# Surgical Management of Intrinsic Tumors of the Facial Nerve

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**BACKGROUND:** Intrinsic tumors of the facial nerve are a rare entity. Dealing with this subset of tumors is challenging both in terms of decision making and surgical intervention. **OBJECTIVE:** To review the outcomes of surgical management of facial nerve tumors and cable nerve graft interpositioning.

**METHODS:** A retrospective analysis was performed at a referral center for skull base pathology. One hundred fifteen patients who were surgically treated for facial nerve tumors were included. In case of nerve interruption during surgery, the cable nerve interpositioning technique was employed wherein the facial nerve palsy lasted for less than 1-yr duration. In cases of facial nerve palsy lasting for greater than 1 yr, the nerve was restituted by a hypoglossal facial coaptation.

**RESULTS:** Various degrees of progressive paralysis were seen in 84 (73%) cases. Sixty nine (60%) of the tumors involved multiple segments of the facial nerve. Sixty-two (53.9%) tumors involved the geniculate ganglion. Seventy four (64.3%) of the cases were schwannomas. Hearing preservation surgeries were performed in 60 (52.1%). Ninety one (79.1%) of the nerves that were sectioned in association with tumor removal were restituted primarily by interposition cable grafting. The mean preoperative House-Brackmann grading of the facial nerve was 3.6. The mean immediate postoperative grading was 5.4, which recovered to a mean of 3.4 at the end of 1 yr.

**CONCLUSION:** In patients with good facial nerve function (House-Brackmann grade I-II), a wait-and-scan approach is recommended. In cases where the facial nerve has been interrupted during surgery, the cable nerve interpositioning technique is a convenient and well-accepted procedure for immediate restitution of the nerve.

**KEY WORDS:** Facial nerve, Facial nerve tumors, House–Brackmann grading, Schwannoma, Meningioma, Subtotal petrosectomy, Transotic approach, Translabyrinthine approach, Cable nerve graft interpositioning, Sural nerve

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acial nerve tumors (FNTs) are a rare subset of tumors that may arise from anywhere along the tortuous intracranial, transtemporal, and an extratemporal course of facial

ABBREVIATIONS: CPA, cerebellopontine angle; FN, facial nerve; FNT, facial nerve tumor; GG, geniculate ganglion; GSPN, greater superficial petrosal nerve; HB, House–Brackmann; HRCT, high-resolution computed tomography; IAC, internal auditory canal; MCF, middle cranial fossa; MRI, magnetic resonance imaging; SN, sural nerve; SNIG, sural nerve interposition grafting.

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nerve (FN). Even though FNTs present with varying grades of facial paralysis, to intervene surgically, in most cases, would mean total loss of facial function, at least temporarily. Hence, the treating practitioner must demonstrate prudence in his decision to intervene surgically, especially in the young and in patients with minimal facial paralysis. Once the nerve is interrupted, reconstruction must be performed immediately to get the best results, either by means of a primary end-to-end coaptation or by a cable nerve graft interposition.1 In this article, we discuss our experience in the management of FNTs in one of the largest published series in world literature and analyze aspects related to clinical features, decision making, and surgical outcomes.

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## **METHODS**

A retrospective chart review was performed of patients operated for intrinsic tumors of the FN between July 1989 and July 2015 at our center, which is a quaternary referral center for skull base pathology. The exclusion criteria included patients with primary malignancies of the FN, less than 1 yr of follow-up and those with inadequate for follow-up. FNTs that presented with other tumors as part of neurofibromatosis were also excluded from the study.

At our center, the FN function is recorded according to the House–Brackmann (HB) grading system.<sup>2</sup> To precisely evaluate FN function pre- and postoperatively, color photographs of the face are taken in 4 positions: facial muscles at rest, tight closure of eyes with a grin, raised eyebrows, pouting lips.<sup>3</sup> All the cases were evaluated preoperatively with high-resolution computed tomography scan and magnetic resonance imaging (MRI) with contrast enhancement. Angiography or angio-MRI was done in cases in which the tumor was in close association with important vasculature. Audiometric data included 4-frequency analysis of pure tone average for bone-conduction, air-conduction, and speech discrimination scores according to the Sanna classification of hearing.<sup>4</sup>

The surgical approaches used to extirpate FNTs included transmastoid, subtotal petrosectomy, middle cranial fossa (MCF), transotic, translabyrinthine, transcochlear, transparotid, or combined approaches. When the FN was interrupted intraoperatively, a sural nerve interposition grafting (SNIG) was done to reconstruct it.<sup>5,6</sup> Details of tumor clearance, intraoperative events, complications, FN, and hearing status at the end of 1-yr follow-up were recorded. All patients were followed by annual MRI scanning.

Data analysis was done by Version 24 SPSS (IBM Corporation, Armonk, New York) statistical package. Chi square tests and Wilcoxon matched pairs test were used for deriving *P* values. A *P* value less than .05 was considered statistically significant.

This study was approved by the institutional review board of the hospital for ethical research. At our center, the patient consent taken before surgery also includes consent for the use of clinical data for scientific purposes.

#### **RESULTS**

One hundred fifteen patients were included in the study. Sixtyone (53%) patients were males and 54 (47%) were females. The ages ranged from 2.5 to 74 yr (mean 42.5 yr). A total of 46.1% were less than 40 yr. The mean follow-up was 34.8 mo (range, 12-300 mo). Fifty-eight (50.4%) lesions were right sided and 57 (49.6%) were left sided.

## **Symptomatology**

Seven (6.0%) patients were referred to our center for surgical intervention after a prior surgery elsewhere. On presentation, FN symptoms were seen in 104 (90.4%) cases including various degrees of progressive paralysis in 84 (73%) cases, hemifacial spasm in 20 (17.4%), and an intermittent paralysis in 5 (5.7%) cases (Table 1). Dizziness/vertigo, hearing loss, and tinnitus were present in 55 (47.8%), 35 (30.4%), and 28 (24.3%) cases, respectively. Twenty four (20.8%) of tumors presented as retrotympanic masses and only 8 (6.9%) presented as external auditory canal

polyps. Apart from VII and VIII nerves, none of our patients presented with any other cranial nerve palsy.

