

Extradural Decompression for the Treatment of Oculomotor and Oropharyngeal Symptoms in Chiari I Malformation: A Case Report

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Abstract

Background and importance: Chiari I malformation (CIM) represents a subset of neurological disorders characterized by herniation of cerebellar tonsils below the foramen magnum. Importantly, it is associated with various motor and sensory dysfunctions. Extradural decompression is one of the most promising surgical approaches for treating CIM. Compared to more invasive techniques, a purely extradural decompression allows for the timely resolution of clinical symptoms as well as reduces the risk of postoperative complications.

Clinical presentation: We report the case of an 8-month old patient presenting with nystagmus, esotropia, laryngomalacia, and sleep apnea secondary to CIM. The patient underwent an extradural decompression, which involved removal of the posterior ring of C1 in addition to the outer dural layer. The patient had a gradual resolution of his symptoms over a several month period. After 4 months, the patient's esotropia and laryngomalacia resolved and his motor nystagmus and sleep apnea also improved significantly. His delayed visual maturation continues to improve.

Conclusion: Extradural decompression has the potential to be used as a first-line treatment for symptomatic CIM. Our case exemplifies how extradural decompression can be used, in a pediatric patient, to successfully treat visual and oropharyngeal deficits associated with CIM.

Keywords: Extradural decompression; Chiari I malformation; Oculomotor symptoms

Background and Importance

First described by Cleland in 1883, Chiari malformation is a group of congenital abnormalities of the cerebellum, brainstem, and craniocervical junction with downward displacement of the cerebellar tonsils into the spinal canal [1]. Chiari later stratified these malformations into four categories based on increasing levels of clinical and radiographic severity. Chiari I malformation (CIM), the most common subtype, is defined as a caudal displacement of the cerebellar tonsils 5 mm or more below the level of the foramen magnum. Clinical symptoms can vary but often include headaches, sleep apnea, tinnitus, vertigo, motor weakness, gait impairment, and incoordination. These symptoms are attributed to altered cerebral spinal fluid (CSF) flow dynamics at the craniocervical junction or alternatively to direct compression of brainstem and spinal cord [2].

While a number of surgical techniques have been described for treatment, there is no consensus on the optimal method for CIM management. The most common surgical approach includes posterior fossa decompression, ranging from decompression with extradural splitting to a more invasive suboccipital craniectomy with duraplasty [3]. While intradural techniques allow for the restoration of CSF flow, they also pose a risk of numerous complications including meningocele, PICA infarct, meningitis, and CSF leaks. Extradural decompression is a minimally invasive technique, which utilizes

unique expansile properties of the inner dural layer. This surgical approach consists of removal of only the outer layer of the dura [4,5], allowing for decompression of the more expandable inner dural layer and subsequent restoration of CSF flow, thereby avoiding these potential complications. Mounting clinical evidence in CIM patients treated with extradural decompression demonstrates significant improvements in symptoms with a minimal reoperation rate. We propose that in addition to treating classic CIM symptoms, this technique can be used for treatment of oculomotor and oropharyngeal symptoms.

In this report we present the case of an 8-month old male with CIM who had complete resolution of his nystagmus, esotropia, and stridor after an extradural decompression. Additionally, we review cases of patients with similar symptoms that resolved through a comparable extradural surgical technique.

Clinical Presentation

An 8-month-old boy presented to the ophthalmologist with a history of a high-frequency horizontal, low-amplitude, rotatory nystagmus, right-sided intermittent esotropia, laryngomalacia, sleep apnea, as well as delayed visual maturation. He was the product of a full-term birth to a 35 year-old mother who had a previous pregnancy with trisomy 14. An MRI of the brain revealed inferior displacement of the cerebellar tonsils 7 mm below the foramen magnum, with no signs of hydrocephalus (Figure 1).

