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Management of Jugular Paragangliomas: The Gruppo Otologico Experience

Mario Sanna, Yogesh Jain, Giuseppe De Donato, Rohit, Lorenzo Lauda, and Abdelkader Taibah

Gruppo Otologico, Piacenza-Rome, Italy

Objective: The objective of this study was to review the outcome of surgical management in patients of jugular paragangliomas.
Study Design: We conducted a retrospective case review.
Setting: Tertiary care otology and skull base center.
Materials and Methods: Fifty-five patients with the diagnosis of a jugular paraganglioma (Fisch Class C and D Glomus Jugulare) were managed over a period of 15 years. All patients with adequate follow up and complete records (53 cases) were reviewed with emphasis on the results of surgical management and the factors influencing them.
Intervention: All 53 patients were managed with a view to surgically extirpate the tumor. The primary approach was the infratemporal fossa approach–Type A used in the majority of the patients. In eight cases, the procedure was staged owing to the presence of large intracranial extension. Three patients required additional procedures to ameliorate the after-effects of lower cranial nerve resection.
Results: Gross total tumor removal was achieved in 49 patients. There were five cases of recurrence. Coupled with the residual tumors in five patients, the surgical control achieved was 83%. There was no perioperative mortality. There were two cases of postoperative cerebrospinal fluid leak, both of which required surgical exploration and closure. The facial nerve was resected in seven patients. The overall preservation rate of clinically uninvolved lower cranial nerves was 75%.
Conclusions: The low level of complications along with a high surgical control achieved makes surgery the primary mode of treatment in the vast majority of these tumors, regardless of the size and location. **Key Words:** Jugular—Paraganglioma—Surgical management.
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Jugular paragangliomas (JP) are slow-growing benign tumors of the temporal bone with the potential to cause extensive morbidity by aggressive involvement of the entire skull base. In the process of this slow and insidious growth, they often invade and erode the bony skull base, infiltrate the regional cranial nerves (CNs), constrict the major blood vessels to the brain either by encasement or direct infiltration, and transgress the dural barrier (1). The ideal primary treatment of these tumors is total surgical extirpation (2,3). The evolution of the present surgical approach to this region was initiated in 1949 with the proposal of the resection of the jugular bulb to treat these tumors (4). However, it took nearly three decades of innovation by a multitude of workers for the surgery of these tumors to evolve to the approach that it is practiced today (5–12). Fisch, in 1977 (13), standardized surgery of this region by presenting the infratemporal fossa approaches. The contribution made by concurrent advances in techniques of imaging, anesthesia, and intensive care in enabling safe and effective surgery of these tumors cannot be overemphasized (14,15). However, surgery in this delicate region is still fraught with danger, requiring the ability to accurately assess varied interrelated factors, their impact on the ultimate result, and their management. The main impediments to achieving a standardized system of surgical cure have been the comparative inaccessibility of these extremely vascular tumors nestling among a cluster of vital structures, their relative rarity, the frequency and severity of surgical complications along with the extremely long period it can sometimes take for a recurrence to occur or manifest. As a result, the data required to validate the surgical therapy of these tumors is still being accumulated (16). In the current study, we report a retrospective analysis of the management of 53 patients with a diagnosis of a JP, the majority of which underwent surgery as the primary treatment, in an attempt to examine the credentials of surgery as the ideal treatment of these tumors.

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Address Correspondence and reprint requests to Dr. Giuseppe De Donato, Gruppo Otologico, Via Emmanueli 42, 29100 Piacenza, Italy; Email: Giuseppe.Dedonato@Gruppootologico.It

MATERIALS AND METHODS

All the charts of patients with a diagnosis of paraganglioma of the temporal bone and attendance at a European tertiary care

referral skull base center between April 1988 and June 2002 were extracted and those with a Class C and/or D JPs (Fisch Classification) (17) as the primary diagnosis were chosen for this study, which yielded a total of 55 cases. All surgical procedures were performed by the senior author (M.S.). Two cases had incomplete records and were excluded. All perioperative data with follow-up records were extracted and analyzed. Each patient had undergone complete radiologic examination, which included high-resolution computed tomography (HRCT) with bone windows, magnetic resonance imaging with gadolinium enhancement (MRI Gd), and four-vessel angiography with manual cross-compression testing. All patients had at least one hearing assessment in the form of pure-tone audiometry and most had a clinical photograph of the otoscopy. The facial nerve (FN) function was recorded using the House-Brackmann (HB) grading system (18) at each visit by the patient, and this assessment was always done by a member of the operating team. We have been maintaining a photographic record of the FN function of our patients since 1996 in the form of four representative postures in each visit of the patient, including the preoperative consultation, and have found this to be a very useful and cost-effective method of increasing the reliability and reproducibility of our data.

The intraoperative records included detailed notes of the procedure, exact location, and extent of the tumor, along with a record of the management of the FN and the internal carotid artery (ICA). The FN monitor has been in routine use in all skull-base cases in our institution since 1994. We do not routinely use lower CN monitoring because we feel its use is yet to be established.

