Child's Nerv Syst (1992) 8:86–91

Child's Nervous System CNS

Anatomical progression of the Chiari II malformation

John R. Ruge [1], Jeff Masciopinto [2], Bruce B. Storrs [3], and David G. McLone [3]

[1] Lutheran General Children's Medical Center and Children's Memorial Hospital, Chicago, Illinois, USA
[2] Rush Presbyterian-St. Luke's Medical Center, Chicago, Illinois, USA
[3] Children's Memorial Hospital, Chicago, Illinois, USA

Received June 12, 1991

Abstract. To evaluate whether anatomic change of the relationship of the Chiari II malformation and the cranial base was occurring, 22 children with meningomyelocele had serial MRI scans reviewed. A ratio (B/A) was established between the distance from the foramen magnum to the caudalmost portion of herniated cerebellum (B) and the diameter of the foramen magnum (A) and this ratio was compared on serial MRI scans. Eighteen children had an increase in the B/A ratio, two children had a decrease, and two had no change. This indicates that continuous anatomic change of the Chiari II malformation and the skull base is occurring. Clinical deterioration in the older child may be explained by a combination of compressive and traction forces due to this change.

Key words: Chiari II – Meningomyelocele – Posterior fossa – Hindbrain dysfunction – Skull base – Magnetic resonance imaging

Hindbrain dysfunction remains the most frequent cause of mortality in children with meningomyelocele (MM) [30]. All children with MM have MRI evidence of the Chiari II malformation. In the Chicago Children's series, a 32% morbidity and an 11% mortality rate could be attributed to hindbrain dysfunction. Hindbrain dysfunction was responsible for 73% of all deaths of children with MM [30].

The Chiari II malformation was first reported by Cleland [12] in 1883 and then described by Chiari [9, 10] in 1891. Since that time many theories have sought to explain why the Chiari II malformation becomes symptomatic [3, 6, 7, 10–12, 21, 26, 29, 32, 41–43, 47, 52]. Theories to explain the pathophysiology must take into account that not all the children become symptomatic, that once they do develop symptoms there can be rapid progressive deterioration, that this deteriorating course can sometimes be reversed by a decompressive procedure, and that the neonate appears to have a different clinical course than the older child with MM [4, 18, 51]. It is currently unclear why an older child with MM and a functional CSF shunt should suddenly deteriorate from symptoms of a congenital hindbrain malformation. This study was performed to address this question and assess whether and to what extent the Chiari II malformation is of a static or a dynamic nature in the older child.

Offprint requests to: J. R. Ruge, Lutheran General Children's Medical Center, 1875 Dempster Street, Suite #605, Park Ridge, IL 60068, USA

Materials and methods

Twenty-two children were selected from 400 patients with MM repaired at Children's Memorial Hospital between June 1979 and December 1989 and represent the study cohort. The selection criteria were for all children who had two or more MRI scans performed that included their posterior fossa and cervical spine. The initial and final MRI scans ($n=44$) were reviewed and compared. The average interval between MRI studies was 25.8 months (range 11 months to 59 months). The average age of the child at first study was 4.9 years (range 7 months to 11.25 years).

The MRI studies were obtained for a variety of indications not necessarily related to hindbrain dysfunction. The children had MRI studies performed after a change of either diminished strength or increased tone was noted in their motor examination. Six children had either lower cranial nerve difficulties, neck pain, or respiratory difficulties. All subjects had functional shunts. This is supported by the fact that only two children had a shunt revision, 1 month and 3 months, respectively, after their last MRI study.

MRI studies were reviewed blinded for date independently by two of the authors (J. M., J. R.). The distance (B) from the foramen magnum to the caudalmost tip of the herniated vermian peg was determined on the midsagittal T_1-weighted MR image. A line was drawn across the foramen magnum, then a perpendicular line was drawn from the foramen magnum to the tip of the vermian peg and the distance (B) measured. The anteroposterior diameter (A) of the foramen magnum was then measured (McRae's line) on the same midsagittal MR image (Fig. 1). The growth rate of the foramen magnum of normal children has previously been published [5, 46]. The ratio B/A was calculated and compared on serial studies.

On those scans with accurate scales the anteroposterior diameter of the foramen magnum was measured at different ages and compared to published norms.

Results

The anteroposterior diameter of the foramen magnum was for the most part larger than published norms but followed similar growth curves (Fig. 2). The age of patients, interval between MRI scans, and ratio changes are shown in Table 1.

