THE SURGICAL TREATMENT OF ARNOLD-CHIARI MALFORMATION IN ADULTS
AN EXPLANATION OF ITS MECHANISM AND IMPORTANCE OF ENCEPHALOGRAPHY IN DIAGNOSIS

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The Arnold-Chiari malformation is a deformity of the hindbrain in which a tongue-like projection of the cerebellar tonsils protrudes through the foramen magnum down onto the cervical cord. The caudal portion of the 4th ventricle likewise is elongated downward. The upper cervical nerve roots pursue a cephalad direction, giving the impression that the brain stem has been drawn down through the foramen magnum.

The malformation was named after Arnold\(^6\) and Chiari\(^6\) who independently described the anomaly in 1894 and 1895 respectively. Schwalbe and Gredig\(^23\) published an embryologic and anatomic treatise on the subject in 1907.

We have operated upon 17 patients, adults and adolescents, with the Arnold-Chiari malformation and our experience suggests that the fundamental mechanism is obstructive hydrocephalus with resulting foraminal herniation of the hindbrain.

Interest in the anomaly was revived in 1935 when Russell and Donald\(^22\) discussed its importance as a possible cause of internal hydrocephalus in spina bifida. They presented 10 examples of myelomeningocele associated with hydrocephalus and Arnold-Chiari malformation in infants. McConnell and Parker\(^16\) reported surgical treatment of 5 patients ranging in age from 10 to 32 years with 2 successful results. All were found to have some degree of hydrocephalus. D'Errico\(^5\) in 1939 described 10 cases of myelomeningocele with hydrocephalus in infants. The myelomeningocele was repaired in 7, and when hydrocephalus followed, operation for the Arnold-Chiari malformation was done. Three remained in good health for a period of 2 years thereafter. He stressed the advisability of doing a suboccipital decompression in all cases of spina bifida associated with myelomeningocele and hydrocephalus. In 1939, McKissock\(^17\) stated that he routinely treated hydrocephalus with associated myelomeningocele by first decompressing the Arnold-Chiari malformation, and that this frequently benefited both the hydrocephalus and the sac protrusion.

Chamberlain\(^5\) in 1939 described concomitant findings of Arnold-Chiari malformation with platybasia and described the latter condition as follows: "The morphologic changes shown by roentgenograms give the impression of softening of the base of the skull and moulding through the force of gravity. It is as though the weight of the head has caused the ears to approach the
shoulders, while the cervical spine, refusing to be shortened, has pushed the floor of the posterior fossa upward into the brain space." Gustafson and Oldberg\textsuperscript{10} in 1940 called attention to the relationship between Arnold-Chiari malformation, Klippel-Feil syndrome, syringomyelia and platybasia. They pointed out the possibility of benefiting syringomyelia by surgically unblocking the fluid pathways between the cranial and spinal cavities.

List\textsuperscript{15} in 1941, in a scholarly discussion of neurologic syndromes accompanying developmental anomalies of the occipital bone, atlas and axis, reported 3 cases of Arnold-Chiari malformation operated upon successfully. He refers to the foraminal herniation of cerebellar tissue as Arnold's deformity, while the associated posterior bulging or kinking of the medulla he calls Chiari's deformity. He stresses the importance of posterior dislocation of the dens as a factor in the production of neurologic signs.

Ogryzlo\textsuperscript{16} in 1942 reported 7 operative cases, 4 in infants associated with spina bifida. Three were in adults, of which 2 improved after operation and 1 died. He suggested that hydrocephalus might be responsible for the Arnold-Chiari deformity and that the hydromyelia could be the result of fluid being forced down the central canal.

Ray\textsuperscript{21} in 1942 described 2 cases of congenital and 2 of acquired platybasia with involvement of the central nervous system. He also commented upon the similarity of the appearance of the brain and cord in cases of platybasia and in Arnold-Chiari malformation.

