52 Surgery of Paraganglioma

Yoichi Nonaka and Takanori Fukushima

Abbreviations

CN	Cranial nerve
EAC	External auditory canal
GJ	Glomus jugulare tumor
GKS	Gamma knife surgery
GT	Glomus tympanicum
GV	Glomus vagale
HC	Hypoglossal canal
ICA	Internal carotid artery
IJV	Internal jugular vein
IPS	Inferior petrosal sinus
JB	Jugular bulb
JF	Jugular foramen
JT	Jugular tubercle
JTP	Jugulotympanic paragangliomas
LCN	Lower cranial nerve
OC	Occipital condyle
RLA	Retrolabyrinthine approach
SCC	Semicircular canals
SRS	Stereotactic radiosurgery
SRT	Stereotactic radiotherapy
SS	Sigmoid sinus
STR	Subtotal resection
TLA	Translabyrinthine approach

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Introduction

Jugulotympanic paragangliomas (JTP) are rare tumors of the autonomic nervous system, accounting for about 0.5% of all head and neck tumors [1, 2]. They are mostly benign and may arise from the paraganglia of the skull base and the neck. JTP are previously known as glomus jugulare tumors (GJ), a name that is still commonly used [3]. Surgery of JTP is difficult and challenging for head and neck surgeons and neurosurgeons due to their location in the vicinity of important blood vessels and cranial nerves. JTP may be classified according to their location and endocrine functionality and involve the head and neck region in 3% of cases [4, 5]. Within the head and neck, these tumors arise in four main locations: the carotid space as carotid body tumors, the middle ear as glomus tympanicum (GT), the jugular foramen (JF) as glomus jugulare (GJ), and along the vagus nerve as glomus vagale (GV).

JTP are benign slow-growing tumors but frequently involve critical neurovascular structures around the jugular bulb (JB) such as lower cranial nerves (LCNs) and the internal carotid artery (ICA). They also often extend into the intradural

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and high cervical spaces. Therefore, JTP continue to pose a significant challenge to skull base surgeons. Despite the development of surgical techniques, approaches, and treatment modalities, management for JTP is still controversial (Chap. 48). They include microsurgery with preoperative feeder embolization and radiotherapy/radiosurgery (SRT/SRS). There has been an increasing interest in SRS for JTP in recent decades, given similar control rates and potentially lower complication rates when compared with surgical excision [6]. This chapter will focus on surgical management of advanced grade GJ extending to the infratemporal fossa.

Operating Room Equipment, Imaging Technology, and Surgical Instruments

The stage of GJ and the degree of ICA involvement are the most important factors to determine the surgical strategy. MRI with and without gadolinium enhancement is performed to delineate the tumor size, location, and extent. Postcontrast MRIs help understand the relationship between the tumor and the sigmoid sinus (SS), JB, and the internal jugular vein (IJV). The tumor's relationship to the JF, cranial nerves, brainstem, temporal bone, and neighboring structures is also examined carefully. Evidence of tumor involvement of ICA can be determined on these images. A thin slice of bone CT scan of the skull base is useful to evaluate the anatomy of the temporal bone and the extent of bony destruction by the tumor. It also helps to understand the relationship between the tumor and the Fallopian canal segment of the facial nerve in the mastoid body.

Angiographic studies are essential and critical to assess the feeding arteries and the venous drainage of the tumor. In patients who have tumor involvement of the ICA, a balloon occlusion test is performed to assess the risk involved with performing an ICA sacrifice and subsequent reconstruction of the ICA with a high-flow bypass. Preoperative feeder embolization is an excellent adjunctive modality for reducing intraoperative blood loss [7, 8]. Arterial feeders most commonly arise from the ascending pharyngeal artery and feeders from the external carotid artery.

The preparation of sophisticated surgical instruments and equipment lead a surgeon to safe surgery. A high-speed electronic drill with various diamond burrs (3 mm, 4 mm, 5 mm size of extra-coarse or coarse diamond burrs) is essential for the mastoidectomy. Micro instruments including rigid skull base microdissectors, thin, medium, and thick blade microscissors, teardrop suctions (Fukushima designed; Fujita Medical Instruments, Tokyo Japan), nonstick bipolar forceps of three types (SilverGlide keyhole type and pro-series, Vesalius, and Tokyo Fujita micro-bipolar), and ultrasonic tumor aspirator are indispensable for tumor resection. Intraoperative electrophysiological monitoring (NIM-Response 3.0 Stimulator™; Medtronic Xomed, Inc. Jacksonville, FL) is also necessary to identify the Fallopian canal segment of the facial nerve in the mastoid body. In surgeries of large or complicated tumors at an unfamiliar location, it is essential to determine the anatomical orientation in every step, but tumor compression disrupts normal anatomy. Once the key structure is identified during the procedure in a limited view, the neuronavigation system should be introduced to ensure the displaced anatomy of surrounding structures by the tumor. The surgical resection safety can be augmented by the adjuvant use of a Doppler ultrasound device to identify the ICA involved by the GJ.

Intraoperative monitoring indispensable for GJ surgery includes somatosensory evoked potentials, motor evoked potentials and facial nerve monitoring. Continuous auditory brainstem response recording should be used for cases with serviceable hearing. The LCNs can also be monitored using an electromyographic endotracheal tube (NIM TriVantage™ EMG Endotracheal Tube; Medtronic Xomed, Inc. Jacksonville, FL). These are strongly recommended as risk-minimizing tools.

Surgical Anatomy

The GJ arise from adventitial chemoreceptor tissue in the JB. The JB is the connection between the SS, and the IJV, which lies in the pars vascularis (pars venosa) occupies the posterolateral aspect of the JF. The pars vascularis also contains the vagus nerve (CN X) and spinal accessory nerve (CN XI). The pars nervosa is the anteromedial portion of the JF and is smaller than the pars vascularis. It contains inferior petrosal sinus (IPS) and glossopharyngeal nerve (CN IX) [9]. The LCNs always pass medially to the JB. Therefore, the GJ arising in the JB displace the LCNs medially by their growth, creating a more favorable position for nerve preservation during resection [10]. The IPS, which often consists of multiple channels, courses from the cavernous sinus and empties into the medial aspect of the JB. The position of the LCNs with respect to the IPS is variable. The IPS will often pass between the CN IX anteriorly and the CN X and XI nerves posteriorly. Therefore, overpacking of the IPS for hemostasis or cautery in this area may cause nerve injuries.

The JB is located beneath the semicircular canals (SCCs) in the mastoid body. The transmastoid approach (mastoidectomy), which includes retrolabyrinthine approach (RLA) and translabyrinthine approach (TLA), is the only way to expose vicinity of the JB. The RLA exposes the SS, presigmoid dura, SCCs, and JB (Fig. 52.1). When hearing preservation is not considered, the region around JB can be more widely exposed by removing SCCs through the TLA.