## **Pathology**

A diagnosis of FNTs was made only on the basis of radiology. We do not perform a preoperative biopsy of any polypoidal lesion of the ear. Definitive diagnosis is made histopathologically after surgery. The most common entity was schwannoma with 74 (64.3%) cases followed by hemangioma with 37 (32.2%) cases (Table 1). There were 2 (1.7%) cases each of meningioma and neurofibroma.

#### **Site of Involvement**

Most often the tumors involved multiple segments of the FN, ie, 69 (60%). The most common site of involvement was the geniculate ganglion (GG) as seen in 62 (53.9%) of the cases followed by the internal auditory canal (IAC) seen in 48 (41.7%) of cases. Twenty-one (18.3%) cases were purely intradural (IAC and cerebellopontine angle [CPA]) and the remaining 94 (81.7%) were extradural. We had no case where the tumor was confined only to the extratemporal (retroparotid and parotid) portions of the FN. Both schwannomas and hemangiomas predominantly involved the GG (52.7% and 48.6%, respectively). Schwannomas predominantly involved multiple segments as seen in 55 (74.3%) cases as compared to 11 (29.7%) cases of hemangioma. This was found to be statistically significant (P = .001). Sixty-three (54.8%) tumors were limited to the confines of the temporal bone, while 10 (8.6%) had extensions into the posterior cranial fossa and 43 (37.3%) into the MCF.

### **Surgical Approaches**

A variety of skull base approaches were used for resection of the tumors depending on the site and extent of the tumor (Table 1). In 61 (53%) cases, hearing preservation surgeries were performed, the most common being the middle cranial fossa approach in isolation or with a transmastoid approach. The translabyrinthine approach was used in 25 (21.7%) cases and was the most common approach that led to a sacrifice of the hearing. Gross total resection was achieved in 101 (87.8%) cases. One surgery had to be stopped due to intraoperative hemorrhage.

Ninety four (81.7%) of the nerves that were sectioned in association with tumor removal were restituted primarily either by SNIG (91 cases) or end-to-end coaptation (3 cases). The sural nerve (SN) was used as a graft material in all but 4 cases where a great auricular nerve was used. Three cases with long-standing preoperative FN palsy (>1 yr) were managed with a facial-hypoglossal and 1 more for a facial-masseteric nerve coaptation. No further treatment was necessary in any of the 17 (14.7%) cases wherein the integrity of the FN was maintained intraoperatively (Table 1).

## **FN Outcomes**

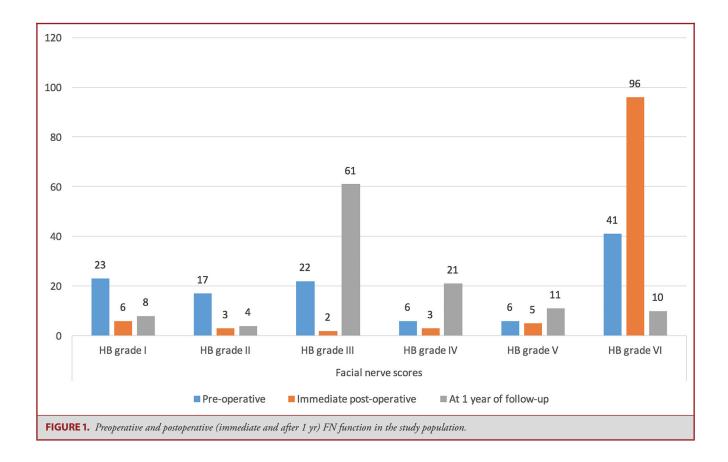
Preoperatively, 53 (46.1%) cases presented with HB grades IV to VI. Immediately after surgery, 96 (83.5%) developed an HB

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| Patient and tumor characteristics  |  | No. (%)             |
|--|--|---------------------|
| Demography and treatment details   |  |                     |
| Patients   |  | 119                 |
| Wait and scan <sup>a</sup>   |  | 4                   |
| Surgery  |  | 115                 |
| Mean age   |  | 42.5 (range 2.5-74) |
| Male: female ratio   |  | 1:0.89              |
| Surgery  |  | 115                 |
| Patients previously operated elsewhere                                   |  | 7 (6)               |
| Symptoms   |  |                     |
| FN weakness  |  | 84 (73)             |
| Hemifacial spasm   |  | 20 (17.4)           |
| Dizziness/vertigo  |  | 55 (47.8)           |
| Hearing loss   |  | 35 (30.4)           |
| Tinnitus   |  | 28 (24.3)           |
| Histopathology   |  |                     |
| Schwannoma   |  | 74 (64.3)           |
| Hemangioma   |  | 37 (32.2)           |
| Meningioma   |  | 2 (1.7)             |
| Neurofibroma   |  | 2 (1.7)             |
| Consistency  |  | ( - )               |
| Hard   |  | 107 (93)            |
| Cystic   |  | 8 (6.9)             |
| Site of involvement  |  | 2 (412)             |
| Single site  |  | 46 (40)             |
| Multisite  |  | 69 (60)             |
| Geniculate ganglion  |  | 62 (53.9)           |
| Internal auditory canal  |  | 48 (41.7)           |
| Mastoid segment  |  | 35 (30.4)           |
| Tympanic segment   |  | 30 (26)             |
| Labyrinthine segment   |  | 23 (20)             |
| Cerebellopontine angle   |  | 12 (10.4)           |
| Stylomastoid foramen and retroparotid segment                            |  | 4 (3.4)             |
| Surgical approaches  |  | 1 (3.1)             |
| Hearing preservation surgeries   | MCFA                                   | 18 (15.6)           |
|  | MCFA + TMA                             | 15 (13)             |
|  | TMA                                    | 14 (12.1)           |
|  | TMA + TPA                              | 7 (6)               |
|  | STP                                    | 5 (4.3)             |
|  | RSA                                    | 2 (1.7)             |
| Hearing destruction surgeries  | TLA                                    | 25 (21.7)           |
|  | TCA                                    | 16 (13.9)           |
|  | TOA                                    | 12 (10.4)           |
|  | STP + drilling out of the otic capsule | 1 (0.8)             |
| FN management  | 31. Talking dat of the one capsule     | . (0.0)             |
| Restitution of the FN after sectioning the nerve intraoperatively        | Interposition cable grafting of the FN | 91 (79.1)           |
| ites at a control of the fire area sectioning the herve intraoperatively | End-to-end coaptation                  | 3 (2.6)             |
|  | XII-VII/V-VII coaptation               | 4 (3.4)             |
| FN integrity maintained intraoperatively (no further treatment)          | Fascicle preservation surgeries        | 3 (2.6)             |
|  | Subtotal resection and decompression   | 7 (6.0)             |
|  | Near total resection                   | 7 (6.0)             |

