Management of Facial Nerve in Surgical Treatment of Previously Untreated Fisch Class C Tympanojugular Paragangliomas: Long-Term Results

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Abstract

The aim of this study was to evaluate the long-term facial nerve outcome according to management of the facial nerve in patients undergoing surgery for Fisch class C tympanojugular paragangliomas. The study population consisted of 122 patients. The infratemporal type A approach was the most common surgical procedure. The facial nerve was left in place in 2 (1.6%) of the 122 patients, anteriorly rerouted in 97 (79.5%), anteriorly rerouted with segmental resection of the epineurium in 7 (5.7%), and sacrificed and reconstructed in 15 (12.3%). One patient underwent cross-face nerve grafting. At last follow-up, House-Brackmann grade I to II was achieved in 51.5% of patients who underwent anterior rerouting and in 28.5% of those who underwent anterior rerouting with resection of the epineurium. A House-Brackmann grade III was achieved in 73.3% of patients who underwent cable nerve graft interposition. The two patients in whom the facial nerve was left in place experienced grade I and grade III, respectively. The patient who underwent cross-face nerve grafting had grade III. Gross total resection was achieved in 105 cases (86%). Management of the facial nerve in tympanojugular paraganglioma surgery can be expected to ensure satisfactory facial function long-term outcome.

Keywords

- tympanojugular paraganglioma
- glomus tumor
- facial nerve
- anterior rerouting

Introduction

Tympanojugular paragangliomas (TJPs) are predominantly benign, slow-growing, highly vascularized tumors that are often locally aggressive and infiltrate the skull base.[1] Fisch classified temporal bone paragangliomas into four categories (Class A, B, C, and D) according to location and extension based on high-resolution computed tomography examination.[2,3] According to Fisch, class C paragangliomas arise from paraganglia cells located within the adventitia of the dome of the jugular bulb and involve the infralabyrinthine temporal bone. They can spread in the following directions: inferiorly along the internal jugular vein and cranial nerves IX to XII, superiorly toward the otic capsule and the internal auditory canal, posteriorly into the sigmoid sinus, anteriorly toward the internal carotid artery (ICA), medially involving the petrous apex and the cavernous sinus, and laterally filling the hypotympanum and middle ear.[3] On the basis of the extent of the ICA involvement, Fisch class C tumors are further subdivided into four subclasses: tumors eroding the carotid foramen without involvement of the ICA (C1), tumors eroding the vertical carotid canal up to the carotid genu (C2), tumors

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involving the horizontal segment of the ICA (C3), and tumors reaching the anterior foramen lacerum with extension into the cavernous sinus (C4).

Because the vertical portion of the facial nerve (FN) is centered and closely related to the jugular bulb, a large proportion of patients with class C TJP require some sort of FN management. Usually, the tumor is found to be eroding the fallopian canal and lying in contact with the epineurium of the FN without infiltrating it. In these cases, permanent anterior rerouting of the FN, which is an integral part of the infratemporal fossa type A approach (IFTA-A), is sufficient management to deal with FN involvement. When the epineurium of the FN is infiltrated by the tumor, segmental resection of the involved epineurium should be performed. If the perineurium is infiltrated by the tumor, the involved segment of the nerve is resected and the nerve repaired by cable grafting.

The objective of this study is to report the long-term FN function results in patients undergoing surgery for Fisch class C TJPs according to FN management.

Methods

The charts of 171 patients with a diagnosis of Fisch class C TJP surgically treated during the period from April 1988 to April 2012 were examined retrospectively. Patients were excluded from the study if no data were available on FN function at 12 months after surgery. The FN follow-up at 1 year was missing for 39 patients, 16 of whom had not completed the postoperative year at the time of this study. The remaining 23 patients were lost to follow-up. Patients who underwent either prior surgery (8 cases) or preoperative radiotherapy (2 cases) for treatment of their paraganglioma were also excluded. Thus, 122 patients who had a follow-up period for postoperative FN function of at least 1 year became the subjects of this study. The collected data were analyzed for age, sex, tumor class, surgical procedure, intraoperative management of the FN, preoperative and postoperative FN function, and radiological follow-up findings. Tumors were staged according to Fisch classification.[3] The preoperative and postoperative FN function was graded according to the House–Brackmann (HB) scale.[4] When there was a doubt in assigning the patient to grade II or III, we preferred to assign the patient to the higher grade. In all the cases, FN function was monitored intraoperatively using an electromyographic system. For the present study, FN results were collected for two specific time intervals: before surgery and after a minimum of 1 year of follow-up. Follow-up was defined as the period of time from surgery to the most recent office visit.

