Pathogenesis and Cerebrospinal Fluid Hydrodynamics of the Chiari I Malformation

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KEYWORDS

• Chiari I malformation • Hydrocephalus • Posterior fossa

KEY POINTS

- The pathogenesis of an anatomic Chiari I malformation can occur with several different mechanisms, including overcrowding from underdevelopment of the posterior fossa bony structures, hemodynamic disturbances of the central nervous system, such as hydrocephalus or bilateral chronic subdural hematomas producing tonsillar herniation, a mass in the posterior fossa causing tonsillar herniation, or, rarely, reduced spinal intrathecal pressure with downward herniation of the cerebellar tonsils associated with low spinal intrathecal pressure from a lumbar-to-peritoneal shunt or a spinal cerebrospinal fluid (CSF) leak.
- Because the abnormal shape and position of the cerebellar tonsils is reversed by surgery that provides unobstructed pulsatile movement of CSF across the foramen magnum, the pathogenesis of a Chiari I malformation is impaction of the tonsils in the foramen magnum, not a result of a congenital brain malformation.
- Evidence provided by anatomic imaging, dynamic imaging with MRI and intraoperative ultrasound, and physiologic studies during, before, and after surgery for Chiari I malformation is consistent with an extramedullary hydrodynamic mechanism in which the cerebellar tonsils are impacted in the foramen magnum and act on a partially entrapped spinal CSF space to increase intrathecal pressure and pulse pressure and produce suboccipital headache, and, in some patients, syringomyelia.
- Thus, the pathophysiology of the Chiari I malformation is simply the obstruction of the normal pulsatile movement of CSF across the foramen magnum.

INTRODUCTION

The pathology that is now known as Chiari malformations of the cerebellar tonsils originated in 1891 with Hans Chiari's manuscript titled "Concerning alterations in the cerebellum resulting from cerebral hydrocephalus."[1–3] In this publication, Chiari described "alterations in the cerebellum resulting from cerebral hydrocephalus."[1–3] In 1896, Chiari described an additional mechanism for the pathogenesis of the malformation; insufficient bone growth and insufficient enlargement of portions of

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the skull during development cause increased intracranial pressure and subsequent tonsillar herniation.[3,4] Since Chiari's initial publications, there have been several hypotheses that attempt to elucidate the pathogenesis of the Chiari I malformation and the pathophysiology associated with it.

This article summarizes the current understanding of the pathophysiology of the Chiari malformation that is based on observations of the anatomy visualized by modern imaging with MRI and prospective studies of the physiology of patients before and after surgery. The pathogenesis of a Chiari I malformation of the cerebellar tonsils is grouped into 4 general mechanisms:

1. Overcrowding caused by underdevelopment of the posterior fossa bony structures
2. Hemodynamic disturbances that increase intracranial pressure
3. Excess tissue in the posterior fossa by a tumor
4. Downward movement of the central nervous system by events that lower intrathecal pressure, such as lumbar-to-peritoneal shunts.

It is noteworthy that each of these mechanisms acts on normal cerebellar tonsils to deform them by impacting them in the foramen magnum, deformation that is consistently reversed by simple surgery to provide extra room at the foramen magnum.

Of note, the authors have limited their comments to Type I Chiari malformation. Even with this limitation, it becomes apparent that unanimity of thought is lacking on the pathophysiology of the type I Chiari malformation. In fact, some hypotheses even contradict each other. For example, hydrocephalus has been proposed as both the etiologic cause and a result of the Chiari malformation.[5]

PATHOGENESIS OF THE CHIARI I MALFORMATION

Limited Embryologic Development of the Skull Base

Several studies have demonstrated that many, but not all, patients with a Chiari I have a small posterior fossa. In 1 study, to investigate overcrowding in the posterior cranial fossa as the pathogenesis of Chiari malformation, Nishikawa and colleagues[6] correlated the morphology of the brainstem and cerebellum with the anatomy of skull base. They used X-Ray tomography to measure 3 occipital enchondral parts: the supraocciput, exocciput, and basiocciput. They found a significant difference in the mean length of the exocciput, from the bottom of the occipital condyle to the top of the jugular tubercle, which measured 16 mm in the Chiari group compared with 20.5 mm in control patients. They further described a significant difference in the length of the supraocciput between the internal occipital protuberance and the opisthion, which measured 38.9 mm in the Chiari group and 48.1 mm in the controls. The axial length of the clivus (the basiocciput and basisphenoid) in the Chiari group was not shorter than that of the control group. On average, the Chiari group had smaller posterior fossa cranial volume (186 cc) compared with the control group (193 cc), although it was not significant, and no significant difference was found in the posterior fossa brain volume (156 cc) compared with the controls (153 cc). As in other studies, there was a significant difference in the ratio of the posterior fossa brain volume to the posterior fossa cranial volume (mean volume ratio 0.833 in the Chiari group and 0.790 in the control group).[6]

Stovner and colleagues measured skull dimensions on lateral skull radiographs in 33 adult patients with MRI-verified Chiari malformations and 40 control subjects. They found that the posterior cranial fossa was significantly smaller and shallower in Chiari patients compared with controls. For Chiari patients, there was a positive correlation between posterior cranial fossa size and cerebellar tonsillar ectopia. Because of this positive correlation, Stovner and colleagues[7] propose that an undersized posterior cranial fossa had been expanded by hindbrain herniation at an early stage in development.

Vega and colleagues[8] studied a series of 42 patients with Chiari malformation compared with 46 control subjects. Their results support the hypothesis that cerebellar tonsillar ectopia is caused by the disproportionate size between the volume of the posterior cranial fossa and the cerebellum. The authors recorded linear, angular, and posterior fossa surface area measurements on lateral skull radiographs. They used computed tomography (CT) to calculate posterior cranial fossa volume. They found that Chiari patients exhibited shorter clival lengths, shorter Twining-opisthion distances, and shorter Chamberlain line. Also, the average size of the posterior cranial fossa was smaller in Chiari I patients compared with control.

Perhaps the most important and often ignored developmental feature that is related directly to the pathophysiology of Chiari malformation is the somatic origin of the occipital bone. Early in embryonic development, the occipital bone forms from at least 3 pairs of sclerotomes. The occipital sclerotomes, which in turn are formed from occipital somites, eventually fuse into a single structure and are incorporated into the developing cranial