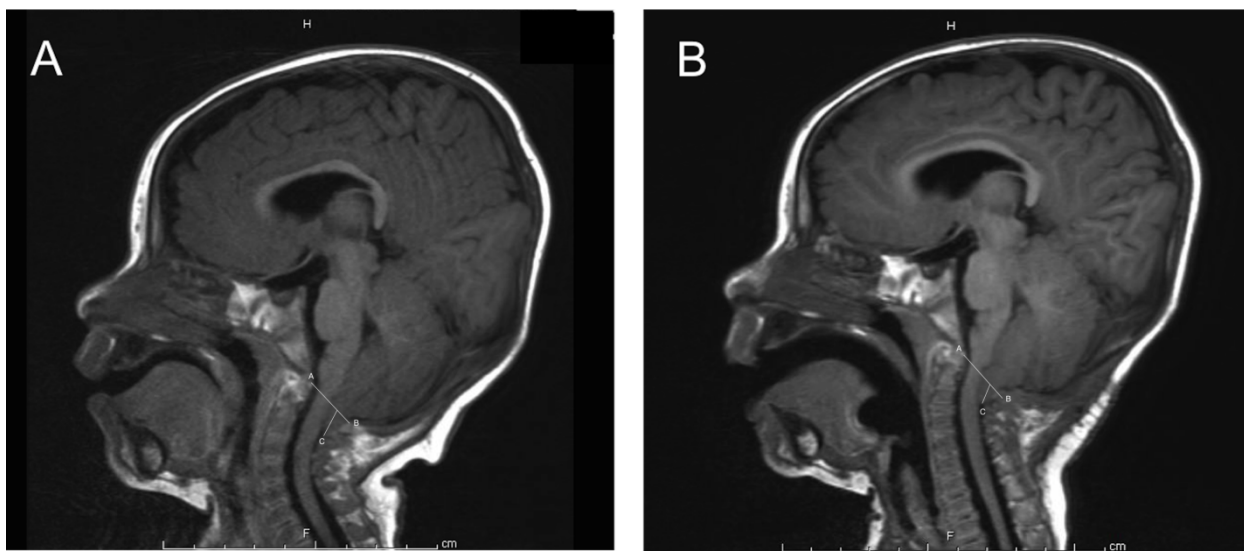


Figure 1: a) Pre-operative sagittal T1 MRI brain demonstrating tonsillar herniation 7 mm below the level of the foramen magnum. The horizontal line between points A and B represents the distance between anterior and posterior margins of the foreman magnum. The vertical line C represents tonsillar herniation. There is no evidence of syringohydromyelia in upper cervical spinal cord. The lateral and third ventricles are larger in size, but consistent with the age of the patient. The fourth ventricle is normal in size. B) Post-operative sagittal T1 MRI brain demonstrating Chiari I malformation with inferior displacement of cerebellar tonsils status post suboccipital craniectomy. Line C represents significant reduction in tonsillar herniation.

No syringohydromyelia was noted and the degree of myelination was consistent with the patient's age. There was no abnormal parenchymal enhancement. An MRI of the orbits showed smaller than normal optic nerves and an optic chiasm, with no other abnormalities noted (Figure 2). Overall the findings were consistent with Chiari I malformation with optic nerve hypoplasia. On neurological examination, he was alert and playful, and had a mild deformational plagiocephaly. His cranial nerves were intact, reflexes were normal, and no head bobbing or abnormal head positions were noted.

Based on the patient's presentation and confirmed diagnosis of CIM, the parents elected for the patient to undergo an extradural Chiari I decompression. A standard suboccipital craniectomy with removal of the posterior ring of C1 was performed. The decompression was carried down to the level of the foramen magnum and the outer portion of the dura was split to allow for restoration of CSF flow. There were no postoperative complications. At 2 and 4 months follow-up, the patient's horizontal nystagmus had significantly improved and his esotropia had resolved. The patient's laryngomalacia also improved and residual symptoms resolved with an antacid. A sleep study confirmed moderate obstructive sleep apnea rather than central apnea, as well as improvement compared to a prior study. The patient continues to be seen at our institution so we can continue to follow his subsequent improvement.

Discussion

Chiari I malformation: Prevalence and causes

Chiari I malformation is classified as neurological anomaly of the craniovertebral junction. Its estimated prevalence rate in the United States is approximately 0.1-0.5% [6,7]. The increased use of

neuroimaging has been useful in identifying asymptomatic or minimally symptomatic patients at much earlier age.

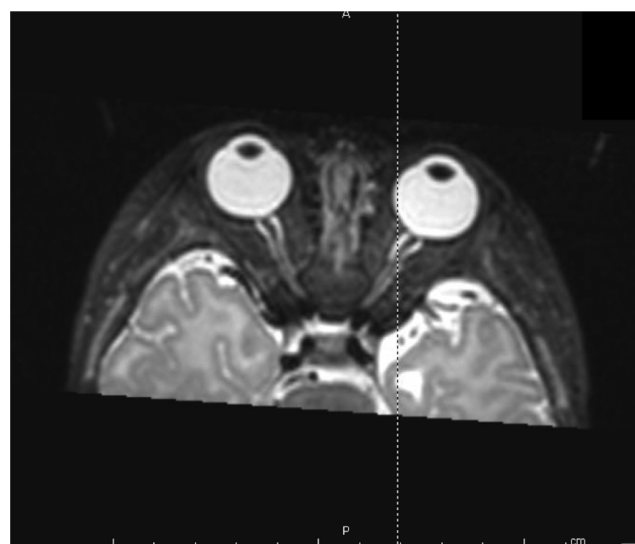


Figure 2: Axial MRI orbits demonstrating bilateral optic nerve hypoplasia. The optic nerves and optic chiasm appear to be smaller; the globes are intact.

