Introduction and Classification of the Chiari Malformations

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Abstract

Once an uncommon clinical finding, the Chiari malformations are now frequently seen with the advent of more sophisticated imaging modalities. With over 100 years of experience with these entities, we currently have a much better understanding of their embryology and pathophysiology. However, many gaps still exist in our knowledge of the Chiari malformations. Long-term outcome studies are becoming more prevalent and patients are commonly operated on with generally favorable results. Since their original classification, the Chiari malformations have grown to include smaller subsets. Herein, we focus on the two most common forms of hindbrain herniation, the Chiari I and II malformations. As with any rapidly changing field, knowledge of the natural history of the untreated condition is essential before recommending intervention.

Since the original description and classification of hindbrain hernias more than 120 years ago, the Chiari malformations have revealed much of their pathophysiology and have become easily diagnosed radiologically. Patients are commonly operated on with generally favorable results. We once thought their clinical presentation was easily understood, but as time has shown the edges of the clinical issue, the indications for surgical intervention in some patient groups have become somewhat clouded and blurred. The natural history of a patient with 5 mm hindbrain hernia and a non-Valsalva-induced frontal headache is unlikely to improve with a Chiari decompression. With the availability of MRI, more and more patients are being labeled with the diagnosis but without symptoms or appropriate symptoms. Because so much progress has been made with our understanding of these conditions, their radiologic definition, details of operative intervention, and prediction of outcome, we feel justified in presenting this text devoted to this fascinating group of conditions.

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Toward the end of the nineteenth century, Professor Hans Chiari developed a classification system that is still used today. He described four types of patients based on postmortem dissection studies. This text will focus on the first two types and their variants. *Type I Chiari malformations* (*CIM*) are patients without neural tube defects who have the cerebellar tonsils move through the foramen magnum. Radiologically, significant caudal displacement is considered to be more than 5 mm. The brain stem remains within the posterior fossa and there may or may not be the development of a syrinx. *Type II Chiari malformations* (*CIIM*) are seen exclusively in patients who have a neural tube defect and have caudal displacement of the cerebellar vermis and lower brain stem. This movement is thought to occur in utero prior to the full development of the cerebellar tonsils. There are many other associated central nervous system anomalies seen with the CIIM. The vast majority of these patients have raised intracranial pressure and hydrocephalus. The relationship of the hindbrain hernia and hydrocephalus in itself is complex. *Type III Chiari malformations* are a rare and extreme form of hindbrain hernia where a portion of the cerebellum and brain stem migrate out of the craniocervical junction through a defect in the dura, skull, and soft tissue layers to present as a mass or sac on the back of the neck. The infants may be viable, and the surgeon will need to amputate nonfunctional nervous tissue without damage to vital medullary function. These lesions will not be discussed in any further detail because of their rarity and limited potential for the surgeon to improve the situation other than the closure of the skin to prevent infection and minimize trauma and drying of vital brain structures. *Type IV Chiari malformations* were originally thought to be part of the hindbrain spectrum where the cerebellum was hypoplastic or absent. This group of patients is also uncommon, usually has no therapeutic options available, and is not appropriate to be considered with the hindbrains hernias. They too will not be considered further.

More recently, two other subtypes of patients have been described. *Chiari 0 malformations* are individuals without a hindbrain hernia or one that is minimal (less than 5 mm) but have a "crowded" appearance of the craniocervical junction and have a syrinx that develops as a consequence of the lack of free and easy CSF movement across this area. Posterior fossa decompression with duraplasty may totally resolve the syrinx and associated symptoms if CSF egress from the fourth ventricle can be reestablished. *Chiari 1.5 malformations* are a hybrid of some aspects of the CIM and some of the CIIM. They are not associated with neural tube defects, but have significant caudal movement of the cerebellar tonsils and brain stem. Their symptoms and outcomes are more challenging than patients without caudal brain stem displacement.

One of the most impressive aspects of these entities has been the evolution of our understanding of the mechanism of their development. As recently as 1979, major neurology texts [1] have grouped the Chiari malformations and syringomyelia into the degenerative section and mentioned possible treatment with radiation therapy. Since that time, a pressure differential theory has been introduced and surgical decompression become commonplace.

We have elected to include a chapter on non-hindbrain hernia related syrinx development. This decision was made to help the reader appreciate the differential diagnosis of any patient with a syrinx and to underscore the large number of other causes, which must be excluded before assuming the syrinx is due to a minimal hindbrain hernia.

As with any rapidly changing field, knowledge of the natural history of the untreated condition is essential before recommending intervention. With the advent of readily available MRI, more and more asymptomatic patients present concerned and worried they have a potentially crippling and debilitating problem. The radiologist confronted with images "on the edge" of normal may report on images and mention an early problem and refer to "borderline changes." Clinicians faced with patients with an encyclopedic array of symptoms, many of which may be seen with the CIM, yield to referral pressure. Sorting through these issues has received significant medical and lay attention [2]. We hope that the experts we

have assembled here will provide light and understanding to these difficult issues. With the advent of MRI, the clinical pendulum has swung from the uncommon CIM patient diagnosed with major neurological problems to many "normal" people with unremarkable neurological examinations who are concerned with their survival.

References

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