



Chiari malformation and atlantoaxial instability: problems of co-existence

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Abstract

Background Association of Chiari malformation and atlantoaxial subluxation varies. There is a complex relationship between the two, bony and soft tissue pathologies.

Methods This is a review of various articles available from the literature on the management of Chiari and its association with atlantoaxial instability.

Results We have an experience of operating on 86 cases of paediatric atlantoaxial subluxation, of which 12 had Chiari malformation diagnosed preoperatively (13.95%). Of the 76 children with Chiari malformations operated on by us, 11 had associated atlantoaxial subluxation diagnosed on imaging (14.47%).

Conclusions Re-alignment and reduction with fixation may be effective in achieving decompression in cases where reduction is possible from posterior approach. In these cases, posterior fixation is all that is required. If reduction is not possible from posterior and there is “fixed” ventral compression, anterior decompression needs to be combined with posterior fixation. In most cases, direct posterior decompression is warranted.

Keywords Chiari · AAD · Instrumentation · Decompression · Fixation

Introduction

The association of Chiari malformation and atlantoaxial subluxation varies from 26 to 38% in quoted literature [1–6]. While it is easy to postulate why the two pathologies—one a bony deficiency and the other a soft tissue herniation at the same level—should occur together, reality makes the association more complex to understand.

Development

The craniovertebral junction arises from the occipital somites and the first three cervical somites [7]. The axis which forms the central pivot in a bony ring, develops from the second spinal sclerotome in three stages—pre-cartilaginous stage, stage of chondrification and finally the stage of ossification. The defect in the posterior elements of the axis may be caused by failure of the centres of chondrification, or by failure of the ossification process [8–10].

There have been significant advances in furthering the understanding of the development of the CVJ (craniovertebral junction). There are two families of regulatory genes which are responsible for the development of the sclerotomal parts of the somites during their re-segmentation to form the specific identity of each vertebra [11–14]. These modulate morphogenesis by influencing the transcription of specific downstream genes. Teratogen-induced disturbance of HOX gene expression and mutation in the HOX genes may alter the process of developments of cervical vertebral somites. Inactivation of HOX-D3 gene results in mutant mice with assimilation of the atlas to the basi-occiput [15]. Disturbances of gene expression might prove to be the

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underlying cause of malformations in this region. PAX genes contribute to the development of the early nervous system. Control of re-segmentation of the sclerotomes to establish vertebral boundaries seems to be independently controlled by regulatory genes in the PAX family [12, 14, 16–19]. Alterations in these genes lead to re-segmentation problems and fused vertebrae.

Malformation of the chorda dorsalis is one of the causes of congenital fusion of cervical vertebrae (CFCV) [20, 21]. We also know that retinoids are one of the key factors responsible for the genesis of abnormalities in the axial skeleton [22, 23], and their effect on the formation of the vertebrae is via regulation of the Hox gene [24, 25]. Some studies suggest that decrease in local blood supply is responsible for the CFCV [26]. Whatever be the exact mechanism, there is little doubt that failure of segmentation controlled by a number of environmental and genetic factors is responsible for CFCV. Pax 1 gene with Pax 9 gene controls fusion between the first and second cervical vertebrae, and between the 4th and 5th cervical vertebrae [27]. The Meox 1 gene [28, 29] and the Cyp26b1 gene [30] have been shown specifically to cause vertebral body fusion. Therefore, it is without debate that the process of embryonic segmentation is a genetically controlled phenomenon with minimal environmental influences, and that this occurs around the 8th week of gestational age [31, 32].

If there are inadequacies in the formation of the occipital sclerotomes (between the 4th and 10th weeks of embryonic life) [4, 33, 34] and this is coupled with a small foramen magnum and a small posterior fossa, there is no doubt that the hindbrain may be pushed out of the small posterior fossa causing the Chiari malformation to develop. Also, vertical movement of the odontoid relative to the foramen magnum (the so-called basilar invagination) can further compromise the space available for the craniocervical junction and impair the flow of cerebrospinal fluid (CSF) resulting in a Chiari malformation and a syrinx associated with maldevelopment of the occipital sclerotomes [35–40].

Some authors have suggested that the assembly of the odontoid process, anterior arch of the atlas and the clivus migrated superiorly in unison results in reduction of the posterior cranial fossa volume, and this is the primary pathology in these patients. The Chiari malformation or herniation of the cerebellar tonsil was considered to be a result of reduction in the posterior cranial fossa volume [41].