CIM is considered a heterogeneous disorder, which arises due to interplay between multiple genetic and environmental factors. Earlier studies using whole genome sequencing were able to link CIM with certain regions on chromosomes 9 and 15 [8]. More recently,

Markunas et al. proposed involvement of growth differentiation factors GDF6 and GDF3 on chromosomes 8 and 12, respectively [9]. The multifactorial nature of CIM makes it difficult to pinpoint the exact genetic cause of this disorder. In our case, the patient's mother underwent chorionic villus sampling with chromosomal microarray analysis during her pregnancy, which yielded normal results and 46XY karyotype. Her past medical history was significant for Trisomy 14 in a previous pregnancy. Trisomy 14 is a rare chromosomal disorder, which is, in most cases, associated with spontaneous abortion, but not CIM. Despite numerous theories to explain the cause of CIM, most cases are sporadic.

Clinical symptoms

CIM is defined by tonsillar herniation of more than 5 mm below the level of the foramen magnum. The condition may also present with hydrocephalus or syringomyelia as the result of CSF pathway obstruction [7]. The amount of herniation, size of the posterior fossa, and degree of posterior fossa crowding, all play a role in the development of symptoms. Symptoms associated with compression of the brainstem and spinal cord may include motor and sensory dysfunction, dysarthria, dysphagia, and incontinence [10]. Occipital headaches and vestibuloocular dysfunctions including nystagmus, esotropia, blurred vision, vertigo, and dizziness are among the most common presentations in symptomatic patients with CIM. In our case, the patient's physical examination revealed normal reflexes and motor functions. He first presented with oropharyngeal and visual deficits, including horizontal nystagmus, esotropia, as well as sleep apnea. His symptoms were among those more commonly seen in children less than 2 years of age and can be attributed to lower cranial nerve compression.

Ocular manifestations of CIM may include different forms of nystagmus (horizontal, downbeat, periodic alternating), esotropia, skew deviation, ocular flutter, and dysmetria. Nystagmus is thought to arise from cerebellar compression, particularly of the uvula, nodulus, and vermis since these structures maintain inhibitory control in response to vestibular and optokinetic stimulations and regulate the time course of eye movements [11]. Esotropia, arising from compression of the abducens nerve, can also be present and signifies an early sign of CIM. These symptoms can result in various degrees of disability and present at any age range. Likewise, our patient is an 8-month-old boy with CIM who presented with similar symptomatology secondary to CIM. Amal Al-Awami et al. reported a similar case of a patient with periodic alternating nystagmus secondary to CIM, that resolved with posterior fossa decompression and removal of the right cerebellar tonsil [11]. Furthermore, in retrospective study by Liebenberg et al. of 12 patients with CIM who underwent posterior fossa decompression, 75% demonstrated improvement and subsequent resolution of oculomotor manifestations [12]. Lastly, successful resolution of esotropia has previously been reported following suboccipital decompression with duraplasty as well [13].

In addition to vestibuloocular dysfunctions, children with CIM, who are younger than 2 years-old, often present with oropharyngeal dysfunction, including snoring, sleep apnea, and stridor. Numerous studies report a 24-63% prevalence of sleep-disordered breathing in children with CIM secondary to both central and obstructive apnea [14-18]. Central sleep apnea may occur as a result of dysfunctional respiratory centers, medullary ischemia, or alternatively from impaired chemoreceptor function. Obstructive apnea could occur due to

reduced pharyngeal muscle tone and loss of upper airway sensation. It is not known why some CIM patients have central and others have obstructive sleep apnea, but it may depend on the degree of tonsillar herniation [16]. Tran et al. report the case of a young patient with obstructive and central sleep apnea secondary to CIM, with symptoms successfully resolving following decompression of the craniocervical junction [19]. Botelho et al. studied 25 patients with CIM and sleep apnea who underwent extraarachnoidal decompression, which included opening of the dura, but leaving the arachnoid and cerebellar tonsils intact. Surgery resulted in an enhanced sleep apnea index and decreased respiratory events during sleep [20]. In the present case, our patient's sleep apnea significantly improved after a purely extradural decompression.

