

## Definition of the adult Chiari malformation: a brief historical overview

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With the widespread use of newer neuroimaging techniques and modalities, significant tonsillar herniation is being diagnosed in more than 0.5% of patients, some of whom are asymptomatic. This puts the definition of the adult Chiari malformation to the test. The author provides a historical review of the evolution of the definition of the adult Chiari malformation in the neurosurgery, radiology, and pathology literature.

**KEY WORDS • adult Chiari malformation • Chiari I malformation • tonsillar ectopia • syringomyelia**

There is confusion in the literature regarding the concept of Chiari I malformation or adult Chiari malformation. Recent advances in neuroimaging modalities and their widespread use has led to an increase in the number of patients with radiological evidence of tonsillar herniation, some of whom are asymptomatic, raising questions as to its true clinical relevance. This is especially true because its incidence has been found to be between 0.56%<sup>37</sup> and 0.77%<sup>65</sup> on MR imaging studies, as well as 0.62% in brain dissection studies.<sup>41</sup> With that in mind, we attempted to review the various historical steps that marked the evolution of the definition of adult Chiari malformation.

### CHIARI'S INITIAL REPORT

The entire concept of these malformations emerged toward the end of the 19th century from Chiari's initial descriptions of "alterations in the cerebellum resulting from cerebral hydrocephalus."<sup>22,30</sup>

Hans Chiari (1851–1916) was born in Vienna, Austria. His father was the famous gynecologist J. B. V. L. Chiari<sup>80</sup> and his brother was the rhinolaryngologist Ottokar Chiari.<sup>55</sup> He graduated from medical school in 1875 and became assistant to the Austrian pathologist Karl Rokitan-sky at the Institute of Pathology in Vienna. Chiari himself became professor of Pathology in Prague, Czechoslo-vakia, in 1882, and in Strasbourg, France, in 1906. His initial work on what would become known as Chiari malformation was published in *Deutsche Medizinische Wo-chenschrift* in 1891 and entitled "Concerning alterations in the cerebellum resulting from cerebral hydrocephalus."<sup>22</sup>

Abbreviations used in this paper: CSF = cerebrospinal fluid; ICP = intracranial pressure; MR = magnetic resonance.

As the title suggests, his goal was to describe "the consecutive changes established in the region of the cerebel-lum by cerebral hydrocephalus." The first type he described, which came to be known as Chiari Type I, was characterized by "elongation of the tonsils and medial divisions of the inferior lobules of the cerebellum into cone shaped projections which accompany the medulla oblongata into the spinal canal" (Fig. 1). These features were demonstrated in a "relatively large percentage of cases of chronic congenital hydrocephalus but never with-out hydrocephalus or in cases of acute or later-developing hydrocephalus .... The elongated portions of the cerebel-lum can show either normal structure, fibrosis or soften-ing...and...extend nearly to the top of the atlas, however in many cases to the undersurface of the axis." Although no clinical symptoms were reported, he speculated that "it is not unlikely that bulbar symptoms could be caused." The case he described was that of a 17-year-old young woman who died of typhoid fever. She suffered from hydrocephalus but "no symptoms referable to the cerebel-lum or medulla." This was probably the first case of Chiari I malformation to be described.

Five years later, in a new study,<sup>21</sup> Chiari described 14 cases with Type I changes (Fig. 2). The grade of hydro-cephalus was not related to the severity of the craniospi-nal changes. He supposed that an additional mechanism played a role in this condition—namely, insufficient bone growth and insufficient enlargement of skull parts result-ing in increased ICP.

