intracranial Arterial Aneurysms in Childhood*

Donald D. Matson, M.D.
Neurosurgical Service, Children’s Hospital Medical Center and Dept. of Surgery, Harvard Medical School, Boston, Mass.

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. Bulx 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 Shelden 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
Intracranial Arterial Aneurysms in Childhood

is not intravascular hypertension, since sac- cular aneurysms notoriously bleed in normoten sive 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, collected 1,125 verified aneurysms from 407 sources prior to 1938. In this group, they reported 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 clipping a middle cerebral artery aneurysm in a 4-week-old infant, and in the same year, 1961, Jones and Shearburn reported clipping 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 cardiovascular 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 spontane- ous intracranial hemorrhage in the pediatric 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 postmortem histological studies as arising in the microscopic subependymal arteriovenous and venous abnormalities termed by Crawford 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- ions 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 bruit was heard over the back; there were palpa- ble intercostal vessels, absent femoral pulses, and the characteristic radiological findings of coarctation 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 contained 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