The ratio B/A increased in 18 children, showed no change in two, and decreased in two. The monthly interval change was similar for those children who had an increase in the B/A ratio (Fig. 1).

Three children in our series had a decompressive procedure performed in the interval between MRI studies. Of these three children, the B/A ratio change increased in one, decreased in one, and remained the same in one.

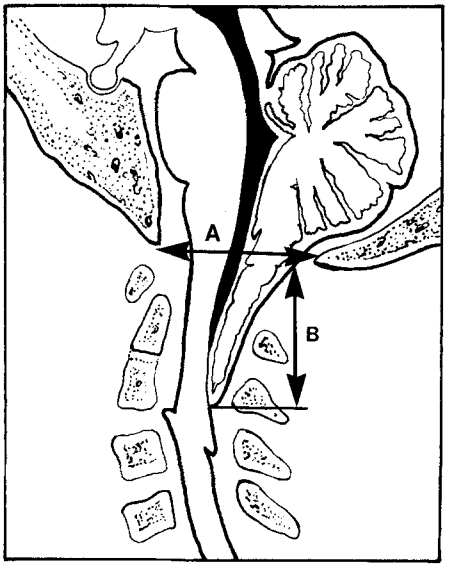

Fig. 1. Ratio B/A established. *B*, Distance from foramen magnum (McRae's line) to vermian peg; *A*, anteroposterior diameter of foramen magnum

Discussion

The Chiari II malformation is a congenital malformation consisting of multiple CNS abnormalities [2, 7–12, 15, 21, 28, 41, 42, 47, 52]. These are characterized by caudal displacement of the brainstem, IV ventricle and cerebellar vermis through an enlarged foramen magnum into the cervical spinal canal with medullary kinking [35, 36, 38]. Cephalad herniation of the cerebellum likewise occurs through an enlarged tentorial incisura [36]. The tectal plate is often "beaked" and the aqueduct of Sylvius is sometimes "forked" [36, 42]. The skull base is malformed in that the petrous bone is scalloped and the basiocciput is concave. The entire posterior fossa is small with the insertion of the tentorium occurring low, often at the foramen magnum [13, 22, 25, 27, 35, 55].

In addition to anatomically displaced CNS structures, much of the CNS is malformed [40–42, 48]. The folia and cytoarchitecture of the cerebellum are abnormal. Brainstem nuclei may be absent or hypoplastic. The thalamic massa intermedia is often large. Polymicrogyria and partial or complete agenesis of the corpus callosum are often present [37].

The neonate has a different clinical course than the infant and older child [17, 51] and usually has the most severe symptoms of these three age groups. The neonate often presents within a week of birth with severe respiratory stridor, apnea, facial nerve paresis, lack of gag reflex, and quadriparesis [1, 17, 19, 20, 23, 24, 39, 44, 49, 50]. There is often a rapid clinical deterioration which may be unaffected by a decompressive procedure. It is speculated on the basis of sparse autopsy studies that these children have dysplastic brainstem nuclei, which would not be

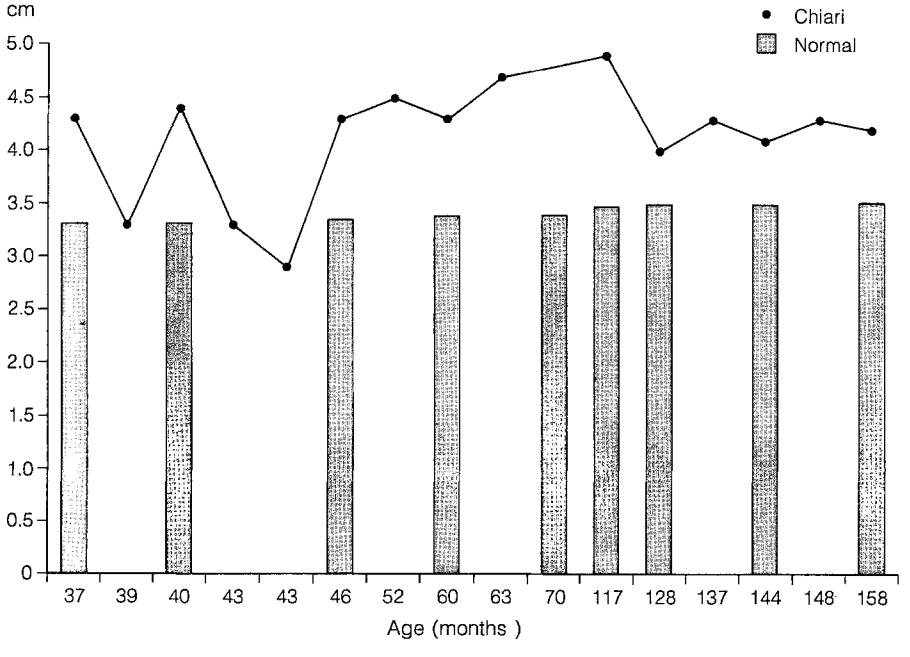

Fig. 2. Comparison of measured foramen magnum diameters with previously published norms [47]

