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MR Imaging of Hindbrain Deformity in Chiari II Patients with and Without Symptoms of Brainstem Compression

John T. Curnes^{1,2}
W. Jerry Oakes³
Orest B. Boyko¹

We examined the MR appearance of the hindbrain deformity, including the upper cervical spinal canal and craniovertebral junction, in 33 patients with Chiari II malformation. In this disorder, there is impaction at birth of the medulla and cerebellar vermis into the upper cervical spine, resulting in obliteration of the subarachnoid space and scalloping of the dens. Spinal canal enlargement during the child's growth, combined with dorsal displacement of neural tissue, eventually causes marked widening of the precervical subarachnoid space. This enlargement may simulate an intradural mass. Our series documents the changes seen at birth and the progression of the widened precervical space through the first and second decades.

Twelve (36%) of the 33 patients studied were symptomatic, with brainstem or long-tract symptomatology, and 11 of these required surgery. This group was compared with the remaining 21 asymptomatic Chiari II patients to identify MR features associated with clinical deterioration. The level of descent of the hindbrain hernia was critical; eight of 12 symptomatic patients had a cervicomedullary kink at C4 or lower, while no asymptomatic patients had a fourth ventricle, medulla, or kink below C3-C4. The precervical cord subarachnoid space was slightly wider in asymptomatic patients, although there was great overlap. In five patients with follow-up scans, this space was seen to increase in width after laminectomy. A CSF flow void was present in the precervical space in about 25% of patients in both groups. In nine of 12 symptomatic patients, C1 arch indentation of the dura (causing significant compression) was confirmed surgically. However, seven (33%) of the 21 asymptomatic patients also had this appearance. Absolute measurement of the anteroposterior diameter of the canal at C1 ranged from 11 to 25 mm in both groups. Retrocollis, which persisted despite sedation for MR, was seen in two patients, both symptomatic.

Recognition of the vermis, medullary kink, cervical cord, C1 arch, fourth ventricle, and precervical space in Chiari II patients is fundamental to the analysis of symptoms in these patients, and is important for preoperative evaluation.

Changes in the size and shape of the craniocervical junction and upper cervical canal in patients with Chiari II malformation were first evaluated with plain films [1], then with CT [2], and most recently with MR imaging [3-5]. MR imaging shows the relationship of the neural structures enclosed in the bony canal, particularly in the sagittal plane.

The MR appearance of the hindbrain deformity in patients of all ages with Chiari II malformation, both asymptomatic and symptomatic from Chiari-related complaints, was the subject of our investigation. More specifically, we tried to identify those MR features that might correlate with symptoms of cranial nerve dysfunction or spasticity, since other tests, such as brainstem auditory evoked responses and CO₂ response curves, do not always provide reliable objective data that confirm the surgeon's clinical impression.

Subjects and Methods

Thirty-one patients with meningocele and two patients with occipital cephaloceles were studied with MR imaging. All patients had treated hydrocephalus with a functioning

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¹ Department of Radiology, Section of Neuroradiology, Duke University Medical Center, Durham, NC 27710.

² Present address: Greensboro Radiological Associates, Box 13005, Greensboro, NC 27415. Address reprint requests to J. T. Curnes.

³ Division of Neurosurgery, Departments of Surgery and Pediatrics, Duke University Medical Center, Durham, NC 27710.

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shunt. Chiari II patients who had previous surgery but no preoperative MR studies were excluded. The age groups and patients in each were <1 month (6), 1–12 months (6), 1–2 years (4), 2–4 years (3), 4–8 years (4), and >8 years (10). Sagittal and axial images of the head and cervical spine were obtained on a GE 1.5-T superconducting system. T1-weighted images, 600/20 (TR/TE), were routinely obtained. Other parameters included a slice thickness of 3 mm for sagittal and 5 mm for axial images, a 1-mm gap, a 256 × 128 matrix, and two averages. If sufficient anatomic detail of the cervical spine was not obtained with the head coil, a 5-in. surface coil was used. In addition, the clinical records, including surgical notes, were reviewed for the 11 patients who required decompressive surgery (Table 1).

The following features were studied: (1) The ratio of the neural tissue in the hindbrain hernia to the maximal sagittal diameter of the canal opposite C2, reflecting the width of the anterior subarachnoid space (Fig. 1). (2) The vertebral level and MR appearance of the vermis and medullary kink within the cervical canal. If no kink was present, the lowermost extent of the fourth ventricle was tabulated (Fig. 2). (3) The presence of a CSF flow void in the anterior subarachnoid space (Fig. 3). (4) The presence of a fibrovascular band or bone at C1 constricting neural tissue, the severity of indentation of the underlying dorsal dura, the narrowest sagittal diameter at that point, and the presence of retrocollis (Fig. 4). (5) The changes in appearance of the fourth ventricle and anterior subarachnoid space before and after laminectomy (Fig. 5).

MR of a neuropathology-museum specimen of a neonate with a severe Chiari II malformation was obtained with a 3-in. surface coil and the identical imaging parameters as before (Figs. 2F and 2G).

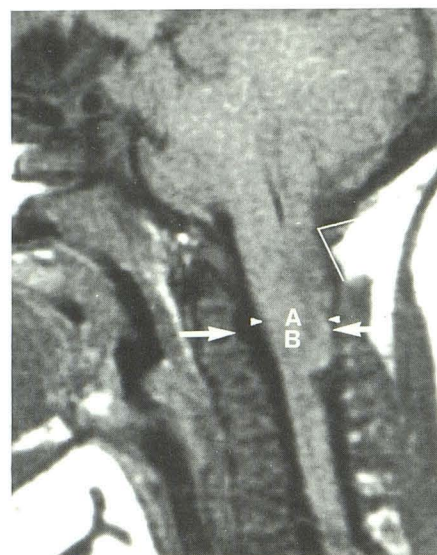


Fig. 1.—Sagittal image illustrates method used to calculate ratio A/B and dural angle. 14-month old, asymptomatic. A = width of neural tissue in hindbrain hernia (arrowheads), B = maximal sagittal diameter of spinal canal at C2 (arrows). A narrow subarachnoid space, therefore, has a ratio closer to 1.0. White lines = dural angulation over arch of C1.

