The Clinical Significance of Hindbrain Herniation and Deformity as Shown on MR Images of Patients with Chiari II Malformation

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This study investigates whether the degree of brainstem herniation and the nature of the cervicomedullary deformity seen on sagittal plane MR images correlates with the clinical syndrome in patients with the Chiari II malformation. The amount of brainstem herniation was assessed by relating the position of the midbrain and pons to the sella turcica and the anterior lip of the foramen magnum, respectively. The cervicomedullary deformity was graded into degrees of increasing severity. We found that the neurologic status of these children was not affected by either the amount of herniation or the characteristics of the cervicomedullary deformities.

Because of these findings, we believe that other factors, such as disorganization of the brainstem nuclei, may be the likely cause for the breathing and swallowing difficulties experienced by children with the Chiari II malformation.

MR imaging has vastly improved visualization of the Chiari II malformation, revealing extensive manifestations of the defect throughout the CNS [1, 2]. We previously reported the MR findings in 24 children with the malformation [1] and found no specific clinical correlation with obliteration of the subarachnoid spaces at the foramen magnum—a fact that we believed was important because of the supposed role of this obliteration in the production of symptoms of lower cranial nerve dysfunction. Surgical decompression of this area has been reported as leading to significant relief of the clinically related symptoms in certain reported series [3]. Others, however, have expressed some skepticism over the efficacy of surgery in this setting, believing that only patients with symptomatology suggesting acute compression of the upper cervical cord such as the acute onset of upper extremity weakness, opisthotonus, or spasticity merit surgery [4].

We speculated that the degree of downward herniation of the brainstem, an anatomic finding that would not be affected by a posterior decompression, might be closely related to the patient's clinical status particularly that due to lower cranial nerve dysfunction. We also questioned whether the type of cervicomedullary herniation as well as the presence of wide subarachnoid spaces in front of the upper cervical cord in many of these patients [1] had any relationship to the clinical status.

We therefore reviewed the MR images of 37 patients with the Chiari II malformation, analyzed the nature and degree of the brainstem and cervicomedullary deformities, and correlated these appearances with the clinical status.

Material and Methods

Thirty-seven patients all initially presenting with myelomeningoceles and in whom MR was subsequently carried out form the basis for this report. Two patients were less than 1 year old, 11 were 1–5 years old, 10 were 6–10 years old, and 14 were 11–26 years old. The patients were classified into two grades. Patients in grade 1 were normal or had problems with lower leg extremities necessitating leg braces or crutches. They had no signs of cranial nerve dysfunction. Patients in grade 2, in addition to lower extremity symptoms, had upper

Received January 13, 1988; accepted after revision April 29, 1988.

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AJNR **9:1075–1078, November/December 1988**
0195–6108/88/0906–1075

extremity symptoms such as progressive weakness or spasticity. Many also had swallowing difficulties such as apnea or stridor, suggesting lower cranial nerve symptomatology.

All the patients were scanned with a 1-T scanner,* and although they were all investigated in three orthogonal planes (axial, sagittal, and coronal), for the purposes of this study only the sagittal plane was used. T1-weighted images were used exclusively in the study. None of the patients had cerebral MR abnormalities other than those associated with the Chiari II malformation.

The position of the pons and midbrain were categorized according to the relationship of the pontomesencephalic and pontomedullary junctions to constructed lines through the sella floor, clivus, and foramen magnum (Fig. 1). As a normal control we assessed the position of the midbrain and pons in a series of 40 patients scanned for reasons other than suspected Chiari II malformation and in whom the scans were all normal. Three of these patients were less than 1 year old, 25 were 1–9 years old, and 12 were 10–15 years old.

The types of cervicomedullary herniation were also grouped into three categories of increasing severity, as previously described [1]. In group 1, the fourth ventricle and medulla did not descend through the foramen magnum, and the sole evidence of a hindbrain deformity was an inferior vermial peg extending through the foramen. In group 2, the fourth ventricle descended vertically through the foramen magnum in front of the vermial peg. In group 3, the medulla was buckled forming a cervicomedullary "kink" or "spur" behind the upper cervical cord and was associated with either a normal-sized or enlarged fourth ventricle [1].

