Hemifacial Spasm

A Comprehensive Guide

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Editors

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ISBN 978-981-15-5416-2 ISBN 978-981-15-5417-9 (eBook)
https://doi.org/10.1007/978-981-15-5417-9

This Springer imprint is published by the registered company Springer Nature Singapore Pte Ltd.
The registered company address is: 152 Beach Road, #21-01/04 Gateway East, Singapore 189721, Singapore

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Overview of Hemifacial Spasm

Jae Sung Park

Definition of Hemifacial Spasm

The term hemifacial spasm (HFS) is self-explanatory: contractions on one side of the face. More specifically, the clinical term HFS refers to involuntary facial contractions that are unilateral, irregular, and tonic or clonic. The twitches usually start with the periorbital muscles, and then they can spread to perinasal, perioral, zygomaticus, and platysma muscles [1]. The diagnosis of HFS is mainly based on clinical history and physical examination, although adjunctive use of electromyographic (EMG) and radiological evaluation methods is commonly acknowledged. Conditions that need to be differentiated from HFS include blepharospasm, facial myokymia, and post-facial palsy synkinesis, etc.

History

Esmail Jorjani (1042–1137), a Persian physician, described syndromes that were probably consistent with trigeminal neuralgia, HFS, and Bell's palsy in his book *Treasure of the Khawarazm Shah* [2]. Also, he implicated an artery–nerve conflict as an etiology of trigeminal neuralgia. In the more modern period, the prototype of HFS was described by Schultze in 1875, when a vertebral artery aneurysm was found to compress the seventh nerve [3]. One of the first descriptions on HFS with a picture of the patient was provided by Edouard Brissaud in 1893 [4]. In 1905, Babinski described a phenomenon called "other Babinski sign" that referred to a paradoxical synkinesis in HFS patients, and this is typically observed in HFS, but not in blepharospasm patients [5, 6]. A modern-day concept of vascular compression syndrome that included trigeminal neuralgia, HFS, and glossopharyngeal neuralgia was introduced by McKenzie in 1936. Based on its pathophysiological background, vascular decompression for HFS was first introduced by Gardner in 1962, following which, a more modern technique with a minimal approach, i.e., microvascular decompression (MVD) via retrosigmoid craniotomy, was first performed by Bremond in 1974 [7, 8]. The current concept of pathophysiology and the surgical treatment of HFS was established and popularized by Jannetta, and it started with his article in 1975, titled as "Neurovascular cross-compression in patients with hyperactive dysfunction symptoms of the eighth cranial nerve" [9].

Epidemiology

According to an epidemiological study based on Norwegian population, the prevalence of HFS was about 9.8 per 100,000 persons [10]. Another study from the USA reported the prevalence rate

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_1

of HFS as 7.4 per 100,000 men and 14.5 per 100,000 women [11]. Data from our own institute revealed the male-to-female ratio being 1:2.28 and the average age of 52.2 years [12]. Concerning the ethnic distribution, HFS has been reported to be more prevalent in Asian population than others [13–15]. Concomitant psychological issues such as anxiety or depression are noticeable and they are thought to influence the prognosis as well [16]. Except for a few familial cases, HFS does not occur in a hereditary manner, and it predominantly occurs to adults [17–19].

Etiology and Pathophysiology

Vascular compression on the root entry zone (REZ) of the facial nerve is acknowledged to be responsible for primary HFS, whereas any impairment of the facial nerve due to a preexisting condition can constitute a secondary HFS: facial palsy, cerebellopontine angle (CPA) tumors, Chiari I malformations, demyelinating diseases, infections, etc. [20]. Primary HFS is 3–4 times more prevalent than secondary HFS [20, 21]. When a vascular curvature causes the compression on the REZ, anterior inferior cerebellar artery (AICA) is most commonly involved one, followed by posterior inferior cerebellar artery (PICA) and the vertebral artery (VA). A single artery could be the sole cause of the neurovascular compression, but it was rather infrequent (4.7%) according to our previous report [22]. In consideration of other additional factors, a total of six compressive patterns in HFS were proposed: loop, arachnoid, perforator, branch, sandwich, and tandem types [22].

Microscopic disruption of myelin in the REZ or its proximal vicinity where an offending vessel compresses has been acknowledged as the pathophysiology of HFS [23]. Regarding a more detailed mechanism of HFS, there are two major hypotheses: central (hyperexcitability of the facial motor nucleus) vs. peripheral (ephaptic transmission between the facial nerve bundles) hypothesis. Increasing number of microanatomical and neurophysiological research is dedicated to elucidate the precise pathway of HFS; but one hypothesis cannot explain all the phenomena without the other.

Diagnosis

Clinical evaluation including history and physical examination is the key to the diagnosis of HFS. The definition of HFS itself is the most important clue; involuntary facial contractions that are unilateral, irregular, and tonic or clonic. In addition to a close observation of patients' face, a physical maneuver called "other Babinski sign" may be handy. This maneuver, also known as Babinski-2 sign, refers to a synchronized activity of the frontalis or orbicularis oculi muscle that is induced by a self-lifting of one's eyebrow while it is closed. This is reported to yield 100% of specificity and 86% of sensitivity for diagnosis of HFS [24]. EMG, magnetic resonance image (MRI), or computed tomography (CT) also can be used to confirm the diagnosis. Time of flight of MR angiography may display the anatomical relationship between the REZ and an offending vessel. More recent studies using 3D MRI volumetric analysis suggested that CSF space in the posterior fossa of HFS patients was smaller than that of the control group [25]. Also, an analysis using color-duplex ultrasound demonstrated that the mean flow velocity of AICA and PICA on the HFS side was greater than that on the contralateral side [26]. EMG in HFS would show spontaneous and high-frequency synchronized firing, and this finding may be helpful to differentiate HFS from other movement disorders, such as myokymia, blepharospasm, post-facial palsy synkinesia, tic disorders, myokymia, partial motor seizures, craniocervical dystonia (Meige syndrome), tardive dyskinesias (TD) and neuromyotonia, as well as phychogenic HFS.

Treatment

Nonsurgical Treatment

No pharmaceutical medicine has succeeded to provide long-term benefits for HFS. Anticonvulsants or GABAergic medicines may

improve symptoms partially and temporarily, but the effectiveness of these is not comparable to botulinum neurotoxin (BTX) injection, not to mention to MVD. BTX injection is the most preferred nonsurgical treatment for HFS, yielding up to 85% of symptomatic relief. Among seven serotypes of BTX, serotypes A and B are currently commercialized. Following injections, symptomatic improvement occurs in 1–3 days and it usually reaches its peak effect in 5 days [27]. The duration of clinical benefit varies from centers to centers by 3–6 months [28, 29]. Repeated injections of BTX is unavoidable, and tolerance can naturally develop in some subjects, although a 10-year multicenter study reported that the average of duration of improvement did not change from the first year of injection to the 10th year of treatment with the similar dose of BTX [30]. Also, they stated that the BTX-induced adverse responses decreased throughout the 10-year course. Local complications of BTX injection include ptosis, blurred vision, and diplopia, but they are rarely permanent [31]. Incidence of any adverse effect is estimated from 20 to 53% (the mode being around 30–40%), and ptosis is universally the most frequent one [28, 29, 32]. Despite its relatively high success rate of symptomatic improvement, one cannot ignore the fact that BTX injection fundamentally requires repeated sessions, which lead to emotionally and financially non-negligible burden on the patients.

Surgical Treatment

MVD is the only curative treatment option for HFS with high success rate and with low incidence of recurrence and complications. According to a systemic review on 22 studies with 5700 patients who underwent MVD, the complete resolution was achieved in 91.1% (95% CI: 90.3–91.8%) of patients [33]. Recurrence occurred in 2.4% (95% CI: 1.9–2.9%) of patients and postoperative complications included transient complications included facial palsy (9.5% [95% CI:8.8–10.3%]), hearing deficit (3.2% [95% CI: 2.7–3.7%]), and cerebrospinal fluid leak (1.4% [95% CI: 1.1–1.7%]). Permanent complications included hearing deficit in 2.3% (95% CI: 1.9–2.7%) and facial palsy in 0.9% (95% CI: 0.7–1.2%) of patients. The risk of stroke was 1 in 1800 and risk of death was 1 in 5500 [33].

The basic technique of MVD is well described in the literature, but the detailed maneuver varies depending on surgeons. Once a lateral retrosigmoid suboccipital craniectomy or craniotomy is performed under a general anesthesia, the dura is incised to reveal the cerebellar cortex. With or without traction of the flocculus, the REZ of the facial nerve is observed. Upon the identification of the compressing vessels, or the offending arteries, they are separated from the seventh nerve, which then can be perpetuated by insertion of Teflon pieces. A few more additional techniques, including transposition of the vessels, snare technique, vascular sling, etc., have been proposed [34–36].

Intraoperative EMG monitoring can be beneficial for improvement of surgical outcomes. Lateral spread response (LSR) is one of the most popularly applied neurophysiologic tests for HFS, since Moller and Jannetta advocated that disappearance of LSR would indicate properly performed decompression [37]. However, persistence of LSR did not necessarily indicate a poor outcome, which precludes LSR from being a reliable predictor for long-term prognosis of HFS after MVD [38]. Also, to properly monitor the integrity of the eighth nerve (CN VIII) during MVD, intraoperative brain stem auditory evoked potential (BAEP) can be employed, which has been accepted by numerous institutions in decreasing the risk of hearing impairment during MVD.

Clinical course following MVD is not identical. According to our own report, 737 (92.8%) of 807 patients who had undergone MVD for HFS became absolutely or nearly spasm-free by the 2-year postoperative follow-up. However, not everyone started to be asymptomatic immediately after the surgery; 140 (19.0%) of 737 patients still experienced residual spasms more than a month, and some of them lasted more than a year [12]. No universally acknowledged explanation on this disparate clinical course is avail-

able so far; therefore, more electrophysiologic microanatomic researches are needed to elucidate it in the future.

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Natural History of Hemifacial Spasm

Jeong-A Lee and Kwan Park

Hemifacial spasm (HFS) is characterized by unilateral, paroxysmal, and involuntary movements of muscles distributed by the ipsilateral facial nerve. Involuntary contractions usually start from the orbicularis oculi muscle and gradually spread to other muscles associated with facial expressions [1, 2]. At present, since HFS rarely improves spontaneously, it has been agreed that most patients need to be treated [3]. However, the natural history of HFS is not well documented yet.

The overall clinical course of HFS is shown in Fig. 1. Previous studies have focused on the clinical course after microvascular decompression (MVD) (I) and botulinum toxin treatment (II). This chapter illustrates the natural history of untreated HFS (III). To illustrate this, we introduce our two previous papers that investigated the clinical course at points IV and V in Fig. 1.

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Natural History of Hemifacial Spasm Until Visit to Hospital [4]

This study was to set an objective parameter for determining the severity of HFS. We investigated the relationship between the severity of spasms and other factors, including the duration of symptoms. A total of 121 HFS patients who visited an outpatient clinic in our hospital between April and August 2010 were enrolled. The following criteria were included: (a) a clinical diagnosis of primary HFS and (b) no evidence of cognitive impairment. Patients with other movement disorders such as myokymia or blepharospasm or chronic debilitating or life-threatening diseases such as malignancy were excluded. Moreover, two patients who treated with botulinum toxin and one patient who lost to follow-up were excluded. The patients were classified into four groups depending on the severity of spasms (Table 1) [5].

Finally, a total of 118 patients were included in the study. There were 90 women (76.3%) and 28 men (23.7%), with a mean age of 51 years ranging from 22 to 79 years. Preoperative evaluation using the SMC grading system for HFS was divided into 25 patients with grade I, 48 patients with grade II, 33 patients with grade III, and 12 patients with grade IV. Overall, the median duration of symptoms was 48 months, with interquartile ranges of 24–90 months. On the basis of the SMC grade, the mean duration of symptoms was 18 months (range

K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_2

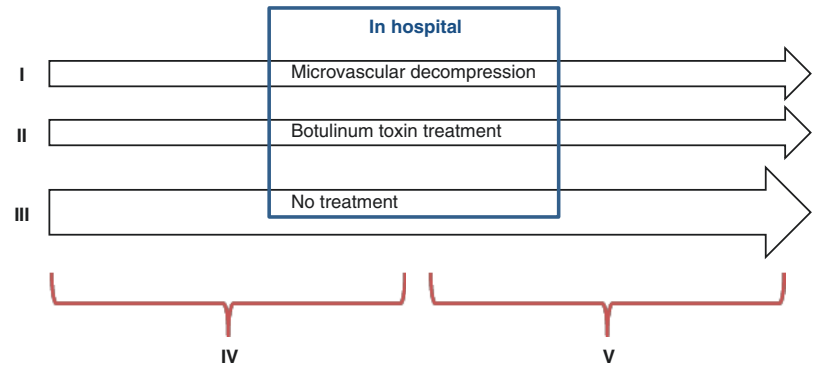

Fig. 1 Research focus on clinical course of HFS. I, clinical course after microvascular decompression (MVD); II, clinical course after botulinum toxin treatment; III, natural history of hemifacial spasm (HFS), IV (until visit to hospital) + V (untreated)

Table 1 SMC grading system for HFS

Grade	Characteristics
I	Localized spasm around the periocular area
II	Involuntary movements spreading to other parts of the ipsilateral face and affecting other muscle groups, including the orbicularis oris, zygomaticus, frontalis, or platysma muscle
III	Frequent tonic spasms affecting vision
IV	Disfiguring asymmetry with continuous contractions of the orbicularis oculi muscle affecting the opening of the eye

Table 2 Spasm severity and symptom duration ($N = 118$)

Spasm severity (n)	Symptom duration, month, mean (range)
Grade I (25)	18 (2–80)
Grade II (48)	39 (2–180)
Grade III (33)	84 (7.5–240)
Grade IV (12)	171 (24–396)

2–80 months) in grade I, 39 months (range 2–180 months) in grade II, 84 months (range 7.5–240 months) in grade III, and 171 months (range 24–396 months) in grade IV patients (Table 2). We observed that the higher the SMC grade, the longer the duration of symptoms that last ($p < 0.05$). This result indicates that the longer the duration of symptoms, the more severe the spasms.

Natural History of Untreated Hemifacial Spasm [6]

This study was to characterize the natural history of untreated HFS over a 5-year period. All 2155 patients initially visited the outpatient clinic of our hospital between 2001 and 2010, and were diagnosed with HFS after a neurological evaluation according to published criteria. Of these patients, 205 patients were selected who met the following criteria: (a) primary HFS diagnosed by one experienced neurosurgeon (K.P.), (b) identification of vascular compression of the facial nerve on magnetic resonance imaging (MRI), and (c) no botulinum toxin or surgical treatment since the initial diagnosis. Other movement disorders such as myokymia or blepharospasm or secondary HFS were excluded. Follow-up was done in 113 of the 205 patients, but the other 92 were not in contact. Nine of these 113 patients were excluded; 6 died and 3 suffered from other diseases such as malignancies and dementia. This is summarized in Fig. 2.

The course of symptoms was divided into four categories: worsened in frequency, duration and intensity, stationary, improved partially, and in remission (little or no spasm). Patients who no longer followed were contacted by phone. These outcomes were determined not by direct medical examination, but by reminding the patients of changes in symptoms since the onset.

Finally, a total of 104 patients were included in the study. There were 62 women (59.6%) and 42 men (60.4%). The mean age of the patients was 62 years (range 34–86 years) at the initial diagnosis of HFS and 50 years (range 22–76 years) at the onset of HFS. The average duration of symptoms was 10.1 years, with a range of 0.2–42.0 years. Changes in the condition were tracked for 5–42 years (mean 12 years) from the onset of symptoms. In 11 out of 104 patients (10.6%), their symptoms worsened from 6 to 42 years (average 16 years). Forty patients

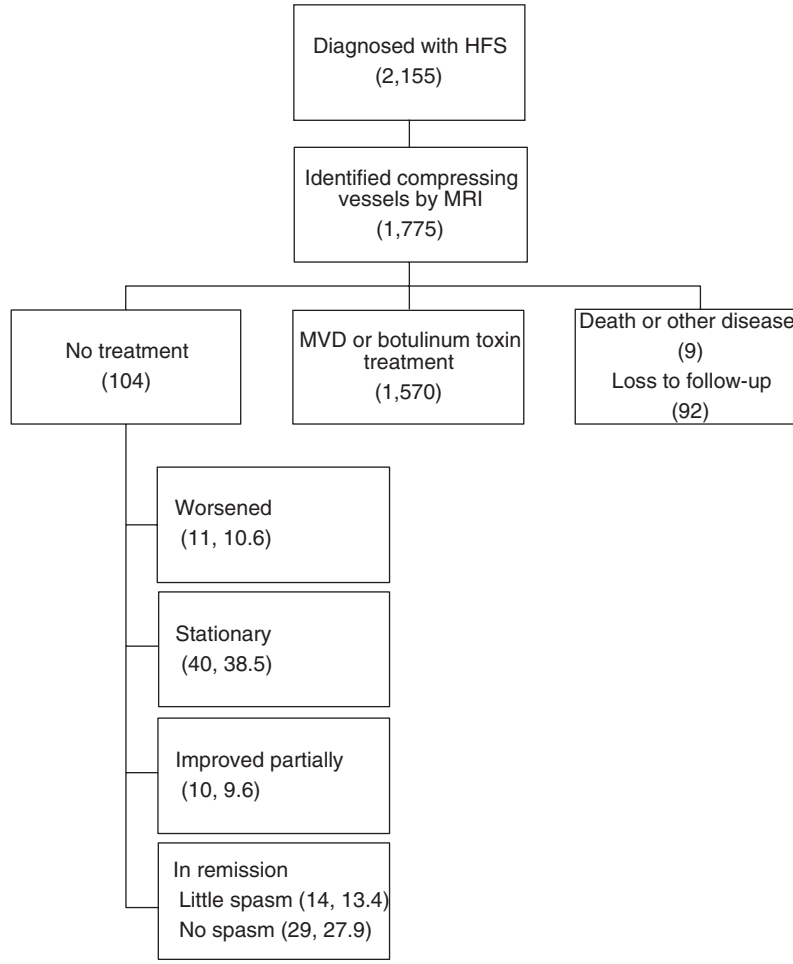

Fig. 2 Study enrollment and follow-up data of untreated HFS patients (*n*, %). *HFS* hemifacial spasm, *MRI* magnetic resonance imaging, *MVD* microvascular decompression

(38.5%) were stationary for 6–23 years (average 12 years). On the other hand, 10 patients (9.6%) improved partially over 7–18 years (average 11 years). Between 2 months and 23 years (mean 6.4 years) after onset, 43 patients (41.3%) were alleviated and no additional treatment was needed for 5 months to 13 years (mean 5.7 years) (Fig. 2). Despite no improvement in symptoms, these patients were conservatively followed due to other health conditions, involved costs, their anxiety and concerns related to risks and limitations, and their adaptation to the symptoms. Thirty-seven patients continued to receive other treatments such as treatments of Traditional Chinese Medicine including acupuncture or herbal remedies, medication, and physiotherapy. Thirty-eight patients did not respond properly and were not treated anymore and 29 patients did not receive any treatment.

However, only 4.8% of 2155 patients diagnosed with HFS were included in this study. Excluded patients may later have surgical intervention due to worsening of symptoms. Given this, the remission rate, or 41% of the population, is expected to decrease.

The follow-up studies after botulinum toxin treatment were conducted by several researchers [3, 7–9]. In the study by Conte et al., spread of spasms to the other facial muscles of the same side of the face existed in 93.4% of HFS patients, and longer latency of spread (the time interval between the onset of muscle spasm and the subsequent involvement of other part of the face) was associated with longer duration of disease and younger age at onset. They concluded that the development of spread likely represents the natural course of HFS. They also assumed that spread in HFS can occur through an ephaptic

mechanism on the facial nerve trunk rather than facial nucleus hyperexcitability, as botulinum toxin injection did not reduce spread rate [7]. Our results are consistent with their findings, which support the debilitating nature of HFS and the need for treatment in a relatively high proportion of patients that HFS persists. In contrast, our results show that the proportion of patients in remission is higher. In other studies, no patients had remission or only 2.3–4.2% experienced symptomatic relief [3, 8, 9].

On the other hand, disease progression is different across patients. In our data, based on the duration and severity of symptoms, patients were classified into rapidly and slowly progressive groups and the factors related to the differences between the two groups were investigated. As a result, the younger age at the time of surgery, the older age at the onset of symptoms, and the absence of indentation on the facial nerve intraoperatively were associated with rapidly progressive HFS. The reason for the young patients in the rapidly progressive group is that if the symptoms progress faster, the patients may choose surgery earlier. Rapid progression of late-onset HFS may be related to aging or the cortical influences of the facial motor nucleus. In addition, we hypothesized that mild vascular compression and consequent absence of nerve indentation, rather than severe vascular compression and consequent nerve indentation, lead to rapid HFS progression. These results may be useful for understanding and informing patients on the differences in disease progression in HFS [10].

In summing up the above results, HFS is often associated with the gradual deterioration of the spasm pattern and the reduction of the spasm-free intervals. Nevertheless, there were no previous studies on why HFS patients have different courses. Even though it is unclear why this happens, our findings suggest that the degree and progression rate of symptoms due to demyelination of the facial nerve [10, 11] or hyperexcitability in the facial motor nucleus [11–13] or the denervation of the affected muscles [14] are diverse, according to individual characteristics. If this inference is correct, further research is needed to clarify the contributing factors. Another issue is whether it is spasm-free interval or whether the symptoms actually improve. Our data showed that 35 of 43 patients (81.4%) in remission were spasm-free for more than 3 years, but this was not clarified. Future trials should explore this evaluation standard.

Quality of Life in Hemifacial Spasm Patients

Although HFS symptoms naturally improve in some patients, patients with HFS have problems with their quality of life (QoL) during their symptoms. First, HFS patients showed a high frequency of social anxiety disorder compared to healthy subjects, especially in young and depressed patients [15]. And HFS patients had higher scores than focal dystonia or healthy-control subjects for "contamination" and "aggressiveness" on the Structured Clinical Interview for Obsessive-Compulsive Spectrum Self-Report [16]. Additionally, sexual function was affected in HFS patients than in healthy controls [17]. Furthermore, the more severe the spasms, the worse are the depression, headache, and QoL [4, 18–20]. Conversely, HFS patients with anxiety reported a significant improvement in symptoms after proper care. Therefore, because stress and anxiety can worsen HFS, the authors concluded that diagnosis and early management of anxiety symptoms can improve QoL in HFS patients [21].

On the other hand, treatments of HFS improved QoL in HFS patients. In many studies of botulinum toxin treatment for HFS, QoL has changed significantly before and after the treatment [22–26]. Also about MVD, many studies have demonstrated that the post-surgery improvements of the HFS symptoms were associated with decreased social anxiety and improved QoL [27–29]. In some cases, the QoL improvement was prolonged due to delayed improvement of symptoms after surgery [30–32]. According to the above findings, patients with HFS benefited from botulinum toxin treatment or MVD in symptoms associated with long-term QoL.

Thus, even if the symptoms improve, appropriate treatment will be necessary for

well-selected patients because such situations do not often occur and QoL is impaired while the symptoms are present.

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Novel Classification Systems for Hemifacial Spasm

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The accumulated experience in performing microvascular decompression (MVD) for hemifacial spasm (HFS) at our institute for more than two decades has allowed us to broaden the scope of the concept in treating HFS patients. During the journey, through more than 5000 cases of MVD for HFS, the results of our observation were quantified and categorized, which are to be introduced in this chapter. We believe that they are intuitive, clinically pertinent, and pragmatic. Also, if used by more and more researchers around the globe, our classification systems can be a foundation for deeper theoretical knowledge and understanding of HFS.

Compression Patterns

Vascular compression on the root entry zone (REZ) of the seventh nerve (CN VII) is the most predominantly accepted etiology of HFS. The term neurovascular compression seems straightforward, but the actual intraoperative findings during MVDs show much more variety of compressing patterns. In terms of the involving arteries, anterior inferior cerebellar artery (AICA) was the most prevalent (51.7%), followed by posterior inferior cerebellar artery (PICA, 21.6%) [1]. The vertebral artery (VA) was rarely responsible for the compression by itself (1.7%), but in combination with AICA, PICA, or both, it could account for 14% of the compressions (Table 1). Compressive patterns that we discuss in this section depict the ways in which a vessel or vessels compressed the REZ of the CN VII, purely based on the microsurgical findings.

Table 1 Number of compression vessels involved in each compression pattern

	VA	AICA	PICA
	($p < 0.001$)	($p = 0.005$)	($p = 0.003$)
Loop type	2	1	8
Arachnoid type	3	40	25
Perforator type	0	49	9
Branch type	0	14	4
Sandwich type	0	26	13
Tandem type	32	43	25
Miscellaneous type	0	2	0
Total	37	175	84

Loop Type (Fig. 1a)

The loop type refers to a type of compression on the REZ caused by a single artery without any other concomitant factors; the excessive curvature of a vessel that directly contacts the REZ

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_3

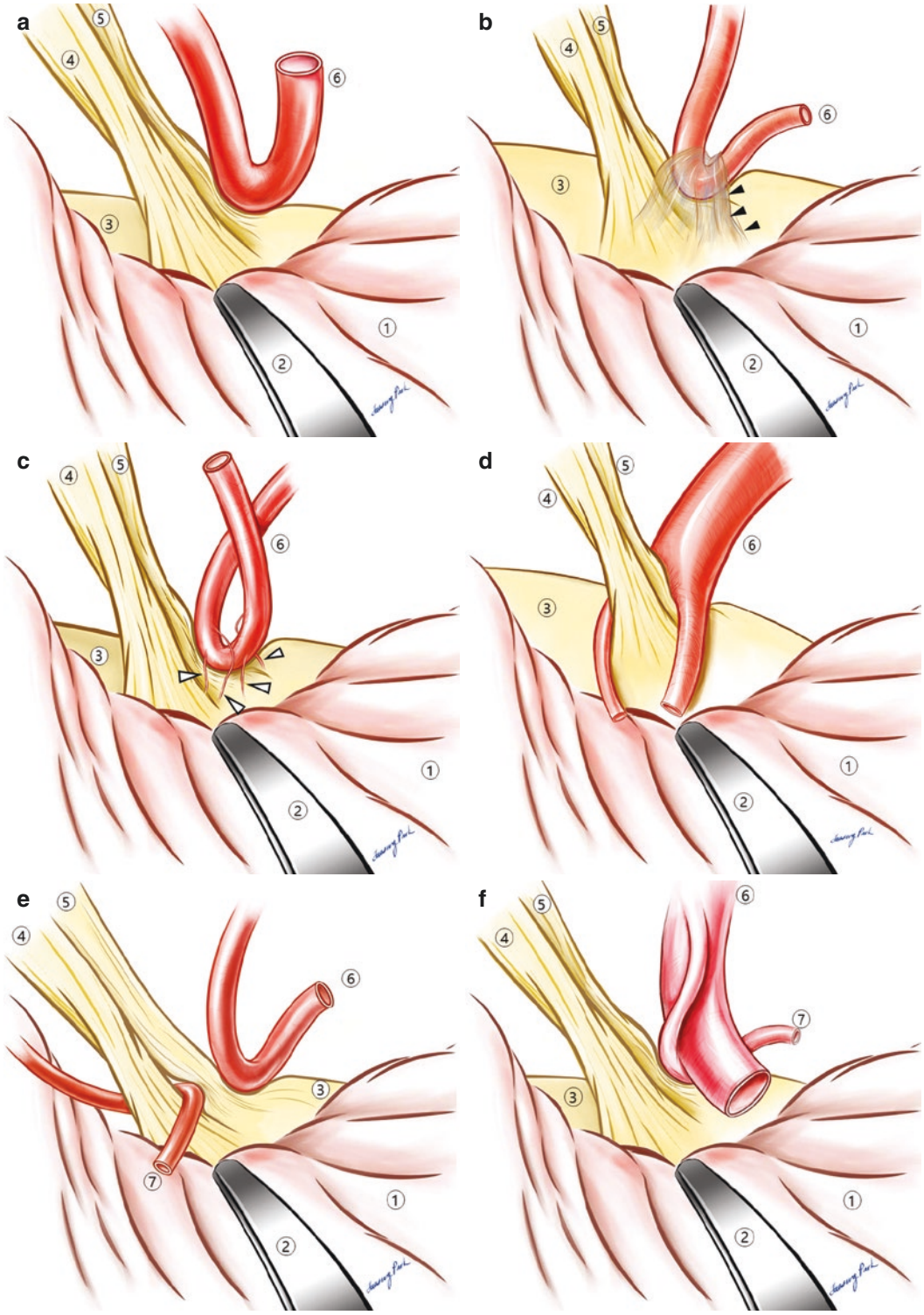

Fig. 1 Various compression patterns of hemifacial spasm: intraoperative view for the right side lesion. (**a**) loop type, (**b**) arachnoid type, (**c**) perforator type, (**d**) branch type, (**e**) sandwich type, and (**f**) tandem type. (1) Cerebellum, (2) retractor, (3) brain stem (pons), (4) CNVIII, the vestibulocochlear nerve, (5) CN VII, the facial nerve, (6) compressing vessel #1, (7) compressing vessel #2; arrowheads, arachnoid membrane; hollow arrowheads, perforators to the brain stem ()

conveys the throbbing pulse to it. This is indeed the simplest type, and yet it is far from the most common one. According to our previous report in 2008, this specific type accounted for only 11 (4.7%) of 236 cases [1]. What interested us was that the involvement of PICA among the loop type compressions (8 of 11, 72.7%) was disproportionately greater than that of the whole cases (51 of 236, 21.6%). Given that there was no additional structure to cause the compression other than the vessel curvature itself, the postoperative outcome for this specific type was excellent; the success rate was 100% with no noticeable complications.

Arachnoid Type (Fig. 1b)

The arachnoid type is defined by a compression where the compressing vessel is tethered tightly to the REZ or brainstem by thickened arachnoid trabeculae, and this was the most common type of all (28.0%). Unlike the loop type, the mobilization of the vessel is hardly achievable without sufficient release of the tethering between the vessel and the REZ. Cautious and meticulous dissection of the thickened arachnoid trabeculae is the key to the successful decompression, and once it is done, the compressing vessel is usually freed from the REZ. More often than not, the insertion of Teflon sponge was not even necessary merely for the purpose of decompression; yet, it was performed for the sake of prevention of recurrence.

Perforator Type (Fig. 1c)

This type is analogous to the arachnoid type in that the tethering of the vessel is attributable to the perforating arteries from the main compressing vessel, instead of the thickened arachnoid trabeculae in the arachnoid type. However, instead of surgical dissection of arachnoid trabeculae in the arachnoid type, much more cautious approach to the REZ is crucial in the perforator type, since any disruption of perforators must be avoided at all costs. In our experience, though, this type was not necessarily related to worse outcome compared to others. To be aware that the neurovascular compression might involve perforators as the tethering mechanism has helped us tremendously, and we believe that many other young neurosurgeons may benefit from it. Perforator type was the second most popular type, following the arachnoid one.

Branch Type (Fig. 1d)

When the REZ is encircled by two branches of an offending artery, this is called a branch type. Although the range of immobilization may be limited compared to other types, no statistically significant difference was noted in terms of employed surgical techniques, postoperative outcomes, and complications.

Sandwich Type (Fig. 1e)

This type depicts a compression where the REZ is caught between two offending arteries: a dorsal and a ventral one. A ventrally compressing artery is easy to miss, unless a thorough inspection around the REZ is carried out. Even after a successful decompression is achieved for the dorsally positioned artery, the patient may end up suffering the unresolved symptom, if the ventral artery is still causing the compression. Existence of this type with the frequency of 11.9% would be the rationale for the necessity of after-decompression examination around the REZ. One of 236 subjects who underwent MVD developed a permanent hearing loss; and she happened to harbor the sandwich type. No statistical analysis was available for this one case, but one could assume that the decompression for a ventrally residing artery may pose technical challenges.

Tandem Type (Fig. 1f)

This type is defined by a compression where a vessel compresses another vessel, and in turn, their combined force affects the REZ. Tandem

type accounted for 22.0% of whole cases, and this had a strong predilection for the VA, and vice versa. The VA was involved in 61.5% of the tandem type and the tandem type accounted for 86.5% of all the cases where the VA was identified as an offending artery. When the VA is suspected to be an offending artery, one must consider the possibility of a tandem type, and an exhaustive exploration around the REZ must be carried out in search of another offending artery.

Pattern of Hearing Loss

Despite a high success rate of MVD for HFS (more than 90%), it is not without postoperative complications, and hearing loss is one of the most serious ones [2–5]. Thus, it is imperative to monitor the functional integrity of the eighth nerve (CN VIII) during the surgery. Intraoperative brain stem auditory evoked potential (BAEP) is one of the most commonly performed monitoring methods, and it is acknowledged to reduce the risk of hearing impairment during a surgery involving the CN VIII [6, 7]. In this section, a new grading system of hearing loss is to be introduced, and its association with intraoperative BAEP is also to be presented.

Hearing Evaluations

Our previous reports on hearing loss following MVD employed pure tone audiometry (PTA) along with speech discrimination score (SDS) [4, 8]. All patients went through both tests prior to and after the surgery; the postoperative one was performed within 10 postoperative days. The average of PTA thresholds on the frequencies of 0.5, 1, 2, and 3 kHz was calculated for each subject, and their postoperative results were compared with the preoperative ones. For those who experienced a change of PTA thresholds greater than 15 dB or 20% or more decrease in SDS, another session of PTA and SDS was performed. The great majority of all patients (1098 of 1144, 96.0%) showed no or negligible change in PTA thresholds, and they were categorized as group 1. Those who developed total deafness fell into group 4 (10, 0.9%), and the remaining 36 belonged to group 2 or 3. Group 2 consisted of subjects whose postoperative hearing had deteriorated by more than 15 dB in PTA threshold, and the decrease in their SDS was proportionate to the degree of PTA. Patients whose SDS was disproportionately worsened compared to the changes of PTA comprised group 3. In accordance with Speech Recognition Interpretation (SPRINT) chart, when SDS score fell in the shaded area, it was considered as "disproportionate decrease" compared to PTA thresholds [8–10] (Fig. 2). Table 2 shows the definition of each group. The pattern of hearing loss of group 2 was considered as "cochlear type," whereas that of group 3 as "retrocochlear type" [4, 8]. In terms of recovery from the hearing loss, the cochlear type was associated with better prognosis than the retrocochlear one.

Intraoperative BAEP in Relation to Postoperative Hearing Loss

Throughout the surgery (from the skin incision to the dural closure), BAEP was monitored for all patients. A decrease in the amplitude in peak V by more than 50% or delayed latency of peak V longer than 1.0 ms was considered as a significant change of BAEP. Whenever either one or both took place, the surgeon was alarmed; the cerebellar retractors were thereupon released or repositioned until the change of BAEP recovered. 280 of 1144 patients experienced decreased amplitude of peak V by 50% or more at some point during the surgery, and upon release of the cerebellar retractor, 268 (95.7%) of 280 recovered their amplitude of peak V above 50% of the reference grand average (Table 3). Whether or not the amplitude of peak V was restored to above 50% of the reference was statistically associated with the four groups of hearing loss based on PTA and SDS. Table 3 shows the relationship of intraoperative BAEP with the categorization of hearing loss.

On the usefulness of intraoperative BAEP, Sindou proposed that an increase in latency of

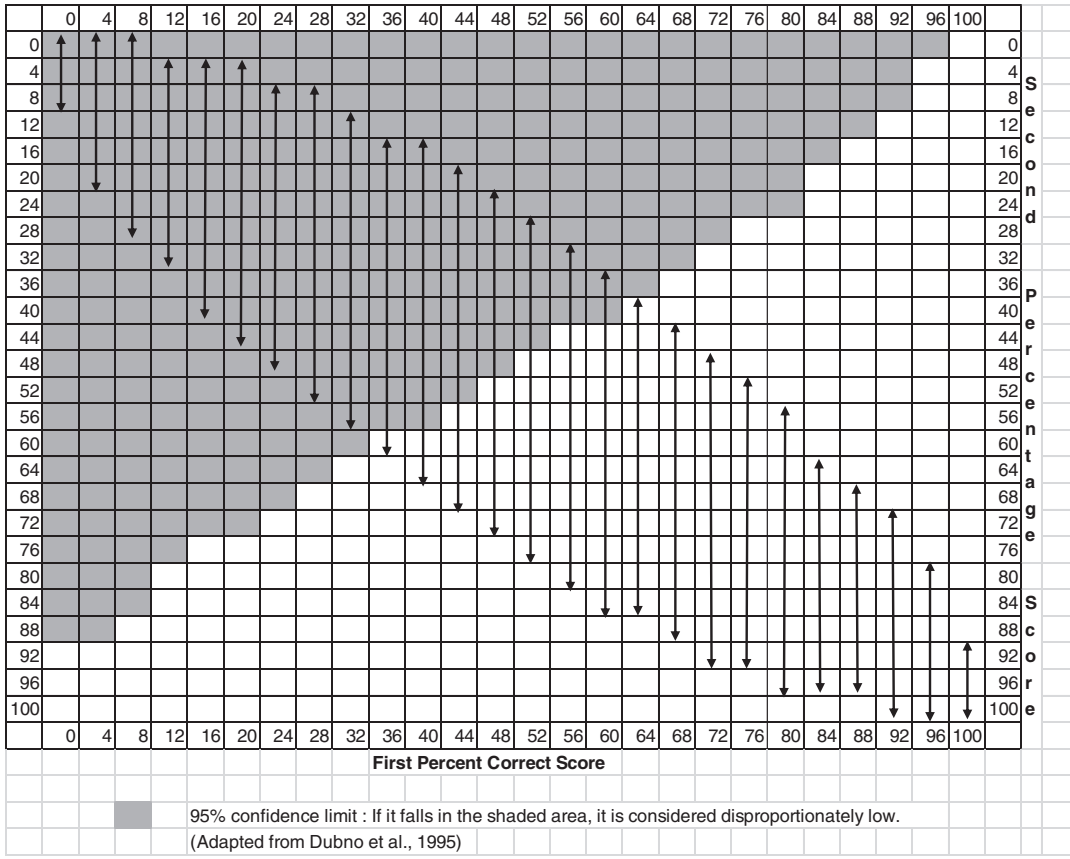

Fig. 2 Speech Recognition Interpretation (SPRINNT) chart

Table 2 Patterns of hearing loss

	Definition
Group 1	None or negligible change
Group 2	Deterioration in PTA >15 dB and proportionate decrease in SDS
Group 3	Deterioration in PTA >15 dB and disproportionate decrease in SDS
Group 4	Total deafness

Table 3 Relationship between intraoperative BAEP change and clinical hearing loss

	No change in BAEP	>50% decrease in peak V amplitude (recovery to >50% vs. no recovery)	
Group 1	841	257	(257 vs. 0)
Group 2	18	8	(7 vs. 1)
Group 3	2	8	(3 vs. 5)
Group 4	3	7	(1 vs. 6)

peak V as well as a decrease in the amplitude of peak I could be a warning sign for excessively stretched cochlear nerve, whereas Polo et al. emphasized the delay in peak V as a watching, warning, or critical sign, depending on the length of the delay [7, 11]. Our previous report suggested that a decrease in the amplitude of peak V was statistically consistent with the clinical severity of hearing loss [4].

Extent of Compression

As previously mentioned, the intraoperative findings greatly vary. One of the diverse features of the compression is the indentation of the REZ. Based on surgical findings of 293 MVDs,

Table 4 Grades of indentation on the REZ

	Definition	Number of patients (%)
Grade 1	No recognizable indentation	64 (21.6)
Grade 2	Indentation without discoloration	114 (39.2)
Grade 3	Indentation with discoloration	115 (39.2)

we proposed a new grading system for the extent of the compression (Table 4) [12].

The grades of indentation were associated with the postoperative outcomes. Our initial hypothesis was that a more severe indentation would yield a poorer outcome; however, the opposite result was drawn. Concerning this rather odd result, a couple of assumptions were suggested. A more severe, that is, more distinct indentation may have enabled the surgeon recognize the optimal site for decompression. Another explanation would be that group 1 (with no or negligible indentation) may have included those with secondary HFS or HFS mimicking conditions such as myokymia, post-facial palsy synkinesis, etc.

Lateral Spread Response

Lateral spread response (LSR) is one of the most frequently researched neurophysiological methods for HFS; and yet there are still many controversies if it actually reflects the long-term outcomes. LSR, a phenomenon observed during the intraoperative facial electromyogram, refers to abnormal responses in a facial muscle group when an irrelevant branch of the facial nerve is stimulated. Given that LSR, in general, disappears during the MVD procedure, numerous researchers postulated that the disappearance of LSR could indicate a successful decompression; this is true in most cases, but not all. There are non-negligible numbers of exceptions: no LSR before a decompression, persistent LSR after a successful decompression, disappearance of LSR followed by an unsatisfactory outcome, etc. No universally accepted consensus is available concerning how to analyze the intraoperative LSR results; factors that can affect the conclusion include the use of neuromuscular blockade (NMB), the dose of NMB, minimal amplitude to constitute LSR, and precise timing of the LSR disappearance. These factors may vary from institute to institute.

Table 5 Definition of LSR grades

LSR grades	Definition
0	Nonexistent LSR
1	Disappearance of LSR before decompression
1a	Upon dural opening
1b	Between dural opening and decompression
2	Disappearance of LSR after decompression
2a	Upon decompression
2b	Delayed disappearance after decompression
3	Persistent LSR throughout MVD

The grading system we used to analyze LSR is presented in Table 5. If this grading system is to be prevalently used among institutes, a less controversial conclusion on LSR may be drawn in the near future.

Clinical Grade: SMC Grading System

HFS is mainly diagnosed by clinical observation. Thus, the severity of the disease is also determined by patients' verbatim and physicians' examination, not by any objective measures including laboratory data, electrophysiological test results, or radiological evaluations. Although changes in symptoms of HFS can vary from person to person, there is a typical pattern of progress. Based on this pattern of progress, we suggested a new grading system for symptoms of HFS, named "SMC grading system" in 2005 [13] (Table 6). This was created in accordance with patients' symptoms and their chronological changes, and our previous report demonstrated that this grading system was consistent with HFS-7 questionnaire, also known as short self-rating quality of life scale ($p < 0.001$) [13]. More and more scholars are adopting this scale to their

Table 6 Description of SMC grading system

Grade	Detailed description
I	Localized spasm around the periocular area
II	Involuntary movement spreads to other parts of the ipsilateral face and affects other muscle groups: the orbicularis oris, zygomaticus, frontalis or platysma muscle
III	Interference with vision because of frequent tonic spasms
IV	Disfiguring asymmetry: continuous contraction of the orbicularis oculi muscles affects opening of the eye

researches, which we wish to be a foundation for a greater understanding of HFS in the future.

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Pathogenesis of Hemifacial Spasm

Min Ho Lee and Jae Sung Park

Hemifacial spasm (HFS), a hyperactive motor dysfunction of the facial nerve, is believed to be caused by vascular compression at the root exit zone (REZ) of the facial nerve, which offered the basis for microvascular decompression (MVD) to become the standard treatment for HFS [1–5]. However, questions have yet to be fully answered about the microscopic pathophysiology of HFS, beyond the compression on the REZ: How come the compression on the REZ causes a hyperactivity instead of hypoactivity of the facial nerve? What can explain disparate clinical courses following a successful MVD? Are there additional contributing factors to the symptoms other than the compression on the REZ?

Anatomy of Facial Nerve

The facial nerve (seventh pair of cranial nerves) is a mixed nerve with efferent (motor) and afferent (general and special sensory) nerve fibers. It emerges from pons of the brainstem and reaches to the internal auditory canal. The segment between the brainstem and the internal auditory canal, that is, intracranial or cisternal segment, is estimated to be 17.93 ± 2.29 mm of length. The proximal end of the cisternal segment where it adheres to the pons is called the REZ. About 0.96 mm (range 2.86–1.9 mm) proximal to the REZ is located the "transition zone." Transition zone, that is, Obersteiner-Redlich zone, refers to the area where the central myelin is gradually replaced by peripheral Schwann cell–derived myelin. The transition zone, not surrounded by perineurium or epineurium, is very susceptible to mechanical disruption when it is compressed by a vessel or vessels [6–10].

Hypotheses for the Pathogenesis of Hemifacial Spasm: Peripheral vs. Central

There are two main hypotheses for the detailed pathogenesis of HFS: peripheral (ephaptic transmission) vs. central (antidromic conduction to the facial motor nucleus) hypothesis (Fig. 1). In 1962, Gardner [1] postulated that the local irritation of the facial nerve caused by vascular compression may lead to ephaptic impulse transmission. The term "ephaptic transmission" depicts a situation where an impulse in a group of nerve fibers initiates additional impulse in the adjacent fibers [1, 11]. On the contrary, the central hypothesis refers to hyperexcitability of the facial motor nucleus (FMN) [12, 13]. In 1987, Møller and Jannetta

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_4

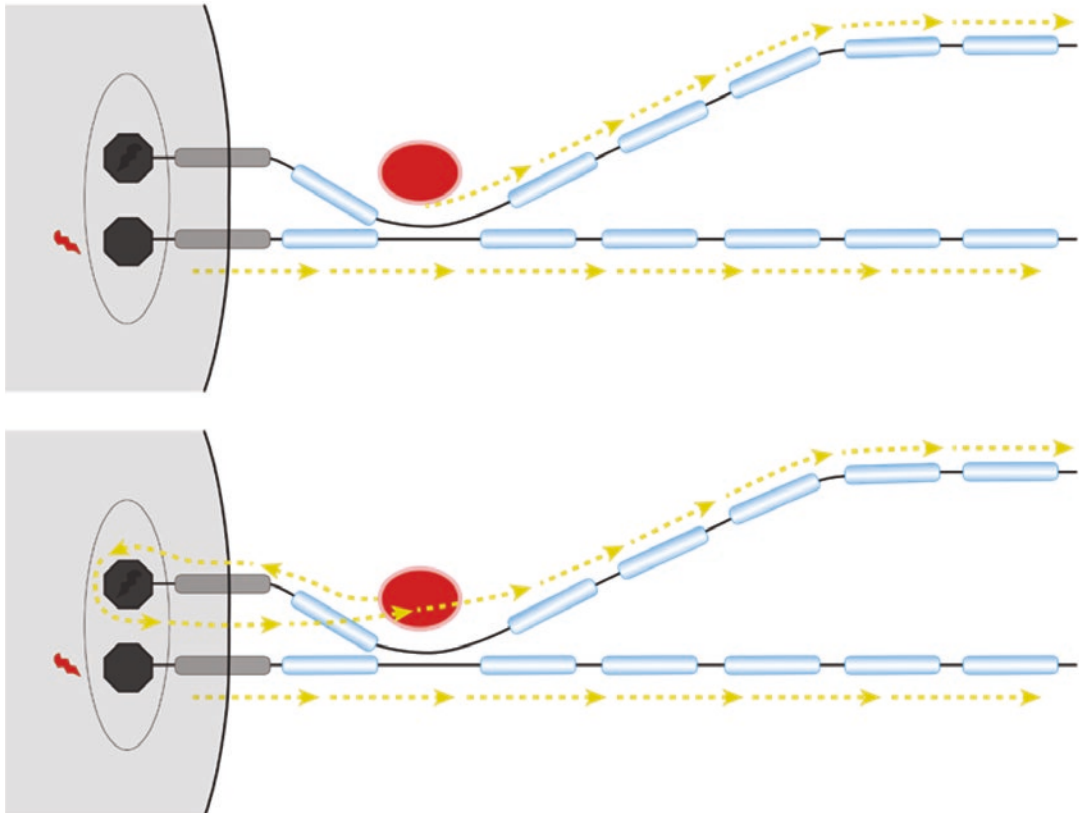

Fig. 1 Hypotheses for the pathogenesis of hemifacial spasm. (**a**) Above: peripheral hypothesis with ephaptic impulse transmission. Adjacent nerve fiber compressed by vessel may transmit the nerve conduction. (**b**) Below: central hypothesis with antidromic nerve conduction to the nucleus. The stimulated signal transmits backward to the FMN, and then it travels forward

reported that there was a characteristic electrophysiological wave that was confined to HFS patients: abnormal muscle response (AMR) or lateral spread response (LSR) [14]. LSR consists of two electrophysiological waves: the initial one followed by a smaller, delayed one. To explain the phenomenon of LSR, the authors advocated that the backfiring of the facial nerve might have antidromically activated the FMN, which, in turn, emitted a delayed signal to the facial nerve [15, 16]. The hyperexcitability of the FMN as a result of the antidromic backfiring constitutes the central hypothesis of HFS. Although the central hypothesis appears to be opposite to the peripheral one, these two are not mutually exclusive; Moller and Jannetta stated that the phenomenon of LSR might be attributable to ephaptic transmission alone or in combination with the hyperexcitability of the FMN [17–20].

Sympathetic Hypothesis

Generally, once the offending artery was removed away from the facial nerve, the LSR wave diminishes immediately and the symptom of spasm ceased postoperatively in most of the cases [2, 21]. This may not be explained by the peripheral or central hypotheses as above, for neither the histological changes at the compression sites nor the hyper-excitability of facial motor neurons was able to repair right after decompression. Of course, if anatomical change or damage is not severe, we can expect a quick improvement, but it is difficult to explain everything with this. Dou et al. proposed that the offending artery may play a more important role than the effect of merely mechanical compression [22]. Anatomically, arteries are covered with adventitia that contains

sympathetic nerve endings as well as vasa vasorum. Normally, the sympathetic endings release neurotransmitters. These neurotransmitters released from the sympathetic nervous endings in the adventitia may spill over from the artery wall and spread to the demyelinated nerve fibers in close contact. And it may induce an ectopia action potential in those demyelinated facial nerve fibers expanding to the neuromuscular conjunction and trigger an attack of HFS. This was revealed using the animal model by Zhou et al. [23]. They called it sympathetic hypothesis. But this still needs more evidence to prove it.

Other Factors That May Be Related with Pathogenesis of Hemifacial Spasm

Vertebrobasilar dolichoectasia may cause vascular crowding in the limited space of the posterior fossa, increasing the risk of vascular compression of the facial nerve. The direction of lateral deviation of the vertebral artery has been reported to be significantly related to the symptomatic side of HFS [24, 25]. Pathogenesis of dolichoectasia is thought to be associated with rarity of the elastic tissue in the tunica media with fragmentation of the internal elastic lamina, and it became elongated and arteriosclerotic. The main causes of such a process are vascular risk factors, like old age, male sex, and hypertension [26, 27]. Several studies have shown a positive correlation between hypertension and HFS [28–34]. Atherosclerosis was reported to be relevant in some studies [35], but it was not found in other studies [36].

In addition to compression of the vessel, another contributing factor leading to HFS is adhesion and traction to vessel of adjacent arachnoid membranes. Some studies have stated that complete separation and dissection of arachnoid membranes can result in sufficient MVD for HFS [5, 37, 38]. Moreover, thickened arachnoid membranes have sometimes been observed during surgery for HFS. Among the classifications suggested by Park et al. [5], it may be regarded as arachnoid type, which thick arachnoid trabeculae between the vessel and the brainstem cause the vessel to be tethered tightly to the nerve.

