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Significance of Cerebellar Tonsillar Position on MR

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It has been noted that a low degree of ectopia of the cerebellar tonsils on MR is of questionable significance. We measured the position of the cerebellar tonsils with respect to the inferior aspect of the foramen magnum in 200 normal patients and in 25 patients with a firm diagnosis of Chiari I malformation. In the normal group, the mean position of the tonsils was 1 mm above the foramen magnum with a range from 8 mm above the foramen magnum to 5 mm below. In the patients with Chiari I malformations, the mean position was 13 mm below the foramen magnum with a range from 3 mm below the foramen magnum to 29 mm below. Fourteen percent of normal patients had tonsils extending slightly below the foramen magnum. If 2 mm below the foramen magnum is taken as the lowest extent for tonsils in a normal patient, our sensitivity in predicting symptomatic patients is 100% and our specificity is 98.5% (three false positives). If 3 mm below the foramen magnum is taken as the lowest normal tonsillar position, our sensitivity is 96% and our specificity is 99.5%. MR demonstration of less than 2 mm of tonsillar ectopia is probably of no clinical significance in the absence of syringomyelia.

The Chiari I malformation is defined as displacement of the cerebellar tonsils into the cervical spinal canal [1]. Unlike many malformations of the central nervous system (CNS), this entity manifests itself in early adulthood and middle age, often with a confusing clinical picture [2–4]. Additionally, a low degree of cerebellar ectopia is often of no clinical significance [6, 7]. The advent of MRI has provided an easy and noninvasive method of imaging the cerebellar tonsils and their relationship to the foramen magnum. The purpose of this study is to establish a normal range for tonsillar position and to determine the significance of various degrees of tonsillar ectopia.

Materials and Methods

The MR studies of 200 patients with clinical signs or symptoms unrelated to the Chiari I malformation or of other cervicocranial junction abnormalities were evaluated retrospectively. The examinations of 25 additional patients with definite Chiari I malformation and associated clinical signs and symptoms were also reviewed as a separate group.

Measurements were made from the lower level of the foramen magnum to the bottom of the lowest lying tonsil. The bottom of the foramen magnum was defined as a line extending from the lowest cortical bone seen (by way of lack of signal) at the posterior lip of the foramen magnum (opisthion) to the lowest cortical bone of the clivus (basion) (Fig. 1).

Measurements were all made on T1-weighted spin-echo images with TR of 250 to 700 msec and TE of 30 to 40 msec. T2-weighted images were difficult to evaluate because of decrease in signal-to-noise ratio with long TE and, more importantly, diminished contrast resolution between CSF and tonsil from the increased relative intensity of the CSF [8]. Proton-density images were advantageous because the slight graying of the CSF creates a sharper contrast with bone while contrast between CSF and tonsils is maintained. Longer imaging times make this less practical, however. All measurements were performed on sagittal images. All the authors agreed that tonsillar position was much more difficult to evaluate on coronal

