

Jugular Fossa Lesions

Kenan I. Arnautović, M.D., and Ossama Al-Mefty, M.D.

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

1. Describe the basic anatomy of the jugular fossa and foramen.
2. Describe the differential diagnosis of jugular fossa lesions.
3. Recall basic preoperative diagnostics and operative approaches to jugular fossa lesions.

The treatment of jugular fossa lesions has been revolutionized by the development of modern diagnostic modalities, refinement of microsurgical techniques, publication of microsurgical anatomic studies, development of skull base approaches, advances in neuroanesthesia and intraoperative neurophysiologic monitoring, and careful multidisciplinary perioperative planning. These lesions now are treated with radical resection, and the rates of permanent surgical morbidities or mortalities are low.

Anatomic Considerations

The jugular foramen is a skull opening, or gap, that connects the posterior cranial fossa and the jugular fossa. It is formed by the jugular incisuras in the temporal and occipital bones. It lies in an oblique position, from the lateral aspect posteriorly toward the medial aspect anteriorly. Classically, it is described as subdivided by a fibrous or bony bridge (the intrajugular septum) into two parts, which serve as a passage for Cranial Nerve IX and the inferior petrosal sinus (pars nervina or nervosa) and for Cranial Nerves X and XI and the jugular vein. More recent anatomic studies have disputed this theory of the anatomic organization of the jugular foramen, and there still is no single accepted view of its anatomy.

The jugular fossa, located at the inferior aspect (inferior surface) of the petrous part of the temporal bone, is a deep depression, the size of which varies from skull to skull. It communicates with the posterior cranial fossa via the jugu-

lar foramen. It lodges the jugular bulb, which continues into the jugular vein inferiorly (Fig. 1).

In the neurosurgical literature, and even in extensive anatomic studies, both the jugular foramen and the jugular fossa often are referred to by the term "jugular foramen." This use of the term may be the result either of simple error or the user's wish to provide a broader anatomic description of the area, and this confusion may be the underlying reason for the current lack of agreement regarding the internal anatomic organization of "the jugular foramen." Regardless of the reason for this mix-up, the jugular foramen and the jugular fossa are two distinct anatomic formations, although they are intimately related.

Differential Diagnosis

The most common differential diagnoses of lesions in the jugular fossa include, in descending order of frequency, glomus jugulare tumors, neurinoma of the lower cranial nerves (Cranial Nerves IX–XI), and meningiomas (Figs. 2–4). The broader differential diagnosis of jugular fossa tumors may include chordoma, chondrosarcoma, primary cholesteatoma, plasmacytoma, epidermoid tumors, choroid plexus papilloma, chondroma, temporal bone carcinoma, salivary gland tumors, aneurysm, metastases, and cerebellar hemangioblastoma.

The preoperative radiologic diagnosis and differential diagnosis are important when jugular fossa lesions are the subject, because preoperative management and operative planning may differ considerably depending on the type of lesion in question, e.g., planning preoperative embolization for glomus jugulare tumors, or determining how much of the tumor involves bone if it is a meningioma.

Category: Tumor, Anatomy

Key Words: Jugular Fossa, Jugular Foramen, Glomus Jugulare, Neurinoma, Meningioma

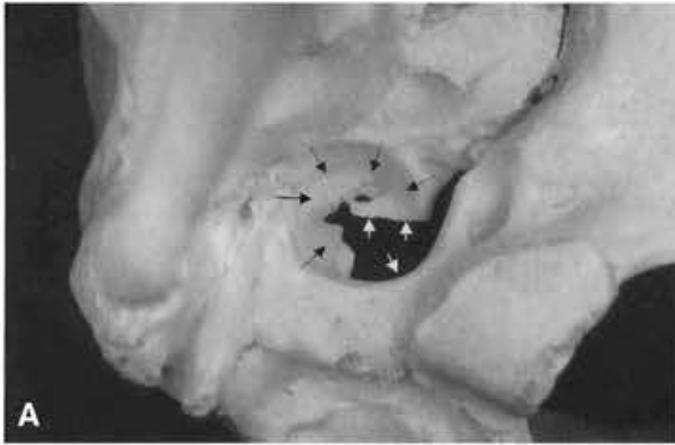


Figure 1. *A*, Right-side, dry anatomic specimen photograph delineating the jugular foramen (*white arrows*) and the jugular fossa (*black arrows*). Outside-inside view from the anterior perspective. *B*, Right-side dry anatomic specimen photograph delineating the jugular foramen (*black arrows*) and the jugular fossa (*white arrows*). Inside-outside view from the posterior perspective. Note the difference between the two.