Previous studies have reported that these abnormal thick arachnoid membranes are caused by inflammation (infection, hemorrhage, etc.) [39–41]. There is no known link between inflammation and HFS yet. Recently, Chen et al. [42] compared inflammatory factors (IL-γ, IL-2 receptor, IL-6, IL-8, IL-10, TNF-α) between patients with HFS, and other control groups (patients with lumbar disc herniation, and healthy control subjects). And they suggested that IL-6 is involved in pathogenesis of HFS.

Meanwhile, a study has been published that HFS is caused by not only simple mechanical compression but also exacerbation by sympathetic activity of autonomic nervous system [43]. They performed simultaneous recordings of the electrocardiography and facial electromyography in patients with HFS and revealed that a transient increase in the heart rate occurred a few seconds before the onset of spasm.

Although these studies still show preliminary results, it is a very interesting approach to the cause of neurovascular conflict.

Secondary Hemifacial Spasm

The incidence of cerebellopontine angle (CPA) tumor-induced HFS ranges from 0.3 to 2.5% of all patients with HFS [44, 45]. HFS can also be induced by not only CPA tumors (meningioma, or epidermoid cyst), even stem glioma, aneurysm, vascular malformation, or infection [46–50]. Some authors suggested that displaced vessels by tumor triggered the HFS [51–53]. And others suggested that tumor may compress the nerve or nucleus directly [37, 54]. It is too primitive for us to conclude here. It is too early to conclude the actual pathogenesis of secondary HFS, yet. But, we should consider various possibilities for these causes of HFS and review the images meticulously. These studies are also expected to be one of the keys to elucidate the pathogenesis of HFS.

Conclusion

As mentioned above, the pathophysiology of HFS is not yet clear. But, the fact is that the compression of the facial nerve by an offending vessel is definitely the origin of the increased excitability of the facial nerve that leads to HFS.

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Clinical Symptoms and Differential Diagnosis of Hemifacial Spasm

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Introduction

Hemifacial spasm (HFS) is characterized by brief, repetitive, and involuntary tonic–clonic contraction of unilateral facial expression muscles. The diagnosis of HFS should be made based on the clinical history and typical symptoms of the disease. At the onset of the disease, HFS is usually insidious; the spasms can be brief and localized in the lower eyelid muscle (orbicularis oculi) before progressing to the upper and lower face [1–3]. In advanced cases, the spasms become more tonic and involve whole unilateral facial expression muscles, such as the frontalis, corrugator, procerus, zygomaticus major, zygomaticus minor, nasalis, levator labii superioris, levator labii superioris alaeque nasi, orbicularis oris, mentalis, depressor labii inferioris, levator anguli oris, risorius, and platysma. HFS is not a life-threatening disease but may induce social embarrassment and withdrawal. Finally, it can decrease the patient's quality of life [3–5]. Sometimes, establishing a diagnosis of HFS is challenging, especially when the symptoms are subtle or before the development of definite symptoms of HFS. Misdiagnosis of HFS as another hyperkinetic facial movement condition can occur and HFS should be differentiated from other hyperkinetic facial movement disorders to ensure the delivery of precise treatment. In these cases, following up on clinical symptoms, performing closed investigations of specific signs of HFS, and taking home videos can help with the precise diagnosis of HFS. In this chapter, we reviewed the clinical features and the differential diagnoses of HFS to ensure a precise diagnosis.

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Clinical Symptoms of Hemifacial Spasm

The facial nerve consists of the temporal, zygomatic, buccal, marginal mandibular, and cervical branches. HFS can involve any of these five branches, although the lateral and lower part of orbicularis oculi muscle, which is innervated by the zygomatic branch [6], is most frequently affected (75–90%) in the beginning. Therefore, visible symptoms of HFS often initiate with twitching of the lower eyelid. In contrast, only 5–11.7% of patients first show the condition in

K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_5

the lower face, while just a few patients reported contractions involving the upper and lower face simultaneously [1, 3–5]. Generally, symptoms beginning in the periocular area spread to the upper and lower facial areas.

With the progression of the disease, the temporal branch, which innervates to the upper part of orbicularis oculi, frontalis, procerus, and corrugator muscles, becomes involved. The contraction of the orbicularis oculi and frontalis muscles lead to repetitive eye-closing movements and elevation of the eyebrows, while the involvement of the procerus and corrugator muscles facilitate spasm in the middle of the forehead.

Also with the progression of HFS, the midface and lower facial muscles also can be affected and the contraction of theses muscles triggers various facial symptoms. Muscles placed at midface are innervated by the zygomatic and buccal branches. The zygomaticus major and minor muscles innervated by the zygomatic and buccal branches support upward and lateral movements of the mouth angle. Separately, the levator labii superioris aleque nasi, levator labii superioris, and levator anguli oris muscles are also innervated by the zygomatic and buccal branches and trigger elevation of the upper lip and spasm in the nasal area. Finally, the risorius muscle, innervated by the buccal branch of the facial nerve, pulls the corner of the mouth laterally.

Lower facial symptoms can also present when HFS involves the marginal mandibular or buccal branches of the facial nerve. Both of these branches innervate to the orbicularis oris and contraction of the orbicularis oris leads to perioral spasm and twitching. Moreover, they also innervate to the depressor labii inferioris, depressor anguli oris, and mentalis muscles, triggering involuntary chin movement. In some patients, the platysma muscle, a neck muscle innervated by the cervical branch, is also involved and presents spasm [7] (Fig. 1).

Usually, HFS involves only one side of the face, with left-sided HFS being slightly more common. However, bilateral HFS is rarely reported (less than 3% of cases) and other hyperkinetic movement disorders such as blepharospasm should be differentiated in this context.

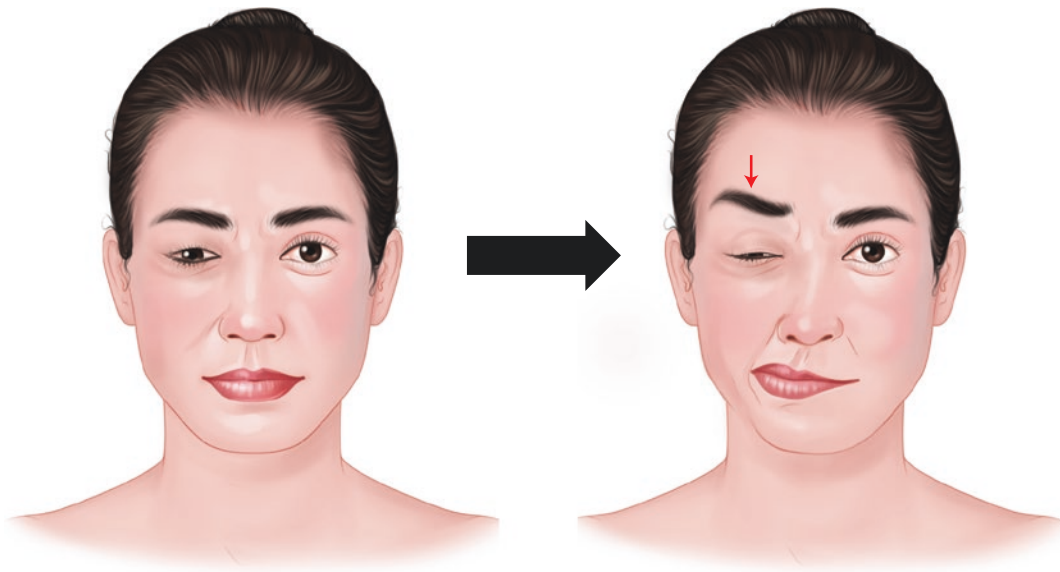

Fig. 1 Symptoms of hemifacial spasm. Visible symptoms of hemifacial spasm often initiate with twitching or spasm of the periocular area without the upper and lower face involvement (Left). With the progression of the disease, the upper, midface, and lower face are also affected (Right). The paradoxical involuntary upward replacement of the ipsilateral eyebrow during involuntary eye closing, known as "the other Babinski sign" or "Babinski sign-2," is observed in HFS patients with the presentation correlated with the severity of symptoms. As such, it is less common in the early stage (Left) of the disease and typically appears as the disease progresses (Right)

About 2% of primary HFS patients report a positive family history of HFS, but genetic factors associated with HFS have not yet been elucidated. The age of onset of familial HFS is variable, and, occasionally, this condition can occur early in life. Vascular decompression is also effective in familial HFS, with study results suggesting that vascular compression is involved in generating HFS even in familial cases [8].

The symptoms of HFS are fluctuated by various factors. Anxiety and stress are the most common aggravating or trigger factors in HFS. Patients have also reported a worsening of symptoms with sleep deprivation, fatigue, reading, light exposure, wind, chewing, reading, or talking. Most patients reported their symptoms were alleviated by relaxation, alcohol intake, touching the area, or facial massage. HFS remains persistent during sleep. One study using polysomnography reported that all the included patients (n = 12) showed persistent HFS during sleep, albeit of a decreased severity [9].

Patients with HFS may complain of various symptoms other than hyperkinetic movement including social embarrassment, depression, interference with vision, dysarthria, and sialorrhea [3]. Along these lines, the presence of a "clicking ear sound" that was simultaneous and synchronous with the involuntary facial muscle contractions was reported by 22.7% of patients with HFS. It is believed that co-contraction of the stapedius muscle leads to rarefaction and compression in the middle ear [10].

In rare cases, trigeminal neuralgia is preceded by or accompanies HFS and is called tic convulsif, which was coined by Cushing in 1920. This rare condition is also induced by vascular compression of the nerve root, with some cases induced by cerebellopontine angle tumor [11].

Provocation Maneuver and Home Video

The diagnosis of HFS is often hampered and contingent by whether patients show the subtle involuntary movement in question at the clinic. Furthermore, the severity of involuntary contraction is variable depending on the situation or the emotional state of the patient. One way to overcome this drawback is by using provocation maneuvers and taking home videos.

Provocation maneuvers such as prolonged voluntary contraction, repetitive forceful contraction of eyes, and repetitive smiling can help observers to visualize involuntary muscle contractions. Forceful, prolonged contraction induces involuntary contraction of the affected muscles and makes it easy to detect the typical features of HFS. Physicians can observe synkinesis of the lower facial muscles during eye closing. Having patients chew or whistle is a simple and practical maneuver inducing synkinesis in the upper facial muscles.

Given the existence of paroxysmal and fluctuating features of HFS, taking home videos is also a helpful tool to facilitate precise initial diagnosis as well as monitor disease progression and the treatment response. It is important to take videos so that physicians can check changes in the treatment response, alterations in symptoms according to various situations, and the progression of the disease over time.

Specific Signs and Red-Flag Signs of HFS

The Other Babinski Sign

The paradoxical involuntary upward replacement of the ipsilateral eyebrow during involuntary eye closing, known as "the other Babinski sign" or "Babinski sign-2," is a specific sign of HFS, with 85–90% of primary HFS patients presenting with such and with the presentation correlated with the severity of symptoms. As such, it is less common in the early stage of the disease and typically appears as the disease progresses [12–15]. Surgical treatment of HFS should be carefully decided if the patient does not show the other Babinski sign. The other Babinski sign may be observed not only during involuntary periocular movement but also in the resting state; therefore, the eyebrow of the affected side is relatively raised more than the unaffected side in the resting state.

Synchronous Contraction of the Upper and Lower Facial Muscles

In HFS, a highly synchronous involuntary contraction in all involved muscles can be observed. This synchronous contraction is a feature of HFS that distinguishes it from other hyperkinetic movements. Patients with postparalysis facial synkinesis (PPFS) can show synchronous contraction, but what is different from the synkinesis of HFS is that the synkinesis in patients with PPFS is seen during voluntary movement of facial muscles, in contrast with that in HFS induced by involuntary contraction [16, 17].

Red-Flag Signs

Despite HFS having characteristic symptoms, sometimes it is difficult to distinguish the condition from other hyperkinetic facial movement disorders, particularly in the early stage of the disease. As such, there are several "red-flag signs" of involuntary facial movements that suggest another hyperkinetic condition other than HFS is at play. The physician should consider the possibility of other hyperactive facial movement disorders if the patient is of a young age at the time of onset, has bilateral involvement, had a past history of peripheral facial palsy, the condition initiated with the lower facial muscles, there are accompanying hyperkinetic movements in body parts other than the face, Charcot's sign is visible, and there is involvement of the masseter muscle or involuntary movement of the tongue. However, the presence of identified red-flag signs does not always indicate a diagnosis than HFS because HFS can also appear paired with unusual, atypical symptoms.

Differential Diagnosis of Hemifacial Spasm

Blepharospasm

Blepharospasm is an involuntary, spasmodic, synchronized contraction of the bilateral orbicularis oculi muscles. Occasionally, involuntary contraction spreads to the lower face as part of a condition known as Meige syndrome. Few patients with blepharospasm show asymmetric symptoms and a small number of patients with HFS present with bilateral symptoms. Therefore, sometimes it can be difficult to distinguish the two conditions. In general, patients with blepharospasm show bilateral symptoms from the beginning of symptom onset, while those with HFS experience a delay between the involvement of one side and the other side. In addition, the contraction of bilateral orbicularis oculi muscles is asynchronous in HFS and is distinct from blepharospasm, which is characterized by synchronized contractions of the bilateral periorbital muscles. The other clinical sign of note is lowering of the eyebrow upon closure of the eyes (Charcot's sign), suggesting blepharospasm, whereas there is a raising of eyebrows with contraction in HFS (the other Babinski sign) [18].

Postparalysis Facial Synkinesis

Postparalysis facial synkinesis (PPFS) aberrant regeneration following peripheral facial palsy triggers synkinetic facial movement and can occur in 9.1% of patients with peripheral facial palsy within 6 months of disease onset [19]. This involuntary co-contraction of the upper and lower facial-expression muscles is often triggered by voluntary, automatic, or emotional movement and can mimic HFS. HFS and PPFS have similar appearances as they involve the same muscle territories of the unilateral face. The most apparent difference between HFS and PPFS is that the synkinesis in PPFS occurs primarily following voluntary contraction of the facial muscles, whereas the synkinesis in HFS is accompanied by involuntary facial movements. Careful history-taking of facial palsy and examination, especially of the frontalis and orbicularis oculi muscles (the other Babinski sign), is important in differential diagnosis. An electrophysiological study can help to differentiate PPFS from HFS.

Facial Motor Tics

Facial tics also can mimic HFS. Facial tics are brief, stereotyped movements that are complex, multifocal, and nonrhythmic, while motor tics can involve limbs and combine with vocalization (vocal tic) and other features of Tourette's syndrome. The mean onset age of tic disorders is 6 years and 93% of patients are symptomatic by the age of 10 years. However, adult-onset tic disorders have also been reported; therefore, they should be carefully investigated in young patients with hyperkinetic facial movement [20]. Abnormal facial movement of tics can affect unilateral or bilateral facial muscles and an urge to perform the movement usually precedes the actual abnormal movement.

Facial Myokymia

Myokymia is a small, undulating, rippling movement of muscles that appears as tiny snakes wriggling just beneath the skin. The majority of cases of facial myokymia involve common benign symptoms in the general population, yet some cases show symptoms of neurologic disease such as multiple sclerosis, Guillain–Barré syndrome, pontine glioma, or episodic ataxia 1 [21]. Needle electromyography is a useful test by which to differentiate myokymia from HFS, where myokymia appears as brief bursts of doublets, triplets, or multiplets of repetitively firing motor-unit potentials without lateral spread responses.

Oromandibular Dystonia

Oromandibular dystonia (OMD) is characterized as sustained, repetitive, stereotypic involuntary contractions of the lower face, tongue, jaw, mouth, and pharynx and can interfere with speaking, chewing, or swallowing. The majority of patients with HFS present with the condition initiating in the upper face, while, in those with OMD, the lower facial muscles are mainly affected and muscles not innervated by facial nerves (e.g., masseter muscle, tongue) may also become involved. In patients with a history of exposure to neuroleptic drugs or dopaminergic antagonists, tardive dyskinesia should be considered. Abnormal movements in tardive dyskinesia are usually irregular, bilateral, and asynchronous, unlike in HFS.

Facial Myoclonus

Facial myoclonus associated with focal motor seizure or cortical myoclonus can mimic HFS. Facial myoclonus is induced by various pathologies including Möebius syndrome, Rasmussen encephalitis, structural lesion of the frontotemporal lobe, olivopontocerebellar atrophy, or vertebrobasilar dolichoectasia. Facial myoclonus can share clinical features with HFS, having brief contractions of the perioral area. Therefore, if patients have uncommon features of HFS, electroencephalography and brain imaging study should be considered for differentiating facial myoclonus.

Hemimasticatory Spasm

Hemimasticatory spasm (HMS) is a rare movement disorder characterized by unilateral, involuntary, paroxysmal contractions of the jaw-closing muscles. Spasms in HMS are painful and triggered by activities like chewing, talking, clenching of teeth, or voluntary tapping of the involved muscles. HMS involves the masseter or temporalis muscles or both, innervated by the trigeminal nerve, unlike in HFS. Electromyography recordings demonstrate spontaneous irregular bursts of high-frequency motor-unit potentials and, in some patients, a loss of inhibition in the form of an absent silent period during spasms in HMS [22].

Myorhythmia

Myorhythmia is an involuntary, hyperkinetic, rhythmic, slow (1–4 Hz) movement, usually affecting the facial muscles but which may also

affect the limb muscles. Isolated facial myorhythmia can occur in various contexts, including in patients with thalamic infarcts or Whipple's disease, patients receiving treatment with interferon alpha-2a, or in correlation with phenytoin intoxication [23]. Myorhythmia induced by Whipple's disease usually appears as oculomasticatory myorhythmia. Isolated facial myorhythmia is more rhythmic, slower, and continuous than HFS.

Functional Facial Spasm

Functional facial spasm is one of the most common presentations of functional movement disorders. Functional facial spasm is usually nonpatterned, varies in intensity, and is distractible. Patients with functional facial spasm demonstrate different clinical features from those of HFS, such as a younger age of onset (30s), more common bilateral involvement, no other Babinski sign, isolated lower facial involvement, downward deviation of the mouth's angle, and the disappearance of symptoms during sleep [24].

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The Electrophysiological Study for Hemifacial Spasm

Byung-Euk Joo

Introduction

Hemifacial spasm (HFS) is an involuntary and irregular spasm of the facial muscles innervated by the facial nerve that typically progresses in severity an extent over time. The etiology of HFS has been attributed to vascular compression of the facial nerve at the root exit zone (REZ) [1]. This led to the development of surgical treatment called microvascular decompression (MVD) for HFS, and MVD has become established as the most effective treatment for HFS now [2]. Although much progress has been made on the cause and treatment for HFS, there has been a debate about the pathogenesis of HFS despite numerous electrophysiological studies on HFS until now. There are two hypotheses for the underlying mechanism (Fig. 1): (1) as the peripheral nerve mechanism, the compression of the facial nerve by a blood vessel causes an injury of the myelin sheath, facilitating ectopic excitation and ephaptic transmission between individual nerve fiber [3–7]; and (2) as the central mechanism, the hyperexcitability of the facial motor nucleus (FMN), triggered by antidromically propagated discharges, induces a spasm [8–14].

Many researchers have studied to elucidate the pathogenesis of HFS in clinical settings using electrophysiological studies involving the lateral spread response (LSR), blink reflex test, facial F-wave, and transcranial electrical stimulation (TES). Due to the efforts of many researchers, much progress has been made in elucidating the pathogenesis of HFS over the last 40 years. In this article, the previous main researches using each electrophysiological study for HFS will be discussed together.

Electrophysiological Study

Lateral Spread Response (LSR)

Lateral spread response (LSR) is an abnormal electromyographic findings in patients with HFS [3]. The LSR is the response of the muscles innervated by the other facial nerve branches by stimulating of one branch of the facial nerve. So, LSR is the most representative electrophysiological findings of HFS and thus has diagnostic value for HFS. Also, the disappearance of LSR usually occur immediately after identifying the offending vessels and performing sufficient decompression during MVD for HFS [15]. Therefore, LSR has been used not only as the diagnostic tool for HFS but also as an indicator of successful MVD. Until now, many electrophysiological studies using LSR have been conducted to elucidate the pathogenesis of HFS and to ensure sufficient MVD for HFS with the development of intraoperative monitoring. Surprisingly, however, there is still

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_6

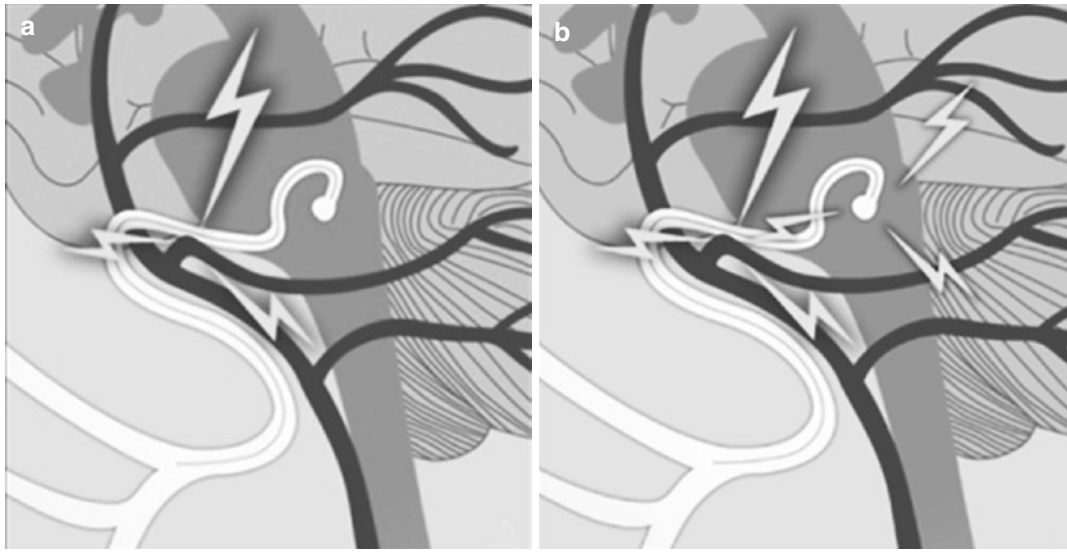

Fig. 1 The pathomegenesis for hemifacial spasm. (**a**) Peripheral ectopic excitation with ephaptic impulse transmission, (**b**) Hyperexcitability of the facial motor nucleus

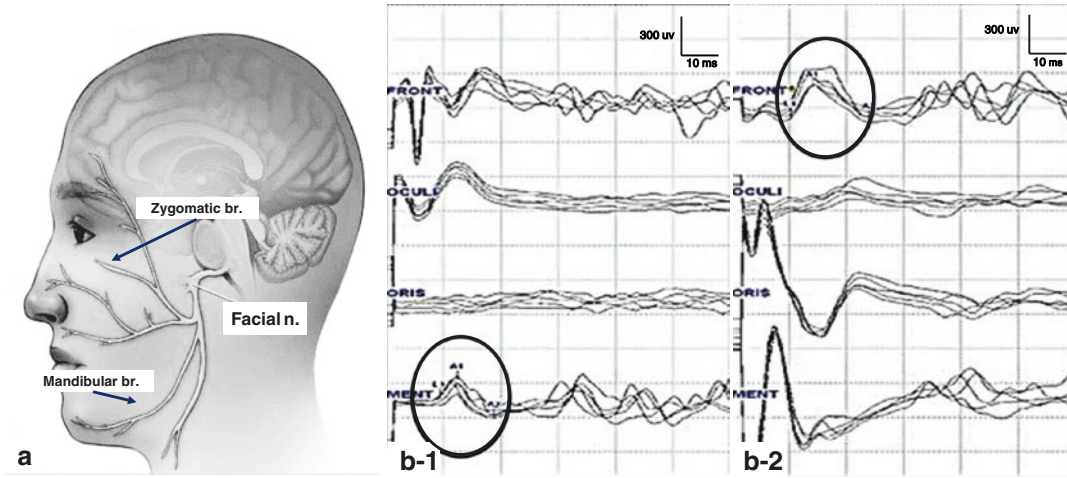

Fig. 2 The lateral spread response (LSR). (**a**) Facial nerve branch. (**b**). (1) LSR from mentalis muscle with stimulation of zygomatic branch of facial nerve. (2) LSR from frontalis muscle with stimulation of mandibular branch of facial nerve

much debate about the origin of the LSR with pathogenesis of HFS.

Methodology

Two types of LSR can be recorded by stimulating the upper and the lower branches of the facial nerve on the symptomatic side of HFS. By stimulating the zygomatic branches of the facial nerve on the symptomatic side, LSR can be recorded from the mentalis muscle. In addition, LSR can be also obtained from the orbicularis oculi muscle or the frontalis muscle by stimulating mandibular branch (Fig. 2). Constant current stimuli are applied for 0.1–0.2 ms with a bar electrode.

Resting motor threshold (rMT) is initially defined as the minimum intensity that could induce the amplitude of LSR of >10 μV in at least five successive trials. After defining the rMT, the LSR can be obtained by stimulating rectangular shock with a suprathreshold strength.

Results and Interpretation

Nielson reported that there was LSR on the symptomatic side of all 62 HFS patients, although there was no LSR on the asymptomatic side of those patients as in the healthy controls [3]. In this study, LSR on mentalis muscle was observed in all 62 patients, and that on the orbicularis oculi muscle in 60 of the 62 patients. The latency of LSR was an average of 9.3 ± 0.13 ms and 9.0 ± 0.13 ms for the orbicularis oculi and the mentalis muscle. Also, the amplitude of LSR was always much smaller about 20–30% than that of the maximal orthodromic response after stimulating the facial nerve. After the previous study, Nielson and Jannetta evaluated LSR for 59 patients with HFS before and after MVD [6]. LSR disappeared in 23% and changed from bidirectional to unidirectional in 45% patients within 1 week after MVD surgery. Within 2–8 months after MVD, LSR was observed in 27%, and unidirectional in 17%. Through these findings, Nielsen insisted that the peripheral mechanism including ephaptic transmission is the main pathogenesis for HFS though the delayed disappearance of LSR after MVD could not exclude the hyperexcitability of FMN as pathogenesis for HFS [4]. To define the origin of LSR in HFS, Møller and Jannetta analyzed the latency of LSR from orbicularis oculi muscle under anesthesia during MVD [10]. After obtaining the latency of the LSR (11.03 ± 0.66 ms) by stimulating the mandibular branch, they simultaneously measured the latency of the response from the facial nerve near the REZ (3.87 ± 0.36 ms). They also measured the latency of the response from the orbicularis oculi muscle by stimulation the facial nerve near the REZ (4.65 ± 0.25 ms). They showed that the latency of the LSR from the orbicularis oculi muscle by stimulating the mandibular branch was larger than the sum of the conduction time from the points of stimulation of the mandibular branch to the REZ of facial nerve and from REZ of the facial nerve to the orbicularis oculi muscle (8.52 ± 0.38 ms) (Fig. 3). Through this difference of the latency, they insisted that the LSR from orbicularis oculi muscle was not a direct result of ephaptic conduction at the site of the lesion, and hyperexcitability of FMN was involved in the synthesis of the LSR. To identify the origin of the LSR, there was the study using double stimulation instead of a

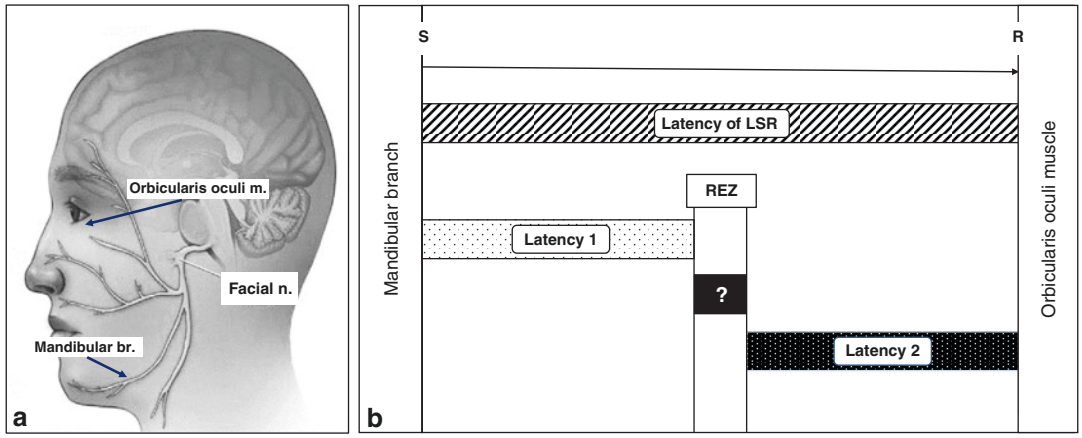

Fig. 3 The results of Møller and Jannetta's study about the origin of lateral spread response (LSR). (**a**) Schematic diagram of LSR method used in this study. The LSR was recorded from orbicularis oculi muscle by stimulating mandibular branch of facial nerve. (**b**) The difference between the actual measured value ant the calculated value of the latency of the LSR. The latency of the LSR from the orbicularis oculi muscle by stimulating the mandibular branch was larger than the sum of the conduction time from the points of stimulation of the mandibular branch to the root exit zone (REZ) of facial nerve and from REZ of the facial nerve to the orbicularis oculi muscle

single stimulus [16]. Yamashita et al. conducted double stimulation at interstimulus intervals (ISIs) ranging from 0.5 to 0.7 ms to evoke the LSR in patients with HFS. By this double stimulation, a total of 15 LSR consisting of two responses (R1 and R2) were obtained. R1 showed a constant latency and amplitude regardless of the ISIs, whereas R2 presented after a fixed refractory period without facilitation or depression in a recovery curve of latency and amplitude. As R2 showed no suppression, they suggested that LSR did not arise from FMN. To elucidate the origin of LSR, there were also studies using the central suppressive effect of anesthetics. Wilkinson et al. defined the changes in amplitude and latency of LSR according to the changes in the concentration of desflurane during MVD in 22 HFS patients [17]. According to their research, the LSR amplitude under 1 MAC desflurane with TIVA was significantly decreased than under TIVA. On the other hand, there was no change on the latency of LSR and on EEG according to the concentration of desflurane.

Blink Reflex Test

The blink reflex is the electrical correlate of the clinically evoked corneal reflex. The blink reflex is a true reflex with a sensory afferent limb, intervening synapses, and a motor efferent. The afferent limb of the blink reflex is mediated by sensory fibers of the supraorbital branch of the ophthalmic division of the trigeminal nerve (cranial nerve V1) and the efferent limb by motor fibers of the facial nerve (cranial nerve VII). Just as the corneal reflex, ipsilateral stimulation of the supraorbital branch of the trigeminal nerve elicits a facial nerve (eye blink) response bilaterally. Stimulation of the ipsilateral supraorbital nerve results in an afferent volley along the trigeminal nerve to both the main sensory nucleus of CN V (mid pons) and the nucleus of the spinal tract of CN V (lower pons and medulla) in the brainstem. Through a series of interneurons in the pons and lateral medulla, the nerve impulse next reaches the ipsilateral and contralateral facial nuclei, from which the efferent signal travels along the facial nerve bilaterally. The blink reflex has two components, an early R1 and late R2 response. The R1 response is only present on the side of stimulation, while the R2 response typically is present bilaterally. The R1 response is thought to represent the disynaptic reflex pathway between the main sensory nucleus of V in the mid pons and the ipsilateral facial nucleus in the lower pontine tegmentum. The R2 responses are mediated by a multisynaptic pathway between the nucleus of the spinal tract of V in the ipsilateral pons and medulla [18]. As mentioned above, because the blink reflex pathway is well known, and the pathway includes the entire facial nerve including FMN, many studies performed blink reflex study to clarify the pathogenesis of HFS.

Methodology

The blink reflex test is basically conducted in the method suggested by Kimura [19]. The cathode of the stimulating electrode is placed at the supraorbital foramen and the anode was placed immediately above (on the forehead), using a bar stimulator. The recording electrodes are placed over the orbicular muscle of both eyes (the mid-lower eyelid and the temple). To avoid stimulation of the nerve during spasm, stimulation was applied when the muscles were electrically at rest. Constant current stimuli of 0.1–0.2 ms in duration were delivered. After defining the threshold that could cause the minimal constant response, a suprathreshold stimulation is applied. To ensure the reproducibility and accurate response, at least five stimuli are applied to each side and then averaged. The latencies of the ipsilateral R1 and R2 responses were defined as the shortest time to the onset of the response. The maximum amplitude and duration of each of R1 and R2 responses were measured.

Results and Interpretation

Nielsen conducted the blink reflex study in 62 patients with HFS [5]. In this study, the latency and amplitude of the R1 response on the symptomatic side were increased as compared with the asymptomatic side and controls ($p < 0.001$). The latency of R1 response on the symptomatic side was increased by 2.1 ms than that of asymptom-

atic side, which was interpreted by focal demyelination over the lesion. Also, all patients showed a synkinetic response in the mental muscle on the symptomatic side, and after-activity and late-activity was observed after the reflex response. Based on these findings, ephaptic/ectopic excitation due to compression and demyelination of the facial nerve was proposed as the primary pathogenesis for HFS [4, 5]. However, Esteban et al. presented other results from the previous study using the blink reflex study. They also measured the values of the blink reflex study in the 53 patients with HFS, and then compared with healthy controls [8]. In this study, the latency of R1 response was not different between groups, and the latency of R2 response was shortened on symptomatic side in HFS patients unlike the results of Nielsen's study. Also, the duration of R2 response was greater on the HFS side when compared with those of asymptomatic side and healthy controls. So, they insisted that the hyperexcitability of FMN was the main pathogenesis for HFS. In performing the blink reflex study for HFS patients, Eekhof et al. showed findings different from the previous studies. In this study, the latency and amplitude of R1 and R2 responses from orbicularis oculi muscle present no significant difference between the HFS patients and healthy controls. However, both R1 and R2 response from the orbicularis oris muscle occurred significantly more often on the symptomatic side in HFS patients, and showed higher amplitude significantly compared to healthy controls [20]. Valls-Sole et al. studied blink reflex response in patients with HFS by applying double stimulation as well as single stimulation [21]. By applying single stimulation, the area of R1 and R2 responses was greater on the symptomatic side in patients with HFS as compared with the asymptomatic side and normal controls. Also, with double stimulation, the inhibitory effect of the conditioning stimuli upon the test stimuli R2 response, which was always observed in healthy controls, was significantly less pronounced at short ISIs in HFS. They reported that this enhanced recovery curve of R2 response was attributed to enhanced excitability of FMN in HFS. Møller and Jannetta conducted the blink reflex study under anesthesia using inhalational anesthetics (isoflurane and nitrous oxide) during MVD [22]. In this study, the R1 response on asymptomatic side was not evoked under anesthesia; however, the R1 response could be observed on the symptomatic side of HFS patients under anesthesia. Also, this R1 response of the symptomatic side was abolished after MVD. Through these findings under anesthesia, they insisted the hyperexcitability of FMN as the main mechanism for HFS.

F-Wave

F-wave is an antidromic pulse that propagates to an alpha motorneuron in the anterior horn cell of the spinal cord and then returns orthodromically down the same axon. So, the F-wave circuitry, both afferent and efferent, is pure motor. There is no synapse, so it is not a true reflex. In the extremities, F-waves have been considered as index of the excitability of anterior horn cell, and have been used as good reflection of lower motoneuron excitability. In the facial muscles, F-waves are also measurable, and those can be used for evaluation of the excitability of the facial motor nucleus. Therefore, there were many studies using facial F-waves to define the pathogenesis of HFS.

Methodology

Though facial F-waves could be obtained from orbicular oculi or the mentalis muscle by stimulating zygomatic branch or mandibular branch of the facial nerve, obtaining them from the mentalis muscle as long as possible by stimulating at the distal marginal mandibular branch is usually recommended to prevent an overlap between M-waves and F-waves. As the amplitude of facial F-wave is relatively small, it is necessary to perform the examination after the spasm has completely disappeared. After obtaining a flat baseline, stimulation was applied at the border or the mandible that was 10 cm from the stylomastoid foramen. Stimulation was performed with a bar electrode and was repeated 10–20 times using

a 0.2 ms square wave at the frequency of 1 Hz. The stimulation intensity was set to supramaximum. When a facial F-wave had a distinct peak and amplitude above 30 μV, it was regarded as F-wave. The parameters that can be analyzed using facial F-waves are as follows [9]: F/M amplitude ratio (the percentage of the peak to peak amplitude of the F-waves to the M-waves), total duration (from the initial deflection from the baseline to the final return of the F-wave), F-wave frequency (the percentage of 10–20 stimuli that produced F-waves with a distinct peak and amplitude above 30 μV), minimum latency (from the onset of the stimulus artifact to the first deflection of the F-wave from the baseline), and F chronodispersion (the difference between minimal and maximal latencies of the F-wave in a series of 10–20 waves).

Result and Interpretation

Ishikawa et al. obtained facial F-wave from the mentalis muscle by stimulating distal mandibular branch in 20 patients with HFS before MVD, and 10 HFS patients after MVD and 10 healthy controls [9]. In their study, F-wave duration F/M amplitude and frequency of F-wave on the symptomatic side of HFS patients were significantly increased when compared with asymptomatic side of HFS patients and health controls before MVD. On the other hand, there was no difference in minimum latency and chronodispersion between groups. They also showed that the enhancement of the facial F-wave eventually decreased at the same time as disappearance with LSR after MVD surgery. In another study, they compared facial F-waves from mentalis muscle and LSR from orbicularis oculi muscle by stimulating the marginal mandibular branch to investigate the origin of LSR in 10 HFS patients [23]. In this study, the LSR showed an afterdischarge after a constant response, and the afterdischarge of LSR with the facial F-wave duration tended to increase on symptomatic side of patients. Also, a lineal correlation between the facial F-wave duration and the afterdischarge duration was observed. (r^2 = 0.961, $p < 0.0001$). So, they insisted that facial F-waves and the LSR would have the same origin. Hai et al. measured LSR and facial F-waves like the previous study after creating an HFS animal model in 10 rabbits [24]. This study also presented that linear correlations between the amplitude ratio of LSR/M-waves and F-waves/M-waves and between the duration of LSR and F-waves. They reported that the peripheral mechanism including ephaptic transmission could not alone explain the increase of facial F-wave duration in HFS as the transmission time of the ephapses between nerve fiber is below 100–200 μs [25]. In another study, Ishikawa et al. conducted facial F-wave study with blink reflexes and LSR before and after MVD in 20 patients with HFS [26]. In this study, the facial F-wave and blink reflex on symptomatic side showed increased values than those of the asymptomatic side before MVD, and facial F-waves and LSR were still recorded in some patients within 1 month after the HFS had disappeared completely. Through these findings, they suggested that hyperexcitability of FMN would be the main cause of HFS.

Transcranial Facial Motor Evoked Potential (TcFMEP)

Transcranial facial motor evoked potentials (TcMEPs) are one of the most powerful tools in the intraoperative monitoring to monitor motor function, particularly for spine surgery. TcMEP are obtained by stimulating the motor pathways rostral to the site surgery. Activation of the motor pathways can be measured by recording waveforms as the impulse descends along the corticobulbar tract and corticospinal tract.

Like the blink reflex, the TcFMEP study can be a tool for examining the complete efferent pathway of the facial nerve.

Methodology

TcFMEPs from the facial muscles are elicited by using transcranial anodal electrical stimulation. Electrodes are placed on the scalp over C3 and C4 according to the international 10–20

system bilaterally. Stimulation electrodes are typically subdermal needle electrodes or corkscrew electrodes. Stimuli are applied as single shocks with a pulse width of 150–200 μs and a voltage range of 90–305 V. For the recording, subdermal needles are placed in pairs in orbicularis oculi, orbicularis oris, and mentalis muscles. Though most types of stimulation in clinical neurophysiology are cathodal, anodal stimulation in TcMEP is more effective, because the cell body and axon hillock, the sites of stimulation for TES, are more sensitive to anodal stimulation. In using TcFMEP, the TcFMEP must be excluded from analysis if the onset latency of TcFMEP is shorter than 10 ms, because they can be thought to be contaminated by direct current spread to the extracranial facial nerve [27]. By using the TcFMEP study, the threshold for FMEP as well as the latency and amplitude of FMEP is usually analyzed. The threshold of FMEP is defined as the minimum voltage required to elicit an FMEP of ≥30 μV in at least 50% of a minimum of several consecutive stimulation trials.

Result and Interpretation

Though not commonly performed, some have proposed that the myogenic facial motor evoked potentials elicited via transcranial electrical stimulation can be used to monitor the functional integrity of the corticobulbar tract, facial motor nucleus, and facial nerve during MVD surgery. Kaufmann et al. measured FMEP with LSR during MVD in 10 HFS patients and conducted FMEP study during MVD for 17 patients with trigeminal neuralgia (TN) [13]. They analyzed latency, amplitude, and duration of the FMEP before and after MVD. They suggested that the amplitude and durations of FMEP significantly decreased on the symptomatic side of HFS patients after MVD, whereas these changes were not observed from the asymptomatic side of HFS patients or TN patients. Also, they presented a dramatic reduction in amplitude and duration of FMEP with disappearance of LSR when decompression of the offending vessel. Otherwise the latency of FMEP revealed no significant change before and after MVD. In other study, they analyzed retrospectively the threshold of FMEP and the incidence of FMEP to the single pule TcMEP during surgery in 65 patients with HFS and 29 patients with skull base tumors [28]. In the study, the threshold of FMEP is significantly lower in HFS compared to skull base tumor patients. Also, FMEP to the single pulse stimulation were observed in 87% of HFS patients, whereas only 10% in patients with skull base tumor showed FMEP response to single pulse stimulation. Recently, Kaufmann et al. prospectively compared FMEP under total intravenous anesthesia (TIVA) with or without desflurane during MVD for HFS patients to define the hyperexcitability of FMN in HFS [14]. As inhalational anesthetics such as desflurane are well known for their suppressive effects on the level of the alpha motor neuron, they expected that there would be a difference in effect of desflurane on FMEP from symptomatic and asymptomatic side of HFS patients. By this study, they suggested that the suppressive effects of desflurane were less on the symptomatic side than on the asymptomatic side (59% vs. 79%, $p = 0.03$), although desflurane (1 minimum alveolar concentration) suppressed FMEPs on both sides. While showing that M-waves recorded from the mentalis muscle remained unchanged together, they also demonstrated that desflurane had no effect on the peripheral facial nerve or neuromuscular junction. Through such a series of research using TcFMEP, they suggested that the hyperexcitability of FMN might be the main pathogenesis for HFS.

Conclusion

There has been a long debate on the main pathogenesis of HFS: ephaptic transmission/ectopic excitation between individual nerve fiber vs. the Hyperexcitability of the FMN. To elucidate the pathogenesis of HFS, many electrophysiological studies have been conducted, including LSR, blink reflex test, facial F-wave, and TcMEP, so far (Table 1). Much progress about the pathogenesis for HFS has been made due to accumulation of knowledge and development of research meth-

Table 1 The summary of the main electrophysiological studies for hemifacial spasm

Researchers	Year	Subjects	The main findings	The pathogenesis[a]
Lateral spread response				
Nielson	1984	62 HFS, 14 TN	LSR was recorded on symptomatic side in all patients with HFS. Also, after-activity and late-activity were recorded on symptomatic side in HFS patients	Peripheral
Nielsen and Jannetta	1984	59 HFS	After MVD, LSR disappeared in 23% and changed from bidirectional to unidirectional in 45% patients in 1 week	Peripheral
Møller and Jannetta	1984	7 HFS	The latency of the LSR from the orbicularis oculi muscle by stimulating the mandibular branch was larger than the sum of the conduction time from the points of stimulation of the mandibular branch to the REZ of facial nerve and from REZ of the facial nerve to the orbicularis oculi muscle	Central
Yamashita et al.	2002	12 HFS	By using double stimulation, the second LSR presented after a fixed refractory period without facilitation or depression in a recovery curve of latency and amplitude regardless of the inter-stimulus intervals	Peripheral
Wilkinson et al.	2014	22 HFS	During MVD for HFS, desflurane with TIVA significantly decreased only LSR amplitude, not LSR latency than under TIVA	Central
Blink reflex test				
Nielson	1984	62 HFS	The latency and amplitude of the R1 response on symptomatic side were increased as compared with the asymptomatic side and controls	Peripheral
Esteban and Molina-Negro	1986	53 HFS, 20 HC	The latency of R2 response was shortened and the duration of R2 response was greater on symptomatic side in HFS	Central
Møller and Jannetta	1986	4 HFS	Under anesthesia using inhalational anesthetics, the R1 response was observed only on the symptomatic side of HFS patients	Central
Valls-Sole and Tolosa	1989	17 HFS	With double stimulation, the inhibitory effect of the conditioning stimuli upon the test stimuli R2 response was significantly less pronounced at short ISIs in HFS patients	Central
Eekhof et al.	2000	23 HFS, 10 PFPS, 22 HC	Both R1 and R2 response from the orbicularis oris muscle occurred significantly more often on the symptomatic side in HFS patients than HC	Central
Facial F-wave				
Ishikawa et al.	1996	20 HFS, 10 HC	On symptomatic side of HFS, F-wave duration, F/M amplitude ration, and frequency of F-wave significantly increased. However, minimum latency and chronodispersion had no difference between groups	Central
Ishikawa et al.	1996	10 HFS	On symptomatic side of HFS, the facial F-wave duration tended to increase, and a lineal correlation between the facial F-wave duration and the afterdischarge duration of LSR was observed	Central
Ishikawa et al.	1997	20 HFS	F-waves and LSR were still recorded in some patients after the HFS had disappeared completely, and then F-waves and LSR disappeared subsequently	Central
Hai and Pan	2007	10 HFS rabbits	There was a linear correlation between the amplitude ratio of LSR/M-waves and F-waves/M-waves and between the duration of LSR and F-waves	Central
Transcranial facial motor evoked potential				
Wilkinson and Kaufmann	2005	10 HFS, 17 TN	The amplitude and durations of Facial MEP significantly decreased on the symptomatic side of HFS patients after MVD. However, the latency of Facial MEP revealed no significant change before and after MVD	Central

Table 1 (continued)

Researchers	Year	Subjects	The main findings	The pathogenesis[a]
Wilkinson and Kaufmann	2014	65 HFS, 29 skull base tumors	The threshold of Facial MEP are significantly lower in HFS compared to skull base tumor patients. Also, FMEP to the single pulse stimulation were observed in 87% of HFS patients and only 10% in patients with skull base tumor	Central
Wilkinson et al.	2016	31 HFS	A significantly lower threshold of facial MEPs on the symptomatic side. Under desflurane during MVD, more less suppressive effects on facial MEPs of the symptomatic side	Central

HFS Hemifacial spasm, *TN* Trigeminal neuralgia, *PFPS* Post-facial palsy synkinesis, *HC* Healthy control, *MVD* Microvascular decompression, *LSR* Lateral spread response, *MEP* Motor-evoked potential
[a]Pathogenesis that the results of the studies favor more between the peripheral mechanism and the central mechanism

ods. Taken all the previous studies together, the hyperexcitability of the FMN is thought as the main pathogenesis of HFS.

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Magnetic Resonance Imaging Evaluation of Hemifacial Spasm

Hyung-Jin Kim and Minjung Seong

Hemifacial spasm (HFS) is one of the most common disease entities in the spectrum of neurovascular compression syndrome (NVCS) which is defined as a direct contact with mechanical irritation of the cranial nerves (CNs) by the blood vessels [1–5]. Although a wide variety of diseases are categorized as NVCS according to the affected CNs and the resulting symptoms, the evidence-based firm cause-and-effect relationship was generally recognized in only three conditions, including trigeminal neuralgia (TN) for the trigeminal nerve, HFS for the facial nerve, and vago-glossopharyngeal neuralgia for the vagus and glossopharyngeal nerves [3, 6].

After the advent of magnetic resonance imaging (MRI), it had rapidly replaced the role of conventional angiography and computed tomography (CT) for evaluation of the patients with HFS and those with other categories of NCVS such as TN. By virtue of its superb contrast resolution, MRI can simultaneously demonstrate both the CNs and blood vessels, and thus inform us of the detailed anatomic relationship between them [7–14]. Now, MRI with the aid of magnetic resonance angiography (MRA) has become the imaging modality of choice in the diagnosis and treatment planning of the patients with HFS. With the various state-of-the-art three-dimensional (3D) fast spin-echo (FSE) or fast gradient-echo (FGE) imaging techniques, we can decode the detailed neurovascular relationship that might be responsible for the clinical symptoms [4–6].

In this chapter, we will discuss the role of MRI in patients with HFS: how to image and what to look. Special attention will be paid to the importance of the anatomy of so-called "root exit zone (REZ)" of the facial nerve on MRI. The imaging issues in patients with failed microvascular decompression (MVD) will be addressed as well.

MR Imaging Techniques Used for Hemifacial Spasm

High-resolution 3D MRI has proved useful and highly accurate in the preoperative evaluation of both primary and secondary types of HFS. Recent advances in hardware and software of MRI enabled us to obtain images of high quality in a reasonably short time. Transversely oriented axial images obtained with 3D heavily T2-weighted fast imaging sequence, so-called magnetic resonance cisternography (MRC), is well suited for evaluation of the complex relationship between the nerves and vessels. On these images, the low signal intensity of the nerves and vessels contrasts with the bright signal intensity of background cerebrospinal fluid (CSF) (Fig. 1a). Based on the isotropic images obtained with the 3D technique, the images can be

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_7

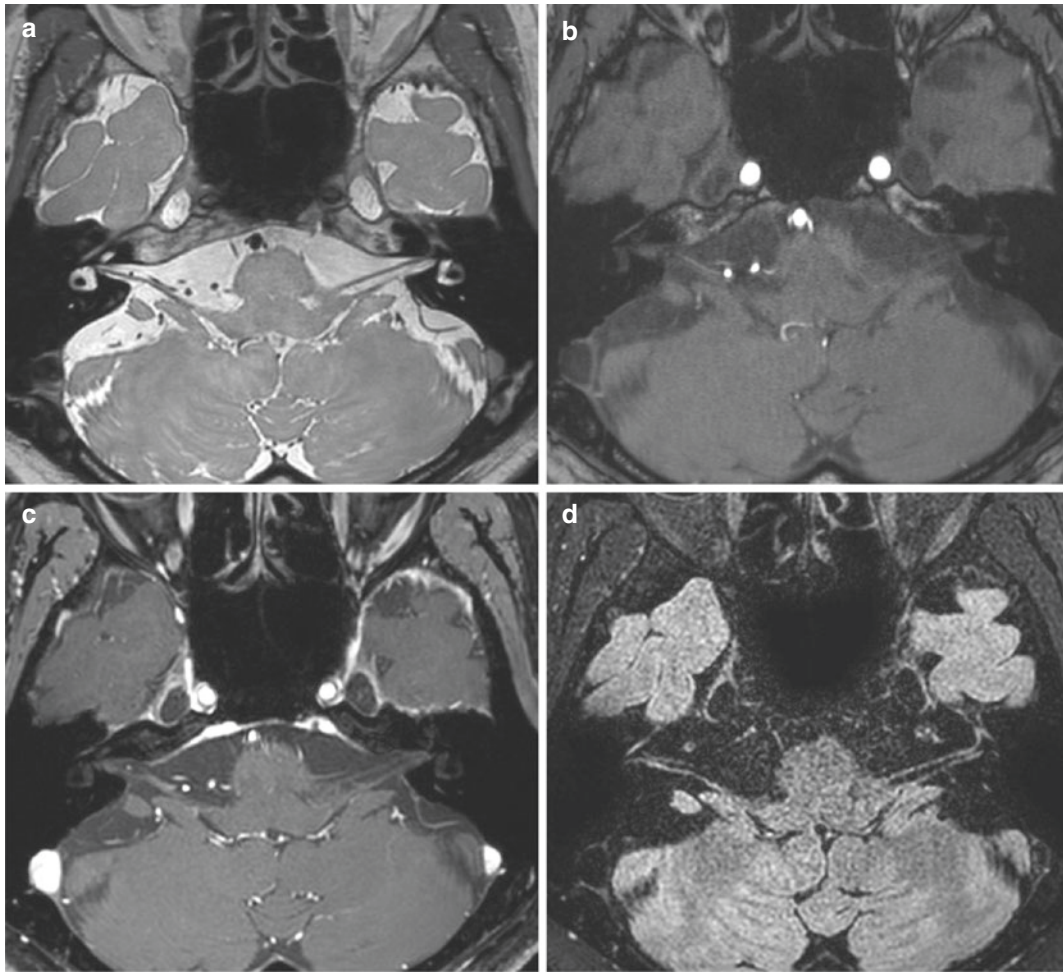

Fig. 1 Various 3D MRI sequences used for evaluation of hemifacial spasm. (**a**) MR cisternography (MRC) using 3D volumetric isotropic turbo spin-echo acquisition (T2 VISTA). (**b**) MR angiography (MRA) using 3D time-of-flight (TOF). (**c**) Contrast-enhanced T1-weighted imaging using 3D balanced turbo field echo (bTFE). (**d**) Fluid-attenuated inversion recovery (FLAIR) image using 3D FLAIR-VISTA

displayed in any planes by multiplanar reformation (MPR) to improve the precision of evaluation [7–14]. Various 3D steady-state FGE and 3D FSE sequences have been used among the different vendors, with the former including FIESTA-C, CISS, and bTFE and the latter including CUBE, SPACE, and VISTA [4–6].