Management

While there is no single definitive treatment for CIM, the most common approach is a suboccipital craniectomy and C1 laminectomy either with or without duraplasty. We performed a literature search of patients with CIM and associated esotropia by searching PUBMED using the terms "Chiari I Malformation and Esotropia" (Table 1). In the majority of cases, patients underwent an intradural decompression with duroplasty and subsequently had resolution of their symptoms [21-30]. Arnautovic et al. reviewed surgical approaches for CIM treatment in pediatric populations from 1965 to 2013 and reported that 92% of decompression procedures involved duraplasty, and in 65% of those cases, the arachnoid was additionally opened and dissected. Overall, neurological symptoms improved in 75% of cases [31]. However, this procedure poses the risk of numerous post-operative complications, including CSF leaks, pseudomeningocele, meningitis, PICA infarct, and neurological deficits. Posterior fossa decompression without duraplasty is a less invasive technique that provides similar clinical improvement rates and lower rates of the complications. According to Litvack's review of 110 cases of pediatric patients with CIM, 9% of the patients who underwent duraplasty experienced CSF-related complications compared to none in the extradural cohort [32]. Furthermore, opening the dura is also associated with longer operative times, prolonged hospital stays, and higher costs for the primary hospitalization [32]. Chotai et al. reviewed and compared the results of the posterior fossa decompression with and without duraplasty in 41 patients who were treated at the University of Toledo Medical Center from 2000 to 2012. Among 41 patients, 12 underwent posterior fossa decompression with duroplasty with 4 of them developing post-operative neurologic complications. In the groups of 29 patients, who were treated without duroplasty, only 1 person developed wound infection. The results of this study indicate favorable surgical outcomes, including decreased duration of preoperative symptoms and decreased complication rate, with extradural decompression in CIM patients without syringomyelia [33].

Another review of surgical series of 11 patients with CIM who underwent extradural decompression at our institution has shown a successful partial or complete resolution of symptoms in 8/11 (72.7%) patients. There was only one case of development of post-operative complication of dysphagia and dysphonia [34,35].

In the case of our patient, a less invasive extradural decompression, involving splitting the outer dura matter and leaving the inner layer intact, was chosen as first line treatment due to its lower risk of complications and the patient's young age.

Reference	Age (gender)	Herniation (mm)	Treatment	Duroplasty	Outcome
Defoort-Dhellemmes et al. [13]	9 (M)	to C3	PFD	Yes	mild esophoria
Baxstrom [21]	14 (F)	-	PFD and prismatic correction	Yes	no esotropia
Biousse et al. [22]	14 (F); 16 (M); 37 (F); 5 (M)	-	PFD; PFD; strabismus surgery; strabismus surgery	-	mild esophoria; orthophoric; recurrence of esotropia; no esotropia
Firth and Burke [23]	16 (F)	12	BT 8 months prior to PFD	-	BT restored BSV prior to PFD
Hentschel et al. [24]	5 (M)	to C3	PFD	Yes	no esotropia
Imes and Quinn [25]	14 (F)	-	PFD and BT	-	no esotropia
Lewis et al. [26]	17 (F); 23 (F); 24 (F); 36 (F); 19 (F)	-	PFD (4 patients); no treatment (19, F)	Yes	no esotropia; improvement of esotropia; no esotropia; no esotropia; n/a
Passo et al. [27]	24 (F)	-	strabismus surgery followed by PFD	-	orthophoric
Pokharel and Statkowski [28]	15 (F)	4	PFD and strabismus surgery	-	no esotropia
Rech et al. [29]	6 (F)	10	PFD followed by strabismus surgery	No	no esotropia
Weeks and Hamed [30]	14 (M); 35 (F)	30; 15	strabismus surgery followed by PFD (both patients)	-	mild esophoria (both patients)

PFD - posterior fossa decompression; BT - botulinum toxin; BSV - binocular single vision

Table 1: Esotropia and Chiari Malformation.

Isu et al. first attempted this technique in 1993, with patients experiencing remarkable improvement in their symptoms [36]. Several studies recommend using an intradural technique when there is a syrinx present, and only resorting to an extradural decompression when there is no syrinx or other abnormality [32,37]. In another large series of 30 pediatric and adult patients, all saw either improvement or resolution of their symptoms, including those who had syrinxes [38].