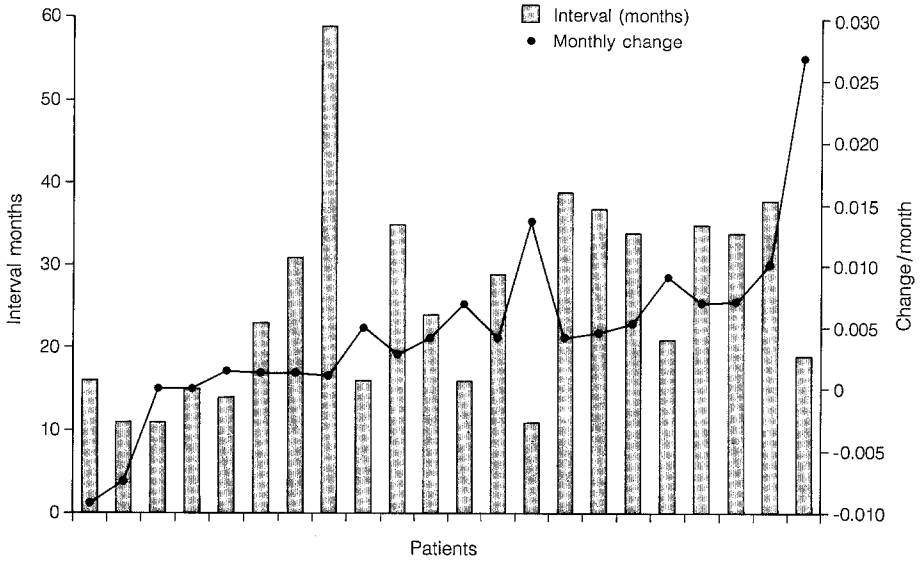

Fig. 3. Monthly interval change (total change/interval) in ratio of B/A compared to interval between MRI scans

Table 1. Patient data

Case no.	Age at first MRI	Interval between MRIs	B/A ratios		Ratio change
			1st MRI	2nd MRI	
1	8 years 1 month	21	0.50	0.69	0.19
2	2 years	19	1.43	1.94	0.51
3	4 years 3 months	31	0.92	0.96	0.04
4	7 years	24	0.40	0.50	0.10
5	3 years	37	0.40	0.57	0.17
6	3 years	34	0.57	0.75	0.18
7	5 years 3 months	35	0.34	0.58	0.24
8	2 years 7 months	16	0.75	0.86	0.11
9	10 years 4 months	35	0.79	0.88	0.09
10	7 years 2 months	29	1.09	1.20	0.11
11	3 years 3 months	23	0.92	1.00	0.08
12	2 years 4 months	38	0.40	0.79	0.39
13	10 years 8 months	16	0.24	0.32	0.08
14	3 years 7 months	39	0.81	1.00	0.19
15	9 years	11	0.63	0.63	0.00
16	11 years 3 months	11	0.47	0.62	0.15
17	1 year 6 months	34	0.63	0.87	0.24
18	2 years 4 months	11	1.02	0.94	−0.08
19	6 years 2 months	16	0.41	0.26	−0.15
20	7 months	14	0.62	0.64	0.02
21	4 years	15	0.52	0.52	0.00
22	16 months	59	0.85	0.91	0.06

affected by surgical decompression [20, 48]. However, Pollack and Pang (unpublished data) have reported dramatic improvement of the neonate with surgical decompression.

