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ORIGINAL ARTICLE

Microvascular decompression for pediatric-onset hemifacial spasm: case series and literature review

Anpei Jia[1] · **Ning Ning Dou**[2] · **Jun Zhong**[3]

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Abstract

Objectives Pediatric hemifacial spasm has been rarely reported in the literature, which contains only 44 cases. Although microvascular decompression (MVD) has been widely regarded as effective therapy for hemifacial spasm, the etiology and surgical treatment of pediatric patients are seldom reported. We report our experience with MVD for pediatric hemifacial spasm patients and review the literature with emphasis on the difference from adults.

Methods This retrospective report included 4 pediatric HFS patients, who underwent MVD in our department between January 2014 and May 2021 and then reviewed all the pediatric hemifacial spasm literature on "pubmed" with emphasis on the clinical data.

Results Our series included 1 boy and 3 girls with an average age of 15.6 ± 3.2 years old; their onset ages were from 7 to 16 years old (11.6 ± 4.3). Three patients achieved immediate excellent outcomes and 1 achieved poor immediately and became good 6 months later. During the operation, all the 4 patients were found compressed by anterior inferior cerebellar artery (AICA). The incidence of pediatric atypical hemifacial spasm patients is 12.5% among the 48 reported cases, which is much higher than adults. Among all the reported 48 cases including ours, the singular artery neurovascular conflictions account for 27/48(56%), the singular vein and combined artery/vein conflictions in 12/48(25%) and the cisternal conflictions in 5/48(10.4%) patients.

Conclusions The etiology of pediatric hemifacial spasm is still neurovascular conflict, of which combined artery/vein and singular venous compression patterns have a higher proportion, which might explain higher incidence of pediatric atypical hemifacial spasm and less favorable postoperative outcome. Sufficient arachnoid release, full exploration and decompression along the facial nerve are necessary, which would help to increase the excellent postoperative cure rate among pediatric patients.

Keywords Pediatric hemifacial spasm · Microvascular decompression · Venous compression · Combined artery/vein compression · Technique

Introduction

Hemifacial spasm (HFS) has a prevalence of approximately 10 per 100,000, which is defined as unilateral, involuntary, irregular clonic or tonic contractions of the facial muscles innervated by the ipsilateral seventh cranial nerve [1, 2]. The high postoperative efficacy of microvascular decompression (MVD) surgery has confirmed the neurovascular conflict theory. HFS is most common in middle-aged and older patients [3–5], which is caused by the elongation and tortuosity of the vascular loop and sagging of the brain accompanying the aging process, however, which could not adequately explain HFS in those pediatric patients, which has been rarely reported in the literature because of its low

✉ Ning Ning Dou
Douningning@XinHuaMed.com.cn

[1] Shidong Hospital, University of Shanghai for Science and Technology, Shanghai 200092, China

[2] Department of Neurosurgery, XinHua Hospital (The Cranial, Nerve Disease Center of Shanghai), Shanghai JiaoTong University School of Medicine, 1665 KongJiang Rd, Shanghai 200092, China

[3] Department of Neurosurgery, XinHua Hospital (The Cranial, Nerve Disease Center of Shanghai), Shanghai JiaoTong University School of Medicine, Shanghai 200092, China

occurrence in childhood and adolescence (Table 1). However, the postoperative outcome of pediatric HFS patients is worse than adults [6–12]. If age is a major determinant of the HFS, there may be clinical differences including etiology, symptom, offending vessels and surgery nuances between the pediatric and elderly HFS patients. Clarification of these questions would help to understand the pathophysiology and improve the cure rate of hemifacial spasm in children. This study was designed to answer the above questions.

Materials and methods

All patients enrolled in this study gave written consent. Here, the 4 HFS patients (≤ 18 years old) who underwent MVD procedures between January 2014 and May 2021 were enrolled in this study. All patients have undergone medical treatment including carbamazepine or oxcarbazepine preoperatively and in vain. Each patient underwent three-dimensional time-of-flight magnetic resonance imaging (3D-TOF MRI) examination before surgery. The medical records including demographics, intraoperative findings, postoperative outcomes and complications were all reviewed. All patients were followed up by telephone interview. The postoperative outcomes were divided into three categories as follows: (1) excellent: complete spasm relief without medication; (2) good: substantial relief but with minimal persistent twitch without medication or complete relief with well-tolerated medication; (3) poor: little or no relief or severe twitch despite medication [13].