All patients were treated with surgery as the primary treatment. The primary approach used was the infratemporal fossa approach–Type A (IFTA–A). The detailed surgical steps have been presented elsewhere (17,19). In some cases, there were supplementary techniques used, especially when the procedure was staged in view of the presence of large intracranial extensions (ICEs). These are mentioned in the appropriate sections.

Much of the dissection of the tumor was done using the operating microscope. The tumor was frequently found attached to the carotid canal with varying degrees of involvement of the ICA, where meticulous and orderly dissection nearly always enabled complete removal of the tumor.

In cases of ICE when this component was not excessively large (Class Di1) or extradural (Classes De1, De2), approximately 1 cm of the posterior fossa dura was opened and the intracranial portion was removed concurrently. The dural opening was closed primarily with sutures intersewn with a plug of free muscle graft. When the size of the intradural intracranial component was greater than 2 cm (Fisch Class Di2), we preferred to stage the resection to a first extradural and a second intradural stage. The second-stage intradural removal of the tumor was generally performed through the same retroauricular approach using the already performed bone removal and sigmoid sinus closure during the first stage and an appropriately sized dural opening to gain access to the posterior fossa.

RESULTS

Epidemiology

There were 53 patients with the youngest 17 years old and the oldest 73 (mean, 47.4 yr). Female sex predominated in a ratio of 1.9 to 1 (35 female). Thirty-two (60.3%) of these tumors were on the left side and 21 (39.6%) on the right side. There were two patients with multicentric tumors. One, aged 17 years, developed a carotid body tumor on the contralateral side 2 years after he was operated for the JP and subsequently underwent successful resection for this second lesion also. This patient has developed a contralateral carotid body tumor (5 yr after the JP was excised) and is scheduled for surgery for this lesion. The other patient had already been operated for a tympanic paraganglioma and a carotid body tumor of the same side and was aged 27 years when he presented to us. There was not a single patient with a catecholamine-secreting or a malignant JP in this series. The radiologic follow-up period (consisting of serial CT and/or MRI scans) of the series ranged from 3 to 144 months (mean, 31.3 mo). Four cases were lost to long-term follow up.

Symptomatology/clinical signs and symptoms

The most common presenting complaint was pulsatile tinnitus in 24 (45.2%) patients, whereas the most common symptom elicited was hearing loss in 41 (77.3%) and tinnitus in 38 (71.7%) cases. The signs and symptoms have been tabulated in Table 1. The most common nonotologic symptom was hoarseness, which was present in 14 (26.4%) of the cases. Three cases were recurrences from previous procedures at other centers. There was one patient who had been treated with radiotherapy in another center.

Class/location of the tumor

The location and the extent of the lesion were determined preoperatively using the radiology reports (HRCT with bone windows and MRI Gd-DTPA), which was subsequently confirmed intraoperatively. The frequency of involvement of the various structures is depicted in Table 2 and according to the Fisch Classification in Table 3. As is evident, the incidence of ICE was directly related to the class of the tumor, with Class C1 having the lowest involvement of the intracranial involvement (0%) and Class C4 the highest (100%). Overall, 33 (62.2%) of the cases had an intracranial component. Twenty-four (72.7%) of these were intradural and the remaining nine (27.2%) extradural.

TABLE 1. *Presenting symptoms/signs*

	No. of patients (%)
Hearing loss	41 (77.3%)
Tinnitus	38 (71.7%)
Pulsatile tinnitus	24 (45.2%)
Hoarseness	14 (26.4%)
Vertigo	11 (20.75%)
Instability	6 (11.3%)
Otorrhagia	3 (5.6%)
Diplopia	3 (5.6%)
Facial weakness	2 (3.7%)
Aural fullness	2 (3.7%)
Facial hypoaesthesia	2 (3.7%)
Dysphagia	1 (1.8%)
Autophony	1 (1.8%)
Anacusia	1 (1.8%)
Otorrhoea	1 (1.8%)

TABLE 2. *Localization of the tumor/sites of involvement*

	No. of patients (%)
Jugular foramen	53 (100%)
Posterior cranial fossa	33 (62.2%)
Petrous apex	13 (24.5%)
Cerebellopontine angle	7 (13.2%)
Clivus	4 (7.5%)
Prepontine cistern	4 (7.5%)
Foramen magnum	3 (5.6%)
Occipital condyle	3 (5.6%)
External auditory canal	2 (3.7%)
Internal auditory canal	2 (3.7%)
Cavernous sinus	2 (3.7%)
Labyrinth	1 (1.8%)
Cochlea	1 (1.8%)
Mandibular condyle	1 (1.8%)
III portion fallopian canal (radiology)	1 (1.8%)
Hypoglossal canal	1 (1.8%)

All tumors underwent embolization of the blood supply using poly vinyl alcohol 1 to 2 days before the procedure.

Cranial nerve status

Twenty-three (43.3%) of the patients were found to be suffering from at least one cranial nerve deficit (CND) in the preoperative evaluation. This number was much higher in the group of patients with ICE (51.5%) than in those without ICE (20%). The most common CN involved was the IX and X CN each in 20 (37.7%) patients (Table 4). Seven (13.2%) patients had paralysis of all four lower CNs on initial diagnosis. We found a positive correlation of the presence of ICE with incidence of preoperative CNDs (Table 5).