The Fallopian segment of the facial nerve is the anterior limit of exposure in the standard transmastoid approach. Therefore, further bone removal anterior to the facial nerve, including a facial recess, the bony external auditory canal (EAC), and hypotympanum area, provides a much wider operative space than the standard transmastoid exposure [11–13]. A thin layer of cortical bone is left to cover the entire Fallopian segment of the facial nerve from the genu to the stylomastoid foramen, creating the "Fallopian bridge" (Fig. 52.2). Careful drilling anteroinferior to the Fallopian bridge results in exposure of the vertical segment of the ICA (C7).

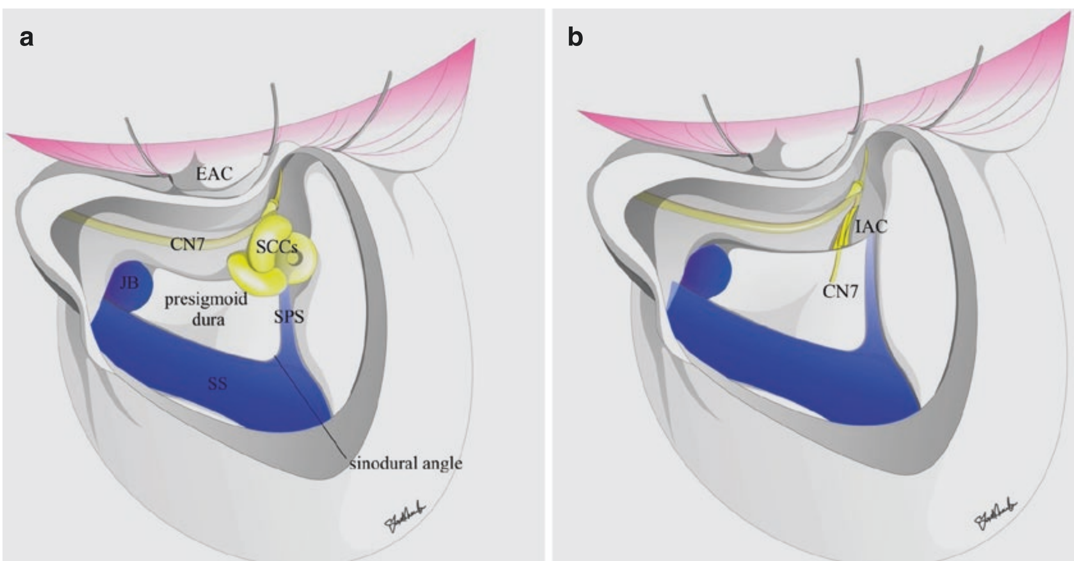

Fig. 52.1 Transmastoid approaches. The schematic illustrations demonstrate the left side transmastoid approach (**a**) retrolabyrinthine approach (RLA), (**b**) translabyrinthine approach (TLA)). The semicircular canals (SCCs) are kept intact to preserve hearing function in the RLA. The presigmoid dura is exposed much wider in the TLA with SCCs removal

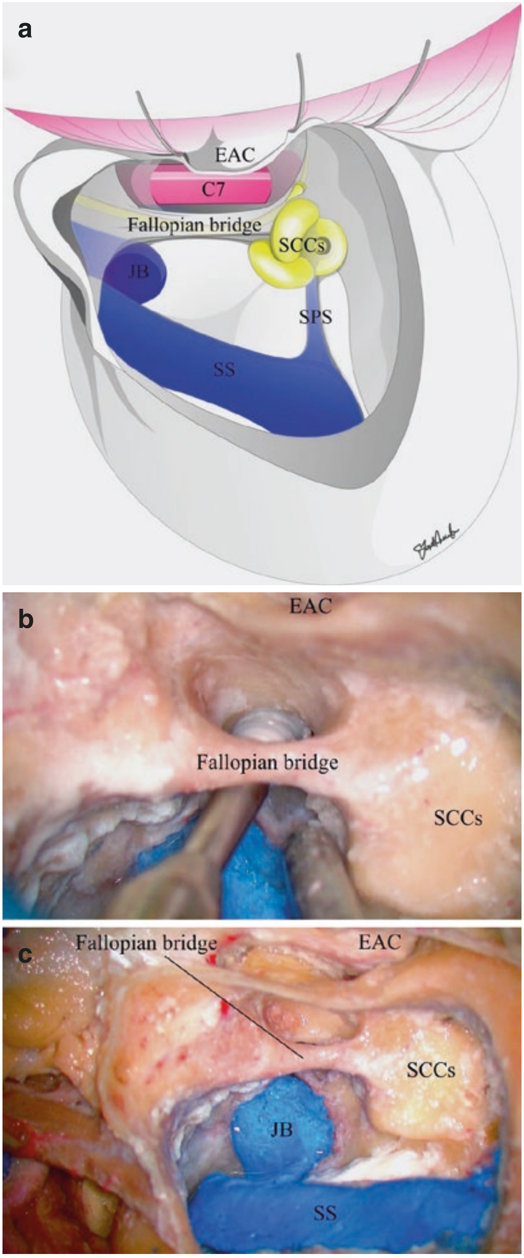

Fig. 52.2 Fallopian bridge technique. (**a**) Left side retrolabyrinthine approach with Fallopian bridge technique. The thin bone around the Fallopian segment of the facial nerve is left to create the Fallopian bridge. (**b**) A photo of cadaver dissection demonstrates bone removal under the Fallopian bridge using a high-speed drill. (**c**) A photo demonstrates the final view of the Fallopian bridge technique

We modified the segmental nomenclature of the ICA described by Fischer in 1938, which numbers the segments beginning from the carotid bifurcation [11]. The horizontal intrapetrous segment is designated as C6, and the infratemporal vertical segment is C7.

Clinical Manifestation and Classification

JTP pathology and variable clinical manifestations are detailed in Chap. 48. JTP frequently involve CNs IX to XII and cause paralysis in the affected nerve distribution. GT can be diagnosed early due to symptoms of pulsatile tinnitus with or without conductive hearing loss. GV causes hoarseness and mass effect due to vocal cord paralysis. It pushes the IJV posterolaterally and the ICA anteromedially.

Two grading systems have been developed to classify JTP based on the location, size, and extent of the disease. Fisch introduced a classification of GJ, which guides the surgical approach depends on tumor size and extension (Table 52.1)

Table 52.1 Fisch classification of glomus tumors

Class	Description
A	Tumors confined to the tympanum and arising from the promontory, without evidence of bone erosion
B	Tumors involving the tympanum, with or without mastoid involvement but always arising from the hypotympanic region; the cortical bone over the jugular bulb must be intact
C	Tumors eroding the bone over the jugular bulb; the tumor may extend into and destroy the bone of the infralabyrinthine and apical compartments of the temporal bone
C1	Tumors involving the foramen caroticum
C2	Tumors involving the vertical segment of the carotid canal
C3	Tumors involving the horizontal segment of the carotid canal
C4	Tumors extending to the ipsilateral foramen lacerum and cavernous sinus
D	Tumors with intracranial extension
De1	Tumors with intracranial extradural extension of up to 2 cm
De2	Tumors with intracranial extradural extension of more than 2 cm
Di1	Tumors with intracranial intradural extension of up to 2 cm
Di2	Tumors with intracranial intradural extension of more than 2 cm
D3	Tumors with inoperable intracranial extension

Fisch [14]

Table 52.2 Glasscock–Jackson classification of glomus jugulare tumors

Grade	Description
I	Small tumor involving jugular bulb, middle ear, and mastoid
II	Tumor extending under internal auditory canal; may have intracranial extension
III	Tumor extending into petrous apex; may have intracranial extension
IV	Tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension

Jackson [15]

[14]. Jackson, et al. also reported a new classification focused upon intracranial extension (Table 52.2) [15].