<sup>&</sup>lt;sup>a</sup> Excluded from analysis, TCA: transcochlear approach, TLA: translabyrinthine approach, TOA: transotic approach, STP: subtotal petrosectomy, TMA: transmastoid approach, MCFA: middle cranial fossa approach, TPA: transparotid approach, RSA: retrosigmoid approach, FN: facial nerve.



grade VI paralysis which recovered to HB grades I to III in 73 (63.5%) cases by 1 yr. The mean preoperative HB grading of the FN was 3.6. The mean immediate postoperative grading was 5.4 which recovered to a mean of 3.4 at the end of 1 yr (Figure 1).

In the subgroup of 17 patients in whom the nerve was anatomically intact after surgery, 15 (88.2%) presented preoperatively with HB grades I to III. Immediately after surgery, 6 (35.3%) developed HB grades IV to VI which recovered to HB grades I to II in 12 (70.6%) and HB grade III in 4 (23.5%) patients. One patient remained at HB grade IV postoperatively (Table 2). Of the 17 cases, 13 were schwannomas and 4 were hemangiomas. We also did a comparison of FN outcomes between schwannomas and hemangiomas (Table 3). There was a positive correlation between pre- and postoperative (1 yr) grades both in schwannomas and hemangiomas.

In the subgroup of 91 patients who underwent SNIG, 45 (49.5%) presented preoperatively with HB grades IV to VI. Immediately after surgery, all of them developed HB grades IV to VI which recovered to HB grade III in 54 (59.3%) of the patients at the end of 1 yr. None of our patients recovered to HB grade I or II after SNIG. Preoperative HB grading of the FN did not have a significant effect on the postoperative FN outcome (P = .273). Histopathology did not have any significant effect on the outcome

(P = .887). Analysis showed that patients who recovered to HB grade III were younger than those who fared worse (38.6 vs 45.5) but this was not significant (P = .188). When the data were analyzed for single site vs multisite involvement for recovery to HB grade III and HB grade IV to VI, results showed that there was statistical significance (P = .001).

Three groups of coaptation were designed<sup>7</sup> and analyzed: *intradural coaptation*, wherein the proximal coaptation was at the brainstem and the distal site was in the IAC, *transdural coaptation*, wherein the proximal site was in the CPA or the IAC and distal site in the part of the temporal bone that remained after excision of the lesion, *extradural coaptation*, wherein both the proximal and distal sites of coaptation were in the temporal bone or extratemporally (pre-parotid and parotid parts of the FN).

Postoperative FN grades were HB grade III in 7 (50%), 12 (50%), and 35 (66%) cases in intradural, transdural, and extradural coaptation, respectively (Table 4). However, there was no statistical significance between the groups (P = .307). When the preoperative FN function was analyzed with the postoperative outcomes, we found that there was an overall improvement in FN function in extradural coaptation (P = .011) and in transdural coaptation (P = .118) but an overall worsening of FN results after an intradural coaptation (P = .027).

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TABLE 2. Comparison Between 1-yr Postoperative FN Outcomes Between SNIG, Hypoglossal-Facial Anastomosis, and in Cases Where the **Anatomical Integrity Was Preserved** 

|                               |               | Postoperative FN function (9 | %)             | Total     |
|-------------------------------|---------------|------------------------------|----------------|-----------|
|                               | HB grade I-II | HB grade III                 | HB grade IV-VI |           |
| FN integrity preserved        | 12 (70.5)     | 4 (23.5)                     | 1 (5.8)        | 17 (100)  |
| SNIG                          | 0 (0)         | 54 (59.3)                    | 37 (40.6)      | 91 (100)  |
| End to end anastamosis        | 1 (33.3)      | 2 (66.7)                     | 0 (0)          | 3 (100)   |
| XII-VII and V-VII anastamosis | 0 (0)         | 1 (25)                       | 3 (75)         | 4 (100)   |
| Total                         | 13            | 61 (59.3%)                   | 41 (40.7%)     | 115 (100) |

HB, House-Brackmann; FN, facial nerve; SNIG, sural nerve interposition grafting.

| ı | TABLE 3. Comparison of FN Outcomes Between Schwannomas and Hemangiomas and Correlation of Preoperative Facial Nerve Outcome on the |
|---|--|
| ı | Postoperative Outcomes   |

| Histology  |  |         |          | Postoper | ative facial ne | rve HB grades | at 1 yr (%) |          | Total    |
|------------|--|---------|----------|----------|-----------------|---------------|-------------|----------|----------|
|            |  |         | T.       | Ш        | III             | IV            | V           | VI       |          |
| Hemangioma | Preoperative facial nerve<br>HB grades (%) | I       | 4 (50)   | 1 (12.5) | 3 (37.5)        | 0 (0)         | 0 (0)       | 0 (0)    | 8 (100)  |
|            |  | II      | 1 (25)   | 1 (25)   | 1 (25)          | 0 (0)         | 1 (25)      | 0 (0)    | 4 (100)  |
|            |  | Ш       | 0 (0)    | 0 (0)    | 3 (60)          | 1 (20)        | 0 (0)       | 1 (20)   | 5 (100)  |
|            |  | IV      | 0 (0)    | 0 (0)    | 1 (33.3)        | 0 (0)         | 2 (66.7)    | 0 (0)    | 3 (100)  |
|            |  | V       | 0 (0)    | 0 (0)    | 3 (100)         | 0 (0)         | 0 (0)       | 0 (0)    | 3 (100)  |
|            |  | VI      | 1 (7.1)  | 0 (0)    | 5 (35.7)        | 4 (28.6)      | 2 (14.3)    | 2 (14.3) | 14 (100) |
|            | Total                                      |         | 6 (16.2) | 2 (5.4)  | 16 (43.2)       | 5 (13.5)      | 5 (13.5)    | 3 (8.1)  | 37 (100) |
|            | Chi-square value = $33.192$ ; $P =$        | = .126  |          |          |                 |               |             |          |          |
| Schwannoma | Preoperative facial nerve<br>HB grades (%) | I       | 2 (13.3) | 1 (6.7)  | 10 (66.7)       | 1 (6.7)       | 0 (0)       | 1 (6.7)  | 15 (100) |
|            |  | II      | 0 (0)    | 1 (7.7)  | 6 (46.2)        | 4 (30.8)      | 1 (7.7)     | 1 (7.7)  | 13 (100) |
|            |  | III     | 0 (0)    | 0 (0)    | 10 (71.4)       | 3 (21.4)      | 1 (7.1)     | 0 (0)    | 14 (100) |
|            |  | IV      | 0 (0)    | 0 (0)    | 1 (33.3)        | 1 (33.3)      | 1 (33.3)    | 0 (0)    | 3 (100)  |
|            |  | V       | 0 (0)    | 0 (0)    | 3 (100)         | 0 (0)         | 0 (0)       | 0 (0)    | 3 (100)  |
|            |  | VI      | 0 (0)    | 0 (0)    | 12 (46.2)       | 6 (23.1)      | 3 (11.5)    | 5 (19.2) | 26 (100) |
|            | Total                                      |         | 2 (2.7)  | 2 (2.7)  | 42 (56.8)       | 15 (20.3)     | 6 (8.1)     | 7 (9.5)  | 74 (100) |
|            | Chi-square value = $25.961$ ; $P$          | = .410. |          |          |                 |               |             |          |          |