He described other malformations that are beyond the scope of this presentation; however, we will discuss brief-ly the origin of the nomenclature "Arnold–Chiari malfor-mation" because there was considerable confusion initial-ly when describing the first two types. In the Type II

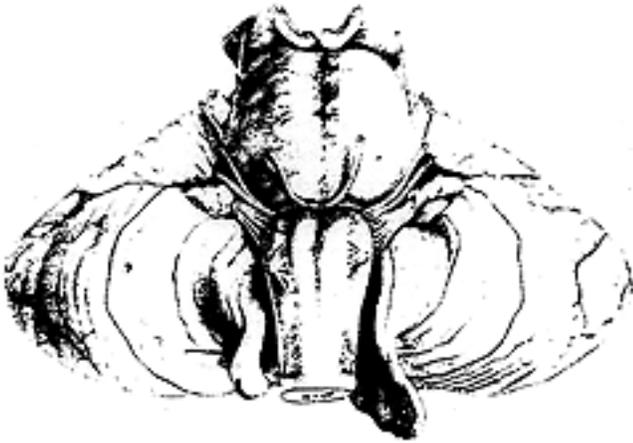


Fig. 1. Illustration of Chiari I malformation in his 1896 publication, showing the tonsillar ectopia. Used with permission from Koehler PJ: Chiari's description of cerebellar ectopia (1891). *J. Neurosurg* 75:823–826, 1991.

anomaly that Chiari described, there were displacements of parts of the inferior vermis, pons, and medulla oblongata as well as elongation of the fourth ventricle into the spinal canal. In the concluding remarks of his 1896 publication, he mentioned a few authors who had already published on this subject, including Cleland and Arnold. In 1883 Cleland<sup>24</sup> reported the case of an infant with spina bifida and hydrocephalus similar to Chiari's type II cases. In 1894 Arnold<sup>7</sup> described an infant with spina bifida in whom there was, elongation of the hindpart of the cerebellum, covering the fourth ventricle and extending into the cervical canal. The observations of these authors, however, were incidental compared with Chiari's thorough study of the malformation. Nonetheless, in 1907, Schwalbe and Gredig,<sup>92</sup> writing from Arnold's lab in Heidelberg, described four cases of myelomeningocele and added Arnold's name to the type II malformation, coining the term Arnold–Chiari malformation. They even referred to the cerebellar malformation as Arnold's deformity and the medullary deformity as Chiari's deformity. Twenty-five years later, in 1932, C. J. Van Houweninge Graftdijk reported the first attempt at surgical correction of this deformity, in his thesis for Doctorate in Medicine entitled "Over hydrocephalus."<sup>88</sup> He intended to relieve the obstruction of CSF flow at the level of the deformity by resecting the tongue of redundant tissue or by resecting the bone over the posterior surface of the malformation and incising the underlying dura. His patients, however, died of the effects of the operation or of postoperative complications.<sup>88</sup>

### INTRODUCTION OF CHIARI MALFORMATIONS INTO THE ENGLISH LITERATURE

The interest in these conditions surged in 1935 after Russell and Donald<sup>88</sup> introduced the notion of Chiari malformations into the English-language literature. They described 10 consecutive infants with Chiari II malformation and used the term Arnold–Chiari malformation



Fig. 2. Title page of Chiari's second paper. Used with permission from De Lotbinière ACJ: Historical considerations, in Anson JA, Benzel EC, Awad IA (eds): Syringomyelia and the Chiari malformations. *Neurosurgical Topics, American Association of Neurological Surgeons*: Chicago 1997, 1–26.

coined by Schwalbe and Gredig.<sup>92</sup> Adult cases of Chiari I malformation were not described until 1938, when McConnell and Parker<sup>64</sup> reported five cases, all with hydrocephalus and neurological symptoms, in whom surgical exploration was performed; an autopsy procedure was performed in three. They used the term "tonsils" to describe the prolapsed cerebellar tissue. The same year, Aring<sup>5</sup> reported a 20-year-old man with an adult Chiari malformation but no hydrocephalus. This was the first reported case of adult Chiari malformation in a patient in whom hydrocephalus was not also present. The condition was diagnosed by cerebellar exploration, and the patient died 18 hours later. The cerebellar tonsils reached down to the axis.

