Microvascular Decompression Surgery

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Preface

Microvascular decompression (MVD) has been used to cure tens of thousands of patients since it was introduced in the 1960s. Now it is still the most effective treatment for cranial nerve syndrome caused by vascular compression, including primary trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia, refractory tinnitus, and dizziness. The cure rate for trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia varies among different centers, but it is over 90 % in most centers and can be as high as 98 %, even though some patients do not have good results, and some patients have recurrence. What's more, MVD has risks such as facial numbness, hearing loss or deafness, peripheral facial paralysis, dysphagia, and hoarseness. This is mostly because the mechanism by which vascular compression causes cranial nerve syndrome is still unknown. In addition, the real cause of ineffectiveness and recurrence is unknown. Therefore, study on the mechanism and refinement of our surgical techniques may help to improve the overall outcome.

As we reviewed the papers in the last 10 years, we found that many experts had been devoted to the study of mechanism, diagnostic criterion, surgical indications, operative techniques, electrophysiological monitoring, outcome evaluation, as well as treatment principles for ineffectiveness and recurrence. The results of these basic and clinical studies have further improved the theoretical bases for MVD, increased the reliability and safety, improved the cure rate, and lowered the incidence of surgery-related complications. The purpose of this book is to summarize these new theories, viewpoints, techniques, and principles, to help clinical practice and improve cure rate. It is exciting that we have invited some of the most famous experts on MVD, including Peter Jannetta, MD, PhD; Albert L. Rhoton, Jr., MD; Raymond F. Sekula, Jr, MD; Aage R. Moller, PhD; Marc P. Sindou, MD; Kwan Park, MD; Anthony M. Kaufmann, MD; and Akinori Kondo, MD. We hope that this book can show the frontiers of MVD and help professionals in this area.

In this book, we used literature review, description, classic case reports, figures, and expert comments to make it illustrated and easy to understand. However, due to the limitations of personal experience, omissions are inevitable. We look forward to your feedbacks and suggestions.

Shanghai, China — Shi-Ting Li

Contents

The History of Microvascular Decompression Surgery

1

Petter Jannatta

Neurosurgery is a very different surgical specialty from the days when this investigator was a research fellow and resident in the years 1957 through 1967. Technological advances have enabled most of these changes from an almost brutal surgical specialty with many poor results to a field wherein a normally sensitive human being can work happily and effectively. These advances, "the four factors," include application of the binocular dissection microscope to surgery, clinical neurophysiology, neuroradiology imaging, and neuroanesthesia. These advances were all necessary for the work to be described below. Two other important applications of technology but not used here include advances in stereotactic surgery (i.e., focused irradiation) and endovascular techniques. Utilization of these four factors has enabled us to see and do things hitherto thought impossible or at least highly dangerous.

Over the years, starting in 1965, neurosurgeons were able to define a number of cranial nerve diseases caused by pulsatile vascular compression of the centrally myelinated (one or two exceptions) cranial nerves (Jannetta 1977). These entities occur as the arteries of the base of the brain elongate and loop about with aging, so that they impact the nerves. The brain also moves caudally in the posterior fossa with aging contributing to arterial and venous pulsatile compression (Jannetta 1967, 1968, 1975, 1997; Rand and Jannetta 1968)

The best known of the cranial nerve vascular compression problems are trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia (Rand and Jannetta 1968). Other entities include Meniere's disease, vertigo, tinnitus (Jannetta 1975), and spasmodic torticollis (Jho and Jannetta 1995).

More recently, beginning in 1973 and first published in 1979, brain stem vascular compression was found to be associated with essential hypertension (Jannetta and Gendell 1979; Segal et al. 1979), type 2 diabetes (Jannetta and Hollihan 2004; Jannetta et al. 2010), and most recently Parkinson's disease (Jannetta et al. 2011). Other entities not yet published include cardiac arrhythmias (left and right heart), hypercholesterolemia and hyperlipidemias, and cerebellomedullary auto-compression syndrome ("hypoplastic posterior fossa" as described by Rosner). The latter is frequently but not always associated with medullary vascular compression.

We will not consider some of these entities in order:

1.1 Trigeminal Neuralgia

Early work by Dandy (1934) and Gardner (1962), concerning vascular compression and other abnormalities of the root entry zone of the

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trigeminal nerve, forms the basis for our current concepts of the etiology and definitive treatments of this disabling symptom called trigeminal neuralgia (TN). Dandy, starting in 1932, noted abnormalities of the dorsal root in a progressively larger percentage of his patients with TN. This innovative surgeon sectioned the portio major at the brain stem in these patients. He was able to treat TN without giving complete numbness by preserving fascicles which he described and called "accessory sensory fascicle" during the nerve section. In 5.6 % of patients where an extra-axial tumor was the cause of TN, he removed the lesion as treatment. He did not treat the TN by vascular compression. Indeed, it was impressive that he was able to see vascular compression by "normal" (although perhaps elongated) arteries and veins in 60 % of his patients without magnification. Gardner further elaborated upon abnormalities of the dorsal root of the trigeminal nerve and lucidly opined upon the pathophysiological mechanisms involved.

Others more recently described gross lesions such as aneurysms, tumors, and other abnormalities in the cerebellopontine angle of patients with TN. Despite these publications, there was little or no acceptance of this concept until recently. Many reasons for this lack of acceptance may be given. Some of them may include relatively primitive technology, lack of verification by others, inadequate documentation of findings, and rare definitive treatment. However, with the development of safer operative and anesthetic techniques for surgery of the cerebellopointine angle, the use of microsurgical techniques, photographic and videotape documentation findings and definitive treatment, and the concepts of root entry zone abnormality as an etiology of TN and microvascular decompression (MVD) as therapy have had wide acceptance in recent years.

The procedure MVD appears to be indicated in patients of any age with intractable tic douloureux who are in reasonably good health and who are not responding to medical therapy (Sekula et al. 2008, 2011). Almost all of our patients have had a course of carbamazepine which has been stopped for one reason or another. It is unfortunate that the elderly and frail patients appear to be more sensitive to the side effects and complications of this drug. The procedure is contraindicated in those who are in poor health, but the usual older person in generally good health appears to tolerate the procedure easily. A prior unsuccessful procedure or a recurrence is not a contraindication to operation.

The most common situation in lower facial TN is that the superior cerebellar artery is found coursing cephalad around the pons and then bifurcating, with the medial and lateral branches impinging upon the anterosuperior aspect of the entry zone of the nerve, the motor proprioceptive fascicle side, as it loops back to the brain stem and cerebellum. After sharp and blunt dissection of the widely opened arachnoid from the nerve and the visible part of the artery, the arterial loops are gently teased out from between the trigeminal nerve and the pons. The loops are usually longer in older patients and especially in those with long-standing tic douloureux. They must be manipulated carefully. The arterial loops may be quite adherent to the nerve or easily separable. Perforating branches to the pons have accommodated in length to the loop and will not tear with gentle manipulation of the vessels over the trigeminal nerve.

In the first division TN, a blood vessel is seen compressing the inferolateral portio of the trigeminal nerve entry zone at the pons. The most common cause of isolated V2 neuralgia is compression by the trigeminal vein coursing alongside the nerve. This may run parallel to the nerve.

Quality of survival is excellent in the vast majority of patients as they remain free of pain with no numbness or paresthesias to remind them of their prior syndrome. This author may be accused of bias regarding quality of survival with this procedure versus a destructive operation. Others, without a vested interest, have compared procedures and state strongly that MVD is the procedure of choice in TN.

1.1.1 Discussion

The concept of neurovascular compression as the common cause of trigeminal neuralgia has

become generally accepted in recent years. The early reports of Gardner (VF) have been well supported but with inadequate technology. Jannetta reported his experience with vascular findings in 1967 (Jannetta 1968, 1975), but at this time, he had performed only one MVD for TN (with Rand), although he had done a similar procedure for hemifacial spasm prior to this experience. The next verification of the vascular etiology and of the treatments by vascular decompression was by Petty in 1976, and others have continued to report their expanding series. Apfelbaum compared percutaneous radiofrequency lesioning (RFL) for trigeminal neuralgia with MVD and in 1977 (Jannetta 1975) published a large series where the quality of survival and results was clearly superior to the destructive procedure. It must be remembered that destructive procedures may be well indicated in many patients, the frail and elderly.

Vascular compression of the trigeminal nerve can now be clearly stated as causal of almost all cases of trigeminal neuralgia. As operative experience in relieving this compression grows, the results of operation should improve so that relief of the pain with little or no sequelae is a standard goal.

Glossopharyngeal neuralgia is similar to TN except that the pain is tonsillar, in ear, or within the throat. Both cranial nerve IX and X must be decompressed at the brain stem. Results are similar to TN (Resnick et al. 1995).

1.2 Hemifacial Spasm

Hemifacial spasm (HFS) is a symptom complex of hyperactive dysfunction of the facial nerve caused by abnormality at the root entry zone (REZ) of the nerve. This abnormality, almost always vascular cross-compression, most commonly occurs as a result of the aging process just as in other cranial nerves which are subject to hyperactive dysfunction. This problem is well known to the neurosurgical community (Jannetta 1968, 1977; Moussa et al. 2006; McLaughlin et al. 1999; Ruby and Jannetta 1975; Nielsen and Jannetta 1984; Moller and Jannetta 1985).

1.2.1 Clinical-Pathological Correlations

Two major points may be made regarding clinical findings with the pathological abnormality. First, the site of the vascular cross-compression is at or proximal to the REX (the Obersteiner-Redlich Zone) of the nerve. At this point, the myelin of the central nervous system provided by oligodendroglia is replaced by peripheral myelin provided by Schwann cells. By ultrastructural techniques, defects in the myelin are noted to be present here. Second, classical HFS is caused by a blood vessel on the anterior-caudal aspect of the nerve REZ. Atypical HFS is almost always caused by a blood vessel on the posterior-rostral aspect of the REZ. This compression may be venous rather than arterial (i.e., on the intramedullary facial nerve) and may be quite caudal on the medullary side of the pontomedullary junction.

Operative findings show that the abnormality is usually arterial. Multiple vessels, often lying parallel and with one pushing another into the REZ, are common. The posterior interior cerebellar artery is the most common causative vessel.

It can be noted that 93 % of patients have had an excellent result, although 12 patients needed a second operation before the vascular compression was relieved, usually because a blood vessel had been missed. The majority of those who have fair results (partially symptomatic <25 % of preoperative level) had prior destructive procedures consisting of nerve crush, partial section, or alcohol injection. Only five patients (2.2 %) have failures of therapy. It should be noted that hearing loss in the ipsilateral ear is the major risk of this procedure, and the patient must have full understanding of this risk before operation is performed. Our incidence of hearing loss is much lower than it was early on.

Gardner (1962) correlated the similarities in trigeminal neuralgia and hemifacial spasm. He did not inspect the root exit on one of the facial nerves in his hemifacial spasm patients. His treatment of hemifacial spasm consisted of application of mild trauma to the peripheral facial nerve or to the nerve in the cerebellopontine angle. Scoville successfully treated hemifacial spasm in one patient by moving a rather

peripheral artery from the facial nerve in the cerebellopontine angle. He, like Gardner, did not use the microscope, nor did he examine the nerve root entry zone in this case, but may have inadvertently decompressed the nerve proximally by moving a more peripheral part of the same looping artery, a situation we have seen a number of times. Recent investigators have shown that vascular cross-compression is the overwhelmingly common cause of HFS and that it can be treated definitively by MVD.

Mild facial weakness is common in hemifacial spasm of long duration, especially if the tonus phenomenon is present. Strength improves gradually after vascular decompression. Experience (Jannetta 1997) with surgical binocular microscope and a thorough knowledge of the normal anatomy of the cerebellopontine angle are vital if observations are to be valid and the procedure safe.

1.3 Vertigo and Tinnitus

The eighth (stato-acoustic) cranial nerve, like the trigeminal, facial, and glossopharyngeal nerves, is subject to symptoms of discorded hyperactivity which may be gradually accompanied by progressive loss of function (Jannetta 1977; Moller and Jannetta 1985; Sekula et al. 2006; Moller et al. 1986). The hyperactive symptoms in this special sensory nerve which carries hearing (cochlear nerve) and balance (superior and inferior vestibular nerves) functions consist of a caricature of those normal functions of the nerve, just as the hyperactive symptoms in a somatic sensory nerve include pain (i.e., trigeminal neuralgia, N V) and in a somatic motor nerve, abnormal muscular movements or twitching (hemifacial spasm, N VII). Patients may first have symptoms of cochlear and/or vestibular nerve involvement. Symptoms which may include tinnitus, hyperacusis, diplacusis, hearing loss, and vertigo may vary in time and intensity. Symptoms may develop in one branch of the eighth cranial nerve and then gradually involve the other. Operative treatment over the years has generally consisted of peripheral denervation. This has indeed been the best available treatment for patients with intractable symptoms. The results regarding quality of life and persistence of relief have not been excellent. Replacing one symptom with another has not proven to be a superior method of treatment.

The concept is based upon continued extrapolation of the findings in trigeminal neuralgia and hemifacial spasm. It seemed reasonable that the same should be true in the eighth cranial nerve. Therefore, starting in December 1972, a series of patients underwent microsurgical exploration of the nerve throughout its intracranial course in the cerebellopontine angle (Moller et al. 1993a, b, 1994).

The operative technique is basically the same as that performed for hemifacial spasm with the exception that the root entry zone of the eighth nerve must be gently dissected free from its caudal contact with the flocculus of the cerebellum more medially than in the hemifacial spasm operation if one is to see clearly the veins at the brain stem which may be causal of the symptoms.

We have progressively better success as our experience has grown. Relief of vertigo is more satisfactory than that of tinnitus. Severe tinnitus of over 2-year duration has a poor prognosis. The use of shredded Teflon has improved the results greatly. The postoperative course is variable, but of interest is the fact that improvement in both symptoms and in testing of function is gradual postoperatively, often taking weeks to months for maximum improvement.

The procedure of MVD appears to be definitive and to reverse both disordered hyperfunction and hypofunction. The extensive studies of changes in cranial nerves V, VII, IX, and X are being carried out (Resnick et al. 1995) and the spinal accessory nerves in spasmodic torticollis (Jho and Jannetta 1995).

1.3.1 Loss of Function Syndromes

It has been shown that Bell's Palsy is commonly due to a shifted arterial loop, usually AICA, stretching the facial nerve in the cerebellopontine angle (Jannetta and Bissonette 1978; Jannetta and Robbins 1980). Other cranial nerves are similarly affected (Jannetta and Robbins 1980) .

1.4 Brain Stem Compression

1.4.1 Essential Hypertension

In 1973 the initial observations were made on anterolateral medullary and adjacent left vagus nerve arterial compression in two patients. The first who had what was thought to be mild hypertension suffered a lethal stroke of the right forebrain after uneventful MVD of the right glossopharyngeal complex for glossopharyngeal neuralgia (GPN). The second patient with normal preoperative BP became severely hypertensive for over 2 weeks after MVD of the left IX–X nerves for GPH (Jannetta and Gendell 1979; Segal et al. 1979). The relationships of the medulla in cadavers and patients who were normotensive and hypertensive and who were operated upon for other problems were then studied. Hypertensives had arterial compression of the left medulla. A prospective controlled study of MVD of the left medulla in hypertensives was performed. The MVD gave prolonged relief of the hypertension, but as in spasmodic torticollis, the improvement was slow, sometimes taking up to a year (Levy et al. 1998, 2001, Jannetta et al. 1998). This work has been verified by a number of German investigators, by the Japanese, and by several Americans.

Essential hypertension was associated almost universally with left medullary compression. The exceptions were a few patients with severe distortion of the medulla due to one or more huge arteries compressing the right side. It became obvious that a right-sided syndrome must exist. Study of the anatomy physiology and distortion of the right vagus nerve led us to select type 2 diabetes (DM2) as a likely problem.

1.4.2 Type 2 Diabetes

Type 2 diabetes. 95% of diabetes mellitus is associated with arterial compression of the right medulla oblongata and adjacent vagus nerve resulting in hyperactive dysfunction presumably by causing increased symptomatic tone in the distribution of the right vagus nerve. Problems develop in the patient only after small arterial abnormalities and occlusions develop. DM2 thus appears to be primarily an arterial deterioration syndrome. Under certain circumstances, progression of type 2 DM can be relieved and in some cases reversed by removing the arterial compression. Our patients all had advanced disease (Levy et al. 2001)

Treatment (MVD) in DM2 (17 subjects). Six were failures with one unchanged preoperatively. In the operative group (11 subjects), four had significant improvement in A_1c, five were unchanged, and two had progressive disease. Only one subject was improved in a group of eight with a BMIO over 32. Two abject failures and three unchanged subjects were in the high BMI group. In the high BMI group, of nine subjects, only one improved with operation. None of the three control subjects improved or remained the same. In the hypertensive group, 24 operative subjects improved, seven were unchanged or failures, seven had progressive disease, and five were unchanged (one needing increased medication). One of the "unchanged" subjects in each group actually had normal blood pressure both pre- and posttreatment.

Extended follow-up. Extended follow-up of ten patients in the DM2 study for 3–8 years showed that the HbA_1c was maintained for this period. Once the new notable improvements developed, they persisted.

Conclusion. Long-term follow-up of subjects treated by MVD and those given best medical therapy showed that operation is helpful, especially more so in hypertension than DM2. Further operative studies are needed especially in DM where early patients without increase in blood glucose should be studied. Operation before pancreatic failure may be more effective. This prospective controlled study must be performed.

1.4.3 Parkinson's Disease (PD)

It has been shown in one PD patient that arterial compression of the anterolateral cerebral peduncle, the region over the substantia nigra, was found and treated. An MRI study of 20 PD patients and two age- and sex-matched controls showed significant midbrain compression in the

PD group (Dunn and Jannetta 1973). The pertinent MRI is an axial T2 study which clearly shows the compression artery (posterior cerebral) and the cerebrospinal fluid. MRI will be useful in patients with PD as they may be amenable to MVD of the midbrain (cerebral peduncle).

In this brief introductory review, we have attempted to bring to the reader some early background material in preparation for the body of contemporary treatises. Here, accomplished investigators will present up-to-date descriptions of the cerebellopontine angle, abnormalities of structures therein, and detailed treatment modalities of the cranial nerve abnormalities.

References

Dandy WE. Concerning cause of trigeminal neuralgia. Am J Surg. 1934;24:447–55.

Dunn DK, Jannetta PJ. Evaluation of chronic pain in children. Curr Prob Surg. 1973:64–72.

Gardner WJ. Concerning the mechanism of trigeminal neuralgia and hemifacial spasm. J Neurosurg. 1962; 19:947–58.

Jannetta PJ. Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neuralgia. J Neurosurg. 1967;107(1):216–9.

Jannetta PJ. The surgical binocular microscope in neurological surgery. Am Surg. 1968;34(1):31–4.

Jannetta PJ. Neurovascular cross-compression in patients with hyperactive dysfunction symptoms of the eighth cranial nerve. Surg Forum. 1975;26:467–9.

Jannetta PJ. Observations on the etiology of trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction and glossopharyngeal neuralgia. Definitive microsurgical treatment and results in 117 patients. Neurochirurgia. 1977;20(5):145–54.

Jannetta PJ. Outcome after microvascular decompression for typical trigeminal neuralgia, hemifacial spasm, tinnitus, disabling positional vertigo, and glossopharyngeal neuralgia (honored guest lecture). Clin Neurosurg. 1997;44:331–83.

Jannetta PJ, Bissonette DJ. Bell's palsy: a theory as to etiology. Observations in six patients. Laryngoscope. 1978;88(5):849–54.

Jannetta PJ, Gendell HM. Clinical observations on etiology of essential hypertension. Surg Forum. 1979;30: 431–2.

Jannetta PJ, Hollihan L. Type 2 diabetes mellitus, etiology and possible treatment: preliminary report. Surg Neurol. 2004;61(5):422–6; discussion 426–8.

Jannetta PJ, Robbins LJ. Trigeminal neuropathy – new observations. Neurosurgery. 1980;7(4):347–51.

Jannetta PJ, Abbasy M, Maroon JC, et al. Etiology and definitive microsurgical treatment of hemifacial spasm. Operative techniques and results in 47 patients. J Neurosurg. 1977;47(3):321–8.

Jannetta PJ, Levy EI, Clyde B, et al. Medullary compression and hypertension. J Neurosurg. 1998;89(1):169–70.

Jannetta PJ, Fletcher LH, Grondziowski PM, et al. Type 2 diabetes mellitus: a central nervous system etiology. Surg Neurol Int. 2010:16;1.

Jannetta PJ, Whiting DM, Fletcher LH, et al. Parkinson's disease: an inquiry into the etiology and treatment. Neurol Int. 2011;3(2):e7.

Jho HD, Jannetta PJ. Microvascular decompression for spasmodic torticollis. Acta Neurochir. 1995;134(1-2): 21–6.

Levy EI, Clyde B, McLaughlin MR, et al. Microvascular decompression of the left lateral medulla oblongata for severe refractory neurogenic hypertension. Neurosurgery. 1998;43(1):1–6; discussion 6–9.

Levy EI, Scarrow AM, Jannetta PJ. Microvascular decompression in the treatment of hypertension: review and update. Surg Neurol. 2001;55(1):2–10; discussion 10–11.

McLaughlin MR, Jannetta PJ, Clyde BL, et al. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999; 90(1):1–8.

Moller AR, Jannetta PJ. Hemifacial spasm: results of electrophysiologic recording during microvascular decompression operations. Neurology. 1985;35(7): 969–74.

Moller MB, Moller AR, Jannetta PJ, et al. Diagnosis and surgical treatment of disabling positional vertigo. J Neurosurg. 1986;64(1):21–8.

Moller MB, Moller AR, Jannetta PJ, et al. Vascular decompression surgery for severe tinnitus: selection criteria and results. Laryngoscope. 1993a;103(4 Pt 1): 421–7.

Moller MB, Moller AR, Jannetta PJ, et al. Microvascular decompression of the eighth nerve in patients with disabling positional vertigo: selection criteria and operative results in 207 patients. Acta Neurochir. 1993b; 125(1-4):75–82.

Moller AR, Jannetta PJ, Jho HD. Click-evoked responses from the cochlear nucleus: a study in human. Electroencephalogr Clin Neurophysiol. 1994;92(3): 215–24.

Moussa R, Harfouche D, Samaha E, et al. Microsurgical facial nerve decompression for hemifacial spasm. J Med Liban. 2006;54(3):146–51. The Lebanese medical journal.

Nielsen VK, Jannetta PJ. Pathophysiology of hemifacial spasm: III. Effects of facial nerve decompression. Neurology. 1984;34(7):891–7.

Rand RW, Jannetta PJ. Microneurosurgery: application of the binocular surgical microscope in brain tumors,

intracranial aneurysms, spinal cord disease, and nerve reconstruction. Clin Neurosurg. 1968;15:319–42.

Resnick DK, Jannetta PJ, Bissonnette D, et al. Microvascular decompression for glossopharyngeal neuralgia. Neurosurgery. 1995;36(1):64–8; discussion 68–9.

Ruby JR, Jannetta PJ. Hemifacial spasm: ultrastructural changes in the facial nerve induced by neurovascular compression. Surg Neurol. 1975;4(4):369–70.

Segal R, Gendell HM, Canfield D, et al. Cardiovascular response to pulsatile pressure applied to ventrolateral medulla. Surg Forum. 1979;30:433–5.

Sekula Jr RF, Marchan EM, Baghai P, et al. Central brain herniation secondary to fulminant acute disseminated encephalomyelitis: implications for neurosurgical management. Case report. J Neurosurg. 2006;105(3):472–4.

Sekula RF, Marchan EM, Fletcher LH, et al. Microvascular decompression for trigeminal neuralgia in elderly patients. J Neurosurg. 2008;108(4):689–91.

Sekula Jr RF, Frederickson AM, Jannetta PJ, et al. Microvascular decompression for elderly patients with trigeminal neuralgia: a prospective study and systematic review with meta-analysis. J Neurosurg. 2011;114(1):172–9.

2 Microsurgical Anatomy for Microvascular Decompression Surgery

Ken Matsushima, Xiaochun Jiang, and Albert L. Rhoton Jr.

Abstract

Microvascular decompression surgery is an operation in which knowledge of normal anatomy is especially important because there are often no mass lesions in the cerebellopontine angle (CPA). There are three neurovascular complexes in the CPA, as described in the "rule of 3" (Hitotsumatsu et al. Neurosurgery 53:1436–1441, 2003). The three neurovascular complexes are associated with the three major neurovascular compression syndromes: the upper neurovascular complex including cranial nerve (CN) V and the superior cerebellar artery (SCA) is related to trigeminal neuralgia, the middle neurovascular complex including CNs VII and VIII and the anterior inferior cerebellar artery (AICA) is related to hemifacial spasm, and the lower neurovascular complex including CNs IX–XII and the posterior inferior cerebellar artery (PICA) is related to glossopharyngeal neuralgia. In this chapter, the microsurgical anatomy of the posterior fossa encountered in microvascular decompression surgery is introduced with the rule of 3.

Keywords

Cerebellopontine angle • Glossopharyngeal neuralgia • Hemifacial spasm • Microsurgical anatomy • Microvascular decompression • Retrosigmoid approach • Trigeminal neuralgia

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Abbreviations

AICA	Anterior inferior cerebellar artery
CN	Cranial nerve
CPA	Cerebellopontine angle
PICA	Posterior inferior cerebellar artery
SCA	Superior cerebellar artery

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2.1 Rule of 3

The structures within the posterior cranial fossa, including the brainstem, cerebellar surfaces, cerebellar peduncles, cerebellar-brainstem fissures, and cerebellar arteries and veins, are organized into three neurovascular complexes, each associated with a major neurovascular compression syndrome (Fig. 2.1) (Hardy et al. 1980; Hitotsumatsu et al. 2003; Lister et al. 1982; Martin et al. 1980; Matsushima et al. 1982, 1983; Rhoton 2000b). The upper neurovascular complex including cranial nerve (CN) V and the superior cerebellar artery (SCA) is related to trigeminal neuralgia, the middle neurovascular complex including CNs VII and VIII and the anterior inferior cerebellar artery (AICA) is related to hemifacial spasm, and the lower neurovascular complex including CNs IX–XII and the posterior inferior cerebellar artery (PICA) is related to glossopharyngeal neuralgia.

2.2 Upper Neurovascular Complex and Trigeminal Neuralgia

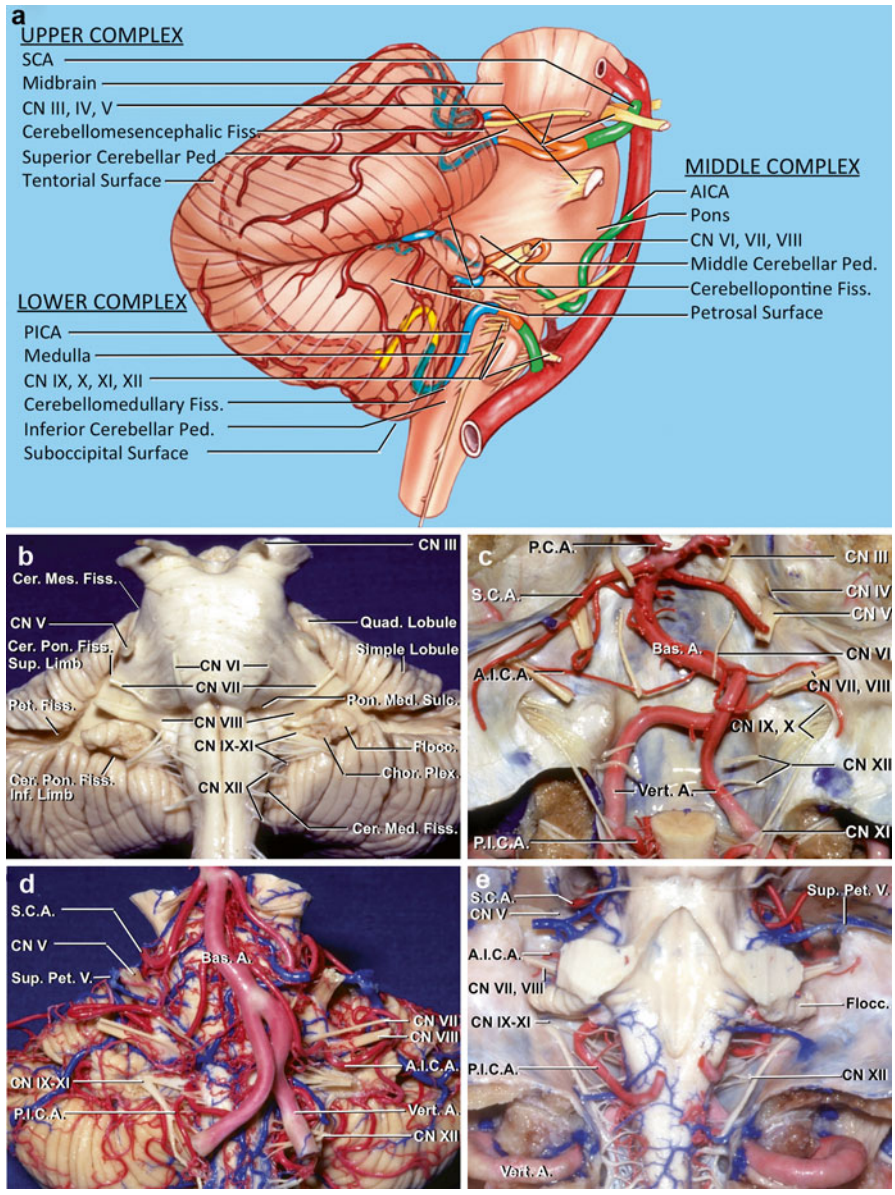
The upper neurovascular complex includes the SCA, midbrain, cerebellomesencephalic fissure, superior cerebellar peduncle, tentorial surface of the cerebellum, and CNs III–V. The SCA arises in front of the midbrain and passes below CNs III and

Fig. 2.1 Three neurovascular complexes. (**a**) The upper complex includes the SCA, midbrain, superior cerebellar peduncle, cerebellomesencephalic fissure, tentorial cerebellar surface, and CNs III–V. The middle complex includes the AICA, pons, middle cerebellar peduncle, cerebellopontine fissure, petrosal surface, and CNs VI–VIII. The lower complex includes the PICA, medulla, inferior cerebellar peduncle, cerebellomedullary fissure, suboccipital surface, and CNs IX–XII. The SCA is divided into four segments: anterior pontomesencephalic (*green*), lateral pontomesencephalic (*orange*), cerebellomesencephalic (*blue*), and cortical (*red*). Each segment may be composed of one or more trunks, depending on the level of bifurcation of the main trunk. The AICA is divided into four segments: anterior pontine (*green*), lateral pontomedullary (*orange*), flocculonodular (*blue*), and cortical (*red*). The PICA is divided into five segments: anterior medullary (*green*), lateral medullary (*orange*), tonsillomedullary (*blue*), telovelotonsillar (*yellow*), and cortical (*red*) (Reprint with permission from Rhoton (2000a)). (**b**) The cerebellopontine fissures are V-shaped fissures formed where the cerebellum wraps around the pons and the middle cerebellar peduncles. These fissures have a superior and an inferior limb, which meet at a lateral apex. The cerebellopontine fissure is continuous with the cerebellomesencephalic fissure superiorly and cerebellomedullary fissure inferiorly. The petrosal fissure extends laterally from the apex of the cerebellopontine fissures. CN V arises from the lateral part of the pons near the point where the superior limb of the cerebellopontine fissure meets the cerebellomesencephalic fissure. CNs VII and VIII arise at the lateral end of the pontomedullary sulcus immediately rostral to the foramen of Luschka and choroid plexus and ventral to the flocculus. CNs IX–XI arise as a line of rootlets that exit the medulla along the posterior edge of the olive in the postolivary sulcus, ventral to the lateral edge of the cerebellomedullary fissure (Reprint with permission from Rhoton (2000c)). (**c**) Posterior view of the cranial base with CNs and arteries preserved. CN V enters into Meckel's cave, CNs VII and VIII enter into the internal acoustic meatus, and CNs IX–XI enter into the jugular foramen. The SCA arises from the basilar artery near the apex at the midbrain level and passes below CNs III and IV and above CN V. The AICA originates from the basilar artery and encircles the pons near CNs VI–VIII. In most cases, the AICA passes below CNs VII and VIII, as seen in this specimen, but it may also pass above or between these nerves. The PICA arises from the vertebral artery at the upper medullary level and passes posteriorly around the medulla, coursing rostral to or between CNs IX and XI. There is slight asymmetry in the level of origin of the AICAs and marked asymmetry in the level of the origin of the PICAs, as is common (Reprint with permission From Rhoton (2000c)). (**d**) Both SCAs arise anterior to the midbrain and loop downward to the level of the junction of CN V with the pons. The right AICA reaches the superior surface of CN VII at its exit, and the left AICA reaches the inferior surface of CN VII at its brainstem exit. The AICA reapproaches CNs VII and VIII near the internal acoustic meatus where it sends the nerve-related branches. The right PICA arises from the vertebral artery at the medullary level and loops upward toward CNs IX and X (Reprint with permission from Rhoton (2013)). (**e**) The cerebellum has been removed to show the fourth ventricle, three cerebellar arteries, and CNs V and VII–XII. The fourth ventricle sits on the posterior surface of the pons and medulla. The flocculus projects laterally into the CPA (Reprint with permission from Rhoton (2007)). *A* artery, *A.I.C.A.* anterior inferior cerebellar artery, *Bas*. basilar, *Cer*. cerebello, *Chor. Plex*. choroid plexus, *CN* cranial nerve, *Fiss*. fissure, *Flocc*. flocculus, *Inf*. inferior, *Med*. medullary, *Mes*. mesencephalic, *P.C.A.* posterior cerebral artery, *P.I.C.A.* posterior inferior cerebellar artery, *Ped*. peduncle, *Pet*. petrosal, *Pon*. pontine, ponto, *Quad*. quadrangular, *S.C.A.* superior cerebral artery, *Sulc*. sulcus, *Sup*. superior, *V*. vein, *Vert*. vertebral

IV and above CN V to reach the cerebellomesencephalic fissure, where it runs on the superior cerebellar peduncle and terminates by supplying the tentorial surface of the cerebellum (Rhoton 2013).

2.2.1 Anatomy of CN V

CN V arises from the lateral part of the pons and runs obliquely upward toward the petrous apex (Fig. 2.2). It exits the posterior fossa to enter the middle cranial fossa by passing forward beneath the tentorial attachment and through Meckel's cave, which sits in the trigeminal impression on the upper surface of the petrous part of the temporal bone (Fig. 2.2a). Throughout the interval from the ganglion to the junction with the pons, the third-division fibers remain in a caudolateral position, second-division fibers in an intermediate position, and first-division fibers in a rostromedial position (Fig. 2.2c) (Emmons and Rhoton 1971). There are anastomoses between the fibers from



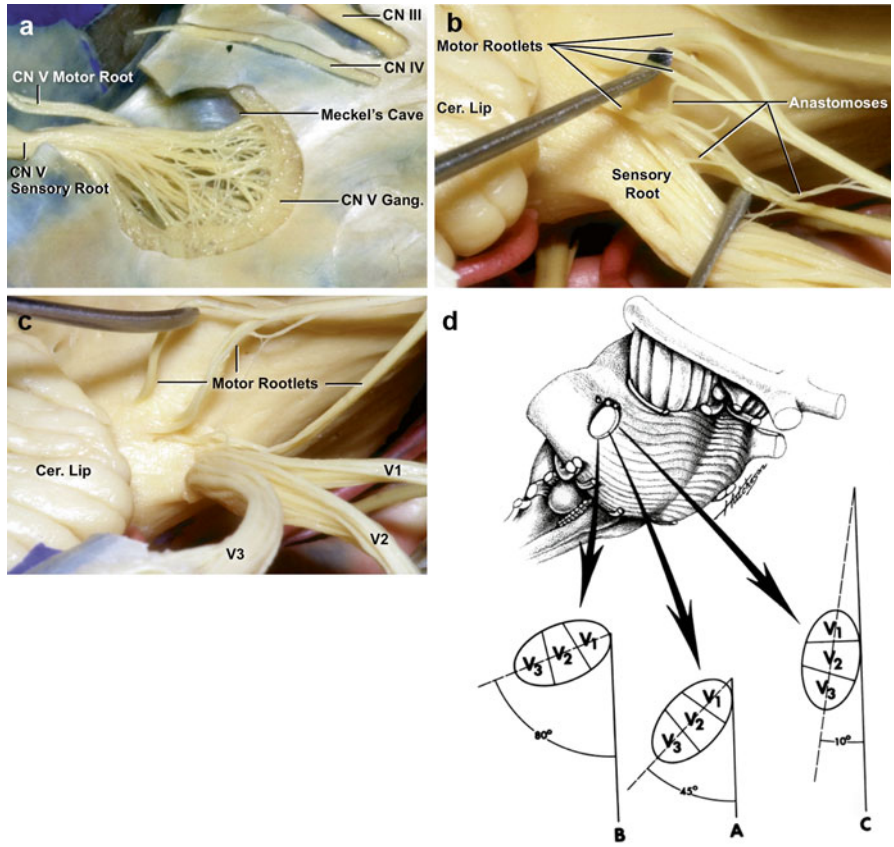


Fig. 2.2 Lateral views, right CN V. (**a**) Meckel's cave, the cistern that extends forward from the posterior fossa along CN V to the level of the midportion of the ganglion, has been exposed by removing the lateral dural wall of the cave. The motor root arises rostral to the sensory root and passes through Meckel's cave on the medial side of the sensory root and ganglion (Reprint with permission from Rhoton (2000b)). (**b**) Four motor rootlets, which arise around the rostral margin of the sensory root, have been elevated to expose the anastomoses between the motor and sensory roots. The cerebellar lip projects forward to form the cerebellomesencephalic fissure and may hide the junction of the sensory root with the pons in the retrosigmoid approach (Reprint with permission from Rhoton (2000b)). (**c**) A cleavage plane between the three trigeminal divisions has been started anteriorly and extended backward to the level of the posterior root. The first-division fibers are rostromedial within the posterior root and the third-division fibers caudolateral with the second division being in an intermediate location (Reprint with permission from Rhoton (2000b)). (**d**) Variability of the longest axis of the elliptical cross section of CN V at the pons (*broken line*) to the longitudinal axis of the body (*solid line*). The long axis of most nerves makes a 40–50° angle with the longitudinal axis of the body (*A*); however, this can vary from 10° (*C*) to 80° (*B*). In (**b**) the third division is almost directly lateral to the first division, and in **c** it is almost directly caudal (Reprint with permission from Gudmundsson et al. (1971)). *CN* cranial nerve, *Cer*. cerebellar, *Gang*. ganglion. *V1* the first trigeminal nerve division, *V2* the second trigeminal division, *V3* the third trigeminal nerve division

each division in the area posterior to the ganglion. The cross section of the sensory root between the pons and the petrous apex is elliptical. In most cases, the angle between the longest diameter of this cross section and the longitudinal axis of the body of CN V is 40–50°, but can vary from 10 to 80° (Fig. 2.2d) (Gudmundsson et al. 1971).

At the junction of the nerve with the pons, as many as 15 motor or aberrant sensory roots may be spread around the rostral half of the junction of the main sensory cone with the pons (Fig. 2.2b) (Gudmundsson et al. 1971). The aberrant sensory fibers penetrate the pons around the rostral two-thirds of the main sensory root and usually join the root a short distance from the brainstem. Of 66 aberrant rootlets found in our previous study of 50 specimens, 49 joined the first division, ten the second division, and seven the third division

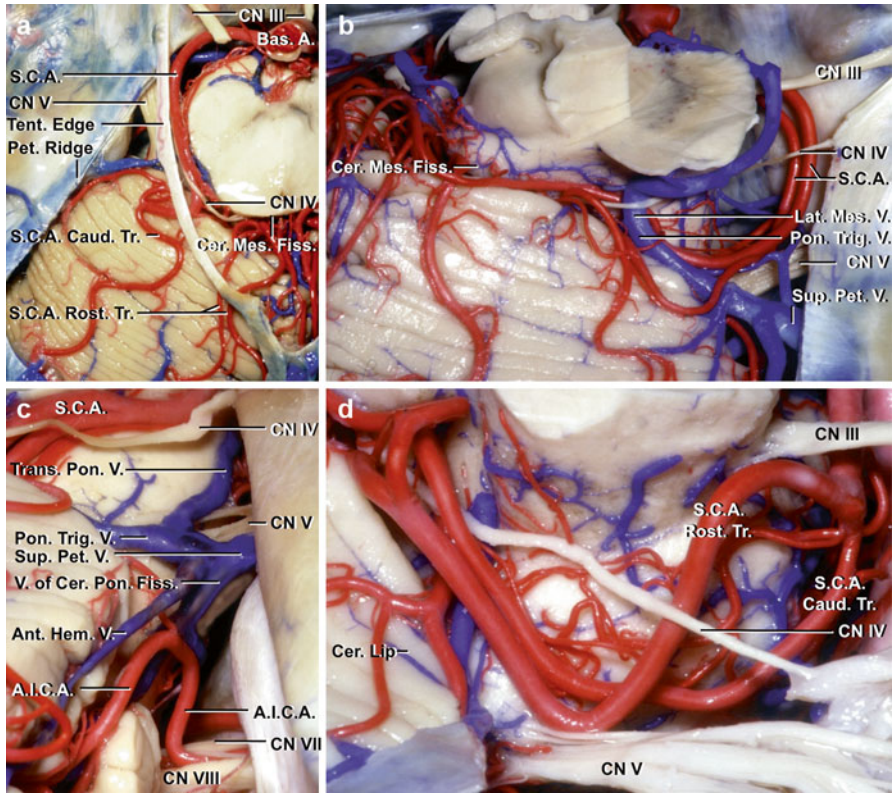


Fig. 2.3 Superior cerebellar artery. (**a**) Superior view. The SCA arises from the basilar artery anterior to the midbrain and encircles the pontomesencephalic junction to enter the cerebellomesencephalic fissure. It usually courses below CNs III and IV and above V and bifurcates into two major trunks, rostral and caudal, near CN V. The rostral trunk supplies the vermian and paravermian areas, and the caudal trunk supplies the hemisphere on the tentorial surface (Reprint with permission from Rhoton (2000a)). (**b**) Superolateral view. The right SCA arises from the basilar artery as a duplicate artery. The rostral duplicate trunk gives rise to vermian branches, and the caudal duplicate trunk gives rise to hemispheric branches, similar to the distribution of the rostral and caudal trunks formed by the bifurcation of a single SCA. The SCA or its major trunks often make a caudal loop to reach CN V ventral to the nerve. The superior petrosal vein and its tributaries also course near CN V. A well-developed pontotrigeminal vein, connecting the lateral mesencephalic vein with the superior petrosal vein, courses above the nerve (Reprint with permission from Rhoton (2000a)). (**c**) Right CPA. A large superior petrosal vein with multiple tributaries, including the pontotrigeminal and transverse pontine veins and the vein of the cerebellopontine fissure, passes above CN V. The AICA passes laterally between CNs VII and VIII and turns medially to course along the middle cerebellar peduncle and cerebellopontine fissure (Reprint with permission from Rhoton (2007)). (**d**) Superolateral view. SCA with an early bifurcation. The rostral trunk loops downward and indents the upper surface of CN V (Reprint with permission from Rhoton (2000b)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Ant*. anterior, *Bas*. basilar, *Caud*. caudal, *Cer*. cerebellar, cerebello, *CN* cranial nerve, *Fiss*. fissure, *Hem*. hemispheric, *Lat*. lateral, *Mes*. mesencephalic, *Pet*. petrosal, petrous, *Pon*. pontine, ponto, *Rost*. rostral, *S.C.A*. superior cerebral artery, *Sup*. superior, *Tent*. tentorial, *Tr*. trunk, *Trans*. transverse, *Trig*. trigeminal, *V*. vein

(Gudmundsson et al. 1971). Motor rootlets also arise around the rostral part of the nerve as 4–14 separate rootlets; however, they tend to arise further from the brainstem exist zone of the sensory root than the accessory sensory rootlets (Gudmundsson et al. 1971). Anastomoses between the motor and sensory roots are present in most nerves.

2.2.2 Anatomy of the SCA

The SCA arises in front of the midbrain, usually from the basilar artery near the apex, and passes below CN III, but may infrequently arise from the proximal posterior cerebral artery and pass above CN III (Fig. 2.3). It dips caudally and encircles the brainstem near the pontomesence-

phalic junction, passing below CN IV and above CN V. Its proximal portion courses medial to the free edge of the tentorium cerebelli, and its distal part passes below the tentorium (Fig. 2.3a). A meningeal branch occasionally originates from the main or rostral trunk near where the artery passes under the tentorium and enters the free edge of the tentorium. The SCA usually arises as a single trunk and bifurcates into two major trunks, rostral and caudal, near CN V. About half of the SCAs have a point of contact with CN V, which is usually on the superior or superomedial aspect of the nerve (Fig. 2.3d) (Hardy and Rhoton 1978). Depending on the site of bifurcation, the SCA trunk reaching the nerve may be the main, rostral, caudal, or both the rostral and caudal trunks, or a marginal hemispheric branch. After passing above CN V, the artery enters the cerebellomesencephalic fissure where its branches give rise to perforating branches to the brainstem and cerebellar peduncles and to precerebellar arteries entering the deep cerebellar white matter and dentate nucleus. On leaving the cerebellomesencephalic fissure, its branches pass posteriorly under the tentorial edge and are distributed to the tentorial cerebellar surface. The rostral trunk supplies the vermian and paravermian areas, and the caudal trunk supplies the hemispheric part of the tentorial surface.

2.2.3 Anatomy of the Superior Petrosal Vein

The superior petrosal veins empty into the superior petrosal sinus as one to three bridging veins (Fig. 2.4) (Rhoton 2000e). This superior petrosal venous complex is among the largest in the posterior fossa. It is frequently encountered in approaches to CN V and occasionally compresses CN V. The superior petrosal veins are usually formed by the union of several veins. The most common tributaries of the superior petrosal veins are the transverse pontine, pontotrigeminal, and anterolateral marginal veins and the veins of the cerebellopontine fissure and middle cerebellar peduncle (Matsushima et al. 1983, 2014). Of 20 superior petrosal sinuses examined in our previous study, eight received one superior petrosal vein, ten received two, and two received three (Matsushima et al. 1983). The draining points of the superior petrosal vein into the superior petrosal sinus were classified into medial, intermediate, or lateral groups based on whether they drained into the superior petrosal sinus in an intermediate location above the internal acoustic meatus or medial or lateral to the meatus. Of 34 superior petrosal veins, 22 (64.7 %) were of the medial type, three (8.8 %) were of the intermediate type, and nine (26.5 %) were of the lateral type (Matsushima et al. 1983).

2.2.4 Offending Vessels of Trigeminal Neuralgia

The SCA, AICA, PICA, the superior petrosal vein and its tributaries, or any combination of these vessels may cause compression of CN V (Figs. 2.5, 2.6, 2.7, and 2.8) (Apfelbaum 2000; Barker et al. 1996; Hong et al. 2011; Jannetta 1980; Li et al. 2004; Lorenzoni et al. 2008; Matsushima et al. 2004; Sekula et al. 2009; Sindou et al. 2008; Thomas and Vilensky 2014). The vertebral and basilar arteries have rarely been reported to be the compressing vessel (Apfelbaum 2000; Barker et al. 1996; Lorenzoni et al. 2008; Thomas and Vilensky 2014; Yang et al. 2012).

The most commonly found offending vessel found in vascular decompression operations for trigeminal neuralgia is the caudal loop of the SCA (Hardy and Rhoton 1978). The SCA was found to be compressing CN V in 52 % of the 50 CPAs that we previously examined and the AICA in 8 % (Hardy and Rhoton 1978). The point of contact between CN V and the SCA is usually on the superior or superomedial aspect of the nerve, and often a few fascicles of the nerve are distorted by an SCA that has looped down into the axilla between the medial side of the nerve and the pons. An arterial loop in the axilla may not be visible from the retrosigmoid view behind CN V if the SCA courses around the brainstem directly in front of the nerve, or if it passes over the rostral aspect of the nerve very close to the brainstem,

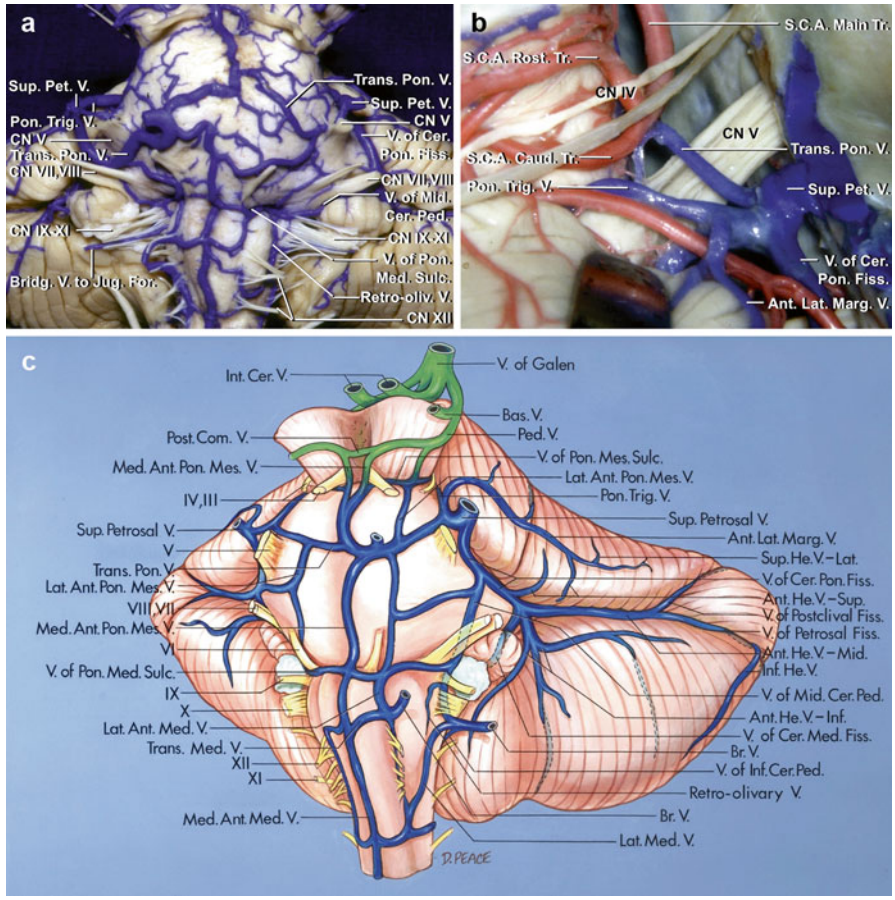


Fig. 2.4 Posterior fossa veins. (**a**) CN V may be compressed by the superior petrosal veins and their tributaries, including the transverse pontine and pontotrigeminal veins and the veins of the cerebellopontine fissure and middle cerebellar peduncle. The right transverse pontine vein makes contact with the lower surface of the right CN V, and the left transverse pontine vein compresses the upper surface of the left CN V. The vein of the pontomedullary sulcus or the middle cerebral peduncle may compress CN VII, as seen in both sides of this specimen. CNs IX and often X may be compressed by surrounding veins, such as the bridging veins emptying into the jugular foramen or the vein of the pontomedullary sulcus, as seen on the right side (Reprint with permission from Rhoton (2000e)). (**b**) Superolateral view. A large superior petrosal vein formed by the union of the transverse pontine, pontotrigeminal, and anterolateral marginal veins and the vein of the cerebellopontine fissure. The superior surface of CN V is indented by the transverse pontine and pontotrigeminal veins. The SCA bifurcates into rostral and caudal trunks medial to CN V (Reprint with permission from Rhoton (2000e)). (**c**) The veins in the posterior fossa are divided into three groups: a galenic group (*green*) that drains into the vein of Galen, a petrosal group (*blue*) that drains into the superior and inferior petrosal sinuses, and a tentorial group that drains into the sinuses near the torcula. The veins surrounding CN V, VII, or IX and X, may be associated with trigeminal neuralgia, hemifacial spasm, or glossopharyngeal neuralgia, respectively (Reprint with permission from Rhoton (2000e)). *Ant*. anterior, *Bas*. basal, *Br. Bridg*. bridging, *Caud*. caudal, *Cer*. cerebellar, cerebello, cerebral, *CN* cranial nerve, *Com*. communicating, *Fiss*. fissure, *For*. foramen, *He*. hemispheric, *Inf*. inferior, *Int*. internal, *Jug*. jugular, *Lat*. lateral, *Marg*. marginal, *Med*. medial, medullary, *Mes*. mesencephalic, *Mid*. middle, *Ped*. peduncle, peduncular, *Pet*. petrosal, *Pon*. pontine, ponto, *Post*. posterior, *Retro-oliv*. retro-olivary, *Rost*. rostral, *S.C.A*. superior cerebral artery, *Sulc*. sulcus, *Sup*. superior, *Tr*. trunk, *Trans*. transverse, *Trig*. trigeminal, *V*. vein

where it may be hidden by the overhanging lip of the cerebellomesencephalic fissure. Exposure directed along the petrosal cerebellar surface, as in the common retrosigmoid approach, but also through the lateral part of the tentorial cerebellar surface, described as the infratentorial lateral supracerebellar approach, may provide better visualization of an arterial loop directly medial to

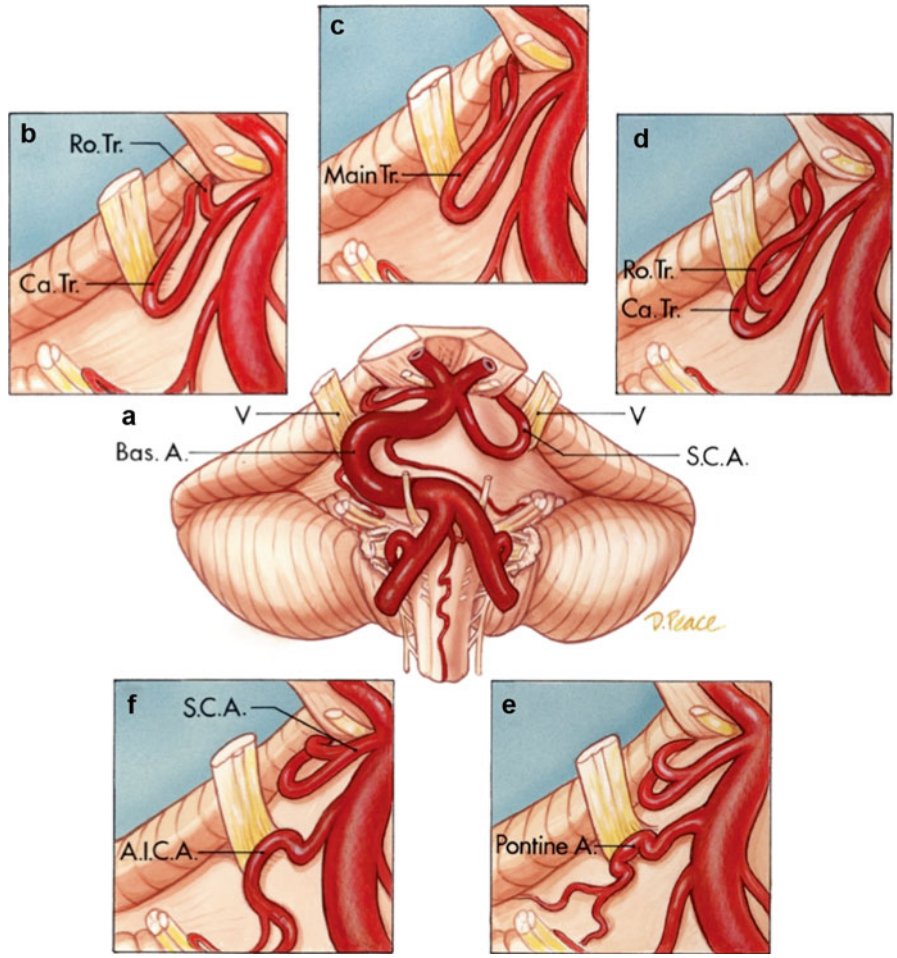


Fig. 2.5 Arterial compression of CN V. Sites of arterial compression of CN V. Orientation as shown in the *central diagram*. (**a**) *Central diagram*. The right CN V is compressed by a tortuous basilar artery, and the left CN V is compressed by the main trunk of the SCA. (**b**) The SCA bifurcates into rostral and caudal trunks prior to reaching CN V. The nerve is compressed by the caudal trunk. (**c**) The SCA bifurcates distal to the nerve. The nerve is compressed by the main trunk. (**d**) The SCA bifurcates prior to reaching the nerve. The nerve is compressed by both the rostral and caudal trunks. (**e**) The nerve is compressed by a large pontine artery. (**f**) The nerve is compressed by an AICA that has a high origin and loops upward into the medial surface of the nerve (Reprint with permission from Rhoton (1990)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Bas*. basilar, *Ca*. caudal, *Ro*. rostral, *S.C.A*. superior cerebral artery, *Tr*. trunk

the nerve (Fujimaki and Kirino 2000; Matsushima et al. 1989). The loop of the SCA may be seen dangling below the lower margin of the nerve, although it is not visible above the nerve. These SCA loops, however, always pass along the medial and rostral surfaces of the nerve to reach the cerebellomesencephalic fissure. This arterial loop may contact CN V more than once, especially if the loop is long enough to overlap. The medial axilla of the nerve must be carefully examined before concluding that there is no arterial loop in the axilla of the nerve. It is important to remember that the trunks do not pass directly from the side of the brainstem to the superior surface of the cerebellum, but, rather, that they dip into the deep fissure between the cerebellum and midbrain at the posterior margin of CN V. The most common site of compression of CN V on the SCA is around the junction of the main trunk with the origin of the rostral and caudal trunks

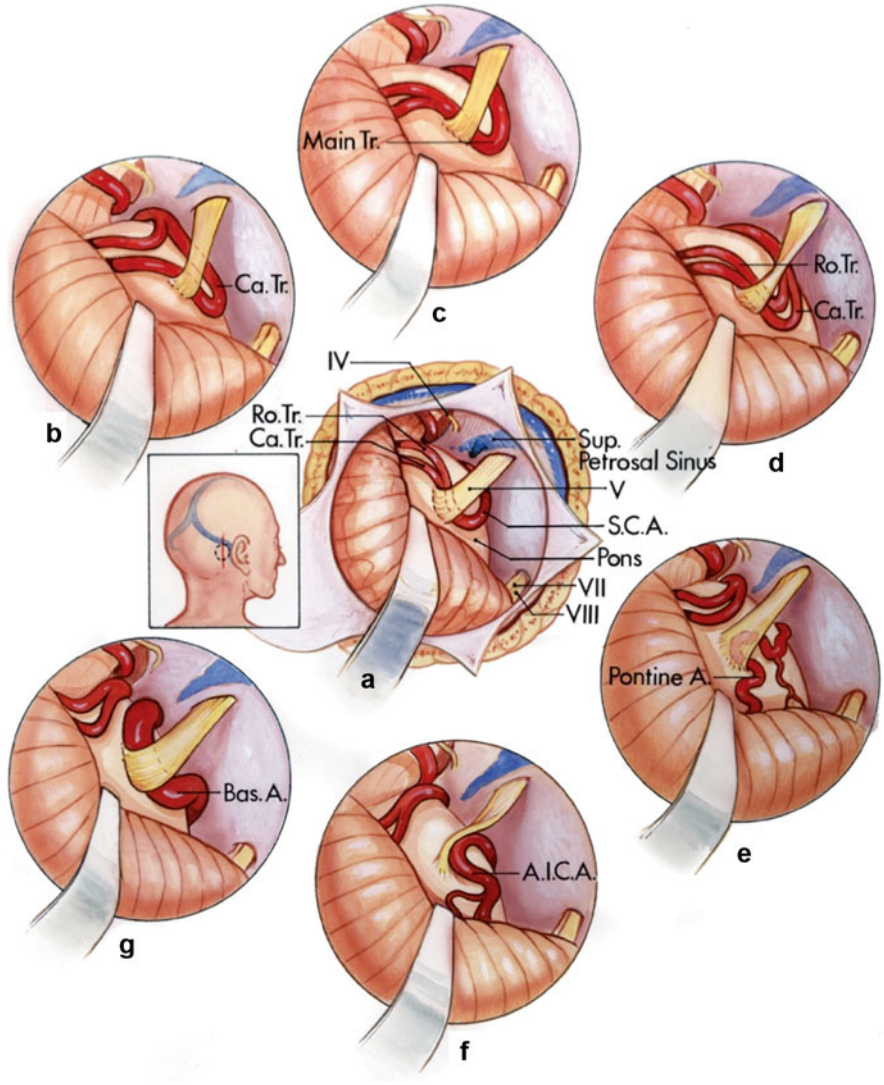


Fig. 2.6 Arterial compression of CN V. Sites of arterial compression of CN V as seen through a suboccipital craniotomy. (**a**) *Central diagram*. The site of the skin incision (*solid line*) and the craniotomy (*interrupted line*), exposing the junction of the sigmoid and transverse sinuses, are shown in the *inset*. The cerebellum has been elevated to expose the junction of CN V with the pons. CN IV is at the superior margin of the exposure and CNs VII and VIII are at the lower margin. CN V is compressed by a loop of the SCA that dangles down into the axilla of the nerve. The site of compression on the artery is at the junction of the main trunk with the rostral and caudal trunks. (**b**) The nerve is compressed by the caudal trunk. (**c**) The nerve is compressed by the main trunk. (**d**) Compression by both the rostral and caudal trunks. (**e**) Compression by a pontine branch of the basilar artery. (**f**) Compression by the AICA from the inferior side. (**g**) Compression by a tortuous basilar artery from the medial side (Reprint with permission from Rhoton (1990)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Bas*. basilar, *Ca*. caudal, *Ro*. rostral, *S.C.A*. superior cerebral artery, *Sup*. superior, *Tr*. trunk

(Rhoton 1990). However, other sites of compression are seen depending on how far distal the artery bifurcates in relation to CN V. If the SCA bifurcates near the basilar artery or if there is a duplicate configuration in which the rostral and caudal trunks arise directly from the basilar artery, both trunks may loop down into the axilla between the pons and CN V to compress the nerve. There are cases in which both the rostral and caudal trunks contact the nerve, so careful

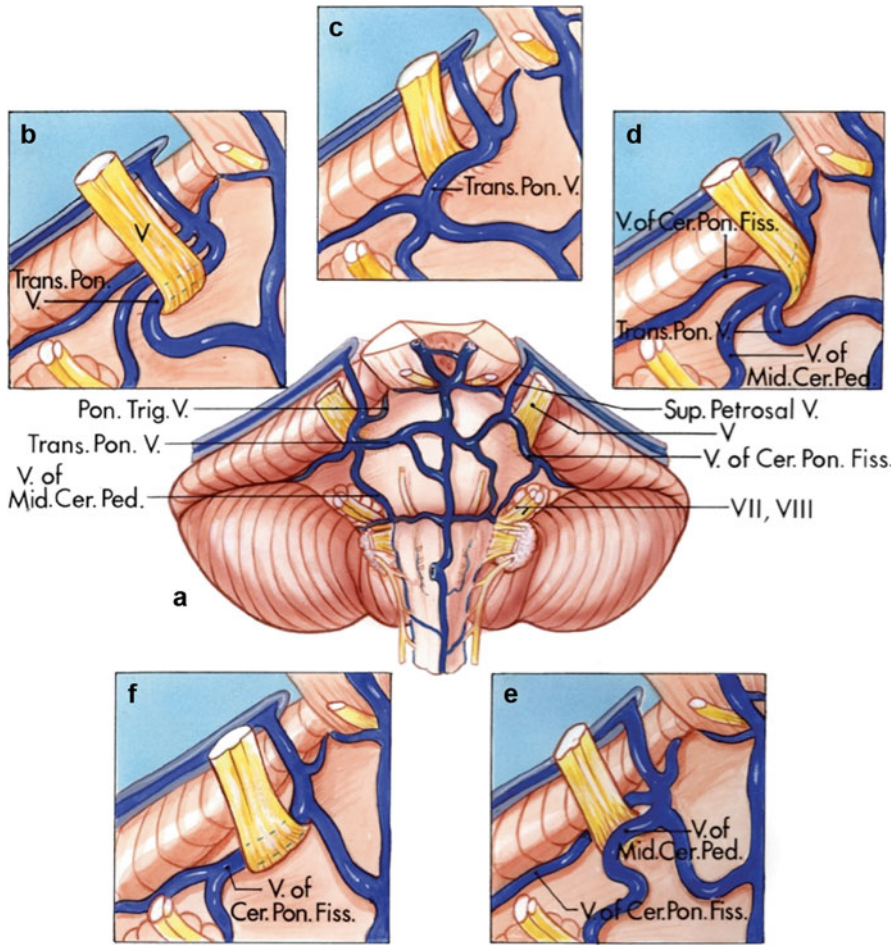


Fig. 2.7 Venous compression of CN V. Sites of venous compression of CN V. (**a**) *Central diagram*. The veins that commonly compress CN V are tributaries of the superior petrosal veins. The tributaries that converge on and may compress the nerve are the transverse pontine and pontotrigeminal veins and the veins of the cerebellopontine fissure and middle cerebellar peduncle. The transverse pontine veins course transversely across the pons. The vein of the middle cerebellar peduncle arises in the region of CNs VII and VIII and ascends on the pons. The vein of the cerebellopontine fissure arises along the superior limb of the cerebellopontine fissure. The pontotrigeminal vein arises on the upper pons and passes above CN V. (**b**) A transverse pontine vein compresses the lateral side of the nerve and joins the veins of the middle cerebellar peduncle and cerebellopontine fissure to empty into a superior petrosal vein. (**c**) The medial side of the nerve is compressed by a tortuous transverse pontine vein. (**d**) The lateral side of the nerve is compressed by the junction of the transverse pontine vein with the veins of the middle cerebellar peduncle and the cerebellopontine fissure. (**e**) The nerve is compressed on the medial side by the vein of the middle cerebellar peduncle and on the lateral side by the vein of the cerebellopontine fissure. (**f**) The lateral side of the nerve is compressed by the vein of the cerebellopontine fissure (Reprint with permission from Rhoton (1990)). *Cer*. cerebellar, cerebello, *Fiss*. fissure, *Mid*. middle, *Ped*. peduncle, *Pon*. pontine, ponto, *Sup*. superior, *Trans*. transverse, *Trig*. trigeminal, *V*. vein

attention is needed to the caudal trunk to determine if it is compressing the junction of the nerve with the pons, which could be hidden by the overhanging lip of the cerebellomesencephalic fissure. Alternatively, if the artery bifurcates before reaching the nerve, the caudal trunk may compress the nerve and the rostral trunk may course well above the nerve. If the artery bifurcates distal to the nerve, only the main trunk will be involved in the compression. The point of bifurcation of the SCA affects the caliber of the vessel that makes contact with CN V; the contacting vessel will be of a smaller caliber if the SCA bifurcates before reaching CN V.

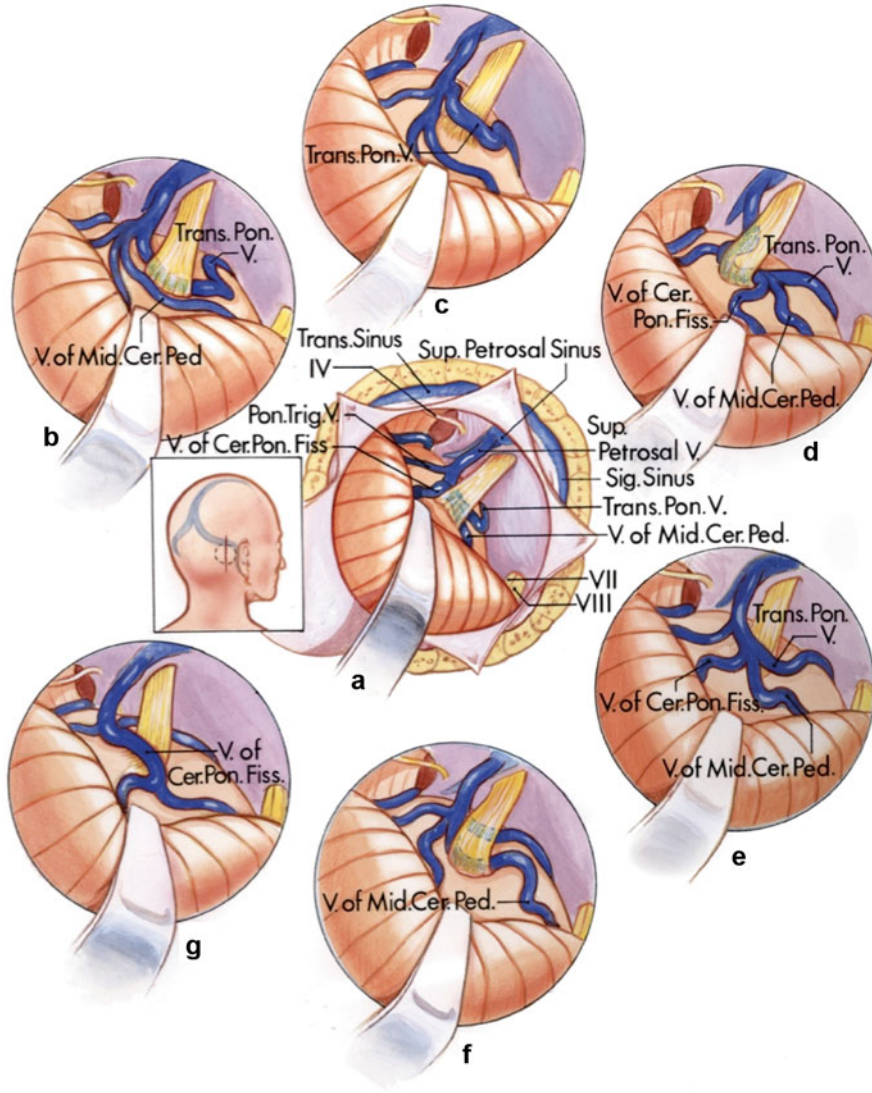


Fig. 2.8 Venous compression of CN V. Sites of venous compression of CN V as seen through a retrosigmoid craniotomy. (**a**) The *inset* shows the site of the scalp incision (*solid line*) and the craniotomy (*interrupted line*). The superior petrosal veins empty into the superior petrosal sinus. CN V is compressed by the junction of a transverse pontine vein and the vein of the middle cerebellar peduncle with the superior petrosal vein. The vein of the cerebellopontine fissure ascends behind the nerve, and the pontotrigeminal vein passes above the nerve. (**b**) CN V is compressed on its medial side by a transverse pontine vein and on its lateral side by the vein of the middle cerebellar peduncle. (**c**) The lateral side of the nerve is compressed by a transverse pontine vein. (**d**) The medial side of the nerve is compressed by the junction of a transverse pontine vein with the veins of the middle cerebellar peduncle and cerebellopontine fissure. (**e**) The lateral side of the nerve is compressed by the junction of the transverse pontine vein with the veins of the middle cerebellar peduncle and cerebellopontine fissure. (**f**) The medial side of the nerve is compressed by the vein of the middle cerebellar peduncle. (**g**) The lateral side of the nerve is compressed by the vein of the cerebellopontine fissure (Reprint with permission from Rhoton (1990)). *Cer*. cerebellar, cerebello, *Fiss*. fissure, *Mid*. middle, *Ped*. peduncle, *Pon*. pontine, ponto, *Sig*. sigmoid, *Sup*. superior, *Trans*. transverse, *Trig*. trigeminal, *V*. vein

A less common source of compression of CN V is by the AICA. The AICA may have a high origin and loop upward to indent the medial or lower surface of CN V prior to passing downward to course with CNs VII and VIII (Figs. 2.5f and 2.6f). A serpentine basilar artery may also wander

laterally and compress the medial side of CN V (Fig. 2.6g) (Apfelbaum 2000; Barker et al. 1996; Lorenzoni et al. 2008; Sunderland 1948; Thomas and Vilensky 2014). This type of basilar artery is often elongated and has a fusiform configuration. More than one artery may compress the nerve, for example, in some cases the SCA compresses the rostral surface of the nerve and the AICA compresses the caudal surface. Infrequently, a tortuous and arteriosclerotic vertebral artery or an upward loop of the PICA may reach and groove the undersurface of CN V. In the latter cases, CN XII may be stretched and thinned by the tortuous artery, and care should be taken to avoid damaging CN XII during the arterial mobilization. CN VI, located medial to the vertebral artery, should also be kept in mind during mobilization from lateral to medial, even though the nerve is difficult to expose sufficiently. CN V may also be compressed by a large pontine branch of the basilar artery, which passes around and penetrates the pons (Figs. 2.5e and 2.6e) (Rhoton 2000b).

Although rarer than arterial compression, compression and distortion of CN V by the surrounding veins also is found in trigeminal neuralgia (Figs. 2.7 and 2.8) (Hong et al. 2011; Matsushima et al. 2004). The superior petrosal veins and their tributaries are the most frequent veins compressing CN V. The transverse pontine veins, which pass transversely near CN V, are the most frequent veins to compress CN V. They may course medially in the axilla of the nerve or pass above, below, or lateral to the nerve and may indent any of its surfaces. The vein of the middle cerebellar peduncle may compress the lateral or medial surface of CN V before joining the superior petrosal veins emptying into the superior petrosal sinus. The vein of the cerebellopontine fissure may indent the lateral margin of CN V as it ascends toward the superior petrosal sinus, and the pontotrigeminal vein may indent the upper margin of the nerve. The junction of these veins, which converge and form a single trunk prior to entering the superior petrosal sinus, is usually lateral to CN V (Matsushima et al. 1983, 2014). This junction, however, may be located medial to CN V, in which case the common trunk must pass around CN V prior to reaching the superior petrosal sinus. These common venous trunks also may compress CN V.

2.3 Middle Neurovascular Complex and Hemifacial Spasm

The middle complex includes the AICA, pons, middle cerebellar peduncle, cerebellopontine fissure, petrosal surface of the cerebellum, and CNs VI–VIII. The AICA arises at the pontine level and courses through the cerebellopontine cistern with CNs VI–VIII to reach the surface of the middle cerebellar peduncle, where it courses along the cerebellopontine fissure and terminates by supplying the petrosal surface of the cerebellum (Rhoton 2013).

2.3.1 Anatomy of CNs VII and VIII

CNs VII and VIII arise at the lateral end of the pontomedullary sulcus immediately rostral to the foramen of Luschka and its rhomboid lip and choroid plexus and ventrocaudal to the flocculus (Fig. 2.9) (Rhoton 2000c). The rhomboid lip is a sheetlike layer of neural tissue attached to the lateral margin of the ventricular floor that joins the tela choroidea to form the ventral edge of a pouch at the outer extremity of the lateral recess. A large rhomboid lip adhering to the dorsal surface of CNs IX and X may block visualization of the brainstem exit of CN VII and would require elevation of the rhomboid lip away from the lower CNs to expose CN VII (Funaki et al. 2010; Nakahara et al. 2013). CN VII arises from the brainstem near the lateral end of the pontomedullary sulcus 1–2 mm anterior to the point at which CN VIII joins the brainstem. The interval between CNs VII and VIII is greatest at the level of the pontomedullary sulcus and decreases as these nerves approach the meatus. The nervus intermedius arises on the anterior surface of CN VIII, has a free segment in the cistern and/or meatus, and joins CN VII distally (Fig. 2.10).

Commonly, the flocculus protrudes behind CNs VII and VIII and blocks their exposure at the junction with the brainstem in the retrosigmoid exposure. It may also be difficult to see CN VII because it is usually hidden anterior to CN VIII. A helpful way of visualizing the point where CN VII will exit from the brainstem is to project an

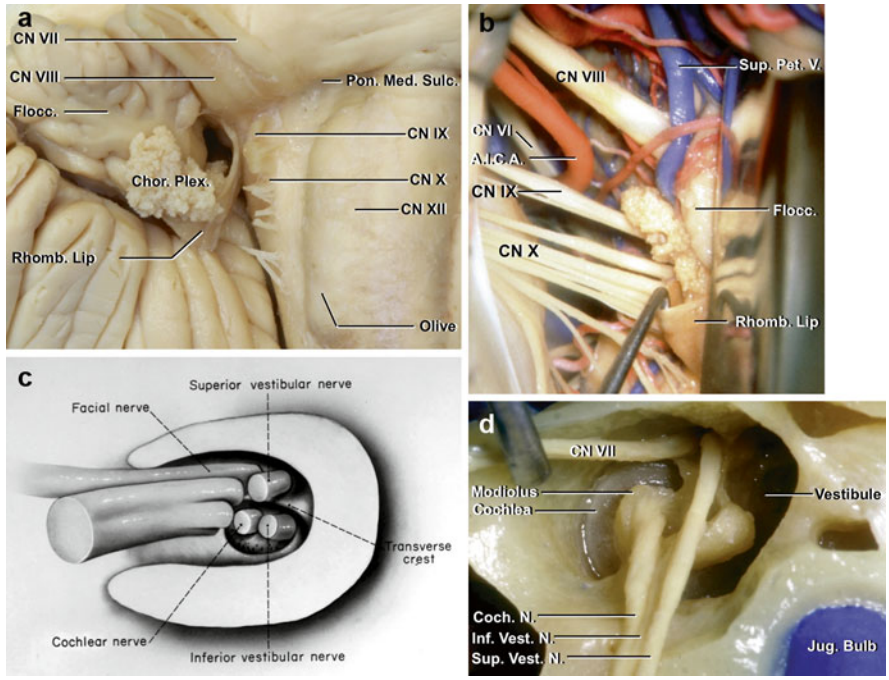


Fig. 2.9 CNs VII and VIII. (**a**) Anterolateral view of the right CPA. CNs VII and VIII arise at the lateral end of the pontomedullary sulcus, ventral to the flocculus and just rostral to a line drawn along the junction of the rootlets of CNs IX–XI with the medulla. The rhomboid lip is a sheetlike layer of neural tissue attached to the lateral margin of the ventricular floor that joins the tela choroidea to form a pouch at the outer extremity of the lateral recess. (**b**) Left retrosigmoid exposure. The choroid plexus protrudes from the foramen of Luschka into the CPA behind CNs VII and VIII. The junction of CN VII with the brainstem is located in front of and is hidden by CN VIII. A nerve hook has been placed inside the rhomboid lip (Reprint with permission from Rhoton (2000b)). (**c**) View of the right internal acoustic meatus with the posterior lip removed to show the four portions of the lateral meatus. The facial and superior vestibular nerves are above the transverse crest with the facial nerve anterior, and the cochlear and inferior vestibular nerves are below with the cochlear nerve anterior. The facial nerve is separated from the superior vestibular nerve by the vertical crest (Reprint with permission from Rhoton (1974)). (**d**) Cadaveric dissection to show the facial, superior and inferior vestibular, and cochlear nerves in the lateral meatus. The cochlear nerve penetrates the modiolus of the cochlea where its fibers are distributed to the turns of the cochlear duct. The basal turn of the cochlea communicates below the modiolus with the vestibule (Reprint with permission from Rhoton (2000f)). *A.I.C.A.* anterior inferior cerebellar artery, *Chor. Plex.* choroid plexus, *CN* cranial nerve, *Coch.* cochlear, *Flocc.* flocculus, *Inf.* inferior, *Jug.* jugular, *N.* nerve, *Pet.* petrosal, *Pon. Med.* pontomedullary, *Rhomb.* rhomboid, *Sulc.* sulcus, *Sup.* superior, *V.* vein, *Vest.* vestibular

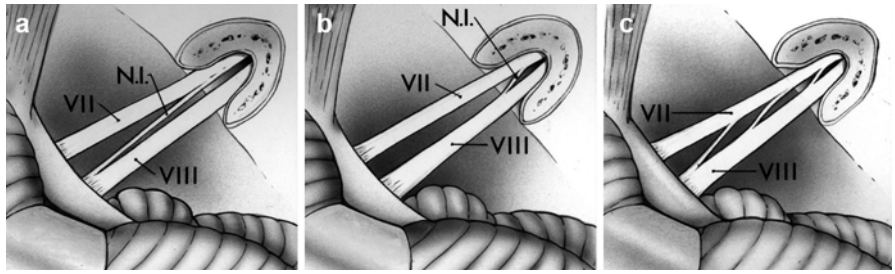


Fig. 2.10 CNs VII and VIII and the nervus intermedius. View of the CPA from above to show the relationship of the nervus intermedius to CNs VII and VIII. (**a**) Most common relationship. The nervus intermedius is joined to the ventral surface of CN VIII for a few millimeters adjacent to the brainstem and then has a free segment in the CPA as it courses to join the facial motor root. (**b**) Pattern presents in 20 % of the nerves studied. The free segment is entirely in the meatus. (**c**) The nervus intermedius consists of three free segments: two are in the CP angle and one is in the meatus. The nervus intermedius in (**a**) could be exposed in the angle without drilling the posterior lip of the meatus. In (**b**) the free segment could not be found in the angle but only in the meatus (Reprint with permission from Rhoton and Tedeschi (1992)). *N.I.* nervus intermedius

imaginary line along the medullary junction of the rootlets forming the lower CNs IX–XI upward through the pontomedullary junction (Rhoton 2000b). This line, at a point 2–3 mm above the junction of CN IX with the medulla, will pass through the pontomedullary junction at the site where CN VII exits from the brainstem. The junction of CN VII with the brainstem is easier to expose in hemifacial spasm from below the flocculus and CN VIII than above, a technique that has been described as an "infrafloccular approach"(Hitotsumatsu et al. 2003). A key step is to elevate the flocculus, rhomboid lip, and choroid plexus after opening the cerebellopontine and cerebellomedullary fissures around the inferior and lateral margin of the flocculus (Fig. 2.11).

The nerves in the lateral part of the internal acoustic meatus are the facial, cochlear, and superior and inferior vestibular nerves (Fig. 2.9c, d) (Rhoton 1974; Rhoton and Tedeschi 1992). The position of the nerves is most constant in the lateral portion of the meatus, which is divided into a superior and an inferior portion by a horizontal ridge, called either the transverse or falciform crest (Rhoton 2007). The facial and the superior vestibular nerves are superior to the crest. The facial nerve is anterior to the superior vestibular nerve and is separated from it at the lateral end of the meatus by a vertical ridge of bone called the vertical crest or "Bill's bar." The cochlear and inferior vestibular nerves run below the transverse crest with the cochlear nerve located anteriorly. In summary, the lateral meatus can be divided into four portions, with the facial nerve anterosuperior, the cochlear nerve anteroinferior, the superior vestibular nerve posterosuperior, and the inferior vestibular nerve posteroinferior (Rhoton 2000f).

2.3.2 Anatomy of the AICA

The AICA originates from the basilar artery, usually as a single trunk, and encircles the pons near CNs VI–VIII (Fig. 2.12). In most cases, the AICA passes below CNs VII and VIII, but it may also pass above or between these nerves in its course around the brainstem. After coursing near and sending branches to the nerves entering the acoustic meatus and to the choroid plexus protruding from the foramen of Luschka, it passes around the flocculus to supply the lips of the cerebellopontine fissure and the petrosal cerebellar surface. It commonly bifurcates near CNs VII and VIII to form a rostral and a caudal trunk. Approximately two-thirds bifurcate before and one-third after crossing CNs VII and VIII (Rhoton 2000a). Duplicate AICAs have a distribution similar to the distribution of the rostral and caudal trunks formed by the bifurcation of a single AICA. The rostral trunk sends its branches laterally along the middle cerebellar peduncle to the superior lip of the cerebellopontine fissure and the adjoining part of the petrosal surface, and the caudal trunk supplies the inferior part of the petrosal surface, including a part of the flocculus and the choroid plexus. The AICA gives rise to perforating arteries to the brainstem, choroidal branches to the tela and choroid plexus, and nerve-related arteries, including the labyrinthine, recurrent perforating, and subarcuate arteries. If the bifurcation is proximal to CNs VII and VIII, either the rostral trunk alone or both of the post-bifurcation trunks may be nerve related.

2.3.3 Offending Vessels in Hemifacial Spasm

The AICA, PICA, vertebral artery, or a combination of these arteries may compress CN VII (Figs. 2.13 and 2.14) (Barker et al. 1995; Huang et al. 1992; Hyun et al. 2010; Jannetta 1980; Mikami et al. 2013; Park et al. 2008). Although rare, the basilar artery, SCA, and the veins around the pontomedullary junction have also been reported as offending vessels (Barker et al. 1995; Campos-Benitez and Kaufmann 2008; Huang et al. 1992; Hyun et al. 2010; Park et al. 2008).