TABLE 1: Summary of Clinical and MR Findings in Symptomatic Chiari II Patients

Case No.	Age	Symptoms	Vermis Level	Medullary Kink Level	Clinical Improvement After Surgery	Dilatation of Fourth Ventricle After Surgery
1	22 months	Breath holding, apnea, retrocollis	C3	C4	Yes	Unknown
2	16 years	Arm spasticity, truncal ataxia, cough	C3	C4	Yes	Unknown
3	18 months	Opisthonos, choking, retrocollis	C2	C3	Yes, but later died	Unknown
4	1 year	Arm spasticity, stridor, apnea	C2	C3	Yes	Unknown
5	6 months	Multiple cranial nerve palsies, failure to thrive	C3–C4	C4–C5	Equivocal	Unknown
6	15 years	Arm spasticity, nystagmus	C2	C2–C3	Yes	No
7	8 years	Arm spasticity, gagging, swallowing difficulty	T1	C6	Equivocal	Yes
8	4 years	Weakness of arms, arm spasticity, ascending sensory level, swallowing difficulty	C3	C3–C4	Yes	Yes
9	4 years	Apnea, bradycardia, weakness of arms, cyanosis	C2	C4	Yes	Yes
10	7 months	Nystagmus, facial weakness	C4	C5	Yes	Unknown
11	1 month	Apnea, bradycardia	C4	C5	Equivocal	No
12	8 months	Retrocollis, choking, nystagmus, failure to thrive, spasticity of arms	T3	T1	No surgery	No surgery

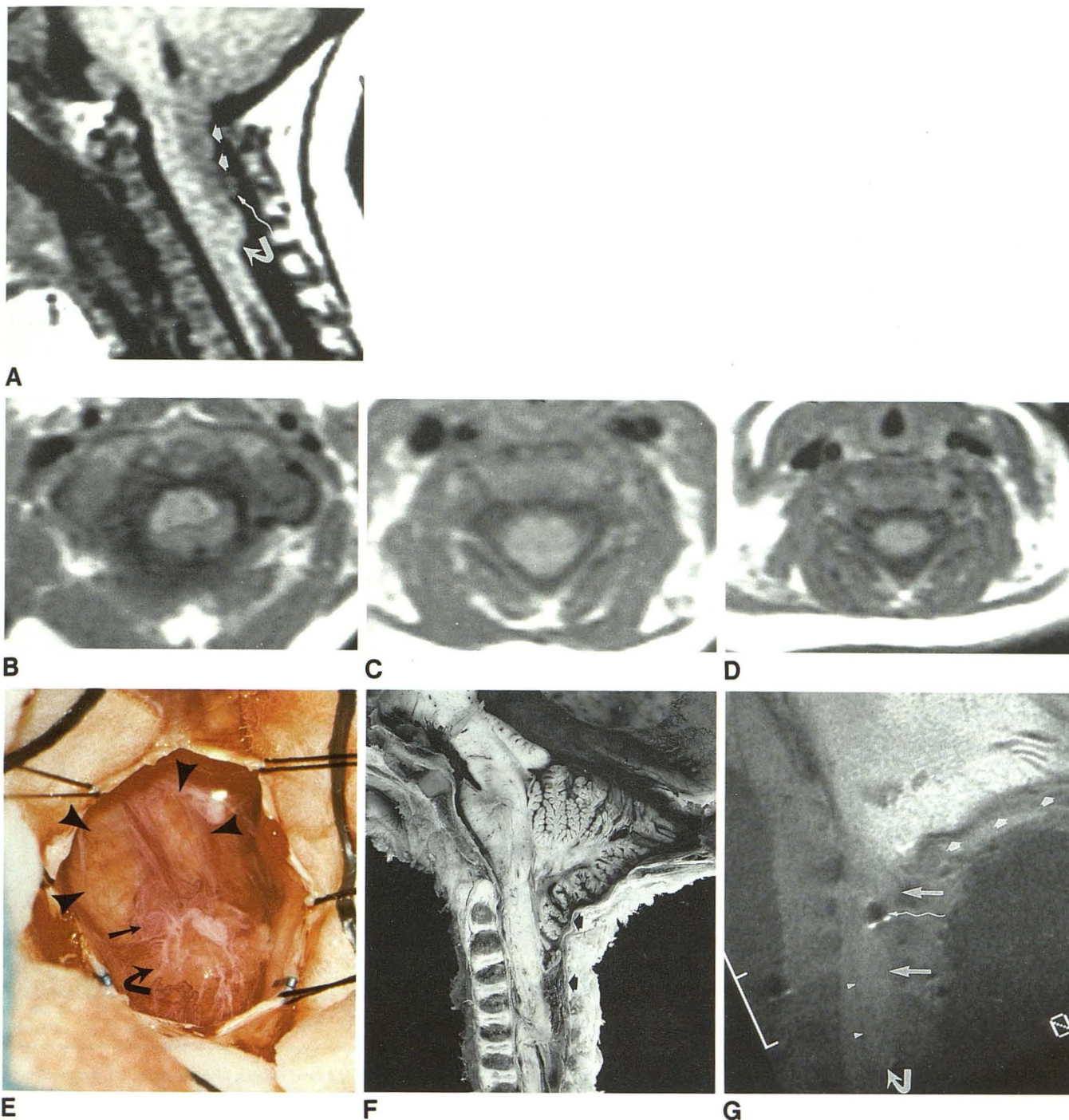


Fig. 2.—Hindbrain hernia analysis.

A–E, 6-month-old, symptomatic.