In 1940 Gustafson and Oldberg<sup>46</sup> described another two patients in whom diagnosis was made intraoperatively. In 1941, Walsh, et al.,<sup>106</sup> described another case in which the diagnosis was made intraoperatively, although radiographs had revealed the presence of basilar invagination. Also in 1941, List<sup>60</sup> described two cases: in one patient (Case 5), the presence of an Arnold–Chiari malformation was suspected preoperatively based on the anomalous foramen magnum and the blockage at the foramen magnum observed on ventriculography; in the patient in Case

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6, however, the neurological picture was “interpreted as being due to an Arnold Chiari deformity rather than to bony compression” based on blockage demonstrated on myelography. This case was the first report of myelographic diagnosis of Chiari malformation. The same year Adams, et al.,<sup>2</sup> described another case in which preoperative myelography demonstrated a block at the level of C-3: they assumed the lesion was a tumor of the cervical cord, but on surgical exploration a diagnosis of a Chiari malformation was made. Adams, et al., delineated the clinical syndrome and classified the symptoms into five groups: increased ICP, involvement of several of the cranial nerves, compression of the brainstem, compression of the spinal cord, and cerebellar signs, and they described the myelographic appearance of the protruding cerebellar tonsils.

Other reports followed, which were mostly isolated case reports and some small series;<sup>3,18,20,43,45,51,58,61,72,95,96</sup> however, it was during the 1970s that the term adult Chiari malformation gained popularity<sup>4,11,33,63,69,85,89</sup> and large series were published. This trend increased considerably over the following two decades.<sup>8,15,19,32,34,35,44,47,54,59,68,78,81,90,98</sup>

### PATHOLOGICAL CLASSIFICATION

In 1949 Russell<sup>87</sup> suggested that “it would appear preferable to restrict the use of the term Arnold Chiari malformation to those cases in which the spina bifida type of abnormality is identified” or Chiari II malformation, rather than using it for “any caudal prolongation of the medulla oblongata or of the cerebellum including tonsillar herniation associated with cerebral tumors.” Later, she “questioned the existence of a genuine Arnold–Chiari malformation in the absence of spina bifida.”<sup>86</sup> This distinction was reemphasized by Peach<sup>79</sup> in 1965 and became widespread over the following decade.

Debate on the classification of the tonsillar herniations was undertaken more recently in the pathology literature. In 1976, Friede and Roessmann<sup>42</sup> reappraised the classification of the Chiari I malformation after reviewing all reported cases of chronic tonsillar herniations at the foramen magnum in the absence of intracranial space-occupying lesions. They believed that “the uncritical propagation of the term ‘malformation’ for Chiari type I lesions has done much harm to obfuscate the issue of chronic tonsillar herniations in the absence of space-occupying supratentorial lesions.” Chiari’s descriptions in 1891 and 1896 addressed cerebellar lesions in infants with hydrocephalus. Two concepts were not known at that time: the relation between the flow of CSF and hydrocephalus and the concept of tonsillar herniation induced by increased ICP. It was only during the following decade that the latter concept had been reported by Cushing<sup>28</sup> and Collier,<sup>27</sup> and it did not become widely known until Meyer’s report<sup>66</sup> in 1920 on the various herniations, including the subfalcine and transtentorial herniations. According to Friede,<sup>41</sup> Chiari’s description of the type I malformation referred mostly to cerebellar pressure cones. Sclerosis of the herniated tonsils might have been the result of the protracted path to death that was common in those days in infantile hydrocephalus. Friede and Roessmann<sup>42</sup> distinguished between two types of cerebellar herniations: a chronic herniation of the cerebellar tonsils in the absence of space-occupying

