Child's Nervous System (2019) 35:1889–1894
https://doi.org/10.1007/s00381-019-04181-5

SPECIAL ANNUAL ISSUE

Management opinions from different centers (Rio de Janeiro)

José Francisco M. Salomão[1] · Tatiana Protzenko Cervante[1] · Antonio Rosa Bellas[1]

Received: 2 April 2019 / Accepted: 28 April 2019 / Published online: 15 May 2019

Abstract

Introduction The surgical treatment of Chiari type 1 (CM1) malformation is controversial and depends largely on the preference of the surgeon. The evolution of neuroimaging resulted in an increased number of asymptomatic patients incidentally diagnosed.
Purpose To study retrospectively a population of 24 symptomatic patients with CM1 operated between 1999 and 2017 in which intraoperative ultrasonography (IOUS)–assisted posterior fossa-C1 decompression was used to decide whether the dura mater should be opened (CVD+) or not (CVD).
Results Most of the patients complained of headache or neck pain, 15 had hydrosyringomyelia and 14 had some spinal cord involvement. Patients were categorized in improved, unchanged, or worse according the preoperative signs and symptoms. Overall, 19 patients improved, 3 deteriorated, and 2 remained unchanged. Among these, 4 out 5 had syringohydromyelia.
Conclusions IOUS-assisted posterior fossa-C1 decompression is our preferred option to treat CM1. Children submitted to intradural procedures, initially or subsequently, had increased postoperative complications. CSF fistula or pseudomeningocele was the major cause of complication. The final result seems to correlate with the preoperative neurological status.

Keywords Chiari 1 malformation · Hydrosyryngomyelia · Craniovertebral decompression

Introduction

Chiari I malformation (CM1) is the ectopy of the cerebellar tonsils through the foramen magnum (FM), congenital or acquired. Aside the small posterior fossa, syringohydromyelia, scoliosis, hydrocephalus, craniosynostosis, craniovertebral junction (CVJ) anomalies such as atlas assimilation, odontoid retroflexion, basilar impression, and other may be part of the malformation [1–5].

The treatment of CM1 remains controversial and options depend largely on the preference of the surgeon. Additionally, with evolution of neuroimaging, the amount of asymptomatic patients increased and further clarification on management and natural history is needed.

This section deals with our current management of the CM1. The so-called Chiari 0 was not included and patients identified as Chiari 1.5 were considered as CM1 [6].

✉ José Francisco M. Salomão
jfsalomao@gmail.com

[1] Pediatric Neurosurgery Division, Fernandes Figueira Institute, Oswaldo Cruz Foundation (IFF-Fiocruz), Av. Rui Barbosa 716, Rio de Janeiro, RJ CEP 22.250-020, Brazil

Signs and symptoms considered typical

a. Posterior headache or neck pain usually lasting less than 5 min and aggravated by Valsalva maneuver such as cough, sneezing, laughing, and screaming [5, 7–9]. Neonates and infants may present with irritability and inconsolable crying.
b. Signs of involvement of brain stem, cerebellum, and cranial nerves such as oropharyngeal dysfunction, sensory and motor deficits, impairment of fine motor control, ataxia, nystagmus, vertigo, weakness, tinnitus, hearing loss, vocal cord paralysis, and tongue atrophy may be found. Sleep-disordered breathing such as snoring and central, obstructive, or mixed sleep apnea may be seen. Polysomnography is an important and useful tool in the follow-up and decision-making process of borderline subjects [10–13].
c. Scoliosis, syringohydromyelia, and spinal cord dysfunction. Scoliosis and syringohydromyelia are associated in up to 70% of the cases of CM1 and these patients are prone to progressive neurological dysfunction [8, 12–14]. Syringomyelia may cause varied spinal cord involvement: motor deficit, spasticity, hyperreflexia, suspended thermoanalgesia, and spine central canal syndrome. Segmental atrophy, fasciculation, and trophic changes may be present [15].