Radiological features

What are the radiological features which occur in these children? Obviously, the fundamental issue is when there is hindbrain herniation documented on sagittal MR images, one has to do lateral digital dynamic films to check for instability, prior to deciding on any management strategy. Failure to pick up an

atlantoaxial instability may have disastrous consequences if radical posterior fossa decompression is done particularly with removal of posterior arch of the atlas. A 7-year-old boy with symptomatic Chiari I type of malformation, and a cervical syrinx was operated on for minimal hyperreflexia in lower limbs combined with tingling numbness in his hands, and 4 months later presented to us with spastic quadriparesis due to a missed atlantoaxial instability (Fig. 1). Occipitocervical fixation was necessary to reverse (though partially only) his symptoms.

The following radiological features in association with a Chiari malformation should alert the physician about the possibility of instability in the atlantoaxial joint: bifid posterior arch of atlas, os odontoideum, occipitalized atlas, C2–3, C5–6 fusion, syringomyelia [42]. Other authors have suggested the following associations with Chiari malformation: occipitalized atlas and cervical 2–3 vertebral body fusion, platybasia, basilar invagination, and C2–C3 anterior listhesis [43]. In Menezes series, atlas assimilation was the commonest bony anomaly in children with Chiari malformation. Segmentation defects of vertebrae C2 C3 and median basilar invagination were also noted [3]. Some authors have suggested an os odontoideum as an associated bony abnormality in patients with Chiari malformation. However, analysis of various series where the two pathologies co-exist has shown incidences which are fairly low (1–8%) [44–47].

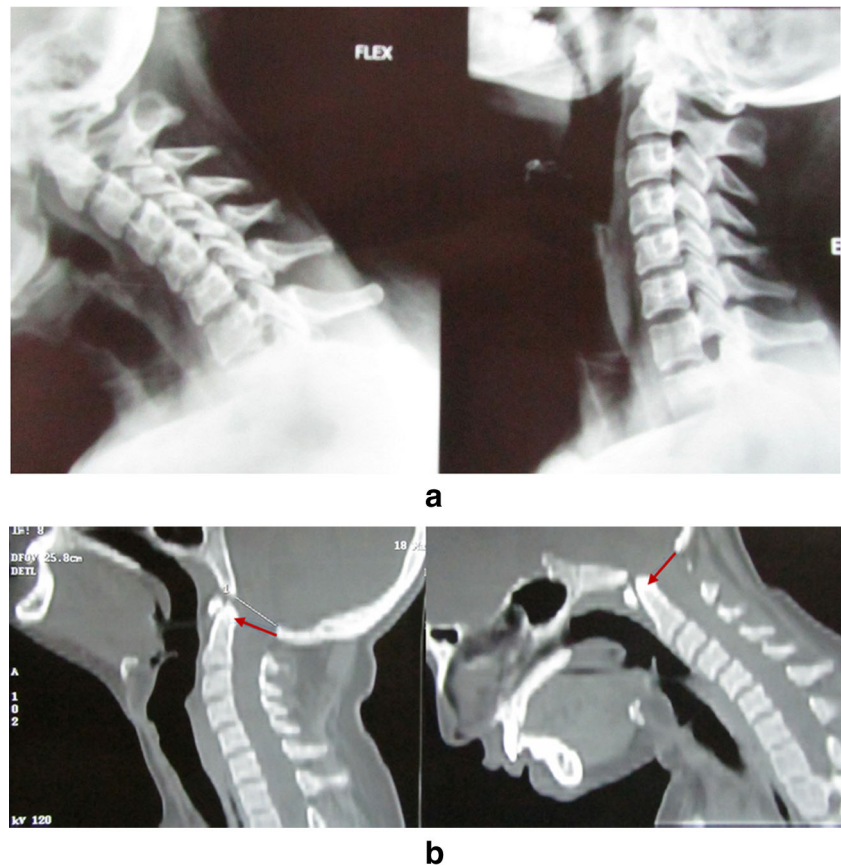
Material and methods: Our experience

We have the experience of dealing with 4 children with missed subluxation, who deteriorated after surgery for Chiari. All of them improved after posterior fixation surgery at a second stage. However, in 2 cases, the improvement was not satisfactory. It is our practice to routinely perform dynamic lateral digital films in all children with Chiari malformation to rule out subluxation prior to decompression for the Chiari. In case of doubt, we perform flexion-extension CT scans, so paranoid are we about