A, Sagittal image demonstrates clearly definable plane between gray vermis posteriorly and myelinated medulla and cord anteriorly. Note that herniated vermis (*short arrows*) has a slightly decreased signal intensity (longer T1) relative to gray matter of cerebellum above foramen magnum, reflecting compressive gliosis. Medullary kink is evident approximately 1 cm below this, at C4–C5 (*curved arrow*), displacing cervical cord very slightly anteriorly. Note lobular choroid plexus (*wavy arrow*). Syringomyelia is present at cervicothoracic junction.

B, Axial image at C2–C3 shows intimate relationship of gray vermis posterior to anteriorly displaced medulla (upper figure of eight).

C, Note lobular, homogeneous appearance of tissue in this axial slice through medullary kink at C4–C5 (lower figure of eight).

D, Cervical cord at C5 is slightly flattened and atrophic.

E, Intraoperative photograph shows flattened, gliotic vermis (*large arrowheads*) and orange choroid plexus (*straight arrow*), below which is the medullary kink (*curved arrow*).

F and G, Chiari II neuropathology-museum specimen.

F, Whole-mount pathologic section. Newborn male with Chiari II malformation who died within hours of birth of respiratory failure. Note gray, gliotic vermis (*arrows*). Compare with normal vermis above foramen magnum. Medullary kink is at C6.

G, T1-weighted sagittal image of specimen in F obtained by using 3-in. surface coil. Note that gliotic vermis below foramen magnum (*long arrows*) is darker than cerebellar vermis tissue above foramen magnum (*short arrows*). Medullary kink is at C6 (*curved arrow*), and cervicomedullary interface runs obliquely upward (*arrowheads*). An artifact from plastic used in mounting of the specimen is seen (*wavy arrow*).

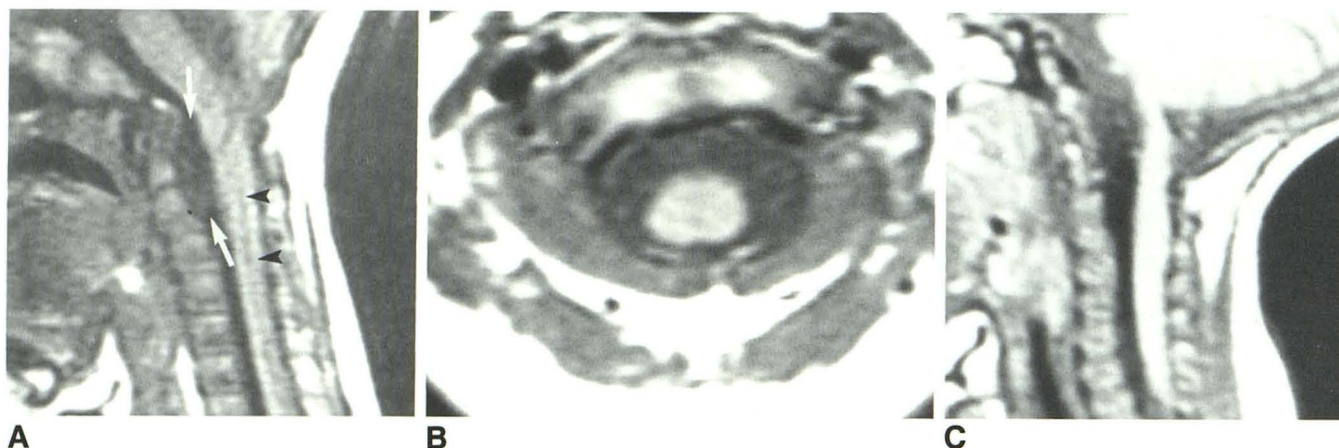


Fig. 3.—CSF flow void.

A and B, 4-year-old girl, symptomatic.

A, Sagittal image through craniocervical junction reveals pseudomass anteriorly (arrows). This CSF appears to have a different signal from that in lower cervical and thoracic subarachnoid space. Vermian tissue is not well seen and cervicomedullary kink lies at approximately C3–C4. Truncation artifact (arrowheads) simulates hydrosyrinx.

B, Axial image at C2 shows no mass anteriorly and CSF with approximately the same intensity anteriorly as laterally.

C, 6-year-old, asymptomatic. Sagittal image shows dark CSF in anterior subarachnoid space opposite C2, compatible with pulsatile or flowing CSF motion.

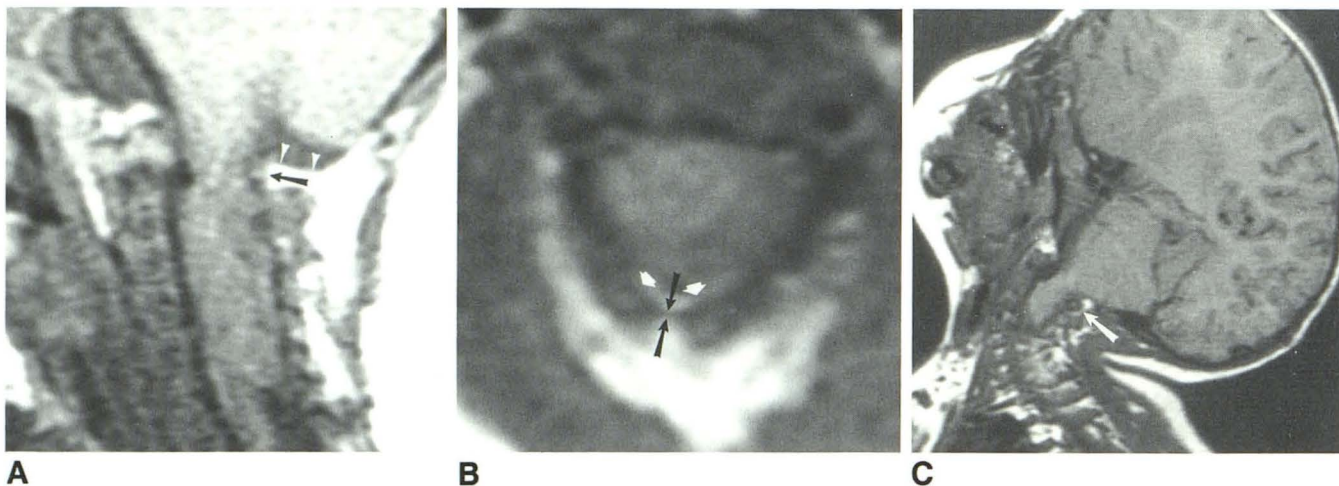


Fig. 4.—C1 arch impingement. See also Fig. 2A.