The final assessment was of the upper cervical canal and its contents. We arbitrarily defined a wide subarachnoid space anterior to the upper cervical cord if the space was equal to or greater than half the diameter of the adjacent vertebral body.

Each of these groups of abnormalities, the positions of the pons and midbrain (both divided into three grades of increasing severity), the type of cervicomedullary herniation, and the presence or absence of a wide space anterior to the upper cervical cord were related to the clinical grades.

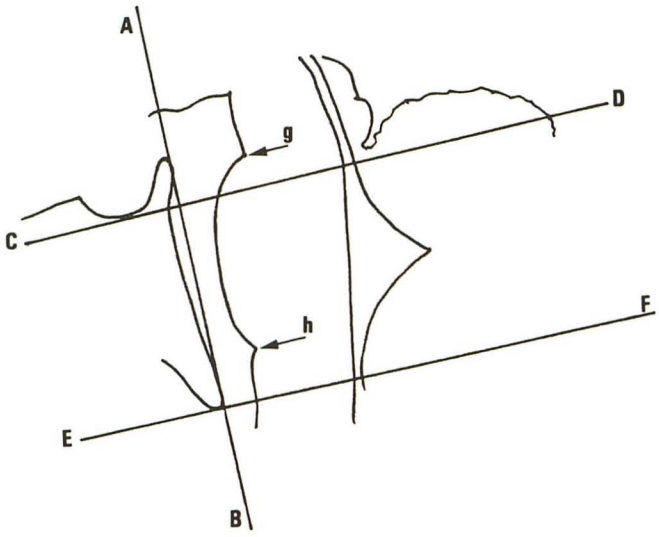

Fig. 1.—Line AB is initially constructed between posterior margin of dorsum sellae and anterior lip of foramen magnum. Line CD is at right angles to AB tangent to the floor of the sella turcica; line EF is at right angles to AB through anterior lip of foramen magnum. The distance between g, the pontomesencephalic junction, and CD is a measure of midbrain displacement; the distance between h, the pontomedullary junction, and EF is a measure of pontine displacement. Measurements of g and h above the lines CD and EF are shown in the tables as positive values; measurements below the lines are shown as negative values. (Based on Weinstein and Newton [11].)

Results

There was no statistical relationship between the degree of caudal displacement of the midbrain or of the pons through the foramen magnum and the clinical status although the data suggest a trend toward a worse status with increasing herniation (Table 1 and Figs. 2, 3, and 4). The types of cervicomedullary deformities and the presence of a wide space in front of the upper cervical cord also did not correlate with the clinical status of the patients. While there was poor correlation between the degree of anatomic deformity and the clinical status, certain correlative anatomic features were noted. Seven of the 11 patients with severe caudal pontine herniations (−1 mm to −14 mm) also had severe caudal midbrain displacement (−7 mm to −14 mm). Seven of the 11 patients with severe pontine herniations also had medullary spurs. The clinical grades did not correlate with the ages of the patients (Table 2).

Discussion

A certain percentage of children with myelomeningoceles will develop signs and symptoms referable to their Chiari

TABLE 1: Relationship Between Amount of Pontine Herniation, Types of Cervicomedullary Deformity, and Presence of Precervical Subarachnoid Spaces, and the Clinical Presentation of Patients

	Clinical Grades		No.
	1	2	
Pontomesencephalic distance (mm)[a]			
Normal 1.5 (2.8) mm			
0 to 7	11	2	13
−1.5 to −6	9	3	12
−7 to −14	8	4	12
Pontomedullary distance (mm)[b]			
Normal 12 (4.7) mm			
1 to 10	9	0	9
0	12	5	17
−1 to −14	7	4	11
Cervicomedullary category			
1	5	1	6
2	12	5	17
3	11	5	16
Wide precervical cord subarachnoid spaces			
Present	15	3	18
Absent	13	6	19

Note.— ( ) = S.D.
[a] $p > .10$.
[b] $p > .10$.

* Siemens Corp., Iselin, NJ.