3D time-of-flight (TOF) MRA is another useful MRI technique to visualize the offending arteries (Fig. 1b) [4, 5]. However, small arteries are difficult to see consistently with this technique. It is also useful for detection of high-flow vascular malformation [5]. Image fusion that combines MRC and 3D TOF MRA has been reported to display the neurovascular relationship more vividly in patients with HFS [15, 16].

3D contrast-enhanced T1-weighted imaging sequence is useful in the evaluation of HFS, especially for exclusion of secondary HFS caused by tumor, inflammation, and demyelinating disease (Fig. 1c) [4, 5]. Venous lesions that may cause HFS can also be demonstrated much better with this technique.

Fluid-attenuated inversion recovery (FLAIR) sequence is useful to evaluate the patients with secondary HFS, as seen in the patients with

demyelinating diseases, such as multiple sclerosis. By using an inversion recovery pulse, the signal intensity of fluid such as CSF is nulled and the lesion other than fluid is demonstrated as high signal intensity on the images (Fig. 1d) [5]. The superiority of the fusion images generated by combined 3D FLAIR and 3D TOF MRA has been reported for more clear delineation of neurovascular relationship in cases where the REZ of the facial nerve is difficult to trace on MRC (Fig. 2) [17]. The value of image fusion of 3D FLAIR and MRC has been reported in patients with TN as well [18].

Diffusion-weighted imaging (DWI) is also helpful in the evaluation of patients with HFS. The lesions showing restricted diffusion are seen as an area of high signal intensity on DWI and best exemplified by the diseases causing secondary HFS such as brainstem infarction and epidermoid cyst in the cerebellopontine angle (CPA) cistern (Fig. 3) [5]. Recently, diffusion tensor imaging with tractography has been

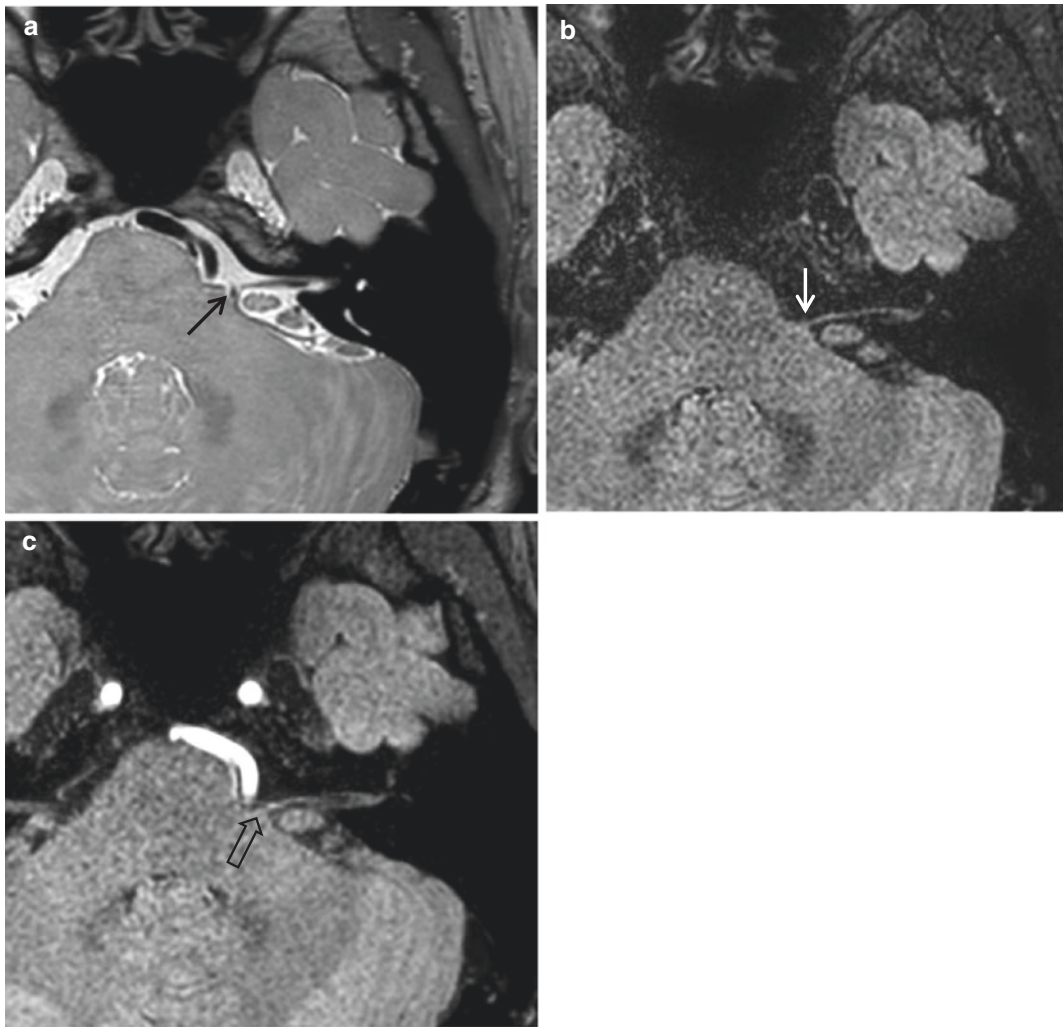

Fig. 2 Usefulness of 3D FLAIR for better demonstration of the proximal course of the facial nerve. (**a**) On MRC, the proximal portion of the facial nerve overlaps with the vascular structure, making it difficult to trace (arrow). (**b**) On 3D FLAIR, the proximal portion of the facial nerve (arrow) is well traced medially thanks to suppression of the signal from the vessels. (**c**) Fusion image of MRA and 3D FLAIR demonstrates more clear relationship between the nerve and offending vessel (open arrow)

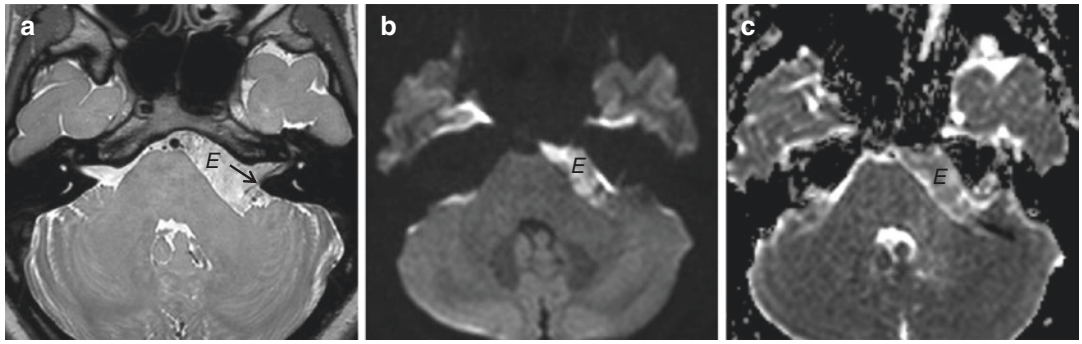

Fig. 3 Hemifacial spasm associated with an epidermoid cyst. (**a**) MRC shows a large ill-defined hyperintense cystic mass (*E*) in the left cerebellopontine angle, displacing the facial nerve laterally (arrow). (**b**) and (**c**) Diffusion-weighted image obtained with $b = 1000$ (**b**) and apparent diffusion coefficient map image (**c**) demonstrate the high signal intensity and the low signal intensity within the mass (*E*), respectively, which is related to restricted diffusion, characteristic of the epidermoid cyst

reported to be useful, especially for patients with TN [6, 19].

Anatomy of the Facial Nerve on MRI and Its Clinical Implication

The most appealing theory as to the pathogenesis of NVCS is that NVC (neurovascular compression) initiates demyelination of the nerve at the REZ, which then causes ephaptic transmission of neural impulse [2, 20, 21]. However, there has been a considerable confusion about the anatomy of the REZ [3, 22, 23]. Unclear definition of the REZ has led to a misconception of the term as synonymous with the term "transition zone (TZ)", also known as Obersteiner–Redlich zone, where a transition occurs between the oligodendrocyte-derived central myelin and the Schwann cell-derived peripheral myelin [6, 22, 23]. Anatomically, however, TZ is only one part of the REZ complex which encompasses the central myelin portion of the nerve root and the subpial portion of the nerve fascicles within the brain stem [4].

Compared to other CNs, the facial nerve is unique for its distinctive course of the REZ. Based on 75 facial nerves in 44 cadaveric brains, Tomii et al. [23] found that unlike the usual cranial nerves, the facial nerve had a long segment of the central glial portion that strongly adhered to the ventral surface of the pons after exiting the brain stem from the pontomedullary sulcus. Based on the works by Tomii et al. [23] and later on by Compos-Benitez and Kaufmann [22], the REZ of the facial nerve can be divided into four parts according to the presence of the central myelin components within the nerve: the root exit point (RExP), where the facial nerve emerges from the brain stem at the upper edge of the supraolivary fossette; the attached segment (AS), where the facial nerve adheres to the ventral surface of the pons; the root detachment point (RDP), where the facial nerve enters the prepontine cistern, separating from the pons; and finally the TZ, where the central myelin transitions into the peripheral myelin (Fig. 4a). Beyond the TZ lies the cisternal portion (CP) of the facial nerve, where the myelin is entirely derived from the Schwann cell. The reported lengths of the AS and the TZ are 8–10 mm and 1–4 mm, respectively [22–24].

Because of this unique anatomy of the REZ of the facial nerve, the oblique coronal reformatted MR images with the plane parallel to the facial nerve on the axial MRC images may be the best plane to appreciate the entire course of the REZ of the facial nerve (Fig. 4b). On the axial images, the portion of the REZ, which is localized at the junction of the facial nerve and the outer surface of the brain stem, may actually represent the RDP and the more proximally located AS cannot

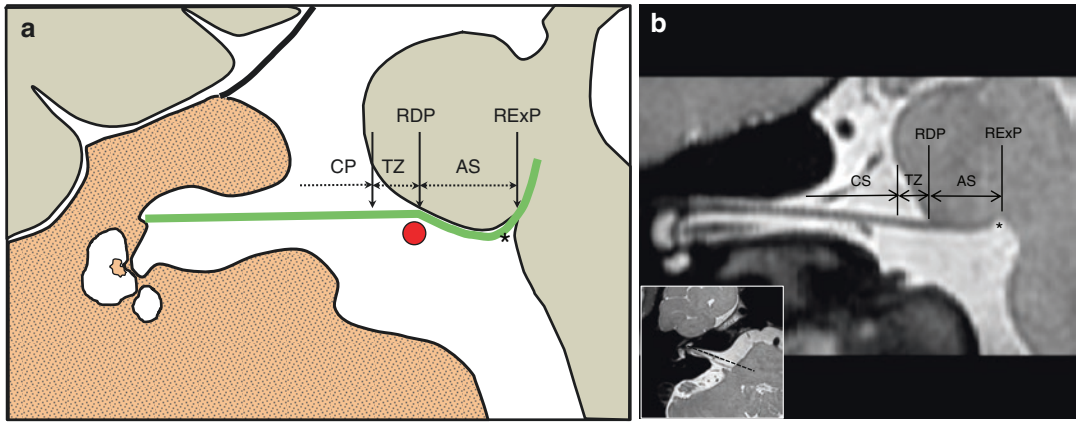

Fig. 4 Anatomy of the root exit zone of the facial nerve. (**a**) and (**b**) A schematic drawing (**a**) and corresponding MRC in oblique coronal plane (**b**) demonstrate the subdivisions of the facial nerve from the root exit point (RExP) at the supraolivary fossette (asterisk) to the cisternal portion (CP). The attached segment (AS) is a relatively long segment that tightly adheres to the ventral surface of the pons before the nerve emerges into the prepontine cistern at the root detachment point (RDP). The transition zone (TZ) is a short segment where the central myelin transitions into the peripheral myelin. The image reformation in oblique coronal plane is obtained with the plane parallel to the facial nerve on the axial MRC as shown in the inset in (**b**)

be well seen as a separate structure. This happens because the tight attachment of the AS to the brain stem frequently makes the distinction between the two structures difficult. A separation of the AS from the brain stem is still not easy even on the oblique coronal reformatted images. However, one can easily imagine the course of the AS underneath the brain stem from the RExP to the RDP on these images (Fig. 4b).

The most important implication of this peculiar anatomy of the REZ of the facial nerve in clinical practice may be that the more the proximal part of the REZ, the greater the responsibility for the site of NVC causing HFS. In a cadaveric study, Tomii et al. [23] demonstrated that NVC occurred proximal to the TZ in more than 80% of the nerves showing vascular compression and suggested that this area might also correspond to the TZ. In their study on 115 patients with MVD for HFS, Campos-Benitez and Kaufmann [22] supported the findings of the study of Tomii et al. They found that 74% of patients with HFS had NVC in the proximal parts of the REZ including AS in 64% and RExP in 10%. In contrast, in only 25% of cases, NVC involved the more distal parts including RDP/TZ in 22% and CP in 3% [22].

The anatomy of the REZ of the facial nerve is also important to evaluate the patients who have persistent symptoms of HFS after MVD. In the study on 18 patients with a failed MVD, Hughes et al. [24] reported that seven of 12 patients (58%), in whom persistent vascular compression was identified on MRI, had NVC at the AS. They also pointed out that the unaddressed vascular compression was typically proximal to the previously placed surgical pledgets in 10 of 12 patients (83%). They recommended a careful scrutiny of the AS on high-resolution MRI to identify a persistent NVC [24].

With the oblique coronal images, one can see the site and severity of NVC at the proximal portion of the REZ of the facial nerve more instantaneously than on the axial images. In a patient with a failed MVD, the NVC at the AS of the facial nerve is also well demonstrated on this oblique coronal image.

MRI Evaluation of Hemifacial Spasm

MRI Evaluation of Primary HFS

The first step of MRI interpretation in patients with HFS is to exclude the secondary causes of HFS, such as tumors, vascular lesions, demyelinating processes, and ischemic changes. If those lesions are not found on MRI, the next step should be carefully focused on the path of the facial nerve to identify the site of vascular compression, if any. To characterize the NVC in patients with primary HFS, there are several things that should be included in the radiologic report on MRI: the type of the offending (culprit) vessels, the site of NVC, and the severity of NVC.

In terms of the type of the offending vessels, NVC in primary HFS is caused by the arteries with the anterior inferior cerebellar artery being the most common vessel in 43–53.2%, followed by the posterior inferior cerebellar artery in 30.9–31% and the vertebral artery in 1.1–23% [22, 25]. HFS can be caused by multiple arteries (Fig. 5). A review of 1174 patients by Hyun et al. [25] identified multiple offending arteries in various combinations in 14.1%. In the study by Campos-Benitez and Kaufmann [22], 38% of patients had multiple vessels compressing the nerve. The venous cause is infrequent with the incidence being reported in 0.3–3% of the cases (Fig. 6) [22, 25].

The site of NVC should be addressed whether it involves the REZ or more distal portion of the facial nerve, such as CP. The reported incidence of the REZ involved in primary HFS nearly approaches 97–100% [13, 22]. To provide more detailed information on the site of NVC to the referring surgeons, it is preferred to comment on the points of neural compression according to the subdivisions of the REZ of the facial nerve, as described previously: the more the proximal site of NVC, the greater the likelihood that the vessel is a real offender (Fig. 7). This approach may help guide surgeons to perform a more optimal surgical management.

The severity of NVC can be classified as simple contact, indentation, and displacement, as the severity increases (Fig. 8). As expected, the greater the severity of NVC, the greater the likelihood that the vessel plays as a real offender [12]. When there are multiple offending vessels, each site of vascular compression should be reported one by one.

MRI Evaluation of Secondary HFS

Secondary or symptomatic HFS refers to the HFS associated with the various diseases that can cause a facial nerve damage anywhere along the facial nerve pathway. By using the various pulse sequences, MRI plays a very important role in the management of secondary HFS not only for the diagnosis of the lesions but also for a precise assessment of the extent of the lesions to help clinicians plan the best treatment for the patients. It is beyond scope of this chapter to deal with the detailed MRI findings of the various lesions that cause secondary HFS, and so only several examples are presented here.

Different kinds of pathology have been implicated as the cause of secondary HFS [5]. In causing HFS, these underlying diseases are collectively thought to induce a neural dysfunction and/or irritation of the facial nerve pathway. The incidence of secondary HFS is approximately one-fourth of that of primary HFS [15]. HFS caused by CPA tumors is rare with the reported incidence in 0.3–2.5% (Fig. 9) [5, 26]. In the study of 2050 patients with HFS by Lee et al. [26], only nine patients (0.44%) had HFS attributable to CPA tumors, including vestibular schwannoma (n = 2), meningioma (n = 5), and epidermoid cyst (n = 2). As previously shown in Fig. 2, DWI is very useful for the diagnosis of the epidermoid cyst. Although HFS associated with the demyelinating diseases, typified by multiple sclerosis, has been reported in the literature, its real incidence is not clear because of its rare occurrence [27]. The vascular lesions can also cause HFS and include the vertebrobasilar artery dolichoectasia, developmental venous anomaly (Fig. 6), arteriovenous malformation, and pial arteriovenous fistula [5]. Other uncommon causes of secondary HFS include the vascular insult, trauma, and infection/inflammation affecting the facial nerve.

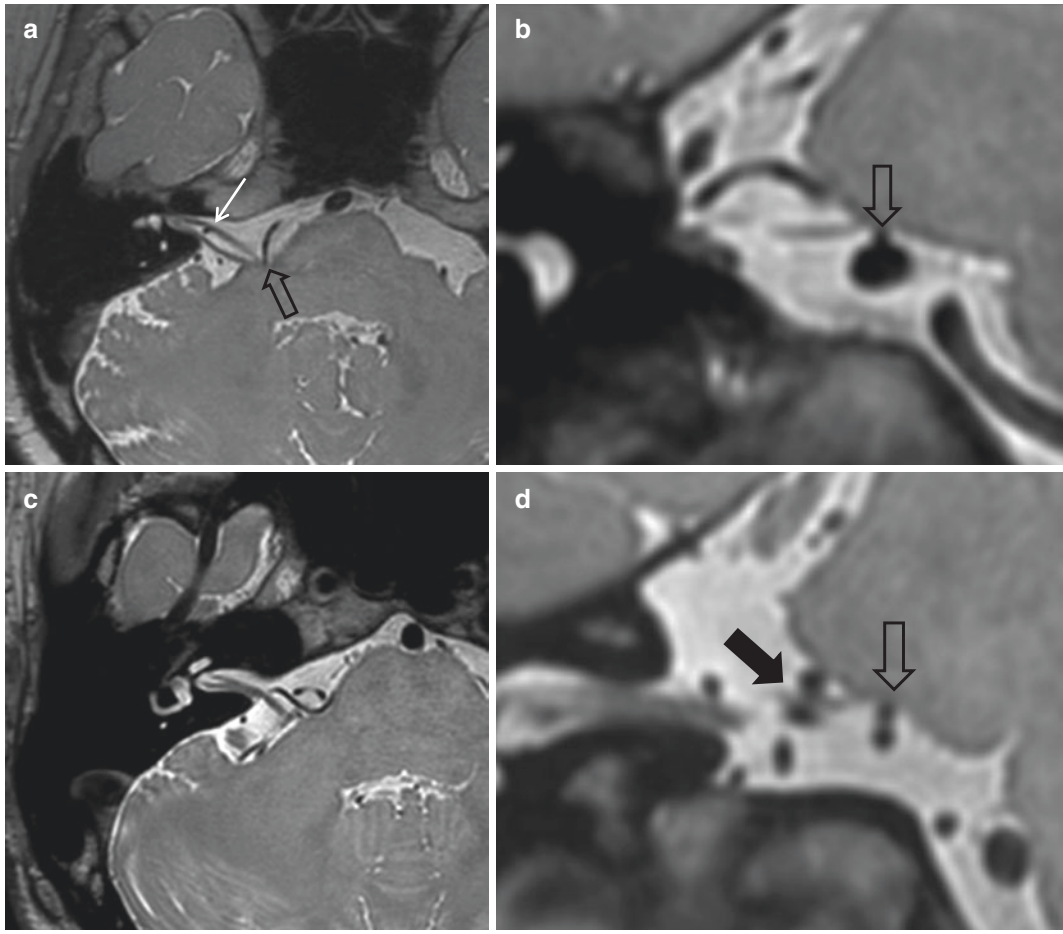

Fig. 5 Examples of various offending vessels in two patients with hemifacial spasm. (**a**) and (**b**) MRCs in axial (**a**) and oblique coronal (**b**) plane demonstrate two separate sites of neurovascular compression (NVC) along the course of the right facial nerve. The proximal site of NVC occurs in the attached segment near the root detachment point with two vessels, the anterior inferior cerebellar artery (upper) and the vertebral artery (lower), showing a tandem type of NVC (open arrow). The distal site of NVC takes place in the cisternal portion near the porus acusticus by the anterior inferior cerebellar artery, causing a mild anterior displacement of the facial nerve (arrow). (**c**) and (**d**) MRCs in axial (**c**) and oblique coronal (**d**) plane demonstrate two separate sites of NVC along the course of the right facial nerve. Proximally, a tandem type of NVC occurs in the attached segment by the anterior inferior cerebellar artery (upper) and the posterior inferior cerebellar artery (lower) in combination (open arrow). More distally, another site of NVC is found at the transition zone where the facial nerve is compressed between the branch of the superior petrosal vein (upper) and the anterior inferior cerebellar artery (lower), showing a sandwich type of NVC (thick arrow)

NVC in Asymptomatic Subjects

It is well known that a vascular contact is not infrequent on MRI in subjects with no clinical signs of NVC. In their works on MRI, Tash et al. [7] and Fukuda et al. [12] reported the incidence of the asymptomatic vascular contact on the facial nerve in 21% and 15%, respectively. According to Kakizawa et al. [28], the incidence was much higher, being reported in 78.6%. In their series, however, there was no severe deviation of the facial nerve. It may be that it is not the existence of NVC itself but the severity of NVC that determines a provocation of the symptoms. Sometimes, as demonstrated in Fig. 10, MRI shows the facial nerve that is compressed more

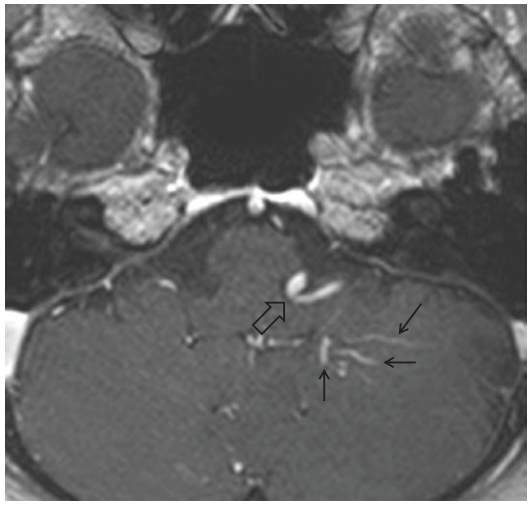

Fig. 6 Hemifacial spasm caused by developmental venous anomaly. 3D axial contrast-enhanced bTFE MRI shows multiple small collecting veins (arrows) in the left cerebellum that drain into a single dilated transcortical vein (open arrow) at the root exit zone of the facial nerve, characteristic of developmental venous anomaly

severely in the asymptomatic side than in the symptomatic side. It is critical to always refer to the clinical information to avoid such pitfalls during image interpretation, because the imaging findings are significant only in patients with the symptoms in the affected side [4].

MRI Evaluation of Persistent or Recurrent HFS After MVD

MVD is a well-established method of treatment for HFS with success rates of 90–95% for initial operation [6, 24, 25]. Despite a successful surgical decompression, however, the recurrence has been reported in up to 25% of the patients [6]. The causes responsible for the persistent or recurrent symptoms post MVD include the missed offending vessels, insufficient decompression, malposition of the surgical implant (Teflon), newly developed vascular compression, Teflon-related adhesion, arachnoid adhesion, and Teflon granuloma [29–32].

In patients with a persistent HFS after MVD, the unaddressed vascular compression is typically proximal to the previously placed surgical material. Bigder and Kaufmann [33] reported that persistent NVC was found proximal to the prior implant material in 11 of 12 patients who underwent a repeat MVD due to persistent NVC. In all 12 patients, NVC involved the REZ, including the AS in 11 patients and the RExP in three patients [33]. In 21 patients who underwent a repeat MVD for recurrent HFS, Lee et al. [30] reported that NVC was found at the REZ in 15 patients and at the cisternal segment in six patients. Reimaging with high-resolution MRI usually identifies the culprit vessels in patients with failed MVD. The predominant proximal location of NVC can be accurately depicted on MRI (Fig. 11). In 12 of 14 patients with evidence of persistent NVC after MVD on MRI, Hughes et al. [24] reported that the locations of NVC were the AS in seven (58%), RDP in one (8%), and TZ in four (33%). In 10 of 12 patients (83%), the contact occurred proximal to the existing surgical implant.

Among others, due to its histocompatibility and absorption resistance, Teflon is currently used as the material of choice in MVD for patients with NVCS including TN and HFS. Since the early 1990s, however, giant cell foreign body reaction induced by Teflon, which causes a granuloma formation, was reported in patients with TN after MVD, with an incidence ranging from 1.1 to 7.3% [34–36]. In contrast, the occurrence of Teflon granuloma after MVD for HFS has been reported much less commonly. As suggested by Chen et al. [36], the greater overall prevalence of Teflon granulomas in MVD for TN may be explained by the fact that due to a longer REZ of the trigeminal nerve, there is a greater chance of Teflon that comes in contact with the tentorium and dura, which then provokes an inflammatory reaction.

Radiologically, Teflon granuloma may present as an enhancing CPA mass on CT and MRI many years or even decades after surgery. It can be con-

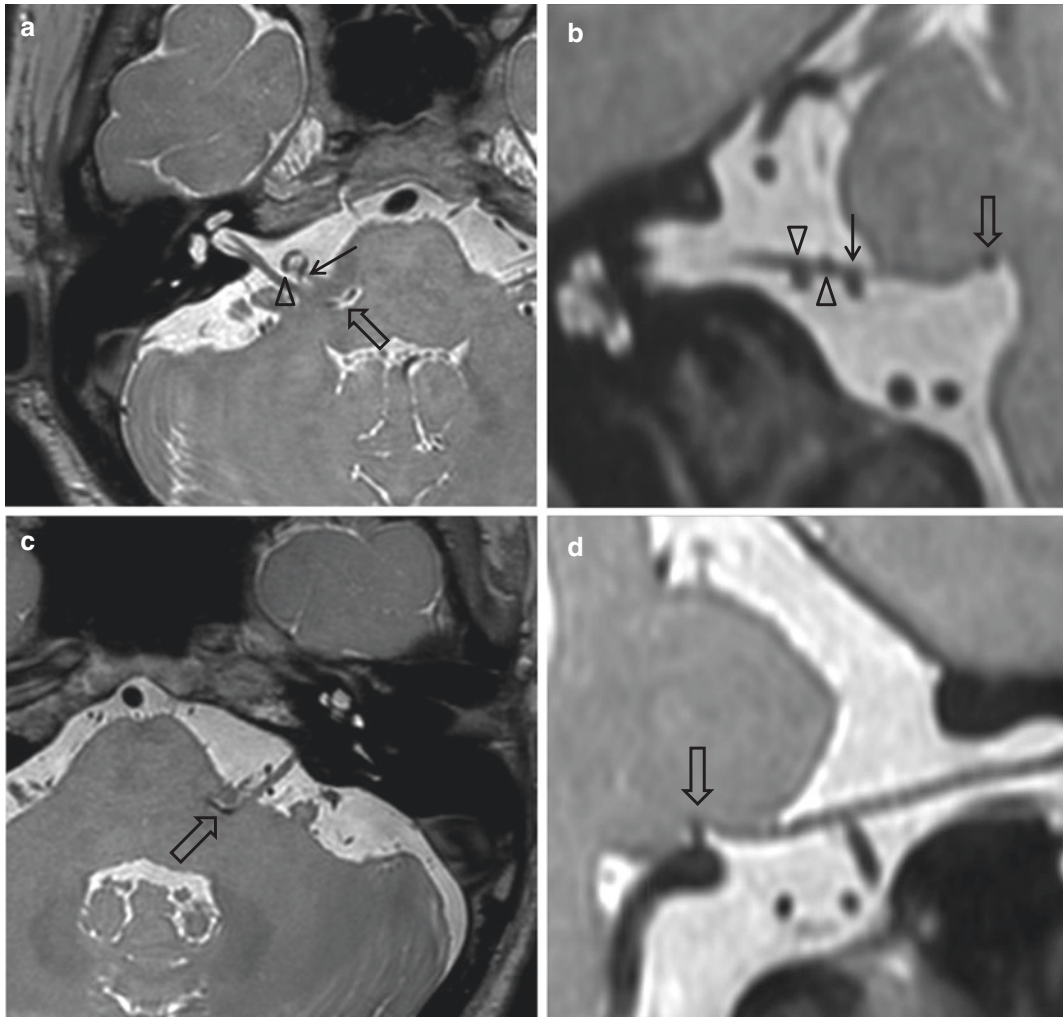

Fig. 7 Examples of various sites of neurovascular compression in two patients with hemifacial spasm. (**a**) and (**b**) MRCs in axial (**a**) and oblique coronal (**b**) plane demonstrate multiple sites of neurovascular contact by the anterior inferior cerebellar artery including the root exit point (open arrows), transition zone (arrows), and cisternal portion (arrowheads). (**c**) and (**d**) MRCs in axial (**c**) and oblique coronal (**d**) plane demonstrate a tandem type of neurovascular compression at the attached segment by the anterior inferior cerebellar artery (upper) and vertebral artery (lower) in combination (open arrow)

fused with other CPA tumors (Fig. 12) [34, 35, 37]. MRI demonstrates an oval to round heterogeneous mass that is hypointense on both T1- and T2-weighted images. Early, actively growing granuloma may demonstrate more avid enhancement, whereas older, quiescent granuloma shows minimal or no enhancement. On CT, focal calcification is often noticed. Although Teflon granuloma frequently continues to grow, associated malignant transformation has not been reported [35].

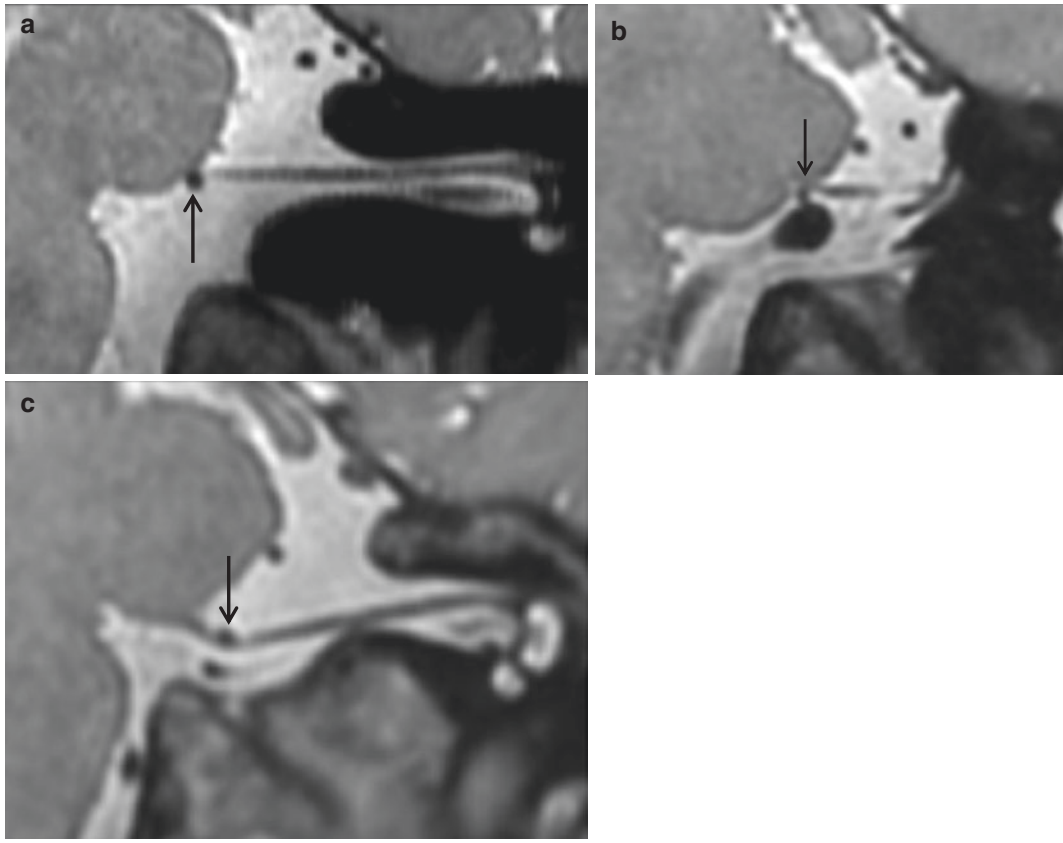

Fig. 8 Examples of the severity of neurovascular compression in two patients with hemifacial spasm. (**a**) Simple contact. MRC in oblique coronal plane demonstrates the anterior inferior cerebellar artery which is in contact with the root detachment point of the facial nerve with no evidence of pressure effect (arrow). (**b**) Indentation. MRC in oblique coronal plane demonstrates a focal indentation of the root detachment point of the facial nerve by the anterior inferior cerebellar artery (upper) and the vertebral artery (lower) in a tandem type of neurovascular compression (arrow). (**c**) Displacement. MRC in oblique coronal plane demonstrates a severe compression on the transition zone of the facial nerve by the anterior inferior cerebellar artery, causing an angulation and displacement of the nerve inferiorly (arrow)

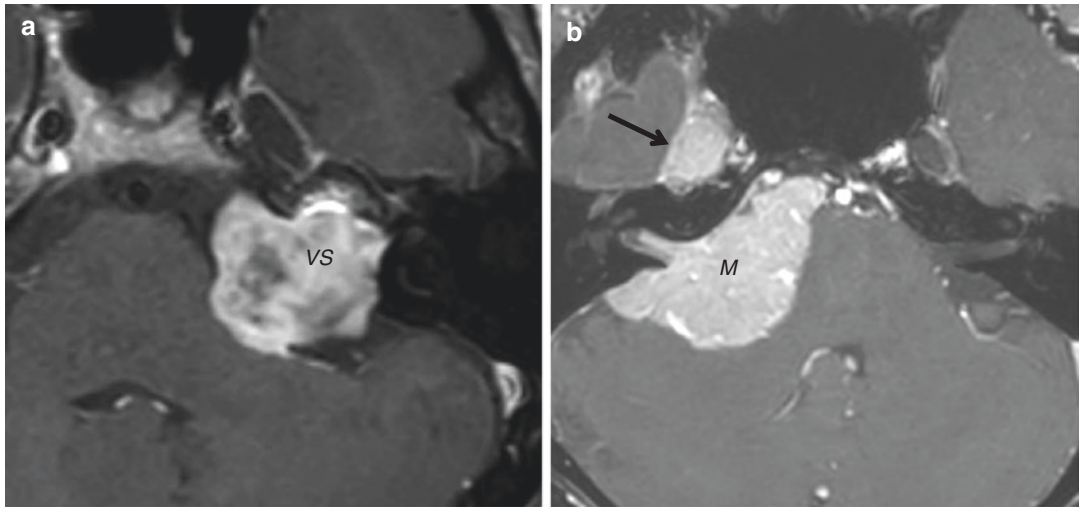

Fig. 9 Examples of secondary hemifacial spasm. (**a**) Vestibular schwannoma. Axial contrast-enhanced T1-weighted MRI shows a large lobulated soft tissue mass (*VS*) with heterogeneous enhancement in the left cerebellopontine angle. The internal auditory canal is replaced and marked widened by the mass. The brain stem is compressed by the mass as well. (**b**) Meningioma. Axial contrast-enhanced T1-weighted MRI shows a large well-enhancing dural-based soft tissue mass (*M*) in the right cerebellopontine angle. The mass partly grows into the internal auditory canal and causes significant compression on the brain stem and cerebellum. The ipsilateral Meckel's cave is also involved by the mass (arrow)

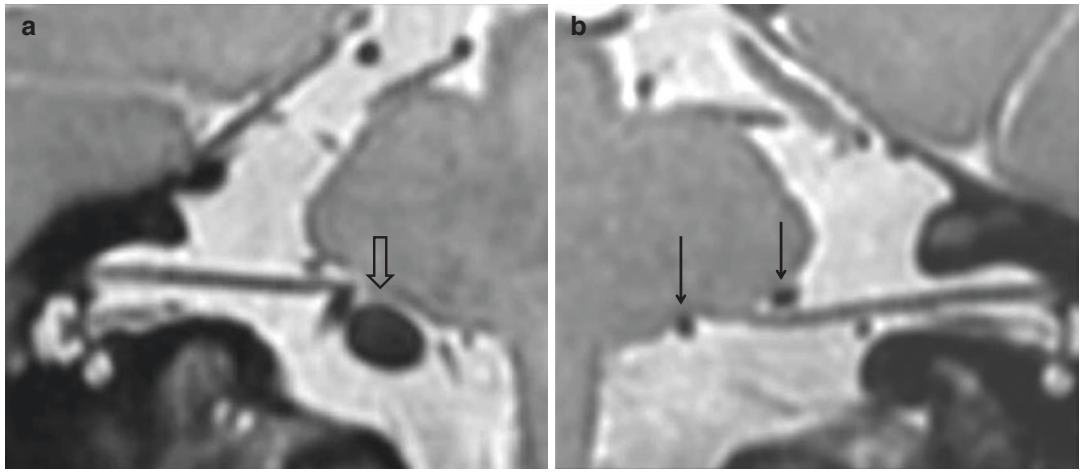

Fig. 10 Asymptomatic NVC. (**a**) and (**b**) MRCs in oblique coronal plane in the same patient demonstrate neurovascular compression in the root exit zone of bilateral facial nerves, more conspicuous on the right (open arrow in **a**) than on the left (arrows in **b**). However, the patient complained of a twitch only on her left face. The image interpretation should always be based on clinical information, because the imaging features are significant only in the symptomatic side

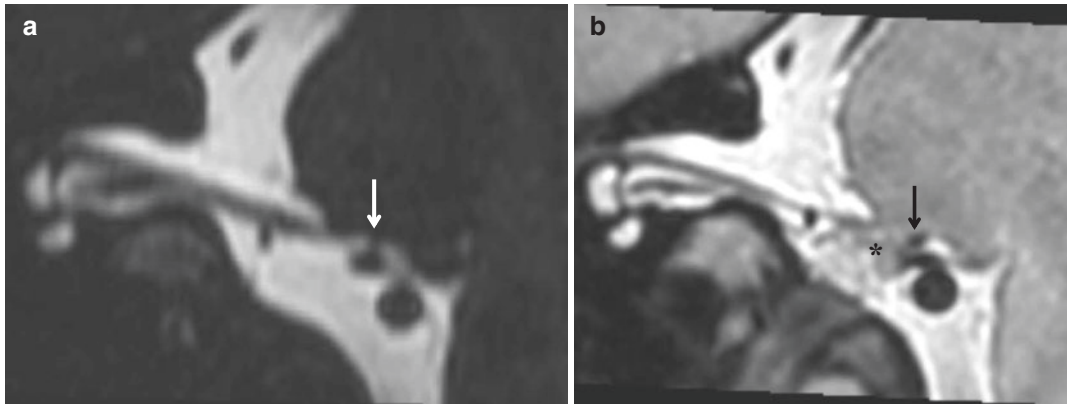

Fig. 11 Recurrent hemifacial spasm after microvascular decompression (MVD). (**a**) Preoperative MRC in oblique coronal plane demonstrates a focal indentation of the attached segment of the facial nerve by the anterior inferior cerebellar artery (arrow). (**b**) MRC in oblique coronal plane obtained 4 years after MVD demonstrates persistent neurovascular compression at the same site by the same offending artery as before (arrow). Note the position of Teflon pledgets (asterisk) which are mostly located distal to the site of neurovascular compression

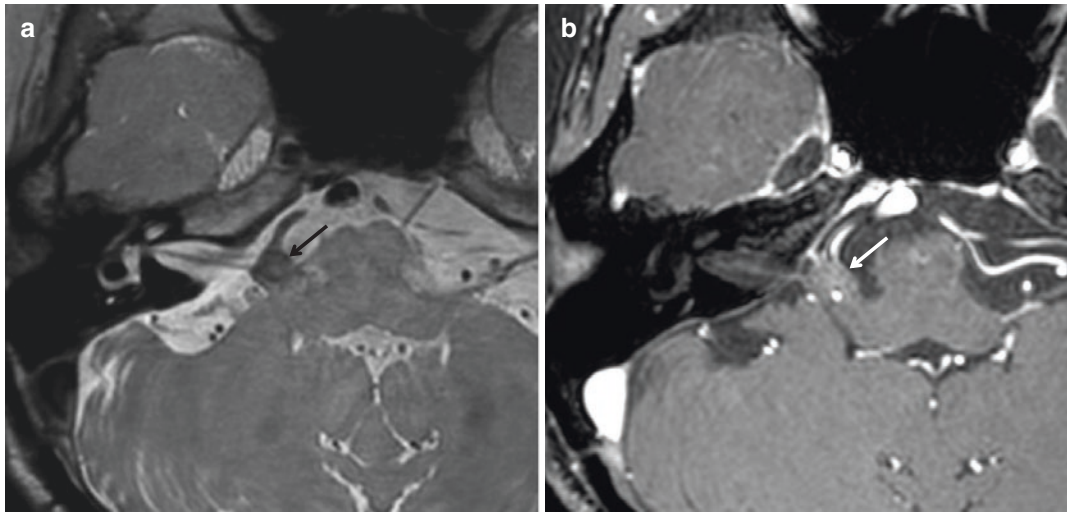

Fig. 12 Recurrent hemifacial spasm caused by Teflon granuloma after microvascular decompression (MVD). (**a**) and (**b**) Axial MRC (**a**) and 3D contrast-enhanced T1-weighted MRI (**b**) obtained 15 years after MVD demonstrate a mass at the root exit zone of the right facial nerve (arrows). The lesion shows low signal intensity on T2-weighted image (**a**) and mild homogenous enhancement after contrast injection (**b**)

Conclusion

In primary HFS, NVC initiates demyelination of the REZ of the facial nerve, which can be divided into four segments: RExP, AS, RDP, and TZ. High-resolution 3D MRI is the imaging modality of choice in the preoperative evaluation of HFS. 3D heavily T2-weighted imaging (MRC), aided by 3D TOF MRI and other pulse sequences, is well suited for evaluation of the complex anatomy at the site of NVC. On MRC, the oblique coronal plane, reformatted parallel to

the facial nerve, is the best plane to appreciate the AS where the facial nerve adheres to the ventral surface of the pons and also where NVC is frequently missed during MVD. The roles of MRI in HFS are to exclude other secondary causes such as tumors and demyelinating diseases and to identify the offending vessels in terms of the type (artery or vein), the site of NVC, and the severity of NVC. Because vascular contact is frequently seen in asymptomatic subjects, it is critical to refer to the clinical information to avoid mistakes during MRI interpretation. By correctly depicting the proximal location of NVC, high-resolution MRI is also effective in patients with HFS who suffer from the persistent or recurrent symptoms after MVD. Rarely, Teflon granuloma can cause recurrent symptoms and may present as an enhancing CPA mass on MRI that should be differentiated from true CPA tumors.

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Surgical Principles of Hemifacial Spasm: How We Do Microvascular Decompression

Seunghoon Lee and Kwan Park

Indication of Microvascular Decompression

The diagnosis of hemifacial spasm (HFS) is made largely based on clinical manifestations of the patient. Recurrent, paroxysmal, involuntary twitching of the facial muscles is the characteristic features of HFS and is manifested mostly unilaterally and less frequently bilaterally [1, 2]. For all patients who show typical symptoms and have offending vessel compressing the facial nerve identified on the MRI, microvascular decompression (MVD) can be performed when general condition permits brain surgery under general anesthesia. If the patient has minimal or atypical symptom and the diagnosis is uncertain, electrophysiologic studies such as electromyography (EMG) or nerve excitability test (abnormal muscle response; AMR) are helpful.

Secondary HFS caused by, for instance, tumors at cerebellopontine angle (CPA) can be identified on the preoperative magnetic resonance image (MRI). Majority of the secondary HFS has its own offending vessel between tumor and the facial nerve. Therefore, offending vessel underneath the brain tumor should be verified and decompressed completely from the facial nerve after tumor resection [3].

It is controversial when to perform MVD in HFS patients. Longer preoperative period might influence on the nucleus of the facial nerve and result in facial nucleus degeneration, which may explain in part the MVD failure cases comprising about 10% of the patients. However, there have been no evidence for this, and no prognostic difference between early versus late MVD was acknowledged to date. We usually recommend the MVD surgery when the patient has been experiencing this disorder to be progressive, and the tonic-clonic spasm is so severe that the patient starts to avoid social interaction and finally is willing to take the risks of brain surgery which has possible complications.

Preoperative Evaluation

The patient is proceeded to review of general condition for general anesthesia, and consulted to relevant department if there is any abnormality. We perform preoperative hearing function review by otologists in all HFS patients. Temporal bone computed tomography (CT), Brain MRI with MR angiography including 3D PD TSE (three-dimensional proton density-weighted turbo spin-echo) images are taken so that the bony structure of the skull, neurovascular relationship, and other

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_8

brain conditions are evaluated. Coronal images which are angled parallel to the facial nerve as well as axial images are of great help to understand the neurovascular relationship. Furthermore, preoperative EMG, nerve conduction study, and nerve excitability test, i.e. AMR, are routinely evaluated in every patient. We published our data that preoperative identification of AMR is helpful for better detection of AMR and higher disappearance of AMR during and after MVD [4].

Patient Position for Lateral Suboccipital Retrosigmoid Approach

We prefer a park-bench position or three quarter prone position. Compared to supine or prone position, a park-bench position requires less neck flexion in any direction, which is beneficial in patients especially with obesity, short neck, or muscular neck. After the patient is in position, we perform three-pin skull fixation with Mayfield® skull clamps; one pin is located on ipsilateral forehead and two are on occipital bone near occipital sinus above transverse sinus. Finally, head is immobilized with the head being rotated 5°–10° to the contralateral side that makes the mastoid tip on top. And a lateral tilt of neck using gravity without further force and an anterior flexion of neck allowing about two-finger breadth space between neck and jaw can make the lateral suboccipital area be widened and adequate for a retrosigmoid approach. Excessive flexion of neck should be avoided for possible airway obstruction, blockage of contralateral jugular venous return, or postoperative neck pain. At last, but not least, immobilization of the body and placing positioners at each pressure position is important to avoid any potential complication related to misposition or malposition such as compressive neuropathy, vascular compromise, and even fall-down during movement of surgical table. Ipsilateral shoulder is pulled in the caudal direction and fixated with medical plaster tape bounded to the surgical table tightly so that that the ipsilateral lateral suboccipital area can be easy to access. The contralateral arm is dropped naturally below the table and wrapped with an arm sling. Body positioners made of soft foam are placed at each point where the body and the surgical bed or instruments are touched. In the end, any additional hazard to the patient should be assessed while the surgical bed is moving; being tilted or in reverse Trendelenburg position (Fig. 1).