Glomus jugulare tumors have a “salt-and-pepper” appearance on noncontrast-enhanced T1-weighted MRI scans, which represent a flow void network from their rich vascularity and enhance nonhomogeneously after the injection of con-



Figure 2. Glomus jugulare tumor. *A*, Axial T1-weighted noncontrast image showing the salt-and-pepper tumor appearance in the right jugular fossa (*). *B*, Sagittal T1-weighted postcontrast MRI scan showing the nonhomogeneous enhancement of the tumor (*).

trast material (Fig. 2). However, they have a propensity to erode and destroy the bone, particularly the jugular spine

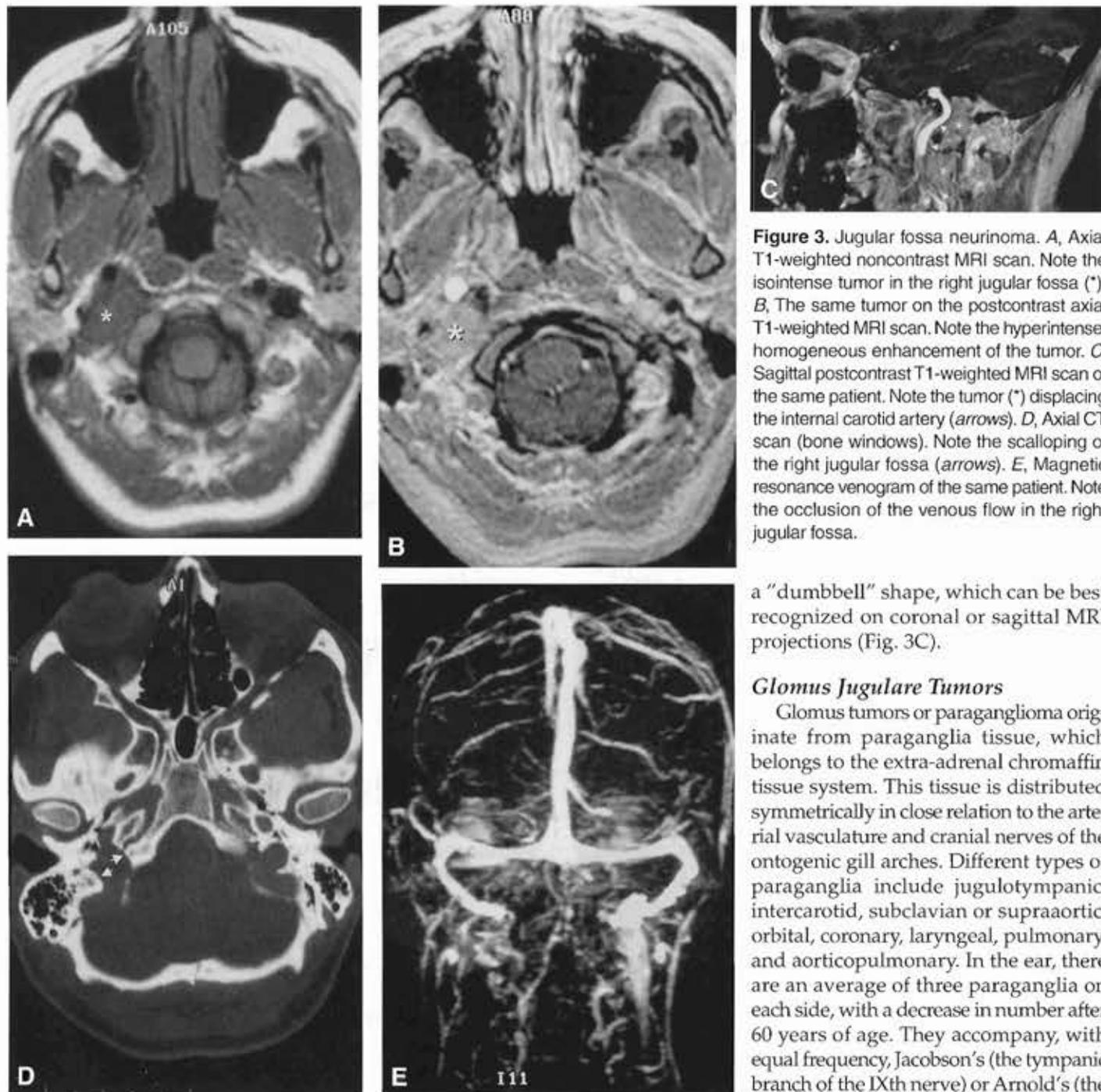


Figure 3. Jugular fossa neurinoma. *A*, Axial T1-weighted noncontrast MRI scan. Note the isointense tumor in the right jugular fossa (*). *B*, The same tumor on the postcontrast axial T1-weighted MRI scan. Note the hyperintense, homogeneous enhancement of the tumor. *C*, Sagittal postcontrast T1-weighted MRI scan of the same patient. Note the tumor (*) displacing the internal carotid artery (arrows). *D*, Axial