Complex Chiari Malformations in Children: Diagnosis and Management

Douglas L. Brockmeyer, MD*, Heather S. Spader, MD

KEYWORDS

• Complex Chiari • Basilar invagination • Clivocervical angle • Retroflexed odontoid

KEY POINTS

- Complex Chiari malformations may be a distinct category from other Chiari malformations.
- These patients are at increased risk for occipitocervical fusions after Chiari decompression or may need upfront decompression and fusion procedures.
- The decision-making algorithm for these patients needs to be modified to account for different outcomes for these patients. This algorithm includes clinical symptoms, extent of Chiari, evaluation of clival-cervical angle, retroflexed odontoid, and basilar invagination.

DEFINITION OF COMPLEX CHIARI MALFORMATION

Chiari malformations were originally classified by Chiari in 1896 into three categories based on anatomic description[1]:

- Chiari 1: Cerebellar tonsil and lower part of the medulla below the foramen magnum without displacement of the fourth ventricle.
- Chiari 2: Caudal migration of the lower part of the cerebellum associated with downward displacement of the fourth ventricle, which appears lengthened; foramina opens into the spinal subarachnoid space; associated with spina bifida.
- Chiari 3: Cerebellum and medulla displaced into the cervical spinal canal associated with an occipital meningocele.

Later, a fourth type was added that describes an incomplete or underdeveloped cerebellum.

These categories were clinically useful for almost 100 years; however, as neurosurgeons began studying outcome data for surgical decompression of Chiari 1 malformations (C1M), it became apparent that there was a subcategory of patients in whom the condition was more complex; these patients required more frequent surgical intervention than the others. Initial examination of this group by Grabb and colleagues[2] in 1999 showed that odontoid retroflexion, manifested by a pBC2 distance (maximum perpendicular distance to the basion-inferoposterior point of the C2 body) greater than 9 mm, defined a patient group that frequently required craniocervical fusion procedures. Further experience with this patient population led to the observation that a subcategory of patients with Chiari malformation had caudal descent of the brainstem and tonsillar ectopia. Therefore, a new category, the Chiari 1.5 malformation (C1.5M), was proposed; it was defined as the presence of obex herniation below the foramen magnum as seen on MRI.[3]

With a growing awareness of the complex nature of disease in some Chiari patients, Bollo and colleagues[4] comprehensively analyzed their experience with patients that manifested a constellation of craniospinal radiographic findings

Division of Pediatric Neurosurgery, Primary Children's Hospital, University of Utah, 100 Mario Capecchi Drive, Salt Lake City, UT 84113, USA
* Corresponding author.
E-mail address: Douglas.Brockmeyer@hsc.utah.edu

Neurosurg Clin N Am 26 (2015) 555–560
http://dx.doi.org/10.1016/j.nec.2015.06.002
1042-3680/15/$ – see front matter

aside from just tonsillar herniation. These findings included

- Brainstem herniation through the foramen magnum (C1.5M)
- Medullary kink
- Retroflexed odontoid
- Abnormal clival-cervical angle (CXA)
- Occipitalization of the atlas
- Basilar invagination (BI)
- Syringomyelia
- Scoliosis

The authors analyzed a group of patients with the previously mentioned radiographic findings and found that the presence of a C1.5M, a CXA less than 125°, and BI placed a patient at a higher risk for requiring a craniocervical fusion than those with a typical C1M. They proposed a new category of Chiari malformation, known as complex Chiari malformation (CCM), that encompasses these radiographic findings, all or in part. Further detail about these measurements is illustrated in **Fig. 1**. Although there are several previous case series in the literature describing the management of complex-type Chiari patients,[5–10] Bollo and colleagues[4] were the first to analyze a large group of patients with CCM and define factors that place patients at a higher risk for craniocervical fusion. Here, we describe the diagnostic criteria for CCM and outline the decision-making process for the optimal treatment of this patient population.

PATIENT SELECTION

Clinical Findings

In general, the clinical presentation of patients with CCM is not much different from that of typical C1M patients. They tend to present with lifestyle-limiting headaches, often in the suboccipital region. Paresthesias or bulbomyelopathic symptoms, such sleep apnea, snoring, dysphagia, or ataxia, are often found. Patients younger than 4 years of age often present with symptoms consistent with oral-motor apraxia (eg, poor feeding, delayed speech) and/or apnea.[4,11]

Radiographic Findings

To identify a CCM, the initial craniospinal MRI must be evaluated for

- Chiari1.5M
- Odontoid retroflexion, measured by pBC2 (**Fig. 2**)
- CXA
- BI
- Assimilation of the atlas
- Medullary kinking

Other findings may include hydrocephalus, associated brain abnormalities, scoliosis, and syringomyelia. Together, these radiographic elements identify a CCM and help the clinician make decisions about proceeding with surgery.

Clinical-Radiographic Integration

Step 1: surgical decision making

First, the surgeon must decide whether the patient's symptoms are severe enough to warrant surgery. A useful distinction is to ask the patient whether the problem is truly lifestyle limiting. For children, this means the symptoms keep them home from school, prevent them from doing something they normally enjoy (such as playing with friends), or cause them to quit early from their

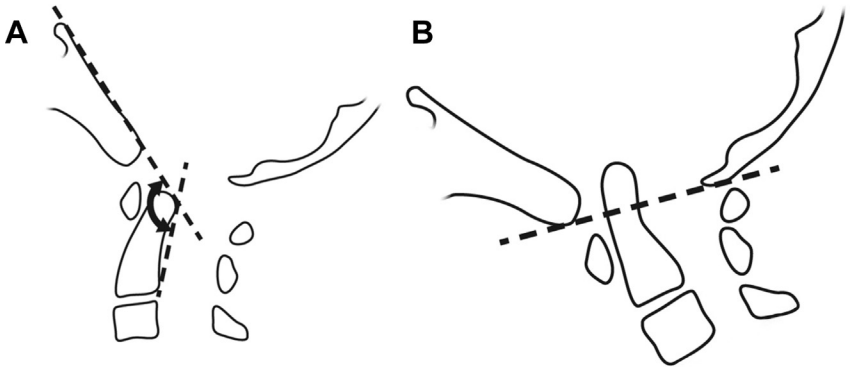

Fig. 1. Illustration of clival-cervical angle. This is the angle (*double arrow*) derived from a line drawn from the inferior two-thirds of the clivus and a second line drawn from the posterior-inferior C2 body to the superior-posterior aspect of the odontoid (*A*). Basilar invagination is seen when the odontoid process of C2 is above the foramen magnum. McRae's line is the line joining the basion and opisthion. The dens should normally be 5 mm below this line (*B*).