Surgical technique

All procedures were performed via a lateral retrosigmoid suboccipital approach, during which a small craniotomy was performed to expose the edges of the sigmoid and transverse sinus. The dura was opened and cerebrospinal fluid was allowed to empty. After careful dissection of the arachnoid membrane and gentle retraction of the flocculus, the root exit zone (REZ) of the facial nerve was observed. Any offending vessel was dissected away from the facial nerve. Several pieces of Teflon sponge were interpositioned between the offending vessel and REZ. During manipulation of the seventh and eighth nerves, brainstem auditory-evoked potentials were closely monitored to assess hearing loss and

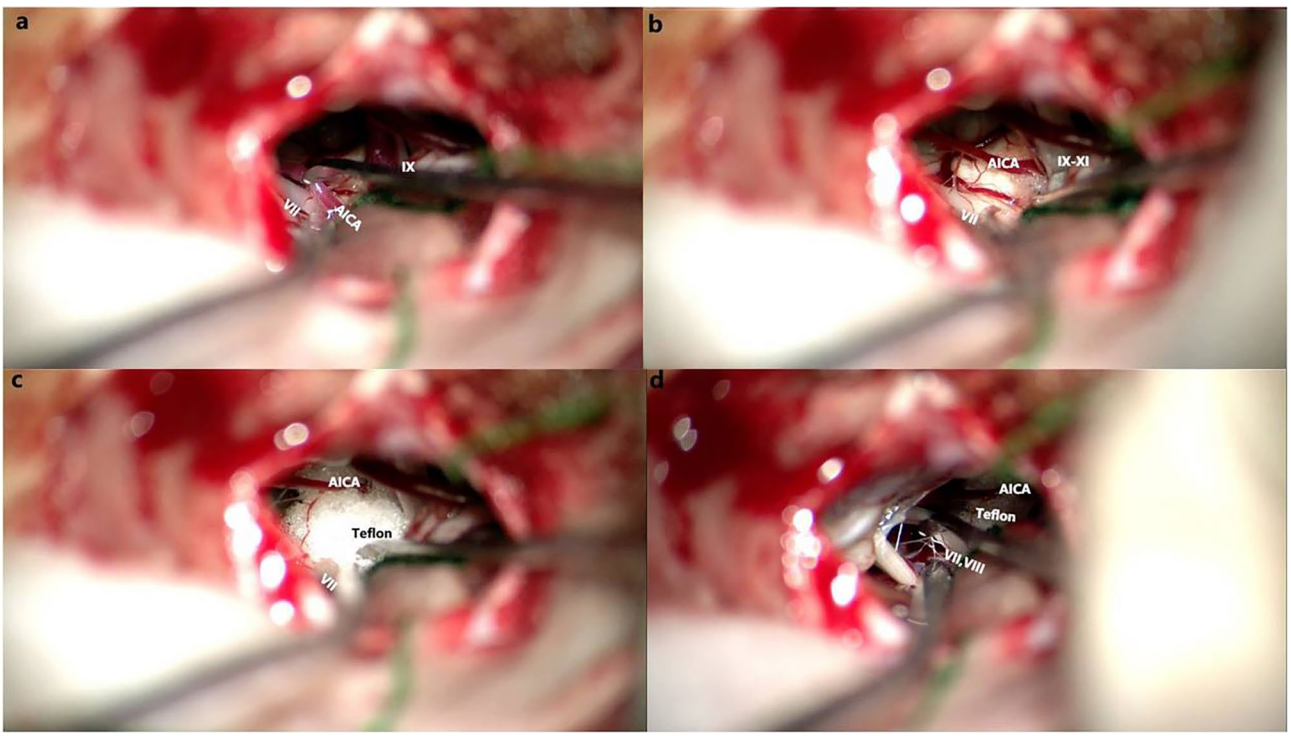

Fig. 1 Intraoperative findings (case 1). (**a**) With dissection of right cranial nerve dissections, the seventh nerve was visualized only compressed by the anterior inferior cerebellar artery at REZ. (**b**) The AICA was mobilized and shredded Teflon was placed between the AICA and the seventh nerve. (**c**) The caudal side of seventh nerve was explored and no other offending vessel was observed. d) We also explored the entire seventh nerve on the cranial side and did not find any other offending vessel