Roadmap to Reach a Decision

Surgical Indication, Strategy, and Complication Avoidance

JTP management strategy is detailed in Chap. 48. Patient's age should be considered the main factor in reducing postoperative morbidity and mortality rates related to pharyngolaryngeal paralysis. In our experience, young and middle-aged patients (<60 years old) are candidates for radical tumor resection for the aim of surgical cure. Patients who had postoperative LCNs deficits require a prolonged rehabilitation, often accompanied by surgical correction such as thyroplasty or Teflon paste injection of the paralyzed vocal cord. These additional procedures are usually well-tolerated. In elderly patients, we should be less aggressive in surgery because LCNs palsies are poorly tolerated and should be avoided, considering postoperative radiotherapy.

"Wait and see" policy is also recommended currently in selected cases, mainly those of elderly patients with intact LCN function or for patient with bilateral lesion. In elderly patients with advanced disease, radiotherapy must be preferred. Size of the tumor, age, clinical symptoms, the physical condition of the patient, social backgrounds, and cooperation should be considered.

Even patients who underwent gross total resection may have some residual infiltration around the JB, ICA, or temporal bone in a minority of cases. We have monitored these residual tumors in follow-up examinations, and in many of these cases, the tumor shows no growth over a follow-up period of 5–10 years. If we see significant growth, the patient is referred for stereotactic radiosurgery (SRS).

Complication Avoidance in GJ Surgery

GJ are locally destructive, highly vascular lesions that are located in one of the most critical skull base regions. Advanced C and D Fisch grade tumors may be both intra- and extradural with engulfment of critical neurovascular structures. The key maneuvers to avoid surgical complications in the management of GJ is how to treat the facial nerve at the fallopian segment, C6 petrous and C7 vertical segments of ICA, and LCNs [7, 8, 16–18].

Facial Nerve Mobilization

The Fallopian bridge technique has been recently advocated because it makes it possible to remove C1 and C2 GJ of Fisch's classification without any need for mobilization of the facial nerve and removal of the EAC and middle ear. In the case of small to large tumors, we specially choose not to reroute the facial nerve to avoid postoperative facial nerve palsy as much as possible. Even in the case with large, aggressive tumors, if necessary, we used to mobilize the descending fallopian and stylomastoid foramen segments of the facial nerve approximately 5 mm anteriorly to provide adequate exposure infratemporal ICA [12].

ICA Engulfment

One of the most critical issues in GJ surgery is the exposure and control of the ICA [7, 9, 16, 17]. GJ frequently adhere or engulf the ICA (C7 or C6) as well as LCNs (Fig. 52.3). For extensive tumors, full exposure of the C7 vertical portion of

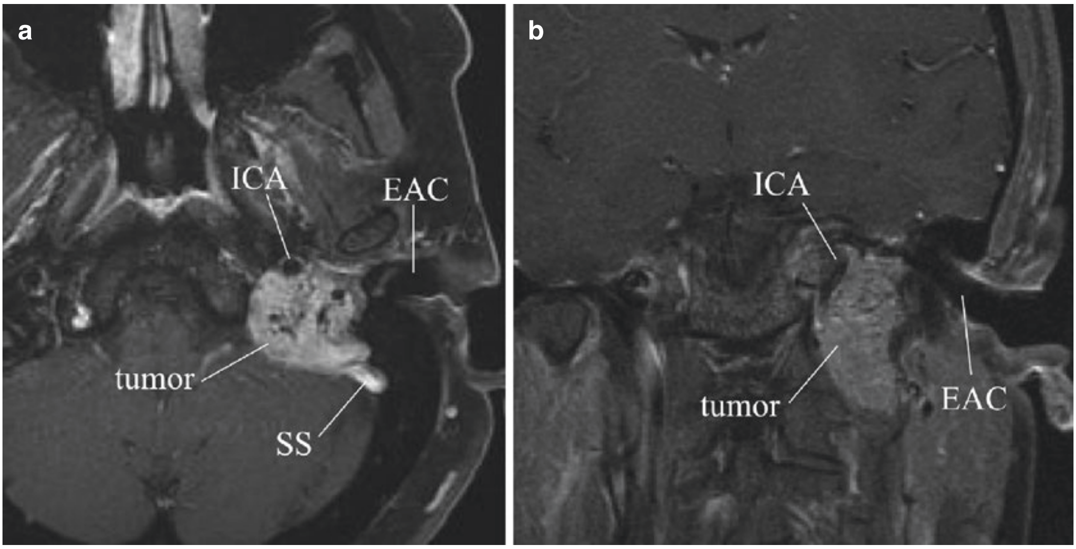

Fig. 52.3 MRI of glomus jugulare. A postcontrast axial (**a**) and coronal MRIs (**b**) demonstrate glomus jugulare (GJ) is occupying the left jugular foramen with engulfment of the internal carotid artery (ICA)

the ICA is mandatory. A complete dissection of the tumor from the C6 is probably the most limiting factor for achieving radical resection of GJ. Moreover, they often receive blood supply from the ICA. Therefore, controlled ICA dissection should be performed. A dissection plane can be developed between the adventitia and the tumor with the aid of the microscope. We do not recommend subadventitial dissection of the ICA because of the high risk of carotid rupture or occlusive complications. When it becomes unsafe to remove more tumors, the residual tumor remnants should be cauterized. Small remnants of GJ left on the ICA wall have a limited potential for generating a recurrence [9]. The risk of permanent ICA occlusion is high, even in patients who passed a balloon occlusion test. In case of extensive carotid involvement, the high-flow carotid bypass should be considered, followed by permanent balloon occlusion of the ICA (Chap. 12). However, it should be kept in mind that reconstruction of the ICA has also been associated with major risks and ICA sacrifice.

Preservation of LCNs

Cranial nerve preservation during the surgical resection of GJ is the main factor in reducing the postoperative morbidity rate. Especially, preservation of the LCN function is one of the major management issues. Worsening or a new deficit of LCNs substantially diminishes the patient's quality of life. There are four possible chances to damage the LCNs in the surgical procedure: (1) drilling of the jugular tubercle (JT), (2) tumor removal at the JB, (3) dissection of the high cervical region, and (4) intradural tumor removal.