Results

Demographic Data

The study population consisted of 37 (30.3%) men and 85 (69.7%) women. The mean age of patients at the time of surgery was 45.5 ± 12.3 years (range, 16 to 67 years). Sixty-seven tumors (54.9%) were on the left side, and 55 (45.1%) were on the right side. Distribution of tumors according to Fisch classification is depicted in ►**Table 1**. The duration of the follow-up period ranged from 12 to 156 months (mean, 39.4 ± 32.6 months).

Treatment

Single-stage tumor removal was adopted in 111 cases. The IFTA-A was chosen in 109 patients. The fallopian bridge technique was used in 2 patients with a Fisch class C1 TJP with predominantly posterior disease. A planned staged procedure was adopted by using the IFTA-A as the initial surgery in 11 patients who had more than a 2 cm intradural tumor extension; a petro-occipital-trans-sigmoid approach was used for the planned second-stage intradural removal of the tumor in 5 cases, a transcochlear approach was used in 5 cases, and an extreme lateral approach in the remaining case. This staging strategy has been adopted to avoid the risk of having postoperative cerebrospinal fluid leak resulting from the wide neck exposure and to reduce intracranial tension during the second stage.

Facial Nerve Management

►**Table 2** shows the outcome of FN function depending on FN management.

Facial Nerve Left in Place

The FN was left intact in 2 (1.6%) of the 122 patients by using the fallopian bridge technique. Preoperatively, both patients had normal FN function, whereas at last follow-up one patient had HB grade I, and the remaining one had HB grade III.

Facial Nerve Grafting

Sixteen patients underwent surgical resection of the mastoid segment of the FN because of tumor infiltration. The defect

Table 1 Distribution of tumors according to Fisch classification[3]

Class C *N* (%)	Class D *N* (%)	ICE *N* (%)
C1 17 (13.9)	0	0
C2 73 (59.8)	De1–12 (16.4) De2–1 (1.3) Di1–14 (19.1) Di2–2 (2.7) De1Di1–3 (4.1) De2 Di1–3 (4.1)	35 (47.9)
C3 29 (23.8)	De1–5 (17.2) De2Di1–2 (6.9) De1Di2–1 (3.5) De2Di2–1 (3.5) Di1–12 (41.3) Di2–6 (20.7)	27 (93.1)
C4 3 (2.4)	De1Di1–2 (66.7) Di2–1 (33.3)	3 (100)

Abbreviation: ICE, intracranial extension.
The incidence of intracranial extension was directly related to the class of tumor, with Class C1 tumors having the lowest involvement of the intracranial involvement (0%) and Class C4 the highest (100%).

Table 2 Facial nerve outcomes based on facial nerve management

Treatment of FN	Number of cases	Preoperative	At last follow-up
Left in place	2	2 (I)	1 (I) 1 (III)
Sural graft	15	4 (I) 3 (II) 1 (V) 7 (VI)	11 (III) 2 (IV) 2 (VI)
Cross face	1	1 (VI)	1 (III)
Anterior rerouting with resection of epineurium	7	5 (II) 1 (III) 1 (IV)	1 (I) 1 (II) 2 (III) 3 (IV)
Anterior rerouting	97	97 (I)	23 (I) 27 (II) 45 (III) 1 (IV) 1 (VI)

Abbreviation: FN, facial nerve.

was repaired using a sural nerve graft in the same sitting in 15 cases. Preoperatively, four patients had normal function, three had HB grade II, one had HB grade V, and seven had complete FN palsy. At last follow-up, 11 of the 15 patients (73.3%) had HB grade III, 2 (13.3%) had HB grade IV, and 2 (13.3%) had grade VI. One of the 16 patients had a long-standing preoperative grade VI palsy and underwent cross-face nerve grafting. This patient reached HB grade III function, at 1-year follow-up.

