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

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(Received for publication August 12, 1940)

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\(^3\) 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 Oldberg10 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.

List15 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.

Ogryzlo19 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.

Ray21 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.

Lichtenstein13,14 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 Scott11 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,2 Penfield and Coburn,20 Adams, Schatzki, and Scoville,1 Ecker and Ferguson,9 Shryock,24 Wagner,26 Moore,18 Bucy and Lichtenstein,4 Steele,25 and Chorobski.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