Fig. 1 Worsening of spastic quadriparesis in a child due to missed AAD and operated Chiari I malformation

Fig. 2 **a** Flexion-extension dynamic films do not clarify presence of AAD. **b** Flexion-extension CT scans establish the diagnosis of AAD



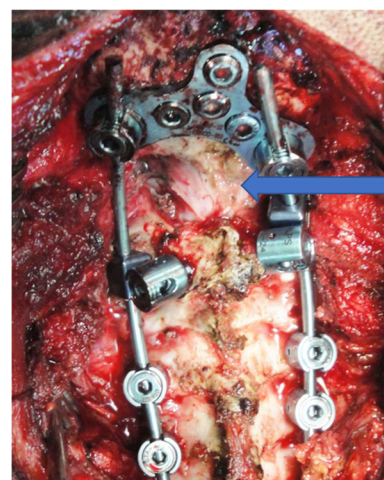
missing subluxation (Fig. 2a and b). We have not encountered subluxation at surgery where we have not been able to demonstrate the same radiologically preoperatively.

Out of 86 cases of paediatric atlantoaxial subluxation operated on by us, 12 had Chiari malformation diagnosed preoperatively (13.95%). Of the 76 children with Chiari malformations operated on by us, 11 had associated atlantoaxial subluxation diagnosed on imaging (14.47%). We have followed the policy of offering posterior decompression alone if no instability is documented preoperatively, and posterior fixation with decompression if instability is documented preoperatively but this is reducible by either position or traction (Fig. 3). We have reserved anterior decompression in addition to posterior decompression and fixation in cases with “fixed” subluxation and significant anterior compression, e.g. in retroverted odontoid compressing the cord with Chiari malformation.

Discussion: Treatment

The fundamental debate in the children who have documented atlantoaxial instability and Chiari malformation revolves around the following questions:

1. Is it necessary to treat both the problems or only the instability?
2. If the instability is addressed alone, is it necessary to address the Chiari malformation at a second stage?
3. If both have to be addressed together, what is the extent of decompression and what indeed the extent of fixation?
4. What is the fate of the associated syrinx in this situation?



Decompression performed

Fig. 3 Posterior decompression and fixation in AAD with Chiari malformation

We shall endeavour to collect the evidences for each of the questions.

The fixers

In his article, Dr. Goel describes how all patients were treated with atlantoaxial plate and screw fixation. He concluded that atlantoaxial stabilization without bone decompression and without duroplasty is the main treatment form for symptomatic tonsillar herniation, whether or not the condition is associated with BI (basilar invagination) or syringomyelia. According to the author's new theory, patients with BI had instability even when it was not demonstrated by dynamic changes. The same theory was applied to CM (Chiari malformations)—all patients had atlantoaxial instability. They have postulated that syringomyelia in CM is primarily because of the C1–2 instability, either subtle or apparent [48]. The resultant tonsillar herniation is the result of a compensatory protective phenomenon, acting as natural “air-bags” that protect the spinal cord from the effects of the atlantoaxial instability [49]. However, the author claimed that in all these cases, he was able to discern instability intra-operatively. Finally, it was reported that CM with or without BI is always associated with atlantoaxial instability even when it is not demonstrable on radiological imaging and that foramen magnum decompression was unnecessary and actually harmful [48]. The hypothesis suggested by Goel et al. is interesting but does require randomization as well as carefully conducted studies to prove its value. It is also limited by significant ethical issues, such as not performing a foramen magnum decompression and/or duroplasty in patients in the presence of a symptomatic tonsillar herniation.

The decompressors

The opinion is challenged by Joaquim AF et al. According to them, routine atlantoaxial fixation without posterior fossa decompression for patients with tonsillar herniation, with or without syringomyelia, and no evident CVJ instability may not be recommended until further evidence is available. Furthermore, fixation of the atlantoaxial complex may result in restriction of the cervical rotation and increase morbidity [50]. Also, this new theory cannot explain the clinical improvement seen in patients in the previously reported series, who underwent posterior fossa decompression alone for treating CM, even though Goel et al. emphasize that the improvement which occurs is due to release of compression by the “air bags-tonsils”, and that the neurological improvement that occurs following posterior fossa decompression is a transient improvement as it does not tackle the primary cause, which is instability at the C1–C2 joints [48].