Anterior Rerouting of the Facial Nerve

Ninety-seven patients with preoperative grade I FN function underwent a permanent anterior transposition of the FN. At long-term follow-up, 23 patients (23.7%) had HB grade I, 27 (27.8%) had HB grade II, 45 (46.4%) had HB grade III, 1 (1%) had HB grade IV, and 1 (1%) had HB grade VI.

Permanent anterior transposition of the FN was also performed in seven patients with preoperative FN dysfunction. Preoperatively, five patients (71.4%) had HB grade II, one (14.3%) had HB grade III, and one (14.3%) had HB grade IV. The patient with a preoperative HB grade IV FN function had developed this paresis as a complication of the preoperative embolization, and the functional status had not improved until the last follow-up visit. The remaining six patients of this group required segmental removal of the epineurium of the mastoid segment of the FN because of tumor infiltration. At final follow-up, one patient recovered to HB grade I, one patient had HB grade II, two had HB grade III, and two had HB grade IV.

Complications, Tumor Removal/Recurrences

There were no perioperative death. The most frequent perioperative complication was cerebrospinal fluid leak, which occurred in two cases (1.6%). Gross total tumor removal was accomplished in 105 cases (86%). From this number, three cases (2.8%) developed tumor recurrence. Subtotal removal was achieved in 17 cases (14%). Fifteen of these residual tumors were intracranial involving the cavernous sinus, posterior fossa dura, and the vertebral and basilar arteries. For extracranial residual tumors, partial resection was performed in two cases due to significant cardiac morbidities.

Discussion

One of the cornerstones of successful TJP surgery is competence in the management of the FN. FN anterior rerouting techniques were developed to enhance exposure of the jugular foramen and ICA.[5,6] In 1952, Capps[7] first described the resection of the jugular bulb and mobilization of the mastoid segment of the FN for removal of a TJP. Subsequently, in the 1960s, Shapiro and Neues[8] described the combination of radical mastoidectomy, anterior rerouting of the FN, and neck exposure with ligation of the internal jugular vein. In 1978, Fisch[2] introduced the IFTA-A, which provides excellent control of the jugular foramen and infralabyrinthine areas, as well as the intratemporal ICA. The IFTA-A includes permanent anterior rerouting of the FN, blind sac closure of the external auditory canal, and closure of the sigmoid sinus and jugular vein. This approach is particularly indicated for removal of Fisch Class C tumors, which are generally considered tumors with an advanced level of extension and ICA involvement.[1] The main disadvantages of the IFTA-A are postoperative conductive hearing loss due to ear canal closure and FN paresis related to the anterior rerouting of the FN.

During the past two decades, there have been increased number of studies describing surgical excision of TJPs without FN mobilization.[9–14] In our opinion, these conservative approaches can be safely considered for nonvascularized jugular foramen tumors that do not infiltrate the ICA walls (i.e., lower cranial nerve schwannomas, meningiomas, and chondrosarcomas) and for Fisch Type C1 TJPs with predominantly posterior disease. The major limitation of these procedures is the limited access to the middle ear, horizontal petrous ICA, and petrous apex, creating the risk of residual tumor and the potential for catastrophic injury to the ICA. Short mobilization of the distal segment of the FN with or without removal of the posterior canal wall can marginally improve access to the vertical segment of the ICA and the tumor in the middle ear.[15–19] However, it is still not sufficient to achieve distal ICA control. Compromising this exposure by keeping the FN in place or by a short FN mobilization would, in our opinion, be equivalent to a compromise of the degree of resection and safety. To minimize the risk of recurrence, all the bone surrounding the tumor should be drilled out aggressively, keeping in mind that the infiltration is often more extensive than that visible on the scans. Infiltrated bone around the jugular fossa cannot safely and adequately be removed without rerouting the FN, particularly if there is anterior extension along the carotid canal.[1,13] The same is true for the tympanic bone, often infiltrated in a medial to lateral direction. Any attempt to keep the FN in place and preserve the external auditory canal and middle ear contents may result in incomplete tumor removal. Our FN results after long anterior rerouting of the FN in patients with preoperative normal facial function show recovery to HB Grade I to III in 98% of cases. These results are consistent with those reported in other series.[2,3,20–26] A summary of FN outcomes in studies reporting 10 or more patients who underwent either short or long anterior rerouting of the FN is reported in ►**Table 3**. As already noticed by Parhizkar et al,[21] it is difficult to summarize the results of FN rerouting objectively because each series is different with regard to tumor histology, preoperative FN function, type of FN management, and descriptions of postoperative FN function. In fact, if we consider the percentages of patients achieving HB grade I to II function, the results of the different series are quite different. Conversely, when we consider the percentages of patients achieving HB grade I to III, the results obtained by different studies are very similar. This can be explained by the interobserver variability, especially for HB grade II and HB grade III.