Clinical series support a difference in outcome between the older child and neonate. Pollack and Pang (unpublished data) reported on 22 children symptomatic from Chiari II malformation. In their study the preoperative neurologic condition seemed to be a more important predictor of postoperative outcome than age. Although the neonate and infant did worse than the older child, they also presented in worse condition. The neonate faired as well as the older child with the same preoperative condition. However, although this study seems to indicate little difference between the neonate and the older child, others more strongly suggest a difference. Hoffman et al. [18] reported in 1975 on 16 patients who were operated on for symptomatic Chiari II malformation. Of the eight infants studied, five recovered completely, one died, and two developed meningitis, while of the eight older children, seven made a full recovery and only one died of meningitis. Bell et al. [4] reported on 19 children with symptomatic Chiari II malformation who underwent surgical decompression. Seventy-one percent (10/14) of the infants who underwent decompression were symptomatic postoperatively, compared to 0% (0/5) of the older children treated similarly. Venes et al. [51] reported on a series of 14 children who underwent surgical decompression; of these, ten improved, two had mild residual symptoms, and five were unchanged. The study showed no difference in outcome between ages.

Older children often present with swallowing difficulties, neck pain, nystagmus (and other ocular motility problems), increased spasticity, and motor weakness [30]. They can also present with vocal cord paralysis, respiratory stridor, and aspiration pneumonia, but this is more typical of the infant [19, 20].

Several theories have attempted to explain the pathophysiology. One theory suggests a vascular mechanism. Morley [33] reported two cases of children who at autopsy were found to have medullary hemorrhages. He speculated that the abnormal anatomy might render the brainstem susceptible to ischemia and that venous congestion may play a role. Brainstem and cerebellar infarcts have been reported by Cameron [7] in 1957. It has been shown by Emery and Levick [14] on post-mortem angiographic studies that there is an abnormal descent and looping of the posterior inferior cerebellar artery which may render it susceptible to compressive injury.

Congenital malformation of the brainstem nuclei may account for their dysfunction. There has been a report of an inherited bilateral vocal cord paralysis [16, 45, 53] and of a mother and son with Charcot-Marie-Tooth disease with bilateral vocal cord paralysis [20].

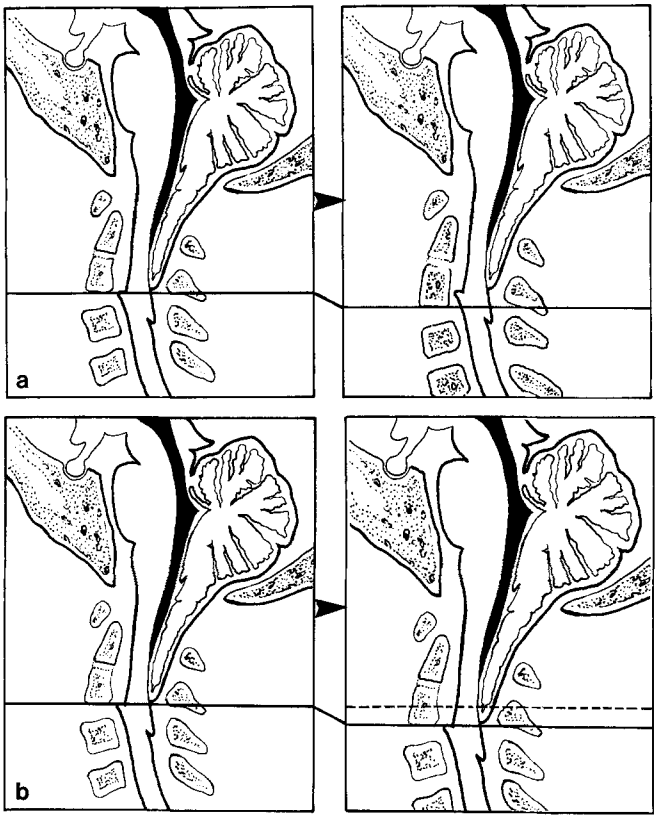

Fig. 4 a, b. Rationale for using a ratio rather than direct measurements or comparison to neighboring spine. **a** Assuming no descent of cerebellum but continued growth of spine, vermian peg appears shorter when compared to C-2. **b** Assuming equal growth of spine and descent of cerebellum, vermian peg appears unchanged in relationship to C-2 despite significant caudal descent of cerebellum