As has also been stated by many other authors, Joaquim also suggested that a congenital shallow posterior fossa is well documented in CM and is the most accepted pathogenic phenomenon for explaining the symptoms in CM [51–53]. Their opinion is supported by Rahman et al. The author pointed out that the tonsils are not “air-bags” since they cause more compression of the neural structures than CSF, suggesting that AAD is not the cause of symptoms in CM [54]. This conventional approach to treatment of Chiari malformations has stood the test of time, but there is no doubt a group of patients deteriorated after posterior fossa decompression. Could underlying undiagnosed instability be the cause?

The fixers and decompressors

Arnold Menezes emphasized that it was the ventral decompression which allowed re-establishment of CSF pathways in fixed dislocations. Patients with irreducible ventral bone abnormalities compressing the medulla and cervicomedullary junction underwent ventral decompression via a transoral or a transpalatopharyngeal route and a dorsal occipitocervical fusion. In reducible craniocervical junction abnormalities, dorsal decompression was performed as needed (foramen magnum-posterior atlas arch) and a limited occipitocervical fusion (occiput to C1 to C2). The presence of syringomyelia and hindbrain herniation did not change this decision [55].

The cause of deterioration after posterior decompression in patients with ventral CVJ abnormalities who undergo a primary PFDD has been investigated by many authors. It is possible that deterioration is due to angulation that takes place at the cervicomedullary junction from the offending ventral pathology during the operation in a prone position, and the fact that instability at the atlantoaxial joint has not been detected [56]. Delayed worsening may be due to a gradual appearance of instability and cranial settling. This latter condition is attributed to the so-called cantilever effect of the cranium. This leads to development of delayed instability [57]. The evidence for this is the significance of the syrinx disappearing after ventral CVJ decompression in the face of hindbrain herniation. It is more than likely that this results from removal of the CSF block at the level of the foramen magnum and also from restoration of the posterior fossa volume. It is possible that there is a reversal of the CSF craniospinal pressure after decompression. However, a combination of events most likely takes place to provide for the significant clinical as well as radiographic improvement [55].

Kohno reported on a case where successful treatment of “adult Arnold-Chiari malformation with associated basilar impression and syringomyelia” used the anterior transoral route. This patient underwent an anterior fusion together with the transoral decompression. Postoperative MR imaging showed that the cerebellar tonsils had ascended and the syringomyelia had regressed; no dorsal decompression was conducted [58].

Klekamp J studied Chiari patients with and without basilar invagination although his series included adult patients. His study presents surgical results for patients with Chiari I malformation with and without additional basilar invagination. Patients with ($n = 46$) basilar invagination in addition to Chiari I malformation were separated into groups: those with ($n = 31$) and without ($n = 15$) ventral compression by the odontoid in the foramen magnum. All craniospinal operations included a foramen magnum decompression with arachnoid dissection, opening of the fourth ventricle and a duroplasty. In all patients with ventral compression, craniospinal stabilization was performed with the foramen magnum decompression, except for 4 patients with mild ventral compression early in the series who underwent posterior decompression only. In the group with ventral compression, 9 patients with caudal cranial nerve dysfunctions underwent a combination of transoral decompression with posterior decompression and fusion. They conclude that Chiari I malformations without invagination and those with invaginations but without ventral compression could be managed by foramen magnum decompression alone. The majority of patients with ventral compression however needed treatment by posterior decompression, re-alignment and stabilization, reserving anterior decompressions for patients with profound, symptomatic brainstem compression [59].

SW Hwang suggests an anterior and posterior circumferential decompression with C1C2 arthrodesis in patients with Chiari malformation and AAD [60]. Jea adopted the middle path [61]. Neurological status scores of patients with Chiari I malformation with irreducible AAD who underwent single-stage transoral decompression with posterior stabilization were significantly better than those patients who underwent the posterior procedure alone. In the presence of CM-I with reducible atlantoaxial dislocation, reduction and stabilization of the joint produced better results. He argued that patients with Chiari I malformation should be investigated for the presence of atlantoaxial dislocation. In case atlantoaxial dislocation co-exists, priority must be given to relieving anterior cervicomedullary compression [60].