Analysis of the table reveals that short mobilization from the second genu results in a 83 to 100% rate of HB grade I to II at long-term follow-up. In contrast, mobilization from the geniculate ganglion results in a 51 to 88% rate of HB grade I to II function at long-term follow-up. On the basis of these data, it seems that the FN outcomes are better after short anterior rerouting. This finding, however, does not permit the conclusion that FN rerouting should be limited to its shortest possible length. Considering the percentage of total tumor removal alters the picture significantly. Pensak and Jackler[11] reported the rate of gross total removal as 71% by keeping the nerve in place with the fallopian bridge technique. Spector et al[16] achieved total tumor removal in 78% of their cases using short anterior rerouting. In contrast, Moe et al[22] achieved total tumor removal in 80% of their cases who underwent long anterior rerouting of the FN. Using the same method, Green et al[27] achieved total tumor removal in 85% of their cases. Total tumor removal was achieved in 86% of our patients.

Table 3 Comparative results of postoperative facial nerve function after anterior rerouting of the FN

Study	Number of patients	Type of anterior rerouting	Pathology	HB grade	HB I-II %	HB I-III %
Spector et al.[16]	35	Short	TJPs	I-II, 29 III-IV, 1 5, VI	82.9	–
Farrior[15]	17	Short	TJPs	16, I 1, 80% recovery	94.1	–
Cece et al.[17]	13	Short	TJPs	I, 13	100	100
Brackmann[20]	22	Long	TJPs and other tumors	I, 10 II, 7 III, 1 IV, 2 V, 1 VI, 1	77.3	81.8
Poe et al.[25]	32	Long	TJPs and other tumors	I, 5 II, 13 III, 9 IV, 3 V,2	56.2	84.4
Woods et al.[26]	37	Long	TJP, glomus vagale	I, 6 II,16 III.11 IV,3 V,1	59.4	89
Pareschi et al.[23]	25	Long	TJPs	I, 3 II, 12 III, 8 IV, 1 V, 1	60	92
Moe et al.[22]	52	Long	TJPs	I, 36 II, 10 III, 4 IV, 1	88	98
This study	97	Long	TJPs	I, 23 II, 27 III, 45 IV, 1 VI, 1	51.5	98

Abbreviations: HB, House-Brackmann; TJP, tympanojugular paraganglioma.

Various factors are crucial for the postoperative results of FN function after long anterior rerouting. These include surgical technique, the degree of surgical trauma, and the vascularization of the FN.[28] To minimize trauma to the nerve, we suggest the following surgical refinements:

1. To obtain extra length of free nerve and minimize traction, the FN should be freed in the parotid gland up to the pes anserinus
2. Skeletonization from the stylomastoid foramen to the geniculate ganglion should be done with large diamond burs under copious irrigation.
3. Preservation of the periosteum and soft tissue of the stylomastoid foramen helps in maintaining a blood supply to the nerve.
4. Removal of the stapes superstructure facilitates dissection while minimizing the risk of sensorineural hearing loss.
5. Sharp dissection using a Beaver knife of firm fibrous attachments of the mastoid segment of the FN, especially in the vertical segment is required.
6. Leaving the pregeniculate portion of the FN in place helps in preserving blood supply.
7. Once the anterior rerouting is accomplished and the nerve is positioned in its new bony bed, the mandibular condyle is anteriorly displaced and taken in place by using a self-retaining retractor. We avoid using the infratemporal fossa retractor so as to not damage the transposed FN.
8. FN monitoring should remain silent throughout the procedure.