CT scan (bone windows). Note the scalloping of the right jugular fossa (arrows). *E*, Magnetic resonance venogram of the same patient. Note the occlusion of the venous flow in the right jugular fossa.

a “dumbbell” shape, which can be best recognized on coronal or sagittal MRI projections (Fig. 3C).

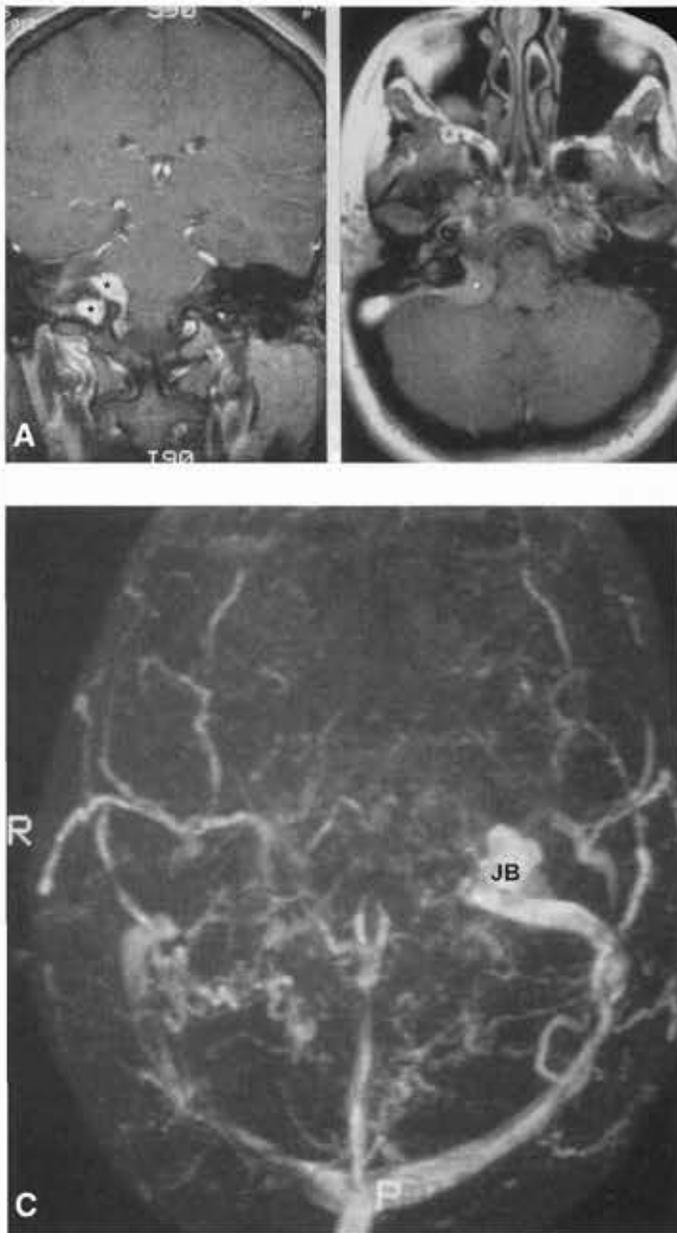
Glomus Jugulare Tumors

Glomus tumors or paraganglioma originate from paraganglia tissue, which belongs to the extra-adrenal chromaffin tissue system. This tissue is distributed symmetrically in close relation to the arterial vasculature and cranial nerves of the ontogenic gill arches. Different types of paraganglia include jugulotympanic, intercarotid, subclavian or supraaortic, orbital, coronary, laryngeal, pulmonary, and aorticopulmonary. In the ear, there are an average of three paraganglia on each side, with a decrease in number after 60 years of age. They accompany, with equal frequency, Jacobson’s (the tympanic branch of the IXth nerve) or Arnold’s (the auricular branch of the Xth nerve) nerves.