A, 7-month-old girl, symptomatic. Sagittal image demonstrates C1 arch (arrow) causing dural indentation and bulging of dura above C1. Angle of dura above C1 compared with that below C1 is acute ($<90^\circ$) (arrowheads). Note medullary kink at C5 level. At surgery, severe indentation of dura was present at C1, which persisted after bone removal.

B, Axial T1-weighted image demonstrates bifid C1 (short arrows) laminar arches. A thin, tough fibrous band (between long arrows) contiguous with inner periosteum of lamina, unites the two arches.

C, 18-month-old boy, symptomatic. Sagittal image demonstrates extreme retrocollis, even while sedated. Both patients in our series with this finding required surgery. Note bony laminar arch posteriorly at C1 indenting neural tissue (arrow). The vermis and medullary kink cannot be identified in this section. Sagittal diameter measured 11 mm at C1.

The gross appearance was correlated with the in vitro imaging features and clinical material.

Results

Results are summarized in Tables 1–4 and in Figure 6. Table 1 is a summary of the 11 surgically treated patients and the one symptomatic patient who did not have decompressive surgery.

Precervical Subarachnoid Space

An examination of Fig. 6 reveals a trend toward a wider anterior subarachnoid space in the asymptomatic group than in the symptomatic group. Both subgroups illustrate the fact that widening of this space occurs with age (Fig. 7). Because of the overlap between the symptomatic and asymptomatic groups, particularly in the first 2 years of life, the presence or absence of a widened precervical space per se does not distinguish these two groups. Of note, however, is the fact

Fig. 5.—Pre- vs postoperative appearance.

A, 8-year-old boy, symptomatic. Preoperative sagittal image demonstrates extreme dorsal herniation of vermis down to T1 level (*curved arrow*). Medulla and fourth ventricle end at C6 (*wavy arrow*).

B, Postoperative sagittal image shows widening of anterior subarachnoid space as well as dilatation of fourth ventricle. A small stent can be seen (*arrows*).

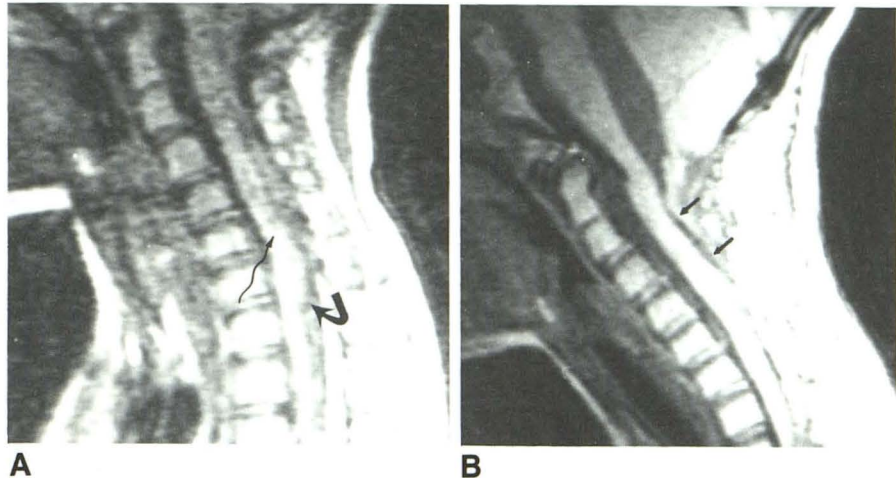


TABLE 2: Level of Medullary Kink or Lowermost Fourth Ventricle in Chiari II Patients

	Asymptomatic (n = 21)	Symptomatic (n = 12)
C1	1	
C1-C2	4	
C2	5	
C2-C3	6	1
C3	3	2
C3-C4	2	1
C4		3
C4-C5		1
C5		2
C5-C6		0
C6		1
C7		0
T1		1

TABLE 3: Appearance of Flow Void in Anterior Subarachnoid Space

	Asymptomatic	Symptomatic
Present	5	3
Absent	16	9

TABLE 4: C1 Impingement on Dorsal Dura and Neural Tissue

	Asymptomatic	Symptomatic
Less than 90°	7	9
Greater than 90°	14	3

that only one of 12 symptomatic patients demonstrated a ratio A/B of 0.5 or less; and in this patient, symptoms from the syrinx predominated over brainstem signs (nystagmus).

Vertebral Body Level and MR Appearance of the Hindbrain Hernia

As shown in Table 2, the level of the medulla or medullary kink in Chiari II patients as determined by MR revealed a subgroup of symptomatic patients in whom there was no

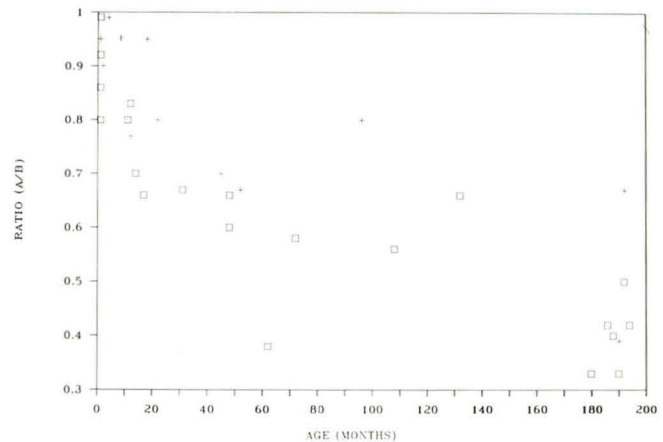


Fig. 6.—Graph of ratio A/B vs age (in months) in Chiari II patients. Asymptomatic = box, symptomatic = +. Both groups illustrate trend toward widening of anterior subarachnoid space with age, with symptomatic patients having a slightly narrower anterior subarachnoid space for a given age.