The CN status at 1 year after the surgery has also been shown in Table 4. The most common CND was the IX, which was present in 31 (58.4%) patients. New CNDs appeared in 14 (26.4%) patients. As compared with 31 (58.4%) patients who did not have a preoperative CND, there were only 17 (32%) who were free from any evidence of CN damage after surgery at the end of 1 year. Of the 14 patients who developed new CNDs, nine (64.2%) had ICE, six of which were intradural. In four patients with no preoperative CND, all four lower CNs needed to be killed because of intraoperative evidence of their involvement. Three (75%) of these cases were tumors with intradural extension. Three of the four lower CNs had to be resected in another six patients. The preservation rates of the lower CNs has been depicted in Table 6 and as a function of the presence of ICE in Table 7.

TABLE 3. *Classification (Fisch)*

Class (%) No. in C class	No. in D class	
C1 (4) (7.5%)	0	(ICE 0/4)
C2 (35) (66%)	Del—7 Dil—9 Di2—4	(ICE—20/35, 57.1%)
C3 (12) (22.6%)	Del—2 Dil—4 Di2—5	(ICE—11/12, 91.6%)
C4- (2) (3.7%)	Dil—1 Di2—1	(ICE—2/2, 100%)

ICE, intracranial extension.

TABLE 4. *Cranial nerve results pre- and postoperative*

	No. of cases with deficits (%)	
Cranial nerve	Preoperative	Postoperative
IV	2 (3.7%)	1 (1.8%)
V	1 (1.8%)	0
VI	3 (5.6%)	2 (3.7%)
IX	20 (37.7%)	31 (58.4%)
X	20 (37.7%)	30 (56.6%)
XI	14 (26.4%)	24 (45.2%)
XII	13 (24.5%)	18 (33.9%)

*At least 1 year after the initial procedure.

Facial nerve status

The preoperative FN status is shown in Table 8. Forty-five patients underwent permanent anterior transposition of the FN. The long-term results (1 yr or more) were available for 44 of these patients (Table 9). One member of this group had a preoperative HB Grade IV paresis, which was maintained at the end of 1 year. This patient had developed this paresis as a complication of the preoperative embolization and the functional status had not improved until the last follow-up visit. In this group, there were no patients who had HB Grade V or VI in the long term. Another seven patients in this series required a resection of the involved segment of the nerve leaving a defect that was repaired using a sural nerve graft in the same sitting in every case. Of these seven cases, three had a preoperative HB Grade VI, two HB Grade II, and the remaining two had a HB Grade I status. We did not encounter a single case with involvement of the epineurium alone. Four of these patients had reached a HB Grade III, one HB Grade IV, and one remained in HB Grade VI at the end of 1 year. One patient was lost to follow up.

Surgical procedures

The IFTA–A was the primary approach used in 52 of the cases, the details of which have been described in

TABLE 5. *Cranial nerve deficits according to ICE*

Cranial nerve	No ICE (n = 20)		ICE (n = 33)	
	Preoperative	Postoperative	Preoperative	Postoperative
IV	0	0	2 (6%)	1 (3%)
V	0	0	1 (3%)	0
VI	0	0	3 (9%)	2 (6%)
IX	3 (15%)	8 (40%)	17 (51.5%)	23 (69.6%)
X	3 (15%)	8 (40%)	17 (51.5%)	22 (66.6%)
XI	3 (15%)	6 (30%)	11 (33.3%)	18 (54.5%)
XII	3 (15%)	3 (15%)	10 (30%)	15 (45.4%)

ICE, intracranial extension; Postoperative, at least 1 year after the initial procedure.

TABLE 6. *Lower cranial nerve preservation*

Cranial nerve	Nerves preserved*/clinically uninvolved nerves†	Percent
IX	22/33	66.7%
X	23/33	69.7%
XI	29/39	74.3%
XII	35/40	87.5%
Overall	109/145	75.1%

*At least 1 year after surgery.
†Preoperative status.

previous publications (17,19). This approach proved sufficient alone in 47 cases and had to be combined with a transotic approach in four tumors and a translabyrinthine exposure in one case. In a single patient, subtotal tumor removal was performed using a subtotal petrosectomy (20) without the need arising for a FN transposition in deference to the patient's wishes and age. The tumor needed to be dissected from the ICA in 43 cases. In one case, the entire circumference of the vertical petrous ICA had to be exposed and the vessel mobilized to remove the tumor, which was completely surrounding it and which could be done without its sacrifice. In an additional five cases, the vessel needed to be killed. The necessity for permanent balloon occlusion (performed at least 3 wk before the planned resection) had been identified preoperatively in each of these patients along with the demonstration of an adequate collateral supply.

In the present series, a total of eight cases required staging to be performed. Six of these were C3 and two were C2 tumors. The average time interval between the initial and the second procedure was 11 months with a range of 3 to 21 months. The modified transcochlear type D approach (21,22) was used in one case, the extreme lateral (23) in another, and a transotic approach (24) was required in a single case. The rest of the second-stage procedures was performed using the same approach as the first stage.