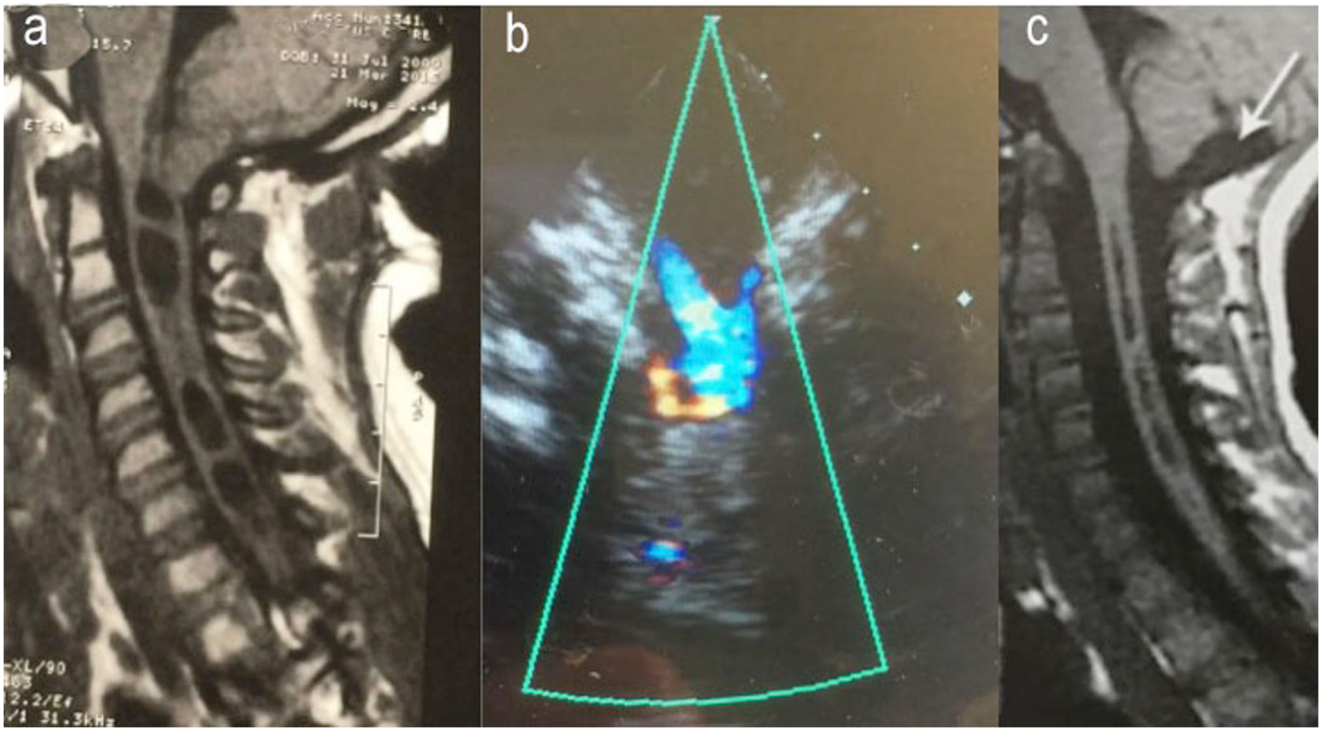

Fig. 1 Tailored posterior fossa-C1 procedure. **a** MRI showing CM1 and hydrosyringomyelia. **b** IOUS detecting flow across the FM. **c** Postoperative MRI showing reduction of the cavities and formation of a retrotonsilar space

d. Macrocrania, ventriculomegaly, and hydrocephalus may be found, especially infants and toddlers [16–18].

Criteria for surgical treatment

There is no agreement regarding criteria for surgical treatment. At our institution, conservative treatment is offered to asymptomatic patients without syringohydromyelia or with cavitation less than 2–3 mm. They are kept under surveillance as well as those with mild or sporadic headache, since data on progression and natural history of these patients are lacking [3, 19–22].

Surgical treatment is offered to the following:

a. Asymptomatic patients with large syrinxes due to its potential devastating effects on the growing spine [17, 22].