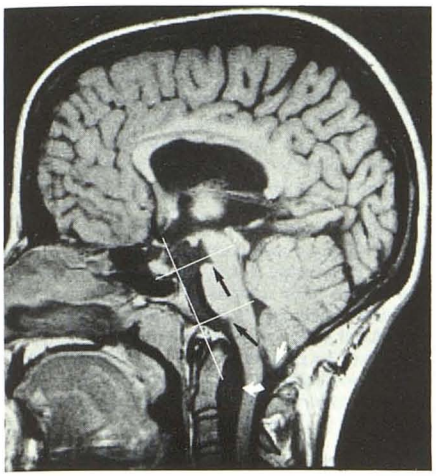

Fig. 2.—17-year-old girl born with lumbar myelomeningocele repaired at birth. Currently, and at time of MR scan, she is walking independently with the use of braces. Her upper extremity function is normal with normal reflexes and no upper extremity spasticity. There is no hoarseness or stridor. Her knee jerks are absent. She is clinically classified as grade 1. The pontomesencephalic junction (*upper black arrow*) is 2 mm below the upper line, and the pontomedullary junction (*lower black arrow*) is 2 mm below the lower line. An inferior vermial peg (*straight white arrow*) is herniating through the foramen magnum (group 1 cervicomedullary herniation). Note the wide precervical cord subarachnoid space (*curved white arrow*).

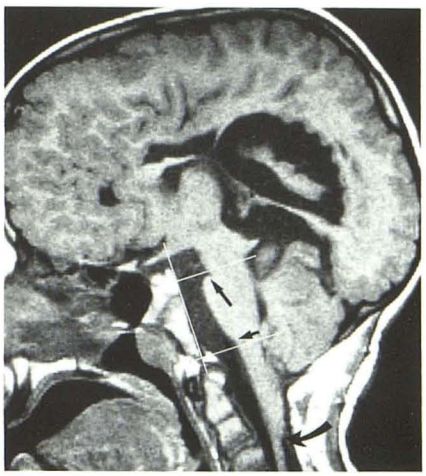

Fig. 3.—7-year-old boy born with large thoracolumbar myelomeningocele repaired at birth. Because of laryngeal stridor a cervical decompression with some improvement was performed at age 2. His upper extremity function was normal but at age 5 he developed slurred speech and progressively increasing spasticity of the upper extremities. He is paraplegic and is clinically classified as grade 2. The MR image shows that the pontomesencephalic junction (*upper black arrow*) is 2 mm below the upper line, and the pontomedullary junction (*lower black arrow*) is 2 mm above the lower constructed line. Note also the normal position of fourth ventricle. The precervical cord subarachnoid space is wide but this could be due to the cervical decompression. The soft-tissue mass (*curved arrow*) is probably an inferior vermial peg, since a medullary kink is usually associated with downward herniation of the fourth ventricle (group 1 cervicomedullary herniation). An interhemispheric cyst with partial agenesis of the corpus callosum is also present.

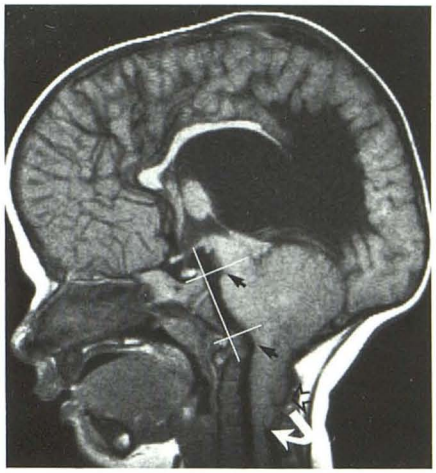

Fig. 4.—1-year-old boy born with lumbar myelomeningocele repaired at birth. At the time of the MR image he is alert, active, with no difficulty swallowing or chewing. His upper extremity function is good without spasticity. Ankle and knee jerks are unobtainable. Clinically he is classified as grade 1. The pontomesencephalic junction (*upper black arrow*) is 2 mm below the upper line, and the pontomedullary junction (*lower black arrow*) is 7 mm below the lower line. The fourth ventricle is not seen. Both an inferior vermial peg (*open arrow*) and a medullary kink (*curved arrow*) are present (group 3 cervicomedullary herniation). The precervical cord subarachnoid space is not wide. Note also the large interhemispheric cyst and absence of the posterior part and splenium of the corpus callosum. (Reprinted from [1].) When seen at age 33 months he was continuing to do extremely well. He was using crutches because of his absent lower extremity function. Verbally he was using complex sentences, able to count to 10, and sing songs.