overlap. Seven of the 11 patients who underwent cervical decompression had their medullary kink at the level of the C4 vertebral body or lower, whereas none in the asymptomatic group had a medullary kink or fourth ventricle descend to this level. One symptomatic patient who did not have surgery had cerebellum and medulla to T1 and was managed conservatively because of other congenital anomalies. Of the symptomatic patients, four were less than 12 months old, two were between 12 and 24 months old, and two were over 3 years old. Overlap occurred between the asymptomatic and symptomatic group at the levels of the C2-C3 disk space through the C3-C4 disk space. Of the five asymptomatic patients who had a medullary kink at C3 or lower in our series, four were less than 1 year old and could develop signs of brainstem dysfunction in the future.

Figure 8 is a receiver operating characteristic (ROC) curve for the predictive rate of medullary level vs symptoms. This curve demonstrates a high true-positive rate with a low false-positive rate over the lower vertebral body levels, indicating a high reliability of vertebral body level as a predictor of symptoms.

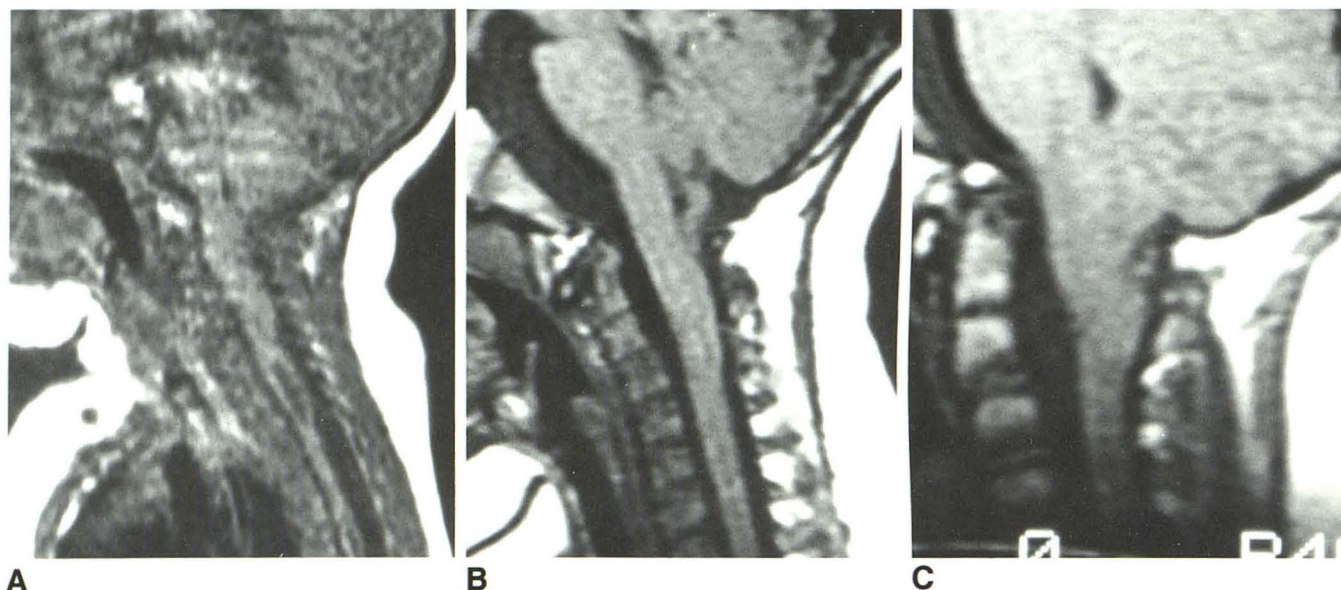


Fig. 7.—Examples of anterior subarachnoid space width at several ages. Note subtle scalloping of dens in each case.

A, 1-month-old symptomatic, ratio = 0.95. Note syringomyelia at cervicothoracic junction.

B, 3-year-old, asymptomatic, ratio = 0.67.

C, 9-year-old, asymptomatic, ratio = 0.57.

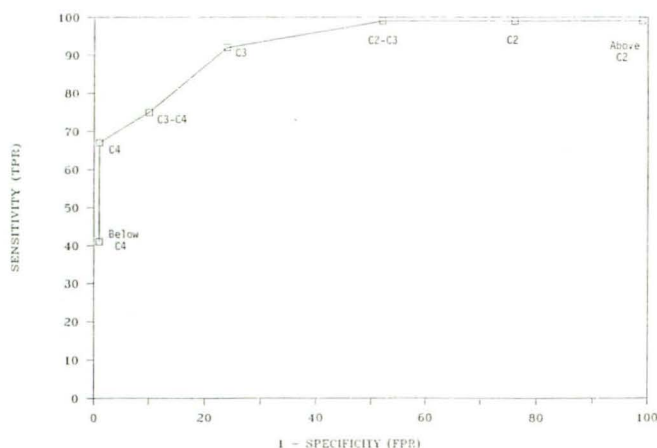


Fig. 8.—Receiver operating characteristic (ROC) curve for the predictive rate of medullary level vs symptoms in Chiari II patients. Curve demonstrates high true-positive rate with low false-positive rate over lower vertebral body levels, indicating high reliability of vertebral body level as a predictor of symptoms.

The herniated vermis was noted to have a lower signal (longer T1) than the vermis above the foramen magnum in five of 12 symptomatic patients and one of 21 asymptomatic patients. This was thought to represent gliosis secondary to long-standing compression. This vermis gliosis was prominently profiled by the higher signal of the myelinated medulla (Figs. 2A and 2B).