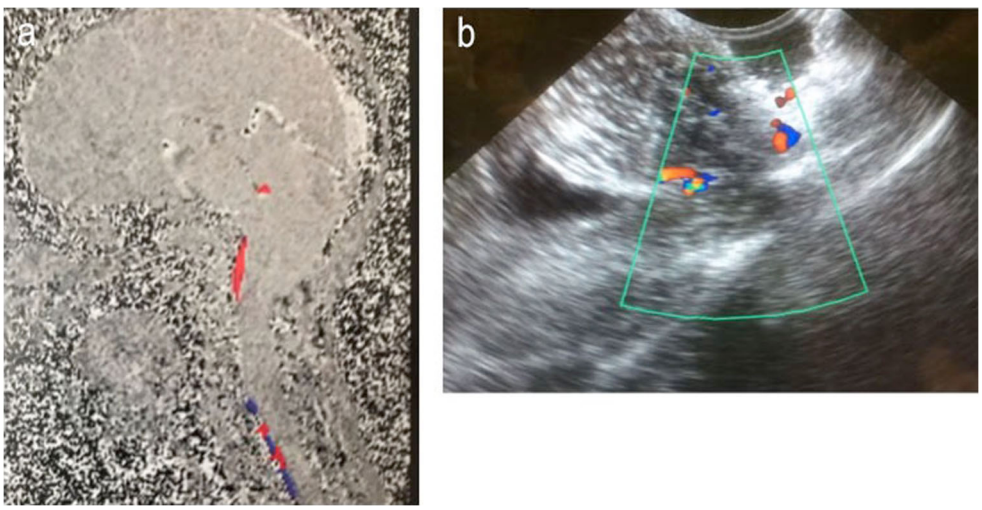

Fig. 2 **a** Preoperative flow MRI showing obstruction at the CVJ. **b** IOUS fails to detect flow across the FM. **c** View through the surgical microscope showing a crowded posterior fossa with tonsils bellow C1. **d** After tonsils shinkrage the cervicomedullary junction (*) and the obex (arrow head) are seen. **e** Postoperative MRI showing flow across the FM

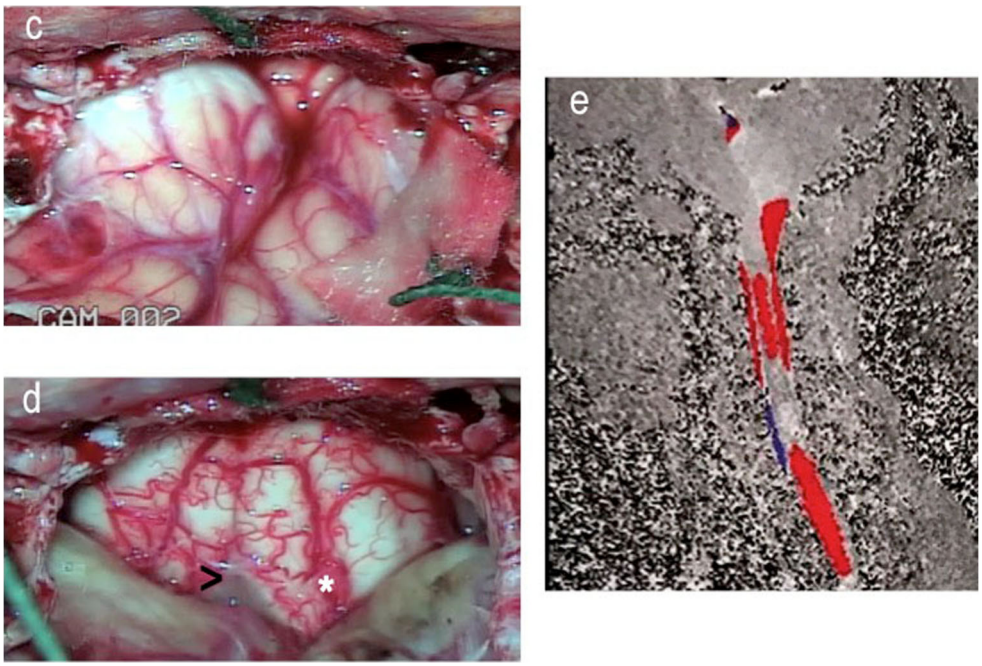

Fig. 2 (continued)

b. Patients with disabling or severe typical headache, with or without syringohydromyelia.
c. Patients with related neurological signs and symptoms.
d. Patients with progressive scoliosis with or without syrinx.