From Skin to Dural Opening

Using landmarks of skull such as mastoid notch, zygomatic arch, and inion, the locations of transverse and sigmoid sinus are presumed and the location of skull opening is determined. Considering the thickness of neck and hair line, a curvilinear lazy “S”-shaped skin incision line is marked. Vertical skin incision is also feasible, but it leaves surgical wound on the neck outside of the hairline and medio-caudal skin tag may block the microscopic view when a neurosurgeon tries to inspect the root exit zone (REZ) of the facial nerve in a caudal-to-cephalic direction. Skin incision starts with the depth to the fascia layer (Fig. 2a), and muscle dissection of sternocleidomastoid muscle and splenius capitis muscle in order is performed. White coarse connective tissue layer is exposed and an occipital artery can be identified below it (Fig. 2b). After coagulation and cut of an occipital artery, dissection of the scalp continues down to the skull, and profuse venous bleeding from mastoid emissary vein at mastoid foramen can be encountered. Hemostasis of bleeding from mastoid emissary vein can be accomplished using monopolar cautery or bone wax according to the size of the vein. When the vein is small, monopolar cautery alone is sufficient to control the bleeding. However, if the vein is bigger and the mastoid foramen is large not to be controlled with monopolar cautery, bone wax is used to stop the bleeding. If the foramen is so large that bone wax keeps being pushed into the foramen, the obstruction of the sinus can be anticipated when the large amount of bone wax is pushed in. Scalp dissection over the skull continues to the posterior margin of the mastoid process

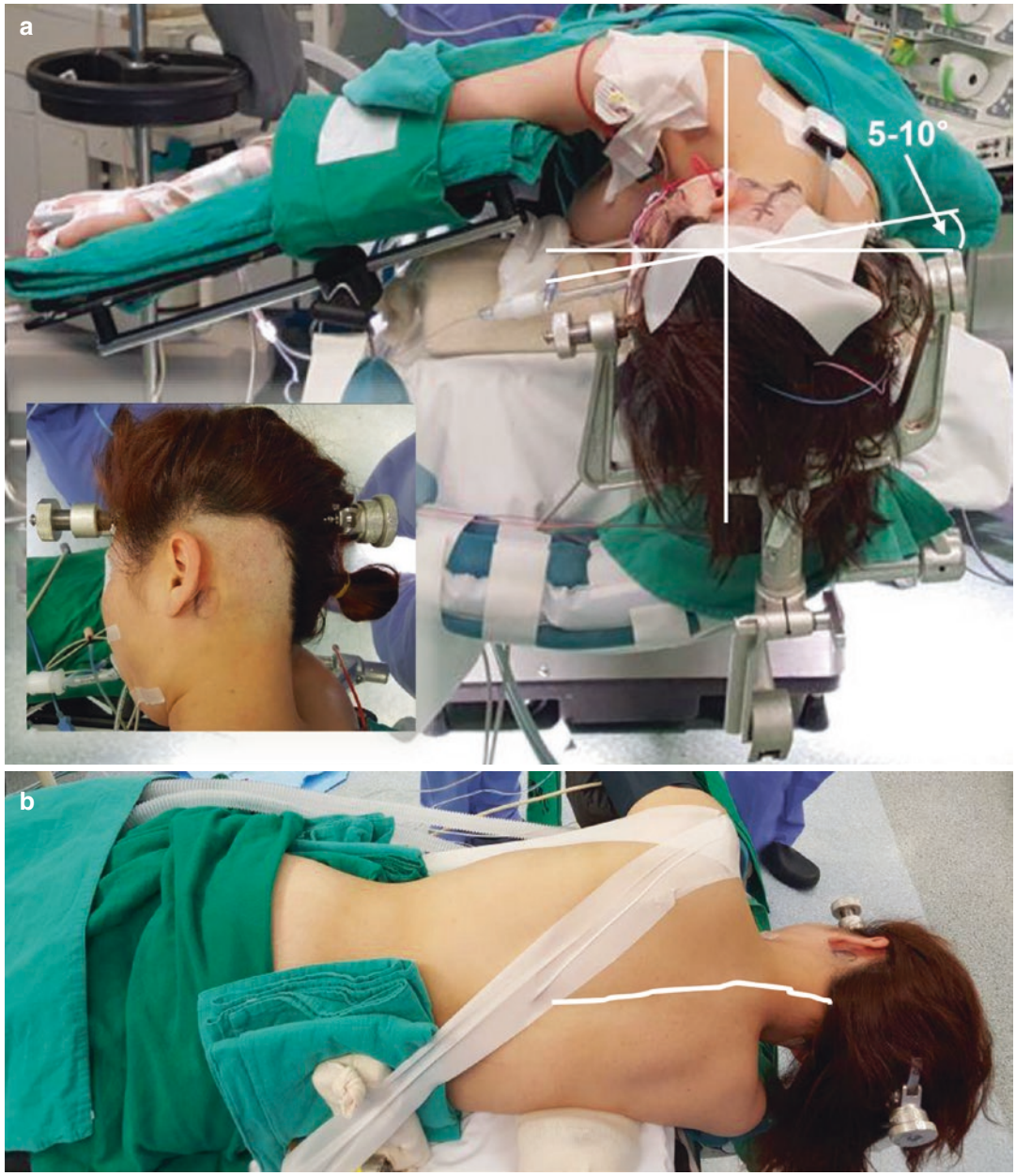

Fig. 1 Positioning of the patient for microvascular decompression. (**a**) The patient is in a park-bench position and the head is fixated with three-pin skull clamps. The head is rotated 5°–10° to the contralateral side that makes the mastoid tip on top, (**b**) and a lateral tilt of neck using gravity without further force and an anterior flexion of neck allowing about two-finger breadth space between neck and jaw can make the lateral suboccipital area be widened and adequate for a retrosigmoid approach. See text for further details

laterally, transverse sinus superiorly, and just below inferior nuchal line inferiorly. And medial limit of dissection depends on the thickness of the scalp, which enables the visualization of CPA without being obstructed by skin tag. Monopolar cautery should be used carefully below inferior nuchal line, or instead, blunt dissection with Penfield dissector #1 in a peel-off fashion is

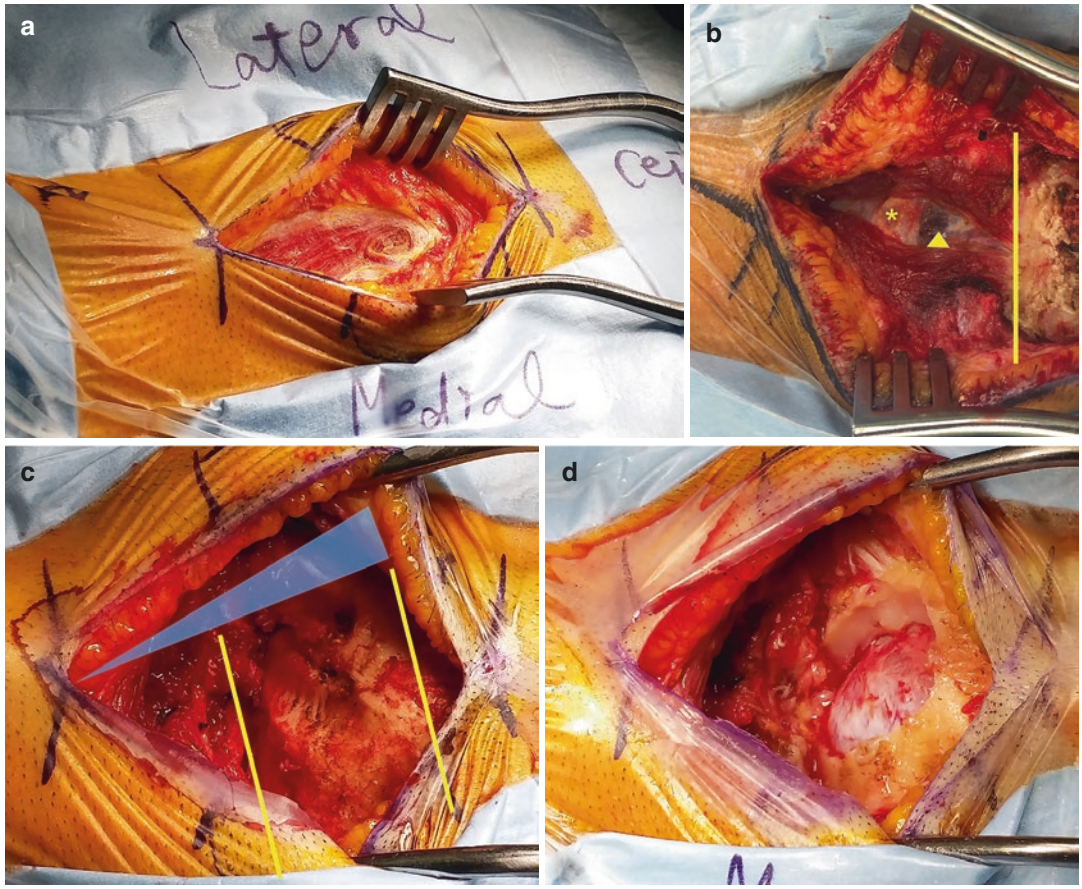

Fig. 2 Scalp and skull opening. (**a**) Skin incision starts with the depth to the fascia layer (asterisk), (**b**) and muscle dissection of sternocleidomastoid muscle and splenius capitis muscle in order is performed. White coarse connective tissue layer (filled star) is exposed and an occipital artery can be identified below it. Sometimes, mastoid foramen emissary vein (filled triangle) is so large that can be seen before an occipital artery is identified. (**c**) Scalp dissection over the skull continues to the posterior margin of the mastoid process laterally, transverse sinus superiorly, and just below inferior nuchal line inferiorly. A mastoid process is located in blue triangle, and superior and inferior nuchal lines are depicted in yellow lines. (**d**) After the scalp dissection, a craniectomy with the average size of 2.5cm × 3.5 cm is performed. (See text)

effective and safe without worrying about damage of vertebral artery or occipital condylar emissary vein (Fig. 2c).

After the scalp dissection and exposure of the adequate extent of skull, a cranial opening is performed. The size of a cranial opening is also customized according to the patient's anatomical characteristics. A 2.5 cm × 3.5 cm of a craniectomy usually gives sufficient operating space. Lateral margin of the craniectomy is to the very edge of sigmoid sinus, and superior margin does not necessarily meet transverse sinus in MVD for HFS; around 0.5 cm above the mastoid notch is enough to explore the cranial nerve 7th and 8th complex. Inferior margin confines to inferior nuchal line, and undercutting of the inner table of the skull at the inferior nuchal line is sufficient for approaching to the cisterna magna in case of difficulty in cerebrospinal fluid (CSF) drainage at cerebellopontine cistern (Fig. 2d).

Management strategy of venous sinus injury is composed of elevation of head and application of hemostatic agents. If there is profuse bleeding from the sinus tear, the position of head needs to

be higher and high-pressure suction of blood clot is required to estimate the size of injury. Use the hemostatic agent such as Surgicel® or TachoSil® as small amount as possible over the injured site and cottonoid patties are used to cover the site and press it gently. If there is larger amount of hemostatic agents used or too much pressure is applied, the size of tear can be increased. After the bleeding is reduced, surgical adhesive is applied over the injury area. Venous bleeding is bound to be ceased, that is why the neurosurgeon should remain calm and use as small amount of hemostatic agent, and as small pressure on the site as possible to control the venous sinus injury.

After cleansing out the bone dusts with saline irrigation, a dural opening is followed. A curvilinear durotomy is performed parallel to the sigmoid sinus and the dura is reflected laterally. It is easy to damage the cortical vessel of the cerebellum during durotomy, and placing a small cottonoid patty is helpful to avoid the injury. A cerebellopontine cistern is punctured and CSF drainage is pursued while the cerebellum is gently retracted using brain spatula. If the CSF is not drained enough, cisterna magna is punctured at the inferomedial direction. If the cerebellum is adequately sunken down, rubber and cottonoid patties are placed over the cerebellum to avoid damaging it when the instruments are placed in and out of posterior fossa.

Exploration Along the CPA to the Neurovascular Compression Site

Approaching to the jugular foramen is performed in the first place. Usually, there is a thick and touch arachnoid membrane covering lower cranial nerves (LCNs), which tightly anchors nerves to the adjacent tissues. When the dissection is insufficient, bradycardia can be induced and cerebellar retraction becomes difficult. Therefore, dissection around LCNs using micro-bayonet or micro-scissors needs to be thorough to the very medial and superior part of LCNs (Fig. 3a). And then, root exit and entry zones of cranial nerve (CN) 7th and 8th are gradually visualized in the cephalic direction. The corridor between the complex of CN 7th and 8th and LCNs are the route to the REZ or neurovascular compression site (Fig. 3b). Usually, broad arachnoid membrane covered over REZ needs to be dissected so that there is no hindrance of visualization. As Jannetta mentioned, retraction of cerebellum using retractor blade or suction should be directed perpendicular to the axis of the CN 7th and 8th complex, not the longitudinal direction of the complex [5]. Frequently, the CN 7th is hard to be visualized even with excessive cerebellar retraction. Head position change by surgical bed movement can be tried and helpful. Under the continuous brainstem auditory evoked potentials (BAEPs) monitoring, frequent release of retraction is required. Prolonged retraction may cause changes in BAEPs and hearing loss. And branches from anterior inferior cerebellar artery or posterior inferior cerebellar artery near internal auditory canal need to be checked repeatedly to see if there is a vasospasm. Stretching causes shrinkage of the vessel diameter, and warm saline or topical vessel dilator such as papaverine needs to be applied. A direct visualization of neurovascular compression site with indentation on the facial nerve is known to be the single-most important factor to determine the prognosis of HFS patients after MVD [6, 7]. Try hard to find the culprit near REZ of the facial nerve, and exploration can be extended to the cisternal portion or medial side of the facial nerve if there is no significant indentation on the REZ. Angled endoscope is helpful to observe the deep neurovascular compression site without excessive cerebellar retraction, especially if the patient has a prominent flocculus or complex vessels blocking the REZ.

Decompression of the Facial Nerve

We have used an interposition method in every HFS patient. Although a transposition method may be, theoretically, more definitive way of decompression of the nerve, there is no direct comparison study of the MVD outcomes between two methods. Moreover, the transposition method cannot be applied in some types of HFS, espe-

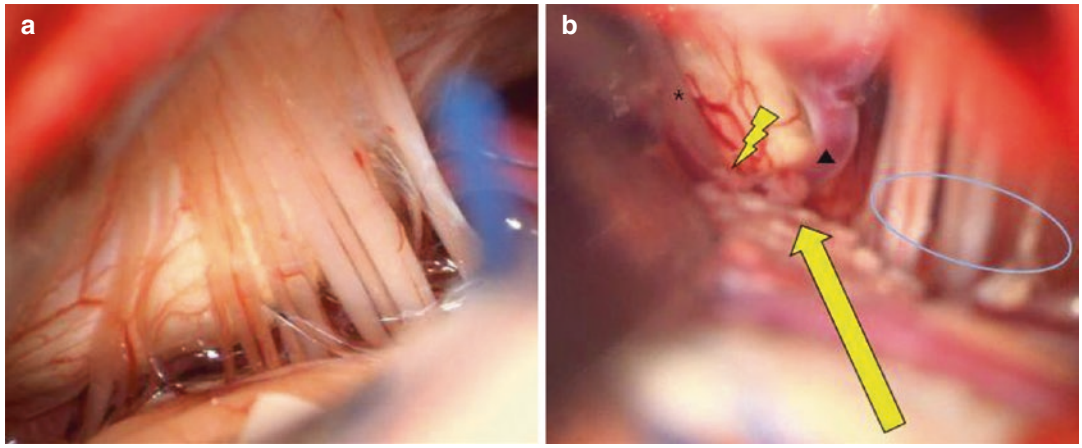

Fig. 3 Approaching to the neurovascular compression site. (**a**) The dissection around LCNs using micro-bayonet or micro-scissors needs to be thorough to the very medial and superior part of LCNs. (**b**) Then, root exit and entry zones of CN 7th (asterisk) and 8th and an offending vessel (filled triangle) are gradually visualized in the cephalic direction. The corridor (yellow arrow) between the complex of CN 7th and 8th and LCNs (circle) are the route to the neurovascular compression site (lightning bolt). *LCN* lower cranial nerve, *CN* cranial nerve

cially HFS with offending vessel having many perforators. And we think that the interposition method is reasonable in the aspect of clinical outcome; overall, 90% chance of spasm-free rate is reported using the interposition method without risk-taking of complications during struggling around narrow CPA [2, 7, 8].

Teflon felts (Teflon felt (BARD® PTFE Felt Pledget, Bard Peripheral Vascular Inc., Tempe, Arizona, USA) are prepared in three sizes (Fig. 4) and used according to the size of the affected offending vessel or anatomical working space. Adequate amount of Teflon felt should be used to decompress the facial nerve completely and to avoid possible Teflon granuloma. After complete decompression, Teflon felt is immobilized using surgical adhesive. Although surgical adhesive is going to be melting away in the end, it will work in effect during the period of brisk change in CSF dynamics during and right after the surgery. And there may be an inflammatory reaction upcoming before the adhesive is melting away, and which can prevent the Teflon felt from slippage.

Fig. 4 Preparation of Teflon felts. Teflon felts are prepared in three sizes, and used according to the size of the affected offending vessel or anatomical working space

CNs should not be manipulated directly in principle. However, some degree of manipulation cannot be avoided. A slight stretching of the CN 7th and 8th is easily observed during MVD even with the infrafloccular approach as Jannetta recommended [5]. And prominent flocculus or medially displaced REZ sometimes needs further retraction than the retraction in typical MVD surgeries. Inevitable manipulation of CNs is antici-

pated in HFS with offending vessels at medial side, in the middle of the facial nerve, or between CN 7th and 8th. For the better recovery of probably damaged CNs during manipulations, prolonged retraction should be avoided, and normal vasculature, even the small arteriole and vein, should be preserved. If there is a warning sign during monitoring of BAEPs or free-running EMG, which implies nerve irritation or damage, the cerebellar retraction should be released. Other measures which can be applied are as follows: head position is to be lowered, warm saline is applied in posterior fossa, and topical papaverine or intravenous steroid can be used.

From Dura to Skin Closure

After the sufficient decompression of the facial nerve, posterior fossa is to be filled with warm saline. If there is any active bleeding from the irrigation, exploration in posterior fossa to find bleeding point should be sought and managed properly. Usually, continuous saline irrigation or gentle compression with cottonoid patty or Teflon felt can manage the bleeding. Thereby, dural closure is initiated. Previously we used muscle plugs between the dural sutures to prevent CSF leakage [9]. Recently we are using DuraGen®, TachoSil®, and surgical adhesive to support the dural closure for the cosmetic problems, "neck depression", which is a common complaint from postoperative patients at the outpatient clinic. Dural sutures are performed with the plugged DuraGen®, TachoSil® is used as an overlay on dura, and finally, surgical adhesive is applied on it. Mastoid air cell sealing needs to be completed in the first place for sure. Then, a cranioplasty using polymethyl methacrylate bone cement is performed to fill the bony defect from a craniectomy, and artificial bone flap is fixed to the nearby skull using plate and screw. One or two surgical knots from dura are tied over the fixating plate which was bridging between the artificial bone and the surrounding skull to suspend the dura and allow the dura to adhere to the overlying bone [10] (Fig. 5).

Massive saline irrigation at lateral suboccipital area is performed to cleanse out the bone dust and to find the bleeding point of whole layers of scalp. After the irrigation and hemostasis of scalp, scalp closure starts with two or three neck muscle sutures. Fascia sutures need to be done in compact intervals. Subcutaneous layer and skin sutures are performed orderly without a surgical drain.

Postoperative Management

After the surgery, the patient is moved to the intensive care unit for immediate postoperative management. If the routine brain CT taken on postoperative day (POD) 1 shows no significant abnormality, the patient is sent to the general ward and continues to be taken care of. We routinely use postoperative steroid, methylprednisolone, for the first 24 h after MVD. The patient is discharged on POD 5, and postoperative hearing function is examined before the discharge. Regular outpatient clinic is scheduled on postoperative 1 month, 3 months, 6 months, and 12 months. After that, follow-up is performed in annual or biannual intervals. We do not have regular postoperative medication.

Use of Endoscope

Facial palsy and hearing loss are the major complications after MVD. Possible causes of them include stretching of CNs, direct trauma, and vasospasm (e.g. labyrinthine artery in case of hearing loss) during cerebellar retraction or nerve manipulation [11]. Moreover, hearing loss after MVD surgery is mainly due to the cerebellar retraction during MVD [12, 13], and we have observed that the risk of hearing loss is increased when the patient had large flocculus or medially deviated brainstem. In those cases, further retraction is required to observe and explore the neurovascular compression site. In case of facing a warning sign [14] in monitoring of BAEPs during exploration at CPA, neurovascular relationship cannot be understood in detail. Teflon felt is inserted to the point where the vessel is visualized and the instrument is reached at best. Surgical

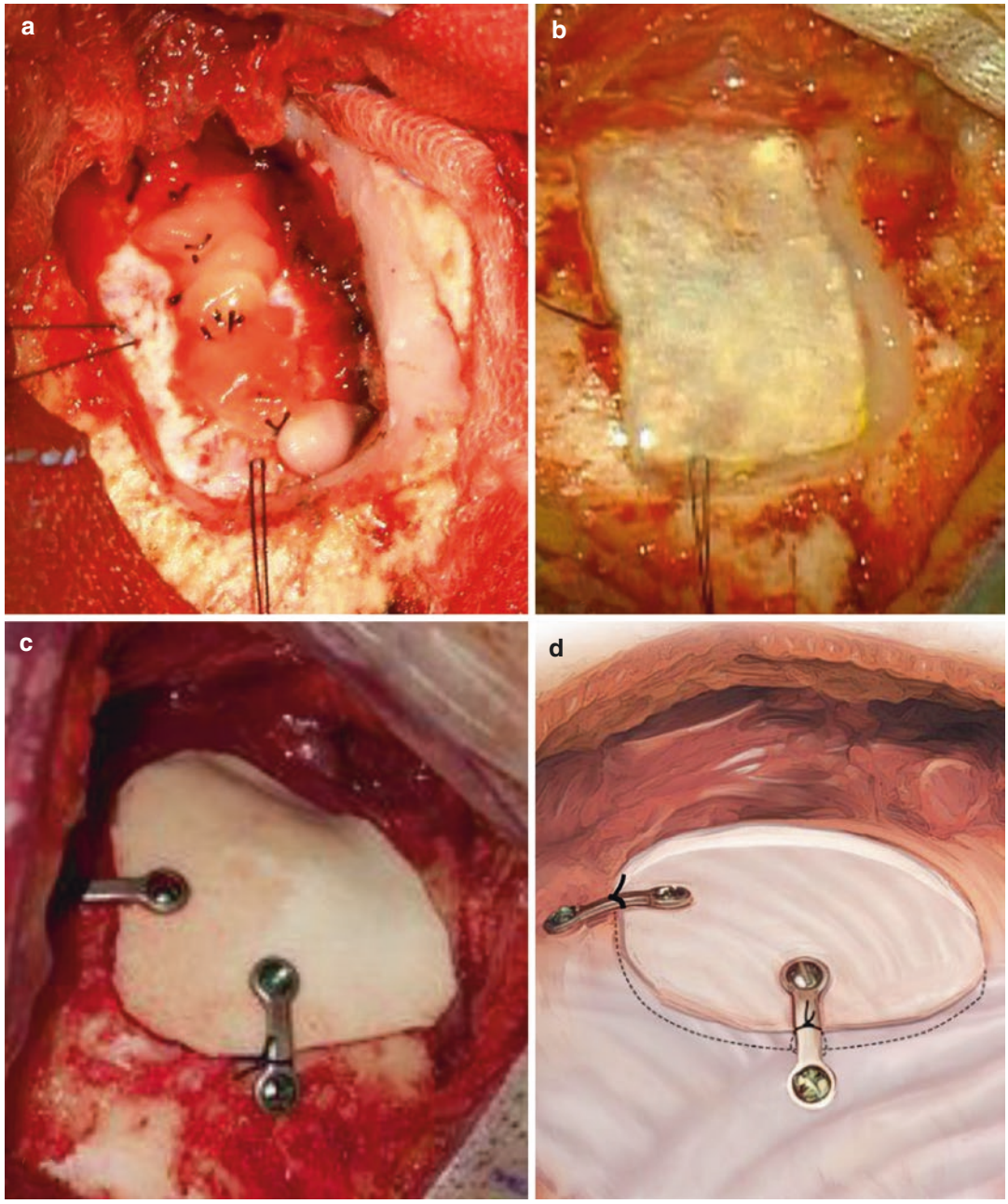

Fig. 5 Dura and skin closure. To prevent CSF leakage, we use three types of barriers. (**a**) Dural sutures are performed with the plugged DuraGen®, (**b**) TachoSil® is used as an overlay on dura, and finally surgical adhesive is applied on it. (**c**) A cranioplasty using polymethyl methacrylate bone cement is performed to fill the bony defect from a craniectomy, and artificial bone flap is fixed to the nearby skull using plate and screw. One or two surgical knots from dura are tied over the fixating plate which was bridging between the artificial bone and the surrounding skull to suspend the dura. (**d**) Illustration demonstrates how knots suspend the dura and allow the dura to adhere to the overlying bone. *CSF* cerebrospinal fluid

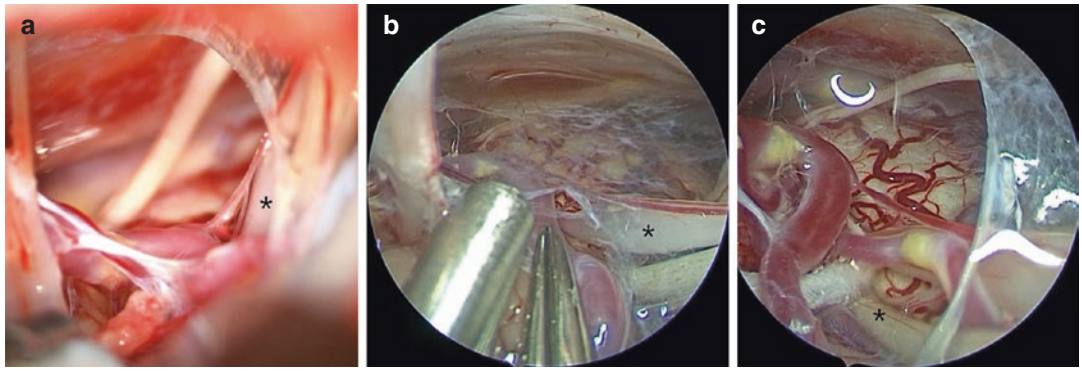

Fig. 6 Endoscopic assistance. Endoscope-assisted MVD has a great advantage for understanding the neurovascular relationship, and also for checking the decompressed facial nerve after releasing retractor or suction device. (**a**) Vessels and prominent flocculus are blocking the root exit zone of the facial nerve (asterisk) from the microscopic view. (**b**) 30°-angled endoscope shows clear neurovascular relationship in the same patient. (**c**) After placement of one Teflon felt, further exploration can be considered for further decompression using more Teflon felts. *MVD* microvascular decompression

outcome from this unclear decompression without identifying the culprit vessel can be inferior [15]. Moreover, the neurovascular relationship can be distorted when the CSF is drained and the cerebellum is retracted either by retractor blade or suction device, so that the offending vessel can be misinterpreted in some cases.

Therefore, initial observation of neurovascular compression site without excessive arachnoid dissection or cerebellar shifting is important to find the culprit offending vessel. It is the endoscope that helps in this respect, especially 30°-angled endoscope. Endoscope-assisted MVD has a great advantage for understanding the neurovascular relationship, and also for checking the decompressed facial nerve after releasing retractor or suction device (Fig. 6). If the neurosurgeon is an expert in endoscope, fully endoscope MVD can be performed, too. Small cranial opening and better visualization can minimize the operation time and maximize the surgical outcome. Although the corridor between the complex of CN 7th and 8th and the LCNs is so narrow that heat from endoscope may cause nerve injury, development of instruments and skillful hands can make up for its present shortcomings. Basic surgical skills are prerequisite for the endoscope users for the possible difficulty in CSF drain, hemostasis, or cerebellar swelling.

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Technical Difficulties of Microvascular Decompression Surgery for Hemifacial Spasm

Kwan Park and Seunghoon Lee

Offending Vessel with Perforators into Brainstem

Branching vessels from anterior inferior cerebellar artery (AICA), posterior inferior cerebellar artery (PICA), or vertebral artery (VA) at pontomedullary junction can go into the brainstem and supply it. These perforating vessels usually anchor the root vessel to the brainstem and prevent the root vessel from being mobilized (Fig. 1). If there is an offending vessel anchored to the brainstem right over the root exit zone (REZ) of the facial nerve, MVD surgery gets difficult. High elevation of the offending vessel as in a typical MVD surgery is dangerous due to brainstem infarction and hemorrhage in the cistern caused by perforator rupture. Therefore, careful inspection of the offending vessel if there is perforator anchoring the offending vessel to the brainstem is advised before elevating it up to insert a Teflon felt. During surgery, careful detachment of the offending vessel from the REZ using dissector, and the Teflon felt is inserted without elevating the offending vessel. Place the Teflon felt first, and then thrust it in between the vessel and the facial nerve. According to the location of the perforators, several small Teflon felt may be required. In this type of HFS, we can hardly observe indentation or discoloration on the facial nerve, and therefore, no other conflicts of neurovascular compression along the whole

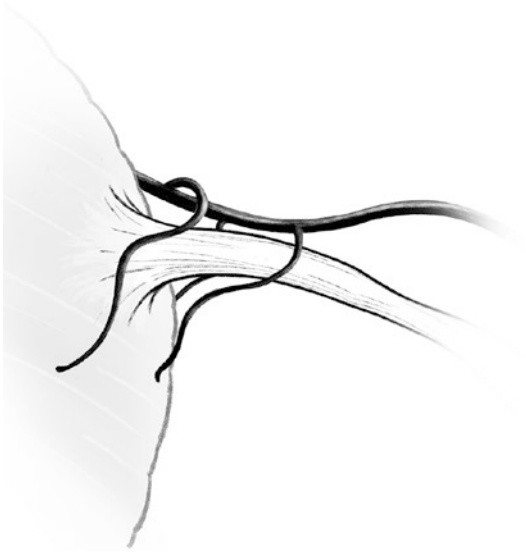

Fig. 1 Offending vessel with perforators. Perforating vessels anchor the root vessel or the offending vessel to the brainstem and prevent the root vessel from being mobilized. High elevation of the offending vessel as in a typical MVD surgery is dangerous due to brainstem infarction and hemorrhage in the cistern caused by perforator rupture. Insertion of the Teflon felt to the space between the nerve and the vessel as shown in the illustration is applicable. *MVD* microvascular decompression

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_9

axis of the facial nerve should be looked into before decompressing the facial nerve from the vessel with perforators.

Offending Vessel Encircling the Facial Nerve

Encircling offending vessel type in HFS is the offending vessel rounding around the facial nerve more than 270° in coil (Fig. 2). It will be the best decompression of the facial nerve if the facial nerve which is caught in the vessel is pulled out of the vessel. However, we could not perform a MVD in this manner in most cases with encircling offending vessel type. Rather, circumferential insertion of the Teflon felt between the nerve and the vessel is the usual method of decompression. Just as how the Teflon felt is inserted in perforator type, careful detachment of the offending vessel from the facial nerve using dissector, and the Teflon felt is inserted piece by piece circumferentially; placing the Teflon felt first, and then thrusting it in between the vessel and the facial nerve. It is better to insert the Teflon felt from the medial side of the facial nerve to the lateral side of it for avoiding the hindrance of visualization of the medial side if the insertion starts with the lateral side first (Fig. 3). The offending vessel in encircling type usually has a small diameter and vasospasm of it can be observed during MVD. Warm saline or papaverine irrigation may help to relieve the spasm.

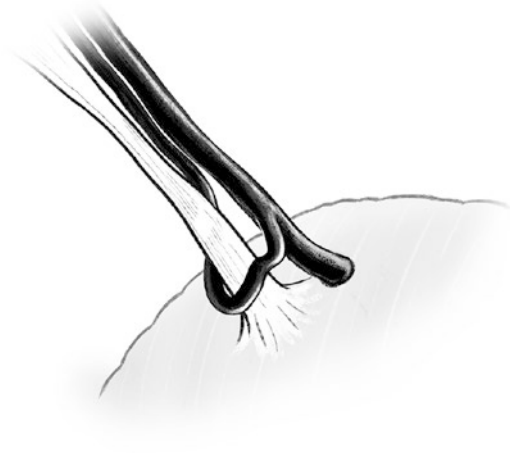

Fig. 2 Encircling offending vessel. Encircling offending vessel is rounding around the facial nerve more than 270° in coil, and compressing the facial nerve

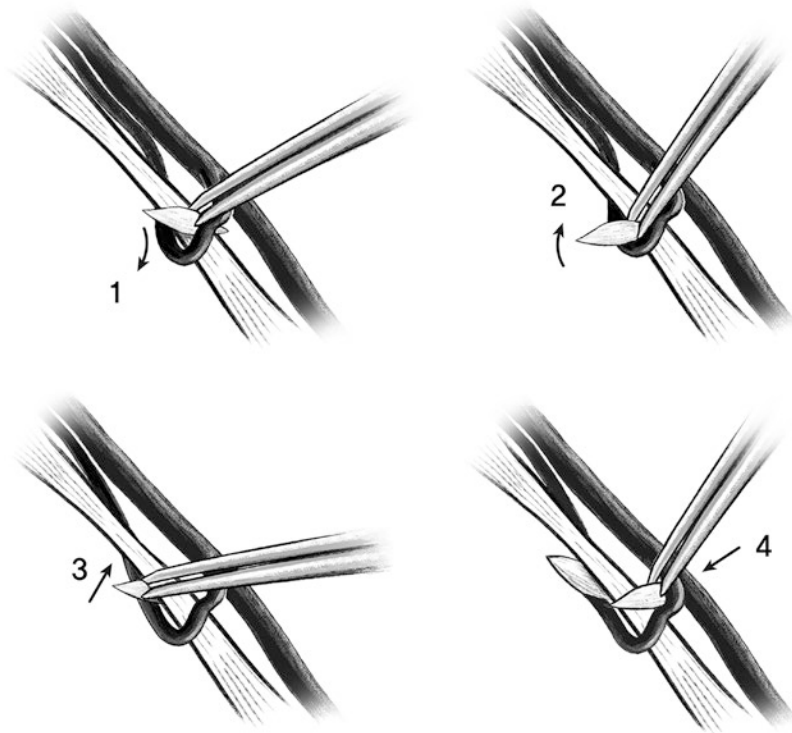

Fig. 3 Illustration of the steps of decompressing the facial nerve from the encircling offending vessel. Careful detachment of the offending vessel from the facial nerve using dissector, and a small Teflon felt is inserted piece by piece circumferentially; placing the Teflon felt first, and then thrusting it in between the vessel and the facial nerve. It is better to insert the Teflon felt from the medial side of the facial nerve to the lateral side of it for avoiding the hindrance of visualization of the medial side if the insertion starts with the lateral side first

Vertebral Artery

The VA as an offending vessel has a potential surgical difficulty during MVD due to its large diameter and stiffness. MVD for VA-associated HFS has difficulties in mobilization of itself and in visualization of the neurovascular compression site near the REZ of the facial nerve. This difficulty accounts for the decreased efficacy of MVD surgery in patients with VA-associated HFS relative to those with non-VA-associated HFS [1, 2]. If the VA has a dolichoectatic change, full REZ visualization can be hardly achieved and surgery gets more difficult. Several previous studies have demonstrated various surgical techniques to completely mobilize the VA away from the facial nerve (transposition method) [3–9]. However, those surgical methods are not universally applicable to all cases and are not easily performed by all neurosurgeons. Moreover, there is no evidence that the transposition method outdoes the interposition method in respect of clinical outcome.

We perform the MVD using the interposition method in every case, the "Fulcrum Teflon method". After dural opening, arachnoid dissection around the lower cranial nerves (LCNs) followed by further cerebellar retraction usually facilitated the visualization of the REZ of the facial nerve at the ventromedial portion of the pontomedullary junction. However, the VA was identified instead in the microscopic surgical field by pushing the LCNs toward the neurosurgeon and obstructing the REZ in VA-associated HFS. Attempts to lift up the VA toward the petrous bone high enough to expose the REZ using a microsurgical instrument often failed due to its large size and stiffness. A large piece of Teflon felt functioning as the fulcrum is placed between the proximal VA and the ventromedial brainstem near the LCNs. With the help of the fulcrum Teflon, the VA could be elevated away from the brainstem and maintained at the height of the Teflon's thickness. A surgical space could then be widened by either moving the VA distal to the fulcrum or pushing the fulcrum together with the overlying VA to further elevate the VA (Fig. 4). As shown in previous reports, majority of the VA-associated HFS had multiple offending vessels including VA [10–12]. The co-offending vessel underneath the VA and the neurovascular compression site could eventually be observed by slightly changing the angle of the microscope. Complete MVD was achieved by inserting one or two more Teflon pieces without inducing further neurovascular damages. Although we technically used the interposition method, we placed the Teflon felt between the VA and the facial nerve proximal or distal to the neurovascular compression site; hence, the site was free of both the offending vessel and Teflon felt.

Offending Vessel Located Medial to or at the Cisternal Segment of the Facial Nerve

In most HFS cases, neurovascular compression site is found at the REZ of the facial nerve. If there is no definitive offending vessel at the REZ,

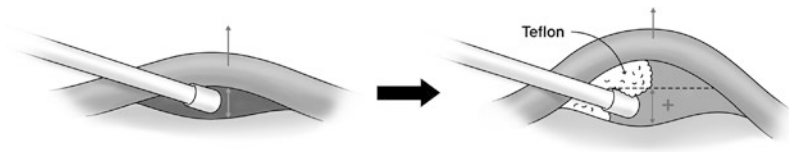

Fig. 4 Fulcrum Teflon method. Illustration of widened surgical space with the help of the fulcrum Teflon felt. Attempts to lift up the VA to expose the REZ using a microsurgical instrument often failed due to its large size and stiffness. A large piece of Teflon felt functioning as the fulcrum is placed between the proximal VA and the ventromedial brainstem near the LCNs. Then, the VA could be elevated away from the brainstem and maintained at the height of the Teflon's thickness. A surgical space could be widened by either moving the VA distal to the fulcrum or pushing the fulcrum together with the overlying VA to further elevate the VA. *VA* vertebral artery, *REZ* root exit zone, *LCN* lower cranial nerve

exploration should be extended to the cisternal segment and medial side of the facial nerve. In our previous report, we could encounter atypical locations of neurovascular compression site, which may be the cause of MVD failure. Those locations include the medial side and cisternal segment of the facial nerve instead of typical locations such as lateral side and REZ of the facial nerve [13]. Neurovascular compression sites at both locations are not easily visualized, and the chances of manipulating of the cranial nerve (CN) 7th and 8th are high during vessel mobilization and decompression of the facial nerve. Therefore, thrusting the Teflon felt between the vessel and the nerve without elevating the vessel or the nerve is the basic surgical skill used in this type of HFS for there being no large space that allows lifting-up. While Teflon felt between the vessel and the medial side of the facial nerve tends to be fixed in position (Fig. 5), it can be slipped out of the neurovascular compression site at cisternal segment if the axis of the nerve and the vessel are perpendicular to each other and conflict area is small. The Teflon felt should be placed more broadly and slippage of the Teflon felt can be prevented.

The offending vessel located at the cisternal segment of the facial nerve sometimes makes the wide-spaced CN 7th and 8th. CN 7th and 8th are located just next to each other and exit or enter through the internal auditory canal. Usually, CN 7th is almost blocked by CN 8th with the routine infrafloccular approach via the retrosigmoid approach because of this proximity of the two CNs. Moving the head position or microscope angle enables the visualization of the lateral portion of CN 7th. However, if the offending vessel is located between CN 7th and 8th, the CN 7th can be moved for, up or downward direction. Preoperative MRI showed elongated and bent CN 7th and large space is shown between CN 7th and 8th (Fig. 6). Unusual location of the CNs may mislead the identification of each CN, and REZ of the facial can also be deviated to the direction where the facial nerve is pushed. During MVD, CN identification can be aided by facial nerve direct stimulation. If the course of the offending vessel can be changed out of the com-

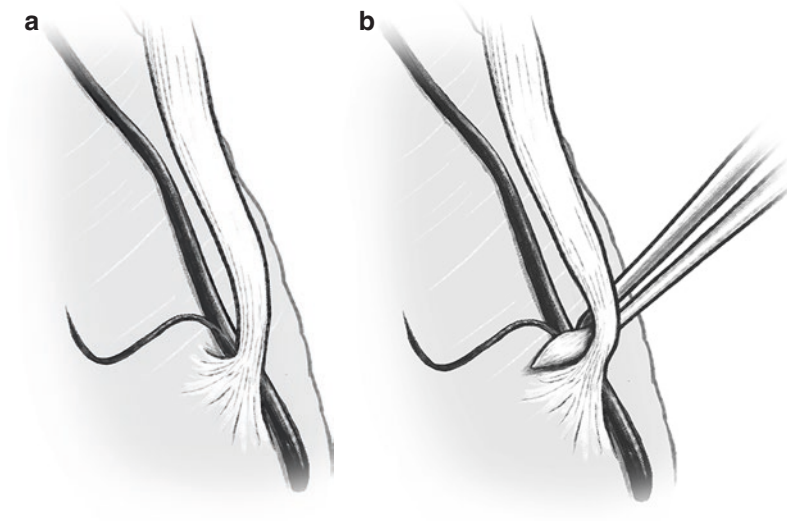

Fig. 5 Offending vessel located at the medial side of the facial nerve. (**a**) Neurovascular compression site is hardly visualized, decompression of the facial nerve cannot be performed without manipulating it, and the chances of postoperative facial palsy increase in this type of HFS. (**b**) Therefore, thrusting the Teflon felt between the vessel and the nerve without elevating the vessel or the nerve is the basic surgical skill for there being no large space that allows lifting-up. *HFS* hemifacial spasm

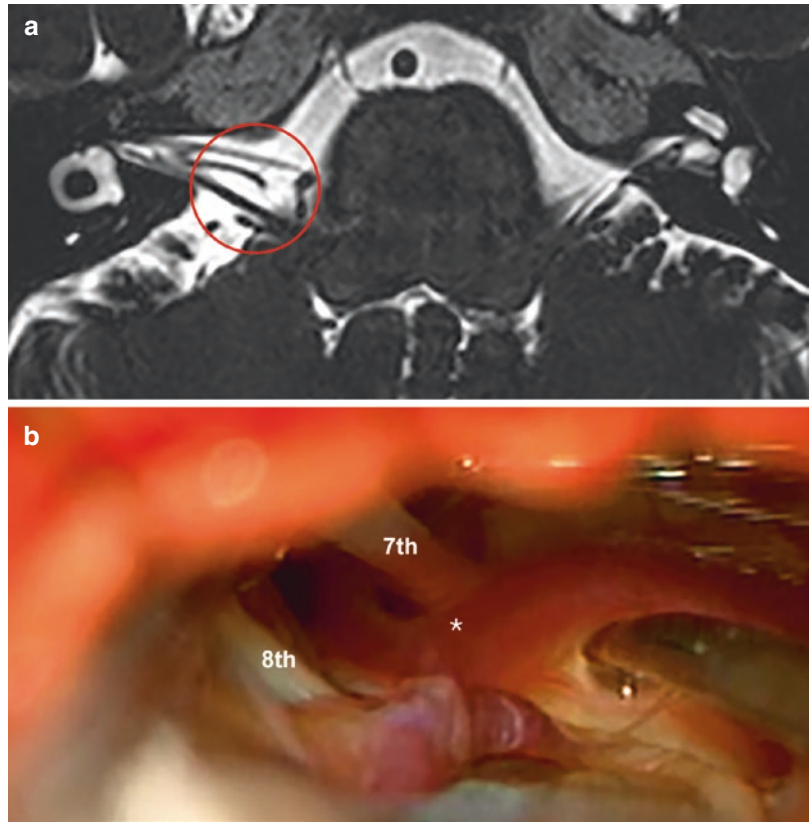

Fig. 6 Wide-spaced CN 7th and 8th due to the offending vessel at the cisternal segment. The offending vessel located at the cisternal segment of the facial nerve sometimes makes the wide-spaced CN 7th and 8th. (**a**) Preoperative MRI showed elongated and bent CN 7th and wide space is shown between CN 7th and 8th. (**b**) During operation, a medially located PICA (asterisk) was identified and it has moved the CN 7th anteriorly and made a wide space between the two CNs. *CN* cranial nerve, *MRI* magnetic resonance image, *PICA* posterior inferior cerebellar artery

plex of CN 7th and 8th, the vessel is moved away out of the complex. However, most of the cases need to be decompressed in situ. As other medial or cisternal segment offending vessels, high chance of CN dysfunction can be anticipated due to possible manipulation of them.

Penetrating Offending Vessel Through the Facial Nerve

In this type of HFS, the offending vessel literally penetrates the facial nerve (Fig. 7). There are cases where blood vessels penetrate the middle of the facial nerve or divide the nerve into main thick portion and thin small portion. It is a very rare type of HFS and only six patients were revealed to have penetrating offending vessel in our more than 4000 MVD surgical cases.

MVD in HFS patients with penetrating offending vessel through the facial nerve is thought to be the most surgically challenging and demands delicate hands. All surface of the facial nerve is theoretically blocked off from the vessel, which is not easy without damaging the facial nerve. Therefore, high possibility of insufficient decompression of the facial nerve may lead to less favorable clinical outcome, and unavoidable facial nerve manipulation has higher chance of facial palsy. Interposition of Teflon felt between the facial nerve and the vessel is pursued, and neurectomy should be avoided as much as possible. Intraoperative monitoring of free-running electromyography and abnormal muscle response is helpful to decide the extent of surgery. As in other types with narrow space between the nerve and the vessel, dissection at the plane of neurovascular conflict, followed by interposition of the Teflon felt is performed.

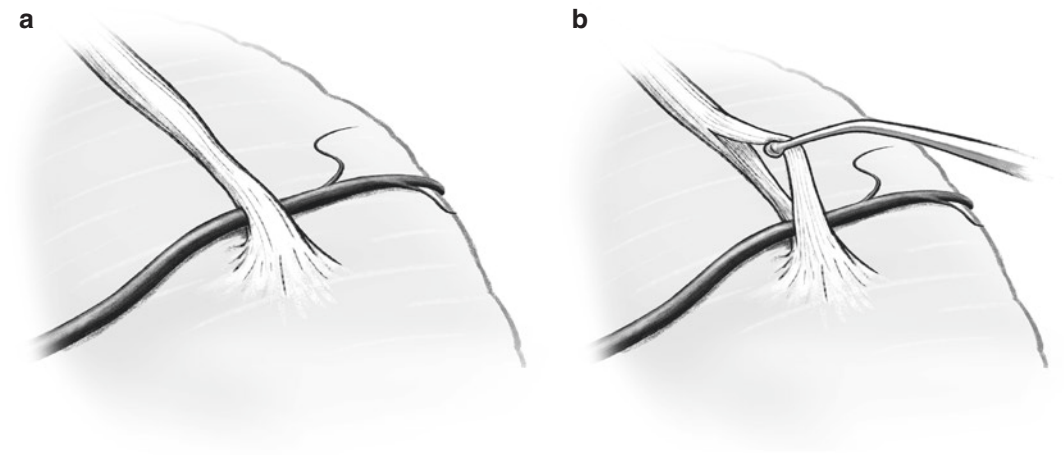

Fig. 7 Penetrating offending vessel. (**a**) Illustration shows the offending vessel literally penetrating the facial nerve. Manipulation of the facial nerve is unavoidable and this yields a high chance of the facial palsy. (**b**) The offending vessel penetrates the middle of the facial nerve

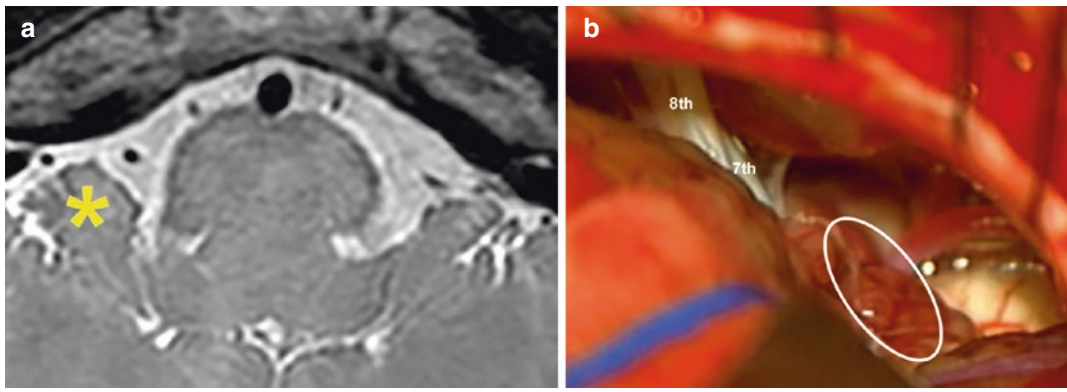

Fig. 8 Flocculus blocking the visualization of the neurovascular compression site. (**a**) Preoperative MRI showed a larger flocculus on the right (asterisk) than left side. (**b**) While approaching the neurovascular compression site, the flocculus blocked the visualization of the site and further retraction of the cerebellum including flocculus was needed. Identification of the indentation on the facial nerve failed in this patient, and frequent changes of the BAEPs were noted without resultant hearing loss after the surgery. *MRI* magnetic resonance image, *BAEP* brainstem auditory evoked potential

Prominent Flocculus Blocking the Neurovascular Compression Site

As Jannetta [14] mentioned earlier, most of the MVD for HFS is performed in infraflocculus corridor. After the dissection of the arachnoid membrane at the level of jugular foramen, we follow the corridor between the complex of CN 7th and 8th and LCNs. At this point, large flocculus can block the visualization of the complex of CN 7th and 8th (Fig. 8). Inevitably, retraction of the cerebellum including flocculus needs to be performed further, which can sometimes result in changes of brainstem auditory evoked potentials and hearing loss. Moreover, difficulty in identification of the neurovascular compression site in detail gives insufficient decompression of the facial nerve and less favorable clinical outcome.

In this regard, angled-endoscopic view provides the neurovascular relationship in situ without retraction, and reduces the retraction time during exploration of the neurovascular compression site. After the neurovascular relationship is fully understood, the culprit vessel is moved away and the Teflon felt is inserted quickly while retracting the flocculus. Flocculus can be resected to visualize REZ and perform MVD. But severe nystagmus or dizziness can be troublesome for patients, and we avoid resecting it.

Redo Surgery

Due to adhesion all along the cerebellopontine angle to the REZ and the possible higher complication rate from it, decision to perform a redo surgery tends to be made reluctantly for all neurosurgeons in general. Wider cranial opening is necessary, cautious arachnoid dissection should be guaranteed before cerebellar retraction, and it is unnecessary to remove all Teflon felt. Unusual location where neurovascular compression occurs should be inspected thoroughly. Further details will be discussed in the chapter "Redo surgery for failed microvascular decompression for hemifacial spasm" of redo surgery for failed MVD.

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Various Applications of Microvascular Decompression Other than for Hemifacial Spasm

Min Ho Lee and Jae Sung Park

Neurovascular compression syndrome (NVC) refers to a group of disorders that are caused by a direct compression on a cranial nerve by a blood vessel or vessels, and hemifacial spasm also belongs to it. After his first microvascular decompression (MVD) for trigeminal neuralgia (TN), Jannetta suggested that MVD might also be applied to other forms of NVC. Indeed, MVD is now established as the treatment of choice for HFS. This chapter presents other disease entities for which MVD may provide cure or improvement.

Trigeminal Neuralgia

Trigeminal neuralgia (TN) is a syndrome characterized by paroxysmal facial pain in the somatosensory distribution of the trigeminal nerve. Studies in the general population show that the prevalence of TN is around 0.3%, peaking in the 1960s, with some preference for female [1]. There are many treatment modalities for TN patients that are unable to obtain sufficient relief with medication. TN is the first case of neurovascular compression syndrome in which Jannetta applied MVD for treatment, and MVD is still the preferred treatment modality [2–4]. There are several large series with long-term follow-up studies have been reported, with pain free rates of 70~80%, with 5~10 years follow-up [5–9]. In addition to MVD, stereotactic radiosurgery (SRS) has also been performed, but according to recent meta-analysis, MVD was associated with greater rate of short- and long-term pain freedom, lower incidences of facial numbness and dysesthesia, and pain recurrence compared to SRS [10].