The AICA or its major trunks may compress CN VII at its exit from the brainstem, before reaching the porus of the internal acoustic meatus. The point of contact with the AICA is usually on the inferior or inferomedial aspect of CN VII's exit from the brainstem. The AICA often reapproaches CN VII near the internal acoustic meatus where it gives rise to the nerve-related branches (Matsushima et al. 1990). It is generally

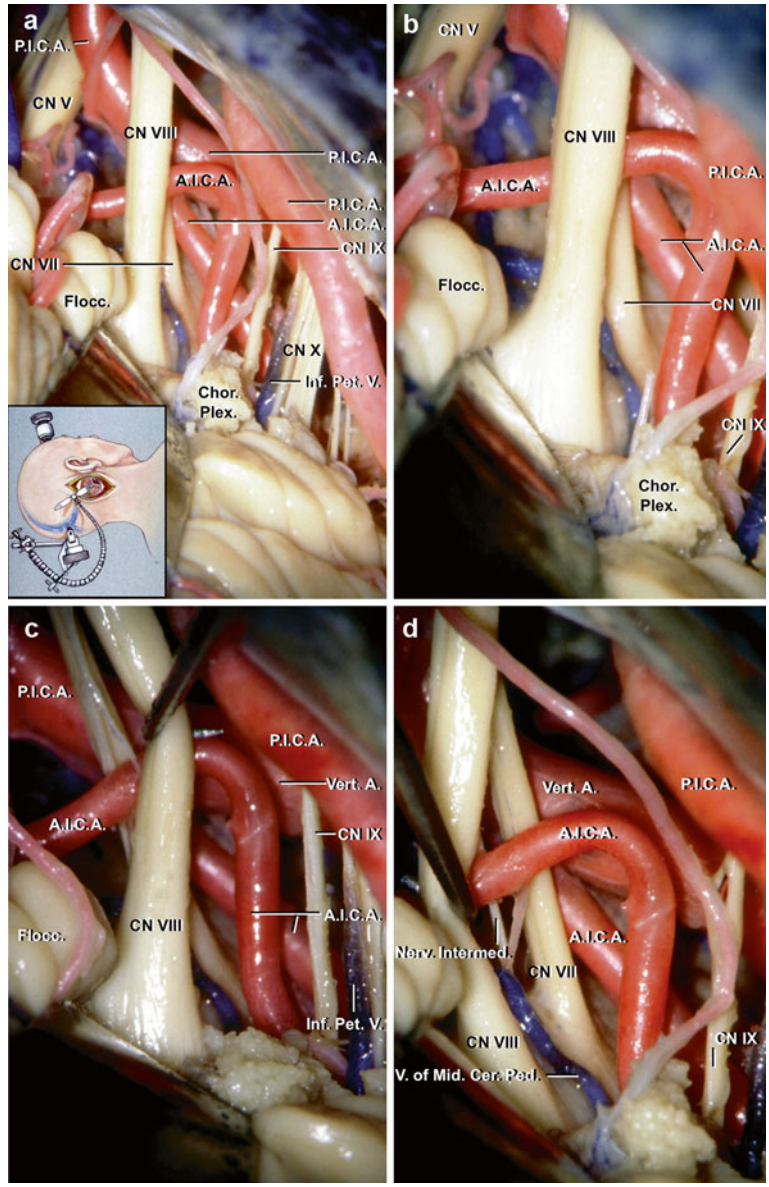


Fig. 2.11 Right infrafloccular exposure of CN VII in hemifacial spasm. (**a**) The *inset* shows the approach along the inferolateral margin of the cerebellum and below the flocculus. The cerebellum has been elevated to expose the right CPA. CN VII's exit zone from the brainstem is exposed along the lower margin of CN VIII. The AICA passes between CNs VII and VIII. A large tortuous PICA loops upward anterior to CNs VII and VIII, before turning downward to reach the cerebellomedullary fissure. The flocculus and choroid plexus protruding from the foramen of Luschka often hide the junction of CNs VII and VIII with the brainstem. In this case, the flocculus has been gently elevated to expose the junction of CNs VII and VIII with the brainstem. (**b**) Enlarged view. Exposing the exit for CN VII from the brainstem is facilitated by directing the exposure along the inferolateral margin of the cerebellum in the area above CN IX and below the lower edge of the flocculus. (**c**) CN VIII has been depressed. This exposes the distal segment of CN VII, but does not provide access to the junction of CN VII with the brainstem, which should be visualized in dealing with hemifacial spasm. (**d**) CN VIII has been gently elevated. This exposes both the rostral and caudal margins of CN VII at the brainstem. A rootlet of the nervus intermedius is also exposed. The vein of the middle cerebellar peduncle passes between CNs VII and VIII (Reprint with permission from Rhoton (2000b)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Cer*. cerebellar, *Chor. Plex*. choroid plexus, *CN* cranial nerve, *Flocc*. flocculus, *Inf*. inferior, *Mid*. middle, *Nerv. Intermed*. nervus intermedius, *P.I.C.A*. posterior inferior cerebellar artery, *Ped*. peduncle, *Pet*. petrosal, *V*. vein, *Vert*. vertebral

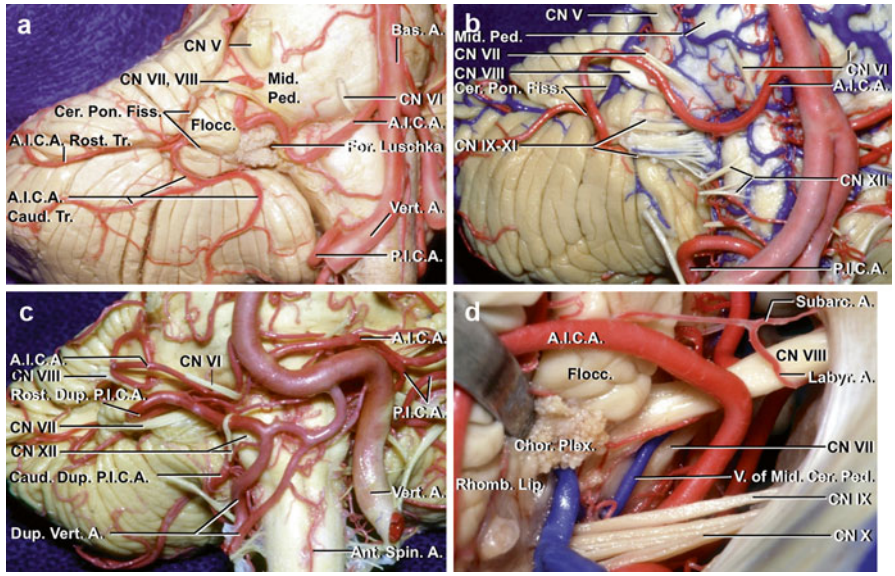


Fig. 2.12 Anterior inferior cerebellar artery. (**a**) The right AICA, originating from the basilar artery as a single trunk, passes below CN VI and bifurcates near CNs VII and VIII to form a rostral and a caudal trunk. The AICA reaches CN VII at its exit from the brainstem and usually reapproaches CNs VII and VIII near the internal acoustic meatus where it gives rise to nerve-related branches (Reprint with permission from Rhoton (2000a)). (**b**) The right AICA passes between CNs VII and VIII before reaching the cerebellopontine fissure and petrosal cerebellar surface (Reprint with permission from Rhoton (2000a)). (**c**) The right vertebral artery is a duplicate artery that gives rise to duplicate PICAs. The AICAs arise from the lower part of the basilar artery. The left AICA is larger than the right. The rostral duplicate PICA loops upward into the CPA. The left vertebral artery loops upward into the left CPA (Reprint with permission from Rhoton (2000a)). (**d**) Right retrosigmoid exposure. The flocculus and choroid plexus, which protrude from the foramen of Luschka, have been elevated to expose the junction of CNs VII and VIII with the brainstem. CN VII is exposed below CN VIII above CN IX. A branch of the AICA gives rise to both the subarcuate and labyrinthine arteries. The subarcuate artery enters the dura and bone superolateral to the meatus. The exit zone of CN VII from the brainstem is easier to expose for decompression of CN VII from below in an "infraflocculer approach" than from above the flocculus and CN VIII (Reprint with permission from Rhoton (2000a)). Anterior inferior cerebellar artery. (**a**) The right AICA, originating from the basilar artery as a single trunk, passes below CN VI, and bifurcates near CNs VII and VIII to form a rostral and a caudal trunk. The AICA reaches CN VII at its exit from the brainstem, and usually reapproaches CNs VII and VIII near the internal acoustic meatus where it gives rise to nerve-related branches (Reprint with permission from Rhoton (2000a)). (**b**) The right AICA passes between CNs VII and VIII before reaching the cerebellopontine fissure and petrosal cerebellar surface (Reprint with permission from Rhoton (2000a)). (**c**) The right vertebral artery is a duplicate artery that gives rise to duplicate PICAs. The AICAs arise from the lower part of the basilar artery. The left AICA is larger than the right. The rostral duplicate PICA loops upward into the CPA. The left vertebral artery loops upward into the left CPA (Reprint with permission from Rhoton (2000a)). (**d**) Right retrosigmoid exposure. The flocculus and choroid plexus, which protrude from the foramen of Luschka, have been elevated to expose the junction of CNs VII and VIII with the brainstem. CN VII is exposed below CN VIII above CN IX. A branch of the AICA gives rise to both the subarcuate and labyrinthine arteries. The subarcuate artery enters the dura and bone superolateral to the meatus. The exit zone of CN VII from the brainstem is easier to expose for decompression of CN VII from below in an "infraflocculer approach" than from above the flocculus and CN VIII (Reprint with permission from Rhoton (2000a)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Ant*. anterior, *Bas*. basilar, *Caud*. caudal, *Cer*. cerebellar, cerebello, *Chor. Plex*. choroid plexus, *CN* cranial nerve, *Dup*. duplicate, *Fiss*. fissure, *Flocc*. flocculus, *For*. foramen, *Labyr*. labyrinthine, *Mid*. middle, *P.I.C.A*. posterior inferior cerebellar artery, *Ped*. peduncle, *Pon*. pontine, *Rhomb*. rhomboid, *Rost*. rostral, *Spin*. spinal, *Subarc*. subarcuate, *Tr*. trunk, *V*. vein, *Vert*. vertebral

accepted that hemifacial spasm is caused by compression of CN VII at its exit from the brainstem rather than near the meatus (Gardner 1966; Moller and Jannetta 1984; Tomii et al. 2003). The AICA often gives off a few perforating arteries near its exit from the brainstem, thus limiting the amount of arterial repositioning that can be done in decompression surgery.

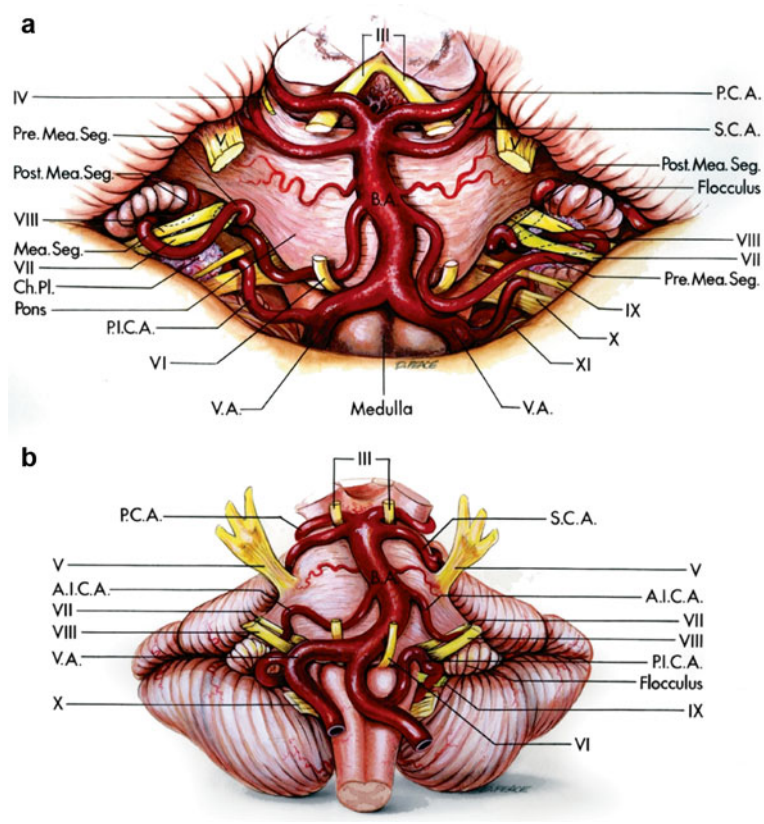


Fig. 2.13 Vascular compression of CN VII. Sites of arterial compression of CN *VII* in hemifacial spasm. (**a**) Anterosuperior view. CNs *VII* and *VIII* are distorted at their junction with the brainstem by the right premeatal and the left postmeatal segments of the AICAs. (**b**) Anterior view. The junction of the right CNs VII and VIII with the brainstem is compressed by a tortuous vertebral artery. The nerves on the left side are compressed by the PICA (Reprint with permission from Martin et al. (1980)). *A.I.C.A.* anterior inferior cerebellar artery, *B.A.* basilar artery, *Ch. Pl.* choroid plexus, *Mea. Seg.* meatal segment, *P.C.A.* posterior cerebral artery, *P.I.C.A.* posterior inferior cerebellar artery, *S.C.A.* superior cerebral artery, *V.A.* vertebral artery. *III* oculomotor nerve, *IV* trochlear nerve, *V* trigeminal nerve, *VI* abducent nerve, *IX* glossopharyngeal nerve, *X* vagus nerve, *XI* accessory nerve

It is expected that the AICA would be the compressing vessel in most cases because CN VII is located in the middle neurovascular complex, yet a tortuous PICA has also been reported to be a frequent compressing artery in hemifacial spasm (Barker et al. 1995). In some CPAs, the proximal part of the PICA, after passing posterior to the hypoglossal rootlets, will loop superiorly toward CNs VII and VIII before descending to pass between CNs IX and XI. The offending arterial loop usually compresses the caudal aspect of CN VII and uncommonly the superior aspect. In some cases, more than one artery will compress the nerve at its exit from the brainstem. The AICA will compress the medial surface of the nerve and the PICA will compress the caudal surface. In cases where a tortuous vertebral artery loops laterally, it may compress CN VII at the brainstem (Guan et al. 2011). The vertebral artery is more often associated with hemifacial spasm as an indirect compressing vessel involving another intervening vessel in tandem fashion and not by direct compression by the vertebral artery (Kim et al. 2008; Mikami et al. 2013). Mobilization of the indirect offending vertebral artery is often required before decompression of the direct compressing vessel. Care is taken to explore the interval between CNs VII and VIII because a vessel compressing CN VII located between the nerves may be missed. Surrounding veins including the vein of the pontomedullary sulcus or the middle cerebral peduncle have also been found to compress CN VII (Fig. 2.4a, c) (Barker et al. 1995; Campos-Benitez and Kaufmann 2008; Huang et al. 1992; Hyun et al. 2010; Park et al. 2008).

2.4 Lower Neurovascular Complex and Glossopharyngeal Neuralgia

The lower neurovascular complex includes the PICA, medulla, inferior cerebellar peduncle, cerebellomedullary fissure, suboccipital surface of

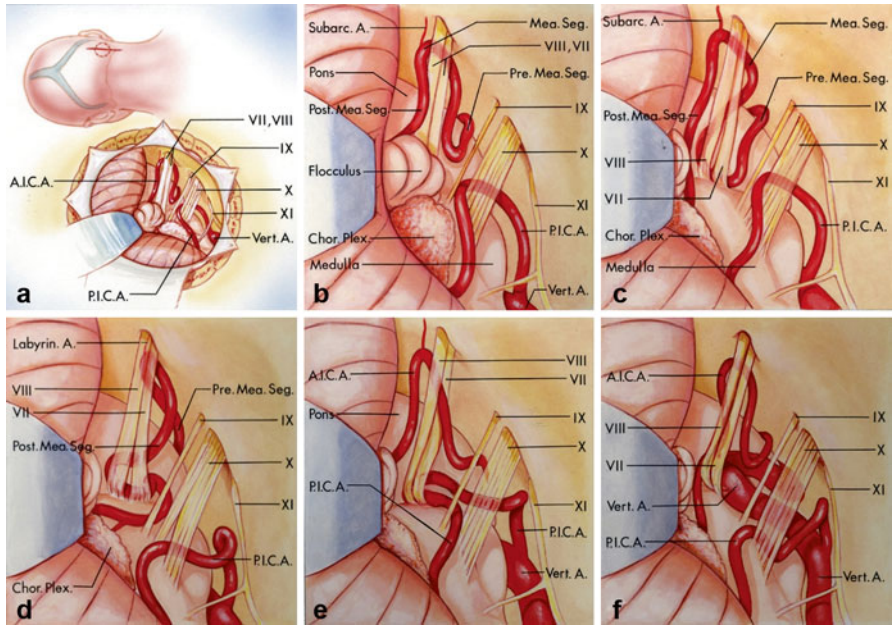


Fig. 2.14 Vascular compression of CN VII through right infraflocculus exposure. Arterial compression of CN VII in hemifacial spasm as viewed through a retrosigmoid craniotomy performed with the patient in the three-quarter prone position. (**a**) The *upper drawing* shows the site of the incision (*straight line*) and the location of the craniotomy for the infraflocculus approach to CN VII (*broken line*). The *lower drawing* shows the surgical exposure obtained with this approach. The AICA and CNs VII and VIII are in the midportion of the exposure. The vertebral artery, PICA, and CNs IX–XI are below. (**b**) The cerebellum is elevated to expose CNs VII and VIII and the premeatal, meatal, and postmeatal segments of the AICA. The flocculus and the choroid plexus block the view of the junction of CNs VII and VIII with the brainstem. (**c**) The flocculus and choroid plexus have been elevated in the infraflocculus approach to the junction of CNs VII and VIII with the brainstem. The premeatal segment compresses the nerves at the junction with the pons and the medulla. (**d**) The junction of CN VII and VIII with the brainstem is compressed by the postmeatal segment. (**e**) A tortuous PICA loops upward to compress the nerves at their junction with the brainstem before turning inferiorly to pass between CNs IX and X. (**f**) A tortuous vertebral artery compresses the junction of the nerves with the brainstem (Reprint with permission from Rhoton (2000b)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Chor. Plex*. choroid plexus, *Labyrin*. labyrinthine, *Mea. Seg*. meatal segment, *P.I.C.A*. posterior inferior cerebellar artery, *Subarc*. subarculate, *Vert*. vertebral

the cerebellum, and CNs IX–XII. The PICA arises at the medullary level and encircles the medulla, passing through the cerebellomedullary cistern with CNs IX–XII to reach the surface of the inferior cerebellar peduncle, where it dips into the cerebellomedullary fissure and terminates by supplying the suboccipital surface of the cerebellum (Rhoton 2013).

2.4.1 Anatomy of CNs IX and X

CNs IX–XI arise as a line of rootlets that exit the medulla along the posterior edge of the olive in the postolivary sulcus, a shallow groove between the olive and the posterolateral surface of the medulla (Fig. 2.15a) (Rhoton 2000d). CN IX arises as one or rarely two rootlets just caudal to the origin of CN VII and ventral to the rhomboid lip of the lateral recess and the choroid plexus protruding from the foramen of Luschka. In cases of two rootlets, a larger dorsal and a smaller ventral component can be seen at the junction with the brainstem (Katsuta et al. 1997; Rhoton and Buza 1975). The smaller ventral rootlets have been demonstrated to be motor and the larger main bundle to be sensory (DuBois and Foley 1936; Tarlov 1937). CN X arises cau-

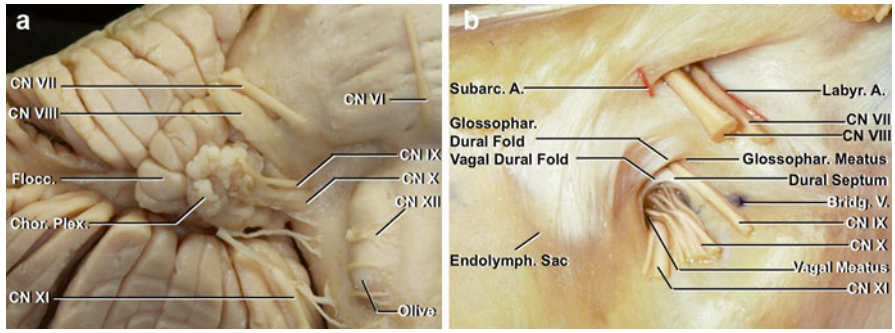
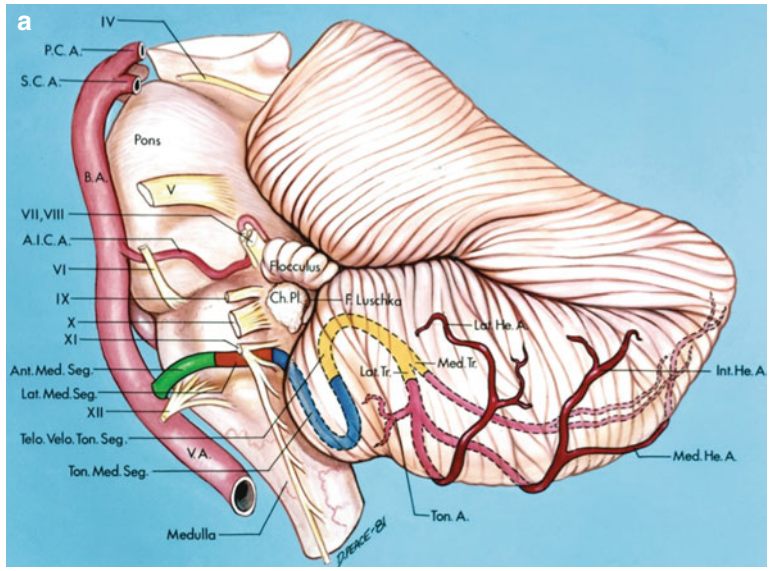


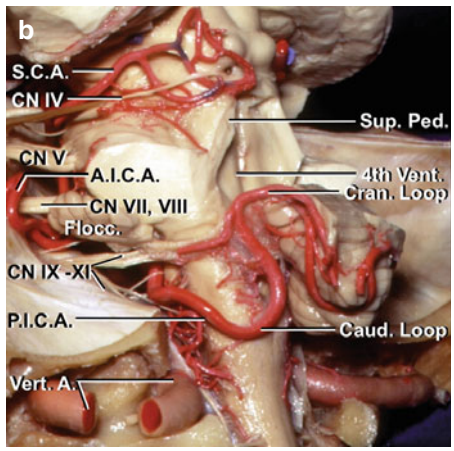
Fig. 2.15 CNs IX and X. (**a**) Anterior view of the cerebellum. CNs IX–XI arise as a line of rootlets that exit the medulla along the posterior edge of the olive in the postolivary sulcus, and CN XII arises anterior to the olive. CN IX arises as one or rarely two rootlets just caudal to the origin of CNs VII and VIII and ventral to the rhomboid lip of the lateral recess and the choroid plexus protruding from the foramen of Luschka. CN X arises inferior to CN IX as a line of tight combinations of large and small rootlets. (**b**) Posterior view of the intracranial aspect of the left jugular foramen. CN IX penetrates the dura covering the jugular foramen below and medial to the glossopharyngeal dural fold. CN X enters the foramen with CN XI below and medial to the vagal dural fold (Reprint with permission from Rhoton (2000d)). *A*. artery, *Bridg*. bridging, *Chor. Plex*. choroid plexus, *CN* cranial nerve, *Endolymph*. endolymphatic, *Flocc*. flocculus, *Glossophar*. glossopharyngeal, *Labyr*. labyrinthine, *Subarc*. subarcuate, *V*. vein

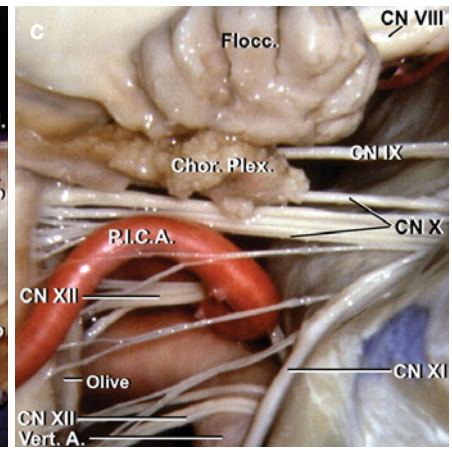
dal to CN IX as a line of tightly packed rootlets posterior to the superior one-third of the olive. The most rostral vagal fibers may be separated from the glossopharyngeal origin by as much as 2 mm. CN X is composed of multiple combinations of large and small rootlets. Several small rootlets, considered to be motor, are occasionally found originating ventral to the majority of the vagal rootlets (DuBois and Foley 1936). CNs IX and X arise rostral to the level of the origin of CN XII, anterior to the olive. CN XI arises as a widely separated series of rootlets that originate from the medulla at the level of the lower two-thirds of the olive and from the upper cervical cord. The cranial rootlets of CN XI arising just caudal to CN X are more properly regarded as inferior vagal rootlets since they arise from vagal nuclei. It may be difficult to separate the lower vagal fibers from the upper accessory rootlets because the vagal and cranial accessory fibers usually enter the vagal meatus as a single bundle. CN IX penetrates the dura of the roof of the jugular foramen below and medial to the glossopharyngeal dural fold, and CN X and XI enter the dura below and medial to the vagal dural fold (Fig. 2.15b). The superior glossopharyngeal and vagal ganglia may be visible intracranially (Katsuta et al. 1997). The superior glossopharyngeal ganglion was intracranial in 32 % of the 50 jugular foramina in our previous study and within or extracranial to the foramen in 68 % (Rhoton and Buza 1975). The superior ganglion of the vagus nerve could be seen intracranially in only 14 % of cases.

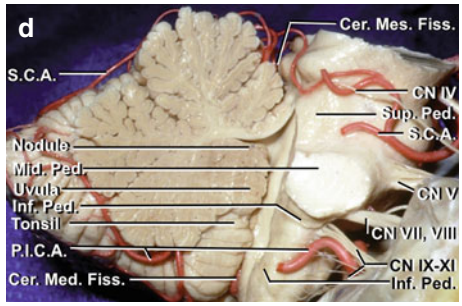
2.4.2 Anatomy of the PICA

The PICA, by definition, arises from the vertebral artery (Figs. 2.16 and 2.17). It arises near the inferior olive and passes posteriorly around the medulla. At the anterolateral margin of the medulla, it passes rostral or caudal to or between the rootlets of CN XII, and at the posterolateral margin of the medulla, it courses rostral to or between the fila of CNs IX–XI. After passing these nerves, it courses near the lateral recess to enter the cerebellomedullary fissure and then caudally to make a caudal loop anterior to the tonsil before ascending along the lower half of the roof of the fourth ventricle to form a convex rostral curve, called the cranial loop. It exits the fissure between the vermis, tonsil, hemisphere, and medulla to reach the suboccipital cerebellar


a
IV
P.C.A.
S.C.A.
Pons
B.A.
V
VII, VIII
A.I.C.A.
VI
IX
X
XI
Ant. Med. Seg.
Lat. Med. Seg.
XII
Telo. Velo. Ton. Seg.
V.A.
Ton. Med. Seg.
Medulla
Flocculus
Ch. Pl.
F. Luschka
Lat. He. A.
Lat. Tr.
Med. Tr.
Int. He. A.
Med. He. A.
Ton. A.


b
S.C.A.
CN IV
Sup. Ped.
CN V
A.I.C.A.
4th Vent.
Cran. Loop
CN VII, VIII
Flocc.
CN IX -XI
P.I.C.A.
Caud. Loop
Vert. A.


c
Flocc.
CN VIII
Chor. Plex.
CN IX
CN X
P.I.C.A.
CN XII
CN XI
Olive
CN XII
Vert. A.


d
Cer. Mes. Fiss.
S.C.A.
CN IV
Sup. Ped.
S.C.A.
Nodule
Mid. Ped.
Uvula
CN V
Inf. Ped.
Tonsil
CN VII, VIII
P.I.C.A.
CN IX-XI
Cer. Med. Fiss.
Inf. Ped.

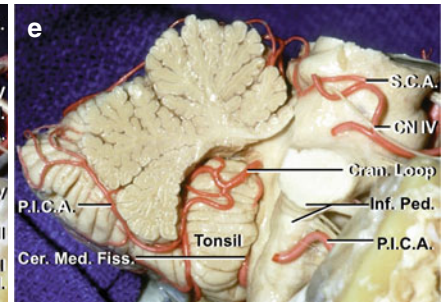

e
S.C.A.
CN IV
Cran. Loop
P.I.C.A.
Inf. Ped.
Tonsil
P.I.C.A.
Cer. Med. Fiss.

Fig. 2.16 Posterior inferior cerebellar artery. (**a**) Lateral view. The PICA arises from the vertebral artery near the inferior olive (anterior medullary segment: *green*) and passes posteriorly around the medulla, passing rostral or caudal to or between the rootlets of CN XII, and rostral to or between the fila of CNs IX–XI (lateral medullary segment: *orange*). After passing these nerves, it enters the cerebellomedullary fissure and descends to make a caudal loop around the tonsil (tonsillomedullary segment: *blue*) and ascends posterior to the lower half of the roof of the fourth ventricle to make a cranial loop (telovelotonsillar segment: *yellow*). Distal to the cranial loop, it exits the fissure between the vermis, tonsil, and hemisphere to reach the suboccipital cerebellar surface (cortical segment: *red*). Most PICAs bifurcate into a medial and a lateral trunk upon exiting the cerebellomedullary fissure (Reprint with permission from Lister et al. (1982)). (**b**) Left posterolateral view. The cerebellum was removed by dividing the cerebellar peduncles. The PICA arises anterior to the inferior olive and courses between CNs X and XI and along the posterolateral medulla and inferior cerebellar peduncle. It forms a caudal loop below the tonsil (Reprint with permission from Rhoton (2000a)). (**c**) Posterior view. The PICA passes posteriorly between the rootlets of CN XII and enters the cerebellomedullary fissure (Reprint with permission from Rhoton (2000a)). (**d**) Lateral view. The right half of the cerebellum has been removed. The right PICA passes between the rootlets of CN X and XI to reach the surface of the inferior cerebellar peduncle. The left PICA, as it courses around the rostral pole of the tonsil, is hidden by the remaining left half of the uvula (Reprint with permission from Rhoton (2000a)). (**e**) The part of the uvula and nodule medial to the tonsil has been removed to expose the PICA's passage through the cerebellomedullary fissure and around the tonsil. The artery frequently forms a caudal loop at the lower margin of the tonsil and a cranial loop that wraps around the rostral pole of the tonsil (Reprint with permission from Rhoton (2000a)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Ant*. anterior, *B.A*. basilar artery, *Caud*. caudal, *Cer*. cerebello, *Ch. Pl. Chor. Plex*. choroid plexus, *CN* cranial nerve, *Cran*. cranial, *F*. foramen, *Fiss*. fissure, *Flocc*. flocculus, *He*. hemispheric, *Inf*. inferior, *Int*., internal, *Lat*. lateral, *Med*. medial, medullary, *Mes*. mesencephalic, *Mid*. middle, *P.C.A*. posterior cerebral artery, *P.I.C.A*. posterior inferior cerebellar artery, *Ped*. peduncle, *S.C.A*. superior cerebral artery, *Seg*. segment, *Sup*. superior, *Ton*. tonsillar, tonsillo, *Tr*. trunk, *V.A., Vert. A*. vertebral artery, *Vent*. ventricle

surface. Most PICAs bifurcate into a medial and a lateral trunk upon exiting the cerebellomedullary fissure. The medial trunk supplies the vermis and adjacent part of the hemisphere, and the lateral trunk supplies the cortical surface of the tonsil and the hemisphere. The PICA gives off perforating, choroidal, and cortical arteries. The cortical arteries are divided into vermian, tonsillar, and hemispheric groups.

2.4.3 Offending Vessels of Glossopharyngeal Neuralgia

The PICA, AICA, vertebral artery, veins around the lower CNs, or a combination of these vessels can cause glossopharyngeal neuralgia (Fig. 2.18) (Ferroli et al. 2009; Gaul et al. 2011; Jannetta 1980; Kandan et al. 2010; Kawashima et al. 2010; Kondo 1998; Laha and Jannetta 1977; Matsushima et al. 2000; Resnick et al. 1995; Sampson et al. 2004).

The PICA is most frequently found to be a cause of glossopharyngeal neuralgia. The site of origin of the PICA from the vertebral artery varies from below the foramen magnum to the vertebrobasilar junction. The PICA, however, may have a high origin and loop upward to indent CNs IX and often X prior to entering the cerebellomedullary fissure (Fig. 2.18) (Kawashima et al. 2010; Matsushima et al. 2000). Of the 42 PICAs found in 50 CPAs in our previous study, 16 passed between the root-

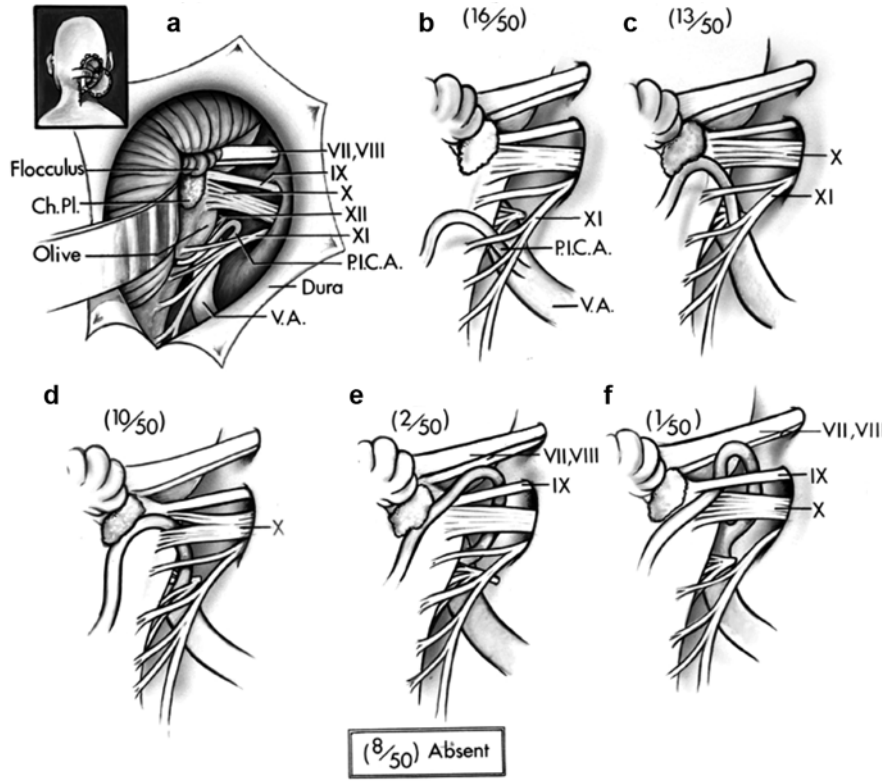


Fig. 2.17 Relationship of the PICA to the rootlets of CNs IX–XI. (**a**) Orientation of illustrations **b–f**. The *inset* shows the site of the scalp flap and the craniotomy. The large illustration shows the cerebellum elevated to expose CNs VII–XII. The PICA arises from the vertebral artery and passes inferior (**b, c**), superior (**e, f**), or between (**d**) the rootlets of CN XII. Of the 42 PICAs found in 50 cerebellar hemispheres in a previous study, 16 passed between the rootlets of CN XI (**b**), 13 passed between CNs X and XI (**c**), ten passed between the rootlets of CN X (**d**), two passed between CNs VIII and IX (**e**), and one passed between CNs IX and X (**f**) (Lister et al. 1982). A tortuous PICA may ascend anterior to CNs IX and X and compress and distort CNs VII and VIII before passing posteriorly between CNs IX–XI (Reprint with permission from Lister et al. (1982)). *Ch. Pl.* choroid plexus, *P.I.C.A.* posterior inferior cerebellar artery, *V.A.* vertebral artery

lets of CN XI, ten passed between the rootlets of CN X, 13 passed between CNs X and XI, two passed above CN IX below CN VIII, and one passed between CNs IX and X (Fig. 2.17) (Lister et al. 1982). The vertebral artery, even less frequently, may also reach and distort CNs IX and often X from the medial side, especially when it is tortuous, ectatic, and/or arteriosclerotic. CNs IX and often X may also be compressed by the AICA or a common trunk of the AICA and PICA. Surrounding veins, including a bridging vein to the jugular foramen and the vein of the pontomedullary sulcus, may also compress CNs IX and often X (Fig. 2.4a, c) (Ferroli et al. 2009; Jannetta 1980; Resnick et al. 1995; Sampson et al. 2004).

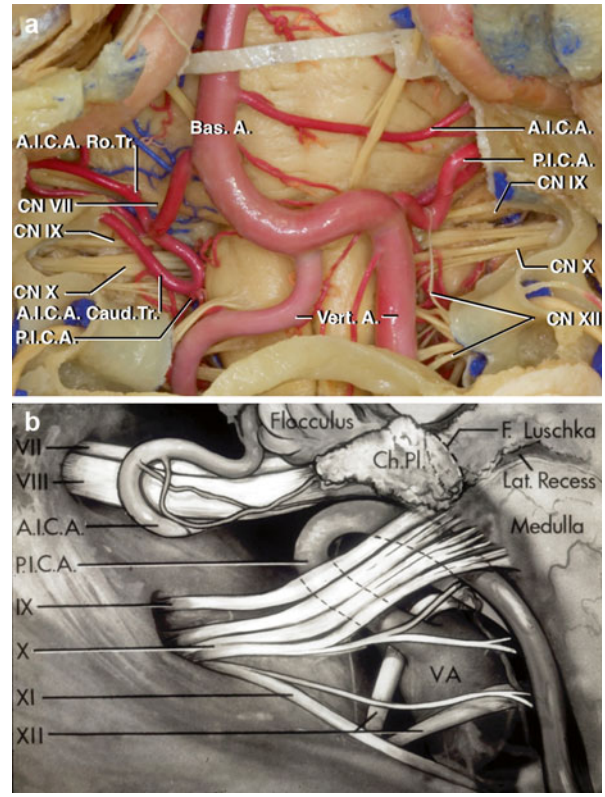


Fig. 2.18 Vascular compression of CN IX. (**a**) Anterior view after removing the clivus and adjacent part of the temporal and occipital bones. The left PICA stretches CN XII, after originating from the caudal part of the vertebral artery. It loops upward and compresses CN IX at its junction with the medulla, before passing posteriorly between the rootlets of CN X. A well-developed right AICA bifurcates in front of the exit of CN VII, and the caudal trunk makes a downward loop in front of CNs IX and X, compressing the nerves. The rostral trunk courses around CNs VII and VIII near the internal acoustic meatus. (**b**) The PICA arises from the vertebral artery, passes between the rootlets of CN XII, and loops superiorly, compressing CNs IX and X before passing posteroinferiorly between the rootlets of CNs X and XI. The vertebral artery stretches the rootlets of CN XII posteriorly. The AICA loops posterior to CNs VII and VIII (Reprint with permission from Lister et al. (1982)). *A*. artery, *A.I.C.A*. anterior inferior cerebellar artery, *Bas*. basilar, *Caud*. caudal, *Ch. Pl*. choroid plexus, *CN* cranial nerve, *F*. foramen, *Lat*. lateral, *P.I.C.A*. posterior inferior cerebellar artery, *Ro*. rostral, *Tr*. trunk, *VA* vertebral artery, *Vert*. vertebral

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References

Apfelbaum RI. Neurovascular decompression: the procedure of choice? Clin Neurosurg. 2000;46:473–98.

Barker 2nd FG, Jannetta PJ, Bissonette DJ, Shields PT, Larkins MV, Jho HD. Microvascular decompression for hemifacial spasm. J Neurosurg. 1995;82:201–10.

Barker 2nd FG, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med. 1996;334:1077–83.

Campos-Benitez M, Kaufmann AM. Neurovascular compression findings in hemifacial spasm. J Neurosurg. 2008;109:416–20.

DuBois FS, Foley JO. Experimental studies on the vagus and spinal accessory nerves in the cat. Anat Rec. 1936;64:285–307.

Emmons WF, Rhoton Jr AL. Subdivision of the trigeminal sensory root. Experimental study in the monkey. J Neurosurg. 1971;35:585–91.

Ferroli P, Fioravanti A, Schiariti M, Tringali G, Franzini A, Calbucci F, et al. Microvascular decompression for glossopharyngeal neuralgia: a long-term retrospectic review of the Milan-Bologna experience in 31 consecutive cases. Acta Neurochir (Wien). 2009;151:1245–50.

Fujimaki T, Kirino T. Combined transhorizontal-supracerebellar approach for microvascular decompression of trigeminal neuralgia. Br J Neurosurg. 2000;14:531–4.

Funaki T, Matsushima T, Masuoka J, Nakahara Y, Takase Y, Kawashima M. Adhesion of rhomboid lip to lower cranial nerves as special consideration in microvascular decompression for hemifacial spasm: report of two cases. Surg Neurol Int. 2010;1:71.

Gardner WJ. Cross talk-the paradoxical transmission of a nerve impulse. Arch Neurol. 1966;14:149–56.

Gaul C, Hastreiter P, Duncker A, Naraghi R. Diagnosis and neurosurgical treatment of glossopharyngeal neuralgia: clinical findings and 3-D visualization of neurovascular compression in 19 consecutive patients. J Headache Pain. 2011;12:527–34.

Guan HX, Zhu J, Zhong J. Correlation between idiopathic hemifacial spasm and the MRI characteristics of the vertebral artery. J Clin Neurosci. 2011;18: 528–30.

Gudmundsson K, Rhoton Jr AL, Rushton JG. Detailed anatomy of the intracranial portion of the trigeminal nerve. J Neurosurg. 1971;35:592–600.

Hardy DG, Rhoton Jr AL. Microsurgical relationships of the superior cerebellar artery and the trigeminal nerve. J Neurosurg. 1978;49:669–78.

Hardy DG, Peace DA, Rhoton Jr AL. Microsurgical anatomy of the superior cerebellar artery. Neurosurgery. 1980;6:10–28.

Hitotsumatsu T, Matsushima T, Inoue T. Microvascular decompression for treatment of trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia: three surgical approach variations: technical note. Neurosurgery. 2003;53:1436–41; discussion 1442–33.

Hong W, Zheng X, Wu Z, Li X, Wang X, Li Y, et al. Clinical features and surgical treatment of trigeminal neuralgia caused solely by venous compression. Acta Neurochir (Wien). 2011;153:1037–42.

Huang CI, Chen IH, Lee LS. Microvascular decompression for hemifacial spasm: analyses of operative findings and results in 310 patients. Neurosurgery. 1992;30:53–6; discussion 56–7.

Hyun SJ, Kong DS, Park K. Microvascular decompression for treating hemifacial spasm: lessons learned from a prospective study of 1,174 operations. Neurosurg Rev. 2010;33:325–34; discussion 334.

Jannetta PJ. Neurovascular compression in cranial nerve and systemic disease. Ann Surg. 1980;192:518–25.

Kandan SR, Khan S, Jeyaretna DS, Lhatoo S, Patel NK, Coakham HB. Neuralgia of the glossopharyngeal and vagal nerves: long-term outcome following surgical treatment and literature review. Br J Neurosurg. 2010;24:441–6.

Katsuta T, Rhoton Jr AL, Matsushima T. The jugular foramen: microsurgical anatomy and operative approaches. Neurosurgery. 1997;41:149–201; discussion 201–2.

Kawashima M, Matsushima T, Inoue T, Mineta T, Masuoka J, Hirakawa N. Microvascular decompression for glossopharyngeal neuralgia through the transcondylar fossa (supracondylar transjugular tubercle) approach. Neurosurgery. 2010;66:275–80; discussion 280.

Kim JP, Park BJ, Choi SK, Rhee BA, Lim YJ. Microvascular decompression for hemifacial spasm associated with vertebrobasilar artery. J Korean Neurosurg Soc. 2008;44:131–5.

Kondo A. Follow-up results of using microvascular decompression for treatment of glossopharyngeal neuralgia. J Neurosurg. 1998;88:221–5.

Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg. 1977;47:316–20.

Li ST, Pan Q, Liu N, Shen F, Liu Z, Guan Y. Trigeminal neuralgia: what are the important factors for good operative outcomes with microvascular decompression. Surg Neurol. 2004;62:400–4; discussion 404–5.

Lister JR, Rhoton Jr AL, Matsushima T, Peace DA. Microsurgical anatomy of the posterior inferior cerebellar artery. Neurosurgery. 1982;10:170–99.

Lorenzoni JG, Massager N, David P, Devriendt D, Desmedt F, Brotchi J, et al. Neurovascular compression anatomy and pain outcome in patients with classic trigeminal neuralgia treated by radiosurgery. Neurosurgery. 2008;62:368–75; discussion 375–6.

Martin RG, Grant JL, Peace D, Theiss C, Rhoton Jr AL. Microsurgical relationships of the anterior inferior cerebellar artery and the facial-vestibulocochlear nerve complex. Neurosurgery. 1980;6:483–507.

Matsushima T, Rhoton Jr AL, Lenkey C. Microsurgery of the fourth ventricle: part 1. Microsurgical anatomy. Neurosurgery. 1982;11:631–67.

Matsushima T, Rhoton Jr AL, de Oliveira E, Peace D. Microsurgical anatomy of the veins of the posterior fossa. J Neurosurg. 1983;59:63–105.

Matsushima T, Fukui M, Suzuki S, Rhoton Jr AL. The microsurgical anatomy of the infratentorial lateral supracerebellar approach to the trigeminal nerve for tic douloureux. Neurosurgery. 1989;24:890–5.

Matsushima T, Inoue T, Fukui M. Arteries in contact with the cisternal portion of the facial nerve in autopsy cases: microsurgical anatomy for neurovascular decompression surgery of hemifacial spasm. Surg Neurol. 1990;34:87–93.

Matsushima T, Goto Y, Natori Y, Matsukado K, Fukui M. Surgical treatment of glossopharyngeal neuralgia as vascular compression syndrome via transcondylar fossa (supracondylar transjugular tubercle) approach. Acta Neurochir (Wien). 2000;142:1359–63.

Matsushima T, Huynh-Le P, Miyazono M. Trigeminal neuralgia caused by venous compression. Neurosurgery. 2004;55:334–7; discussion 338–9.

Matsushima K, Matsushima T, Kuga Y, Kodama Y, Inoue K, Ohnishi H, et al. Classification of the superior petrosal veins and sinus based on drainage pattern. Neurosurgery. 2014;10 Suppl 2:357–67; discussion 367.

Mikami T, Minamida Y, Akiyama Y, Wanibuchi M, Sugino T, Houkin K, et al. Microvascular decompression for hemifacial spasm associated with the vertebral artery. Neurosurg Rev. 2013;36:303–8; discussion 308–9.

Moller AR, Jannetta PJ. On the origin of synkinesis in hemifacial spasm: results of intracranial recordings. J Neurosurg. 1984;61:569–76.

Nakahara Y, Matsushima T, Hiraishi T, Takao T, Funaki T, Masuoka J, et al. Importance of awareness of the rhomboid lip in microvascular decompression surgery for hemifacial spasm. J Neurosurg. 2013;119:1038–42.

Park JS, Kong DS, Lee JA, Park K. Hemifacial spasm: neurovascular compressive patterns and surgical significance. Acta Neurochir (Wien). 2008;150:235–41; discussion 241.

Resnick DK, Jannetta PJ, Bissonnette D, Jho HD, Lanzino G. Microvascular decompression for glossopharyngeal neuralgia. Neurosurgery. 1995;36:64–8; discussion 68–9.

Rhoton Jr AL. Microsurgery of the internal acoustic meatus. Surg Neurol. 1974;2:311–8.

Rhoton Jr AL. Microsurgical anatomy of decompression operations on the trigeminal nerve. In: Rovit RL, Murali R, Jannetta PJ, editors. Trigeminal neuralgia. Baltimore: Williams and Wilkins; 1990. p. 165–200.

Rhoton Jr AL. The cerebellar arteries. Neurosurgery. 2000a;47(3 Suppl):S29–68.

Rhoton Jr AL. The cerebellopontine angle and posterior fossa cranial nerves by the retrosigmoid approach. Neurosurgery. 2000b;47:S93–129.

Rhoton Jr AL. Cerebellum and fourth ventricle. Neurosurgery. 2000c;47(3 Suppl):S7–27.

Rhoton Jr AL. Jugular foramen. Neurosurgery. 2000d;47(3 Suppl):S267–85.

Rhoton Jr AL. The posterior fossa veins. Neurosurgery. 2000e;47(3 Suppl):S69–92.

Rhoton Jr AL. The temporal bone and transtemporal approaches. Neurosurgery. 2000f;47(3 Suppl):S211–65.

Rhoton Jr AL. Cerebellopontine angle and retrosigmoid approach. Neurosurgery. 2007;61(4 Suppl):S175–92.

Rhoton Jr AL. Microsurgical anatomy of the cerebellopontine angle by the retrosigmoid approach. In: Samii M, Gerganov V, editors. Surgery of cerebellopontine lesions. Berlin: Springer; 2013. p. 9–72.

Rhoton Jr AL, Buza R. Microsurgical anatomy of the jugular foramen. J Neurosurg. 1975;42:541–50.

Rhoton Jr AL, Tedeschi H. Microsurgical anatomy of acoustic neuroma. Otolaryngol Clin North Am. 1992;25(2):257–94.

Sampson JH, Grossi PM, Asaoka K, Fukushima T. Microvascular decompression for glossopharyngeal neuralgia: long-term effectiveness and complication avoidance. Neurosurgery. 2004;54:884–9; discussion 889–90.

Sekula RF, Frederickson AM, Jannetta PJ, Bhatia S, Quigley MR, Abdel Aziz KM. Microvascular decompression in patients with isolated maxillary division trigeminal neuralgia, with particular attention to venous pathology. Neurosurg Focus. 2009;27:E10.

Sindou M, Leston JM, Decullier E, Chapuis F. Microvascular decompression for trigeminal neuralgia: the importance of a noncompressive technique--Kaplan-Meier analysis in a consecutive series of 330 patients. Neurosurgery. 2008;63:341–50; discussion 350–1.

Sunderland S. Neurovascular relations and anomalies at the base of the brain. J Neurol Neurosurg Psychiatry. 1948;11:243–57.

Tarlov I. Structure of the nerve root. II. Differentiation of sensory from motor roots; observations on identification of function in roots of mixed cranial nerves. Arch Neurol Psychiatry. 1937;37:1338–55.

Thomas KL, Vilensky JA. The anatomy of vascular compression in trigeminal neuralgia. Clin Anat. 2014;27:89–93.

Tomii M, Onoue H, Yasue M, Tokudome S, Abe T. Microscopic measurement of the facial nerve root exit zone from central glial myelin to peripheral Schwann cell myelin. J Neurosurg. 2003;99:121–4.

Yang XS, Li ST, Zhong J, Zhu J, Du Q, Zhou QM, et al. Microvascular decompression on patients with trigeminal neuralgia caused by ectatic vertebrobasilar artery complex: technique notes. Acta Neurochir (Wien). 2012;154:793–7; discussion 797.

Pathogenesis and Treatment of Hemifacial Spasm

3

Aage R. Møller

Abstract

Hemifacial spasm can be cured by microvascular decompression (MVD) operations of the root exit zone of the facial nerve. This fact was the basis for the ("ephaptic") hypothesis stating that the anatomical location of the pathology that generates the signs of HFS, spasm in the mimic muscle on one side of the face and synkinesis, was the root exit zone of the facial nerve. However, later intracranial recording from the facial nerve provided strong experimental support of a different hypothesis about the pathology of HFS, namely, that the anatomical location of the pathology is the facial motonucleus.

Intraoperative measurements of neural conduction times provided evidence against the "ephaptic" hypothesis and showed evidence that hyperactivity of the facial motonucleus could explain the symptoms of HFS. Studies of the blink reflex supported the hypothesis that the facial motonucleus is hyperactive in people with HFS. The results of animal experiments showed that signs of HFS could be caused by facial motonucleus hyperactivity.

It was hypothesized that the abnormalities in the facial motonucleus in HFS were caused by activation of maladaptive neuroplasticity that was activated by the irritation of the root of the facial nerve by a blood vessel. These findings were supported by the results of animal studies.

Since a similar close contact with a blood vessel is present in at least 50 % of individuals who do not have any symptoms of spasm, it was concluded that a second factor in addition to vascular contact with the facial nerve root must be present in order to create the signs of HFS.

MVD operations have a success rate of over 85 %, and when combined with monitoring of the abnormal muscle contraction, success rates of 97 % have been reported. No other treatment has been shown to have noticeable success in relieving the signs of HFS.

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Keywords
Hemifacial spasm • Vascular compression • Neuroplasticity • Facial nucleus • Hyperactivity

3.1 Introduction

Hemifacial spasm (HFS) is a rare disease (incidence: 0.74 per 100,000 in white men and 0.81 per 100,000 in white women; (Auger 1979)) that has its onset relatively late in life. HFS is characterized by episodes of spasm separated by periods of essentially normal function of the mimic musculature (Ehni and Woltman 1945). The disorder can be cured by moving a blood vessel (artery or vein) off of the intracranial portion of the facial nerve using a surgical technique known as microvascular decompression (MVD) (Jannetta 1970).

Microvascular decompression (MVD) operations have been shown to be effective in treating hemifacial spasm (HFS), trigeminal neuralgia (tic douloureux) (TGN), and glossopharyngeal neuralgia (GPN). Symptoms of some disorders associated the eighth cranial nerve, such as some forms of tinnitus and disabling positional vertigo (DPV), a balance disorder, can also be alleviated by MVD. In the following Chap. 8 in this book, we will discuss the forms of tinnitus and DPV that are treatable by MVD. Vascular contact with the hypoglossal nerve seems to be associated with a rare disorder, hemilingual spasm (Osburn et al. 2010). Some forms of spasmodic torticollis also seem to be associated with close contact between a blood vessel and a cranial nerve (CNXI) (Freckmann et al. 1981, 931; Nakai et al. 1989).

In this chapter, we will discuss HFS and its treatment with MVD of the intracranial portion of the facial nerve. For a review of history of the MVD operation, see Møller (1998).

Vascular contact with a cranial nerve is common, occurring in at least 50 % of people without causing any symptoms (Sunderland 1948). This means that symptoms of disease only occur when another factor (pathology) is present. This other factor is unknown (Møller and Jannetta 1984), but since it does not seem to cause symptoms on its own and symptoms only occur when vascular contact is also present, the disease can be cured by eliminating the vascular contact. This explains why MVD operations are effective in eliminating the symptoms of diseases such HFS, TGN, and GPN.

3.2 Results of MVD Operations for HFS

Large studies have shown that the cure rate for MVD operations for HFS (Barker et al. 1995) is commonly 85 %. This is similar to MVD operations for TGN (Barker et al. 1996) and GPN (Laha and Jannetta 1977) and probably some forms of spasmodic torticollis (Nagata et al. 1989). Intermediate nerve neuralgia (geniculate neuralgia) can also be cured by MVD operations (Tang et al. 2014). (Treatment of vascular compression disorders of the eighth cranial nerves is discussed in Chaper 8 in this book.)

A study of 1174 operations with 1 year or more follow-up (Hyun et al. 2010) showed that at the 1-year follow-up of 1105 patients, 5.9 % had residual spasm, thus a cure rate of 94.1 %. This study also reported that transient hearing loss occurred in 31 patients (2.6 % of all the patients operated upon), permanent hearing loss in 13 (1.1 %), transient facial weakness in 86 (7.3 %), permanent facial weakness in nine (0.7 %), cerebrospinal fluid leak in three (0.25 %), and cerebellar infarction or hemorrhage in two (0.17 %).

The precise location of the vascular contact along the intracranial portion of the facial nerve occurs varies among people with HFS. De Ridder et al. (2002) showed evidence that vascular compression syndromes arise from vascular contact along the CNS segment of the cranial nerves. Campos-Benitez and Kaufmann (2008) studied 115 patients who were operated upon for HFS (Campos-Benitez and Kaufmann 2008). Thirty-eight percent of these patients had multiple vessels in contact with the facial nerve. When the

intracranial portion was examined, it was found that the prevalence of vascular contact along the cranial portion of the nerve was not distributed evenly. The root exit point was the primary offending area in 10 %, the attached segment in 64 %, the Obersteiner–Redlich zone (transition between central and peripheral axonal myelination) in 22 %, and the distal cisternal portion in 3 %. As judged during the operation, the severity of compression was mild in 27 %, moderate in 61 %, and severe in 12 % of patients. Failure to alleviate the HFS occurred in nine cases, but this failure was not related to compression location, severity, or vessel type. The vessels that were in close contact with the facial nerve were the anterior inferior cerebellar artery (in 43 %), posterior inferior cerebellar artery (in 31 %), vertebral artery (in 23 %), or a large vein (in 3 %).

3.3 Symptoms and Signs of Hemifacial Spasm

HFS is characterized by periods of spasm engaging only the mimic muscles of one side of the face. In some people with HFS, more or less pronounced degrees of synkinesis are present. The signs typically develop gradually, beginning with brief contractions of the muscles around the eye. The condition progresses over several years. The intensity of the spasm around the eye increases and spreads to other mimic muscles lower down on the face. Typically, after many years (10–15), most mimic muscles have been recruited, including the platysma, though the muscles of the forehead are excluded as they have a different innervation pattern (bilateral facial nerve innervation).

In addition to the episodes of spasm and synkinesis, HFS is characterized by an abnormal muscle response (AMR) which can be elicited from mimic muscles by electrical stimulation of a branch of the facial nerve (Esslen 1957; Nielsen 1984a; Møller and Jannetta 1984, 1985; Møller 1993). This AMR can be demonstrated by EMG recordings from mimic muscles that are innervated by a different branch of the facial nerve. The initial component of the AMR has a latency of approximately 10 ms when, for example, elicited from the temporal or zygomatic branch of the facial nerve and recorded from the mentalis muscle (Fig. 3.1).

It is not known how specific the AMR is for HFS, but it can normally not be elicited in people who do not have HFS, and in HFS, it disappears after successful treatment with MVD. The AMR is also known as the "lateral spread response."

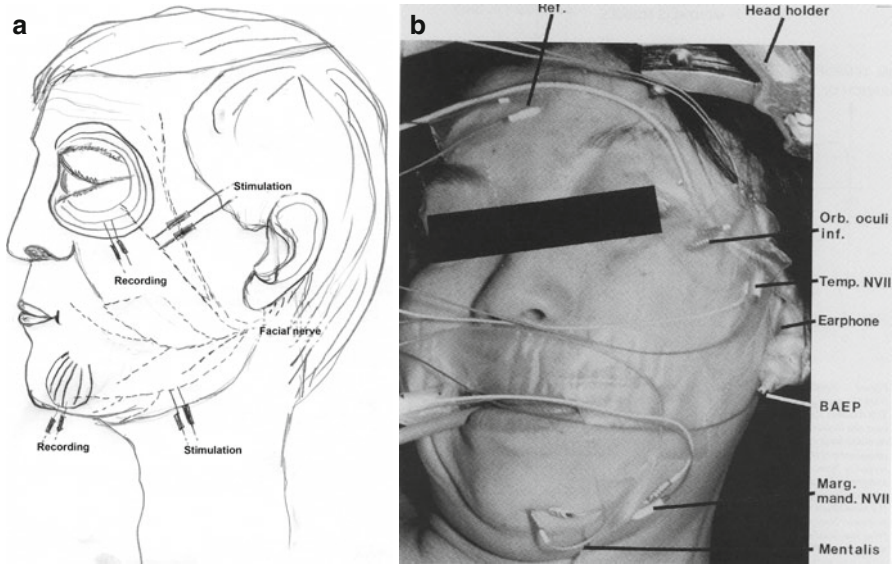

Fig. 3.1 (**a**) Electrode montage for recording the abnormal muscle response (Artwork by Zahra Akhavi). (**b**) Placement of needle electrodes for recording the AMR in a patient to undergo MVD for HFS

The AMR can normally be recorded under surgical anesthesia although the amplitude of the AMR will be slightly lower than when the patient was awake. The AMR has some similarities with the F-response. (The F-response can be elicited by electrical stimulation of a peripheral motor nerve (Møller and Jannetta 1987). The response is caused by firing of alpha motoneurons by antidromic activity in their motor nerves. Like the F-response, the AMR is assumed to be a measure of excitability of the facial motor nucleus.

The initial component of the AMR that can be recorded in response to electrical stimulation of a peripheral branch of the facial nerve is followed by EMG potentials that may last 100 ms or more (Fig. 3.2). The prolonged response reflects repetitive firings of motoneurons that produce repetitive muscle contractions (spasm).

The AMR usually persisted until the blood vessel that was in close contact with the facial nerve lifted off the facial nerve. In some patients, especially patients who had had their spasm for a short time only, the AMR decreased and even disappeared before the facial nerve was exposed (Møller and Jannetta 1985). When that happens, the AMR can almost always be activated again by stimulation of the facial nerve branch at a high rate (50 pps) for a short time (Fig. 3.3). This procedure sometimes has to be repeated in some MVD operations to maintain the AMR before the facial nerve is exposed.

In the example shown in Fig. 3.3, the amplitude of the AMR decreased before the facial nerve was exposed. Increasing the stimulus rate from 5 pp to 50 pp for a brief period brought the

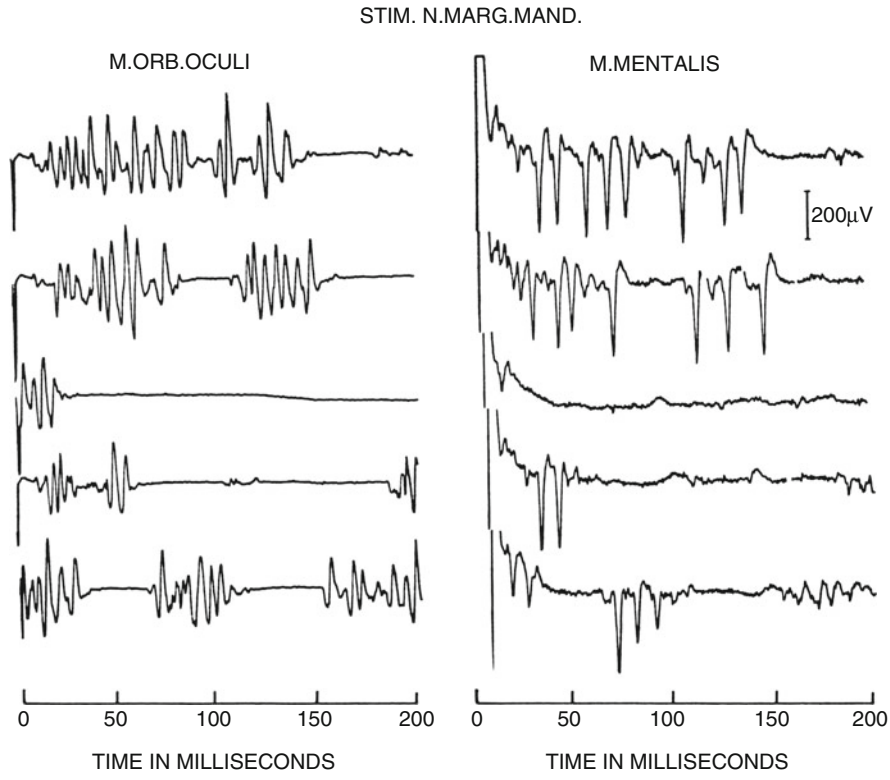

Fig. 3.2 Typical recordings of the abnormal muscle response in a patient with HFS who is to undergo an MVD operation. The response was recorded from the mentalis muscle and elicited by stimulation of the zygomatic branch of the facial nerve (Reprint with permission from Møller (1997); originally published in Møller and Jannetta (1985)

AMR back to the amplitude it had at the beginning of the operation.

Many patients have multiple vessels in contact with the facial nerve, but often times, only one vessel needs to be moved off the facial nerve in order to achieve complete relief from the spasm. A visual inspection is not sufficient; the only way to discern which vessels need to be moved is through intraoperative monitoring of the AMR. The AMR disappears when the blood vessel that is associated with the symptoms is moved off the intracranial portion of the facial nerve (Møller and Jannetta 1985) (Figs. 3.4 and 3.5). This principle has led to the development of a monitoring method that can help identify which vessel should be moved off the facial nerve to eliminate the symptoms (Møller and Jannetta 1987).

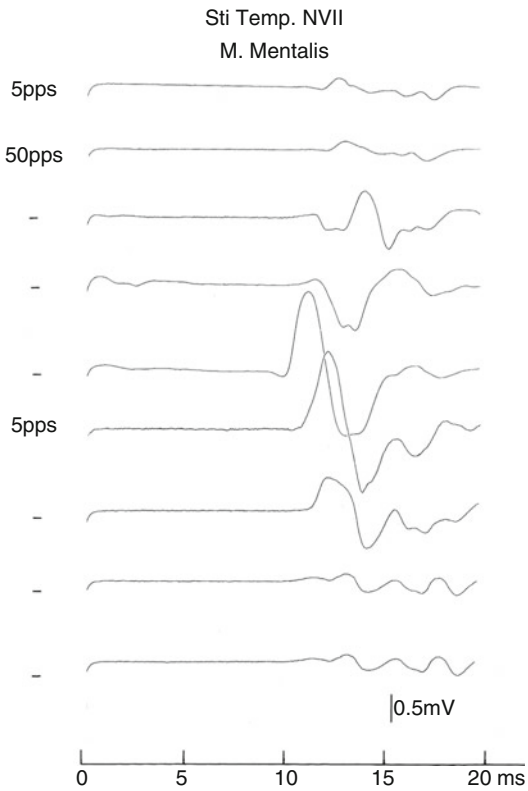

Fig. 3.3 Recordings of the AMR in a patient undergoing an MVD operation for HFS. Consecutive recordings (beginning at top) from the mentalis muscle in response to electrical stimulation of the temporal branch of the facial nerve are shown. The effect on the abnormal muscle response from increasing the stimulus rate from 5 to 50 pps for a short period of stimulation is shown in a patient in whom the abnormal muscle response was very small when recorded in the beginning of the operation (Reprint with permission from Møller (2011) Fig. 15.8; originally from Møller and Jannetta (1985))

The blink reflex (also known as the supraorbital nerve reflex (Nielsen 1984b)) elicited by stimulation of the supraorbital nerve is abnormal in people with HFS in that it engages several muscle groups including the mentalis muscle, while it normally only involves the orbicularis oculi muscles.

When a blink reflex response is elicited in people with HFS through electrical stimulation of the supraorbital nerve, the reflex response not only includes the muscles around the eye but also other muscles of the face (Kim and Fukushima 1984; Nielsen 1984b; Auger 1979). This spread of blink reflex activity to other parts of the face is a sign of the synkinesis of mimic muscles often observed in people with HFS. This abnormality has been named the "lateral spread response" (Nielsen 1984b).

Valls-Sole and Tolosa (1989) found that the R_1 component of the blink reflex response recorded from the mentalis or the orbicularis oris muscle has a slightly longer latency than the response from the orbicularis oculi in the normal blink reflex response (12.6 ± 1.3 ms). This prolongation of the R_1 component of the response recorded from muscles of the lower face could be explained by the fact that facial nerves travel a greater distance to the muscles of the lower face than to the muscles of the upper face. Nielsen reported similar results (Nielsen 1984b).

Despite having seemingly distinct and clear symptoms, hemifacial spasm is frequently reported to be misdiagnosed in the primary care setting (Martinez et al. 2014).

Spasm of mimic muscles that resembles hemifacial spasm occasionally occurs together with trigeminal neuralgia, known as tic convulsive (Yeh and Tew 1984).

An atypical form of HFS where the spasm progresses upward over the face has been described (Jannetta 1990).

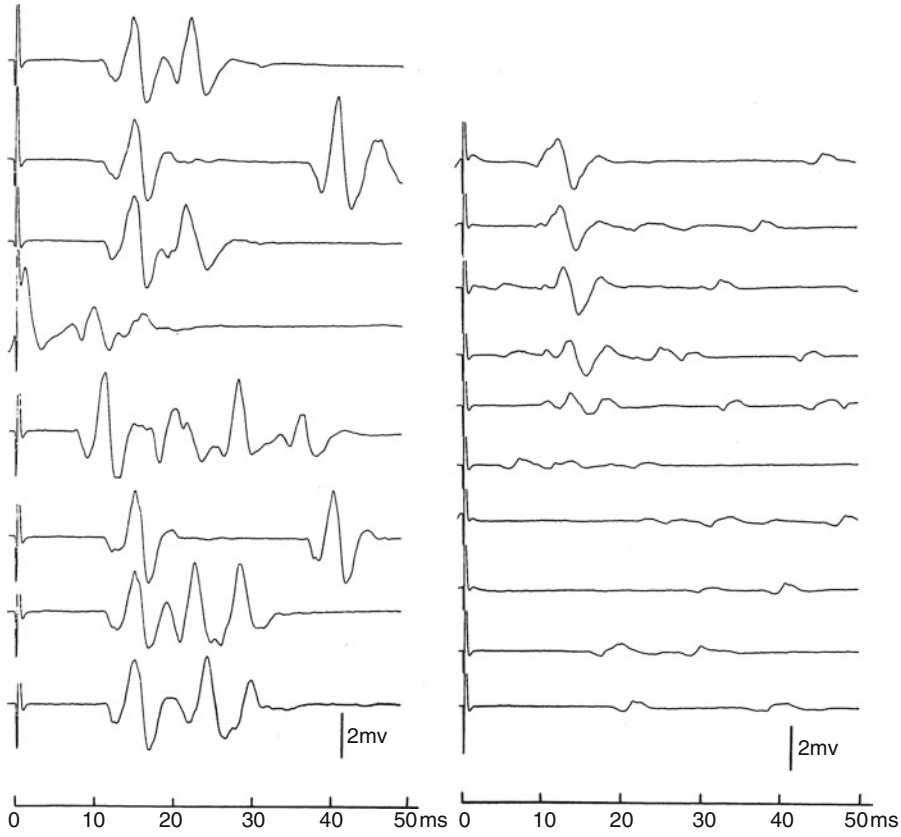

Fig. 3.4 AMR recorded from the mentalis muscle in response to consecutive stimulations of the zygomatic branch of the facial nerve in a patient undergoing an MVD operation for HFS. Then recordings started at the top of the left-hand column. A blood vessel was lifted off the root of the facial nerve at the time the top recording of the second column was done (Reprint with permission from Møller (2011), Fig. 15.9; originally from Møller and Jannetta (1985))

3.4 Pathophysiology of HFS

It was probably Dandy (1934) who first recognized and described how vascular conflict of cranial nerves could cause a specific disorder, in his case, trigeminal neuralgia (TGN). When Dandy (1929) sectioned the trigeminal nerve in the cerebellopontine angle (CPA) to treat patients with TGN, he observed vascular compression of the trigeminal nerve, and he later reported that he believed this compression to be the cause of tic douloureux (Dandy 1934). Cushing (1920) had earlier hypothesized that TGN could be caused by pressure from a tumor on the trigeminal root, but it was probably Taarnhøj (1952, 1956) who first described the beneficial effect of decompressing the trigeminal nerve root to treat trigeminal neuralgia. Much later, Gardner and Sava (1962) reported the presence of vascular compression of the seventh cranial nerve root in patients with HFS (Møller 1998).

Several theories have been presented explaining the pathology of those disorders that can be cured by moving a blood vessel off the intracranial portion of the fifth, seventh, and eighth cranial nerves (for a review, see Møller (1991). It was earlier believed that it was the pounding of an artery into a nerve root that injured the nerve and thereby caused the symptoms. It has also been assumed that arteries in close contact with a nerve root were important, but it is now known that veins (Wang et al. 2013) and very small arteries are important to move off the facial nerve to effectively treat HFS in some patients (Jannetta 1984).

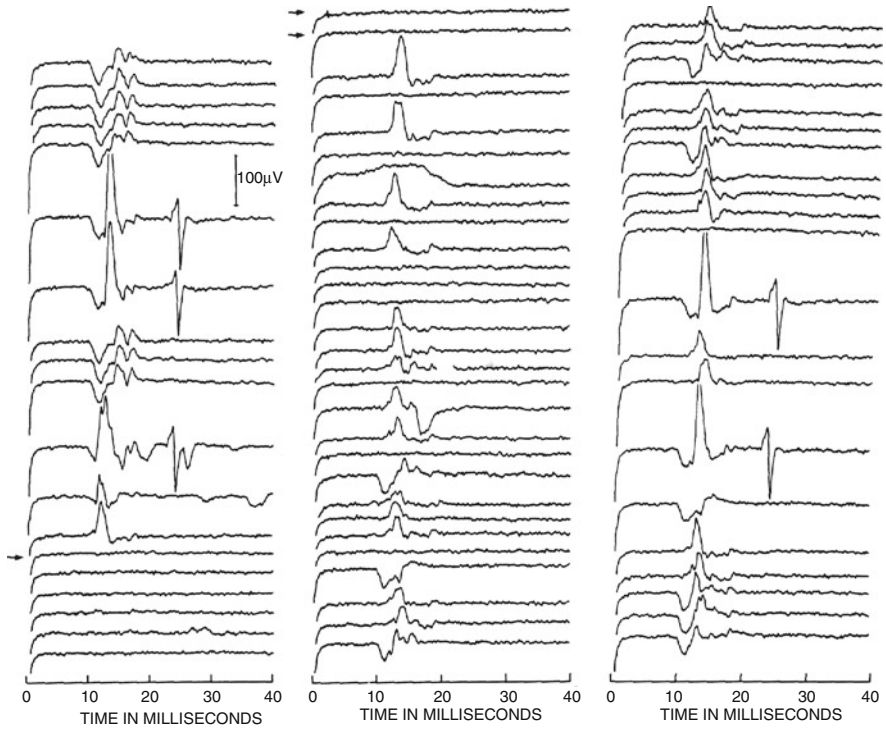

Fig. 3.5 Recordings of the AMR in a patient undergoing an MVD operation for HFS. The responses were recorded from the mentalis muscle in response to stimulation of the zygomatic branch of the facial nerve. The response to consecutive stimulation beginning at the top of the left-hand column, continuing from the top of the middle column. The *single arrow* near the bottom of the left-hand column indicates when a vessel was lifted off the root of the facial nerve. The *double arrows* at the top of the middle column indicate when the vessel was allowed to fall back on the nerve root (Reprint with permission from Møller (2011), Fig. 15.7; originally from Møller and Jannetta (1985))

It is also known that symptoms of HFS may be caused by close contact between a vessel and other parts of the intracranial portion of the facial nerve than the Obersteiner–Redlich zone (Kondo et al. 1980; Møller and Jannetta 1987).

Investigators in different fields of medicine have been fascinated by the pathology of diseases that are so effectively cured by MVD. One of the unanswered questions has been the anatomical location of the site of the pathology; another question has regarded an explanation for the fact that both HFS and TGN are very rare disorders, while vascular contact (compression) is commonly found in asymptomatic people.

Two hypotheses explaining the role of the close vascular contact have prevailed. One suggests that hyperactivity of the facial motonucleus is the cause of the spasm and synkinesis (Ferguson 1978). The other hypothesis states that cross talk (ephaptic transmission) between individual nerve axons of the facial nerve occurs (ephaptic communication) where it is in contact with a blood vessel causing ectopic excitation that could explain the signs of HFS (Esslen 1957; Williams et al. 1952; Gardner 1966; Gardner and Sava 1962; Nielsen 1984a).

That close contact between injured nerve fibers could cause direct communication ("cross talk") between axons of nerve was first suggested by Granit et al. (1944).

Ephaptic transmission seems a logical explanation of many different pathologies including the synkinesis in HFS. Ephaptic transmission, however, is a rare phenomenon (Granit et al. 1944) and has only been verified in very few instances. The ephaptic transmission hypothesis was nonetheless favored for many years as an explanation for HFS (Nielsen 1984a, b).

Ferguson (1978), however, was one of the first investigators to point out that ephaptic transmission between denuded facial nerve axons is not sufficient to explain the symptoms and signs of HFS. More recently, Esteban and Molina-Negro (1986) arrived at similar conclusions on the basis of preoperative studies of people with HFS. These investigators questioned whether the ephaptic transmission hypothesis could explain HFS because it seemed unlikely that the nerve fibers could be in contact with each other on a scale sufficient to cause the characteristically massive contraction of nearly all the facial muscles (Møller 1999).

Results of studies using electrophysiological recording during MVD operations of patients with HFS reported by Møller and Jannetta (1984) provided physiological evidence against the ephaptic hypothesis and in favor of the hyperactivity hypothesis.

That stimulation of one branch of the facial nerve produced EMG responses from not only the muscles that are innervated by the stimulated nerve branch but also from muscles innervated by other branches of the facial nerve indicates that there is an abnormal cross-transmission somewhere in the path of the antidromic facial motor nerve. The branch that is stimulated must come in contact with the branch that innervates the muscles from which the EMG potentials are recorded.

Studies using the recording of the AMR during MVD operations for HFS supported the hypothesis that the anatomical location of the physiological abnormalities in HFS that causes spasm and synkinesis is the facial motonucleus. Measurements of the conduction time in the individual segments of the assumed path of the AMR in these intraoperative studies revealed that the latency of the AMR was approximately 2 ms longer that it would have been if the cross talk occurred in the facial nerve at the location of the vascular contact (Møller and Jannetta 1984) (Fig. 3.6).

The neural activity generated by stimulation of a branch of the facial nerve thus had to travel further towards the facial motonucleus before it activated descending facial structures. The magnitude (approximately 2 ms) of the estimated increase in

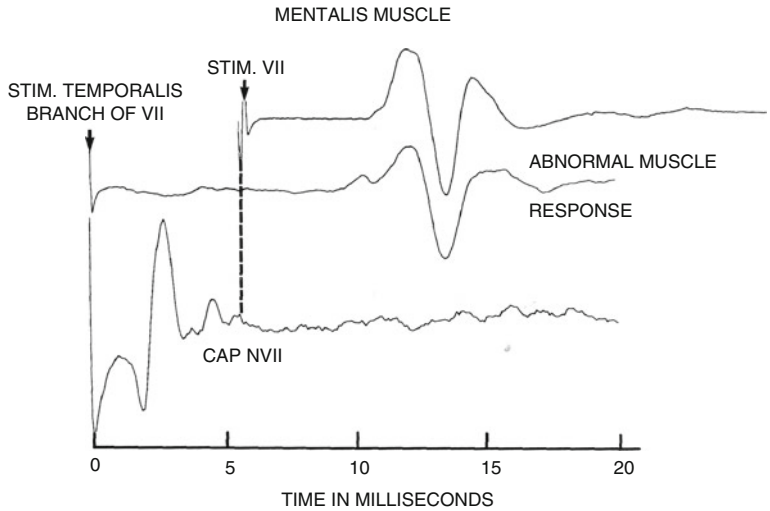

Fig. 3.6 Comparison between the AMR elicited by stimulation of the temporal branch of the facial nerve recorded from the mentalis muscle (middle tracing) and the response from the intracranial portion of the facial nerve at the location of the vascular contract in response to stimulation of the temporal branch of the facial nerve (bottom tracing). The top tracing is the response from the mentalis muscle to electrical stimulation of the intracranial portion of the facial nerve at the location of the vascular contact. If the cross talk that causes the AMR occurred in the facial nerve at the location of the vascular contact, the tracing of the response from the mentalis muscle to stimulation of the facial nerve would have been shifted to the left coinciding with the occurrence of the negative peak of the recording from the facial nerve

travel time supported the hypothesis that the anatomical location of the cross talk that is the basis for the AMR is the facial motonucleus. We further suggested that the facial motonucleus was also the site of the generation of the spasm and the cross talk (causing the synkinesis that is present in HFS) (Møller and Jannetta 1984).

These results of intraoperative recordings thus provided evidence that the anatomical location of the cross talk along the facial nerve was not the location of the vascular contact, and the results of these studies supported the hypothesis that the facial motonucleus is the anatomical site for the cross talk that causes synkinesis and is a basis for the AMR (Møller and Jannetta 1984). The results were later confirmed in other studies (Itagaki et al. 1988; Møller and Jannetta 1986).

We have proposed that plastic changes in the facial motonucleus and the resulting increase in synaptic efficacy could activate dormant pathways that connect different groups of motoneurons thereby causing the synkinesis that is observed especially in the blink reflex as well as the AMR (Møller 1997, 2014). The increased synaptic efficacy could also explain the spasm.

In all of these conduction time studies, there was only one patient undergoing MVD for HFS who exhibited some signs of ephaptic activity (Møller 1987) The results of intraoperative recordings indicated that the cross talk occurred where a blood vessel came into close contact with the facial nerve root. This shortening of the latency of the AMR occurred after signs that the facial nerve had been injured by surgical manipulations. The shortening of the latency of the AMR was only present for a few minutes; it was assumed to be caused by the cross talk (caused by ephaptic transmission) between facial nerve fibers at the location of the injury.

These observed transient signs of cross talk in the injured nerve root are in good agreement with the early description of abnormal cross talk between nerve fibers by Granit et al. 1944 (Granit et al. 1944), occurring for a brief period after injury. This author who has studied many patients during MVD operations for HFS has only seen that to occur in a single patient and only for a few minutes.

The facial motonucleus has also been identified as the likely site of the cross talk that creates the AMR through preoperative HFS studies (Valls-Sole and Tolosa 1989).

It was hypothesized (Møller and Jannetta 1984) that the facial motonucleus became hyperactive through a process similar to the kindling phenomenon described by Goddard (1964). In fact, it was later shown in animal experiments that it is possible to create HFS-like signs in a rat through electrical stimulation of the facial nerve using a kindle paradigm (Sen and Møller 1987; Saito and Møller 1993). In these animal studies, the facial nerve was stimulated electrically twice per day for several weeks near its entrance into the skull. After 4–6 weeks of such stimulation, the animals presented with signs similar to human HFS (Fig. 3.7). The AMR in the rat appear with a latency of approximately 6.5 ms thus shorter than that in humans because of the smaller size of the head of a rat.

There are other signs that the facial motonucleus is hyperactive in people with HFS. During normal surgical anesthesia, it is not possible to elicit the blink reflex by electrical stimulation of

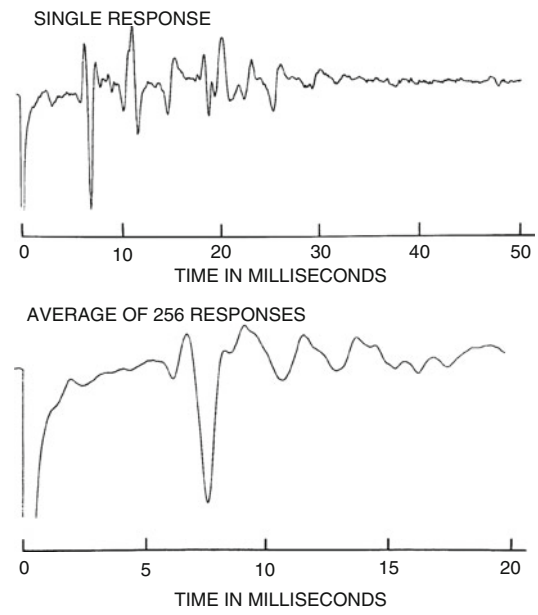

Fig. 3.7 AMR recorded from a rat that has had daily electrical stimulation of the facial nerve for 6 weeks (Reprint with permission from Sen and Møller 1987)

the supraorbital nerve. In patients with HFS, however, the R_1 component of the blink reflex can be electrically elicited by stimulation of the supraorbital nerve on the affected side (but not on the unaffected side), despite the suppressive effect of general anesthesia (Møller and Jannetta 1986). This means that the suppressive effect of the anesthesia on the blink reflex was counteracted on the spasm side by the effect of the pathology of HFS and the blink reflex can be elicited even when a patient with HFS is anesthetized. When the offending vessel is moved off the facial nerve and the AMR disappears, it is no longer possible to elicit the blink reflex under anesthesia.

The R_1 component of the blink reflex response on the spasm side has a latency that is slightly longer than what is the normal value of the latency, most likely because of the suppressive effect of anesthetic agents. The suppressive effect of the anesthesia was also evident from the fact that the R_2 component was not present.

Additionally, the F-response is stronger than normal in people with HFS (Hai and Pan 2007), supporting the hypothesis that the facial motonucleus is hyperactive in HFS. The F-response is the response of a motonucleus in response to antidromic activity in its motor nerve. The F-response thus reflects the excitability of the motonucleus.

As revealed by postmortem studies, close contact between the intracranial portions of the trigeminal and the facial nerves occurs in more than 50 % of asymptomatic individuals (Sunderland) (Sunderland 1948), but only very few have signs of diseases (HFS and TGN). This can only be explained by assuming that a second factor, in addition to the vascular contact, is necessary for creating AMR symptoms. Both vascular contact and this second factor must be present at the same time for AMR to occur. The nature of this second factor is unknown as is it anatomical location.

In some MVD operations, the AMR reappeared when the vessel was allowed to slip back onto the nerve (Fig. 13.2, Møller and Jannetta 1985). This is another critical piece of evidence supporting the indication that close contact between a blood vessel and the intracranial portion of the facial nerve is crucial for generating the symptoms and signs of HFS. Furthermore, it demonstrates that MVD works by removing the actual cause of the disorder rather than simply causing local injury to the nerve.

Since HFS can be effectively treated by MVD of the intracranial portion of the facial nerve, the symptoms must be linked to the effect of a blood vessel being in close contact with the facial nerve. This means that the presence of a blood vessel on the facial motor nerve somehow changes the function of the cells in the facial motonucleus.

We have suggested that the changes in the function of the facial motonucleus are caused by plastic changes in the motonucleus and that the activation of neuroplasticity is caused the irritation of the facial nerve from the close contact with blood vessel (Møller 1997). Activation of neuroplasticity may cause increased efficacy of synapses in the facial motonuclei including activating dormant synapses that connect cells in different parts of the nucleus (Møller 1997). This can explain the hyperactivity that causes the spasm and the synkinesis of the supraorbital nerve reflex as well as the AMR. The hypothesis that neuroplasticity causes the functional abnormalities in the facial motonucleus also can explain the anatomical extension of the spasm from a few muscles around the eye to engaging all mimic muscles except the muscles of the forehead without the vascular contact with the facial nerve extends.

We suggested that the lateral spread of activity in the facial nerve system that causes the AMR and the abnormal blink reflex is the result of creation of functional connections between alpha motoneurons in different parts of the facial motonucleus (Møller 1987). These are normally dormant but may be activated through plastic changes as a part of the pathology in HFS. The synapses that are normally dormant in adults may be active in infants and explain the mass facial movements often seen in infants. Very strong stimulation of the supraorbital nerve has been reported to cause similar spread of muscle activity (Kugelberg 1946).