supratentorial lesions and an adult Arnold–Chiari malformation in which at least one hindbrain deformity is associated with tonsillar herniation. The hindbrain deformities are similar to some of those encountered in the infantile form and would include medullary deformities comparable with those found in the infantile form (typical hump, beak, or kinking of its dorsal portion at the cervicomedullary junction) and an upward course of the cervical rootlets. In contrast to the infantile form, however, the cerebellar tonsils, but not the vermis, herniate through the foramen magnum, although the most caudal portion of the fourth ventricle, particularly the choroid plexus, may herniate, and there is no associated myelomeningocele and spina bifida. Friede and Roessmann supported their proposition by reviewing all the cases reported in the literature for which there was appropriate documentation as well as 13 of their own cases. Twenty-five cases of adult Arnold–Chiari malformation and 39 cases of chronic tonsillar herniation were found.<sup>13,25,26,29,38,46,57,75,83,91,99,100,102</sup> It is worth mentioning that both groups had associated osseous anomalies and syringomyelia, indicating a possible difference in degree only between the groups. This issue remains unanswered, however.

### *A Small Posterior Fossa, Craniocephalic Disproportion, and Tonsillar Ectopia*

Perhaps tonsillar herniation can be viewed as a craniocephalic disproportion or disproportion between the container (skull) and the contents. This is confirmed by examples of acquired tonsillar descent. Acquired tonsillar ectopia is seen in various conditions; in some cases the volume of the cranial cavity is reduced, and in others the volume of the intracranial contents is increased. Disorders associated with the former condition include severe craniosynostosis<sup>23</sup> and lesions that cause calvarial thickening such as Paget disease,<sup>36</sup> rickets,<sup>56</sup> and erythroid hyperplasia.<sup>84</sup> Disorders that cause an increase in the volume of the intracranial contents and induce tonsillar descent are associated with supratentorial tumors<sup>77,94</sup> and infratentorial mass lesions.<sup>93,103</sup> The tonsillar herniation resolved after tumor resection. Acute tonsillar herniation is a common finding in cases of rapidly enlarging intracranial processes such as acute brain edema, intracerebral hemorrhage and acute hydrocephalus. Mikulis, et al.,<sup>67</sup> have shown that with increasing patient age there is tonsillar ascent linked to neuronal dropout, volume loss, and brain atrophy.

Similarly, in patients with adult Chiari malformation, recent studies have revealed the presence of a small posterior fossa. Using the posterior fossa ratio method, Nyland and Krogness<sup>73</sup> found that the posterior fossa was small in five patients with adult Chiari malformation. Based on examination of lateral skull radiographs, Stovner, et al.,<sup>97</sup> found that the posterior fossa was smaller and shallower in patients with Chiari I malformation than in controls. Badie, et al.,<sup>8</sup> compared the ratio of the posterior fossa with supratentorial volumes on MR images obtained in 20 patients with Chiari I malformation and 20 control patients. The ratio was smaller in the former patients, and those with smaller posterior fossa developed symptoms earlier and were more likely to respond to decompressive surgery. Nishikawa, et al.,<sup>71</sup> found a higher volume ratio of the posterior fossa brain compared with that of the posterior fossa cranium. Milhorat, et al.,<sup>68</sup> have also reported

similar findings, and they concluded that Chiari I malformation is a disorder of the mesoderm leading to a volumetrically small posterior fossa that predisposes patients to hindbrain overcrowding. Experimentally-induced small posterior fossa was also found to lead to tonsillar herniation.<sup>62</sup>

### **RADIOLOGICAL CRITERIA FOR CHIARI MALFORMATION: THE PRE-MR IMAGING ERA**

Attempts at finding radiological criteria for the Chiari malformations were published mainly in the radiology literature.