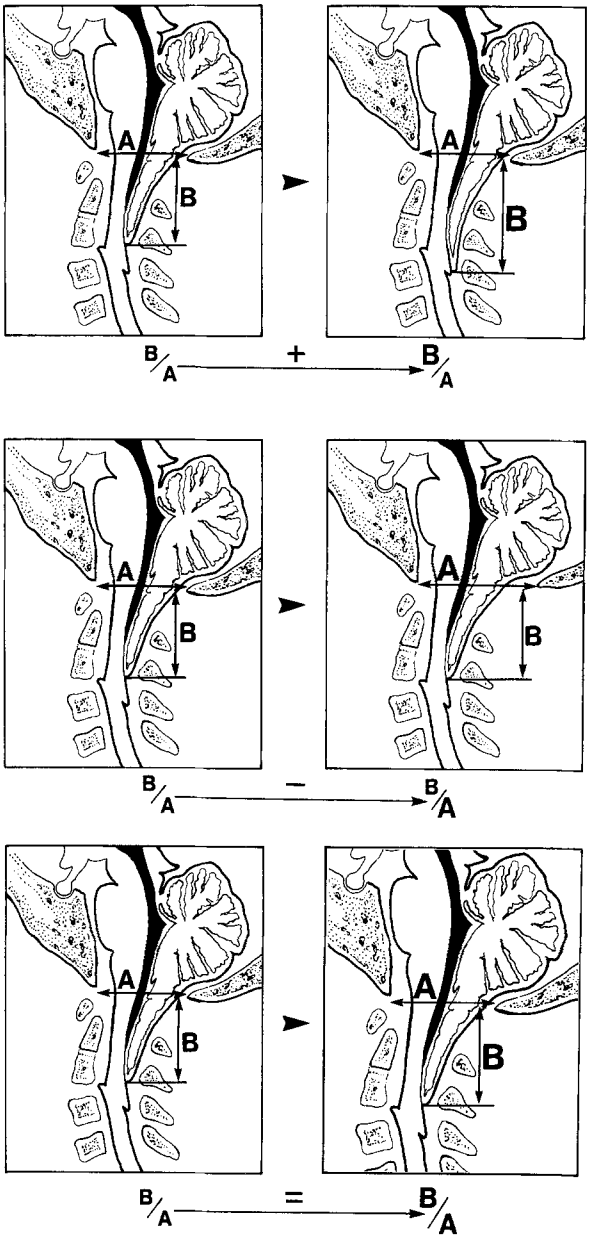

Fig. 5. Interpretation of changes in the ratio B/A. A "+" change necessarily means that B has increased since there is no evidence to suggest A decreasing. A "−" change means either B has decreased or A has increased. No change in ratio implies either no change in the absolute measurements of A, B or equivalent changes

Vascular injury and congenital malformation theories may help to explain the occurrence of the neonate who is born with severe hindbrain dysfunction and who continues to deteriorate despite surgical intervention, but they fail to explain the late onset of symptoms in many children and teenagers. They also fail to account for the reversibility of the symptoms after a surgical decompression.

Direct traction on the lower cranial nerves as they course upward through the foramen magnum has also been implicated in the pathophysiology [48]. Because of the herniated hindbrain contents and crowded foramen magnum, the lower cranial nerves are at increased risk of compressive injury or traction injury. This may account for the autopsy findings of cranial nerve nuclei hypoplasia. Traction injury to the cranial nerves is an attractive theory because of the reversibility of the injury. Whereas one would not expect an infarcted brainstem to recover, a mild compression or traction injury to the cranial nerves would be expected to recover once the tension was relieved. Traction may account for the lower cranial nerve involvement but does not explain the myelopathic findings or central apnea.

It is the general experience that hydrocephalus may cause many of these symptoms and that once this is corrected, hindbrain symptoms improve [30, 31]. The presumed mechanism of this is of temporary distention either of a dilated IV ventricle or syrinx, or caudal displacement of the hindbrain structures from non-communicating hydrocephalus.

Direct compression of the hindbrain at the foramen magnum and cervical canal causing neural dysfunction is the most plausible theory. Wickramasinghe et al. [54], in 1968, first reported improvement after surgical decompression in a subgroup of these children. Since then, others [4, 18, 30, 51] have confirmed his findings. Mullan and Raimondi [34] reported that at surgery flexion of the head of a patient who has the Chiari II malformation can cause apnea, which responds to repositioning, presumably because of hindbrain compression. Just as the neural deficits caused by CNS compression from intracranial mass lesions are often reversible if decompressed early, so