Complications

A single patient developed a HB Grade IV FN paresis after embolization. There were no other complications related to embolization. There was no perioperative mortality in this series. One patient died 4 years after the surgery of unrelated causes. There were two cases of cerebrospinal fluid leakage in the whole series, both of which required an additional procedure for resolution. There were three patients who had varying degrees of aspiration. A feeding jejunostomy had to be performed in one of these patients after she developed pneumonitis in the postoperative period. The same patient developed peritonitis as a consequence of the jejunostomy, from which she recovered with conservative treatment. The jejunostomy was successfully closed after she developed good compensation of the contralateral vocal cord. A medialization thyroplasty was performed in the other two patients with dysphonia and complaints of a disturbance in deglutition. There is one patient with persistent dysphonia with no complaints in swallowing. She has refused any intervention for the rehabilitation of the voice, being satisfied with the present quality. A mean of 1.1 IUs (range, 0–4 IUs) of blood was transfused in this series.

TABLE 7. *Lower cranial nerve preservation as a function of presence of ICE*

	No ICE	ICE
Cranial nerve	Preserved*/clinically uninvolved† (%)	Preserved*/clinically uninvolved† (%)
IX	12/17 (70.6%)	10/16 (62.5%)
X	12/17 (70.6%)	11/16 (68.8%)
XI	14/17 (82.4%)	15/22 (68.2%)
XII	17/17 (100%)	18/23 (78.3%)
Overall	55/68 (80.8%)	54/77 (70.1%)

*Preserved-function of the cranial nerve at least 1 year after surgery.
†No clinical evidence of involvement in the preoperative period.
ICE, intracranial extension.

TABLE 8. *Preoperative facial nerve status*

HB Gr	No. of patients (%)	Distribution of class
I	44 (83%)	
II	2 (3.7%)	C2Di2, C3Di2
III	2 (3.7%)	C1, C2Di2
IV	2 (3.7%)	C2, C3Di2
VI	3 (5.6%)	C2, C2, C2De1

HB Gr, House Brackmann Grade.

Tumor removal/recurrence

Gross total removal was achieved in 49 (90.7%) of the tumors of which we have been able to document a recurrence in five (10.2%) cases. Staged tumor removal had been performed in two of the cases with a recurrence. The time interval between the surgery and the recurrence to occur, in years, was 3, 3, 6, 6, and 10. One of these occurred in a Class C4 tumor, another in a C3, and the rest were C2 tumors. There were two cases with a large intradural component (C4Di2, C3Di2) and three (C2, C2, C2De2) without. One patient had an absence of

TABLE 9. *Anterior rerouting was done in 45 cases of which 44 have follow up of 1 year or more: long-term facial nerve (FN) results*

Pr HB Gr	Preoperative	Postoperative	
I	41	10 (22.7%)	Long Term Postoperative HB Gr I/II—65.8%
II	1	19 (43.1%)	
III	1	14 (31.8%)	
IV	1	1 (2.2%)	

Anterior rerouting of the FN was performed on cases with no evidence of infiltration of the nerve. Patients with an infiltrated FN are not included. FN resection and grafting was performed in all 7 such patients.

HB Gr, House-Brackmann Grade; Postoperative, at least 1 year or more after the initial procedure.

collateral circulation on preoperative balloon occlusion of the ICA, and the recurrence was detected around the horizontal portion of the petrous ICA after 10 years. Owing to the noncompensatory ICA status, he has had two therapeutic embolizations with plans afoot for another for continued growth of the recurrence. No growth has been detected on annual MRI scanning in the rest of the patients. In five (9.2%) cases, subtotal tumor removal was done with minute remnants being left behind in strategically vulnerable locations. These belonged to classes C4Di1, C3Di2 (2), C2Di2, and C1. The C1 tumor occurred in a patient with a dominant sigmoid sinus, which precluded its closure, thereby necessitating a partial removal. One patient with a C2Di2 tumor and a HB Grade III FN function was aged 66 years when she came to our center. She had a history of an operation 7 years ago at another center. In view of her age and unwillingness to risk her lower CN status, we performed a conservative excision using a subtotal petrosectomy approach. The third case was an extremely aggressive C3Di2 tumor, which involved the jugular foramen, posterior cranial fossa dura, and the occipital condyle. He underwent the first stage in the form of an IFTA–A, a second-stage transcochlear D approach after 4 months, and still had evidence of tumor in the region of the foramen magnum and inferior clivus at the last follow up. This patient has opted in favor of intratumoral therapeutic embolization at another center, of which he has had several so far with evidence of continued, albeit slow, growth. There was another C3Di2 tumor, which beginning in the jugular foramen, extended to the prepontine cistern, clivus, occipital condyle, and the cavernous sinus. A small piece was left behind in the region of the cavernous sinus, which was managed by stereotactic radiosurgery. However, a recurrence was detected recently at the region of the jugular foramen, which is being managed with regular radiologic follow up. The patient has been informed of the possibility of the need arising for another surgical procedure if there is demonstrable growth of the recurrence. The fifth tumor undergoing partial removal belonged to the Class C4Di1 and extended up to the foramen magnum and the cavernous sinus. Tumor was left behind in the region of the cavernous sinus, which was effectively managed with stereotactic radiosurgery.