Perhaps Baker<sup>9</sup> was the first to attempt defining criteria for the extent of tonsillar ectopia needed for a diagnosis of the Chiari malformation. He considered, by definition, the cerebellar tonsils to lie in a normal position if the lower border was above a line joining the posterior rim of the foramen magnum and the tip of the clivus on lateral view. He described 28 cases of developmental anomalies found in a pool of 179 patients in whom symptoms and signs were referable to the upper cervical posterior fossa region and who underwent myelography. Of three patients with marked constriction of the foramen magnum, two underwent decompressive surgery and their neurological status improved postoperatively. Herniation of the tonsils down to the arch of C-1 was present in 11 patients, in whom Arnold-Chiari malformation was diagnosed. In the remaining 14 patients myelography demonstrated normal findings except that the cerebellar tonsils were located 2 to 5 mm below the foramen magnum. The author wrote, "Since there was no obstruction to the free flow of fluid through the foramen magnum it was difficult to justify operative decompression.... However, three such patients with severe intermittent symptoms, which were thought to be due to a ball-valve type of obstruction, were surgically treated." Baker concluded that "these cases represent a mild form of Arnold-Chiari malformation [Chiari type I] and that further observation is necessary to establish the clinical significance of the low grade tonsillar herniation."<sup>9</sup>

In 1973 O'Connor, et al.,<sup>74</sup> emphasized the "need to establish the normal location of the tonsils...so that an abnormally low position may be recognized." They studied 100 myelograms, excluding cases of foramen magnum or intracranial mass space-occupying lesions. These studies were obtained in patients with various conditions including disseminated sclerosis, neck or back pain of unknown origin, and motor neuron disease. The tonsils were always above the basion-opisthion line. The authors concluded that "demonstration of the tonsils at the level of and through the foramen magnum is a strong evidence of a Chiari malformation," but they cautioned that "coincidental disease of the central nervous system [with] demonstration of tonsillar herniation..." could confuse the diagnosis.

In 1974 Bloch, et al.,<sup>16</sup> measured the distance of the tonsillar tip from the upper lip of the foramen magnum on the anteroposterior view of myelograms obtained in 60 normal volunteers and 19 patients with Arnold-Chiari malformation. The tonsils ranged from 7 mm above to 8 mm below the upper lip in the normal cases, and from 3 mm above to 25 mm below the inner rim in patients with Chi-

ari malformation. Magnification was not accounted for. They concluded that marked tonsillar herniation can be encountered as an incidental finding.

Computerized tomography with or without injection of intrathecal metrizamide has been used to establish a diagnosis of the Chiari malformation, but there have been no attempts to find definite criteria by which to diagnose adult Chiari malformation.<sup>17,31,39,40,49,70</sup>

### **RADIOLOGICAL CRITERIA FOR ADULT CHIARI MALFORMATION: THE MR IMAGING ERA**

In 1985, Aboulezz, et al.,<sup>1</sup> used MR imaging to study the position of the cerebellar tonsils in the normal population (82 individuals) and in 13 patients with Chiari malformations (11 with type I and two with type II). In the normal population the position of the tonsils varied from 2.8 mm below the foramen magnum to 20 mm above, whereas in patients with Chiari malformations the tonsils were located 5.2 to 17.7 mm below the foramen magnum and in all cases the tonsils were pointed. The authors concluded that the tonsils may extend up to 3 mm below the foramen magnum in the normal population, and in patients with Chiari malformation they extend 5 mm or greater below the foramen magnum.

The following year Barkovich, et al.,<sup>12</sup> compared the MR images obtained in 200 patients with clinical symptoms unrelated to the Chiari I malformation or other cervicocranial anomalies with those obtained in 25 patients with the Chiari I malformation. Patients for whom imaging studies could not clearly be classified into one of these categories were excluded from the study, raising the possibility that some minimally symptomatic or as yet asymptomatic patients were missed. The tonsils were located from 8 mm above to 5 mm below the foramen magnum in the controls and from 3 to 29 mm below the foramen magnum in patients with Chiari malformation. Peglike tonsils were found in both groups. Narrowing or complete effacement of the CSF spaces of the foramen magnum and cisterna magna were documented in all patients with more than 1 mm tonsillar ectopia. The authors concluded that, in terms of the most accurate diagnosis, a herniation 2 mm below the foramen magnum should be the cutoff because no symptomatic patient had less than 3-mm herniation and only one asymptomatic patient had more than 3-mm ectopia.