Spearman's correlation coefficient between preoperative and 1 yr postoperative facial nerve grades using in Schwannoma and Hemangioma are 0.331 and 0.504 respectively, and this was statistically significant at .01 level (2-tailed).

HB, House-Brackmann.

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|---------|------------------------|------------------------|-------------------------|-----------------|

|                       |               | Preoperative | FN function    |           |               | Postoperative | e FN function  |           |
|-----------------------|---------------|--------------|----------------|-----------|---------------|---------------|----------------|-----------|
|                       | HB grade I-II | HB grade III | HB grade IV-VI | Total     | HB grade I-II | HB grade III  | HB grade IV-VI | Total     |
| Intradural coaptation | 9             | 1            | 4              | 14        | 0             | 7             | 7              | 14        |
| Transdural coaptation | 5             | 5            | 14             | 24        | 0             | 12            | 12             | 24        |
| Extradural coaptation | 12            | 14           | 27             | 52        | 0             | 35            | 18             | 53        |
| Total                 | 26 (28.6%)    | 20 (21.9%)   | 45 (49.4%)     | 91 (100%) | 0 (0%)        | 54 (59.3%)    | 37 (40.7%)     | 91 (100%) |

HB, House-Brackmann; FN, facial nerve.

TABLE 5. Pre- and Postoperative Hearing Status in Hearing Preservation Surgeries at the End of 1 yr of Follow-up.

|                         | Preope          | rative hearing (dB); n | o (%)           | Postope         | erative hearing (dB); r | no (%)          |
|-------------------------|-----------------|------------------------|-----------------|-----------------|-------------------------|-----------------|
|                         | Mean PTA AC     | Mean PTA BC            | Mean ABG        | Mean PTA AC     | Mean PTA BC             | Mean ABG        |
| All patients (n = 115)  | 32.0 ± 26.5     | 21.0 ± 7.6             | 16.5 ± 14.1     | 37.6 ± 20.7     | 27.9 ± 14.7†            | 18.5 ± 14.7     |
| MCFA (n = 18)           | 25.9 ± 15       | 17.1 ± 7.1             | 8.8 ± 11.2      | $30.7 \pm 15.7$ | 27.1 ± 14.1             | $7.6 \pm 10.7$  |
| MCFA + TMA (n = 15)     | $27.3 \pm 18.6$ | $16.3 \pm 7.2$         | $11.0 \pm 15.4$ | $37.8 \pm 21.2$ | 27.0 ± 15.1             | $12.3 \pm 15.0$ |
| TMA ( $+$ TPA) (n = 21) | $34.7 \pm 22.0$ | 22.6 ± 11.6            | $12.1 \pm 16.3$ | $40.8 \pm 23.5$ | $28.5 \pm 15.5$         | $13.5 \pm 16.6$ |
| STP $(n = 5)$           | $28.0 \pm 16.0$ | $18.0 \pm 13.0$        | $10.0 \pm 17.0$ | 53.3 ± 25.2     | 32.5 ± 17.1             | $26.7 \pm 23.1$ |
| RSA $(n = 2)$           | $10.0 \pm 0.0$  | 20.0 ± 7.1             | $10.0 \pm 7.1$  | $25.0\pm0.0$    | $10.0 \pm 0.0$          | $15.0\pm0.0$    |

PTA, pure tone audiogram, AC, air conduction, BC, bone conduction, ABG: air bone gap, n, number.

We did an XII to VII or V to VII anastomosis in 4 cases. All of these patients had preoperative grade VI for greater than 1 yr. Postoperatively, the FN outcome was HB III, IV to VI in 1 and 3 cases, respectively. For long-standing FN paralysis, a gold weight implant was done in 5 cases, tarsorraphy in 4 cases, and a dynamic facial reanimation technique using temporalis muscle in 1 patient. The results of the different types of surgical nerve management have been compared in Table 2.

## **Hearing Outcomes**

Pre- and postoperatively, 17 (14.9%) and 53 (60.4%) ears were profoundly deaf respectively (Table 5). Of the 60 hearing preservation surgeries, the mean pre- and postoperative air bone gap were  $16.5 \pm 14.1$  and  $18.5 \pm 14.7$ , respectively.

## **Follow-up and Complications**

All patients were followed-up for at least 1 yr. Of the 14 cases with subtotal or near-total resections, 12 cases have shown no growth or slow growth (less than 3 mm/yr). Two cases showed growth of greater than 3 mm/yr after 2 yr of follow-up, and they were subjected to a second surgery wherein total tumor removal was achieved. SNIG was performed in both these cases but were not included in the analysis. There were no major complications in our series. There were no incidences of meningitis, intractable headaches, seizures, or cerebrospinal fluid leakages. The minor complications included abdominal hematoma (2 cases, 1.8%) and postauricular fistula with associated fat infection (2 cases, 1.8%). They were managed by additional local intervention. None of our patients underwent conventional or stereotactic radiotherapy.