and the carotid crest (caroticojugular spine)—the bone that separates the petrous carotid artery from the jugular bulb. Meningiomas often invade the bone, including the jugular spine and particularly the jugular tubercle, producing “hyperostosis” and bone thickening, but not bone erosion (Fig. 4B).

Jugular fossa neurinomas originate from the lower cranial nerves. They enlarge the jugular fossa with smooth, distinct sclerotic margins seen on CT scans with bone window (“bone scalloping”) (Fig. 3D). They may contain cysts, which appear as hypointensities on T1-weighted MRI scans. Frank bony invasion is very rare. Although they may mimic the appearance of meningiomas on both T1- and T2-weighted MRI scans, the enhancement seen in meningiomas usually is considerably greater. Neurinomas often assume

More than 50% are located in the jugular fossa; the rest are located in the middle ear. They appear encapsulated, lobulated, and tan-gray to purple-red in color. Their vascular supply is derived from the postauricular, occipital, internal maxillary, and internal carotid arteries. Although every glomus tumor may secrete catecholamines and other neuropeptides, only 1% to 3% of these tumors present with clinically detectable symptoms, because the serum norepinephrine must be elevated to at least four to five times the normal level before symptoms (e.g., headaches, excessive perspiration, tachycardia, pallor, and nausea) are produced. Thus, evaluation of serum and urine levels of catecholamines is part of the routine preoperative evaluation. In these cases, preoperative administration of alpha- and beta-blockers is important.



Although the incidence of glomus jugulare tumor is low, it is the most common lesion in the jugular fossa. It tends to extend into the middle ear, neck, and posterior cranial fossa. Superselective embolization with absorbable materials may be used as adjuvant therapy in the treatment of large tumors. Complications of embolic therapy may include skin necrosis, cranial nerve palsies, and strokes. Multicentricity and metastases are possible in these tumors, particularly in cases with the familial father-to-daughter inheritance pattern. Females are more commonly affected than males, by a ratio of 6 to 1. The incidence peaks in the fifth and sixth decades of life. The most common clinical presentation of these tumors includes lower cranial nerve and hearing deficits. Pulsatile tinnitus, facial nerve weakness, and vertigo also are common. The most common postoperative complications include deficits of the lower cranial nerves (Cranial Nerves IX–XII) and Cranial Nerve VII (Fig. 2).