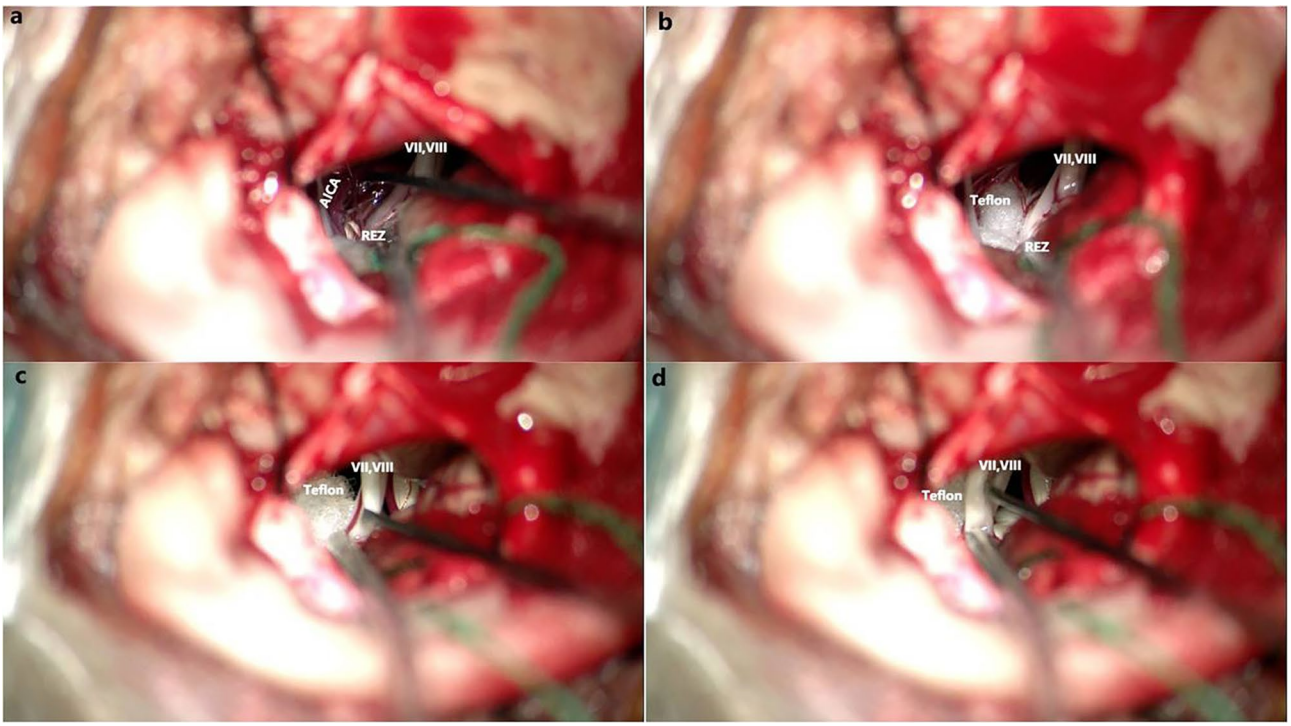

Fig. 2 Intraoperative findings (case 2). (**a**) With dissection of left cranial nerve dissections, the seventh nerve was visualized compressed by the anterior inferior cerebellar artery at REZ and CP. (**b**) The vessel was mobilized and small pieces of shredded Teflon were placed between the anterior inferior cerebellar artery and the nerve to keep the artery free from the nerve. (**c**) The seventh nerve was checked along the cranial side, no other offending vessel was found. (**d**) The caudal side of the seventh nerve was also checked, no offending vessel was found, either. After the surgery, the spasm was totally relieved

facial electromyography to assess the lateral spread response phenomenon, thereby achieving complete decompression. The dura was closed, with several muscle pieces interposed between the interrupted sutures to prevent cerebrospinal fluid leakage [14].