The presence of the AMR and the synkinesis of the blink reflex response are signs that different motoneurons communicate with each other in an abnormal way in people with HFS.

The irritation of the facial nerve from a blood vessel may be what activates neuroplasticity that causes these functional changes in the facial motonucleus (Møller 1997). HFS may thus be yet another example of maladaptive neuroplasticity that causes signs and symptoms of disease (Møller 2014; Engineer et al. 2013).

3.5 Treatment of HFS

The symptoms of HFS (muscle spasm and synkinesis) can be successfully treated by MVD. MVD operations are traditionally performed using the retromastoid approach described by Jannetta (1970) and later by Kondo et al. (1980). This allowed for good visibility of the cerebellopontine angle region and provided access to the intracranial portion of the facial nerve. Unfortunately, the procedure involved retraction of the cerebellum, which involved certain risks.

The success rate of MVD operations for HFS as reported in studies of large populations is approximately 85 % freedom of symptoms for HFS (Barker et al. 1995, #767). This is similar to the results of MVD operations for TGN (Barker et al. 1996).

Endoscopic techniques have recently been described for MVD operations. One study involved 80 patients showed a success rate of 92.5 % (Badr-El-Dine et al. 2002).

406 patients treated by MVD using the traditional surgical techniques with the longest follow-up of 12 years and 3 months; 374 were cured (92.12 % cure rate). These investigators reported a recurrence rate of 7.88 %. Complications were sensorineural hearing loss in 31 patients (23 temporary, 8 permanent).

Introduction of intraoperative monitoring of AMR has made it possible to identify the vessel removal of which eliminated postoperative spasm. Before the introduction of monitoring of the AMR in operations for HFS, some patients were not relieved of their symptoms of HFS and had facial spasm after the operation. Such patients often were reoperated upon to achieve relieve of their facial spasm.

The value of monitoring of the AMR in predicting the outcome of MVD operations for HFS was documented in a study published 1987 (Møller and Jannetta 1987).

For intraoperative monitoring in MVD operations for HFS, the AMR were recorded. The stimuli used to elicit the AMR were 100 microsecond long rectangular impulses presented to the temporal branch of the facial nerve at a rate of 5 per second. The response was recorded from the mentalis muscle. The strength was adjusted individually to get a stable response.

If lifting a vessel off the facial nerve root does not abolish the AMR, it can be concluded that this particular vessel was not involved in causing the symptoms of the spasm in the particular patient. Only when moving a vessel off the intracranial portion of the facial nerve eliminated the AMR could it be concluded that the particular vessel was involved in the patient's symptoms. However, the AMR must remain absent when the stimulus rate is increased to 50 impulses per second. The operation should only be concluded when the AMR was absent after the stimulus rate had been raised to 50 pps.

In this study of 67 patients undergoing MVD for HFS, EMG activity was recorded from the mentalis muscle on the spasm side as AMR was elicited through electrical stimulation of the temporal or the zygomatic branch of the facial nerve. Before a blood vessel was moved off of the facial nerve, all patients demonstrated recordable activity in the mentalis muscle following stimulation. The latency of the initial component of the response was approximately 10 msec.

In 58 of the 67 patients, the AMR was eliminated completely after recovery from the operation. Of the remaining seven in which the AMR persisted at its original strength after the operation, only four experienced little or no improvement in the 2–6 months after the operation. These four underwent reoperation. In two of the remaining three patients, the spasm was absent at the 3- and 7-month follow-up examination, respectively, and one had mild spasm. Eighty-three percent of patients were thus completely cured by the first operation and 94 % of the patients experienced significant relief without a second operation. Monitoring of the AMR is now used routinely during operations to relieve hemifacial spasm.

Recent studies have confirmed the benefit from monitoring the AMR in MVD operations for HFS (Fukuda et al. 2012). These authors recorded the AMR, and the facial motor evoked potential elicited by transcranial electrical stimulation. They found that disappearance of AMR or a 50 % or more reduction in their amplitude and reduction in the amplitude of the facial motor evoked potential amplitude of the orbicularis oculi muscle after MVD were predictive of postoperative relief of HFS. Of the 42 patients in whom the amplitude of the facial motor evoked potentials was reduced from the baseline, 40 patients (95 %) had complete relief of the symptoms. AMRs disappeared in 20 of the 42 patients, and 19 (95 %) of these 20 patients experienced immediate relief of their symptoms after the MVD operation. Of the 20 patients in whom the AMRs persisted at the final recordings, 14 (70 %) had symptoms of HFS immediately after the operation but improved over time and eventually subsided (P <0.001).

This study showed that both intraoperative recorded AMR and facial evoked motor potentials are valuable in predicting the immediate outcome of an MVD operation for classical HFS. If the abnormal muscle response persists at the end of the operation conversely, there is a high likelihood of symptoms after the operation, but these symptoms may in some patients abate over time.

Surgical manipulations in connection with MVD of the facial nerve can cause injuries to the cochlear nerve, resulting in hearing loss and tinnitus. It is therefore also essential to monitor the cochlear nerve during the procedure. The usefulness of monitoring both auditory brainstem evoked responses (ABR) recorded from electrodes placed on the scalp and compound action potentials recorded directly from the eighth cranial nerve or from the cochlear nucleus has been evaluated in several studies (Møller and Jannetta 1987; Møller and Møller 1989; Sindou 2005) for monitoring the function of the cochlear nerve during MVD operations.

The risk of hearing loss was noticeably reduced after introduction of intraoperative monitoring of the cochlear nerve. In a study of 140 operations for HFS, only one patient lost hearing (became deaf) (Møller and Møller 1989), and one patient suffered noticeable hearing loss postoperatively but had recovered nearly normal hearing by 4 months after the operation; 6.4 % had an average elevation of the hearing threshold in the speech frequency range (500–2000 Hz) of 11 dB when tested 4–5 days after the operation; eight of these had fluid in their middle ears that most likely contributed to the hearing loss. Threshold elevations occurred at 4000 Hz in 13.6 % and at 8000 Hz and 20.7 %. Speech discrimination scores decreased by 15 % or more in 5 % of the operated patients, and it increased in 4 patients by 15 % or more, in 2 patients by as much as 52 %. What has been reported as "hearing loss" can vary between surgeons, some have reported only those who became deaf on one ear, or others have reported just noticeable change in the pure tone audiogram such as 20 dB decrease from preoperative value for two frequencies.

In 1983, Loeser and Chen summarized the complications in 450 MVD operations in 16 patient series (Loeser and Chen 1983). They showed that 13 % of the patients in their series had "cochlear nerve dysfunction." Studies by Wilkins (1993) showed in a series of MVD operations before introduction of intraoperative monitoring 6.6 % "profound ipsilateral hearing loss of deafness" (Wilkins 1993).

A later study by McLaughlin et al (1999) reports the number of complications in 4400 operations performed by a single surgeon over 29 years. During this period, the procedure has undergone many modifications that have contributed to the observed decrease in complications. Introduction of intraoperative neurophysiological monitoring (IONM) of hearing in the early 1980s has contributed to the decline in occurrence of hearing loss. Of the 2420 MVD operations performed for TGN, HFS, and GPA before 1990, cerebellar injury occurred in 0.87 %, hearing loss in 1.98 %, and cerebrospinal fluid (CSF) leakage in 2.44 %. Of the 1995 operations performed since 1990, cerebellar injuries declined 0.45 %, hearing loss to 0.8 %, and CSF leakage to 1.85 %.

Partial sectioning of the extratemporal branches of the facial nerve (Dobie and Fisch 1986; Fisch and Esslen 1972), injections of

phenol or alcohol in mimic muscles # 4205 and, more recently, injection of botulinum toxin (Botox) (Kraft and Lang 1988; Marion 1997; Vial and Vighetto 1997; Kollewe et al. 2010) into the affected muscles have had some success.

The beneficial effect of Botox injections changes (abate) over time, and the injections have to be repeated with intervals of 3–6 months. After using this treatment for several years, the beneficial effect decreases, and many patients will seek MVD operations. While some investigators have found some effect of baclofen, in some cases in conjunction with imipramine, on HFS (Sandyk and Gillman 1987 2096), the patients in this study may have had a peculiar form of HFS since the symptoms tended only to be precipitated by emotional events.

Conclusion

Microvascular decompression operations are effective in treatment of HFS and TGN. Intraoperative neurophysiological studies of patients undergoing MVD operations for HFS has shown evidence that the anatomical location of the pathologies that cause the spasm and the synkinesis of facial mimic muscles is the facial motor nucleus. These studies also showed evidence that vascular close contact with the intracranial portion of the facial nerve is only one of at least two factors that must be present in order that the symptoms of HFS are created and maintained.

The symptoms and signs of disorders such as HFS are caused by functional changes in specific parts of the nervous system without any morphological changes that can be detected by the imaging methods that are presently available. Few functional tests are available, but EMG recordings are helpful in diagnosis of HFS.

The functional changes in the central nervous system facilitate hyperexcitability and possibly rerouting of information; it is that hyperexcitability that causes the symptoms and signs. These functional changes are believed to be brought about by novel input to the central nervous system and perhaps slight injury to the respective cranial nerve activating neuroplasticity. Other factors in addition to the close contact between a cranial nerve and a blood vessel are most likely necessary to cause the symptoms of HFS. The fact that the incidence of these disorders increases with increasing age may be a result of a general decrease in inhibition in the central nervous system, caused by an age-related loss of the common inhibitory neurotransmitter substance (GABA) (Caspary et al. 1990).

References

Auger RG. Hemifacial spasm: clinical and electrophysiologic observations. Neurology. 1979;29:1261–72.

Badr-El-Dine M, El-Garem H, Talaat A, Magnan J. Endoscopically assisted minimally invasive microvascular decompression of hemifacial spasm. Otol Neurotol. 2002;23:122–8.

Barker FG, Jannetta PJ, Bissonette DJ, Shields PT, Larkins MV. Microvascular decompression for hemifacial spasm. J Neurosurg. 1995;82:201–10.

Barker FG, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Eng J Med. 1996;334:1077–83.

Campos-Benitez M, Kaufmann A. Neurovascular compression findings in hemifacial spasm. J Neurosurg. 2008;109:416–20.

Caspary DM, Raza A, Lawhorn Armour BA et al. Immunocytochemical and neurochemical evidence for age-related loss of GABA in the inferior colliculus: implications for neural presbycusis. The Journal of neuroscience:the official journal of the Society for Neuroscience 1990;10(7):2363–72.

Cushing H. The major trigeminal neuralgias and their surgical treatment based on experience with 332 gasserian operations. Am J Med Sci. 1920;160:158–84.

Dandy W. An operation for the cure of tic douloureux. Partial section of the sensory root at the pons. Arch Surg. 1929;18:687–734.

Dandy W. Concerning the cause of trigeminal neuralgia. Am J Surg. 1934;24:447–55.

DE Ridder D, Møller AR, Verlooy J, Cornelissen M, DE Ridder L. Is the root entry/exit zone important in microvascular compression syndromes? Neurosurgery. 2002;51:427–34.

Dobie R, Fisch U. Primary and revision surgery (selective neurectomy) for facial hyperkinesis. Arch Otolaryngol Head Neck Surg. 1986;112:154–63.

Ehni G, Woltman H. Hemifacial spasm. Arch Neurol Psychiatry. 1945;53:205–11.

Engineer ND, Møller AR, Kilgard MP. Directing neural plasticity to understand and treat tinnitus. Hear Res. 2013;295:58–66.

Esslen E. Der Spasmus facialis -- eine Parabiosserscheinung: Elektrophysiologische Untersuchnungen zum Enstehungsmechanismus des Facialisspasmus. Dtsch Z Nervenheil. 1957;176:149–72.

Esteban A, Molina-Negro P. Primary hemifacial spasm: a neurophysiological study. JNNP. 1986;49:58–63.

Ferguson JH. Hemifacial spasm and the facial nucleus. Ann Neurol. 1978;4(97):103.

Fisch U, Esslen E. The surgical treatment of facial hyperkinesis. Arch Otolaryngol Head Neck Surg. 1972;5:400–5.

Freckmann N, Hagenah R, Herrmann HD, Muller D. Treatment of neurogenic torticollis by microvascular lysis of the accessory nerve roots: indication, technique, and first results. Acta Neurochir (Wien). 1981;59:167–75.

Fukuda M, Oishi M, Takao T, Hiraishi T, Sato Y, Fujii Y. Monitoring of abnormal muscle response and facial motor evoked potential during microvascular decompression for hemifacial spasm. Surg Neurol Int. 2012;3:118.

Gardner WJ. Crosstalk -- The paradoxical transmission of a nerve impulse. Arch Neurol. 1966;14:149–56.

Gardner WJ, Sava GA. Hemifacial spasm -- a reversible pathophysiologic state. J Neurosurg. 1962;19:240–7.

Goddard GV. Amygdaloid stimulation and learning in the rat. J Comp Physiol Psychol. 1964;58:23–30.

Granit R, Leksell L, Skoglund CR. Fibre interaction in injured or compressed region of nerve. Brain. 1944;67:125–40.

Hai J, Pan Q. Experimental study on the correlation between abnormal muscle responses and F waves in hemifacial spasm. Neurol Res. 2007;29:553–6.

Hyun S, Kong D, Park K. Microvascular decompression for treating hemifacial spasm: lessons learned from a prospective study of 1,174 operations. Neurosurg Rev. 2010;33:325–34.

Itagaki S, Saito S, Nakai O. Intraoperative recording of evoked EMG in patients with hemifacial spasm – possible physiological mechanism. Facial N Res Jpn (Tokyo). 1988;8:143–6.

Jannetta PJ. Microsurgical exploration and decompression of the facial nerve in hemifacial spasm. Curr Top Surg Res. 1970;2:217–22.

Jannetta PJ. Hemifacial spasm caused by a venule: case report. Neurosurgery. 1984;14:89–92.

Jannetta PJ. Cranial rhizopathies. In: Yomans JR, editor. Neurological surgery. Philadelphia: W.B. Saunders; 1990.

Kim P, Fukushima T. Observations on synkinesis in patients with hemifacial spasm. Effect of microvascular decompression and etiological considerations. J Neurosurg. 1984;60(4):821–827.

Kraft SP, Lang AE. Botulinum toxin injections in the treatment of blepharospasm, hemifacial spasm, and eyelid fasciculations. Can J Neurol Sci. 1988;15(3):276–80.

Kollewe K, Mohammad IB, Dengler R, Dressler D. Hemifacial spasm and reinnervation synkinesias: long-term treatment with either Botox or Dysport. J Neural Transm. 2010;117:759–63.

Kondo A, Ishikawa J, Yamasaki T, Konishi T. Microvascular decompression of cranial nerves, particularly of the seventh cranial nerve. Neurol Med Chir (Tokyo). 1980;20:739–51.

Kugelberg E. “Injury activity” and “trigger zones” in human nerves. Brain. 1946;69:310–24.

Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg. 1977;47:316–20.

Loeser J, Chen J. Hemifacial spasm treatment by microsurgical facial nerve decompression. Neurosurg. 1983;13:141–6.

Marion M-H. Hemifacial spasm: treatment with botulinum toxin (long term results). In: Sindou M, Keravel Y, Møller AR, editors. Hemifacial spasm. A multidisiplinary approach. Wien: Springer; 1997. p. 141–4.

Martinez ARM, Nunes MB, Immich ND, Piovesana L, França Jr M, Campos L, D’abreu A. Misdiagnosis of hemifacial spasm is a frequent event in the primary care setting. Arq Neuro Psiquiatr. 2014;72:119–22.

Mclaughlin M, Jannetta P, Clyde B, Subach B, Comey C, Resnick D. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999;90:1–8.

Møller AR. Hemifacial spasm: ephaptic transmission or hyperexcitability of the facial motor nucleus? Exp Neurol. 1987;98:110–9.

Møller AR. The cranial nerve vascular compression syndrome: II. A review of pathophysiology. Acta Neurochir (Wien). 1991;113:24–30.

Møller AR. Cranial nerve dysfunction syndromes: pathophysiology of microvascular compression, chap. 2. In: Barrow DL, editor. Neurosurgical topics book 13, ‘Surgery of cranial nerves of the posterior fossa’. Park Ridge: American Association of Neurological Surgeons; 1993.

Møller AR. Pathophysiology of hemifacial spasm. In: Sindou M, Keravel Y, Møller AR, editors. Hemifacial spasm: a multidisciplinary approach. Wien: Springer; 1997.

Møller AR. Vascular compression of cranial nerves. I: History of the microvascular decompression operation. Neurol Res. 1998;20:727–31.

Møller AR. Vascular compression of cranial nerves. II. Pathophysiology. Neurol Res. 1999;21:439–43.

Møller AR. Intraoperative neurophysiological monitoring. 3rd ed. New York: Springer; 2011.

Møller AR. Neuroplasticity and its dark sides: disorders of the nervous system. Dallas: Aage R. Møller; 2014.

Møller AR, Jannetta PJ. On the origin of synkinesis in hemifacial spasm: results of intracranial recordings. J Neurosurg. 1984;61:569–76.

Møller AR, Jannetta PJ. Microvascular decompression in hemifacial spasm: intraoperative electrophysiological observations. Neurosurgery. 1985;16:612–8.

Møller AR, Jannetta PJ. Blink reflex in patients with hemifacial spasm: observations during microvascular decompression operations. J Neurol Sci. 1986;72:171–82.

Møller AR, Jannetta PJ. Monitoring facial EMG during microvascular decompression operations for hemifacial spasm. J Neurosurg. 1987;66:681–5.

Møller AR, Møller MB. Does intraoperative monitoring of auditory evoked potentials reduce incidence of hearing loss as a complication of microvascular decompression of cranial nerves? Neurosurgery. 1989;24:257–63.

Nagata K, Matsui T, Joshita H, Shigeno T, Asano T. Surgical treatment of spasmodic torticollis: effectiveness of microvascular decompression. Brain Nerve (Tokyo). 1989;41:97–102.

Nakai O, Itagaki S, Saito S. Electromyographic analysis of spasmodic torticollis. Tenth meeting of the World Society for Stereotactic and Functional Neurosurgery. Abstract. 1989.

Nielsen VK. Pathophysiological aspects of hemifacial spasm. Part. I Evidence of ectopic excitation and ephaptic transmission. Neurology. 1984a;34:418–26.

Nielsen VK. Pathophysiology of hemifacial spasm: II. Lateral spread of the supraorbital nerve reflex. Neurology. 1984b;34:427–31.

Osburn LL, Møller AR, Bhatt JR, Cohen-Gadol AA. Hemilingual spasm: defining a new entity, its electrophysiological correlates and surgical treatment through microvascular decompression. Neurosurgery. 2010;67:192–5.

Saito S, Møller AR. Chronic electrical stimulation of the facial nerve causes signs of facial nucleus hyperactivity. Neurol Res. 1993;15:225–31.

Sandyk R, Gillman MA. Baclofen in hemifacial spasm. Int J Neurosci. 1987;33:261–4.

Sen CN, Møller AR. Signs of hemifacial spasm created by chronic periodic stimulation of the facial nerve in the rat. Exp Neurol. 1987;98:336–49.

Sindou M. Microvascular decompression for primary hemifacial spasm. Importance of intraoperative neurophysiological monitoring. Acta Neurochir (Wien). 2005;147:1019–26.

Sunderland S. Microvascular relations and anomalies at the base of the brain. J Neurol Neurosurg Psychiatry. 1948;11:243–57.

Taarnhøj P. Decompression of the trigeminal root and the posterior part of the ganglion as treatment in trigeminal neuralgia; preliminary communication. J Neurosurg. 1952;9(3):288–90.

Taarnhøj P. Trigeminal neuralgia and decompression of the trigeminal root. Surg Clin North Am. 1956; 1145–57.

Tang I, Freeman S, Kontorinis G, Tang M, Rutherford S, King A, Lloyd S. Geniculate neuralgia: a systematic review. J Laryngol Otol. 2014;128:394–9.

Valls-Sole J, Tolosa ES. Blink reflex excitability cycle in hemifacial spasm. Neurology. 1989;39:1061–6.

Vial C, Vighetto A. Hemifacial spasm: treatment with botulinum toxin (a report of 50 patients). In: Sindou M, Keravel Y, Møller AR, editors. Hemifacial spasm. A multidisciplinary approach. Wien: Springer; 1997.

Wang X, Thirumala P, Shah A, Gardner P, Habeych M, Crammond D, Balzer J, Burkhart L, Horowitz M. The role of vein in microvascular decompression for hemifacial spasm: a clinical analysis of 15 cases. Neurol Res. 2013;35:389–94.

Wilkins R. Hemifacial spsam and other facial nerve dysfunction syndromes. In: Barrow D, editor. Surgery of the cranial nerves of the posterior fossa. Park Ridge: American Association of Neurological Surgeons; 1993.

Williams H, Lambert E, Woltman H. The problem of synkinesis and contracture in cases of hemifacial spasm and Bell's palsy. Ann Otol Rhinol Laryngol. 1952;40:857–70.

Yeh HSH, Tew JM. Tic convulsif, the combination of geniculate neuralgia and hemifacial spasm relieved by vascular decompression. Neurology. 1984;34: 682–3.

A Novel Hypothesis on the Mechanism of Hemifacial Spasm

4

Jun Zhong and Ning-Ning Dou

Abstract

Regardless neurovascular conflict has been believed to be the cause of hemifacial spasm, the mechanism of the disorder remains unclear to date. Current theories, merely focusing on the facial nerve, failed to explain the clinical phenomenon of immediate relief following a successful microvascular decompression (MVD) surgery. With experience of thousands MVDs and preliminary investigations, we have learnt that the offending artery may play a more important role rather than the effect of mechanical compression in the pathogenesis of the disease. Due to the mutual friction of nerve and artery with pulsation, the surfaces in contact are abraded. Neurotransmitters released from the sympathetic nervous endings in the adventitia may spillover from the artery wall and spread to the demyelinated nerve fibers in close contact. As these neurotransmitters bind with the transmembrane receptor proteins, ectopic action potentials are triggered from those nerve fibers with lower excitability threshold caused by vascular compression. When those messy impulses expand to the neuromuscular junctions, involuntary contractions of facial muscles occur. In this chapter, this "sympathetic hypothesis" was evaluated with logical and theoretical evidences as well as our experimental data.

Keywords

Hemifacial spasm • Mechanism • Offending artery • Sympathetic nerves • Ectopic action potentials • Transmembrane receptor proteins • Neurotransmitters

4.1 Introduction

Hemifacial spasm (HFS) is a common disorder of intracranial nerve hyperexcitability, which is caused by vascular compression of the seventh nerve root (Campbell and Keedy 1947; Gardner

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1953; Wartenberg 1950; Iwai et al. 2001; Marneffe et al. 2003; Miller and Miller 2012; Chung et al. 2001). Although the neurovascular conflict theory has been verified by successful microvascular decompression (MVD) surgery (Jannetta 1970, 1980, 1981; Jannetta et al. 1977), the underlying pathogenesis of HFS has been debated extensively for more than a century since Gowers first described (Valls-Solé 2007; Gowers 1892). Until now, many scholars have contributed their researches on the mechanism of the disease, and there are two main hypotheses so far, which were referred as the peripheral and the central.

4.1.1 The Peripheral Hypothesis

In 1962, Gardner (1962) postulated the symptom of HFS was an unstable and reversible pathophysiologic state caused by a mild compression of the nerve root which permitted transaxonal excitation while not interfering with axonal conduction. This local irritation of the nerve may facilitate the initiation of impulses in active fibers by impulses traveling over adjacent fibers or, in other words, ectopic excitation and ephaptic impulse transmission. Several experiments observed some histological changes at the site of compression, such as demyelination, vacuolization of the myelin sheath, and partial degeneration of axons (Nielsen 1984a, b; Nielsen and Jannetta 1984; Sanders 1989). However, researches have not involved the detail concerning the ectopic excitability emersion from the facial nerve fibers yet.

4.1.2 The Central Hypothesis

With development of electrophysiology, a characteristic wave of HFS has been recorded (Moller and Jannetta 1987). It was called abnormal muscle response (AMR). The wave could only be monitored in HFS patients by stimulating one branch of the facial nerve while recording from the muscle innervated by the other branch within approximately 10 msec (Kuroki and Moller 1994; Moller and Jannetta 1986). If the peripheral hypothesis was correct, the latency for AMR should theoretically equal the latency of a stimulus delivered to the facial nerve branch and recorded at the site of vascular compression plus the latency of a direct facial root stimulation at the compressed site. However, it was found that the sum of these latencies consistently fall short of the actual latency (Moller and Jannetta 1984). This extra time was then assumed to be consumed in the facial motor nucleus. Whereas this central hypothesis did not explain how a vascular compression results in central changes.

Whatever, the above hypotheses failed to explain the clinical phenomenon of immediate relief following a successful MVD operation. Nevertheless, it is hard to answer the question: why vascular compression of the facial nerve root results in neural hyperactivity (spasm) rather than hypoactivity (palsy)?

4.2 A New Hypothesis

With experience of thousands MVDs (Zhong et al. 2012, 2014), we have learnt that the offending artery may play a more important role other than the effect of mechanical compression in the pathogenesis of the disease. Eventually, a novel hypothesis was then proposed.

When the facial root is compressed by an artery, the neurovascular interfaces could be abraded with pulsation. As the adventitia is worn out, neurotransmitters that released from sympathetic endings in the offending artery wall may spillover and spread to the contact facial nerve. Meanwhile, the excitability threshold of the compressed nerve drops down due to transmembrane proteins (iron channels and receptors) occurs in the damaged axons. With the neurotransmitter-receptor interaction, G-protein-coupled Na^+ channels are activated, which induces ectopic action potentials on the facial nerve fibers. As these irregular impulses expand to the neuron-muscle junctions, involuntary contractions of facial muscles occur.

4.3 Evidences

4.3.1 Logics

During the MVD processes (Nielsen 1984b; Zhong et al. 2015; Xia et al. 2015; Ying et al. 2011, 2013; Zhou et al. 2012a), it was observed that once the offending artery was removed away from the nerve, the AMR wave was diminished immediately and the symptom of spasm ceased postoperatively in most of the cases (Gowers 1892; Ying et al. 2011, 2013; Zhou et al. 2012a; Martin et al. 1980; Habibi et al. 2011; Zheng et al. 2012a, b; Wang et al. 2014). This could not be explained by the peripheral or central hypotheses, for neither the histological changes at the conflict sites nor the hyperexcitability of facial motor neurons was able to repair at once after decompression (Zhong et al. 2010, 2011a, b, 2012; Kim et al. 2008; Kurokawa et al. 2004). Moreover, it was noticed that the episode of HFS is likely to occur when the patient is excited. Based on the fact that the symptom occurs with emotions and disappears with transposition of the offending artery, we guessed that the attack may relate to sympathetic nerves and the offending artery seemed to be the hinge (Dou et al. 2015). Given that the neurovascular conflict has been widely accepted as the etiology of the disease, it does not make sense to put emphasis on the nerve and to ignore the artery for investigation of the pathogenesis.

4.3.2 Animal Model

Møller's classical HFS mode in SD rats was adapted (Kuroki and Moller 1994; Zhou et al. 2012b). With a post-auricular skin, the main trunk of the facial nerve distal to stylomastoid foramen and the ipsilateral superficial temporal artery were exposed, which were then put in close contact. A 2/0 thread of chromic suture was squeezed in between them in order to induce lesions at the interfaces (Fig. 4.1). Two weeks later, the chromic suture was withdrawn and the artery and nerve were still kept in tighten contact. Another 2 weeks later, the animal was ready for electrophysiology. Finally, a stable AMR wave was monitored in 60 % of the experimental animals (Fig. 4.2). The result implied that HFS could be developed from vascular compression of the facial nerve root, but this neurovascular contact may not always lead to HFS.

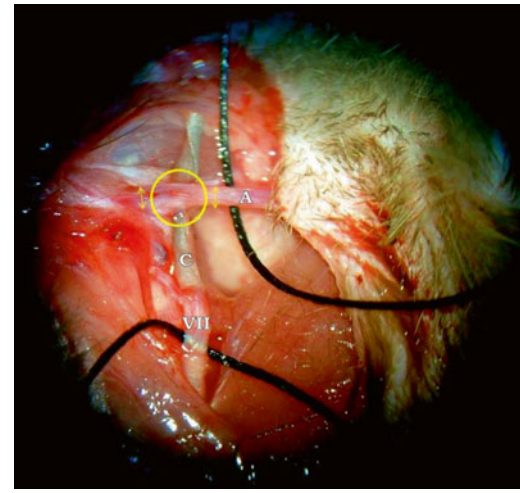

Fig. 4.1 A microscopic view of the HFS animal model. The SD rat was adopted in the animal model of hemifacial spasm. Under microscope, the superficial temporal artery (*A*) and extracranial facial nerve (*VII*) were dissected and put together in tight contact (*circle*). A chromic thread (*C*) was squeezed in between the nerve and the artery in order to induce lesions. To evaluate the effect of offending artery, a segment of the offending artery was cut off (*double arrows*) at both sides of the nerve, which yet was still in close contact with the facial nerve

4.3.3 Pathology

4.3.3.1 Attrition of the Neurovascular Interface Is the Precordium of HFS

The pathology demonstrated lesions of epineuria and/or adventitia at the neurovascular interfaces. However, only those with both lesions of the epineuria and the adventitia were monitored a stable AMR wave. As a result, we concluded that the precondition of HFS is the abrasion of neurovascular interfaces much than the vascular compression of

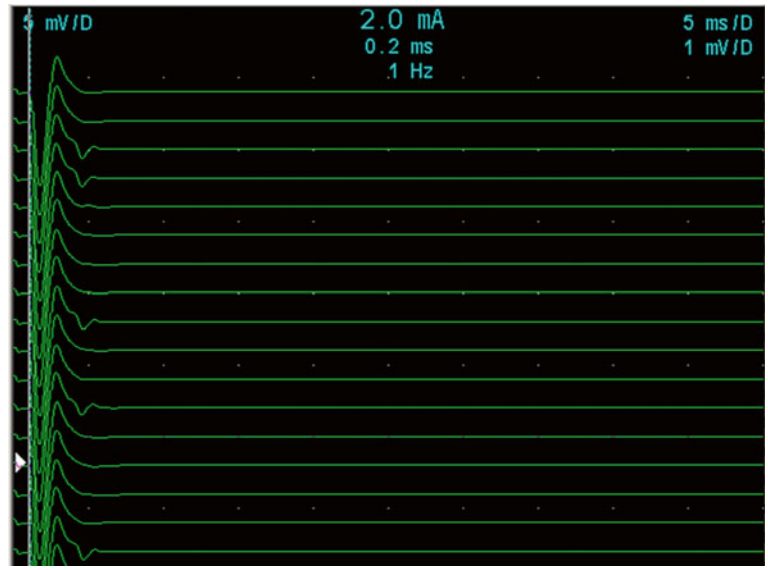

Fig. 4.2 A stable AMR wave

facial nerve root. This conclusion can explain why so many neurovascular contact cases were found in cadavers who had no history of HFS (Martin et al. 1980; Habibi et al. 2011).

4.3.4 Effect of Offending Artery

4.3.4.1 The Offending Artery May Play a Role More than Mechanical Compressions

HFS-mode rats were used to evaluate the effect of offending artery. After coagulation, a segment of the offending artery crossing the facial nerve was cut off at both sides of the nerve (Fig. 4.1). For the sham surgery group, the animal underwent the same operation except for cutting of the offending artery. Thirty HFS rats with positive AMR were randomly grouped, 20 for treatment and 10 for sham operation. Postoperatively, the AMR disappeared in 14 from the offending artery excluded group, while in three from the sham surgery group (p <0.05) (Zhou et al. 2012a). This experiment implied that the vascular connection rather than the vessel per se has some effect on the facial nerve root to trigger an attack of HFS.

4.3.4.2 The Sympathetic Nerve in the Artery Wall Might Be Involved in Generation of HFS

Anatomically, arteries are coated by adventitia which contains sympathetic nerve endings as well as vasa vasorum. Normally, the sympathetic endings release neurotransmitters that act on the nerve-muscle junctions to control contraction and dilation of the vascular smooth muscles (to regulate the vascular diameter). Accordingly, we made a denervation of the offending artery to assess the sympathetic effect in the HFS rats. With microscopy, the supper cervical ganglion was identified, and the ganglionectomy was completed. Twenty-four HFS rats were used in this series, 16 for treatment and eight for sham surgery. Postoperatively, the AMR disappeared in 12 of the treatment group, while in two of the sham surgery group (p <0.05) (Zhou et al. 2012b). For the fact that sympathetic denervation of the artery resulted in AMR vanish, we presumed that the sympathetic nerves may be involved for the pathogenesis of HFS. This explains why the attack often occurs when the patient is nervous.

4.3.5 Electrophysiology

4.3.5.1 Biological Connection between the Artery and the Nerve

In order to investigate how the sympathetic endings act upon the damaged nerve and induce an impulse, we conducted a clinical study. During the MVD for patients with HFS, we monitored a typical AMR wave with a latency of 10.7±0.5 ms (Zhong et al. 2012; Zheng et al. 2012a, b; Ying et al. 2011). When we directly stimulated the facial nerve root, we recorded a waveform with a latency of 7.3±0.8 ms, which disappeared when the offending was moved away from the nerve (Zheng et al. 2012a, b). Based on the latency difference, we deduced that something must have happened before an action potential emerged from the compressed nerve, as a physical current spread in light velocity with little time consumed in conduction.

4.3.5.2 An Irregular Impulse Could Be Induced by Neurotransmitters

As norepinephrine is the predominant neurotransmitter released from the sympathetic endings in the adventitia, we dripped norepinephrine onto the neurovascular conflict site in the animal experiment. Twelve HFS rats following exclusion of the offending artery were randomized into two groups according to drip with norepinephrine or normal saline. Postoperatively, the AMR reappeared in 4/6 animals of the norepinephrine group, while 0/6 in the normal saline group (p <0.05). The result demonstrated that the sympathetic effect may be executed through neurotransmitters (Zhou et al. 2012b).

4.4 Analysis

4.4.1 A Low Excitation Threshold in a Traumatic Nerve Fiber

Basically, functional proteins that synthesized intracellularly would accurately migrate to proper sites of the cell membrane (Wang et al. 2011). When the nerve fibers are injured, this protein synthesis and migration process could be out of control, and ectopic proteins may occur in the cell membranes. With sialylated extracellular domains of the injured neuron, negative charges are present surrounding the damaged neuron. This makes the resting transmembrane potential moves toward the polarization direction. As extracellular positive charges tend to neutralize these negative charges, the membrane potential is fluctuating. This phenomenon is called subthreshold membrane potential oscillation (SMPO) (Xing et al. 2001) (Fig. 4.3). It means the excitability threshold decreases in a traumatic nerve.

4.4.2 Ectopic Excitabilities

The amplitude and frequency of SMPO depends on voltage, which can be affected by a variety of factors, especially the opening and closing of the Na^+ channel. When this potential fluctuation reaches the threshold level, an action potential emerges (Xing et al. 2001, 2003; Xie et al. 2011). Recently, some transmembrane proteins have been found, such as α-adrenergic (Taylor and Ribeiro-da-Silva 2011), cholinergic (Moalem et al. 2005), and ATP (Coddou et al. 2011) receptors. It was reported in a chronic dorsal root ganglion crush injury experiment, stimulation of ATP receptors could increase the excitability (Xiang et al. 2008). While in a peripheral nerve injury study, the excitability reduced with the α_2-adrenergic receptor being blocked (Tulleuda et al. 2011). Our recent study showed that AMR could be recorded with a drip of norepinephrine (Zhou et al. 2012a). This implied that the relevant ligand may exist on the demyelinated facial nerve fibers. With a combination of norepinephrine and its receptor, the electrical voltage across the membrane decreases, and the membrane potential shifts from −90 mV toward 0 level, which allows the G-protein-coupled sodium channels ($Na_v1.8$) to open and induces occurrence of a propagable action potential (Xia et al. 2014) (Fig. 4.3).

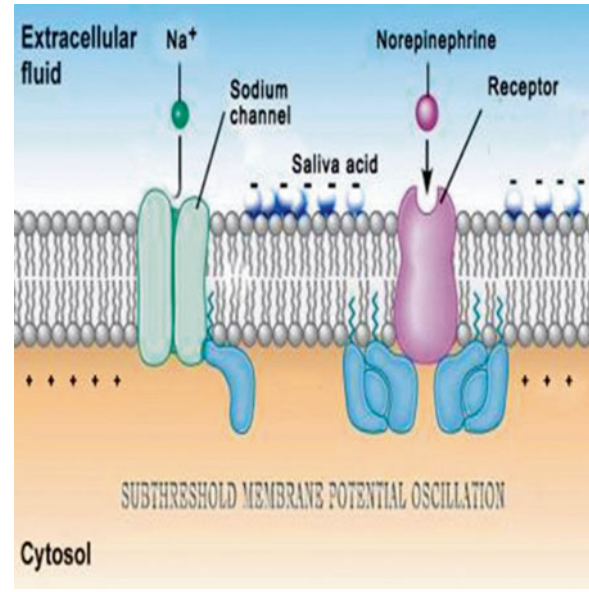

Fig. 4.3 An illustration of ectopic action potential emerged in a traumatic nerve. While the nerve fibers are damaged, the process of protein synthesis and migration can be out of control and ectopic proteins may emerge in the cell membranes, which make the extracellular domains of the injured neuron sialylated. Because of negative charges carried by sialic acids, the resting transmembrane potential moves toward the polarization direction. With combination of norepinephrine and its receptor, the electrical voltage across the membrane decreases further, which allows the G-protein-coupled sodium channels to open and finally induces a propagable action potential

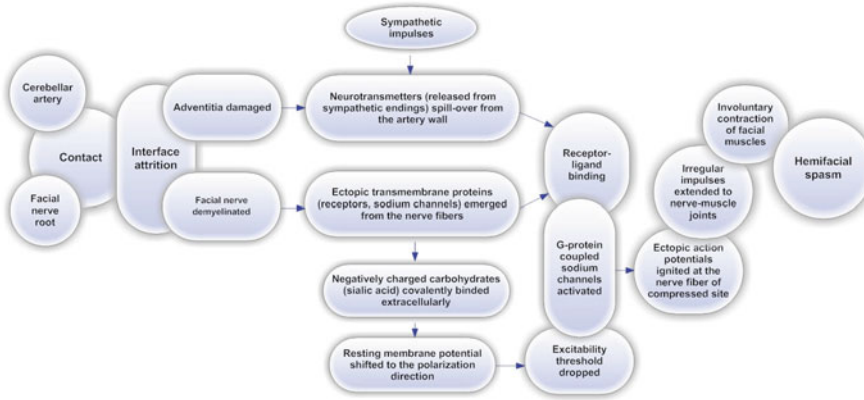

Fig. 4.4 An illustrative summary of the sympathetic hypothesis. Due to mutual friction with pulsation, the neurovascular interfaces are abraded. When the adventitia is worn out, neurotransmitters that released from sympathetic endings in the offending artery wall may spillover and transmit to the demyelinated facial nerve in close contact. Meanwhile, the excitability threshold of the nerve drops due to transmembrane proteins (iron channels and receptors) occurs in the abraded axons. With neurotransmitter-receptor interaction, G-protein-coupled Na+channels are activated, which induces ectopic action potentials on the facial nerve fibers. When those irregular impulses expand to the neuromuscular junctions, involuntary contractions of facial muscles occur

4.5 Summary

Eventually, we regard the essence of HFS attack as ectopic excitabilities that generated from the nerve fibers of facial root at the site where the artery compressed rather than from the central nucleus. While these irregular impulses propagate to the never-muscle junction, involuntary contractions of facial muscles happen. The pathological basis of the disease is the lesions of both epineurium and adventitia at the nerve-artery interface caused by mutual abrasion with pulsation in the posterior fossa. Since it has been found that the resting transmembrane potential in an injured nerve fiber may arise from transmembrane proteins (including receptors and channels), it make sense that neurotransmitters could

trigger an ectopic action potential on the compressed facial nerve fibers via receptor-ligand interaction (Fig. 4.4). However, we still need evidences to prove the factor that norepinephrine can spill over from an arterial wall in case of adventitia attrition.

References

Campbell E, Keedy C. Hemifacial spasm; a note on the etiology in two cases. J Neurosurg. 1947;4(4):342–7.

Chung SS, Chang JH, Choi JY, Chang JW, Park YG. Microvascular decompression for hemifacial spasm: a long-term follow-up of 1,169 consecutive cases. Stereotact Funct Neurosurg. 2001;77(1–4):190–3.

Coddou C, Yan Z, Obsil T, Huidobro-Toro JP, Stojilkovic SS. Activation and regulation of purinergic P2X receptor channels. Pharmacol Rev. 2011;63(3):641–83. Epub 2011 Jul 2017.

Dou NN, Zhong J, Zhou QM, Zhu J, Wang YN, Xia L, et al. The mechanism of hemifacial spasm: a new understanding of the offending artery. Neurol Res. 2015;37(2):184–8.

Gardner WJ. The mechanism of tic douloureux. Trans Am Neurol Assoc. 1953;3(78th Meeting):168–71; discussion, 171–163.

Gardner WJ. Concerning the mechanism of trigeminal neuralgia and hemifacial spasm. J Neurosurg. 1962;19:947–58.

Gowers WR. A manual of diseases of the nervous system, vol. 1. Philadelphia: Blakiston; 1892.

Habibi Z, Meybodi AT, Maleki F, Tabatabai S. Superior and anterior inferior cerebellar arteries and their relationship with cerebello-pontine angle cranial nerves revisited in the light of cranial cephalometric indexes: a cadaveric study. Turk Neurosurg. 2011;21(4):504–15.

Iwai Y, Yamanaka K, Nakajima H. Hemifacial spasm due to cerebellopontine angle meningiomas--two case reports. Neurol Med Chir (Tokyo). 2001;41(2):87–9.

Jannetta P. Microsurgical exploration and decompression of the facial nerve in hemifacial spasm. Curr Top Surg Res. 1970;2(21):220.

Jannetta PJ. Neurovascular compression in cranial nerve and systemic disease. Ann Surg. 1980;192(4):518.

Jannetta PJ (1981) Hemifacial spasm. In: The cranial nerves. Springer, pp 484–493.

Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS. Etiology and definitive microsurgical treatment of hemifacial spasm: operative techniques and results in 47 patients. J Neurosurg. 1977;47(3):321–8.

Kim JP, Park BJ, Choi SK, Rhee BA, Lim YJ. Microvascular decompression for hemifacial spasm associated with vertebrobasilar artery. J Korean Neurosurg Soc. 2008;44(3):131–5.

Kurokawa Y, Maeda Y, Toyooka T, Inaba K. Microvascular decompression for hemifacial spasm caused by the vertebral artery: a simple and effective transposition method using surgical glue. Surg Neurol. 2004; 61(4):398–403.

Kuroki A, Moller AR. Facial nerve demyelination and vascular compression are both needed to induce facial hyperactivity: a study in rats. Acta Neurochir (Wien). 1994;126(2–4):149–57.

Marneffe V, Polo G, Fischer C, Sindou M. Microsurgical vascular decompression for hemifacial spasm. Follow-up over one year, clinical results and prognostic factors. Study of a series of 100 cases. Neurochirurgie. 2003;49(5):527–35.

Martin RG, Grant JL, Peace D, Theiss C, Rhoton Jr AL. Microsurgical relationships of the anterior inferior cerebellar artery and the facial-vestibulocochlear nerve complex. Neurosurgery. 1980;6(5):483–507.

Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for treatment of hemifacial spasm: a systematic review. Br J Neurosurg. 2012; 26(4):438–44.

Moalem G, Grafe P, Tracey DJ. Chemical mediators enhance the excitability of unmyelinated sensory axons in normal and injured peripheral nerve of the rat. Neuroscience. 2005;134(4):1399–411.

Moller AR, Jannetta PJ. On the origin of synkinesis in hemifacial spasm: results of intracranial recordings. J Neurosurg. 1984;61(3):569–76.

Moller AR, Jannetta PJ. Physiological abnormalities in hemifacial spasm studied during microvascular decompression operations. Exp Neurol. 1986;93(3): 584–600.

Moller AR, Jannetta PJ. Monitoring facial EMG responses during microvascular decompression operations for hemifacial spasm. J Neurosurg. 1987;66(5):681–5.

Nielsen VK. Pathophysiology of hemifacial spasm: I. Ephaptic transmission and ectopic excitation. Neurology. 1984a;34(4):418–26.

Nielsen VK. Pathophysiology of hemifacial spasm: II. Lateral spread of the supraorbital nerve reflex. Neurology. 1984b;34(4):427–31.

Nielsen VK, Jannetta PJ. Pathophysiology of hemifacial spasm: III. Effects of facial nerve decompression. Neurology. 1984;34(7):891–7.

Sanders DB. Ephaptic transmission in hemifacial spasm: a single-fiber EMG study. Muscle Nerve. 1989;12(8): 690–4.

Taylor AM, Ribeiro-da-Silva A. GDNF levels in the lower lip skin in a rat model of trigeminal neuropathic pain: implications for nonpeptidergic fiber reinnervation and parasympathetic sprouting. Pain. 2011; 152(7):1502–10. Epub 2011 Mar 1512.

Tulleuda A, Cokic B, Callejo G, Saiani B, Serra J, Gasull X. TRESK channel contribution to nociceptive sensory neurons excitability: modulation by nerve injury. Mol Pain. 2011;7:30.

Valls-Solé J. Electrodiagnostic studies of the facial nerve in peripheral facial palsy and hemifacial spasm. Muscle Nerve. 2007;36(1):14–20.

Wang Q, Cao J, Zhu Q, Luan C, Chen X, Yi X, et al. Inhibition of voltage-gated sodium channels by

bisphenol A in mouse dorsal root ganglion neurons. Brain Res. 2011;1378:1–8. Epub 2011 Jan 2015.

Wang YN, Zhong J, Zhu J, Dou NN, Xia L, Visocchi M, et al. Microvascular decompression in patients with coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Acta Neurochir. 2014;156(6):1167–71.

Wartenberg R. Hemifacial spasm. Trans Am Neurol Assoc. 1950;51:276–7.

Xia L, Dou NN, Zhong J, Zhu J, Wang YN, Liu MX, et al. Upregulation of Nav1.8 in demyelinated facial nerves might be relevant to the generation of hemifacial spasm. J Craniofac Surg. 2014;25(4):1334–6.

Xia L, Zhong J, Zhu J, Dou NN, Liu MX, Li ST. Delayed relief of hemifacial spasm after microvascular decompression. J Craniofac Surg. 2015;26(2):408–10.

Xiang Z, Xiong Y, Yan N, Li X, Mao Y, Ni X, et al. Functional up-regulation of P2X 3 receptors in the chronically compressed dorsal root ganglion. Pain. 2008;140(1):23–34. Epub 2008 Aug 2019.

Xie RG, Zheng DW, Xing JL, Zhang XJ, Song Y, Xie YB, et al. Blockade of persistent sodium currents contributes to the riluzole-induced inhibition of spontaneous activity and oscillations in injured DRG neurons. PLoS One. 2011;6(4):e18681.

Xing JL, Hu SJ, Long KP. Subthreshold membrane potential oscillations of type A neurons in injured DRG. Brain Res. 2001;901(1–2):128–36.

Xing JL, Hu SJ, Jian Z, Duan JH. Subthreshold membrane potential oscillation mediates the excitatory effect of norepinephrine in chronically compressed dorsal root ganglion neurons in the rat. Pain. 2003;105(1–2):177–83.

Ying TT, Li ST, Zhong J, Li XY, Wang XH, Zhu J. The value of abnormal muscle response monitoring during microvascular decompression surgery for hemifacial spasm. Int J Surg. 2011;9(4):347–51. Epub 2011 Mar 2015.

Ying T, Thirumala P, Shah A, Nikonow T, Wichman K, Holmes M, et al. Incidence of high-frequency hearing loss after microvascular decompression for hemifacial spasm. J Neurosurg. 2013;118(4):719–24.

Zheng X, Hong W, Tang Y, Ying T, Wu Z, Shang M, et al. Discovery of a new waveform for intraoperative monitoring of hemifacial spasms. Acta Neurochir (Wien). 2012a;154(5):799–805. Epub 2012 Feb 2022.

Zheng X, Hong W, Tang Y, Wu Z, Shang M, Zhang W, et al. Sympathetic nerves bridge the cross-transmission in hemifacial spasm. Neurosci Lett. 2012b;517(1):52–5. Epub 2012 Apr 2015.

Zhong J, Zhu J, Li ST, Li XY, Wang XH, Yang M, et al. An analysis of failed microvascular decompression in patients with hemifacial spasm: focused on the early reoperative findings. Acta Neurochir. 2010;152(12):2119–23.

Zhong J, Zhu J, Li ST, Guan HX. Microvascular decompressions in patients with coexistent hemifacial spasm and trigeminal neuralgia. Neurosurgery. 2011a;68(4):916–20; discussion 920.

Zhong J, Li ST, Zhu J, Guan HX. Is entire nerve root decompression necessary for hemifacial spasm? Int J Surg. 2011b;9(3):254–7.

Zhong J, Li ST, Zhu J, Guan HX, Zhou QM, Jiao W, et al. A clinical analysis on microvascular decompression surgery in a series of 3000 cases. Clin Neurol Neurosurg. 2012;114(7):846–51. doi:10.1016/j.clineuro.2012.01.021. Epub 2012 Feb 5.

Zhong J, Zhu J, Sun H, Dou NN, Wang YN, Ying TT, et al. Microvascular decompression surgery: surgical principles and technical nuances based on 4000 cases. Neurol Res. 2014;36(10):882–93.

Zhong J, Xia L, Dou NN, Ying TT, Zhu J, Liu MX, et al. Delayed relief of hemifacial spasm after microvascular decompression: can it be avoided? Acta Neurochir. 2015;157(1):93–8; discussion 98–99.

Zhou QM, Zhong J, Jiao W, Zhu J, Yang XS, Ying TT, et al. The role of autonomic nervous system in the pathophysiology of hemifacial spasm. Neurol Res. 2012a;34(7):643–8.

Zhou QM, Zhong J, Jiao W, Zhu J, Yang XS, Ying TT, et al. The role of autonomic nervous system in the pathophysiology of hemifacial spasm. Neurol Res. 2012b;30:30.

Pathogenesis of Trigeminal Neuralgia

5

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5.1 Introduction

Facial pain is a common and nonspecific symptom that is associated with known and unknown etiologies. Because the most effective therapeutic interventions address a disorder's etiopathogenesis, it is important, when possible, to properly classify patients with different etiologies of facial pain. This is particularly true for trigeminal neuralgia (TN), because of the intensity of the pain associated with this disorder. Historically, the term TN has been used to refer to several different conditions. Taken in its most literal and general form, trigeminal neuralgia denotes pain that occurs within the dermatomal distribution of the trigeminal nerve. Many clinicians, however, reserve the term, TN, to signify a more specific disorder, which manifests as attacks of sudden, unilateral, and lancinating facial pain with characteristic triggers (e.g., light touch, cold air). These attacks may result from vascular compression of the trigeminal nerve near its entry into the brainstem (Jannetta 1967; Gardner and Miklos 1959). Vascular compression as the etiopathogenesis of TN, however, occurs in a minority of patients with facial pain. Furthermore, facial pain that does not fit this description completely may also be associated with probable incidental vascular compression.

Despite the ambiguity about the causal relationship between vascular compression and facial pain, the separation of intermittent facial pain, associated with "typical TN," and constant facial pain, associated with "atypical TN," is important because the presence of intermittent facial pain implies that the pain is more likely to be associated with vascular compression. This association, in turn, portends a higher chance of response to microvascular decompression (MVD) (Barker et al. 1996; Tyler-Kabara et al. 2002; Li et al. 2004).

5.2 Classification

The updated criteria for headaches, published in 2013 by the International Headache Society (IHS) (2013), consider primary headaches, secondary headaches, cranial neuralgias, central

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causes of facial pain, and all other headaches. Within this classification scheme, facial pain that is intermittent and shock-like, "recurring in paroxysmal attacks lasting from a fraction of a second to 2 min…without persistent background facial pain…without apparent cause other than neurovascular compression," is considered classical trigeminal neuralgia, purely paroxysmal type. Facial pain meeting the criteria for classic trigeminal neuralgia, "with persistent facial pain of moderate intensity in the affected area," is considered classic trigeminal neuralgia with concomitant persistent facial pain. Facial pain that occurs for more than 2 h each day for more than 3 months is considered persistent idiopathic facial pain. Thus, while this system enables some distinction between TN patients with and without vascular compression, further classification of idiopathic facial pain is needed.

5.3 Vascular Compression as the Cause of TN

Scrupulous examination of the results of MVD to treat TN reveals why classification of a patient's clinical symptoms is germane to any discussion of TN's etiopathogenesis. Because it relieves facial pain by eliminating vascular compression of the trigeminal nerve, MVD enables confirmation of the diagnosis of "vascular" TN. The results of several large case series by very experienced surgical teams yielding different success rates for the MVD operation, however, underscore the need for further refinement of the criteria used for classifying this disorder (Lee et al. 2014). Close examination of the inclusion and exclusion criteria reveal why this is so. In 2003 patients who underwent MVD for "typical TN" and 672 patients who underwent MVD for "atypical TN," the proportions of patients who were free of facial pain at last long-term follow-up (i.e., more than one half of patients were followed for greater than 5 years) were 73.7 and 34.7 % in the typical and atypical groups, respectively. While the stark difference in outcomes between the two groups supports a causal link between typical TN and vascular compression and suggests other mechanisms must account for the majority of cases with constant facial pain, the association between clinical presentation and underlying mechanisms is far from perfect.

Another classification criteria of facial pain is the Burchiel classification system, wherein type 1 TN corresponds to pain that is constant less than 50 % of the time and type 2 TN corresponds to TN that is constant greater than 50 % of the time. Following publication of the Burchiel classification criteria (Burchiel 2003; Miller et al. 2009), however, Heros suggested in an editorial (Heros 2009) that type 2 TN ought to be subdivided into type 2a and type 2b. Type 2a TN begins intermittently and, over time, transitions toward developing a constant component. Type 2b TN, alternatively, begins insidiously as a constant or aching pain (Sekula et al. 2011). This implies that type 1 and 2a are pathologically related, while type 2b may arise via an independent mechanism. One speculative explanation for the rare responses to MVD in patients with type 2b TN includes iatrogenic injury to the trigeminal nerve during surgical manipulation resulting in a rhizotomy-like effect (Adams 1989).

The first evidence that there may be a link between vascular compression and TN was published by Dandy (1934), who observed an intracranial portion of the trigeminal nerve impinged by a blood vessel. The first evidence of a causal link between vascular compression and TN was provided by Gardner who performed, in 1959 (Gardner and Miklos 1959), the first vascular decompression resulting in symptomatic resolution of trigeminal neuralgia. This operation, however, was substantially developed and refined along with the use of the operating microscope by Jannetta. The results of MVD reported by Jannetta (and subsequently others) showed that treatment of the vascular compression resulted in resolution of a patient's symptoms in some patients (Barker et al. 1996).

Further evidence of a causal link between vascular compression and TN comes from an analogous cranial nerve disorder, hemifacial spasm (HFS). The pathophysiology of

hemifacial spasm, a syndrome of unilateral episodic twitching of the muscles around the eye and the lower face, is thought to be analogous to trigeminal neuralgia. Because the seventh cranial nerve is a motor nerve, however, intraoperative electromyography is routinely performed during MVD for HFS. This technique has been employed to show that immediate or delayed cessation of HFS' signature "abnormal motor response" is often correlated with decompression of offending blood vessels providing indirect evidence that MVD is unlikely to work merely by causing trauma to the nerve.

5.4 Mechanisms Underlying Vascular Compression and TN

While it is generally assumed that mechanical stimulation of the trigeminal nerve by the impinging blood vessel is ultimately responsible for the pain of TN, the close association between blood vessels and nerves throughout the body in addition to the relatively large mechanical forces regularly impinging on many peripheral nerves in the absence of pain suggests that mechanical stimulation alone cannot account for the pain of TN. Consequently, investigators have searched for additional underlying mechanisms. Most striking among these is the demyelination observed in the nerve at the site of compression.

Preclinical evidence in support of a link between demyelination and TN-like changes was obtained from Burchiel and colleagues who demonstrated that following iatrogenic demylination of cat and *Macaca mulatta* monkey trigeminal nerve roots, trigeminal nerve roots from both animals produced extra action potentials in response to stimulation (Burchiel 1980). Suture placement resulted in focal nerve injury and demyelination. Electrophysiological recordings were then performed at intervals of several weeks following suture implantation. The appearance of abnormal action potentials was correlated with the appearance of focal demyelination, which took at least 1 week to occur. Subsequent data from rodent models of peripheral nerve compression confirmed that any compression of a peripheral nerve of sufficient intensity to occlude the local blood supply to the nerve was capable for producing signs of neuropathic pain (Bennett and Xie 1988) and spontaneous activity in the compressed nerve (Kajander et al. 1992).

Clinical evidence of demyelination associated with TN was originally reported by Hilton and colleagues. Staining followed by electron microscopy revealed degraded central myelin at the site of arterial compression in a single patient (Hilton et al. 1994). This observation was subsequently replicated by Rappaport et al. (1997) who published an analysis of biopsy samples taken from the site of compression in 12 patients with trigeminal neuralgia undergoing microvascular decompression. Interestingly, in 11 of 12 patients, both demyelination and axonopathy were noted by biopsy even though only 7 of the 12 patients had intraoperatively confirmed arterial compression of the trigeminal nerve. Furthermore, the authors introduced the concept of the "ignition hypothesis" which holds that damage to the root entry zone of the trigeminal nerve induces parts of the trigeminal ganglion to develop autorhythmicity (Devor et al. 2002).

Jannetta hypothesized that compression-induced demyelination allowed the spread of ephaptic impulse transmission at these sites. These ephaptic impulses, in turn, are perceived as pain (Jannetta 1967). In addition to sites of dymelination at the site of vascular compression, demyelinating plaques of the trigeminal system have instead been implicated as the causative lesion in TN. In an MR study of six patients with symptomatic trigeminal neuralgia, Gass et al. (1997) demonstrated the presence of demyelinating plaques in the trigeminal fibers of all six patients, with the lesions ranging anywhere from pontine trigeminal nuclei to the junction of the central and peripheral myelin, midway through the cisternal segment of the trigeminal nerve. Furthermore, Cruccu et al. showed that patients with symptomatic TN were more likely to have a plaque within trigeminal afferents (i.e., mostly intrapontine) than patients with MS and trigeminal sensory dysfunction other than TN (Cruccu et al. 2009).

In addition to these imaging findings, there exists both intraoperative (Lazar and Kirkpatrick 1979; Love et al. 1998) and postmortem (Rushton

and Olafson 1965) evidence for the presence of demyelinating lesions within the trigeminal afferent fibers. Lazar examined a biopsy specimen taken during a complete sensory rhizotomy performed on a patient with trigeminal neuralgia secondary to multiple sclerosis. Histological examination of the specimen demonstrated myelin degeneration consistent with an MS plaque occurring at the root entry zone of the trigeminal nerve. This finding was expanded upon by Love who examined six biopsy samples from patients with TN secondary to MS who underwent a partial sensory rhizotomy. Samples were taken from the most proximal part of the trigeminal nerve near the entry of afferent fibers into the pons. Histological examination showed numerous areas of directly abutting axons and a dearth of normal myelin. While all of these lesions were confined to the centrally myelinated portion of the trigeminal nerve, the area of demyelination extended proximally along the cisternal portion of the nerve to the transition between the distal and proximal portions of the trigeminal nerve (Love et al. 2001). Additionally, Olafson demonstrated, upon autopsy of deceased patient with right-sided symptomatic TN, a plaque "At the junction of the right fifth nerve and the pons" and again in the brainstem near the forth ventricle (Olafson et al. 1966).

One final line of evidence in support of the link between TN and demyelination comes from the observation that the incidence of TN is higher in patients with multiple sclerosis (MS) than in the general population (O'Connor et al. 2008). Additionally, when compared to TN patients without MS, TN patients with MS are more likely both to develop facial pain at a younger age and to develop bilateral facial pain. These facts raise the possibility that it is demyelination per se, if not ultimately plaque formation, rather than the combination of demyelination and vascular compression that is necessary for the manifestation of TN, because in contrast to patients with classical TN, most patients with MS and TN are much less likely to have a compressive vessel and, therefore, much less likely to respond to microvascular decompression (Resnick et al. 1996). Further evidence that the causal factor for the pain associated with TN is dymelination rather than demyelination with vascular compression comes from the observation that while MVD has a high cure rate with a low risk of sensory side effects in patients with type 1 or classical paroxysmal type TN or type 2a, the operation is largely ineffective in patients with other types of TN even when clear neurovascular compression is present.

5.5 Evidence against Demylination and/or Demyelinating Plaques as a Mechanism of TN

While evidence in support of a link between vascular compression and demyelination/demyelinating plaques and TN is compelling, there are several lines of evidence suggesting that additional mechanisms are necessary for the manifestation of pain. Most prominent among these is the timing of the pain relief produced by MVD: the resolution of symptoms is almost instantaneous following MVD. In a landmark study, Bunge demonstrated that spontaneous central remyelination can occur in cats following spinal cord injury (Bunge et al. 1961). The authors, however, noted that the very first evidence of new myelin sheaths did not appear until 19 days following injury. If removal of the offending blood vessel does promote remyelination, it would not be expected to produce a clinical effect within seconds to hours. Additionally, as Adams suggests, demyelination is unlikely to account for the characteristic periods of remission and recurrence experienced by most patients with type 1 TN (Adams 1989). Myelin is simply too slow to regenerate in order to account for an episode of spontaneous remission. Additionally, it is unclear why myelin would spontaneously regenerate in the presence of unrelenting vascular compression.

Additional evidence in support of the suggestion that neither MVD nor demyelination alone is sufficient for the generation of pain comes from the relatively high incidence of asymptomatic vascular compression. Cadaveric observations have demonstrated vascular compression of the trigeminal nerve in 16–58 % of asymptomatic individuals, compared to 90–100 % of patients with TN (Haines et al. 1980; Hamlyn 1997a; b). These postmortem data must, of course, be viewed with caution, given the potential for structural

changes that occur during the immediate postmortem period including loss of blood pressure and atrophy of the intracranial tissues that make identification of compression more difficult. Nevertheless, this relatively high incidence of neurovascular compression in asymptomatic individuals is consistent with findings in MRI studies. That is, recent technological and methodological advances have given rise to the ability to detect vascular compression of the cranial nerves radiographically, with a high degree of sensitivity and specificity (Sekula et al. 2014). During the dedicated examination of 200 trigeminal nerves in 100 unaffected individuals, vascular compression was observed in 87.5 % of the nerves studied. Moreover, 86 % of these compressive vessels were arteries and the compression occurred along the most proximal portion of the trigeminal nerve 58 % of the time (Peker et al. 2009).

Similarly, despite the numerous reports of demyelinating lesions in the trigeminal fibers of patients with symptomatic TN, similar lesions appear in many MS patients who do not have TN-like pain. Da Silva examined MRIs from 275 patients with MS but without TN and found that 2.9 % of these TN-free patients had demyelinating lesions at the area where the trigeminal afferents enter the pons (da Silva et al. 2005). The existence of incidental demyelinating plaques along the trigeminal fiber distribution in patients with MS but without TN was confirmed by Mills et al. (2010). Using high resolution (3 T) MRI, Mills showed that 23 % of the 47 MS patients examined had a plaque between the trigeminal pontine nuclei and the cisternal portion of the nerve despite no symptoms of TN. This shows that the presence of demyelination alone is not sufficient to produce TN in all individuals.

5.6 Mechanisms That May Work in Concert with Vascular Compression and Demyelination

As suggested by the ICHD or Burchiel classification of TN and the incomplete overlap between either vascular compression or demyelination, the spectrum of symptoms associated with TN and the immediacy of relief afforded by MVD suggest other mechanisms must contribute to the pain of TN. One such mechanism would be the redistribution of voltage-gated Na^+ channels at points of demyelination. The distribution of ion channels in myelinated nerves is tightly regulated with voltage-gated Na^+ channels clustered at nodes of Ranvier and K^+ channels targeted to the paranodal regions (Buttermore et al. 2013). Demyelination results in a dramatic disruption in this organization, with Na^+ channels becoming broadly distributed across the demyelinated region (Henry et al. 2006; 2009). The result may be a disruption in the balance between excitatory and inhibitory channels and the emergence of spontaneous activity. Such changes have been most clearly documented in preclinical models of traumatic nerve injury, where such a shift in the balance results in oscillations that can result in bursts of activity comparable to what one would anticipate in classical TN (Amir et al. 2002). Low threshold tetrodotoxin (TTX)-sensitive Na^+ channels appear to play a particularly important role in the emergence of this spontaneous activity, a mechanism that may account for the efficacy of the use-dependent Na^+ channel blocker, carbamazepine, in the pharmacological treatment of TN (Amir et al. 2006).

Consistent with the suggestion that a redistribution of voltage-gated Na^+ channels may contribute to the emergence of TN, not all portions of the trigeminal nerve are equally susceptible to neurovascular compression (Sindou et al. 2002; Peker et al. 2006). From his early experience with MVD, Jannetta described the importance of the trigeminal nerve's "root entry zone" (REZ), which he believed extended to the porous trigeminus (McLaughlin et al. 1999). The existence of this so-called transition zone has been demonstrated empirically, and its dimensions have been quantified by Peker et al. (2006). Upon staining histological sections of 100 trigeminal nerves with Luxol fast blue in order to detect oligodendrocyte-derived (i.e., central) myelin, the authors showed that the end of the transition zone never extended beyond the halfway point between the nerve's exit point from the pons and Meckel's cave. A report by Sindou, however, documented the location of compression identi-

fied intraoperatively during MVD, showing that compression occurred along the most distal portion of the trigeminal nerve (i.e., near the porous trigeminus) only 9.8 % of the time.

The correlation between decompression of the trigeminal nerve's proximal segment and a pain-free outcome following MVD implies differential susceptibility of the central and peripheral myelin to vascular insult. While the exact nature of this variable susceptibility remains uncharacterized, there are a number of known differences between central and peripheral myelin, which may explain why compression of the central myelin is more likely to be associated with TN. The ratio of cerebroside and sulfatide lipids to sphingomyelin is significantly lower in the peripherally derived myelin than in the centrally derived myelin. Potentially more importantly, however, is the evidence in support of the unique distribution of channels along a peripheral nerve. Sites of transition, such as that associated with the "T-junction" of the bifurcated peripheral axon may be associated with particularly high densities of voltage-gated Na^+ channels to ensure action potential propagation through this zone, as well as the presence of channels with unique biophysical properties (Amir et al. 2006). Both of these factors may make this zone particularly sensitive to the demyelinating effect of vascular compression.

An additional mechanism that may account for the emergence of ectopic activity at the site of vascular compression is the ectopic expression of chemo- (Chen et al. 1996; Shinder et al. 1999), thermo- (Kirillova et al. 2011), and, potentially most importantly, mechanotransducers at the site of compression (Kirillova et al. 2011). In an elegant preclinical study, Janig and colleagues demonstrated that following nerve injury, transducers are inserted in the membrane of axons. The result is the emergence of sensitivity of chemicals, changes in temperature (heat and cold), and mechanical stimuli (Kirillova et al. 2011; Grossmann et al. 2009). The presence of mechanotransducers would enable the trigeminal nerve to become responsive to the changes in blood pressure in the compressing nerve and, consequently, serve as a trigger for a painful attack.

A third mechanism for the bursts of pain associated with TN was proposed by Devor, based on the somatotopic organization of the trigeminal ganglion (Devor et al. 2002). In contrast to spinal ganglia, there is a loose somatotopic organization of the trigeminal ganglia such that the cell bodies of neurons giving rise to innervation of particular cranial structures are likely to reside in close approximation (Chai et al. 2014; Oyagi et al. 1989; Gregg and Dixon 1973). This fact, in combination that sensory afferents are not only able to release transmitter within the ganglion but response to transmitter released within the ganglia (Matsuka et al. 2001; Neubert et al. 2000), could account for the observation that pain attacks may be triggered with innocuous stimulation of overlaying tissue (Devor et al. 2002). In a neuron already hyperexcitable from the redistribution of ion channels at sites of demyelination, the additional depolarization associated with the actions of transmitters released within the ganglia may be all that is needed to trigger an attack.

Conclusion

While vascular compression clearly plays a part in some patients' trigeminal neuralgia, other mechanisms almost certainly contribute as well. The possibility that these other factors contribute to the manifestation of TN could account for the variability in both the clinical presentation of the disorder and the response to pharmacological interventions and MVD. On the one hand, classical TN would depend primarily on the changes in the excitability of the peripheral nerve and aberrant expression of transducers, such that MVD eliminates a primary drive for the attack, and relief is obtained. Conversely, in patients with more widespread demyelination, the loss of membrane stability associated with ion channel distribution and the emergence of ectopic chemotransducers as a primary driver for aberrant afferent activity, patients may experience ongoing pain and limited responsiveness to MVD. A final possibility is a loss of homeostatic balance as a result of a focal loss of non-nociceptive input to the CNS. Under this scenario, focal block or loss of propagation of action potentials in low

threshold afferents that normally inhibit input of nociceptive afferents would enable the manifestation of ongoing pain that may also be unresponsive to MVD.

References

Adams CB. Microvascular compression: an alternative view and hypothesis. J Neurosurg. 1989;70(1):1–12.

Amir R, Michaelis M, Devor M. Burst discharge in primary sensory neurons: triggered by subthreshold oscillations, maintained by depolarizing after potentials. J Neurosci. 2002;22(3):1187–98.

Amir R, Argoff CE, Bennett GJ, et al. The role of sodium channels in chronic inflammatory and neuropathic pain. J Pain. 2006;7(5 Suppl 3):S1–29.

Barker 2nd FG, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med. 1996;334(17):1077–83.

Bennett GJ, Xie YK. A peripheral mononeuropathy in rat that produces disorders of pain sensation like those seen in man. Pain. 1988;33(1):87–107.

Bunge MB, Bunge RP, Ris H. Ultrastructural study of remyelination in an experimental lesion in adult cat spinal cord. J Biophys Biochem Cytol. 1961;10:67–94.

Burchiel KJ. Abnormal impulse generation in focally demyelinated trigeminal roots. J Neurosurg. 1980; 53(5):674–83.

Burchiel KJ. A new classification for facial pain. Neurosurgery. 2003;53(5):1164–6; discussion 1166–7.

Buttermore ED, Thaxton CL, Bhat MA. Organization and maintenance of molecular domains in myelinated axons. J Neurosci Res. 2013;91(5):603–22.

Chai Y, Chen M, Zhang W. Somatotopic organization of trigeminal ganglion: three-dimensional reconstruction of three divisions. J Craniofac Surg. 2014;25(5):1882–4.

Chen Y, Michaelis M, Janig W, Devor M. Adrenoreceptor subtype mediating sympathetic-sensory coupling in injured sensory neurons. J Neurophysiol. 1996;76(6):3721–30.

Cruccu G, Biasiotta A, Di Rezze S, et al. Trigeminal neuralgia and pain related to multiple sclerosis. Pain. 2009;143(3):186–91.

da Silva CJ, da Rocha AJ, Mendes MF, Maia Jr AC, Braga FT, Tilbery CP. Trigeminal involvement in multiple sclerosis: magnetic resonance imaging findings with clinical correlation in a series of patients. Mult Scler. 2005;11(3):282–5.

Dandy W. Concerning the cause of trigeminal neuralgia. Am J Surg. 1934;24:447–55.

Devor M, Amir R, Rappaport ZH. Pathophysiology of trigeminal neuralgia: the ignition hypothesis. Clin J Pain. 2002;18(1):4–13.

Gardner WJ, Miklos MV. Response of trigeminal neuralgia to decompression of sensory root; discussion of cause of trigeminal neuralgia. J Am Med Assoc. 1959;170(15):1773–6.

Gass A, Kitchen N, MacManus DG, Moseley IF, Hennerici MG, Miller DH. Trigeminal neuralgia in patients with multiple sclerosis: lesion localization with magnetic resonance imaging. Neurology. 1997;49(4):1142–4.

Gregg JM, Dixon AD. Somatotopic organization of the trigeminal ganglion in the rat. Arch Oral Biol. 1973;18(4):487–98.

Grossmann L, Gorodetskaya N, Baron R, Janig W. Enhancement of ectopic discharge in regenerating A- and C-fibers by inflammatory mediators. J Neurophysiol. 2009;101(6):2762–74.

Haines SJ, Jannetta PJ, Zorub DS. Microvascular relations of the trigeminal nerve. An anatomical study with clinical correlation. J Neurosurg. 1980;52(3):381–6.

Hamlyn PJ. Neurovascular relationships in the posterior cranial fossa, with special reference to trigeminal neuralgia. 1. Review of the literature and development of a new method of vascular injection-filling in cadaveric controls. Clin Anat. 1997a;10(6):371–9.

Hamlyn PJ. Neurovascular relationships in the posterior cranial fossa, with special reference to trigeminal neuralgia. 2. Neurovascular compression of the trigeminal nerve in cadaveric controls and patients with trigeminal neuralgia: quantification and influence of method. Clin Anat. 1997b;10(6):380–8.

Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalgia. 2013;33(9):629–808.

Henry MA, Freking AR, Johnson LR, Levinson SR. Increased sodium channel immunofluorescence at myelinated and demyelinated sites following an inflammatory and partial axotomy lesion of the rat infraorbital nerve. Pain. 2006;124(1–2):222–33.

Henry MA, Luo S, Foley BD, Rzasa RS, Johnson LR, Levinson SR. Sodium channel expression and localization at demyelinated sites in painful human dental pulp. J Pain. 2009;10(7):750–8.

Heros RC. Results of microvascular decompression for trigeminal neuralgia. J Neurosurg. 2009;110(4): 617–8; author reply 618–9.

Hilton DA, Love S, Gradidge T, Coakham HB. Pathological findings associated with trigeminal neuralgia caused by vascular compression. Neurosurgery. 1994;35(2): 299–303; discussion 303.

Jannetta PJ. Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neuralgia. J Neurosurg. 1967;26(1):Suppl:159–62.

Kajander KC, Wakisaka S, Bennett GJ. Spontaneous discharge originates in the dorsal root ganglion at the onset of a painful peripheral neuropathy in the rat. Neurosci Lett. 1992;138(2):225–8.

Kirillova I, Rausch VH, Tode J, Baron R, Janig W. Mechano- and thermosensitivity of injured muscle afferents. J Neurophysiol. 2011;105(5):2058–73.

Lazar ML, Kirkpatrick JB. Trigeminal neuralgia and multiple sclerosis: demonstration of the plaque in an operative case. Neurosurgery. 1979;5(6):711–7.

Lee A, McCartney S, Burbidge C, Raslan AM, Burchiel KJ. Trigeminal neuralgia occurs and recurs in the

absence of neurovascular compression. J Neurosurg. 2014;120(5):1048–54.

Li ST, Pan Q, Liu N, Shen F, Liu Z, Guan Y. Trigeminal neuralgia: what are the important factors for good operative outcomes with microvascular decompression. Surg Neurol. 2004;62(5):400–4; discussion 404–5.

Love S, Hilton DA, Coakham HB. Central demyelination of the Vth nerve root in trigeminal neuralgia associated with vascular compression. Brain Pathol. 1998;8(1):1–11; discussion 11–2.

Love S, Gradidge T, Coakham HB. Trigeminal neuralgia due to multiple sclerosis: ultrastructural findings in trigeminal rhizotomy specimens. Neuropathol Appl Neurobiol. 2001;27(3):238–44.

Matsuka Y, Neubert JK, Maidment NT, Spigelman I. Concurrent release of ATP and substance P within guinea pig trigeminal ganglia in vivo. Brain Res. 2001;915(2):248–55.

McLaughlin MR, Jannetta PJ, Clyde BL, Subach BR, Comey CH, Resnick DK. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999;90(1):1–8.

Miller JP, Acar F, Burchiel KJ. Classification of trigeminal neuralgia: clinical, therapeutic, and prognostic implications in a series of 144 patients undergoing microvascular decompression. J Neurosurg. 2009; 111(6):1231–4.

Mills RJ, Young CA, Smith ET. Central trigeminal involvement in multiple sclerosis using high-resolution MRI at 3 T. Br J Radiol. 2010;83(990):493–8.

Neubert JK, Maidment NT, Matsuka Y, Adelson DW, Kruger L, Spigelman I. Inflammation-induced changes in primary afferent-evoked release of substance P within trigeminal ganglia in vivo. Brain Res. 2000;871(2):181–91.

O'Connor AB, Schwid SR, Herrmann DN, Markman JD, Dworkin RH. Pain associated with multiple sclerosis: systematic review and proposed classification. Pain. 2008;137(1):96–111.

Olafson RA, Rushton JG, Sayre GP. Trigeminal neuralgia in a patient with multiple sclerosis. An autopsy report. J Neurosurg. 1966;24(4):755–9.

Oyagi S, Ito J, Honjo I. Topographic study of the feline trigeminal ganglion via the horseradish peroxidase tracer method. Brain Res. 1989;476(2):382–3.

Peker S, Kurtkaya O, Uzun I, Pamir MN. Microanatomy of the central myelin-peripheral myelin transition zone of the trigeminal nerve. Neurosurgery. 2006;59(2):354–9; discussion 354–9.

Peker S, Dincer A, Necmettin Pamir M. Vascular compression of the trigeminal nerve is a frequent finding in asymptomatic individuals: 3-T MR imaging of 200 trigeminal nerves using 3D CISS sequences. Acta Neurochir (Wien). 2009;151(9):1081–8.

Rappaport ZH, Govrin-Lippmann R, Devor M. An electron-microscopic analysis of biopsy samples of the trigeminal root taken during microvascular decompressive surgery. Stereotact Funct Neurosurg. 1997; 68(1–4 Pt 1):182–6.

Resnick DK, Jannetta PJ, Lunsford LD, Bissonette DJ. Microvascular decompression for trigeminal neuralgia in patients with multiple sclerosis. Surg Neurol. 1996;46(4):358–61; discussion 361–52.

Rushton JG, Olafson RA. Trigeminal neuralgia associated with multiple sclerosis. A case report. Arch Neurol. 1965;13(4):383–6.

Sekula Jr RF, Frederickson AM, Jannetta PJ, Quigley MR, Aziz KM, Arnone GD. Microvascular decompression for elderly patients with trigeminal neuralgia: a prospective study and systematic review with meta-analysis. J Neurosurg. 2011;114(1):172–9.

Sekula Jr RF, Frederickson AM, Branstetter BF, et al. Thin-slice T2 MRI imaging predicts vascular pathology in hemifacial spasm: a case–control study. Mov Disord. 2014;29(10):1299–303.

Shinder V, Govrin-Lippmann R, Cohen S, et al. Structural basis of sympathetic-sensory coupling in rat and human dorsal root ganglia following peripheral nerve injury. J Neurocytol. 1999;28(9):743–61.

Sindou M, Howeidy T, Acevedo G. Anatomical observations during microvascular decompression for idiopathic trigeminal neuralgia (with correlations between topography of pain and site of the neurovascular conflict). Prospective study in a series of 579 patients. Acta Neurochir (Wien). 2002;144(1):1–12; discussion 12–3.

Tyler-Kabara EC, Kassam AB, Horowitz MH, et al. Predictors of outcome in surgically managed patients with typical and atypical trigeminal neuralgia: comparison of results following microvascular decompression. J Neurosurg. 2002;96(3):527–31.

6 Surgical Technique of Microvascular Decompression Surgery for Trigeminal Neuralgia

Jun Zhong and Hui Sun

Abstract

As an etiological treatment of trigeminal neuralgia, the microvascular decompression (MVD) surgery has been popularized around the world for more than half a century. However, as a functional operation in the cerebellopontine angle, this process should be refined to enhance the cure rate and minimize the complication. After accomplishment of more than 6000 MVDs, we've learned something concerning the operative technique: (1) the principle of MVD is to separate the neurovascular confliction rather than isolation with prostheses; (2) identification of the conflict site is important, which relies upon a good exposure; (3) a satisfactory working space can be established by an appropriate positioning and a close-to-the-sigmoid craniectomy as well as a caudorostral approach; (4) a sharp dissection of arachnoids leads to a maximal visualization of the entire intracranial course of the nerve root; (5) all the vessels contacting the trigeminal nerve root should be treated; and (6) the dura should be closed with watertight stitches at the end. In this chapter, every single step of the procedure was detailed.

Keywords

Microvascular decompression • Surgical technique • Trigeminal neuralgia

According to the classification of *the International Headache Society*, trigeminal neuralgia can be classified as idiopathic (primary) and symptomatic (secondary) (Olesen and Steiner 2004). Whatever, the diseases are etiologically caused by cerebellovascular compression of the trigeminal root, no matter directly or indirectly pushed by neoplasms or adhesions in the cerebellopontine angle (Shulev et al. 2011; Zhong et al. 2008; de Lange et al. 1986). Therefore, separation of the nerve from the offending vessel(s) seems to be an ideal treatment. Since microvascular decompression (MVD) was first introduced by Dandy and then popularized by Jannetta in the last century, it has been thought to be the most

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reasonable technique for treatment of trigeminal neuralgia (Kellogg et al. 2010; Jannetta 2007; Haines et al. 1979; McLaughlin et al. 1999; Zhong et al. 2012). Nevertheless, this sort of surgical process is still with risk because of those delicate cerebellopontine structures, and some patients cannot totally relieve their symptoms postoperatively (Zhong et al. 2012; Sindou et al. 2002, 2007; Xia et al. 2014; Barker et al. 1996). As a result, the surgical techniques of MVD need to be further discussed (Devor et al. 2002; Hong et al. 2011; Zhong et al. 2014).

6.1 Indications

MVD is appropriate for most of the patients with trigeminal neuralgia.

Generally, MVD is indicated for all the patients suffering from drug-resistant trigeminal neuralgia as long as their general conditions do not contraindicate general anesthesia. We would like to point out that old age is not an unconditional contraindication for MVD. Instead, it is relatively easier to operate on the aged for they have a wider subarachnoid space as a result of brain atrophy (Sekula et al. 2008, 2011). Only those with decompensated dysfunction of vital organs should be evaluated cautiously. Hence, we suggest performing the surgery in the early stage before the patient's quality of life is awfully influenced. Especially, for those undernourished because of less eating to avoid an attack of severe pain induced by oral movement, a prompt surgery is encouraged (Lemos et al. 2011; El-Ghandour 2010). In addition, MVD is indicated in patients with coexistent trigeminal neuralgia and hemifacial spasm (Zhong et al. 2011; Cook and Jannetta 1984), glossopharyngeal neuralgia, (Wang et al. 2014) or Bell palsy (Jiao et al. 2013).

6.2 Anesthesia

Intracranial pressure and brain pulsation should be well controlled.

General anesthesia is used for the process. Besides imperceptions and relaxations, decrease of intracranial pressure (ICP) should not be neglected. The dura should not be opened until a satisfactory ICP is ready. Otherwise, the brain tissue might be squeezed out and lead to cerebellar contusions. Hyperventilation can be used temporarily to reduce brain volume and decrease ICP when necessary. Meanwhile, the brain pulsation should be well controlled as it may generate a terrible cerebrospinal fluid (CSF) tide. The microscopic imagings of the trigeminal root and offending artery as well as petrosal veins will be transferred ceaselessly due to the changing optic refraction arose by CSF fluctuation. Hence, it will be a challenge to dissect the trigeminal root in such an SCF rapid – not to mention the deep and narrow operation field. As brain pulsation is generated by the heartbeat via the transmission of the arterial pulse, ventricular tachyarrhythmia should be avoided. Therefore, β-adrenergic blockers will be helpful. Basically, small doses of esmolol can be used repeatedly to control the perioperative tachycardia or hypertension.

6.3 Positioning

A proper positioning contributes to a satisfactory exposure.

We place the patient in a park bench position (3/4 lateral prone decubitus). This position is superior to supine or full prone position because it obviates the need to turn the patient's head into an uncomfortable position and therefore decreases the risk of postoperative neck pain, especially for

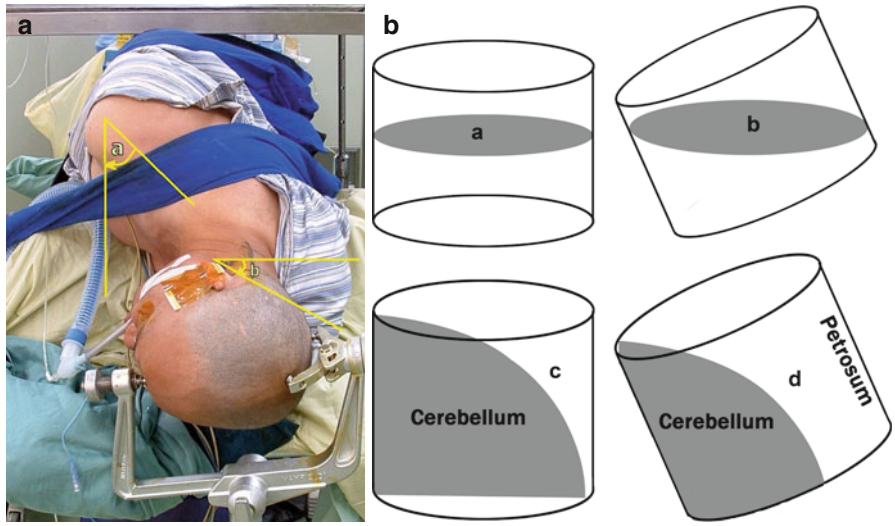

Fig. 6.1 Positioning. (**a**) The patient is placed in a 3/4 lateral prone decubitus position with the ipsilateral shoulder being slanted forward (∠*a*) and pulled away from the head. The patient's head is turned back 15° from the horizontal plane (∠*b*). (**b**) The diagrammatic drawing exhibits the benefit of the head-inclined position. It offers a bigger visualizable area (*B*) than a flat position (*A*) does. Because of its own gravity, the cerebellum falls away from the petrosal bone and provides more working space (*D*) than a flat position (*C*) does

obese patients with generous supraclavicular fat pads. It is necessary to point out that the contralateral shoulder should be close to the edge of the bed so that the surgeon can easily reach the surgical site. Meanwhile, the ipsilateral shoulder should be pulled away from the head by a shoulder belt so as to create a satisfactory working space, which facilitates the instruments getting in and out of the surgical field. A fixation frame is used to hold the patient's head stable and make it possible to apply retractor system when necessary. However, we no longer use retractors even in cases of secondary trigeminal neuralgia resulting from CPA masses since we have chosen an oblique position of the patients' head. This inclined position with the patient's head turning back 15° from the level surface facilitates the cerebellum to fall away under its own gravity from the petrosal bone and obviates the need of retractors. Therefore, a good exposure is achieved by rational positioning of the patient's head rather than retracting of cerebellum (Fig. 6.1) (Zhong et al. 2012, 2014).