Extradural bone removal of the JT is a key maneuver in transcondylar-transtubercular exposure because it reduces an obstructed view of the ventral side of the brainstem and clivus anterior to the LCNs during intradural procedure. The JT should be drilled away as much as possible with extreme care because the LCNs which cross over the posterior aspect of the JT into the JF are in very close proximity and may be at risk of damage by direct trauma, stretching of the dura, and heat generated by the drill.

GJ consistently arise in the lateral aspect of the JF, displacing the LCNs medially. This positioning accounts for the high rate of neural preservation in small and medium-size GJ that have not invaded the JF central partition [10]. After complete ligation of the SS, tumor within the JB can be removed with the lateral wall of the JB. The tumor should be elevated sharply to leaving the medial wall of the JB intact to preserve the LCNs at the pars nervosa. This procedure has already been introduced as "Intrabulbar dissection technique" by Al-Mefty and Teixeira [7]. If there is apparent tumor invasion into the pars nervosa through the medial wall of the JB, it is controversial to leave or remove the tumor. Sanna et al. advocated that it is not advisable to attempt to preserve the anatomic integrity of the LCNs when there is tumor involvement, which usually occurs when the lesion infiltrates the medial wall of the JB [16]. In their experience, dissection of the involved nerves has always resulted in a complete palsy, with the additional risk of leaving some tumor remnants medial to the JF. They concluded that possible dural infiltrations managed conservatively might result in intradural recurrences after some years.

Sen et al. studied the histopathological features of 11 glomus tumors involving the temporal bone, with respect to nerve invasion, associated fibrosis, and carotid artery adventitial invasion [19]. Within the JF, the cranial nerves lie anteromedial to the JB and maintain a multifascicular histoarchitecture. GJ of the temporal bone can invade the cranial nerve fascicles, and infiltration of these nerves can occur despite normal function. In these situations, total resection may not be possible without the sacrifice of these nerves.

Even the patients who underwent gross total resection may have microscopic residual infiltration around the JB. Therefore, a long-term follow-up examination is necessary for such a slow-growing neoplasm to monitor if the residual infiltration may grow.

Preservation of Key Neurovascular Structures

Basic Settings of Surgical Materials for Microsurgery

Simple, clear anatomical orientation leads to safe surgery. Additionally, it also helps understand the complex relationships between the lesion and the surrounding compressed neurovascular structures. The majority of neurosurgical procedures are performed under an operating microscope. Meticulous and delicate microsurgical technique requires clean and bloodless operative fields (Chap. 6). An unclear operative field with the spread of blood clot or uncontrollable bleeding makes surgical procedure problematic. Several types of absorbable hemostat (Surgicel®; sheet type, cotton ball type, fibrillary type) or gelatine sponge (Gelfoam®) are used for hemostasis at cavernous sinus, pterygoid plexus, and network between them with or without in combination with fibrin glue (Bolheal®). Micropatties (Delicot®) have been used for a variety of purposes. They can be used not only to protect critical structures but also as markers, dissectors, dividers, spacers, stabilizers, and holders in all type of microsurgery [20]. A noticeable micropatty, such as one colored by dye or cut into a unique shape, which is easy to recognize, can be used as a warning sign of a danger zone or region of interest. Micropatties should be removed when the high-speed drill is used to avoid critical injury to the surrounding structures. These surgical materials play an important role in all microsurgical procedures to achieve safe and successful neurovascular protective surgery.

Surgicel® (Ethicon US, LLC, Cincinnati, OH, USA)
Delicot® (American Surgical Company, Salem, MA, USA)
Gelfoam® (Pfizer, New York, NY, USA)
Bolheal® (Teijin, Tokyo, Japan)

Surgical Technique

Surgical Approach Variation

The surgical approach to GJ is crucial to obtain a radical removal. The surgical technique should be tailored to each case, mainly depending on the tumor size and its location (Fig. 52.4). The preoperative symptoms are also important factors in deciding the surgical approach. The key anatomical issues for surgical management of GJ are the Fallopian segment of the facial nerve, ICA and LCNs [7, 8, 16–18, 21]. The conventional surgical technique is based on the temporal bone-

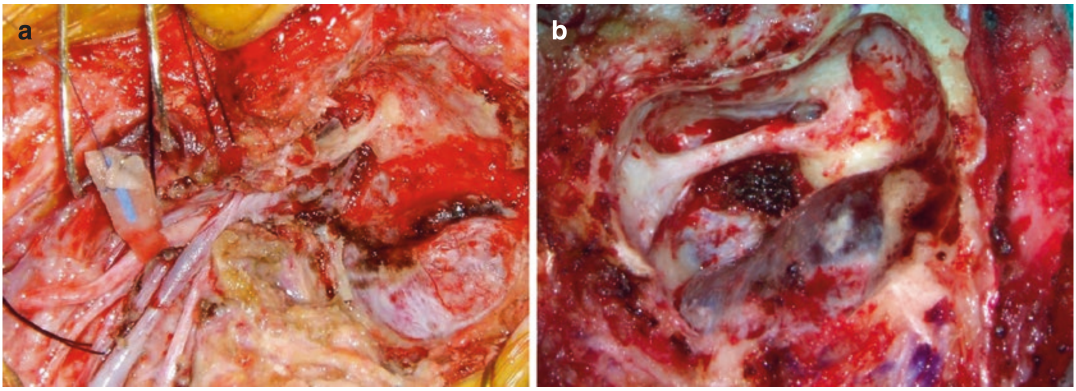

Fig. 52.4 Surgical variations for glomus jugulare. (**a**) Conventional transmastoid–transjugular approach with high cervical exposure. (**b**) Less invasive transmastoid approach with Fallopian bridge technique

transmastoid drilling, exposure of the JB and ICA with or without rerouting the facial nerve and high cervical exposure. Over the past three decades, these extensive postauricular approaches introduced by Fisch have been the basis for GJ surgery [15] (Chap. 44). This technique has been recommended by many authors with some modifications, and is advocated as a two-stage operation in some cases [12, 22, 23].

The transjugular approach is used for GJ, which entails the sacrifice of the SS. GJ are slow-growing lesions, so there is usually time for a rerouting of the venous drainage through collaterals and through intralesional shunting [24]. In most of these cases, the JB is already occupied by the tumor, and venous flow around the JB is decreased or blocked. Thus, the occlusion of the SS by the double-ligation technique will not further compromise the venous drainage.

For large or extensive GJ, we perform one-stage transjugular posterior infratemporal fossa approaches that allow radical resection of tumors located around the JF, lower clivus, and the high cervical region. This approach is a combination of the transmastoid, retro- and infralabyrinthine, transjugular, extreme lateral infrajugular transcondylar transtubercular, and high cervical approaches [11, 12]. The bony labyrinth should be kept intact in a case with a serviceable hearing. The total exposure of the JF can be achieved, and multidirectional approaches can be performed from an anterolateral direction, including suprajugular, transjugular, and infrajugular exposures. Both intracranial and extracranial tumors can be removed in a one-stage procedure. Transection of the EAC and permanent rerouting of the facial nerve are not necessary; instead, slight anterior transposition of the facial nerve, in select cases, can provide adequate exposure of the infratemporal ICA (C7 segment), without anterior dislocation of the mandible. This complex approach for total JF exposure can be simplified in a stepwise fashion; (1) retroauricular curvilinear skin incision; (2) high cervical exposure; (3) retrolabyrinthine mastoidectomy; (4) skeletonization and anterior mobilization of the facial nerve; (5) lateral suboccipital craniotomy and transcondylar-transtubercular exposure; (6) ligation and removal of the IJV, JB, and SS; and (7) intradural exposure if necessary.