Results

This series included 1 boy and 3 girls with an average age of 15.6 ± 3.2 years old; their onset ages were from 7 to 16 years old(11.6 ± 4.3). Three patients achieved immediate excellent outcomes and 1 achieved poor immediately and became good 6 months later. Through the review, 48 patients including our 4 patients were enrolled in this study. The incidence of pediatric atypical hemifacial spasm was 12.5% among the 48 reported cases, which is much higher than adults. Among all the 48 cases, no serious complications occurred. In our 4 cases, no complications occurred. Among the reported 44 cases, the complications included 1 cerebrospinal fluid leak, 1 chemical meningitis, 3 temporary mild facial paresis, 1 temporary moderate delayed facial paralysis and 1 moderate hearing impairment.

Intraoperative findings in MVDs

During the operation, all the 4 patients were found compressed by anterior inferior cerebellar artery (AICA), 2 at REZ (Fig. 1), 2 at REZ and cisternal part (CP) intraoperatively (Fig. 2 and Table 1). Among all the reported 48 cases including ours, the singular artery conflictions account for 27/48 (56%), the singular vein and combined artery/vein conflictions in 12/48(25%), the cisternal conflictions in 5/48(10.4%) patients (Table 2).

Discussion

HFS is primarily a problem of middle-aged and older patients [3], in which the offending vessels tend to be elongated and redundant. However, such age-related arterial vasculature changes are absent in children. Thus, the etiology of HFS caused by vascular compression in pediatric and young adolescents is still controversial, which has been debated for a long time. It has been 77 years since Ehni and WOLTMAN [15] firstly report pediatric HFS in 1945, since then several theories have

Table 1 Reported cases of pediatric hemifacial spasm treated by MVD surgery

Author	Year	symptom	cases	Offending vessel		Re-occurrence	Immediate excellent Outcome	Complication
				cases	Vessel + site			
Jho and Jannetta [6]	1987	5ty 2aty	7	3	vein + REZ	2 times in 1 case	42.9%	28.6%
				2	artery + REZ			
				1	arteriole + CP			
				1	multiple artery + REZ			
Milani et al. [7]	1991	aty	1	1	artery + REZ	NA	100%	0
Kobata et al. [8]	1995	ty	2	2	artery + REZ	0	100%	0
Levy et al. [9]	1997	ty	12	2	artery + REZ	2	66.7%	25%
				3	vein + REZ			
				5	(artery + vein) + REZ			
				2	multiple artery + REZ			
Feng et al. [10]	2011	4ty 1aty	5	3	artery + REZ	0	75%	0
				1	artery + REZ + CP			
				1	(artery + vein) + REZ			
Liang et al. [11]	2014	16ty	16	10	artery + REZ	0	75%	12.5%
				4	multiple artery + REZ			
				1	petrous bone crest			
				1	adhesion of the arachnoid membrane			
Vasquez et al. [12]	2016	aty	1	1	artery + CP	1	100%	0
This study	2021	3ty 1at	4	2	artery + REZ	0	75%	0
				2	artery + REZ + CP			

REZ; root exit zone, *ty*: typical, *aty*: atypical, *CP*: cisternal portion, *NA*: not available

been raised. The anatomical variations of the vessels and thickening of the arachnoid membrane at the cerebellopontine angle were both considered be the main cause possibly [6–12]. The thickened-arachnoid theory could explain the neuro-vascular theory despite the absence of atherosclerotic changes, which was adhered to the arteries and nerves. We all know that the arachnoidal thickening could be caused by encephalitis, meningitis or subarachnoid hemorrhage. However, it is unusual in the pediatric HFS patients according to the literature [6–12]. Although the other reasons such as trauma, CPA occupation, infections and vascular abnormalities could also cause HFS, the neurovascular confliction is still the main reason [16] (Table 1). Among our cases, we questioned every patient and found no infection or trauma.