Surgical procedure commonly used

We prefer the intraoperative ultrasonography (IOUS)–assisted posterior fossa-C1 decompression. The aim of this surgery is, whenever possible, to decompress the CVJ without opening the dura (CVD).

Table 1 Demography, signs, symptoms, and image findings in 24 patients

Demography	
Males: 14; Females: 10	
Age: 4 months - 18 years	
Mean: 8.6 year; Median: 9 years	
Signs and symptoms	
Headache/neck pain	16
Spinal cord involvement	14
Posterior fossa, cerebellum, cranial nerves	12
Scoliosis	12
Fenotipical characteristics	7
Syringomyelia	15
Skeletal anomalies	13

a. Patients are positioned prone with the neck discreetly flexed and the head slightly elevated from the trunk. Intraoperative neurophysiological monitoring (IONM) is important at this step.
b. Occipital craniectomy is limited to 3 cm around the FM that is widely opened. The posterior arch of C1 is always resected.
c. Thickened fibrous bands at the CVJ are removed.
d. If IOUS shows flow across the CVJ, the procedure is terminated after osseous decompression (Fig. 1).
e. Otherwise, the dura mater is opened in a Y fashion and the tonsils reduced with bipolar coagulation (Fig. 2). The dura is then expanded with a patch usually harvested from the pericranium and closed in a watertight fashion. A stitch is applied to the graft and tied to the superficial aponeurosis, trying to expand the retrotonsilar space.

When results were considered good

The results were assessed according the perception of the examiners and information given by the patients and/or their parents. Thus, the results were categorized in improved (or resolved), unchanged, or worse compared with the preoperative status.

Surgical complications CSF fistula or pseudomeningocele was reported in 4 patients and 2 required reoperation. One had aseptic meningitis and another a transient hoarseness and

Table 2 Details of the surgical procedures, reoperations and complications

	Surgery	Syrinx	Reoperation	Complication	Result	
1	VPS/CVD				Improved	
2	CVD	YES			Improved	
3	CVD	YES			Improved	
4	CVD	YES	CVD+	Deterioration	Worse	
5	CVD/ODT/PA	YES			Improved	
6	CVD+		DURAPL/PA	CSFF + HC	Improved	
7	CVD	YES			Improved	
8	VPS/CVD	YES			Improved	
9	CVD+	YES		PMEN	Improved	
10	CVD	YES			Improved	
11	CVD	YES			Improved	
12	CVD+		DURAPL/VPS	CSFF	Unchanged	
13	CVD/PA	YES			Unchanged	
14	CVD+	YES	CVD+/PA/SPS	Deterioration	Worse*	
15	CVD	YES			Improved	
16	CVD+	YES		PMEN	Improved	
17	CVD				Improved	
18	CVD				Improved	
19	CVD				Improved	
20	VPS/CVD				Improved	
21	CVD	YES			Improved	
22	CVD				Improved	
23	CVD		CVD+		Improved	
24	CVD+	YES			Worse**	Worse**

AA, atlas assimilation; *BI*, basilar impression; *CVD*, craniovertebral decompression; *CVD+*, craniovertebral decompression plus dura opening and tonsil coagulation; *CSFF*, CSF fistula; *DURAPL*, duraplasty; *KF*, Klippel-Feil; *OR*, odontoid retroflexion; *PA*, posterior arthrodesis; *PMEN*, pseudomeningocele; *VPS*, ventriculoperitoneal shunt; *SPS*, syringopleural shunt

*Arachnoiditis

**Initially operated elsewhere (syringostomy)

swallowing difficulties. There was one superficial wound infection.

