Intracranial Arterial Aneurysms in Childhood*

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The present paper is concerned with aneurysms of the cerebral arterial circulation in very young patients. One of the very earliest significant reports which provided any clinical data in addition to a description of the postmortem anatomy was that of a German pathologist named Eppinger in 1871. He discussed the case of a 15-year-old boy who was unusually active in gymnastics. This boy suddenly collapsed in the midst of physical exertion and died 3 days later without improving. Autopsy revealed stenosis of the isthmus of the aorta. Indeed, the principal subject of Eppinger's paper was the aortic malformation. However, he also reported free blood at the base of the brain at autopsy, and presented a lucid anatomical picture of a ruptured saccular aneurysm of the right anterior cerebral artery.

According to McDonald and Korb, ruptured intracranial aneurysm was first reported in 1778 by Biumi, of Milan, "... who gave a clear clinical description of the disease now called 'spontaneous subarachnoid hemorrhage' and described the ruptured aneurysm observed at autopsy." However, it was not until a number of years later, in 1887, that Eppinger clearly stated his opinion that rupture of a saccular aneurysm of the cerebral arteries could occasionally be a cause of so-called "cerebral hemorrhage of unknown etiology" in children. Bull made similar observations in Norway in 1877. The therapeutic implications of these opinions, however, remained unrealized for virtually half a century.

In 1922, Strassman, also from Germany, reported the case of a 13-year-old boy who dropped dead while playing football. Autopsy in this instance revealed coarctation of the aorta and a ruptured aneurysm of the middle cerebral artery. Again, this was reported as rather an unfortunate complication of coarctation of the aorta. In 1927, Woltman and Shedlin in the United States, reported postmortem findings on 32 patients with coarctation of the aorta who had manifested some abnormal neurological symptoms during life. They found that the cause of death in 37.5 per cent of these patients was cerebral hemorrhage, but they did not know how many might have had intracranial arterial aneurysms.

In 1928, Abbott collected 20 cases of coarctation of the aorta in which cerebral hemorrhage was the cause of death. In no less than 7 of these a ruptured intracranial aneurysm was definitely demonstrated to be the source of the fatal hemorrhage.

Although there has been general consensus that saccular intracranial aneurysms involving the circle of Willis and its major branches represent a congenital defect of the media of the arterial wall, it is uncommon to find these lesions becoming symptomatic neurologically, or providing the source of spontaneous intracranial hemorrhage, until adult life. It is not clear what causes such aneurysms to bleed. Certainly, lesions which remain very small indeed may be the source of fatal hemorrhage in middle or late adult life; the cause, therefore, is not always size in itself. It is not necessarily acute stress that causes hemorrhage, since arterial aneurysms seem to bleed frequently when patients are at ease as well as when they are in the midst of strenuous physical or emotional exertion. It

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is not intravascular hypertension, since sac-
cular aneurysms notoriously bleed in normo-
tensive young adults. It has always been an
enigma, therefore, why these lesions, appar-
ently of prenatal origin, so rarely become
manifest during childhood.

McDonald and Korb, for instance, col-
lected 1,125 verified aneurysms from 407
sources prior to 1938. In this group, they re-
ported one 1½-year-old infant, 2 under 5
years of age, and only 28 patients or 2.5 per
cent of this large group of aneurysms under
15 years of age. In 1964, Laitinen reported
from Finland that among 1,175 patients with
verified subarachnoid hemorrhage, 35, or 3
per cent, were 15 years of age or younger.
However, a sacular aneurysm was demon-
strated in only 9 of these children, that is, in
1.3 per cent of all the patients with aneu-
rysms. It is of interest that only 1 of Lai-
tinen’s patients was under 10 years of age.

Isolated case reports of aneurysms in early
childhood have appeared in the surgical
literature. Thus, a 16-month-old infant with
hemorrhage from a middle cerebral aneu-
rysm was reported in 1960 by Kimbell et al. In
1961, Jones and Shearburn described clippin
a middle cerebral artery aneurysm in a 4-week-old infant, and in the same year
Jane reported a ruptured aneurysm of the
posterior inferior cerebellar artery causing
death in a 1-year-old child.

Because rupture of an arterial aneurysm
as the cause of spontaneous intracranial
bleeding in childhood is so rare, the experi-
ence of any one clinic or any one surgeon has
necessarily been limited. The infrequency of
such lesions on our pediatric neurosurgical
service at The Children’s Hospital Medical
Center has certainly been impressive, par-
ticularly when one realizes the intensive
arteriographic studies now performed in the
presence of any suggestion of non-traumatic
intracranial hemorrhage, the meticulous
postmortem examinations routinely carried
out in all fatal intracranial hemorrhages, and
the large number of children seen with car-
diovascular anomalies. The latter group in-
cludes more than 700 patients with coarcta-
tion of the aorta who have been studied and
treated in our hospital.

