Foramen Magnum Meningiomas

Part I: Definition, Clinical Features, Diagnostic Approach, and Perioperative Protocol

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Learning Objectives: *After reading this article, the participant should be able to:*

1. *Recall the incidence and differential diagnosis of foramen magnum meningiomas.*
2. *Describe the clinical presentation and the pre-operative evaluation of patients with foramen magnum meningiomas, including the radiologic features and the different types of foramen magnum meningiomas.*
3. *Explain the pre-operative evaluation, including the anesthetic considerations and issues related to intra-operative monitoring.*

Meningiomas of the foramen magnum (FMM) account for 0.3% to 3.2% of all meningiomas, between 4.2% and 20% of all posterior fossa meningiomas, and 8.6% of all spinal meningiomas. Benign, extramedullary tumors of the foramen magnum account for one third of all tumors found at the craniospinal junction; meningiomas are the most common, accounting for 60% to 77% of these tumors. Meningiomas occur three times more often than neurinomas, which are the next most common type of benign tumors among those that occur at the foramen magnum. In addition to neurinomas, the differential diagnosis of a foramen magnum meningioma can include dermoids, epidermoids, teratomas, lipomas, hemangioblastomas, cavernous angiomas, giant thrombosed aneurysms of the vertebral artery, intramedullary cervical spinal cord tumors, and syringomyelia. Also included in the differential diagnosis are primarily extradural lesions such as chordomas, synovial cysts, osseous tumors, sarcomas, infections, congenital malformations of the craniocervical junction, rheumatoid arthritis, and metastases. Meningiomas affect females two to three times as often as males. Patients most commonly are in the fourth through sixth decades of life, but these tumors have been reported in patients of almost every age. Although a foramen magnum meningioma has been reported in a 4-year-old child, this tumor rarely occurs in children. The most common histologic type is meningothelial, followed by psammomatous and fibrous type of meningioma.

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Meningiomas of the foramen magnum arise at the dura of the craniocervical junction. The zone from which they arise corresponds anteriorly to the area between the lower third of the clivus and the upper edge of the axis, laterally from the jugular tubercle to the upper aspect of the C2 lamina, and posteriorly from the anterior edge of the squamous occipital bone to the C2 spinous process. There are two main types: (1) craniospinal meningioma, which originates from the basal groove at the lower third of the clivus, anterior or anterolateral to the medulla, and projects downward toward the foramen magnum; and (2) spinocranial meningioma, which originates at the upper cervical area, usually posterior or posterolateral to the spinal cord, and projects upward intracranially into the cerebellomedullary cistern. Those meningiomas that arise anterior to the ascending coronal plane between the first dentate ligament and the 9th through 12th cranial nerves on both sides are defined as ventral (anterior) FMM (VFMM). This type accounts for 68% to 98% of all FMM and should be clearly distinguished from meningiomas of the dorsal (posterior) FMM (DFMM), because there are differences in operative approach, postoperative results, complications, and mortality. Ventral FMM is one of the most challenging types of meningioma to treat because a large number of vital neurovascular structures, which are sensitive to injury, are crowded together in this deeply hidden central area. These neurovascular elements drape the tumor, making radical removal of the tumor difficult and leading to a relatively high rate of complications.

The rare incidence of FMM limits experience with their treatment. However, the development of modern diagnostic

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techniques, refinement of microsurgical techniques, advances in microsurgical anatomic studies, development of skull base approaches, advances in neuroanesthesia, availability of intraoperative neurophysiologic monitoring, and careful multidisciplinary perioperative planning have advanced the treatment of FMM, allowing for more radical tumor resection, decreasing the rate of complications, and improving patient survival. This issue of *Contemporary Neurosurgery* presents our experiences and describes the management protocol that we have developed. It also takes into consideration the experiences of others in treating FMM, general perioperative skull base management principles, and lessons learned from each case.

Clinical Features

Meningiomas at the foramen magnum cause symptoms that are unusual, atypical, bizarre, confusing, slowly progressing, and, often, remitting. Before magnetic resonance imaging (MRI) was available, these tumors often were diagnosed at an advanced stage, because the cerebrospinal cisterns at the foramen magnum apparently allow enough room for the tumor to grow steadily for some time before clinical signs appear. In other cases, they were misdiagnosed as multiple sclerosis, cervical spondylosis, a herniated cervical disk, amyotrophic lateral sclerosis, or a carpal tunnel syndrome. Trauma to the head or neck occasionally caused these tumors to manifest themselves before they otherwise would have been detected. Neurofibromatosis type 2 and syringomyelia sometimes were reported to be associated with this type of tumor.