DISCUSSION

The present series is composed of 53 patients of JPs. The mode of presentation of these tumors has been extensively described in the literature, and our series did not present any significant variation (Table 1) (1,25). The protocol of radiologic investigation in this series consisted of a HRCT with bone windows, MRI Gd-DTPA, and four-vessel angiography in every case. The various advantages of these investigations have been detailed elsewhere (25). We would like to emphasize, however, that all of these three investigations are essential to accurately delineate the extent of the tumor preoperatively and optimize its management. This is typified by the case with an ipsilateral dominant sigmoid sinus (and an absence of contralateral sinus). As a result of the tumor slowly obstructing the jugular bulb over a long period, much of the venous drainage was being handled by the emissary vein. Thus, we could carry out the IFTA–A just as long as we made sure to perform the closure of the sinus proximal to the junction of the emissary vein with the sigmoid sinus. Although the ICA is no longer the limiting factor in JP surgery, an accurate preoperative identification of the extent of its involvement is imperative for a safe resection of the tumor. This successful detection of the advanced level of involvement of the ICA in five patients along with its subsequent permanent balloon occlusion leading to its resection has been well demonstrated in this series. It is our opinion that achieving surgical control in these five patients would have been an unattainable goal without this maneuver. Some authors have advocated preoperative or concurrent reconstruction/replacement of the vessel in every case requiring its sacrifice (16). However, none of the five patients in this series have experienced any untoward effects from resection of the ICA in a mean follow-up period of 96 months (range, 60–156 mo) substantiating the contention that it can be safely resected in carefully selected cases. Preoperative embolization of the vascular supply to the tumor with PVA was used in every case. The low rate of complications of embolization in this series, in our opinion, underlines the importance of a sufficiently experienced and competent interventional neuroradiologist in the management team. The role played by this procedure in management of these tumors is ably demonstrated by the low level of blood replacement required (Table 11).

The presence of ICE in JPs has been reported as 14% to 72% (1,25). The present series had an overall intracranial involvement of 62%. This figure lends weight to the observation that a majority of these tumors are advanced at the time of initial diagnosis (1). The insidious and subtle nature of growth of these tumors is also ably demonstrated by this figure. The presence of ICE correlated well with the increasing class of tumor, i.e., higher the C class of tumor, more the probability of ICE. We believe the advanced level of extension in this series warranted the use of the IFTA–A in the majority of the cases. There are some points in the surgical technique that we would like to highlight. First, all cancellous bone adjacent to the tumor should be drilled until "healthy" cortical bone is reached to lessen the possibility of residual tumor. Second, the technique of closure of the sigmoid sinus by extra- and intraluminal compression instead of the traditional ligature method is an easier alternative of dealing with the sigmoid sinus, which does not violate the subarachnoid space without in any way compromising the operative time, eventual resection, and safety of the surgical procedure.

In the presence of large ICE, there are conflicting opinions in the literature regarding the practice of staging, which we have been after for the past 25 years (16,26–28). We believe this is the principal reason for

the extremely low level of postoperative complications, especially cerebrospinal fluid leak in this series. The unreliability and technical difficulty of achieving a watertight closure of a large dural opening along with extensive removal of the adjacent bone and soft tissues and the possibility of a potentially catastrophic communication between the subarachnoid space and the spaces of the neck in a single-stage procedure has prevented us from using it. A distinct advantage we have observed over the years has been the relative ease and technical simplicity of the intradural tumor removal in the second stage. Because the remaining tumor has frequently been devascularized during the initial stage, dissection and preservation of the lower CNs in the second intradural stage have been simplified to a great degree, which is further aided by the embolization of the blood supply before the second stage to further reduce intraoperative bleeding.

The involvement and resection of the lower CNs has been the principal source of postoperative morbidity and was positively correlated to the presence of ICE of the tumor, an observation supported by other series (16). We are in agreement with the observation that the lack of preoperative CNDs does not correlate well with the intraoperative neural involvement (29). The overwhelming majority of CN resections were performed because of intraoperative evidence of involvement. The presence of ICE had a positive correlation with the appearance of new CNDs underlining the importance of appropriate counseling of the patients with ICE and clinically uninvolved lower CNs. At this juncture, we would like to distinguish between the types of ICEs. In our experience, the presence of intradural extension usually indicates infiltration of the lower CNs leading to their resection. This can be explained by the observation that the most common route of the intradural spread is through the medial wall of the jugular bulb. The proximity of this structure to the lower CNs makes their preservation in the setting of an infiltrated medial wall a disappointing endeavor. We have not found extradural extension to be as indicative of lower CN infiltration. The preservation of the CNs in the extradural ICE group of patients was similar to the group without ICE, whereas it was significantly worse in the group with intradural ICE. This is in contrast to the results reported by other workers (30). The overall results demonstrate that 75% of the clinically uninvolved nerves can be preserved during surgery. These results compare favorably with the other major series reported in the literature (Table 10) (1,16,30,31). Only three patients in the whole series required further surgery to ameliorate the effects of the lower CN resection. It has been noted earlier that patients with preexisting CNDs have a better prognosis with regard to postoperative compensation (32). Interestingly, all three patients (aged 40, 55, and 61 yr) in this series requiring further intervention had a preexisting paralysis of the IX and X CNs. None of our patients developing a new CN paralysis required surgery for the relief of symptoms as a result of these deficits. These results compel us to disagree with the authors recommending early rehabilitative surgery after major skull base surgery (33). Most of these patients compensated very well with prolonged and intensive physiotherapy, which in our experience, obviates the need for any additional surgery for their CNDs.