Less frequently, the FN is found to be infiltrated by the tumor and the surgeon has to evaluate the degree of nerve involvement. The vertical segment of the FN is the most common site of infiltration in TJPs; however, involvement in the tympanic segment, stylomastoid foramen, internal auditory canal, or cerebellopontine angle can also occur.[28,] In this study, the presence of a preoperative facial paresis often precluded preservation of the nerve. In fact, we observed that those patients with preoperative FN weakness were more likely to have an infiltrated FN. In our series, 7 (5.7%) patients necessitated segmental resection of the epineurium and 16 patients (13.1%) had FN interruption because of infiltration of both perineurium and endoneurium. It is interesting to notice that 19 (82.6%) of these patients had preoperative facial weakness. ►**Fig. 1** shows the grading scheme for neural invasion proposed by Makek et al.[29]

Segmental resection of the infiltrated epineurium can be performed when the perineurium remains intact.[3,30] In such a case, the nerve is rerouted anteriorly before beginning to remove the tumor, and definitive management of the nerve is performed at the end of the surgery when the field is clear of tumor and blood. This strategy reduces the risk of nerve interruption during the rerouting of a fragile nerve deprived of its epineurium. When tumor removal is accomplished, incision of the sheath is started, either proximally or distally, at a site that is free of tumor infiltration. Dissection of the sheath then proceeds by a combination of sharp and blunt dissection under high magnification until the whole infiltrated epineurium is dissected away from the perineurium. If the

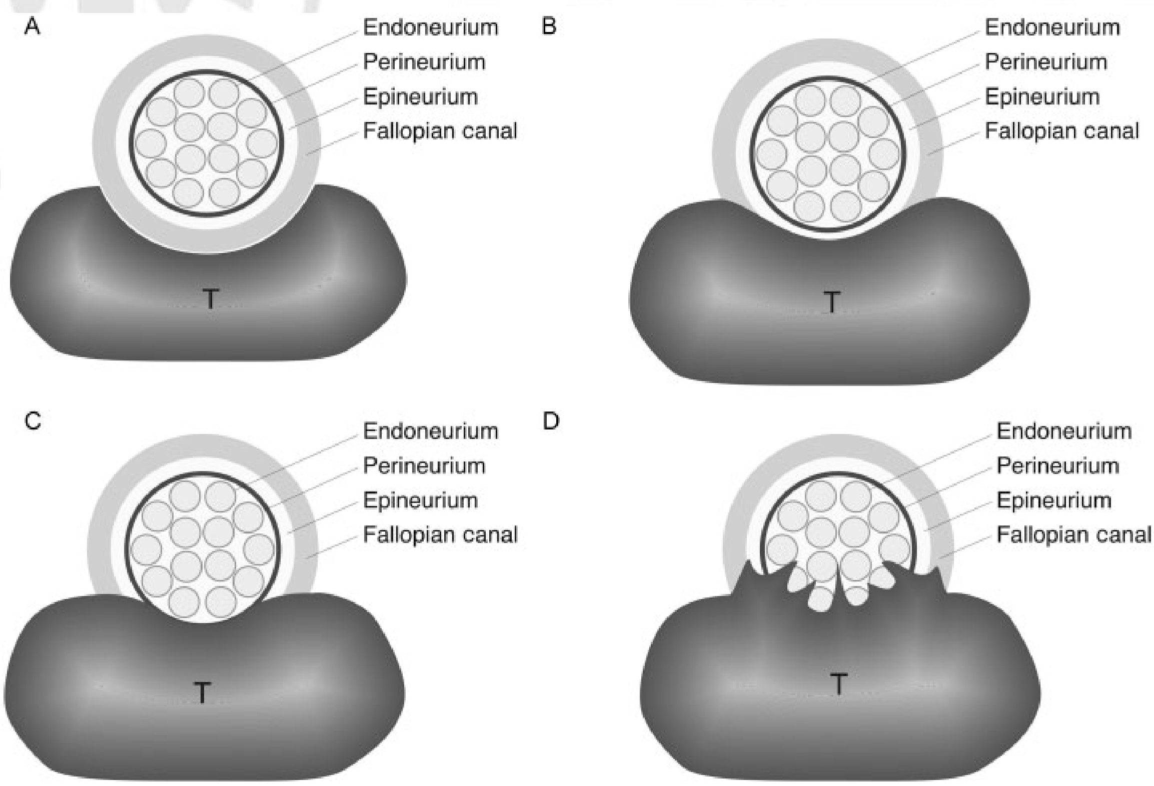

Fig. 1 Degrees of facial nerve involvement according to Makek et al.[29] (A) Grade I, tumor remains 1 mm or more from the epineurium. (B) Grade II, invasion of the epineurium. (C) Grade III, invasion of the perineurium. (D) Invasion of the endoneurium. T, tumor.