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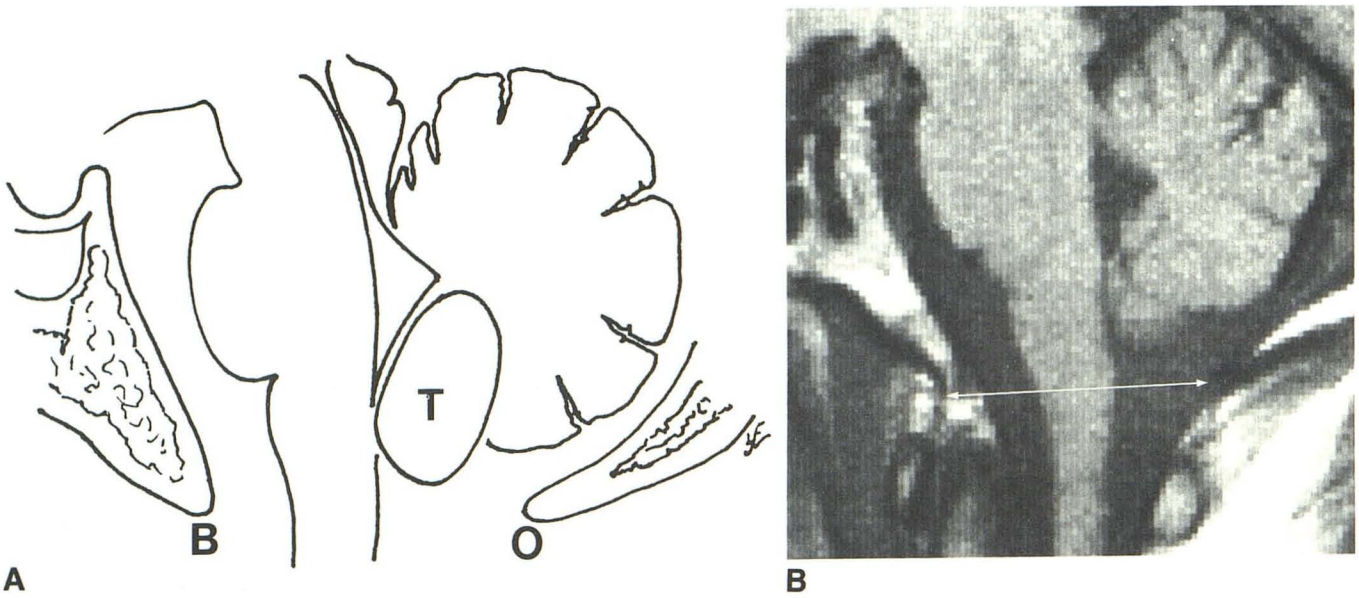


Fig. 1.—**A**, Drawing showing basion (B), opisthion (O), and cerebellar tonsil (T) in a normal patient. **B**, Midline sagittal section, SE 500/40, showing line from basion to opisthion in a nomal patient. Measurements were from this reference line. Bottoms of tonsils have a normal, rounded appearance and CSF is seen in a normal cisterna magna.

images (see discussion and Fig. 2). Distances were measured with calipers from the hard copy of the images. Measurements were to the nearest millimeter.

Patients with lesions that might affect tonsillar position were eliminated from the study. Specifically, this included any posterior fossa lesion, hydrocephalus of any origin, and supratentorial lesions causing any evidence of transtentorial herniation.

One hundred eighty-three images had a slice thickness of 1 cm and 42 images had a slice thickness of 5 mm. Ten patients had a 1-cm midline "scout" image as well as 5-mm images during a multislice sequence; measurements were identical, implying no difference in the methods.

Two scanners were used. One hundred forty-four studies were done on a 0.5-T Picker International superconducting scanner (pixel size 1.1 × 1.1 mm) and 81 were done on a 0.6-T Technicare superconducting scanner (pixel size 0.9 × 0.9 mm). Four signal acquisitions, a 256 × 256 matrix, and 2DFT reconstruction algorithms were used on all scans.

Results

Histogram results of the studies on the 200 normal and 25 Chiari I patients are given in Figure 3. The tonsillar position in the normal patients ranged from 8 mm above the foramen magnum (+8 mm) to 5 mm below the foramen magnum (−5 mm) with a mean value of +1 mm and standard deviation of 1.9 mm. In the 25 Chiari I patients, the range was from 29 mm below the foramen magnum (−29 mm) to 3 mm below the foramen magnum (−3 mm) with a mean value of −13 mm and standard deviation of 7.1 mm. As expected, the difference in means was significant ($p = 0.0001$ using the unequal variance T-test) (Table 1).

Sensitivities and specificities were calculated using 0, −1, −2, −3, −4, and −5 mm as the cutpoint (lowermost limit for normal tonsil position [See Table 2]). High sensitivities and specificities were obtained at all cutpoints from −1 to −4. The greatest overall accuracy was obtained at cutpoints of −2, −3, and −4.