In the series by Hankinson, posterior decompression alone was sufficient in the majority of cases but where anterior decompression was required this was always supplemented by posterior fixation [62].

Conclusions: The decompression/fixation summary—our philosophy

So, here is a summary of the problem:

1. The children have an atlantoaxial instability and a hind-brain herniation with or without a syrinx.
2. There is no doubt that the CSF circulation is impeded by the herniation as well as the anterior compression caused by the AAD.
3. What is clear is that, for symptomatic improvement, CSF circulation at the foramen magnum needs to be re-established.
4. Re-alignment and reduction with fixation may be effective in achieving decompression in cases where reduction is possible from posterior approach. Here, posterior fixation is all that is required.
5. If reduction is not possible from posterior and there is “fixed” ventral compression, anterior decompression needs to be combined with posterior fixation, although some authors have advocated anterior fixation.
6. There is enough evidence including our own data to suggest that if atlantoaxial subluxation is missed, and posterior decompression alone is done, these children deteriorate and require fixation. Hence, the question of two-stage approach to this problem cannot arise.
7. Authors have reported using ultrasound to assess CSF flow across the foramen magnum. If one were to belong to the Goel school, one would need to demonstrate evidence of CSF flow after re-alignment and fixation alone. If this was in doubt, additional posterior decompression should be the sensible approach. Moreover, whether decompression should be bony or dural can also be decided on the basis of ultrasound evidence of flow.

Perhaps if one looks at the literature dispassionately, there is less disagreement among the groups than it would seem at first look.

Compliance with ethical standards

Conflict of interest None.

References

1. Levy WJ, Mason L, Hahn JF (1983) Chiari malformation presenting in adults: a surgical experience in 127 cases. *Neurosurgery* 12: 377–390
2. Dyste GN, Menezes AH, VanGilder JC (1989) Symptomatic Chiari malformations. *J Neurosurg* 71:159–168
3. Menezes AH (1995) Primary craniovertebral anomalies and the hindbrain herniation syndrome (Chiari I): data base analysis. *Pediatr Neurosurg* 23:260–269
4. Milhorat TH, Chou MW, Trinidad EM et al (1999) Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery* 44:1005–1017
5. Menezes AH, Greenlee JDW, Donovan KA (2005) Honored guest presentation: lifetime experiences and where we are going: Chiari I with syringohydromyelia—controversies and development of decision trees. *Clin Neurosurg* 52:297–305