In 1988 Ishikawa, et al.,<sup>52</sup> studied 50 control patients and found that the cerebellar tonsils were always located above the line of the foramen magnum.

In 1992 Elster and Chen<sup>37</sup> found that Chiari I malformation was documented in 0.56% of patients who underwent MR imaging in a tertiary care center and in another 0.16% this diagnosis was also possible. A 5-mm cutoff for one tonsil, or 3 to 5 mm cutoff for both tonsils was used as the diagnostic criterion. A female preponderance (2:1) was reported for the first time. In 12% only subjective findings were demonstrated, and 31% (0.17% of the total population examined) were asymptomatic (ectopia from 5–12 mm). The authors concluded that a "careful clinical assessment remains the cornerstone for proper diagnosis and management."

The same year Mikulis, et al.,<sup>67</sup> studied how tonsillar

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position varies with patient age in 220 patients (age range 5 months–89 years). They found that the cerebellar tonsils ascend with increasing age and suggested the following criteria for ectopia: first decade, 6 mm below the foramen magnum; second to third decades, 5 mm; fourth to eighth decades, 4 mm; and ninth decade, 3 mm below the foramen magnum. These criteria were based on the distance greater than two standard deviations from the normal range for each decade.

In 1998 Iskandar, et al.,<sup>53</sup> described five children with syringohydromyelia in whom clinical and radiological resolution were demonstrated after posterior fossa decompression. None of the patients had hindbrain herniation. Their course was similar to that of patients with Chiari malformation. In cases in which preoperative cine-MR imaging was performed, decreased or normal-to-decreased flow was demonstrated. On review of the published MR images, the cerebellar tonsils were found at the level of the foramen magnum (Fig. 3). Three years later, Tubbs, et al.,<sup>105</sup> added a sixth case and analyzed various radiological indices, finding that the contents of the posterior fossa were indeed compromised in these patients, even in the absence of tonsillar ectopia, and that the obices were located more than two standard deviations below normal. “Chiari zero malformation” was the name they chose to characterize the disorder.

In their seminal paper of 1999, Milhorat, et al.,<sup>68</sup> studied 364 symptomatic patients with Chiari I malformation. They found that in 32 (9%) of the 364 symptomatic patients tonsillar ectopia of less than 5 mm was demonstrated, but compression of the CSF spaces posterior and lateral to the cerebellum was present in all cases. In fact the tonsils were at the level of the foramen magnum in the illustrative case they presented (Fig. 4).

In 2000 Meadows, et al.,<sup>65</sup> published a review of 22,591 patients in whom MR imaging was performed. Tonsillar herniation extending more than 5 mm below the foramen magnum was found in 0.77%. Fourteen percent of those patients were asymptomatic (with 7–25 mm of ectopia) and 25% of those had peglike tonsils. The authors concluded that “isolated tonsillar herniation is of limited



Fig. 4. The illustrative case of adult Chiari malformation reported by Milhorat, et al., in which the tonsillar herniation extended less than 5 mm below the foramen magnum. In fact the tonsils are at the level of the foramen magnum. The patient had typical Chiari symptoms that resolved immediately postoperatively. *Neurosurgery* 44:1005–1017, 1999.

prognostic utility, and should be considered in the context of all available clinical and imaging data....”.

Based on the review of all these papers the following remarks can be made. 1) Radiologically significant tonsillar ectopia may be completely asymptomatic. 2) There are patients in whom the tonsillar herniation extends less than 5 mm from the foramen magnum, whose clinical behavior is similar to the remainder of the Chiari population, including even those with radiological evidence of syringohydromyelia.