6.4 Incision

An extra-large incision may not be good for exposure.

Although transversal, arc, or reverse 'U' shape incisions have been reported (Cohen-Gadol 2011), we choose a vertical linear incision. It is laterally parallel to the hairline and crosses the inion-zygomatic line with 1/3 above and 2/3 below. Nowadays, we have adopted a mastoid retractor to hold the incision and no scalp clip is needed. Owing to the limitation of the retractor's open angle, an incision longer than 7 cm is not really necessary. To save a good blood supply, undue coagulation should be avoided during the incision making and a quick retraction of the incision is recommended. As the operator is sitting behind the patient, a more medial peeling will provide a good sight (Fig. 6.2).

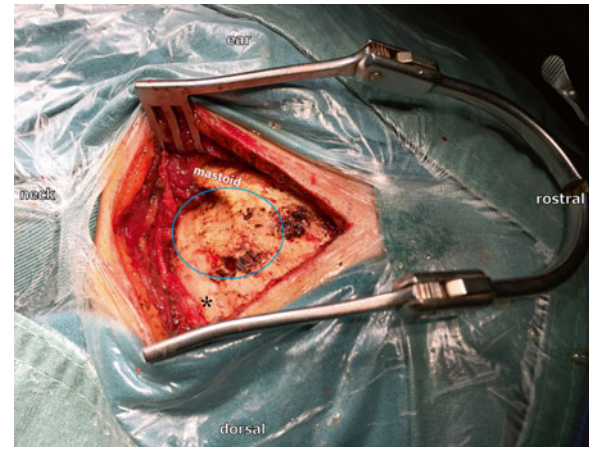

Fig. 6.2 Incision and craniectomy. A vertical linear incision is suggested, which parallels laterally to the hairline. The incision is held by a mastoid retractor without scalp clip. Owing to the limitation of the retractor's open angle, an incision longer than 7 cm is not really necessary. As the operator is sitting behind the patient, a more medial peeling (*) will provide a good sight. A *circle* marks the location of craniectomy, which should be very close to the mastoid and lateral enough to the sigmoid sinus

6.5 Craniectomy

A much lateral craniectomy close to the sigmoid sinus is recommended.

Basically, a craniectomy of <3 cm in diameter is enough for most cases. The edge of the sigmoidal sinus should be exposed to ensure an ideal surgical corridor. To avoid dural sinus injury, we prefer craniectomy with pneumatic drill and Kerrison rongeur to craniotomy with milling cutters. Bone dust and chips were preserved for the later cranioplasty. The bone over the sigmoid sinus should be removed in small pieces. Bone wax is effective for homeostasis at the edge of dural sinuses. To obtain a good working angle, the mastoid antrum could be opened if necessary, but it should be immediately waxed to prevent infection and CSF leak (Fig. 6.2).

6.6 Durotomy

A good durotomy provides a maximal surgical corridor toward the conflict site.

Before dural opening, a thorough irrigation is necessary. It is not merely for bone scraps cleaning but mainly for double checks of bleeding. Even a little oozing of blood may flow in your surgical field continuously, which may interfere with your vision and mood terribly. In spite of a variety of dural tailoring, we now prefer an arc-shaped cutting with its chord paralleling to the lateral rim of the craniectomy. After the dural mater is sutured back with double knotting, the suture thread remains in place (without cutting off), which facilitates tightening when necessary during the procedure. This pattern of dural opening leaves the majority of the dura on the cerebellum, which avails the protection of the cerebellar hemisphere during the process (Fig. 6.3). To avoid shrinkage of the dura due to the heat generated by the operating microscope's lamp aimed at the surgical field during the intradural portion of the operation, pieces of wet Gelfoams are placed over the dura.

6.7 Exposure

A satisfactory exposure is obtained by sharp microdissection as well as proper position and craniectomy rather than harsh retraction.

In our series of 6000 MVDs, the main reason of a failed surgery was that the exact offending vessel(s) were not recognized or the neurovascular conflict site was inaccessible intraoperatively. So

we believe a full exposure of the entire trigeminal nerve root course is the key to obtain a good result. Advancing toward the target starts with a Cottonoid placed over the cerebellum and draining CSF slowly. Usually, an unhurried suction drainage of CSF and an ample adhesiolysis are effective enough to achieve brain relaxation, and no mannitol or lumbar puncture is needed for most of the cases. We do not use retracting blades because a narrow suction tube allows more mobility and can actually afford more working space than a wider spatula does during the procedure at a moment when a specific area is dissected. As a matter of fact, with a good knowledge of the regional anatomy, one does not have to visualize the whole area of CPA while operating at a particular site (but those surrounding structures should always be in mind). Basically, a Fukushima teardrop suction tube, a pair of microscissors, a microdissector, and a pair of bipolar coagulation forceps are enough to complete all the intracranial manipulation. Instead of the ordinary gun-shape forceps, a self-irrigating bipolar forceps can keep the Cottonoid over the cerebellum moist all the time and avoid cerebellar contusion. The action of clamping an artery should be avoided, which may cause vasospasm (Fig. 6.4).

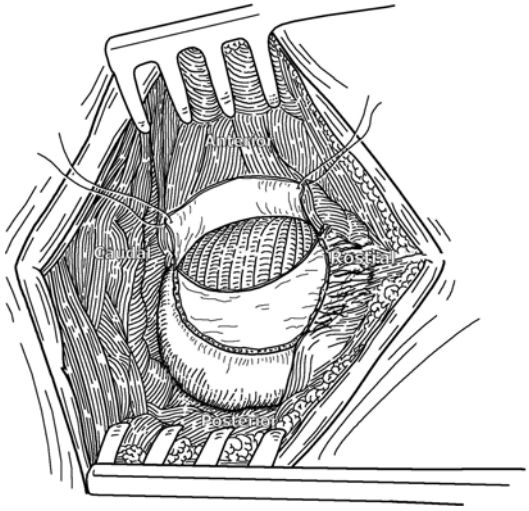

Fig. 6.3 Durotomy. The dural membrane is cut in an arc shape with its chord paralleling to the lateral rim of the craniectomy. This pattern of dural opening leaves the majority of the dura on the cerebellum, which avails the protection of the cerebellar hemisphere during the process. After the dural mater is sutured back with double knotting, the suture thread remains in place (without cutting off), which facilitates tightening when necessary during the procedure

6.8 Approach

A caudorostral (via cerebellar fissures) approach is suggested.

The protection and management of petrosal veins poses the main challenges and risks during the process. In early cases, we approached the

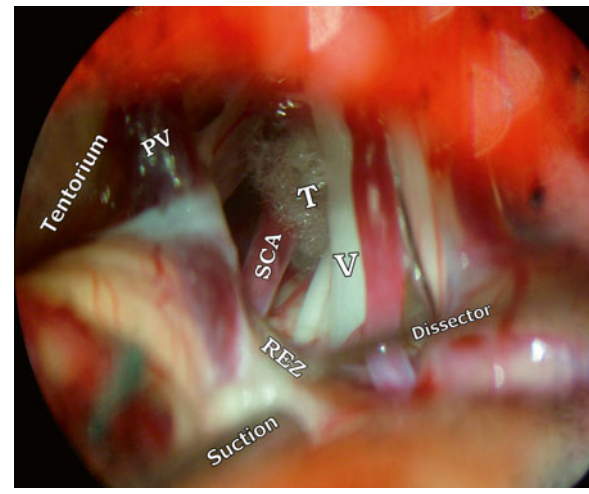

Fig. 6.4 Exposure. A satisfactory exposure can be achieved without using retracting blades. With a slow drainage of CSF and an ample adhesiolysis, a surgical corridor can be established. Actually, a narrow suction tube affords more working space than a wider spatula does at a moment when a specific area is dissected. *V* trigeminal nerve, *PV* petrosal vein, *SCA* superior cerebellar artery, *T* Teflon waddings, *REZ* root entrance zone

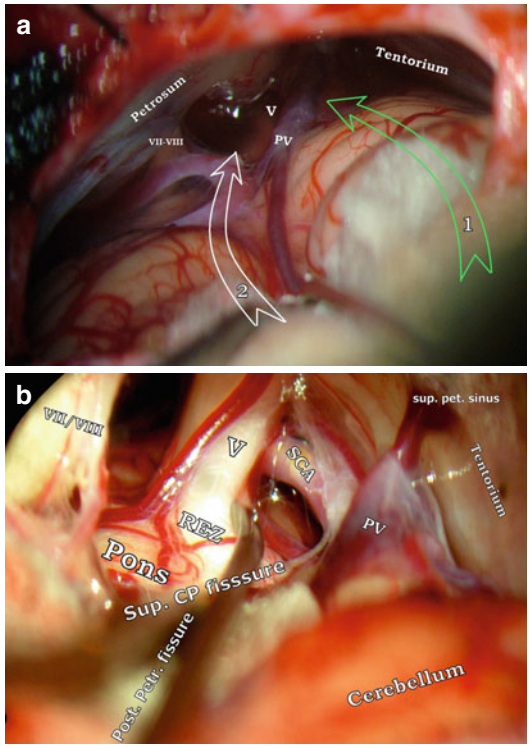

Fig. 6.5 A caudorostral approach. (**a**) To yield petrosal veins (*PV*), we choose the via-cerebellar-fissure approach (*2*) instead of the infratentorial superior-lateral cerebellar approach (*1*). (**b**) After dissection of the arachnoid membranes around the posterior petrous and the superior cerebellopontine fissures, usually the REZ of the trigeminal nerve (*V*) could be turned up in your sight directly. *VII-VIII* facial and vestibulocochlear nerves

trigeminal nerve from the superolateral aspect of the cerebellum, and petrosal veins were often obstructing the access path. We used to sacrifice these veins to prevent unintentionally tearing at its entry to the superior petrosal sinus, which was tough to manage. In that case, compression with Gelfoam was the only way for homeostasis, while coagulation could only make things worse. However, when a Gelfoam was used to cover the bleeding point, working space became much narrower and the following procedures could be very difficult. To detour off those petrosal veins, we now dissect the arachnoid membranes around the petrous and the superior cerebellopontine fissures. Basically, the REZ of the V nerve will be directly in your sight with the cerebellar fissures opened thoroughly (Fig. 6.5) (Zhu et al. 2014).

6.9 Decompression

Identification of the neurovascular confliction: a full-way inspection of the nerve root increases the chance.

The trigeminal nerve is then circumferentially inspected along its entire intracranial course from its REZ at the brainstem laterally to its entrance into the Meckel's cave. The neurovascular relationship is carefully studied, and any vessel related to the V nerve is moved away, followed by placement of soft Teflon wadding between them. Those venules adhered to or even go through the trigeminal nerve can be coagulated and cut. Due to structural difference, the central portion of trigeminal nerve is more vulnerable to compression. A study found that the average length of the central portion of the trigeminal nerve is 2.6 mm, while for facial nerve, it is only 1.7 mm (De Ridder et al. 2002). Accordingly, the patients with trigeminal neuralgia have more chances of lateral confliction than those with hemifacial spasm (Katusic et al. 1990; Auger and Whisnant 1990). Nevertheless, to achieve a high cure rate, a thorough exposure of the whole intracranial nerve course is recommended. Comparing to hemifacial spasm, the neurovascular confliction in trigeminal cases is more complicated (Sindou et al. 2002). A superior cerebellar artery along the shoulder of the root entry zone is commonly seen compressing the caudal side of the trigeminal nerve ventromedially. Mobilization of the arterial loop often discloses a site of discoloration (or even an indent) along the nerve, which confirms that the intended pathology is found and is predictive of a good outcome after surgery. The possibility of multiple offending vessels (arterial and/or venous loops) should be excluded with careful inspection.

The Principle of Decompression: separation of the neurovascular conflict rather than insertion of prosthesis between them

As a matter of fact, a skillful driving of a microdissector in assistance with a fine suction tube by the operator's two hands is enough to complete the decompression process, including removal of the offending vessel(s) and placement of Teflon wadding piece by piece, and no other instruments controlled by a third hand are needed in such a small room. Moving the vessel with forceps is not recommended. The substance of decompression should be separation of the neurovascular confliction rather than insertion of prosthesis between the nerve and the vessel. Actually, the role of the Teflon is to keep the offending vessel from rebounding; therefore, it is unnecessary to be always put at conflict site (Fig. 6.6). Furthermore, those offending arteries are supposed to be positioned anatomically. As the SCA or AICA is the most common culprit, they should be transposed rostrally or caudally though they may occur any way in the nerve root (Jannetta 2007). We advocate simple and safe techniques in MVD surgeries (Zhong 2012). As the ordinary decompression process is usually not so complicated, we merely discuss some extreme cases here.

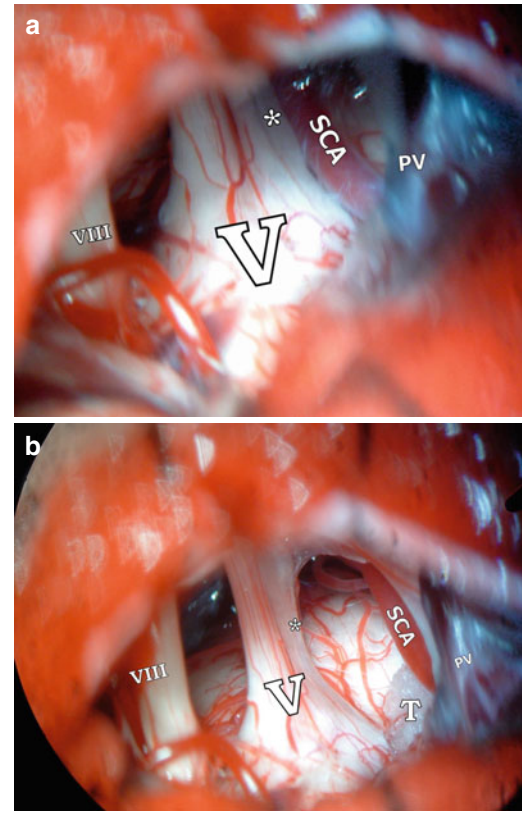

Fig. 6.6 The decompression process. The principle of decompression is separation of the neurovascular conflict rather than insertion of prosthesis between them. The Teflon may be placed anyway as long as it could keep the vessel from rebounding. (**a**) In this case, the superior cerebellar artery (*SCA*) was identified as the culprit, which contacted with trigeminal nerve (*V*) rostrally. (**b**) After the offending artery was separated from the nerve, a Teflon wadding (*T*) was not placed at the original conflict site (*). VIII the vestibulocochlear nerve, PV petrosal vein

6.9.1 Ectatic Vertebrobasilar Artery Complex

It will be tough when the surgeon encounters an incompliant and ecstatic vertebrobasilar artery complex (VBA) during the MVD (Linskey et al. 1994; Mittal and Mittal 2011; Ma et al. 2013). In that case, the point of the surgery is to move the VBA proximally. Basically, a tortuous vertebral artery is usually found ventrally to the caudal cranial nerves in these cases. Through the interstices between the caudal nerves, the proximal segment of VBA is mobilized caudolaterally, and then small pieces of shredded Teflon are gradually placed between the VBA and the medulla oblongata to keep the artery free from the brainstem. With the arachnoid membrane around the nerves opened thoroughly, the cerebellum is gradually raised until the pontomedullary sulcus is visualized. Then, ventrally to the facial nerve, more pieces of Teflon wadding are added between the VBA and the pons in order to further remove the VBA caudolaterally. By that time, the neurovascular confliction can be distinguished without difficulty – sometimes, the offending artery may have left from the trigeminal nerve root spontaneously (Yang et al. 2012). This lateroinferior cerebellar approach has the advantage of evading veins that may block the surgical corridor when the infratentorial approach was adapted (Fig. 6.7) (Sekula et al. 2011; Lemos et al. 2011).

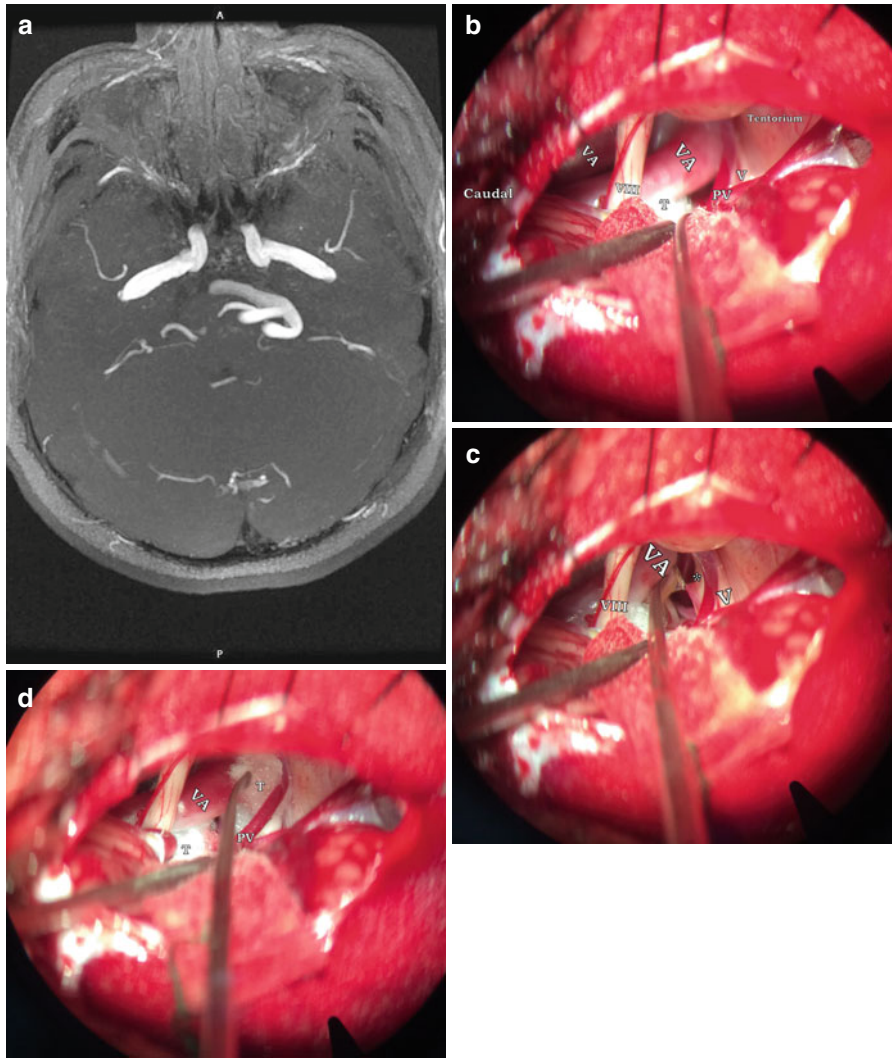

Fig. 6.7 Decompression of an ectatic vertebrobasilar artery. (**a**) The MRI delineated an ectatic vertebrobasilar artery that shifted to the ipsilateral side. (**b**) As the cerebellum being raised, both the vertebral arteries (*VA*) were visualized in the surgical field and the trigeminal nerve (*V*) was covered by a tortuous VA. Before dissection of the trigeminal root, the covering VA should be freed beginning from the proximal segment. In this photo, the proximal VA had been mobilized caudolaterally and held by Teflon waddings. Without spatula, these waddings had been buried by the cerebellar hemisphere while the section tuber moved rostrally, but only those Teflon (*T*) in the level of acoustic nerve (*VIII*) were visible. (**c**) With the VA being moved caudally, a dent (*) was found in the caudal V nerve. (**d**) Teflon waddings were inserted between the nerve and VA and the dorsal petrosal vein (*PV*) as well as a small artery

6.9.2 Venous Compression

As a high recurrence rate has been reported, the venous compression is worth addressing here (Hong et al. 2011; Helbig et al. 2009; Kimura et al. 1999; Sato et al. 1979; Lee et al. 2000; Li et al. 2014; Matsushima et al. 2004). A thorough understanding of the venous anatomy around the trigeminal nerve is crucial to identify the culprit veins accurately. The veins that commonly compress the trigeminal nerve are tributaries of the superior petrosal vein, namely, the transverse pontine vein, the pontotrigeminal vein and the vein of cerebellopontine fissure, and the vein of middle

cerebellar peduncle (Choudhari 2007). The transverse pontine vein is usually attached to the superior-medial or inferior surface of the trigeminal nerve just in front of the Meckel's cave. When the culprit vein adheres firmly to the brain stem and cranial nerve, coagulating and cutting techniques are preferred in the decompressing process. However, large veins ($\Phi > 2$ mm) should avoid sacrificing unless they are surely not related to deep venous drainage of the brain stem (Zhong et al. 2008; Hong et al. 2011; Masuoka et al. 2009).

6.9.3 Intraneural Vessels

In very rare instances, a vessel may cross through the trigeminal nerve (Helbig et al. 2009; Kimura et al. 1999; Tashiro et al. 1991). For those very small arteries (arterioles) or veins (venules), coagulation and cutoff could be used. For those that situate very laterally to the edge, some nerve fibers of the trigeminal nerve root could be cut off to release the offending vessel. However, improper management of big middle intraneural vessels may lead to trigeminal nerve injury and result in uncomfortable neuropathy and numbness (including corneal hypoesthesia). In that case, we suggest fully dissecting to mobilize the artery or vein, followed by wrapping with tiny Teflon wadding (Zheng et al. 2012).

6.10 Closure

The dura should be closed in a watertight pattern.

At the end of the operation, the surgical field should be well irrigated with 37 °C normal saline to vent air as well as to assure that there is no bleeding. Meanwhile, attention should be paid to make sure the implanted Teflon is stable under the flow of CSF. Before dural closure, it is suggested to place a piece of wet Gelfoam over the cerebellum, which prevents not only epidural hemorrhage from flowing into the subdural space but also accidental injury to the brain tissue while suturing. In order to prevent CSF leakage, the dura should be closed tightly. The surgeon should begin the stitch process from the lowest level to prevent blood inflow from outside. Sometimes, it is difficult to complete a watertight closure due to dura defect. In that case, a muscle patch can be used to repair the defect (Fig. 6.8). Then, the dura is sealed with glue and covered by an artificial dura plus bone chips to reconstruct the normal anatomy. Finally, the muscles, subcutaneous tissues, and the scalp are sutured layer by layer, and no drainage is required.

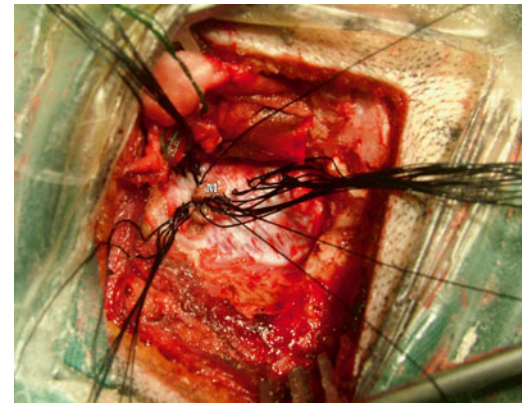

Fig. 6.8 Closure. The dural membrane should be always stitched in a watertight pattern as far as possible. In case of dura defect, a muscle patch (*M*) is the first choice to be used to repair the defect. Artificial materials are not suggested

References

Auger RG, Whisnant JP. Hemifacial spasm in rochester and olmsted county, minnesota, 1960 to 1984. Arch Neurol. 1990;47:1233–4.

Barker 2nd FG, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med. 1996;334:1077–83.

Choudhari KA. Superior petrosal vein in trigeminal neuralgia. Br J Neurosurg. 2007;21:288–92.

Cohen-Gadol AA. Microvascular decompression surgery for trigeminal neuralgia and hemifacial spasm: naunces of the technique based on experiences with 100 patients and review of the literature. Clin Neurol Neurosurg. 2011;113:844–53.

Cook BR, Jannetta PJ. Tic convulsif: results in 11 cases treated with microvascular decompression of the fifth

and seventh cranial nerves. J Neurosurg. 1984;61: 949–51.

de Lange EE, Vielvoye GJ, Voormolen JH. Arterial compression of the fifth cranial nerve causing trigeminal neuralgia: angiographic findings. Radiology. 1986; 158:721–7.

De Ridder D, Moller A, Verlooy J, Cornelissen M, De Ridder L. Is the root entry/exit zone important in microvascular compression syndromes? Neurosurgery. 2002;51:427–33; discussion 433–4.

Devor M, Amir R, Rappaport ZH. Pathophysiology of trigeminal neuralgia: the ignition hypothesis. Clin J Pain. 2002;18:4–13.

El-Ghandour NM. Microvascular decompression in the treatment of trigeminal neuralgia caused by vertebrobasilar ectasia. Neurosurgery. 2010;67:330–7.

Haines SJ, Martinez AJ, Jannetta PJ. Arterial cross compression of the trigeminal nerve at the pons in trigeminal neuralgia. Case report with autopsy findings. J Neurosurg. 1979;50:257–9.

Helbig GM, Callahan JD, Cohen-Gadol AA. Variant intraneural vein-trigeminal nerve relationships: an observation during microvascular decompression surgery for trigeminal neuralgia. Neurosurgery. 2009;65: 958–61; discussion 961.

Hong W, Zheng X, Wu Z, Li X, Wang X, Li Y, Zhang W, Zhong J, Hua X, Li S. Clinical features and surgical treatment of trigeminal neuralgia caused solely by venous compression. Acta Neurochir (Wien). 2011;153:1037–42.

Jannetta PJ. Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neuralgia. 1967. J Neurosurg. 2007;107:216–9.

Jiao W, Zhong J, Sun H, Zhu J, Zhou QM, Yang XS, Li ST. Microvascular decompression for the patient with painful tic convulsif after bell palsy. J Craniofac Surg. 2013;24:e286–9.

Katusic S, Beard CM, Bergstralh E, Kurland LT. Incidence and clinical features of trigeminal neuralgia, rochester, minnesota, 1945–1984. Ann Neurol. 1990;27:89–95.

Kellogg R, Pendleton C, Quinones-Hinojosa A, Cohen-Gadol AA. Surgical treatment of trigeminal neuralgia: a history of early strides toward curing a "Cancerous acrimony". Neurosurgery. 2010;67:1419–25; discussion 1425.

Kimura T, Sako K, Tohyama Y, Yonemasu Y. Trigeminal neuralgia caused by compression from petrosal vein transfixing the nerve. Acta Neurochir (Wien). 1999; 141:437–8.

Lee SH, Levy EI, Scarrow AM, Kassam A, Jannetta PJ. Recurrent trigeminal neuralgia attributable to veins after microvascular decompression. Neurosurgery. 2000;46:356–61; discussion 361–2.

Lemos L, Alegria C, Oliveira J, Machado A, Oliveira P, Almeida A. Pharmacological versus microvascular decompression approaches for the treatment of trigeminal neuralgia: clinical outcomes and direct costs. J Pain Res. 2011;4:233–44.

Li GW, Zhang WC, Yang M, Ma QF, Zhong WX. Clinical characteristics and surgical techniques of trigeminal neuralgia caused simply by venous compression. J Craniofac Surg. 2014;25:481–4.

Linskey ME, Jho HD, Jannetta PJ. Microvascular decompression for trigeminal neuralgia caused by vertebrobasilar compression. J Neurosurg. 1994;81:1–9.

Ma X, Sun X, Yao J, Ni S, Gong J, Wang J, Li X. Clinical analysis of trigeminal neuralgia caused by vertebrobasilar dolichoectasia. Neurosurg Rev. 2013;36:573–7; discussion 577–8.

Masuoka J, Matsushima T, Hikita T, Inoue E. Cerebellar swelling after sacrifice of the superior petrosal vein during microvascular decompression for trigeminal neuralgia. J Clin Neurosci. 2009;16:1342–4.

Matsushima T, Huynh-Le P, Miyazono M. Trigeminal neuralgia caused by venous compression. Neurosurgery. 2004;55:334–7; discussion 338–9.

McLaughlin MR, Jannetta PJ, Clyde BL, Subach BR, Comey CH, Resnick DK. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999;90:1–8.

Mittal P, Mittal G. Painful tic convulsif syndrome due to vertebrobasilar dolichoectasia. J Neurosci Rural Pract. 2011;2:71–3.

Olesen J, Steiner TJ. The international classification of headache disorders, 2nd edn (icdh-ii). J Neurol Neurosurg Psychiatry. 2004;75:808–11.

Sato O, Kanazawa I, Kokunai T. Trigeminal neuralgia caused by compression of trigeminal nerve by pontine vein. Surg Neurol. 1979;11:285–6.

Sekula RF, Marchan EM, Fletcher LH, Casey KF, Jannetta PJ. Microvascular decompression for trigeminal neuralgia in elderly patients. J Neurosurg. 2008;108: 689–91.

Sekula Jr RF, Frederickson AM, Jannetta PJ, Quigley MR, Aziz KM, Arnone GD. Microvascular decompression for elderly patients with trigeminal neuralgia: a prospective study and systematic review with meta-analysis. J Neurosurg. 2011;114:172–9.

Shulev Y, Trashin A, Gordienko K. Secondary trigeminal neuralgia in cerebellopontine angle tumors. Skull Base. 2011;21:287–94.

Sindou M, Howeidy T, Acevedo G. Anatomical observations during microvascular decompression for idiopathic trigeminal neuralgia (with correlations between topography of pain and site of the neurovascular conflict). Prospective study in a series of 579 patients. Acta Neurochir (Wien). 2002;144:1–12; discussion 12–3.

Sindou M, Leston J, Decullier E, Chapuis F. Microvascular decompression for primary trigeminal neuralgia: long-term effectiveness and prognostic factors in a series of 362 consecutive patients with clear-cut neurovascular conflicts who underwent pure decompression. J Neurosurg. 2007;107:1144–53.

Tashiro H, Kondo A, Aoyama I, Nin K, Shimotake K, Nishioka T, Ikai Y, Takahashi J. Trigeminal neuralgia caused by compression from arteries transfixing the nerve. Report of three cases. J Neurosurg. 1991;75: 783–6.

Wang YN, Zhong J, Zhu J, Dou NN, Xia L, Visocchi M, Li ST. Microvascular decompression in patients with

coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Acta Neurochir (Wien). 2014;156:1167–71.

Xia L, Zhong J, Zhu J, Wang YN, Dou NN, Liu MX, Visocchi M, Li ST. Effectiveness and safety of microvascular decompression surgery for treatment of trigeminal neuralgia: a systematic review. J Craniofac Surg. 2014;25:1413–7.

Yang XS, Li ST, Zhong J, Zhu J, Du Q, Zhou QM, Jiao W, Guan HX. Microvascular decompression on patients with trigeminal neuralgia caused by ectatic vertebrobasilar artery complex: technique notes. Acta Neurochir (Wien). 2012;154:793–7; discussion 797.

Zheng X, Feng B, Hong W, Zhang W, Yang M, Tang Y, Zhong J, Hua X, Li S. Management of intraneural vessels during microvascular decompression surgery for trigeminal neuralgia. World Neurosurg. 2012;77:771–4.

Zhong J. An ideal microvascular decompression technique should be simple and safe. Neurosurg Rev. 2012;35:137–40; author reply 140.

Zhong J, Li ST, Xu SQ, Wan L, Wang X. Management of petrosal veins during microvascular decompression for trigeminal neuralgia. Neurol Res. 2008;30:697–700.

Zhong J, Zhu J, Li ST, Guan HX. Microvascular decompressions in patients with coexistent hemifacial spasm and trigeminal neuralgia. Neurosurgery. 2011;68:916–20; discussion 920.

Zhong J, Li ST, Zhu J, Guan HX, Zhou QM, Jiao W, Ying TT, Yang XS, Zhan WC, Hua XM. A clinical analysis on microvascular decompression surgery in a series of 3000 cases. Clin Neurol Neurosurg. 2012;114:846–51.

Zhong J, Zhu J, Sun H, Dou NN, Wang YN, Ying TT, Xia L, Liu MX, Tao BB, Li ST. Microvascular decompression surgery: surgical principles and technical nuances based on 4000 cases. Neurol Res. 2014;36:882–93.

Zhu J, Zhong J, Jiao W, Zhou QM, Guan HX, Dou NN, Wang YN, Xia L, Li ST. Via-cerebellar-fissures approach for microvascular decompression of trigeminal nerve. J Craniofac Surg. 2014;25:1438–40.

7 Surgical Techniques of Microvascular Decompression for Hemifacial Spasm

Shi-Ting Li and Hui Sun

Abstract

Microvascular decompression (MVD) has been accepted as the most effective treatment for hemifacial spasm (HFS). Many surgeons from different countries and areas have concentrated on this procedure, aiming to increase the cure rate and lower the risks. In this chapter we introduced our viewpoints on MVD surgery, based on our experience from about 1200 MVD surgeries each year. We discussed the whole course of management, from diagnosis to patient selection, from anesthesia to surgical procedure, and from preoperative evaluation to postoperative management. We put our emphasis on the techniques of MVD, in which part we discussed the approaches, exploration range, and decompression method. We described some challenging situations, such as large sclerosed vertebral artery, degeneration of facial nerve, offending vessels between facial nerve and acoustic nerve, as well as offending vessels going through the fibers of facial nerve. We also discussed some special cases, including persistent AMR waves, no vascular compression found, and dual side HFS. We developed a new criterion for ending an MVD surgery. We hope our experiences would be helpful for surgeons who are specialized in cranial nerve surgeries.

Keywords

Microvascular decompression (MVD) • Hemifacial spasm (HFS) • Diagnosis • Surgical treatment • Complication

7.1 Diagnosis and Differential Diagnosis of HFS

Hemifacial spasm (HFS) refers to recurrent paroxysmal, involuntary twitching of the facial muscles (orbicularis oculi muscle, expression of muscle, orbicularis oris) on one side or both.

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It usually aggravates when the patient is emotional or nervous. Severe HFS even leads to difficulties in eye opening, mouth askew, and twitching-like noise in the ear. HFS can be classified into two groups: typical and atypical. Typical HFS means the symptom begins at the eyelid and spreads to the lower part of the face. Atypical HFS means the spasm starts at the lower part of the face and spreads upward to the eyelid and frontal muscle. Atypical HFS is relatively rare, while most are typical. HFS occurs mostly in elder patients, with females more than males. However, the ages tend to be younger and younger. Although in most patients HFS occurs on one side, bilateral HFS is not rare.

7.1.1 Diagnosis of HFS

The diagnosis of HFS relies mostly on its characteristic manifestations. When the manifestations are not typical, examinations may help, including electrophysiological examinations, radiology, and carbamazepine trial treatment. Electrophysiologic examinations include electromyography (EMG) and abnormal muscle response (AMR) (also known as lateral spread response, LSR). EMG can show a high-frequency spontaneous potential (the frequency can be as high as 150 Hz). AMR is a HFS-specific reaction. Positive AMR supports the diagnosis of HFS. Radiology includes CT and MRI, which can reveal possible intracranial mass lesions. 3D-TOF-MRA is helpful to show the vascular distributions around the facial nerve. HFS patients are generally responsive to carbamazepine therapy at the beginning of the disease (some few patients are not), so the trial can help diagnose.

7.1.2 Differential Diagnosis of HFS

HFS needs to be differentiated from bilateral blepharospasm, major syndrome, masseter spasm, sequelae of facial paralysis, and other facial dystonia diseases.

7.1.2.1 Bilateral Blepharospasm

Patients usually present with bilateral recurrent involuntary eye closing and tear reduction. At onset, symptoms often occur simultaneously on both sides. Patients may have difficulty in opening eyes with the progress of the disease, but symptoms are always limited to bilateral eyelids as time goes.

7.1.2.2 Major Syndrome

Patients often present with bilateral recurrent involuntary eye closing at onset. But as time goes, involuntary muscle twitching occurs below the eye fissure, showing bilateral abnormal involuntary facial movements. As the condition deteriorates, muscle spasms gradually expand, even involving the neck and the muscles of the limbs and trunk.

7.1.2.3 Masseter Spasm

Patients present with unilateral or bilateral masticatory muscle spasm. Varying degrees of mandibular occlusal disorders and teeth and mouth opening problems may occur. The motor branch of trigeminal nerve is likely the cause.

7.1.2.4 Sequelae of Facial Paralysis

Patients present with paralysis of the ipsilateral facial muscle, ipsilateral mouth twitching, and synkinetic movement of the mouth and eyelid. It can be diagnosed according to the exact history of facial paralysis.

7.2 Indications and Contraindications of MVD

7.2.1 Indications of MVD

MVD is suggested for all HFS patients once diagnosed. As not all HFS patients are primary HFS, CT/MRI is essential to exclude secondary HFS. Diagnosis of primary HFS is the essential requirement for MVD. As the rate of progress of hemifacial spasm varies, and oral carbamazepine may be effective in most patients at early stage or with mild symptoms, MVD for primary HFS is indicated for the following conditions: (1)

Symptoms are severe and affect daily life and work, and the patient has a strong willingness for surgery. (2) The patient has tried medicine therapy or botulinum toxin treatment, but the result is poor or invalid, or there are drug allergies or severe side effects. (3) Recurrence after an MVD surgery. (4) The first MVD is invalid and it is believed that the decompression was inadequate. AMR remains positive. (5) Symptoms do not relieve or even deteriorate during follow-up.

7.2.2 Contraindications of MVD

MVD requires general anesthesia, so if the patient cannot tolerate general anesthesia, MVD is contraindicated, such as suffering from serious blood disorders or significant organ dysfunction (heart, lung, kidney, or liver). For vascular decompression surgery itself, there is no limit of age, but literatures suggest that in patients over 70 years old, complications of MVD is relatively high, and choosing MVD should be made with caution.

7.3 Preoperative Evaluation and Preparation

7.3.1 Evaluation of General Conditions

General physical condition assessment includes routine preoperative chest X-ray examination, ECG, blood coagulation, and liver and kidney function. In addition, the patient's blood pressure, blood sugar, and the potential risk of stroke should be assessed. Adjust blood pressure and blood sugar to normal range before surgery. Preoperative assessment of facial nerve function and hearing must also be done. For patients who have hearing impairments, the auditory pathways should be examined to exclude diseases of external auditory canal, middle ear, and inner ear. ENT doctors should be consulted when necessary. For patients who have loss of hearing on the contralateral side, preoperative assessment of hearing on the surgical side is even more important.

7.3.2 Imaging Evaluation

All patients with HFS must receive radiographic assessment before MVD. The best choice is magnetic resonance imaging (MRI). For patients who cannot take magnetic resonance imaging, a head CT scan should be performed. The purpose of MRI is to exclude intracranial lesions, such as tumors, cavernous hemangioma, cerebral arteriovenous malformation (AVM), posterior circulation aneurysms, cysts, and skull deformities. MRI examination is also valuable to show the presence of vascular contact to the facial nerve. It can even show the vessel's category, thickness, and extent of oppression to the facial nerve (Fig. 7.1). Especially, the three-dimensional time of flight magnetic resonance angiography (3D-TOF-MRA) has become a routine examina-

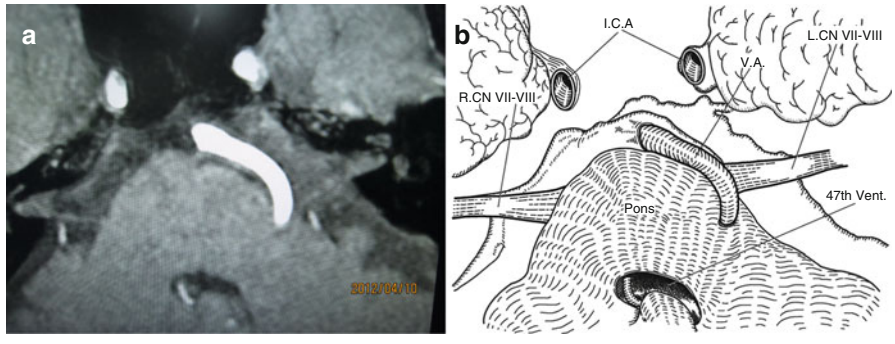

Fig. 7.1 MRI shows the vertebral artery shifts to the left side and compresses the facial nerve (The label "47th Vent." should be "4th Vent." in the schematic picture)

tion before MVD. 3D-TOF-MRA-based MRI imaging technology has been able to show the presence of all vascular contact to the facial nerve from 360° (picture). But it must be pointed out that the vessels MRI shows are not necessarily the real offending vessels. Meanwhile, a negative result of 3D-TOF-MRA does not necessarily mean that MVD surgery is contraindicated. However, the decision of performing MVD needs to be more careful. Therefore, it is necessary to check again whether the diagnosis of HFS is exact, and electrophysiological assessment should be referred when necessary.

7.3.3 Preoperative Electrophysiological Evaluation

Preoperative electrophysiological evaluation helps differential diagnosis of HFS and objective assessment of the level of facial nerve and vestibular-cochlear nerve function. It is strongly suggested when available. Electrophysiological assessment includes AMR (LSR), EMG, and brainstem acoustic evoked potential (BAEP). AMR is specific to HFS. Its usual latency is generally about 10 ms. It has supplementary value for the diagnosis of HFS. It has special significance for patients who have recurrent HFS or prepare to receive reoperation after the first MVD is invalid. AMR recording is achieved from the mentalis muscle by electrically stimulating the temporal branch of the facial nerve or from the orbicularis oculi muscles by stimulating the marginal mandibular branch. Electrical stimulation consists of square-wave pulses. The duration is 0.2 ms, the frequency is 0.5–1.0 Hz, and the intensity level is 5–20 mA. Preoperative positive AMR often prompts significant value of AMR during the operation. Disappearance of AMR after decompression indicates good results. Conversely, if preoperative AMR is negative, choosing MVD should be particularly cautious. In such case, the surgeon's experience may be an important factor which determines the efficacy of surgery.

Preoperative EMG examination is mainly used to evaluate the functional status of the facial nerve and help diagnosis. Concentric needle electrode is generally inserted into the frontal muscle, orbicularis oculi, and orbicularis oris to record changes in motor units. In HFS patients a paroxysmal high-frequency spontaneous potential (up to 150 times per second maximum) can be recorded.

Preoperative BAEP reflects the function of the whole acoustic pathway. I, III, and V are the main waves. A prolonged latency implies disturbance of neural conduction. Since each wave has its specific origin, it is valuable in localization diagnosis. When combined with pure tone audiometry, it can comprehensively evaluate the function of the vestibular-cochlear nerve.

7.3.4 Preparation of Anesthesia

The anesthetist must know the patient's respiratory and surgical history and the allergic history to anesthetic drugs. To prevent interfering with intraoperative electrophysiological monitoring, the anesthesia plan should be optimized, especially on the choice of muscle relaxants, the time of use, and amount used.

7.4 Techniques of MVD

7.4.1 Position

Select appropriate surgical position according to the surgeon's preference. Usually select lateral position with head fixed by frame. Head is up 15–20° and flexed. The chin is about two fingers from the sternum. The upper shoulder is pulled to the caudal end with a strap to keep the head hyperextended while avoiding excessive traction-induced brachial plexus injury. The mastoid root should be at the highest point. In case the head frame is not available, a ring pillow can be used to fix the head.

7.4.2 Methods of Monitoring

Electrophysiological monitoring plays a very important role in the operations on cranial nerves. It not only makes the surgeries safer, but also makes the surgeries more effective. Electrophysiological monitoring mentioned here refers to monitoring of the brainstem and cranial nerves. Specific methods include SEP, BAEP, EMG, AMR, LSR, and ZLR.

Surgeries of different cranial nerve diseases have their specific methods of monitoring. For instance, in a MVD surgery for HFS, SEP, BAEP, and EMG of the facial nerve and abduction nerve, AMR and ZLR should be applied simultaneously, while for TN, SEP, BAEP, and EMG of oculomotor nerve and trochlear nerve, BTEP of the trigeminal nerve should be conducted. Brachial plexus may be injured by excessive traction on the operated side, so it is routinely monitored during all MVD surgeries. We would like to point out that the value of monitoring requires that the monitoring physician closely observe and identify various types of waves and keep in close coordination with the surgeon. Usually it is considered a sign of nerve injury when the amplitude decreased more than 20 % or more than 1 ms in latency. Otherwise, changes within this range can fully recover in most cases. The consultation and cooperation of the monitoring physician and the surgeon is important.

7.4.3 Anesthesia

Intravenous anesthesia with endotracheal intubation is the most commonly used method. Except for the induction phase of anesthesia, muscle relaxant drugs are restricted so as not to interfere with neurophysiological monitoring. During the surgery, the amount of infusion should be strictly controlled, and the carbon dioxide partial pressure should be around 26 mmHg. When the cerebrospinal fluid fluctuates significantly and affects surgical manipulation, ß-blockers may help. Usually Exelon 20–40 mg is given intravenously. At the same time observe changes in heart rate and blood pressure.

7.4.4 Incision

There are no stringent requirements for the scalp incision in MVD surgeries for HFS. For aesthetic purpose, oblique incision in the hairline or transverse incision behind the ear is usually used. The incision is 4–6 cm long, with the middle point at 1 cm below the mastoid root. With a burr, rongeur, or milling drill, a craniectomy of about 2.5 cm in diameter is made. When making the craniectomy, the mastoid air cell should be closely sealed to prevent fluid and blood from flowing in which affects hearing. It is unnecessary to expose transverse sinus or its junction with sigmoid sinus, but the bone window must be low enough, and the sigmoid sinus must be visualized from the outer edge of the bone window. There are many ways to cut the dura, including semicircular, triangular, cross, T-shaped, etc. The purpose is to facilitate the surgical procedure and reduce stretch of the brain tissue. We recommend a V-shape incision with the base adjacent to the sigmoid sinus (Fig. 7.2). This incision not only exposes the area between the auditory nerve and vagus nerve, but also facilitates exposing the zone 1–4 of the facial nerve from the caudal end to the rostral end. In this way, traction of the nerve is minimized. At the end of surgery, dural closure is convenient, and the incidence of cerebrospinal fluid leakage is low.

7.4.5 Approaches

There are two approaches of MVD for HFS: the lateral cerebellum approach and the lateral retrocerebellum approach. In the lateral cerebellum approach, after opening the dura, the cerebellar hemispheres are pulled inward directly to expose the facial and acoustic nerve. The arachnoid is dissected further to reveal zone 2 and the surrounding blood vessels, and then decompres-

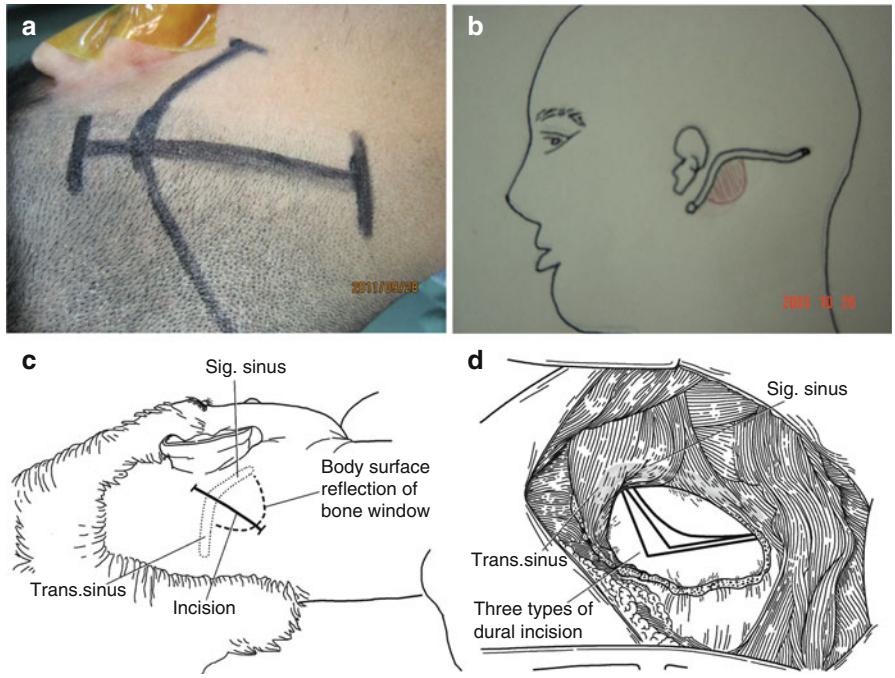

Fig. 7.2 (**a**) Incision (*straight*) with its relation to the transverse sinus and sigmoid sinus (*curved*); (**b**) bone window (*red shadow*); (**c**) bone window with its relation to the incision to the incision; (**d**) dura incision

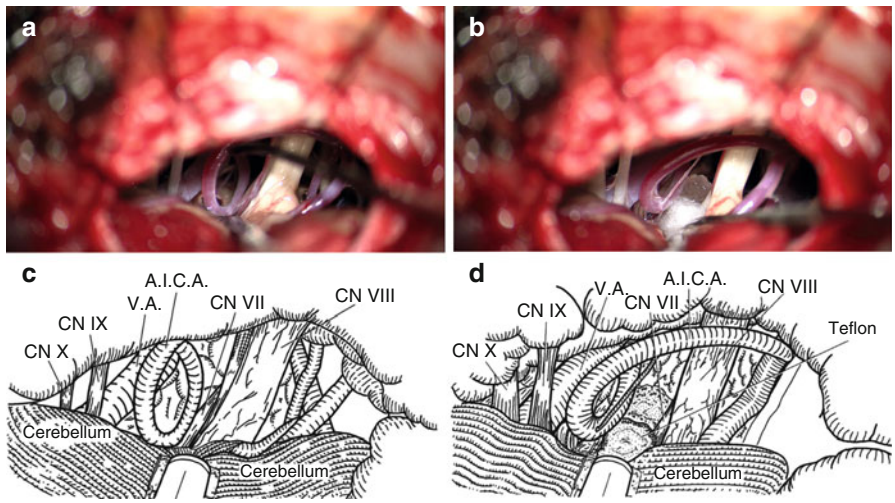

Fig. 7.3 Intraoperative images of lateral cerebellum approach (**a**, **c**) before implanting teflon; (**b**, **d**) after implanting teflon

sion is executed (Fig. 7.3). The advantage of this surgical approach is to expose the facial-auditory nerve directly so as to expose zone 2 and the surrounding blood vessels. When the offending vessels are located in zone 2, this approach can shorten the operation time. However, this approach also has obvious shortcomings. It often requires excessive traction of the cerebellar hemisphere. Because the arachnoid has not been dissected, this stretch may injure the vestibular-cochlear nerve, glossopharyngeal nerve, and vagus nerve. It can also lead to cer-

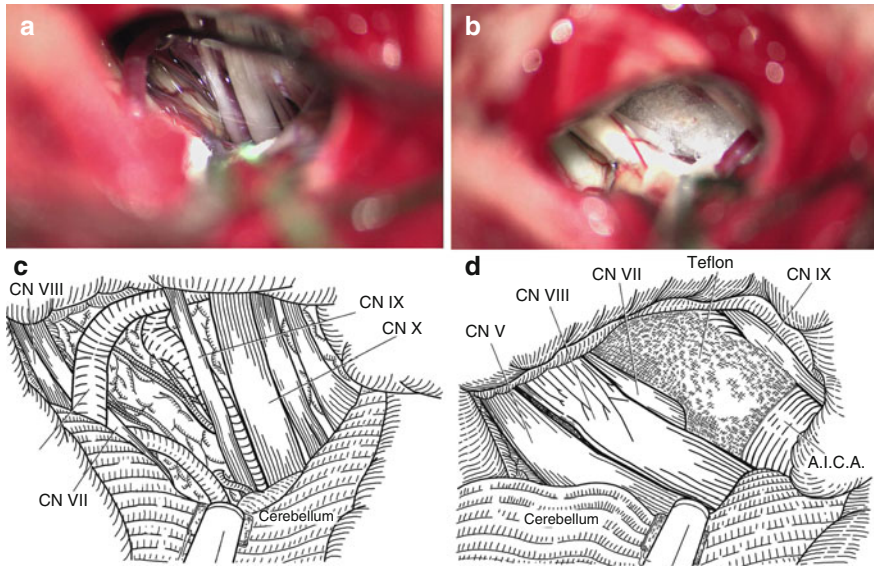

Fig. 7.4 Intraoperative images of lateral retro-cerebellum approach (**a**, **c**) before implanting teflon; (**b**, **d**) after implanting teflon

ebellar contusion. In case there are petrous veins on the dorsal side of the vagus nerve, it may lead to venous bleeding and even petrous vein tear. In addition, although this approach can expose zone 2, it is difficult to expose zone 1, 3, and 4. In case the offending vessels are located between the facial nerve and the acoustic nerve, vascular decompression will be very difficult. So this surgical approach is now rarely used.

Currently we recommend lateral retro-cerebellum approach (Zhong et al. 2010, 2014; Zhu et al. 2012a; Liang et al. 2012; Li et al. 2010, 2013a) (Fig. 7.4). The key of this approach is that, after dura opening, the cerebellum is pulled forward and medially, so as to expose the arachnoid at the bottom of the cerebello-medullar fissure. The arachnoid is first opened with a pointed blade, which allows slow release of cerebrospinal fluid. Then separate the arachnoid from the caudal end to the rostral end with spring scissors until the vagus nerve, glossopharyngeal nerve, and vestibular-cochlear nerve are completely released. In some patients there are petrous vein going on the dorsal side of vagal nerve. We suggest that this vein cannot be coagulated or cut. We recommend preserving this vein by carefully separating the adhesions surrounding the petrous vein. In case the AICA or PICA goes on the dorsal side of the vagal nerve, use the same method to preserve the arteries.

In some patients, the flocculus cerebelli is abnormally hypertrophied. In such case, it is very difficult to expose the whole facial nerve only by dissection of the arachnoid. Excessive traction of the flocculus cerebelli is not recommended. Instead, we suggest resection of the flocculus cerebelli. Then, we can easily expose the facial nerve and the offending vessels, and perform MVD safely.

7.4.6 Exploration Range

The success of MVD for HFS relies on identifying all the offending vessels and full decompression. Therefore, we need to know common offending vessels and their distribution. Our experiences show that the offending vessel can be a big artery like the vertebral artery or small vessels like the perforating arteries. It can be a common artery, or vein, or mixed compression consisting of arteries and veins. In fact, multiple vascular compressions are very common.

Common offending vessels include AICA, PICA, vertebral artery, and perforating artery (Zhu et al. 2012b; Feng et al. 2011; Zheng et al.

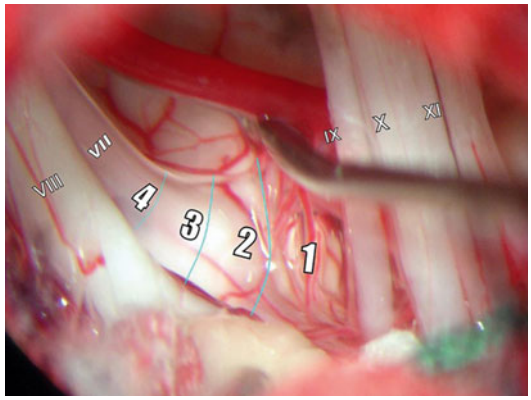

Fig. 7.5 Zone 1-4 of facial nerve

2011; Dou et al. 2014). It has been reported that in more than 50 % of patients, bilateral vertebral arteries shift to the affected side (Wang et al. 2014; Zhong et al. 2011a). The contact point of compression varies among different patients. It is reported that the contact point can be distributed in the full length of the facial nerve, from the pontine-medullar sulcus to the inner ear meatus. That is to say, there may be offending vessels on any part of the facial nerve.

In order to guide surgical manipulation and avoid omissions of offending vessels, we suggest that the facial nerve be divided into 4 zones (Li et al. 2013b; Zhong et al. 2011b) (Fig. 7.5). Zone 1, also known as REZ, is where the nerve emerges to the brainstem surface from the parenchyma and goes through the pontomedullary sulcus; zone 2 is where the root attaches to the surface of the pons; zone 3, the shortest segment among the four zones, is where the nerve gradually transits to the subarachnoidal segment; and zone 4, the longest segment, is where the nerve fibrins separate from the brainstem and extend to the internal meatus.

It is reported that the location of the offending vessels are as follows: 15 % in zone 1, 36 % in zone 2, 37 % in zone 3, and 12 % in zone 4 (Li et al. 2013b; Zhong et al. 2011b). We have similar findings in our clinical practice. Therefore, zone 4 must be inspected thoroughly during MVD for HFS. By this way we are able to find all vessels that are in contact with the facial nerve and then separate and properly fix them and ensure that all existing and potential vascular compression are entirely treated.

7.4.7 Decompression Method

Because vascular compression is the main cause of HFS, anatomical contact of blood vessels with facial nerve is a necessary condition for the occurrence of hemifacial spasm. Therefore, the core mission of MVD is to separate the offending vessels from the facial nerve and then properly remove and fix the vessels to avoid recurrence. Various decompression methods can be selected according to the thickness, elasticity, tortuosity, and length of the offending vessels.

There are three most commonly used decompression methods: isolation, suspension, and biological glue adhesion. The so-called isolation is to separate the offending vessels from the facial nerve and then implant Teflon cotton in between to ensure the vessels are no longer touching the facial nerve (Zhong et al. 2012, 2014). It is currently the most commonly used method. The advantage of this method includes simplicity of manipulation, high effectiveness and univeral applicability. However, its biggest drawback is that implanted Teflon cotton may adhere to the vessels and facial nerve, which is the leading cause of recurrence (Kureshi and Wilkins 1998; Payner and Tew 1996).

The so-called suspension is to separate the offending vessels from the facial nerve and then use muscle, fascia, or artificial straps to wrap around the offending vessels and fix them to the sidewall of the posterior fossa or tentorium (Swiątnicki et al. 2014; Khoo et al. 2012). In this type of method, Teflon cotton is not required. Therefore, the biggest advantage of this type of method is that it is suitable for long and tortuous offending vessels, and perineural adhesions do not occur, which makes reoperation relatively easy. But this method has its drawbacks, which is that the surgical procedure is very difficult and the surgical risk is significantly increased. Most clinicians are reluctant to use this method.

Similarly, biological glue adhesion method is to separate the offending vessels from the facial

nerve and stick the vessels to the sidewall of the posterior fossa or tentorium with biological glue (Kurokawa et al. 2004; Ichikawa et al. 2011). There is no Teflon cotton implanted between the facial nerve and the vessels. Biological glue adhesion method is mainly used for long and tortuous vessels, as well as complementary measures for thick vessels. The advantage of this method is similar to suspension, but the drawback is that reoperation will be very difficult due to glue-induced adhesion. In addition, in some patients, the glue may be unstable and lead to recurrence, and the glue itself can cause cranial nerve injuries. In fact, similar to a suspension, glue adhesion method is rarely used in clinical practice.

Although MVD may be simple in typical cases, it can be extremely difficult in complicated cases, such as compression of large vessels, facial nerve degeneration, offending vessels in between the facial nerve and acoustic nerve, offending vessels going through the fibers of facial nerve, persistent AMR waves, as well as no vascular compression found. We would like to brief the principles according to our clinical experience.

7.4.7.1 Large Sclerosed Vertebral Artery

In case the vertebral artery directly or indirectly acts as an offending vessel, the posterior fossa is usually small and the cerebellar-medullary cistern is narrow, which makes either separation or displacement of vertebral artery very difficult. If Teflon cotton is forcedly inserted in between the vertebral artery and the facial nerve, it will only increase compression and does not help to achieve satisfactory results. In this case, it is difficult to decompress the facial nerve even if using the method of suspension or biological glue adhesion (Suzuki et al. 1990; Mikami et al. 2013). For such cases, we suggest to open the lower part of the cerebellar-medullary cistern, pull the vertebral artery caudally, and then implant Teflon cotton in between the vertebral artery and the medulla from the caudal side to the rostral side. By this way, we can remove the vertebral artery from the surface of the facial nerve, which creates conditions for implanting Teflon cotton in between the vertebral artery and the facial nerve (Wang et al. 2014; Zhong et al. 2011a) (Fig. 7.6).

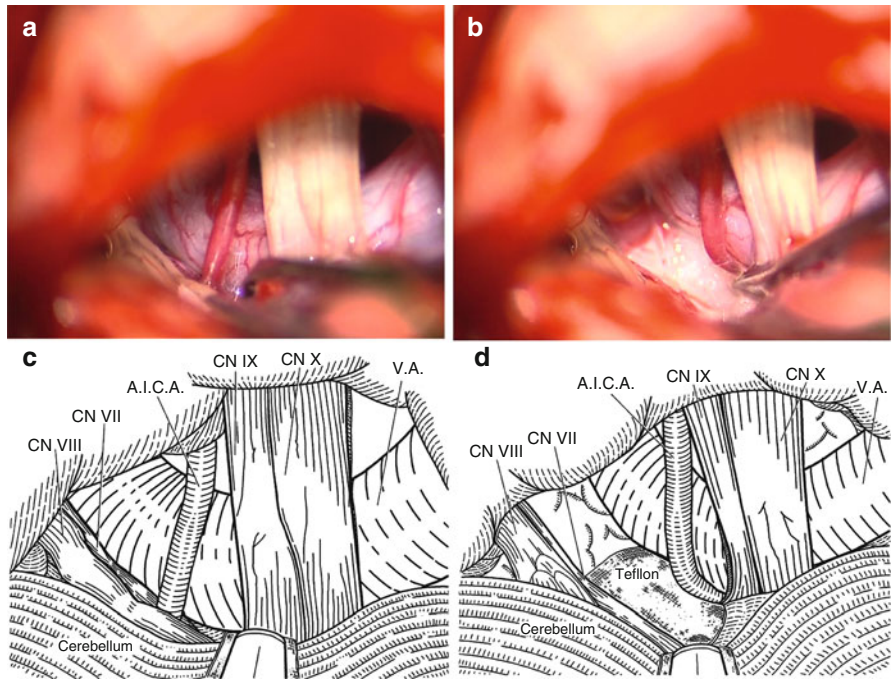

Fig. 7.6 Management of large sclerosed vertebral artery

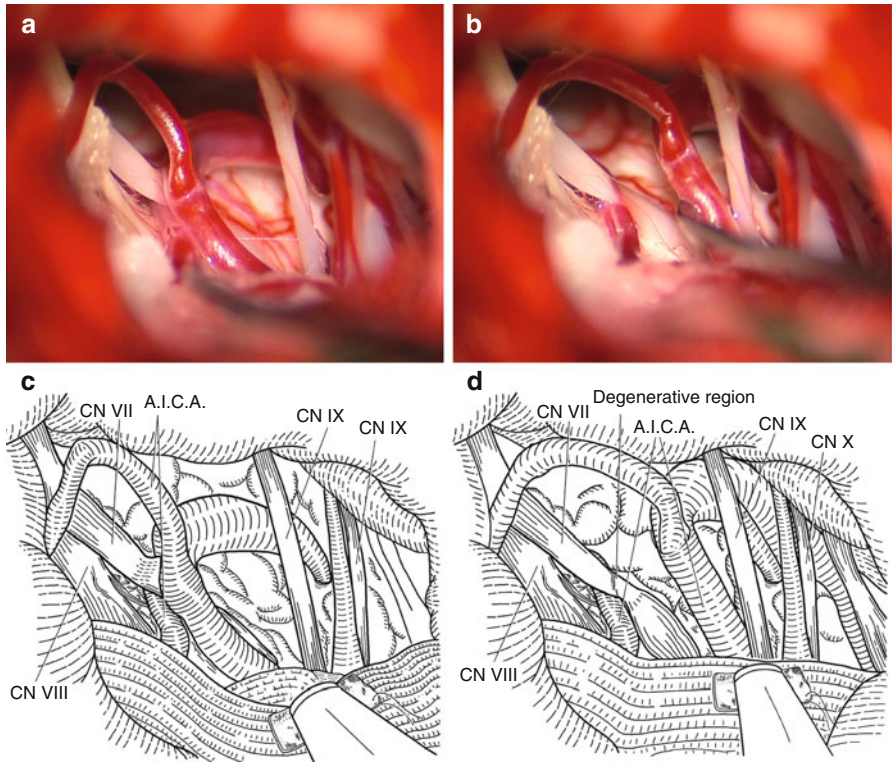

Fig. 7.7 Degeneration of facial nerve caused by severe compression

7.4.7.2 Degeneration of Facial Nerve

In some patients, severe compression may cause obvious pressure traces on facial nerves (Fig. 7.7). Other signs include thinning, pigmentation, and nerve fiber degeneration. It may be easy to separate the offending vessels from the facial nerve. However, if Teflon is simply inserted in between the facial nerve and the vessels, it will probably produce severe adhesion, which is a common cause of recurrence. In addition, it will also affect the functional recovery of facial nerve. For this case, we suggest to place some gelatin sponge between the Teflon cotton and facial nerve. It may well solve the above problems.

7.4.7.3 Offending Vessels between Facial Nerve and Acoustic Nerve

In about 8 % of HFS patients, AICA between facial nerve and acoustic nerve acts as offending vessels of HFS. In such case, neither is the AICA possible to remove, nor is there enough space for decompression. MVD is therefore very difficult. Currently, we suggest to use "sandwich" decompression (Fig. 7.8). First, separate AICA safely from the facial nerve and vestibular nerve on each side. Then, implant the right amount of Teflon in between AICA and facial nerve, as well as AICA and vestibular nerve. Finally, implant the right amount of gelatin sponge in between Teflon and both nerves. By this method, we cannot only achieve effective separation, but also avoid adhesion between Teflon cotton and nerves.

7.4.7.4 Offending Vessels Going through the Fibers of the Facial Nerve (Fig. 7.9)

It is rare that offending vessels go through the fibers of facial nerve. In such cases, decompression is very difficult, mainly due to conflicts of protecting facial nerve from injury and full decompression. We also suggest "sandwich" decompression method. Since the gap between the two bundles of the facial nerve fibers is much

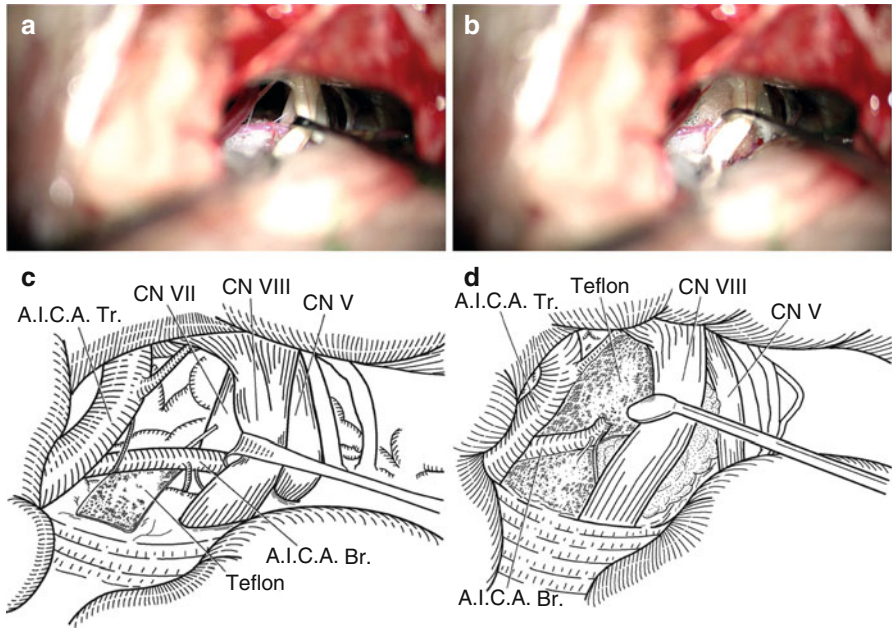

Fig. 7.8 "Sandwich" decompression of the facial nerve when AICA goes between the facial nerve and acoustic nerve

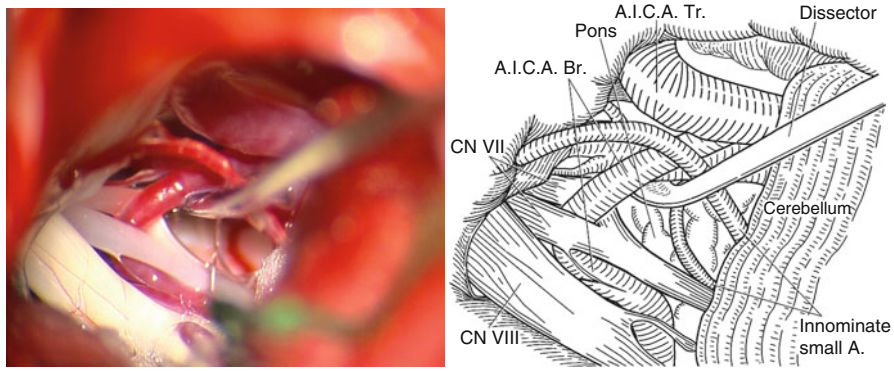

Fig. 7.9 Management of offending vessel which goes through the fibers of facial nerve

smaller than the gap between the facial nerve and vestibular nerve, separation and decompression should be done carefully. First, you need to shift the offending vessels to one side and then expand the gap by blunt dissection. It is best to implant a small amount of Teflon and gelatin sponge to complete the procedure.

7.4.7.5 Persistent AMR Waves

AMR wave is specific to typical hemifacial spasm. Positive AMR often prompted the existence of offending vessels, and AMR disappearance indicates sufficient decompression. However, the biggest drawback of AMR monitoring is that it is not stable enough and easily affected by many factors, which in turn undermines its value.

In some patients, AMR waves persist no matter what steps are taken. In such cases, it is difficult for the surgeon to decide whether to end the surgery or continue the investigation. We believe that if the monitoring electrodes are in correct position, persistent AMR usually implies the real offending vessels have not been found or the decompression is insufficient. Therefore, we

suggest a multiple monitoring method consisting of AMR, ZLR, and EMG. By this way, the surgeon can often find small vessels which were missed. After separation and decompression, AMR will disappear and the patient may get good results. In a words, we don't suggest to end an MVD surgery when AMR wave persists (Ying et al. 2011; Yang et al. 2014; Zheng et al. 2012).

7.4.7.6 No Vascular Compression Found

If no obvious vascular compression is found, the surgeon should first review the diagnosis of HFS and then comprehensively judge from the results of preoperative MRTA and intraoperative electrophysiological monitoring. The first possibility is that in the process of arachnoid dissection, the original offending vessels are removed from the facial nerve; a second possibility is that the thickened arachnoid itself is the cause of HFS. A third possibility is that the offending vessels are small perforating arteries, which require using ZLR monitoring to identify and judge. It should be noted that in these cases, the surgeon must not rashly judge that vascular compression does not exist. Instead, the surgeon should judge these various possibilities and take appropriate measures to ensure the effectiveness of operation.

7.4.7.7 Dual Side HFS

We suggest operating on the severer side first and then, according to the degree of relief and the patient's general conditions, operating on the other side at proper time. It is not recommended to perform bilateral MVD surgeries at one time. However, the time interval between the two operations is not particularly defined.

7.4.8 Assessment of MVD Progress

Accurately assessing the progress of MVD is the key to ensure full decompression and complete remission of symptoms and reduce unnecessary manipulation and surgical complications. According to the principles of evidence-based medicine, there are three classes of evidence for determining the endpoint of MVD surgery: full-length evidence, vessel evidence, and electrophysiological evidence (Sun et al. 2014).

Full-length evidence means the full length of the facial nerve (zone 1–4) has been examined under microscope and confirmed to be clear of offending vessels. Full-length evidence is the basic requirement for MVD (Zhong et al. 2014). Vessel evidence means all the vessels that have anatomical contact with facial nerve have been sufficiently dealt with (removed and decompressed). All offending vessels and potential offending vessels must be removed and decompressed to ensure no recurrence (Zhu et al. 2012a). Electrophysiological evidence means AMR and ZLR, which exist before decompression, have completely disappeared. MVD can be ended only after AMR and ZLR completely disappeared (Liang et al. 2012).

These principles must be adhered to in all MVD for HFS patients. When full-length evidence, vessel evidence, and electrophysiological evidence all prompt sufficient decompression, satisfactory clinical results are expected. But surgeons may sometimes find that while full-length evidence and vessel evidence both suggest sufficient decompression, electrophysiological evidence does not. AMR waves persistently exist. In such situation, do not doubt the reliability of AMR, but double check the reliability of full-length evidence and vessel evidence, because the probability of AMR false positive is very low.

7.4.9 Closure

Use warm saline to rinse the surgical field thoroughly to make sure there is no bleeding before closure. The dura should be sutured tightly. Inject warm saline repeatedly to exhaust gases before closing the dura. Use artificial dura and biological glue when necessary. Use the original bone flap, artificial skull, or metal plate to repair skull defects. Close the wound layer by layer.

7.4.10 Evaluation of Outcome

The results of evaluation are closely related to time of follow-up because there are delay remissions and recurrences. So time must be indicated when evaluating the outcome.

The outcome is defined as four grades (Zhong et al. 2010, 2014; Li et al. 2010). (1) Excellent means symptoms of HFS totally disappear, and the patients are subjectively very satisfied with no auxiliary drug needed. (2) Good means symptoms disappear, but will occasionally reappear when the patient is agitated or under stress or making certain facial movements. The patients are subjectively satisfied with no auxiliary drug needed. The above two grades are "effective." (3) Fair, or partial remission, means symptoms are partly relieved, but still frequent. The patients are not satisfied and need auxiliary drugs. (4) Poor, or invalid, means symptoms remain unchanged or even become worse. For patients with invalid results or partial remission, we suggest repeating AMR examination. If AMR is positive, a repeat surgery at early stage is recommended. Otherwise, we suggest follow-up, supplementary drugs, or Botox treatment.

7.4.11 Postoperative Management

Postoperatively, it is required to observe vital signs, consciousness, facial paralysis, hoarseness, cough, and vomiting. Head CT is routinely carried out within 24 h. In case of postoperative intracranial hypotension, put the patient in supine and head low-foot high position. When nausea and vomiting occurs, rotate the head to one side to avoid aspiration, and give symptomatic treatment. If the patient has postoperative facial paralysis, the cornea and oral care should be paid attention to. We suggest giving hormones, neurotrophic drugs, and microcirculation drugs, providing functional exercise, and keeping the patient warm. In case of drinking and swallowing dysfunction, aspiration pneumonitis should be avoided. If there is cerebrospinal fluid leak, the patient should take supine position with head up 30 °. Auristilla, blockage or flushing of the nasal and ear canal is strictly forbidden. Find out the cause and take proper measures.

7.4.12 Prevention of Complications

Common complications of MVD include cranial nerve dysfunction, cerebellum and brain stem injury, cerebrospinal fluid leakage, and intracranial hypotension syndrome.

7.4.12.1 Cranial Nerve Dysfunction

Cranial nerve dysfunction mainly includes facial paralysis, tinnitus, and hearing loss. In a minority of patients, there can be facial numbness, hoarseness, choking, and diplopia. Cranial nerve dysfunction is divided into two classes, acute and delayed. Acute dysfunction means it occurs within 3 days postoperatively, while delayed dysfunction means it occurs 3 days after surgery. Most delayed cranial nerve dysfunction occurred within 30 days after surgery. For example, more than 90 % of delayed facial paralysis occurs in 1 month after surgery. It may be related to operative procedure or postoperative virus infection. Therefore, we suggest that patients should keep warm within a month to reduce the risk of delayed facial paralysis. Once it occurs, use hormone therapy, antiviral drugs, and complimentary neurotrophic drugs (Lee et al. 2015; Ying et al. 2014).

Note that the following measures can effectively reduce the incidence of cranial nerve dysfunction. Avoid coagulating perforating vessels on the surface of cranial nerves or surrounding cranial nerves; avoid over-stretching cranial nerves; minimize direct stimulation of the cranial nerves to avoid feeding vessels spasm; fully dissect arachnoid around cranial nerves; routinely perform intraoperative electrophysiological monitoring; and use vasodilator drugs, hormones, and neurotrophic drugs from the day of surgery (Zhong et al. 2012, 2014).

7.4.12.2 Injuries of the Cerebellum and Brainstem

MVD for hemifacial spasm has 0.1 % mortality, which is mainly due to injuries of cerebellum and brain stem, including infarct and hemorrhage (Sandel and Eide 2013). The key to avoid injuries is to reduce time and strength of stretch. Use mannitol 30 min before surgery starts to reduce intracranial pressure. Use hyperventilation. Make the bone window close to the sigmoid sinus. Avoid using brain plate. Slowly drain CSF from the cerebellopontine cistern before exploring CPA. Avoid coagulation of vessels on the surface of the cerebellum and brainstem.

Postoperatively, 24 h continuous monitoring of blood pressure, pulse, respiration, and blood oxygen saturation is conducted using multiparameter ECG monitor. Observe consciousness and pupillary changes closely. In the following situations, infarction, swelling, or bleeding in the cerebellum or brainstem should be considered: sudden increase of blood pressure accompanied by pulse slowing down and unconsciousness, slow and deep breathing or even arrest, decreased oxygen saturation, dilated pupils, and light reflex diminished or disappearing. An emergent head CT scan should be performed. According to the result, measures include suboccipital decompression craniectomy or ventricular drainage.

7.4.12.3 CSF Leakage

CSF leakage occurs mainly because the mastoid air cells are not tightly sealed or the dura is not tightly sutured during closure (Miller and Miller 2012; Hyun et al. 2010). Therefore, the key to prevent cerebrospinal fluid leakage is to standardize surgical manipulations, including using bone wax to seal mastoid air cells and suturing the dura tightly. The dura may be repaired using muscle fascia when it cannot be tightly sutured. Meanwhile, use biological glue to paste a piece of artificial dura to the dura mater. Suture the incision in four layers, the muscles, fascia, subcutaneous tissue, and the skin, and avoid leaving dead space in between.

In case of cerebrospinal fluid rhinorrhea, ask the patient to keep supine without using pillow, keep the nose clean, and not to dig or block nose. Observe body temperature changes, and use antibiotics to prevent infection. Prevent constipation, coughing, and hard-stool-induced increased intracranial pressure. Use dehydrating agent or lumbar drainage to reduce intracranial pressure when necessary. A repair surgery is indicated for persistent leakage and multiple recurrent leakage.

7.4.12.4 Intracranial Hypotension Syndrome

Intracranial hypotension syndrome may be due to long time exposure of the surgical field, loss of large amounts of cerebrospinal fluid, reduced postoperative cerebrospinal fluid secretion, as well as inadequate infusion. Common symptoms include headache, dizziness, nausea, and non-jet-like vomiting, accompanied by low blood pressure and increased pulse rate. The symptoms can be alleviated by lowering the head position. Injecting saline to exhaust air before dura closure is important to prevent intracranial hypotension.

References

Dou NN, Hua XM, Zhong J, Li ST. A successful treatment of coexistent hemifacial spasm and trigeminal neuralgia caused by a huge cerebral arteriovenous malformation: a case report. J Craniofac Surg. 2014; 25:907–10.

Feng B, Zheng X, Zhang W, Yang M, Tang Y, Zhong J, Hua X, Ying T, Li S. Surgical treatment of pediatric hemifacial spasm patients. Acta Neurochir (Wien). 2011;153(5):1031–5; discussion 1035.

Hyun SJ, Kong DS, Park K. Microvascular decompression for treating hemifacial spasm: lessons learned from a prospective study of 1,174 operations. Neurosurg Rev. 2010;33(3):325–34; discussion 334.

Ichikawa T, Agari T, Kurozumi K, Maruo T, Satoh T, Date I. "Double-stick tape" technique for transposition of an offending vessel in microvascular decompression: technical case report. Neurosurgery. 2011;68 (2 Suppl Operative):377–82; discussion 382.

Khoo HM, Yoshimine T, Taki T. A "sling swing transposition" technique with pedicled dural flap for microvascular decompression in hemifacial spasm. Neurosurgery. 2012;71(1 Suppl Operative):25–30; discussion 30–1.

Kureshi SA, Wilkins RH. Posterior fossa reexploration for persistent or recurrent trigeminal neuralgia or hemifacial spasm: surgical findings and therapeutic implications. Neurosurgery. 1998;43(5):1111–7.

Kurokawa Y, Maeda Y, Toyooka T, Inaba K. Microvascular decompression for hemifacial spasm caused by the

vertebral artery: a simple and effective transposition method using surgical glue. Surg Neurol. 2004;61(4): 398–403.

Lee MH, Lee HS, Jee TK, Jo KI, Kong DS, Lee JA, Park K. Cerebellar retraction and hearing loss after microvascular decompression for hemifacial spasm. Acta Neurochir. 2015;157(2):337–43.

Li S, Hong W, Tang Y, Ying T, Zhang W, Li X, Zhu J, Zhong J, Hua X, Xu S, Liang W, Wang X, Yang M, Li Y, Zheng X. Re-operation for persistent hemifacial spasm after microvascular decompression with the aid of intraoperative monitoring of abnormal muscle response. Acta Neurochir. 2010;152:2113–8.

Li X, Zheng X, Wang X, Li B, Ying T, Li Y, Li S. Microvascular decompression treatment for post-Bells palsy hemifacial spasm. Neurol Res. 2013a; 35(2):187–92.

Li Y, Zheng X, Hua X, Ying T, Zhong J, Zhang W, Li S. Surgical treatment of hemifacial spasm with zone-4 offending vessel. Acta Neurochir (Wien). 2013b; 155(5):849–53.

Liang Q, Shi X, Wang Y, Sun Y, Wang R, Li S. Microvascular decompression for hemifacial spasm: technical notes on pontomedullary sulcus decompression. Acta Neurochir (Wien). 2012;154(9): 1621–6.

Mikami T, Minamida Y, Akiyama Y, Wanibuchi M, Sugino T, Houkin K, Mikuni N. Microvascular decompression for hemifacial spasm associated with the vertebral artery. Neurosurg Rev. 2013;36:303–8.

Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for treatment of hemifacial spasm: a systematic review. Br J Neurosurg. 2012; 26(4):438–44.

Payner TD, Tew JM. Recurrence of hemifacial spasm after microvascular decompression. Neurosurgery. 1996;38(4):686–90.

Sandel T, Eide PK. Long-term results of microvascular decompression for trigeminal neuralgia and hemifacial spasms according to preoperative symptomatology. Acta Neurochir. 2013;155(9):1681–92; discussion 1692.

Sun H, Li ST, Zhong J, Zhang WC, Hua XM, Wan L, Zheng XS. The strategy of microvascular decompression for hemifacial spasm: how to decide the endpoint of an MVD surgery. Acta Neurochir (Wien). 2014;156(6):1155–9.

Suzuki S, Tsuchita H, Kurokawa Y, Kitami K, Sohma T, Takeda T. New method of MVD using a vascular tape for neurovascular compression involving the vertebrobasilar artery – report of two cases. Neurol Med Chir. 1990;30(13):1020–3.

Swiątnicki W, Heleniak M, Komuński P. Hemifacial spasm caused by the cross-compression of the vertebral artery loop – a case-centered report of a stitched sling retraction technique. Bull Soc Sci Med Grand Duche Luxemb. 2014;3:73–82.

Wang YN, Zhong J, Zhu J, Dou NN, Xia L, Visocchi M, Li ST. Microvascular decompression in patients with coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Acta Neurochir (Wien). 2014;156(6):1167–71.

Yang M, Zheng X, Ying T, Zhu J, Zhang W, Yang X, Li S. Combined intraoperative monitoring of abnormal muscle response and Z-L response for hemifacial spasm with tandem compression type. Acta Neurochir (Wien). 2014;156(6):1161–6.

Ying TT, Li ST, Zhong J, Li XY, Wang XH, Zhu J. The value of abnormal muscle response monitoring during microvascular decompression surgery for hemifacial spasm. Int J Surg. 2011;9(4):347–51.

Ying T, Thirumala P, Chang Y, Habeych M, Crammond D, Balzer J. Empirical factors associated with brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm and its correlation to hearing loss. Acta Neurochir. 2014; 156:571–5.

Zheng XS, Feng BH, Zhang WC, Ying TT, Li ST. Hemifacial spasm caused by cross type vascular compression. Neurol Res. 2011;33(9):965–8.

Zheng X, Hong W, Tang Y, Ying T, Wu Z, Shang M, Feng B, Zhang W, Hua X, Zhong J, Li S. Discovery of a new waveform for intraoperative monitoring of hemifacial spasms. Acta Neurochir (Wien). 2012;154(5): 799–805. Epub 22 Feb 2012.

Zhong J, Zhu J, Li S-T, Li X-Y, Wang X-H, Yang M, Liang W, Guan H-X. An analysis of failed microvascular decompression in patients with hemifacial spasm: focused on the early reoperative findings. Acta Neurochir. 2010;152:2119–23.

Zhong J, Zhu J, Li S-T, Guan H-X. Microvascular decompressions in patients with coexistent hemifacial spasm and trigeminal neuralgia. Neurosurgery. 2011a;68: 916–20.