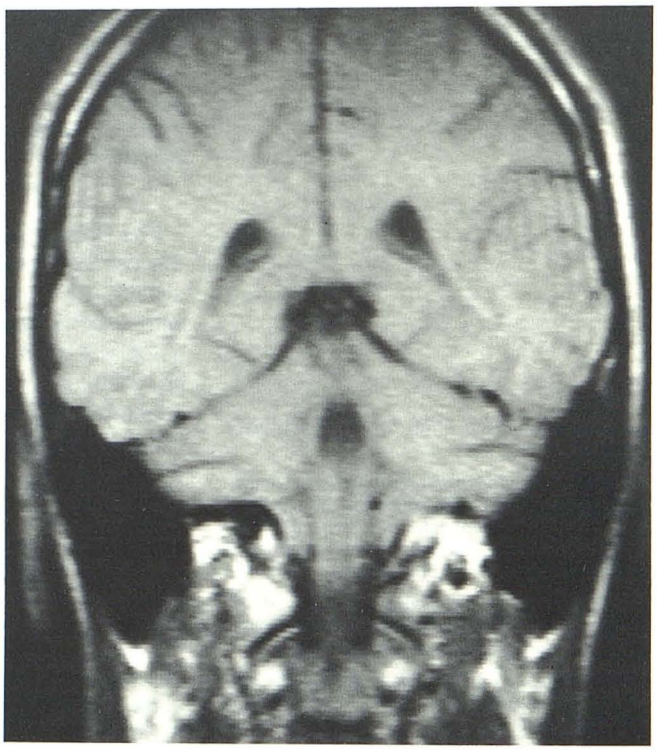


Fig. 2.—Coronal SE 700/40 through medulla and tonsils. The lower limits of foramen magnum are difficult to identify. Coronal scans were not used for this reason.

Scans were also examined for evidence of narrowing of the

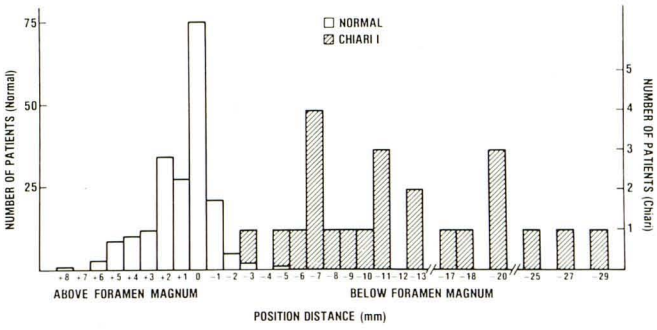


Fig. 3.—Histogram showing tonsillar positions in 200 normal and 25 Chiari I patients. Open boxes represent normal patients. Cross-hatched boxes represent patients with Chiari malformation.

TABLE 1: **Tonsil Position**

	No. of Patients (n = 225)	P (mm)	SD
Normal Pts	200	+1.0	1.90
Chiari I Pts	25	−13.12	7.08

Note.—P = average position of tip of tonsil with respect to lower border of the foramen magnum; SD = standard deviation.

TABLE 2: **Accuracy of Tonsil Position in Determination of Chiari I Malformations**

Cutpoint	Sensitivity	Specificity
0	100	85.43
−1	100	95.98
−2	100	98.49
−3	96.00	99.50
−4	96.00	99.50
−5	92.00	100

Note.—Cutpoint is the highest position considered a positive result; i.e., if zero is the cutpoint, any patient with tonsils at zero or below is considered to have Chiari I.

subarachnoid space or compression of the tonsils at the foramen magnum. All patients with ectopia of more than 1 mm showed narrowing or complete loss of the CSF spaces of the foramen magnum and cisterna magna. Pointed or "peglike" tonsils, as previously described in Chiari I malformations [8], were seen in both the symptomatic and asymptomatic patients with tonsillar ectopia. One patient had pointed tonsils prior to a suboccipital craniectomy but normally rounded tonsils on a postoperative scan (Fig. 4). Cervicomedullary kinking was present in only three of the 25 symptomatic patients (12%) and in none of the asymptomatic patients.