CSF Flow Void

No correlation between asymptomatic and symptomatic patients could be made with the presence or absence of a

flow void in the anterior subarachnoid space (Table 3). It was absent (76%) more than it was present (24%) in both groups (Fig. 3). When absent, the appearance was either mottled or homogeneous.

C1 Arch Impingement

Impingement of the C1 arch upon the dorsal dura and underlying neural tissue caused a greater than 90° angle between the dura above and below C1 in 17 patients and less than 90° in 16 patients. The trend was that a majority of symptomatic patients appeared to have a tight C1 arch, whereas asymptomatic patients displayed less severe dural compression (Fig. 4). In that subgroup of asymptomatic patients in whom less than a 90° angle over the arch of C1 was formed by the dura, all but one was less than 1 year old. The presence of significant C1 compression in nine of 12 patients was confirmed at surgery, and often a visible groove remained in the dura after C1 laminectomy, reflecting long-standing impingement.

Actual measurement of the sagittal diameter of C1 to the tip of the dens revealed a range of 11 to 25 mm. In children 6 months old or younger, the diameter measured between 11 and 16 mm, with considerable overlap between asymptomatic and symptomatic patients.

Changes After Surgery

Five patients had both pre- and postoperative MR imaging. In each patient the precervical space widened postoperatively (Fig. 5). No change in the presence or absence of a flow void was observed on follow-up scans. In three of the five patients, the fourth ventricle increased in size postoperatively, despite

the presence of a fourth ventricular to subarachnoid shunt (Fig. 5). In one of these patients a separate fourth ventricular to peritoneal shunt connection was inserted for brainstem compressive symptoms. With resolution of the enlargement of the fourth ventricle, the symptoms resolved.

In Vitro Analysis of a Museum Specimen

T1-weighted images of the Chiari II specimen (Figs. 2F and 2G) showed the entire hindbrain hernia severely impacted in the upper cervical canal, with obliteration of the precervical cord subarachnoid space and mild scalloping of the dens.

The vermis was compressed, devoid of its delicate folial pattern, and extended down to C5. Its signal characteristics were similar to the in vivo symptomatic patients, with brighter signal of vermis above the foramen magnum than in the hernia below. Lack of mature medullary myelination in this newborn provided little contrast with the gliotic vermis.

The medullary kink extended to C5–C6, consistent with the brainstem compression and resultant respiratory death within hours of birth, despite ventricular drainage. The rounded kink and the cervicomedullary interface are well seen on images of the specimen.

Clinical Outcome

Clinical improvement occurred in eight of the 11 patients who had surgery, with an average of 2 years of follow-up. One patient with severe retrocollis, diffuse cervical hypoplasia, and truncus arteriosus was followed conservatively. Further posterior decompression was not thought to be beneficial, since the patient had marked enlargement of the foramen magnum and wide laminal defects from C1 through C5. Another patient with life-threatening brainstem symptomatology initially improved after surgery both clinically and by physiologic testing only to die as a result of infantile Gaucher's disease.

Of the 11 surgical patients, five had progressive upper extremity spasticity as their primary clinical problem while six had lower cranial nerve disturbances. The three patients with little or no functional improvement and the one who died from Gaucher's disease were all symptomatic from lower cranial nerve disturbances. Therefore, only two patients with major medullary disturbances had clear long-term improvement from surgical intervention. A larger clinical series including these patients has been previously published [6].

Discussion

Naidich et al. [7–9] contributed much to our understanding of the Chiari II malformation through CT, and more recently elucidated the radiographic features of the hindbrain deformity [2]. Wolpert et al. [4] and El Gammal et al. [5] established that MR is now the procedure of choice in the evaluation of Chiari II patients. Our study focused on the changes seen in the upper cervical spine and craniocervical junction as shown with MR imaging.

Upper Cervical Canal and Precervical Cord Space

Using plain films, Naik and Emory [1] first showed that the upper cervical canal (C2–C4) was wider in Chiari II patients than in normal infants. They attributed this to the Chiari deformity, which acts as a "space-occupying lesion." MR confirms these early observations, and shows that the increased sagittal diameter persists over the length of the hindbrain descent (Fig. 7). In the past, surgeons used the number of segments widened on lateral plain films to estimate the level of hindbrain herniation, and therefore the lowermost laminectomy level [10].

Furthermore, the impaction of neural tissue in the upper cervical spine in newborn patients with myelodysplasia was a consistent finding in our series, and differs markedly from the normal appearance in infancy. The subarachnoid space normally present in front of and behind the cord was obliterated at birth in our Chiari II patients (Figs. 2A, 4A, and 7A). Therefore, we believe that the overall enlargement of the upper cervical canal, with scalloping of the dens (Fig. 7), is a pressure phenomenon, analogous to the clival and petrous scalloping seen above the foramen magnum [11]. If the canal enlargement were due to cervical cord hypogenesis with resultant mesodermal dysplasia of the canal, as postulated by Wolpert et al. [4], one would expect enlargement of the lower cervical canal, where the cord is small. However, the reverse is true: the canal is small below the vermis and medullary kink [1].

Enlargement of the upper precervical cord space was described by Wolpert et al. [4] in nine of 24 patients with myelodysplasia. As we have shown, at birth this wide subarachnoid space opposite C2 is nonexistent, but does increase with age to the point that most patients over 10 years old showed a ratio A/B (Fig. 6) of 0.5 or less.

In addition to the overall increase in the sagittal diameter of the canal, as described by Naik and Emory [1], two other factors contribute to a widened MR appearance of the precervical space: First, the dens is scalloped secondary to the impaction of tissues present in utero. Second, the vermis, medulla, and cord remain dorsally displaced in Chiari II patients. While in some cases this dorsal location is due to adhesions, other patients may have no demonstrable adhesions at surgery.

As shown in Figure 6, the decrease in ratio A/B is more or less continuous with age. If one defines widening as a ratio A/B of 0.5 or less, then in only one of 11 symptomatic patients was it found to be wide, whereas in seven of 22 asymptomatic patients it was wide.