Furthermore, half of those with syrinxes saw a reduction in size. We recently published a large series of adults with Chiari I malformation who saw improvement following extradural decompression [34]. Nevertheless, most of the reports of extradural decompression in CIM patients suggest improvement of headaches, syringomyelia, and hydrocephalus symptoms. None have shown the successful resolution of visual and oropharyngeal deficits, such as nystagmus, esotropia,

laryngomalacia, and sleep apnea as in the case of our patient. We have since turned to the extradural splitting technique as first line for patients at our institution because extradural decompression appears to be safer, faster, more cost effective, and have fewer surgical complications.

Conclusion

In summary, CIM is the most common abnormality of the craniovertebral junction, often presenting with visual, oropharyngeal, and cerebellar dysfunctions. Our case serves as an excellent example of successful resolution of CIM symptoms following extradural decompression. While a number of other techniques could be used to treat the CIM symptoms of nystagmus, esotropia, and sleep apnea, most are highly invasive and associated with numerous complications. Extradural decompression is a less invasive technique with the potential to become first line treatment for patients with CIM.

Consent

Written informed consent was obtained from the patient's family before the procedure.

References

1. Sarnat HB (2008) Disorders of segmentation of the neural tube: Chiari malformations. *Handb Clin Neurol* 87: 89-103.
2. Ventureyra EC, Aziz HA, Vassilyadi M (2003) The role of cine flow MRI in children with Chiari I malformation. *Childs Nerv Syst* 19: 109-113.
3. Oró JJ, Mueller DM (2011) Posterior fossa decompression and reconstruction in adolescents and adults with the Chiari I malformation. *Neurol Res* 33: 261-271.
4. Chauvet D, Carpentier A, George B (2009) Dura splitting decompression in Chiari type 1 malformation: clinical experience and radiological findings. *Neurosurg Rev* 32: 465-470.
5. Baisden J (2012) Controversies in Chiari I malformations. *Surg Neurol Int* 3: S232-237.
6. Speer MC, George TM, Enterline DS, Franklin A, Wolpert CM, et al. (2000) A genetic hypothesis for Chiari I malformation with or without syringomyelia. *Neurosurg Focus* 8: E12.
7. Schanker BD, Walcott BP, Nahed BV, Kahle KT, Li YM, et al. (2011) Familial Chiari malformation: case series. *Neurosurg Focus* 31: E1.
8. Boyles AL, Enterline DS, Hammock PH, Siegel DG, Slifer SH, et al. (2006) Phenotypic definition of Chiari type I malformation coupled with high-density SNP genome screen shows significant evidence for linkage to regions on chromosomes 9 and 15. *Am J Med Genet A* 140: 2776-2785.
9. Markunas CA, Lock E, Soldano K, Cope H, Ding CK, et al. (2014) Identification of Chiari Type I Malformation subtypes using whole genome expression profiles and cranial base morphometrics. *BMC Med Genomics* 7: 39.
10. Albert GW, Menezes AH, Hansen DR, Greenlee JD, Weinstein SL (2010) Chiari malformation Type I in children younger than age 6 years: presentation and surgical outcome. *J Neurosurg Pediatr* 5: 554-561.
11. Al-Awami A, Flanders ME, Andermann F, Polomeno RC (2005) Resolution of periodic alternating nystagmus after decompression for Chiari malformation. *Can J Ophthalmol* 40: 778-780.
12. Liebenberg WA, Georges H, Demetriades AK, Hardwidge C (2005) Does posterior fossa decompression improve oculomotor and vestibulo-ocular manifestations in Chiari I malformation? *Acta Neurochir (Wien)* 147: 1239-1240.
13. Defoort-Dhellemmes S, Denion E, Arndt CF, Bouvet-Drumare I, Hache JC, et al. (2002) Resolution of acute acquired comitant esotropia after suboccipital decompression for Chiari I malformation. *Am J Ophthalmol* 133: 723-725.
14. Dauvilliers Y, Stal V, Abril B, Coubes P, Bobin S, et al. (2007) Chiari malformation and sleep related breathing disorders. *J Neurol Neurosurg Psychiatry* 78: 1344-1348.
15. Henriques-Filho PS, Pratesi R (2008) Sleep apnea and REM sleep behavior disorder in patients with Chiari malformations. *Arq Neuropsiquiatr* 66: 344-349.