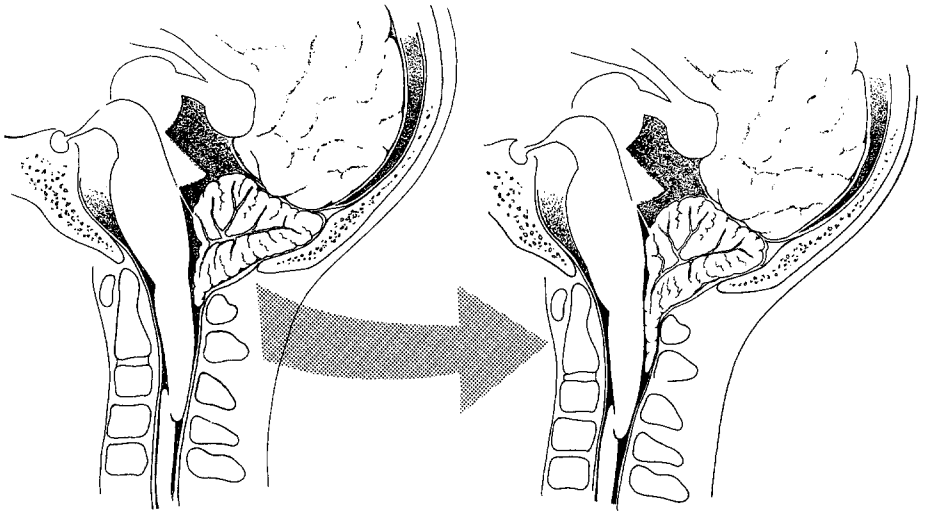

Fig. 6. Continual change in the relationship of the CNS to the cranial base may account for delayed neurologic deterioration in the older child with Chiari II malformation

likewise the deficits from hindbrain compression often reverse after a decompressive procedure. However, this theory fails to explain why this compression should develop at variable times, often years after birth.

Findings of arachnoidal adhesions and sometimes "open" CSF spaces at surgery have led some to speculate that symptomatology may be secondary to a cervical "tethering" mechanism. Just as releasing a tethered spinal cord improves symptoms, cervical decompression may be accomplishing a similar result (Rekate, personal communication, 1989).

Our results indicate that even in the older child, anatomical changes are occurring in the relationship of the vermian peg to the diameter of the foramen magnum, which could explain the late onset of hindbrain compression. The ratio B/A was established for several reasons (Fig. 4). First, an internal control was necessary to eliminate the variability between scans performed on different machines with different scales and to increase the accuracy of measurements, since scale factors were unnecessary. Secondly, simply comparing the herniated vermian peg to neighboring bone structures requires many assumptions. The spine would have to not change between studies, or change at a different rate between studies, in order to make any change in the herniated vermian peg noticeable, if one were using the spine as the measure of comparison. Thirdly, the child's brain and skull are rapidly developing and growing. An absolute measurement may simply be reflecting normal growth – not pathophysiology.

In this study, 18 of 22 children had an increase in the ratio B/A between serial MRI scans. An increase in B/A means that either A (foramen magnum) got smaller or that B (herniated cerebellar peg) increased. We were able to show that the foramen magnum in these children starts larger than published norms, but grows at approximately the same rate. The only other possibility is that B increased, meaning that the vermian peg descended between studies.

In two children there was no change in the ratio – which may indicate no change in growth or that the vermian peg descended at the same rate of growth as the foramen magnum diameter change. In two children the ratio got smaller – which probably means the vermian peg did not change whereas the foramen magnum continued to grow (Fig. 5).

This study cohort was not necessarily symptomatic from hindbrain dysfunction. Some underwent release of their tethered cord between or after the MRI scans. Some simply improved spontaneously. In one child a shunt revision was performed 1 month after the last MRI scan and at 3 months in another child. This gives credence to the assumption that the changes observed in the B/A ratio did not reflect a shunt malfunction.

Hindbrain dysfunction secondary to the Chiari II malformation occurs as the result of direct compression of the hindbrain structures at a variable age during development. Our study supports the theory that there is a continual change in the relationship of the hindbrain to the cranial base (Fig. 6). Individual characteristics of the child's cranial base and cervical canal along with other factors such as CSF dynamics would dictate when the child would become symptomatic. Surgical decompression is most effective if performed early. Delay may lead to irreversible hindbrain injury, perhaps on a vascular basis.

Most of the children in our experience who are symptomatic from hindbrain dysfunction are evaluated with a single MRI and then operated upon and are thus excluded from the present analysis. Further study is necessary to evaluate whether the child clearly symptomatic from hindbrain dysfunction has a more dramatic change in the ratio and whether this ratio change can be altered with surgical decompression.