We truly find some difference in clinical symptoms and surgery details between pediatric and adult HFS patients. According to our study and literature review, we had two valuable findings. Although the limited reported cases, we found a higher incidence of atypical hemifacial spasm than adults, whose incidence rate was reported around 1.3% [17]. However, the incidence of pediatric atypical hemifacial spasm is 12.5% among the 48 reported cases. We guess it might be related to the different offending vessel and neuro-vascular confliction on site, for the reason that the incidence of alone/combined venous compression was much higher than adults, which was 25% of the pediatric hemifacial patients. The incidence of cistern neurovascular confliction was also higher, which was 10.4%.

Table 2 Features of pediatric hemifacial spasm patients

Case	Onset/surgery age(years)	Sex	Symptom	Offending vessel/site	Immediate outcome	Complication
1	7/18	M	ty	AICA/REZ	Excellent	None
2	11/17	F	aty	AICA/REZ + CP	Fair	None
3	16/17	F	ty	AICA/REZ + CP	Excellent	None
4	16/18	F	ty	AICA/REZ	Excellent	None

F: female, *M*: male, *ty*: typical, *aty*: atypical, *CP*: cistern portion of facial nerve, *REZ*: root exit zone

Through our study, we believe that two factors including crowed cerebellopontine angle (CPA) and easy attrition of the neurovascular interfaces may play important roles in the young HFS patients. Our 3D-TOF MRI delineated a crowded CPA space in the posterior fossa, which increased the chance of neurovascular confliction. Our study regarding the mechanism of HFS demonstrated that attrition of the neurovascular interfaces is necessary for HFS. We believe that the precondition for generating this hyperexcitability is the attrition of the neurovascular interfaces due to mutual friction with pulsation, which is the hinged between the sympathetic nerves in the offending artery wall and the demyelinated nerve root in close contact with each other [18].

The MVD surgery in pediatric HFS patients needs more skills because of the narrow operative space and complex vascular compression. During the operation, excessive stretching of the cerebellum could easily increase the incidence of postoperative complications. A caudorostral approach seems to be an alternative to reduce traction and increase exposure. Combined artery/vein and singular venous compression patterns play an important role in the pediatric patients. The postoperative outcome of pediatric hemifacial spasm seemed to be less favorable because of the different conflict sites and patterns from adults. We should pay more attention to venous and combined vascular decompression during the MVD operation.

Conclusion

The etiology of pediatric hemifacial spasm is still neurovascular conflict, of which combined artery/vein and singular venous compression patterns have a higher proportion, which might explain higher incidence of pediatric atypical hemifacial spasm and less favorable postoperative outcome. Sufficient arachnoid release, full exploration and decompression along the facial nerve are necessary, which would help to increase the excellent postoperative cure rate among pediatric patients.

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Declarations

Conflict of interest No conflict of interest.