Presentation of the series

We retrospectively studied the records of 24 symptomatic patients with CM1 operated between 1999 and 2017. We excluded 8 patients operated in the preceding years plus another 5 during the study period, due to insufficient data recording. Patients with tonsilar ectopia related to craniosynostosis, cerebral spinal fluid (CSF) diversions, and space-occupaying lesions were also excluded. There was a predominance of males ($n = 14$) and the age at presentation ranged from 4 months to 18 years, with mean age of 8.6 years and median of 9 years.

Imaging findings All patients had at least one preoperative MRI showing peg-like cerebellar tonsils protruding 5 mm or more bellow the FM. Syringohydromyelia was found in 15 patients. CT scan showed 13 skeletal anomalies of the CVJ and cervical spine in 10 patients (Table 1).

Follow-up and outcomes The minimum follow-up was 1 year, with a mean of 5.9 years. Preoperative ventriculoperitoneal shunt (VPS) was required in 3 patients. In 18, the initial spinal procedure was restricted to CVD. In 5 children, we decided for CVD+ according IOUS findings (Fig. 2). Another one underwent a CVD+ due to a previous isolated syringostomy unsuccessfully performed elsewhere. One patient had a simultaneous posterior arthrodesis (PA) and another one a circumferential surgery including transoral odontoidectomy. Remission or improvement of the signs and symptoms was observed in 19 patients. Five patients deteriorated or remained unchanged. In this context, 3 patients previously submitted to CVD undervent a CVD+. In another, a severe arachnoiditis was found and she underwent sequentially a dural expansion, a ventriculoperitoneal shunt, and finally a syringopleural diversion. The parents of the fifth patient refused any further treatment. None of these patients improved and they all had in common a severe preoperative neurological deficit. Details concerning the results of this series are seen in Table 2.

For more than 20 years, we have abandoned extensive posterior fossa decompression and obex plugging in favor of a less invasive tailored posterior fossa-C1 procedure. Extensive craniectomy is not advisable because the target area is limited to the FM-C1 region. Moreover, patients who underwent more extensive procedures may have a longer hospital stay and late deterioration due to cerebellar ptosis. IONM is important for positioning at the surgical table, although its

value is questionable during surgical manipulation [23–25]. The posterior arch of C1 is always resected and laminectomy below this level is not needed. IOUS is an important tool to estimate CSF flow at the CVJ [12, 18, 19, 26–29].

In patients with progressive hydrocephalus, we consider shunt diversion or ETV as initial treatment. Craniocervical malformations such as odontoid retroflexion, occipitalization of the atlas, basilar impression, Klippel-Feil syndrome, and other predispose to CVJ instability and its incidence ranges according different series [4, 12, 30, 31]. In this scenario, a few of our patients needed simultaneous surgical stabilization.

It is also debatable how much the surgical technique impacts the outcome of patients with syringohydromyelia [15, 16, 18, 27, 32]. In our series, we observed that children submitted to intradural procedures, initially or subsequently, had increased postoperative complications. However, it should also be considered that these had a more severe and long-standing preoperative deficit.

The presented series has some limitations. Firstly, data collected previously to the last 20 years were inaccurate and these patients were excluded, decreasing the size of the sample. Secondly, a more effective outcome scale [33] was not applied due to the retrospective design of this study. The "gestalt method" applied comparing preoperative and postoperative resulted in some bias. Moreover, pain improvement is usually reported more quickly, while neurological deficits may take longer to be evaluated. Scoliosis and CVJ instability may eventually require additional interventions and this may further impair the results here presented.

In our opinion, IOUS-assisted tailored posterior fossa-C1 decompression is a good treatment option for CM1, especially when intradural manipulation is not needed. Children submitted to intradural procedures, initially or subsequently, had increased postoperative complications. The final result seems to correlate with the preoperative neurological status.

Compliance with ethical standards

Conflict of interest The authors declare no conflict of interest.