malformation [4]. The spectrum of signs and symptoms suggests dysfunction of the lower cranial nerves, caudal brainstem, and cerebellum. A number of theories has been postulated to account for these observations. The displacement of the caudal brainstem may put traction or pressure on the lower cranial nerves [5]. Abnormal descent and looping of the basilar-vertebral circulation have been demonstrated in patients with the Chiari II malformation [6], and it has been postulated that the cause of the symptoms may be vascular compression [5]. Medullary hemorrhages possibly due to the abnormal configuration of the medulla and foramen magnum or to venous congestion have been invoked as causes for laryngeal stridor in Chiari II patients [7].

Our data do not support the theory of traction on the lower cranial nerves. As shown in Table 1 and in Figs. 2, 3, and 4, neither downward herniation of the midbrain nor of the pons correlates with poor outcome. Similarly, the degree and nature of the hindbrain deformity and the presence of wide precervical subarachnoid spaces did not correlate with outcome. We are unable to comment about compression of the vertebral and basilar arteries since these are not reliably seen by MR. However, other studies in which angiography was performed to define the Chiari malformation [8], while demonstrating deformity of the vertebrobasilar system, failed to demonstrate occlusion of these vessels. We cannot with certainty exclude medullary hemorrhage, since T2-weighted images were not obtained so that the presence of hemosiderin, a sign of chronic hemorrhage, could not be determined; however, on the T1-weighted images, the bright signal of methemoglobin was not seen.

TABLE 2: Relationship Between Age and Clinical Presentation of Patients

Age	Clinical Grades		No.
	1	2	
Less than 1 year	2	0	2
1–5 years	8	3	11
6–10 years	6	4	10
11–26 years	12	2	14

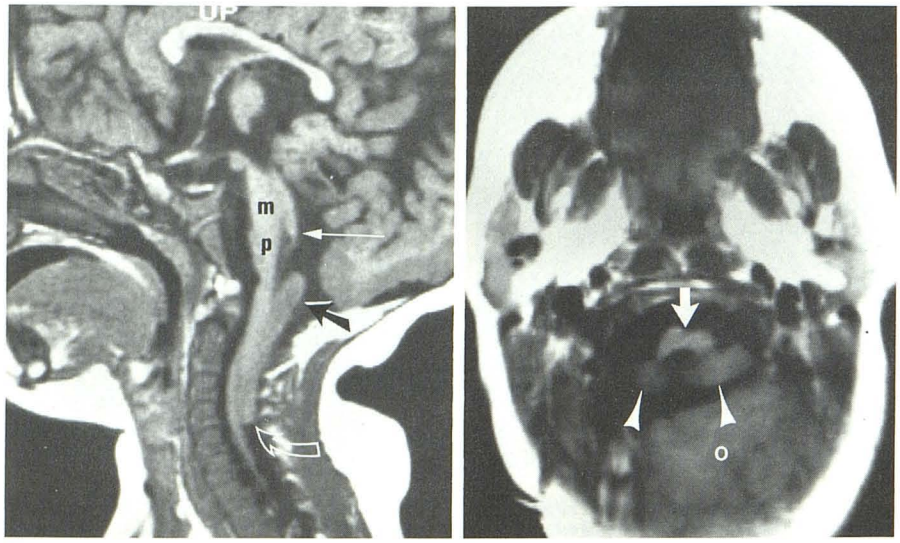

Fig. 5.—17-month-old girl born with lumbar meningomyelocele closed at birth. Episodes of stridor and cyanosis on crying were noted at the age of 1 week. At the time of the MR image her chest showed inspiratory stridor and she also had a left sixth nerve palsy. The motor movement and coordination of the upper extremities were poor with slightly depressed reflexes. The knee jerks were present but the ankle jerks were absent.