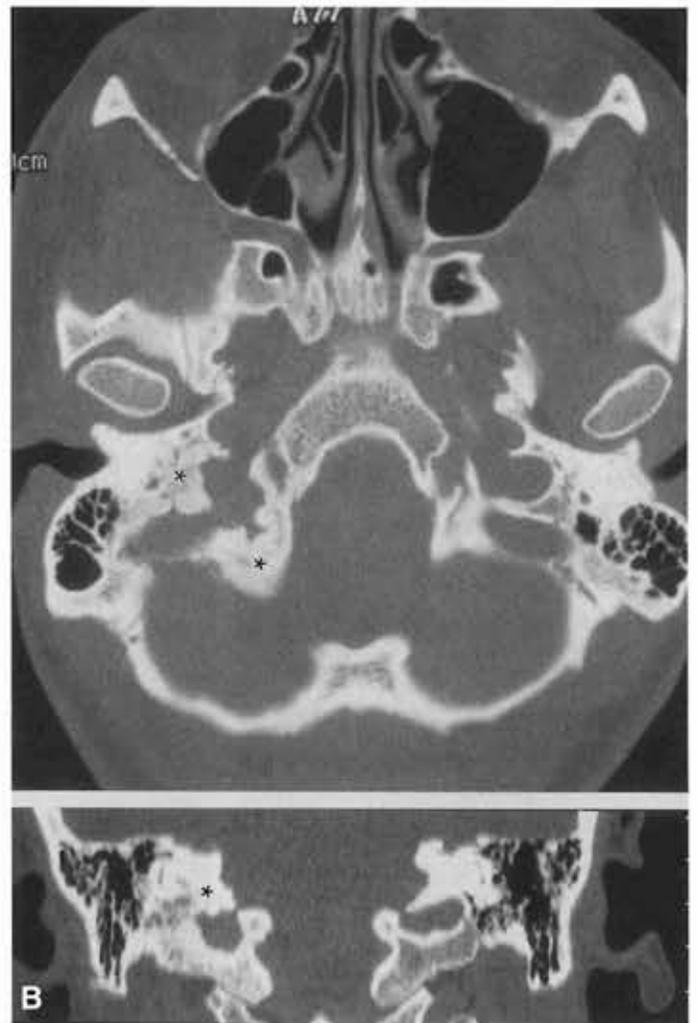


Figure 4. Jugular fossa meningioma. *A*, Postcontrast T1-weighted MRI scan showing the right jugular fossa tumor homogeneously enhancing (*). *Left*, coronal projection; *right*, axial projection. *B*, CT scan ("bone window") showing the bone invasion ("hyperostosis") by the tumor (*). *Top*, axial reconstruction; *Bottom*, coronal reconstruction. *C*, Axial reprojection of the magnetic resonance venogram. Note the occluded jugular bulb on the right (tumor) side and the patent one (*JB*) on the left side.

Jugular Fossa Neurinomas (Lower Cranial Nerve Schwannomas)

Jugular fossa neurinomas arise, in descending order of frequency, from Cranial Nerves IX, X, and XI. They account for 2.9% of all intracranial neurinomas, and are the fourth most common type, after neurinomas of Cranial Nerves VIII, V, and VII. The most common clinical presentations include palsies of Cranial Nerves IX, X, and XI; hearing loss; and facial weakness. Other presentations include altered facial sensation, tongue atrophy, and cerebellar symptoms. These lesions are more common in females between the ages of 14 and 63 years (Fig. 3).

Jugular fossa neurinomas may compress or occlude the jugular bulb. The most common postoperative complications include deficits of the lower cranial nerves (Cranial Nerves IX–XII), cerebrospinal fluid leak, and meningitis.

Primary Jugular Fossa Meningiomas

Primary jugular fossa meningiomas are rare. They are believed to arise from the arachnoid cells lining the jugular

bulb. They may compress the lower cranial nerves; invade the temporal bone; invade, compress, obstruct, or narrow the jugular bulb; or extend into the posterior cranial fossa or extracranially. To date, about 50 cases of jugular fossa meningioma have been reported. Women account for between 67% and 100% of the patients in the published series, and the mean age of patients in these studies ranges from 32 to 49 years. The most common presentation is deficit of the lower cranial nerves (Cranial Nerves IX–XII), followed by hearing deficits and facial numbness resulting from compression of the spinal trigeminal tract. Meningothelial meningiomas, followed closely by transitional meningiomas, are the most common histological variant. Atypical and malignant forms also have been reported (Fig. 4).

As many as 20% of all intracranial meningiomas eventually extend extracranially, although extensions in other locations (e.g., the orbit and the external table of the calvaria, followed by the nasal cavity and the paranasal sinuses) are more common. In cases of extracranial extension, the meningioma invades the parapharyngeal space through either the foramen lacerum or the jugular fossa.

As with other lesions in this location, the main postoperative concern pertaining to jugular fossa meningioma is a deficit of the lower cranial nerves (Cranial Nerves IX and X) resulting in swallowing deficits. If the nerves are anatomically preserved and "gently" manipulated, recovery of or compensation for swallowing deficits may be expected within 30 days. Other postoperative complications may include weakness of Cranial Nerves XI and XII.

Perioperative Protocol

The perioperative protocol described here has been developed over the years of our experience and is based on the general principles of perioperative skull base and posterior fossa management. Treatment is tailored to each patient. The extensive preoperative diagnostic neuroradiologic work-up includes MRI with and without contrast, magnetic resonance angiography in both arterial (MRA) and venous (MRV) phases, and CT. Imaging studies are used to define the local anatomy of the jugular foramen and fossa, the tem-



Figure 5. The operative position of the patient for a right-sided jugular fossa tumor. Note the skin incision behind and inferior to the auricle, the frame for image-guided navigation in position, and the intraoperative neurophysiologic electrodes being placed in position.