Over the initial 10 years of our experience, we modified Fisch A technique to make it a smaller, less invasive exposure and minimize cranial nerve morbidity. First, we moved away from performing a two-stage operation. Second, in patients with functional hearing, we did not close the EAC. Then, we rarely perform extensive neck dissection in favor of limited high cervical exposure. In our recent cases of small to large tumors with minimal extension to the high cervical region, we performed a transmastoid–transsigmoid–transjugular approach with slight anterior translocation of the fallopian segment of the facial nerve. We recognized that even though the less invasive combined transmastoid and high cervical approach, we experienced significant morbidity with facial weakness, hearing loss, and LCN dysfunction [12].

Moreover, in recent years, we have developed a less invasive transjugular approach with the Fallopian bridge technique, allowing tumor resection while preserving hearing and protecting the facial nerve and the pars nervosa [13]. The fallopian bridge technique was first used by Pensak and Jackler for smaller tumors with no erosion of the EAC, and no extension anterosuperior to the carotid genu [25]. In the majority of their cases, satisfactory exposure of the region of the JF could be provided without facial nerve mobilization with a favorable result.

Transjugular Approach

The patient is placed in a supine position with the head turned laterally away from the lesion's side. A shoulder roll is used to elevate the ipsilateral side of the shoulder. For obese patients with short necks, a lateral position may be used. A retroauricular C-shape skin incision is made to fully expose the mastoid body. Transmastoid drilling is performed to expose the infralabyrinthine-suprajugular area bounded superiorly by the posterior semicircular canal (SCC), inferiorly by the JB, laterally by the Fallopian segment of the facial nerve, and medially by the presigmoid dura. Adequate drilling of this area allows a surgeon to expose an extradural component of GJ, which often extends to the anterior aspect of the JB. The SS is double-ligated at a point halfway between the sinodural angle and JB, then incised (Fig. 52.5). The dissection plane between the intraluminal tumor and the medial wall of the JB is established proximally in the SS to preserve the LCNs.

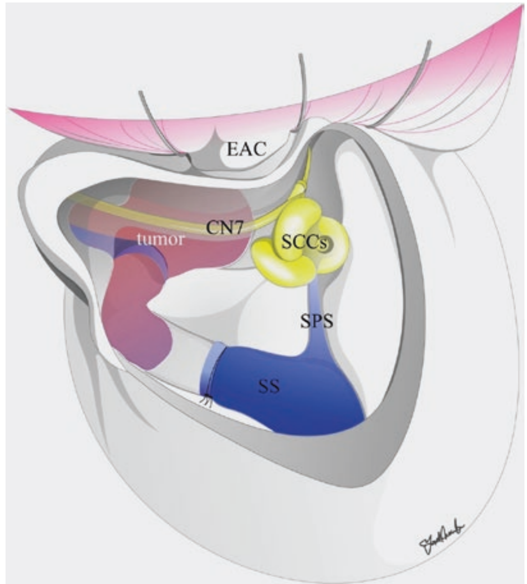

Fig. 52.5 Retrolabyrinthine – transjugular approach. The tumor occupies the infralabyrinthine suprajugular area (caudal to the semicircular canals (SCCs)) with extension into the SS. The sigmoid sinus (SS) is double-ligated to remove the intraluminal tumor

Transmastoid Approach with High Cervical Exposure (The Combined Transmastoid Retro- and Infralabyrinthine Transjugular Transcondylar Transtubercular High Cervical Approach)

The patient is placed in a supine position with the head rotated to the contralateral side (Fig. 52.6a). A retroauricular curvilinear sickle skin incision is started 2 cm posterior to the upper border of the ear. It continues posteroinferiorly into the neck over the anterior border of the sternocleidomastoid muscle and under the mandibular angle (Fig. 52.7a). The suboccipital muscles, including the sternocleidomastoid muscle, longissimus capitis, and splenius capitis muscles, are reflected posteroinferiorly to expose the mastoid body, inferolateral part of the occipital bone, suboccipital triangle, and the transverse process of C1. The superior oblique muscle, which is composed of the upper part of the suboccipital triangle, is also elevated from the inferior nuchal line of the occipital bone then reflected inferiorly. The posterior belly of the digastric muscle is also detached from the digastric groove behind the mastoid tip and reflected anteriorly to protect the extracranial portion of the facial nerve. The transverse process of C1 is an important landmark to identify IJV, ICA, and the extracranial portion of the LCNs.

Transmastoid drilling is performed by using a diamond burr. The SS down to the JB is fully skeletonized, and the mastoid air cells are removed to

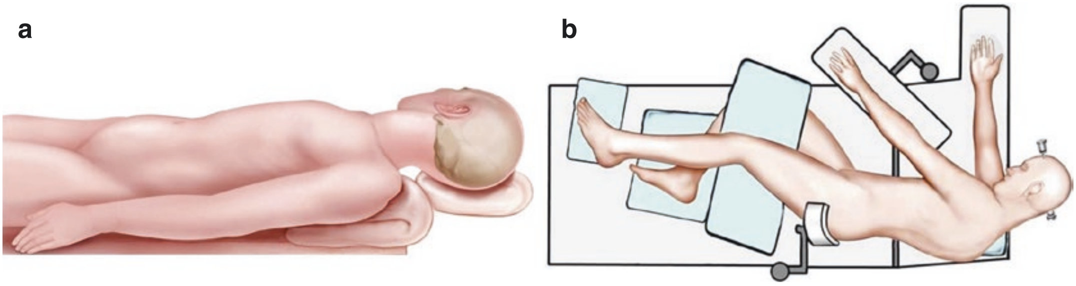

Fig. 52.6 Patient Positioning. (**a**) Supine head-lateral position. (**b**) Lateral position