Zhong J, Li ST, Zhu J, Guan HX. Is entire nerve root decompression necessary for hemifacial spasm? Int J Surg. 2011b;9(3):254–7.

Zhong J, Li ST, Zhu J, Guan HX, Zhou QM, Jiao W, Ying TT, Yang XS, Zhan WC, Hua XM. A clinical analysis on microvascular decompression surgery in a series of 3000 cases. Clin Neurol Neurosurg. 2012;114(7): 846–51.

Zhong J, Zhu J, Sun H, Dou NN, Wang YN, Ying TT, Xia L, Liu MX, Tao BB, Li ST. Microvascular decompression surgery: surgical principles and technical nuances based on 4000 cases. Neurol Res. 2014;36(10): 882–93.

Zhu J, Li ST, Zhong J, Ying TT, Guan HX, Yang XS, Zhou QM, Jiao W. Microvascular decompression for hemifacial spasm. J Craniofac Surg. 2012a;23(5): 1385–7.

Zhu J, Li ST, Zhong J, Guan HX, Ying TT, Yang M, Yang X, Zhou Q, Jiao W. Role of arterioles in management of microvascular decompression in patients with hemifacial spasm. J Clin Neurosci. 2012b;19(3):375–9. Epub 25 Jan 2012.

Microvascular Decompression Surgery for Disabling Positional Vertigo and Tinnitus

8

Aage R. Møller

Abstract

Disabling positional vertigo (DPV) and some forms of tinnitus can be treated successfully with microvascular decompression (MVD) of the root of the auditory-vestibular nerve. These two diseases have many different forms and the operation is more complex than MVD for trigeminal neuralgia or hemifacial spasm. Success of DPV depends on correct selection of candidates for the treatments. In a study of 41 patients operated upon for severe DPV, 73.2 % were totally free of DPV symptoms or experienced significant improvements that allowed them to return to nearly normal life, 4.9 % had minor relief of symptoms, and 22 % had no noticeable improvement. Other studies have shown that treatment with medications of the benzodiazepine family such as Valium is effective in some individuals who have DPV symptoms.

In a study of 72 patients who underwent MVD operations for severe tinnitus, 18.1 % had total relief from tinnitus, 22.2 % had marked improvement, 11.1 % slight improvement, 45.8 % no improvement, and 2.8 % experienced a worsening of symptoms.

The success of MVD operations as treatment of severe tinnitus was inversely related to the time the patients had had their tinnitus. Those who experienced total relief or marked improvement had had their tinnitus for an average of 2.9 and 2.7 years, respectively; those who showed slight or no improvement had prior experienced with tinnitus for 5.2 and 7.9 years, respectively.

There was a strong gender effect; of the 32 women in the study, 54.8 % experienced total relief or marked improvement, while of the 40 men, only 29.3 % had favorable outcome.

Studies have shown that administration of benzodiazepines such as alprazolam can have beneficial effect on some forms of tinnitus.

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Keywords
Microvascular decompression operations • Disabling positional vertigo • Tinnitus • Vertigo • Nausea

8.1 Introduction

Treatment of hemifacial spasm (HFS) using microvascular decompression (MVD) surgery was discussed in Chaps. 3, 7, and 14. In this chapter we will discuss two other diseases that can be treated in a similar way, namely, some forms of tinnitus and a vestibular disorder known as disabling positional vertigo (DPV) (Jannetta et al. 1984) (Møller 1987).

There are considerable differences between the diagnosis and treatment of diseases that are associated with the vestibulocochlear nerve (DPV and tinnitus). The selection of people who may benefit from MVD operations is more complex for patients with tinnitus and patients with DPV than what it is for people with HFS, trigeminal neuralgia (TGN), or glossopharyngeal neuralgia (GPN).

8.2 Disabling Positional Vertigo

Disabling positional vertigo (DPV) is a rare form of a balance disorder that is characterized by constant positional vertigo and nausea (Møller et al. 1993b; Jannetta et al. 1984). The symptoms of DPV are constant and people with DPV experience a state of nausea to a disabling degree. As symptoms become more intense with changes in head position, people with DPV are most comfortable lying in bed. In addition to abnormal vestibular test results, people with DPV also exhibit signs of functional changes in the cochlear nerve resulting in subtle but distinct changes in their auditory brain stem responses (ABR). There is a prolonged interval between peaks I and III (IPL I–III) and peak II is also abnormal (Møller et al. 1993b).

MVD operations of the vestibular nerve are in use for treating DPV. Other treatments are medications with benzodiazepines such as diazepam (Valium) (Møller 1997). The results of MVD operations for DPV are similar to those of HFS and TGN (Møller et al. 1993b). The operation is more difficult because of the risk of injuring the cochlear nerve.

Studies by De Ridder et al. (2002) produced evidence that vascular compression syndromes may arise from vascular contact along the entire CNS segment of the cranial nerves and not only at the root entry zones (Obersteiner-Redlich zone). De Ridder and Møller (2010) described the results of studies that showed the correlation between the length of the centrally myelinated (oligodendrocyte) portion of the vestibular nerve and the incidence of the microvascular compression syndrome. Furthermore, decompression of the CNS segment of the vestibular nerve is effective in eliminating DPV symptoms without decompression of the root entry zone. De Ridder and colleagues interpreted this to corroborate the hypothesis that close contact with a blood vessel anywhere along the entire length of the central portion of the vestibular nerve could be associated with symptoms.

8.3 Tinnitus

Tinnitus is hearing meaningless sounds that are not caused by a physical sound reaching the ear. In addition to hearing annoying sounds, many people with tinnitus also experience suffering or distress that often is independent of the phantom sound of tinnitus. Severe chronic tinnitus has many similarities to what has been described for chronic neuropathic pain (Møller 2014b).

Many different treatments have been studied but few have reached routine clinical use. The reason for the inconsistent success in treatment of people with tinnitus is mainly of lack of understanding of the abnormal functions that generate the phantom sounds of tinnitus and their connection to distress and suffering. It was a major

progress in understanding of the pathophysiology of tinnitus when it was discovered that chronic tinnitus is rarely caused by pathologies of the ear but rather often pathologies of the nervous system.

MVD operations directed to the cochlear-vestibular nerve are one form of treatment that is in use. However, tinnitus takes a plethora of forms (Møller 2010a) and only a few people with a certain form of tinnitus benefit from MVD operations on the cochlear nerve. The form of tinnitus that can be alleviated by MVD of the cochlear nerve is associated with ABR changes similar to those that occur in people with DPV (De Ridder et al. 2007; Møller et al. 1993a). The success rate of MVD operations for tinnitus is considerably lower than those of HFS and TGN.

8.4 Pathophysiology of DPV

The pathophysiology of DPV is unknown. The fact that the symptoms of DPV can be relieved by MVD suggests that there may be similarities between the pathology of DPV and those of the other disorders that can be cured effectively by MVD (HFS, TGN, and GPN). This means that it is likely that the anatomical location of the pathology is the brain and not the vestibular organ (inner ear).

It also seems likely that activation of neuroplasticity is involved in the creation of the symptoms of DPV similar to what is the case for, for example, chronic neuropathic pain and tinnitus (Møller 2014a).

The fact that head movements trigger the symptoms indicates that they are evoked by signals from the vestibular organ in the ear. As such signals normally do not reach consciousness, it can be inferred that, in people with DPV, the signals from the balance organ are abnormally routed to reach regions of the brain that they do not typically reach. The feeling of vertigo and nausea indicates involvement of structures in the autonomic nervous system and possibly the insula. Such rerouting can occur because of unmasking of dormant synapses that connect axons from the vestibular nuclei to different regions of the brain (Møller 2014a).

Dormant synapses may become unmasked by activation of neuroplasticity. Synapses that do not conduct because of insufficient input may also become activated if the excitatory input is increased or if inhibition is decreased. This means that the rerouting of information that causes the symptoms of DPV may be caused by hyperactivity at some level of the ascending vestibular pathways. This hypothesis is supported by evidence that the symptoms of DPV can be lessened by administration of drugs of the benzodiazepine family, which are known to be $GABA_A$ receptor agonists and thereby enhancers of inhibition.

8.5 Pathophysiology of Tinnitus That Can Be Treated Successfully with MVD

Little is known about the pathophysiology of those forms of tinnitus where the symptoms can be relieved by MVD of the cochlear nerve. In general, studies of patients who are undergoing MVD operations for tinnitus have shown that evoked potentials that are generated by the cochlear nerve, the nuclei, and the fiber tracts of the ascending auditory pathway of the brain stem in people with incapacitating tinnitus are not significantly different from those that can be recorded from individuals with the same degree of hearing loss but no tinnitus (Møller et al. 1992a). This suggests that severe tinnitus might be the result of abnormalities in more rostral brain structures. The abnormalities that occur in people with tinnitus may not be limited to structures that are normally associated with auditory functions, but many other structures may be involved (Schlee et al. 2009, 2010; Vanneste and De Ridder 2012).

That injury to the peripheral auditory system can result in hyperactive changes in more central auditory structures was demonstrated by Gerken et al. (1991). They found that overexposure to sound (noise impairment) affects the function of central structures in the ascending auditory pathways. Earlier, Syka and colleagues showed that noise-induced hearing loss was associated with abnormalities in the function of central auditory structures (Syka and Popelar 1982). The fact that

some people with tinnitus experience hyperacusis has been taken to indicate that the physiological abnormality is located at higher brain levels and that perhaps the prefrontal cortex is involved (Hazell 1990).

Rerouting of auditory information through such activation of neural plasticity may activate an ascending auditory pathway that uses the dorsal-medial thalamic auditory nucleus. This hypothesis is supported by the finding that the nonclassical ascending pathways in adults with tinnitus may be active (Møller et al. 1992b). Other studies have shown indications that sound activated the nonclassical pathways in children but normally not in adults (Møller and Rollins 2002). This route has subcortical connections to limbic structures (Eggermont 2007; Møller 2010b, 2014a) and may explain why affective signs and symptoms such as phonophobia and depression often accompany severe tinnitus. Other studies have shown evidence of increased activation of limbic structures in people with tinnitus (Lockwood et al. 1998).

Many forms of tinnitus have similarities with central neuropathic pain (Møller 2010b, c).

8.6 Selection of Candidates for MVD for DPV

The process for selection of candidates for MVD of CNVII, CNV, CNIX, and CNVIII depends almost entirely on the patient's history and assessment of the patients' symptoms. While HFS, TGN, and GPN have distinct diagnostic characteristics, the symptoms of DPV are similar to those of several other vestibular disorders, which may not respond to MVD. It is an additional challenge to determine which side of the patient is affected by the DPV. Due to these variables, the process to select candidates for MVD operations needs to be more rigorous for potential DPV cases than for other MVD procedures. Not all people with the symptoms of DPV are candidates for MVD operations.

In addition to a review of the patients' histories, two different objective methods have been described for selecting patients with DPV and tinnitus for MVD operations. One makes use of electrophysiological methods (ABR) (Møller et al. 1993b) and the other makes use of imaging methods (Brackmann et al. 2001; De Ridder et al. 2004). The fact that vascular contact may occur asymptomatically (Sunderland 1948), however, indicates that imaging studies are less important in the diagnosis of the disorders that can be treated successfully with DPV. Other tests may be negative; one study reported that 43 % of patients with severe DPV had normal electronystagmography (ENG) (Møller et al. 1993b).

It has been reported that approximately 80 % of patients with DPV were successfully treated with a small dosage of Valium® (Møller 1997).

8.7 Selection of Candidates for MVD for Tinnitus

In contrast to other diseases that can be successfully treated by MVD operations such as HFS and TGN, tinnitus represents a large group of disorders with diverse and poorly defined pathophysiology. Only people with certain forms of unilateral tinnitus will benefit from MVD operations (Møller et al. 1993a; Vasama et al. 1998).

Imaging studies such as MRI are of limited value because vascular contact frequently occurs without producing any symptoms (Sunderland 1948).

The functional effect of close contact between a blood vessel and at the root of the auditory-vestibular nerve may be reflected by subtle but typical abnormalities of the ABR. Abnormalities in the form of prolonged interpeak latency (IPL) of peaks I–III and absence of peak II are the most important in the diagnosis of people who may benefit from MVD for tinnitus. Especially important is the difference between IPL I–III on the side with tinnitus and the other side.

More recently De Ridder et al. (2007) and De Ridder and Møller (2010) have described more specific and refined selection criteria for MVD operations for tinnitus regarding abnormalities in the ABR.

Even with careful selection of patients for MVD, operations for tinnitus have much lower success rate than MVD for HFS and TGN (Møller

et al. 1993a; De Ridder and Møller 2010). That must be considered when advising patients about MVD operations for tinnitus.

8.8 MVD Operations for DPV and for Tinnitus

The surgical techniques for MVD operations for DPV and tinnitus (Jannetta et al. 1984) are similar to those used for treating other MVD disorders such as HFS, TGN, and GPN (Møller 1998).

However, operations on the auditory-vestibular nerve require substantial experience by the surgeon, and such operations carry higher risk of hearing loss than MVD operations of other cranial nerves. It is therefore critical to use intraoperative neurophysiological monitoring (IONM) of neural conduction in the cochlear nerve during MVD operations of the cochlear-vestibular nerve (Møller 2011), preferably by direct recording from the cochlear nucleus (Møller et al. 1994). In the hands of experienced surgeons, other risks such as brain stem strokes are small but do exist.

8.9 Results of MVD for DPV

With proper selection of patients, the cure rate for MVD operations of selected patients with DPV is approximately 73 % (Møller et al. 1993b; Brackmann et al. 2001), only slightly lower than that of HFS and TGN operations where the cure rate is approximately 85 % (Barker et al. 1995, 1996).

In one study 41 DPV patients belonging to a group of 200 consecutive patients with vestibular problems were followed for 2½ to 3½ years after their MVD operation (Møller and Møller 1990). These patients all had significant vascular contact with the vestibular nerve at the time of the operation. Before the operation, only 4 had ever been employed and the other 37 had been unable to work because of their DPV. Approximately half were housebound or had to use a wheelchair for all activities including housework. By the follow-up, 30 out of 41 (73 %) were either totally free of DPV symptoms or had made significant improvement that allowed them to return to full-time work or perform meaningful activities.

Two of the patients reported minor relief and 9 had no noticeable improvement after the operation. Nonetheless, for 73 % of the patients, the operation was a tremendous success.

The patients in this study were operated upon using the techniques described by Jannetta et al. (1984), and the patients were selected according to the criteria described by MB Møller et al. (1993b).

8.10 Results of MVD for Tinnitus

The rate of favorable outcome (great improvement or complete elimination of tinnitus) for MVD of the intracranial portion of the cochlear nerve in patients with severe chronic tinnitus has been reported to be between 44 and 50 % (Okamura et al. 2000; Ko and Park 1997) (Møller et al. 1993b).

In a study of 72 patients who underwent MVD of the cochlear nerve, 29 (40.3 %) had marked improvement or complete elimination of their tinnitus (Møller et al. 1993b). These patients complained of incapacitating tinnitus and were operated upon using an MVD technique similar to that described by Jannetta for treating DPV (Jannetta et al. 1984) but modified for MVD of the intracranial portion of the cochlear nerve. Of the 72 patients, 13 (18.1 %) had total relief from tinnitus, 16 (22.2 %) had marked improvement, 8 (11.1 %) slight improvement, 33 (45.8 %) no improvement, but 2 (2.8 %) experienced a worsening of symptoms (Møller et al. 1993b).

The patients in this study were selected according to the criteria described by Møller et al. (1993b) using the patients' history and auditory test results (ABR, acoustic middle ear reflex, and the patients' audiograms).

The success of the operations was inversely related to the time the patients had endured their tinnitus. The most favorable results were obtained in patients who had had their tinnitus only for a short period. Those who experienced total relief or marked improvement had had their tinnitus for an average of 2.9 and 2.7 years, respectively. Those who showed slight or no improvement had prior experience with tinnitus for 5.2 and 7.9 years, respectively.

There was also a gender effect. Of the 32 women in the study, 54.8 % experienced total

relief or marked improvement. Of the 40 men, only 29.3 % had that favorable outcome.

This study thus showed that the outcome of MVD operations for patients who have had tinnitus for more than 4 years was less favorable than it was for patients who have had tinnitus for a shorter time (Møller et al. 1993b). It is also interesting that there is a noticeable difference in the success rate of MVD for tinnitus in men and women. The reason for this difference is unknown, but it is an example of differences between the efficacy of MVD operations for tinnitus and for other diseases that can be cured by MVD operations.

Estrogen may play a role in creating this difference in the success rate for MVD operations in men and women. It has been shown that estrogen interacts with the $GABA_A$ receptor (N-Wihlbäck et al. 2006; Pinaud and Tremere 2012).

More recently De Ridder et al. (2007) and De Ridder and Møller (2010) have shown indications that MVD operations to treat tinnitus also could improve hearing thresholds.

Conclusion

It is more difficult to select DPV or tinnitus patients for MVD operations. Fewer people with DPV or tinnitus experience benefit from MVD operations than do HFS or TGN patients. Even with the best possible selection of patients for MVD, the success rate of MVD is slightly lower for DPV than it is for HFS and TGN. For tinnitus, the success rate is much lower. MVD operations of the cochlear-vestibular nerve for treatment of HFS and TGN are more technically demanding than those of the facial or trigeminal nerve, and the risks of complications in the form of hearing loss and tinnitus (or worsened tinnitus) are greater.

References

Barker FG, Jannetta PJ, Bissonette DJ, Shields PT, Larkins MV. Microvascular decompression for hemifacial spasm. J Neurosurg. 1995;82:201–10.

Barker FG, Jannetta PJ, Bissonette DJ, Larkins MV, Jho HD. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med. 1996;334:1077–83.

Brackmann DE, Kesser DW, Day JD. Microvascular decompression of the vestibulocochlear nerve for disabling positional vertigo: the House Ear Clinic experience. Otol Neurotol. 2001;22:882–7.

De Ridder D, Møller AR. Microvascular compression of the vestibulocochlear nerve. In: Møller AR, Langguth B, De Ridder D, Kleinjung T, editors. Textbook of tinnitus. New York: Springer; 2010.

De Ridder D, Møller AR, Verlooy J, Cornelissen M, De Ridder L. Is the root entry/exit zone important in microvascular compression syndromes? Neurosurgery. 2002;51:427–34.

De Ridder D, Ryu H, Møller AR, Nowe V, Van De Heyning P, Verlooy J. Functional anatomy of the human cochlear nerve and its role in microvascular decompressions for tinnitus. Neurosurgery. 2004;54:381–8.

De Ridder D, Heijneman K, Haarman B, Van Der Loo E. Tinnitus in vascular conflict of the eighth cranial nerve: a surgical pathophysiological approach to ABR changes. Prog Brain Res. 2007;166:401–11.

Eggermont JJ. Pathophysiology of tinnitus. In: Langguth B, Hajak G, Kleinjung T, Cacace A, Møller AR, editors. Tinnitus: pathophysiology and treatment, progress in brain research. Amsterdam: Elsevier; 2007.

Gerken GM, Solecki JM, Boettcher FA. Temporal integration of electrical stimulation of auditory nuclei in normal hearing and hearing-impaired cat. Hear Res. 1991;53:101–12.

Hazell JW, Jastreboff PJ. Tinnitus. I: Auditory mechanisms: a model for tinnitus and hearing impairment. J Otolaryngol. 1990;19(1):1–5.

Jannetta PJ, Møller MB, Møller AR. Disabling positional vertigo. N Engl J Med. 1984;310:1700–5.

Ko Y, Park CW. Microvascular decompression for tinnitus. Stereotact Funct Neurosurg. 1997;68:266–9.

Lockwood A, Salvi R, Coad M, Towsley M, Wack D, Murphy B. The functional neuroanatomy of tinnitus. Evidence for limbic system links and neural plasticity. Neurology. 1998;50:114–20.

Møller MB. Vascular compression of the eighth nerve as a cause of tinnitus. In: Feldmann H, editor. Proceedings of the III international tinnitus seminar, Munster, Western Germany, June 11–13, 1987. Karlsruhe: Harsch Verlag; 1987.

Møller MB. Selection criteria and treatment of patients with disabling positional vertigo. Stereotact Funct Neurosurg. 1997;68:270–3.

Møller AR. Vascular compression of cranial nerves. I: history of the microvascular decompression operation. Neurol Res. 1998;20:727–31.

Møller AR. Different forms of tinnitus. In: Møller AR, Langguth B, De Ridder D, Kleinjung T, editors. Textbook of tinnitus. New York: Springer; 2010a.

Møller AR. Pathology of the auditory system that can cause tinnitus. In: Møller AR, Langguth B, De Ridder D, Kleinjung T, editors. Textbook of tinnitus. New York: Springer; 2010b.

Møller AR. Similarities between tinnitus and pain. In: Møller AR, Langguth B, De Ridder D, Kleinjung T, editors. Textbook of tinnitus. New York: Springer; 2010c.

Møller AR. Intraoperative neurophysiological monitoring. Intraoperative neurophysiology. 3rd ed. New York: Springer; 2011.

Møller AR. Neuroplasticity and its dark sides: disorders of the nervous system. Dallas: Aage R. Møller; 2014a.

Møller AR. Pain: its anatomy, physiology and treatment. Dallas: Aage R Møller Publishing; 2014b.

Møller MB, Møller AR. Vascular compression syndrome of the eighth nerve: clinical correlations and surgical findings. In: Arenberg IK, Smith DB, editors. Neurologic clinics: diagnostic neurotology and otoneurology. Philadelphia: WB Saunders Publishing Co.; 1990.

Møller AR, Rollins P. The non-classical auditory system is active in children but not in adults. Neurosci Lett. 2002;319:41–4.

Møller AR, Møller MB, Jannetta PJ, Jho HD. Compound action potentials recorded from the exposed eighth nerve in patients with intractable tinnitus. Laryngoscope. 1992a;102:187–97.

Møller AR, Møller MB, Yokota M. Some forms of tinnitus may involve the extralemniscal auditory pathway. Laryngoscope. 1992b;102:1165–71.

Møller MB, Møller AR, Jannetta PJ, Jho HD. Vascular decompression surgery for severe tinnitus: selection criteria and results. Laryngoscope. 1993a;103:421–7.

Møller MB, Møller AR, Jannetta PJ, Jho HD, Sekhar LN. Microvascular decompression of the eighth nerve in patients with disabling positional vertigo: selection criteria and operative results in 207 patients. Acta Neurochir (Wien). 1993b;125:75–82.

Møller AR, Jannetta PJ, Jho HD. Click-evoked responses from the cochlear nucleus: a study in human. Electroencephalogr Clin Neurophysiol. 1994;92:215–24.

N-Wihlbäck A, Sundström-Poromaa I, Bäckström T. Action by and sensitivity to neuroactive steroids in menstrual cycle related CNS disorders. Psychopharmacology (Berl). 2006;186:388–401.

Okamura T, Kurokawa Y, Ikeda N, Abiko S, Ideguchi M, Watanabe K, Kido T. Microvascular decompression for cochlear symptoms. J Neurosurg. 2000;93:421–6.

Pinaud R, Tremere L. Control of central auditory processing by a brain-generated oestrogen. Nat Rev Neurosci. 2012;13:520–7.

Schlee W, Hartmann T, Langguth B, Weisz N. Abnormal resting-state cortical coupling in chronic tinnitus. BMC Neurosci. 2009;10:11.

Schlee W, Lorenz I, Hartmann T, Müller N, Schulz H, Weisz N. A global brain model of tinnitus. In: Møller AR, Langguth B, De Ridder D, Kleinjung T, editors. Textbook of tinnitus. New York: Springer; 2010.

Sunderland S. Microvascular relations and anomalies at the base of the brain. J Neurol Neurosurg Psychiatry. 1948;11:243–57.

Syka J, Popelar J. Noise impairment in the guinea pig. I. Changes in electrical evoked activity along the auditory pathway. Hear Res. 1982;8:263–72.

Vanneste S, De Ridder D. The auditory and non-auditory brain areas involved in tinnitus. An emergent property of multiple parallel overlapping subnetworks. Front Syst Neurosci. 2012;6:31.

Vasama JP, Møller MB, Møller AR. Microvascular decompression of the cochlear nerve in patients with severe tinnitus. Preoperative findings and operative outcome in 22 patients. Neurol Res. 1998;20:242–8.

Microvascular Decompression Surgery for Glossopharyngeal Neuralgia

9

Anthony M. Kaufmann and Behzad Sabit

Abstract

Glossopharyngeal neuralgia is characterized by paroxysmal attacks of severe pain involving the sensory distributions of the glossopharyngeal and vagus nerves. A rare manifestation is hemodynamic disturbance or syncope related to excessive vagal outflow. Analogous to trigeminal neuralgia, classical glossopharyngeal neuralgia is caused by pulsatile vascular compression upon the ninth and tenth cranial nerve rootlets as they emerge from the medulla, and microvascular decompression surgery is potentially curative. The surgery is particularly challenging due to the narrow operative corridor, complex neurovascular relationships, and relatively small caseload experience. Optimal surgical outcomes are, however, achieved with careful patient selection, thorough understanding of the microsurgical anatomy, and adherence to the well-established operative techniques.

Keywords

Glossopharyngeal neuralgia • Vagoglossopharyngeal neuralgia • Microvascular decompression • Root entry zone • Anatomy • Surgery

9.1 Introduction

Glossopharyngeal neuralgia is a rare pain condition analogous to trigeminal neuralgia in regard to both clinical manifestations and therapeutic alternatives. It typically consists of brief attacks of severe, sharp pain involving the sensory distributions of the glossopharyngeal and vagus nerves (Nakahara et al. 2013, Tarlov 1942, and Teixeira et al. 2008). The condition was first described by Weisenberg in 1910, in a patient with a cerebellopontine angle tumor (Weisenberg 1910), and the term glossopharyngeal neuralgia was coined by

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Harris in 1921 (Harris 1921). White and Sweet suggested the condition may be more appropriately called vagoglossopharyngeal neuralgia, acknowledging the common involvement of both the vagus and glossopharyngeal nerves (White and Sweet 1969). As in trigeminal neuralgia, classical glossopharyngeal neuralgia is caused by pulsatile neurovascular compression on the cranial nerve root entry zone, and microvascular decompression surgery is potentially curative. The operative technique, however, is more challenging due to anatomical considerations and the relatively limited surgical experience with this condition.

9.2 Clinical Considerations

Glossopharyngeal neuralgia is rare with an incidence estimated at 0.2–0.7/100,000/year (Katusic et al. 1991; Koopman 2009; Manzoni and Torelli 2005). The pain is characterized by paroxysmal brief attacks, similar in character to trigeminal neuralgia (Cephalalgia 2013; Dandy 1927; Evans et al. 2006; Rozen 2004; Rushton et al. 1981). These come on suddenly and are described as sharp, stabbing, shooting, or likened to electric shock or pin prick. Attacks last seconds to 2 min and are localized to the posterior tongue, tonsil, throat, and sometimes the deep ear or beneath the angle of the jaw. They may occur spontaneously or be triggered by mechanical stimulations such as swallowing, coughing, yawning, or touching the back of the throat. Attacks may occur repeatedly in clusters, especially later in the disease course. There are often periods of remission, although a general progression over time is expected for most sufferers. Analogous to trigeminal neuralgia, glossopharyngeal neuralgia also occurs mainly in adults with a slight predilection for females and those over 50 years old and is almost always unilateral. Clinical examination is normal between pain attacks, although trigger points may be demonstrated in the oropharynx during exacerbations. The diagnosis is also supported by temporary relief from pain attacks with topical application of anesthetic to the posterior oropharynx. Anticonvulsants are not as consistently effective for glossopharyngeal neuralgia as for trigeminal neuralgia, and therefore response to such medical treatment cannot be considered a diagnostic test.

Glossopharyngeal neuralgia may rarely coexist with trigeminal neuralgia (Adams 1957). Differentiating between the conditions may be problematic if the predominant pain occurs in the shared sensory regions of the ear, tongue, and angle of the jaw (Kobata et al. 1998; Tubbs et al. 2011; Yoshioka et al. 1985). Glossopharyngeal neuralgia pain may also be associated with some symptoms referable to the trigeminal distribution, perhaps on the physiologic basis of shared input to the spinal nucleus of the trigeminal nerve (Rushton et al. 1981). Rare manifestations are episodes of bradycardia, hypotension, asystole, or syncope that occur in conjunction with the paroxysmal pain attacks or separately and are likely caused by excessive vagal outflow (Acosta and Clark 1970; Barbash et al. 1986; Esaki et al. 2007; Ferrante et al. 1995; Kong et al. 1964; Reddy et al. 1987; St John 1982; Thomson 1954; Wallin et al. 1984). Such cardiac syncope was found in only four of the 217 glossopharyngeal neuralgia cases reported in the Mayo Clinic series between 1922 and 1977 (Rushton et al. 1981).

Classical glossopharyngeal neuralgia is typically caused by neurovascular compression on the cranial nerve root entry zone. The area of transition from a central oligodendrocyte-derived myelin to peripheral Schwann cell-derived myelin, the Obersteiner-Redlich zone, occurs within 1–2 mm of the brainstem surface (Obersteiner and Redlich 1894). This most proximal portion of the nerve is particularly sensitive to pulsatile vascular compression where an elongated arterial loop is prone to become tightly lodged against the nerve rootlets emerging from the brainstem. High-resolution diagnostic imaging is useful to demonstrate such culprit neurovascular compression (Gaul et al. 2011). Glossopharyngeal neuralgia may also arise secondary to other pathologies including extracranial malignant tumors, trauma or infection involving the pharynx and larynx, an elongated styloid process, trauma, multiple sclerosis, or mass lesions in the posterior fossa. Such symptomatic glossopharyngeal neuralgia often

manifests with an element of constant, aching pain and associated deficits related to involvement of the cranial nerves in addition to the more typical lancinating pain attacks.

9.3 Management Considerations

The mainstay of initial treatment for glossopharyngeal neuralgia is anticonvulsant medication (De Simone et al. 2008; Evans et al. 2006; Garcia-Callejo et al. 1999; Kitchener 2006; Koopman et al. 2010; Luef and Poewe 2004; Ringel and Roy 1987; Rozen 2004; Titlic et al. 2006). Baclofen and antidepressants such as amitriptyline may also be helpful (Ringel and Roy 1987), while common analgesics are generally ineffective. Recurring pain may, however, progress to become excruciating and refractory to medical therapies. Surgical interventions should then be considered, as well as for those manifesting with cardiovascular symptoms.

The first surgical treatments for glossopharyngeal neuralgia were extracranial ablation procedures. These were associated with significant morbidity and proved relatively ineffective in achieving long-term relief (Adson 1924; Bruyn 1983; Doyle 1923; Mairs and Stewart 1990; Sicard and Robineau 1920; Wilson and McAIpine 1946). Supraganglionic sectioning of the glossopharyngeal nerve performed by an intracranial approach proved a more effective treatment option (Ceylan et al. 1997; Dandy 1927; Fraioli et al. 1989; Peet 1935; Taha et al. 1994; Rushton et al. 1981; Uihlein et al. 1955). Even higher rates of long-term pain control are achieved when a portion of vagal nerve rootlets are additionally sectioned (Bohm and Strang 1962; Love 1944). The combined sacrifice of glossopharyngeal and upper vagal rootlets usually results in relatively benign complaints of irritative cough, foreign body sensation in the throat, and transient hoarseness or dysphagia (Chawla and Falconer 1967; Giorgi and Broggi 1984; King 1987; Rushton et al. 1981). There is, however, an associated risk of more serious complications such as lost gag reflex, dysphagia, and vocal cord paralysis (Rey-Dios and Cohen-Gadol 2013).

Percutaneous radiofrequency rhizotomy directed at the jugular foramen was developed as an alternative approach to intracranial nerve sectioning, particularly for patients with symptomatic glossopharyngeal neuralgia due to malignancy (Arbit and Krol 1991; Barrow 1993; Esaki et al. 2007; Giorgi and Broggi 1984; Isamat et al. 1981; Lazorthes and Verdie 1974; Salar et al. 1983; Tew 1988). Other ablative procedures include peripheral glycerol injections (Strajcic 1989; Yue and Zhang 2013; Yue and Zhang 2014) and trigeminal tractotomy (Crue et al. 1972; Kanpolat et al. 1998; Kunc 1965). Stereotactic radiosurgery targeting the glossopharyngeal meatus at the jugular foramen has most recently been reported to provide good rates of early success, although lacks long-term follow-up (Martinez-Alvarez et al. 2014; Pollock and Boes 2011; Stanic et al. 2012; Stieber et al. 2005; Yomo et al. 2009).

Jannetta pioneered microvascular decompression surgery for the treatment of glossopharyngeal neuralgia (Laha and Jannetta 1977), following his earlier successes with trigeminal neuralgia (Jannetta 1967) and hemifacial spasm (Jannetta et al. 1977). Subsequent series have further demonstrated the safety and efficacy of this nondestructive procedure (Ferroli et al. 2009; Gaul et al. 2011; Kawashima et al. 2010; Kondo 1998; Matsushima et al. 2000; Michelucci et al. 1986; Olds et al. 1995; Patel et al. 2002; Resnick et al. 1995; Sampson et al. 2004; Sindou et al. 1991; Sindou and Mertens 1993; Wakiya et al. 1989). A comparison of recent reports has demonstrated the rates of long-term pain relief equal or exceed those achieved with rhizotomy procedures, with a three- to fourfold reduction in the risk of causing permanent cranial nerve deficits (Rey-Dios and Cohen-Gadol 2013).

9.4 Anatomical Considerations

There are notable challenges in performing microvascular decompression for glossopharyngeal neuralgia. The operative space is narrow, the neural elements are particularly delicate, the neurovascular relationships are complex, and the

volume of surgical experience is limited. These considerations help to emphasize the importance of a thorough understanding of the microsurgical anatomy (Fig. 9.1).

9.4.1 Cranial Nerves

The glossopharyngeal nerve is predominately a sensory nerve, but also carries secretomotor fibers. The functional components include (1) general somatic afferents from small areas of the postauricular skin and posterior fossa meninges terminating in the spinal tract and spinal nucleus of the trigeminal nerve; (2) general visceral afferents of touch, pain, and temperature sensation from the mucous membranes of the posterior third of the tongue, tonsil, posterior wall of the upper pharynx, and eustachian tube; (3) special visceral afferents from the carotid sinus and body as well as taste from the posterior third of the tongue (the visceral afferents terminate in the solitary tract nucleus); (4) general visceral efferents arising from the inferior salivary nucleus provide parasympathetic input to the otic ganglion and parotid gland; and (5) special visceral efferents arising from the nucleus ambiguus innervate the stylopharyngeus muscle (Carpenter 1985).

Rootlets of the glossopharyngeal nerve arising and terminating at the intramedullary nuclei emerge from the brainstem at the upper third of the postolivary sulcus of the medulla and converge into one or two bundles. These arise immediately inferior to the root exit point of the facial nerve that emerges from the pontomedullary sulcus. The proximal glossopharyngeal nerve is closely associated with the flocculus and choroid plexus that protrudes from the foramen of Luschka. It then traverses the subarachnoid

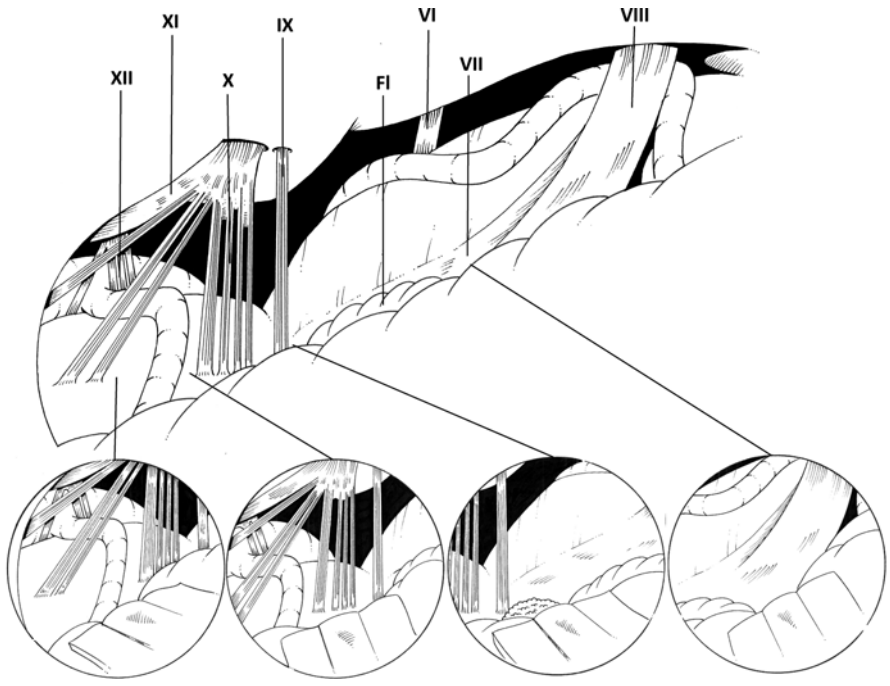

Fig. 9.1 A panoramic representation of anatomical relationships seen through a left retrosigmoid approach. The medulla is associated with vertebral and posterior-inferior cerebellar arteries, while the anterior-inferior cerebellar artery courses over the pons and in relation to the vestibulocochlear (*VIII*) and facial nerves (*VII*). The hypoglossal nerve (*XII*) arises from the anteriolateral medulla and courses over the posterior-inferior cerebellar artery origin. The glossopharyngeal (*IX*), vagal (*X*), and accessory (*XI*) nerves pass over the jugular tubercle and enter the jugular foramen. The flocculus (*Fl*) and choroid plexus (not shown) overlie the lateral portion of the pontomedullary sulcus. *Circles:* Surgical views gained by variations of approach trajectory and retraction

space, crosses over the jugular tubercle, and enters at the jugular foramen. The glossopharyngeal nerve may traverse the subarachnoid space adjacent to the vagus nerve fibers or be separated by several millimeters. At the jugular foramen, the glossopharyngeal nerve enters a dural meatus, separated from the more inferiorly situated vagal meatus by a dural septum that measures between 0.5 and 4.9 mm in width (Katsuta et al. 1997). Other components of the jugular foramen are not visible from the retrosigmoid approach, but include the inferior petrosal sinus situated superomedially and the sigmoid sinus situated inferior laterally from the neural elements. Injury to the glossopharyngeal nerve alone causes minimal deficits of reduced oropharyngeal sensation and diminished gag reflex.

The vagus nerve is closely related to the glossopharyngeal nerve sharing common nuclei and also containing a variety of both afferent and efferent fiber types. The functional components include (1) general somatic afferents from the cutaneous areas, back of the ear, and external auditory meatus terminating in the spinal tract and spinal nucleus of the trigeminal nerve; (2) general visceral afferents from the pharynx, larynx, trachea, esophagus, and thoracoabdominal viscera terminating in the solitary tract nucleus; (3) special visceral afferents from taste buds in the region of the epiglottis terminating in the solitary tract nucleus; (4) general visceral efferents from the dorsal vagal nucleus distributed to parasympathetic ganglia of the thoracic and abdominal viscera; and (5) special visceral efferents arising from the nucleus ambiguus that innervate the striated muscles of the pharynx and larynx.

The 8–10 vagal rootlets emerge from the postolivary sulcus, in a straight vertical line that is in continuity with the glossopharyngeal rootlets above and accessory nerve rootlets below. These enter the vagal meatus of the jugular foramen together with the accessory nerve. Dysfunction of the vagus nerve unilaterally will lead to uvular deviation to the normal side, hoarseness, dysphagia, and dyspnea related to a paralysis of the ipsilateral soft palate, pharynx, and larynx. An ipsilateral loss of cough reflex is related to anesthesia of the pharynx and larynx, while loss of visceral motor fibers of the vagus nerve will decrease the carotid sinus reflex.

The accessory nerve has two distinct parts. The cranial portion is a special visceral efferent arising from the nucleus ambiguus and innervates intrinsic muscles of the pharynx and larynx. These fibers emerge from the postolivary sulcus of the medulla, join the vagus nerve rootlets entering the jugular foramen, and continue as an "accessory" to the vagus nerve. The spinal portion of the accessory nerve is a general somatic efferent that arises from a column of anterior horn cells of the cervical spinal cord and innervate the sternocleidomastoid and upper portion of the trapezius muscles. The fibers emerge from the lateral aspect of the upper cervical cord segments and come together as a common trunk posterior to the dentate ligament. The nerve enters the posterior fossa through the foramen magnum and continues superiorly along the dural surface to the jugular foramen. Unilateral dysfunction of the accessory nerve produces weakness in turning the head against resistance and sagging of the shoulder with downward and outward rotation of the scapula.

The hypoglossal nerve is a general somatic efferent that provides motor innervation to the tongue. Fibers arise from the hypoglossal nucleus beneath the floor of the fourth ventricle and emerge as a series of rootlets from a groove between the lower two-thirds of the olive and medullary pyramid, the ventrolateral medullary sulcus. These fibers arise in the same vertical line as the ventral spinal roots. The rootlets course anterolaterally through the subarachnoid space and enter the hypoglossal canal. Dysfunction of the hypoglossal nerve produces a lower motor neuron paralysis of the ipsilateral tongue.

There is a close anatomical relationship between the lower cranial nerves arising from the medulla and the vestibulocochlear and facial nerves that arise from the pontomedullary sulcus. The vestibulocochlear nerve is most laterally situated and tethered to the anterior cerebellum and flocculus with fine arachnoid. Retraction of the cerebellum may therefore exert mechanical stress on the vestibulocochlear nerve that is particularly sensitive to even minor degrees of manipulation

and stretching (Sekiya et al. 1986). Its axons are invested with the central oligodendrocyte-derived myelin throughout the cisternal course. Furthermore, the cochlear nerve separates into multiple tiny filaments that traverse the *lamina cribrosa* at the fundus of the internal auditory canal, where they may be torn with minor degrees of traction (Sekiya and Moller 1987; Sekiya and Moller 1988).

The facial nerve root exit point also emerges from the pontomedullary sulcus, immediately medial to the vestibulocochlear nerve and directly superior to the glossopharyngeal nerve. It then courses superiorly, adherent to the bulging pontine surface before detaching from the pons and continuing across the prepontine cistern toward the internal auditory meatus. Its Obersteiner-Redlich zone is situated 1–2 mm distal to the root detachment point, such that the entire attached segment has myelin derived from oligodendrocytes. This long root exit zone of the facial nerve, from root exit point to the transition zone, is an important consideration during microvascular decompression for hemifacial spasm (Campos-Benitez and Kaufmann 2008; Kaufmann and Wilkinson 2005) but also highlights the relative sensitivity of the nerve to mechanical injury during surgery for glossopharyngeal neuralgia.

9.4.2 Arteries

The arteries of the posterior fossa have usual anatomical relationships with the cranial nerves, although there exist many variations and asymmetries. The length of these vessels, also, may vary considerably and predispose to vascular loops impinging on the cranial nerve roots as culprit neurovascular compression (Rhoton 2003). A thorough examination of the preoperative diagnostic imaging should allow the neurosurgeon to anticipate the course of these vessels (Fig. 9.2).

The distal cervical portion of the vertebral artery penetrates the dura lateral to the cervicomedullary junction and continues superiorly and anteromedially before merging with its contralateral mate at the level of the pontomedullary sulcus, becoming the basilar artery. The vertebral artery typically courses medial to the hypoglossal nerve and vertebrobasilar junction is between the abducens nerves. Branches of the vertebral artery include the anterior and posterior spinal arteries, meningeal arteries, medullary perforating arteries, and the posterior-inferior cerebellar artery origin.

The posterior-inferior cerebellar artery arises from the posterior lateral surface of the vertebral

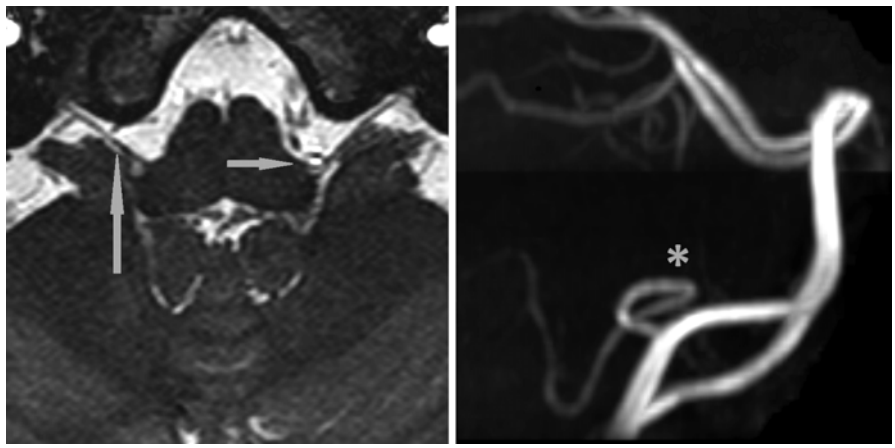

Fig. 9.2 Magnetic resonance imaging of a patient with left-sided glossopharyngeal neuralgia. *Left panel:* An axial CISS imaging sequence through the medulla. The glossopharyngeal nerve (*vertical arrow*) courses from the postolivary sulcus, across the jugular tubercle, and enters the jugular foramen. Culprit neurovascular compression is seen at the root entry zone (*horizontal arrow*). *Right panel:* A magnetic resonance angiography reconstruction with the elongated culprit posterior-inferior cerebellar artery loop (*asterisk*)

artery and usually courses laterally around the inferior end of the olive. This anterior medullary segment is inferior to most of the hypoglossal nerve rootlets but sometimes may go above or between them. The lateral segment then courses posteriorly between rootlets of the cranial nerves emerging from the postolivary sulcus or rarely rostral to the glossopharyngeal nerve. The vessel then continues posteriorly and inferiorly in the tonsillomedullary fissure, forms a caudal loop, and then ascends along the medial surface of the tonsil. It then forms a cranial loop before dividing over the inferior cerebellar surface.

The course of the posterior-inferior cerebellar artery varies considerably, as the length of each segment is commonly greater than required. Similarly, the vertebral arteries are often also asymmetrical and elongated. A common finding is a lateral and superior displacement of the vertebral artery with the posterior-inferior cerebellar artery origin situated well superior to the olive. In this configuration, fibers of the hypoglossal nerves may be stretched superiorly and cross the vertebral artery at the posterior-inferior cerebellar artery origin before looping inferiorly again toward the hypoglossal foramen. Elongated loops of the posterior-inferior cerebellar artery are the typical culprit in glossopharyngeal neuralgia, usually impinging on the medial aspect of the root entry zone but occasionally on the lateral side.

The anterior-inferior cerebellar artery is a rare cause of culprit neurovascular compression in glossopharyngeal neuralgia. The vessel arises from the basilar artery, usually between its inferior and middle thirds. The vessel loops around the inferior pons and through the cerebellopontine angle in close association with the facial and vestibulocochlear nerves. A loop toward the internal auditory meatus gives rise to the internal auditory artery. The distal vessel bifurcates with a superior and inferior division supplying the anterior cerebellum. An anterior-inferior cerebellar artery loop may rarely come into contact with the glossopharyngeal nerve root and its root entry zone, particularly when associated with an aberrant or hypoplastic posterior-inferior cerebellar artery.

The brainstem perforating arteries require special attention during microvascular decompression surgery. Both direct and circumflex vessels arise from the vertebral artery, anterior spinal artery, first three segments of the posterior-inferior cerebellar artery, as well as anterior-inferior cerebellar artery. In the setting of an elongated parent vessel, the perforating arteries will also be elongated between their origin and perforating point on the brainstem surface.

9.5 Surgical Technique

9.5.1 Opening

The patient is positioned lateral decubitus with the knees slightly bent and positioned such that the lumbar region is perpendicular to the floor. Rigid cranial fixation is employed with the head drawn posteriorly, while the chin is flexed, preserving a space of two fingerbreadths between the chin and thyroid cartilage. Additional adjustments include a 10–15° drop of the vertex and contralateral rotation to bring the retrosigmoid bone toward a horizontal plane.

The incision is planned to provide maximal access to the most anterior-inferior aspect of the retrosigmoid space, at the junction of the posterior fossa floor and sigmoid sinus. Landmarks include the mastoid tip that extends a couple of millimeters caudal to the posterior fossa floor, and the posterior edge of the mastoid process parallels the underlying sigmoid sinus. A straight line drawn between the lateral canthus, root of zygoma, and inion approximates the inferior edge of the distal transverse sinus behind the ear. A linear incision is marked 1 cm behind the mastoid process, superiorly to just above the transverse sinus line and inferiorly across from the mastoid tip (Fig. 9.3).

The incision is carried down through successive layers and a single self-retaining retractor placed to maintain the exposure. The occipital artery coursing between the splenius capitis and underlying longus capitis muscles may cross the field. Superiorly, the suboccipital muscle origin at the nuchal line is elevated in the subperiosteal

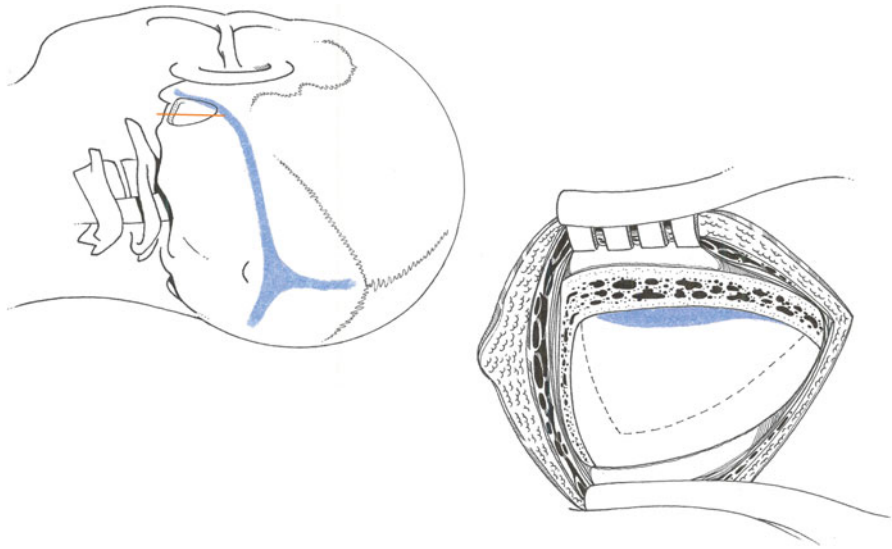

Fig. 9.3 Left retromastoid craniectomy. *Left panel:* The course of the transverse and sigmoid sinuses (*blue*) is determined form surface landmarks (see text). A linear skin incision (*orange*) provides the required exposure for a bony opening that extends to the sigmoid sinus laterally and posterior fossa floor inferiorly. *Right panel*: Soft tissue is held with a self-retaining retractor and bony opening completed. The planned dural flap (*dotted line*) provides maximal anterior and inferior access

plane. Inferiorly, the exposure should extend to the inferior edge of the occipital squamous bone. The posterior aspect of the mastoid process and digastric groove is cleared. An emissary vein usually coursing anterolaterally through the mastoid foramen toward the mid-sigmoid sinus is controlled with wax.

A low retrosigmoid craniectomy measuring up to 3×2 cm is fashioned with a high-speed pneumatic burr and 3–5-mm angulated rongeurs, exposing the posterior edge of the sigmoid sinus along its length. The occipital squamous bone is removed inferiorly until the floor is seen end-on (Fig. 9.4). Maximizing the anterolateral and inferior extent of this exposure provides direct access to the deep arachnoid cisterns and a clear surgical trajectory to the lower cranial nerve roots and medulla (Ferroli et al. 2009; Gaul et al. 2011; Olds et al. 1995; Patel et al. 2002; Resnick et al. 1995; Sindou and Mertens 1993). Some surgeons have advocated for additional anterior-inferior exposure through a transcondylar approach (Hitotsumatsu et al. 2003; Kawashima et al. 2010; Matsushima et al. 2000; Sampson et al. 2004), although we have used this only in cases with more complex pathologies such as aneurysms and cranial base tumors.

An L-shaped dural opening is reflected anteriorly overtop a saline-soaked Gelfoam. The inferior dural edge is retracted with a stitch or can be incised inferiorly if required to visualize the posterior fossa floor. Cerebrospinal fluid may spontaneously drain from the subdural space, or gentle compression of the cerebellum may facilitate its release, resulting in relaxation of the cerebellum. If the cerebellum remains full, the microscope is brought into use, and the cerebellum is elevated from the posterior fossa floor to access and open the deep arachnoid cisterns (Fig. 9.5). Access to cerebrospinal fluid can also be achieved by intraoperative lumbar puncture performed by the anesthesiologist using a 25-gauge spinal needle, while the surgeon remains attentive to the operative field. This is rarely required but helpful particularly for repeat microvascular decompression surgeries if early access to CSF is impaired by scarring of dura to the cerebellum.

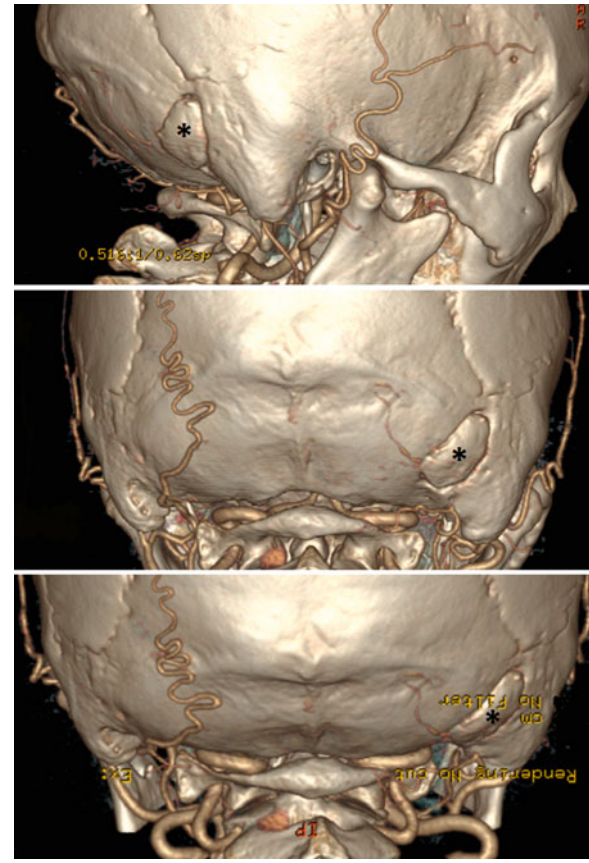

Fig. 9.4 Three-dimensional computerized tomographic reconstructions demonstrating a right retromastoid craniectomy, with cranioplasty closure (*asterisk*). The bony opening extends to the sigmoid sinus laterally and posterior fossa floor inferiorly

9.5.2 Approach

The anterior-inferior aspect of the cerebellum is elevated a couple of millimeters, and retraction is applied by suction and bayonet overtop a 1/2×3-inch cottonoid and latex sheet "slider" (Fig. 9.6). Bridging veins are identified as retraction is advanced. These can be released for the cerebellum with arachnoid dissection and preserved or divided after coagulation. The cervical portion of the accessory nerve courses on the dura from the foramen magnum toward the jugular foramen and should be identified to mechanical or thermal injury, particularly if coagulating a bridging vein.

The arachnoid exposed inferior to the jugular foramen provides the most direct access to the deep cistern, and when opened the cerebrospinal fluid release will further "deflate" the cerebellum. Further sharp dissection opens the arachnoid between the lower cranial nerves and cerebellum and is continued superiorly across the cerebellopontine angle. Separation of the cerebellum from the vestibulocochlear nerve can be achieved with careful dissection of fine arachnoid tethering these structures. This will facilitate brainstem exposure with minimal cerebellar retraction. Finally, exposure of the facial, glossopharyngeal, and upper vagal root exit/entry zones is achieved with elevation of the flocculus and closely associated choroid plexus (Fig. 9.7).

Throughout surgery, the degree of retraction should always be minimized as much as possible in order to avoid cerebellar compression and cranial nerve traction injuries. Each repositioning of retraction is gauged by observation of any resultant distortion or traction on neurovascular elements as well as intraoperative monitoring feedback, as further discussed below. Focused

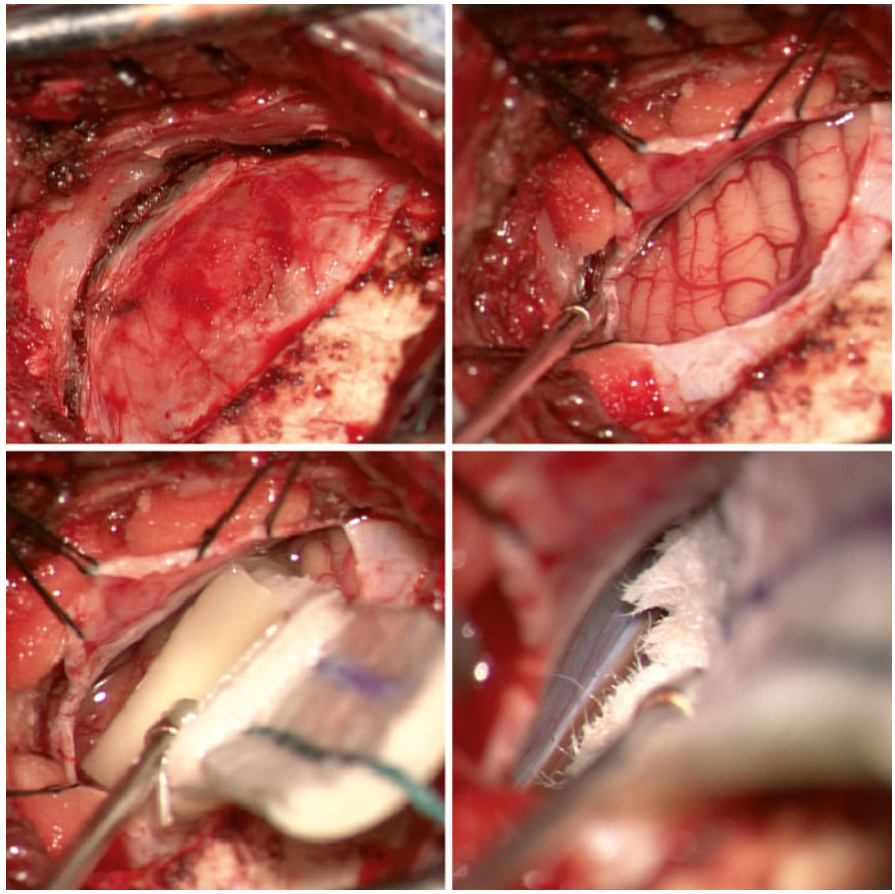

Fig. 9.5 Approach through a left retromastoid craniectomy. *Top left:* Mastoid bone edges have been sealed with wax and the sigmoid sinus edge is visible laterally. *Top right:* The dura has been opened in a curved fashion with an anterior flap reflected over moistened Gelfoam; a retraction stitch placed inferiorly. *Bottom left:* The cerebellum is gently depressed with a folded "slider," fashioned with a sheet of latex beneath a 1/2 × 3-inch cottonoid. *Bottom right:* The slider has been unfolded and advanced beneath the cerebellum parallel to the posterior fossa floor. This corridor will provide access to the deep arachnoid cistern

retraction is directed to provide clear exposure of one anatomical region at a time, rather than an all encompassing single panoramic view. On rare occasions we have utilized an angulated mirror to view behind the cranial nerves, as can be achieved also with an angulated endoscope (Ferroli et al. 2009). More commonly, all neural and vascular elements can be visualized by varying the microscope trajectory and adjusting retraction (Fig. 9.1).

9.5.3 Decompression

The most common culprit vessel in glossopharyngeal neuralgia is the posterior-inferior cerebellar artery, with the apex of a loop impinging against the glossopharyngeal and vagal nerve root entry zone. Displacement of the proximal nerve rootlets or indention of the medulla is often apparent, although even gentle pulsating compression is a sufficient cause. Conversely, vessels in simple contact with the cisternal portion of the nerve roots are incidental. The vertebral artery, anterior-inferior cerebellar artery, or a large vein may also contribute to neurovascular compression or rarely may be the sole culprit.

The technical objective of microvascular decompression surgery is to reorient the axis of the offending artery loop away from the target root entry zone, rather than simply cushioning

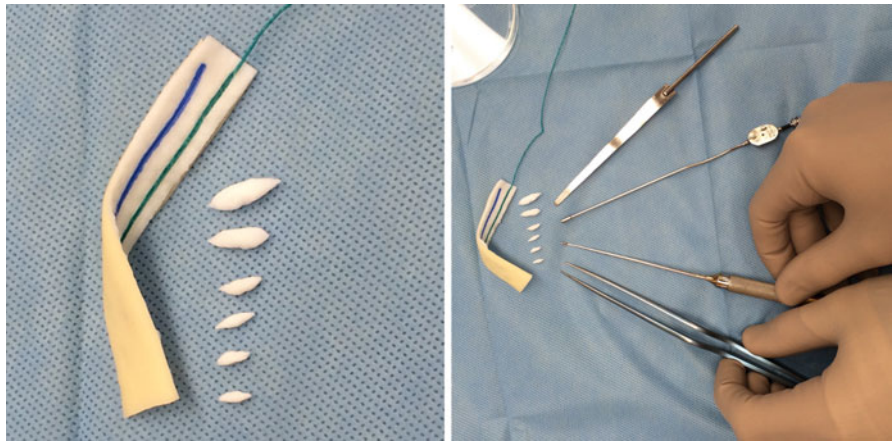

Fig. 9.6 Microsurgical materials. *Left panel:* A "slider" fashioned from a sheet of latex beneath a 1/2×3-inch cottonoid protects the brain. Various sizes of shredded Teflon felt implants are prepared (see text). *Left panel:* Microsurgical instruments include a tapered 3 mm self-retaining retractor blade, 5F smooth tipped suction, microdissectors and fine forceps, as well as microscissors and bipolar (not shown)

the neurovascular conflict. Beginning proximally and distally, away from the point of maximum compression, the vessel is lifted off the brainstem. Working toward the apex of the loop in a stepwise fashion, the entire vessel is shifted distally along the nerves, alleviating compression on the root entry zone. Care is taken to avoid tension on perforating vessels, and all arachnoid tethers are sharply divided. Implanted material is used to maintain the vessel in its new orientation, positioned between the vessels and brainstem, without pressure on the root entry zone or proximal rootlets. The tail of larger implants may also extend over the cerebellum to be further leveraged when retraction is eased (Fig. 9.8). When the offending artery forms a long loop, it is often possible to entirely transpose the vessel to a new position relative to the lower cranial nerves (Fig. 9.9). There is potential to kink particularly long vessel loops during transposition, and this can be recognized by visual inspection and checked with a Doppler flow probe. Rarely culprit veins may be encountered and considered for coagulation, although mechanical or thermal injury to brainstem rootlets and perforating vessels must be avoided.

We use shredded Teflon felt as an inert implant material. A variety of sizes are prepared at the beginning of each surgery. Forceps or hemostat is used to tease fibers from the felt that are then rolled between moistened fingers into the cigar shapes measuring approximately 1×3 to 6×15 mm (Fig. 9.6). Non-shredded Teflon felt pieces are not used, as these may densely scar against the dura and neurovascular structures, rarely inciting inflammatory reactions (Bobek and Sagher 1999; Chen et al. 2000; Premsagar et al. 1997). A variety of sling techniques have also been described to maintain a mobilized vessel in new position away from the target root entry zone (Bejjani and Sekhar 1997; Fukushima 1982; Kondo 1998; Kyoshima et al. 1999; Rawlinson and Coakham 1988; Sampson et al. 2004; Shigeno et al. 2002). On occasion, we have also utilized a sling technique utilizing a fenestrated aneurysm clip applied around the artery and then securing this with suture to the petroclival dura (Attabib and Kaufmann 2007).

Special care is required to avoid tension on the perforators, either during vessel mobilization or implant placement. These fine vessels will often have been elongated together with the parent vessel and thereby provide some latitude in vessel mobilization or transposition. Alternatively, short perforating vessels will occasionally limit a desired extent of decompression. The manipulated vessels are also prone to mechanically induced vasospasm that can be mitigated with topical papaverine applied on a soaked Gelfoam sponge or shredded Teflon implant.

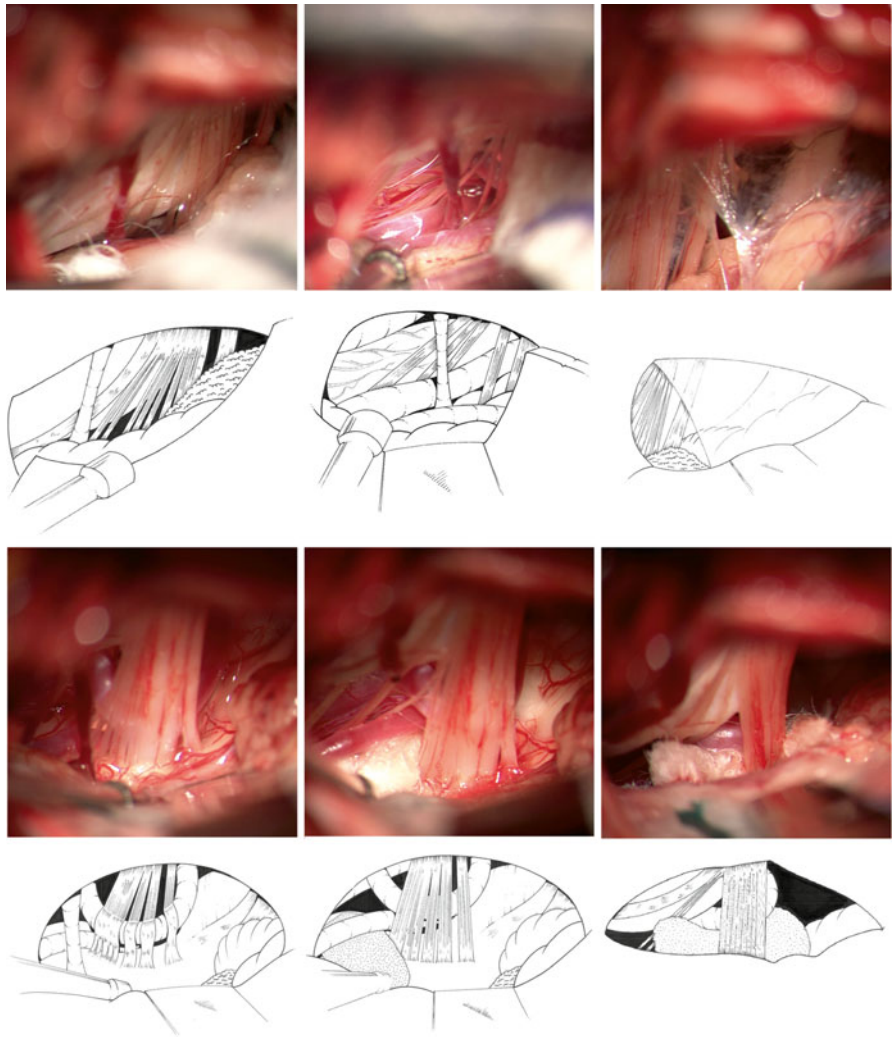

Fig. 9.7 Paired operative photographs and illustrations of microvascular decompression for left-sided glossopharyngeal neuralgia. *Top left:* Arachnoid has been opened over the lower cranial nerves entering the jugular foramen. A bridging vein is seen to cross over the cervical portion of accessory nerve, and the choroid plexus overlies the superior vagal and glossopharyngeal nerve roots. *Top middle:* The posterior-inferior cerebellar artery lateral segment courses tightly against the medulla, between the lower vagal rootlets and associated with a bridging vein. *Top right:* Superiorly in the surgical field, the arachnoid covers the cerebellum and flocculus. This will be widely opened and the flocculus further separated from the vestibulocochlear and glossopharyngeal nerves. *Lower left:* The flocculus is retracted superiorly to expose the glossopharyngeal and vagal rootlets emerging from the medulla. Immediately superior to this, the attached segment of the facial nerve emerges from the pontomedullary sulcus. A loop of the proximal posterior-inferior cerebellar artery is seen to impinge and distort the vagal and glossopharyngeal nerve root entry zone. *Bottom middle:* Mobilization is initiated away from the point of maximum compression, in this case elevating the lateral segment of the distal posterior-inferior cerebellum artery anteriorly off the medulla and maintaining this new position with a shredded Teflon felt implant placed between the nerve rootlets. This results in a shift of the more proximal culprit vascular loop distally along the lower cranial nerve rootlets, alleviating compression at the root entry zone. *Bottom right:* The culprit vascular loop is further mobilized distally toward the jugular tubercle and additional shredded Teflon felt implants placed beneath this vessel to maintain its new position

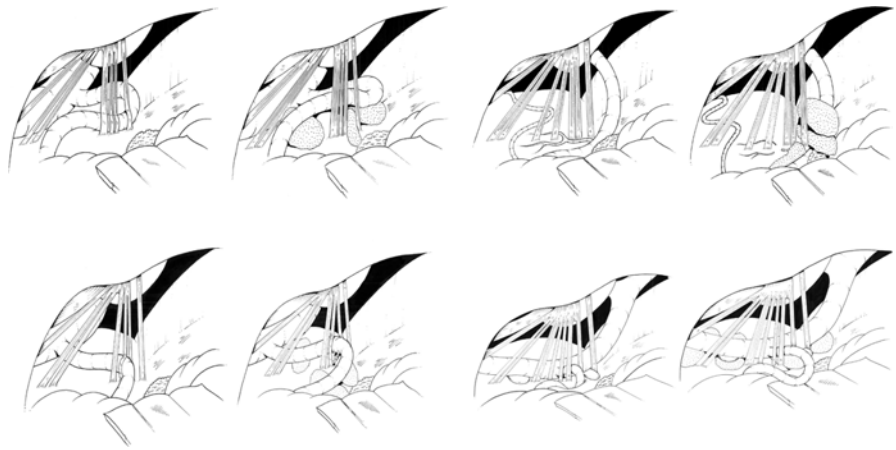

Fig. 9.8 A series of illustrations from four microvascular decompression surgeries for glossopharyngeal neuralgia showing culprit vessel position before and after mobilization away from the root entry zone. *Top left:* An elongated loop of the culprit posterior-inferior cerebellar artery compresses and distorts the glossopharyngeal and vagal root entry zone. This vessel is mobilized anteriorly from the brainstem and distally along the nerve roots, with shredded Teflon felt implants (speckled) maintaining the new vessel position. Complete pain relief was achieved. *Top right:* A prominent anterior-inferior cerebellar artery courses in close proximity to the glossopharyngeal root entry zone, as seen after elevation of the flocculus and choroid plexus. This vessel is mobilized further superiorly and shredded Teflon felt implants positioned. A small vein tracking around the vagal root entry zone is coagulated and divided, while the hypoplastic posterior-inferior cerebellar artery appears coincidental. This patient had nonclassical symptoms of glossopharyngeal neuralgia that did not improve after surgery. *Bottom left:* The posterior-inferior cerebellar artery courses between vagal nerve rootlets, causing a distortion at the root entry zone. Mobilization is limited by a small direct perforating branch. This patient did not improve after surgery and underwent subsequent sectioning of the glossopharyngeal nerve root and upper vagal rootlets at a second operation with complete pain relief but moderate laryngeal and pharyngeal weakness as a consequent to the ablative procedure. *Bottom right:* This patient had undergone a prior surgery elsewhere with sponge implants placed between the elongated vertebral artery and underlying medulla (clear ovals). At reoperation, the posterior-inferior cerebellar artery was seen to deeply impinge into the medulla, immediately lateral to the vagal root entry zone. This vessel loop was mobilized away from the brainstem and placed over the anterior cerebellum, with additional shredded Teflon felt implants placed to elevate the vertebral artery more distally along the nerve root and maintain separation of vessels from the vagal and glossopharyngeal root entry zone. Complete pain relief was achieved

The importance of hemostasis cannot be overemphasized, both for optimal microsurgical visualization and complication avoidance. Extradural hemostasis will prevent "run in" that may lead to distraction and obscure fine anatomical details. Bridging veins must also be thoroughly secured with bipolar coagulation before dividing. Protection of the brain surface is achieved with the "slider," gentle retraction, as well as avoiding unnecessarily large craniectomies and dural openings. If intradural bleeding is encountered, suctioning and directed irrigation should precisely locate the source and control attained with low-power bipolar coagulation. Heavy packing and indiscriminant coagulation are to be avoided in order to prevent collateral damage and potentially initiate additional bleeding. The optimal means to ensure hemostasis are, of course, gentle technique and avoidance of tissue and vessel injury.

9.5.4 Closing

A watertight closure can usually be achieved with primary suturing if the dura has remained moist throughout the surgery. The edges of the bone are sealed with a layer of wax, taking care not to "stuff" any exposed mastoid air cells. We routinely reconstruct the small craniectomy defect with a cranioplasty of methylmethacrylate, without the need for any rigid fixation. The

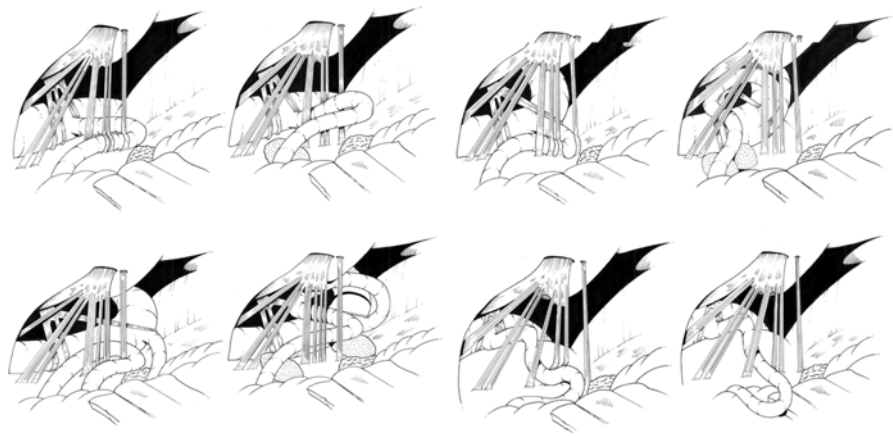

Fig. 9.9 A series of illustrations from four microvascular decompression surgeries for glossopharyngeal neuralgia showing culprit vessel position before and after transposition away from the root entry zone. *Top left:* The elongated posterior-inferior cerebellar artery arising from the laterally displaced vertebral artery is impinging on the medial aspect of the glossopharyngeal and upper vagal root entry zone, with displacement of the proximal nerve roots. The vessel is transposed anteriorly and laterally between the tenth nerve rootlets with shredded Teflon felt (speckled) elevating the vessel from the medulla. *Top right:* Similar neurovascular compression with indentation of the medulla at the root entry zone. The culprit loop is transposed inferiorly, as accommodated by long perforating vessels (not shown). Shredded Teflon felt is used to maintain the distal vessel and vertebral artery off the brainstem. *Bottom left:* The posterior-inferior cerebellar artery arises from an elongated vertebral artery at the pontomedullary sulcus, and a long loop impinges on a lateral aspect glossopharyngeal and vagal root entry zone. The vascular loop is mobilized medial to the lower cranial nerves. *Lower right:* A loop of the posterior-inferior cerebellar artery, posterior segment, impinges upon the lateral aspect of the vagal root entry zone. This culprit loop is mobilized off the medulla and transposed to reposition over the anterior cerebellum, elevated from the posteriolateral medulla. Complete pain relief was achieved in all of these patients

soft tissues are closed in multiple layers and antibacterial ointment is applied in lieu of a bandage.

9.6 Intraoperative Neurophysiological Monitoring

The monitoring of brainstem auditory-evoked potentials has been associated with complication avoidance in microvascular decompression surgery (Brock et al. 2004; Kaufmann and Wilkinson 2005; Lovely 1998; McLaughlin et al. 1999; Moller and Jannetta 1984; Moller and Moller 1989; Polo et al. 2004; Sato et al. 2009; Wilkinson et al. 2003). Attention to minimizing and correcting intraoperative latency shifts will not only help prevent cochlear nerve damage and hearing loss but also reduce the risk of other mechanical- or retractor-related injuries. Increased waveform latencies can often be corrected by releasing arachnoid tethers between the vestibulocochlear nerve and cerebellum or adjusting of retraction to reduce stretch or compression on the nerve. The degree and duration of latency shifts should be minimized as much as possible as delays beyond 1.5–2.0 milliseconds are associated with an increased risk of permanent hearing loss, especially when waveform amplitude diminishes (Moller and Moller 1989). Conversely, the loss of brainstem auditory-evoked potentials is not a definite evidence of a permanent hearing loss, and therefore measures to preserve and protect the vestibulocochlear nerve should be continued throughout the surgery (Wahlig et al. 1999).

The electromyography monitored during surgery is a helpful means to gauge the response of muscle groups innervated by the facial and lower cranial nerves. Inadvertent nerve contact by instruments or implants may generate a short burst of high-frequency responses (>50 Hz).

These responses are rarely neurotonic in nature, but should still be avoided using careful microsurgical technique. Conversely, prolonged high-pitched, high-frequency responses of up to 200 Hz are sometimes referred to as "mosquito buzzes" or "injury potentials" and probably indicate a compromise of nerve membrane integrity. This is a classic early warning of potential injury and should prompt measures to avoid further nerve irritation.

The glossopharyngeal nerve motor recordings can be obtained from needle electrodes inserted into the ipsilateral soft palate. The vagus nerve is monitored with either percutaneous insertion of needles into the cricothyroid membrane or a commercially available laryngeal surface electrode. Our preference is the latter, which easily attaches to the endotracheal tube prior to intubation. The correct placement of the recording electrode can be confirmed by intraoperative stimulation of the corresponding nerves intracranially. We also obtain recordings from facial and accessory nerve-innervated muscles. Most recently, facial motor-evoked potentials have proven valuable in microvascular decompression for hemifacial spasm (Wilkinson and Kaufmann 2005) (Kaufmann chapter) and now are routinely used in all our microvascular decompression surgeries.

9.7 Postoperative Course

Patients are observed in a neurosurgical step-down unit following microvascular decompression surgery, maintaining close attention to any alteration of vital signs or neurological examination. Approximately half of the patients are ambulatory and eating well on the first postoperative day and discharged from the hospital. Other patients have moderate headaches and nausea that typically subsides over a couple of days, such that the overall average length of stay is 2 days. As soon as postoperative fatigue and intermittent headaches resolve, usually over 2–8 weeks, patients may resume full levels of activity. At the time of hospital discharge, patients should be advised of potential delayed complications such as cerebrospinal fluid leak or wound infection. Meningitis is a rare complication, but more often aseptic rather than bacterial. Most patients experience immediate pain relief following surgery, and therefore microvascular decompression for glossopharyngeal neuralgia is useful under emergency conditions for medical refractory and severe pain flare-ups. Preoperatively prescribed anticonvulsants are generally tapered over 2 weeks to minimize any potential drug withdrawal effects.

9.8 Outcomes

Jannetta pioneered microvascular decompression surgery, and results from his center have included the original report of three patients undergoing this surgery for glossopharyngeal neuralgia (Laha and Jannetta 1977), then a series of the first 40 patients since 1971 with long-term follow-up (Resnick et al. 1995), and most recently a retrospective database review of the 217 patients treated from 1973 to 2000 (Patel et al. 2002). Although the latter report provided limited long-term follow-up results, it did clearly demonstrate that surgical outcomes and safety improved over time. This positive learning curve was driven by the volume of experience as well as advances in microsurgical anatomy knowledge and operative techniques including intraoperative monitoring (McLaughlin et al. 1999).

The recent series of microvascular decompression for glossopharyngeal neuralgia published in the current millennium have further exemplified these advances, with high rates of symptom relief and complication avoidance. In the largest of these series, neurovascular compression was evident and alleviated with complete pain relief in 46 of 47 patients (Sampson et al. 2004). Similar overall success was reported in the other series, with results of long-term pain relief of medications ranging from 89 to 95% (Ferroli et al. 2009; Gaul et al. 2011; Kawashima et al. 2010). Transient postoperative dysfunction of the vagus nerve was common, manifesting with hoarseness and/or dysphagia in 10–33 %. Most, however, recovered, and only mild

permanent vagal nerve deficits were reported in six of the combined 110 patients from these four series. Other complications included one mild permanent facial paresis, one CSF leak treated with three days of external lumbar drainage, one pulmonary embolism, and two cases of self-limited aseptic meningitis. There were no complications of deafness, death, or other major morbidity. Neurovascular findings in these series demonstrated the most common finding of a culprit posterior-inferior cerebellar artery loop (85 %), either alone (70 %) or in combination with other vessels (15 %). The vertebral artery and anterior-inferior cerebellar artery were the sole culprit in 7 and 3 %, respectively, or together in 1 %. Isolated venous compression was found in only 4 % and associated with failure to achieve pain relief.

As an alternative to microvascular decompression for glossopharyngeal neuralgia, some surgeons have instead recommended sectioning the glossopharyngeal nerve root or sectioning the glossopharyngeal nerve root together with some upper vagal nerve rootlets, either alone (Kandan et al. 2010; Xiong et al. 2012) or in combination with microvascular decompression (Brzustowicz 1955; Ceylan et al. 1997; Fraioli et al. 1989; Kondo 1998; Rushton et al. 1981; Taha and Tew 1995; Uihlein et al. 1955). These rhizotomy procedures performed through a retrosigmoid craniectomy do not require root entry zone exposure and vessel mobilization. Compared to microvascular decompression alone, the ablative procedures have similar potential to achieve permanent pain relief, particularly when upper vagal rootlets are sacrificed. There is, however, at least a threefold increase in permanent vagal nerve deficits caused by rhizotomy procedures compared to microvascular decompression surgery (Rey-Dios and Cohen-Gadol 2013). While the extent of vagal sectioning is directly related to long-term pain relief, it is similarly related to the risk and severity of resultant pharyngeal and laryngeal weakness. There is, also, no clear demarcation of how many of the upper vagal rootlets to divide to reach an optimal outcome.

A special consideration involves those presenting with glossopharyngeal neuralgia and associated cardiac symptoms or syncope. A comprehensive review of such cases included 68 patients plus another five added by the authors (Ferrante et al. 1995). A variety of treatments were summarized including anticonvulsants, cardiac pacemakers, rhizotomies, and microvascular decompression surgery of the glossopharyngeal and vagal nerves. Microvascular decompression surgery was the most effective and preferred treatment when arterial cross-compression on the glossopharyngeal and vagal nerves was evident. Alternatively, a Dandy procedure was advised if neurovascular compression was not present, as others have also suggested (Esaki et al. 2007; Ozenci et al. 2003). Our own practice is to perform microvascular decompression alone for patients with classical glossopharyngeal neuralgia or glossopharyngeal neuralgia with cardiac syncope and resort to rhizotomy of the glossopharyngeal nerve and upper vagal rootlets if repeat surgery is required.

9.9 Summary

Classical glossopharyngeal neuralgia is caused by pulsatile neurovascular compression of the glossopharyngeal and vagus rootlets emerging from the medulla. Surgical treatment is indicated when pain attacks become severe and medically refractory or whenever there are associated cardiac symptoms or syncope. The surgical options include a variety of ablative procedures or microvascular decompression. The latter has the potential advantage of minimizing surgically induced morbidity while achieving permanent pain relief. The surgery is challenging and very few neurosurgeons will gain a sizable volume of experience with this procedure. The impressive outcomes of published series can, however, be reached with a thorough understanding of the microsurgical anatomy and adherence to well-tested operative techniques.

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References

Acosta C, Clark K. Glossopharyngeal neuralgia associated with cardiac arrest. Case report. J Neurosurg. 1970;32(6):706–7.

Adams JE. Combined trigeminal and glossopharyngeal neuralgia. Neurology. 1957;7:72–4.

Adson AW. The surgical treatment of glossopharyngeal neuralgia. Arch NeurPsych. 1924;12:487–506.

Arbit E, Krol G. Percutaneous radiofrequency neurolysis guided by computer tomography for the treatment of glossopharyngeal neuralgia. Neurosurgery. 1991;29: 580–2.

Attabib N, Kaufmann AM. Use of fenestrated aneurysm clips in microvascular decompression surgery. J Neurosurg. 2007;106:929–31.

Barbash GI, Keren G, Korczyn AD, et al. Mechanisms of syncope in glossopharyngeal neuralgia. Electroencephalogr Clin Neurophysiol. 1986;63:231–5.

Barrow DL, editor. Surgery of the cranial nerves of the posterior fossa. New York: Thieme; 1993.

Bejjani GK, Sekhar LN. Repositioning of the vertebral artery as treatment for neurovascular compression syndromes. Technical note. J Neurosurg. 1997;86: 728–32.

Bobek MP, Sagher O. Aseptic meningitis caused by Teflon implantation for microvascular decompression. J Neurosurg. 1999;90:145–7.

Bohm E, Strang RR. Glossopharyngeal neuralgia. Brain. 1962;85:371–88.

Brock S, Scaioli V, Ferroli P, et al. Neurovascular decompression in trigeminal neuralgia: role of intraoperative neurophysiological monitoring in the learning period. Stereotact Funct Neurosurg. 2004;82:199–206.

Bruyn GW. General substantial review of the clinical features, etiology, differential diagnosis, and available treatment options. Glossopharyngeal neuralgia. Cephalalgia. 1983;3(3):143–57.

Brzustowicz RJ. Combined trigeminal and glossopharyngeal neuralgia. Neurology. 1955;5:1–10.

Campos-Benitez M, Kaufmann AM. Neurovascular compression findings in hemifacial spasm. J Neurosurg. 2008;109:416–20.

Carpenter M. Core text of neuroanatomy. Baltimore: Williams & Wilkins; 1985.