References

1. Adeloye A, Singh SP, Odeku EL (1970) Stridor, myelomeningocele, and hydrocephalus in a child. Arch Neurol 23:271
2. Arnold J (1894) Myelocyste, Transposition von Gewebskeimen und Symbodie. Beitr Pathol Anat 16:1
3. Barry A, Patten BM, Steward BH (1957) Possible factors in the development of the Arnold-Chiari malformation. J Neurosurg 14:285
4. Bell WO, Charney EB, Bruce DA, Sutton LN, Schut L (1987) Symptomatic Arnold-Chiari malformation: review of experience with 22 cases. J Neurosurg 66:812–816

5. Bliesener JA, Schmidt LR (1980) Normal and pathological growth of the foramen occipitale magnum shown in the plain radiograph. Pediatr Radiol 10:65–69
6. Bloch S, Rensburg MJ van, Danziger J (1974) The Arnold-Chiari malformation. Clin Radiol 25:335–341
7. Cameron AH (1957) The Arnold-Chiari and other neuroanatomical malformations associated with spina bifida. J Pathol Bact 73:195
8. Cameron AH, Osman Hill WC (1955) The Arnold-Chiari malformation in a sacred baboon (*Papio hamadryas*). J Pathol Bact 70:552
9. Chiari H (1891) Über Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von kongenitaler Hydrocephalie des Großhirns. Dtsch Med Wochenschr 27:1172
10. Chiari H (1891) Über Veränderungen des Kleinhirns in Folge von Hydrocephalie des Großhirns. Dtsch Med Wochenschr 27:1172
11. Chiari H (1896) Über Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von kongenitaler Hydrocephalie des Großhirns. Denkschr Akad Wiss Wien 63:71
12. Cleland (1883) Contribution to the study of spina bifida, encephalocele, and anencephalus. J Anat Physiol 17:257
13. Davies HW (1967) Radiological changes associated with Arnold-Chiari malformation. Br J Radiol 40:262–269
14. Emery JL, Levick R (1966) Movement of brainstem and vessels around the brainstem in children with hydrocephalus and Arnold-Chiari deformity. Dev Med Child Neurol Suppl 11:49
15. Frazer JE (1931) A manual of embryology. Baillière, Tindall and Cox, London
16. Gacek RR (1976) Hereditary abductor vocal cord paralysis. Ann Otol Rhinol Laryngol 85:90
17. Graham MD (1963) Bilateral vocal cord paralysis associated with meningomyelocele and the Arnold-Chiari malformation. Laryngoscope 73:85
18. Hoffman JH, Hendrick EB, Humphreys RP (1975) Manifestations and management of Arnold-Chiari malformation in patients with myelomeningocele. Child's Brain 1:255
19. Holinger LD, Holinger PC, Holinger PH (1976) Etiology of bilateral abductor vocal cord paralysis: a review of 389 cases. Ann Otol Rhinol Laryngol 85:428
20. Holinger PC, Holinger LD, Reichert TJ, Holinger PH (1978) Respiratory obstruction and apnea in infants with bilateral abductor vocal cord paralysis, meningomyelocele, hydrocephalus and Arnold-Chiari malformation. J Pediatr 123:368–373
21. Ingraham FD, Scott HW Jr (1943) Spina bifida and cranium bifidum. V. Arnold-Chiari malformation: a study of 20 cases. N Engl J Med 229:108
22. Karshner RG, Reeves DL (1947) Lacunar skull (Lückenschädel) of the newborn. Am J Roentgenol 57:321
23. Kirsch WM, Duncan BR, Black FO, et al (1969) Laryngeal palsy in association with meningomyelocele, hydrocephalus, and the Arnold-Chiari malformation. J Neurosurg 28:207
24. Krieger AJ, Detwiler JS, Trooskin SZ (1976) Respiratory function in infants with Arnold-Chiari malformation. Laryngoscope 86:718
25. Kruyff E, Jeffs R (1966) Skull abnormalities associated with the Arnold-Chiari malformation. Acta Radiol (Diagn) 5:9–24
26. Lichtenstein BW (1942) Distant neuroanatomic complications of spina bifida (spinal dysraphism). Arch Neurol Psychiatr 47:195
27. List CF (1941) Neurologic syndromes accompanying developmental anomalies of the occipital bone, atlas and axis. Arch Neurol Psychiatr 45:577–616
28. Malis LI, Cohen I, Gross SW (1951) Arnold-Chiari malformation. Arch Surg 63:783