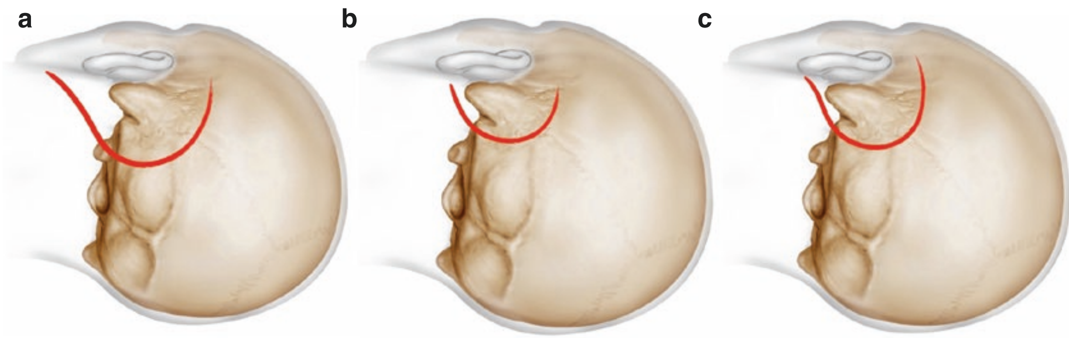

Fig. 52.7 Variation of retroauricular skin incision. (**a**) Retroauricular curvilinear sickle skin incision for combined transmastoid-high cervical approach. (**b**) Retroauricular C-shape incision for transmastoid approach (retrolabyrinthine, translabyrinthine approaches). (**c**) Retroauricular C-shape incision for transotic approach

expose the bony labyrinth, presigmoid dura, the superior petrosal sinus, sinodural angle, the temporal tegmen, and the retrosigmoid dura (Fig. 52.1a). The facial nerve at the fallopian canal segment, which is located 12–15 mm deep from the cortical surface of the mastoid body, can be identified with the aid of a facial nerve stimulator. The fallopian canal is carefully skeletonized from the genu to the stylomastoid foramen with a diamond burr under constant irrigation to avoid thermal injury of the facial nerve (Fig. 52.8a). In most cases, the tumor will be exposed during infralabyrinthine drilling around the JB. The mastoid tip is removed carefully to decompress the facial nerve at the level of the stylomastoid foramen. If necessary, the fallopian segment of the facial nerve can be slightly transpositioned anteriorly to provide added exposure to the C7. This maneuver should be used selectively and produces less risk of facial nerve palsy in contrast with permanent facial nerve rerouting.

After transmastoid retrolabyrinthine drilling with the mastoid tip removal and anterior reflection of the digastric muscle, the rectus capitis lateralis, which overlies the IJV, is exposed (Fig. 52.8b, c). Removal of the rectus capitis lateralis finally allows full exposure from the SS to the IJV (Fig. 52.8d). A lateral suboccipital craniotomy is then performed to expose retrosigmoid posterior fossa dura. After the craniotomy, additional bone removal toward the lateral inferior to the SS is mandatory. Further bone removal, which means an extradural reduction of the occipital condyle (OC) and jugular tubercle (JT), are the key maneuvers in this step. Removal of the posteromedial one-third of the OC is adequate to increase the surgical corridor to the brainstem's ventral side while preserving the atlanto-occipital stability. During this procedure, the posterior condyle emissary vein will be encountered as it travels from the JB and exits the condylar fossa via the condylar canal to join the extradural venous

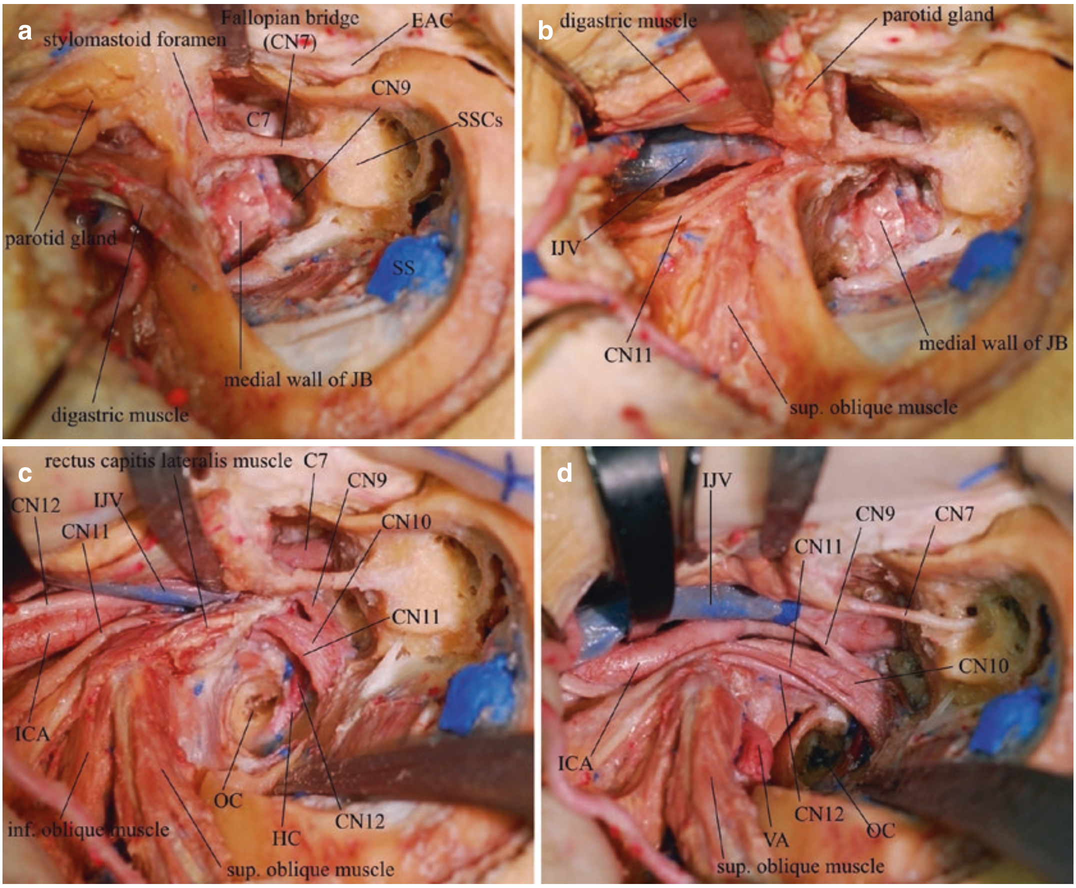

Fig. 52.8 Combined transmastoid transjugular approach with high cervical exposure in cadaver dissection. (**a**) Left side retrolabyrinthine approach with Fallopian bridge technique is presented. The C7 is exposed in-depth medial to the Fallopian bridge. The medial wall of the jugular bulb (JB) and the sigmoid sinus (SS) is exposed. (**b**) The digastric muscle is detached from the digastric groove and retracted anteriorly to expose the internal jugular vein (IJV). (**c**) Additional bone removal was performed. The medial wall of the JB is removed to expose pars vascularis and pars nervosa. The mastoid tip is removed, and the stylomastoid foramen is opened to expose IJV, internal carotid artery (ICA), CN 11, and CN 12 at the high cervical area. The occipital condyle (OC) is also drilled out to expose the hypoglossal nerve (CN12) in the hypoglossal canal (HC). (**d**) Removal of the rectus capitis lateralis muscle allows exposure of the lateral aspect of the craniovertebral junction

plexus. Bone removal of the posteromedial aspect of the OC by using a drill should be done until the cortical layer of bone covering the hypoglossal canal (HC) is exposed (Fig. 52.8c). The HC situated superior to the OC and inferior to the JT, is an important landmark for condyle bone removal. Bone removal is next directed superiorly toward the JT. It is situated slightly medial and inferior to the JB, superior to the HC, and medial to the JF. The bone removal of the JT is one of the difficult procedures in this surgical approach because mechanical damage of the posterior fossa dura just superior to the JT may directly damage LCNs.