Ceylan S, Karakus A, Duru S, et al. Glossopharyngeal neuralgia: a study of 6 cases. Neurosurg Rev. 1997; 20:196–2000.

Chawla JC, Falconer MA. Glossopharyngeal and vagal neuralgia. Br Med J. 1967;3:529–31.

Chen J, Lee S, Lui T, et al. Teflon granuloma after microvascular decompression for trigeminal neuralgia. Surg Neurol. 2000;53:281–7.

Crue BL, Carregal JA, Felsoory A. Percutaneous stereotactic radiofrequency: Trigeminal tractotomy with neurophysiological recordings. Confin Neurol. 1972; 34:389–97.

Dandy W. Glossopharyngeal neuralgia (tic douloureux): its diagnosis and treatment. Arch Surg. 1927;15:198–214.

De Simone R, Ranieri A, Bilo L, et al. Cranial neuralgias: from physiopathology to pharmacological treatment. Neurol Sci. 2008;29(1):S69–78.

Doyle JB. A study of four cases of glossopharyngeal neuralgia. Arch Neur Psych. 1923;9:34–46.

Esaki T, Osada H, Nakao Y, et al. Surgical management for glossopharyngeal neuralgia associated with cardiac syncope: two case reports. Br J Neurosurg. 2007;21:599–602.

Evans RW, Torelli P, Manzoni GC. Glossopharyngeal neuralgia. Headache. 2006;46:1200–2.

Ferrante L, Artico M, Nardacci B, et al. Glossopharyngeal neuralgia with cardiac syncope. Neurosurgery. 1995;36: 58–63.

Ferroli P, Fioravanti A, Schiarti M, et al. Microvascular decompression for glossopharyngeal neuralgia: a long term retrospective review of the milan-bologna experience in 31 consecutive cases. Acta Neurochir. 2009;151:1245–50.

Fraioli B, Esposito V, Ferrant L, et al. Microsurgical treatment of glossopharyngeal neuralgia: case reports. Neurosurgery. 1989;25:630–2.

Fukushima T. Posterior fossa microvascular decompression in the management of hemifacial spasm and trigeminal neuralgia. No Shinkei Geka. 1982;10:1257–61.

Garcia-Callejo FJ, Velert-Vila MM, Talamantes-Escriba F, et al. Clinical response of gabapentin for glossopharyngeal neuralgia. Rev Neurol. 1999;28(4): 380–4.

Gaul C, Hastreiter P, Duncker A, et al. Diagnosis and neurosurgical treatment of glossopharyngeal neuralgia: clinical findings and 3-D visualization of neurovascular compression in 19 consecutive patients. J Headache Pain. 2011;12(5):527–34.

Giorgi C, Broggi G. Surgical treatment of glossopharyngeal neuralgia and pain from cancer of the nasopharynx. J Neurosurg. 1984;61:952–5.

Harris W. Persistent pain in lesions of the peripheral and central nervous system. Brain. 1921;44:557–71.

Headache classification committee of the international headache society. The international classification of headache disorders, 3rd edition (beta version). Cephalalgia. 2013;33:629–808.

Hitotsumatsu T, Matsushima T, Inoue T. Microvascular decompression for treatment of trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia: three surgical approach variations: technical note. Neurosurgery. 2003;53(6):1436–41.

Isamat F, Ferran E, Acebes JJ. Selective percutaneous thermocoagulation rhizotomy in essential glossopharyngeal neuralgia. J Neurosurg. 1981;55:575–80.

Jannetta PJ. Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neuralgia. J Neurosurg. 1967;26:159–62.

Jannetta PJ, Abassy M, Maroon JC, et al. Etiology and definitive microsurgical treatment of hemifacial spasm: operative techniques and results in 47 patients. J Neurosurg. 1977;47:321–8.

Kandan SR, Khan S, Jeyaretna DS, et al. Neuralgia of the glossopharyngeal and vagal nerves: long-term out-

come following surgical treatment and literature review. Br J Neurosurg. 2010;24(4):441–6.
Kanpolat Y, Savas A, Batay F, et al. Computer tomography-guided trigeminal tractotomy-nucleotomy in the management of vagoglossopharyngeal and geniculate neuralgias. Neurosurgery. 1998;43:484–9.
Katsuta T, Rhoton Jr AL, Matsushima T. The jugular foramen: microsurgical anatomy and operative approaches. Neurosurgery. 1997;41:149–202.
Katusic S, Williams DB, Beard CM, et al. Incidence and clinical features of glossopharyngeal neuralgia, Rochester, Minnesota, 1945-1984. Neuroepidemiology. 1991;10(5-6):266–75.
Kaufmann AM, Wilkinson MF. Microvascular decompression surgery for hemifacial spasm. In: Schmidek HH, editor. Schmidek & sweet operative neurosurgical techniques: indications, methods, and results. 5th ed. Philadelphia: Elsevier; 2005. p. 1473–90.
Kawashima M, Matsushima T, Inoue T, et al. Microvascular decompression for glossopharyngeal neuralgia through the transcondylar fossa (supracondylar transjugular tubercle) approach. Neurosurgery. 2010;66:275–80.
King J. Glossopharyngeal neuralgia. Clin Exp Neurol. 1987;24:113–21.
Kitchener JM. Glossopharyngeal neuralgia responding to pregabalin. Headache. 2006;46:1307–8.
Kobata H, Kondo A, Iwasaki K, et al. Combined hyperactive dysfunction syndrome of the cranial nerves: trigeminal neuralgia, hemifacial spasm, and glossopharyngeal neuralgia: 11-year experience and review. Neurosurgery. 1998;43:1351–62.
Kondo A. Follow-up results of using microvascular decompression for treatment of glossopharyngeal neuralgia. J Neurosurg. 1998;88:221–5.
Kong Y, Heyman A, Entman ML, et al. Glossopharyngeal neuralgia associated with bradycardia, syncope, and seizures. Circulation. 1964;30:109–13.
Koopman JS, Dieleman JP, Huygen FJ et al. Incidence of facial pain in the general population. Pain. 2009; 147(1-3):122–7.
Koopman JSHA, Huygen FJ, Dieleman JP, et al. Pharmacological treatment of neuropathic facial pain in the dutch general population. J Pain. 2010;11:264–72.
Kunc Z. Treatment of essential neuralgia of the 9th nerve by selective tractotomy. J Neurosurg. 1965;23: 494–500.
Kyoshima K, Watanabe A, Toba Y, et al. Anchoring method for hemifacial spasm associated with vertebral artery: technical note. Neurosurgery. 1999;45:1487–91.
Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg. 1977;47:316–20.
Lazorthes Y, Verdie JC. Radiofrequency coagulation of the petrous ganglion in glossopharyngeal neuralgia. J Neurosurg. 1974;4(6):512–6.
Love JG. Diagnosis and surgical treatment of glossopharyngeal neuralgia. Surg Clin North Am. 1944;24:959–62.
Lovely T. Efficacy and complications of microvascular decompression: a review. Neurosurg Quarterly. 1998;8:92–106.
Luef G, Poewe W. Oxcarbazepine in glossopharyngeal neuralgia: clinical response and effect on serum lipids. Neurology. 2004;63:2447–8.
Mairs AP, Stewart TJ. Surgical treatment of glossopharyngeal neuralgia via the pharyngeal approach. J Laryngol Otol. 1990;104:12–6.
Manzoni GC, Torelli P. Epidemiology of typical and atypical craniofacial neuralgias. Neurol Sci. 2005;26(2):65–7.
Martinez-Alvarez R, Martinez-Moreno N, Kusak ME, et al. Glossopharyngeal neuralgia and radiosurgery. J Neurosurg. 2014;121:222–5.
Matsushima T, Goto Y, Natori Y, et al. Surgical treatment of glossopharyngeal neuralgia as vascular compression syndrome via transcondylar fossa (supracondylar transjugular tubercle) approach. Acta Neurochir (Wien). 2000;142(12):1359–63.
McLaughlin MR, Jannetta PJ, Clyde BL, et al. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999;90:1–8.
Michelucci R, Tassinari CA, Samoggia G. Intracranial microvascular decompression for "cryptogenic" hemifacial spasm, trigeminal and glossopharyngeal neuralgia, paroxysmal vertigo and tinnitus: II. clinical study and long-term follow up. Ital J Neurol Sci. 1986;7:367–74.
Moller AR, Jannetta PJ. Monitoring auditory nerve potentials during operations in the cerebellopontine angle. Otolaryngol Head Neck Surg. 1984;92(4):434–9.
Moller AR, Moller MB. Does intraoperative monitoring of auditory evoked potentials reduce incidence of hearing loss as a complication of microvascular decompression of cranial nerves? Neurosurgery. 1989;24:257–63.
Nakahara Y, Matsushima T, Hiraishi T, et al. Importance of awareness of the rhomboid lip in microvascular decompression surgery for hemifacial spasm. J Neurosurg. 2013;119:1038–42.
Obersteiner H, Redlich E. Uber wesen und pathogenese der tabischen hinterstrangsdegeneration. Arb Neurol Inst Univ Wiein. 1894;1–3:158–72.
Olds MJ, Woods CI, Winfield A. Microvascular decompression in glossopharyngeal neuralgia. Am J Otol. 1995;16:326–30.
Ozenci M, Karaguz R, Conkbayir C, et al. Glossopharyngeal neuralgia with cardiac syncope treated by glossopharyngeal rhizotomy and microvascular decompression. Europace. 2003;5:149–52.
Patel A, Kassam A, Horowitz M, et al. Microvascular decompression in the management of glossopharyngeal neuralgia: analysis of 217 cases. Neurosurgery. 2002;50:705–11.
Peet MM. Glossopharyngeal neuralgia. Ann Surg. 1935;101:256–68.
Pollock BE, Boes CJ. Stereotactic radiosurgery for glossopharyngeal neuralgia: preliminary report of 5 cases. J Neurosurg. 2011. doi:10.3171/2011.5.JNS1133.
Polo G, Fischer C, Sindou MP, et al. Brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm: intraoperative

brainstem auditory evoked potential changes and warning values to prevent hearing loss – prospective study in a consecutive series of 84 patients. Neurosurgery. 2004;54(1):97–104.

Premsagar IC, Moss T, Coakham HB. Teflon-induced granuloma following treatment of trigeminal neuralgia by microvascular decompression: report of two cases. J Neurosurg. 1997;87:454–7.

Rawlinson JN, Coakham HB. The treatment of hemifacial spasm by sling retraction. Br J Neurosurg. 1988;2: 173–8.

Reddy K, Hobson D, Gomori A, et al. Painless glossopharyngeal "neuralgia" with syncope: a case report and literature review. Neurosurgery. 1987;21:916–9.

Resnick DK, Jannetta PJ, Bissonnette D, et al. Microvascular decompression for glossopharyngeal neuralgia. Neurosurgery. 1995;36(1):64–8.

Rey-Dios R, Cohen-Gadol A. Current neurosurgical management of glossopharyngeal neuralgia and technical nuances for microvascular decompression surgery. Neurosurg Focus. 2013;34(3):E8.

Rhoton Jr A. The cerebellar arteries. In: Apuzzo M, editor. Cranial anatomy and surgical approaches, vol. 53. Hagerstown: Lippincott Williams & Wilkins; 2003. p. 461–500.

Ringel RA, Roy III EP. Glossopharyngeal neuralgia: successful treatment with baclofen. Ann Neurol. 1987;21:514–5.

Rozen TD. Trigeminal neuralgia and glossopharyngeal neuralgia. Neurol Clin. 2004;22:185–206.

Rushton JG, Stevens JC, Miller RH. Glossopharyngeal (vagoglossopharyngeal) neuralgia: a study of 217 cases. Arch Neurol. 1981;38:201–5.

Salar G, Ori C, Baratto V, et al. Selective percutaneous thermolesions of the ninth cranial nerve by lateral cervical approach: report of eight cases. Surg Neurol. 1983;20:276–9.

Sampson JH, Grossi PM, Asaoka K, et al. Microvascular decompression for glossopharyngeal neuralgia: long-term effectiveness and complication avoidance. Neurosurgery. 2004;54:884–9.

Sato S, Yamada M, Koizumi H, et al. Neurophysiological mechanisms of conduction impairment of the auditory nerve during cerebellopontine angle surgery. Clin Neurophysiol. 2009;120(2):329–35.

Sekiya T, Moller AR. Avulsion rupture of the internal auditory artery during operations in the cerebellopontine angle manipulations: an electrophysiological and morphological study in dogs. Neurosurgery. 1987;21:631–7.

Sekiya T, Moller AR. Effects of cerebellar retractions on the cochlear nerve: an experimental study on rhesus monkeys. Acta Neurochir. 1988;90(1):45–52.

Sekiya T, Moller AR, Janetta PJ. Pathophysiological mechanisms of intraoperative and postoperative hearing deficits in cerebellopontine angle surgery: an experimental study. Acta Neurochir. 1986;81(3): 142–51.

Shigeno T, Kumai J, Endo M, et al. Snare technique of vascular transposition for microvascular decompression. Neurol Med Chir. 2002;42(4):184–9.

Sicard R, Robineau J. Communications et presentations: part I: algie véloparyngée essentielle: traitement chirugical. Rev Neurol. 1920;36:256–7.

Sindou M, Mertens P. Microsurgical vascular decompression in trigeminal and glossopharyngeal neuralgias: A 20 year experience. Acta Neurochir. 1993;58:168–70.

Sindou M, Henry JF, Blanchard P. Idiopathic neuralgia of the glossopharyngeal nerve: Study of a series of 14 cases and review of the literature. Neurochirurgie. 1991;37(1):18–25.

St. John JN. Glossopharyngeal neuralgia associated with syncope and seizures. Neurosurgery. 1982;10:380–3.

Stanic S, Franklin SD, Pappas CT. Gamma knife radiosurgery for recurrent glossopharyngeal neuralgia after microvascular decompression. Stereotact Funct Neurosurg. 2012;90:188–91.

Stieber VW, Bourland JD, Ellis TL. Glossopharyngeal neuralgia treated with gamma knife surgery: treatment outcome and failure analysis. Case report. J Neurosurg. 2005;102(Suppl):155–7.

Strajcic SS. Peripheral glycerol injection for the treatment of trigeminal neuralgia. Int J Oral Maxillofac Surg. 1989;18:255–7.

Taha JM, Tew Jr JM. Long-term results of surgical treatment of idiopathic neuralgias of the glossopharyngeal and vagal nerves. Neurosurgery. 1995;36:926–31.

Taha JM, Tew Jr JM, Keith RW, et al. Intraoperative monitoring of the vagus and upper vagal rhizotomy: technical note. Neurosurgery. 1994;35:775–7.

Tarlov IM. Section of the cephalic third of the vagus-spinal accessory complex: clinical and histologic results. Arch Neurol Psychiatry. 1942;47:141–8.

Teixeira MJ, de Siqueira SR, Bor-Seng-Shu E. Glossopharyngeal neuralgia: neurosurgical treatment and different diagnosis. Acta Neurochir. 2008;150: 471–5.

Tew Jr JM. Percutaneous rhizotomy in the treatment of intractable facial pain (trigeminal, glossopharyngeal, and vagal nerves). In: Schmidek HH, Sweet WH, editors. Operative neurosurgical techniques: indications, methods, and results. New York: Grune & Stratton; 1988. p. 1083–106.

Thomson JL. Glossopharyngeal neuralgia accompanied by unconsciousness. J Neurosurg. 1954;11:511–5.

Titlic M, Jukic I, Tonkic A, et al. Use of lamotrigine in glossopharyngeal neuralgia: a case report. Headache. 2006;46:167–9.

Tubbs RS, Mortazavi MM, Loukas M, et al. Intraoperative and anatomical descriptions of intracranial connections between the glossopharyngeal and vagus nerves: clinical implications. Laboratory investigation. J Neurosurg. 2011;115:179–81.

Uihlein A, Love JG, Corbin KB. Intracranial section of the glossopharyngeal nerve; sensory changes observed postoperatively. AMA Arch Neurol Psychiatry. 1955;74:320–4.

Wahlig JB, Kaufmann AM, Balzer J, et al. Intraoperative loss of auditory function relieved by microvascular decompression of the cochlear nerve. Can J Neurol Sci. 1999;26(1):44–7.

Wakiya K, Fukushima T, Miyazaki S. Results of microvascular decompression in 16 cases of glossopharyngeal neuralgia. Neurol Med Chir. 1989;29(12):1113–8.

Wallin BG, Westerberg CE, Sundolf G. Syncope induced by glossopharyngeal neuralgia: sympathetic outflow to muscle. Neurology. 1984;34:522–4.

Weisenberg TH. Cerebello-pontine tumor diagnosed for six years as tic douloureux. The symptoms of irritation of the ninth and twelfth cranial nerves. JAMA. 1910;54:1600–4.

White K, Sweet WH. Idiopathic vagoglossopharyngeal neuralgia. In: Thomas CC, editor. Pain and the neurosurgeon: a forty year experience. Springfield: C.C. Thomas; 1969. p. 265–302.

Wilkinson MF, Kaufmann AM. Monitoring of facial muscle motor evoked potentials during microvascular decompression for hemifacial spasm: Evidence of changes in motor neuron excitability. J Neurosurg. 2005;103(1):64–9.

Wilkinson M, Chan S, Kozey-Rodgers M, et al. Routine intra-operative monitoring improves neurological outcome. Can J Neurolog Sci. 2003;30:I07.

Wilson C, McAIpine D. Glossopharyngeal neuralgia treated by trans-tonsillar section of the nerve. Proc R Soc Med. 1946;40(2):82–3.

Xiong N, Zhao H, Zhang F. Vagoglossopharyngeal neuralgia treated by microvascular decompression and glossopharyngeal rhizotomy: clinical results of 21 cases. Stereotact Funct Neurosurg. 2012;90: 45–50.

Yomo S, Arkha Y, Donnet A, et al. Gamma knife surgery for glossopharyngeal neuralgia. J Neurosurg. 2009; 110:559–63.

Yoshioka J, Ueta K, Ohmoto T, et al. Combined trigeminal and glossopharyngeal neuralgia. Surg Neurol. 1985;24:416–20.

Yue WL, Zhang Y. Treatment for glossopharyngeal neuralgia with peripheral glycerol injection: Our experience in twenty-one older patients. Clin Otolaryngol. 2013;38(6):533–58.

Yue W, Zhang Y. Peripheral glycerol injection: an alternative treatment of children with glossopharyngeal neuralgia. Int J Pediatr Otorhinolaryngol. 2014;78: 559–61.

MVD for Neurogenic Hypertension: A Review

10

Marc Sindou and Andrei Brînzeu

Abstract

Experimental and clinical arguments to estimate that vascular compression of ventrolateral medulla at level of IX–Xth root entry/exit zone (REZ) seem convincing enough to consider vascular decompression as a possible measure for the treatment of apparently essential HT, likely to be of neurogenic origin.

The main problem remains patient selection, even when high-resolution MRI exploration shows images revealing megadolicho-artery(ies) likely to contribute to the pathology. Are these images of elongated arteries the cause or the consequence of the elevated blood pressure? Beyond such uncertainty, it could however be justified to consider decompressive surgery in the few selected patients in whom, in spite of a well-conducted medical treatment, blood pressure remains high, unstable, and thus life threatening.

Keywords

Neurovascular conflict • Vagus and glossopharyngeal nerves • Hemifacial spasm • Ventrolateral medulla • Functional neurosurgery • Microvascular decompression • Essential hypertension

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10.1 Introduction

Hypertension (HT) is a worldwide major problem that causes 7.1 million premature deaths and represents 4.5 % of the disease burden, namely, 64 million disability-adjusted life years (DALYs) according to the WHO International Society of Hypertension Writing Group (2003). Primary, in other words essential, HT is applied to the 95 % of cases in whom no specific etiology can be identified. Because not all essential HT cases can be controlled by medications, search for identifying eventual causative factors, among those, neurogenic dysfunction, is most justified. Among various factors responsible for neurogenic dysfunction, one cause may be vascular compression by an ectactic megadolicho-artery located at the rostro-ventro-lateral (RVL) aspect of the medulla, especially on the left side, as postulated by Jannetta and his group in the late 1970s (Jannetta et al. 1985a; Segal et al. 1982).

Since then, only few studies, either clinical or experimental (Sindou and Brinzeu 2015), were reported in the literature, despite such a neurogenic mechanism might potentially concern a large number of patients. As a matter of fact, there are robust anatomical/physiological evidences that compression of the IX–Xth root entry/exit zone (REZ) and adjacent RVL medulla can be at the origin of systemic HT (Morise et al. 2000; Smith et al. 2004). Related publications are reviewed in a recently published article (Sindou 2015).

Our contribution in the field was an assessment of the effects obtained on blood pressure (BP) by microsurgical vascular decompression (MVD) of the IX–Xth REZ and adjacent VL medulla (in addition to MVD of the facial REZ) in a group of patients operated on for their HFS and who presented with an associated apparent essential HT. Long-term effects of the MVD on those patients' HT was promising (Sindou and Brinzeu 2015).

10.2 Background

10.2.1 First Surgical Attempts on HT

The first clinical report on vascular compression as an etiology of essential HT was by Jannetta and Gendell in (1979). They observed a high rate of neurovascular compression of the VL medulla on the left side in their patients affected with trigeminal neuralgia or hemifacial spasm when there was an associated hypertension, compared to the patients who did not have HT. Since then, only two surgical studies were published: one, retrospective, by Jannetta et al. (1985b) and one, prospective, by Geiger et al. (1998). In Jannetta et al. series, of the 53 patients with essential HT, 51 had vascular compression at the left VL medulla. Among the 36 patients who benefited from a decompression considered "adequate," 32 (i.e., 89 %) had subsequent blood pressure normalization (see Table 10.1 for values). In Geiger et al. series, of the ten patients followed from 5 to 66 months, eight (80 %) were improved, three of them requiring no further antihypertensive medications. Improvement in blood pressure occurred more often in the patients who underwent surgery on the left side (Geiger et al. 1998; Jannetta et al. 1985b; Levy et al. 1998; Naraghi and Fahlbusch 1999). The predominant left-sided lateralization of blood pressure control is concordant with the prevalence of the left IX–Xth nerve complex in conveying the afferences originating from the baroreceptors of the (left-sided) cardiac atrium. Meantime, experiments in baboons were performed by the group of Jannetta to induce arterial hypertension by mimicking neurovascular pulsating compression in the VL medulla; authors used a pulsatile inflated balloon device. Hemodynamic changes were observed after pulsatile compression performed on left side, but not on the right side (Sindou 2015).

10.2.2 MRI Studies on HT

Because all patients affected with essential HT not-medically controlled cannot be candidates for surgical operation, a large number of studies

Table 10.1 WHO grading system for HTN

Grading	Systolic BP values	Diastolic BP values
Normal	≤140	≥90
Grade 1	140–159	90–99
Grade 2	160–179	100–109
Grade 3	≥180	≥110

were launched to evaluate the validity of MRI exploration to find out the eventual role of a vascular compression at brainstem in the genesis of the HT. Literature on MRI findings is reviewed elsewhere (Sindou 2015). Briefly, most publications report a higher rate of images of vascular contact/compression (vc/c) at the VL medulla in the group of patients with essential HT, compared to the group of patients with normal blood pressure. In a majority of those publications, rate of left-sided vc/c was higher than right-sided vc/c. However, significance was absent in the prospective studies ($p=0.178$) vs $p=0.001$ for retrospective studies.

A major limitation for MRI screening is the difficulty to discriminate whether a detected elongated arterial loop in relation with the VL medulla and the adjacent IXth–Xth REZ is the cause or the consequence of the raised blood pressure. Studies comparing image findings between patients with apparent primary HT and patients with secondary HT showed that in the group of patients with secondary HT, rate of NVC at brainstem was less than in the group of patients with primary – i.e., essential – HT. However those studies did not reach statistical significance.

10.2.3 Value of MRI Studies for Depicting Neurovascular Compressions

High-resolution sequences – 3DT2 MRI, 3D TOF MRA, and 3D T1 with gadolinium, in association – allow to detect the presence or absence of a vascular compression with high sensitivity and specificity, 96.7 % and 100 %, respectively (Leal et al. 2010).

To reach high level of reliability association of the three high-resolution sequences is important because of their complementarity (Leal et al. 2011). *3D T2 high-resolution* sequence obtains fine images of the root allowing to define its trajectory, caliber, and focal indentations. The cerebrospinal fluid has a hyperintense signal, while the other structures of the cerebellopontine angle have a hypointense signal, achieving a cisternography-like exploration. The limitation of this sequence is the absence of differentiation of signals between vessels and nerves. *3D TOF MRA*, if performed with a presaturation filter, visualizes only high-flow vessels, which are principally the arteries. *3D T1 sequence with gadolinium* is able to depict all vascular structures – that is, both arteries and veins – because they are both enhanced by contrast medium. The combination of the latter two sequences is indispensable to differentiate veins from arteries. Figures 10.1, 10.2, and 10.3 show examples of MRI studies of patients with vascular conflict at the level of the IX–Xth REZ and ventrolateral medulla in whom microvascular decompression improved their hypertension.

10.3 Surgical Technique

Installation and approach are approximately same as for HFS and VGPN MVD (Barker et al. 1995; Jianqing and Sindou 2015). The patient is placed in the contralateral decubitus position, the neck contralateral flexed to approach lower cranial nerves and VL medulla inferolaterally (Fig. 10.4).

A retromastoid craniectomy is performed posteriorly to the tip of the mastoid process with a semilunar shape of 2 cm in length and 1.5 cm in width, just posterior to the sigmoid sinus. The burr hole must not be turned too laterally onto the sigmoid sinus as this could endanger the external wall of the sinus, which is often reduced to a thin endothelial layer adhesive to the bone (Fig. 10.5). The use of a Doppler microprobe may help in detecting the posterior border of the sigmoid sinus. The dura is opened by making a small flap retracted along the sigmoid sinus. Then a self-retaining retractor, of the Yasargil type, mounted with a very thin blade of Sugita – Fukushima type – is placed on the inferolateral aspect of the cerebellum down to the cerebellomedullary fissure, to maintain the fissure opened (Sindou et al. 1992). The arachnoid is incised from XIth up to VIIIth nerves, and then the X–IXth root entry/exit zones are reached and the ventrolateral aspect of the medulla seen. The choroid plexus emerging from the lateral foramen of Luschka, which frequently covers the lower cranial nerves, is gently retracted to expose REZ and vessels at the brainstem (Fig. 10.6).

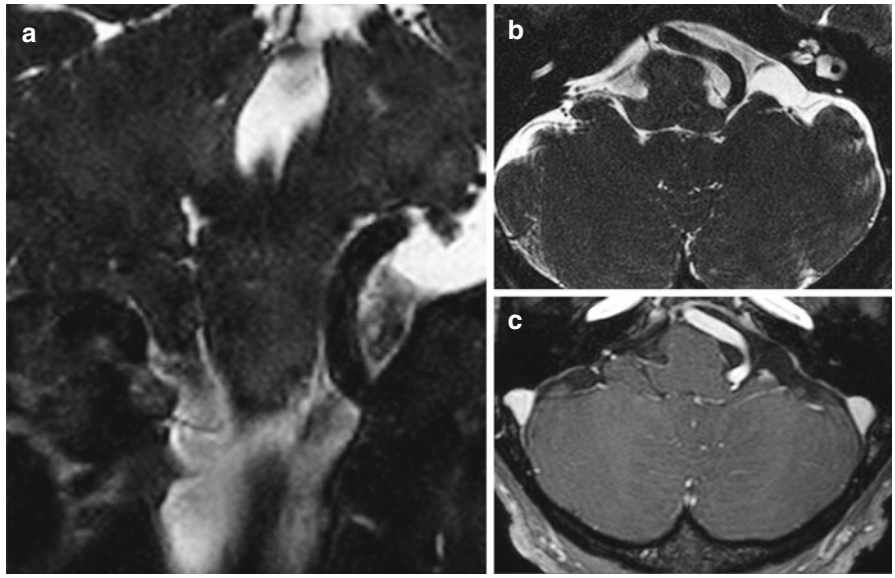


Fig. 10.1 MRI in a patient harboring left vagoglossopharyngeal neuralgia together with apparent essential arterial hypertension. (**a**) Coronal view of T2 high-resolution sequence showing compression of the ventrolateral aspect of medulla by vertebral artery (VA) on left side (*arrow*). Axial T2 (**b**) and T1 with gadolinium (**c**) showing VA and posterior inferior cerebellar artery conflicting, in addition to the brainstem, the root entry/exit zone of the IXth and Xth cranial nerves on left side

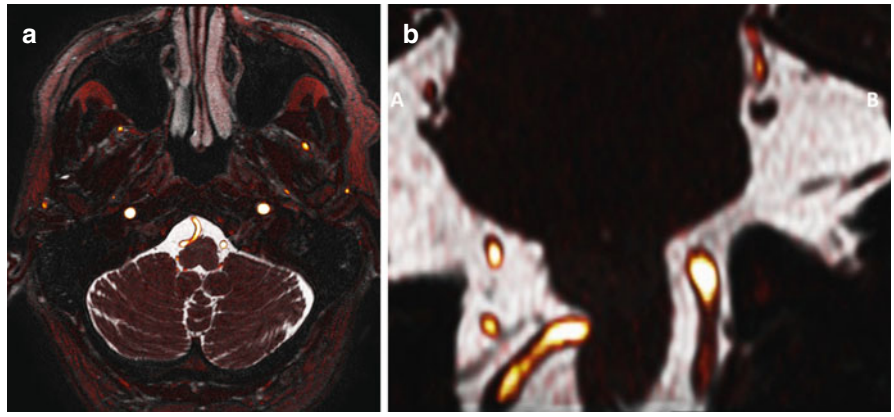


Fig. 10.2 MRI with fusion of T2 and TOF-angio in a patient with apparent essential hypertension. Axial (**a**) and coronal (**b**) views of the vertebral artery compressing the ventrolateral aspect of the medulla on the right side

The compressive vessels most frequently found in the group of patients affected with HT were the posterior cerebellar artery (PICA) and the vertebrobasilar artery (VBA) and frequently the association of both as shown in Table 10.2. Compression was almost always ventral to the Xth–IXth rootlets. This implies that maneuvers on the conflicting vessels be done passing between the rootlets at the several interspaces. During mobilization of the compressive arteries, care should be taken to respect the tiny perforating collaterals and not to generate mechanical vasospasm. Throughout vascular manipulations, irrigation with warm saline and application of a few droplets of papaverine in solution (1 in 10 mL saline) are important measures to limit vasospastic reactions. Not too much of papaverine should be used because of its very acid PH.

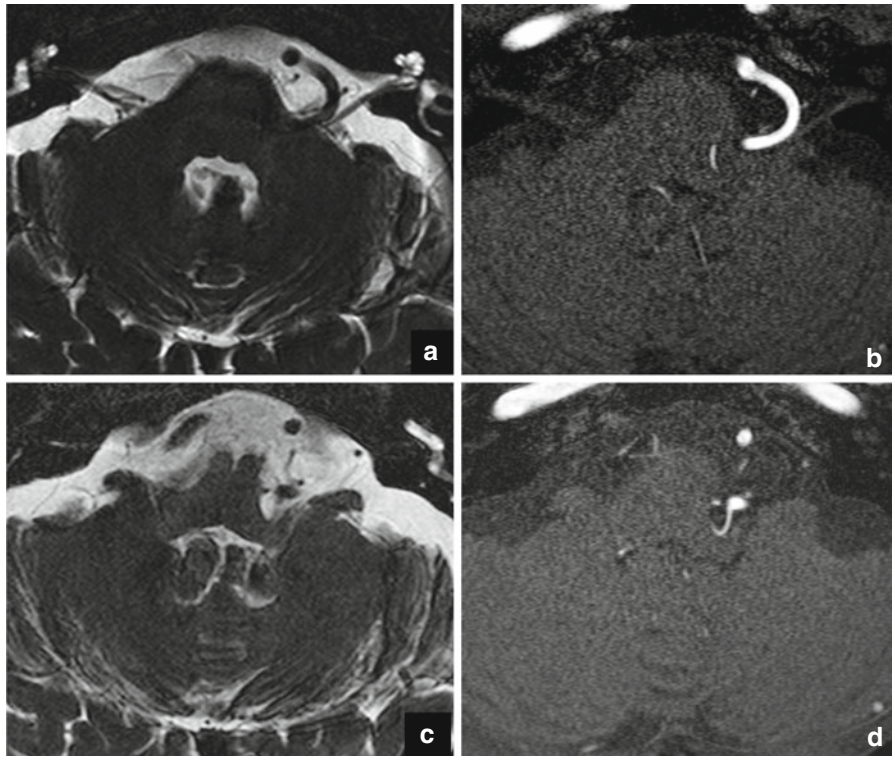


Fig. 10.3 MRI in a patient afflicted with left hemifacial spasm and apparent essential hypertension. *Upper row*: axial T2 (**a**) and TOF-angio (**b**) sequences showing megadolicho-vertebral artery (VA) and posterior inferior cerebellar artery (PICA) compressing the REZ of the VIIth cranial nerve (CN) on the *left side*. *Lower row*: axial T2 (**c**) and TOF-angio (**d**) sequences showing VA and PICA loops stretching and compressing the IXth and Xth CN on the left side

After the compressive arteries have been identified and dislodged from their conflicting situation, often marked by engrooving the ventrolateral aspect of the medulla, the vessels have to be maintained apart in such a way not to return to the previous compressive location.

When an arterial loop has a sufficient laxity, it can be kept apart by means of slings approximately 3–4 cm in length and 2–3 mm in width, made of shredded fibers of Teflon felt. Slings are passed around the vessel to exert a pulling effect and are blocked to avoid recurrence of malposition.

When an arteriosclerotic/atheromatous megadolicho-artery (PICA or VBA or the complex of VBA + PICA, as seen in Fig. 10.3) is in cause, its rigidity makes transposition difficult if not somewhat dangerous. Dissection should start at the brainstem from caudal to rostral and the vessel maintained away by inserting a piece of semirigid prosthesis (Dacron or Teflon) and/or a cushion of Teflon fibers in between REZ/brainstem and the artery. Care must be taken not to exert a (too strong) "neurocompressive" effect.

Closure should be tight to avoid CSF fistula. For tightness, dural suturing may need additional patch of fascia lata and/or fatty tissue affixed onto the mastoid cells if opened.

10.4 Results

Few studies have directly addressed the clinical problem of HT as a consequence of a vascular conflict at the level of the IX–X REZ or ventrolateral medulla. Overall results, while promising, are still far from conclusive; yet no randomized controlled trial has been published on the issue.

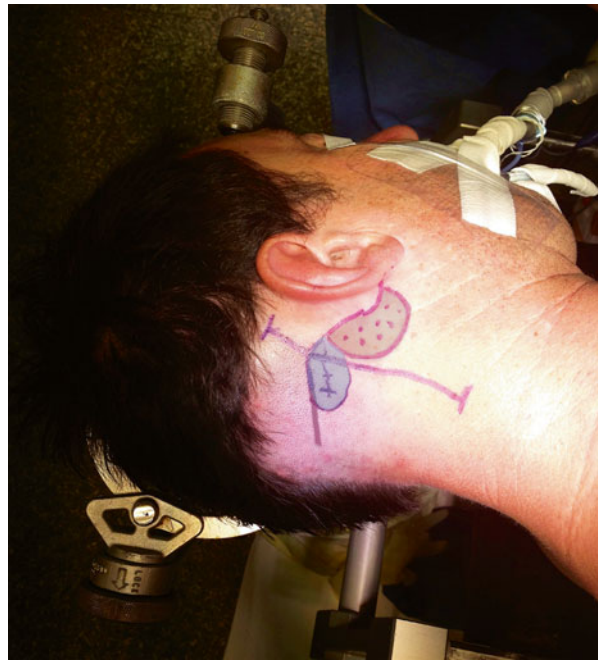


Fig. 10.4 Access to the IX–Xth REZ and ventrolateral aspect of the medulla should be following an infrafloccular trajectory along the cerebellomedullary fissure. Main concern is to avoid hearing loss. The lesser the exposure of the cochleovestibular nerve complex, the better for preservation of hearing and vestibular function. The patient is placed in the contralateral decubitus position, the head in a three-pin holder, slightly flexed and rotated 15° toward the contralateral side. The neck is laterally flexed toward the contralateral side to expose the retromastoid-retrocondylar region without the view obstructed by the shoulder taking care not to stretch the brachial plexus especially in patients with a gracile neck. The ipsilateral shoulder is tapped and pulled caudally and posteriorly. Landmarks of skin incision and craniectomy are drawn posteriorly to the tip of the mastoid process. Keyhole (retromastoid, retrosigmoid, infrafloccular) approach for accessing IXth and Xth cranial nerves (CN) and the ventrolateral aspect of medulla on the *right side*. Landmarks of mastoid tip (M), transverse (T) and sigmoid (S) sinuses. Craniectomy for infrafloccular, access to IXth and Xth CN (*dotted*), and for comparison, craniectomy for infratentorial supracerebellar access to trigeminal CN (*crosses*)

Studies published so far are case series, usually by one center and by a single surgeon. Table 10.4 summarizes the published series:

1. First observations of an effect of MVD of the ventrolateral medulla were published by Jannetta in 1979 in a series of patients operated on for glossopharyngeal neuralgia (Jannetta and Gendell 1979). This was later followed by the largest series yet published, 53 patients, benefiting from MVD for various reasons (mostly trigeminal neuralgia and HSF) but also having hypertension at the time of decompression (Jannetta et al. 1985b). Out of the 53, in 42, decompression of the ventrolateral medulla (exclusively on the left side) was performed. The compressing vessel was either the VBA or PICA or a combination of both in a vast majority (see Table 10.2). After a follow-up of at least 6 months (maximum FU 7 years), 31 patients had normalized their BP. In the normalized group, 13 patients had no treatment at all at latest FU, 12 had decreased their treatment, and 6 had kept the same treatment as preoperatively.

   The same group went on to publish in 1998 a series of patients in whom the indication for decompression was the refractory hypertension itself (Levy et al. 1998). In this retrospective series of 12 patients, all had only severe refractory HT with a major lability of BP values. All patients underwent preoperative MRI; however, existence of a conflict was determined at surgery.

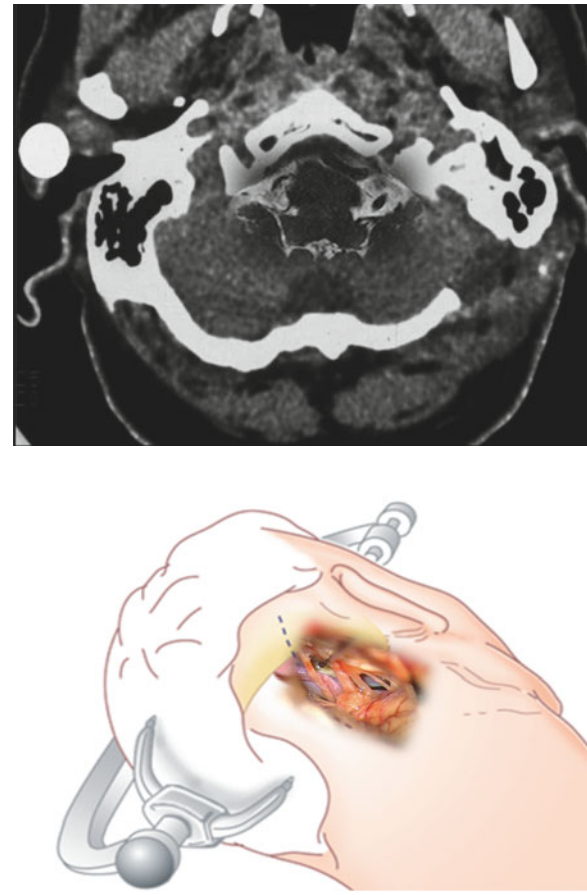


Fig. 10.5 Schematic views of IXth and Xth cranial nerves and ventrolateral (VL) aspect of the medulla on the *right side. Upper view*: superimposition of CT and MRI showing VL compression of brainstem by megadolicho-vertebrobasilar (VB) artery on the right side and retromastoid craniectomy to perform an inferolateral approach to the lower cranial nerves (CN). *Lower view*: superimposition of the lower part of the cerebellopontine angle to a drawing of the patient's head in the lateral decubitus position

Vessels were found to contact the REZ of the IXth–Xth pair or the ventrolateral medulla in all patients. In 11 the compressive vessel was PICA, whereas in the remaining one it was the VBA (see Table 10.2). Successful decompression was achieved in all patients. After a mean FU of 51.7 months, 8 patients had improved their BP with a mean drop of the systolic of 42.3 mmHg (20 mmHg was considered a minimum for declaring improvement). One patient had worsening of his HT, and the others showed no change.

2. A number of papers, also directly addressing refractory HT due to vascular conflict at the ventrolateral medulla, has been published by the team of Erlangen in Germany under the coordination of Pr. Fahlbusch. In the initial publication in The Lancet in 1998, eight patients had been prospectively studied (Geiger et al. 1998). Patients were included if MRI showed a conflict at the level of the VL medulla or IXth–Xth REZ. In six the conflict was caused by PICA on the left side, in one by PICA bilaterally, and in the last one by PICA and the VBA. At 3 months seven patients had improved their HT to the point of normalization with a decrease in treatment. Only five patients were followed up to 1 year, and out of these, four had normal BP values. In none of the patients, it was possible to halt the antihypertensive treatment. The series was followed for another 36 months for a mean FU of 3.5 years (Frank et al. 2001). At the end of this period, three patients had normal BP with little or no medication, and two had the same status as preoperatively. In the remaining three, severe complications of uncontrolled HT occurred, with two deaths related to high BP values.

In a parallel publication, the same team attempted to provide mechanistic data by following the evolution of BP and sympathetic activity in parallel in 14 patients after MVD

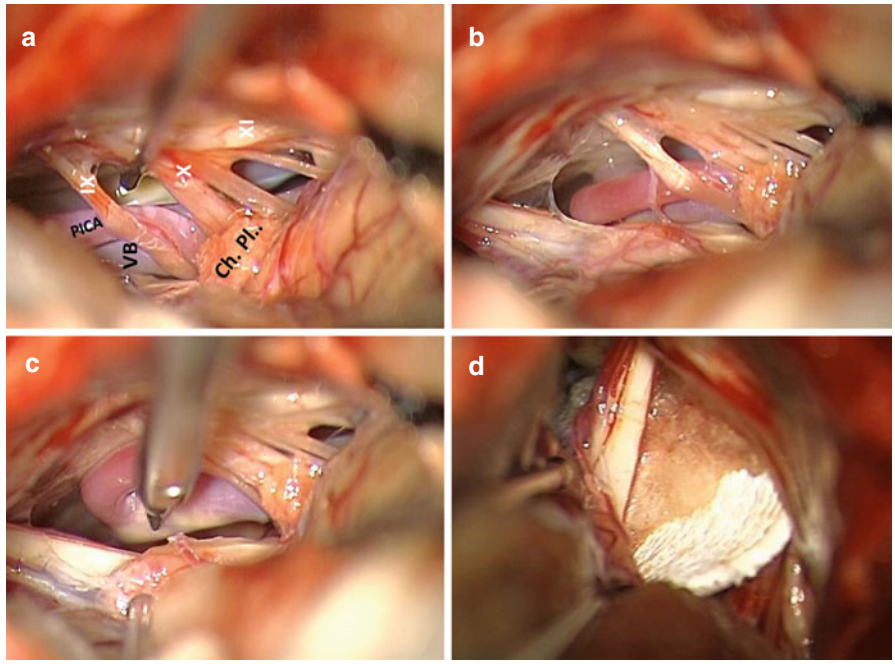


Fig. 10.6 Step-by-step technique for microvascular decompression of *IX*th and *X*th root entry/exit zone and ventrolateral (VL) aspect of the medulla on the right side. (**a**) Inferolateral floccular trajectory to *XI*th and *X*th–*IX*th cranial nerves, with REZ masked by choroid plexus (*ch. pl.*) REZ and VL medulla are compressed by a megadolicho-vertebrobasilar (*VB*) and posterior inferior cerebellar artery (*PICA*). (**b**) Access to the REZ after retraction of the choroid plexus. (**c**) Decompression after dissection of neural/vascular structures and translation of VB-PICA complex. (**d**) VB-PICA complex maintained apart after insertion of Teflon material

Table 10.2 Patient demographics and anatomical findings

		HT with HSF [*n* = 48] *n* (%)
Sex	Female	22 (46)
	Male	26 (54)
Age		35–82 years (60 years)
Side	Right side	18 (37.5)
	Left side	30 (62.5)
Compressive vessel(s)	VBA	2 (4)
	PICA	12 (25)
	AICA	15 (31)
	AICA + PICA	5 (10)
	VBA + PICA	10 (21)
	VBA + AICA	1 (2)
	VBA + PICA + AICA	3 (6)
Implication (alone or in association) of	VBA	16 (33)
	PICA	30 (63)
	AICA	24 (50)
Presence of multiple conflicts		19 (40)

VBA vertebrobasilar artery, *PICA* posterior inferior cerebellar artery, *AICA* anterior inferior cerebellar artery

for severe refractory HT. The series was published in Stroke in 2009 (Frank et al. 2009). Mean FU was 26 months. At 1 year patients showed a significant decrease in blood pressure (monitored over 24 h) from 179 mmHg on average preoperatively to 139 mmHg. Furthermore sympathetic activity (monitored by peroneal microneurography) had a similar evolution. However, BP values returned to above normal at 2 years postoperatively (average 152) and the same held true for sympathetic activity. This led the authors to believe that neurogenic HT was mediated through sympathetic hyperactivity and that although their results were limited further investigations were warranted.

3. Other small series and case studies were published by several authors with effects of HT being studied in patients undergoing MVD principally for other pathology (mostly HSF) (Morimoto et al. 1999; Van Ouwerkerk et al. 1989). These are detailed in Table 10.4.

Table 10.3 Evolution of long-term effects after MVD for HT

Grading	Pre MVD on medication *n* (%)	Post MVD at latest FU *n* (%)			*p* value
Normal	1 (2)	28 (58)	No treatment	14	<0.0001*
			Reduced treatment	10	
			Same treatment	4	
Grade 1	38 (79)	18 (38)			
Grade 2	9 (19)	1 (2)			
Grade 3	0 (0)	1 (2)			
Average grade	1.19	0.48			<0.0001*

Table 10.4 Results of MVD for HTN in the literature

Investigator	Date	Number of patients	Offending vessel	Indication for MVD	Mean FU	Results
Jannetta et al.	1979	16 patients	N/A	IXth neuralgia	N/A	12 improved
Jannetta et al.	1985	53 patients (42 with actual decompression of the medulla)	23 PICA L 20 VBA L 8 VBA & PICA L 1 SCA L 1 AICA L	HSF, GN and TN	8 years – 6 months	32 improved (20 mmHg decrease in BP)
van Ouwerkerk et al.	1989	7 patients	N/A	HFS	24 months	2 off medication 2 with half doses 3 no change
Levy et al.	1998	12 patients (an additional 7 unpublished)	PICA L VBA L	Only HT	51.7 months	8 improved (20 mmHg decrease in BP with a mean decrease of 42.3 mmHg) 2 no change 1 worsened (of the additional 7–2 normalized their BP)
Geiger et al.	1998	8 patients	6 PICA L 1 PICA Bilat 1 PICA + VB L	Only HT	12 months	At 12 months 4 normalized
Morimoto et al.	1999	1 patient	VBA L	HFS	5 months	Decrease in BP from 152/110 to 108/74 with elimination of 2 out of 3 drugs
Frank et al.	2001	8 patients	(Same as Geiger)	Only HT	3.5 years	3 normalized 2 had little change 3 had severe long-term HT with 2 deaths
Frank et al.	2008	14 patients	Left side Vessel not reported	Only HT	26 months	At 12 months average BP decreased from 162/98 to 133/85 at 24 months average BP was 158/96
Sindou	2015	48 patients	See Table 10.2	HFS	7 years (2–16 years)	At latest FU average systolic BP had decreased from 141 to 132 BP had normalized in 28 patients with 14 having no treatment after MVD

VBA vertebrobasilar artery, *PICA* posterior inferior cerebellar artery, *AICA* anterior inferior cerebellar artery, *SCA* superior cerebellar artery, *HFS* hemifacial spasm, *TN* trigeminal neuralgia, *GN* glossopharyngeal neuralgia, *L* left-sided conflict

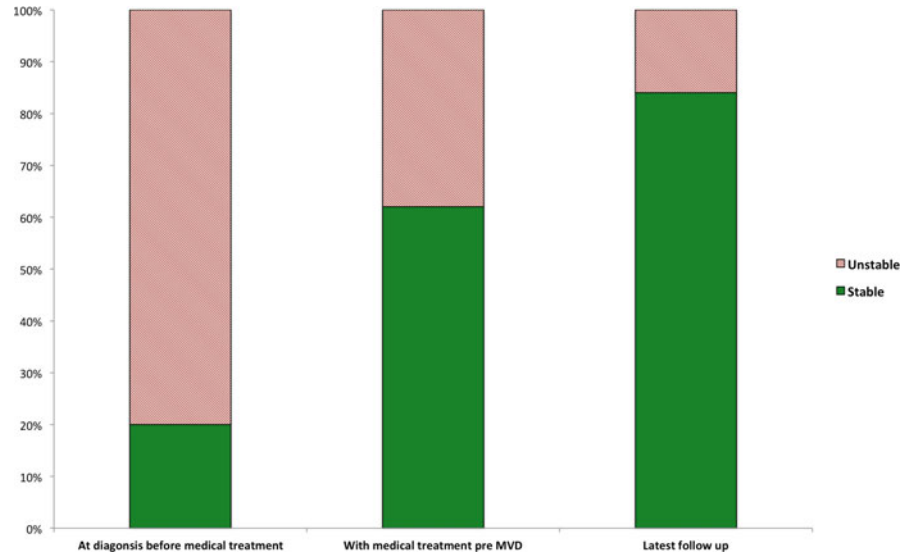


Fig. 10.7 Evolution of the stable/unstable character of the hypertension (at diagnosis, before and after MVD) [in a personal series of 48 patients]. A significant reduction in the number of patients having an unstable BP was noted after introduction of the medical treatment ($p<0.0001$ Bhapkar test for homogeneity). Similarly the number of patients with a stable blood pressure was significantly higher after MVD ($p=0.0003$ Bhapkar test for homogeneity)

4. The authors of this chapter have studied MVD effects on HT in a series of patients operated on for HFS (Sindou and Brinzeu 2015) and followed at least 2 years, up to 16 years with an average of 7 years after the surgery. Of 201 patients referred for HFS, 48 (23.8 %) had associated HT considered essential. All had high-resolution MRI that visualized a NVC (see Table 10.2). HT was severe in all of the patients with one exception, as in 47 BP was still WHO grade I or II and in 18 still unstable in spite of a medical treatment conducted by specialists in HT. Decision was taken not to discontinue nor modify medications for MVD surgery for safety (and also methodological) reasons. All underwent MVD of the IX–Xth root exit/entry zone (REZ) and adjacent ventrolateral medulla, in addition to VIIth REZ. Effects on HT, graded using the WHO classification (see Table 10.1), were studied up to latest follow-up, ranging from at least 2 years up to 16 years, 7 years on average. Also, effects of MVD on blood pressure (BP) according to the side of vascular compression were evaluated.

After MVD, at latest FU, BP returned to normal, i.e., below 140 mmHg of systolic, in 28 patients, of whom 14 (29.10 % of the whole series) without the need for any antihypertensive treatment. The 14 others still necessitated some medication to have their BP below 140 mmHg ($p<0.0001$) (Table 10.3). At latest FU, BP remained unstable in only 8 out of the 18 patients with instability prior to MVD ($p<0.02$) (Fig. 10.7).

Effects of MVD on BP along time were evaluated by comparing WHO grades at 1 year and at latest FU. The study shows a favorable delayed effect in a number of patients; in eight patients BP shifted to normal after the first year of FU (Fig. 10.8).

According to side, of the 30 patients with left-sided compression, 17 had BP normalized, 11 of whom without the need for any medical treatment. Of the 18 patients with right-sided compression, 11 had BP normalized, 3 of whom without medication. Difference between sides was not globally significant. However, when comparing groups of patients with BP normalized without the need for treatment, patients with

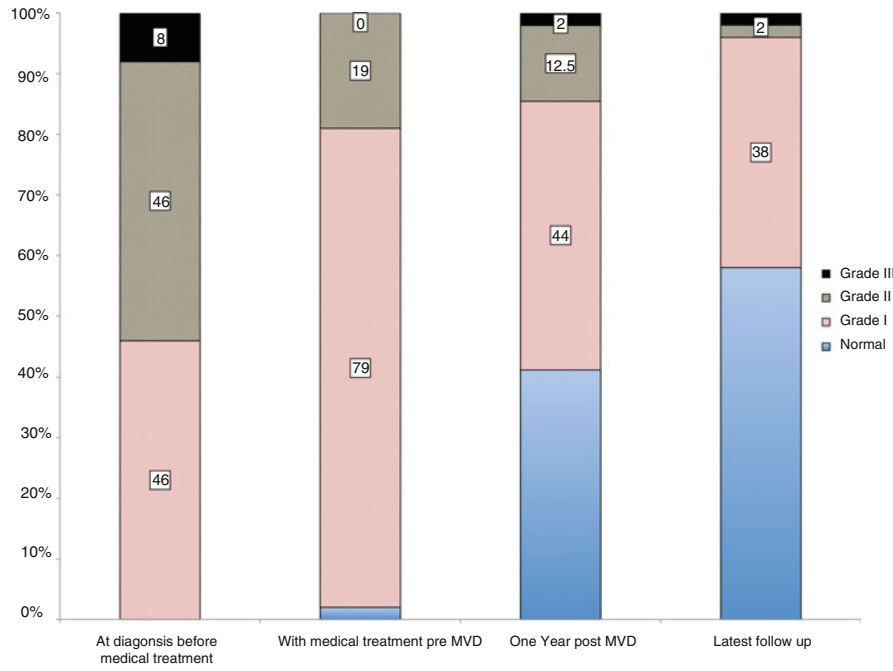


Fig. 10.8 Repartition of patients according to grades of WHO classification for hypertension [in a personal series of 48 patients] with evolution of outcomes along time. From *left* to *right*:

At diagnosis before introduction of antihypertensive drugs.

Before microvascular decompression (*MVD*) under medical treatment.

1 year after MVD.

At latest follow-up (FU) (inserted: respective percentages for each grade).

A significant difference was noted after the MVD procedure both at 1 year and at the latest FU (χ^2 test for both $p<0.01$)

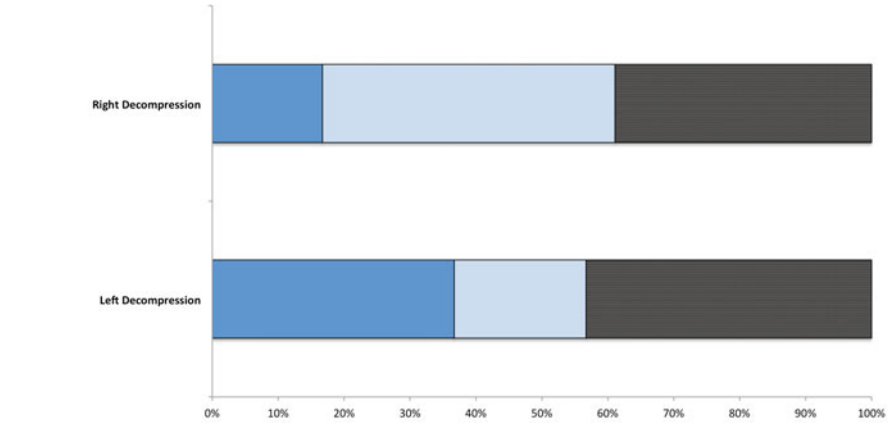
left-sided compression had better outcome (38 %) than right sided (16.7 %) (Fig. 10.9).

Complications of surgery in our 48 patients series were as follows: there was no mortality, permanent neurological disturbances amounted to 2 % for swallowing and vocal cord paresis, 4 % for hearing loss, and 2 % for balance disturbances.

5. Critics of these publications have pointed out that the studies were uncontrolled and that patients might have been undertreated for their hypertension preoperatively. While this is probably true in some cases, it is also true that at least in our series, medical treatment was independently managed by specialists in HT outside the surgical team and that a clear reduction in treatment could be shown. While no definitive conclusion can yet be drawn, these results encourage to consider MVD in severe cases of refractory HT if a conflict is discovered and most certainly encourage the build-up toward clinical randomized trials on the issue.

10.5 Discussion

Neurogenic hypertension by vascular compression of the IX–Xth REZ and the ventrolateral medulla has solid scientific bases from previous animal studies. However, transposing experimental data into clinical practice may be hazardous, and a clear-cut role of a neurovascular conflict in the genesis of hypertension is still to be demonstrated. Several problems remain unsolved, but all revolve around one evidence: hypertension is notoriously multifactorial; therefore, singling out one key factor in one specialty studies is at least questionable.



	Left Decompression	Right Decompression
Normalisation - No treatment	36.7	16.7
Normalisation - Still treated	20	44.4
No normalisation	43.3	38.9

Fig. 10.9 Repartition of patients' outcome according to the side of compression/surgery [in a personal series of 48 patients]. There was no statistically significant difference between right and left side of pathology/decompression, except when comparing groups with blood pressure normalized. In those decompressed on the left side, there was a significantly higher proportion of patients not requiring any treatment (36.7 % on the left side versus 15.7 % on the right side $p = 0.05$ χ^2)

Firstly, if the problem is addressed from a pathogenetic point of view, as shown previously, MRI studies are not able to differentiate a subpopulation with hypertension in whom an image of vascular conflict can be ascertained as the real cause. It remains a matter of debate whether a conflict on the ventrolateral medulla is the cause or the consequence of the hypertension. One cannot therefore rely solely on imaging studies to identify patients in whom the hypertension is neurogenic and amenable to surgery.

Secondly, benefit of decompression has yet to be demonstrated. Clinical studies are few and far between. Furthermore most of them make observations in series of patients operated on for other pathology. Moreover, these observations often lack the rigor of controlled clinical trials. Main weak points are related to the diagnosis of refractory hypertension as well as the way follow-up was ensured. Thus, it would be difficult to recommend straight forward microvascular decompression to a patient for solely the treatment of hypertension, albeit severe, and even facing clear-cut images of neurovascular conflicts.

However, in spite of these many unresolved issues, it may be admissible to consider MVD for some selected groups of patients with severe, unstable, and refractory hypertension. As a matter of fact, literature reports changes in the clinical situation after decompression of a marked vascular conflict of the ventrolateral medulla and IXth and X REZ.

The key problem is therefore patient selection. Research efforts should be oriented to investigations on the impact of the vascular compressions at the level of the IX–Xth REZ and ventrolateral medulla in the clinical setting of severe, refractory, unstable hypertension. The search of physiological "markers" of sympathetic dysfunction – likely to be the preeminent mechanism in the pathogenesis of hypertension through vascular conflict – could provide the necessary link between imaging and clinical manifestations. Additionally pharmacological tests of sympathetic blockade could help predict the effects of surgery.

When solid criteria for selecting potential candidates have been established, microvascular decompression can be proposed for the treatment of neurogenic hypertension likely to be generated by a vascular compression.

References

Barker FG, Jannetta PJ, Bissonette DJ, Shields PT, Larkins MV, Jho HD. Microvascular decompression for hemifacial spasm. J Neurosurg. 1995;82:201–10.

Frank H, Schobel HP, Heusser K, Geiger H, Fahlbusch R, Naraghi R. Long-term results after microvascular decompression in essential hypertension. Stroke J Cereb Circ. 2001;32:2950–5.

Frank H, Heusser K, Geiger H, Fahlbusch R, Naraghi R, Schobel HP. Temporary reduction of blood pressure and sympathetic nerve activity in hypertensive patients after microvascular decompression. Stroke J Cereb Circ. 2009;40:47–51.

Geiger H, Naraghi R, Schobel HP, Frank H, Sterzel RB, Fahlbusch R. Decrease of blood pressure by ventrolateral medullary decompression in essential hypertension. Lancet. 1998;352:446–9.

Jannetta PJ, Gendell HM. Clinical observations on etiology of essential hypertension. Surg Forum. 1979;30: 431–2.

Jannetta PJ, Segal R, Wolfson SK, Dujovny M, Semba A, Cook EE. Neurogenic hypertension: etiology and surgical treatment. II. Observations in an experimental nonhuman primate model. Ann Surg. 1985a;202:253–61.

Jannetta PJ, Segal R, Wolfson SK. Neurogenic hypertension: etiology and surgical treatment. I. Observations in 53 patients. Ann Surg. 1985b;201:391–8.

Jianqing C, Sindou M. Treatment of Vago-glossopharyngeal neuralgia with MVD and other neurosurgical procedures. In: Shi-Ting Li, Jun Zhong, Raymond F Sekula jr (editors). Microvascular decompression. Wien: Springer; 2015.

Leal PRL, Hermier M, Froment JC, Souza MA, Cristino-Filho G, Sindou M. Preoperative demonstration of the neurovascular compression characteristics with special emphasis on the degree of compression, using high-resolution magnetic resonance imaging: a prospective study, with comparison to surgical findings, in 100 consecutive patients who underwent microvascular decompression for trigeminal neuralgia. Acta Neurochir (Wien). 2010;152:817–25.

Leal PRL, Hermier M, Souza MA, Cristino-Filho G, Froment JC, Sindou M. Visualization of vascular compression of the trigeminal nerve with high-resolution 3T MRI: a prospective study comparing preoperative imaging analysis to surgical findings in 40 consecutive patients who underwent microvascular decompression for trigeminal neuralgia. Neurosurgery. 2011;69: 15–25; discussion 26.

Levy EI, Clyde B, McLaughlin MR, Jannetta PJ. Microvascular decompression of the left lateral medulla oblongata for severe refractory neurogenic hypertension. Neurosurgery. 1998;43:1–6; discussion 6–9.

Morimoto S, Sasaki S, Takeda K, Furuya S, Naruse S, Matsumoto K, Higuchi T, Saito M, Nakagawa M. Decreases in blood pressure and sympathetic nerve activity by microvascular decompression of the rostral ventrolateral medulla in essential hypertension. Stroke J Cereb Circ. 1999;30:1707–10.

Morise T, Horita M, Kitagawa I, Shinzato R, Hoshiba Y, Masuya H, Suzuki M, Takekoshi N. The potent role of increased sympathetic tone in pathogenesis of essential hypertension with neurovascular compression. J Hum Hypertens. 2000;14:807–11.

Naraghi R, Fahlbusch R. Microvascular decompression of the left lateral medulla oblongata for severe refractory neurogenic hypertension. Neurosurgery. 1999;45:955.

Segal R, Jannetta PJ, Wolfson SK, Dujovny M, Cook EE. Implanted pulsatile balloon device for simulation of neurovascular compression syndromes in animals. J Neurosurg. 1982;57:646–50.

Sindou M. Is there a place for microsurgical vascular decompression of the brainstem for apparent essential blood hypertension? A review. Adv Tech Stand Neurosurg. 2015;42:69–76.

Sindou M, Fobé JL, Ciriano D, Fischer C. Hearing prognosis and intraoperative guidance of brainstem auditory evoked potential in microvascular decompression. Laryngoscope. 1992;102:678–82.

Sindou M, Mahmoudi M, Brînzeu A : Hypertension of neurogenic origin: effect of microvascular decompression of the CNIX-X root entry/exit zone and ventro-lateral medulla on blood pressure in a prospective series of 48 patients with hemifacial spasm associated with essential hypertension. J Neurosurg. 2015:1–9.

Smith PA, Meaney JFM, Graham LN, Stoker JB, Mackintosh AF, Mary DASG, Ball SG. Relationship of neurovascular compression to central sympathetic discharge and essential hypertension. J Am Coll Cardiol. 2004;43:1453–8.

Van Ouwerkerk W, Samii M, Ammitatia M. Essential hypertension in patients with hemifacial spasm or trigeminal neuralgia. Adv Neurosurg. 1989;17:188–93.

WHO, ISH Writing Group. World Health Organization (WHO)/International Society of Hypertension (ISH) statement on management of hypertension. J Hypertens. 2003;21:1983–92.

11 Treatment of Vagoglossopharyngeal Neuralgia with MVD and Other Neurosurgical Procedures

Marc Sindou and Jianqing Chen

11.1 Introduction

Vagoglossopharyngeal neuralgia (VGPN), named as such (White and Sweet 1969) because the pain frequently affects not only the sensory territory corresponding to the glossopharyngeal nerve but also the one tributary of the sensory vagus nerve, represents about 0.2–1.3 % of all facial pain syndromes (Chawla and Falconer 1967). VGPN consists of paroxysmal, transient, severe, sharp pain in the back of the throat, the tonsillar fossa, the base of the tongue, the depth of the ear canal, and the area beneath the angle of the jaw. It usually lasts seconds to minutes and is often triggered by swallowing, eating, chewing, coughing, yawning, talking, etc. (Headache Classification Committee of the International Headache Society (IHS) 2013; Sindou et al. 2014). VGPN may also be associated with cardiovascular manifestations (Esaki et al. 2007; Ferrante et al. 1995; Greeson and Linden 1981; Jamshidi and Masroor 1976; Ozenci et al. 2003; Svien et al. 1957; Tew et al. 1982), and because of its association with cardiac dysrhythmias, some may be fatal.

11.2 Etiologies

VGPN may be secondary, due to tumors or various pathologies in the cerebellopontine angle (aneurysms, arachnoiditis, persistent hypoglossal artery, or petrositis, etc.). Also VGPN may be due to extracranial causes, like tumors in the oropharynx, elongated styloid process, ossification of the styloid ligament, tonsillitis, peritonsillar abscess, trauma, and even vertebral artery dissection (Hamada et al. 2013). An MR imaging exploration is recommended to rule out tumors, vascular lesions, Chiari malformations, etc.

VGPN may be idiopathic; in this type no demonstrable lesion can be found by definition. However, since last decades, these cases are mainly attributed to a IXth–Xth cranial nerve compression caused by a vessel, predominantly but not always at the root entry zone (REZ) of the brainstem. With modern MR imaging, these vessels can be evidenced with a high sensibility and a high specificity (Leal et al. 2010).

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11.3 Clinical Presentation and Diagnosis of Idiopathic VGPN

Incidence is estimated at 1 % compared to the one of trigeminal neuralgia (TN). VGPN is more common on the left side (left-to-right ratio of 3:2), TN being more common on the right (right-to-left ratio 5:3).

According to the International Headache Society, the diagnosis of VGPN is clinical; criteria are cited as follows (Headache Classification Committee of the International Headache Society (IHS) 2013; Sindou et al. 2014):

A. At least three attacks of unilateral pain fulfilling criteria B and C.
B. Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw, and/or in the ear.
C. Pain has at least three of the following four characteristics:
   1. Recurring in paroxysmal attacks lasting from a few seconds to 2 min
   2. Severe intensity
   3. Shooting, stabbing, or sharp in quality
   4. Precipitated by swallowing, coughing, talking, or yawning
D. No clinically evident neurological deficit.
E. Not better accounted for by another ICHD-3 diagnosis.

11.4 Treatment of Idiopathic VGPN

Management of idiopathic VGPN includes medical and surgical treatments. The medical treatment is the same as for trigeminal neuralgia, mainly based on anticonvulsant medications (Headache Classification Committee of the International Headache Society (IHS) 2013; Sindou et al. 2014). As for TN, carbamazepine is the first option; when insufficient or not tolerated, other anticonvulsants may be administered, namely, diphenylhydantoin, oxcarbazepine, clobazam, sodium valproate, topiramate, gabapentin, pregabalin, lamotrigine, and levetiracetam. As regards to non-antiepileptic drugs, association of baclofen may be useful. When the medical treatment fails or drug intolerance, allergies, or side effects become a problem, surgical treatment should be taken into consideration. At present microvascular decompression (MVD) is the first option, as the method is directed to the most frequent anatomical cause of VGPN and because MVD is of conservative nature.

11.4.1 Treatment with MVD

Dandy (1934) and Gardner (1953) raised the theory that vascular compression of the root entry/exit zone of the cranial nerves might be at the origin of hyperactivity syndromes. In 1977 Jannetta further investigated this mechanism and published the first series of patients with VGPN treated with microvascular decompression (MVD) (Jannetta 1977). Since then MVD gained greater acceptance than the traditional rhizotomy procedure, and a number of series of MVD were published regarding its satisfactory efficacy (Table 11.1). Modern MR imaging, using the association of high-resolution 3D T2, 3D TOF angiography, and 3D T1 + gadolinium, is able to predict with a high sensibility and a high specificity, presence or not, type, topography, and degree of compression of the cranial nerves (Leal et al. 2010). Figures 11.1 and 11.2 show high-resolution MR imaging in a patient with VGPN.

A recent review of literature included 28 series and regrouped 515 patients (Chen and Sindou 2015). Overall total relief rate ranges from 50 to 100 % according to the series (Table 11.1). In the more recent series, total relief rate was higher than 90 %. MVD has a lower recurrence rate, compared to the percutaneous RF thermocoagulation procedure. As a matter of fact, in well-experienced centers, MVD provides a good outcome with a high rate of pain relief (80–90 % according to main series). Because MVD is a delicate open intervention, it carries a risk of nerve deficit, variable according to the published series. In well-trained surgical team, these risks are statistically rare.

Table 11.1 Literature series with MVD of IXth and/or Xth cranial nerves, from 1977 to 2014

Authors and Year	No. of patients	Mean age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complications (%)
Laha and Jannetta (1977)	3	44.3	1 (33.3)	0.7	1(50)	1(50)	0	0
Jannetta (1980)	9	30–69	1 (11.1)	NR	6 (75)		2 (25)	T. decreased palatal and gag reflexes 2 (22.2)
Murasawa et al. (1985)	1	46	0	0.5	1	0	0	0
Tsuboi et al. (1985)	1	39	0	1	1 (100)			0
Yoshioka et al. (1985)	1	62	0	1	1 (100)			0
Michelucci et al. (1986)	3	56.9	0	1.8	3 (100)			T. headache 2 (66.6)
Wakiya et al. (1989)	16	54.7	0	2	15 (93.7)	1 (6.3)		
Sindou et al. (1991)	9	66	0	3.5	9	0	0	
Ferrante et al. (1995)	3	58.3	0	2.2	2	0	1	
Resnick et al. (1995)	40	55	2 (5)	4	28 (76)	6 (15)	3 (8)	P. paresis of IX and X 3 (8) T. paresis of IX and X 4 (10) infection1 (2) T. conjunctivitis 1 (2) T. hypertension 2 (5)
Platania et al. (1997)	1	58	0	NR	1	0	0	
Kondo (1998)	17	59.3	1 (5.9)	11.6	16 (94.1)			P. mild hoarseness 2 (11.8) T. coughing 2 (11.8)
Nishikawa et al. (2000)	1	47	0	NR	1	0	0	
Matsushima et al. (2000)	3	59.3	0	1.3	3 (100)			0
Patel et al. (2002)	217	50.2	3 (5.8)	4	29 (58)	9 (18)	12 (24)	Brainstem infarction 2 (0.9) CN palsy 15 (6.9) CFL 6 (2.8) Dysphagia (0.9)
Sampson et al. (2004)	47	56.4	0	12.7	28 (96.5)			T. hoarseness/ dysphagia 13 (28) T. facial paresis 3 (6) P. hoarseness/ dysphagia 4 (6) P. facial paresis 1 (2)
Ohyama et al. (2006)	1	61	0	NR	1			
Esaki et al. (2007)	2	NR	0	NR	2	0	0	0

(continued)

Table 11.1 (continued)

Authors and Year	No. of patients	Mean age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complications (%)
Ferroli et al. (2009)	31	55.8	0	7.5	28 (90.3)	3 (9.7)		T. dysphonia/ dysphagia 3 (9.7) T. hypoacousia 4 (12.9) T. VI/VII palsy 3 (9.6) CSFL 1 (3.2)
Sindou and Keravel (2009)	23	NR	0	9	21 (91 %)	0		P. CN palsy 2 (8.7)
Munch et al. (2009)	1	63	0	63	1 (100)	0	0	NR
Kawashima et al. (2010)	14	59.2	0	6.5	20 (95.2)	1 (4.7)		T. hoarseness/ dysphagia 4 (28) P. hoarseness/ dysphagia 2 (14)
Kandan et al. (2010)	15	52.5	0	4	14 (93.3)	1 (6.7)		T. hoarseness/ dysphagia 4 (28) P. hoarseness/ dysphagia 2 (14)
Ma et al. (2010)	4	61.5	0	2	4 (100)			0
Gaul et al. (2011)	18	54.5	0	NR	16 (88.9)	1 (5.5)	1 (5.5)	T. IX/X CN deficit 6 (33.3)
Martínez-González et al. (2011)	7	58	0	NR	7 (100)			NR
Xiong et al. (2012)	21	50.4	0	3.4	21 (100)			T. VIII deficit 5 (23.8) T. facial palsy 1 (4.7)
Wang et al. (2014)	6	60.1	0	NR	4 (66)	2 (33)	0	0

NR not reported, *T* transient, *P* permanent, *CSFL* cerebral spinal fluid leakage

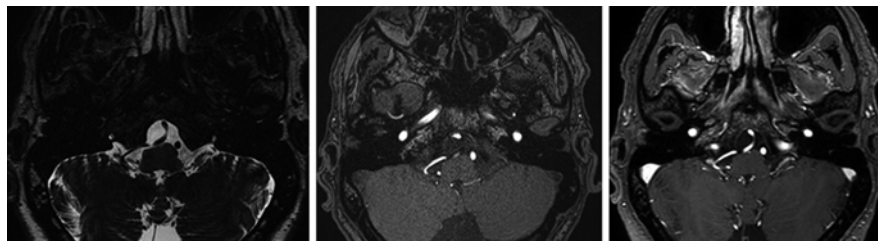

Fig. 11.1 From left to right, high-resolution T2, TOF angiography, and T1 + gadolinium MRI sequences showing neurovascular conflict between the IXth nerve (*arrow head*) and posterior inferior cerebellar artery [PICA] (*arrow*)

11.4.2 Technique for VGPN Decompression at Lyon University

11.4.2.1 Basic Principles

Indication for surgery is based on imaging. High-resolution MRI allows to predict neurovascular compression (NVC) and, if MVD is decided, to help surgical planning and design microsurgical approach that should be tailored according to individual features of every patient. To achieve high sensibility and high specificity, MRI exploration should associate the three following high-resolution sequences: 3D T2 to finely depict the neural structures and vessels and degree of compression, 3D T1 with gadolinium to show both

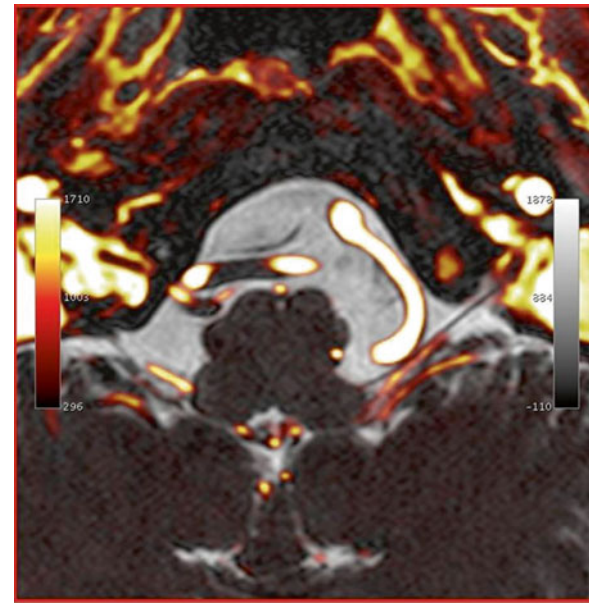

Fig. 11.2 T2 high-resolution MRI sequence showing the IXth nerve (*arrow head*) compressed by vertebrobasilar artery ventrally (*small arrow*) and posterior inferior cerebellar artery vascular loop dorsally (*large arrow*)

the arteries and veins, and 3D TOF angiography to differentiate the arteries from veins. As a matter of fact, the later sequence preferentially, if not exclusively, shows vessels with high velocity, i.e., the arteries, in almost an exclusive way when a superior band of presaturation filter is put to mask veins.

The main complication of MVD after surgery is hearing loss. As demonstrated by intraoperative BAEP recordings, principal cause is stretching of cochlear nerve due to lateral-to-medial retraction of the cerebellar hemisphere, Therefore, approach of the IXth–Xth REZ should be from below, passing inferolaterally to the cerebellar hemisphere and tonsil, i.e., following an infraflocculare trajectory along the cerebellomedullary fissure. The lesser the exposure of the cochleovestibular nerve complex, the better for hearing preservation. Figure 11.3 shows a schematic anatomic drawing of the lower part of the cerebellopontine angle, approached via an infraflocculare trajectory. Our current approach is as follows.

Under general anesthesia, the patient is placed in the contralateral decubitus position, the head in a three-pin holder, slightly flexed and rotated 15° toward the contralateral side. The neck is flexed to the contralateral side to access the retrocondylar region without view obstructed by the shoulder, but not too much to avoid stretching of the brachial plexus especially in patients with a gracile neck. The ipsilateral shoulder is taped and moderately pulled caudally and posteriorly. Hair is shaved in the retromastoid region. Then mastoid borders are identified by palpation with the index finger and landmarks of craniectomy drawn posteriorly to the tip of the mastoid process (Fig. 11.4).

The skin incision – 5 cm in length – is made obliquely, 1 cm medial to the bisector of the angle formed by the nuchal line and the posterior aspect of the mastoid process. The underlying subcutaneous tissue and muscles are divided using electrocautery, not too extensively to avoid damaging the occipital nerve. If the occipital artery is encountered, which is frequent with this approach, it is divided between two silk ligatures. The posterior aspect of the mastoid process together with the digastric groove and the retrocondylar fossa are cleared of soft tissue. The mastoid emissary bony vein is obliterated first with a small pledget of Surgicel and then waxed.

Then, a retromastoid craniectomy of the keyhole type is performed, posterior to the tip of the mastoid process, next to the retrocondylar fossa, with a semilunar shape of 2 cm in length and

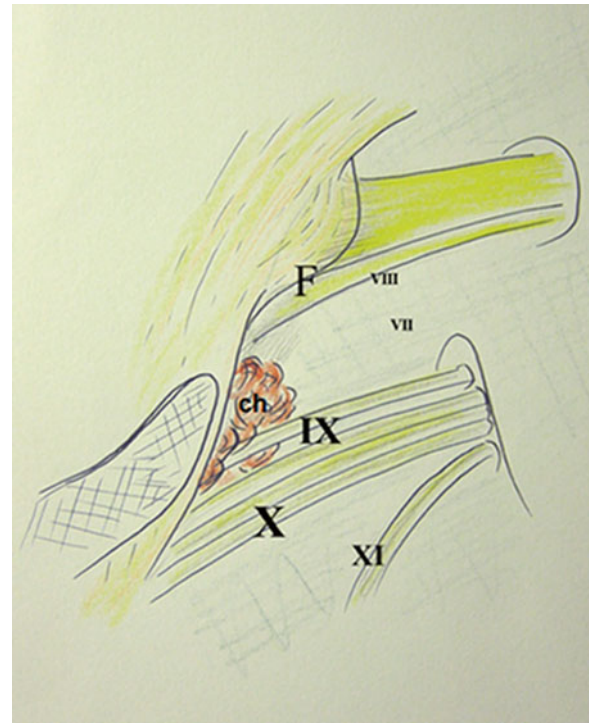

Fig. 11.3 Schematic view of lower part of cerebellopontine angle with lower cranial nerves (*IXth*, *Xth*, *XIth*), on the right side. Approach via infrafloccular (*F*) trajectory. Note choroid plexus (*ch*) covering the IXth–Xth REZ

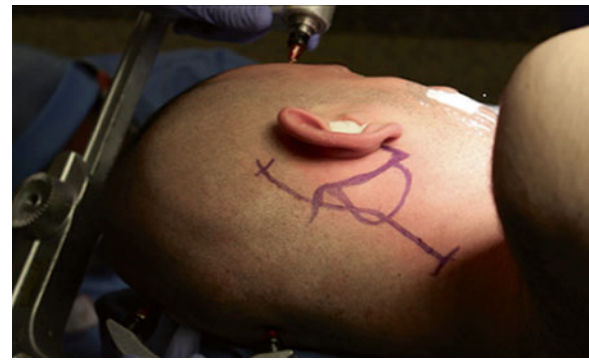

Fig. 11.4 Keyhole (retromastoid, retrosigmoid, infrafloccular) approach for vagal and glossopharyngeal nerve microvascular decompression (*right side*). Landmarks of *M* mastoid tip, *T* transverse, *S* sigmoid sinus, *I* skin incision, and *C* keyhole craniectomy

1.5 cm in width, just posterior to the sigmoid sinus. Goal is to expose the inferolateral aspect of the cerebellum and in the depth cistern of the IXth and Xth nerves. The burr hole must not be turned too laterally onto the sigmoid sinus as this could endanger the external wall of the sinus, which is often reduced to a thin endothelial layer adhesive to the bone. If bleeding occurs, suturing would not be possible owing to its friable texture. Use of a Doppler microprobe may help in detecting the posterior border of the sigmoid sinus. If mastoid cells are opened – which is common – they are occluded by affixing a piece of subcutaneous tissue, e.g., fat plus aponeurosis (fascia lata harvested from the thigh). The dura is opened by making a small flap retracted along the sigmoid sinus. A self-retaining retractor – of the Yasargil type – mounted with a very thin blade (Sugita-Fukushima type) is placed on the inferolateral aspect of the cerebellum. No vein is on the way, excepted sometimes a (tiny) inferior petrosal vein that can be coagulated and divided without consequence, at least in our experience.

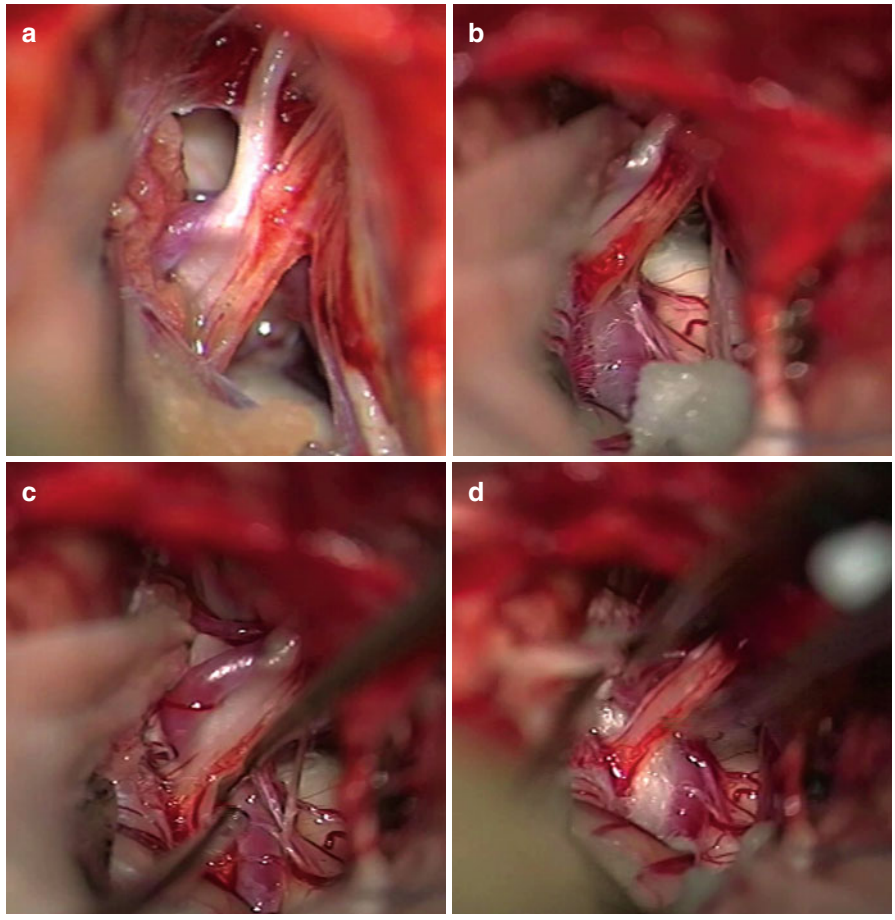

Fig. 11.5 Patient affected with a right vagoglossopharyngeal neuralgia due to a posterior inferior cerebellar artery (PICA); infra- and latero-floccular microsurgical approach on the right side. (**a**) Shows the offending vessel PICA ventral to the root entry zone of the IXth (*asterisk*) and the Xth (*triangle*) nerves, note the atrophic and grayish aspect of the IXth and Xth rootlets testifying of focal demyelination. (**b**) Shows the compressive PICA (*star*), ventro-caudally to the IXth and Xth rootlets. (**c**) Shows dissection and freeing of the rootlets from the PICA loop. (**d**) Shows Teflon felt bundle (*T*) maintaining artery apart from REZ of IXth and Xth rootlets

Then microscope is installed; and microsurgical steps start (Fig. 11.5). After incision of the arachnoid from XIth up to VIIIth nerve, the nerves are approached inferolaterally to the tonsil, to reach their root entry/exit zones (REZ) at the ventrolateral aspect of the medulla. REZ are often covered by the choroid plexus emerging from the lateral foramen of Luschka that must be gently retracted to expose REZ and vessels at the brainstem.

Not infrequently arachnoid is found thickened and strongly adhesive to the IXth–Xth rootlets. Dissection of rootlets needs to be meticulous, sometimes one by one, and as atraumatic as possible. Arachnoid chordae have to be divided using fine microscissors to free rootlets from vessels. Their sharp dissection should be completed before their mobilization.