A much more common source of sponta-
neous intracranial hemorrhage in the pediat-
ric age group, in our experience as well as
that of others, has been hemorrhage from
cortical arteriovenous malformations. In
addition, we have been perplexed, as have
others, by the large group of children with
massive spontaneous intracerebral hemor-
rhage of unknown etiology. A few of these
catastrophic hemorrhages have been iden-
tified in more recent years by careful post-
mortem histological studies as arising in the
microscopic subependymal arteriovenous
and venous abnormalities termed by Craw-
ford and Russell “cryptic vascular hamar-
tomas.” Many others have remained unex-
plained.

Clinical Material and Results

Because of the rarity of arterial aneurysms in
early life, it seemed worthwhile to place on record
briefly a personal experience with 13 of these les-
sions in children treated during the last 12 years.

In 1958, I first saw a very active 13-year-old boy from Nova Scotia, who is of interest in view
of Eppinger’s original description referred to
above. This boy while playing in the snow had the sudden onset of headache, vomiting, stiff neck and
dizziness. He improved after 4 days of bed-rest
until a month later the same findings recurred
after straining at stool. This time he developed a
left hemiparesis. On examination, a loud systolic bruin was heard over the back; there were palpable
intercostal vessels, absent femoral pulses, and
the characteristic radiological findings of coarcta-
tion of the aorta. Carotid arteriograms revealed
an aneurysm arising at the junction of the right
anterior cerebral and the anterior communicating
arteries. The cerebrospinal fluid collected during
lumbar puncture was xanthochromic and con-
tained crenated red blood cells.

Under hypothermia and induced hypotension,
with the medial tip of the right frontal lobe re-
sected, the aneurysm was exposed and ligated.
The postoperative course was uneventful and
followed by complete recovery. Approximately 6
months later this boy returned to our hospital and
had the coarctation repaired without difficulty
(Dr. Samuel Schuster). He has remained well and
is now neurologically and intellectually within
normal limits 6 years later. I like to think that
Eppinger would have been thrilled if he could
have lived 90 more years to witness the successful
modern treatment of a boy so similar to his original case.

During the 12-year period covered by the present series, not a single additional child was found to have an incidental arterial aneurysm demonstrated at autopsy for any cause, nor by arteriography carried out in the study of epi-

lepsy, increased intracranial pressure, syncope, or any other neurological abnormality.

As will be noted from Table 1, the age range of this group, with the exception of 1 slightly older patient, was from 1 to 13 years. It is an interesting but probably entirely unimportant fact in such a small group, that 12 of these children were boys and only 1 a girl.

All but 1 of these children exhibited spontaneous hemorrhage as the initial symptom in a previously healthy child. None of these 12 children had had earlier convulsions, cranial nerve abnormalities, evidences of increased intracranial pressure, or abnormal neurological symptoms or signs of any kind. The initial hemorrhage in 11 of the patients was a massive one, accompanied by coma and grossly bloody spinal fluid.

The distribution of the lesions in this small series of childhood aneurysms is probably no different than might be encountered with this number of consecutive aneurysms in any age group (Fig. 1). There were 11 above the tentorium and 2 in the posterior fossa; 5 arose in the anterior communicating artery complex, 4 from the internal carotid, 2 from the middle cerebral, 1 from the bifurcation of the basilar, and 1 from the left posterior inferior cerebellar artery. To our knowledge, there were no patients with multiple aneurysms in this series, although total angiography was not performed in each patient.

In 5 of the 12 children whose aneurysms had ruptured, there was in addition to subarachnoid hemorrhage, an intracerebral clot. Indeed, 2 of