MVD for TN is quite analogous to that for HFS. With the park-bench position (3/4 lateral prone decubitus), retromastoid suboccipital craniectomy can be performed as described by McLaughlin and colleagues [11]. In general, to gain access to the trigeminal nerve, it is relatively easy to approach via superolateral aspect of the cerebellum. In the process, one should be cautious not to injure the petrosal vein, for the iatrogenic obliteration of it may cause complications including venous infarction, sigmoid thrombosis, cerebellar hemorrhage, etc., and the incidence of the complications was reported up to 6.2% [12].

TN is the condition for which MVD is most frequently performed, followed by HFS. According to a recent study [8], the patients in the TN group were older than those in the HFS group. There was no predilection as to which side of the face was affected by HFS; however, TN was reported to be present more frequently on the right side. The offending vessels were mainly the

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_10

AICA and/or PICA in the HFS group, as opposed to the SCA in the TN group. The initial response to MVD for TN did not differ from that for HFS, although the recurrence rate following the former was significantly lower than the latter. MVD is an effective treatment for both HFS and TN. MVD is a very promising intervention for HFS but is associated with a risk of recurrence when used to treat TN. The application of MVD surgery should be carefully considered in the context of these specific conditions.

Glossopharyngeal Neuralgia

Glossopharyngeal neuralgia (GPN) is a rare pain syndrome in the sensory distribution of the IX cranial nerve with a brief episodic unilateral, sharp, and stabbing pain. The glossopharyngeal nerve distribution includes the angle of the jaw, ear, tonsillar fossa, and the tongue base, and GPN accounts for 0.2–1.3% of all types of cranial neuralgias. The prevalence of GPN is estimated to be approximately 0.8/100,000 populations/year, which is far less common than TN (4.7/100,000) [13]. GPN shows a predilection for the left side of the body, whereas TN is more commonly observed on the right side [14]. The etiology of glossopharyngeal neuralgia is not entirely elucidated, but the neurovascular compression appears to be one of the main causes [15, 16].

The glossopharyngeal nerve is a mixture of cranial nerves that contain the somatic sensory fibers from the oropharynx, mastoid, middle ear, and Eustachian tube, as well as the posterior third of the tongue. The middle ear and mastoid areas are also innervated by the glossopharyngeal nerve via the tympanic branch or Jacobson's nerve [15, 17]. Like TN, carbamazepine, gabapentin, and pregabalin are first-line pharmacological treatments for glossopharyngeal neuralgia. Nerve block may be an option for GPN when refractory to medications; however, one must not trivialize the possibility of complications including dysphagia or hoarseness, not to mention arrhythmia and syncope [18, 19].

Jannetta performed MVD for six GPN patients in 1977 and reported the pain control rate of 80% [20]. Zhao et al. reported 94.3% of pain control rate based on their performance of MVD for 35 GPN patients [21]. MVD has been established as an effective and safe treatment option for glossopharyngeal neuralgia [22].

Geniculate Neuralgia

Geniculate neuralgia, also known as nervus intermedius neuralgia, is a pain in the ear triggered by sensory or mechanical stimuli at the posterior wall of the auditory canal without any pathology. The causative nerve is intermediate nerve (Wrisberg's nerve). It is a sensory component of the CN VII, containing sensory and parasympathetic fibers. The intermediate nerve joins the motor root of the facial nerve in the internal auditory canal (meatal or intracanalicular segment) to form a common trunk. The sensory auricular branch of the facial nerve arises from the vertical segment of the facial nerve, between the second genu and the chorda tympani nerve origin. This auricular sensory nerve usually arcs laterally and courses inferiorly to supply the posterior and inferior external ear canal at the region of the osseous–cartilaginous junction, as well as the inferior portions of the pinna [23].

In addition to the aforementioned TN and glossopharyngeal neuralgia, geniculate neuralgia may also be treated with medications such as carbamazepine.

Jannetta reported in 1997 that 13 patients had good results in 14 patients by sectioning intermediate nerves [24]. However, Holste et al. also had good results in 13 of 15 patients, but recurred in 6 patients with 4.8 year of median pain-free interval, which was interpreted somewhat negatively [25]. Other researchers tried surgical treatments, but geniculate neuralgia seems to have performed transection rather than MVD [26, 27]. Other researchers reported at the case report level that MVD had good results [28–31]. On the other hand, there was a case series of chorda tympani was identified and preserved through postauricular mastoidectomy and sensory branches of the facial nerve were identified and transected [32]. Taken all together, the initially, transection

of intratemporal division of the cutaneous branches of the facial nerve can be tried. If this is unsuccessful, resection of the nervus intermedius is advised; simultaneous microvascular decompression should be performed if there is also vascular compression [23].

Hemimasticatory Spasm

Hemimasticatory spasm is a rare clinical entity characterized by involuntary and paroxysmal contractions of the masseteric muscles on one side of the face. Reported results show hemimasticatory spasm more commonly presents in females in the third and fourth decade [33, 34]. The cause is the motor branch of the trigeminal nerve. Hemimasticatory spasm involves the masseter and the temporalis muscles, with the medial pterygoid muscle also rarely being involved. There is usually no involvement of the jaw-opening muscles. Motor branch travels inferior to the trigeminal ganglion and inferomedial to the mandibular branch when reaching the foramen ovale. Pathophysiologic examination can be used to distinguish oromandibular dystonia, etc. The characteristic EMG findings of hemimasticatory spasm include irregular bursts of motor unit potentials that correlate with the involuntary masseter spasms [33, 34].

It is relatively effective for botulinum toxin type A injection. However, this is not as fundamental a treatment as HFS is. Several studies with relatively good results have been reported [35–39]. Among them, Wang et al. reported a case series. Of the six cases, four cases showed improvement immediately after surgery and no recurrence [38]. If there is contraindication to MVD, or if the patient is hesitant about surgical treatment, botulinum toxin injection may be tried first.

Tinnitus, Vertigo

Tinnitus is a very common disease, reported by 10–20% in adults. In most cases, mild symptoms are controlled by medication, but in some cases, they interfere with daily life, which greatly affects the quality of life. The American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS) recently issued a clinical practice guideline for tinnitus in 2014. But, use of MVD in the treatment for tinnitus is not addressed, yet [40]. However, some groups have performed MVD on medically refractory tinnitus, and report relatively good results. In 1975, Jannetta attempted to explain hyperactive CN VIII symptoms such as intractable tinnitus and/or vertigo [41]. Since then, MVD has been attempted by many surgeons [42–44]. Recently, there was a systemic literature review with previously reported 43 cases. The authors reported MVD for CN VIII showed 70% success rate, which is lower than that of HFS or TN. The tinnitus-only patients had an even lower success rate of 60%, suggesting the importance of accurate diagnosis. The shorter the duration of symptoms, 5-year or less, showed the better the results.

Hypertension

Refractory hypertension is believed to affect 5–30% of the general population [45]. In the 1970s, Jannetta mentioned the possibility for neurogenic etiology in refractory hypertension [46]. They discovered a relationship between arterial compressions of the brainstem, in particular the rostral ventrolateral medulla (RVLM) and its impact on the regulation of cardiovascular activities. Since then, the idea has been established through anatomy studies. RVLM contains sympathoexcitatory bulbospinal neurons that play an important role in the control of blood pressure [47, 48]. And effectiveness of MVD has been demonstrated in several case series [49–52]. Compression was due to dolicoectatic vertebral artery or basilar artery at the medulla, which showed improvement after decompression. Sandou et al. reported the result of MVD for HFS with hypertension [53]. They mobilized the low cranial nerve in patients with HTN while operating MVD for HFS patients. At last follow-up, blood pressure was normalized in 28 patients

(58.33%), and medication is withdrawn in 14 patients (50%).

There must be some patients with refractory hypertension who could potentially benefit from MVD operation to normalize their BP. However, the long-term impact of this procedure remains debatable. Despite questionable long-term outcomes, the observed positive short-term outcomes indicate potential for future interventions to improve hypertension in this population.

Spasmodic Torticollis

Spasmodic torticollis is atonic and clonic neuromuscular disorder characterized by continuous or intermittent involuntary spasm of the cervical musculature. The exact cause of spasmodic torticollis has not yet been identified. Various hypotheses are suggested, such as abnormal neurotransmitters concentration [54], or maybe genetics [55]. Various surgical treatments are suggested—myotomy, neurectomy, selective peripheral denervation, MVD, and even deep brain stimulation. In 1995, Jannetta and Jho performed MVD on accessory nerves in 20 patients and reported cure in 13 patients and symptom improvement in 4 patients [56].

Compared to other diseases mentioned in this article, relatively many operations were performed. Li et al. [57] compared MVD (80 patients) and neurectomy (41 patients), and reported better improvement of symptoms in the MVD group than neurectomy. And they reported improvement of symptoms by 6 months.

The American Academy of Neurology recommends the Botulinum toxin as first-line treatment for primary spasmodic torticollis [58]. However, the limitation of Botulinum toxin is also obvious. Thus, MVD could be chosen in spasmodic torticollis patients with confirmed accessory nerve compression.

Others

In addition, MVD has been tried for a wide variety of disease entities. Table 1 describes the neurovascular conflict syndromes that can occur depending on the each cranial nerve. Some of these are hypoactivity. This is far from the general definition of neurovascular conflict. However, it may be also caused by neurovascular compression with the cranial nerve, which is included in the table.

Olfactory nerve has not been reported with disease due to neurovascular conflict. In the case

Table 1 Periodic table of neurovascular compression syndromes

Cranial nerve		Pathology	References
I	Olfactory nerve		none
II	Optic nerve	Paroxysmal phosphenes	[59, 60]
III	Oculomotor nerve	Oculomotor nerve palsy[a] Ocular neuromyotonia[a]	[61–64]
IV	Trochlear nerve	Superior oblique myokymia	[65–67]
V	Trigeminal nerve	Trigeminal neuralgia	[b]
	Mandibular branch	Hemimasticatory spasm	[35–39]
VI	Abducens nerve	Abducens nerve palsy[a]	[68, 69]
VII	Facial nerve	Hemifacial spasm	[b]
	Intermediate nerve	Geniculate neuralgia	[23, 70–72]
VIII	Cochlear, Vestibular nerve	Tinnitus, Vertigo	[41, 42, 44, 73]
IX	Glossopharyngeal nerve	Glossopharyngeal neuralgia	[20, 22, 74–77]
X	Vagus nerve	Essential hypertension	[46, 49–51]
XI	Accessory nerve	Spasmodic torticollis	[56, 57]
XII	Hypoglossal nerve	Hypoglossal palsy[a] Hemilingual spasm	[78–80]

[a]Hypoactivity is not hyperactivity. This is far from the general definition of neurovascular conflict
[b]Too many references and not mentioned

of olfactory nerve, it is unlikely to cause any special symptoms because there is no main vessel around the olfactory bulb. In addition, olfaction is a very subjective symptom, which may be one reason why the patient does not actively complain about it.

In the case of optic nerve, MVD was reported to be effective when paroxysmal phosphenes occurred in some case reports [59, 60]. The patient, reported by De Ridder D [59], complained light flash with visual field deterioration. An MRI scan showed an ectatic distal right internal carotid artery (ICA) abutting the undersurface of the right optic nerve and a compression of the optic chiasm by the anterior communicating artery. They were approached by orbitozygomatic craniotomy and Teflon was inserted between the A1 and optic chiasm. After surgery, symptoms improved mostly. It is a very rare case, but it is a good example of how MVD can work in a disease we overlook.

The cranial nerves (oculomotor and abducens nerves), involved in extra ocular movement, developed palsy by compression [61–64]. In the case of trochlear nerves, there were reports of superior oblique myokymia treated with MVD [65–67]. Superior oblique myokymia is a rare condition of unclear etiology. Since 1906, about 116 cases have been reported. Although meta-analysis has been published, there is no established treatment [81]. Bringewald [82] first presented the possibility of neurovascular compression and was confirmed in some cases. However, some studies have not found compression. Yet, superior oblique myokymia cannot be truly defined as a neurovascular compression syndrome.

Hypoglossal nerve has been reported with hypoactivity (hypoglossal palsy) and hyperactivity (hemilingual spasm) [78–80, 83, 84]. De Ridder et al. reported a case of hemilingual spasm caused by an arachnoid cyst [83]. Subsequently, it is suggested that hemilingual spasm could be caused by the tortuosity of the extracranial internal carotid artery [85]. Afterward, treatment of hemilingual spasm with MVD by decompression of cranial nerve XII at the lower brainstem was reported [78].

Conclusion

Even with preliminary results, MVD has been tried in a wide variety of diseases. From cranial nerve I to XII (although not found in CN I), neurovascular compression syndrome for each cranial nerve has been discovered and treated. This neurovascular compression syndrome can occur not only in the cranial nerve, but also in other autonomic nervous systems. Much research is still needed.

MVD surgery, started from Jannetta, is not an old technique but is still changing and developing. It is expected that the experiences and efforts of neurosurgeons will help more patients in the future.

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Intraoperative Neurophysiological Monitoring in Microvascular Decompression for Hemifacial Spasm

Sang-Ku Park

Introduction

Globally, HFS is not a common disease. However, it is a relatively common disease in Korea, China, and Japan. We have had many experiences and many thoughts in touching more than 4000 cases. 'How can I proceed more safely?', 'How can I detect nerve damage at a faster rate?' Etc. In this chapter, we will look at the tests performed during HFS surgery. There are so many opinions on the warning criteria that determine the BAEPs test and hearing loss. There is also a question about whether the lateral spread response measured only in patients with HFS is satisfactory if it is lost during surgery, and if it does not disappear during surgery. We hope that you will have a closer look at the tests used in MVD surgeries and understand them correctly to help you become safer and more successful.

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Conventional Intraoperative Neurophysiological Monitoring

Brainstem Auditory Evoked Potentials (BAEPs)

BAEPs is an electrical stimulus induced by stimulation from cochlear to brainstem by sound stimuli, which normally form 5–7 waves [1] and can evaluate the auditory pathway function. Jewett and Williston reported this noninvasive method for the first time in 1971 [2].

The Origin of Waveforms

The origin of each waveform is as follows: the I wave is the distal portion of the cochlear nerve; the II wave is the frequency, intensity, and phase of the wave sound originating from the proximal portion of the cochlear nerve. It plays a role of transferring information about the phase. Wave III performs the information processing of time and frequency, the control role of sending the wave neuron information from the cochlear nucleus to the appropriate part of the auditory nerve system. The wave IV functions to analyze the difference in the time and the intensity of the wave generated in the superior olivary complex. Therefore, it plays an important role in locating the sound source. Wave V is the most important path of the auditory system in the lateral lemniscus. It responds sensitively to the change of the negative stimulus by time and stimulus size. Wave VI and wave VII occur in the inferior

K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_11

colliculus, which also deal with auditory and somatosensory information. Wave VI and wave VII are not all observed, so they are generally excluded from the waveform evaluation [3].

Filtering

The BAEPs waveform is a very small waveform, so it is strong at 150 Hz to block the waveform from shaking even a little. HFF 3 KHz using a filter to make fine and fast waveforms visible, Use a notch filter to prevent the incorporation of electrical artifacts.

Electrode Position

Surface disc electrodes are used for recording and are placed on both mastoid processes and the vertex. A two-channel montage is used: Cz-Ai (vertex to ipsilateral mastoid) and Cz-Ac (vertex to contralateral mastoid). The auditory stimulus is a broadband click delivered through a tubal insert phone sponge. The operative ear is stimulated with an intensity of 85 dB normal hearing level (nHL), and a masking stimulus of 55 dB nHL or intensity of 120 dB sound pressure level (SPL), and a masking stimulus of 80 dB SPL are delivered to the contralateral ear.

Stimulation Rate and Averaging Time

Several hospitals around the world are doing a little different test for BAEPs. This is because the test has not yet been fully established, and experience with the surgery is different. Based on the paper, Thirumala [4], the stimulation rate was 17.5 Hz, averaging 256 (at least) times, and Amagasaki [5], the stimulation rate 10 Hz and averaging 1000 times. In Damaty [6], the stimulation rate was 11.1 Hz, averaging 1000 times, and so on, it is slightly different. James [7] reported that the test time was longer than 1 min, but the results should be reported within 15 s, depending on the stimulation rate 31.1 Hz, averaging 1000 and 4000 times.

During BAEPs monitoring, investigating patterns of BAEPs changes, analyzing correlations with surgical techniques, and investigating possible technical factors can identify the cause of BAEPs changes and provide appropriate information to the rest of the surgical team [8]. However, if the test method is different, the analysis of the information that can be provided will be different, so further research will be carried out in the future.

Cause of Waveform Changes

Sindou and Fernández-Conejero explain the causes of BAEPs waveform changes: (1) Extension of the VIIIth nerve caused by excessive cerebellar traction. (2) Manipulation of the labyrinthine artery or the motorized cerebellar artery that can cause a vasospastic reaction. (3) Direct trauma due to coagulation or heating due to appliances. (4) Classification of new CN compression of the eighth CN at the end of surgery by material inserted between VIIth and VIIIth complexes [9, 10].

Legatt studied the waveforms I, III, and V in detail. He concluded that cochlear ischemia or infarction during infarction or infarction and temporal bone drilling due to internal auditory artery injury affects all BAEPs components including wave I, delayed and attenuated direct mechanical or thermal trauma to the eighth nerve, and removed III and V waves, but waves I from the cochlea of the eighth nerve can be preserved. And extending the I–III interpeak interval during the retraction of the cerebellum and brainstem reflects stretching of the eighth nerve and is often reversible [8].

The origin of wave I is the cochlear junction at the distal end of the cochlea, and vasospasm in the eighth nerve may cause potentially reversible BAEPs changes. The changes in BAEPs, which all waveforms disappear without Wave I, may reflect direct mechanical or thermal damage of the brainstem, brainstem compression, or ischemia due to vascular injury [4, 9].

Post Operation Hearing Loss

In general, postoperative hearing loss is 2.3~21.2% [11–13]. This postoperative hearing disorder is a good test for hearing conservation in the BAEPs test in MVD surgery, and loss of wave V during MVD is a specific indicator of postoperative hearing loss. Current alarm criteria used to alert surgeons are a sensitive indicator of impending hearing loss after surgery [4].

Other studies have also reported that hearing impairment occurs when the offending vessel is present between 7 and 8 CN [14, 15].

According to several studies, the high-frequency (4–8 kHz) hearing loss (HFHL) associated with the cochlea, such as the impaired appearance of the cellular dysfunction caused by the acoustic trauma created by the drill, also occurs [16, 17].

Warning Criteria

In the past, the BAEPs warning criteria showed an interest in the latency change of wave V. After a while, both the latency and the amplitude of the V waveform are considered to be important. Recently, it has no relation with latency, the decrease or loss of amplitude is reported to be closely related to postoperative hearing.

In the case of classification based on the latency of V waveform, a study defined warning criteria by dividing by 0.4 ms (watching) signal, 0.6 ms (risk "warning" signal), and 1 ms [18, 19] , and some studies have focused on the latency of the wave V [20–22].

The American Clinical Neurophysiology Society reported that V latency longer than 1 ms or 10% increase in latency and/or with a 50% reduction in amplitude as a warning criterion [23, 24].

James studied hearing loss in cerebellopontine angle (CPA) surgery, where the 1 ms prolongation of the wave V or a 50% amplitude reduction was considered to be an indicator of influencing [7]. Although the BAEPs waveform is poor in patients with normal hearing, the BAEPs loss during surgery may be normal after hearing, so further studies are needed.

Hatayama was not associated with postoperative hearing loss when the latency period of the wave V was extended, and all patients who showed a decrease in amplitude more than 40% of the wave V were accompanied by prolongation of the latency and it was reported to be highly related to postoperative hearing loss [25].

Legget [8], Jo [26], Jung [27], and Park [28] have little association with the latency of wave V and hearing loss but the reduction of amplitude by more than 50% was highly related to hearing loss.

Thirumala [4] and Park [28] reported that the disappearance of wave V was an indicator of postoperative hearing loss.

Damaty showed a New Score that predicted postoperative hearing impairment by analyzing the scores of the changes of amplitude of BAEPs wave I, amplitude of wave V, change of latency, and IPL change of wave I–III [6].

The optimal stimulation rate and averaging time should be studied in the future, and the optimum warning criteria will be derived if the optimal test method is established based on these studies.

Lateral Spread Response (LSR)

In hemifacial spasm, the LSR test was performed on all surgeries, beginning with the study of ephaptic transmission and ectopic excitation of Nielsen [29].

Moller and Jannetta played a major role in the study of MVD surgery in earnest. LSR tests were made with bandpass from 0.3 to 3 KHz and latency was observed at an average of 11.03 + 0.66 ms [30].

Electrode Position

A dermis needle was inserted at the junction of the facial nerve at the midpoint between the trauma of the ipsilateral side and the outer side of the eyeball. Electrical stimulation is performed with two electrodes spaced 3 cm apart from the temporal or zygomatic branch, and the direction of the stimulation is toward the brainstem and a 0.2–0.3 ms stimulus with an intensity of 5–25 mA [31]. It is also very effective to stimulate the lower branch (buccal or mandibular), such as stimulating the upper branch of the facial nerve, to measure the LSR in the upper branch. Most of the reactions in both LSRs are measured, but only in one place.

Meaning of LSR

It is difficult to explain precisely whether LSR is a central mechanism or a peripheral mechanism.

But there are more claims of central mechanism [10, 32, 33]. If the origin of the facial nerve is affected by the arterial blood vessels, an anomaly occurs between the facial nerve strands. It is hypothesis that this phenomenon affects the facial motor nuclei in the brainstem and that the brainstem function becomes hyperexcitic state, resulting in hemifacial spasm.

A specific electrophysiologic study has shown that the disappearance of the LSR signifies good separation of facial nerve and blood vessels [10, 34–40]. Thirumala predicted that if LSR remains in operation in 2011, it may be possible to have spasm after surgery. LSR and the outcome are not statistically related, but long-term effects are predictable [31]. In addition, the case reported that the LSR did not disappear until the end of the operation, and that the HFS disappeared a year later [33], adding to the expectation and concentration of LSR measurement during surgery. However, questions are now being raised about what LSR measurements mean. Because we have learned from experience that LSR and spasm are not necessarily proportional.

Prognosis of LSR

LSR is not measured in normal persons [34, 35, 37, 41]. The relationship between long-term prognosis and LSR that does not disappear even after adequate decompression. There are many studies that LSR is useful during surgery, and studies have shown that the loss of LSR during surgery is a good surgical outcome after surgery [10, 32, 33, 35, 38, 42], showed that patients who did not disappear during surgery had a good outcome after surgery. Even if the LSR does not disappear during surgery, surgery is terminated when a change in shape is observed [34–37]. Because of these various studies, the LSR has been controversial. LSR was reported to be associated with spasm at 1 week postoperatively, but not at 1 year [43], and reported a different result from that reported in association with long-term outcome [31].

Some researchers believe LSR monitoring during surgery is useful for predicting facial outcomes of MVD-reported surgeries for HFS. It has not completely disappeared, but some research has shown that a reduction in LSR amplitude is associated with favorable outcomes [35, 44, 45]. However, some reports claim that the prognostic value of LSR monitoring for predicting the long-term outcome of facial seizures is still questionable [34, 46, 47]. In addition, some authors have reported that the loss of LSR after decompression is not always correlated with favorable postoperative results [44, 48].

Hatem et al. [47] reported complete improvement of spasm at the last follow-up for 10 of 10 patients who continued until the end of the operation. They suggested that uninjured LSRs are not always a bad prognosis. This is because motor nuclei and excitability can take months or years to normalize.

Persistence (Residual) of LSR

If the LSR response remains unchanged despite all surgical interventions, additional irradiation and/or decompression may be performed during surgery to relieve all vascular compression of the facial nerve [35, 44, 49]. However, this method is not recommended because he or she may have additional hearing loss or facial paralysis after extensive surgery for the purpose of LSR loss. Eckardstein et al. reported that 90% or more postoperative spasm is resolved if LSR is lost during surgery. However, it is not a bad thing to remain, and even if it does not disappear, it takes no further action [50].

Damaty reported that the LSR did not predict whether the operation was successful or not, and reported that LSR did not cause bad results even if it did not disappear during surgery. In particular, it has been suggested that endoscopy may be a good way to confirm decompression of peripheral blood vessels [51].

Lee et al. reported that the disappearance of LSR is helpful in the operation. Patients with symptoms less than 1 year had a significantly higher LSR loss rate after decompression than those with symptoms for 10 years or more. EMG monitoring was useful in confirming proper insertion of Teflon, especially when multiple vessels were involved. The LSR was suitable for short-term follow-up but not long-term follow-up for more than 1 year [52].

Reoperation

Park suggests that it is better to judge after 1 year of reoperation if more than 1 year of observation is needed because some people have improved after 10 months [53].

Thirumala reported on residual LSRs, suggesting that long-term outcomes may be seen even if LSR is not lost if decompression is appropriate, and intraoperative communication between surgeons and testers is important in producing adequate decompression [54]. It was advised to consider reoperation if the LSR did not disappear during surgery but there were signs of trembling for more than 1 year after surgery [31].

Optimal Method of LSR Test

LSR does not necessarily disappear after decompression, but when the CSF flows out after the dura open, the brain is sunken down and naturally the facial nerve and offender are separated and disappear [42]. It is therefore very important to accurately assess at which point the LSR is lost. Therefore, it is recommended that the LSR measurement be divided into at least six sections. (1) Before the dura open (2) After the dura open (3) Before the Teflon insertion (4) After the Teflon insertion (5) After the dura closed (6) After the operation is done. If the LSR is lost after dura open, it may be lost by retraction before Teflon insertion. It may slowly disappear after MVD, and may be measured again, so the measurement should be checked in at least six steps [42].

The pattern of loss of LSR varies greatly depending on the patient and most is suddenly lost. Sometimes it gets smaller and then disappears, In some cases, the small amplitude does not disappear and the surgery is terminated. Some studies have categorized these different cases [55], In our experience, it is rather a larger measure, there was a case where it was maintained without being reduced at all. There is a need for further studies on various aspects of these LSR.

It is very important to detect the moment when the LSR disappears during surgery. Knowing quickly what has been lost in any operation gives a lot of confidence in surgery, so informing the moment when it disappears gives a very useful help to the flow of surgery. Although LSR has a very significant role as an indicator during surgery, the meaning of the residual LSR still needs further research. It is still uncertain to predict LSR symptoms during surgery over the long term of more than 1 year.

Use a filter such as free-running EMG with LFF 10 Hz, HFF 3 KHz, and notch filter off so that the LSR waveform similar to 50~60 Hz will not be measured by the filter.

When the test is performed during the operation rather than the preoperative waveform, the amplitude of the LSR is measured as small as 100 μV due to anesthesia [56]. Therefore, it should be taken into consideration that LSR, which is measured very small in preoperative examination, may not be measured during surgery.

Zhong-Lee Response (ZLR)

Measurements should be made at the same site as the LSR measurement site, and stimulation should be performed prior to decompression of the nerve and blood vessels within 5 mm of the nerve, followed by stimulation after decompression. Stimulate with a bipolar stimulator (0.2 ms, 2 mA). Concurrent surgical monitoring of LSR + ZLR monitoring provides more valuable neurosurgical guidance than LSR for HFS, regardless of whether the facial nerve compression vessel is simple or complex [57].

There is LSR that is lost before decompression or LSR that is not lost after decompression. If this is the case, ZLR can be used effectively to check for contact between the vessel and the facial nerve [41]. However, other studies have shown that MVD was successful in 20 patients without HFS, before decompression ZLR was observed in 19 patients, 13 patients were lost after decompression, 6 patients were measured, three of these six were weak, while the other three had waveforms similar to those before decompression. The reason for this is that, in addition to being attached to the distal part of the facial nerve, if there is more part of the distal part of the facial nerve, it can be induced by electrical stimulation [58].

A monopolar stimulator is effective for mapping localization of the facial nerve. However, if the intensity of the stimulation is too high, facial neuro-EMG responses are measured in a wide area around the facial nerve, and therefore should be examined with low electrical stimulation. When direct electrical stimulation is applied to the facial nerve, it is stimulated to less than 1 V or 0.5 mA. However, if electrical stimulation is applied to the facial nerve in the presence of CSF or blood, "current shunting" may occur and a false-positive reaction may occur and so it is advantageous to check with Voltage rather than Current [59].

Facial Motor Neuron Function Test

Facial F-Wave

Facial F-wave is possible to record changes in elicitability of the facial F-wave during MVD. Immediate changes in hyperexcitability of the FMN can be observed by monitoring changes in F-wave elicitability. Facial F-waves can be obtained from the mentalis muscle by stimulating the mandibular branch of the facial nerve (Fig. 1).

The facial F-wave represents the backfiring of the facial motor neurons after being activated antidromically. F-wave activity was shown to be an index of motor neuron excitability [60].

F-wave persistence appearance was reported in patients with HFS [61] and found to decrease after adequate MVD, albeit with delay as long as 2 years.

Facial MEP

Facial MEP is a hyperexcitability facial motor nuclei in hemifacial spasm patients. Therefore, a large amplitude waveform is formed compared to the normal side. There is no change in the amplitude of the normal side after decompression during surgery but surgical site can be used as an indicator of good surgical outcome with decreased amplitude [62].

However, there are some drawbacks to using Facial MEP as a test.

First, electrical stimulation should be given to the facial motor cortex. However, since patients are slightly different, it is difficult to find the exact location. Therefore, the brain cortex area should be extensively stimulated to include the facial motor cortex site within the stimulus range. In other words, it is difficult to know whether the facial motor cortex is fully excited every time it is inspected to achieve the optimal waveform. It is doubtful whether the facial motor cortex was sufficiently stimulated if a waveform was observed in only one of the Oris or mentalis (Fig. 2c).

Second, the amplitude of the waveform varies with each test. Because the MEP waveform is the response of the muscle twitching, the waveform is observed slightly differently each time it is examined, as the stimulus intensity increases, the amplitude increases, in some cases, the waveform

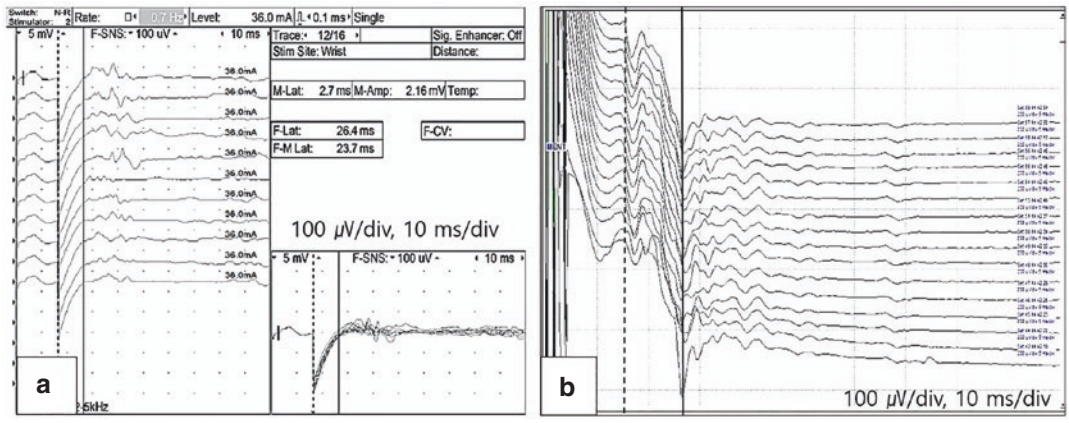

Fig. 1 Preoperative facial F-wave test waveform (**a**) and intraoperative facial F-wave test waveform (**b**)

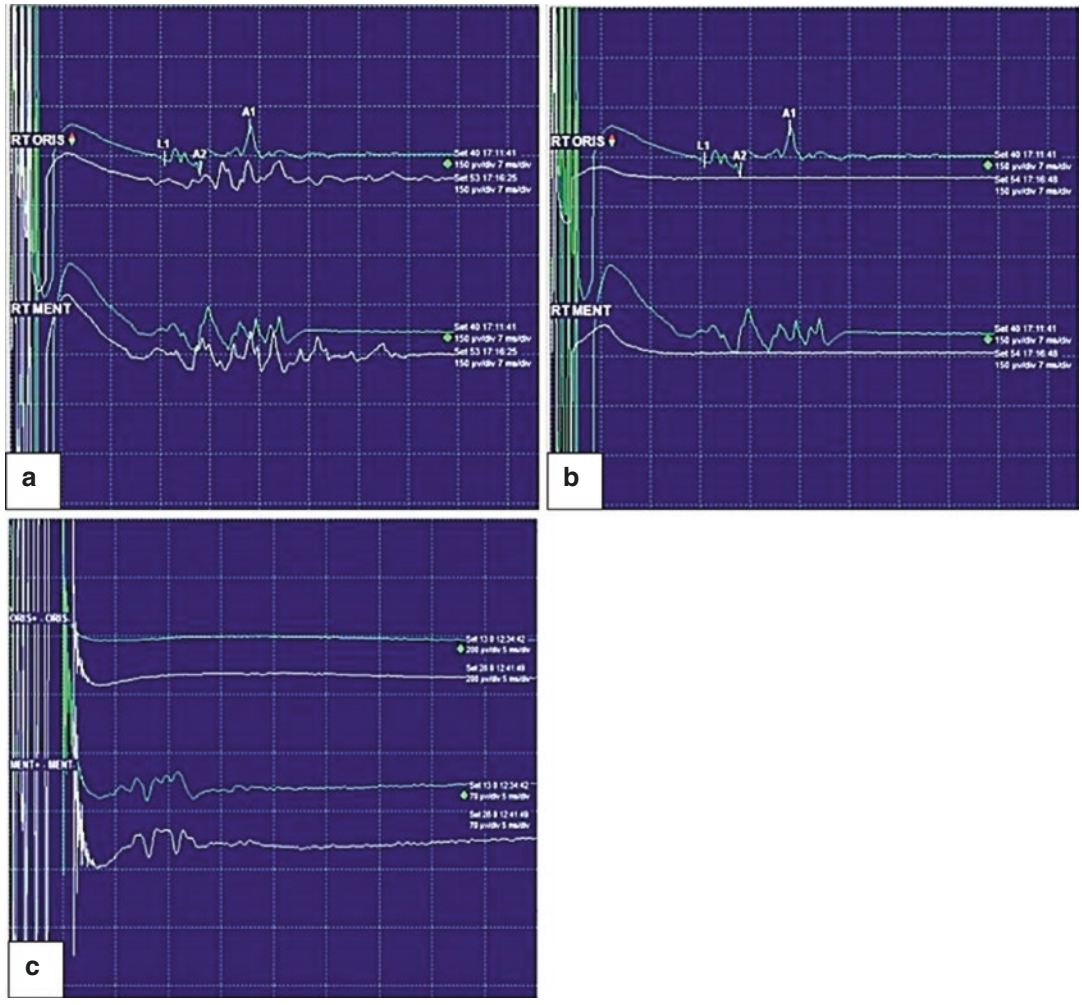

Fig. 2 (**a**) Typical waveforms of facial MEP measured during surgery, (**b**) Facial MEP waveform lost during surgery, (**c**) Waveform measured only in mentalis but not in oris

may disappear even if the muscle relaxant anesthesia is slightly deeper (Fig. 2b).

Third, there is no clear criterion that the amplitude of the waveform should be decreased after the decompression during the operation to make the operation successful. In addition, each patient has a different magnitude of amplitude reduction after decompression and so it is difficult to evaluate whether the operation is clearly done.

Fourth, since it is necessary to compare with the waveform of the normal side as well as the operation site, it is troublesome to further install the electrode.

In this way, it is inconvenient to use four Facial MEPs as an indicator during surgery, in particular, since the presence of a waveform like the LSR is not all or not, it may be helpful for surgery. I think it is difficult to predict the success or failure of surgery with only Facial MEP.

It has the disadvantages as above, but there is a unique advantage. It is very useful for distinguishing facial palsy after surgery due to facial nerve damage during MVD surgery and when facial MEP is performed in a cerebella-pontine angle tumor with a trans-labyrinthine approach, the surgical approach to the distal portion of the

facial nerve, it is very useful to distinguish proximal lesions of facial nerve during tumor removal. In vascular surgery, it is very useful to distinguish the damage of the facial motor cortex due to the blood supply problem [62].

New Advanced INM in MVD

Brainstem Auditory Evoked Potentials (BAEPs)

Electrode Position

If the electrode is placed on the back of the ear, it is close to the surgical site and there is a risk of contamination (Fig. 3a). So, there are problems to think about when choosing under earlobe or choosing in front of ear (Fig. 3b, c). Facial nerves are frequently touched during the process of separating facial nerve from blood vessels in surgery. However, under the earlobe or the frontal part of the ear is also affected by facial nerve, touching the facial nerve during surgery affects the BAEPs waveform, which hinders smooth waveform analysis. Therefore, when electrodes are attached to the tragus or antitragus of the auricle, it is completely separated from the facial nerve, and a very stable waveform can be obtained (Fig. 3d).

BAEPs Ipsilateral and Contralateral Wave Forms

The BAEPs waveform is generally characterized by 5–7 waveforms. Wave I, III, and V are always well seen, wave VI, VII are measured or not measured. A contralateral wave III was observed in the latency between ipsilateral wave II and III, the latency of the contralateral wave IV was observed more rapidly than the ipsilateral wave IV, the latency of the contralateral wave V is later than that of the ipsilateral wave V [63] (Fig. 4a).

In the BAEPs test, only the A1-Cz test is performed in the left operative case, on the right side, only the A2-Cz test is performed. Care

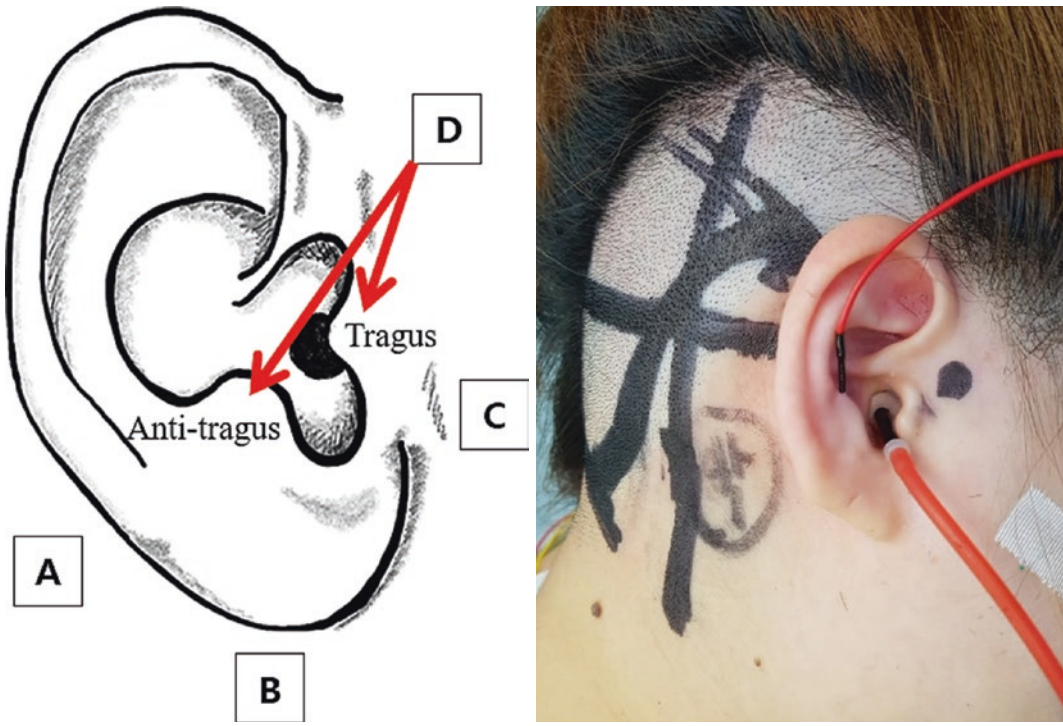

Fig. 3 Photographs of electrodes inserted on the Tagus, anti-tragus. The facial nerve can only be touched by manipulation during surgery. In this case, if the BAEPs electrode is installed in the facial nerve peripheral branch, it can be affected and not measured smoothly

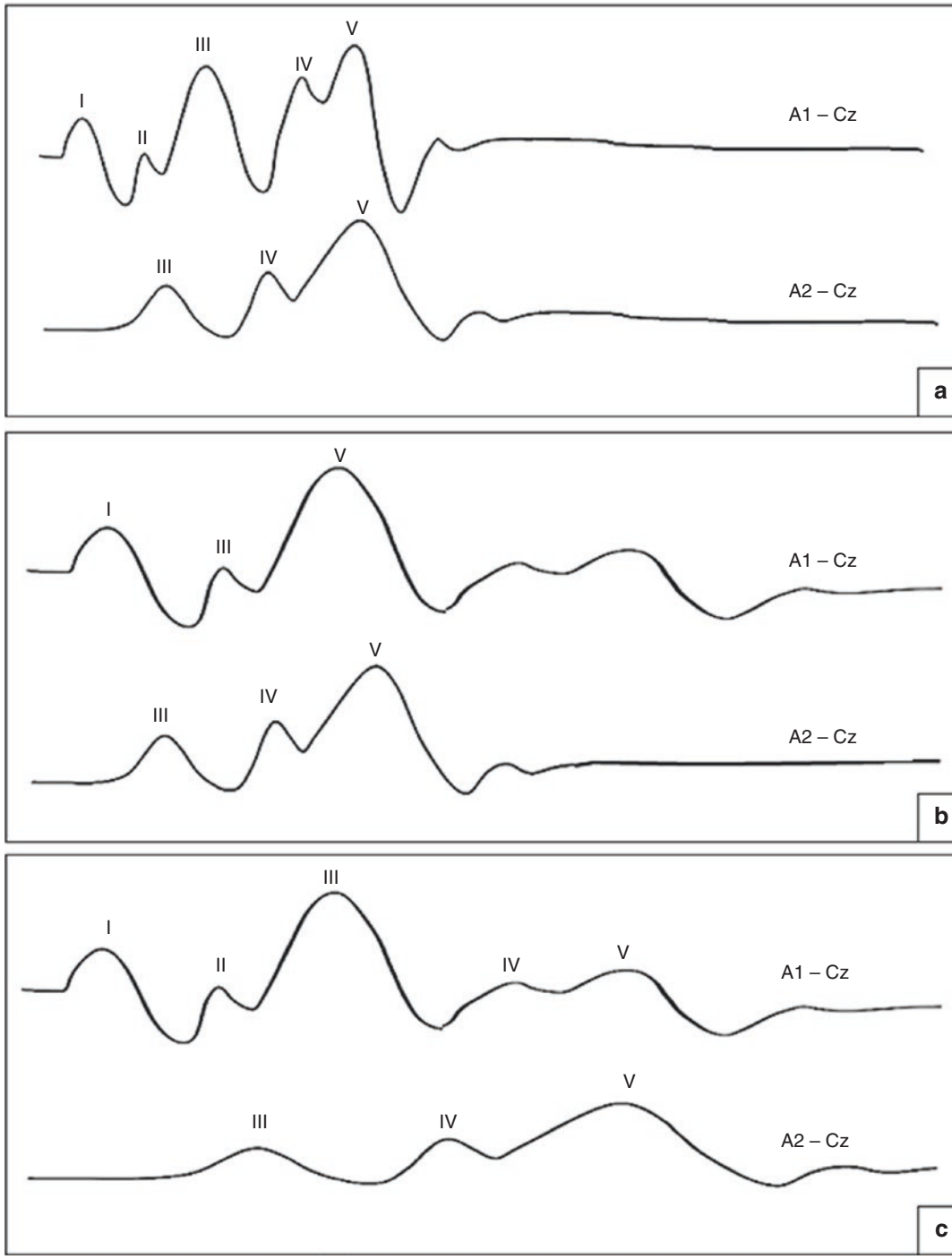

Fig. 4 Conventional BAEPs waveforms (**a**), contralateral waveforms were observed (**b**), and (**c**) judged ipsilateral waveforms differently. If the BAEPs test is carried out on the ipsilateral side only, it may not be possible to correctly judge various changes in the waveform. Changes in the contralateral side waveform can be observed at the same time to help with these problems

should be taken when performing this test, as it can cause errors in correct identification of waveforms. For example, if the cochlear nerve experiences very weak damage, the amplitude of wave III is often reduced or lost. In this case, the loss of wave III or delayed wave III is clearly distinguished. If there is a loss of wave III and the latency of wave V is maintained, there is no big problem, but if the wave III is extended and the wave V is extended, it is very severely damaged it must be clearly distinguished.

When such a situation occurs, the BAEPs test can be helpful in observing the A1-Cz and A2-Cz waveforms in a single window of both ipsilateral and contralateral test of the surgical site. Compared with the contralateral wave, it is easily discernible that the ipsilateral wave III disappears and the latency of the wave V is maintained (Fig. 4b). In addition, the determination of whether wave III and wave V are simultaneously extended can be easily distinguished from the contralateral wave (Fig. 4c). Therefore, in the operating room, it is recommended to observe the waveform changes by measuring both ipsilateral and contralateral when performing the BAEPs test.

Real-Time BAEPs

A typical BAEPs test is performed more than 2000 times at 10 Hz [64]. The reason for this test is to get the best waveform that the patient has. When the patient was awake, the examination required at least 2000 s to remove movement artifacts.

However, there is no reason for the movement artifact to be incorporated because there is no movement of the patient, a faster test time was needed to determine if the surgery affected the auditory nerve.

Our previous test required a stimulation rate of 26.9 Hz and averaging time of 1000–2000, which was about 37.1–74.3 s. We have also observed that the BAEPs waveform has been shown to have very little wave V latency and a decrease in amplitude of less than 50% during surgery, immediately following the next test, I had experienced BAEPs loss (Fig. 5).

This phenomenon is thought to be caused by the fact that abnormal waveforms are mixed in normal waveform due to too much averaging time, or that the stimulation rate is too slow to be appropriate for discriminating between auditory nerve damage.

So we reduce the averaging time from 1000 and proceed to 100 times. I looked to see if a waveform like 1000 would be maintained at a certain level, and I found that 400 times was enough for averaging time 1000. The stimulation rate was tested from 10 to 100 Hz. It was found that if the stimulation was too fast, the effect of the artifact caused by the surgery would be reflected more severely on the waveform. Thus, we conclude that testing at 40–50 Hz is very stable for discriminating waveforms (Fig. 6).

We think that it is the best BAEPs test to perform with the setting of waveform completion at 9.1 s with stimulation rate 43.9 Hz, 400 averag-

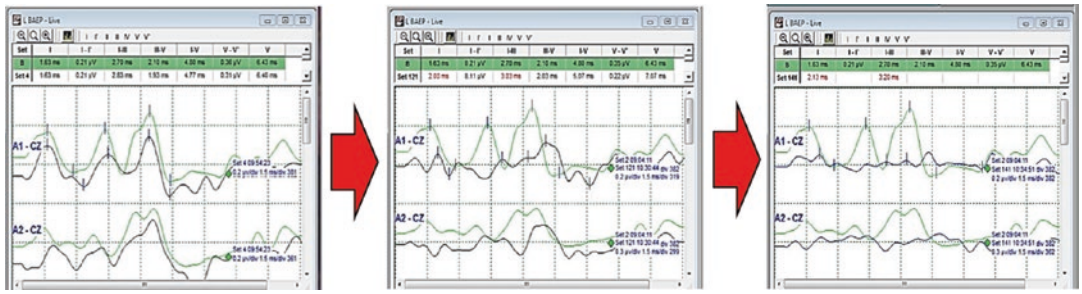

Fig. 5 Each waveform change step at stimulation rate of 26.9 Hz and averaging time of 1000–2000. If the averaging time is prolonged, it is difficult to determine whether the nerve is damaged because the normal waveform and the abnormal waveform are mixed. We have experienced a lot of sudden loss of waveforms during the next test after a fine change in the waveform was observed during the long time average

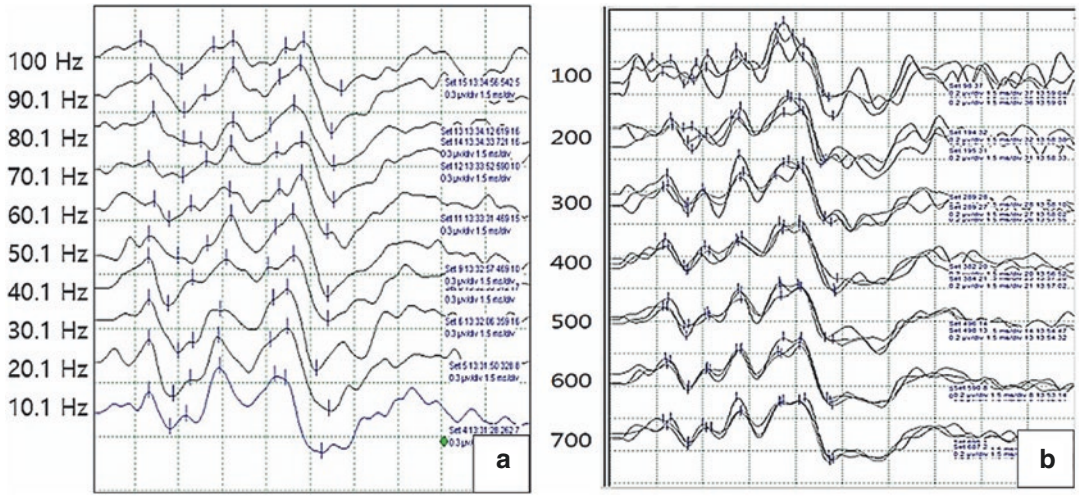

Fig. 6 (**a**) Waveform change according to stimulation rate (with averaging of 1000 trials). (**b**) Averaging trials (with 43.9 Hz/s stimulation rate) from IOM of BAEPs. Although the waveform was formed from 10 to 100 Hz, the faster the test above 50 Hz, the more the waveform was affected by the surgical operation. Thus, when the test was performed at a speed of about 40 Hz, stable waveforms were not affected by the surgical operation. Reproducibility was enough about 400 times to achieve the same effect as 1000 times of averaging time. (These results are from Joo et al. [65])

ing time. With the development of medical equipment, waveforms that were not possible with analog equipment in the past have become possible, and testing in less than 10 s is possible on most equipment. Indeed, a continuous test was repeated every 9.1 s and the amplitude of the waveform was reduced or lost within 10 s [65].

When we used Wave V amplitude loss >50% on an alert basis, we were able to reduce the incidence of postoperative hearing impairment to less than 0.4% (Table 1). We believe that obtaining a stable BAEPs within 10 s is a real-time IOM procedure of BAEPs and very important for preventing CN VIII damage during MVD surgery (Fig. 7).

New Warning Criteria

If the auditory nerve is instantaneously damaged, averaging is performed for a long period of time, and abnormal waveforms are mixed in the conventional normal waveform, so that the waveform is very small. Therefore, we observe the finely changed waveform. That is, the continuous and slowly varying latency will be reflected in the averaging test.

So, sudden decrease or disappearance of amplitude within 10 s will not be reflected.