16. Khatwa U, Ramgopal S, Mylavarapu A, Prabhu SP, Smith E, et al. (2013) MRI findings and sleep apnea in children with Chiari I malformation. *Pediatr Neurol* 48: 299-307.
17. Losurdo A, Dittoni S, Testani E, Di Blasi C, Scarano E, et al. (2013) Sleep disordered breathing in children and adolescents with Chiari malformation type I. *J Clin Sleep Med* 9: 371-377.
18. Chartier A, Martinot A, Dhellemmes P, Vallée L, Lamblin MD, et al. (2002) [Chiari type I malformation in children: presentation in 34 cases]. *Arch Pediatr* 9: 789-796.
19. Tran K, Hukins CA (2011) Obstructive and central sleep apnoea in Arnold-Chiari malformation: resolution following surgical decompression. *Sleep Breath* 15: 611-613.
20. Botelho RV, Bittencourt LR, Rotta JM, Tufik S (2010) The effects of posterior fossa decompressive surgery in adult patients with Chiari malformation and sleep apnea. *J Neurosurg* 112: 800-807.
21. Baxstrom CR (2009) Nonsurgical treatment for esotropia secondary to Arnold-Chiari I malformation: A case report. *Optometry* 80: 472-478.
22. Biousse V, Newman NJ, Petermann SH, Lambert SR (2000) Isolated comitant esotropia and Chiari I malformation. *Am J Ophthalmol* 130: 216-220.
23. Firth AY, Burke JP (2007) Botulinum toxin for the treatment of acute-onset comitant esotropia in Chiari I malformation. *Br J Ophthalmol* 91: 1094.
24. Hentschel SJ, Yen KG, Lang FF (2005) Chiari I malformation and acute acquired comitant esotropia: case report and review of the literature. *J Neurosurg* 102: 407-412.
25. Imes RK, Quinn TA (2001) Acute comitant esotropia in Chiari I malformation. *Ophthalmology* 108: 834.
26. Lewis AR, Kline LB, Sharpe JA (1996) Acquired esotropia due to Arnold-Chiari I malformation. *J Neuroophthalmol* 16: 49-54.
27. Passo M, Shults WT, Talbot T, Palmer EA (1984) Acquired esotropia. A manifestation of Chiari I malformation. *J Clin Neuroophthalmol* 4: 151-154.
28. Pokharel D, Siatkowski RM (2004) Progressive cerebellar tonsillar herniation with recurrent divergence insufficiency esotropia. *J AAPOS* 8: 286-287.
29. Rech L, Mehta V, Mac Donald IM (2013) Esotropia and Chiari I malformation: report of a case and review of outcomes of posterior fossa decompression and strabismus surgery. *Can J Ophthalmol* 48: e90-92.
30. Weeks CL, Hamed LM (1999) Treatment of acute comitant esotropia in Chiari I malformation. *Ophthalmology* 106: 2368-2371.
31. Arnautovic A, Splavski B, Boop FA, Arnautovic KI (2015) Pediatric and adult Chiari malformation Type I surgical series 1965-2013: a review of demographics, operative treatment, and outcomes. *J Neurosurg Pediatr* 15: 161-177.
32. Litvack ZN, Lindsay RA, Selden NR (2013) Dura splitting decompression for Chiari I malformation in pediatric patients: clinical outcomes, healthcare costs, and resource utilization. *Neurosurgery*. 72: 922-928
33. Chotai S, Medhkour A (2014) Surgical outcomes after posterior fossa decompression with and without duraplasty in Chiari malformation-I. *Clin Neurol Neurosurg* 125: 182-188.
34. Quon J, Grant R, Diluna M (2014) Multimodal evaluation of CSF dynamics following extradural Chiari I decompression. *Journal of Neurosurgery: Spine*, accepted.
35. Genitori L, Peretta P, Nurisso C, Macinante L, Mussa F (2000) Chiari type I anomalies in children and adolescents: minimally invasive management in a series of 53 cases. *Childs Nerv Syst* 16: 707-718.

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36. Isu T, Sasaki H, Takamura H, Kobayashi N (1993) Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. *Neurosurgery* 33: 845-849.
37. Valentini L, Visintini S, Saletti V, Chiapparini L, Estienne M, et al. (2011) Treatment for Chiari I malformation (CIM): analysis of a pediatric surgical series. *Neurol Sci* 32 Suppl 3: S321-324.
38. Caldarelli M, Novegno F, Vassimi L, Romani R, Tamburrini G, et al. (2007) The role of limited posterior fossa craniectomy in the surgical treatment of Chiari malformation Type I: experience with a pediatric series. *J Neurosurg* 106: 187-195.