### Table 1

<table>
<thead>
<tr>
<th>No.</th>
<th>Patient</th>
<th>Age</th>
<th>Location of Aneurysm</th>
<th>Operation</th>
<th>Result</th>
<th>Time after Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M.N.</td>
<td>1 yr. 4 mos.</td>
<td>Rt. middle cerebral</td>
<td>Excision</td>
<td>Dead</td>
<td>2 days</td>
</tr>
<tr>
<td>2</td>
<td>D.B.</td>
<td>1 yr. 11 mos.</td>
<td>Lt. posterior inferior cerebellar</td>
<td>Excision</td>
<td>Alive, well</td>
<td>5 mos.</td>
</tr>
<tr>
<td>3</td>
<td>S.W.</td>
<td>4 yrs. 2 mos.</td>
<td>Rt. middle cerebral</td>
<td>Excision</td>
<td>Dead</td>
<td>3 mos.</td>
</tr>
<tr>
<td>4</td>
<td>S.B.</td>
<td>6 yrs. 4 mos.</td>
<td>Lt. internal carotid</td>
<td>Ligation of aneurysm</td>
<td>Alive, well</td>
<td>5 yrs.</td>
</tr>
<tr>
<td>5</td>
<td>V.T.</td>
<td>8 yrs. 6 mos.</td>
<td>Rt. internal carotid bifurcation</td>
<td>Clipping of aneurysm</td>
<td>Alive, well</td>
<td>7 yrs.</td>
</tr>
<tr>
<td>6</td>
<td>W.M.*</td>
<td>10 yrs. 5 mos.</td>
<td>Lt. internal carotid</td>
<td>Ligation in neck</td>
<td>Alive, well</td>
<td>2 yrs.</td>
</tr>
<tr>
<td>7</td>
<td>T.I.*</td>
<td>11 yrs. 9 mos.</td>
<td>Bifurcation of basilar</td>
<td>None</td>
<td>Dead, 2 days after bleeding</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>G.R.</td>
<td>12 yrs. 6 mos.</td>
<td>Anterior communicating</td>
<td>Ligation of aneurysm</td>
<td>Alive, well</td>
<td>4 yrs.</td>
</tr>
<tr>
<td>9</td>
<td>G.J.</td>
<td>13 yrs.</td>
<td>Anterior communicating</td>
<td>Clipping of aneurysm</td>
<td>Alive, well, retarded</td>
<td>12 yrs.</td>
</tr>
<tr>
<td>10</td>
<td>R.Q.*</td>
<td>13 yrs.</td>
<td>Anterior communicating</td>
<td>Ligation of aneurysm</td>
<td>Alive, well</td>
<td>6 yrs.</td>
</tr>
<tr>
<td>11</td>
<td>J.B.</td>
<td>13 yrs.</td>
<td>Anterior communicating</td>
<td>Clipping of aneurysm</td>
<td>Alive, well</td>
<td>7 yrs.</td>
</tr>
<tr>
<td>12</td>
<td>W.G.</td>
<td>13 yrs.</td>
<td>Lt. internal carotid</td>
<td>Ligation of carotid</td>
<td>Alive, well</td>
<td>10 yrs.</td>
</tr>
<tr>
<td>13</td>
<td>C.P.</td>
<td>17 yrs. 7 mos.</td>
<td>Anterior communicating</td>
<td>Clipping of aneurysm</td>
<td>Alive, well, slow</td>
<td>5 mos.</td>
</tr>
</tbody>
</table>

* Also had coarctation of the aorta.

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Fig. 1. Distribution of intracranial aneurysms in this series of 13 children.
these patients, children of 1 and 8 years of age, were operated upon as emergencies; large clots were evacuated before the aneurysm itself was diagnosed. In one of these, a middle cerebral artery aneurysm was found and excised at the time of the emergency operation for clot (Fig. 2), and in the other an aneurysm at the bifurcation of the internal carotid protruding into the frontal lobe was revealed by arteriography 1 week after a large intracerebral blood clot had been evacuated. This aneurysm was then clipped successfully and the patient has developed well in the succeeding 7 years.

One characteristic of this group of children with aneurysms, other than the 2 just mentioned with large intracerebral clots, appeared to be their ability to improve spontaneously and progressively and not to bleed again in the early period after a severe hemorrhage. Thus, 8 children in spite of rather desperate condition on admission to the hospital, were improved to a "good" to "excellent" general status by the time of intracranial operation 7 days to 6 weeks later. These 8 children all survived surgery and are still alive and well, although at least 3 show evidence of some mental retardation.

All of these aneurysms were demonstrated satisfactorily by preoperative arteriography except the one already mentioned that was encountered at emergency operation for clot, and one other 11-year-old child who was admitted to the hospital moribund and decerebrate, with grossly bloody cerebrospinal fluid. Carotid arteriograms were done on this child promptly, showing no aneurysm but suggesting spasm of all vessels in the anterior part of the circle of Willis; the basilar system was not visualized. This child died in 6 days without improvement, and autopsy revealed rupture of a saccular aneurysm at the crotch of the basilar artery. Incidentally, this child also had a mild degree of coarctation of the aorta.

Operative treatment was carried out in all of the patients in this series except the child with the basilar aneurysm just described. One 10-year-old child with a left internal carotid aneurysm arising at the origin of the posterior communicating artery had internal carotid ligation in the neck performed in another clinic. This was followed by severe hemiparesis which gradually recovered. Investigation of subsequent headaches and vascular hypertension led to the discovery of coarctation of the aorta. This was repaired uneventfully in our clinic (Dr. Robert Gross) with an excellent result to date. Another 13-year-old boy with an unusual internal carotid aneurysm extending both intra- and extracranially, had trap ligation of the internal carotid artery in the neck and intracranially. He has remained well with only minimal dysphasia but normal intelligence over 10 years. Each of the other 10 patients had either intracranial clipping, ligation, or excision of the aneurysms.