The compressive vessels are the posterior inferior cerebellar artery (PICA), the vertebrobasilar artery (VBA), and frequently the association of both. Compression is most often ventral to the rootlets, which implies that maneuvers on the conflicting vessels be done passing in between rootlets at several interspaces. During mobilization of the compressive artery(ies), care should

be taken to respect their tiny perforating collaterals and not to generate vasospasm.

Throughout vascular manipulations, gentle irrigation with saline and, time to time, application of a few drops of papaverine in solution (1 ml in 10 ml saline) are important measures to limit the mechanical vasospastic reactions. However, not too much of papaverine should be used because of its very acid PH.

After the offending artery(ies) has(have) been dislodged from its(their) conflicting situation, often marked by an invagination into the ventrolateral surface of the medulla, the vessel(s) has(have) to be maintained apart in such a way not to go back to the previous compressive location.

In cases when arterial loops have a sufficient laxity, transposition is relatively easy. The loop is kept apart by means of sling(s), approximately 3–4 cm in length and 2–3 mm in width, made of shredded fibers of Teflon Felt, passed around the artery to exert a pulling effect, and blocked to avoid recurrence of malposition. Conversely, in cases where the compressive vessel is rigid, transposition is usually rather difficult. The vessel cannot simply be pulled away. A small piece of semirigid prosthesis (Dacron or Teflon) and/or a small balled cushion of Teflon fibers is interposed between the REZ of the brainstem and the artery. Care must be taken not to exert any "neurocompressive effect".

Before the microscope is taken out, the surgeon must verify that arteries have no kinking or twisting. Irrigation of the vessels with warm saline and a few droplets of papaverine in solution is a wise precaution to suppress possible spasms due to surgical manipulations. Venous hemostasis is checked by asking the anesthesiologist to perform sustained digital compression at the neck of both jugular veins or, if this is not possible, by carrying out a Valsalva maneuver with the ventilation machine.

Then the dura is closed either with single stitches or to better achieve watertight closure with a small patch of fascia lata. Additional fatty tissue is affixed onto the mastoid cells if opened. Bone chips are packed over the craniectomy defect only if no mastoid cells were opened. Finally, the muscular, subcutaneous, and cutaneous layers are closed with interrupted sutures and a compressive dressing is applied to avoid pseudomeningocele and decrease the risk of cerebrospinal fluid fistula or of rhinorrhea through the Eustachian tube if mastoid cells were opened.

11.4.2.2 Results of the Authors' Series

In our series of 36 MVD for VGPN, there was complete pain relief in the long-term follow-up, i.e., with more than 2 years of follow-up, in more than 94.4 % of the cases. There was no mortality, no general morbidity, and no wound complication. As regards to neurological outcome, there was no postoperative permanent deficits, except two cases with a disabling IXth–Xth sensory and motor deficits.

11.4.3 Treatment with Lesioning Techniques

Lesioning techniques for treating idiopathic VGPN should not be first option. A summary of them is given in the following section.

11.4.3.1 Intracranial Rhizotomy

The first attempted procedures for treating VGPN were the extracranial section of the glossopharyngeal nerve (Sicard and Robineau 1920), soon after the avulsion of the nerve at the jugular foramen via a high cervical approach (Adson 1924). On account of a high morbidity and a high incidence of recurrence of pain, these methods were not long propagated. It was Dandy in 1927 who gave the explanation of the low efficacy of the extracranial procedure and popularized the intracranial section of the glossopharyngeal nerve (Dandy 1927). Table 11.2 includes the series of patients who underwent rhizotomy of the IXth and/or Xth cranial nerves.

11.4.3.2 Percutaneous RF Thermocoagulation

The first percutaneous radiofrequency (RF) thermocoagulation procedures for VGPN were reported in 1974 (Lazorthes and Verdie 1979).

Table 11.2 Literature series with intracranial rhizotomy of IXth and/or Xth cranial nerves, from 1931 to 2014

Authors and Year	No. of patients	Mean age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complication (%)
Jefferson (1931)	1	34	0	1.25	1	0	0	
Keith (1932)	1	46	0	10 days	1	0	0	
Reichert (1933)	1	31	0	0.33	1	0	0	
Lillie and Craig (1936)	1	15	0	6 weeks	1	0	0	
Cuneo (1943)	1	33	0	NR	1	0	0	
Svien et al. (1957)	1	37	0	0.5	1	0	0	0
Bohm and Strang (1962)	4	48	0	5	2	2	0	
Laha and Jannetta (1977)	3	59.3	0	3	3 (100)			0
Jannetta (1980)	2	30-69	0	NR	2 (100)			Decreased palatal and gag reflex 2 (100 %)
Rushton et al. (1981)	129	NR	7	NR	110 (85.3)		13 (10)	
King (1987)	2	29	0	1.6	1	1	0	
Fraioli et al. (1989)	3	62.3	0	NR	3 (100)			
Sindou et al. (1991)	3	66	0	3.5	3	0	0	0
Ferrante et al. (1995)	2	63.5	0	6.5	2	0	0	
Taha and Tew (1995)	12	42.7	0	10	12 (100)			T. dysphagia 2 (16.7) T. hoarseness 1 (8.3) P. dysphagia 1 (8.3) T. cough 3 (25)
Ceylan et al. (1997)	2	55.5	0	2	2 (100)			Diminished gag reflux 2 (100)
Ozenci et al. (2003)	1	51	0	NR	1	0	0	0
Lou et al. (2008)	12	NR	0	15 months	11	1	0	
Kandan et al. (2010)	6	52.5	0	4	5 (83.3)	1 (16.7)		
Ma et al. (2010)	3	61.5	0	2	2	1	0	0
Martínez-González et al. (2011)	3	60	0	NR	3	0	0	
Zhang et al. (2014)	8	48.2	0	9–39 months	7	1	0	0

NR not reported, *T* transient, *P* permanent

Compared to the procedure for trigeminal neuralgia, it is far more difficult to perform it on the IXth–Xth sensory nerves due to their location and the high risk of lesioning the adjacent vessels, namely, the jugular vein and the internal carotid artery. Because of the high incidence of side effects, such as a diminished gag reflex, dysphagia, and vocal cord paralysis, Tew recommended that this procedure be reserved for patients whose condition is secondary to cancer of the oropharynx (Taha and Tew 1995; Tew et al. 1982). Table 11.3 includes the series of patients who underwent percutaneous RF thermocoagulation of the ninth and/or tenth cranial nerves. Whereas craniotomy with posterior fossa exploration had a significant mortality rate in some series, reported mortality with percutaneous RF rhizotomy was none. However, complications were frequently noticed including cardiovascular disturbances occurring during the procedure (Ori et al. 1983).

11.4.3.3 Trigeminal Tractotomy-Nucleotomy

Trigeminal tractotomy, defined by the section of the descending trigeminal tractus in the medulla via posterior fossa craniectomy, was first described by Sjöqvist in 1938. Kunc applied this method to treat glossopharyngeal neuralgia in six patients in 1954 (Kunc 1965); for doing so, he used a simple light mechanical stimulation with a thin needle to localize the target fibers and called it "selective tractotomy." Later on the method was revisited by others with technical modifications (Crue et al. 1972; Hitchock and Schvarcz 1972; Nashold et al. 1992). More recently Kanpolat et al. developed the CT-guided trigeminal tractotomy-nucleotomy which has more accuracy and safety; this method is mainly applied to treat intractable facial pain and malignant pain (Kanpolat et al. 1998, 2008). Literature data are summarized in Table 11.4.

Table 11.3 Literature series of percutaneous RF thermocoagulation of IXth or Xth from 1965 to 1991

Authors and Year	N. of patients	Mean Age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complications (%)
Lazorthes and Verdie (1979)	1	45	0	NR	1 (100)			
Isamat et al. (1981)	3	57	0	1.6	3 (100)	0		0
Salar et al. (1983)	1	NR	0	0.6	1 (100)			
Oris (Ori et al. 1983)	1	34	0	NR	1 (100)			
Giorgi and Broggi (1984)	9	54.7	0	NR	6 (67)	3 (33)		P. dysphagia 1 (11.1 %)
Arias (1986)	2	60.5	0	1.5	2 (100)			0
Sindou et al. (1991)	3	66	0	3.5	2 (67)	1 (33)		0

NR not reported, *P* permanent

Table 11.4 Literature series with trigeminal tractotomy-nucleotomy from 1961 to 1998

Authors and Year	N. of patients	Mean age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complications (%)
Bues (1961)	7	NR	0	NR	7 (100)			
Kunc (1965)	6	65	0	NR	6 (100)			0
Kanpolat et al. (1998)	6	50.5	0	49.5 m	6 (100)			0

NR not reported

Table 11.5 Literature series with SRS of IXth and/or Xth cranial nerves, from 2005 to 2013

Authors and Year	N. of patients	Mean age (years)	Deaths (%)	Mean follow-up (years)	Total relief (%)	Partial relief (%)	No relief (%)	Complications (%)
Stieber et al. (2005)	1	NR	0	6 months	1 (100)			0
Yomo et al. (2009)	2	45	0	NR	1 (50)	1 (50)		0
Williams et al. (2010)	1	47	0	11 month	1 (100)			0
Pollock and Boes (2011)	5	61	0	NR	3 (60)	0	2 (60)	0
Lévêque et al. (2011)	7	62	0	1.5	5 (71)	2 (29)		0
O'Connor and Bidiwala (2013)	1	99	0	1.3	1 (100)			0

NR not reported

11.4.3.4 Stereotactic Radio Surgery

Stereotactic radio surgery (SRS) was intended to provide a solution for those who cannot tolerate or who refuse open intervention. Dhople AA in 2009 reported a study which owns one of the longest median follow-up periods (Dhople et al. 2009); the lesion maker was Gamma Knife. Targets can be REZ, cistern segment, or pars nervosa of the jugular foramen (i.e., vagoglossopharyngeal meatus) (Lévêque et al. 2011), with a dose ranging from 60 to 80 Gy (Pollock and Boes 2011). Literature data are summarized in Table 11.5.

Although rate of pain relief after 1-year of follow-up was lower with SRS (64 %) than with MVD (84 %), SRS may be considered a potential viable option.

11.5 Discussion and Conclusions

In spite of important disparities between reports collected for the present literature review on neurosurgical procedures for treating VGPN, and lack of precisions for some of the publications, the following may be concluded.

Rhizotomies, whatever their type, open or percutaneous, basically produce sensory deficits responsible for disabling dysphagia and hoarseness; rhizotomies should not be the first option. Tractotomies-nucleotomies, due to their potential risks linked to brainstem location, should remain exceptionally indicated for essential VGPN. SRS, recently introduced for VGPN, needs more long-term studies to confirm efficacy and innocuity.

Because of its curative and conservative nature, MVD should be proposed as the first option, when high-resolution MR imaging evidences clear-cut neurovascular conflict at the IXth–Xth cranial nerve REZ. Being a delicate surgery, MVD should be performed within the frame of a well-experienced neurosurgical team.

References

Adson A. The surgical treatment of glossopharyngeal neuralgia. Arch Neurol Psychiatry. 1924;12:487–506.

Arias MJ. Percutaneous radio-frequency thermocoagulation with low temperature in the treatment of essential glossopharyngeal neuralgia. Surg Neurol. 1986;25:94–6.

Bohm E, Strang RR. Glossopharyngeal neuralgia. Brain. 1962;85:371–88.

Bues E (1961) Long-term results after tractotomy for glossopharyngeal neuralgia. 2nd International. Congress. Neurology. Surgery, vol. 36. Excerpta Medica International Congress Series. N Washington, DC. p. E60–1.

Ceylan S, Karakus A, Duru S, Baykal S, Koca O. Glossopharyngeal neuralgia: a study of 6 cases. Neurosurg Rev. 1997;20:196–200.

Chawla JC, Falconer MA. Glossopharyngeal and vagal neuralgia. Br Med J. 1967;3:529–31.

Chen J, Sindou M. Vago-glossopharyngeal neuralgia: a literature review of neurosurgical experience. Acta Neurochir. 2015;157:311–21.

Crue BL, Carregal JA, Felsoory A. Percutaneous stereotactic radiofrequency. Trigeminal tractotomy with neurophysiological recordings. Confin Neurol. 1972;34:389–97.

Cuneo HM. Glossopharyngeal neuralgia. Bull Los Angeles Neurol Soc. 1943;8:143–5.
Dandy WE. Glossopharyngeal neuralgia (tic douloureux): its diagnosis and treatment. Arch Surg. 1927;15:198–214.
Dandy WE. Concerning the cause of trigeminal neuralgia. Am J Surg. 1934;24:447–55.
Dhople AA, Adams JR, Maggio WW, Naqvi SA, Regine WF, Kwok Y. Long-term outcomes of Gamma Knife radiosurgery for classic trigeminal neuralgia: implications of treatment and critical review of the literature. J Neurosurg. 2009;111:351–8.
Esaki T, Osada H, Nakao Y, Yamamoto T, Maeda M, Miyazaki T, Sumiyoshi M, Mori K. Surgical management for glossopharyngeal neuralgia associated with cardiac syncope: two case reports. Br J Neurosurg. 2007;21:599–602.
Ferrante L, Artico M, Nardacci B, Fraioli B, Cosentino F, Fortuna A. Glossopharyngeal neuralgia with cardiac syncope. Neurosurgery. 1995;36:58–63. discussion 63.
Ferroli P, Fioravanti A, Schiariti M, Tringali G, Franzini A, Calbucci F, Broggi G. Microvascular decompression for glossopharyngeal neuralgia: a long-term retrospectic review of the Milan-Bologna experience in 31 consecutive cases. Acta Neurochir. 2009;151:1245–50.
Fraioli B, Esposito V, Ferrante L, Trubiani L, Lunardi P. Microsurgical treatment of glossopharyngeal neuralgia: case reports. Neurosurgery. 1989;25:630–2.
Gardner WJ. The mechanism of tic douloureux. Trans Am Neurol Assoc. 1953;3:168–71; discussion, 171–163.
Gaul C, Hastreiter P, Duncker A, Naraghi R. Diagnosis and neurosurgical treatment of glossopharyngeal neuralgia: clinical findings and 3-D visualization of neurovascular compression in 19 consecutive patients. J Headache Pain. 2011;12:527–34.
Giorgi C, Broggi G. Surgical treatment of glossopharyngeal neuralgia and pain from cancer of the nasopharynx. A 20-year experience. J Neurosurg. 1984;61:952–5.
Greeson SD, Linden CH. Glossopharyngeal neuralgia. Ann Emerg Med. 1981;10:656–8.
Hamada O, Ogata T, Abe H, Okawa M, Higashi T, Matsumoto J, Takano K, Inoue T. A patient with vertebral artery dissection who initially suffered from pharyngeal pain. No Shinkei Geka. 2013;41:1081–5.
Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia. 2013;33:629–808.
Hitchock ER, Schvarcz JR. Stereotaxic trigeminal tractotomy for post-herpetic facial pain. J Neurosurg. 1972;37:412–7.
Isamat F, Ferran E, Acebes JJ. Selective percutaneous thermocoagulation rhizotomy in essential glossopharyngeal neuralgia. J Neurosurg. 1981;55:575–80.
Jamshidi A, Masroor MA. Glossopharyngeal neuralgia with cardiac syncope: treatment with a permanent cardiac pacemaker and carbamazepine. Arch Intern Med. 1976;136:843–5.
Jannetta PJ. Observations on the etiology of trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction and glossopharyngeal neuralgia. Definitive microsurgical treatment and results in 117 patients. Neurochirurgia (Stuttg). 1977;20:145–54.
Jannetta PJ. Neurovascular compression in cranial nerve and systemic disease. Ann Surg. 1980;192:518–25.
Jefferson G. Glosso-pharyngeal neuralgia. Lancet. 1931;221:397–9.
Kandan SR, Khan S, Jeyaretna DS, Lhatoo S, Patel NK, Coakham HB. Neuralgia of the glossopharyngeal and vagal nerves: long-term outcome following surgical treatment and literature review. Br J Neurosurg. 2010; 24:441–6.
Kanpolat Y, Savas A, Batay F, Sinav A. Computed tomography-guided trigeminal tractotomy-nucleotomy in the management of vagoglossopharyngeal and geniculate neuralgias. Neurosurgery. 1998;43:484–9; discussion 490.
Kanpolat Y, Kahilogullari G, Ugur HC, Elhan AH. Computed tomography-guided percutaneous trigeminal tractotomy-nucleotomy. Neurosurgery. 2008;63:ONS147–53; discussion ONS153-145.
Kawashima M, Matsushima T, Inoue T, Mineta T, Masuoka J, Hirakawa N. Microvascular decompression for glossopharyngeal neuralgia through the transcondylar fossa (supracondylar transjugular tubercle) approach. Neurosurgery. 2010;66:275–80; discussion 280.
Keith WS. Glossopharyngeal neuralgia. Brain. 1932;55:357–66.
King J. Glossopharyngeal neuralgia. Clin Exp Neurol. 1987;24:113–21.
Kondo A. Follow-up results of using microvascular decompression for treatment of glossopharyngeal neuralgia. J Neurosurg. 1998;88:221–5.
Kunc Z. Treatment of essential neuralgia of the 9th nerve by selective tractotomy. J Neurosurg. 1965;23: 494–500.
Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. J Neurosurg. 1977;47:316–20.
Lazorthes Y, Verdie JC. Radiofrequency coagulation of the petrous ganglion in glossopharyngeal neuralgia. Neurosurgery. 1979;4:512–6.
Leal PR, Hermier M, Froment JC, Souza MA, Cristino-Filho G, Sindou M. Preoperative demonstration of the neurovascular compression characteristics with special emphasis on the degree of compression, using high-resolution magnetic resonance imaging: a prospective study, with comparison to surgical findings, in 100 consecutive patients who underwent microvascular decompression for trigeminal neuralgia. Acta Neurochir. 2010;152:817–25.
Lévêque M, Park MC, Melhaoul A, Yomo S, Donnet A, Regis J. Gamma Knife radiosurgery for glossopharyngeal neuralgia, Marseille: experience. J Radiosurg SBRT. 2011;1:41–6.
Lillie HI, Craig WM. Anomalous vascular lesion in cerebellopontine angle. Severe neuralgic pain in the ear and profound nervous disturbance; operation and recovery. Arch Otolaryngol. 1936;23:642–5.
Lou WH, Lu WH, Sun HR. [Glossopharyngeal neurotomy through laterocervical approach to treat idiopathic glossopharyngeal neuralgia]. Zhonghua Er Bi

Yan Hou Tou Jing Wai Ke Za Zhi. 2008;43: 818–21.

Ma Z, Li M, Cao Y, Chen X. Keyhole microsurgery for trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Eur Arch Otorhinolaryngol. 2010;267:449–54.

Martinez-Gonzalez JM, Martinez-Rodriguez N, Calvo-Guirado JL, Brinkmann JC, Dorado CB. Glossopharyngeal neuralgia: a presentation of 14 cases. J Oral Maxillofac Surg. 2011;69:e38–41.

Matsushima T, Goto Y, Natori Y, Matsukado K, Fukui M. Surgical treatment of glossopharyngeal neuralgia as vascular compression syndrome via transcondylar fossa (supracondylar transjugular tubercle) approach. Acta Neurochir. 2000;142:1359–63.

Michelucci R, Tassinari CA, Samoggia G, Tognetti F, Calbucci F. Intracranial microvascular decompression for "cryptogenic" hemifacial spasm, trigeminal and glossopharyngeal neuralgia, paroxysmal vertigo and tinnitus: II. Clinical study and long-term follow up. Ital J Neurol Sci. 1986;7:367–74.

Munch TN, Rochat P, Astrup J. Vagoglossopharyngeal neuralgia treated with vascular decompression. Ugeskr Laeger. 2009;171:2654–5.

Murasawa A, Yamada K, Hayakawa T, Aragaki Y, Yoshimine T. Glossopharyngeal neuralgia treated by microvascular decompression--case report. Neurol Med Chir. 1985;25:551–3.

Nashold Jr BS, El-Naggar A, Mawaffak Abdulhak M, Ovelmen-Levitt J, Cosman E. Trigeminal nucleus caudalis dorsal root entry zone: a new surgical approach. Stereotact Funct Neurosurg. 1992;59:45–51.

Nishikawa Y, Suzuki M, Kuwata N, Ogawa A. Microvascular decompression for treating glossopharyngeal neuralgia complicated by sick sinus syndrome. Acta Neurochir. 2000;142:351–2.

O'Connor JK, Bidiwala S. Effectiveness and safety of Gamma Knife radiosurgery for glossopharyngeal neuralgia. Proc (Bayl Univ Med Cent). 2013;26:262–4.

Ohyama S, Oki S, Sumida M, Isobe N, Kureshima M, Kurokawa Y. [Microvascular decompression for glossopharyngeal neuralgia: case report]. No Shinkei Geka. 2006;34:169–73.

Ori C, Salar G, Giron G. Percutaneous glossopharyngeal thermocoagulation complicated by syncope and seizures. Neurosurgery. 1983;13:427–9.

Ozenci M, Karaoguz R, Conkbayir C, Altin T, Kanpolat Y. Glossopharyngeal neuralgia with cardiac syncope treated by glossopharyngeal rhizotomy and microvascular decompression. Europace. 2003;5:149–52.

Patel A, Kassam A, Horowitz M, Chang YF. Microvascular decompression in the management of glossopharyngeal neuralgia: analysis of 217 cases. Neurosurgery. 2002;50:705–10; discussion 710–701.

Platania N, Nicoletti GF, Barbagallo G, Albanese V. Concurrent trigeminal and glossopharyngeal neuralgia, hemifacial spasm and hypertension by neurovascular compression. Case report. J Neurosurg Sci. 1997;41:303–7.

Pollock BE, Boes CJ. Stereotactic radiosurgery for glossopharyngeal neuralgia: preliminary report of 5 cases. J Neurosurg. 2011;115:936–9.

Reichert FL. True tic douloureux of the ear or so-called geniculate ganglion neuralgia: cure effected by intracranial section of the glossopharyngeal nerve. JAMA. 1933;100:1744–6.

Resnick DK, Jannetta PJ, Bissonnette D, Jho HD, Lanzino G. Microvascular decompression for glossopharyngeal neuralgia. Neurosurgery. 1995;36:64–8; discussion 68–69.

Rushton JG, Stevens JC, Miller RH. Glossopharyngeal (vagoglossopharyngeal) neuralgia: a study of 217 cases. Arch Neurol. 1981;38:201–5.

Salar G, Ori C, Baratto V, Iob I, Mingrino S. Selective percutaneous thermolesions of the ninth cranial nerve by lateral cervical approach: report of eight cases. Surg Neurol. 1983;20:276–9.

Sampson JH, Grossi PM, Asaoka K, Fukushima T. Microvascular decompression for glossopharyngeal neuralgia: long-term effectiveness and complication avoidance. Neurosurgery. 2004;54:884–9; discussion 889–890.

Sicard R, Robineau J. Communications et presentations: algie vélo-pharyngée essentielle. Traitement chirururgical. Rev Neurol. 1920;36:256–7.

Sindou M, Keravel Y. Neurosurgical treatment of vagoglossopharyngeal neuralgia. Neurochirurgie. 2009;55:231–5.

Sindou M, Henry JF, Blanchard P. Idiopathic neuralgia of the glossopharyngeal nerve. Study of a series of 14 cases and review of the literature. Neurochirurgie. 1991;37:18–25.

Sindou M, Keravel Y, Laurent B. Aspects cliniques et thérapeutiques des névralgies essentielles du trijumeau et du glossopharyngien. EMC-Neurologie. 2014;11:1–21.

Sjöqvist O. Studies on pain conduction in the trigeminal nerve. A contribution to the surgical treatment of facial pain. Acta Psychiatr Neurol Scand Suppl. 1938;17:1–139.

Stieber VW, Bourland JD, Ellis TL. Glossopharyngeal neuralgia treated with gamma knife surgery: treatment outcome and failure analysis. Case report. J Neurosurg. 2005;102(Suppl):155–7.

Svien HJ, Hill NC, Daly DD. Partial glossopharyngeal neuralgia associated with syncope. J Neurosurg. 1957;14:452–7.

Taha JM, Tew Jr JM. Long-term results of surgical treatment of idiopathic neuralgias of the glossopharyngeal and vagal nerves. Neurosurgery. 1995;36:926–30. discussion 930–921.

Tew JMJ, Tobler WD, Van Loveren H. Percutaneous rhizotomy in the treatment of intractable facial pain (trigeminal, glossopharyngeal, and vagal nerves). In: Schmidek HH, Sweet WH, editors. Operative neurosurgical techniques, vol. 2. Philadelphia: W.B. Saunders; 1982. p. 1083–106.

Tsuboi M, Suzuki K, Nagao S, Nishimoto A. Glossopharyngeal neuralgia with cardiac syncope. A case successfully treated by microvascular decompression. Surg Neuro. 1985;24:279–83.

Wakiya K, Fukushima T, Miyazaki S. Results of microvascular decompression in 16 cases of glossopharyngeal neuralgia. Neurol Med Chir. 1989;29: 1113–8.

Wang YN, Zhong J, Zhu J, Dou NN, Xia L, Visocchi M, Li ST. Microvascular decompression in patients with coexistent trigeminal neuralgia, hemifacial spasm and glossopharyngeal neuralgia. Acta Neurochir. 2014;156:1167–71.

White J, Sweet WH. Pain and the neurosurgeon, a forty-year experience. Springfield: C.C. Thomas; 1969.

Williams BJ, Schlesinger D, Sheehan J. Glossopharyngeal neuralgia treated with gamma knife radiosurgery. World Neurosurg. 2010;73:413–7.

Xiong NX, Zhao HY, Zhang FC, Liu RE. Vagoglossopharyngeal neuralgia treated by microvascular decompression and glossopharyngeal rhizotomy: clinical results of 21 cases. Stereotact Funct Neurosurg. 2012;90:45–50.

Yomo S, Arkha Y, Donnet A, Regis J. Gamma Knife surgery for glossopharyngeal neuralgia. J Neurosurg. 2009;110:559–63.

Yoshioka J, Ueta K, Ohmoto T, Fujiwara T, Tabuchi K. Combined trigeminal and glossopharyngeal neuralgia. Surg Neurol. 1985;24:416–20.

Zhang W, Chen M, Zhang W, Chai Y. Use of electrophysiological monitoring in selective rhizotomy treating glossopharyngeal neuralgia. J Craniomaxillofac Surg. 2014;42:e182–5.

Intraoperative Monitoring

12

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Abstract

This chapter provides an overview of the tests that constitute the tools in intraoperative neurophysiology of cranial nerves. The general introduction and equipment of monitoring are described. This chapter gives a practical presentation of each neurophysiological method regarding the following: (1) the principle on which it is based, (2) the methodology for stimulation and recording, (3) the intraoperative interpretation, and (4) the various types of anesthetics and their effects on neurophysiological monitoring. At the end of the chapter, a detailed example of the combined use of intraoperative monitoring for MVD surgery is discussed.

Keywords

Intraoperative monitoring • Cranial nerve • Evoked potentials • Electromyography • ZLR • F wave • Blink reflexes • Anesthetics

12.1 Introduction

With the rapid developments in neuroscience, intraoperative neurophysiological monitoring (IOM or IONM), as a new and exciting field, has grown rapidly over the past two decades. IOM is gradually becoming part of standard medical practice, for it can provide information regarding the functional integrity of the nervous system of a patient who is anesthetized and therefore cannot be neurologically examined. Common IOM techniques include electroencephalography (EEG), electromyography (EMG), evoked potentials (EPs), and nerve conduction velocity (NCV).

Cranial nerves are at risk of being injured during various kinds of neurosurgical operations. By offering early detection of reversible neurophysiological dysfunction during surgery, neuro-monitoring can provide prognostic information about clinical outcomes and prevent the occurrence of permanent neurological damage. Neurophysiological methods are increasingly used for diagnostic support in operations such as those involving peripheral nerves. In certain operations, intraoperative neurophysiology can

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increase the likelihood of achieving the therapeutic goal of an operation. Intraoperative neurophysiological recordings have shown to be of help in identifying the offending blood vessel in a cranial nerve disorder (hemifacial spasm, HFS) (Moller and Jannetta 1986).

12.2 Equipment and Electrodes

The choice of equipment for intraoperative monitoring is very important. The equipment should have several desirable features which, although not absolutely necessary for routine clinical recordings, are of special importance for intraoperative recordings. For instance, it should allow for simultaneous multimodality recordings, such as EMG and evoked responses, to meet the needs of specific operations. A typical system includes one or more programmable devices that can deliver auditory, visual, and electrical stimuli of variable amplitude, duration, and rate; various types of electrodes for delivering electrical stimuli and for recording electrophysiological activity from the scalp, nerves, and muscles; a head box for selecting among different electrode groups to be connected to the amplifiers; a set of amplifiers; a display monitor; a printer; and often a modem or network connection for remote monitoring (Zouridakis and Papanicolaou 2000). All parts are connected to a computer which, depending on the kind of recordings, controls stimulus delivery, data collection, filtering, averaging, display, printing, as well as remote transmission and permanent storage of the data (Zouridakis and Papanicolaou 2000). However, it should allow modifications in the recording protocol and display parameters, if necessary, thus permitting fast interpretation of the results.

There are several types of stimulation electrodes for both stimulation and recording, including stick-on electrodes, metal cup electrode, subdermal needles, corkscrew electrode, and metal probes, which can be monopolar or bipolar. The efficiency of stimulus delivery or recording is determined by selection of the appropriate electrode type and its correct placement.

The subdermal needles are more practical in the operating room as they can be applied quickly after the patient has been anesthetized without the need for skin preparation. They are usually easy to secure and provide stable and steady low electrode-skin impedances. If possible, the addition of more "redundant" electrodes at both recording and ground sites can be useful in case electrodes get dislodged during surgery.

12.3 The Effects of Anesthetics on Intraoperative Neurophysiology Studies

Various types of anesthetics were found to affect neurophysiology studies used in intraoperative monitoring because of the effects they have on cerebral blood flow, perfusion, and metabolic rate. General anesthesia consists of several components, including analgesia (suppression of response to pain), sedation (induction of sleep), amnesia (suppression of recollection of the intraoperative experience), and muscle relaxation (suppression of muscle contraction). It may also include hypotension (decreased blood pressure) and hypothermia (decreased body temperature) which might also affect the neurophysiological signals (Zouridakis and Papanicolaou 2000). Thus, interpretation of all neurophysiological recordings should always take into account the effects of changes in anesthesia regime which are very similar to the changes from surgical intervention. Pulse oximetry, ECG, arterial blood pressure, capnography, body temperature, and muscle twitch response should be routinely monitored and recorded during the surgeries. Communication with the anesthesiologist is very critical. Successful neurophysiological intraoperative monitoring requires a team approach between the anesthesiology, surgical, and NIOM teams (Galloway et al. 2010). A detailed discussion of the various types of anesthetics and their effects on neurophysiological monitoring will be described at the end of each method of monitoring.

12.4 Monitoring Techniques and Exclusionary Criteria

12.4.1 Brain Stem Auditory Evoked Potentials

Hearing loss (HL) is one of the most common complications after microvascular decompression (MVD) (Møller and Møller 1989; Samii et al. 2002).

Brain stem auditory evoked potentials (BAEPs) monitoring, also named auditory brain stem responses (ABRs), has been widely used as a classical and noninvasive technique to reduce the risk of hearing impairment with surgery in the region of the cerebellopontine angle (CPA), including MVD (López 2004). The normal BAEPs consist of 5–7 vertex-negative peaks that occur between 2 and 10 ms after the presentation of a high-intensity transient sound such as a click or a short, high-intensity tone burst (Fig. 12.1). Specific anatomic locations have been proposed as the predominant source for the generation of each of these waveforms: wave I, distal auditory nerve; wave II, cochlear nucleus and proximal auditory nerve; wave III, superior olivary nucleus (lower pons); wave IV, lateral lemniscus; wave V, inferior colliculus; wave VI, medial geniculate body; and wave VII, auditory radiations. In addition to reflecting the conduction in the auditory pathways, BAEPs also provide valuable information about the general function of the brain stem.

The cochlear nerve may be damaged for the following reasons during the surgery: (1) stretching of the cochlear nerve while retracting the cerebellum, (2) manipulation of the labyrinthine artery and/or the anteroinferior cerebellar artery, (3) direct trauma by instruments or a nearby coagulation, and (4) new compression of the cochlear nerve at end of surgery by the interposed Teflon between the compressive vessel and the VIIth-cochlear nerve complex (Polo et al. 2004; Sindou 2005). During BAEP monitoring, careful assessment of the changes in the pattern of BAEPs and analysis of their relationship to the surgical maneuvers applied should determine whether the changes reflect damage to or potentially reversible dysfunction of the auditory pathways in the ear, auditory nerve, or brain stem. Thus, this test can be used in surgical procedures

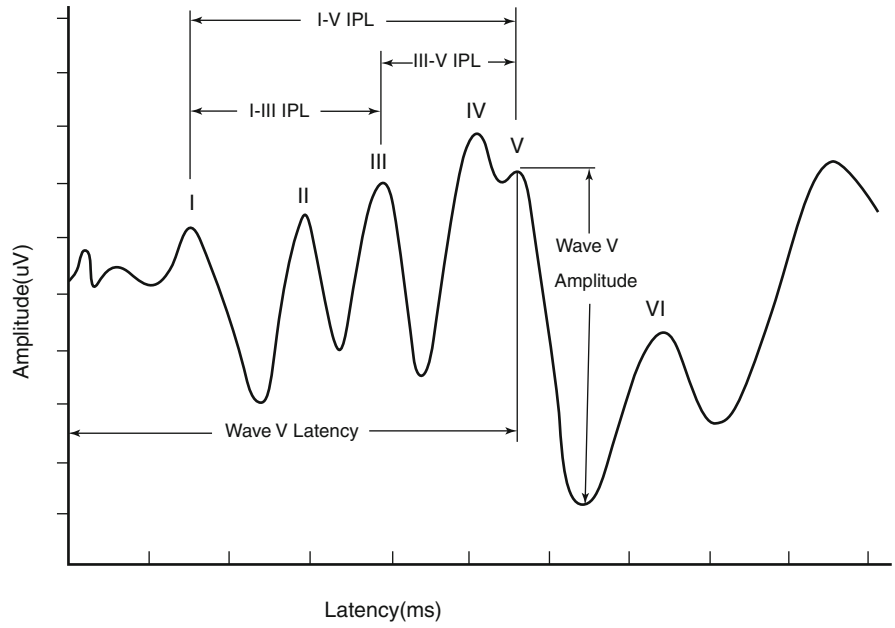

Fig 12.1 Typical recording of brain stem auditory evoked potentials

such as microvascular decompression of the facial nerve or trigeminal nerve or tumor removal at CPA.

12.4.1.1 Stimulation and Recording

Stimulation Sound stimulus is delivered through miniature electromagnetic earphones inserted in the ear canals and secured though sponge (molded foam) ear inserts connected to a transducer by plastic tubing. Dislodgment or kinking of this tubing can lead to complete absence of all BAEP components or reduction in amplitudes and delay in latencies.

Sound Intensity Clicks of alternating polarity are commonly used for intraoperative monitoring to cancel the stimulus artifact. Usually, the stimulus intensity needed is at or above 70 dB nHL, but higher intensities are often needed if there is a preexisting hearing loss.

Stimulus Rate To eliminate contamination with electrical artifacts, exactly 10 Hz or multiples of the used power frequency should be avoided. Usually, a rate between 20 and 40 Hz (e.g., 33.1 Hz) is used, and the selected rate should be used consistently for the rest of the surgery. In order to eliminate the electrical noises in the operating room, several hundreds of averaged trials are often necessary to obtain reliable signals.

Recording Electrodes and Band Pass Electrode placement follows the EEG 10–20 international system. The standard setup of recording electrodes should positioned as follows: Channel (1) vertex to ipsilateral earlobe/mastoid (Cz-Ai/Mi) and Channel (2) vertex to contralateral earlobe/mastoid (Cz-Ac/Mc). Two additional channels could also be used: Channel (3) ipsilateral earlobe/mastoid to contralateral earlobe/mastoid (Ai-Ac or Mi-Mc) and Channel (4) vertex to noncephalic reference (Cz-Nc), e.g., vertex to cervical (Cz-Cv2). Typical filter settings are a low-frequency filter of 100 Hz and a high-frequency filter of 3 kHz. As BAEPs are of very short latencies and small amplitudes, time base of 1 ms (sweep of 10 ms) and sensitivity of 0.1–0.2 uV/division are used.

12.4.1.2 Intraoperative Interpretation

A set of baselines should be obtained after anesthesia induction and patient positioning. Baseline responses should contain clear and reliable components and also be compared with throughout the surgery using the same acquisition and stimulus parameters.

As with the 2006 criteria published by the American Clinical Neurophysiology Society for recording standard BAEPs, analysis typically involves monitoring for the presence of waves I, III, and V. The measurements must include the following: (1) wave I peak latency, (2) wave III peak latency, (3) wave V peak latency, (4) I–III interpeak interval, (5) III–V interpeak interval, (6) I–V interpeak interval, (7) wave I amplitude, (8) wave V amplitude, and (9) wave IV–V/I amplitude ratio (Society 2006).

Traditionally, the most important criterion involves the latency and the amplitude of peak V. Some authors have suggested that a latency prolongation of as little as 0.5 ms of the wave V is significant (Acevedo et al. 1997). It has been found that a reduction of amplitude more than 50 % in wave V was a stronger indicator of hearing than latency (Hatayama and Mollar 1998; Legatt 2002; Jo et al. 2011). However, Polo et al. (2004) reported that the I–V interpeak latency, or the latency of wave V, was an effective and predictive indicator of postoperative hearing, whereas others have suggested that hearing loss occurs when wave V is completely lost (Schlake et al. 2001; Lee et al. 2009). It has been also proposed that patients with HL had higher rates of loss in the amplitude of wave V and prolongation in the interpeak latency of peaks I–V during MVD (Ying et al. 2014). The criteria for significant intraoperative BAEP change are still not universally accepted. It has been suggested that no single value can be used to either predict when hearing will be preserved or lost, and a sliding scale approach should be used (Polo et al. 2004).

12.4.1.3 Anesthesia Requirements

The effects of various drugs most commonly used in anesthesia on the BAEPs are summarized and listed in Table 12.1.

Table 12.1 Effects of anesthetic agents on BAEP amplitude and latency

Agent	Amplitude	Latency
Nitrous oxide (N_2O)	↓	–
Inhalational anesthetics	–	↑
Propofol	–	↑
Barbiturates	–	↑
Ketamine	–	↑
Opiates	–	–
Benzodiazepines	–	–
Muscle relaxants	–	–

Hypothermia increases the latency and decreases the BAEP amplitude, whereas hyperthermia decreases the amplitude and the latency of the responses (Markand et al. 1987)

12.4.2 Brain Stem Trigeminal Evoked Potentials

The trigeminal somatosensory system, which provides sensation to the face and the anterior two-thirds of the tongue, can be monitored using standard SSEPs. It has been shown that monitoring of SSEP is useful in intraoperative monitoring of the medulla oblongata and in trigeminal rhizotomy in patients with trigeminal neuralgia in whom it may be of value to monitor neural conduction in the trigeminal nerve (Stechison and Kralick 1993). Recordings from electrodes on the scalp can be used for monitoring the ascending trigeminal sensory pathways when elicited by electrical stimulation of the peripheral trigeminal nerve.

12.4.2.1 Stimulation and Recording

Stimulation Sites Stimulation of the trigeminal nerve is performed with needle electrodes introduced subcutaneously into the upper (anode) and lower (cathode) lips (Malcharek et al. 2011).

Stimulation Intensity and Rate Each set of the BTEP was stimulated with 300 trials to confirm the repeatability of the obtained cortical response. The frequency of the stimulation was 4.7 Hz, and the duration of each stimulus was 0.2 ms. The polarity of the stimulation was alternating to avoid large baseline shifts. The intensity of stimulation during each set of trials varied from 7 to 16 mA (Malcharek et al. 2011).

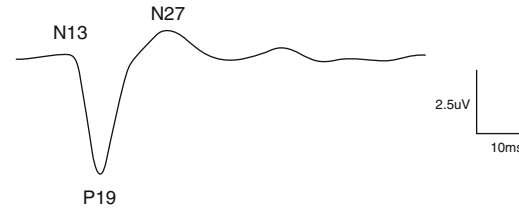

Fig 12.2 Characteristic wave of trigeminal SSEP under general anesthesia

Recording Electrodes and Band Pass The cortical response is recorded from electrodes situated at the scalp. Recordings were performed with needle electrodes placed at C5 or C6 (the contralateral side of the scalp) with Fpz as reference, according to the International 10/20 system (Malcharek et al. 2011). The waveform pattern was maintained irrespective of whether the reference site was Fpz or Cv7 (Fagade and Wastell 1990). Signals are filtered with band-pass filter from 0.1 to 1000 Hz. Sampling rate is 5000 Hz.

12.4.2.2 Intraoperative Interpretation

Short-latency, negative components elicited by electrical stimulation of branches of the trigeminal nerve have latencies of 0.9, 1.6, and 2.6 ms when recorded from the trigeminal nerve where it enters the brain stem (Stechison and Kralick 1993). These potentials represent neural activity in the trigeminal nerve, not in any other rostral structures, and such recordings can, thus, only be used to monitor the trigeminal (sensory) nerve (Oikawa et al. 2000). Long-latency components are also followed. Latencies and amplitudes of the N13 and P19 peaks were measured (Malcharek et al. 2011) (Fig. 12.2).

Different trigeminal evoked potentials showed significantly increased latencies and statistically significant threshold elevations on the affected sides (Bennett and Jannetta 1983); tSSEP could represent a fast and safe way of determining trigeminal afferent function in a laboratory setting (Adamec et al. 2014).

A significant difference in BTEP latencies was found between the normal side and the affected side before the surgery. While in the patients who were released from pain after the MVD, the BTEP latencies of the two sides did not differ significantly (Vriens and Pasman 1994). The latencies after MVD became shorter than before the surgery, and the intensity of stimulation necessary to reach the threshold was lower after the surgery (Adamec et al. 2014).

12.4.3 Somatosensory Evoked Potentials of Upper Extremity

Somatosensory evoked potentials (SSEPs) can be elicited by electrical stimulation of a peripheral nerve, such as the median/ulnar nerve at the wrist or the posterior tibial nerve at the ankle. It is used intraoperatively to monitor blood perfusion of the cortex or the spinal cord and the structural and functional integrity of peripheral nerves and spinal nerve roots.

Injury of the brachial plexus secondary to malposition of the patient during surgery is a significant perioperative problem and one of the common complications after MVD. In most cases, damage to the plexus can be prevented by monitoring the SSEPs generated in this region elicited by stimulation of the ulnar nerve and the median nerve.

12.4.3.1 Stimulation and Recording

Stimulation Sites Stimulation to the upper extremity is delivered to the median or ulnar nerve at the wrist. Stimulation to the lower extremity is delivered to the posterior tibial nerve at the ankle.

Stimulus Intensity and Rate Typical intensity values are 25 mA for arm stimulation and 50 mA for leg stimulation. The stimulus duration is set at 0.3 ms. A noninteger stimulation rat, such as 4.7/5.1 Hz, is used to avoid synchronization with power line interference.

Recording Electrodes and Band Pass The electrodes are placed on the scalp on specific locations according to the 10–20 international placement system used in clinical applications. For upper extremity stimulation, recordings are taken from the shoulder, cervical spine, and scalp. Three cortical channels (C'3-Fpz, C'4-Fpz, C'3-C'4), one cervical channel (Cs2-Fpz/Cs5-Fpz), and one peripheral channel (Erb's ipsilateral-contralateral) are needed for recording the median/ulnar nerve. Reference electrodes may be at the forehead or mastoid reference sites. The optimal low filter setting usually is 30 Hz. The optimal high filter setting is 1500–3000 Hz. The 60 and 50 Hz notch filter should be kept off. About 300–500 stimulations can produce a well-defined SSEP.

12.4.3.2 Intraoperative Interpretation

The recording channel Erbi-Erbc displays the first recorded potential as the Erb's point or N9. This consists of a negative upward deflection, occurring at about 9 ms after stimulation of the median/ulnar nerve at the wrist and represents the electronegativity caused by the electrical volley reaching the region of the EP (Chiappa 1990).

The recording channel Cs2-Fz/Cs5-Fz displays the second recorded potential, the cervicomedullary junction potential, represented by P/N13 complex. This potential is represented by a negative upward deflection, occurring at about 13 ms after the stimulation of the median nerve at the wrist.

The most important scalp-recorded component has a negative peak at about 20 cosec which is followed by a positive peak at about 25 ms, forming the N20-P25 complex. The N20 probably originates from the cortical area contralateral to the side of stimulation (C'3-Fpz/C'4-Fpz) (Fig. 12.3).

SSEPs have become an important adjunct in a wide range of surgical procedures. Thus, they are used for monitoring the function of the large-fiber sensory system during a variety of surgical procedures that could result in its damage. A greater than or equal to 50 % decrease in SSEP amplitude and/or a greater than or equal to 20 %

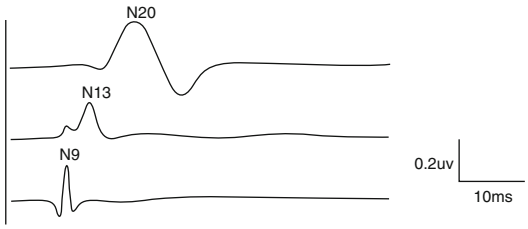

Fig 12.3 Somatosensory evoked potentials from median nerve stimulation are shown. Typical peaks are shown in each of the three recording channels

increase in latency is considered as significant change to produce potential postoperative neurological deficit. Amplitudes of the different components of evoked potentials are more susceptible to random changes than are the latencies of specific components.

During MVD surgery, the patient was placed in the park-bench position (lateral position with neck flexed and head rotated toward the floor). In order to get a better exposure and access, the upper shoulder and arm were depressed and fixed by means of tape attached to the shoulder and arm, while pronation and flexion of the lower arm can compress the ulnar nerve between the table and the cubital tunnel. Continuous intraoperative SSEP monitoring of ulnar/median nerve function intraoperatively as an indicator of brachial plexus function is a valid and useful technique to minimize intraoperative neurological injuries during MVD surgeries.

12.4.3.3 Anesthesia Requirements

Inhalational anesthetics, such as isoflurane, halothane, and enflurane, all reduce the amplitude and increase the latency of cortical components, and they are directly correlated to the concentration in which such agents are administrated in a dose-dependent fashion (Wang et al. 1985; Onofrj et al. 1990).

Intravenous agents, propofol, benzodiazepines, barbiturates, etomidate, ketamine, and opiates, all affect the amplitude and latency of SSEPs. The effects of various drugs most commonly used in anesthesia on the SSEPs are summarized and listed in Table 12.2.

Table 12.2 Effects of anesthetic agents on SSEP amplitude and latency

Agent	Amplitude	Latency
Nitrous oxide (N_2O)	↓	↑
Inhalational anesthetics	↓	↑
Propofol	–	↑
Barbiturates	↓	↑
Etomidate	↓	↑
Ketamine	–	↑
Opiates: morphine, fentanyl, alfentanil, sufentanil	–	↑
Benzodiazepines	↓	↑
Muscle relaxants	–	–
Hypotensive agents	↓	↑

Hypothermia might increase the latency and slightly decrease the amplitude of SSEPs, while hyperthermia will decrease the latency and decrease the amplitude. Severe hypotension can also result in a drastic decrease or even total loss of the cervical and cortical responses

12.4.4 Free-Run and Triggered Electromyography

Free-run EMG (fEMG) consists of recording spontaneous muscle activity. In intraoperative neurophysiology, it helps identify motor nerve depolarization from surgically driven irritation, hopefully before irreversible damage to these structures had occurred.

Triggered electromyographic (tEMG) activity can be recorded from a corresponding muscle after direct electrical stimulation of the motor nerve or nerve root. These signals are also known as compound muscle action potentials (CMAPs). Thus, it can be used to identify specific cranial nerves or nerve roots that may be difficult to distinguish from tumoral, fibrous, and fatty tissues and protect structural and functional integrity of cranial nerves and spinal roots.

12.4.4.1 Stimulation and Recording

Stimulation Sites No stimulation is required for fEMG recording. It is based entirely on recording spontaneous and detecting irritation-driven muscle activity, while an electrical stimulus directly on the nervous structures is required for triggered

EMG. For cranial nerve stimulation, monopolar technique is generally used. With bipolar or monopolar handheld probe, triggered EMG can be done by direct electrical stimulation of the nerve.

Stimulus Intensity and Rate The stimulation technique consists of delivering constant current, with repetitive square wave pulses of 0.1–0.2 ms, frequency 4 Hz, averaging 4–8 trials. Stimulus intensity for direct nerve or nerve root stimulation is gradually increased from 0 mA until an EMG response is seen up to a maximum of about 2 mA.

Recording Electrodes and Band Pass For both these methods, electrodes are placed in the target muscles corresponding to the nerve of interest. Most commonly, pairs of monopolar needles are used. The muscles typically used for monitoring cranial nerves are listed in the table. Relatively wide filter settings are used: low-frequency filter at 5 Hz and the high-frequency filter at 5 kHz. The sweep speed is usually set at 200 ms per division for recording continuous EMG and 2–5 ms/division for recording electrically evoked EMG potentials. The sensitivity is set at 50–200 uV per division.

12.4.4.2 Intraoperative Interpretation

Monitoring of both spontaneous and triggered EMG is strongly recommended also in microvascular decompressions. Continuous recording of spontaneous EMG can be used to provide early warnings of irritation of the nerve.

Free-Run EMG Activity

During free-run EMG monitoring, muscle activity is assessed continuously during portions of the procedure where the associated motor nerves are at risk. Manipulation of motor nerves may elicit depolarization of motor axons which then activates corresponding motor units within the monitored muscle. The recorded motor unit potentials (MUPs) may exhibit a wide range of patterns, which are characterized and interpreted to understand and predict the consequence of surgical actions that may elicit the activity. Making the EMG signal audible can greatly facilitate EMG monitoring, so that both the neurophysiologist and the surgeon can hear it. The following three criteria might be the signal of possible nerve injury: (1) sustained firing of a high-frequency train lasting for tens of seconds, (2) several large bursts of activity of complex morphology, and (3) sudden bursts of high-amplitude spikes followed by complete silence (Moller 1995). As a general rule, greater numbers and higher rates of recurrent MUPs correlate to higher levels of nerve irritation. But in the extreme cases, nerve injury may occur with a clean transaction of the nerve; there may be no EMG activity (Nelson and Vasconez 1995).

Triggered EMG Activity

Electrical stimulus is represented by CMAPs, which are a sum of motor action potentials that arise in several muscle fibers. During the surgery, the monitoring with EMG of muscles innervated by CNs VII, IX, and X can prevent facial palsies, dysphonia, and/or dysphagia (Kartush et al. 1991; Mishler and Smith 1995; Schlake et al. 1999; Harper 2004; Minahan and Mandir 2011; Singh and Husain 2011). Though these CN palsies are usually transient, they are directly attributable to these nerves' cauterization, pulling, retraction, or section that can happen during these procedures (Habeych et al. 2014). The identity of the cranial nerve can be resolved by simply determining the muscle on which a response has been obtained. If the same recording electrode detects activity from a muscle corresponding to two different nerves, then the latency of the response will determine its origin, for example, identifying the trigeminal from the facial nerve, since stimulation of the trigeminal nerve elicits a CMAP onset latency ranging between 3.5 and 5 ms (Moller 2011), while facial nerve stimulation generally produces a CMAP with an onset latency between 6 and 8 ms (Fig. 12.4).

During a surgery of CPA tumor resection, tEMG can be used to identify and thus avoid damage to the motor branch of the nerve which must be preserved. A baseline threshold should

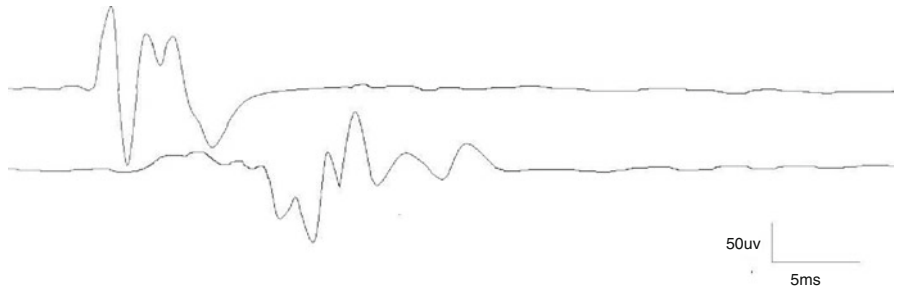

Fig 12.4 *Upper curve*: EMG potentials recorded from electrodes placed in the masseter muscles. *Lower curve*: EMG potentials recorded from electrodes placed in the orbicularis oculi muscles

be taken before the dissection. In addition to tEMG, continuous recording of fEMG can be used to provide early warnings of mechanical or thermal irritation of the nerve. Periodic stimulation of the nerve and recording of tEMG can inform the surgeon of the functional integrity of the nerve at any given point in time and then at the end of the surgery, by stimulating the nerve proximal to the tumor first. A prolonged reduction in response amplitude that is not due to perisurgical events, such as an increased level of muscle relaxation, is associated with an increased likelihood of postoperative muscle weakness (Zouridakis and Papanicolaou 2000; Moller 2011).

12.4.4.3 Anesthesia Requirements

The level of muscle relaxation or pharmacologically induced paralysis is the most crucial factor affecting EMG monitoring. It is recommended that the patient is free of their effects during the time when EMG monitoring is performed, or the neuromuscular blocking agents should be kept to the minimum that the clinical situation allows. Inhaled anesthetic agents have little or no effect on EMG monitoring. Depth of muscle relaxation is monitored by delivering a train of four electrical stimuli, with an intensity of about 25 mA, usually to the median or facial nerve, and measuring the resulting number of muscle contractions. Zero twitches indicate complete paralysis, whereas at least three twitches indicate that the patient remains minimally relaxed or practically no paralysis (Zouridakis and Papanicolaou 2000). The effects of anesthesia and of the patient's blood pressure and temperature must all be considered when applying EMG in the operating room.

12.4.5 Abnormal Muscle Response

In 1985, Moller and Jannetta first showed that in patients with hemifacial spasm, stimulation of one branch of the facial nerve may result in the activation of facial muscles innervated by other branches, due to the hyperexcitability of the facial nerve. This abnormal muscle response has been termed "lateral spread" or "lateral spread response" (LSR) and is thought to be related to ephaptic transmission, though the specific mechanism of lateral spread is unclear. Due to the fact that LSR disappears instantly in most of the patients when the offending vessel is moved off the facial nerve, monitoring the abnormal muscle response can guide the surgeon during MVD which results in a better postoperative outcome (Moller and Jannetta 1987).

Neuro-monitoring of the lateral spread response and, more specifically, its disappearance may help predict short- and long-term success of microvascular decompression (MVD) for hemifacial spasm.

12.4.5.1 Stimulation and Recording

Stimulation Sites Stimulation needle electrodes are most commonly placed over the temporal branch or the marginal mandibular branch of the facial nerve (Moller 1991).

Stimulation Intensity and Rate A single constant current stimulus of 0.1–0.2 ms duration, at a stimulating rate of 1 Hz, is used for eliciting lateral spread. Usually, a stable AMR was recorded at a stimulation intensity level of 5–15 mA. During MVD, the threshold intensity is adjusted for continuously eliciting the lateral spread. If AMR disappeared, the intensity was increased up to 50 mA for a few seconds in order to confirm the permanent LSR disappearance (Moller and Jannetta 1986).

Recording Electrodes and Band Pass Recording needles are inserted 0.5–1 cm apart subcutaneously into the mentalis muscle if the temporal branch of the facial nerve is stimulated or into the orbicularis oculi muscles if the marginal mandibular branch is stimulated (Fig. 12.5).

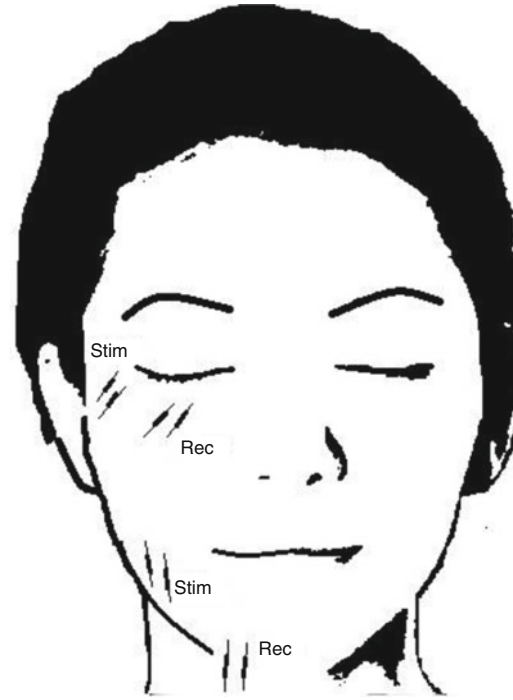

Fig 12.5 Placement of stimulating and recording electrodes for monitoring the abnormal muscle response in patients with hemifacial spasm

12.4.5.2 Intraoperative Interpretation

With intraoperative monitoring, an AMR wave with a latency of around 10 ms after stimulation was recorded. The baseline threshold for eliciting the AMR is determined, and stimulation may proceed near this level. When the offending vasculature is moved off the facial nerve, the LSR is known to disappear or become markedly attenuated (Thirumala et al. 2011) (Fig. 12.6). However, the AMR may dissipate before the MVD is performed as sometimes occurs, for instance, when the dura is opened. It is suggested that stimulation intensity and/or rate should be increased in these circumstances to reestablish the presence of AMR (Sekula et al. 2009).

The practical value of AMR disappearance as a method to evaluate MVD efficacy is still controversial (Joo et al. 2008). There are some patients where AMR does not fully disappear despite an apparent effective decompression and they are free of spasm with a delay up to 1 year after surgery. However, in most cases, LSR monitoring is an effective tool to predict outcome after MVD for HFS (Kong and Park 2007; Kim et al. 2010; Thirumala et al. 2011; Ying et al. 2011).

12.4.5.3 Anesthesia Requirements

As AMR is a kind of triggered electromyographic response, the level of muscle relaxation played a very important role in the monitoring. No muscle relaxant is used after the induction of anesthesia.

12.4.6 ZLR

ZLR was first reported by Zheng et al. (2012) for intraoperative monitoring of HFS. It is useful when an AMR is absent before decompression or persists after all vascular compressions are properly treated. Particularly, the ZLR response may help neurosurgeons determine the real culprit when multiple offending vessels exist.

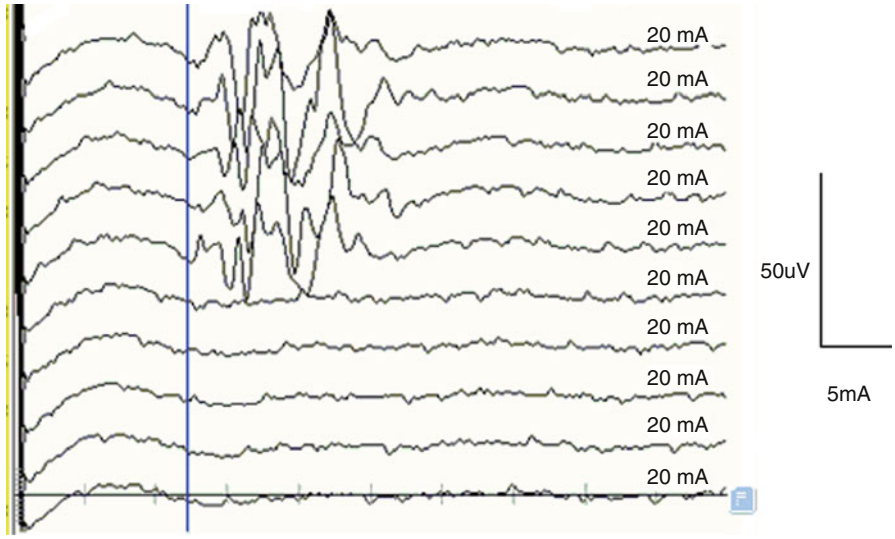

Fig 12.6 Typical changes of AMR during MVD for HFS

12.4.6.1 Stimulation and Recording

Stimulation Sites The stimulating electrode was placed on the offending artery wall near the compression site (within 5 mm) during MVD for hemifacial spasm (Zheng et al. 2012).

Stimulation Intensity and Rate A single constant current stimulus of 0.1–0.2 ms duration, at a stimulating rate of 1 Hz, is used for eliciting ZLR. The intensity of stimulation is 1–2 mA.

Recording Electrodes and Band Pass The ZLR was recorded from the orbicularis oculi, orbicularis oris, and mentalis muscles. Needle electrodes are used to record ZLR from the facial muscles including orbicularis oculi, orbicularis oris, and mentalis. Filter settings are the same as that of the AMR.

12.4.6.2 Intraoperative Interpretation

The ZLR looked similar to an AMR. The latency of ZLR is 7.3 ± 0.8 ms, which is significantly shorter than the AMR latency (Fig. 12.7).

The ZLR can only be detached on the offending artery, when more than one artery compressed the facial nerve. In the majority of patients, in which one offending vessel compressed the facial nerve at one offending site, ZLR and AMR provided the same information for the surgery: both ZLR and AMR disappeared immediately when the offending artery is removed from the facial nerve. But when AMR was absent from the beginning or persisted after the offending vessel was transposed with Teflon sponges, ZLR played a crucial role to identify whether there was sufficient decompression (Yang et al. 2014). The combination of AMR and ZMR might provide more useful information than does the AMR alone, and ZLR may be the only useful intraoperative EMG for MVD surgery in some cases (Zheng et al. 2012).

12.4.6.3 Anesthesia Requirements

The anesthesia requirements of ZLR monitoring are the same as that of AMR. No muscle relaxant is supposed to be used after the induction of anesthesia.

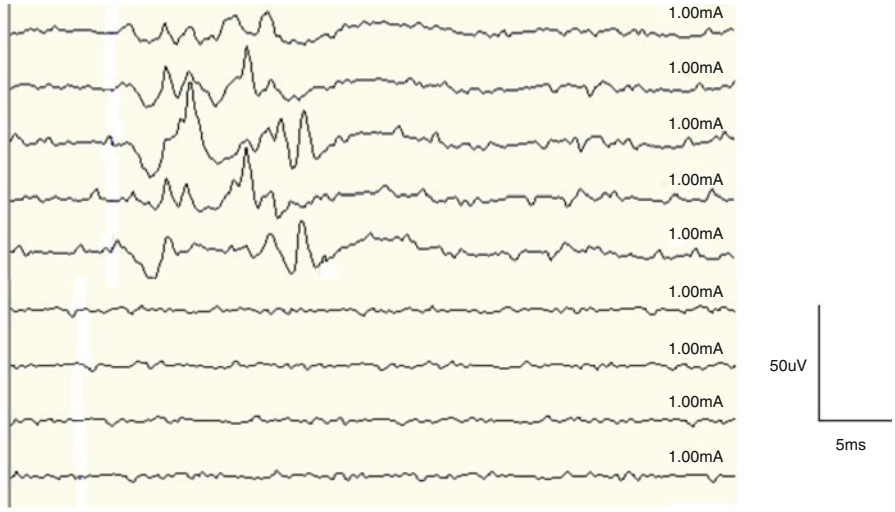

Fig 12.7 ZLR recorded from the facial muscles when stimulating the offending artery wall

12.4.7 Facial F Wave

The F wave is an antidromic pulse that propagates to an alpha motor neuron in the anterior horn of the spinal cord and then returns orthodromically down the same axon. It is a standard electrophysiological means to reveal lesions in the proximal segments of peripheral nerves.

The facial F-wave findings in various diseases involving the intracranial and intracanalicular portion of the nerve have recently been reported to demonstrate the diagnostic utility of this method (Wedekind and Klug 1998).

12.4.7.1 Stimulation and Recording

Stimulation Sites The stimulating electrodes are placed in proximal position over the zygomatic branch of the facial nerve.

Stimulus Intensity and Rate The facial nerve was stimulated with constant current rectangular monophasic pulses (duration of 0.1–0.2 ms) transmitted transcutaneously (for extraoperative studies) or by subdermal needle electrodes (for intraoperative measurements). Stimulation frequency was 1 Hz (Wedekind and Klug 2003). The stimulation strength was set to supramaximum to elicit stable M waves with minimum stimulus intensity (usually 3–10 mA).

Recording Electrodes and Band Pass For recording of muscle activity, silver chloride cup electrodes filled with conducting gel were placed on both alae nasi to record the compound action potential of the pars alaris of the nasal muscle. These active electrodes were referenced to an electrode on the nose tip or the glabella (Wedekind and Klug 2000; Wedekind et al. 2001). F waves, which varied according to each stimulus, were analyzed for F/M amplitude ratio (i.e., the percentage of the peak to peak amplitude of the F wave to the M wave) and duration (from the initial deflection from the baseline to the final return of the F wave) (Ishikawa et al. 1996). Recordings were passed through a preamplifier and amplifier (100 u/div) and then filtered (band pass of 20 and 3000 Hz) without averaging.

12.4.7.2 Intraoperative Interpretation

The F waves are useful indicators of lower motor neuron excitability (Hai and Pan 2007). In the ipsilateral mentalis muscles, the F wave through stimulating the marginal mandibular branch of the facial nerve is also an antidromic pulse that propagates to the facial motor nucleus and returns orthodromically down the same axon. Thus, the F wave in facial muscles may indicate the excitability of the facial motor nucleus (Fig. 12.8).

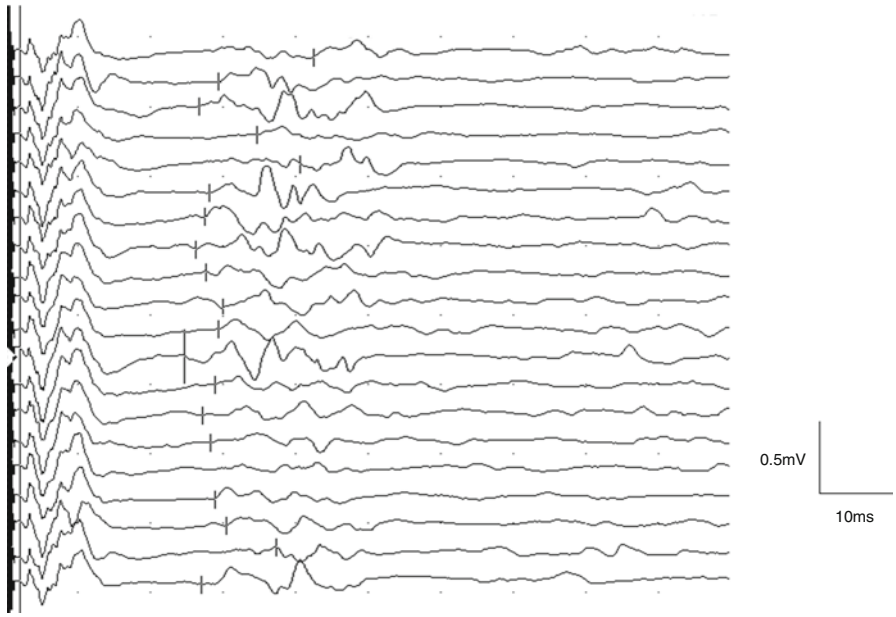

Fig 12.8 Typical F wave recorded from facial nerve

Electrophysiological study demonstrated that the F-wave appearance is more persistent in patients with HFS. Neurophysiological investigations have been done in patients with HFS, and F wave has been recorded in facial muscles before and after MVD (Moller 1991; Ishikawa et al. 1997). It has been shown that there are significant increased F waves in the amplitude, duration, and frequency in patients with HFS (Hai and Pan 2007). Immediate changes in excitability on facial motor nucleus are observed by monitoring changes in F-wave elicitability. These responses are monitored together with LSR, and there is evidence that whenever LSR disappears, F wave disappears as well (Fernandez-Conejero et al. 2012).

Intraoperative recordings of facial nerve F waves recorded from nasal muscles have been studied as an effective measure of facial nerve function in surgery. In the surgery of CPA tumors, facial F-wave monitoring provides continuous and valid real-time information concerning the functional status of the nerve under the strain of dissection. Intraoperative F wave changes closely parallel the actual strain exerted on the nerve. Intraoperative monitoring of F waves further provides a feature (transient loss of the F wave) which heralds an imminent danger of severe facial dysfunction to occur. Wedekind et al. (2001) have investigated the use of F waves during vestibular schwannoma resection and report that a permanent loss of F-wave results in 91 % sensitivity and 100 % specificity for poor outcome. In addition, the transient loss of signals was detected more frequently in patients with moderate outcomes. Facial F-wave recording provides valuable information on the functional status of the nerve intra- and extraoperatively (Wedekind and Klug 2003).

12.4.7.3 Anesthesia Requirements

The intraoperative use of facial nerve F waves is limited by its sensitivity to anesthesia and its absence in some healthy adults (Wedekind et al. 2001; Wedekind and Klug 2003).

12.4.8 Blink Reflexes

In 1952, Kugelberg was the first to report the presence of two electrically induced blink reflex (BR) components, R1 and R2 (Kugelberg 1952).

The R1 response corresponds to the oligosynaptic reflex arc, which includes trigeminal afferents, brain stem connections between the sensory part of the trigeminal nucleus and the motor nucleus of the facial nerve, the facial nerve proper, and the orbicularis oculi muscle. The R2 component is more complex in its central, polysynaptic connections within the brain stem, but it has the same afferent and efferent pathways as the R1.

The blink reflex would be suitable for continuous monitoring of the function of both trigeminal and facial nerve intraoperatively. Unfortunately, BR was typically absent in anesthetized patients (Mourisse et al. 2004). It was recently shown that it is possible to elicit the blink reflex in surgically anesthetized patients using a train of four to seven stimulus impulses applied to the supraorbital nerve when suitable modern anesthesia techniques are used (Deletis et al. 2009). Thus, the use of this test for the use for intraoperative neuro-monitoring appears feasible.

12.4.8.1 Stimulation and Recording

Stimulation Sites Stimulation of the supraorbital nerve was performed using a pair of electroencephalographic needle electrodes inserted subcutaneously over the supraorbital nerve.

Stimulus Intensity and Rate Four to seven rectangular constant current stimuli with an interstimulus interval (ISI) of 2 ms, intensity of 20–40 mA, and train repetition rate of 0.4 H z were used.

Recording Electrodes and Band Pass Recording was done with needle electrodes identical to those used for stimulation. The electrodes were inserted in the low lateral part of the orbicularis oculi muscle ipsilateral and contralateral to the stimulating side. Recordings were made by using a 50-ms epoch and band-pass digital filters of 70 and 1219 Hz. Recording of the BR was attempted after intubation, in the middle of surgery, and after starting skin closure (Deletis et al. 2009).

12.4.8.2 Intraoperative Interpretation

Characteristically, an electrical stimulus on the supraorbital nerve (VI) induces two recordable responses in the orbicularis oculi muscles: an early one, the so-called R1, ipsilateral to the stimulated side, and a later one, the R2, which is bilaterally expressed. The R2 response ipsilateral to the stimulus is frequently cited as R2i, and the R2c is the one obtained on the contralateral side, also denominated "consensual response," in analogy to the photomotor pupillary reflex (Esteban 1999).

The R1 component of the BR has a rather stable latency of approximately 10 ms. R2 typically shows relative variable latencies and larger magnitudes than R1 and its threshold is lower (Sanes et al. 1982).

In symptomatic trigeminal pains, the trigeminal reflexes have a very high sensitivity, probably because they allow examination of all three divisions. The most sensitive reflex is the R1 of the blink reflex (Cruccu et al. 1990). Mild reflex abnormalities occur occasionally, but in most patients with idiopathic trigeminal neuralgia, all reflexes are normal (Aramideh 2002).

BR can be also used to evaluate hyperexcitability of the facial motor nucleus in patients with HFS by measuring the latency and duration of the R2 component as well as the R1 and R2 recovery curves (Valls-Sole and Tolosa 1989). A larger R1 and R2 response is usually obtained on stimulating the affected side, rather than the contralateral side (R2c), or after stimulating the unaffected side, the ratio R2c/R2 may be increased (Eekhof et al. 2000; Oge et al. 2005; Sekula et al. 2009). After decompression of the facial nerve during the MVD surgery for HFS, it is necessary to increase the number of stimuli within the train in order to reproduce BR. It is presumed that this phenomenon results in the immediate decrease in excitability of the facial motor nucleus after an effective MVD (Fernandez-Conejero et al. 2012).

The possibility to achieve continuous monitoring of the sensory part of the trigeminal nerve, brain stem, and facial nerve is a further important development.

12.4.8.3 Anesthesia Requirements

It is feasible to record the BR in patients under general anesthesia using low doses of desflurane or sevoflurane during surgery. If total intravenous anesthesia is used, boluses of propofol should be avoided, because they prevent elicitation of the BR. Administration of muscle relaxants should also be avoided.

12.4.9 Motor Evoked Potentials

Transcranial electrical stimulation was first applied to the human brain by Merton and Morton (1980). It can be generated by electrical stimulation of the cortex or the spinal cord. It involves applying an electrical current with the purpose of depolarizing the corticospinal system proximal to the level of the surgery, above its threshold. This method assesses the integrity of the motor pathways, from the cortex to the muscles. The method was found to be practical and effective intraoperatively, and it is now widely accepted as necessary in the neurosurgical field (MacDonald 2002; Motoyama et al. 2011). For neurophysiological IOM of the motor pathways, the use of MEPs triggered by electrical cortical stimulation is a technique done successfully under anesthesia.

12.4.9.1 Stimulation and Recording

Stimulation Sites The standard 10–20 EEG electrode placement is used. Scalp electrodes are often placed more lateral than the standard montage for spinal surgeries (Fernandez-Conejero et al. 2012): C3-Cz for left hemispheric stimulation and C4-Cz for right hemispheric stimulation (Dong et al. 2005). The use of a Cz cathode may help to minimize the chance of extracranial activation of the facial CSound stimulus is delivered through miniature.

Stimulus Intensity and Rate Transcranial stimulation may be constant voltage (range of 180–600 V) or constant current (range of 50–220 mA) with varying durations of 0.5–3 ms and interstimulus interval of 1–5 ms, typically using three or more pulses with a train repetition rate of 2 Hz (Akagami et al. 2005; Dong et al. 2005; Sala et al. 2007; Fukuda et al. 2008; Acioly et al. 2010).

Table 12.3 Cranial nerves and the corresponding muscles used in IOM

Cranial nerve	Nerve name	Muscles
II	Oculomotor	Inferior rectus, inferior oblique
IV	Trochlear	Superior oblique
V	Trigeminal	Masseter, temporalis
VI	Abducens	Lateral rectus
VII	Facial	Frontalis, orbicularis oculi, nasalis, orbicularis oris, mentalis
IX	Glossopharyngeal	Lateral soft palate
X	Vagus	Vocal cords, cricothyroid
XI	Accessory	Trapezius/ sternocleidomastoid
XII	Hypoglossal	Tongue/genioglossus

Recording Electrodes and Band Pass Electrodes are placed in the target muscles corresponding to the nerve of interest. Most commonly, pairs of monopolar needles are used. The muscles typically used for monitoring cranial nerves are listed in Table 12.3. Filter settings vary between 5 and 10 kHz (Akagami et al. 2005; Dong et al. 2005; Sala et al. 2007; Fukuda et al. 2008; Acioly et al. 2010).

12.4.9.2 Intraoperative Interpretation

Preservation of postoperative facial nerve function is an important goal of microvascular decompression. To estimate postoperative cranial nerve function, MEP monitoring was reported to be of value (Fukuda et al. 2008; Matthies et al. 2011). It facilitates observation of the function of both the first and second motor neuron and helps to predict postoperative nerve function. However, MEP cannot be used for continuous intraoperative monitoring because of body movement artifacts. Operative manipulations must be stopped during recording of the MEP (MacDonald 2002; Motoyama et al. 2011).

MEP can provide a beneficial impact on the prevention of paraplegia and paralysis during surgeries

in which the motor pathways may be compromised (Kodama et al. 2014) The intraoperative increase in stimulation threshold and decrease in amplitude were closely correlated to postoperative nerve dysfunction (Cosetti et al. 2012; Macdonald et al. 2013; Sarnthein et al. 2013).

More recently, MEPs have also been used during MVD as a method to identify and avoid lesions to the facial nerve when manipulating the vessels compressing the facial nerve. Facial CMAPs in TcMEPs appear at a short latency around 15 milliseconds (Fukuda et al. 2008). However, the most sensitive criterion for intraoperative interpretation is based only on amplitude. Fujiki et al. suggest that a decline ≥35 % relative to baseline amplitude is associated with minimum transient postoperative motor deficits (Fujiki et al. 2006; Fukuda et al. 2008).

Fernandez-Conejero et al. (2012) reported that MEP monitoring in patients with HFS has remarkable characteristics due to the hyperexcitability of the facial nerve and facial motor nucleus. The FCoMEPs are continuously recorded before, during, and after MVD together with LSR. The major excitability changes observed in FCoMEPs are increased by the number of stimuli required to obtain responses or intensity of current compared to the baselines. Thus, TcMEPs have also been investigated as adjuvant monitoring during MVD for hemifacial spasm (Fig. 12.9).

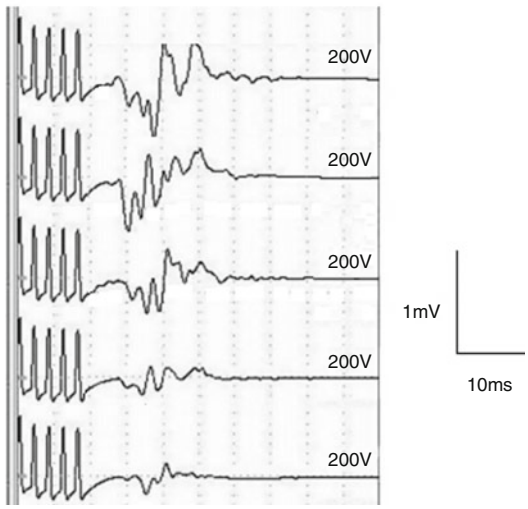

Fig 12.9 It takes a short train stimulus of 200 V to elicit MEP before surgery. After MVD is completed, the amplitude of MEP decreased significantly

12.4.9.3 Anesthesia Requirements

In general anesthetic techniques should be used with minimized inhalational agents, such as nitrous oxide and isoflurane (Kalkman et al. 1993), and minimized neuromuscular blockade. Anesthesia techniques using a combination of less than 50 % nitrous oxide and narcotics, etomidate, or ketamine allow the recording of reliable MEPs (Kalkman et al. 1993).

12.5 Case

The patient was diagnosed as a typical left HFS based on the clinical history of typical symptoms and electrophysiological examination. BAEP, AMR, ZLR, and SSEPs were recorded during the MVD operation.

The operation was performed under general anesthesia. No muscle relaxant is supposed to be used after the induction of anesthesia. To ensure the integrity of the stimulating and recording electrodes, an initial recording was made as baseline following induction, before the patient had been positioned. After the patient was positioned, another SSEP was recorded. Then, subsequent recordings continued throughout the case.

During the entire operation, the root exit zone (REZ) of the facial nerve was compressed by both the vertebral artery (VA) and anterior inferior cerebellar artery (AICA) (a). A typical AMR was recorded on this operation (b). The Z-L response (ZLR) was identified from AICA (c, d), but not from VA (e, f). Both AMR and ZLR disappeared after decompression (g, h, i). The patient achieved immediate "excellent" resolution of spasms after surgery.

The latency of wave V and the IPL I–V was found delayed when the VIIIth nerve was retracted. Then the surgeon was informed and the retracting stopped. The BAEPs came back to normal until the end of the surgery (j).

This patient achieved excellent resolution of spasm. No significant change was recorded from SSEPs. Postoperatively, there were no complications observed, and the patient denied any symptoms such as hearing loss, tingling, or numbness of upper extremities (Fig. 12.10).

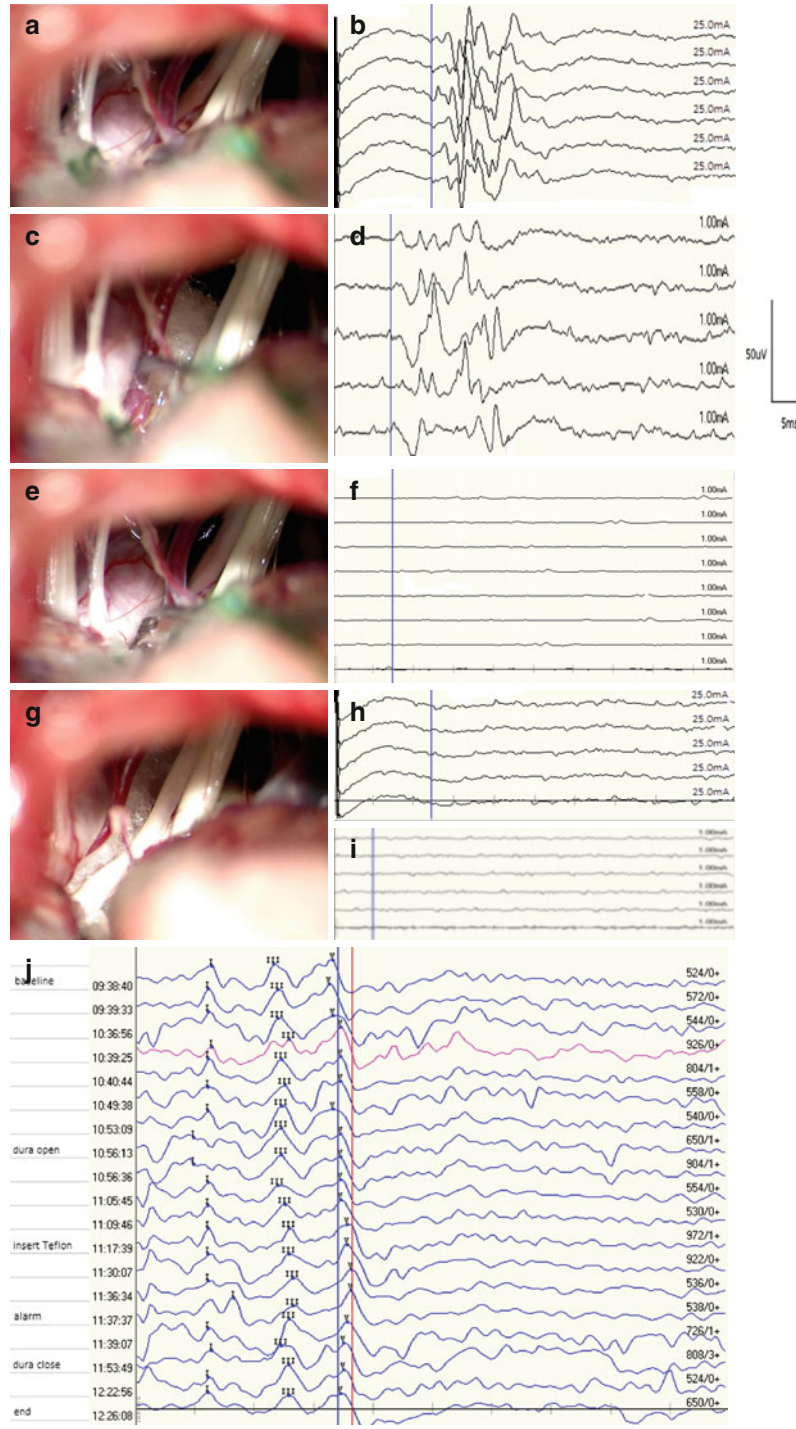

Fig 12.10 Electrophysiological monitoring change and intraoperative pictures of the typical HFS patient

References

Acevedo JC, Sindou M, Fischer C, Vial C. Microvascular decompression for the treatment of hemifacial spasm. Retrospective study of a consecutive series of 75 operated patients--electrophysiologic and anatomical surgical analysis. Stereotact Funct Neurosurg. 1997;68(1–4 Pt 1):260–5.

Acioly MA, Liebsch M, Carvalho CH, Gharabaghi A, Tatagiba M. Transcranial electrocortical stimulation to monitor the facial nerve motor function during cerebellopontine angle surgery. Neurosurgery. 2010;66:354–61; discussion 362.

Adamec I, Grahovac G, Krbot Skoric M, Chudy D, Hajnsek S, Habek M. Tongue somatosensory-evoked potentials in microvascular decompression treated trigeminal neuralgia. Acta Neurol Belg. 2014;114:55–8.

Akagami R, Dong CC, Westerberg BD. Localized transcranial electrical motor evoked potentials for monitoring cranial nerves in cranial base surgery. Neurosurgery. 2005;57:78–85; discussion 78–85.

Aramideh M, Ongerboer de Visser BW. Brainstem reflexes: electrodiagnostic techniques, physiology, normative data, and clinical applications. Muscle Nerve. 2002;26:14–30.

Bennett MH, Jannetta PJ. Evoked potentials in trigeminal neuralgia. Neurosurgery. 1983;13:242–7.

Chiappa KH. Evoked potentials in clinical medicine. Philadelphia: Lippincott Williams & Wilkins; 1990.

Cosetti MK, Xu M, Rivera A, Jethanamest D, Kuhn MA, Beric A, et al. Intraoperative transcranial motor-evoked potential monitoring of the facial nerve during cerebellopontine angle tumor resection. J Neurol Surg B Skull Base. 2012;73:308–15.

Cruccu G, Leandri M, Feliciani M, Manfredi M. Idiopathic and symptomatic trigeminal pain. J Neurol Neurosurg Psychiatry. 1990;53:1034–42.