After complete exposure of the SS, JB, and IJV, the tumor mass can be palpated within these venous structures through the venous wall. After all arterial feeders are coagulated, the IJV is then ligated just inferior to the tumor. The SS is occluded just above the tumor with a suture ligature. Then, the lateral wall of the SS is incised and removed with the tumor down to the JB (Figs. 52.8c, d and 52.9a). The inferior portion of the tumor also can be removed through an incision on the lateral wall of the IJV. As the tumor is removed from the intraluminal space, bleeding will be encountered from remaining feeding

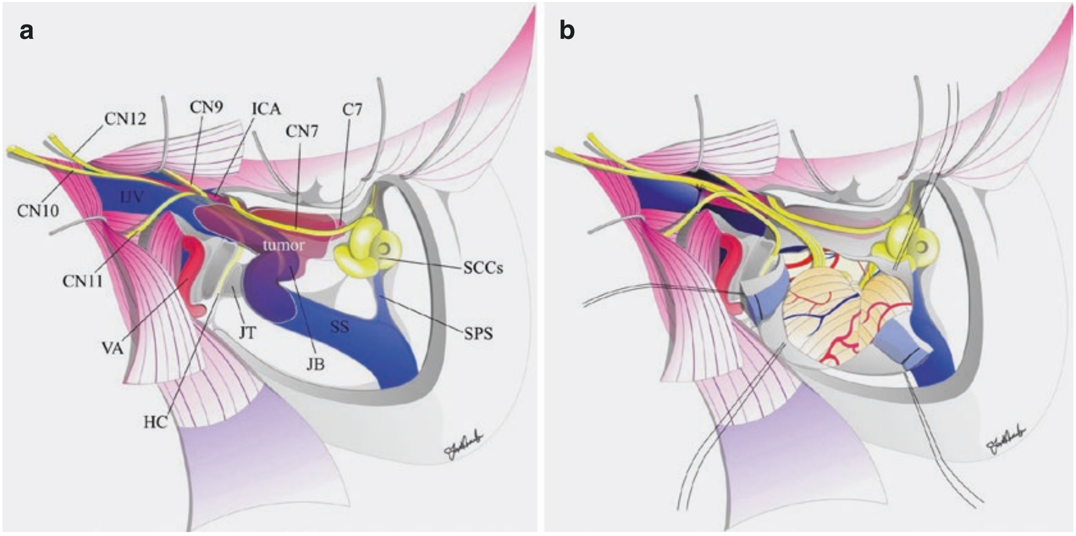

Fig. 52.9 Combined transmastoid transjugular approach with high cervical exposure. (**a**) Left side sigmoid sinus (SS) down to the internal jugular vein (IJV) is fully exposed. (**b**) The IJV is ligated to remove the jugular bulb component of the tumor. The SS is ligated and transected to expose the cerebellopontine angle to remove the intradural tumor

vessels, venous channels around the JB, like the IPS. It sometimes disturbs the surgeon to identify the correct dissection plane between the tumor and the medial wall of the JB overlying pars nervosa, which may lead to unexpected LCNs damage. This bleeding can be controlled with Surgicel packing and micropatties. The dissection plane between the tumor and the medial wall of the JB is established proximally in the SS to decrease the risk of LCNs damage.

If there is tumor invasion into the intradural space, the retrosigmoid dura needs to be incised. The dural incision is extended toward the presigmoid dura through the SS for wide exposure of the cerebellopontine angle (Fig. 52.9b). For extensive GJ, the high cervical ICA should be exposed to proximal control. Then C7, in front of the JB, is exposed by drilling the inferior bony tympanic ring to remove the tumor around the C7.

Less Invasive Transjugular Approach with Fallopian Bridge Technique

The patient is positioned in a supine position with the head supported on an ENT pillow (Fig. 52.6a). If there is a limitation in the head rotation due to a short neck or obesity, the lateral position should be chosen (Fig. 52.6b). A standard retroauricular C-shaped incision covering the whole mastoid body is made (Fig. 52.7b). An extended mastoidectomy is performed with total skeletonization of the SS, exposing both the retrosigmoid and presigmoid dura. Maximum shaving of the SCCs and the Fallopian canal and full exposure of the JB is done next. A thin layer of cortical bone, like an eggshell, is left to cover the entire Fallopian segment of the facial nerve from the genu to the stylomastoid foramen, creating the "Fallopian bridge" (Fig. 52.2). The mastoid tip is maximally skeletonized, the digastric groove is removed, and infrajugular drilling is carried down into the OC until the HC is exposed.

After creating the Fallopian bridge, further bone removal from the facial recess exposes the tumor in the inner ear and the hypotympanum area. This allows the identification and preservation of the annulus of the tympanic membrane [9, 21]. While protecting the EAC's thin wall, the inferior bony tympanic ring is maximally drilled away (Fig. 52.10). This removes the styloid process base and exposes the vertical C7 segment of the ICA, which is located anterior to the JB. At this point, the microscope's viewing angle can be manipulated allowing the surgeon to fully visualize the jugular structures, the C7, and the hearing

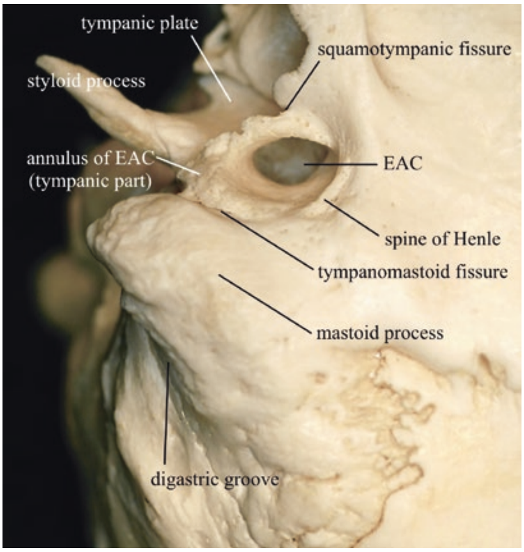

Fig. 52.10 Bony structure around the mastoid process and external auditory canal. A photo demonstrates the bony structure around the left side mastoid process

apparatus, looking from above or below the "Fallopian bridge." The SS should be ligated to remove the JB portion of the tumor.

In a majority of GJs, the retrofacial air cell space is filled with the tumor. Resection of tumor from this suprajugular area must be done with extreme caution to prevent damage to the posterior semicircular canal SCC and the basal turn of the cochlea. Additionally, care must be used while resecting tumor from around the ICA to prevent damage to the crossing CN IX or the arterial adventitia. In intradural extension cases, maximum effort is made to preserve anatomical continuity of the LCNs, unless the patient presents with LCN deficits. Tumor extension into the IJV through the JB can be removed with dissectors, ring curettes, and tumor forceps because this portion of the tumor usually does not adhere to the intimal surface of the IJV (Fig. 52.11) [13, 26].