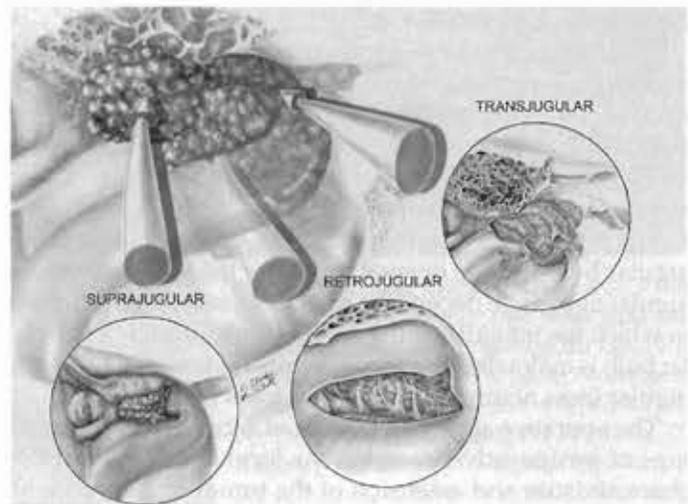


Figure 6. Artistic rendering of the three approaches used for the resection of a jugular fossa tumor on the right side.

poral bone, and the condyles. We also study the tumor's involvement of the bone (e.g., erosion, "scalloping," or hyperostosis) and calcification within the tumor using CT; and the tumor's features, extensions, and consistency, using CT and MRI with and without contrast. Studies also are performed to evaluate the regional vascular anatomy, the vertebrobasilar arterial system, the size and dominance of the transverse and sigmoid sinuses, the superior and inferior petrosal sinuses, the bilateral vein of Labbe, and the position, patency, and size of the jugular bulb using MRA and MRV. We abandoned the routine use of four-vessel angiography in neurinomas and meningiomas, because MRA and MRV provide most of the necessary preoperative information regarding the vascular anatomy. Four-vessel angiography still is important in glomus jugulare tumors, however, especially for identifying possible concomitant glomus tumor on the other side and when preoperative embolization is required. Five years ago, we started to use intraoperative image-guided frameless stereotaxy. Postoperative imaging includes a CT scan 24 hours after the surgery and MRI in the early postoperative period; at 3, 6, and 12 months; and annually thereafter.

Perioperative multidisciplinary evaluations include audiologic evaluation, both before and after surgery; speech pathology and otolaryngological evaluations; and pre- and postoperative swallowing studies. Patients are given a "nothing by mouth" (NPO) regimen postoperatively, with parenteral nutrition until swallowing studies show satisfactory results. If a patient has difficulty swallowing, appropriate actions—such as an NPO regimen with parenteral nutrition, swallowing exercises, soft mechanical diet with swallowing precautions, and vocal cord medialization—are taken immediately, as needed.

Intraoperative neurophysiologic monitoring includes somatosensory evoked potentials and brain stem auditory evoked potentials bilaterally. Cranial Nerve X is monitored through an electromyography endotracheal tube. Cranial Nerves VII, XI, and XII are monitored unilaterally by needle insertion in the corresponding muscle.

Operative Approach

The approach to a jugular fossa tumor should be tailored to each patient. Because these tumors involve the jugular fossa and beyond, the skull base approach is required for their removal. However, the patency and dominance of the involved jugular bulb dictate the surgical approach. Three approaches are possible: the transjugular approach, commonly practiced in resection of glomus tumors, in which the jugular bulb itself is opened; or either the supra- or retrojugular approach, depending on the extension of the tumor, in which the integrity of the venous flow through the jugular bulb is maintained, as is commonly done in patients with jugular fossa neurinoma or jugular fossa meningioma.

The operative approach is tailored according to the findings of preoperative imaging, the local anatomy, and the characteristics and extension of the tumor in each patient (Figs. 3–9). Three different routes are used:

1. The suprajugular approach. This presigmoid, infralabyrinthine route is chosen if the jugular bulb is patent and the tumor extends primarily anteriorly (Fig. 7).
2. The retrojugular approach. This transcondylar, transtuberacle route is used if the jugular bulb is patent and the tumor was extending predominantly behind it (Fig. 8).
3. The transjugular approach. This is an infratemporal route, used in cases in which the jugular bulb is totally occluded by a tumor (Fig. 9).