Deletis V, Urriza J, Ulkatan S, Fernandez-Conejero I, Lesser J, Misita D. The feasibility of recording blink reflexes under general anesthesia. Muscle Nerve. 2009;39:642–6.

Dong CC, Akagami R, Westerberg B, Alkhani A, Kanaan I, Hassounah M. Intraoperative facial motor evoked potential monitoring with transcranial electrical stimulation during skull base surgery. Clin Neurophysiol. 2005;116(3):588–96.

Eekhof JL, Aramideh M, Speelman JD, Devriese PP, De Ongerboer VBW. Blink reflexes and lateral spreading in patients with synkinesia after Bell's palsy and in hemifacial spasm. Eur Neurol. 2000;43:141–6.

Esteban A. A neurophysiological approach to brainstem reflexes. Blink reflex. Neurophysiol Clin. 1999;29:7–38.

Fagade OO, Wastell DG. Trigeminal somatosensory evoked potentials: technical parameters, reliability and potential in clinical dentistry. J Dent. 1990;18:137–41.

Fernandez-Conejero I, Ulkatan S, Sen C, Deletis V. Intraoperative neurophysiology during microvascular decompression for hemifacial spasm. Clin Neurophysiol. 2012;123:78–83.

Fujiki M, Furukawa Y, Kamida T, Anan M, Inoue R, Abe T, Kobayashi H. Intraoperative corticomuscular motor evoked potentials for evaluation of motor function: a comparison with corticospinal D and I waves. J Neurosurg. 2006;104(1):85–92.

Fukuda M, Oishi M, Takao T, Saito A, Fujii Y. Facial nerve motor-evoked potential monitoring during skull base surgery predicts facial nerve outcome. J Neurol Neurosurg Psychiatry. 2008;79:1066–70.

Galloway GM, Nuwer MR, Lopez JR, Zamel KM. Intraoperative neurophysiologic monitoring. Cambridge: Cambridge University Press; 2010.

Habeych ME, Crammond DJ, Gardner P, Thirumala PD, Horowitz MB, Balzer JR. Intraoperative neurophysiological monitoring of microvascular decompression for glossopharyngeal neuralgia. J Clin Neurophysiol. 2014;31(4):337–43.

Hai J, Pan QG. Experimental study on the correlation between abnormal muscle responses and F waves in hemifacial spasm. Neurol Res. 2007;29(6):553–6.

Harper CM. Intraoperative cranial nerve monitoring. Muscle Nerve. 2004;29:339–51.

Hatayama T, Mollar AR. Correlation between latency and amplitude of peak V in the brainstem auditory evoked potentials: intraoperative recordings in microvascular decompression operations. Acta Neurochir (Wien). 1998;140(7):681–7.

Ishikawa M, Namiki J, Takase M, Ohira T, Nakamura A, Toya S. Effect of repetitive stimulation on lateral spreads and F-waves in hemifacial spasm. J Neurol Sci. 1996;142:99–106.

Ishikawa M, Ohira T, Namiki J, Kobayashi M, Takase M, Kawase T, et al. Electrophysiological investigation of hemifacial spasm after microvascular decompression: F waves of the facial muscles, blink reflexes, and abnormal muscle responses. J Neurosurg. 1997;86:654–61.

Jo KW, Kim JW, Kong DS, Hong SH, Park K. The patterns and risk factors of hearing loss following microvascular decompression for hemifacial spasm. Acta Neurochir (Wien). 2011;153:1023–30.

Joo WI, Lee KJ, Park HK, Chough CK, Rha HK. Prognostic value of intra-operative lateral spread response monitoring during microvascular decompression in patients with hemifacial spasm. J Clin Neurosci. 2008;15:1335–9.

Kalkman CJ, Been HD, Ongerboer de Visser BW. Intraoperative monitoring of spinal cord function. A review. Acta Orthop Scand. 1993;64:114–23.

Kartush JM, Larouere MJ, Graham MD, Bouchard KR, Audet BV. Intraoperative cranial nerve monitoring during posterior skull base surgery. Skull Base Surg. 1991;1:85–92.

Kim CH, Kong DS, Lee JA, Kwan P. The potential value of the disappearance of the lateral spread response during microvascular decompression for predicting the clinical outcome of hemifacial spasms: a prospective study. Neurosurgery. 2010;67:1581–7; discussion 1587–8.

Kodama K, Javadi M, Seifert V, Szelenyi A. Conjunct SEP and MEP monitoring in resection of infratentorial lesions: lessons learned in a cohort of 210 patients. J Neurosurg. 2014;121:1453–61.

Kong DS, Park K. Hemifacial spasm: a neurosurgical perspective. J Korean Neurosurg Soc. 2007;42(5): 355–62.

Kugelberg E. [Facial reflexes]. Brain. 1952;75:385–96.

Lee SH, Song DG, Kim S, Lee JH, Kang DG. Results of auditory brainstem response monitoring of microvascular decompression: a prospective study of 22 patients with hemifacial spasm. Laryngoscope. 2009; 119:1887–92.

Legatt AD. Mechanisms of intraoperative brainstem auditory evoked potential changes. J Clin Neurophysiol. 2002;19:396–408.

López JR. The use of evoked potentials in intraoperative neurophysiologic monitoring. Phys Med Rehabil Clin N Am. 2004;15(1):63–84.

MacDonald DB. Safety of intraoperative transcranial electrical stimulation motor evoked potential monitoring. J Clin Neurophysiol. 2002;19:416–29.

Macdonald DB, Skinner S, Shils J, Yingling C, American Society of Neurophysiological M. Intraoperative motor evoked potential monitoring – a position statement by the American Society of Neurophysiological Monitoring. Clin Neurophysiol. 2013;124:2291–316.

Malcharek MJ, Landgraf J, Hennig G, Sorge O, Aschermann J, Sablotzki A. Recordings of long-latency trigeminal somatosensory-evoked potentials in patients under general anaesthesia. Clin Neurophysiol. 2011;122:1048–54.

Markand ON, Lee BI, Warren C, Stoelting RK, King RD, Brown JW, et al. Effects of hypothermia on brainstem auditory evoked potentials in humans. Ann Neurol. 1987;22:507–13.

Matthies C, Raslan F, Schweitzer T, Hagen R, Roosen K, Reiners K. Facial motor evoked potentials in cerebellopontine angle surgery: technique, pitfalls and predictive value. Clin Neurol Neurosurg. 2011;113: 872–9.

Merton PA, Morton HB. Stimulation of the cerebral cortex in the intact human subject. Nature. 1980; 285(5762):227.

Minahan RE, Mandir AS. Neurophysiologic intraoperative monitoring of trigeminal and facial nerves. J Clin Neurophysiol. 2011;28:551–65.

Mishler ET, Smith PG. Technical aspects of intraoperative monitoring of lower cranial nerve function. Skull Base Surg. 1995;5:245–50.

Moller AR. Interaction between the blink reflex and the abnormal muscle response in patients with hemifacial spasm: results of intraoperative recordings. J Neurol Sci. 1991;101(1):114–23.

Moller AR. Intraoperative neurophysiologic monitoring. Am J Otol. 1995;16:115–7.

Moller AR. Intraoperative neurophysiological monitoring. 3rd ed. New York: Springer-Verlag New York Inc; 2011.

Moller AR, Jannetta PJ. Physiological abnormalities in hemifacial spasm studied during microvascular decompression operations. Exp Neurol. 1986;93:584–600.

Moller AR, Jannetta PJ. Monitoring facial EMG responses during microvascular decompression operations for hemifacial spasm. J Neurosurg. 1987;66:681–5.

Møller AR, Møller MB. Does intraoperative monitoring of auditory evoked potentials reduce incidence of hearing loss as a complication of microvascular decompression of cranial nerves? Neurosurgery. 1989;24(2):257–63.

Motoyama Y, Kawaguchi M, Yamada S, Nakagawa I, Nishimura F, Hironaka Y, Park YS, Hayashi H, Abe R, Nakase H. Evaluation of combined use of transcranial and direct cortical motor evoked potential monitoring during unruptured aneurysm surgery. Neurol Med Chir (Tokyo). 2011;51(1):15–22.

Mourisse J, Lerou J, Zwarts M, Booij L. Electromyographic assessment of blink reflexes correlates with a clinical scale of depth of sedation/anaesthesia and BIS during propofol administration. Acta Anaesthesiol Scand. 2004;48:1174–9.

Nelson KR, Vasconez HC. Nerve transection without neurotonic discharges during intraoperative electromyographic monitoring. Muscle Nerve. 1995;18(2): 236–8.

Oge AE, Yayla V, Demir GA, Eraksoy M. Excitability of facial nucleus and related brain-stem reflexes in hemifacial spasm, post-facial palsy synkinesis and facial myokymia. Clin Neurophysiol. 2005;116:1542–54.

Oikawa T, Matsumoto M, Sasaki T, Kodama N. Experimental study of medullary trigeminal evoked potentials: development of a new method of intraoperative monitoring of the medulla oblongata. J Neurosurg. 2000;93:68–76.

Onofrj M, Basciani M, Fulgente T, Bazzano S, Malatesta G, Curatola L. Maps of Somatosensory Evoked Potentials (SEPs) to mechanical (tapping) stimuli: comparison with P14, N20, P22, N30 of electrically elicited SEPs. Electroencephalogr Clin Neurophysiol. 1990;77:314–9.

Polo G, Fischer C, Sindou MP, Marneffe V. Brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm: intraoperative brainstem auditory evoked potential changes and warning values to prevent hearing loss--prospective study in a consecutive series of 84 patients. Neurosurgery. 2004;54(1):97–104; discussion 104–6.

Sala F, Manganotti P, Tramontano V, Bricolo A, Gerosa M. Monitoring of motor pathways during brain stem surgery: what we have achieved and what we still miss? Neurophysiol Clin. 2007;37(6):399–406.

Samii M, Gunther T, Iaconetta G, Muehling M, Vorkapic P, Samii A. Microvascular decompression to treat hemifacial spasm: long-term results for a consecutive series of 143 patients. Neurosurgery. 2002;50:712–8; discussion 718–9.

Sanes JN, Foss JA, Ison JR. Conditions that affect the thresholds of the components of the eyeblink reflex in humans. J Neurol Neurosurg Psychiatry. 1982;45(6): 543–9.

Sarnthein J, Hejrati N, Neidert MC, Huber AM, Krayenbühl N. Facial nerve motor evoked potentials during skull base surgery to monitor facial nerve function using the threshold-level method. Neurosurg Focus. 2013;34(3):E7.

Schlake HP, Goldbrunner R, Milewski C, Siebert M, Behr R, Riemann R, et al. Technical developments in intraoperative monitoring for the preservation of cranial motor nerves and hearing in skull base surgery. Neurol Res. 1999;21:11–24.

Schlake HP, Milewski C, Goldbrunner RH, Kindgen A, Riemann R, Helms J, et al. Combined intra-operative monitoring of hearing by means of auditory brainstem responses (ABR) and transtympanic electrocochleography (ECochG) during surgery of intra- and extrameatal acoustic neurinomas. Acta Neurochir (Wien). 2001;143:985–95; discussion 995–6.

Sekula Jr RF, Bhatia S, Frederickson AM, Jannetta PJ, Quigley MR, Small GA, Breisinger R. Utility of intraoperative electromyography in microvascular decompression for hemifacial spasm: a meta-analysis. Neurosurg Focus. 2009;27(4):E10.

Sindou MP. Microvascular decompression for primary hemifacial spasm. Importance of intraoperative neurophysiological monitoring. Acta Neurochir (Wien). 2005;147(10):1019–26; discussion 1026.

Singh R, Husain AM. Neurophysiologic intraoperative monitoring of the glossopharyngeal and vagus nerves. J Clin Neurophysiol. 2011;28:582–6.

Society ACN. Guideline 9C: guidelines on short-latency auditory evoked potentials. Am J Electroneurodiagnostic Technol. 2006;46(3):275–86.

Stechison MT, Kralick FJ. The trigeminal evoked potential: Part I. Long-latency responses in awake or anesthetized subjects. Neurosurgery. 1993;33:633–8.

Thirumala PD, Shah AC, Nikonow TN, Habeych ME, Balzer JR, Crammond DJ, et al. Microvascular decompression for hemifacial spasm: evaluating outcome prognosticators including the value of intraoperative lateral spread response monitoring and clinical characteristics in 293 patients. J Clin Neurophysiol. 2011;28:56–66.

Valls-Sole J, Tolosa ES. Blink reflex excitability cycle in hemifacial spasm. Neurology. 1989;39:1061–6.

Vriens JP, Pasman JW. Assessment of trigeminal nerve function by means of short-latency somatosensory evoked potentials after microneurosurgical repair. J Craniomaxillofac Surg. 1994;22:156–62.

Wang AD, Costa e Silva I, Symon L, Jewkes D. The effects of halothane on somatosensory and flash visual evoked potentials during operations. Neurol Res. 1985;7:58–62.

Wedekind C, Klug N. Nasal muscle F-wave for peri- and intraoperative diagnosis of facial nerve function. Electromyogr Clin Neurophysiol. 1998;38:481–90.

Wedekind C, Klug N. Assessment of facial nerve function in acoustic tumor disease by nasal muscle F waves and transcranial magnetic stimulation. Muscle Nerve. 2000;23:58–62.

Wedekind C, Klug N. Facial F wave recording: a novel and effective technique for extra- and intraoperative diagnosis of facial nerve function in acoustic tumor disease. Otolaryngol Head Neck Surg. 2003;129: 114–20.

Wedekind C, Stauten W, Klug N. A normative study on human facial F waves. Muscle Nerve. 2001;24: 900–4.

Yang M, Zheng X, Ying T, Zhu J, Zhang W, Yang X, et al. Combined intraoperative monitoring of abnormal muscle response and Z-L response for hemifacial spasm with tandem compression type. Acta Neurochir (Wien). 2014;156:1161–6; discussion 1166.

Ying TT, Li ST, Zhong J, Li XY, Wang XH, Zhu J. The value of abnormal muscle response monitoring during microvascular decompression surgery for hemifacial spasm. Int J Surg. 2011;9:347–51.

Ying T, Thirumala P, Chang Y, Habeych M, Crammond D, Balzer J. Emprical factors associated with Brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm and its correlation to hearing loss. Acta Neurochir (Wien). 2014;156:571–5.

Zheng X, Hong W, Tang Y, Ying T, Wu Z, Shang M, et al. Discovery of a new waveform for intraoperative monitoring of hemifacial spasms. Acta Neurochir (Wien). 2012;154:799–805.

Zouridakis G, Papanicolaou AC. A concise guide to intraoperative monitoring. Hoboken: CRC Press; 2000.

Outcome Evaluation and Postoperative Management

13

Akinori Kondo

Abstract

Objective To obtain objective surgical results from microvascular decompression (MVD) for trigeminal neuralgia (TN) and hemifacial spasm (HFS), a method to evaluate and analyze overall postoperative results from MVD by combining the cure rate of symptoms with the complication rate is proposed. In addition, tactics to prevent surgical complications and of the postoperative management are described.

A new standardized scoring system using consistent criteria to document treatment results of MVD is needed to allow individual surgeons to correlate and compare results with other institutes using the common criteria.

Method and Findings Surgical results were obtained from a questionnaire sent to 233 patients who had undergone surgery and had been followed up for more than 1 year after surgery (TN patients, 95; HFS patients, 138). When surgical outcome is complete cure of symptoms, the efficacy of surgery (E) is designated E-0, but when moderate symptoms are still persist postoperatively, the score is designated E-2. When no complications are reported after surgery, the complication score (C) is C-0; if troublesome complications remain, the score is designated as C-2.

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Total evaluation of the results (T) is judged by combining the (E) and (C) scores. For example, when E is 0, and C is C-2, the total evaluation is scored as T-2, which is diagnosed as fair. Analysis of the collected data revealed an outcome of T-0 was 70 % (59/85 patients), T-1 was 19 % (16/85), T-2 was 8 % (7/85), T-3 was 1 % (1/85), and T- was 2 % (2/85) in TN, whereas in HFS, T-0 was 61 % (62/102), T-1 was 28 % (29/102), T-2 was 7 % (7/102), and T-3 was 4 % (4/102).

Conclusion The total results of MVD should be evaluated and analyzed by combining the cure rate of symptoms together with the complication rate. Successful MVD involves not only to obtain good cure rate but also minimizing surgical complications. Therefore, to realize various methods how to avoid complications and to study postoperative management is mandatory. This new scoring system could allow much more objective analysis of the results of following MVD, and by adopting this scoring system, surgeons can compare their own overall surgical results with those of other institutes.

Keywords

Microvascular decompression • Trigeminal neuralgia • Hemifacial spasm • New scoring system • Postoperative management

List of Abbreviation

ABR	Auditory brainstem response
BNI	Barrow Neurological Institute
CSF	Cerebrospinal fluid
HFS	Hemifacial spasm
MVD	Microvascular decompression
TN	Trigeminal neuralgia
VBA	Vertebrobasilar artery

13.1 Introduction

It is well known that microvascular decompression (MVD) is a safe and definitive treatment for trigeminal neuralgia (TN) and hemifacial spasm (HFS) with proven long-term efficacy (Barker et al. 1955, 1966; Henson et al. 2005; Jannetta 1977; Kalkanis et al. 2003; Little et al. 2008; McLaughlin et al. 1999; Rogers et al. 2000). However, surgical results are not always reliable, if surgery is not appropriately performed. Since MVD is a functional neurosurgery, satisfactory results should entail a complete and permanent cure of symptoms without any postoperative sequelae, such as complications or recurrence of symptoms. To realize various tactics to prevent complications and to study postoperative management are, therefore, mandatory. The results of MVD should thus be evaluated and analyzed objectively by combining the cure rate of symptoms with complication rates to score the overall treatment results using consistent criteria in a standardized manner.

13.2 Methods and Materials

The questionnaires were sent to 233 patients (TN patients 95, HFS patients 138) who had undergone surgery for TN or HFS within the last 7 years (2006–2012) and had been followed up more than 1 year after surgery, and an evaluation of the overall results of MVD was performed more than 1 year after surgery. Total response rate was 80.3 % (187/233 patients), TN patients, 89.5 % (85/95), and HFS patients, 73.9 % (102/138), respectively.

13.2.1 Evaluation of MVD Results for TN

Evaluation of postoperative pain (E)

E-0: Completely pain-free
E-1: Occasional slight pain, self-controllable, without medication
E-2: Moderate pain, controllable by medication
E-3: Persistent pain, not controllable by medication, not cured

Evaluation of complications (C)

C-0: No deficits, or only slight subjective complaints
C-1: Slight cranial nerve or cerebellar dysfunction, not bothersome for daily life
C-2: Both subjective and objective cranial nerve or cerebellar dysfunction, problematic for daily life

Total evaluation grade of the results (T)

(T) is determined as the sum of the (E) and (C) grades. For example, when the postoperative pain grade is E-0 and complication grade is C-0, the total evaluation is graded as T-0, representing an excellent result. If the postoperative pain grade is E-0, but postoperative complication grade is C-2, then the total evaluation is graded as T-2, representing a fair result. Total evaluation of results (T) and clinical outcome of TN
T-0: Excellent, T-1: Good, T-2: Fair, T-3 to T-5: Poor

13.2.2 Evaluation of MVD Results for HFS

Evaluation of postoperative grade of involuntary movement (E)

E-0: Complete disappearance of spasm
E-1: Occasional slight spasm
E-2: Moderate spasm, apparently persisting
E-3: Not cured

Evaluation of postoperative complications (C)

The same method used to judge TN is applied to evaluate postoperative results of HFS. Total evaluation of surgical results (T) and clinical outcome of HFS: The same method used to judge TN is applied.

13.2.2.1 Overall Surgical Results

Overall data were evaluated and analyzed using the new scoring system. Analysis of the collected data revealed an outcome of T-0 was 70 % (59/85 patients), T-1 was 19 % (16/85), T-2 was 8 % (7/85), T-3 was 1 % (1/85), and T-4 was 2 % (2/85) in TN, whereas in HFS, T-0 was 61 % (62/102), T-1 was 28 % (29/102), T-2 was 7 % (7/102), and T-3 was 4 % (4/102) (Fig. 13.1).

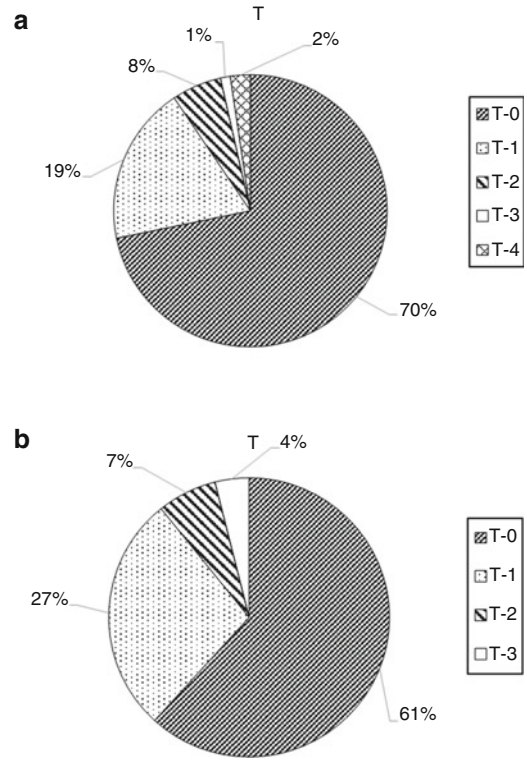

Fig. 13.1 (**a**) TN cases. (**b**) HFS cases. The percentages of patients judged by the new scoring system, documented using both cure rate and surgical complication rate. Total evaluation rate (T = E + C) is shown as T-0 to T-3 (Reprint with permission from Kondo et al. (2012))

13.3 Prevention of Complications and Postoperative Management

13.3.1 Postoperative Cerebellar Edema

Postoperative cerebellar edema, which is commonly induced by either inappropriate retraction of the cerebellum or intraoperative traumatization of superior petrosal veins, is the most severe and potentially fatal surgical complication in MVD. Cerebrospinal fluid (CSF) should be suctioned as much as possible by full opening of cerebellar cisterns. Cerebellar retraction should be as gentle as possible, and duration of every each retraction time should preferably be limited less than 5 min. A suction tube is preferably used intermittently instead of retractors to avoid overly strong retraction. To prevent accidental laceration of petrosal veins, the arachnoid membranes covering the veins should be cut meticulously to allow vessels movable, and the outlet (draining site) of veins to the major sinus should be reinforced using fibrin glue.

Once acute postoperative cerebellar edema is encountered, its therapy and management should be urgent and appropriate. Either ventricular drainage or urgent posterior fossa decompression surgery is needed for severe cerebellar edema.

13.3.2 Postoperative Dysfunctions of Cranial Nerves

Cranial nerve dysfunction that is not rarely seen postoperatively is cochlear and/or vestibular dysfunction. During MVD, the direction of traction should be perpendicular to the axis of the acoustic nerve. Monitoring of auditory brainstem response (ABR) is inevitable. When judging the data of ABR, clinicians should keep in mind that warning sign of adverse changes of ABR during surgery is a >1 ms delay of the latency of the 5th wave and 40 % reduction of amplitude at most.

To prevent postoperative facial nerve hypofunction, care should be taken when handling an offending artery, mostly anterior inferior cerebellar artery, with small perforators. Since hypofunction of abducent nerve can be caused particularly when handling the vertebrobasilar artery (VBA), care should be taken not to compromise the nerve by too much amount of prosthesis when the VBA is replaced. In terms of lower cranial nerve hypofunction, as these nerves are also vulnerable, impairment is mostly caused by excessive manipulation of the nerves or heat by a bipolar coagulator during the procedure. A bipolar coagulator should be carefully used near the nerves by covering the nerves with a wet cottonoid for protection.

13.3.3 Postoperative Liquorrhea

Postoperative liquorrhea is also a very troublesome complication after MVD. For prevention, watertight closure of dura mater should be achieved using fascia or muscle piece, if necessary. The opening of the mastoid air cell can be packed and closed as tight as possible with the use of bone wax together with a section of muscle piece, bone dusts, and fibrin glue. If the opening of bony defect is ignored during procedure, postoperative CSF leak is inevitable, and its repair will be much more troublesome after surgery.

13.4 Discussion

In experienced hands, MVD for TN and HFS offers the highest likelihood of long-term successful cure of cranial nerve dysfunction symptoms along with low rates of morbidity and recurrence (Barker et al. 1966; Kalkanis et al. 2003). The objective of the present analysis of overall surgical results is to set quality criteria and standards for outcome reports following MVD to identify and assess the surgical results appropriately. The same criteria obtained from standardized method should be used to compare

each data of treatment results from MVD at each neurosurgical institute (Kondo et al. 2012). And also surgeons should well realize and study the various methods how to lessen surgical complications, together with postoperative management of MVD (Taha and Tew 1966).

Little et al. (2008) described the pain response and quality of life in patients with TN treated using gamma knife surgery by assessing outcomes using the Barrow Neurological Institute (BNI) Pain Intensity Score and Brief Score Inventory. With that score, post-gamma knife treatment status was clearly analyzed and documented from score I (no pain) to V (no relief). Rogers et al. (2000) assessed not only the efficacy of gamma knife radiosurgery for TN but also complications. They presented a BNI pain scoring system and BNI facial numbness score, which was classified after treatment into four scores. Henson et al. (2005) also described treatment results such as pain response, pain recurrence, and sensory neuropathy after both glycerol rhizotomy and gamma knife treatment for TN. Although they evaluated rates of pain relief or complications, they did not combine both rates to judge overall results. Accurate diagnosis of chronic pain is, however, crucial for determining the efficacy of surgical therapy, as measurement of such subjective phenomena is difficult. Although Chen et al. recently proposed an overview of psychometric testing, such methods of evaluating surgical results for TN are too complicated to be useful in daily clinical situations (Chen and Lee 2010). Our grading system of classification is not made too detailed but is instead simplified as much as possible. On the other hand, judgment of postoperative status of HFS is relatively straight forward, as the symptoms are mainly objective in nature.

According to the report by Kondo (1997), symptom recurrence mostly occurs 3–6 years after disappearance of initial symptoms after MVD. However, the definition of the term "recurrence" has not been clearly defined and remains controversial. A factor that may lead to errors in judging outcomes is that the course of postoperative recovery for cranial nerve symptoms is variable. Based on daily clinical experience, the definition of surgical results should allow a postoperative period of more than 12 months to lapse. The immediate postoperative period is therefore too soon to judge whether the patient has been cured. The other problem in judging postoperative symptoms is that the perception of follow-up results for MVD sometimes much differed between doctors and patients, concerning cure rate and complication rate. When analyzing data obtained from questionnaire, the opinion of the patient should typically hold sway when a difference is seen. Jannetta once cautioned that when diagnosing patient symptoms, a common error is "the assumption that patients you have not heard from are doing well (personal communication).

The overall results obtained from this study were not as satisfactory as had been expected. Because with the present method, the score should be defined either one or two ranks downgraded when postoperative incomplete cure symptoms or surgical complications are encountered, even if the initial symptoms are successfully resolved.

But the total results of MVD should be evaluated and analyzed by combining the cure rate of symptoms together with the complication rate. And to perform successful MVD, it is necessary not only to obtain good cure rate but also to minimize surgical complications

Although a scoring system of the results of MVD for TN has been more documented, the trial for HFS is quite a few. Differences are known to exist between Caucasians and Asians in terms of the incidence of cranial nerve dysfunction symptoms; TN is much more frequent in Caucasians, while HFS is much more frequent in Asians (Jannetta 1977). It is assumed that difference in the shape and volume of the posterior cranial fossa may play a role (Yamamoto et al. 1987).

Steps must be taken to allow standardized analysis of results to become much more objective, and standardization of surgical results would also allow individual surgeons to correlate and compare results with other institutes using the common criteria.

References

Barker II FG, et al. Microvascular decompression for hemifacial spasm. J Neurosurg. 1955;82:201–10.

Barker II FG, et al. The long-term outcome of microvascular decompression for trigeminal neuralgia. N Engl J Med. 1966;334:1077–83.

Chen HI, Lee JY. The measurement of pain in patients with trigeminal neuralgia. Clin Neurosurg. 2010;57:129–31.

Henson CF, et al. Glycerol rhizotomy versus gamma knife radiosurgery for the treatment of trigeminal neuralgia; an analysis of patients treated at one institution. Int J Radiat Oncol Biol Phys. 2005;63(1):82–90.

Jannetta PJ. Observations on the etiology of trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction and glossopharyngeal neuralgia: definitive microsurgical treatment and results in 117 patients. Neurochirurgie. 1977;20:145–54.

Kalkanis SN, et al. Microvascular decompression surgery in the United States, 1966 to 2000: mortality rates, morbidity rates, and the effects of hospital and surgeon volumes. Neurosurgery. 2003;52(6):1251–62.

Kondo A. Follow-up results of microvascular decompression in trigeminal neuralgia and hemifacial spasm. Neurosurgery. 1997;40(1):46–52.

Kondo A, et al. A proposal for standardized analysis of the results of microvascular decompression for trigeminal neuralgia and hemifacial spasm. Acta Neurochir. 2012;154:773–8.

Little AS, et al. Long–term pain response and quality of life in patients with typical trigeminal neuralgia treated with Gamma knife stereotactic radiosurgery. Neurosurgery. 2008;63(5):915–24.

McLaughlin MR, et al. Microvascular decompression of cranial nerves: lessons learned after 4400 operations. J Neurosurg. 1999;90:1–8.

Rogers CL, et al. Gamma knife radiosurgery for trigeminal neuralgia; the initial experience of The Barrow Neurological Institute. Int J Radiat Oncol Biol Phys. 2000;47(4):1013–9.

Taha JM, Tew Jr JM. Comparison of surgical treatment for trigeminal neuralgia: reevaluation of radiofrequency rhizotomy. Neurosurgery. 1996;38(5):865–71.

Yamamoto Y, et al. Measurement and clinical significance of the posterior fossa volume of patients with hemifacial spasm. No Shinkei Geka. 1987;15:243–8.

Reoperation for Hemifacial Spasm

14

Shi-Ting Li and Xue-Sheng Zheng

Abstract

Despite the high success rate of microvascular decompression procedures for hemifacial spasm and trigeminal neuralgia, failure and recurrence are unavoidable. If the spasm or pain does not resolve after surgery and the surgeon thinks perhaps some culprit vessels were neglected in the first operation, early reoperation is indicated. If the symptoms persist for a long period after surgery and delayed resolution does not take place, late reoperation should be performed. Besides, late reoperation is indicated for recurrence. The uncertainty of "delayed resolution" and the high risk of complications of the reoperation make the decision very difficult.

Keywords

Reoperation • Hemifacial spasm • Trigeminal neuralgia

A "failed" patient is a signal that we are not perfect and that the forces of nature have again outwitted us. We cannot hide these failures, avoid them, or ignore them. Rather, we can learn from them and, frequently, can make the patients feel better or even cure them.

PETER J. JANNETTA, 1985

14.1 Introduction

Many papers have been published regarding the high efficacy of MVD for HFS, with immediate cure rates ranging from about 82 to 92 %. Those patients who are not immediately cured following MVD are likely to be cured in 1–3 years without additional operations, namely, delayed resolution (Ishikawa et al. 2001). In contrast, some of the patients who are cured by MVD may gradually recur in several months or years (Chung et al. 2001). Patients with persistent or recurrent spasms have three options: live with their symptoms, take Botox therapy, or undergo repeat MVD. As Engh et al. proposed (2005), there are four kinds of situations after the original MVD procedure.

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The first kind consisted of those patients who had neurophysiological monitoring abnormalities, for example, an abnormal BAEP alert during their first MVD, so the MVD procedure was aborted to preserve the neural function. For this kind, reoperation is surely needed after a short-time rest. The second kind of patients still had significant spasm after their first MVD surgery or a significant recurrence in the immediate postoperative period. These patients should undergo early reoperation. The third kind of patients had recurrence of spasm more than 1 month after their first MVD. The fourth kind of patients had already failed at least two MVD operations, required a third or fourth MVD. The latter two kinds of patients undergo late reoperation.

Although the abovementioned principle seems clear and simple, the practice is not easy at all, because success is not guaranteed for the second, third, or even more operations, but operative complications are indeed more frequent after the reoperation than the first MVD (Li et al. 2010). While the prospect of possible "delayed resolution" add the surgeon's hesitation, the fact that late reoperation is less effective and more dangerous than early reoperation, however, encourages the surgeon to react earlier. As a result, the issues as to if and when the reoperation should be performed for persistent spasm have been under debate for long time.

14.2 Evaluation of the First MVD and the Decision of an Early Reoperation

Early reoperation means repeated MVD procedure within 1 month of the first MVD.

For experienced neurosurgeons, spasm resolution rates immediately after MVD are about 90–94 %, in other words, the fail rate is about 6–10 % (Engh et al. 2005). It's very difficult to persuade the patient that the hemifacial spasm may automatically resolve after discharge, especially when the spasm was not relieved at all or even worsened after the first surgery.

It's still controversial concerning about the treatment of such kind of patients. For example, Hyun et al. (Oh et al. 2008; Hyun et al. 2010) believed that any decision regarding retreatments should be drawn after postoperative 12 months or later, no matter how the outcomes of first MVD is. In contrast, Li (2005) reported that a second operation was indicated for patients with significant persisting or even worsening HFS after the first MVD; and if the spasm became less severe and less frequent after MVD, it's better to wait for delayed resolution. Engh et al. (2005) reported that the patients with significant postoperative spasm undergo facial EMG; and if the postoperative abnormal muscle response (AMR) is still significant, they usually offered re-exploration, provided that the patient is well aware of the high risk of operative complications; otherwise, the patient is not offered a repeat MVD. Li et al. (2010) also considered AMR as an important reference to make the decision of early reoperation. It was observed that the long-term clinical outcomes of the intraoperative AMR disappearance before and after MVD were correlated. Thus, AMR may be considered an objective factor in the indication for reoperation (Ying et al. 2011).

Hughes et al. (2015) reported that re-imaging with high-resolution T2-weighted MRI will identify the missing culprit vessel after the failed MVD for hemifacial spasm. The imaging interpretation was concordant with the surgical results regarding artery versus vein in 86 % of cases and regarding the segment of the nerve contacted in 92 %. They found the unaddressed vascular compression is typically proximal to the previously placed Teflon felts.

Li Shi-ting team divided the facial nerve root into 4 zones (Zhong et al. 2010) and insisted that all zones should be explored carefully and decompressed adequately (Zhong et al. 2011). Otherwise, multiple neurovascular conflicts might be neglected and lead to failure. Especially, arteriole compression in zone 1 and "cross-type" AICA compression in zone 4 are the typical types of missing culprits (Li et al. 2010, 2013; Zheng et al. 2011).

In fact, the different percentages of delayed relief underlie this controversy. However, the delayed relief rate reported in literatures ranges from 90 % (Ishikawa et al. 2001) to <4 % (Zhong et al. 2015). The authors advocating early reoperation generally reported very low percentage of

delay relief. And the authors reporting high percentage of delay resolution would accordingly recommend waiting. Therefore, unless an agreement as to the exact delayed relief percentage is reached in the future by multicenter prospective clinic trial, the debate will continue.

The different answers to if early reoperation should be performed reflect the surgeons' different belief of the mechanism of hemifacial spasm. According to Moller et al., primary HFS may be due to hyperactivity of the facial nucleus, progressively induced by the continuous pulsation of the neurovascular conflict. If follows logically that the effect of surgical decompression takes time to decrease and normalize the clinical spasm, and the delayed spasm resolution may be due to the time required for remyelination of the compressed axons, as well as the recovery of normal excitability of the facial nucleus (Moller and Jannetta 1984, 1985a, b). Of course, a surgeon believing this hyperactivity hypothesis will firmly insist on the "waiting" principle. The hypothesis is not consistence with the fact that in most patients AMR vanished immediately when the compressing vessel was detached. It's also challenged by Park's report (Kim et al. 2009) that the severe indentation on the facial nerve were associated with good surgical outcomes, because the deduction of this hypothesis is that the more severe the compression is, the more hyperactivity there should be in the nucleus. In contrast, Zheng et al. hypothesized that the cross-transmission between the facial nerve fibers is bridged by sympathetic nerve fibers on the offending artery wall (Zheng et al. 2012a, b). Hence, this might explain most of patients who were relieved immediately after facial nerve and offending vessels wall were separated by Teflon. And they believe that the reasons for failure may include missing the multiple vessels, Teflon pled get movement after closure, or incomplete decompression. Accordingly, a neurosurgeon who believes the sympathetic bridge hypothesis tends to do early reoperation to find the missing offending vessels or the inadequate decompression. However, neither hypothesis has been fully proven by substantial direct evidence. Probably, some patients are of hyperactivity, but the others are mediated by sympathetic bridge. Thus, the debate as to whether early reoperation is necessary will continue until the mechanism of hemifacial spasm is clearly addressed.

Based on the literatures mentioned above and our own experience, here we propose indications for early reoperation.

1. The spasm did not resolve at all or get even worse.
2. Review the surgery video and find that four zones were not explored fully; especially if the zone-1 or the zone-4 was not dissected in the first MVD, reoperation should be recommended.
3. Review the surgery video and find that some arteriole in contact with facial nerve root was not detached in the first MVD.
4. AMR persisted after final nerve decompression, or AMR disappeared before the beginning of decompression, or AMR reemerge after the operation. AMR is positive at the postoperative day 2.
5. Postoperative high-resolution T2-weighted MRI identifies one or more vessels still in contact with the facial nerve.

If the above listed five criteria are matched, an early reoperation is recommended.

On the contrary, if there is initial relief after surgery and then subsequent recurrence, the decision to reoperate should be considered very carefully (Park et al. 2006).

If the first MVD surgery was aborted because of neuroelectrophysiological monitoring alert, a reoperation should be scheduled within 1 week.

14.3 Early Reoperation for Fail or Abortion

As described in our previously reported papers (Zhong et al. 2010), the early reoperation was performed via the original approach. Craniectomy is very easy by removing sutures and titanium wire mesh. There is no or very little adhesion.

If arachnoid membrane is not dissected adequately, additional dissection is needed in reoperation. The dissection was started from the caudal cranial nerves. While the arachnoid membrane between facial nerve and the caudal cranial nerves being opened fully, the cerebellum as well

as flocculus was gently raised until the pontomedullary sulcus was exposed.

For reoperation, the previously decompressed site was double checked at first to confirm that a satisfied decompression was completed. Then the full course of facial nerve was inspected through different angles. Exploration began from zone 2 and then moved to zones 3, 1, and 4 (zone 1, where the nerve root emerges to the brainstem surface from the parenchymal and goes through the pontomedullary sulcus; zone 2, where the nerve root attaches to the surface of the pons; zone 3, where it gradually detached from the brain stem; zone 4, where the nerve runs free in the subarachnoid cistern and extends to the internal meatus).

Any arteries or big veins in contact with the facial nerve were detached. After the offending vessel was moved away from the nerve, pieces of Teflon sponge were gently placed between the vessel and the brainstem, so as to transpose the course of the vessel. However, when transposition is impossible, Teflon felt can be inserted into the gap between the nerve and the vessel. The suspicious venulein contact with the nerve was coagulated with low power.

According to Hughes MRI (Hughes et al. 2015) study and Zhong clinical observation (Zhong et al. 2015), the missing culprit vessels usually lie proximal to the previous Teflon felt, especially at zone 1. Therefore, early reoperation should be mainly focused on this portion; sometimes, endoscope may help find small vessels in the pontomedullary sulcus.

With the aid of intraoperative AMR and ZLR monitoring, we found a new type of vascular compression pattern, the cross type, which involves the AICA passing through the gap between cranial nerves VII and VIII and compressing the cisternal portion of facial nerve, that is, zone 4. For this type, the treatment is to interpose Teflon felt between the facial nerve and AICA (Li et al. 2013; Zheng et al. 2011).

If new vascular compression was not found in reoperation after careful scrutinizing, it's very likely that the shape and tension of Teflon felt are not suitable. The old Teflon felt should be replaced by a new one (Li et al. 2010). If the Teflon felts previously inserted should be pulled out, great caution should be paid as pulling large sponges may break some perforating arterioles.

In the early reoperation, some small perforator arteries turned out to be the culprit vessels, which were hidden beneath the big artery. To this subset, determination of the culprit vessels in the reoperation was completely relied on the intraoperative AMR monitoring. These small vessels looked just ordinary, and did not seem "compressing" in appearance; however, when they were lifted with a nerve dissector, the AMR disappeared in no time. After these small perforator arteries were decompressed by small piece of Teflon sponges, these patients were cured (Li et al. 2010).

There are higher risks of operative complications related to the early second MVD (Li et al. 2010). Of course, there is nothing with adhesion. When performing the early reoperation, the surgeon is under great psychological stress and always tries his best to find additional culprit vessels; thus, the entire course of facial nerve will be explored over and over. We think this is the main reason for high risk of early reoperation (Li et al. 2010). Therefore, each step should be performed carefully. For example, the dissection of the arachnoid membrane, exploration of the facial nerve, detachment of the offending vessel, and insertion of the Teflon sponge all should be done gently under direct vision. While dissecting the arachnoids around the nerve, the feeding arterioles for the nerve should be preserved. While mobilizing the offending artery, retraction of the nerve should be avoided. When vasospasm was observed, the operation should be paused and sometimes narceine was administered on the vessel surface. When the coagulation of the small vein is strongly required, it should be kept far away from the nerve. Meanwhile, real-time neuroelectrophysiological monitoring of the nerve function is mandatory (Zhong et al. 2010).

14.4 Follow-Up of Delayed Resolution and Recurrence and the Decision of a Late Reoperation

If a patient achieved no or little improvement after the first MVD, but did not undergo early reoperation, he should be closely followed up with the focus of possible delayed resolution. Most delayed resolution would take place within 1 year, so reoperation is indicated if persistent spasm did not disappear or

diminish after 1-year postoperative follow-up. However, if severe compression indentation on the facial nerve root had been found in the first MVD, the possibility of delayed resolution is much higher, and the decision of reoperation should be postponed 3 years after the first MVD (Oh et al. 2008; Park et al. 2006). It's worth of emphasis that patients undergoing early reoperation were significantly more likely to be cured or improved than patients undergoing late reoperation.

Those patients cured by the first MVD should be followed up too, because there is a chance of spasm recurrence in these subsets. Recurrence is defined as reemerging facial spasm after a period (>1 months) of complete resolution after MVD. The mean incidence of recurrence is 7.0 % (range, 1.1–55.5 %); reported recurrences occurred 1–60 months after surgery. Among all patients initially cured and in cases in which the timing of recurrence was given, only 1.0 % had a recurrence after 24 months; therefore, patients who are symptom-free 2 years after MVD can be considered cured. Unlike controversy as to delayed resolution, it's now generally accepted that recurrence means failure of the first operation or newly developed vascular compression to the facial nerve, thus, reoperation is indicated as soon as recurrence emerge (Payner and Tew 1996).

The causes of the failed first MVD include misjudgment of offenders, improper selection, improper insertion, or inadequate Teflon volume. Li et al. (2010) reported one case of late reoperation for recurrence; he noted dense adhesion of the Teflon felt to the offending vessel and the facial nerve; the patient was cured through the revised MVD. Arterial hypertension contributes to late recurrence (Oliveira et al. 1999), and there was a trend toward an association of recurrence with coexistence of male gender, vein or VA offender, and experience of transient facial weakness.

14.5 Late Reoperation for Fail and Recurrence

The procedures of late reoperation are similar to that of the early reoperation. The abovementioned principles such as full-course exploration, adequate decompression, thorough sharp dissection, and caudal-to-rostral exploration should be followed as well. However, there are many other features and difficulties in late reoperation:

1. The craniectomy should be enlarged, because the bone tissues regenerate and gradually occupy the margin of the hole. Otherwise, intracranial exposure will be very difficult.
2. There is adhesion between the dura and the cerebellum, which should be dissected sharply and sufficiently, so as to produce enough operating space.
3. There is adhesion between the cranial nerves and the cerebellum and the brainstem; the adhesion should be cut sharply. Pull of cranial nerves and blunt dissection should be avoided.
4. There is severe adhesion among Teflon graft and the cranial nerves and the vessels. Pulling out the old Teflon is generally impossible and sometimes dangerous. If necessary, a small part of these Teflon sponges could be cut with microscissors to make a space for new grafts, and this kind of process is dangerous too. Sometimes, the Teflon bolus itself may be the culprit of spasm recurrence.
5. Due to extensive adhesion, full-course exploration is usually very difficult, sometimes impossible.

As a result, late reoperation is much less effective and with higher complications than early reoperation. Engh et al. (2005) reported that the effective rate (excellent plus good relief) of late reoperation is only about 75 %, while the effective rate of the early reoperation by the same team is almost 100 %. Moreover, in the late reoperation grout, some spasm relief may result from facial nerve injury rather than nerve decompression. This is the main reason that some surgeons preferred early reoperation.

14.6 Reoperation for Trigeminal Neuralgia

14.6.1 Early Reoperation for Failed MVD

Unlike in hemifacial spasm, MVD procedures for trigeminal neuralgia seldom disturb the VIII cranial nerve, and abortion due to intraoperative

electrophysiology monitoring alert rarely takes place. Therefore, reoperation for aborted MVD of trigeminal neuralgia will not be discussed here.

Immediate postoperative pain relief is reportedly obtained in up to 98 % of cases after MVD, i.e., only a very small subset of patients with trigeminal neuralgia failed the first MVD (Kondo 1997). "Delayed relief," which is very common in hemifacial spasm, however, is rare in trigeminal spasm; so the surgeons do not need to be entangled in the difficult choice of waiting or acting right now. What they have to choose is the dilemma: should these failed patients undergo less invasive procedures, such as gamma-knife and radiofrequency trigeminal rhizotomy, which are associated with a higher risk for sensory loss, or should they undergo repeat MVD in the hope of relieving their facial pain (van Loveren et al. 1982; Zhang et al. 2011)?

Here we propose some advice as to if choosing to do a repeat MVD:

1. If the whole course of the intracranial trigeminal root was not fully exposed in the first MVD, a repeat MVD is indicated.
2. If a vein was found in contact with the V CN, but neglected to treat in the first MVD, a repeat MVD is indicated, because veins as a culprit vessel are much more common in trigeminal neuralgia than in hemifacial spasm (Hong et al. 2011).
3. If a patient was initially cured by MVD but recur within several days, please review the video of the first MVD, and if the Teflon interpositor is small, a repeat MVD is indicated, as interpositor displacement may cause a very acute recurrence (Amador and Pollock 2008).
4. If the first MVD was performed by a less experienced surgeon and surgical video is unavailable, a repeat MVD is indicated, as sometimes the VIII and VII cranial nerves may be mistaken as the trigeminal sensory and motor roots (Amador and Pollock 2008).
5. Otherwise, if the first MVD is regarded as proper, a negative re-exploration is very likely to take place, it's better to choose less invasive procedures. As Amador reported, when no addition neurovascular conflict was found in repeat MVD, the surgeon usually performed a partial nerve section, which leads to comparable sensory loss than that from gamma-knife and radiofrequency trigeminal rhizotomy (Amador and Pollock 2008).

Techniques for early repeat MVD: The previous incision is used. If necessary, the craniectomy should be enlarged to the edge for the transverse sinus and sigmoid sinus. If arachnoid membrane is not opened sufficiently, additional dissection is needed in reoperation. Sometimes, one or more giant veins attach firmly to the cerebellar surface and drain to the superior petrosal vein, making it very difficult to expose the REZ of trigeminal nerve. The cerebellar fissure approach is useful in this situation (Zhu et al. 2014). The arachnoid membranes covering the petrosal fissure and superior cerebellopontine fissures were dissected sharply. After these fissures were dissected, a pyramid-shaped space was opened, with the bottom triangle facing the REZ of trigeminal nerve root. The large veins are the lateral edge of this triangle. We could perform decompression procedure of the REZ though this pyramid-shaped space, while the major venous drainage was preserved. In reoperation, the previously inserted Teflon felt was checked at first in order to confirm if it had moved or if the decompression was sufficient. Afterward the whole course of the V cranial nerve was inspected thoroughly. The nerve root was divided into five zones. Zone 1–4 are the rostral, caudal, medial, and lateral aspects of the REZ; and zone 5 is the free cisternal portion. In reoperation, more attention should be paid to the following vascular compression.

1. Intraneural vessels: small vein can be coagulated and cut; big vein or artery should be wrapped by thin pieces of Teflon felt. In our experience, vessel in contact with the motor root may also cause pain, since there are some small communications between the sensory and motor roots (Zheng et al. 2012c).
2. Vessels in zone 3 tend to be neglected in the first MVD, because this area cannot be seen directly. The nerve should be pulled gently

rostrally and then caudally to expose zone 3, and Teflon interpositor inserted from one side should be viewable from the other side.
3. Small or medium vein located at zone 4 can be exposed via the cerebellar fissure approach; otherwise, it may hide in the cerebellopontine fissure (Zhu et al. 2014).
4. Vessels near or in the Meckel's cave.

Small compressing veins can be coagulated and cut. Arteries or large veins should be interposed with Teflon felts. Sometimes, the vein is attached firmly to the nerve root and brainstem, and mobilizing is impossible and dangerous. In this situation, we can gently dissect a small gap between the vein and the nerve root using a fine dissector; then small pieces of wet gelatin sponge were filled into the gap one by one, so as to expand the gap. When the vein was completely detached from nerve, a piece of Teflon sponge was interposed into this gap.

If the re-exploration is negative, a partial nerve section is indicated, according to the distribution of affected branches (Amador and Pollock 2008).

14.6.2 Late Reoperation for Persist or Recurrent Trigeminal Neuralgia

For those patients cured by the MVD, the rate of recurrence of facial pain was 6–30 % in follow-up studies, with the annual rate of recurrence between 1 and 3.5 % (Sun et al. 1994). TN involving more than two branches of distribution tends to have recurrences, probably due to the broader sensitive area over the root entry zone in those cases involving multiple branches, so that the chance of recurrence was higher.

The most frequent causes of recurrence of symptoms are incomplete decompression, migration of the inserted Teflon felt, adhesion, or fibrosis between the offending artery and the nerve. Venous compression is an important factor that contributes to recurrence of trigeminal neuralgia (Lee et al. 2000). In addition, Teflon compression is a common course of recurrent trigeminal neuralgia. Teflon felt may cause severe adhesion, and granulomatous formations. The time necessary for Teflon felt to cause a fibrotic change ranges from 10 months to 5 years. Almost 25 % of recurrent TN patients had significant compression resulting from excessive Teflon placement at their first MVD. Teflon seems to have the tendency to expand and adhere to the tentorium rather than the brainstem over time, interpreting that Teflon granule compression is more common in TN than in HFS (Chen et al. 2000).

Amador reported surgical findings of 29 cases of repeat MVD (Amador and Pollock 2008). Compression of the nerve was found in 24 patients (83 %) by an artery (13 patients, 45 %), vein (4 patients, 14 %), or Teflon (7 patients, 24 %). Four patients (14 %) who underwent operations in other hospital had incorrect cranial nerves decompressed at their first surgery. The surgical outcomes of repeat MVD are comparable with percutaneous needle-based techniques and stereotactic radiosurgery. Therefore, they supposed that patients with persistent or recurrent TN should be considered for repeat MVD. Lee suggested that MVD remains the optimal treatment for the recurrent TN attributable to vein regrowth. Barker stated that repeat MVD is a feasible therapeutic option with good chances of success, even in patients who have undergone neurodestructive procedures. Complication rates, particularly facial numbness, can be avoided if only a limited neurolysis is performed (Bakker et al. 2014).

In contrast, some authors declared that patients with persist or recurrent TN should be firstly recommended to undergo less invasive procedures to achieve pain relief for two reasons. For one reason, the success rate of repeat MVD is significantly lower than primary operations. Complete pain relief at 10 years in 64 % of patients after their initial MVD compared with 42 % after undergoing a second operation. Secondly, the risk of facial numbness is greater for repeat MVD. Surgeons frequently have to perform a partial nerve section at the time of repeat MVD if there was no compelling compressive lesion. Thus, the chance of trigeminal injury is similar for repeat MVD compared with destructive procedures. As a result, the factor of preserving sensory function that makes MVD the best option as

a primary procedure for idiopathic TN is largely negated when discussing the results of repeat MVD (Zhang et al. 2011).

To sum up, our opinion is that repeat MVD is better for those patients involving venous compression in the first operation, previously treated in a less experienced hospital, with young age and with good sensory function. And the destructive procedures are indicated for those patients probably involving non-vascular compression, with elder age, with poor sensory function, or with poor physical conditions.

Techniques for late repeat MVD: Operation is performed via suboccipital retromastoid approach. The skin incision is the same as the previous scar. After subcutaneous dissection and removal of the titanium wire mash, the previous craniectomy is enlarged a little until the angle between the transverse sinus and sigmoid sinus was reached. Then the dura mater is opened. After sharp dissection of the adhesion between the dura and the cerebellum, the cerebellar hemisphere is retracted gently while CSF is drained away. The adhesive tissue among the cranial nerves, vessels, and the cerebellar surface is dissected sharply. Decompression of the 5th cranial nerve for many offending vessel or Teflon felt is the goal of repeat MVD. To this end, exploration of the full course of the nerve root is necessary, including the REZ (zone 1–4) and the cisternal portion (zone 5). Exposing sequence and technique are similar to that in early reoperation, however, with much more effort in managing adhesion and bleeding. If the recurrence was caused by new vascular compression, Teflon felt was used to interpose the nerve root and the vascular loop. Otherwise, if adhesion was induced by Teflon felt, then the previous Teflon felt and the granulomatous tissue were removed. No more Teflon felt was inserted. If no compressing lesions were noted at the time of reoperation, a partial nerve section was performed (Amador and Pollock 2008).

References

Amador N, Pollock BE. Repeat posterior fossa exploration for patients with persistent or recurrent idiopathic trigeminal neuralgia. J Neurosurg. 2008;108(5):916–20.

Bakker NA, Van Dijk JM, Immenga S, Wagemakers M, Metzemaekers JD. Repeat microvascular decompression for recurrent idiopathic trigeminal neuralgia. J Neurosurg. 2014;121(4):936–9.

Chen J, Lee S, Lui T, Yeh Y, Chen T, Tzaan W. Teflon granuloma after microvascular decompression for trigeminal neuralgia. Surg Neurol. 2000;53(3):281–7.

Chung SS, Chang JH, Choi JY, Chang JW, Park YG. Microvascular decompression for hemifacial spasm: a long-term follow-up of 1,169 consecutive cases. Stereotact Funct Neurosurg. 2001;77(1–4):190–3.

Engh JA, Horowitz M, Burkhart L, Chang YF, Kassam A. Repeat microvascular decompression for hemifacial spasm. J Neurol Neurosurg Psychiatry. 2005;76(11):1574–80.

Hong W, Zheng X, Wu Z, Li X, Wang X, Li Y, et al. Clinical features and surgical treatment of trigeminal neuralgia caused solely by venous compression. Acta Neurochir (Wien). 2011;153(5):1037–42.

Hughes MA, Branstetter BF, Taylor CT, Fakhran S, Delfyett WT, Frederickson AM, Sekula RF Jr. MRI findings in patients with a history of failed prior microvascular decompression for hemifacial spasm: how to image and where to look. AJNR Am J Neuroradiol. 2015;36(4):768–73. doi:10.3174/ajnr.A4174.

Hyun SJ, Kong DS, Park K. Microvascular decompression for treating hemifacial spasm: lessons learned from a prospective study of 1,174 operations. Neurosurg Rev. 2010;33(3):325–34; discussion 34.

Ishikawa M, Nakanishi T, Takamiya Y, Namiki J. Delayed resolution of residual hemifacial spasm after microvascular decompression operations. Neurosurgery. 2001;49(4):847–54; discussion 54–6.

Kim HR, Rhee DJ, Kong DS, Park K. Prognostic factors of hemifacial spasm after microvascular decompression. J Korean Neurosurg Soc. 2009;45(6):336–40.

Kondo A. Follow-up results of microvascular decompression in trigeminal neuralgia and hemifacial spasm. Neurosurgery. 1997;40(1):46–51; discussion 51–2.

Lee SH, Levy EI, Scarrow AM, Kassam A, Jannetta PJ. Recurrent trigeminal neuralgia attributable to veins after microvascular decompression. Neurosurgery. 2000;46(2):356–61; discussion 61–2.

Li CS. Varied patterns of postoperative course of disappearance of hemifacial spasm after microvascular decompression. Acta Neurochir (Wien). 2005;147(6):617–20; discussion 20.

Li S, Hong W, Tang Y, Ying T, Zhang W, Li X, et al. Re-operation for persistent hemifacial spasm after microvascular decompression with the aid of intraoperative monitoring of abnormal muscle response. Acta Neurochir (Wien). 2010;152(12):2113–8.

Li Y, Zheng X, Hua X, Ying T, Zhong J, Zhang W, et al. Surgical treatment of hemifacial spasm with zone-4 offending vessel. Acta Neurochir (Wien). 2013;155(5):849–53.

Moller AR, Jannetta PJ. On the origin of synkinesis in hemifacial spasm: results of intracranial recordings. J Neurosurg. 1984;61(3):569–76.

Moller AR, Jannetta PJ. Hemifacial spasm: results of electrophysiologic recording during microvascular decompression operations. Neurology. 1985a;35(7):969–74.

Moller AR, Jannetta PJ. Synkinesis in hemifacial spasm: results of recording intracranially from the facial nerve. Experientia. 1985b;41(3):415–7.

Oh ET, Kim E, Hyun DK, Yoon SH, Park H, Park HC. Time course of symptom disappearance after microvascular decompression for hemifacial spasm. J Korean Neurosurg Soc. 2008;44(4):245–8.

Oliveira LD, Cardoso F, Vargas AP. Hemifacial spasm and arterial hypertension. Mov Disord. 1999;14(5):832–5.

Park YS, Chang JH, Cho J, Park YG, Chung SS, Chang JW. Reoperation for persistent or recurrent hemifacial spasm after microvascular decompression. Neurosurgery. 2006;58(6):1162–7; discussion 1162–7.

Payner TD, Tew Jr JM. Recurrence of hemifacial spasm after microvascular decompression. Neurosurgery. 1996;38(4):686–90; discussion 90–1.

Sun T, Saito S, Nakai O, Ando T. Long-term results of microvascular decompression for trigeminal neuralgia with reference to probability of recurrence. Acta Neurochir (Wien). 1994;126(2–4):144–8.

van Loveren H, Tew Jr JM, Keller JT, Nurre MA. A 10-year experience in the treatment of trigeminal neuralgia. Comparison of percutaneous stereotaxic rhizotomy and posterior fossa exploration. J Neurosurg. 1982;57(6):757–64.

Ying TT, Li ST, Zhong J, Li XY, Wang XH, Zhu J. The value of abnormal muscle response monitoring during microvascular decompression surgery for hemifacial spasm. Int J Surg. 2011;9(4):347–51.

Zhang LW, Liu YG, Wu CY, Xu SJ, Zhu SG. Radiofrequency thermocoagulation rhizotomy for recurrent trigeminal neuralgia after microvascular decompression. Chin Med J (Engl). 2011; 124(22):3726–30.

Zheng X, Feng B, Zhang W, Ying T, Li S. Hemifacial spasm caused by cross type vascular compression. Neurol Res. 2011;33(9):965–9.

Zheng X, Hong W, Tang Y, Wu Z, Shang M, Zhang W, et al. Sympathetic nerves bridge the cross-transmission in hemifacial spasm. Neurosci Lett. 2012a;517(1): 52–5.

Zheng X, Hong W, Tang Y, Ying T, Wu Z, Shang M, et al. Discovery of a new waveform for intraoperative monitoring of hemifacial spasms. Acta Neurochir (Wien). 2012b. doi:10.1007/s00701-012-1304-6.

Zheng X, Feng B, Hong W, Zhang W, Yang M, Tang Y, et al. Management of intraneural vessels during microvascular decompression surgery for trigeminal neuralgia. World Neurosurg. 2012c;77(5–6):771–4.

Zhong J, Zhu J, Li ST, Li XY, Wang XH, Yang M, et al. An analysis of failed microvascular decompression in patients with hemifacial spasm: focused on the early reoperative findings. Acta Neurochir (Wien). 2010;152(12):2119–23.

Zhong J, Li ST, Zhu J, Guan HX. Is entire nerve root decompression necessary for hemifacial spasm? Int J Surg. 2011;9(3):254–7.

Zhong J, Xia L, Dou NN, Ying TT, Zhu J, Liu MX, et al. Delayed relief of hemifacial spasm after microvascular decompression: can it be avoided? Acta Neurochir (Wien). 2015;157(1):93–8; discussion 8–9.

Zhu J, Zhong J, Jiao W, Zhou QM, Guan HX, Dou NN, et al. Via-cerebellar-fissures approach for microvascular decompression of trigeminal nerve. J Craniofac Surg. 2014;25(4):1438–40.

15 Perioperative Adverse Events of Microvascular Decompression: Review and a Personal Experience of 2263 Cases

Doo-Sik Kong and Kwan Park

Abstract

Background Microvascular decompression (MVD) is effective treatment modality for the treatment of trigeminal neuralgia and hemifacial spasm. However, there are potential risks of MVD generally related to the surgical technique and particularly to manipulation around the cranial nerve. The purpose of this study was to identify possible adverse events after MVD and to establish the strategies to minimize the complications after MVD.

Methods We reviewed the literatures focusing on the adverse events after MVD and analyzed our personal series of 2263 cases.

Results The complications after MVD include facial palsy, middle ear effusion, hearing loss, dysgeusia, other cranial nerve deficits, thromboembolic or hemorrhage complication, etc. The complications can be classified into transient and permanent deficits. For transient complications, patient's reassurance is important, and for permanent form, we should keep in mind the disciplined surgical technique, collaborative intraoperative monitoring, and development of modern equipments.

Conclusion We believe that a significant proportion of these complications can be avoidable through development and testing of standardized protocols to incorporate monitoring technologies and specific technical practices, teamwork and communication, and concentrated volume and specialization.

Keywords

MVD • Complications • Avoidable • Standard protocols • Techniques

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Microvascular decompression (MVD) is considered the treatment of choice for trigeminal neuralgia and hemifacial spasm and is a safe and effective treatment option for definitive cure. However, there are potential risks of MVD

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generally related to the surgical technique and particularly to manipulation around the cranial nerve. Common complications included facial palsy, hearing impairment, cerebrospinal fluid leaks and fistulas, cranial nerve deficits, and cerebrovascular events. In particular, cerebrovascular events, although relatively rare, remains a significant problem. Hemorrhagic complications carry by far the highest risk of devastating neurological outcome in functional neurosurgery. Estimating the predictive risk factors of postoperative complications is a challenging issue. Increased regionalization, more stringent volume requirements, and greater standardization and more rapid assimilation of standardized devices may prove beneficial for the more technical complications of surgery, but these strategies have not been adequately evaluated. This study reviews the factors that may predict susceptibility to post-MVD complications and addresses therapeutic choices, adjunctive therapies, and technological applications that may help with complication avoidance.

15.1 Transient and Permanent Deficits

Postoperative complications after MVD can be classified into transient and permanent deficits. Transient complications include facial palsy, hearing deficit, and cerebrospinal fluid leak. Permanent complications include hearing loss, facial palsy, and hemorrhagic/thromboembolic events (Miller and Miller 2012; Huh et al. 2008). Transient complications can be overcome by close follow-up of symptom change, appropriate warning for possible conversion to the permanent deficits, and simultaneously reassuring the patients. Huh et al. published in 2008 that postoperative complications were noted in 545 (35.8 %) patients. Among them, facial palsy, hearing deficit, and low cranial nerve palsies were found in 18.6 % ($n=283$), 7.2 % ($n=109$), and 2.8 % ($n=43$), respectively. However, permanent facial weakness, hearing deficit, and lower cranial nerve palsies such as hoarseness and dysphagia were encountered in 1.2 % ($n=18$), 2.1 % ($n=32$), and 0.1 % ($n=2$), respectively (Huh et al. 2008). Therefore, 1-year or longer follow-up monitoring is required before determination of "permanence" with respect to cranial nerve dysfunction (Huh et al. 2008).

There are some differences of the incidence of complications after MVD between trigeminal neuralgia and hemifacial spasm. In our series of 2263 patients undergoing MVD (under submission), the most common complications after MVD for trigeminal neuralgia was middle ear effusion (MEE) (5.38 %), followed by hearing impairment (1.35 %), dysgeusia (1.35 %), and wound complications (1.35 %). After MVD for hemifacial spasm, the most common complications were facial nerve palsy (8.19 %), followed by MEE (5.38 %) and hearing deficits (3.63 %). In the following sections, we are to discuss each complication of MVD, which we can encounter commonly.

15.1.1 Facial Palsy

Facial palsy is the most common complication of MVD for hemifacial spasm. The incidence of facial palsy is reported to be 14.3–18.6 % (Miller and Miller 2012; Huh et al. 2008). Facial palsy can be classified into two groups based on the mode of onset. Immediate facial palsy occurs immediately after surgery within 24 h, where delayed facial palsy usually occurs over a 24-h to 7-day period after the operation (Hongo et al. 1985). Recent literatures showed that the rate for incurring delayed facial palsy following MVD is 2.8–8.3 % (Lovely et al. 1998; Rhee et al. 2006; Furukawa et al. 2003; Han et al. 2012) and was reversible in almost all the cases. Recently, in our 2263 cases series, we found that facial palsy occurred in 169 (7.47 %) cases. Eighteen patients (0.80 %) exhibited immediately after surgery, and 151 patients (6.67 %) had facial palsy after a delayed period. Severity of immediate-onset facial palsy was higher than delayed-onset facial palsy. In cases of trigeminal neuralgia, there was no incidence of immediate-onset facial palsy, but two cases had delayed-onset facial palsy.

The accurate mechanism of postoperative facial palsy remains unclear. Possible etiologies

of facial palsy include direct injury in the exit zone of facial nerve injury by the Teflon felt, delayed facial nerve edema by manipulation, or disturbance of microcirculation due to vasospasm (Huh et al. 2008; Lovely et al. 1998; Rhee et al. 2006). Finding that most cases of facial palsy occur in the hemifacial spasm rather than trigeminal neuralgia showed evidence that immediate-onset facial palsy is caused by direct injury to facial nerve. During the surgical field, any degree of stretching force on the nerve can be added by cerebellar retraction to secure operation field. In general, the facial weakness can resolve spontaneously, with excellent outcomes (Lovely et al. 1998; Rhee et al. 2006; Samii et al. 2002; Kondo 1998). Cause of delayed-onset facial palsy is unknown. The theory of viral origin may have reliable rationale (Lovely et al. 1998; Rhee et al. 2006; Furukawa et al. 2003; Hung et al. 2002; Badr-El-Dine et al. 2002). The manipulation of the nerve could stimulate a dormant virus, possibly localized in the geniculate ganglion. Postoperative facial palsy is usually managed with extensive rehabilitation, and reoperations are rarely required (Lovely et al. 1998; Rhee et al. 2006; Furukawa et al. 2003; Han et al. 2012; Dannenbaum et al. 2008; Jeon et al. 2010).

15.1.2 Hearing Impairment

Hearing impairment is a relatively infrequent but significant risk of the MVD caused by damage to the vestibulocochlear nerve. In general, hearing impairment is defined as either an increase of more than 15 dB of average pure tone audiogram (0.5, 1, 2 K) threshold according to bone conduction or an increase of more than 20 % of speech discrimination scale related to baseline hearing. Excessive and forceful retraction of the cerebellum is supposed to be a major reason of hearing impairment after MVD. Profound sensorineural hearing loss is a recognized complication of MVD for hemifacial spasm with a reported incidence of 1.9–20 % (Miller and Miller 2012; Dannenbaum et al. 2008; Chung et al. 2001; Fritz et al. 1988; Hatayama and Moller 1998; Hyun et al. 2010; Jannetta and Hirsch 1993; Jannetta et al. 1986; Jo et al. 2011, 2013; Moffat et al. 2005; Moller and Jannetta 1983; Park et al. 2009; Rosseau et al. 1993; Sekiya et al. 1991; Shah et al. 2012; Vasama et al. 1998). These reports may underestimate the true incidence of hearing loss because preoperative and postoperative audiograms are not always available for comparison.

15.2 Possible Mechanism of Hearing Loss

Hearing loss after MVD may occur for the following reasons (Miller and Miller 2012; Dannenbaum et al. 2008; Chung et al. 2001; Fritz et al. 1988; Hatayama and Moller 1998; Hyun et al. 2010; Jannetta and Hirsch 1993; Jannetta et al. 1986; Jo et al. 2011, 2013; Moffat et al. 2005; Moller and Jannetta 1983; Park et al. 2009; Rosseau et al. 1993; Sekiya et al. 1991; Shah et al. 2012; Vasama et al. 1998):

1. Stretching of the 8th nerve when retracting the cerebellum; the manipulation of the nerve is likely to lead to the stretching or spasm of the vasa nervorum and consequent ischemic phenomena.
2. Manipulation of the labyrinthine artery and/or the anteroinferior cerebellar artery; the cochlea receives its blood supply from the internal auditory artery. Damage to the artery leads to cochlear ischemia or infarction which may affect wave I.
3. Direct trauma to the nerve by instruments or nearby coagulation.
4. Neo-compression of the nerve by the prosthesis interposed between the offending vessel and the 7th nerve complex at the end of surgery (Polo et al. 2004; Sindou 2005).
5. Recently, we reported a new possible mechanism of hearing loss due to increased intracranial pressure from overinfusion of saline with dural closure (Jo et al. 2013); intradural compression due to overinfusion of saline may lead to postoperative hearing loss, although the incidence is low, and immediate decompression by drainage may be required.

Recently, Ying et al. (2013) suggested that high-frequency hearing loss occurs in a significant number of patients following MVD surgery for hemifacial spasm. Drill-induced noise and transient loss of CSF during surgery may impair hearing in the high-frequency ranges on both the ipsilateral and contralateral sides.

Considering the flocculus covering the VII–VIII nerves complex, it is difficult to directly approach in a dorsoventral direction and completely observe the root exit zone of facial nerve. Cerebellar retraction may be mandatory to observe the exit zone of facial nerve, and it is sometimes closely associated with postoperative hearing deterioration. In the recent literature, Lee et al. (2015) suggested that preoperative assessment of pathological anatomy by MR imaging can predict the degree of cerebellar retraction intraoperatively and attempted to show indirectly the correlation of cerebellar retraction with BAEP change.

Rarely, sudden deterioration in brainstem auditory evoked potentials (BAEP) occurs during or immediately after craniectomy, probably caused by an edema in the intracanalicular tract of the nerve. The vibrations caused by drilling for craniotomy is transmitted to the rocca petrosa and may lead to edema and subsequent ischemia of the cochlear nerve, resulting in a reversible blockade of its transmission capacities (Lee et al. 2015). Grundy et al. suggested that hypoperfusion should be considered as a contributing factor, when decreasing of the amplitude without prolongation of the latency (Grundy et al. 1982). The amplitude of BAEP can depend upon the individual situation such as recording conditions, electrode impedance, and a number of unknown factors (Moller 1995).

15.3 Role of BAEP

Intraoperative monitoring of brainstem auditory evoked potentials (BAEP) is useful for reducing the risk of hearing impairment in patients undergoing the cerebellopontine angle (CPA) surgery including MVD (Hatayama and Moller 1998; Lee et al. 2009; Sindou et al. 1992). The surgical procedure most frequently associated with BAEP deterioration is traction and/or compression of the nerve during the course of the maneuvers to release the nerve structures from the vascular loop. These BAEP changes may reflect direct mechanical or thermal damage to the brainstem, brainstem compression, or ischemia or infarction resulting from vascular compromise. Our experience supported that during MVD surgery, if the decrease of amplitude and prolongation of latency in wave V are rapid and the changes do not recover with correction, the possibility of hearing loss is high (Ying et al. 2013).

Some authors suggested that 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 (Hatayama and Moller 1998; Jo et al. 2011; Sindou et al. 1992; Acevedo et al. 1997). However, Polo et al. (2004) suggested that the I–V interpeak latency, or the latency of wave V, was an effective and predictive indicator of postoperative hearing. Ying et al. (2014) demonstrated that hearing impairment has earlier change in the amplitude of wave V and peak I–V latency during intraoperative BAEP monitoring for MVD of hemifacial spasm. To date, there is no consensus regarding the criteria for significant intraoperative BAEP change. In our series of 2165 cases, incidence of hearing impairment was 2.74 % with no BAEP change, 6.17 % with change of the amplitude <50 %, 11.76 % with change of the amplitude >50 %, and 35.29 % with loss of wave V (under submission).

15.4 Strategies to Minimize the Hearing Impairment during or after MVD

Intraoperative BAEP monitoring has an essential role of alarming for preservation of hearing. Comprehensive understanding of surgical anatomy around the structures and optimal approach can reduce the risk of possible hearing impairment. In addition, minimal retraction of the cerebellum is very critical, and the endoscope may be helpful if it can provide a second look to iden-

tify the nerve root entry zone and confirms the position of the Teflon felt (Badr-El-Dine et al. 2002; Lee et al. 2014; Bohman et al. 2014; Liang et al. 2009; Cheng et al. 2008; Kabil et al. 2005).

15.4.1 Other Cranial Nerve Deficits

As a rare complication following MVD, some patients sometimes complain of various nature of dysgeusia (impairment of the sense of taste), including tongue numbness, continuous sour taste, and hypogeusia. In our series of 2263 series, 11 patients had persistent dysgeusia. 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 (Grant et al. 1987; Natarajan 2000; Nicol et al. 2000). Unlike other complications, the prognosis of dysguesia was poor. It is very uncommon that there is sporadically trochlear or abducens nerve palsy after MVD (Barker et al. 1995). Sometimes vocal cord palsy occurs. As a result, patients showed asymmetric soft palate elevation, uvula deviation, or phonation difficulty.

For MVD for hemifacial spasm, lower cranial nerve palsy should be considered as an uncommon complication. The clinical manifestations of lower cranial nerve palsies include hoarseness, dysphagia, and swallowing difficulty. However, those symptoms can be more commonly caused by other perioperative complication such as intubation-related injury to pharynx or vocal cord. In most cases, lower cranial nerve palsy has a self-limiting course.

15.4.2 Cerebrospinal Fluid (CSF) Leakage

Middle ear effusion (MEE) was frequently found in 5.38 % of our series. MEE does not always mean active leakage of CSF. It can be sometimes caused by saline irrigated or blood through exposed mastoid air cells during surgery. As another result of CSF leak, patients can have CSF rhinorrhea. Fortunately, most patients recover spontaneously. For intractable CSF leaks, more active management such as CSF drainage via lumbar puncture, myringotomy, or revision surgery should be considered. To avoid postoperative CSF leak, meticulous sealing with bone wax, when mastoid air cells are open, should be performed.

15.4.3 Hemorrhagic or Thromboembolic Complications

Hemorrhagic complication is the most serious complication after MVD and is life-threatening. Representative cerebrovascular events include subdural hematoma, cerebral infarction from disturbance of venous outflow, dural arteriovenous fistula, etc. To reduce the retraction injury, overdrainage of CSF can be often performed. However, because overdrainage of CSF is often associated with the tear to bridging vein, leading to supratentorial subdural hematoma, it should be kept in mind to infuse saline adequately before the dural closure. It will be also helpful to minimize the pneumocephalus.

There is no definitive evidence that the sacrifice of the inferior petrosal vein does not carry the risk of venous hypertension or infarction. Considering the rare incidence of events, it is very difficult to draw any conclusion regarding the safety of the sacrifice of petrosal vein; however, we believe that unnecessary sacrifice of inferior petrosal vein should be avoided. Rarely, disturbance of blood flow through transverse sinus by air embolism can contribute the ipsilateral temporal lobe infarction. Treatments for cerebrovascular events must be selected carefully based on the etiology of the condition.

15.4.4 Complications after MVD in the Elderly Patients

Recently, Youn et al. (2013) demonstrated the incidence of complications associated with MVD in the elderly patients over 60 years. The authors showed that complications were more common in the elderly patients than in the younger group.

In the elderly patients, significant difference in the incidence of permanent hearing deficits was found (3.9 % vs. 1.9 %, $p=0.042$). Comorbid stroke history and multiple offenders were the risk factors for hearing loss after surgery. The authors speculated that more common hearing loss after MVD occurred, because the elderly patients were more vulnerable to the damage to the microcirculation. However, there was no difference in serious complications such as hemorrhage, thromboembolic infarct, and death between both groups. Here, the authors recommended that MVD was also effective in elderly patients, but surgeons should be more careful for relatively higher complications rate.

Conclusions

In conclusion, in an attempt to reduce the major morbidities associated with MVD, the disciplined implementation of the surgical techniques collectively contributes to lower incidence of morbidities: (1) proper direction and adequate retraction of cerebellum, (2) gradual retraction of the cerebellum as CSF is allowed to drain, and (3) preservation of arachnoid sheath over the CN VIII and CN VII complex (Bond et al. 2010). The above-described surgical technique and development of modern equipments can reduce complications and will ensure more favorable outcome (Barker et al. 1995). In addition, collaborative monitoring and evaluation of such protocols are likely necessary for the advancement of MVD. We believe that a significant proportion of these complications may be avoidable through development and testing of standardized protocols to incorporate monitoring technologies and specific technical practices, teamwork and communication, and concentrated volume and specialization.

References

Acevedo JC, et al. Microvascular decompression for the treatment of hemifacial spasm. Retrospective study of a consecutive series of 75 operated patients – electrophysiologic and anatomical surgical analysis. Stereotact Funct Neurosurg. 1997;68(1–4 Pt 1):260–5.

Badr-El-Dine M, et al. Endoscopically assisted minimally invasive microvascular decompression of hemifacial spasm. Otol Neurotol. 2002;23(2):122–8.

Barker 2nd FG, et al. Microvascular decompression for hemifacial spasm. J Neurosurg. 1995;82(2):201–10.

Bohman LE, et al. Fully endoscopic microvascular decompression for trigeminal neuralgia: technique review and early outcomes. Neurosurg Focus. 2014;37(4):E18.

Bond AE, et al. Operative strategies for minimizing hearing loss and other major complications associated with microvascular decompression for trigeminal neuralgia. World Neurosurg. 2010;74(1):172–7.

Cheng WY, Chao SC, Shen CC. Endoscopic microvascular decompression of the hemifacial spasm. Surg Neurol 2008;70 Suppl 1:S1:40–6.

Chung SS, et al. Microvascular decompression for hemifacial spasm: a long-term follow-up of 1,169 consecutive cases. Stereotact Funct Neurosurg. 2001;77(1–4):190–3.

Dannenbaum M, et al. Microvascular decompression for hemifacial spasm: long-term results from 114 operations performed without neurophysiological monitoring. J Neurosurg. 2008;109(3):410–5.

Fritz W, Schafer J, Klein HJ. Hearing loss after microvascular decompression for trigeminal neuralgia. J Neurosurg. 1988;69(3):367–70.

Furukawa K, et al. Delayed facial palsy after microvascular decompression for hemifacial spasm due to reactivation of varicella-zoster virus. No Shinkei Geka. 2003;31(8):899–902.

Grant R, et al. Evoked taste thresholds in a normal population and the application of electrogustometry to trigeminal nerve disease. J Neurol Neurosurg Psychiatry. 1987;50(1):12–21.

Grundy BL, et al. Intraoperative monitoring of brain-stem auditory evoked potentials. J Neurosurg. 1982;57(5):674–81.

Han JS, et al. Delayed cranial nerve palsy after microvascular decompression for hemifacial spasm. J Korean Neurosurg Soc. 2012;52(4):288–92.

Hatayama T, Moller AR. Correlation between latency and amplitude of peak V in the brainstem auditory evoked potentials: intraoperative recordings in microvascular decompression operations. Acta Neurochir (Wien). 1998;140(7):681–7.

Hongo K, et al. Posterior fossa microvascular decompression for hemifacial spasm and trigeminal neuralgia – some improvements on operative devices and technique. No Shinkei Geka. 1985;13(12):1291–6.

Huh R, et al. Microvascular decompression for hemifacial spasm: analyses of operative complications in 1582 consecutive patients. Surg Neurol. 2008;69(2):153–7; discussion 157.

Hung CM, et al. Contralateral neurologic deficits following microvascular decompression surgery – a case report. Acta Anaesthesiol Sin. 2002;40(2):91–5.

Hyun SJ, Kong DS, Park K. Microvascular decompression for treating hemifacial spasm: lessons learned from a prospective study of 1,174 operations. Neurosurg Rev. 2010;33(3):325–34; discussion 334.

Jannetta PJ, Hirsch BE. Restoration of useful hearing following microvascular decompression of cochlear nerve. Am J Otol. 1993;14(6):627.

Jannetta PJ, et al. Neurosurgical treatment of vertigo by microvascular decompression of the eighth cranial nerve. Clin Neurosurg. 1986;33:645–65.

Jeon CJ, et al. The efficacy and safety of microvascular decompression for hemifacial spasm in elderly patients. J Korean Neurosurg Soc. 2010;47(6):442–5.

Jo KW, et al. The patterns and risk factors of hearing loss following microvascular decompression for hemifacial spasm. Acta Neurochir (Wien). 2011;153(5): 1023–30.

Jo KW, et al. A new possible mechanism of hearing loss after microvascular decompression for hemifacial spasm. Otol Neurotol. 2013;34(7):1247–52.

Kabil MS, Eby JB, Shahinian HK. Endoscopic vascular decompression versus microvascular decompression of the trigeminal nerve. Minim Invasive Neurosurg. 2005;48(4):207–12.

Kondo A. Follow-up results of using microvascular decompression for treatment of glossopharyngeal neuralgia. J Neurosurg. 1998;88(2):221–5.

Lee SH, et al. Results of auditory brainstem response monitoring of microvascular decompression: a prospective study of 22 patients with hemifacial spasm. Laryngoscope. 2009;119(10):1887–92.

Lee CC, et al. Brainstem auditory evoked potential monitoring and neuro-endoscopy: two tools to ensure hearing preservation and surgical success during microvascular decompression. J Chin Med Assoc. 2014;77(6):308–16.

Lee MH, et al. Cerebellar retraction and hearing loss after microvascular decompression for hemifacial spasm. Acta Neurochir (Wien). 2015;157:337–43.

Liang J, et al. Microvascular decompression for the hemifacial spasm with endoscopy. Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2009;23(4):145–8.

Lovely TJ, Getch CC, Jannetta PJ. Delayed facial weakness after microvascular decompression of cranial nerve VII. Surg Neurol. 1998;50(5):449–52.

Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for treatment of hemifacial spasm: a systematic review. Br J Neurosurg. 2012; 26(4):438–44.

Moffat DA, et al. Outcome following retrosigmoid microvascular decompression of the facial nerve for hemifacial spasm. J Laryngol Otol. 2005;119(10):779–83.

Moller AR. Intra-operative neurophysiologic monitoring in neurosurgery: benefits, efficacy, and cost-effectiveness. Clin Neurosurg. 1995;42:171–9.

Moller AR, Jannetta PJ. Monitoring auditory functions during cranial nerve microvascular decompression operations by direct recording from the eighth nerve. J Neurosurg. 1983;59(3):493–9.

Natarajan M. Percutaneous trigeminal ganglion balloon compression: experience in 40 patients. Neurol India. 2000;48(4):330–2.

Nicol B, et al. Gamma knife radiosurgery using 90 Gy for trigeminal neuralgia. J Neurosurg. 2000;93 Suppl 3:152–4.

Park K, et al. Patterns of hearing loss after microvascular decompression for hemifacial spasm. J Neurol Neurosurg Psychiatry. 2009;80(10):1165–7.

Polo G, et al. Brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm: intraoperative brainstem auditory evoked potential changes and warning values to prevent hearing loss – prospective study in a consecutive series of 84 patients. Neurosurgery. 2004;54(1):97–104; discussion 104–6.

Rhee DJ, et al. Frequency and prognosis of delayed facial palsy after microvascular decompression for hemifacial spasm. Acta Neurochir (Wien). 2006;148(8):839–43; discussion 843.

Rosseau GL, et al. Restoration of useful hearing after microvascular decompression of the cochlear nerve. Am J Otol. 1993;14(4):392–7.

Samii M, et al. Microvascular decompression to treat hemifacial spasm: long-term results for a consecutive series of 143 patients. Neurosurgery. 2002;50(4):712–8; discussion 718–9.

Sekiya T, et al. Vestibular nerve injury as a complication of microvascular decompression. Neurosurgery. 1991;29(5):773–5; discussion 775–6.

Shah A, et al. Hearing outcomes following microvascular decompression for hemifacial spasm. Clin Neurol Neurosurg. 2012;114(6):673–7.

Sindou MP. Microvascular decompression for primary hemifacial spasm. Importance of intraoperative neurophysiological monitoring. Acta Neurochir (Wien). 2005;147(10):1019–26; discussion 1026.

Sindou M, et al. Hearing prognosis and intraoperative guidance of brainstem auditory evoked potential in microvascular decompression. Laryngoscope. 1992; 102(6):678–82.

Vasama JP, Moller MB, Moller AR. Microvascular decompression of the cochlear nerve in patients with severe tinnitus. Preoperative findings and operative outcome in 22 patients. Neurol Res. 1998;20(3):242–8.

Ying T, et al. Incidence of high-frequency hearing loss after microvascular decompression for hemifacial spasm. J Neurosurg. 2013;118(4):719–24.

Ying T, et al. Empirical factors associated with Brainstem auditory evoked potential monitoring during microvascular decompression for hemifacial spasm and its correlation to hearing loss. Acta Neurochir (Wien). 2014;156(3):571–5.

Youn J, et al. Safety and effectiveness of microvascular decompression for the treatment of hemifacial spasm in the elderly. Eur Neurol. 2013;70(3–4):165–71.