Patients now usually undergo surgery 2 to 3.5 years after symptoms are first noted; with the craniospinal type of FMM the preoperative period is somewhat shorter than in those with spinocranial type of lesion.

The symptoms that most commonly appear in patients with FMMs can be summarized as a triad for ease in memorization. The first category of the triad includes occipital and posterior cervical pain and hyperesthesia in the C2 dermatome. This pain is the most salient initial manifestation of the tumor. The pain is unilateral, commonly is present when the patient awakens, and can be aggravated by extension or lateral flexion of the neck or by maneuvers that raise the intracranial pressure, such as coughing, straining, or sneezing. The second category of the triad of symptoms includes motor deficits and sensory symptoms in the hands, a state known as "cold clumsy hand," which patients often notice as a decreased ability to write or button clothes. Various combinations of limb weakness and sensory deficits in the third category ultimately may advance to a spastic quadriplegia and marked loss of sensation. Other symptoms include dizziness, dysarthria, dysphagia, nystagmus, respiratory difficulties, atrophy of the sternocleidomastoid or trapezius muscles, nausea and vomiting, and hoarseness. Overlapping or combinations of two or more symptoms are more the rule than the exception.

Neurological findings are not always present at the time of admission. The most common neurological sign is hyperreflexia, followed by weakness of the extremities in all combinations. The Babinski sign, a spastic gait, or lower cranial nerve (IX–XI) palsies are present in approximately half of patients with this tumor. Dissociated sensory loss, loss of coordination in the hands, Brown-Séquard syndrome, downbeating nystagmus, and nuchal rigidity and tenderness occur in one quarter to one third of patients. Papilledema, Horner's syndrome, and dysarthria are less common neurological findings.

Pre- and Postoperative Tests

The development of modern diagnostic tools has made the diagnosis of FMMs easier. The surgeon should use all available diagnostic techniques pre-, peri-, and postoperatively, and evaluate all images thoroughly when planning the surgical procedure. One of the primary preoperative diagnostic methods is MRI, including contrast enhancement with gadolinium (Fig. 1). Views in axial, coronal, and sagittal planes delineate an extension of the tumor laterally, vertically, or anteroposteriorly. These planes also show the degree of compression or displacement of the adjacent brain stem and spinal cord, as well as encasement or narrowing of the vertebral artery. MRI also shows accompanying hydrocephalus or other associated lesions. Multiple relaxation techniques (T1-weighted, T2-weighted, and proton