Suprajugular Approach

In the suprajugular approach, the patient is placed supine with a shoulder roll under the shoulder and the head elevated, turned away from the lesion, and fixed in a Mayfield headholder. The skin incision circles behind the ear, starting at the temporal area anteriorly and superiorly and extending down into the upper neck horizontally along the skin crease. The skin flap is then retracted anteriorly and inferiorly. The sternocleidomastoid muscle is retracted inferiorly and posteriorly. A mastoidectomy is then performed, followed by complete skeletonization of the sigmoid sinus, the jugular bulb, and the jugular vein. The jugular fossa is accessed in the presigmoid, infralabyrinthine space.

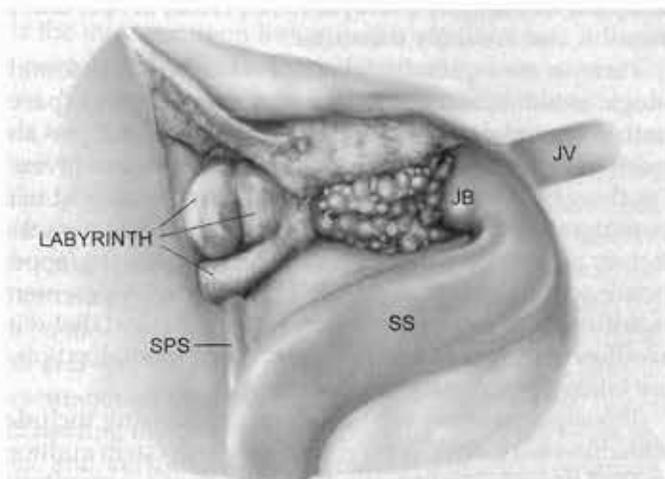


Figure 7. The suprajugular approach. Note the infralabyrinthine position of the tumor, the sigmoid sinus (SS); the jugular bulb (JB), which is patent, the jugular vein (JV); the superior petrosal sinus (SPS); and the labyrinth (right side).

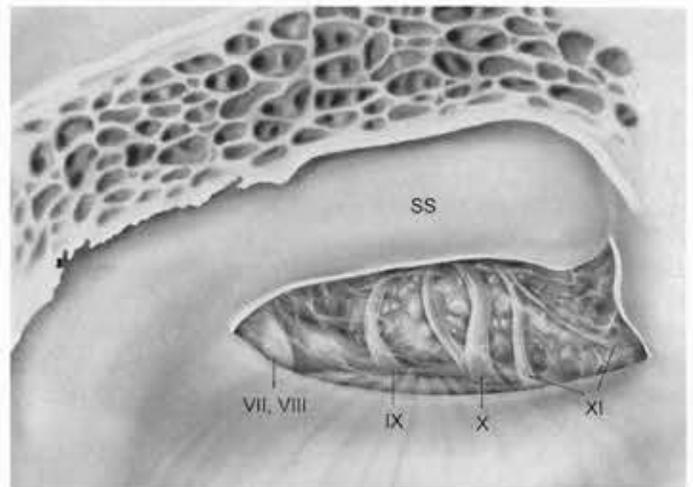


Figure 8. The retrojugular approach. Note the retrojugular position of the tumor covered by the arachnoid membrane, the sigmoid sinus (SS), Cranial Nerves VII and VIII, and the lower cranial nerves (IX–XI) stretched over the tumor (right side).

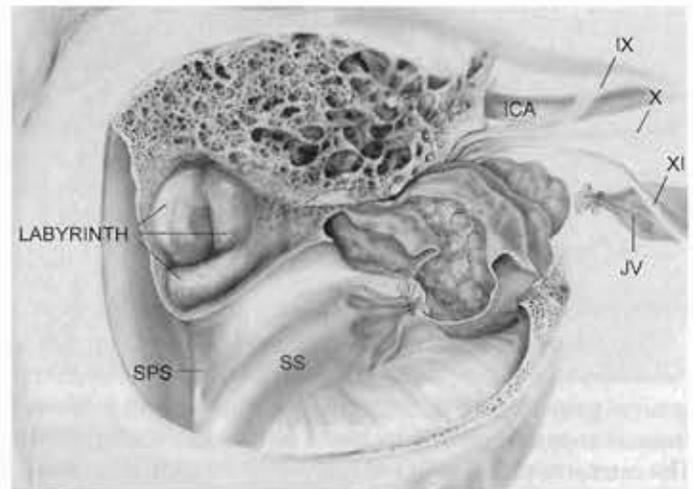


Figure 9. The transjugular approach. Note the tumor occluding the jugular bulb, which is open, the sigmoid sinus (SS) and the jugular vein (JV), both of which are ligated; the superior petrosal sinus (SPS); the internal carotid artery (ICA); and the lower cranial nerves extracranially (IX–XI).