Lichtenstein\textsuperscript{13,14} in 1943 likewise called attention to the similarity of the appearance of the cerebellar tonsils in platybasia and in Arnold-Chiari malformation. He believes that the accumulating cerebrospinal fluid in these cases may dilate the central canal with resulting hydromyelia and may also perforate the ependymal lining with the formation of a syrinx.

Ingraham and Scott\textsuperscript{11} in 1943 presented an excellent review of the literature and described 20 cases of Arnold-Chiari malformation in infants associated with myelomeningocele. The majority of their patients had associated microgyria and cranialacumia but only 2 had platybasia.

There are several other case reports of this interesting malformation, including those of Aring,\textsuperscript{2} Penfield and Coburn,\textsuperscript{20} Adams, Schatzki, and Scoville,\textsuperscript{1} Ecker and Ferguson,\textsuperscript{9} Shryock,\textsuperscript{24} Wagner,\textsuperscript{26} Moore,\textsuperscript{18} Bucy and Lichtenstein,\textsuperscript{4} Steele,\textsuperscript{25} and Chorobski.\textsuperscript{7}

We have reviewed the cases of Arnold-Chiari malformation reported in the literature and, exclusive of infants, found 17 cases verified and treated surgically. There are several other probable cases classified under other diagnoses, particularly platybasia.

Exclusive of infants, the files of the Cleveland Clinic Hospital of the last 10 years contain the records of 17 patients with Arnold-Chiari malformation who were treated surgically. The ages of these patients varied from 14 to 54 years. The mean duration of symptoms was 3 years, but it varied from 26 years to 7 months. The onset of symptoms followed a febrile illness in 2 cases and a fall in 2 cases. In the remaining 13 instances there was no
history of a factor that precipitated the symptoms. The complaint that the patients stressed was weakness and/or numbness of one or more extremities in 12 cases, ringing in the ears and loss of hearing in 2 cases, headache, girdle pain, and difficulty in breathing in 1 case each. Other complaints prof- fered by these patients consisted of double vision, loss of balance, pain in the arm, curvature of the spine, incontinence of urine, hoarseness, difficulty in swallowing, and ulceration of the finger tips.

The most common finding was weakness of one or more extremities; in only 2 of the 17 cases this was not demonstrable. It involved the upper more than the lower extremity. There were 5 cases of hemiparesis and 3 of quadripareisis. In 1 case the weakness involved both legs and in another one leg. There was a syringomyelic type of sensory loss in 12 cases, although in 2 of these, no cyst was found at operation.* The sensory loss included the 2nd cervical dermatome in 10 of the cases. Ten patients exhibited nystagmus, and the Romberg test was positive in 7. Two patients had astereognosis. In only 1 case was there papilledema. The preoperative diagnoses in these 17 cases were cord tumor, 3; platybasia, 2; acoustic tumor, 2; syringomyelia, 2; brain tumor, 1; multiple sclerosis, 1. The last 6 cases were observed within a period of 6 months and the preoperative diagnosis of Arnold-Chiari malformation was made correctly in each instance.

A careful review of the roentgenograms in these 17 cases disclosed definite evidence of platybasia in 8 (Fig. 1). In these 8 cases the tip of the dens projected above Chamberlain’s line† for from 6 to 30 mm. In 3 additional cases the appearance suggested a mild degree of platybasia while in only 4 instances could the x-rays of the skull and cervical spine be said to be entirely normal. There were 2 cases of Klippel-Feil syndrome and 1 of hemi-vertebra at the level of the 12th dorsal and 1st lumbar vertebrae. In 8 of these 17 cases, there was some degree of scoliosis of the dorsal spine. In none of these patients were there any physical signs suggesting an occult spina bifida although in 2 cases a failure of fusion of the 5th lumbar arch was demonstrated by x-ray.