References

1. Mizobuchi Y, Nagahiro S, Kondo A, Arita K, Date I, Fujii Y, Fujimaki T, Hanaya R, Hasegawa M, Hatayama T, Hongo K, Inoue T, Kasuya H, Kobayashi M, Kohmura E, Matsushima T, Masuoka J, Morita A, Munemoto S, Nishizawa S, Okayama Y, Sato K, Shigeno T, Shimano H, Takeshima H, Tanabe H, Yamakami I (2021) Prospective, Multicenter Clinical Study of Microvascular Decompression for Hemifacial Spasm. Neurosurgery 88(846–854):8. https://doi.org/10.1093/neuros/nyaa1549
2. Al Menabbawy A, Refaee EE, Elwy R, Salem AA, Lehmann S, Vollmer M, Matthes M, Fleck S, Baldauf J, Schroeder HWS (2022) A multivariable prediction model for recovery patterns and time course of symptoms improvement in hemifacial spasm following microvascular decompression. Acta Neurochir 1:022–05133
3. Jeon CJ, Kong DS, Lee JA, Park K (2010) The efficacy and safety of microvascular decompression for hemifacial spasm in elderly patients. J Korean Neurosurg Soc 47(442–445):4. https://doi.org/10.3340/jkns.2010.3347.3346.3442 (Epub 2010 Jun 3330)
4. Jo KW, Kong DS, Park K (2013) Microvascular decompression for hemifacial spasm: long-term outcome and prognostic factors, with emphasis on delayed cure. Neurosurg Rev 36: 297–301; discussion 301–292. https://doi.org/10.1007/s10143-10012-10420-10143. Epub 12012 Sep 10142
5. Li J, Lyu L, Chen C, Yin S, Jiang S, Zhou P (2022) The outcome of microvascular decompression for hemifacial spasm: a systematic review and meta-analysis. Neurosurg Rev 20:022–01739
6. Jho HD, Jannetta PJ (1987) Hemifacial spasm in young people treated with microvascular decompression of the facial nerve. Neurosurgery 20(767–770):7. https://doi.org/10.1227/00006123-198705000-198700015
7. Milani N, Scaioli V, Giombini S, Grisoli M, Angelini L (1991) Hemifacial spasm in a child. Childs Nerv Syst 7(466–468):4. https://doi.org/10.1007/BF00263191
8. Kobata H, Kondo A, Kinuta Y, Iwasaki K, Nishioka T, Hasegawa K (1995) Hemifacial spasm in childhood and adolescence. Neurosurgery 36(710–714):7. https://doi.org/10.1227/00006123-199504000-199500011
9. Levy EI, Resnick DK, Jannetta PJ, Lovely T, Bissonette DJ (1997) Pediatric hemifacial spasm: the efficacy of microvascular decompression. Pediatr Neurosurg 27(238–241):2. https://doi.org/10.1159/000121260
10. Feng B, Zheng X, Zhang W, Yang M, Tang Y, Zhong J, Hua X, Ying T, Li S (2011) Surgical treatment of pediatric hemifacial spasm patients. Acta Neurochir (Wien) 153: 1031–1035; discussion 1035. https://doi.org/10.1007/s00701-00011-00956-y. Epub 02011 Feb 00706
11. Liang J, Guo Z, Zhang L, Yu Y (2014) Adolescent-onset idiopathic hemifacial spasm. Neurol India 62(175–177):1. https://doi.org/10.4103/0028-3886.132367
12. Vasquez PC, Torres AR, Silvera VM, Scott RM (2016) Hemifacial Spasm in a Child Treated With Microvascular Decompression of the Facial Nerve. Pediatr Neurol 57:84–87.: https://doi.org/10.1016/j.pediatrneurol.2016.1001.1007. Epub 2016 Jan 1020
13. Dou NN, Zhong J, Xia L, Li ST (2022) Microvascular decompression for pediatric onset trigeminal neuralgia: patterns and variation. Childs Nerv Syst 15:021–05432
14. Dou NN, Zhong J, Liu MX, Xia L, Sun H, Li B, Li ST (2016) Teflon Might Be a Factor Accounting for a Failed Microvascular Decompression in Hemifacial Spasm: A Technical Note. Stereotact Funct Neurosurg 94(154–158):1. https://doi.org/10.1159/000446192 (Epub 000442016 Jun 000446192)
15. Ehni G, WOLTMAN HW (1945) Hemifacial spasm: review of one hundred and six cases. Arch Neurol Psychiatry 53:205
16. Cai Y, Ge M, Qi X, Sun H, Zhang D (2019) Ocular Dyskinesia and Hemifacial Spasm Secondary to Fourth Ventricular Tumor: Report of 4 Cases and Review of the Literature. Pediatr Neurosurg 54(359–366):3. https://doi.org/10.1159/000501915 (Epub 000502019 Aug 000501922)

17. Ryu H, Yamamoto S, Miyamoto T (1998) Atypical hemifacial spasm. Acta Neurochir (Wien) 140(1173–1176):11. https://doi.org/10.1007/s007010050233
18. Dou NN, Zhong J, Zhou QM, Zhu J, Wang YN, Xia L, Yang XS, Ying TT, Zheng XS, Li ST (2015) The mechanism of hemifacial spasm: a new understanding of the offending artery. Neurol Res 37(184–188):1. https://doi.org/10.1179/1743132814Y.0000000424 (Epub 1743132014 Jul 1743132818)