The dura superior to the patent jugular bulb and inferior to the labyrinth is opened. The cerebrospinal fluid is then released from the cerebellomedullary cistern and tumor is dissected away from the lower cranial nerves (Cranial Nerves IX – XI), the posteroinferior (PICA) and anteroinferior (AICA) cerebellar arteries, and the vertebral artery (VA) respecting the arachnoidal planes. Debulking of the tumor is done by suction and bipolar coagulation or by an ultrasonic aspirator and is completed with microsurgical radical resection of the tumor (Fig. 7).

Retrojugular Approach

In the retrojugular approach, the patient is placed supine and rotated about 40 degrees, keeping the head and neck elevated 30 degrees in neutral position and fixing it in the Mayfield headholder. The skin incision is curved around the ear two fingerbreadths behind the mastoid, extending down transversely along the horizontal neck crease. The muscles of the neck are dissected in three layers, beginning with detachment

of the sternocleidomastoid muscle from the superior nuchal line on the occipital bone. After the muscles are dissected and reflected medially and inferiorly, the VA, surrounded by a complex venous structure, is encountered. Transposition of the VA complex usually is not needed for the jugular fossa tumor surgery. The jugular vein and the lower cranial nerves (IX–XII) are exposed in the neck at the lower aspect of incision. A lateral, posterior fossa craniotomy is then performed, and the mastoid tip drilled to expose the occipital condyle. Approximately one-third of the condyle drilling usually suffices for the exposure, and stabilization is not necessary. Attention is then turned to the jugular tubercle, which is completely drilled away, thus facilitating the opening of the jugular fossa lodging the jugular bulb. Bone work is completed with a partial mastoidectomy, completely skeletonizing the sigmoid sinus, the jugular bulb, and the jugular vein. The dura is incised under the microscope along the posterior border of the sigmoid sinus, which then is tented by the suture and gently retracted laterally. The tumor is then carefully separated from the medulla, the lower cranial nerves, the VA, and the PICA along arachnoidal planes, and followed toward the jugular fossa and anteriorly. Careful, meticulous dissection of the tumor from the jugular bulb and vein wall is essential. Ultrasonic aspirator or suction and bipolar coagulation are used. The involved dura and the bone of the jugular fossa should be resected to minimize the chances for future tumor recurrence (Fig. 8).

Transjugular Approach

The patient is placed supine, with the head elevated, turned away from the lesion, and secured in the Mayfield head rest.

The skin incision is curved around the ear superiorly to the temporal area and down transversely along the natural skin crest in the neck. If the hearing has been lost preoperatively, the external ear canal is transected at the bony cartilaginous junction, and the skin of the external ear canal is everted and closed as a blind sac. The sternocleidomastoid muscle is detached from its mastoid insertion. The neurovascular structures in the neck (Cranial Nerves IX–XII, the jugular vein, and the carotid artery) are dissected and exposed.

A radical mastoidectomy exposes the sigmoid sinus down to the jugular bulb and is followed by a posterior fossa craniotomy. In some cases, it may be necessary to skeletonize and transpose the facial nerve. The jugular vein is followed superiorly to the jugular bulb, and the bone is skeletonized completely. To enlarge the exposure, the posterior belly of the digastric muscle and the stylohyoid muscle are transected and the styloid process removed. The sigmoid sinus and the jugular vein are then ligated proximally to the mastoid emissary vein and distally to the tumor obstruction of the jugular bulb. The inferior pole of the tumor is then dissected off the internal carotid artery and the jugular vein, and the extradural tumor is thus completely exposed. Bleeding from the inferior petrosal sinus, which may be profuse, is controlled by packing with Gelfoam. Using the microscope, the dura mater is then opened posteriorly to the sigmoid sinus and carried forward. The intradural tumor extension is then exposed. Meticulous intradural dissection of the tumor respecting the arachnoidal planes helps functional preservation of the lower cranial nerves, the VA, the PICA, the AICA, and the anterolateral surface of the medulla (Fig. 9).

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