These findings concur with the conclusion of Meadows, et al.,<sup>65</sup> in that the radiological criteria for the extent of tonsillar herniation are not absolute and should be considered within the clinicopathological context.

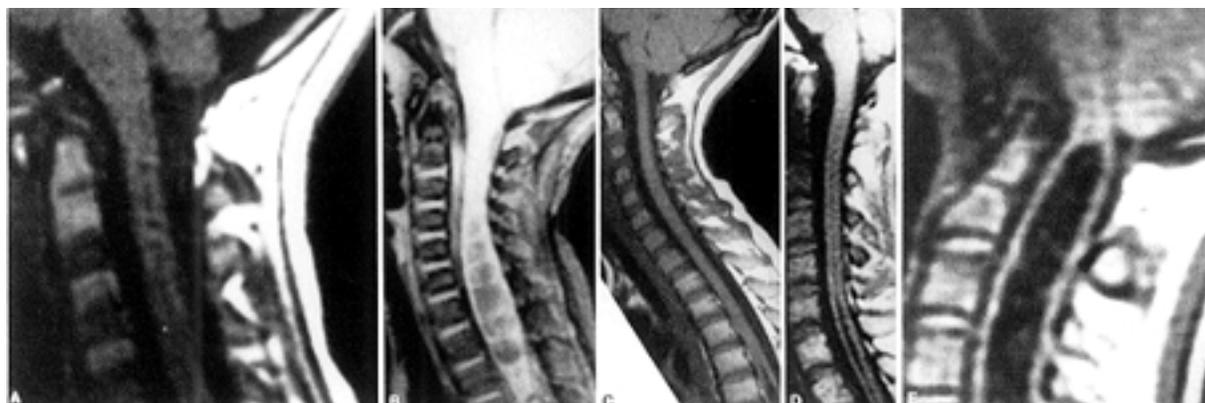


Fig. 3. Magnetic resonance images obtained in the five cases of Chiari zero malformations reported by Iskandar, et al. The tonsils are at the level of the foramen magnum. All five patients had syringomyelia that resolved after posterior fossa decompression. Used with permission from Iskandar, et al.:The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. *J Neurosurg* 89:212–216, 1998.

## DYNAMIC MR IMAGING STUDIES OF ADULT CHIARI MALFORMATION

The use of dynamic MR imaging studies to diagnose CSF flow obstruction in patients with Chiari malformation was first reported in 1991 by Tominaga, et al.<sup>104</sup> They described CSF flow obstruction at the level of the foramen magnum in a patient with basilar impression accompanied by Chiari malformation and its resolution after transoral decompressive surgery. In 1994 Oldfield, et al.,<sup>76</sup> reported three patients with Chiari malformation in whom cine-MR studies were performed. Later the same year Armonda, et al.,<sup>6</sup> reported the first series of patients with Chiari malformation who underwent cine-MR imaging. Other studies followed in which the authors described the characteristics of the Chiari malformation on dynamic MR imaging;<sup>14,48,50,82,101,107</sup> however, there was no attempt to define criteria that would clearly delineate the normal control individuals from symptomatic patients.

### CONCLUSIONS

Perhaps it is the opening statement written by Ball and Crone<sup>10</sup> in their editorial that seems appropriate as a concluding remark: "Ever since the initial postmortem description by Chiari in 1891 of the group of malformations that bears his name, it seems there have always been more questions on this subject than answers." As we have seen, the definition of the adult Chiari malformation has varied with the evolution of our neurodiagnostic capabilities and knowledge of physiopathology. There is no single test that allows a clear-cut distinction between clinically significant tonsillar ectopia and incidental tonsillar descent. With the fluidity of the definition of the adult Chiari malformation, as well as the increasing number of asymptomatic patients with significant radiological tonsillar herniation, it is the clinical judgment of the physicians evaluating this disorder that is of the utmost importance to avoid the therapeutic extremes of pursuing unnecessary surgery or withholding necessary treatment from patients.

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