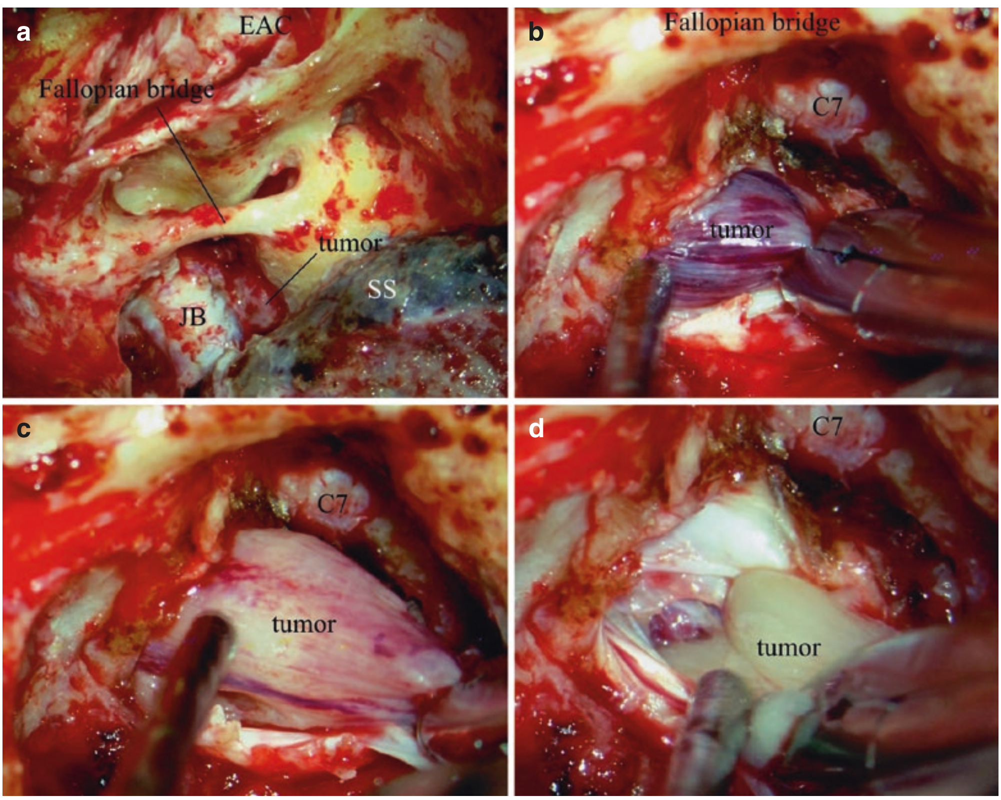

Fig. 52.11 Tumor removal via the transjugular approach with Fallopian bridge technique. (**a**) Left side retrolabyrinthine approach with Fallopian bridge technique is performed to resect the tumor occupies the jugular bulb (JB) and infralabyrinthine area. (**b**–**d**) After removal of the outer wall of the JB, the internal jugular vein (IJV) component of the tumor is pulled out with the tumor forceps. No adhesion between the tumor and the endothelial surface of the IJV exists

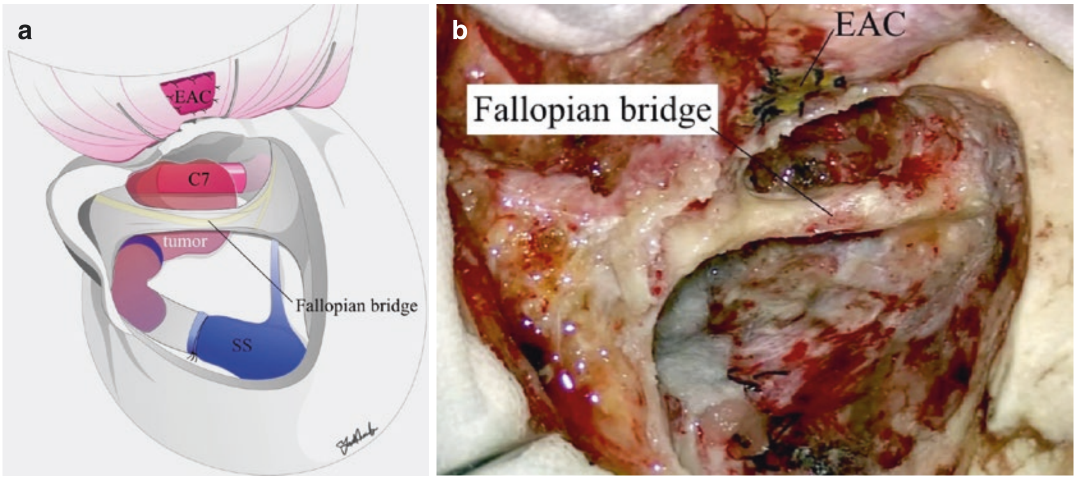

Fig. 52.12 Transotic approach with Fallopian bridge technique. (**a**) The tumor occupies the infralabyrinthine suprajugular area with engulfment of the C7 and extension into the SS. The sigmoid sinus (SS) is double-ligated to remove the intraluminal tumor. The external auditory canal (EAC) is closed by a piece of fascia. The bony EAC is drilled away to expose the tumor around the C7 by creating the Fallopian bridge. (**b**) An intraoperative photo demonstrates the left side transotic approach with the Fallopian bridge technique. The SS is ligated, and the tumor located at the jugular bulb (JB) is already removed

Transotic Approach

If the patient had already lost hearing function or the tumor has invaded the ventral side of the C7, the EAC can be closed, followed by bony EAC drilling (transotic drilling) to obtain wider operative field (Fig. 52.12). The transotic drilling provides a full anterior exposure. This advanced technique allows radical resection of extensive GJ located around the JF, with expansion toward the upper clivus and the intrapetrous region. One of this approach's critical maneuvers is the blind sac closure of the EAC to avoid postoperative cerebrospinal fluid leak and infection. The EAC's soft tissue should be closed tightly by multiple-layer technique using fascia to avoid postoperative infection [11, 26].

A postauricular C-shaped skin incision covering whole mastoid body is made (Fig. 52.7c). A standard translabyrinthine drilling is done with creation of the Fallopian bridge. The EAC's bone edge is drilled off medially to expose about 10 mm of the EAC periosteum overlying the skin of the cartilaginous canal. The EAC skin is carefully elevated off the bony canal and transected. This EAC fish mouth skin is everted into the EAC outer opening and sutured. A fascial patch covers the sutured EAC to reinforce it. Then, the ear canal, tympanic membrane, auditory ossicles, and SCCs are entirely drilled away, whereas the facial nerve's tympanic and Fallopian segment is left in situ.

Conclusion

Surgery of JTPs is still challenging. However, advances in microneurosurgery, and modern technique of skull base surgery allow safe resection of these tumors with lower morbidity and mortality rates. For the best outcome, the surgeon must take a very individualized approach to each patient.

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