References

1. Krieger MD, Mccomb JG, Ml L (1999) Toward a simpler surgical management of Chiari I malformation in a pediatric population. Pediatr Neurosurg 30:113–121. https://doi.org/10.1159/000028777
2. Galarza M, Martínez-Lage JF, Ham S, Sood S (2010) Cerebral anomalies and Chiari type 1 malformation. Pediatr Neurosurg 46: 442–449. https://doi.org/10.1159/000327220
3. Aitken LA, Linda CE, Sidney S, Gupta N, Barkovich AJ, Sorel M et al (2009) Chiari type I malformation in a pediatric population. Pediatr Neurol 40:449–454. https://doi.org/10.1016/j.pediatrneurol.2009.01.003
4. 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. https://doi.org/10.3171/2014.10.SPINE14176
5. Poretti A, Ashmawy AR, Garzon-Muvdi AT, Jallo GI, Huisman TA, Raybaud C (2016) Chiari type 1 deformity in children : pathogenetic , clinical , neuroimaging , and management aspects. Neuropediatrics 47:293–307. https://doi.org/10.1055/s-0036-1584563
6. Azahraa Haddad F, Qaisi I, Joudeh N, Dajani H, Jumah F, Elmashala A, Adeeb N, Chern JJ, Tubbs RS (2018) The newer classifications of the chiari malformations with clarifications: an anatomical review. Clin Anat 31:314–322. https://doi.org/10.1002/ca.23051
7. International Headache Society (2018) The international classification of headache disorders, 3rd edition. Cephalalgia 38(1):1–211. https://doi.org/10.1177/0333102417738202
8. Pindrik J, Johnston JM (2015) Clinical presentation of Chiari I malformation and syringomyelia in children. Neurosurg Clin N Am 26:509–514. https://doi.org/10.1016/j.nec.2015.06.004
9. Mehta A, Chilakamarri P, Zubair A, Kuruvilla D (2018) Chiari headache. Curr Pain Headache Rep 22(49):49. https://doi.org/10.1007/s11916-018-0702-8
10. Zaffanello M, Sala F, Sacchetto L, Gasperi E, Piacentini G, Sala F (2017) Evaluation of the central sleep apnea in asymptomatic children with Chiari 1 malformation : an open question. Childs Nerv Syst 33:829–832. https://doi.org/10.1007/s00381-017-3399-9
11. Losurdo A, Testani E, Scarano E, Massimi L, Della MG (2013) What causes sleep-disordered breathing in Chiari I malformation ? Comment on: "MRI findings and sleep apnea in children with Chiari I malformation". Pediatr Neurol 49:11–13. https://doi.org/10.1016/j.pediatrneurol.2013.04.016
12. Milhorat TH (2003) Tailored operative technique for Chiari type I. Neurosurgery 53:899–906
13. Godzik J, Dardas A, Kelly MP, Holekamp TF, Lenke LG, Smyth MD, Park TS, Leonard JR, Limbrick DD (2016) Comparison of spinal deformity in children with Chiari I malformation with and without syringomyelia: matched cohort study. Eur Spine J 25:619–626. https://doi.org/10.1007/s00586-015-4011-1
14. Eule JM, Erickson MA, Brien MFO, Handler M (2002) Chiari I malformation associated with syringomyelia and scoliosis a twenty-year review of surgical and nonsurgical treatment in a pediatric population. Spine 27:1451–1455
15. Navarro R, Olavarria G, Seshadri R, Gonzales-Portillo G, McLone DG, Tomita T (2004) Surgical results of posterior fossa decompression for patients with Chiari I malformation. Childs Nerv Syst 20: 349–356. https://doi.org/10.1007/s00381-003-0883-1
16. Lee A, Yarbrough CK, Greenberg JK, Barber J, Limbrick DD, Smyth MD (2014) Comparison of posterior fossa decompression with or without duraplasty in children with type I Chiari malformation. Childs Nerv Syst 30:1419–1424. https://doi.org/10.1007/s00381-014-2424-5
17. Brockmeyer DL, Spader HS (2015) Complex Chiari malformations in children. Diagnosis and Management Neurosurg Clin N Am 26: 555–560. https://doi.org/10.1016/j.nec.2015.06.002
18. Entezami P, Gooch MR, Poggi J, Perloff E, Dupin M, Adamo MA (2019) Current management of pediatric chiari type 1 malformations. Clin Neurol Neurosurg 176:122–126. https://doi.org/10.1016/j.clineuro.2018.12.007
19. Macinante GL (2000) Chiari type I anomalies in children and adolescents : minimally invasive management in a series of 53 cases. Childs Nerv Syst 16:707–718. https://doi.org/10.1007/s003810000338
20. Massimi L, Caldarelli M (2011) Natural history of Chiari type I malformation in children. Neurol Sci 32:275–277. https://doi.org/10.1007/s10072-011-0684-3
21. Schijman E (2004) Steinbok P. International survey on the management of Chiari I malformation and syringomyelia 2004:341–348. https://doi.org/10.1007/s00381-003-0882-2