Unfortunately, there is much overlap in these two groups when examined with MR imaging, particularly from birth to 2 years (Figs. 1 and 7). This was disappointing because it is this age group that most often develops severe brainstem symptomatology. Symptomatic Chiari II malformation is the leading cause of death in treated myelodysplastics up to age 2. While the precervical space tended to be narrower in symptomatic children older than 2 years, again, overlap with asymptomatic Chiari II cases limits the usefulness of this ratio as a predictor of brainstem dysfunction or long-tract signs.

Appearance and Level of Hindbrain Descent

An understanding of the MR appearance of the Chiari II hindbrain malformation is fundamental to the analysis of vermian and medullary herniation. Correlation of MR with recognized pathologic anatomy and appearance at surgical decompression formed the basis of our analysis.

Naidich et al. [2] emphasized the "cascade" of protrusions involving cervical cord, medulla, and vermis into the upper cervical canal, each of which causes anterior displacement and compression of structures ventrally. Vermian herniation and compression often involve the nodulus, uvula, and pyramis. Pathologically, this tissue is gliotic, edematous, and devascularized below the level of compression [2]. At the level of maximum compression (usually C1), there is frequently a plethora of vascularity.

The vermis was recognized in six patients (five symptomatic) on MR as a dark gray area (long T1) on T1-weighted images, comprising the dorsal one-third of the hindbrain hernia. Its distinct appearance related both to the compressive edema and gliosis (long T1), as well as to the contrast produced by its juxtaposition to the myelinated medullary kink (short T1) (Fig. 2). The low-signal vermis should not be confused with low-signal CSF in the fourth ventricle. Axial images will confirm the intimate relationship of the vermis as it envelopes the cervicomedullary kink (Fig. 2B). This appearance of the floor of the fourth ventricle as seen on axial CT with metrizamide was termed the "upper figure of eight" by Naidich et al. [2]. In cases in which the vermis was not gliotic, it appeared as the typical vermian peg, isointense to gray matter, dorsal to the medulla. The fourth ventricle at lower levels may be so compressed that it is a potential space, which may be stented open at the time of surgery. At other times, the fourth ventricle is dilated at its upper or lower end, and is visible ventral to the vermis.

The term "lower figure of eight" was used by Naidich et al. [2] to describe axial sections through the cervicomedullary kink. On sagittal MR, the kink is well seen as a rounded protuberance, dorsal to the cord, which may often extend one or two vertebral bodies below the gray vermis. On axial MR, it can be easily identified by the lack of gray vermian tissue dorsally, signifying a level below the vermis (Fig. 2C). The cervical cord below the kink is usually flattened and atrophic, and the transition is abrupt (Fig. 2D).

The choroid plexus was occasionally identified on MR. The choroid plexus is most often located at the tip of the vermis and, in vivo, appears as an orange nodular structure below the arachnoidal adhesions. When seen on MR, it may appear on sagittal images as a small cluster of grapes just dorsal to the lowest extent of the vermis (Figs. 2A and 2E). It has signal characteristics similar to the vermis. It is an important surgical landmark, for once it is identified, the floor of the fourth ventricle is nearby.

Review of the literature fails to reveal any consistent relationship between the level of the vermis or medulla and symptomatology. Hoffman et al. [12] found that in 15 patients undergoing decompression, the lower limit of the cerebellar vermis varied from C2 down to C7. Emery and MacKenzie [13] analyzed 100 children with myelomeningocele coming to

necropsy, classifying the deformities of the fourth ventricle and spinal cord. Seven of 10 patients displayed a medullary kink with or without cystic dilatation of the fourth ventricle. The vertebral body level at which the medullary kink descended was not tabulated. These investigators could not correlate the severity of medullary deformity and overlap with age at death. No attempt was made to differentiate between death due to brainstem symptoms versus death due to other causes, such as meningitis.

Naidich et al. [2] state that the length of the cerebellar tail correlates inversely with the age of the child at time of death. This, however, does not correlate with the length or level of medullary hernia [2].

In a larger series, Park et al. [14] described their results in 45 children younger than 3 months old who had posterior fossa decompression. They described the lowermost level of the cerebellar tongue or medullary kink at C1–C4 in 28 cases and C5–T1 in 17 cases. They state that the medullary kink typically extended caudal to the vermian peg, but they did not specifically distinguish between these two in stating the exact level of descent. They, and others, make the important point that failure to extend the laminectomy and dural opening below the lowermost level of the cerebellar tongue and medulla invites continued clinical deterioration postoperatively, requiring reexploration and extension of the laminectomy.

Venes et al. [3] correlated their surgical findings in 14 Chiari II patients with findings on preoperative MR imaging and intraoperative sonography. Their surgical approach varied depending on the presence or absence of fourth ventricular enlargement and hydromyelia. While these authors do not specifically state exactly at what levels the medullary kink occurred in each of their patients, in one figure the enlarged cystic fourth ventricle can be seen to extend down to C6. Wolpert et al. [4] show one child with brainstem compression in whom the medullary kink was at C4–C5 (see Fig. 7 of their article). Other asymptomatic patients appeared to have kinks above C3–C4.

Our results would suggest that if medullary kink is at the C3–C4 disk space or lower, there is a significant chance of the patient developing symptomatic brainstem or corticospinal tract dysfunction (Fig. 8). Of the 12 symptomatic patients (of whom 11 underwent surgery) in this series, eight were at C4 or lower (Table 2 and Fig. 2). While these results need to be confirmed in a larger series of patients, it seems intuitively logical that the lower the hernia, the more severe the medullary deformity and compression.

Identification of the vermis and cervicomedullary kink allows for a more limited and expeditious surgical procedure. The laminectomy need not be carried below the inferior extent of the cervicomedullary kink. Extensive laminectomy in young children, particularly if syringomyelia is present, may lead to a deforming kyphosis. By dissecting under the lower aspect of the vermis, the floor of the fourth ventricle can be identified in the majority of cases, and a small Silastic tube can be inserted to stent the fourth ventricle open and serve as a conduit for CSF into the cervical subarachnoid space.