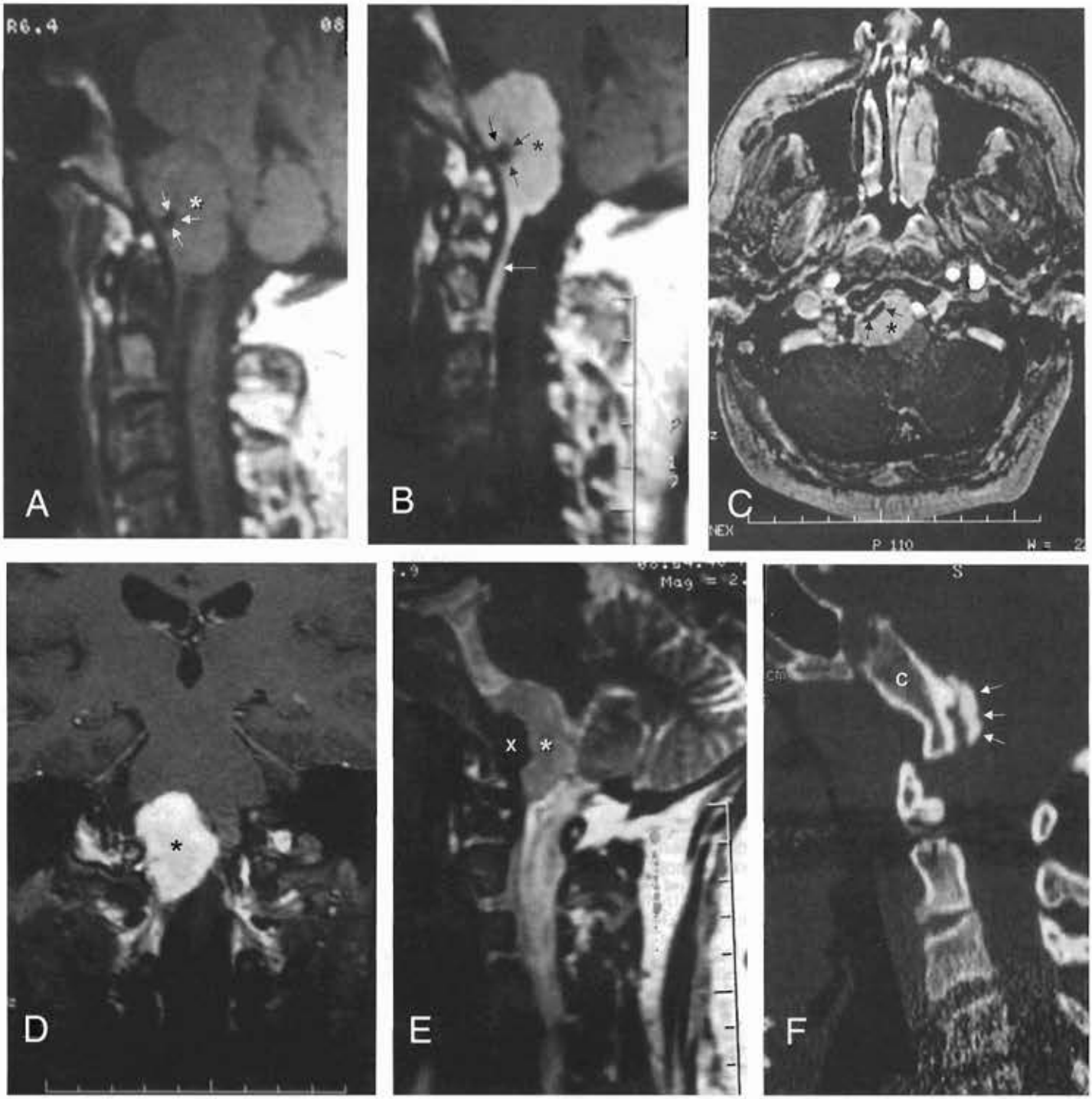

Figure 1. A: Sagittal T1-weighted MRI shows an isointense VFMM (*). Note the vertebral artery (arrows) encased by a tumor. B: The same MRI projection after contrast enhancement. Note the tumor enhancement (hyperintense) and dural enhancement (white arrow). C: Axial T1-weighted MRI after contrast enhancement. Note the hyperintense tumor (*) and tumor encasement of the vertebral artery (arrows). D: Coronal T1-weighted MRI after contrast enhancement. Note the tumor (*) and its extension superiorly, inferiorly, and across the midline. E: Sagittal T2-weighted MRI. Note the hyperdense tumor (*) and the hypodense core of the tumor (X), which proved to be "hyperostotic" tumor infiltration of the clivus. F: Sagittal CT scan ("bone window"). Note the clivus (c) and its tumor infiltration (hyperostosis) (*arrows*). G: Coronal CT scan ("bone window"). Note the hyperostosis (*arrows*). Note also the anatomy of condyles (C) and the location of jugular bulbs (JB) on both sides, all of which are important preoperative diagnostic features.

density-weighted) are necessary. Contrast-enhanced MRI also is important for preoperative evaluation of the venous structure named the suboccipital cavernous sinus. This venous structure cushions the horizontal part of the suboccipital vertebral artery that is encountered during the surgical approach. This venous sinus may be the cause of bothersome intraoperative bleeding as well as a source of air embolism.

On T1-weighted MRI, FMMs appear isointense (see Fig. 1A) or slightly hypointense relative to normal brain tissue. These tumors enhance markedly with the injection of contrast medium (see Fig. 1B–D). On proton density-weighted images, FMMs are hyperintense or isointense, whereas on T2-weighted images, they appear isointense or hyperintense (see Fig. 1E). T2-weighted images are particularly important for delineating the degree of edema in adjacent brain tissue and, often, the consistency ("suckability") of the meningioma (ranging from "soft" and "more suckable" to "hard"). This information is particularly important for planning the surgical exposure and the technique that will be used to remove the tumor (suction aspiration, Cavitron ultrasonic aspiration, etc.).