Facial nerve management is an intrinsic part of JP surgery. Long and permanent anterior FN rerouting is the generally accepted method of its management during surgery of these tumors. The long-term outcome of such a manipulation in experienced hands has been excellent with an overall incidence of 72% achieving HB Grades I and II in the long term (34). Our results have been comparable (Table 9). In recent years, there have been doubts expressed concerning the need for long rerouting of the nerve in the majority of JPs (35–37). The pivotal role played by this manipulation in the management of these tumors can be put in stark perspective by the frequency of ICA manipulation in our series. Forty-eight patients (90.5%) in the present series required active management of the ICA (ranging from dissection of tumor from the vessel wall, partial and total exposure, mobilization to occlusion and resection of the ICA) with no adverse cerebrovascular sequelae in any of the patients. In our opinion, FN rerouting is essential to obtain adequate control of the distal petrous ICA and the entire extent of the

TABLE 10. *Comparative review of the major jugular paraganglioma series in the last 15 years*

Report	ICE incidence	CN preservation				FN status (House-Brackmann Grade I/II)	TTR	Recurrence	Surgical control
		IX	X	XI	XII				
Fisch[1] (n = 119)	72	22.3	54	61	73	87	83	1.6	74.7
Green[31] (n = 28)*	54	66.6	81.8	68.2	93	95	85	—	—
Jackson[16] (n = 152)†	36	61	75	74	74	—	88.8	5.5	85
Manolidis[30] (n = 139)†	33	—	—	—	—	66	—	—	81
Present series (n = 54)	62	66.7	69.7	74.3	87.5	65.8	90.7	10	83

All figures are percentages.

*24 tumors in series of 52 were Class B paragangliomas. Thus, the results might not be fully representative of the C/D tumors.

†These two series represent the same institution and we assume considerable overlap of case material. The earlier series was included as the later report does not have the facial nerve results.

ICE, intracranial extention; FN status, facial nerve functional status at 1 year or more in cases with anterior rerouting performed, fraction of patients in House-Brackmann Grade I or II; TTR, total tumor removal percentage; surgical control, percent of patients with no disease at the time of the study; CN, cranial nerve.

TABLE 11. *Complications*

	Fisch[1]	Green[31]	Jackson[16]	Present series
Cerebrospinal fluid leak	11	4	4.5	3.7
Aspiration	10	4	12.5	6
Pneumonitis	0	6	3.2	1.8
Pulmonary embolism	2	2	2.6	0
Meningitis	1	4	2.6	0
Wound infection	6	6	4	0
Seroma/hematoma	0	4	1.9	0
Stroke/cerebrovascular accident	1	0	2.6	0
Ileus	0	0	3.9	0
Pneumothorax	0	0	1.3	0
Mortality	1	0	2.7	0
Intraoperative blood loss (average)	2 L	2.9 IU used		1.1 IU used

IU, international units.

tumor. This extremely delicate maneuver (dissection of the tumor from the ICA wall) requires maximal surgical room, which in our experience, is best provided by the anterior transposition of the FN. Compromising this exposure by the use of a "short" FN rerouting or the fallopian bridge technique would, in our opinion, be equivalent to a compromise of the degree of resection and safety of the lower CNs and indeed, the patient in the vast majority of the JPs (35).

Gross total tumor removal in this series approaches the 91% mark, which compares favorably with the other reported series (Table 10). There were five cases of subtotal tumor removal in the present series. In each case, all possible outcomes had been extensively discussed with the individual patient and a final decision made keeping in mind his or her wishes. Small tumor remnants in the cavernous sinus were deliberately left behind to avoid compromising CNs III, IV, and VI function. A dominant sigmoid sinus, absence of collateral flow on temporary occlusion of the ICA, and advanced age with a poor general condition were other factors coercing us in performing a subtotal resection. We believe the small, often miniscule residuum is best managed with wait and watch and with serial MRI scans. If growth can be demonstrated, which is uncommon as a result of devascularization of the remnant, radiosurgery is a viable option in such a scenario. Recurrences in this series accounted for 10.2% of the cases in which a gross total tumor had been achieved. One of these was detected 10 years after the initial procedure. Other authors have commented on the length of time it may take for a recurrence to occur or manifest, which has been documented to be as long as 23 years (16). We fully endorse Jackson's recommendation of the required follow up to be lifelong while attesting to the practical difficulty in ensuring it (16). The recurrence rate in this and the other series highlights the infiltrative nature of the JPs, their capacity for microscopic invasion of the neighboring structures, and their ability to recur after a long time even after "total" tumor removal has been performed. We would reiterate the importance of wide removal of adjacent seemingly "uninvolved" cancellous bone to minimize recurrence (19,38).