Table 1 Comparison the protocol for INM of BAEPs and postoperative hearing loss

INM BAEPs	Previous protocol[a]	Real-time protocol[b]	*p* value
Number of averaging trials	1000–2000 times	400 times	
Stimulation rate	26.9 Hz	43.9 Hz	
Time to obtain BAEPs	About 37.1–74.3 s	about 9.1 s	
Warning criteria (of the wave V)	1 ms latency prolongation or a 50% decrease in amplitude	A 50% decrease in amplitude	
Postoperative hearing loss	4.02%	0.39%	0.002

[a]Means the protocol that used in our previous study (Jo et al. [26])
[b]Means the protocol that used in our recent study (Joo et al. [65])

So far we have been doing BAEPs tests with slightly different settings, but we have not thought that we cannot detect real-time nerve changes because the averaging process is too long.

The real-time BAEPs method with a stimulus frequency of 9.1 s is characterized in that the

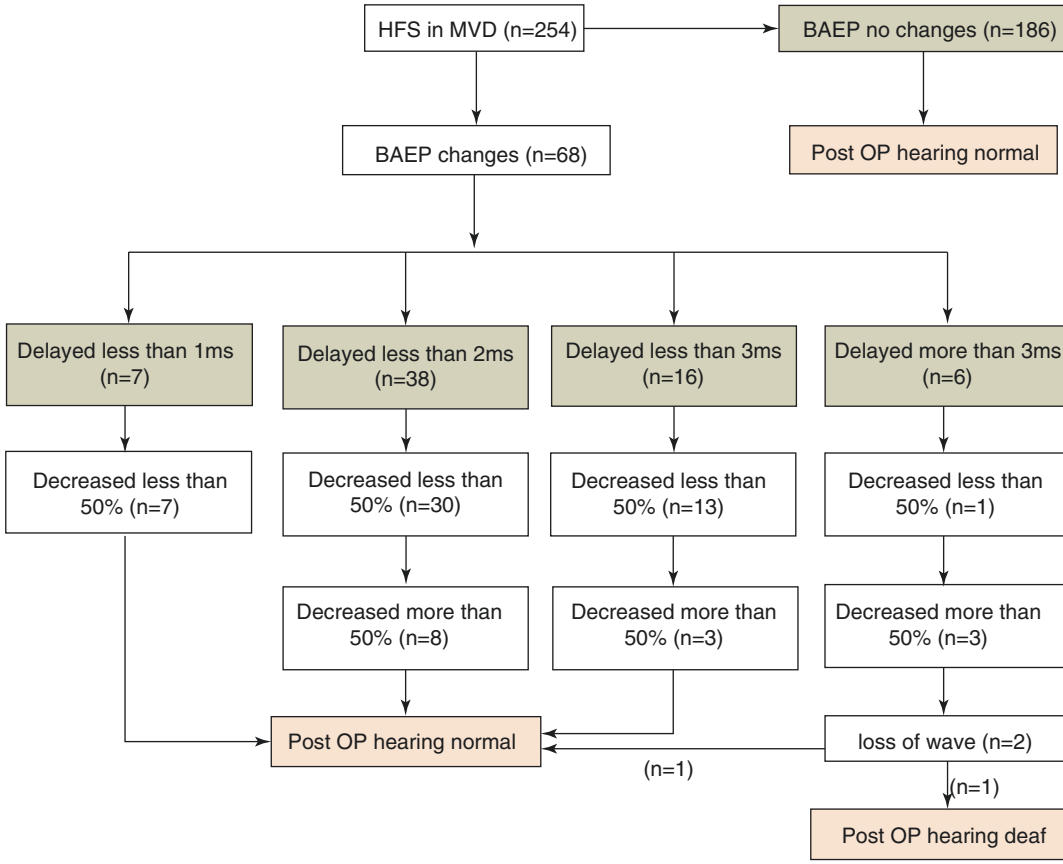

Fig. 7 Examination with real-time BAEPs allows you to determine the state of the auditory nerve in 9.1 s. In particular, we noticed that the latency changes very slowly and continuously, and the amplitude changes very quickly. Even within 9.1 s, the waveforms were lost. The prolongation of latency by about 1 ms was observed very frequently and was not related to postoperative hearing. However, as latency was extended by more than 2 ms, the amplitude decreased more often, leading to waveform loss. (These results are Joo et al. [65])

latency changes gradually and slowly, and the amplitude changes suddenly decrease or disappear within 10 s. Observe clearly. In other words, latency and amplitude change independently, latency changes are observed very often, and amplitude changes are observed very rare. In addition, changes in latency only have no effect on postoperative hearing changes (Fig. 8). Therefore, the change in amplitude should be observed much more important than the change in latency delayed [28].

For warning criteria of BAEPs, we used a "sliding scale" as follows: (1) the observation sign: latency prolongation of 1 ms without an amplitude reduction of at least 50%; (2) the warning sign: latency prolongation of 1 ms with a reduction in amplitude of at least 50%; (3) and the critical sign: loss of wave V. When the neurophysiologist detected the observation sign during MVD surgery, he notified the surgeon immediately, but the surgeon did not perform any corrective maneuvers. However, when the warning or critical sign happened, the neurophysiologist immediately notified the surgeon, the surgeon halted the operation that he was doing, and did surgical corrective measures (Table 2).

However, when the BAEPs wave V latency is prolonged by more than 2 ms, amplitude is often reduced by 50% or more. In particular, prolongation of more than 3 ms may result in more than 50% decrease in amplitude and BAEPs loss more frequently, so prolonged BAEPs wave V latency

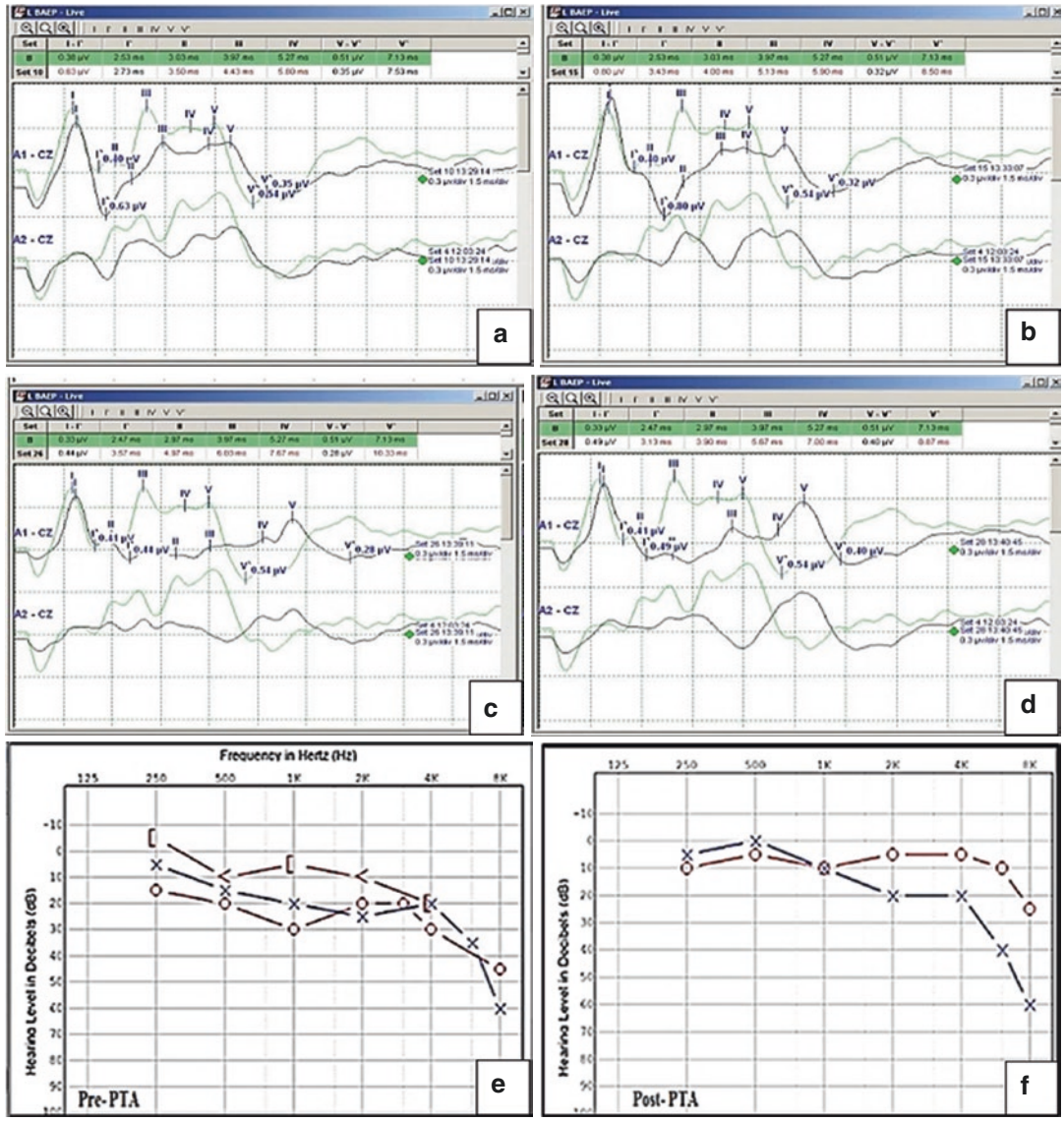

Fig. 8 (**a**–**d**) representative case showing only latency prolongation (≥1 ms) without a significant change in amplitude. (The latency of wave V was delayed by 3.20 ms from 7.13 to 10.33 ms with a minimal decrease in the amplitude.) The patient in this example did not experience postoperative hearing loss. (The average pre-PTA threshold was 22.5 dB, and the average post-PTA threshold was 6.25 dB.) (**e**) The INM of BAEPs during MVD surgery (white arrow = baseline of wave V; black arrow = wave V showing maximal prolongation in latency), (**f**) Pure tone audiometry of the patients obtained prior to surgery and 7 days postoperatively

should also be observed carefully. Therefore, we want to present the relationship between latency and amplitude as follows (Fig. 9).

If you go through the process in real-time BAEPs in less than 10 s, there are many cases where the amplitude of the wave V suddenly changes within 10 s. Most of the reasons for the decrease in wave V amplitude were due to brain retraction. It is important to quickly detect when the waveform has decreased and notify the neurosurgeon, as soon as the retractor that affects the cochlear nerve is removed, the waveform is observed to recover. We conducted the BAEPs test in real-time in this way. In 2016, 417 patients who underwent surgery did not have any HL after surgery (Fig. 10).

However, these criteria are applicable only if the waveform results can be derived within 10 s. In the case of a test that takes more than 1 min, very minute changes in the latency period or minute changes in amplitude should be observed. The trend of the warning criteria in the past is that it is important to change the latency of the V waveform, but now it is shifting to the direction that amplitude change is more important. It is also important to observe changes in waveform I due to ischemia associated with auditory nerve as well as changes in waveform V, and studies of changes in I-III IPL are also needed in the future [4, 9].

Table 2 The validity of the warning criteria of BAEPs to predict postoperative hearing loss

Warning criteria (of the wave V)	Latency prolongation (≥ 1ms) with amplitude decrement (≥ 50%)	Transient loss	Permanent loss
Positive predictable value	0.075	0.211	0.545
Negative predictable value	0.999	0.996	0.995
Sensitivity	0.909	0.727	0.545
Specificity	0.865	0.967	0.994

These results are from Park et al. [28]

Hearing Loss Patterns

Cerebellar Retraction

From January 2001 to January 2011, in 1518 patients with MVD due to HFS, the causes of BAEPs changes during surgery and hearing loss after surgery are considered retractors used in patients during surgery. An anatomical analysis was performed on how it affected depending on the degree of withdrawal.

All patients underwent preoperative evaluation by computed tomography (CT) and MRI with the addition of a contrast agent. MRI was obtained at 3.0 T (Achieva; Philips Medical Systems, Best, The Netherlands) with an eight-channel sensitivity-encoding (SENSE) head coil. Imaging protocol included three-dimensional time-of-flight (3-D TOF; TR/TE/flip angle, 25 ms/3.5 ms/20 section thickness 1.6 mm, slice spacing 0.8 mm,

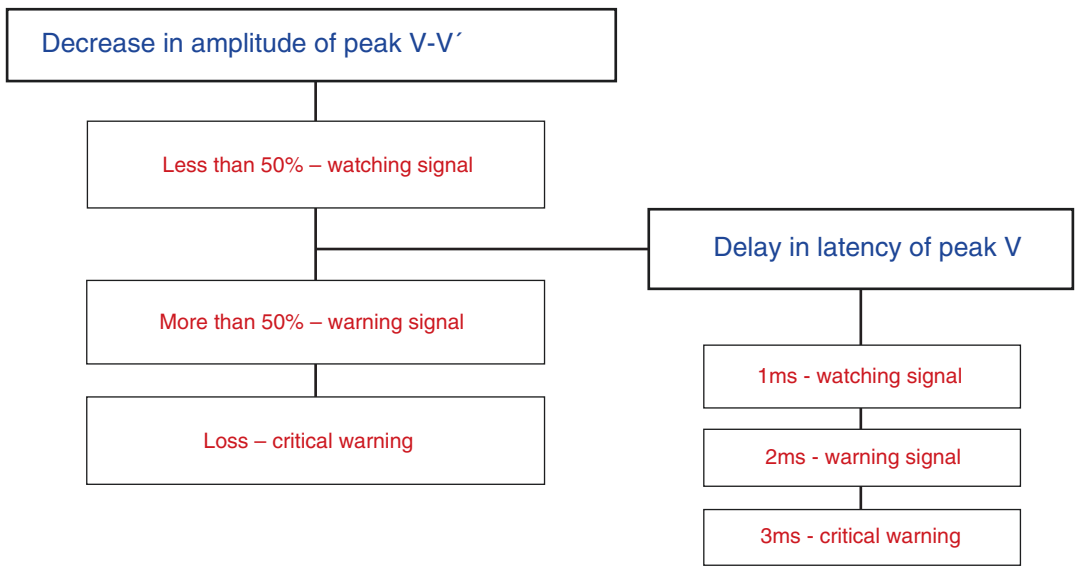

Fig. 9 Warning criteria applied when using real-time BAEPs. The relationship between latency and amplitude is presented separately based on wave V. In real-time BAEPs tests, latency changes were always observed first, followed by amplitude changes. In addition, latency change was observed slowly and continuously, and amplitude change suddenly decreased by more than 50% within 10 s

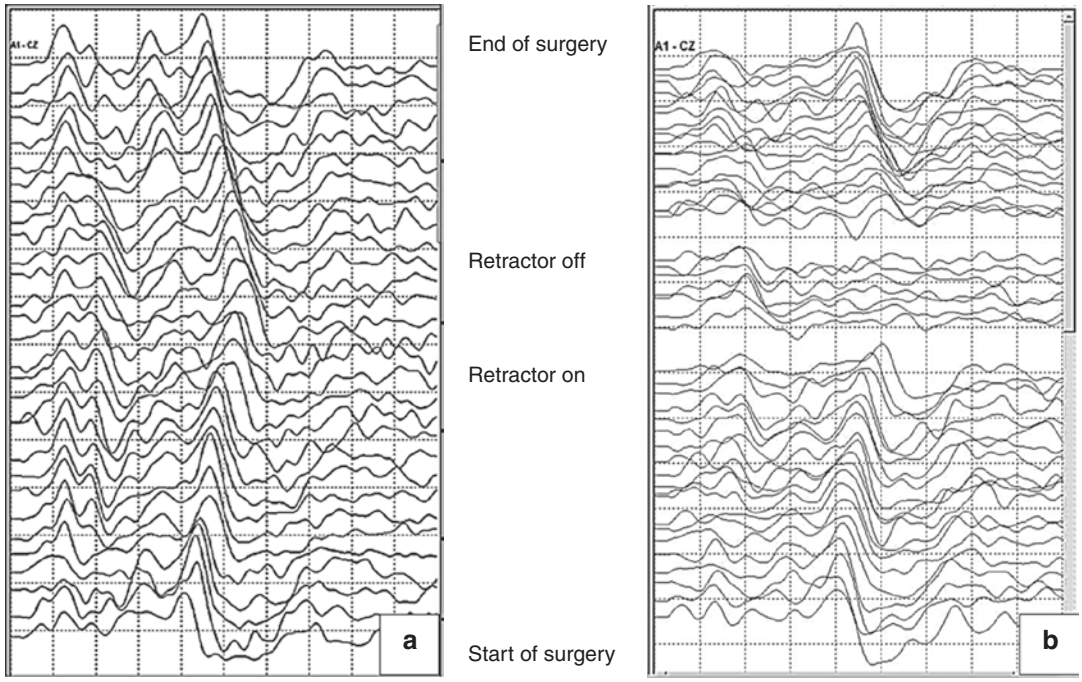

Fig. 10 If the wave V waveform is delayed (**a**) or lost (**b**) by the retractor, the retractor is removed, immediately testing and reporting the recovered waveform in real time. If you remove the retractor immediately after quickly detecting nerve damage, it will recover even if the waveform is lost

50048 matrix, acquisition time = 5 min 29 s), and 3-D T2 VISTA (volumetric isotropic T2-weighted acquisition; TR/TE/flip angle, 2000 ms/228 ms/20 NSA = 2, mm3, ETL=70, 24850 matrix, voxel size = 0.8.8.8 acquisition time=4 min 10 s). Neurovascular compression was determined by experienced neuroradiologists.

Among the 1518 patients, 106 (6.98%) displayed functional hearing changes. Hearing loss was permanent in 12 patients (0.79%). Of the 1412 patients with stationary hearing compared with preoperative audiometry, 96 patients were selected who were individually matched with respect to sex, age, and degree of spasm. BAEPs changed immediately after cerebellar retraction in 7 of 12 hearing-loss patients, suggesting the importance of retraction on hearing outcomes. The distance from the cerebellar surface of the petrous temporal bone to the neurovascular compression point was measured. The median distance of cerebellar retraction in the hearing loss group was 13.77 mm, which was longer than the median distance in the control group (Fig. 11).

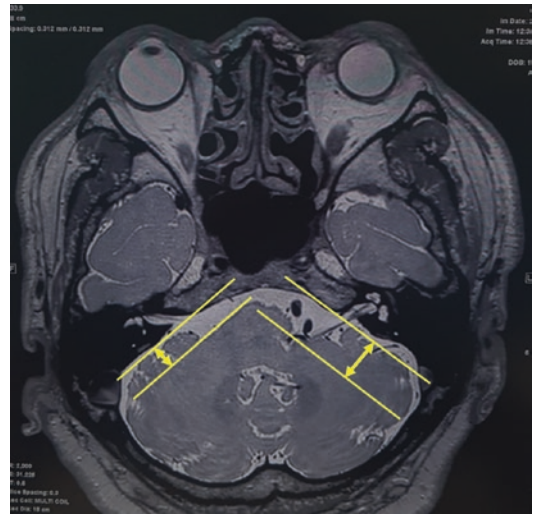

Fig. 11 A preoperative MRI showed the left vertebral artery and posterior inferior cerebellar artery in close proximity to the root exit zone of the ipsilateral facial nerve, and the vertical distance of cerebellar retraction was more than 10 mm. View of measured vertical length from the petrous temporal bone to neurovascular compression point. The greater the amount of retraction, the greater the damage to the cochlear nerve

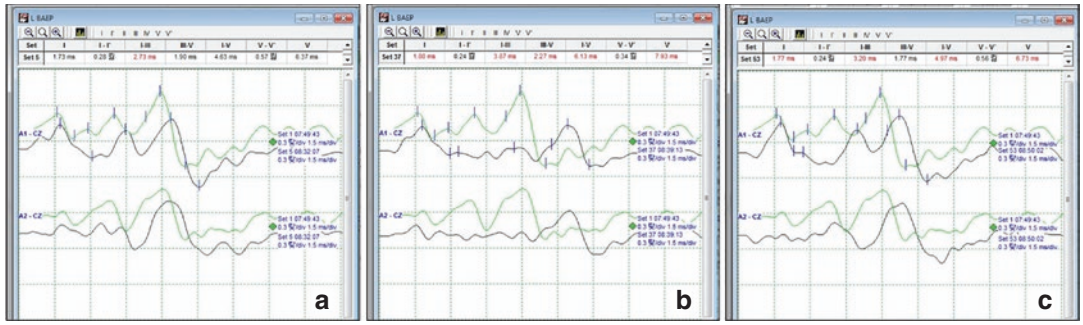

Fig. 12 During surgery, the BAEPs waveform prolonged from wave III (**a**), wave of III was lost (**b**), and unrecovered picture with the procedure extended to the end of surgery (**c**)

We retrospectively investigated attempting to correlate the distance of cerebellar retraction and BAEPs change with the occurrence of hearing loss after MVD for HFS. BAEPs monitoring is a valuable intraoperative indicator for preservation of auditory function, as in previous reported studies [26]. Therefore, we should design the most appropriate approach by knowing individual pathologic anatomy before surgery and the retraction should be done minimally, and in a direction perpendicular, rather than longitudinal, to the axis of the eighth nerve, to minimize the risk of postoperative hearing impairment. Furthermore, according to our experience, individual pathologic anatomy before surgery assessed by MRI is crucial to adapt and design the most appropriate approach for every patient.

Delayed Hearing Loss

We retrospectively reviewed the medical records of 3462 patients who received MVD for reflective spasms from January 1998 to August 2017. Five of these were normal immediately after surgery, but there were five cases of delayed hearing loss. After a while (middle, 22 days, 10–45 days) they suddenly developed hearing problems.

All of these patients showed postoperative sensorineural hearing loss, a common phenomenon was observed, and in the brainstem auditory evoked potentials (BAEPs), the inter-peak latency of waves III was prolonged during surgery, but recovered within a short time.

We have not yet identified the incidence of prolonged inter-peak latency of waves I–III. Therefore, it is difficult to conclude that this is a characteristic feature of delayed hearing loss. But we think about prolongation of the inter-peak latency of waves I–III seems to be associated with the occurrence of delayed hearing loss. During surgery, the BAEPs waveform prolonged from III. If you stay prolonged without recovery even at the end of surgery, hearing problems may occur after surgery (Fig. 12). Hence, it is possible that BAEPs changes may predict delayed hearing loss, but this issue requires further investigation. Analysis of more cases will be necessary to identify the exact cause of delayed hearing loss and to determine whether BAEPs monitoring can be used to predict delayed hearing loss after MVD for HFS [66].

The Significance of Wave I Loss of Brainstem Auditory Evoked Potentials

We looked closely at the loss of BAEPs waveforms during surgery. We enrolled 670 patients with primary HFS who underwent MVD surgery with IOM of BAEPs from January 2015 to December 2016. We distinguish between the case where wave I is observed when the BAEPs waveform is lost and when the wave I is lost and not all waveforms are observed (Fig. 13).

If the remaining waveforms are lost while wave I is maintained, it is mainly caused by retractor due to direct nerve damage. After retractor removing, we know that the waveform recovers normally. However, the loss of all waveforms without wave I is thought to be caused by a problem in the blood supply, which is observed as a delayed change phenomenon in which the waveform disappears after 10 min without any change in the waveform immediately after damage. In particular, the postopera-

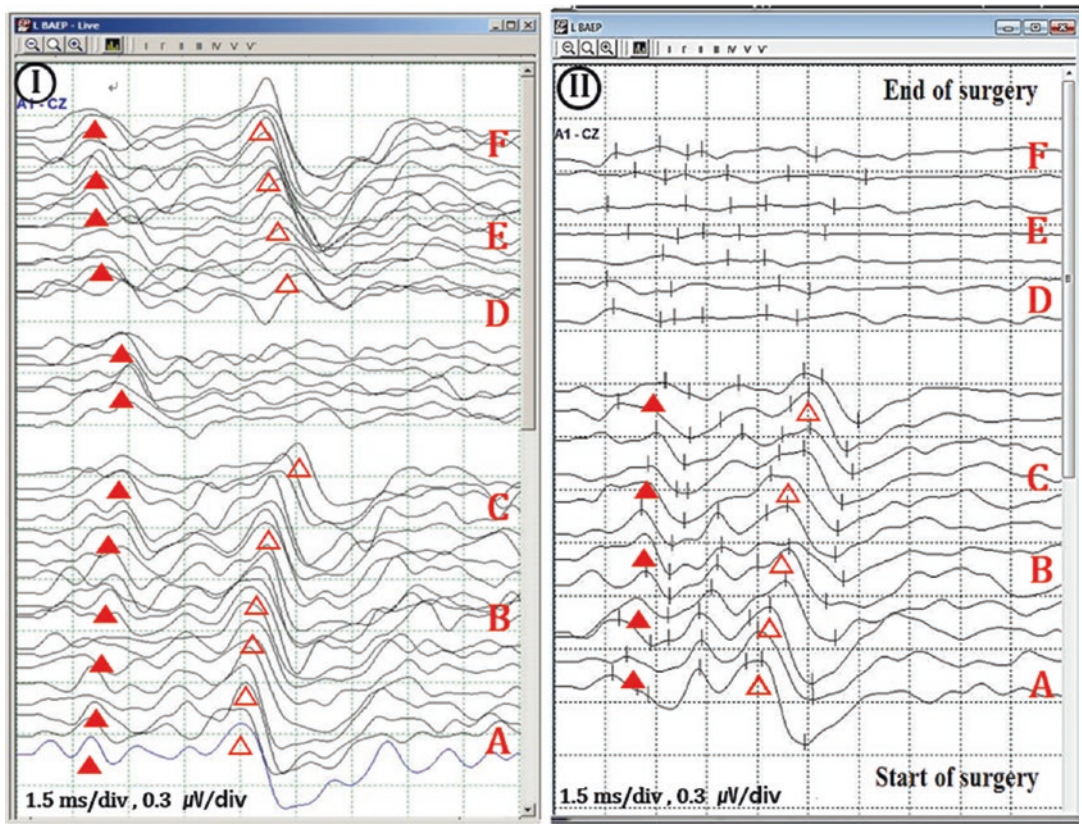

Fig. 13 Example of brainstem auditory evoked potentials (BAEPs) according to persistence of wave I in patients with wave V loss during microvascular decompression surgery. (I: BAEPs with persistence of wave I; II: BAEPs without persistence of wave I; A: Surgery start; B: Dural opening; C: Direct microvascular decompression procedure start; D: Direct microvascular decompression procedure end; E: Dural closure; F: Surgery end; filled triangle: wave I; unfilled triangle: wave V.)

tive patient's condition in which all waveforms are lost and not recovered without wave I is considered to be the result of vasospasm-like artery affecting the vestibular cochlear, as severe damage to the entire vestibular system occurs (Table 3, Fig. 14).

In the case of failure to recover due to loss of waveform during surgery, hearing loss occurs after surgery (Table 4). Especially if all waveforms are lost without wave I, much higher postoperative hearing loss may occur (Fig. 15).

Table 3 The proportion of postoperative complications according to persistence of wave I among the patients showing wave V loss

	w/i persistence of wave I	w/o persistence of wave I	*p* value
Patients, *n*	24	12	
Dizziness, *n* (%)	0	5 (41.67%)	0.002
Tinnitus, *n* (%)	0	3 (25.00%)	0.031
Diplopia, *n* (%)	0	1 (8.34%)	0.333
Hoarseness, *n* (%)	0	1 (8.34%)	0.333
Hearing loss, *n* (%)	2 (8.33%)	6 (50.00%)	0.009
Subtype of hearing loss, *n* (Low: High: Total)	2:0:0	0: 0: 6	

New Method LSR

We have used LSR as an indicator of good surgical outcome during surgery.

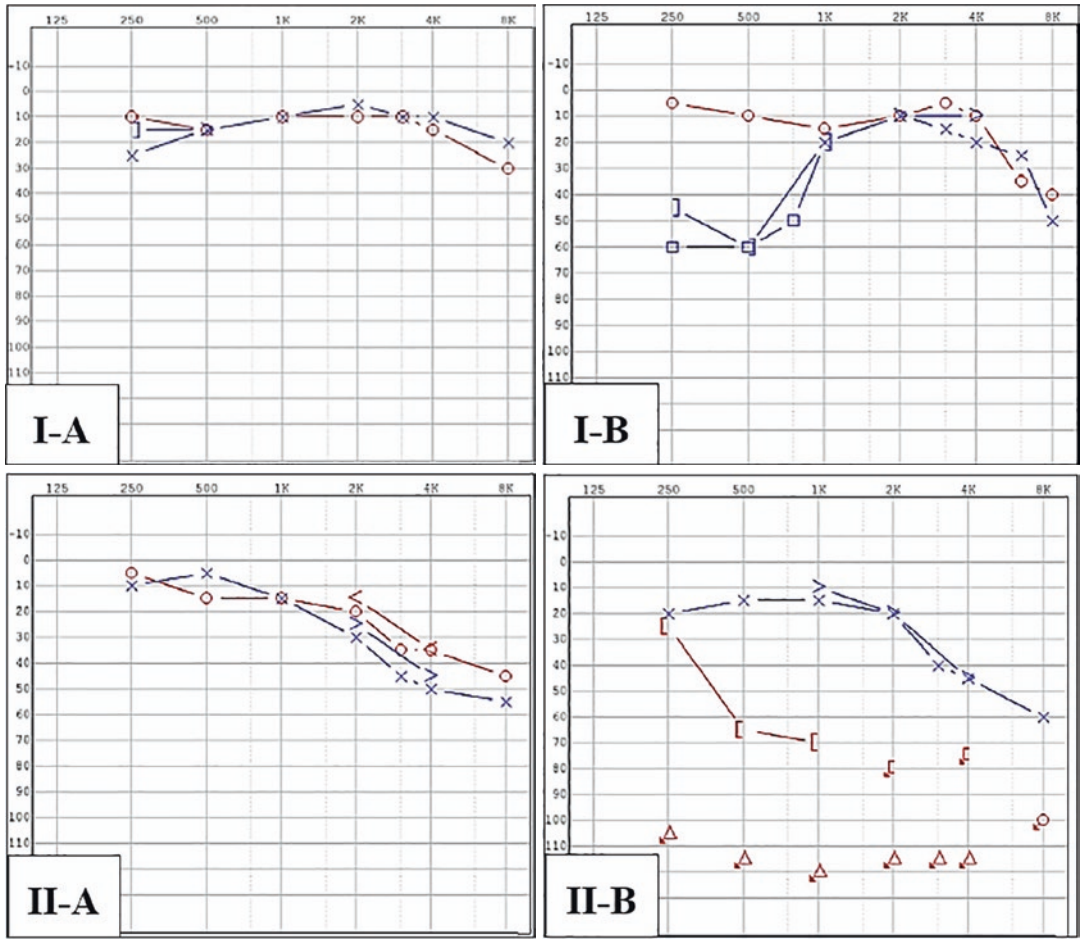

Fig. 14 Example of Pre-/Post-PTA according to timing of wave V loss during microvascular decompression surgery for hemifacial spasm. (I-A/B: Pre/Post-PTA of the immediate-change group; II-A/B: Pre/Post-PTA of the delayed-change; PTA: pure tone audiometry)

Table 4 The comparison of postoperative hearing loss according to BAEPs changes

BAEPs change (Wave V)		Patients, *n* (%)	Postoperative hearing loss, *n*
Only amplitude decrement (≥50%)		12 (1.8%)	0
Only latency prolongation (>1 ms) (the observation sign)		9 (1.3%)	0
Latency prolongation (>1 ms) with amplitude decrement (>50%) (the warning sign)		93 (13.9%)	0
No change		520 (77.6%)	0
Wave V loss (the critical sign)	Transient loss	26 (3.9%)	0
	Permanent loss	10 (1.5%)	8
Total		670	8

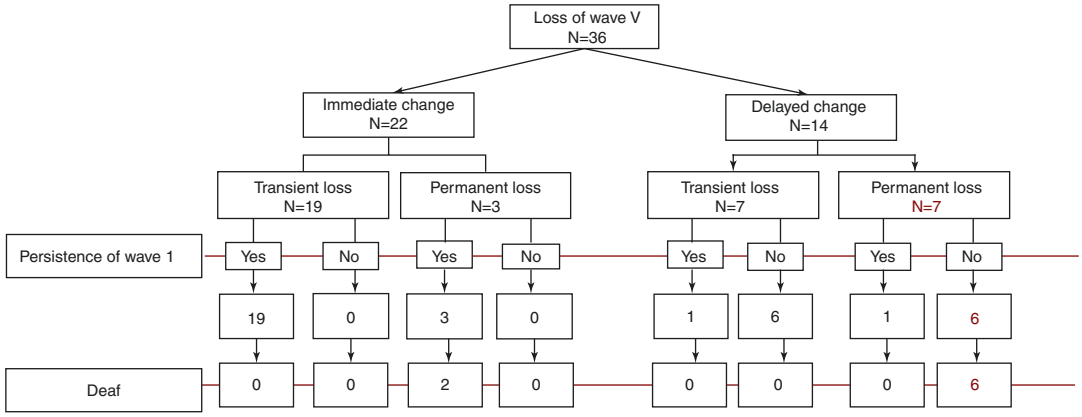

Fig. 15 The differences in postoperative hearing loss according to persistence of wave I in patients with wave V loss during microvascular decompression surgery for hemifacial spasm: Immediate change: Immediate-change group; Delayed change: Delayed-change group; Immediate phase: During the decompressive procedure; Delayed phase: After the decompressive procedure

Most institutions stimulate the temporal or zygomatic branch of the facial nerve about 3 cm lateral to the lateral margin of the orbit during LSR monitoring. The direction of stimulation with paired needles or surface electrodes is centripetal toward the brainstem with the cathode positioned proximally [67, 68].

To our knowledge, there have been no attempts to develop or apply different LSR monitoring methods to increase efficacy. A new LSR monitoring method including facial nerve mapping and centrifugal stimulation of the facial nerve before MVD was studied.

Recording Site

Orbicularis oculi is placed on the eyebrows around the orbit by touching the hand with the electrode so that the electrode does not enter the eye. If you plug in parallel 3~4 cm above the Oculi electrode location, it is in Frontalis position. Orbicularis oris is inserted in the upper lip because it is difficult to plug under the lip because of the intubatio tube. Because the thickness of the upper lip is thin, care should be taken that the electrode does not penetrate through the flesh and into the mouth. If the electrode penetrates into the mouth, noise may be mixed into the electromyogram waveform due to the saliva of the mouth. The mentalis position must be between

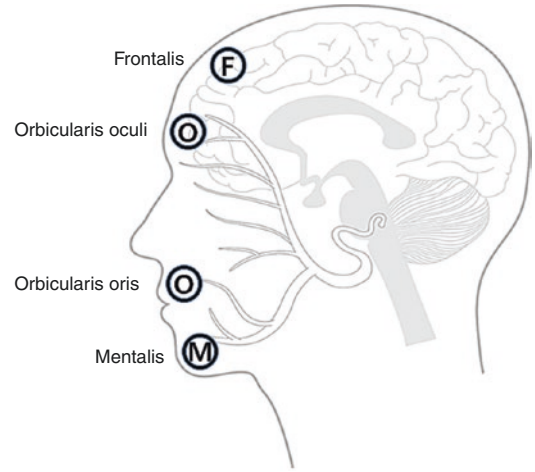

Fig. 16 Illustration of electrode position according to facial nerve branch. The position of the orbicularis oculi is placed in the eye, and the position of the orbicularis oris is located between the nose and the upper lip. The location of the orbicularis oculi is placed at the eye level to eliminate adjacent microscopic connections between the zygomatic branch and the buccal branch. The location of the orbicularis oris is placed between the nose and the upper lip to solve the difficulty of electrode installation because the lower lip is opened due to the intubation tube

the jawbone and the lower lip with the jawbone in the palm of your hand. A person with a lot of flesh is mistaken for the shape of the jaw mixed with the neck, and it is not a mentalis but a mistake of putting the electrode in the neck (Fig. 16).

Stimulation Response

We sought to determine whether electrical stimulation occurs after the facial nerve is fully excited when performing LSR measurements. In other words, we looked at which part of the Upper branch should give the electric stimulus enough to excite the entire Upper branch. We looked at whether the measured LSR after electrical stimulation was better measured so that the upper branch was fully excited [56].

Preoperative facial nerve mapping was conducted for a total of 486 consecutive patients with HFS who underwent surgery at the Samsung Medical Center between February 2015 and August 2016. Patients were monitored for an LSR using centrifugal stimulation of the facial nerve during MVD with the aid of preoperative mapping data.

We observed the response of the muscles that responded to the frontalis and oculi muscles that occurred before the LSR to assess whether the electrical stimulating upper branch was fully excited.

In the same way as the conventional method, stimulation in the stem direction was very difficult to confirm whether or not all the upper branches were fully excited by stimulating several facial parts.

We thus gave electrical stimulation to the peripheral nerve (Fig. 17), as opposed to conventional testing. The response of the muscles in the frontalis and oculi muscles, which are electrically stimulated, was observed to be very responsive. Especially, frontalis and oculi muscular reactions were observed when stimulating various parts of the body by moving the electric stimulating facial region. So when I was stimulating somewhere I was able to clearly see if the Upper branch was fully excited (Fig. 18).

It is also known that the upper branch is fully excited by the well-observed muscle response in both frontalis and oculi muscle reactions, and that LSR measurements in this case also measure a larger amplitude at lower intensities it was.

Stimulation Direction and Intensity

In contrast to the conventional method, we applied electrical stimulation to the frontalis and oculi muscles to detect where the Upper Branch was fully excited.

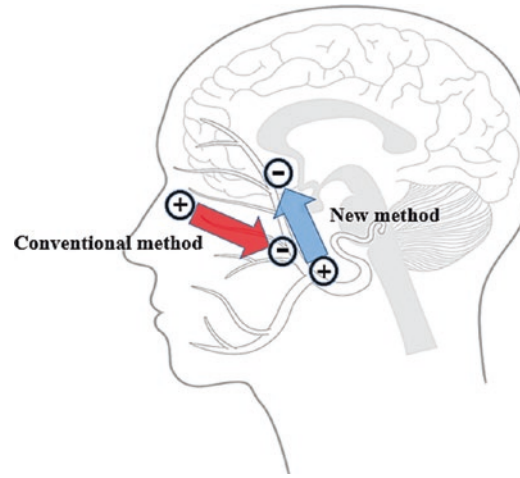

Fig. 17 The direction of stimulation in the conventional and new methods. In the conventional method, electrodes are placed in the temporal or zygomatic branch of the facial nerve, about 3 cm lateral to the lateral margin of the orbit, and centripetal impulses are transmitted toward the brainstem with the cathode positioned proximally. In contrast, electrodes were inserted intradermally with the anode located proximally over the area just anterior to the mandibular fossa and the cathode located distally in the temporal branch of the facial nerve in the new method. The direction of stimulation was centrifugal outward from the brainstem

We used a preoperative test to fix the temporomandibular joint to the anodic electrode in order to use the LSR in the same area as the well-measured area. In addition, the cathode electrode was divided into three large areas and stimulated.

Three directions were designated as 'F' for the frontalis electrode, 'O' for the oculi attached to the oculi, and 'F-O' for the midpoint between the frontalis and oculi (Fig. 19).

LSR was measured in 428 out of 486 patients. Direction F was the most frequent in 325 (66.9%), direction F-O was in 91 (18.7%), and direction O was in 12 (2.5%), and this is not a zygomatic branch but a temporal branch (Table 5).

We have experienced the patient's condition good after surgery even if the LSR is not lost. It is also known that the time of LSR disappears during surgery is very diverse.

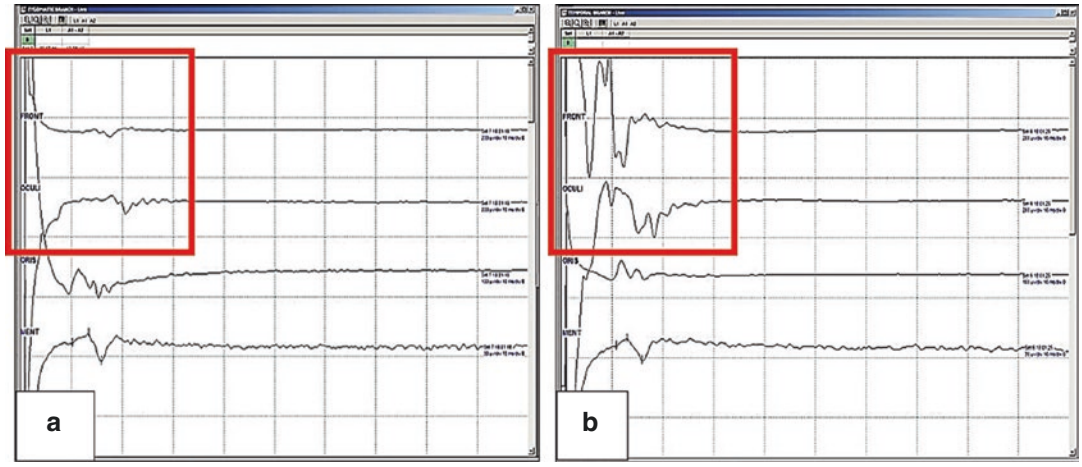

Fig. 18 The response measured in the frontalis and oculi muscles when the upper branch was stimulated by the conventional stimulation method was very small (**a**). The response measured in the frontalis and oculi muscles when the upper branch was stimulated by the new method stimulation was very large, making it easy to assess whether the upper branch was sufficiently excited (**b**)

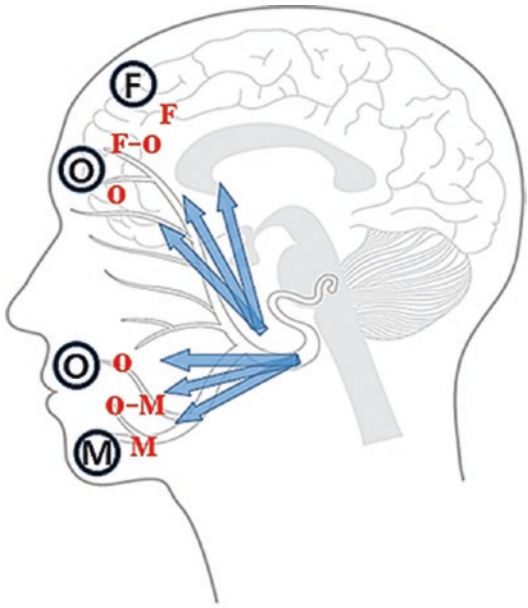

Fig. 19 Facial nerve mapping performed preoperatively. The anode was the reference point or pivot and was placed over the location just anterior to the mandibular fossa. The cathode was first placed in the direction of the frontalis muscle, vertical with respect to the anode, and then moved fanwise toward the direction of the orbicularis oculi muscle while stimulating the facial nerve. The locations of maximal LSR elicitation were divided into three regions: F (the direction toward the frontalis muscle, which was almost vertical with respect to the anode), O (the direction toward the orbicularis oculi muscle), and F-O (in between F and O). In the upper branch stimulation, the LSR measurement was the highest when temporal branch stimulation was performed, and in the lower branch stimulation, the LSR measurement was the highest during the mandibular branch stimulation

Table 5 LSR, lateral spread response; F, direction which was almost vertical to stylomastoid foramen; F-O, in between F and O; O, direction toward orbicularis oculi muscle

n (%)	
Location of maximal LSR	
F	325 (66.9)
F-O	91 (18.7)
O	12 (2.5)
Preoperative LSR	428 (88.1)
Intraoperative LSR positive	419 (86.2)
Post-decompression disappeared	404 (96.4)
Post-decompression persistent	15 (3.6)

These results are from Lee et al. [56]

We do not know the exact meaning of the LSR, but I think it is very important to observe that the LSR is lost due to any manipulation during surgery.

So every time we measure the LSR, we give the electric stimulus to the lowest intensity at which the LSR is measured. For example, if the LSR starts to be measured from 5 mA, the electric stimulus intensity at which the amplitude of the LSR waveform becomes the maximum is found by increasing the electric stimulus intensity by 1 mA.

Then, in the reverse order, the stimulus intensity is gradually lowered and the test progresses. Then, the minimum intensity of the electric stimulation in which the waveform is formed in

proximity to the maximum amplitude is searched for, and the stimulus intensity is continuously tested during the operation.

When the offending vessel is slightly removed from the facial nerve during surgery, the LSR disappears immediately. Once the vessel is in place again, the LSR is measured again. In this way, it is very advantageous to discriminate whether it is a true offending vessel because LSR changes are observed very sensitively with the minimum electric stimulus intensity.

If you do not do this and do your tests with strong electrical stimulation, after the decompression, the LSR is not lost or the amplitude of the waveform is slightly decreased. Therefore, additional procedures that are not necessarily required are performed, resulting in hearing loss or facial palsy [35, 44, 49].

We were able to obtain a very sensitive response by testing with minimal electrical stimulus intensity in the LSR test and when it is judged that the decompression is perfect even if the LSR is not lost, additional procedures are not performed [50].

Compare Conventional and New Method

We performed LSR monitoring using centrifugal and conventional, centripetal methods simultaneously in 62 patients and compared the outcomes of the methods.

The conventional LSR measurement was performed in 34 patients (61.8%) before decompression, after decompression, 16 (29.1%) LSRs were observed without loss. In the new method, LSR measurement was 54 (98.2%) before decompression, after decompression, 1 (1.8%) LSR was observed without disappearance. In other words, the LSR measurement was smoother and the LSR was lost even after decompression (Table 6).

Among 419 patients, in LSR patients, 404 patients (96.4%) lost LSR after decompression, 15 patients (3.6%) remained unresolved (Table 5).

New methods of testing than the conventional method had to give a stronger electrical stimulus intensity of 0.476 mA on average, the latency of the waveform was measured an average of 0.725 ms later, the amplitude of the waveform was observed to be 7.892 μV small (Table 7).

Table 6 Comparison of efficacy of LSR monitoring by using the conventional and new methods

	Conventional method (%)	New method (%)	*p* value
No LSR	9.1	0.0	<0.0001
Disappearance of LSR	61.8	98.2	0.0012
Persistence of LSR	29.1	1.8	0.0051

LSR lateral spread response
These results are from Lee et al. [56]

Table 7 Parameters of intraoperative LSR monitoring (mean ± SD)

	Previous	New	*p*-value
Latency (ms)	11.255 ± 2.145	11.980 ± 1.567	0.0017
Amplitude (mV)	46.659 ± 50.081	38.767 ± 45.163	0.0600
Stimulation intensity (mA)	11.633 ± 5.594	12.109 ± 5.051	0.4305

LSR lateral spread response, *SD* standard deviation
These results are from Lee et al. [56]

Facial Nerve Innervation

The conventional LSR measurement method gives the stimulus to the stem direction in the zygomatic branch, in this case, it is impossible to exclude that the zygomatic branch and the buccal branch are connected to each other [69, 70].

Comprehensive analysis of the branching pattern of the facial nerve is classified into six types, and zygomatic branch and buccal branch are connected to each other, accounting for more than 60% of C, E, and F. therefore, The facial nerve branch used for LSR measurement is the most effective temporal branch and mandibular branch. If the zygomatic branch or buccal branch is selected as the stimulation site for LSR measurement, residual LSR may be present because the upper and lower portions of the facial nerve branch can be stimulated at the same time (Fig. 20).

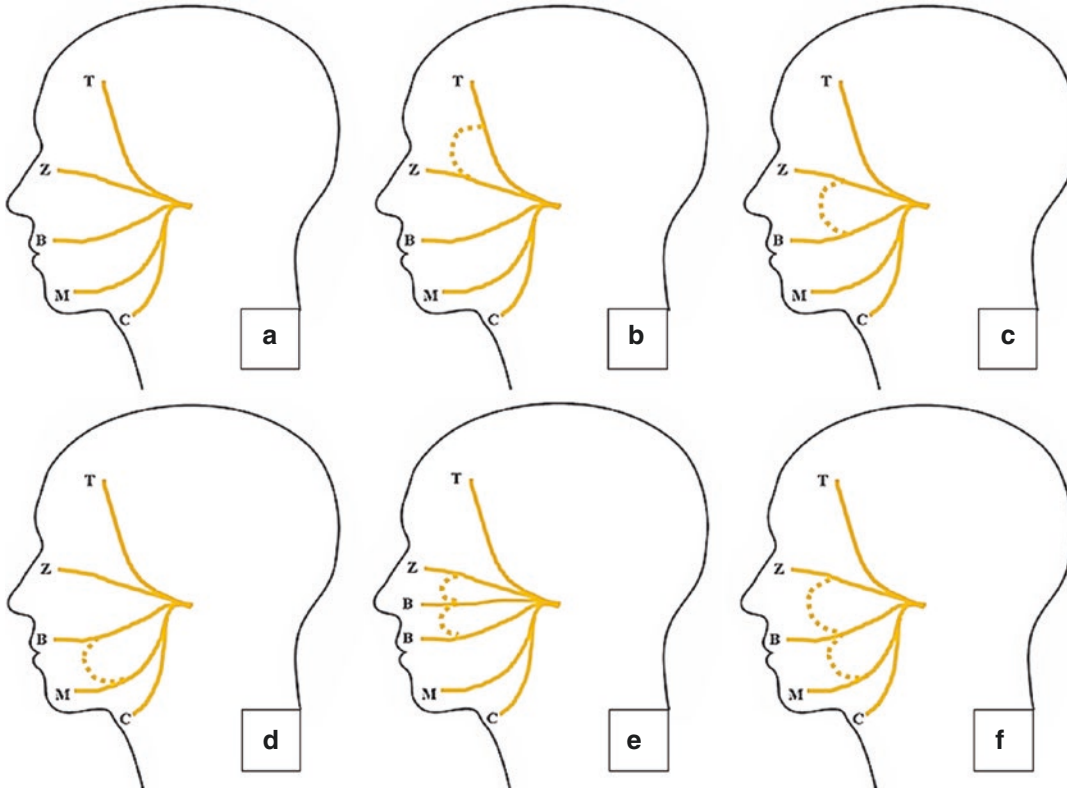

Fig. 20 The phenomenon in which the facial nerves are finely connected to each other is divided into six major categories. (**a**) is the basic shape of the branches of the facial nerve. (**b**) is a shape in which temporal branch and zygomatic branch are finely connected to each other. (**c**) is the shape that the zygomatic branch and the buccal branch are finely connected to each other. (**d**) is a shape in which the buccal branch and mandibular branch are finely connected to each other. (**e**) is the shape that the buccal branch is very developed and is finely connected to the zygomatic branch. (**f**) is a shape in which zygomatic branch, buccal branch, and mandibular branch are finely connected to each other. T, indicates temporal branch; Z, zygomatic branch; B, buccal branch; M, mandibular branch; and C, cervical branch

In general, the residual LSR measurement rate is observed in Thirumala 17.0% [54], Damaty 24.0% [51], Lee 25.6% [52].

However, if we stimulate the upper stimulation to the temporal branch in the opposite direction, the stimulation response of the frontalis and oculi was significantly increased, indicating that the upper branch was fully stimulated.

The LSR that occurs in this state, even if a patient with a Zygomatic branch and a Buccal branch are connected to each other, they can stimulate only the upper branch, which can help to eliminate residual LSR caused by insufficient stimulus.

According to the study, LSR loss rate during postoperative decompression was 96.4%, only 57 of 419 patients remained undisturbed and the measurements were very low at 3.6% [56].