Although difficult to identify at surgery because of gliosis, ischemia, and loss of surface anatomy, all cerebellar tissue involved in the caudalmost portion of the hindbrain hernia was

cerebellar vermis and not tonsil. Descent of the cerebellar tonsils through the foramen magnum can be seen, but it is almost never as severe as the degree of movement of the vermis in the malformations associated with myelodysplasia.

Correlation of the *in vitro* museum-specimen images with the clinical images was surprisingly good (Figs. 2F and 2G). Specimen detail was improved by using a 3-in. surface coil rather than a head coil. No significant artifact was generated from the preservative fluid or the mounting device. One would have expected better contrast between the vermis and medulla had the patient lived to be at least 6 months old, because of the normal myelinization that would have occurred. We believe this postmortem MR technique can be applied successfully to other museum specimens. Pathologic correlation can be performed in investigations such as this, where the pathology displayed may become available only once every few years. We are currently developing such a data base.

Retrocollis

One apparent correlate with clinical signs of brainstem compression was demonstration of retrocollis on MR images. Fixed retrocollis is an ominous sign of medullary compression [10]. We observed two patients whose retrocollis persisted throughout MR imaging, despite sedation and sleep (Fig. 4C). This was not seen in the asymptomatic group. An infant's neck should never be forcibly flexed to accommodate the MR procedure; the child should be scanned without manipulation, on his or her side if necessary.

CSF Flow Void

Although the presence or absence of a CSF flow void in the precervical space did not correlate with age, ratio, or symptomatology, it is important to point out that the pseudomass appearance caused by nonflowing, or flowing, dephased CSF in this wide precervical space may simulate a mass (Fig. 3). This pseudomass can be isointense with gray matter and resemble an intradural extramedullary process. GRASS or T2-weighted sequences, or CT-myelography, could be used to confirm the CSF characteristics if a serious question is raised.

C1 Arch

If the foramen magnum is enlarged in the Chiari II population, what is the cause of symptomatic neural compression of the herniated hindbrain? In many cases, the structural cause is a transverse band attached to the bifid lamina arches of C1, or the inner periosteum of the arch itself [15]. Naidich et al. [2] found that C1 is bifid in 70% of cases. In the large series reported by Park et al. [14] of 85 patients requiring surgery for apnea, stridor, and motor weakness, 41% had a transverse C1 band constricting the dural sac. These authors comment that division of the transverse dural band allowed the dural sac to expand immediately. Prompt reversal of bradycardia has also followed division of this band.

Surgical identification of a significant fibrovascular band or bony arch at C1 in nine patients prompted our correlation of this arch and its effects in both the symptomatic and asymptomatic group. Each arch of C1 could be identified by its bright marrow signal on T1-weighted parasagittal images (Figs. 2A and 4B). The fibrotic band, when present, appeared as an area of absent signal indenting the dorsal dura (Fig. 4). Axial images confirm the bifid laminar arches, if present (Figs. 2B and 4B). In nine of 12 symptomatic patients, the dura and neural tissues over C1 formed a $<90^\circ$ angle with the dura below (Figs. 1, 2A, and 4). Postoperatively, once the bone is removed the dura assumes a more normal contour, and dorsal migration of the neural elements is associated with widening of the precervical space (Fig. 5B).

The majority of asymptomatic patients with C1 dural indentation $<90^\circ$ were infants less than 1 year old, and this reflects again the overlap in appearance between symptomatic and asymptomatic children in the younger age groups (Fig. 7A).

Postoperative Changes

Of interest were the changes in the precervical space postoperatively. In five of 11 patients in whom postoperative scans were obtained, the precervical subarachnoid space widened postoperatively as compared with preoperative scans (Fig. 5). As stated earlier, this reflects canal widening secondary to the upper cervical laminectomy with relief of neural compression.

No change in the appearance of CSF flow void could be seen between studies. Further work is planned to elucidate the changes in CSF dynamics around the hindbrain in the postoperative state.

One puzzling observation was the enlargement of the fourth ventricle after decompression in three of five patients, despite placement of a fourth ventricle to subarachnoid space shunt. This enlargement was severe enough to cause symptoms in one case, requiring a separate decompression. Venes et al. [3] have written about their experience with encystment of the fourth ventricle after surgery. However, their suggestion of placing an internal shunt from the fourth ventricle into the subarachnoid space was not always successful in our series. The progression and outcome of this subset of Chiari II children is an area of ongoing investigation and interest. It should be emphasized that the recurrence of brainstem compressive symptoms postoperatively should prompt MR imaging or CT to exclude a trapped fourth ventricle.

Follow-up of operated patients had mixed results. The older patients who presented primarily with increasing spasticity of the upper extremities were improved by surgery. The infants with lower cranial nerve dysfunction did less well, with half (three of six) showing marginal to no improvement. The criteria for selecting patients for surgery were strict [6] and chosen to identify patients with life-threatening symptoms. It may be that those infants identified for surgery were so severely affected by their deformity that to expect substantial improvement may be unrealistic. If this is the case, then earlier selection of patients by imaging or physiologic criteria may improve this group's long-term functional status. Earlier sur-

gery, decided on the basis of additional selection criteria, is currently under investigation.

Conclusions

The spectrum of MR features characterizing the hindbrain deformity reflects the variable clinical expression in Chiari II patients, ranging from normal brainstem function to severe spasticity and apnea. A medullary kink at C4 or lower was seen only in symptomatic patients. The precervical cord space is obliterated at birth in all Chiari II patients, increases with age and following surgery, and tends to be greater in asymptomatic patients, although great overlap exists, particularly in infancy. A constrictive fibrous or bony C1 arch and its relation to the dura can be identified preoperatively in most patients. MR analysis provides the pediatric neurosurgeon additional important data upon which to base a decision to operate on these children.

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