinical workup starts with a noncontrast CT scan of the head. In case of a negative CT head, but clinical suspicion of a SAH, a lumbar puncture should be performed. Positivity should prompt a CT angiogram or MR angiogram to identify the source of the SAH and to decide on the best treatment.

Recommendations for treatment:

In our institution, all treatment options are discussed for each individual patient in a multidisciplinary team setting including interventional neuroradiologists, neurosurgeons and neurologists. The decision is based on the presumed aetiology of the aneurysm, its morphology including size, shape, difficulty of access and the clinical condition of the patient. Despite emerging treatment methods and a focus on reconstructive endovascular treatment over deconstructive surgical or endovascular treatment, we consider all techniques as can be seen from our cohort.

Major differences are seen to the adult population when planning treatment, such as the significant size difference of intracranial vessels and the reduced availability of suitable treatment devices.

Another difference that needs to be considered are possible ischaemic events, especially when planning deconstructive techniques such as endovascular or surgical large parent vessel occlusion. Prior to large vessel occlusion in adults, a balloon test occlusion as described by Günther et al in 1981 (18) is performed under local anaesthesia to assess the response to the vessel sacrifice and predict possible ischaemic events.

Preoperative knowledge of the feasibility of permanent occlusion of the internal carotid artery is crucial in children as well, but as the procedure would not be tolerated under
local anaesthetics, an angiographic balloon occlusion under GA assessing the venous delay is performed (19).

**Recommendations for follow up:**

Children with intracranial aneurysm require appropriate follow up regardless of the aneurysm aetiology. This is crucial due to their long life expectancy and the risk of further aneurysm. For every patient, an individual follow up plan is created after discussion within the multidisciplinary team using general considerations and the individual patient history.

The time frame for follow up depends largely on the type of aneurysm and the treatment modality.

For a coiled, adult type saccular aneurysm, a cerebral angiogram and noninvasive cross-sectional MRA are performed 6 months post treatment. If the aneurysm remains occluded, further noninvasive imaging is performed at 2 years post treatment and then in decreasing intervals from 2 to 5 yearly well into their adult life.

The follow up imaging for clipped saccular aneurysm is with MRA and a cerebral angiogram performed at 6 months post treatment. If occlusion is confirmed, the patient can be discharged and no further follow up imaging is needed.

A similar follow up regime is performed for single mycotic aneurysms. The neurovascular team at the authors’ institution tends to occlude single mycotic aneurysm using glue and performs a cerebral angiogram and MRA at 6 months post treatment. Assuming no radiological complication is evident, the patient can be subsequently discharged from radiological follow up.

In case of multiple mycotic aneurysms, the consensus is not to treat the aneurysms by endovascular or surgical means, but to treat the underlying condition. More frequent cross-sectional imaging (preferably MRI/MRA) should be performed, the frequency of which is decided on an individual basis.

In case of a dissecting aneurysm and parent vessel occlusion, follow up imaging at 6 months is performed (MRA and cerebral angiogram) and the patients are subsequently discharged. In case of a coiled dissecting aneurysm, further follow up is needed at 2 years post treatment and then 2 to 5 yearly.
Fusiform aneurysms treated with flow diverting stents are followed up differently. They are initially imaged at 6 months post treatment with both MRA and cerebral angiogram. Further MRA and cerebral angiogram is performed at 12 months post treatment and assuming no complications, the patient then receives noninvasive MRI imaging at 2 years post treatment and then 2-5 yearly. The difference to the follow up protocol stems from the relative lack of data for long term flow diverting stent performance. Another point that needs to be considered in patients treated with flow diverters is the anticoagulation regime and when to stop it. Generally, at Great Ormond Street Hospital we tend to stop one of the antiplatelet agents, Clopidogrel, after 6 months post procedure and the second one, Aspirin, after 1 year post procedure, although this largely depends on the imaging findings and the clinical condition of the patient.
**Figure 1:** This figure demonstrates the different aneurysm aetiologies identified in our cohort and the locations of the aneurysm. ICA: internal carotid artery, MCA: middle cerebral artery, ACA: anterior cerebral artery, A1: A1 part of anterior cerebral artery; PCA: posterior cerebral artery, Pcom: posterior communicating artery, VA: vertebral artery