In comparison, neurinomas are mainly hypointense on T1-weighted images (less intense than meningiomas), isointense or hyperintense on proton density-weighted images (less intense than meningiomas), and hyperintense or isointense on T2-weighted images (more intense than meningiomas). Like FMMs, neurinomas enhance prominently on T1-weighted images with the injection of contrast material. In addition, on contrast-enhanced images, they often have a meningeal sign–enhancement of the dural tail adjacent to the main portion of the tumor (see Fig. 1B). In contrast, neurinomas can have a "pseudomeningeal" sign. This sign is similar and is seen rarely; when it is seen, it appears on MRI both before and after contrast enhancement.

Postoperative MRI (both without and with contrast, again in all three projections) is done within a few days after the surgery, to minimize the amount of postsurgical enhancement. It evaluates the degree of tumor resection, checks for possible postoperative cerebellar and brain stem changes, and serves as a baseline. Even after radical resection of the FMM, persistent contrast enhancement may remain at the site of tumor attachment for several years, and then gradually diminish. To minimize artifacts on follow-up images, any instrumentation or skull flap plating done in this area should be of titanium. Follow-up MRI scans are done 3, 6, and 12 months after the surgery, and yearly thereafter.

Frameless image-guided stereotaxic navigation using MRI (more often) or CT images has proved to be very useful tool in VFMM surgery. Scanning usually is done the day before surgery, with or without fiducials placed on the patient's head. This navigation may help to localize important neurovascular structures (for example, the vertebral artery completely encased by the tumor) and to achieve radical ("gross total)" tumor resection.

On computed tomographic (CT) scans, FMMs usually appear hyperdense or isodense and enhance markedly after the injection of contrast (hyperdense). A CT scan imaging protocol with 1-mm slices provides axial, sagittal, and coronal "bone window" reconstructions and is a very important modality for preoperative diagnosis of adjacent bony changes secondary to tumor invasion ("hyperostosis)" (see Fig. 1F and G) and the degree of calcification within the tumor. It also depicts local bony anatomy such as the shape and size of condyles, the jugular tubercle, and the position of the jugular fossa and the jugular bulb within it (see Fig. 1G). CT scanning also is important in the postoperative evaluation. It usually is done within 24 hours after the surgery and evaluates immediate postoperative changes of both the cerebellum and the brain stem, as well as the bone structures.

Angiographic studies provide a thorough analysis of the posterior circulation. Magnetic resonance angiography (MRA) can detect many of the features that formerly could be found solely with classic angiography and has largely replaced classic angiography (Fig. 2 [*top*]). This diagnostic tool also is simpler, easier to perform, and noninvasive. The analysis should evaluate the basilar and vertebral arteries, including their branches and perforators. It also should determine which vertebral artery is dominant and should detect any of the following information: displacement and narrowing of these