In 3 of these 17 cases a subarachnoid block was demonstrated at spinal puncture but there was no increase in the total protein content. In 2 instances without subarachnoid block, the total protein content of the spinal fluid was elevated. Pneumo-encephalography was carried out in 11 cases and little or no air entered the ventricles (Fig. 2). The shape of the corpus callosum and the lateral ventricles, when outlined, indicated the presence of hydrocephalus. The pontine cisterns were flattened and air could not be demonstrated in the cisterna magna. In 1 case in which a cisternal pantopaque injection was attempted, the oil was found on x-ray to have been introduced into the caudally displaced 4th ventricle.

In the majority of these cases the symptoms began during adulthood.

* Of the 13 patients who had cystic spinal cords at operation, 8 exhibited no sensory loss.
† Chamberlain’s line: A line drawn between the hard palate and the posterior rim of the foramen magnum.
A possible explanation of this late development of signs in cases of a congenital disorder is as follows. Mild and unrecognized hydrocephalus with Arnold-Chiari deformity is present in infancy, but becomes compensated so that the growth of the brain catches up with that of the skull. Thus, when adulthood is attained, the circumference of the skull may be normal, although some degree of platybasia may be present. Then, due to the increasing density of aging connecting tissues, aggravated perhaps by infection or trauma, the outlets of the 4th ventricle become less pervious to fluid, so that an increased head of pressure is required to filter the fluid through to the subarachnoid cisterna. The resulting progression in the foraminal herniation is responsible for the gradually developing symptoms.

**SURGICAL TREATMENT**

These patients were operated upon in the sitting position. Through a midline incision, an opening was made in the occipital bone including the posterior rim of the foramen magnum. The laminae of the upper cervical vertebrae were removed. As the dura was incised, the cerebellar tonsils were found to fill the cisterna magna and to extend through the foramen magnum for a variable distance. The dura was usually adherent to a thickened arachnoid membrane at the level of the foramen magnum. The foramen of Magendie was closed by a dense membrane or by adhesion of the cerebellar tonsils to each other. The 2nd cervical nerve roots pursued a cephalad course.
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The exposed portion of the cervical cord was cystic in 13 instances and in each case the aspirated fluid was clear and colorless. In 2 cases the total protein content of the cyst fluid was estimated and found to be identical with that in the spinal fluid. An attempt was made to reopen the foramen of Magendie in each case and the dura, of course, was left open (Figs. 3, 4, and 5).

Figs. 3 and 4. Left, Operative exposure illustrating for the sake of comparison a normal brain stem. Right, A 44-year-old woman with progressive 8th nerve and cerebellar symptoms for many years. The tip of the left cerebellar tonsil reached the level of the 3rd cervical lamina.

In this series of 17 patients, 13 were definitely benefited by the operation, 3 were made worse because of aggravation of a quadriparesis, and 1 died 18 hours after operation of respiratory failure. In retrospect we believe that postoperative aggravation of symptoms may be due to a squeezing of the cord produced by flexion of the head during the positioning of the patient for operation.

A CASE OF ACQUIRED ARNOLD-CHIARI MALFORMATION

The following additional case of an Arnold-Chiari malformation, sub-

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* This stretching of C-2 is probably responsible for the occipital headache so frequently complained of by these patients and it may explain the fact that a pin prick usually feels less sharp on the posterior portion of the scalp than on the anterior.
sequently proved to be due to an acoustic tumor, offers some interesting ground for speculation in regard to the mechanics of hydrocephalus with associated hydromyelia.

The patient was a woman of 50 who presented herself on Mar. 29, 1945 because of headaches of 4 months’ duration. She exhibited bilateral papilledema, complete deafness in the right ear with absence of caloric response, and partial deafness in the left ear. There were definite cerebellar signs and impairment of sensation in the distribution of both 5th nerves. An acoustic tumor was diagnosed, but when spinal puncture disclosed normal pressure with complete spinal subarachnoid block the diagnosis was changed to tumor at the foramen magnum.