#### **DISCUSSION**

Intrinsic tumors of the FN are a rare entity. While the global average ranged between 0.004% and 1.1% of ear cases, our series showed an incidence of 0.38% of all admissions in our center for otological conditions—2.7% of all skull base tumors over a 35-yr period. 8-10 In accordance with other studies, the mean age of our

patients also fell into the fourth decade and there was a slight male preponderance. <sup>11,12-18</sup>

## **Etiopathology**

FNTs are mostly benign. Schwannomas, followed by hemangiomas, <sup>19-24</sup> are the most common intrinsic tumors of the FN but a variety of other lesions have been reported in the literature including meningioma, <sup>25-28</sup> neurofibroma, <sup>29-32</sup> angioma, <sup>33-35</sup> paraganglioma, <sup>36-39</sup> chondromyxoid fibroma, <sup>40</sup> rhabdomyoma, <sup>41</sup> granular cell tumors, <sup>42,43</sup> and epineurial pseudocysts. <sup>44</sup> While a vast majority of FNTs appear spontaneously, there are studies that attribute the occurrence of such tumors following trauma, <sup>45</sup> surgery, <sup>46</sup> otomastoiditis, <sup>47</sup> and cholesteatoma. <sup>48</sup>

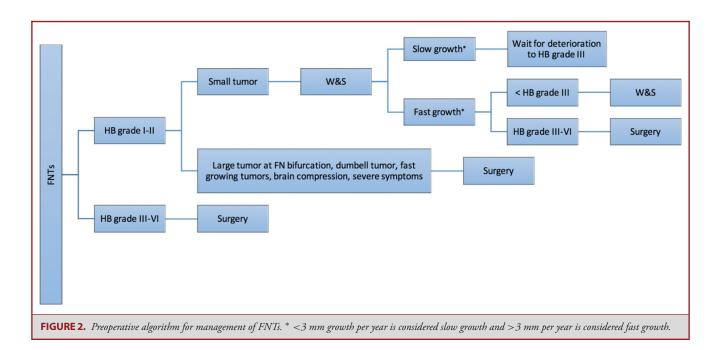
### **Clinical Features**

Because FNTs are commonly misdiagnosed as Bell's palsy by referring physicians, it is not surprising that multisite involvement indicative of a progressive tumor was found to be more common (60.5%) in our series. In addition to FN paralysis, tumors involving the vertical segment present with conductive hearing loss and a mass in the ear. Tumors involving the tympanic segment, GG, labyrinthine segment, and the IAC cause sensorineural hearing loss, tinnitus, and vertigo. Any mass that is visualized in these segments is usually retrotympanic and rarely do they present as transtympanic masses. Tumors involving the extratemporal FN present with a mass in the upper neck. Involvement of the greater superficial petrosal nerve (GSPN) may lead to unilateral lacrimal insufficiency.

## Radiology

A classic radiological description of FNTs cannot be described due to the convoluted course of the FN. On HRCT, schwannomas appear as local enlargement of the fallopian canal with well-defined margins, while hemangiomas show a honey-comb appearance with irregular borders and bony spicules due to intralesional calcification. On MRI, both schwannomas and

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hemangiomas show enhancement after contrast but the enhancements of hemangiomas are stronger and heterogenous due to calcifications.<sup>49</sup>

It must be borne in mind that normal individuals may show a mild or moderate enhancement in MRI around the anterior tympanic portion and GG due to the presence of a rich perivenous plexus surrounding the nerve in the fallopian canal and this is also the most common site of origin of FNTs. Also, enhancement of the distal IAC and labyrinthine segment of the FN is a common finding in Bell's palsy. <sup>50,51</sup> Enhancement in MRI with a widening of the fallopian canal is diagnostic of FNTs.

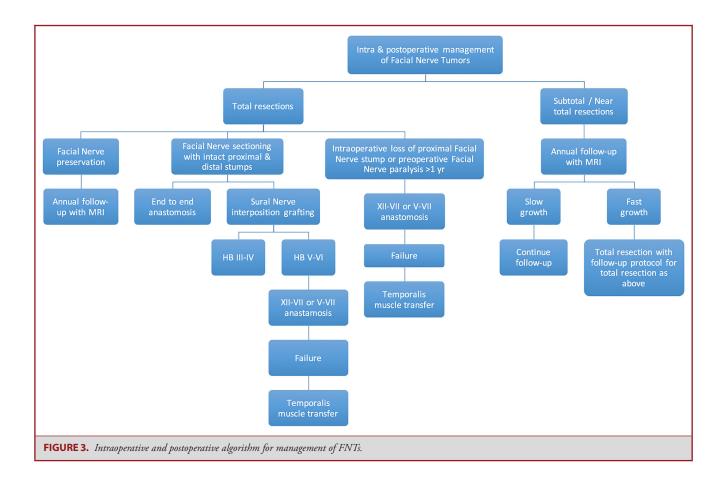
FNTs isolated to the IAC/CPA may pose a unique problem as they are often radiologically indistinguishable from a vestibular schwannoma. The presence of FN symptoms in such a case must make one suspect an FNT. A differentiating feature in radiology may be an enhancement along the labyrinthine segment (labyrinthine tail) that may sometimes be seen. 52-54 Rare but of interest are FNTs that are cystic. Eight (6.9%) of our tumors had cystic components.

## **Site of Tumor**

All parts of the nerve have been reported to be involved by tumors including the chorda tympani, <sup>55-57</sup> GSPN, <sup>58-60</sup> and the nervus intermedius. <sup>61-63</sup> Reports have increasingly supported the fact that the area around the GG is the most common site of involvement. <sup>12-15</sup>, <sup>18</sup>, <sup>64</sup>, <sup>65</sup> This area is flush with venous plexus and arterial supply from the petrosal artery. Further studies could be directed to determine any possible relationship between the vascularity and the origin of the tumor.