In addition, the LSR measured by upper branch stimulation only observes changes measured in mentalis among the lower branch responses, and the LSR measured by lower branch stimulation only observes changes measured in frontalis among the upper branch responses. You can increase your sensitivity in evaluating whether you have (Fig. 21).

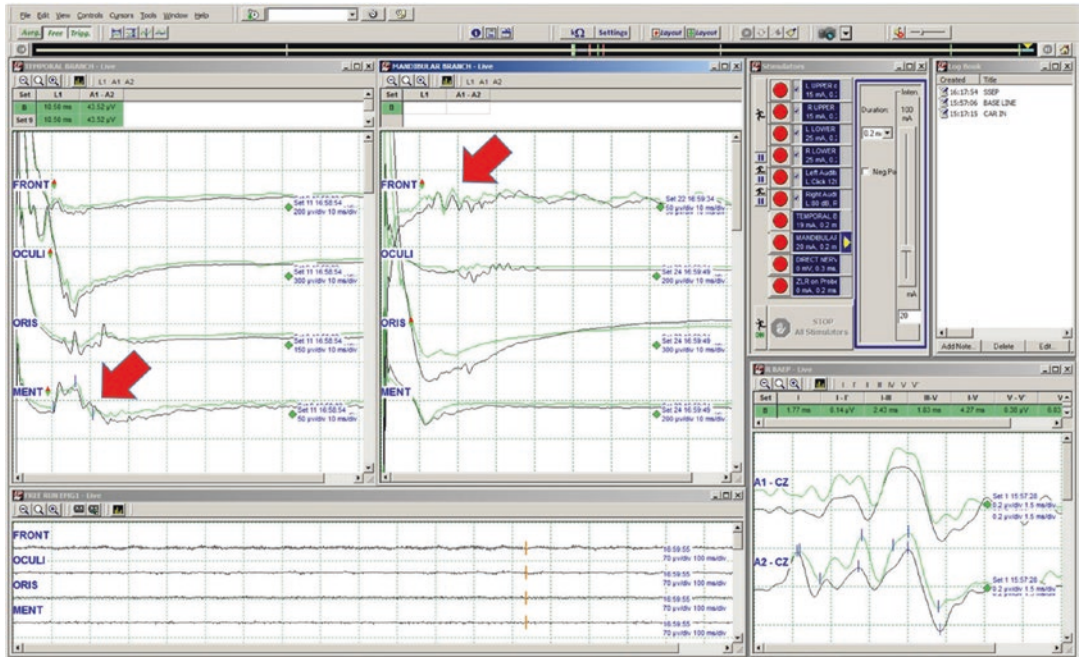

Fig. 21 LSR measurements observed changes in mentalis at upper branch stimulation and changes in frontalis at lower branch stimulation. It is advantageous to configure on the same screen in order to discriminate with the free-running EMG how much touching the facial nerve during surgery. The LSR should also be measured while observing the changes in the very sensitive BAEPs waveforms, so it is best to configure the same screen

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Anesthetic Management of MVD

Jeong Jin Lee

Current anesthetic management during surgery for hemifacial spasm follows standardized anesthetic protocols. Most patients present with a good general condition and the surgery is often performed electively. Regardless, anesthesiologists should keep in mind potential problems that may arise during posterior fossa surgery, including possible injury to vital brainstem centers; pneumocephalus; and, with unusual patient positioning, C-spine injury and upper airway swelling or decubitus injury of the dependent portion [1–3]. Other considerations include the potential usage of intraoperative neurophysiologic monitoring (IONM). In this chapter, anesthetic management during posterior fossa surgery will be discussed, focusing on the completion of microvascular decompression (MVD) for the resolution of hemifacial spasm.

Preoperative Management

During the process of medical care, patient records and laboratory data are reviewed for the presence of hypertension, diabetes mellitus, cardiovascular disease, and other medical problems. If uncontrolled or active disease is found, elective surgery should be postponed to ensure a good physical state. Airway examination should be performed. In cases of high Mallampati scores, limited neck extension, or loose teeth, the anesthesiologist should anticipate the likelihood of difficult intubation and prepare rescue intubation modalities. Patients should fast for at least 8 h beforehand.

Intraoperative Management

When the patient arrives in the operating room, electrocardiography, oxygen saturation monitoring, and noninvasive blood pressure (BP) monitoring begin and a neuromuscular monitoring probe is applied to the ulnar nerve. The induction of anesthesia begins with an intravenous bolus injection of thiopental or propofol and opioid analgesic agents such as remifentanil, fentanyl, or sufentanil. After confirmation of an abolished eyelid reflex, baseline twitch height (T_0) measurements should be collected and the injection of a neuromuscular-blockade (NMB) agent (NMBA) such as rocuronium or vecuronium should be completed to establish an optimal intubation condition. Anesthesia is maintained with total intravenous anesthesia (TIVA) using propofol and remifentanil, or balanced anesthesia with volatile anesthetics and an opioid. Without IONM, there is no significant restriction on the choice of anesthetic agents.

Arterial catheterization can detect sudden changes in BP and identify blood gases and

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_12

electrolytes as needed. A bispectral index (BIS) monitor should be attached to the forehead to measure the depth of anesthesia, and the propofol infusion rate should be titrated to maintain a target BIS value of 40–60. Importantly, the patient must not move during microsurgery; unanticipated bucking or straining may induce a catastrophic outcome. Such can be prevented with a cautious titration of appropriate anesthetic agents including NMBA and analgesics. Spontaneous activity as seen via facial nerve electromyographic (FNEMG) monitoring may precede physical movement. Volatile anesthetic agents had more hemodynamic variability and spontaneous activity on FNEMG, whereas TIVA was proven to be a more effective anesthetic for preventing patient physical movement when clinically titrated to produce stable operation conditions [4]. When spontaneous activity on FNEMG occurs, increasing the dose of NMBA or opioid can prevent the patient's movement. If NMBA is not used at all during surgery, deeper anesthesia is needed, but, with such, extreme hypotension and bradycardia can develop. This problem can be mitigated by infusing inotropic agents. Arterial BP should be controlled to within 30% of the preoperative value measured on the day before surgery.

Positioning

Most MVD surgeries can be performed using the park-bench position (three-fourths lateral prone decubitus) (Fig. 1), although the prone position or the sitting position has been preferred by some surgeons [1]. The assurance of careful positioning and padding can help to avoid injuries; for example, a gel roll should be placed under the axilla of the dependent side, while other pressure points such as the elbows, wrists, ischial spines, and heels should be protected. Excessive neck flexion may induce venous obstruction or cervical spine injury, which causes increased intracranial pressure (ICP), upper airway edema, or quadriplegia. Preexisting cervical spinal stenosis may predispose patients to cervical spine injury. Confirming that the airway pressure under controlled ventilation with fixed tidal volume has not risen during head fixation can avoid problems of excessive neck flection. Avoid compression or protrusion of tongue to prevent ischemia or venous congestion of the tongue. After positioning, patients should be checked carefully for signs of obstruction such as discoloration of the face, lips, or tongue, especially given that, during the operation, the patient's face can be more difficult to access because the operating table rotates 90 or 180 degrees. The corrugated tube and monitoring lines should be carefully secured before the start of the operation.

Anesthesia for Intraoperative Neuromonitoring

Various electrophysiologic monitoring techniques may be used during surgery for the central nervous

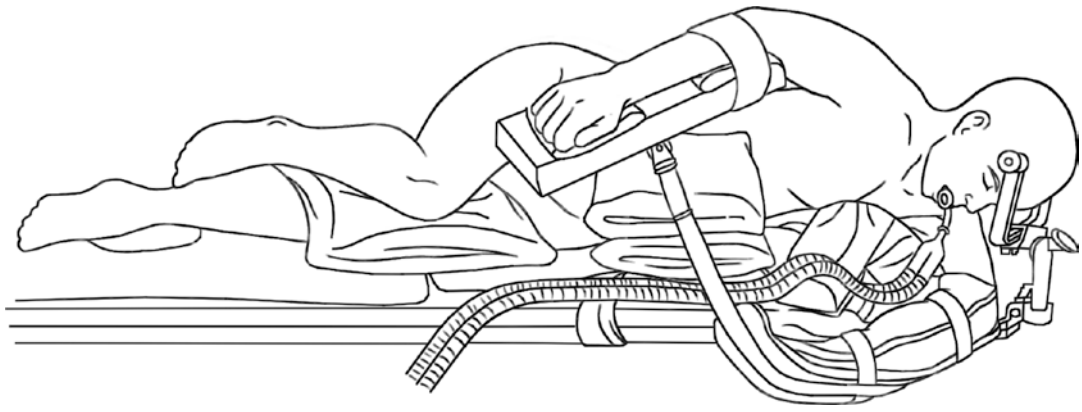

Fig. 1 Park-bench position

system. These include somatosensory evoked potentials (SSEPs), visual evoked potentials (VEPs), motor evoked potentials (MEPs), brainstem auditory evoked potentials (BAEPs), and electromyographic (EMG) monitoring of the cranial nerves (e.g., facial nerve, glossopharyngeal nerve). In general, during MVD surgery, SSEPs, BAEPs, and facial nerve EMG (FNEMG) are essential intraoperative monitoring. Most anesthetics affect IONM to varying degrees (Table 1).

Sensory evoked potential (SEP) monitoring affects the selection of anesthetics. In general, volatile anesthetics affect SEPs more than intravenous anesthetics. Evoked potentials (Eps) of cortical origin (i.e., the cortical portion of SSEPs and VEPs) are considered more prone to modification by anesthetics than brainstem potentials (i.e., BAEPs and subcortical portions of SSEPs). However, the neurologist can receive an interpretable SSEP wave under volatile anesthetics. In contrast, BAEPs are almost always entirely unaffected by anesthetics. It is essential to maintain constant anesthetic drug levels during the recording of Eps [5].

MEPs are affected by both the anesthetic agent and NMBA. Myogenic MEPs are easily suppressed by anesthesia, especially by volatile anesthetics [6]. Without NMBA, MEP waves can be monitored with volatile anesthetic agents but, with partial NMB, they are significantly abolished by a small dose of volatile anesthetics [7]. Intravenous anesthetics such as propofol and dexmedetomidine also affect myogenic MEPs, albeit to a lesser extent. Anesthesiologists prefer to use propofol in conjunction with an opioid during MEPs. NMBA does not affect D-waves but will affect myogenic MEPs. A complete withdraw of NMB can easily ensure a clear myogenic MEP wave, but the patient can move during transcranial electrical stimulation, which can interfere with the surgery and often puts the patient in a dangerous situation. Partial NMB may reduce but not abolish these movements and can complicate the interpretation of myogenic MEPs. Some centers generally omit NMB, whereas others tend to use partial NMB. If employed, partial NMB should be achieved with a continuous infusion of the NMBA with close titration under neuromuscular monitoring such as the assessment of the responses to train-of-four stimulation [8] to assess the degree of NMB. Bolus injections should be avoided because they induce too variable NMB levels [9]. Kim et al. reported that, under TIVA with propofol and remifentanil, partial NMB with a target T2/Tc of more than 50% can achieve acceptable MEP waves, but complete withdrawal of the NMBA can lead to more steady MEP waves [10].

Facial nerve-triggered EMG is essential monitoring during MVD. The lateral spreading response (LSR) is considered to be an effective diagnostic tool for complete decompression and also to be an important prognostic factor in MVD surgery [11]. Because EMG waves are influenced by NMBA, this requires that the patient not be paralyzed or been in a constant state of incomplete paralysis. In general, a TOF count of more than two has been recommended. It is well-known that facial muscles are more resistant to NMBA than the adductor pollicis muscle. However, Chung et al. reported that the maintenance of partial NMB with a target T1/Tc ratio of 50% rather than a TOF count of two resulted in a clinically acceptable success rate for LSR monitoring and surgical condition during MVD [12]. When NMB was completely removed, the success rate of LSR monitoring was not significantly different from when the target T1/Tc ratio was maintained at 50%, but the LSR response after decompression was not as easily defined in the patients without NMBA because of the delayed abolishment of LSR and spontaneous free-run

Table 1 Influence of anesthetic agents on IONM

	SSEP	BAEP	MEP	EMG	Facial LSR
Sevoflurane	+	–	++	–	N/A
Desflurane	+	–	++	–	+
Remifentanil	+	–	±	–	N/A
Dexmedetomidine	±	–	+	–	N/A
Propofol	±	–	+	–	±
Muscle relaxant	–	–	++	++	++

SSEP somatosensory evoked potential, *BAEP* brainstem auditory evoked potential, *MEP* motor evoked potential, *EMG* electromyography, *Facial LSR* facial lateral spreading response

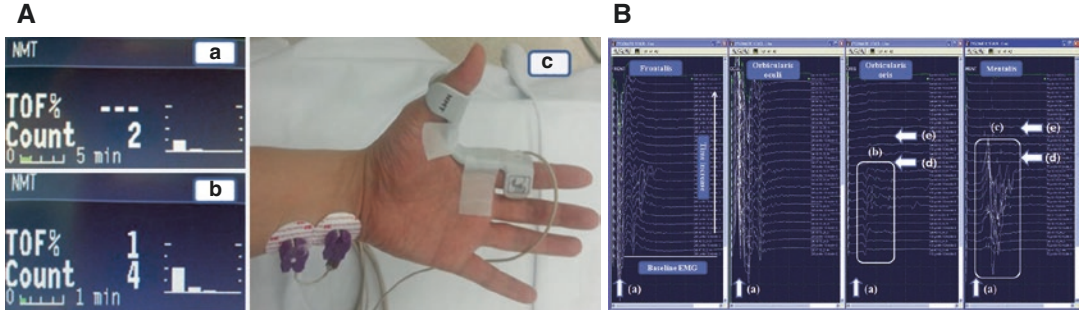

Fig. 2 (**A**) Images of neuromuscular blockade (NMB) monitoring using NMT module. The target of two counts of train-of-four (TOF) in TOF group (a), T1/Tc ratio of 50%inT1 group (b), and set up image (c). (**B**) Stack wave image of an intraoperative facial EMG upon stimulation of the zygomatic branch in a patient with partial NMBA maintained with a T1/Tc of 50%. Electrical stimulation of the zygomatic branch (a) produced an abnormal muscle response, called the LSR, in the orbicularis oris (b) and mentalis (c) muscles. This abnormal response disappeared (e) after decompression (d) [13]

EMG activity. Maintenance of partial NMB with a target T1/Tc ratio of 50% during LSR monitoring for MVD can therefore be recommended (Fig. 2) [13]. In general, facial nerve EMG is known to be affected by NMBA alone. However, a recent report suggests that desflurane (1 MAC) suppresses the LSR amplitude by 43% compared to TIVA alone, providing direct evidence that there is a central mechanism of action inherent in the origin of the LSR [14]. Most of all, close conversation between the neurosurgeon, neurologist, and anesthesiologist to optimize IONM, attain hemodynamic stability, and avoid unexpected movement of the patient is key.

Trigeminal Arrhythmia

Manipulation of the sensory branches of the trigeminal nerve is known to cause autonomic changes, such as bradycardia or asystole, known as the trigeminocardiac reflex (TCR). A risk for TCR should be considered in any craniomaxillofacial surgery, especially during surgery performed at or near the cerebellopontine angle (e.g., acoustic neuromas; microvascular decompression of the trigeminal, facial, and glossopharyngeal cranial nerves). One retrospective study showed that TCR occurred in 18% of microvascular trigeminal decompression patients: of note, their heart rate decreased by 46% and their mean arterial blood pressure decreased by 57% during their operative procedures performed near the trigeminal nerve compared to the levels recorded immediately before the stimulus [15]. Knowledge of the applied anatomy will help in predicting this risk and choosing the appropriate prevention. If the risk of TCR is high, an intravenous anticholinergic agent like atropine or glycopyrrolate may be used to prevent such. Intraoperative predisposing factors are light plane of anesthesia, hypercarbia, hypoxia, and acidosis. If a TCR is suspected, anesthesiologists should check and correct those factors and inform the surgeon to stop stimulating and wait for the pulse to return to normal. Administration of intravenous anticholinergic agents after heart rate recovery can be used to prevent the recurrence of TCR. The surgeon then should continue the operation with gentle manipulation [16].

Cranial Nerve Dysfunction and Respiratory Center Injury

Operations in the posterior fossa can injure vital circulatory and respiratory brainstem centers and cranial nerves or their nuclei. Such injuries may occur as a result of direct surgical trauma or ischemia from retraction or other interruptions of the blood supply. Cranial nerve dysfunction, particularly of nerves IX, X, and XII, can result in a loss

of control and patency of the upper airway. EMG of lower cranial nerves during surgery can help detecting the risk of cranial nerve injury ahead. Abnormal cardiovascular responses can result from irritation of the lower portion of the pons and upper medulla. These areas are most often stimulated during procedures on the floor of the fourth ventricle. Isolated damage to respiratory centers may rarely occur without preceding circulatory signs during surgery. Therefore, abrupt onset of bradycardia and hypotension, tachycardia and hypertension, or bradycardia and hypertension as well as ventricular dysrhythmias should alert the anesthesiologist to the possibility of such an injury [3, 17]. Meticulous attention to the electrocardiogram and a directly transduced arterial pressure during manipulation in this region are necessary to provide the surgeon with an immediate warning of the risk of damage to the adjacent cranial nerve nuclei and respiratory centers. The posterior fossa is a relatively small space, and its compensatory latitudes are even more limited than those of the supratentorial space. Relatively little swelling can result in disorders of consciousness, respiratory drive, and cardiomotor function. Abnormal respiration after MVD is more likely to occur when the brainstem injury is the result of vessel occlusion or hematoma (which can develop in a delayed manner) rather than direct mechanical damage caused by retraction of or dissection in the brainstem [18].

Macroglossia

An abnormal swelling of the tongue is a rare postoperative complication often associated with serious airway obstruction and prolonged intubation. Macroglossia has been most commonly reported after suboccipital and/or posterior fossa craniotomies and spine surgeries performed with the patient in the prone or park-bench positions. Risk factors for macroglossia include a surgery duration of more than 8 h, type of surgery (i.e., suboccipital and/or posterior fossa craniotomies and spine surgeries), and patient position (i.e., prone, park-bench, or sitting). The etiology of macroglossia is multifactorial and possible mechanisms include local mechanical tongue compression interfering with venous and/or lymphatic drainage, regional venous thrombosis, and/or local trauma. Prevention, awareness of the possibility, and early recognition are the best forms of treatment (see *Positioning*) [19, 20].

Pneumocephalus and Cerebrospinal Fluid Leakage

Although the sitting position notably increases the likelihood of pneumocephalus, this can also occur in the park-bench or prone positions [2]. Air enters the subarachnoid space, as cerebrospinal fluid (CSF) is lost during surgery. In patients with cerebral atrophy, drainage of CSF is marked; air can replace the CSF on the surface of the brain and in the lateral ventricles. Expansion of a pneumocephalus following dural closure can compress the brain. Postoperative pneumocephalus can cause delayed awakening and continued impairment of neurological function [21]. Because of these concerns, nitrous oxide is rarely used for sitting craniotomies [3]. Sometimes the surgeon asks for induction of a Valsalva maneuver (VM) to confirm a watertight dural closure. If the dural closure is not tight, CSF is expelled following a rise in ICP and air is sucked inside the cranium to equalize the ICP, opening up the possibility of pneumocephalus. In addition, forceful or prolonged VM can be associated with profound and complicated physiological responses and significant complications can occur. Therefore, clinicians should be well-acquainted with its applied physiology and use it judiciously to avoid the complications associated with it [22]. When we used high-dose volatile anesthetics, which were potent cerebral vasodilators, the brain could bulge out after dura opening and an infusion of mannitol or application of hyperventilation was mandatory. However, during TIVA anesthesia, infusing mannitol or applying hyperventilation in order to reduce the ICP during surgery should be done in consultation with the surgeon [23].

Venous Air Embolism

Venous air embolism is a very rare complication which can occur when the pressure within an open vein is subatmospheric. This scenario may occur in the context of any surgical position (and during any procedure) whenever the wound is above the level of the heart. The incidence of venous air embolism is greater during sitting craniotomy (20–40%) than in any other position. Air entry into large cerebral venous sinuses increases the risk. The most sensitive monitoring modalities are precordial Doppler and transesophageal echocardiography. However, these technologies are not necessary if the patient is not in a sitting position. An abrupt decrease in end-tidal CO_2 is another indicator of an air embolism. When an air embolism is detected, anesthesiologists should warn surgeons to stop surgery and apply saline or water-soaked gauze. Change the patient's position to head-down and right-side-up to prevent the air from traveling to the coronary or cerebral artery is another step [3, 17, 23].

Postoperative Management

Extubation and Airway Management

Most patients awake immediately after surgery and extubation is possible. However, in rare cases, respiratory center injury or macroglossia may occur. If there is evidence of macroglossia at the end of surgery, the patient should not be extubated. If there is suspicion of a circulatory center injury (i.e., severe hypertension, bradycardia, or arrhythmia during the operation) this should be taken into account during planning for extubation and postoperative care. If macroglossia develops after extubation, early airway management should be considered [24]. Airway management may be difficult and a surgical airway may be needed. Other supportive treatments may be required, such as placing the patient in a head-up position, preventing further tongue compression, steroids, keeping the tongue moist to prevent desiccation, and administering analgesics as needed [19, 20].

Nausea, Vomiting, and Headache

Patients, after MVD of the cranial nerves, frequently experience postoperative nausea and vomiting (PONV). MVD and acoustic neuroma resection were associated with an increased likelihood of PONV as compared with craniotomy performed for other tumor resections [25]. The risk was also higher among patients who underwent MVD of the trigeminal neuralgia than those treated for hemifacial spasm. Female sex was more susceptible to PONV and the use of a volatile agent increase the risk of PONV. Using intravenous anesthesia and prophylactic antiemetic agents like ondansetron, ramosetron, and metoclopramide can prevent PONV. However, despite the use of intraoperative prophylactic ondansetron, the overall incidence of PONV (nausea, emesis, or both) was reported to be 60% during the first 24 h after surgery. Application of a prophylactic transdermal scopolamine patch may also help to prevent PONV. It may be necessary to administer a combination of antiemetics to decrease the incidence of PONV after retromastoid craniotomy [26]. When vomiting occurs, the patient's head should be rotated to one side to avoid aspiration and symptomatic treatment should be provided.

Other Considerations

MVD surgery is known to be less invasive and safe. However, it is still challenging to perform and may result in a fatal outcome. In a retrospective analysis of 46 patients (0.66% of 6974 cases of MVD), who presented with a decline in consciousness after waking up from the anesthesia, 15 patients (0.2%) died. The authors reported that mortality is significantly higher in trigeminal cases with cerebellar hematoma and an immediate hematoma evacuation plus ventricular drainage could give the patient a greater chance of survival [27]. Most of the patients go to the intensive care unit after surgery. However, in case the patient is transferred to the postoperative care unit, continuous monitoring of blood pressure, pulse, respiration, and blood oxy-

gen saturation is mandatory. The patient's consciousness and pupillary changes should also be observed closely. If the patient shows a sudden increase in BP accompanied by bradycardia and unconsciousness, slow and deep breathing or even arrest, decreased oxygen saturation, dilated pupils, and light reflex diminished or disappearing, the possibility of infarction, swelling, or bleeding in the cerebellum or brainstem should be considered [28]. Immediate communication with the neurosurgeon is required, an emergent head computed tomography scan should be considered, and other immediate actions as necessary should be taken.

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Botulinum Toxin Injection in Hemifacial Spasm

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Hemifacial spasm (HFS) is a movement disorder characterized by involuntary, irregular, and recurring contractions of the muscles innervated by the facial nerve [1]. HFS is not a life-threatening disease, but can severely affect the quality of life [2–4]. Moreover, HFS could be associated with various psychologic symptoms, including low self-esteem, social embarrassment, social isolation, and depression [4, 5]. Although HFS could improve or even resolve without any treatment [6], HFS rarely resolves spontaneously because it mainly results from vascular compression of the facial nerve at the root exit zone [7]. Therefore, effective management is important for the quality of life, even tough HFS is not a fatal disease. Accordingly, the majority of HFS patients undergo treatment, such as symptomatic or curative options, and 1570 (88.5%) of 1775 HFS patients performed microvascular decompression or botulinum toxin injection therapy in previous study [6]. Additionally, the severity of HFS means severe indentation of facial nerve [8], and severe spasm is related with more impaired quality of life [2]. HFS patients with severe spasm might have even less chance for improvement or remission; thus, symptomatic or curative treatment is strongly recommended in severe HFS. Although the only curative option is surgical relief of the neurovascular compression in HFS, botulinum toxin injection is an evident, effective and safe treatment options to control the spasm, because HFS is a benign disease and the spasm itself could be well-controlled with injection therapy.

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Botulinum Toxin Injection as Management Option for Hemifacial Spasm

History of Botulinum Toxin Injection in Hemifacial Spasm

The initial trial with botulinum toxin was done in patients with strabismus [9], and botulinum toxin tested was unreliable, short-acting, or necrotizing at the early period [10]. Finally, with the purifying techniques and extensive animal experiments, botulinum toxin showed desired long-lasting, localized, dose-dependent muscle weakening without any previous side effects. By

K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_13

the early 1980s, botulinum toxin was injected for various diseases such as strabismus, blepharospasm, hemifacial spasm, cervical dystonia, and thigh adductor spasm. At Samsung Medical Center (Seoul, Korea), botulinum toxin injection clinic is run by the Neurology Department, and we usually perform more than 1500 injections annually. Among the patients with botulinum toxin treatment at Samsung Medical Center on 2017, 371 (50%) of total 744 patients were HFS patients. For all indications illustrated in Figure 1, HFS was the most commonly injected indication, and blepharospasm wa the second most common indications among various movement disorders for botulinum toxin injection therapy.

With various previous studies during two decades, botulinum toxin treatment has emerged as the first-choice treatment option for hemifacial spam, as well as microvascular decompression surgery [11–13]. Two randomized controlled trials and more than 30 open label studies, encompassing more than 2200 patients, have been already published, and botulinum toxin injection demonstrated excellent improvement in terms of symptom control [14, 15]. Additionally, evidence-based review by the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology concluded that botulinum toxin injection is possibly effective with minimal side effects for the treatment of hemifacial spasm (one Class II and one Class III study) [16]. In addition, various botulinum toxin agents that are commercially available were also studied, and regarded to have similar effects for HFS [17].

For long-term efficacy, treatment with botulinum toxin appears to remain effective over long-term use of several years (from 4 to 20 years). In most cases, botulinum toxin treatmant will not require dosage increase, and even if required, the dosage increase usually occurs within the first 2 years of treatment [18].

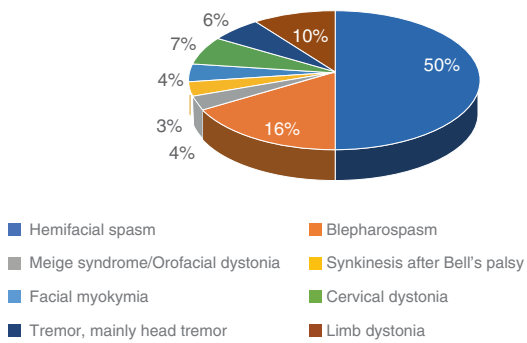

Fig. 1 The indications for botulinum toxin treatment at Samsung Medical Center for 1 year (2017). Total number of patients was 744, and almost half of the whole patients (n = 371) were injected because of HFS

Strategy to Decide Botulinum Toxin Injection in Patients with Hemifacial Spasm

Even though botulinum toxin treatment is an effective and safe treatment option in hemifacial spasm, botulinum toxin injection therapy is symptomatic management option unlike surgical relief of neurovascular compression. Considering the different characteristics of each treatment option, treatment modality should be decided with discussion based on three considerations [19].

- Patient-related factors: age at surgery, age at symptom onset, occupation, alcohol use, smoking history, family history, history of facial palsy, contralateral hearing loss and comorbidities, including, hypertension, diabetes, hyperlipidemia, and cardiovascular disease
- Disease-related factors: affected side, duration of symptoms, severity of spasm, associated symptoms (tinnitus and headache), preoperative facial palsy, and previous botulinum toxin treatment
- Surgery-related factors: offending vessel, number of offending vessels, compressive pattern, indentation on the facial nerve, discoloration of nerve, abnormal muscle responses, postoperative delayed facial palsy, and operation year

Among the patient-related factors, age should be considered. Although surgical decompression is effective in elderly patients as well as young patients, more complications were reported in

elderly patients [20]. Higher surgical risk and more comorbidities in elderly patients has to be also considered the severity of spasm. Additionally, severe spasm is associated with severe indentation of facial nerve [8]; thus, there might be less chance for spontaneous improvement or remission. Although we reported improvement or remission without any treatments in half of the HFS patients [6], 86.0% of the enrolled subjects in this study had mild HFS (SMC grade 1–2), thus this result should be interpreted cautiously. Therefore, for the patients with severe HFS, treatment, such as macrovascular decompression or botulinum toxin injection, is strongly recommended, whereas we can follow up the patients with mild spasm without any treatment.

Interestingly, all these patient-related, disease-related, and surgery-related factors are not independent, but connected with each other. For example, severe spasm (disease-related factor) is marker for indentation of facial nerve (surgery-related factor), and associated with comorbidities, like hypertension or diabetes (patient-related factor), and disease duration (another disease-related factor). Additionally, associated factors with surgical outcome is also important during the decision of treatment option for HFS. For example, botulinum toxin treatment should be considered prior to surgical treatment in the people with the contralateral hearing loss, because ipsilateral hearing loss, possible complication from MVD, can make the patient deaf. The contents about the prognosis and complication for surgical decompression will be discussed in other chapters (Chapters "Possible complications of Microvascular Decompression" and "Prognosis of symptoms after microvascular decompression for hemifacial spasm").

Considerations Before Start Botulinum Toxin Injection in Hemifacial Spasm

In spite of the efficay and safety of botulinum toxin treatment in HFS, botulinum toxin injection is symptomatic management option, and not without side effects. Therefore, discussion with detailed information, such as the duration and latency for response, and possible side effects, should be done before to perform botulinum toxin treatment in HFS patients. Mostly, the effects and side effects happen based on the injected dose and target muscles. Possible side effects of botulinum toxin injections are erythema and ecchymosis of the injected site, dry eyes, mouth droop, ptosis, lid edema, and facial muscle weakness [15, 21]. Among the side effects, ptosis and facial muscle weakness tends to be transient and will resolve within 1–4 weeks. In terms of efficacy, the onset of effect occurs within 3 days to 2 weeks, generally with a peak effect at approximately 2 weeks. The beneficial effects of botulinum toxin injections are also transient with a mean duration of improvement of approximately from 2.8 to 3.1 months [13, 15]. At Samsung Medical Center, the mean duration of response was 3.46 months, and the mean frequency of injection was 2.15 per year for HFS patients. However, for the duration and onset of effect, there is a high variability of duration of the beneficial effect, thus all injections should be personalized for each patient.

The other consideration is the selection of botulinum toxin agent. Onabotulinumtoxin A (Botox, Allergan, Irvine, CA) is the most commonly used among the commercially available preparations. A large number of trials have validated the successful outcomes of botulinum toxin injection therapy with improvements in as many as 75–100% of individuals with hemifacial spasm [11, 22, 23].

Preparation for Botulinum Toxin Injection

Botulinum toxin injections in HFS are usually performed with the patient lying supine. We use specialized chair for botulinum injection on the face (Fig. 2), but it is not mandatory. If it is comfortable for both the patients and doctors for injection, any position is acceptable.

The toxin is diluted to minimum concentration of 10–50 Botox U/ml, 50–200 U/ml Dysport (Ipsen, Milford, MA) or 5000–10,000 U/ml Neurobloc/myobloc (Solstice Neurisciences, Malvern, PA) to minimize diffusion. At Samsung

Fig. 2 The chair and position for botulinum toxin injection on HFS patients' face at Samsung Medical Center

Medical Center, Botox 100 U is diluted in 2 ml of normal saline (50 Botox U/ml), and Dysport 500 U in 3.3cc (Dysport 151.5 U/ml). However, the amount of normal saline depends on the physician's experiences and the responses from each patient. For the muscles that need higher dose of botulinum toxin, like limb or neck muscles, botulinum toxin could be diluted in smaller amount of normal saline. On the contrary, if the patient easily showed weakness from botulinum toxin treatment or had already weakness even before injection, botulinum toxin could be mixed with a larger amount of normal saline.

Other materials for injection treatment should be prepared before starts treatment (Fig. 3). The injections are performed usually with 1-ml syringe with fine needle (30 gauge). Alcohol swab to sterilize injection site and gauge to compression in case of bleeding are also needed.

Other Specific Considerations

Usually, there is no need for management for injection pain, but if patients worry about pain during injection, the pain can be reduced either

Fig. 3 The materials that are prepared before starting botulinum toxin injection at Samsung Medical Center. Botulinum toxins (Botox and Dysport), alcohol swab, syringes, needle, normal saline, and icepack

with skin cooling using ice or with EMLA Cream (lidocaine 2.5% and prilocaine 2.5%) [24, 25]. When we checked our patients at Samsung Medical Center, only 2.4% of HFS patients with botulinum toxin injection needed EMLA Cream for the pain during injections. Unlike in spasmodic torticollis, EMG recordings during injection are not necessary in HFS.

Botulinum Toxin Injection: Injection Site and Dose Selection

HFS usually begins in the periocular region and then progresses to involve the cheek and perioral muscles. The natural course is heterogeneous among the patients, and especially the time to spread to hemi-face and the time to visit clinic from the onset, is very different depending on the patients. The goal of botulinum toxin injection is a symptomatic management, not cure of HFS, thus the injection sites should be decided with careful observation of patients' symptoms (involved muscles) and history taking (main muscles that patients complain of). Sometimes, even though patients have spasm in hemi-face, they just complain of periocular spasm. In these cases, injections in perioral area are not necessary because this injection might cause side effects without any benefit. Additionally, the injection dose should be also personalized. With the same

severity of spasm, some patients may experience severely impaired quality of life, but not for the others. Same for the side effects; it could be tolerable for some patients, but totally not tolerable for the others. Therefore, both injection site and dose should be decided based on full discussion with patients.

Facial Muscles and Injection Sites

For successful injection, the physicians should know the muscle anatomy and function in the face. The main muscles injected for HFS tend to be the orbicularis oculi, corrugator, frontalis, risorius, buccinator, and depressor anguli oris (Fig. 4).

The orbicularis oculi is composed of two parts: the pars palpebralis, which opens (with the help of the levator muscle) and closes the eyelid, and the pars orbitalis, which squeezes the eye shut. The pars palpebralis is composed of two parts: the preseptal and the pretarsal region. Typically, the injection sites in the orbicularis oculi at Samsung Medical Center are medial and lateral part of upper lid, and middle and lateral part of lower lid (Fig. 5), and medial part of lower lid could be added based on the severity of symptoms. For the injection at pars palpebralis, pretarsal injection shows more effects with longer duration and less side effects compared to preseptal injection [26, 27]. Additionally, injection at pars orbicularis could be added. The most important rule is not to inject at the midline of upper eyelid to avoid ptosis. Ptosis could be due to local diffusion of the botulinum toxin affecting the levator palpebrae [28]; thus, too much dilution is not recommended especially for the patients with HFS.

The injection at orbicularis oris should be done carefully, because this injection could result in paralysis of the mouth producing further disability. Even though perioral spasm is controlled with botulinum toxin injection at perioral muscles, ipsilateral upper lip droop could be seen [29]. If patients complain of spasm more than the weakness of perioral muscles, bilateral injection to minimize asymmetry could be another option. However, considering that most HFS patients complain of periocular spasm rather than perioral spasm, the sites of injection should be fully

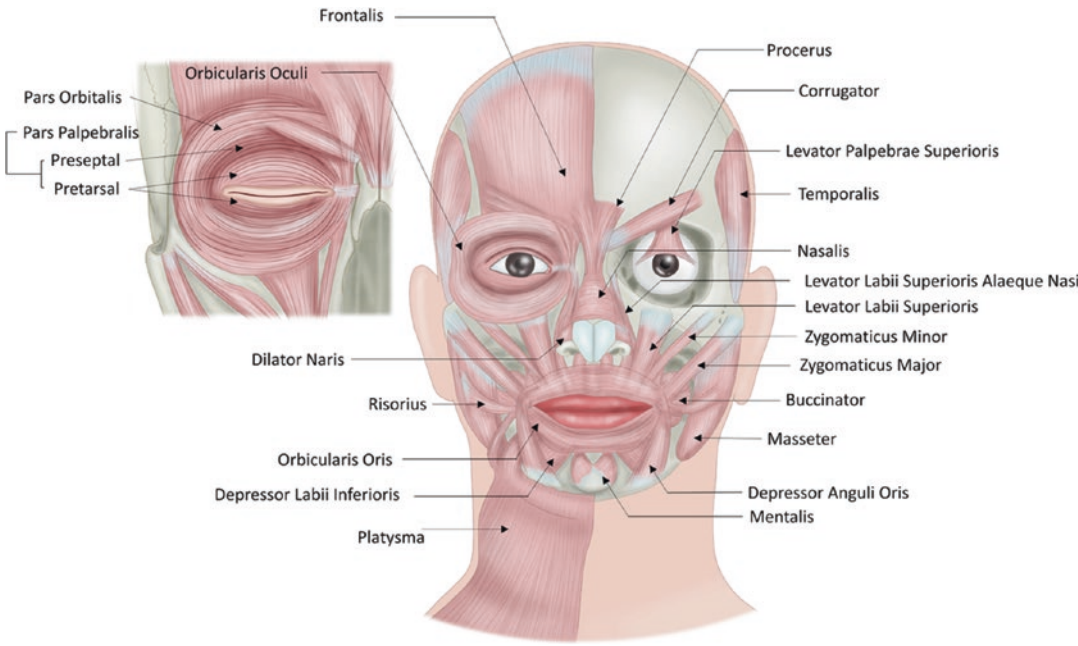

Fig. 4 Illustration of facial muscle anatomy for botulinum toxin injection. The injection at right muscle is the best way to maximize the efficacy and minimize the side effects

discussed with patients. Not all spasm needs to be controlled with botulinum toxin injection.

Injection Doses for Facial Muscles in HFS

The usual doses for commonly injected facial muscle are illustrated in Table 1 [30]. The average dose of botulinum toxin varies from 10 to 46 Botox U [12, 31], from 53 to 160 Dysport U [18, 32–34], and from 1250 to 9000 NeuroBloc/Myobloc U [35, 36]. Based on our experience at Samsung Medical Center, a small dose of botulinum toxin injection is recommended for the first injection, and the dose could be adjusted at follow-up injections based on the response at the previous injection. In particular, the dose for injection should be assessed by each muscle.

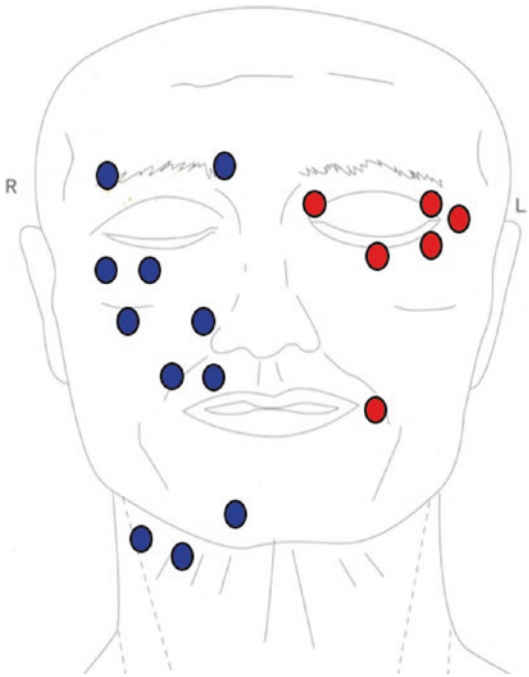

Fig. 5 Injections sites for HFS at Samsung Medical Center. Red dots are typical injection sites and blue dots are sites for additional injections

Unilateral Injection vs. Bilateral Injection

HFS is an unilaterally involved disorder, even though there are some cases with bilateral involvements. Therefore, botulinum toxin injection could be performed unilaterally. However, with unilateral injection, some patients could suffer from the asymmetry (sometimes subjective asymmetry) and bilateral injection could be helpful in these cases. When we compared bilateral injection (n = 33) with unilateral injection (n = 45), asymmetry in lower face was more common in unilateral injection than in bilateral injection (75.6 vs. 48.5%, unilateral and bilateral injection respectively, p = 0.033). Asymmetry was more prominent during voluntary movement compared to resting status. However, there was no difference in asymmetry of upper face between unilateral and bilateral injections. Based on our experience, bilateral injection in lower face is recommended, but not usually in upper face.

Table 1 The usual doses for commonly injected facial muscle in HFS patients [30]

	Frontalis	Corrugator	Orbicularis oculi	Zygomaticus major	Buccinator	Depressor angularis oris
Botox U	10	1	15–20	1	2	1
Dysport U	30	3	45–60	3	6	3
NeuroBloc/Myobloc U	500	50	1000	50	100	50

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Medical Treatment of Hemifacial Spasm and Other Involuntary Facial Movement Disorders

Wooyoung Jang and Jinyoung Youn

Involuntary movement of facial musculature encompasses various etiology such as hemifacial spasm, blepharospasm, facial myokymia, oromandibular dystonia, tardive dyskinesia, and psychogenic origin [1]. Before establishing the treatment strategy, physicians should be cautious about what etiology could be involved. Therefore, careful history taking including medication, co-morbidity as well as delicate description for characteristics of involuntary movement could be essential [2]. For example, unilateral presentation is more likely organic etiology than psychogenic origin. While hemifacial spasm has usually a peripherally derived movement by ipsilateral facial nerve, involvement of masticatory muscle or tongue could designate central origin [3]. However, differentiation for each etiology for facial involuntary movement is beyond the scope of this chapter and we review what kinds of medical option are available for each facial involuntary movement disorders.

Hemifacial Spasm

Hemifacial spasm (HFS) can be defined as a movement disorder showing involuntary synchronous tonic or clonic muscle contraction of facial muscles that is innervated by ipsilateral facial nerve. HFS is usually unilateral, but bilateral HFS was reported and essential blepharospasm, Meige syndrome, Tardive dyskinesia, facial tic and myokymia should be differentiated [1, 4, 5].

Blepharospasm usually manifests with bilateral, symmetrical contractions of the eyelids [6]. In HFS, famous description by Joseph Babinski that internal part of frontalis muscle contract, when orbicularis oculi contracts, so called, the other babinski sign could help to differentiate HFS from blepharospasm [7]. Oromandibular dystonia could be differentiated from HFS in the point of involuntary muscle contractions that involve primarily the lower part and structures of the face, the mouth, the tongue, and the pharynx [8]. Facial tic presents with more bizarre and multifocal involvement patterns and alternating propagation from side to side of the face [9]. Compared to other involuntary movement

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_14

disorders, the tics can typically be suppressible and the patients feel urge to move.

As a treatment, four kinds of options could be suggested. First, if patient does not complain about symptoms and not particularly bothered by HFS, let HFS go by with no treatment at all. In this case, clinicians should exclude structural lesions such as mass (except compression or indentation of intracranial artery of facial nerve root entry or exit zone) and reassure their patients as it is a benign condition [4]. Lee et al. reported 5-year follow-up data of 104 untreated HFS revealing that 38.5% of patients remained stationary, 9.6% of patients showed partial improvement, and 41.3% experienced complete remission [10]. However, in another series, 93.4% of HFS showed spreading of muscle spasm [11]. Therefore, non-treatment option should be carefully considered. As second and tertiary options, local injection of botulinum toxin and microvascular surgery are the treatment of choice for HFS based on evidence with high success rate and low complication rate [2, 12, 13]. However, there could be HFS patients who do not want these types of treatment options or who are not tolerable of botulinum toxin. In the aspect of oral medication, there are several options available for exceptional patients who do not fit botulinum toxin or surgical intervention. A variety of oral medications have been suggested for treatment of HFS, but almost in non-controlled studies. Therefore, evidence data is hardly lacking. Wang and Jankovic reported that only 8% with oral medication treatment revealed meaningful benefit [14]. Most of employed medication for treatment of HFS are anti-epileptic drugs, which are regarded as decreasing nerve excitability and can reduce the spasm and provide symptomatic benefit.

Carbamazapine

In 1982, one case series including three patients reported efficacy of carbamazepine with a dose of 600–1200 mg for HFS [15]. In this series, authors suggest that 50% of 46 patients, including their patients, showed improvement.

Baclofen

There is a single case report showing a 58-year-old woman with baclofen (daily dose of 37.5 mg) experienced a dramatic effect and remained asymptomatic during 12 months [16]. No side effect was reported in this report. However, there were no controlled studies or further evidence.

Clonazepam

In 1985, one non-controlled series was reported. Standard dosage of clonazepam ranges from 0.5 to 4 mg for daily dose [17].

Zonisamide

Siniscalchi et al. reported a single case regarding complete remission of HFS in a 65-year-old woman by adding zonisamide (150 mg twice a day, 6 weeks) followed by failure of clonazepam treatment [18]. There were no further studies regarding the effectiveness of zonisamide for HFS.

Gabapentin

There are three case series that gabapentin was reported to be effective for HFS [19–21]. Each series included 1, 5, 23 patients and show its efficacy from 600 to 2400 mg per day. Caravaglios et al.'s series including 23 patients demonstrated that 69.6% of subjects showed a significant clinical reduction, defined as reduction above 70%. Gabapentin is usually well tolerated, but clinicians should be cautious about its side effects such as somnolence and dizziness avoiding non-compliance. Because gabapentin showed its efficacy with a relatively high dose, it should be introduced at the lowest dose possible and gradually escalated. In Caravaglios's series, transient weakness and marked reduction of anxiety were presented as a side effect.

Pizotifen

Pizotifen, which is a 5HT receptor antagonist, was reported as showing improvement of HFS with continual efficacy in two patients [22]. The dose of pizotifen was 1 and 1.5 mg per day for respective patients.

Levetiracetam

Relatively levetiracetam was recently reported as an option of HFS treatment. There are three case series showing efficacy of levetiracetam for HFS [23–25]. First, in 2004, Deleu described two patients with HFS showing improvement with levetiracetam (1500 mg per day). In this series, both patients also underwent treatment with botulinum toxin type A, but showed only short-term relief and disabling side effect. Second, in 2005, Biagio Carrieri also reported complete remission of HFS with 2 weeks' levetiracetam treatment (500 mg per day). Finally, in 2016, Kuroda et al. suggested mechanism of levetiracetam for HFS as anti-kindling effect with two case reports showing dramatic improvement after levetiracetam introduction without significant adverse effect (500 mg per day). Therefore, till now, levetiracetam could be good candidate of oral medication for HFS treatment, warranting well designed clinical trials.

Blepharospasm and Other Involuntary Facial Movement Disorders

Blepharospasm

Essential blepharospasm is regarded as focal dystonia, which is characterized by forced eyelid closure due to dystonia of orbicularis oculi and other periorbital muscles [6]. Besides medical treatment, chemodenervation using botulinum toxin is a treatment of choice for blepharospasm. The American Academy of Neurology suggests Level B recommendation for onabotulinumtoxin-A and incobotulinum toxin-A and level C recommendation for abobotulinum toxin-A for treatment of blepharospasm [26].

There has been increasing evidence that deep brain stimulation could be effective for blepharospasm, although this beneficial effect is reported from studies including patients with blepharospasm combined with other types of dystonia. Santos et al. reported successful treatment of isolated blepharospasm by pallidal stimulation, and there is also a single case report for successful symptomatic relief after bilateral pallidal deep brain stimulation in intractable blepharospasm [27–29]. Further studies of DBS as treatment options of blepharospasm are strongly warranted. On the other hand, repetitive transcranial magnetic simulation (rTMS) shows a beneficial effect on blepharospasm. One randomized control study including 15 patients with low-frequency rTMS provides Class II evidence for blepharospasm in the aspects of safety and efficacy [30]. Recently, rTMS combined botulinum toxin injection revealed enhanced efficacy and prolongation of the effect of botulinum toxin treatment on blepharospasm [31].

Oral medication as a treatment of blepharospasm encompasses anticholinergics, GABAergic, antidopaminergics, and mexiletine. Owing to unwanted detrimental side effects and lack of evidence in clinical trials, oral medication has many limitations for symptomatic relief of blepharospasm.

At first, there is only one double-blinded crossover study for investigating the efficacy of anticholinergics such as trihexyphenidyl and benztropine for blepharospasm [32]. In this study, anticholinergic side effect was presented with high incidence such as confusion, sedation, and dry mouth, and in eight patients, there was no significant difference among peripheral, central anticholinergics and placebo. Baclofen and clonazepam have been reported in single case reports showing efficacy, and mexiletine was reported in case series including three patients showing meaningful efficacy [33–35]. Tetrabenazine, which is a presynaptic monoamine-depleting drug, has also been reported as a treatment option for blepharospasm in one double-blinded study [36]. Finally, apraclonidine, an alpha-2 adrenergic

agonist, was introduced revealing lid elevation through sympathetic activation of Muller muscle [37]. However, its therapeutic availability is known to be limited due to tachyphylaxis.

Other Facial Involuntary Movement Disorders

Meige's syndrome is a combination of two different forms of dystonia including blepharospasm and oromandibular dystonia [8]. The pathogenesis of Meige's syndrome is identical to other types of dystonia including abnormal plasticity and cortical disinhibition. Therefore, management of Meige's syndrome is also identical with treatment options of dystonia including blepharospasm. There are several clinical trials and case reports for botulinum toxin and pallidal deep brain stimulation as a treatment option for Meige's syndrome [38–40]. In the aspect of oral medication, anticholinergics, benzodiazepine, levodopa, haloperidol, tetrabenazine, and baclofen have been used for symptomatic relief. Marsden investigated many drugs in 39 subjects with Meige's syndrome, and found that anticholinergics and benzodiazepine were beneficial [41]. Taner et al. also reported beneficial effect of centrally acting anticholinergics in 13 patients with blepharospasm and oromandibular dystonia [42]. In another study, similar findings are reported and they also reported that lithium could be an alternative [43]. Jankovic and Ford suggested long-term effectiveness in 26% of tetrabenazine, 37% of trihexyphenidyl, and 26% of lithium [44]. Recently, levetiracetam and zolpidem monotherapy were reported being beneficial in some case reports [45, 46]. Facial myokymia is defined as involuntary contraction resulting in wavelike or vermicular propagation involving facial muscles, especially orbicularis oculi [47]. This phenomenon is characterized by electromyography findings, spontaneous muscle activity with different motor units and showing brief, repetitive discharges with rhythmic burst [47]. Usually, facial myokymia occurs in healthy subjects with no associated disease entity and most cases are transient and self-limiting [48, 49]. However, in some reports, facial myokymia with persistence and involvement of entire facial muscles could be suggested as ipsilateral pontine tegmentum lesion such as multiple sclerosis, timorous condition, and cysticercosis [50–53]. Therefore, atypical nature or long-standing facial myokymia should be regarded as potential structural lesion and cranial MRI could be considered.