Traditionally, the three main complications in JP surgery have been bleeding, cerebrospinal fluid leak, and aspiration. As a result of the sweeping use of embolization and a precise microsurgical technique, we have been able to reduce the need for blood transfusions to an acceptable level. There were two cases of postoperative cerebrospinal fluid leak, one of which was the result of inadequate closure of the eustachian tube (in the case requiring a second-stage transcochlear approach) and was certainly avoidable. The other instance of cerebrospinal fluid leak was in a single-stage procedure for a <2 cm ICE. The small dural opening, on reoperation, was found to be inadequately closed with the free muscle graft. A bigger piece of muscle was used to reinforce this closure with cessation of the leak. The reasons for this extremely low rate have already been explained here. The problem of aspiration has also not been a major issue in most of our patients, mainly as a result of our practice of early and intensive physiotherapy of the patients with a postoperative CN X or IX/X deficit. The detractors of surgery of JPs have often relied on the frequency and severity of these complications to advocate alternative forms of therapy. A closer look at the large series reported in the past decade reveals the low rate of complications along with a high rate of surgical control achieved when experienced and suitably trained personnel perform the surgery (Tables 10 and 11) (39).

We do not consider radiotherapy as an adequate primary treatment of these tumors. The reasons for this have been sufficiently explored in the literature (1,3). We are also in agreement with Jackson that with the technical advances made in the past 2 decades, no JP is inoperable (16). However, surgery, although being the preferable mode of treatment in the vast majority of these tumors, is not always the most prudent choice. In elderly patients with minimal clinical symptoms and a poor general condition, a conservative approach in the form of observation with serial MRI scanning is preferred. If there is growth of the tumor or more significantly, the neurologic status of the patient worsens, a partial resection of the tumor can be undertaken with a very high probability of preserving the preoperative CN functional status. In case of a very large tumor in a medically infirm and anesthetically high-risk patient with a significant brainstem compression, we would advise reversing the order of our surgical stages and performing the intracranial stage initially to sufficiently decompress the brainstem. Depending on the individual case, the second stage can then be performed or we can revert to our initial policy of observation with serial MRI scans. Again, in such patients, we favor performing partial resections to preserve clinically uninvolved CNs because the postoperative quality of life can be greatly and often irrevocably affected by the sacrifice of these nerves in this group of patients. Attempted total resections are the rule in younger individuals who have a greater capacity for compensation for the loss of CN functions. One exception to this rule is the

area of the cavernous sinus where we do not hesitate to leave behind a small remnant to preserve the III, IV, and VI CNs. In this area, the risk of damaging these CNs outweighs the potential benefit of removal of this generally minute portion, which, in our experience, can be adequately managed with observation/stereotactic radiosurgery.

CONCLUSION

We would advocate surgery in general and the infratemporal fossa type A approach in particular as the preferred treatment of choice in the management of a majority of the jugular paragangliomas. The high surgical control achieved in the majority of these large tumors with the lowest reported complication rate and no mortality in our series of 54 patients supports this conclusion. There are instances, however, in which other forms of management play an equally, if not more, important role in the treatment protocol of these extremely complex and difficult-to-treat tumors. The present series demonstrates the interplay of the various factors related to optimum treatment of these tumors and the need for this management to be concentrated in experienced hands.