Discussion

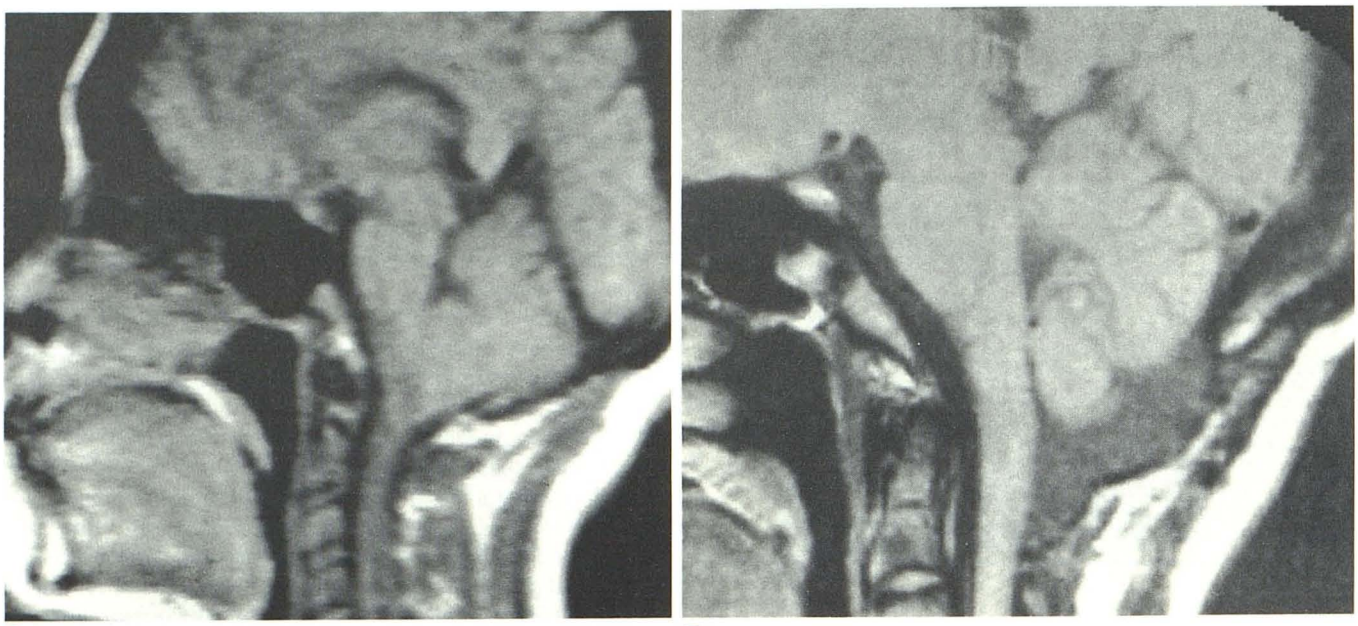
In 1891, Chiari [1] published his initial paper on the hindbrain malformations that now bear his name. He divided the malformations into three types: Type I—Displacement of the cerebellar tonsils, and sometimes the lower vermis, into the cervical spinal canal. Type II—Displacement of the brainstem and lower cerebellum into the cervical spinal canal such that the elongated fourth ventricle also extends below the foramen magnum. Type III—Downward displacement of the medulla with herniation of the cerebellum first through the foramen magnum and then dorsally through a cervical spina bifida, forming a cervical encephalocele. (Only a single case of this type was described.)

The Chiari III anomalies are clinically obvious and are diagnosed at birth. Chiari II anomalies are associated with multiple other CNS malformations [9], most notably hydrocephalus and meningomyelocele, and are usually diagnosed in infancy or childhood. In Chiari I malformations, the anatomic abnormalities are less obvious, consisting of tonsillar ectopia with variable syringohydromyelia and bony craniocervical junction anomalies [2–5]. The clinical findings are diverse and inconsistent and tend to be manifested in early to middle adulthood; indeed, the patients are frequently misdiagnosed as having multiple sclerosis [2, 3]. Cranial nerve and brainstem compression can cause vertigo, facial pain, sensorineural deafness, or bulbar palsy. Cerebellar signs include ataxia, nystagmus, and oscillopsia. Also seen are spastic paraparesis secondary to compression of the pyramidal tracts, impaired proprioception, and vibratory sense from dorsal column involvement and muscle wasting with pain in the upper extremities secondary to syringomyelia [3].