A midline exposure of the cerebellum was carried out, together with laminectomy of the upper four cervical vertebrae. As the dura was opened, the cervical cord was

![Image](https://via.placeholder.com/150)

Fig. 5. A 44-year-old woman with progressive weakness and numbness of arms for 2 years. The cerebellar tonsils are protruding through the foramen magnum, the 2nd cervical nerve roots pursue a cephalad course, there is a posterior bulging of the medulla (Chiari deformity), and below that a cystic dilatation of the cervical cord.

found to be distended with fluid. There was a pronounced herniation of the cerebellar tonsils. A needle was introduced into the spinal cord and clear, colorless fluid was aspirated. An incision was made into the hydromyelic cyst, after which the distended cord collapsed. The cerebellar tonsils were separated by retractors and the floor of the 4th ventricle exposed. It was deformed by a firm swelling at the level of the calamus scriptorius. This led the surgeon to suspect a tumor of the medulla with associated hydromyelia. The course of the 2nd cervical nerve roots was not described.

The patient improved, but 14 months following this operation, she returned with unmistakable signs of a right 8th nerve tumor. She was operated upon and a typical acoustic tumor was found and removed. The patient again improved, but died suddenly 5 months later, presumably of a cerebral vascular accident. Post-mortem examination was not obtained.

In retrospect we believe that the hydromyelia in this case, as in the case
of congenital Arnold-Chiari malformation, was the result of extrusion of ventricular fluid into the patent central canal of the cord from the obstructed 4th ventricle, and that the swelling of the medulla was due to a crowding downward of the brain stem (Chiari deformity). We believe that hydromyelia is commonly associated with the congenital form of Arnold-Chiari malformation and but rarely seen in the acquired form produced by cerebellar tumor because, as pointed out by Kramer, the central canal is always patent in infancy and relatively rarely so in the adult.

CONCLUSIONS

Although fully cognizant of the assailability of deductions drawn from an experience which has been almost purely surgical, nevertheless the following conclusions seem inescapable.

1. The Arnold-Chiari malformation is merely a foraminal herniation of intracranial structures produced by an obstructive hydrocephalus. Cord traction resulting from myelomeningocele, when present, may contribute to the deformity. The associated hydromyelia is due to the extrusion of fluid from the obstructed 4th ventricle downward into the central canal of the cord. A descriptive term for the condition is congenital obstructive hydrocephalomyelia with foraminal hernia.

2. The foraminal hernia which we call Arnold-Chiari malformation differs from the foraminal hernia produced by a cerebellar tumor because the former is a more chronic condition, produced by a relatively small increase in intraventricular pressure, which has its beginning during intrauterine life.

3. The "congenital" form of platybasia is produced by the increased weight of the head and malleability of the skull bones in cases of congenital obstructive hydrocephalus. On the other hand, if the infant is unable to assume the erect posture because of an associated myelomeningocele, platybasia should not develop.

4. In acquired platybasia of Paget's disease, osteogenesis imperfecta, and other bone-softening disorders, an Arnold-Chiari-like deformity results from the extrusion of the hindbrain through the foramen magnum due to the enroachment on the volume of the brain case occasioned by the deformity. While this may resemble the congenital variety of true Arnold-Chiari deformity it is not accompanied by hydrocephalomyelia.

5. Arnold-Chiari malformation should be suspected in any patient with signs suggesting syringomyelia, syringobulbia, platybasia, Klippel-Feil syndrome, and also in cases of unexplained scoliosis. It should be particularly suspected in the above cases if cerebellar signs are present or if there is impairment of pain perception in the distribution of the 2nd cervical nerve.

6. The diagnosis may be confirmed in the above cases by encephalography. The films will disclose evidence of obstructive hydrocephalus with absence of air in the cisterna magna. Since the intraventricular pressure is
usually only slightly increased, this procedure is safe provided the surgeon is prepared to operate at once if the films confirm the diagnosis.

7. Surgical treatment should be aimed at relieving the obstructive hydrocephalus.

REFERENCES

17. McKESSON. Personal communication.