REFERENCES

1. Moe KS, Li D, Linder TE, et al. An update on the surgical treatment of temporal bone paraganglioma. *Skull Base Surgery* 1999; 9:185–94.
2. Jackson CG, Haynes DS, Walker PA, et al. Hearing conservation in surgery for glomus jugulare tumors. *Am J Otol* 1996;17:425–37.
3. Jackson CG. Surgery for benign tumors of the temporal bone. In: Glasscock ME III, Gulya AJ, eds. *Surgery of the Ear*. Hamilton, Ontario: BC Decker Inc, 2003:;714–42.
4. Lundgren M. Tympanic body tumors in the middle ear—tumors of the carotid body type. *Acta Otolaryngol (Stockh)* 1949;37:366–79.
5. Capps FCW. Glomus jugulare tumors of the middle ear. *J Laryngol Otol* 1952;66:302–14.
6. Brown LA. Glomus jugulare tumor of the middle ear—clinical aspects. *Laryngoscope* 1953;63:281–92.
7. Gejrot T. Surgical treatment of glomus jugulare tumors with special reference to the diagnostic value of retrograde jugulography. *Acta Otolaryngol (Stockh)* 1965;60:150–68.
8. Shapiro MJ, Neues DK. Technique for removal of glomus jugulare tumors. *Arch Otolaryngol* 1964;79:219–24.
9. House WF, Glasscock ME. Glomus tympanicum tumors. *Arch Otolaryngol* 1968;87:550–4.
10. Mazzoni A. Jugulo-petrosectomy [Italian]. *Arch Ital Otol Rhinol Laringol* 1974;2:20–35.
11. Glasscock ME III, Harris PF, Newsome G. Glomus tumors: diagnosis and treatment. *Laryngoscope* 1974;84:2006–32.
12. Gardner G, Cocke EW Jr, Robertson JT, et al. Combined approach surgery for removal of glomus jugulare tumors. *Laryngoscope* 1977;87:665–88.
13. Fisch U. Infratemporal approach for extensive tumors of the temporal bone and base of skull. In: Siverstein H, Norrel H, eds. *Neurological Surgery of the Ear,* vol II. Birmingham: Aesculapius, 1977:34–53.
14. Gejrot T, Lindbom A. Venography of the internal jugular vein and the transverse sinuses (retrograde jugulography). *Acta Otolaryngol [Suppl] (Stockh)* 1960;158:180–6.
15. Hekster REM, Luyendijk W, Matricali B. Transfemoral catheter embolization: a method of treatment of glomus tumors. *Neuroradiology* 1973;5:208–14.
16. Jackson CG, McGrew BM, Forest JA, et al. Lateral skull base surgery for glomus tumors: long-term control. *Otol Neurotol* 2001; 22:377–82.
17. Fisch U, Mattox D. *Microsurgery of the Skull Base.* Stuttgart, New York: Georg Thieme Verlag, 1988:148–281.
18. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93:146–7.
19. Sanna M, Saleh E, Russo A, et al. *Atlas of Lateral Skullbase Surgery.* Stuttgart, New York: Georg Thieme Verlag, 1995: 132–45.
20. Fisch U, Mattox D. *Microsurgery of the Skull Base.* Stuttgart, New York: Georg Thieme Verlag, 1988:4–69.
21. Sanna M, Mazzoni A. The Modified Transcochlear Approach to the Tumors of the Petroclival Area and Prepontine Cistern. Presented at the Third Annual Meeting of the North American Skull Base Society; Acapulco, Mexico; February 15–20, 1992.
22. Sanna M, Mazzoni A, Saleh EA, et al. Lateral approaches to the median skull base through the petrous bone: the system of the modified transcochlear approach. *J Laryngol Otol* 1994;108: 1036–44.
23. Sen CN, Sekhar LN. An extreme lateral approach to intradural lesions of the cervical spine and foramen magnum. *Neurosurgery* 1990;27:197–204.
24. Fisch U, Mattox D. *Microsurgery of the Skull Base.* Stuttgart, New York: Georg Thieme Verlag, 1988:74–131.
25. Rigby PL, Jackler RK. Clinicopathologic presentation and diagnostic imaging of jugular foramen tumors. *Operative Tehniques in Otolaryngology–Head Neck Surgery* 1996;7:99–105.
26. Anand VK, Leonetti JP, al-Mefty O. Neurovascular considerations in surgery of glomus tumors with intracranial extensions. *Laryngoscope* 1993;103:722–8.
27. Kinney SE. Glomus jugulare tumor surgery with intracranial extension. *Otolaryngol Head Neck Surg* 1980t;88:531–5.
28. Gjuric M, Rudiger Wolf S, Wigand ME, et al. Cranial nerve and hearing function after combined-approach surgery for glomus jugulare tumors. *Ann Otol Rhinol Laryngol* 1996;105:949–54.
29. Makek M, Franklin DJ, Zhao JC, et al. Neural infiltration of glomus temporale tumors. *Am J Otol* 1990;11:1–5.
30. Manolidis S, Jackson CG, Von Doersten PG. Lateral skull base surgery: the Otology Group experience. *Skull Base Surgery* 1997; 7:129–37.
31. Green JD Jr, Brackmann DE, Nguyen CD, et al. Surgical management of previously untreated glomus jugulare tumors. *Laryngoscope* 1994;104:917–21.
32. Sen C, Syderman CH, Sekhar LN. Complications of skullbase operations. In: Sekhar LN, Janecka IP, eds. *Surgery of Cranial Base Tumors*. New York: Raven Press, 1993:831–39.
33. Bielamowicz S, Gupta A, Sekhar LN. Early arytenoid adduction for vagal paralysis after skull base surgery. *Laryngoscope* 2000; 110:346–51.
34. Selesnick SH, Abraham MT, Carew JF. Rerouting of the intratemporal facial nerve: an analysis of the literature. *Am J Otol* 1996; 17:793–805.
35. Pensak ML, Jackler RK. Removal of jugular foramen tumors: the fallopian bridge technique. *Otolaryngol Head Neck Surg* 1997; 117:586–91.
36. Martin CH, Prades JM. Removal of selected infralabyrinthine lesions without facial nerve mobilization. *Skull Base Surgery* 1992; 2:220–6.
37. Maniglia AJ, Sprecher RC, Megerian CA, et al. Inferior mastoidectomy-hypotympanic approach for surgical removal of glomus jugulare tumors: an anatomical and radiologic study emphasizing distances between critical structures. *Laryngoscope* 1992;102: 407–14.
38. Fisch U. Invited comments. *Am J Otol* 1996;17:806–9.
39. Briner HR, Linder TE, Pauw B, et al. Paragangliomas of the temporal bone: long term subjective assessment of the results of surgery. *Laryngoscope* 1999;109:577–83.