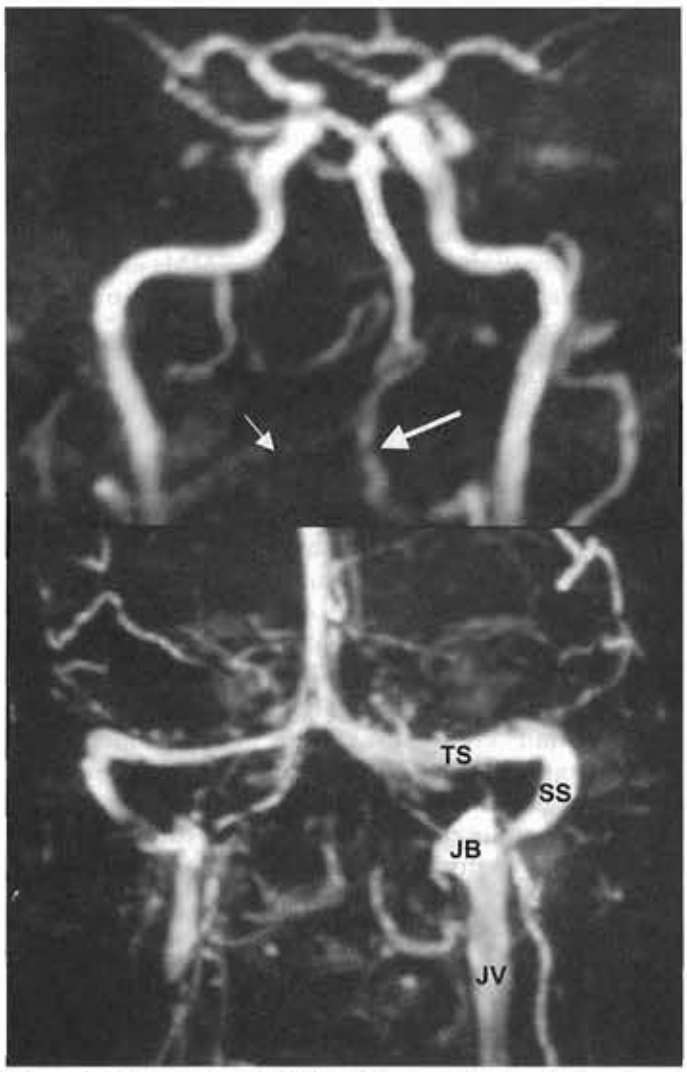

Figure 2. MRA (top) and MRV (bottom). Note the dominant and nondominant vertebral arteries (large and small arrows, top) and the dominant transverse (TS) and sigmoid (SS) sinuses, jugular bulb (JB), and vein (JV).

arteries due to tumor encasement; the change in the distance between the contralateral vertebral arteries; the presence and level of any displacement of the vertebrobasilar junction; displacement of the posterior inferior cerebellar artery (PICA) or the anterior inferior cerebellar artery (AICA); vascularization of the tumor; the presence of associated lesions (aneurysm, arteriovenous malformation); and the status of collateral circulation. For example, it would be very important in preoperative planning to know that the dominant vertebral artery on the side of approach was completely encased by a tumor. Such information is obtained by comparing the MRI and MRA studies (see Fig. 1A–C; Fig. 2). The venous phase of the MRA study (MRV) (see Fig. 2 [*bottom*]) is equally important to determine the venous drainage from the intracranium; the status and dominance of the transverse-sigmoid dural venous sinuses; the level, size and position of the jugular bulb; and other features of the associated venous anatomy. For example, it is important to know preoperatively about the existence of a single, dominant transverse-sigmoid venous sinus and a jugular bulb on the side of approach. In certain cases, the "classic" four-vessel angiography still has a role in delineating particular anatomy that was depicted on MRA.

Multidisciplinary Evaluations

Regardless of the preoperative function of cranial nerves IX through XII, preoperative evaluation should include swallowing studies (for example, bedside and barium swallow) both for an assessment and as a baseline. These studies are coupled with otolaryngological evaluation of larynx and vocal cords. The most common and serious postoperative complication after resection of VFMMs is the deficit of lower cranial nerves (IX and X in particular), so a patient is postoperatively kept on a "nothing per mouth" regimen until swallowing is cleared, in anticipation of this possible or likely deficit. Deficits of these lower cranial nerves are critical due to the risk of aspiration, pneumonia, and death, and contribute to the prolonged hospital stay. As long as the nerves are anatomically preserved during the surgery and "cautiously" manipulated, most (if not all) patients recover or compensate their swallowing, progressing from dysphasia to a regular diet. Meanwhile, appropriate actions such as parenteral nutrition, an orogastric or a gastric tube, vocal cord injections and medialization, and tracheostomy are taken as needed—and as early as possible, to avoid the risk of aspiration pneumonia. Aggressive respiratory management, daily chest radiographs, physical and occupational therapy, speech pathology therapy, and consultations for rehabilitation are involved early in the postoperative period. Depending on the extent of the tumor, ophthalmology, audiometry, electromyography, and other necessary studies may be added in the preoperative work-up.

Operative Indications and Aims of Operation

Regardless of the size of a FMM, the surgical goal should be to preserve and improve neurological function by radical tumor removal. Such removal should be attempted at the first operation, which is the best time to achieve a "cure." Radical resection of a recurrent tumor, although less likely to be achieved than radical resection of a "virgin" tumor, still provides a relatively long and stable follow-up without a recurrence. At times, the surgeon may be forced to accept subtotal removal. In such situations, when a small piece of tumor, adherent to vital neurovascular structures, is left behind, additional radiosurgical treatment may be considered.