perineurium is free of tumor, dissecting the sheath is considered sufficient management and the nerve is returned to the new canal and fixed in place using fibrin glue. Among the seven patients who underwent segmental resection of infiltrated epineurium, four patients (57%) achieved good FN function (HB grade I-II), and three patients (43%) experienced HB grade IV.

If the perineurium or nerve fibers are infiltrated by the tumor, the involved segment is resected, and reconstruction is performed. The type of reconstruction depends as usual on the amount of nerve available. If there is enough to create a tension-free reconstruction, end-to-end anastomosis is the choice of reconstruction. Alternatively, an interposition graft is used. The sural nerve or the greater auricular nerve are the most commonly used donor nerves. We prefer the sural nerve because of its length, better size matching and neural content, and the fact that it can be obtained by an assistant while the surgeon works on the primary lesion.[31]

Numerous techniques have been described both in neural preparation and stabilization of anastomoses.[32] No significant differences have been reported between single or multiple sutures, nor the use of fibrin glue to stabilize the anastomosis. It is the tension-free nature of the repair on a stable bed, and prevention of soft tissue interposition, that have an impact on the outcome.

The approximation of the proximal FN and the graft is best stabilized in a bony groove supplemented by fibrin glue and fascia. The extratemporal component is stabilized using 8.0 monofilament epineurial sutures.

Leonetti et al[33] reported in their series of nine patients who underwent FN grafting after TJP removal that 89% achieved HB grade III. In the series of eight patients who underwent grafting reported by Moe et al,[22] 75% had HB grade III. In the present study, 11 (73.3%) of the 15 cases who underwent grafting achieved HB grade III, whereas 2 patients (13.3%) experienced HB grade IV. The remaining two patients experienced poor outcome (HB grade VI). These two patients had a pre-existing deficit of longer than 1 year. From the data reported in the literature, the factors that adversely affect the successful postoperative recovery (HB grade III) are the presence and duration of the preoperative FN deficit, increasing age, length of the graft, degree of mobilization of the nerve, and number of anastomoses.[31] Based on the fact that the preoperative presence and duration of the FN deficit represent the main prognostic factors, it is clear that early diagnosis and proper timing of surgery are fundamental to increase the chances of a good recovery. In a previous publication, the duration of the preoperative deficit as an indicator of the final prognosis was closely analyzed.[31] To achieve a high rate of good postoperative recovery, the cutoff point should be considered to be 1 year after the occurrence of the preoperative clinical deficit.

The use of a facial hypoglossal anastomosis in the setting of TJPs is limited. A high proportion of tumors that result in FN paralysis are also associated with compound lower cranial nerves palsies. Although the hypoglossal nerve is less commonly paralyzed, the combination of a vagal and hypoglossal paralysis would lead to a significantly higher rate of swallowing difficulties and aspiration. Therefore, the situation of a patient with a grafted FN who achieves a poor result is a difficult one to manage. The options include a facial to hypoglossal jump graft anastomosis, with preservation of hypoglossal function or non-neural rehabilitation techniques.[28]

Conclusion

In our opinion, the IFTA-A is the approach of choice for resecting tympanojugular class C paragangliomas. Anterior rerouting of the FN is essential to obtain adequate exposure and control of the ICA and allows bone infiltrated by the pathology of the jugular foramen to be removed. Temporary FN palsy, with a high percentage of recovery to grade I to III at 1 year, is the price to be paid for attempting safe and total tumor removal, which the authors consider the correct goal of TJP surgery. FN reconstruction is rarely required in TJP surgery because tumors that invade the FN represent an uncommon category.

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