One patient, a boy of 6 years and 4 months, may be of special significance in that he proved at operation to have a large atherosclerotic plaque in the internal carotid artery at the origin of a saccular aneurysm arising near the posterior communicating artery. The child did not seem to have any evidence of generalized atherosclerosis or other metabolic disturbance.

One of the most interesting lesions among this group of patients was in a child of 23 months admitted moribund and opisthotonic with a diagnosis of meningitis. Grossly bloody cerebrospinal fluid was recovered at lumbar puncture. Right brachial arteriograms showed normal right carotid and vertebral vessels but suggested spasm of the basilar artery; left carotid arteriogram revealed normal vessels. The possibility of an intraspinal vascular malformation was entertained but myelography showed that the entire spinal canal was normal. Finally, because of continued concern about this evidence of basilar artery spasm and the history of a really massive subarachnoid hemorrhage, a left brachial arteriogram was also carried out. This revealed an arterial aneurysm arising from the left posterior inferior cerebellar artery. At operation the aneurysm lay within the lumen of the 4th ventricle and was easy to excise (Fig. 3). The baby made an excellent recovery and seems to be completely normal.
cerebral clot from rupture of a middle cerebral artery aneurysm (Fig. 2). The operation was conducted under assisted respiration and the child never improved, succumbing within 36 hours of respiratory and cardiac failure. The other fatality was also a patient with a middle cerebral artery lesion; this was a 4-year-old child, stuporous and hemiplegic on admission. The aneurysm was excised and a large intracerebral clot evacuated (Fig. 4). In this child also, a yellow, atherosclerotic plaque was grossly visible in the parent vessel at the origin of the aneurysm, just as if the patient had been 64 years old instead of 4. This child made an excellent postoperative recovery, but subsequently re-entered the medical service with fever and stupor. He was a known congenital cardiac patient with a ventricular septal defect but no cyanosis. He died 3 months after the intracranial operation without responding to treatment for severe endocarditis accompanied by repeated cultures of alpha streptococci from the blood stream. Total cerebral angiography after the operation had confirmed obliteration of the aneurysm and showed no evidence of other intracranial vascular lesions. This was felt to be a true mycotic aneurysm.

Summary

Perhaps the only deduction to be drawn from this brief clinical review is that arterial
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Intracranial aneurysms do rupture in children with production of both subarachnoid and intracerebral hemorrhage and should therefore be looked for in the presence of spontaneous intracranial bleeding. Operative treatment of these lesions in children seems on the whole to be well tolerated, and the surgical results in our hands have been as good or better than in a comparable group of adults.

In the absence of arteriographic evidence of a large intracranial clot, there is probably justification for waiting until substantial general improvement in the child’s condition has taken place, since in this group of young patients with presumably good vessels elsewhere, there has not seemed to be any evidence of early recurrent bleeding. Moreover, the hazards of major intracranial surgery in the comatose child with increased intracranial pressure are many. It also appears that the common adult complication of distant cerebral infarction secondary to the vascular spasm associated with rupture of the aneurysm may not occur as often in children with their more resilient cerebral arteries.

Postoperative mortality was limited to 1 infant who was operated upon as an emergency because of profound coma and cessation of spontaneous respirations due to a large intracerebral clot from rupture of a middle cerebral aneurysm. Hypothermia was used in only 3 of the older children; it is not clear that it contributed anything.

We have reached no particular conclusions as to why some arterial aneurysms become symptomatic and bleed in childhood. Among this group of 13 patients, 3 had associated coarctation of the aorta with obvious intracranial hypertension, 1 had a mycotic aneurysm associated with a ventricular septal defect and endocarditis, 1 had extension of an aneurysm into the extracranial portion of the internal carotid artery, and 2 had grossly demonstrable segmental atherosclerotic plaques in the parent artery near the neck of the aneurysm.

Addendum

One case, a child of 11 years with an aneurysm of the anterior communicating artery, treated during this same time interval, was unintentionally omitted from the original manuscript. This aneurysm was ligated intracranially and the child has remained well for 5 years. The total number of childhood intracranial aneurysms in this series, therefore, should be 14.

References

1. Abbott, M. E. Coarctation of the aorta of the adult type. II. A statistical study and historical retrospect of 200 recorded cases, with autopsy, of stenosis or obliteration of descending arch in subjects above the age of two years. Amer. Heart J., 1928, 3:392–421; 574–618.