Anesthetic Considerations and Intraoperative Monitoring

Good anesthetic technique is essential for the successful removal of foramen magnum meningiomas. Awake endoscopic intubation is done to avoid neck overextension. Systemic monitoring is done through an arterial line, a central venous line (with the position of the tip confirmed radiologically) in the right atrium, a Foley catheter, a Doppler monitor, an electrocardiogram, and an oximeter. End-expiratory PCO_2 pressures, arterial blood gases, electrolytes, hemoglobin and hematocrit values, and a coagulation profile are determined as necessary. We recently have adopted a methylprednisolone spinal cord injury protocol that is initiated on induction of anesthesia and lasts for 24 hours. Antibiotics (usually vancomycin and ceftazidime) usually are initiated at the same time and usually are discontinued after 72 hours. The use of intraoperative neurophysiological monitoring necessitates the use of certain anesthetic agents or intraoperative switching from one agent to another (for example, no muscle relaxation medications in some phases of the surgery). In some instances, when temporary occlusion of a major blood vessel is necessary, an ultrashort barbiturate (thiopental) and burst suppression on electroencephalography may be used.

Intraoperative neurophysiological monitoring is an important adjunct in the surgery of FMMs (Fig. 3). This monitoring helps prevent operative injury of cranial nerves, the brain stem, and the cervical spinal cord. It also helps decrease postoperative complications. Somatosensory evoked potentials and brain stem auditory evoked potentials are monitored bilaterally. Electromyographic monitoring of the vagal, accessory, and hypoglossal nerves should be done ipsilateral to the lesion, or bilaterally when necessary. The vagus nerve is monitored with an EMG endotracheal tube or by the laryngeal surface electrode that is placed preoperatively after the intubation. The accessory and

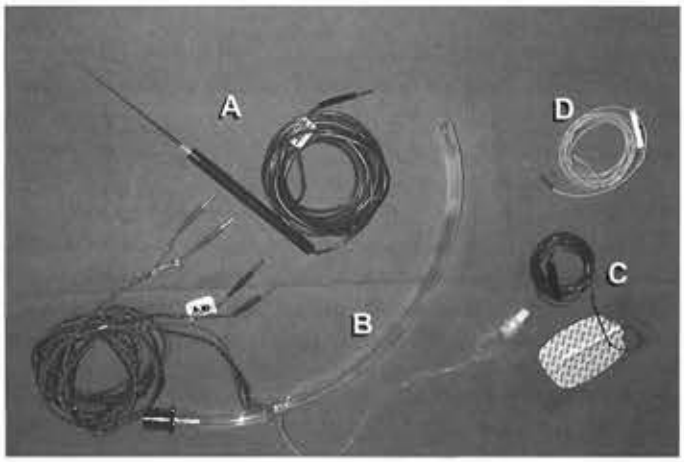

Figure 3. Some important tools for intraoperative diagnostic monitoring. A: A nerve stimulator. B: EMG endotracheal tube for monitoring cranial nerves IX and X. C and D: EMG electrodes.

hypoglossal nerves are monitored with electrodes placed directly into the trapezius muscle and the inferolateral aspect of the tongue, respectively. Any change in the monitoring signal warns the surgeon to take appropriate action. Such extensive intraoperative neurophysiologic monitoring helps to preserve important neurovascular structures, particularly lower cranial nerves; this preservation may, in turn, decrease the rate of complications or the time of the deficit recovery.

Readings

Al-Mefty O: Part 4, Meningiomas of the craniovertebral junction, in Al-Mefty O (ed): *Operative Atlas of Meningiomas*. Philadelphia: Lippincott-Raven, 1997, pp 349–382.

Al-Mefty O, Borba LAB, Aoki N, et al: The transcondylar approach to extradural nonneoplastic lesions of the craniovertebral junction. *J Neurosurg* 84: 1, 1996

Arnautovic KI, Al-Mefty O: Ventral foramen magnum meningiomas. *J Neurosurg* (Spine 1) 92:71, 2000

Arnautovic KI, Al-Mefty O: Foramen magnum meningiomas, in Kaye A, Black PMcL, (eds): *Operative Neurosurgery*. London: Harcourt Brace, 2000, pp 623–634

Arnautovic KI, Al-Mefty O, Pait TG, et al: The suboccipital cavernous sinus. *J Neurosurg* 86:252, 1997

Blom S, Ekbom KA: Early clinical signs of meningiomas of the foramen magnum. *J Neurosurg* 29:661, 1962

Caruso RD, Rosenbaum AE, Chang JK, et al: Craniocervical junction venous anatomy on enhanced MRI images: the suboccipital cavernous sinus. *AJNR* 20:1127, 1999