Pharmacology of Botulinum Toxin

Botulinum neurotoxin is produced by Gram-positive anaerobic spore–forming bacteria called *Clostridium botulinum*. Botulinum toxin is divided into 7 serotype of neurotoxins from A to G. Since 1973, first application of treatment for strabismus, FDA approved its indication for hemifacial spasm or blepharospasm in 1989 and botulinum toxin is regarded as treatment of choice of hemifacial spasm [54, 55]. A detailed explanation of its application is beyond the scope of this chapter and the next chapter handles this issue. In this section, mechanism and basic pharmacology of botulism toxin will be discussed.

Mechanism

Botulinum toxin is produced by *Clostridium botulinum* in an anaerobic circumstance which is targeting intracellular substrate. The mechanism of botulinum toxin is divided into five major steps. (1) binding to membrane, (2) internalization or endocytosis, (3) low pH–driven membrane translocation, (4) secretion of the L chain in the cytosol, (5) cleavage of SNARE protein with following inhibition of neurotransmitter release and paralysis of targeted muscle [56, 57].

Synaptic vesicle is filled with neurotransmitters, including acetylcholine. A heavy chain of various types of botulinum toxin is binding to each receptors of ganglioside and translocated into cytosol. Subsequently, disulfide bond is cleaved between heavy chain and light chain and a light chain of each types of botulinum toxin

binds each targeted proteins within SNARE complex, which result in inhibiting the release of acetylcholine. For example, a light chain of botulinum toxin A, E cleaves synaptosome-associated protein (SNAP25), which is necessary for fusion of vesicles, while botulinum toxin B,D,F,G target vesicle-associated membrane protein (VAMP, synaptobrevin).

Pharmacology

Botulinum toxin has some unique characteristics of pharmacological activity [56]. First, the mechanism of action of botulinum toxin is derived by modification of single target protein within neuron. Second, there is still no available information of pharmacokinetics of botulinum toxin at the doses for treatment. Finally, botulinum toxin cannot be reversed of its action once the toxin has arrived within the neuron [58].

There are many preparations for clinical use, and almost are based on A1 serotype botulinum toxin, while only one type is based on B1 serotype [59]. Three major brands are commercially available using A1 type toxin, including onabotulinumtoxinA known as Botox by Allergan (Irvine, CA), abobotulinumtoxinA as Dysport by Ipsen (Paris, France), and icobutulinumtoxinA as Xeomin by Merz Pharmaceuticals (Frankfurt, Germany). Three other manufacturers based in South Korea and China also produce further serotype A botulism toxin, which are Meditoxin by Medy-Tox (Korea), Botulax by Hugel Inc. (Korea), and Prosigne by Lanzhou Institute for Biologic product (China). One serotype B1 toxin, Myobloc, using rimabotulinumtoxinB, is available. Table 1 demonstrates the characteristics of each product.

Botulinum toxins are freeze-dried or vacuum-dried preparation. However, all toxin products show the similar range of diffusion and efficacy from the site of injection, which is dependent on the amount [60]. Usually, greater doses of serotype B1 toxin are necessary to acquire a comparable effect of serotype A1 toxin and the duration of action is shorter in B1 toxin in skeletal muscle [61]. Usually, the comparable dose ratio of A1 and B1 toxin is 1:25–30 [61]. The potency of botulinum toxin preparation is estimated by units. 1U in Botulinum Toxin A1 correspond to 1 lethal dose (LD) for 3 days in mice [62]. In case of male human with 70 kg, LD50 of botulinum toxin A is 2500~3000 U. Dose conversion ratio between Botox and Dysport are reported as 1:2.5–3, while Botox and Xeomin are reported as having near-equivalence in potency [63, 64]. Recently, Yun et al. suggested that a 1:2.5 conversion ratio between Botox and Dysport could be appropriate in treatment of cervical dystonia [65].

Owing that botulinum neurotoxin and related protein could act like antigen, injection into patient could elicit antibody formation, which could interfere its biological activity [66]. However, the frequency of developing neutralizing antibody for serotype A1 is reported to be very low (0–3%) [67]. Whereas, for botulinum toxin B1, the frequency of neutralizing immune response is reported to be relatively high (10–44%) [68]. Common factors influencing the immunogenicity of botulinum toxin are produced related factors such as protein load, presence of inactive toxins and treatment associated factors including dose, frequency of injections, and sites of injection [69].

Table 1 Comparison of various representative commercial botulinum toxin products

Product	Botox	Dysport	Xeomin	Myobloc	Prosigne	Meditoxin
Company	Allergan (USA)	Ipsen(UK)	Merz(Germany)	Solstice Neurosciences (USA)	Lanzhou Institute of Biological Products(China)	Medy-Tox (Korea)
Unit	100 or 200 units	300 or 500 units	50, 100, 200 units	2500 units per 0.5 mL, 5000 units per 1 mL, 10,000 units per 2 mL	50, 100 units	50, 100, 150, 200 units
Serotype	A	A	A	B	A	A
Molecular weight	900 kDa	<900 kDa	150 kDa		900 kDa	940 kDa
Pharmaceutical form	Powder	Powder	Powder	Solution	Powder	Powder
Storage after dilution	24 h/2–8 °C	4 h/2–8 °C	24 h/2–8 °C	4 h (if diluted)/2–8 °C	4 h/2–8 °C	4 h/2–8 °C
Storage before dilution	−20~−5 °C (Freeze), 2~8 °C	2~8 °C	2~8 °C		2~8 °C	−20~−5 °C (Freeze), 2~8 °C
pH	7.4	7.4	7.4	5.6		6.8
Generic	Onabotulinumtoxina (ONA)	Abobotulinumtoxina (ABO)	Icobutulinumtoxina (INCO)	Rimabotulinumtoxinb (RIMA)	CBTX-A	BONTA
Conversion ratio	ONA:INCO = 1:1	ABO:ONA = 3:1 or even lower		ONA:RIMA = 1:50 ~100		

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Possible Complications of Microvascular Decompression

Doo-Sik Kong

Background Microvascular decompression (MVD) is a safe and effective treatment modality for the treatment of trigeminal neuralgia and hemifacial spasm. Postoperative complications may be either general or specific to the MVD and should be managed with care. In this chapter, we aimed to address possible complications during or following MVD. The possible complications associated with MVD include associated cranial deficits such as facial palsy and hearing loss, hemorrhagic complications, etc. Considering a rare incidence of the events, it cannot be overemphasized that care should be taken to avoid and reduce the major morbidities associated with MVD.

Microvascular decompression (MVD) has been recommended as a major safe and definitive treatment option for trigeminal neuralgia and hemifacial spasm. But in general, brain surgery itself poses a potential risk associated with vascular manipulation, especially around the cranial nerve. Operative morbidity and mortality are often defined as events within 30 days of operation.

Common complications during or after MVD include the postoperative facial paralysis, newly developed hearing deficits, cerebrospinal fluid leaks, cranial nervous deficits, and cerebrovascular events. The frequency of operative and postoperative complications naturally increases with the complexity of the procedure. In most cases, incidence of postoperative complications is between 1 and 7 days after the operation, specific complications occur throughout the postoperative period and in the late postoperative period. Among them, cerebrovascular complications are relatively rare, but once they occur, they can be fatal and are very significant complications. These complications associated with cerebral hemorrhage can leave very serious side effects, requiring more advanced techniques and standardized surgical procedures to prevent them from occurring. This study examines factors that can predict vulnerabilities to complications with MVD. There is a slight difference in the incidence of post-MVD complications between trigeminal neuralgia and hemifacial spasm. In our series over 4000 patients undergoing MVD, middle ear effusion (MEE), hearing impairment, dysgeusia, etc., were common adverse events post-MVD for trigeminal neuralgia. In contrast, after MVD for hemifacial spasm, facial nerve palsy, MEE, and hearing deficits [1] were more common adverse events. Surgeons have to know that morbidities associated with MVD is transient form or permanent form. As transient complications, temporary facial palsy, transient hearing deficit, and cerebrospinal fluid leak can occur. Permanent complications such as permanent hearing loss, complete facial palsy, and hemorrhagic/thromboembolic events, despite a

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_15

rare incidence, should be carefully avoided [2, 3]. In the case of transient complications, it is important to keep a close eye on the changes in the symptoms, which can be overcome wisely by reassuring the patient sufficiently while realizing that the symptoms may last forever.

Facial Palsy

Facial palsy is relatively common complication of MVD for hemifacial spasm, of which incidence has been reported to be 14.3–18.6% [2, 3]. According to the mode of onset, early-onset facial palsy occurs immediately after surgery within 24 h, where late-onset facial palsy usually occurs over a 24-h to 7-day period after the operation [4]. The exact pathogenesis of newly developed facial palsy after MVD is not known yet. Early-onset facial palsy seems to be associated with the inserted Teflon between offending vessels and facial nerve. Direct facial nerve injury by the compression of Teflon felt, delayed facial nerve edema by long-standing manipulation, or disturbance of microcirculation can explain the occurrence of facial palsy [2, 5, 6]. Because most cases of early-onset facial palsy were common in hemifacial spasms rather than trigeminal neuralgia, direct impairment of the facial nerve may be closely associated with early-onset facial palsy. In terms of surgical technique, Teflon-interpositional technique may contribute the occurrence of early-onset facial palsy after MVD more than arterial transpositional technique. Main cause of late-onset facial palsy remains unclear. The theory of viral origin may have plausible hypothesis [5–9]. The incidence of late-onset facial palsy following MVD is known to be 2.8–8.3% [5, 6, 10] and most of them could be improved spontaneously.

Most facial weakness can be resolved without special treatment. However, extensive rehabilitation can be required for excellent outcomes in some cases [11, 12]. Careful attention to the preoperative MR images, complete decompression of the facial nerve, less manipulation of the facial nerve, and intraoperative monitoring are the best ways to avoid this complication.

Hearing Impairment

Hearing loss is a relatively rare but significant risk of MVD. Hearing impairment is generally defined as increasing more than 15 dB depending on the bone conduction or by more than 20% of speech discrimination associated with the reference hearing. In most cases, hearing impairment after MVD is caused by an excessive retraction of the cerebellum. Several reasons can explain the postoperative hearing loss after MVD [3, 13–23]. Most plausible cause of hearing loss is attributable to the stretch of the eighth nerve during retracting the cerebellum. As a result, spasm of the vasa nervorum or ischemic injury to the nerve can develop. Taking into account the flocculus overlying the VII–VIII nerve complex, it is challenging to access directly with the dorsal direction and to observe the root exit area of the facial nerve. Thus, during the search for the root exit zone of the facial nerve, an excessive cerebellar retraction with inappropriate direction may lead to the postoperative hearing deficits. Our series showed the correlation between cerebellar retraction and BAEP changes.

Another possible cause includes the ischemic injury from the block of the labyrinthine artery and/or the anteroinferior cerebellar artery, resulting in the cochlear ischemic damages. In addition, direct trauma by coagulation or instruments, over-compression with Teflon felt interposed between the offending vessel and 7–8th nerve complex [24, 25]. Rarely, there happens a sudden decrease of brainstem auditory evoked potentials (BAEP) during or immediately after craniectomy. It may be attributable to an edema in the intracanalicular tract of the nerve, sudden change of intracranial pressure [26], or drill-induced noise or transient loss of CSF during surgery. There are some differences in the amplitude of BAEP under individual situation including recording conditions or electrode impedance [27].

In some cases, hearing impairment can show a gradual improvement when it is caused by ischemic injury due to local vasospasm. However, if hearing impairment is caused by a direct retraction injury, postoperative hearing

impairment remains persistent and constant [3, 13–23].

Intraoperative monitoring of BAEP during surgery is helpful to minimize the risk of hearing impairment in patients undergoing the cerebellopontine angle (CPA) surgery including MVD [28, 29]. The traction or compression of the nerve is closely associated with BAEP deterioration. These BAEP changes are caused by direct mechanical damage to the brainstem, vascular ischemia, or infarction. Our experience supported the high probability of hearing loss if the amplitude reduction and delay time of wave V during MVD surgery are fast and the changes are not recovered with modification [30]. In the literature, although there is no consensus regarding the criteria for significant intraoperative BAEP change, a latency prolongation of as little as 0.5 ms of the wave V or a reduction of amplitude more than 50% in wave V was a strong indicator of hearing impairment [23, 31, 32]. During surgery, BAEP monitoring plays an essential role in alarms for hearing preservation. Thus, we need to understand the surrounding surgical anatomy to minimize the risk of hearing loss. In addition, the minimum retraction of the cerebellum is of great importance, and can be helpful if the endoscope can provide a second look to identify nerve root entry areas and identify the location of the Teflon felt [9, 33–35]. Whether the hearing loss or impairment is identified intraoperatively or postoperatively, the rehabilitation protocol should be modified appropriately.

Lower Cranial Nerve Deficits

Clinical manifestations of lower cranial nerve deficits include hoarseness, dysphagia, and swallowing difficulty. In most cases, lower cranial deficits have a self-limiting course and are not fatal to the quality of life in patients. Despite a rare incidence, they should be carefully monitored, because their natural prognosis is sometimes poor. Considering that lower cranial nerve deficits are more common complications in hemifacial spasm than trigeminal neuralgia, manipulation of lower cranial nerves during the arachnoid dissection may be a major contributing factor. In review of our series, some patients can have various natures of dysgeusia (impairment of the sense of taste), continuous sour taste, and hypogeusia. In the literature, some authors suggested the hypothesis of this complication as injury to nervus intermedius or the trigeminal nerve can develop abnormal sensation of taste involving anterior two-thirds of the tongue and floor of the mouth and the palate [36, 37]. In general, the prognosis of dysgeusia seemed to be poor. Other cranial deficits include trochlear or abducens nerve palsy after MVD, vocal cord palsy occurs, resulting in asymmetric soft palate elevation or uvula deviation.

Hemorrhagic or Thromboembolic Complications

Hemorrhage or thromboembolic complications are relatively rare but serious complications of MVD. Hemorrhagic risk after MVD is similar to the risk inherent in the general cranial surgery. It must be the most serious and life-threatening complication. Cerebral infarction and dural arteriovenous fistula can be candidates of reasons of hemorrhagic risks. During operation, the sacrifice of the inferior petrosal vein does not always carry the risk of hemorrhagic events. Even though it must be rare, it is recommended that the inferior petrosal vein should be preserved. Managements for cerebrovascular events must be modified personally based on each situation. Sometimes, drainage of CSF during MVD may be associated with sudden slack-down of the cerebellum, leading to the tearing of bridging vein, resulting in subdural hematoma.

Conclusions

In conclusion, MVD is known to be a safe and effective treatment modality for hemifacial spasm and trigeminal neuralgia. Considering a rare incidence of the events, it should not be overemphasized that care should be taken to avoid and reduce the major morbidities associated with

MVD. It cannot be overemphasized that care should be taken to maintain the disciplined implementation of the surgical, such as proper direction and adequate retraction of cerebellum and preservation of arachnoid sheath over the CN VIII and CN VII complex, etc. Most of these complications can be prevented through standardized protocols to incorporate monitoring technologies and specific technical practices, team approach, and accumulating volume.

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Prognosis of Symptoms After Microvascular Decompression for Hemifacial Spasm

Jeong-A Lee and Kwan Park

Patients with hemifacial spasm (HFS) are more likely to have obsessive-compulsive disorders, anxiety, and sexual dysfunction [1–3]. The more severe the spasms, the worse are the quality of life (QoL), headache, and depression [4–6]. Microvascular decompression (MVD) has been recognized as an effective and reliable treatment for HFS [7–8]. The post-surgery improvements of the HFS symptoms were associated with decreased social anxiety and improved QoL [9]. However, the postoperative course of MVD is variable for each patient. The QoL improvements were delayed in operated patients due to such postoperative course [10]. The endpoint to confirm whether the surgical outcomes are successful is still unclear. The decision for reoperation is difficult. Surgeons should be careful when deciding indications and timing of reoperation [11–13]. Predicting outcomes as accurately/soon as possible is needed to minimize patient discomfort and anxiety by providing specific and practical information and to determine the proper timing for reoperation and reduce unnecessary reoperation. This chapter aims to investigate the predictors and the optimal prediction time of the surgical outcome through literature review.

Postoperative Course of Hemifacial Spasm

The postoperative course of MVD for HFS varies. In a systematic review for 12 articles with 2727 patients, the mean follow-up duration after surgery was 49 months (range 6.4–121.6 months). An average postoperative success rate was 85.1% with a range of 76.5–93.5%, but the immediate success rate after surgery is only 71.8% with a range of 59.5–84.0%. The mean rate of delayed improvement was 25.4% with a range of 18.8–37.1% [14]. Another review analyzed 82 publications counting more than 10,000 operating cases. The proportion of patients with total relief was between 85 and 90%. In many series, delayed relief was obtained in 33 ± 8% of patients and 12% of them were delayed for approximately 1 year. When the effect of MVD was considered to have been achieved, relief retained permanently, with the exception of 1–2% of the long-term followed patients [8].

The various postoperative courses of HFS were categorized into several patterns. This is an extreme representation of the postoperative course (Fig. 1). Patients were simply divided into two groups, with or without postoperative spasms [15]. Patients were divided into three groups

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_16

according to the postoperative course: immediate cure, delayed cure, and failure [16]. Different outcomes were divided into four groups depending on the variable recovery period: immediate cure, spasms lasted with milder degree and slowly disappeared from 7 days to 2 years, spasms stopped immediately but relapsed 3 days later and ran the same course as in the second group, and failure [17]. The five groups included the following: immediate cure without recurrence, temporary relapse followed by cure, slow but steady improvement that leads to cure after 1 month or more, recurrence with persistent symptoms, and no improvement or improvement to some extent that does not lead to cure [18]. On the other hand, disappearance of symptoms over time was classified into three groups: immediate disappearance of symptoms after surgery, delayed disappearance of symptoms, and unusual disappearance of symptoms [19].

This figure is a polarized representation of the remaining spasms after surgery compared to the preoperative spasms.

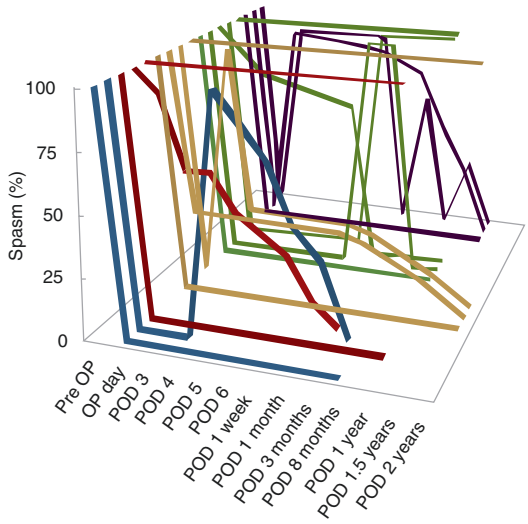

Fig. 1 Postoperative course of MVD for HFS. This figure is a polarized representation of the remaining spasms after surgery compared to the preoperative spasms

Predictors of Postoperative Outcome

English-language studies on MVD for HFS published since 2000 to the present have been retrieved from PubMed. Keywords of interest were prognosis, outcome, and result. Variables used in relation to the surgical outcome were arbitrarily classified into three factors: personal factors, diagnostic factors, and surgical factors (Fig. 2).

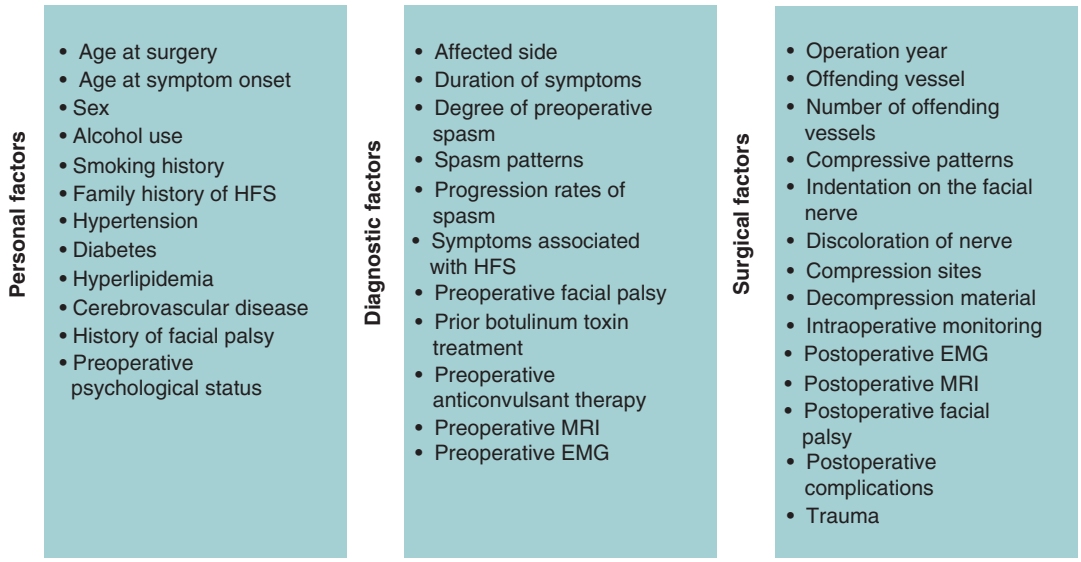

Fig. 2 Three factors used in relation to the surgical outcome. *EMG* electromyography, *HFS* hemifacial spasm, *MRI* magnetic resonance imaging

Personal Factors

Personal factors are potentially causative factors before diagnosis or surgery in patients with HFS, which are an individual-related trait that affects the postoperative course due to the general characteristics of the subject. These factors include the following: age at surgery, age at symptom onset, sex, alcohol use, smoking history, family history of HFS, hypertension, diabetes, hyperlipidemia, cerebrovascular disease, history of facial palsy, and preoperative psychological status. Although most variables were not related to the surgical outcome, there are a few studies that have described the association between age at surgery, sex, and the surgical outcome.

A few studies have shown that age at surgery considered being predictor, which significantly predicted the clinical outcome of patients following MVD [20–23]. The reason for this result is not clear. Engh et al. proposed that perhaps older patients who have failed MVD are more likely to have chronic, irreversible neuropathy of the facial nerve than younger patients [20]. Lv et al. presented that there is no doubt that (a) younger patients will have a relatively better operation condition for craniotomy, and (b) also indicate a relatively rapid recovery process than those of elderly patients. (c) With the extension of the duration of the disease and with aging, the incidence of microvascular complications may significantly increase accordingly, and hence associated with a relatively poor prognostic outcome [22].

Outcome of patients with HFS after MVD was predicted by sex [23–24]. Cheng et al. reported that female patients with HFS showed larger posterior fossa crowdedness and smaller posterior fossa cerebrospinal fluid (CSF) volume than male. These researchers suggested that posterior fossa crowdedness might increase the risk of developing HFS by causing congestion of the nervous and vascular structures [25]. Our study proposed that these results might be due to differences in hyperexcitability of facial motor nuclei in women and men [24].

Diagnostic Factors

Diagnostic factors are causative factors related to the disease or test result during the diagnosis of HFS, which are a disease-related trait that affects the postoperative course due to the characteristics of the disease. These factors include the following: affected side, duration of symptoms, characteristics of preoperative spasm (degree, patterns, and associated symptoms), preoperative facial palsy, prior botulinum toxin treatment, preoperative anticonvulsant therapy, preoperative magnetic resonance imaging (MRI), and preoperative electromyography (EMG). Although most variables were not related to the surgical outcome, there are a few studies that have described the association between duration of symptoms, prior botulinum toxin treatment, degree of preoperative spasm, and the surgical outcome.

Duration of symptom was the influential variable which may be useful for the prediction of prognosis in the patients who underwent MVD [22, 26]. Jin et al. proposed that delayed improvement is more likely to occur in patients with severe indentation of the facial nerve and/or longer duration of preoperative disease. The reasons they explained were as follows: (a) It could cause the pathological demyelinating changes in the root exit zone (REZ) or hyperexcitability of the facial motor nucleus; (b) MVD itself can lead to edema of the facial nerve, thus causing microinjury of the facial nerve and generating residual HFS; (c) Although the REZ is decompressed, it may still be compressed by pulsatile decompression materials and CSF, which may result in delayed resolution [26].

Regarding the relationship between prior botulinum toxin treatment and the surgical outcome, there have been different results. Ishikawa et al. suggested that the reason why this treatment produces good results is because it causes facial nerve paresis [15]. On the contrary, Zhao et al. explained that the possible reasons why this treatment produces bad results are as follows: (a) The neurotoxic effect of botulinum toxin may cause demyelination at different location in the same facial nerve; (b) Among patients, the incidence of

hypertension was high, which can result in torturing and prolongation of blood vessels [27].

Some researchers have found that the degree of spasms was an important classifier that can predict patients' outcomes [23, 24]. However, different classification based on the tendency [16] or severity and extent [28] of spasms did not show a significant association with the outcomes. Such discrepancies in the results might be due to inconsistent criteria with regard to the degree of preoperative spasms. In our previous study, the degree of preoperative spasms based on the grading system had significant correlation with the duration of preoperative symptoms and QoL [28]. Furthermore, the preoperative duration of symptoms and psychological status have previously been reported to significantly affect the postoperative outcome [26]. These results imply a close relationship between the degree of preoperative spasms and the postoperative outcome.

Surgical Factors

Surgical factors are the factors related to the surgical findings or surgical outcome of MVD in the HFS, which are a surgery-related trait that affects the postoperative course due to the surgical findings or surgical outcome. These factors include the following: operation year, offending vessel, number of offending vessels, compressive patterns, indentation on the facial nerve, discoloration of nerve, compression sites, decompression material, intraoperative monitoring (IOM), postoperative EMG, postoperative MRI, postoperative facial palsy, postoperative complications, and trauma. Many studies have described the association between indentation on the facial nerve, offending vessel, postoperative facial palsy, and the surgical outcome.

Regarding intraoperative indentation on the facial nerve, most previous studies have proven that severe indentation on the REZ is closely related to postoperative improvement [22, 24, 25, 29–31]. The improved results observed in the case of indentation on the nerve might be attributed to the fact that the indentation is evidence of definitive compression by an offending vessel. Thus, it is necessary to identify indentation on the facial nerve during the surgery. Whereas some researchers described the reason for negative results may be because it produces ischemia of the nerve and gradually causes nerve degeneration [32]. Also, it is explained that severe indentation of the REZ could lead to the pathological demyelinating changes of the REZ or hyperexcitability of the facial motor nucleus [26].

Previous studies have demonstrated that venous compression may be related to a worse prognosis, despite thorough decompression in HFS [31, 33, 34]. Wang et al. mentioned that a vein can play an important role and can be the offending vessel in MVD for HFS, although HFS caused by a vein is rare [35]. Toda et al. guided that the surgical issues regarding venous compression are its resolution and the distinction of venous compression from venous contact [34]. Therefore, we suggest that it is necessary to carefully explore offending vessels to prevent surgical failure.

Previous studies have shown that significantly better results were observed in terms of overall disappearance of HFS in the patients with delayed facial palsy than in patients without delayed facial palsy [24, 36]. Lee et al. proposed that the occurrence of delayed facial palsy is due to manipulations or the gradual development of postoperative edema [36]. Although the causes of delayed facial palsy after surgery are unknown, 6.5–14.5% of patients who received MVD experienced it, and most patients have recovered completely [36, 37]. Therefore, it is important to inform patients of the possibility of delayed facial palsy postoperatively and to improve psychological stability by reassuring patients that it can be completely resolved and become an advantageous prognostic factor of the postoperative spasm pattern.

Numerous studies have been published on neurophysiological monitoring during surgery. However, the effectiveness of IOM on the surgical outcome was the most controversial. It is not yet conclusive, but it may be helpful if it is well utilized.

Models

The models were developed by analyzing various factors in a multidimensional manner. A risk assessment model was first created consisting of significant preoperative variables (Model 1) and intraoperative variables demonstrated little additive value (Model 2). This model demonstrated predictive value for persistent or recurrent spasm (Table 1) [23].

We established a prediction model for determining MVD outcome in HFS patients using decision tree analysis technique, a data-mining method. This prediction model was based on six categories including four items (postoperative delayed facial palsy, degree of preoperative spasm, indentation on the facial nerve, and sex) (Fig. 3). All patients of the first category consisted of postoperative delayed facial palsy (yes) and the degree of preoperative spasm (grade I) showed improvement of spasm including delayed improvement. Meanwhile, 58.8% of the third category consisted of postoperative delayed facial palsy (no), indentation on the facial nerve (no or unknown), and the degree of preoperative spasm (grade IV) showed persistence of spasm [24].

Optimal Time for Outcome Prediction

It is necessary to find the earliest optimal time to determine the outcome of the surgery. The optimal time for the outcome prediction is divided as follows: POD 3 months, 6 months, 12 months, and 3 years.

Table 1 Risk scoring model of persistent HFS after MVD

Model		Variables			
		OR (95%CI)	*p*	AUC (95%CI)	*p*
Model 1		Age >50 + female gender + history of botulinum toxin use + platysma muscle involvement			
	Discharge	1.50 (1.03–2.18)	0.035	0.60 (0.50–0.70)	0.045
	Follow-up	3.01 (1.52–5.95)	0.002	0.75 (0.64–0.85)	0.001
Model 2		Age >50 + female gender + history of botulinum toxin use + platysma muscle involvement + LSR aberrancy after MVD + 2-stage surgery			
	Discharge	1.51 (1.16–1.97)	0.003	0.65 (0.56–0.75)	0.003
	Follow-up	1.61 (1.08–2.40)	0.019	0.67 (0.54–0.80)	0.023

LSR lateral spread response

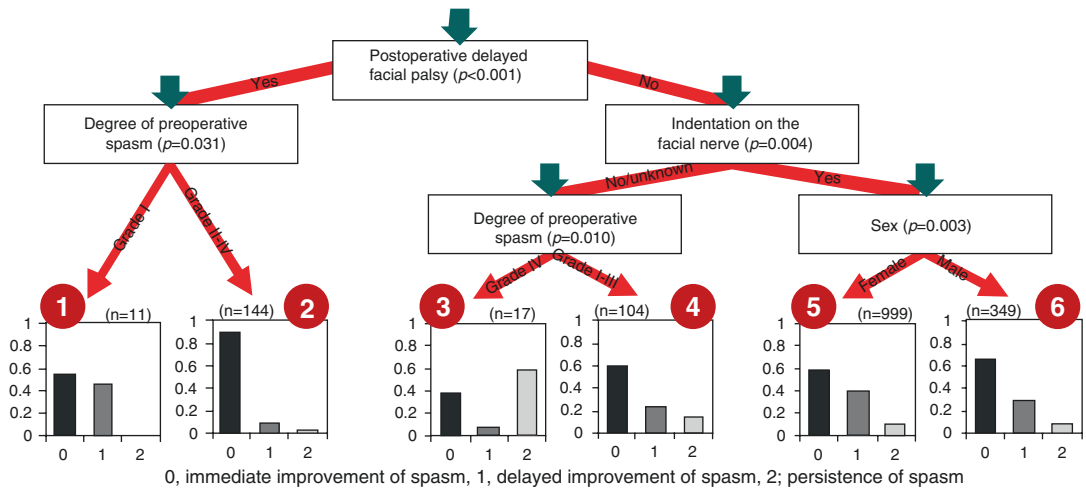

Fig. 3 Prediction model of outcome after MVD for HFS ($N = 1624$)

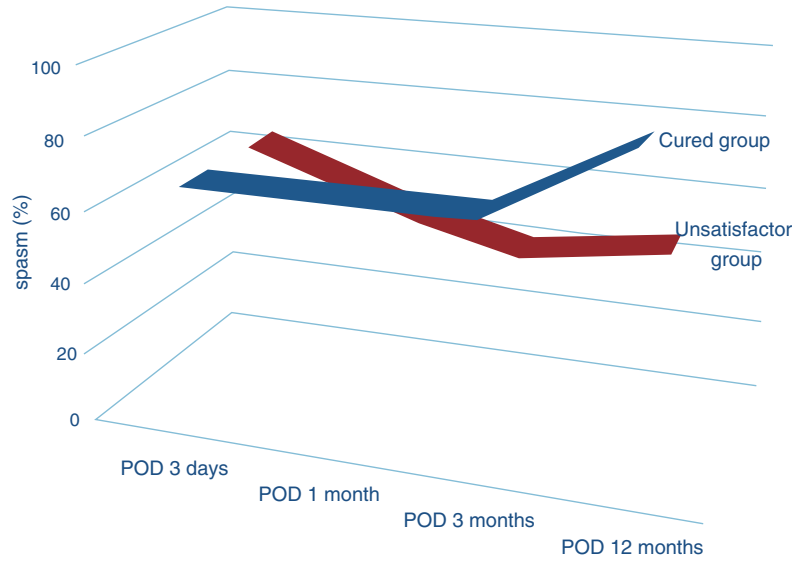

Fig. 4 Pattern of clinical improvement in cured and unsatisfactory group. Significant difference in symptom change between cured group and unsatisfactory group began to appear at 3 months postoperatively

POD 3 Months

In our previous studies, a chronological analysis of the changes in symptoms revealed that 3 months after surgery, symptoms of the cured group and the unsatisfactory group began to differentiate significantly (Fig. 4). This result implies that it might be possible to predict the outcome at least 3 months after surgery [18, 30].

Significant difference in symptom change between cured group and unsatisfactory group began to appear at 3 months postoperatively.

POD 6 Months

In our data, short-term and long-term outcomes of MVD in HFS patients were different, but the outcomes at 6 and 9 months were similar to those at 12 and more months (Fig. 5). Patients whose intraoperative offending vessel was not a vein, patients with intraoperative indentation on the facial nerve and patients with postoperative delayed facial palsy could predict good outcomes after 6 months of surgery [31].

The outcomes at 1, 3, 6, and 9 months were individually compared with the outcome after 12 months of surgery using the McNemar test with Bonferroni's correction.

POD 12 Months

Many authors commented that most cases were completely recovered within 1 year of observation and delayed resolution is uncommon after 1 year postoperatively. Therefore, even if MVD was expected to fail, it would be wise to wait 1 year before deciding on a reoperation [8, 11, 38]. Additionally, the patterns of postoperative outcome over time were divided into 10 groups. Of 267 patients in group 5–10, who showed persistence of spasm at the first month postoperatively (early non-responders), 198 patients (74.2%) in group 5, 7, 8, and 9 showed improvement of spasm over 12 months (late responders) and the spasms improved at 3 months in 89 patients in group 5 (Table 2) [31]. This result can support the above opinions.

POD 3 Years

In our study, 12 out of 70 patients (17.1%) who experienced residual or recurrent spasms for more than 1 year postoperatively gradually improved after 1 year to even 3 years (Fig. 6). If the lateral spread response is resolved and severe indentation on the nerve is identified during surgery, reoperation may be delayed up to 3 years after MVD [29].

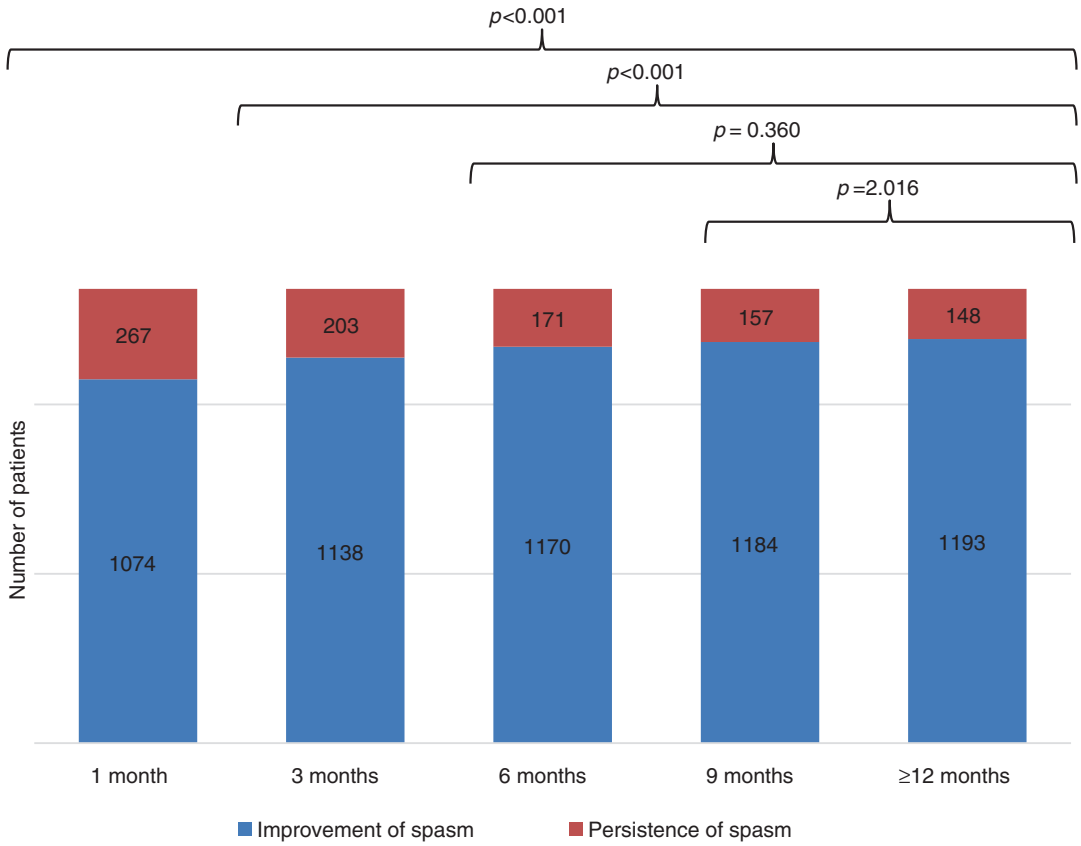

Fig. 5 Comparison of short-term and long-term outcomes (N = 1341). The outcomes at 1, 3, 6, and 9 months were individually compared with the outcome after 12 months of surgery using the McNemar test with Bonferroni's correction

Table 2 Patterns of postoperative outcome over time (N = 1341)

	Postoperative outcome over time					
Group	1 month	3 months	6 months	9 months	≥12 months	No. of patients
1	N	N	N	N	N	935
2	N	Y/N	Y/N	Y/N	N	60
3	N	N	N	N	Y	49
4	N	Y/N	Y/N	Y/N	Y	30
5	Y	N	N	N	N	89
6	Y	Y	Y	Y	Y	51
7	Y	Y	N	N	N	44
8	Y	Y	Y	Y	N	39
9	Y	Y/N	Y/N	Y/N	N	26
10	Y	Y/N	Y/N	Y/N	Y	18

N, improvement of spasm; Y, persistence of spasm; Y/N, improvement or persistence of spasm, with discontinuous patterns

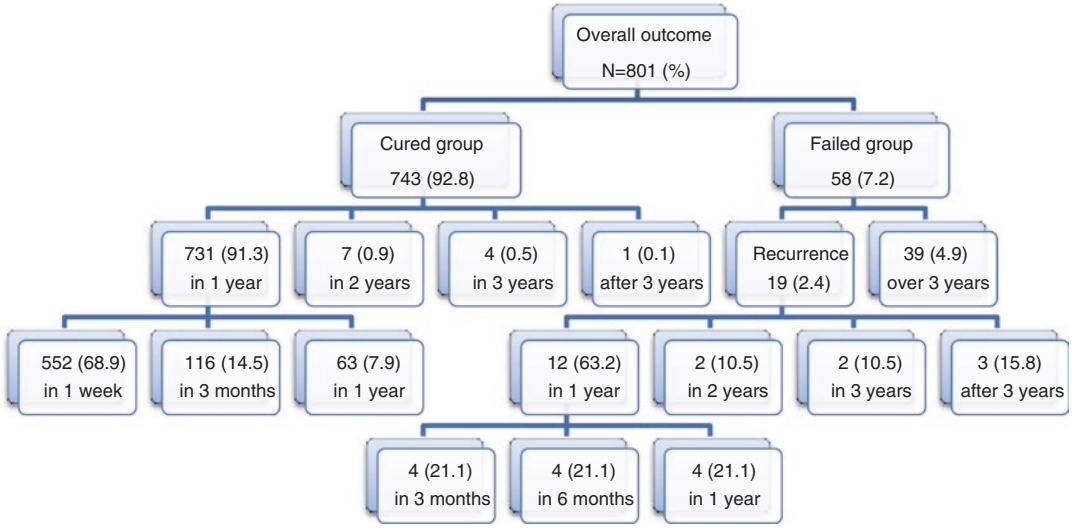

Fig. 6 Flowchart showing the postoperative course

Table 3 Summary of predictors and prediction time

			Prognosis	
			Better	Worse
Predictors	Weak	Pre-op	− Age at surgery Male − Symptom duration ± Botulinum toxin treatment − Spasm severity	+ Age at surgery Female + Symptom duration ± Botulinum toxin treatment + Spasm severity
	Moderate	Intra-op	Neurophysiologic findings	Neurophysiologic findings
	Strong	Intra-op	− Vein as offending vessel + Indentation on the nerve	+ Vein as offending vessel − Indentation on the nerve
		Post-op	+ Facial palsy	− Facial palsy
Prediction time	POD 3 months		− Spasm	+ Spasm
	POD 6 months		− Vein as offending vessel + Indentation on the nerve + Facial palsy	+ Vein as offending vessel − Indentation on the nerve − Facial palsy
	POD 12 months		− Spasm	+ Spasm

+/–, yes/no or increase/decrease

The above-mentioned points can be arbitrarily summarized in Table 3.

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Redo Surgery for Failed Microvascular Decompression for Hemifacial Spasm

Seunghoon Lee and Kwan Park

Failed Microvascular Decompression and Patient Selection for Redo Surgery

Sindou [1] reported successful results after microvascular decompression (MVD) in 87% of hemifacial spasm (HFS) patients. The recurrence rate was 2.4% and about 50% of patients who experience recurrent spasm proceeded to redo surgery from a meta-analysis comprising 5685 HFS patients from 22 reports [2]. What we can understand from these figures are (1) approximate 10% of the patients could not be spasm-free after MVD, (2) recurrence rate after MVD has been underreported so far, and the number of redo MVDs is much more limited. Therefore, we need to acknowledge that many patients after failed MVD are still suffering from spasm and a redo MVD can be considered for them.

Variable clinical responses are encountered after MVD and sometimes doctors as well as patients are confused about whether HFS recurs. For now, there is no established consensus on the observation period of variable responses after surgery or the timing of redo MVD. Sindou [1] and Hatayama [3] recommended at least 1 year of observation because no difference was observed in the MVD success rate between studies with a follow-up duration of less than 1 year and those with a follow-up duration of more than 1 year. Jiang [4] even advocated 2 years of observation before considering redo MVD. On the other hand, several other groups prefer early redo MVD. Engh [5] reported less favorable results in patients with late redo surgery and Zhong [6] showed a delayed spasm relief rate of <5% and recommended reoperation as early as possible. Liu [7] also compared results of early and late redo MVD and demonstrated the superiority of the early redo surgery. Our previously published data showed that 17.1% of patients with residual spasms at 1 year after MVD experienced gradual improvement, mostly within 3 years of MVD [8]. Therefore, the observation period required after MVD at our institution is at least 1 year and up to 3 years if the initial surgery was considered sufficient, with an indentation or discoloration of the facial nerve fully assessed, the offending vessel completely mobilized, and the facial nerve decompressed in the prior surgery. Referred patients, who had undergone surgery and usually spent several months postoperatively at other hospitals, were also followed up for at least 1 year and up to 3 years. However, earlier redo MVD was recommended in patients who did not undergo thorough exploration, had insufficient

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K. Park, J. S. Park (eds.), *Hemifacial Spasm*, https://doi.org/10.1007/978-981-15-5417-9_17

decompression during initial surgery, had no period of spasm relief or experienced worse spasm after the initial surgery, or had an evident neurovascular conflict revealed on MRI after the initial surgery.

However, there are many factors that make a decision to be reluctant in considering redo MVD. First of all, patient may not want to experience painful recovery time after surgery all over again. Many patients often complain of headache, dizziness, nausea, or vomiting at least couple of days after MVD. Neurosurgeons may be reluctant of any redo surgery, even more in surgery like MVD involving vulnerable cranial nerves (CNs) that are tangled with Teflon felt. Higher complication rates such as facial palsy or hearing loss can be easily anticipated. And magnetic resonance image (MRI) or electrophysiologic studies can hardly unveil the possible cause of failed MVD. Moreover, patient–doctor relationship has been somewhat compromised after the recurrence.

Intraoperative Considerations in Redo MVD: Surgical Guidance and Possible Causes of MVD Failure

Redo surgery is performed via the same route as the initial MVD. The cranial opening is usually larger than in the initial surgery, extending to the edge of the sigmoid sinus laterally and in the cephalic and caudal directions, too. Continuous intraoperative monitoring of brainstem auditory evoked potential (BAEP) and facial electromyography are performed to prevent CN dysfunction and to check abnormal muscle response (AMR). Due to postoperative adhesions, cautious arachnoid dissection should be guaranteed before cerebellar retraction to release the nerves and vessels from one another. An infrafloccular approach is used as usual to visualize the root exit zone (REZ) from the inferior aspects of the CN 7th–8th complex; however, if exploration is not available due to adhesions, another corridor is chosen from above the CN 7th–8th complex. The Teflon felt is withdrawn as much as needed to thoroughly inspect the neurovascular compression site, and we prefer not to remove all the Teflon felt. Because we have not experienced Teflon granuloma, and complete removal of the Teflon felt may injure surrounding tissue. The neurovascular compression site is explored from REZ to internal auditory canal (IAC) until the offending vessel that triggered the spasm, even a small arteriole or vein, is detected. A 360° view of the facial nerve, including the medial and lateral sides, should be inspected. When exploration is performed near the IAC, the labyrinthine artery should be preserved; iatrogenic injury and vasospasm often occur. A 360° and whole-segment decompression of the facial nerve is performed during redo MVD. Disappearance of AMR is monitored during surgery to determine appropriate decompression of the culprit vessel. The change of BAEPs is also monitored to determine the degree of surgical manipulation, such as cerebellar retraction. If the BAEP changes remain abnormal or a vasospasm occurs at the exposed vessel, intravenous steroids or topical papaverine irrigation can be applied.

Regarding the causes of MVD failure in HFS patients, Zhong [6] postulated that insufficient decompression and a missed offending vessel located distal to the REZ can be the reason. Some reports have identified the growth of Teflon granuloma as the cause of recurrence [9, 10]. Furthermore, de novo artery compression may cause recurrent HFS [11]. We published our evaluation on causes of initial MVD failures based on redo MVD cases [12]. Possible causes can be classified into three categories. A neglected offending vessel refers to a causative vessel that was overlooked and not decompressed during the initial MVD, even though there are traces of attempted exploration of the REZ and decompression of other nearby vessels. Insufficient decompression results from surgical difficulties despite locating the offending vessel. Offending vessels with multiple perforators, encircling the facial nerve, or located deep in the brainstem were the major causes of surgical difficulty and resulted in insufficient decompression. An untouched neurovascular compression site is an area with no traces of Teflon felt or arachnoid adhesion after previous surgical attempts near the REZ or neurovascular compression site; instead, Teflon felt was found at a location far from the REZ. And noticeable findings were as follows; a

Table 1 Possible causes of initial failure after exploration during redo MVD

Cause	Description
Neglected offending vessel Medially located artery or vein at the REZ at the cisternal segment	Traces of prior MVD at the REZ or Overlooked causative vessel at unusual location
Insufficient decompression	Traces of causative vessel manipulation Surgical difficulty
Untouched NVC site	No traces of prior attempts near the REZ or NVC sites

MVD microvascular decompression, *REZ* root exit zone, *NVC* neurovascular compression

vein was found to be the causative offending vessel, and cisternal segment or medial side of the facial nerve could be the location of compression. This is why a 360° and whole-segment inspection and decompression of the facial nerve are performed during redo MVD. Both a neglected offending vessel and insufficient decompression can cause MVD failure even in the hands of an experienced neurosurgeon, owing to the unusual location and surgical difficulty, whereas an untouched NVC site may be the result of inexperience (Table 1).

Clinical Outcomes of Redo MVD

In previous report by Jannetta [13], the spasm-free rate was inferior to the initial MVD at the postoperative 1-year follow-up. However, long-term spasm-free rate after redo MVD is similar to the rate after initial MVD, and complications such as hearing loss and immediate facial palsy occur more frequently after redo MVD than after initial MVD. Wang et al. [14] showed no significant difference in the relief rate (85.0 vs. 92.6%) and in the incidence of complications between repeat and first MVD. Bigder et al. [15] reported 91.7% of complete spasm resolution after reoperation for failed MVD without increasing complication rates. And most recently, three different Chinese groups reported their good outcomes after reoperations, ranging from 92.3 to 100%. However, their complications including facial palsy or hearing loss were higher than initial MVD, ranging from 7.7 to 24.1% [4, 16, 17]. From our registry, spasm-free rates of redo MVD were 81.0% at immediate post-redo MVD, 81.0% at 1 month, and 80.5% at 1 year. At the last follow-up, 90.5% were spasm-free. Hearing loss and facial palsy were observed in 14.3% and 38.1% patients, respectively, and were persistent in 9.5% and 9.5% of these patients, respectively, at the last follow-up. Two patients with permanent hearing loss remained in unserviceable status. Two patients who presented with House–Brackmann (H–B) grade IV facial palsy after MVD still had H–B grade III palsy at the last follow-up. In one patient, cerebellar infarction due to an injury to the cortical branch of the posterior inferior cerebellar artery during dura opening was noticed on routine postoperative CT. One patient developed hoarseness postoperatively, and vocal cord palsy was noted but improved without any further management. There was no infection, hemorrhage, or mortality [12].

Feasibility of Redo MVD and Future Directions of MVD

From our experiences, spasm-free rates of redo MVD at the follow-ups earlier than postoperative 1 month showed higher than those of initial MVD due to the higher occurrence of postoperative facial palsy and masking residual or improving spasm. As facial palsy improves at around 1 year after redo surgery, the spasm-free rate is decreasing, whereas the spasm-free rate after the initial MVD is increasing. And thereby, at the last follow-up, spasm-free rates in redo MVD is comparable to those after the initial MVD. Therefore, it can be concluded that the long-term spasm-free rate after redo MVD is similar to that after initial MVD despite the different course of clinical symptoms.

In terms of complications, more frequent changes in BAEPs or permanent hearing loss and more facial palsy indicate an inherent higher risk in redo MVD. Although the frequent changes in BAEPs do not necessarily lead to permanent

hearing loss, there is a high risk of nerve damage. Further manipulation of the facial nerve cannot be avoided and causes facial nerve damage in patients with an offending vessel located medial to or at the cisternal segment of the facial nerve. Moreover, overall hearing loss and immediate facial palsy occur more frequently after redo MVD than after initial MVD in our series as well as in other groups. Other severe complications, such as vascular complications or infection, can occur, although those do not develop significantly higher in redo MVD.

Based on prior studies including ours, redo MVD in HFS is a feasible treatment option for failed MVD in that spasm-free rates are comparable to those after the initial MVD. However, it is associated with higher risks of cranial nerve and vascular injuries. With the help of initial surgical records, images, and electrophysiologic data, patient selection as well as redo MVD surgery should be performed deliberately and carefully.

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