29. McLone DG, Kuepper PA (1988) The cause of Chiari II malformation: a unified theory. Pediatr Neurosci 13:1–11
30. McLone DG, Naidich TP (1989) Myelomeningocele: outcome and late complications. In: McLaurin R, Schut L, Venes J, Epstein F (eds) Pediatric neurosurgery, 2nd edn. Saunders, Philadelphia, pp 53–70
31. McLone DG, Raimondi AJ, Sommers MW (1981) The results of early treatment of 100 consecutive newborns with myelomeningocele. Z Kinderchir 2:115–117
32. McLone DG, Suwa J, Collins JA, Poznanski S, Knepper PA (1983) Neurolation: biochemical and morphological studies on primary and secondary neural tube defects. In: Humphreys RP (ed) Concepts in pediatric neurosurgery, vol 4. Karger, Basel, pp 15–29
33. Morley AR (1969) Laryngeal stridor, Arnold-Chiari malformation and medullary hemorrhages. Dev Med Child Neurol 11:471
34. Mullan S, Raimondi AJ (1962) Respiratory hazards of the surgical treatment of the Arnold-Chiari malformation. J Neurosurg 19:675
35. Naidich TP, Pudlowski RM, Naidich JB, Gornish M, Rodriguez FJ (1980) Computed tomographic signs of the Chiari II malformation. I. Skull and dural partitions. Radiology 134:65–71
36. Naidich TP, Pudlowski RM, Naidich JB (1980) Computed tomographic signs of the Chiari II malformation. II. Midbrain and cerebellum. Radiology 134:391–398
37. Naidich TP, Pudlowski RM, Naidich JB (1980) Computed tomographic signs of the Chiari II malformation. III. Ventricles and cisterns. Radiology 134:657–663
38. Naidich TP, McLone DG, Fulling KH (1983) The Chiari II malformation. IV. The hindbrain deformity. Neuroradiology 25:179–197
39. Paul KS, Lye RH, Strang FA, Dutton J (1983) Arnold-Chiari malformation: review of 71 cases. J Neurosurg 58:183–187
40. Peach B (1964) Arnold-Chiari malformation with normal spine. Arch Neurol 10:497–500
41. Peach B (1965) Arnold-Chiari malformation: anatomical features of 20 cases. Arch Neurol 12:613–621
42. Peach B (1965) Arnold-Chiari malformation: morphogenesis. Arch Neurol 12:527–535
43. Penfield W, Coburn DF (1938) Arnold-Chiari malformation and its operative treatment. Arch Neurol Psychiatr 40:328
44. Reichert TJ, Schild JA, Holinger PH (1975) Respiratory obstruction in infants with meningomyelocele and hydrocephalus. Presented at the Third Annual Meeting of the Society for Ear, Nose and Throat Advances in Children, Mexico City
45. Richter RB (1960) Unilateral congenital hypoplasia of facial nucleus. J Neuropathol Exp Neurol 19:33
46. Schmeltzer A, Babin E, Wenger JJ (1971) Foramen magnum in children: measurements of the antero-posterior diameter on midsagittal pneumotomograms. Neuroradiology 2:162–163
47. Schwalbe E, Gredig M (1907) Über Entwicklungsstörungen des Kleinhirns, Hirnstamms und Halsmarks bei Spina bifida (Arnold'sche und Chiari'sche Mißbildung). Beitr Pathol Anat 40:132
48. Sieben RL, Hamida MB, Shulman K (1971) Multiple cranial nerve deficits associated with the Arnold-Chiari malformation. Neurology 21:673–681
49. Smith MEN (1959) The association of laryngeal stridor with meningomyelocele. J Laryngol Otol 73:188
50. Snow JB, Rogers KA Jr (1965) Bilateral abductor paralysis of the vocal cords secondary to the Arnold-Chiari malformation and its management. Laryngoscope 75:316
51. Venes JL, Black KL, Latuck JT (1986) Pre-operative evaluation and surgical management of the Arnold-Chiari II malformation. J Neurosurg 64:363–370
52. Verbiest H (1953) The Arnold-Chiari malformation. J Neurol Neurosurg Psychiatry 16:227
53. Watters GV, Fitch N (1973) Familial laryngeal abductor paralysis and psychomotor retardation. Clin Genet 4:429
54. Wickramasinghe SF, Eckstein HB, Nixon HH (1968) Posterior fossa decompression in shunt-treated hydrocephalic children. Dev Med Child Neurol Suppl 15:11
55. Yu HC, Deck MDF (1971) The clivus deformity of the Arnold-Chiari malformation. Radiology 101:613–615