## **Decision Making in FNTs**

Surgery is the mainstay of treatment for FNTs but in most cases immediate postoperative FN paralysis of various degrees is almost certain. Hence, therapeutic options must be balanced and discussed well with the patient before taking a decision. Studies show that FN schwannomas grow at a rate of 1.4 mm/yr (range 0.7-2.6 mm/yr)<sup>64</sup> and hence wait-and-scan can be an effective policy in the management of these tumors. Since FNTs present in a younger age group (fourth decade of life) and they grow slowly, it is our policy to observe patients with an FN function of HB grade I and II, and opting for surgery only when the paralysis worsens to HB grade III and above (Figure 2). We recommend early surgical intervention regardless of the preoperative facial and hearing functions in the following cases: extratemporal FNTs extending into the bifurcation in the parotid, multisegment FNTs extending into both the CPA and the MCF, fast growing FNTs with worsening FN function, and large FNTs with temporal lobe compression. In this series, 23 (20%) patients with HB grade I and II were operated due to the abovementioned factors and also honoring the patients' choice. Surgery is done when the tumor is diagnosed near the bifurcation irrespective of the FN grading, because when the tumor involves the bifurcation, reconstruction requires splitting of the SN and 3 anastomoses: 1 to the main trunk and 1 each for the peripheral branches. This would lead to an unfavorable outcome.<sup>66</sup> However, one could argue that small tumors with good preoperative HB grades yield good postoperative outcomes due to 2 reasons. First, there is a very high possibility of intraoperative FN preservation. Secondly, even if a portion of the nerve was to be sacrificed, SNIG gives better results over smaller segments and due to better preoperative HB grades.



Stripping of tumors from the FN is an alternative to total resection in small tumors, especially hemangiomas. 17,67 This is possible because hemangiomas do not arise from the nerve itself but from the vascular plexus around the nerve. A small hemangioma usually does not infiltrate the nerve and can be easily peeled off it. However, we do not agree with the practice of stripping of the tumor from the nerve when it is frankly infiltrated or in large tumors. Another practice is subtotal resection with decompression of the fallopian canal. In both FN sparing surgeries and decompression, <sup>16</sup>, <sup>17</sup>, <sup>68</sup>-<sup>71</sup> authors advocate early resection of tumors in an attempt to save FN function. However, a study by McRackan et al. 16 has shown that subtotal resections and FN preservation surgeries are not associated with superior FN function that is statistically significant. Furthermore, since the tumor is not completely removed, there is a high chance of recurrence. Li et al.<sup>72</sup> reported a recurrence rate of 10% of cases with 95% of resection and 60% of cases with a 70% to 80% resection.

When the decision is taken to operate, we go for total tumor clearance or preplanned near/subtotal resection. The intraoperative and postoperative algorithm is depicted in Figure 3. In CPA tumors like vestibular schwannomas, the FN function is

rarely disrupted (despite stretching) even in large tumors. Hence, the facial muscle function is not affected in such cases and performing a XII to VII or V to VII anastomosis when the proximal stump is unavailable after tumor removal would yield a good result. However, this is not the case in FNTs because there is an invariable paralysis in large FNTs. Also, in any FN paralysis of more than 1 yr the facial muscles would have begun to atrophy. The response to reinnervation after a year of paralysis diminishes and becomes unpredictable. In cases of incomplete denervation, the facial muscle can remain viable for longer periods and respond to further reinnervation techniques.<sup>73</sup> In FN paralysis of greater than 1 yr, if an SNIG is done, it would take an additional 6 mo to 1 yr for the proximal axonal stump to grow through the grafted nerve (that acts as a conduit) to reach the distal neuromuscular junction by which time the muscles would have atrophied further yielding un orable results. However, if a XII to VII or a V to VII anastomosis is done in such cases, the distance and the time taken for the nerve to regenerate is shorter and hence innervation of the residual functioning muscles is better. Hence, this must be the first choice of reconstruction. If this fails, then a dynamic facial reanimation technique using temporalis muscle (Labbe's surgery)

| TABLE 6. F                                   | eview of Li | TABLE 6. Review of Literature and Comparison | d Comparis            | on of Outcomes of Important Series From 1995 to 2017   | mportant Serie   | s From 19 | 995 to 20        | 17       |                 |                                      |          |           |            |                   |           |   |
|--|-------------|--|-----------------------|--|--|-----------|------------------|----------|-----------------|--------------------------------------|----------|-----------|------------|-------------------|-----------|---|
|  |             | Most   |                       |  |  |           |                  |          | FN functi       | FN function in operated patients (%) | rated pa | tients (% |            |                   |           |   |
|  |             | common<br>site of                            | Treatment             | Surgical<br>approach   | FN<br>management   | 뚝         | HB1&II           |          | HBIII           | HB IV                                | 2        | Ī         | HB V       | Ξ                 | HB VI     |   |
| Study  | Pathology   | tumor (%)                                    | options               | (%)  | (%)  | Preop     | Postop           | Preop    | Postop          | Preop                                | Postop   | Preop     | Postop     | Preop             | Postop    | Complications   |
| Liu et al <sup>87</sup> , $n = 22$           | S           | IAC (82)                                     | Sx, W&S               | TLA (59), RSA (27), TMA (9)  | NG (46),<br>VII-XII (9)                                  | 3 (25)    | (0) 0            | 0 (0)    | 3 (25)          | 3 (25)                               | 5 (42)   | 1(1)      | 0 (0)      | 5 (42)            | 2 (17)    | NA  |
| Perez et al <sup>64</sup> , $n = 24$         | S           | TS (33.3)                                    | SX                    | CA (25.0), TMA (16.7),<br>TLA (4.2)  | NG (12.5),<br>VII-XII (4.2)                              | 3 (12.5)  | 3 (12.5)         | 2 (8.3)  | 6 (25)          | 2 (8.3)                              | 1 (4.2)  | 1 (4.2)   | (0) 0      | 3 (12.5) 1 (4.2)  | 1(42)     | NA  |
| Shirazi et al $^{15}$ , $n = 16$             | v           | GG (69)                                      | Sx, D                 | TLA (25), TMA (25),<br>TMA + MCFA (18.8),<br>TMA + TPA (18.8), TPA<br>(6.3), ST + ELA (6.3)              | NG (81.3),<br>VII-XII (6.3)                              | 3 (18.8)  | 1 (6.3)          | 3 (18.8) | 12 (75)         | 5 (31.3)                             | 2 (12.5) | 5 (31.3)  | (0) 0      | 0)0               | (0) 0     | NA  |
| Liu et al <sup>88</sup> , $n = 22$           | S, N, H     | <b>∀</b><br>Z                                | Sx                    | TMA (54.5),<br>MCFA + TMA (22.7),<br>MCFA (13.6), RSA (9.0)  | NG (45.4)  | 4 (18.2)  | 8 (36.4)         | 4 (18.2) | 7 (31.8)        | 4 (182)                              | 2 (9.1)  | 6 (27.3)  | (0) 0      | 4 (18.2) 5 (22.7) | 5 (22.7)  | NA  |
| McMonagle<br>et al <sup>14</sup> ,<br>n = 53 | v           | GG (45.3)                                    | Sx, W&S               | TLA (32), RSA (113),<br>TMA + TPA (5.7),<br>TMA + MCFA (5.7),<br>TMA + MCFA (3.8), TMA (1.9), MCFA (1.9) | NR (41.7),<br>NG (36.1),<br>VII-XII (16.7),<br>EEA (5.6) | 35 (66)   | 23 (43.4) 9 (17) | (71) 6   | 17 (32.1) 0 (0) |                                      | 6 (11.3) | 2 (3.8)   | (0) 0      | 6 (11.3)          | 4 (7.6)   | Wound infection  & CSF leak (2.78), vertigo (2.78), headache (2.78), EAC stenosis (2.78), tongue atrophy (2.78) |
| Wilkinson et al <sup>69</sup> , $n = 79$     | s           | GG (65.8)                                    | Sx, D,<br>W&S,<br>GKS | NA   | NA   | (63.3)    | N<br>A           | (16.5)   | NA              | (6.3)                                | Y<br>Y   | (10.1)    | NA         | (3.8)             | NA        | NA  |
| McRackan et al $^{16}$ , $n = 56$            | S           | IAC (81.8)                                   | Sx W&S,<br>GKS        | TLA (54.7), TMA (11.3),<br>TMA + MCFA (11.3),<br>MCFA (9.4), RSA (9.4),<br>TLA + MCFA (3.8)              | NG (17.8),<br>EEA (5.3)                                  | 37 (58.9) | (0) 0            | 7 (12.5) | (0) 0           | 4 (7.1)                              | (0) 0    | 2 (3.6)   | 29 (51.8)* | 6 (10.7)          | 27 (482)* | N A   |
| Lee and<br>Kim <sup>n</sup> , n = 25         | S           | MS (40)                                      | Sx, D                 | TMA (40.0), MCFA (16.0),<br>TLA (16.0), MCFA + TMA<br>(12.0), TMA + TPA (8.0),<br>TPA (8.0)              | NG (28),<br>EEA (4)                                      | 16 (64)   | 14 (56)          | 2 (8)    | 2 (8)           | 1 (4.0)                              | 5 (20)   | 4 (16.0)  | 1(4)       | 2 (8)             | 3 (12)    |   |