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### Aneurysm patients with underlying conditions

<table>
<thead>
<tr>
<th>Underlying disease</th>
<th>Location of Aneurysm</th>
<th>Type</th>
<th>Multiple Aneurysm</th>
<th>Clinical Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Noonan Syndrome</td>
<td>R MCA</td>
<td>saccular</td>
<td>no</td>
<td>SAH</td>
</tr>
<tr>
<td>HIV</td>
<td>R ICA/ACA</td>
<td>segmental ectasia</td>
<td>yes</td>
<td>incidental finding</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(immunodeficiency)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aortic stenosis with Aspergillus endocarditis</td>
<td>L/R MCA/ACA</td>
<td>fusiform (mycrobial)</td>
<td>yes</td>
<td>SAH</td>
</tr>
<tr>
<td>Hemimegalencephaly with S aureus endocarditis</td>
<td>R ICA</td>
<td>fusiform (mycrobial)</td>
<td>no</td>
<td>irritable</td>
</tr>
</tbody>
</table>

**Figure 2**: Aneurysm patients with underlying conditions. This figure demonstrates all underlying pathologies identified in our cohort and visualizes the most important aneurysm features. MCA: middle cerebral artery, ICA: internal cerebral artery, ACA: anterior cerebral artery, R: right, L: left.

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<table>
<thead>
<tr>
<th>GOS</th>
<th>Description</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Death</td>
<td>2 (9.6%)</td>
</tr>
<tr>
<td>2</td>
<td>Persistent vegetative state</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>3</td>
<td>Severe disability</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>4</td>
<td>Moderate disability</td>
<td>1 (4.8%)</td>
</tr>
<tr>
<td>5</td>
<td>Low disability/Intact</td>
<td>17 (81%)</td>
</tr>
<tr>
<td></td>
<td>Lost to follow up</td>
<td>1 (4.8%)</td>
</tr>
</tbody>
</table>

**Figure 3**: Outcome of paediatric aneurysm rated by Glasgow Outcome Scale (GOS)

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Figure 4: 14 year old patient presenting with 2 year history of bitemporal headache and a 5 week history of vomiting and dizziness. Examination was unremarkable other than mild nystagmus on left lateral gaze. A) CT angiogram revealed a large aneurysm arising from the left vertebral artery causing compression of the brain stem (B) and mild hydrocephalus (not visualised). The subsequent angiogram (C,D) again demonstrated a giant fusiform left intradural vertebral artery aneurysm extending to the vertebro-basilar confluence with both antegrade and retrograde filling. No PICA or anterior spinal contribution from the distal vertebral artery was identified and the decision was made to coil occlude the left vertebral artery to minimise antegrade flow. E) demonstrates the location of the coil cast and F) shows minimal residual filling of the aneurysm neck during right vertebral injection. CTA and MRA imaging on the 2nd postoperative day confirm near exclusion of the aneurysm, although there remained mass effect of the aneurysm. The patient experienced transient bulbar signs postoperatively, which resolved with physiotherapy during the admission.

Fig. 4

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Conclusion

Paediatric intracranial present a very rare pathology and due to the high variability of aneurysm sizes, shapes and location, their epidemiology is still poorly understood.

Underlying diseases are frequently diagnosed and children are not exposed to the usual risk factors accounting for aneurysm in the adult population. Endovascular treatment of paediatric aneurysms is the preferred approach because it offers both reconstructive and deconstructive techniques, however all treatment choices need to be discussed for each individual patient to ensure the best outcome. As long term durability of endovascular repair is still uncertain, long term radiological and clinical follow up is pertinent (20).
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