6. Caetano de Barros M, Farias W, Ataíde L, Lins S (1968) Basilar impression and Arnold-Chiari malformation. A study of 66 cases. *J Neurol Neurosurg Psychiatry* 31:596–605
7. Pang D, Thompson DNP (2011) Embryology and bony malformations of the craniovertebral junction. *Childs Nerv Syst* 27:523–564
8. van Gilder JCMA (1983) Craniovertebral junction abnormalities. *Clin Neurosurg* 30:514–530
9. Asakawa H, Yanaka K, Narushima K et al (1999) Anomaly of the axis causing cervical myelopathy. *J Neurosurg Spine* 91:121–123
10. Behari S, Kiran Kumar MV, Banerji D et al (2004) Case report atlantoaxial dislocation associated with the maldevelopment of the posterior neural arch of axis causing compressive myelopathy. *Neurol India* 52(4):489
11. Lufkin T, Mark M, Hart CP et al (1992) Homeotic transformation of the occipital bones of the skull by ectopic expression of a homeobox gene. *Nature* 359:835–841
12. Condie BG, Capecchi MR (1993) Mice homozygous for a targeted disruption of *Hoxd-3* (*Hox-4.1*) exhibit anterior transformations of the first and second cervical vertebrae, the atlas and the axis. *Development* 119:579–595
13. Dietrich S, Kessel M (1997) The vertebral column. In: Thorogood P (ed) *Embryos, genes and birth defects*. Wiley, Chichester, pp 281–30214
14. Menezes AH (2008) Craniocervical developmental anatomy and its implications. *Childs Nerv Syst* 24:1109–1122
15. Chisaka O, Capecchi MR (1991) Regionally restricted developmental defects resulting from targeted disruption of the mouse homeobox gene *hox-1.5*. *Nature* 350:473–479
16. Kessel M (1993) Reversal of axonal pathways from rhombomere 3 correlates with extra *hox* expression domains. *Neuron* 10:379–393
17. Koseki H, Wallin J, Wilting J, Mizutani Y, Kispert A, Ebensperger C, Herrmann BG, Christ B, Balling R (1993) A role for *Pax-1* as a mediator of notochordal signals during the dorsoventral specification of vertebrae. *Development* 119:649–660
18. Kimura Y, Matsunami H, Inoue T et al (1995) *Cadherin-11* expressed in association with mesenchymal morphogenesis in the head, somite, and limb bud of early mouse embryos. *Dev Biol* 169:347–358
19. Keynes RJ, Stern CD (1988) Mechanisms of vertebrate segmentation. *Development* 103:413–429
20. Erdil H, Yildiz N, Cimen M (2003) Congenital fusion of cervical vertebrae and its clinical significance. *J Anat Soc India* 52:125–127
21. Saraga-Babić M, Saraga M (1993) Role of the notochord in the development of cephalic structures in normal and anencephalic human fetuses. *Virchows Arch A Pathol Anat Histopathol* 422:161–168
22. Sakai Y, Meno C, Fujii H et al (2001) The retinoic acid-inactivating enzyme *CYP26* is essential for establishing an uneven distribution of retinoic acid along the antero-posterior axis within the mouse. *Genes Dev* 15:213–225
23. Kashyap V, Gudas L, Brenet F et al (2011) Epigenomic reorganization of the clustered *Hox* genes in embryonic stem cells induced by retinoic acid. *J Biol Chem* 286(8):3250–3265
24. Wellik DM, Capecchi MR (2003) *Hox10* and *Hox11* genes are required to globally pattern the mammalian skeleton. *Science* (80-) 301:363–367
25. Wellik DM (2009) *Hox* genes and vertebrate axial pattern. *Curr Top Dev Biol* 88:257–284
26. Mallo M, Wellik D, Deschamps J (2010) *Hox* genes and regional patterning of the vertebrate body plan. *Dev Biol* 344:7–15
27. McGaughan JM, Oates A, Donnai D et al (2003) Mutations in *PAX1* may be associated with Klippel–Feil syndrome. *Eur J Hum Genet* 11:468–474
28. Soshnikova N, Dewaele R, Janvier P et al (2013) Duplications of *hox* gene clusters and the emergence of vertebrates. *Dev Biol* 378:194–199
29. Jukkola T, Trokovic R, Maj P et al (2005) *Meox1Cre*: a mouse line expressing *Cre* recombinase in somitic mesoderm. *Genesis* 43:148–153
30. Abu-Abed S, Dollé P, Metzger D et al (2001) The retinoic acid-metabolizing enzyme, *CYP26A1*, is essential for normal hindbrain patterning, vertebral identity, and development of posterior structures. *Genes Dev* 15:226–240
31. Mardani M, Borujeni MJS, Esfandiary E (2016) Congenital fusion of cervical vertebrae: a review on embryological etiology. *Rev Clin Med* 3:148–153
32. Chatterjee S (2018) The chicken or the egg - pancervical fusion with atlantoaxial dislocation. *Neurol India* 66:153
33. Nishikawa M, Sakamoto H, Hakuba A et al (1997) Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. *J Neurosurg* 86:40–47
34. Moore KL, Persaud TVN, Torchia MG (1998) *The developing human: clinically oriented embryology*, 6th edn. WB Saunders, Philadelphia, pp 405–426
35. Wackenheim A (1974) Roentgen diagnosis of the craniovertebral region. Springer-Verlag, Berlin, pp 353–379
36. Shapiro R, Robinson F (1976) Anomalies of the craniovertebral border. *Am J Roentgenol* 127:281–287
37. Di Lorenzo N, Fortuna A, Guidetti B (1982) Craniovertebral junction malformations. *J Neurosurg* 57:603–608
38. Mesiwala AH, Shaffrey CI, Gruss JS, Ellenbogen RG (2001) Atypical hemifacial microsomia associated with Chiari I malformation and syrinx: further evidence indicating that Chiari I malformation is a disorder of the paraaxial mesoderm. *J Neurosurg* 95:1034–1039
39. Kagawa M, Jinnai T, Matsumoto Y et al (2006) Chiari I malformation accompanied by assimilation of the atlas, Klippel–Feil syndrome, and syringomyelia: case report. *Surg Neurol* 65:497–502
40. Fenoy AJ, Menezes AH, Fenoy KA (2008) Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia. *J Neurosurg Spine* 9:1–9
41. Goel A (2009) Basilar invagination, Chiari malformation, syringomyelia: a review. *Neurol India* 57:235
42. Achawal AGS, Goel SA (1995) The surgical treatment of chiari malformation associated with atlantoaxial dislocation. *Br J Neurosurg* 9:67–72
43. Behari S, Kalra SK, Kiran Kumar MV et al (2007) Chiari I malformation associated with atlanto-axial dislocation: focussing on the anterior cervico-medullary compression. *Acta Neurochir* 149:41–50
44. Kumar R, Kalra SK, Vaid VK, Mahapatra AK (2008) Chiari I malformation: surgical experience over a decade of management. *Br J Neurosurg* 22:409–414
45. Galarza M, Martínez-Lage JF, Ham S, Sood S (2011) Cerebral anomalies and Chiari type I malformation. *Pediatr Neurosurg* 46:442–449
46. Dlouhy BJ, Policeni BA, Menezes AH (2017) Reduction of atlantoaxial dislocation prevented by pathological position of the transverse ligament in fixed, irreducible os odontoideum: operative illustrations and radiographic correlates in 41 patients. *J Neurosurg Spine* 27:20–28
47. Goel A, Gore S, Shah A et al (2018) Atlantoaxial fixation for Chiari I formation in pediatric age-group patients: report of treatment in 33 patients. *World Neurosurg* 111:e668–e677
48. Goel A (2015) Is atlantoaxial instability the cause of Chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. *J Neurosurg Spine* 22:116–127