22. Nishizawa S, Yokoyama T, Yokota N, Tokuyama T, Ohta S (2001) Incidentally identified syringomyelia associated with Chiari I malformations: is early interventional surgery necessary? Neurosurgery 49:637–640; discussion 640-1
23. Sala F, Squintani G, Tramontano V, Coppola A, Gerosa M (2011) Intraoperative neurophysiological monitoring during surgery for Chiari malformations. Neurol Sci 32(Suppl 3):317–319. https://doi.org/10.1007/s10072-011-0688-z
24. Kawasaki Y, Uchida S, Onishi K, Toyokuni M, Okanari K, Fujiki M (2017) Intraoperative neurophysiologic monitoring for prediction of postoperative neurological improvement in a child with Chiari type I malformation. J Craniofac Surg 28:1837–1841. https://doi.org/10.1097/SCS.0000000000003926
25. Barzilai O, Roth J, Korn A, Constantini S (2016) The value of multimodality intraoperative neurophysiological monitoring in treating pediatric Chiari malformation type I. Acta Neurochir 158:335–340. https://doi.org/10.1007/s00701-015-2664-5
26. James HE, Brant A (2002) Treatment of the Chiari malformation with bone decompression without durotomy in children and young adults. Childs Nerv Syst 18:202–206. https://doi.org/10.1007/s00381-002-0574-3
27. Gallo P, Sokol D, Kaliaperumal C, Kandasamy J (2017) Comparison of three different cranio-cervical decompression procedures in children with Chiari malformation type I: does the surgical technique matter? Pediatr Neurosurg 52:289–297. https://doi.org/10.1159/000479327
28. Narenthiran G, Parks C, Pettorini B (2015) Management of Chiari I malformation in children: effectiveness of intra-operative ultrasound for tailoring foramen magnum decompression. Childs Nerv Syst 31:1371–1376. https://doi.org/10.1007/s00381-015-2699-1
29. Pisapia JM, Merkow MB, Brewington D, Henn RE, Sutton LN, Storm PB et al (2017) External validity of the Chiari severity index and outcomes among pediatric Chiari I patients treated with intra- or extra-dural decompression. Childs Nerv Syst 33:313–320. https://doi.org/10.1007/s00381-016-3300-2
30. Klekamp J (2015) Chiari I malformation with and without basilar invagination: a comparative study. Neurosurg Focus 38(4):E12. https://doi.org/10.3171/2015.1.FOCUS14783
31. Menezes AH, Vogel TW (2008) Specific entities affecting the craniocervical region: syndromes affecting the craniocervical junction. Childs Nerv Syst 24:1155–1163. https://doi.org/10.1007/s00381-008-0608-6
32. Valentini L, Visintini S, Saletti V, Solero CL (2011) Treatment for Chiari 1 malformation (CIM): analysis of a pediatric surgical series. Neurol Sci 32(Suppl 3):321–324. https://doi.org/10.1007/s10072-011-0731-0
33. Yarbrough CK, Greenberg JK, Park TS (2015) Clinical outcome measures in Chiari I malformation. Neurosurg Clin N Am 26:533–541. https://doi.org/10.1016/j.nec.2015.06.008