| TABLE 6.                              | TABLE 6. Continued |                    |           |   |   |           |                                      |          |           |              |                                      |                  |          |                    |          |                           |
|---------------------------------------|--------------------|--------------------|-----------|---|---|-----------|--------------------------------------|----------|-----------|--------------|--------------------------------------|------------------|----------|--------------------|----------|---------------------------|
|                                       |                    | Most               |           |   |   |           |                                      | "        | N functio | n in oper    | FN function in operated patients (%) | ints (%)         |          |                    |          |                           |
|                                       |                    | common             | Treatment | Surgical<br>approach  | FN  | HB        | HBI&II                               | HB       | =         | HB IV        | 2                                    | HB V             | >        | HB VI              |          |                           |
| Study                                 | Pathology          | tumor (%)          | options   | (%)   | (%)   | Preop     | Postop                               | Preop    | Postop    | Preop Postop | Postop                               | Preop            | Postop   | Preop              | Postop   | Complications             |
| Mowry et al <sup>61</sup> , $n = 16$  | S                  | IAC + CPA<br>(100) | ×         | MCFA (50), TLA (43.8)   | NG (12.5)   | 16 (100)  | 13 (81.3)                            | (0) 0    | 1(63)     | (0) 0        | (0) 0                                | (0) 0            | (0) 0    | (0) 0              | (0) 0    | ı                         |
| Wang et al <sup>89</sup> ,<br>n = 16  | I                  | GG (100)           | š         | MCFA (87.5),<br>MCFA + TMA (6.3), ELA<br>(6.3)  | NG (56.3)   | (0) 0     | 3 (18.8)                             | 2 (12.5) | 8 (50)    | 4 (25)       | 2 (12.5)                             | 4 (25)           | 3 (18.8) | 6 (37.5)           | (0) 0    | Ψ.                        |
| Park et al <sup>17</sup> ,<br>n = 28  | v                  | TS (60.7)          | ×         | TMA (50), TLA (14.3),<br>MCFA (10.7),<br>MCFA + TMA (10.7),<br>TMA + TPA (3.6),<br>TPA (3.6)  | NG (21.4),<br>VII-XII<br>(7.1),<br>EEA (3.6)              | 17 (60.7) | 16 (57.1)                            | 4 (14.3) | 1 (3.6)   | 2 (7.1)      | 8 (28.6)                             | 3 (10.7) 1 (3.6) |          | 2 (7.1)            | 2 (7.1)  | N A                       |
| Li et al <sup>72</sup> , $n = 15$     | S                  | TS (53.3)          | Sx        | TMA (66.6), MCFA (33.3)****   | NA  | 10 (66.6) | 10 (66.6) 14 (93.3) 4 (26.7) 1 (6.7) | 4 (26.7) | 1 (6.7)   | (0) 0        | 0 (0)                                | (0) 0            | (0) 0    | (0) 0              | (0) 0    | NA                        |
| Watson et al <sup>90</sup> , $n = 15$ | S, H, M            | GG (40)            | Sx, D     | MCFA (20), TMA (20),<br>TLA (6.7), CA (20)  | NG (46.7)   | 4 (26.7)  | 2 (13.3)                             | 7 (46.7) | 5 (33.3)  | (0) 0        | 2 (13.3)                             | 1 (6.7)          | 1 (6.7)  | 3 (20)             | 2 (13.3) | ۷<br>۷                    |
| Present<br>study n = 115              | S, H, M, N         | GG (53.9)          | ×         | FPS: 3 (2.6), MCFA 18<br>(15.6), MCFA + TMA 15<br>(13), TMA 14 (12.1),<br>TMA + TPA 7 (6), STP 6<br>(5.2), RSA 2 (1.7) TLA 25<br>(21.7), TCA 16 (13.9), TOA | NG 91 (79.1),<br>EEA 3 (2.6),<br>VII-XII/VII-V<br>4 (3.4) | 40 (34.8) | 12 (10.4) 22 (19.1)                  |          | 61 (53)   | 6 (5.2)      | 21 (18.3)                            | 6 (52)           | 11 (9.6) | 41 (35.7) 10 (8.7) |          | Wound infection<br>(1.8%) |