Castellano F, Ruggiero G: Meningiomas of the posterior fossa. *Acta Radiol* (Suppl) 104:1, 1953

Cohen L, Macrae D: Tumors in the region of the foramen magnum. *J Neurosurg* 19: 462, 1962

Cushing H, Eisenhardt L: *Meningiomas: Their Classification, Regional Behavior, Life History and Surgical End Results*. Springfield, IL: Charles C. Thomas, 1938

Dodge HW, Love JG, Gottlieb CM: Benign tumors of the region of the foramen magnum. *J Neurosurg* 13:603, 1956

George B, Lot G, Boissonnet H: Meningioma of the foramen magnum: A series of 40 cases. *Surg Neurol* 47: 371, 1997

Guidetti B, Spallone A: Benign extramedullary tumors of the foramen magnum. *Adv Tech Stand Neurosurg* 16:83, 1988

Haddad GF, Al-Mefty O: Infratentorial and foramen magnum meningiomas, in Wilkins RH, Rengachary SS (eds): *Neurosurgery*. 2nd ed. New York: McGraw-Hill, 1996, pp 951–958

Menezes AH, Traynelis VC: Tumors of the craniovertebral junction, in Youmans JR (ed): *Neurological Surgery*. Philadelphia: WB Saunders, 1996, pp 3041–3072

Meyer FB, Ebersold MJ, Reese DF:. Benign tumors of the foramen magnum. *J Neurosurg* 61:136, 1984

Pieper DR, Al-Mefty O, Hanada Y, et al: Hyperostosis associated with meningioma of the cranial base: secondary changes or tumor invasion. *Neurosurgery* 44: 742, 1999

Russell JR, Buery PC: Meningiomas of the posterior fossa. *Surg Gynecol Obstet* 96: 183, 1953

Scott EW, Rhoton AL Jr: Foramen magnum meningiomas, in Al-Mefty O (ed): *Meningiomas*. New York: Raven Press, 1991, pp 543–568

Scott M: The surgical management of meningioma of the cerebellar fossa. *Surg Gynecol Obstet* 135: 545, 1972

Simpson D: The recurrence of intracranial meningiomas after surgical treatment. *J Neurol Neurosurg* Psychiatry 20:22, 1957

Stein B, Leeds NE, Taveras JM, et al: Meningiomas of the foramen magnum. *J Neurosurg* 20:740, 1963

Welling B, Al-Mefty O: Foramen magnum tumors, in Cohen AR (ed): *Surgical Disorders of the Fourth Ventricle*. Cambridge: Blackwell Science, 1996, pp 251-261

Welling B, Park Y K, Al-Mefty O: Primary extramedullary tumors of the craniovertebral junction, in Dickman CA, Spetzler RF, Sonntag VKH (eds): *Surgery of the Craniovertebral Junction*. New York: Thieme, 1998, pp 239–252

Yasargil MG, Mortara RW, Curcic M: Meningiomas of basal posterior cranial fossa. *Adv Tech Stand Neurosurg* 7:1, 1980

Yasuoka S, Okazaki H, Daube JR, et al: Foramen magnum tumors: analysis of 57 cases of benign extramedullary tumors. *J Neurosurg* 49: 828, 1978

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1. Craniospinal foramen magnum meningiomas are more medial than spinocranial foramen magnum meningiomas.

True or False?

2. Spinocranial meningiomas are thought to arise from the upper spinal dura and grow upward.

True or False?

3. Spinocranial meningiomas are much more challenging surgically than are craniospinal meningiomas.

True or False?

4. Meningiomas are the most common extramedullary tumors of the foramen magnum.

True or False?

5. A foramen magnum meningioma may mimic multiple sclerosis in the cervical disc on presentation.

True or False?

6. The motor and sensory hand deficit known as "cold clumsy hand" is a common presentation in foramen magnum meningiomas.

True or False?

7. Intraoperative neurophysiological monitoring is not essential in the surgery of foramen magnum meningiomas.

True or False?

8. Postoperative MRI enhancement may persist for several years after resection of foramen magnum meningiomas before the enhancement diminishes.

True or False?

9. Lower cranial nerve dysfunction is the source of the most serious morbidity after resection of foramen magnum meningiomas.

True or False?

10. Unlike most other meningiomas, foramen magnum meningiomas are more common in males.

True or False?