Compounding the difficulty in clinical diagnosis is the question of the significance of a low degree of tonsillar ectopia. In 1963, Baker [6] noted 14 cases in which all myelographic findings were normal except that the tips of the tonsils extended 2 to 5 mm below the foramen magnum. Significantly, there was no obstruction to the free flow of contrast through the foramen magnum, implying no compression at that site. In contrast, O'Connor et al. [10], in 1973, measured tonsillar position retrospectively in 100 patients without posterior fossa or foramen magnum lesions and found that the position of the tonsillar tips was above the foramen magnum in all cases. In 1974, Bloch et al. [7] measured the position of the tonsillar tips with respect to the upper lip of the foramen magnum on myelograms of 60 "normal" and 19 "pathologic" patients. Using his results, the mean position for a tonsil is 1.3 mm below the upper lip of the foramen magnum. Considering that he counted each tonsil separately and that one tonsil is commonly 2–4 mm lower than the other [7], his results compare favorably with our mean value of 1 mm above the lower foramen magnum. The depth of the foramen magnum is usually 4–10 mm. Unfortunately, Bloch did not define his "normal" or "pathologic" groups, so there may be some variation in the populations studied.

Patients who did not clearly fall into the "normal" or "Chiari I" category were eliminated from our study. By this process, some minimally symptomatic or as yet asymptomatic patients may have been excluded, possibly yielding a somewhat low mean tonsillar position for this group.

The position of the tonsils was determined from sagittal images for several reasons. First, short TR, short TE sagittal



A B

Fig. 4.—**A**, Midline sagittal SE 500/40 scan in a symptomatic patient. Tonsils extend 1.1 cm below foramen magnum. Tonsils are "pointed" and cisterna magna is obliterated. The latter two findings were seen in both asymptomatic and symptomatic patients with low tonsils. **B**, Midline sagittal SE 500/40 scan after decompression via suboccipital craniectomy. Tonsils now have a normal rounded appearance.

midline spin-echo images are routinely obtained at our institution as a guide for axial and coronal slice positioning, so it was easy to obtain these images with no added examination time or discomfort for the patient. Moreover, it was more difficult to identify the foramen magnum on the coronal images (Fig. 2). The reasons for this include the lack of a high-signal-intensity diploic space to highlight the lack of signal from the cortical bone and the undulations and multiple foramina of the skull base lateral to the foramen magnum.

We could not find a significant difference in the appearance of the tonsils or basilar cisterns in the patients with asymptomatic cerebellar ectopia when compared with those with symptomatic Chiari malformations. The cisterna magna was small or nonexistent and the foramen magnum was filled with the medulla and "pointed" tonsils in both groups. The pointed appearance of the tonsils is most likely secondary to their compression within the foramen magnum. This is demonstrated by the patient illustrated in Fig. 4, in whom markedly pointed tonsils reverted to a more normally rounded appearance after decompression by a suboccipital craniectomy. The fact that compression of the tonsils is seen in asymptomatic patients would suggest that compression itself is not the primary determinant of the clinical manifestations of low-lying tonsils.

The question of why some patients with mild tonsillar ectopia are asymptomatic has been addressed by previous authors [3, 4, 11, 12]. The symptoms most likely result from the build-up of arachnoidal scarring and adhesions near the foramen magnum, possibly secondary to the repetitive rubbing of the tonsils against the bone over many years. These adhesions can cause additional or new compression of the tonsils and brainstem. They can also obstruct the flow of CSF from the basilar cisterns into the cervical subarachnoid space, which has been implicated as a possible factor in the development of syringomyelia [11].

We found cervicomedullary kinking in only three of our 25 Chiari I patients (12%). This frequency is identical to that found by Paul et al. [5] but far less than that found by Spinos et al. [8]. We did not observe such "kinking" in the normal group. This finding is, therefore, helpful when present.

A striking demarcation was noted in the degree of tonsillar ectopia between the symptomatic and asymptomatic patients. If the tonsillar tips extended 3 mm or more below the foramen magnum, all patients except one were symptomatic. No symptomatic patients had less than 3 mm of tonsillar ectopia. This was the major difference between the groups on MR.

Perhaps the greatest danger in evaluating patients for Chiari malformations is to call a positive patient's MR findings normal, and thus to end the work-up for foramen magnum lesions. It is probably better to err on the side of a false positive diagnosis, whereby further evaluation for cmpression at the foramen magnum (by myelography, for example) would reveal a lack of compression and direct the work-up elsewhere. Using this logic, a cut-off of −2 mm gives the best overall yield with no false negatives and a minimum of false positives.

In summary, there are multiple MR findings in patients with Chiari I malformations. The degree of tonsillar ectopia is the finding that most closely corresponds to the presence of clinical symptoms. Ectopia of 2 mm or less is unlikely to be of clinical significance.

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