***A*, There is marked hypogenesis of the cerebellum (*oblique* and *curved arrows*), which lies almost completely in the upper cervical canal. The tectum (*thin straight arrow*) is beaked; the midbrain (m) and pons (p) are hypogenetic.**

***B*, Axial image at level of pons shows hypogenesis of the pons anteriorly (*arrow*), cerebellar hemispheric lobules (*arrowheads*) on either side of an open fourth ventricle, and the occipital lobe posteriorly (o). (Reprinted from [1].)**

We therefore agree that the most likely cause for the symptomatic Chiari patient with breathing and swallowing deficits is probably the disorganization of brainstem nuclei and other associated anomalies described by Gilbert et al. [9] and by Cameron [10]. Gilbert et al. demonstrated absence or reduction in the cell numbers in the cranial nerve, basal pontine, or olivary nuclei or in the tegmentum in almost half their patients [9]. Although we previously demonstrated pyramidal hypogenesis in four of 24 patients with Chiari II malformation (Fig. 5), we were unable to further analyze the cranial nerve nuclei since we cannot with confidence prospectively define these nuclei in normal patients either with T1- or T2-weighted images.

Gilbert et al. [9] also described cerebral cortical and cerebellar dysplasia consisting of polymicrogyria and heterotopia in a majority of their patients. Cameron [10] in a study of 26 patients with the Chiari II malformation found cerebellar and cerebral dysplasia in the majority, with microgyria in some. We found no patient with subependymal nodules in the wall of the ventricle (a common manifestation of cortical heterotopias), although in many of our patients cortical gyri indented the surfaces of interhemispheric cysts [1]. Gilbert et al. [9] described microgyria (small gyri with a normal six-layered pattern) in three of their 25 patients. Stenogyria (small closely placed gyral folds of histologically normal cortex) were seen in seven of our initial 24 patients. Agenesis of the corpus callosum, seen in three of the 25 patients studied by Gilbert et al. [9], was seen in eight of our 24 patients [1].

We concluded, therefore, that although MR imaging can provide exquisite detail about the neuroanatomy of the Chiari II patient, unfortunately it cannot provide data to assess prognosis or to select patients for upper cervical decompression. It is possible that operations for patients with symptomatic Chiari II malformation should be withheld if MR fails to demonstrate a surgically correctable finding.

REFERENCES

1. Wolpert SM, Anderson M, Scott RM, Kwan ESK, Runge VM. Chiari II malformation: MR imaging evaluation. *AJNR* **1987**;8:783–792
2. El Gammal T, Mark EK, Brooks BS. Mr imaging of Chiari II malformation. *AJNR* **1987**;8:1037–1044
3. Park TS, Hoffman HJ, Hendrick EB, Humphreys RP. Experience with surgical decompression of the Arnold-Chiari malformation in young infants with myelomeningoceles. *Neurosurgery* **1983**;13:147–152
4. Bell WO, Charney EB, Bruce DA, Sutton LN, Schut L. Symptomatic Arnold-Chiari malformation: review of experience with 22 cases. *J Neurosurg* **1987**;66:812–816
5. Sieben RL, Hamida MB, Shulman K. Multiple cranial nerve deficits associated with the Arnold-Chiari malformation. *Neurology* **1971**;21:673–681
6. Emery JL, Levick RK. The movement of the brain stem and vessels around the brain stem in children with hydrocephalus and the Arnold-Chiari deformity. *Dev Med Child Neurol* **1966**;8 (suppl):11:49–60
7. Morley AR. Laryngeal stridor, Arnold-Chiari malformation and medullary haemorrhages. *Dev Med Child Neurol* **1969**;11:471–474
8. Gabrielsen TO, Seeger JF, Amundsen P. Some new angiographic observations in patients with Chiari type I and II malformations. *Radiology* **1975**;115:627–634
9. Gilbert JN, Jones KL, Rorke LB, Chernoff GF, James HE. Central nervous system anomalies associated with meningomyelocele, hydrocephalus, and the Arnold-Chiari malformation: reappraisal of theories regarding the pathogenesis of posterior neural tube closure defects. *Neurosurgery* **1986**;18:559–564
10. Cameron AH. The Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. *J Pathol Bacteriol* **1957**;73:195–211
11. Weinstein M, Newton TH. Caudal dislocation of the pons in the adult Arnold-Chiari malformation: an angiographic evaluation. *AJR* **1976**: 126:798–801