49. Goel A (2014) Is Chiari malformation nature's protective "air-bag" ? Is its presence diagnostic of atlantoaxial instability? *J Craniovertebr Junction Spine* 5:107–109
50. Joaquim A, Tedeschi H, Ps C (2019) Controversies in the surgical management of congenital craniocervical junction disorders – a critical review. *Neurol India* 66:1003
51. Menezes AH (2011) Current opinions for treatment of symptomatic hindbrain herniation or Chiari type I malformation. *World Neurosurg* 75:226–228
52. Bollo R, Riva-Cambrin J, ... Brockmeyer DL (2012) Complex Chiari malformations in children: an analysis of preoperative risk factors for occipitocervical fusion. *Pediatrics* 10:134–41
53. Salunke P, Sura S, Futane S et al (2012) Ventral compression in adult patients with Chiari I malformation sans basilar invagination: cause and management. *Acta Neurochir* 154:147–152
54. Rahman A (2015) Letter to the editor: does atlantoaxial dislocation really cause Chiari? *J Neurosurg Spine* 23:393–394
55. Menezes AH (2012) Craniovertebral junction abnormalities with hindbrain herniation and syringomyelia: regression of syringomyelia after removal of ventral craniovertebral junction compression. *J Neurosurg* 116:301–309
56. Menezes AH, VanGilder JC, Graf CJ, McDonnell DE (1980) Craniocervical abnormalities. A comprehensive surgical approach. *J Neurosurg* 53:444–455
57. Müller F, O'Rahilly R (1991) Development of anencephaly and its variants. *Am J Anat* 190:193–218
58. Kohno K, Sakaki S, Shiraishi T et al (1990) Successful treatment of adult Arnold-Chiari malformation associated with basilar impression and syringomyelia by the transoral anterior approach. *Surg Neurol* 33:284–287
59. Klekamp J (2015) Chiari I malformation with and without basilar invagination: a comparative study. *Neurosurg Focus* 38:E12
60. Hwang SW, Heilman CB, Riesenburger RI, Kryzanski J (2008) C1–C2 arthrodesis after transoral odontoidectomy and suboccipital craniectomy for ventral brain stem compression in Chiari I patients. *Eur Spine J* 17:1211–1217
61. Jea A (2015) Editorial: Chiari malformation I surgically treated with atlantoaxial fixation. *J Neurosurg Spine* 22:113–115
62. Hankinson TC, Klimo P, Feldstein NA et al (2007) Chiari malformations, syringohydromyelia and scoliosis. *Neurosurg Clin N Am* 18:549–568

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