n, number of cases; 5, schwannoma; H, hemangioma; M, meningioma; NA, neurofibroma; IAC, internal acoustic canal; CPA, cerebellopontine angle; GG, geniculate ganglion; TS, tympanic segment; MS, mastoid segment; MS, wait-and-scan; GK, gamma knife radiosurgery; TLA, translabyrinthine approach; MCFA, middle cranial fossa approach; RSA, retrosigmoid approach; TOA, transparotid approach; TTA, translabyrinthine approach; ELA, extralabyrinthine approach; CA, combined approach; CRA, cervical approach; EEA, end-to-end anastomosis; NG, nerve graft; VII-XII, facial hypoglossal anastomosis; VII-V, facial trigeminal (masseteric) anastomosis; PS, fascicle; NA, not available. '12 mo after surgery.

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We do not recommend any form of radiation in these patients as they present in a young age group.

#### **Sural Nerve Graft Interpositioning**

Cable nerve graft interpositioning, using either the SN or the great auricular nerve, is a standard procedure for interruptions of the FN in the skull base. Recent studies have shown that a cable nerve graft interpositioning provides as good a result as a primary end-to-end coaptation.<sup>74-77</sup> However, the best possible postoperative outcome is HB grade III regardless of the graft material used or the technique employed, because the frontal muscle function rarely recovers and a certain degree of synkinesis is unavoidable after grafting.<sup>76,78,79</sup>

The cable nerve graft acts as a nerve conduit with empty endoneurial tubes, a reserve of viable Schwann cells and nerve growth factors through which the regenerating axons can be directed. One of the most key factors that determines the ultimate success of any reinnervation procedure is the duration of facial paralysis. Facial muscles completely denervated for less than 1 yr respond to nerve grafting. The response to reinnervation after a year of paralysis diminishes significantly and becomes unpredictable. There are other factors that play a role in the success of a graft like loss of blood supply to the nerve, insult to the nerve due to the tumor or the inflammation. Studies have suggested that the more proximal the site of transection of nerve to the cell body, the more intense is the damage to the cell body located within the central nervous system and hence lesser chance of survival of the nerve. 77,80,81

Delicate maneuvers of nerve approximation like perineurial, group funicular, and epineurial suturing are very difficult to perform in the depths of a pulsating CPA and even more so in the setting of a tired surgeon after a long and frequently laborious procedure. The Furthermore, the FN lacks a true fascicular organization till the GG<sup>82,15,18,83</sup> and this makes it practically impossible to perform any kind of epi- or perineurial suturing till the GG. Hence, we prefer to use the stitchless fibrin glue for coaptation in intradural and transdural coaptation. The success of this technique has been researched and replicated by other authors. Suturing is reserved only for extradural coaptation.

In our analysis, postoperative FN grades were better (HB grade III) with extradural coaptation (66%) compared to 50% each in intradural and transdural respectively. We also found that there was an overall improvement in FN function in extradural and transdural coaptation, but an overall worsening of FN results after an intradural coaptation.

## **Review of Literature**

All case series in the literature with more than 15 cases that reported on surgical intervention of intrinsic tumors of the FN from January 1995 to March 2017 were reviewed.

With 115 cases, ours was the largest series of FNTs (Table 6). Six (42.8%) series reported that the GG was the most common site of involvement. Thirteen (92.9%) of the series reported

using lateral skull base procedures for tumor removal. Intraoperative FN grafting was employed in 12 (85.7%) series. Seven (50%) of the authors preferred to intervene in cases where there was an advanced FN paralysis ( $\geq$ HB grade III). Postoperatively, 9 (64.3%) of the authors managed to achieve  $\geq$ HB grade III in >60% of their cases. The results of this study were consistent with the literature, with 63.5% cases achieving a postoperative  $\geq$ HB grade III.

#### CONCLUSION

FNTs are a difficult pathology, and a thorough knowledge of skull base surgery is essential to effectively deal with such tumors. Since FNTs present in a younger age group and they grow slowly, it is rational to observe patients with an FN function up to HB grade II, and opt for surgery only when the paralysis worsens or when the symptoms are severe. When the decision is taken to operate, a gross-total resection is desirable with SNIG in cases of recent onset paralysis.

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## **COMMENT**

e congratulate the authors for their tremendous work collecting this largest series of relatively infrequent tumors (115 patients in 26 years, a mean of 4.4 patients per year in a quaternary referral center). We also strongly adhere with them that the "wait and scan" approach is a very good option in patients with limited facial nerve affection (House & Brackmann 1 or 2), mainly because after the tumor resection surgery the risk of severe facial nerve palsy is almost secured. In the best case, an interposed sural nerve graft will lead to a House & Brackmann 2 or generally 3, with severe dyskinesias and poor reinervation of the frontal muscles.

Having said that, it is important to point out that in the first place on the podium for facial nerve reconstructive surgery remains the facial to facial (with or without interposed nerve graft) stump coaptation, as used by the authors. It is also important to point out that the use of fibrin glue to perform the nerve coaptation is a good technique for avoiding a difficult suture in a very deep surgical field. Even though we usually use nerve sutures except in the aforementioned settings and also in obstetric brachial plexus surgery, it has been reported that the results of fibrin glue are similar to those achieved by a direct nerve suture, even in unexperienced surgeons.1

Finally, we would like to highlight that the direct hypoglossal to facial or masseter to facial nerve transfer, both give acceptable results (House & Brackmann 3) when a proximal facial nerve stump is not available for reconstruction, occupying the second place on the podium. Nevertheless, is has been largely demonstrated that these techniques give the best results when performed within one year of palsy, when compared to later attempts.<sup>2</sup> In that sense, an early and aggressive surgical facial nerve reconstruction -whenever possible- will derive in a better outcome for the patient.

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<sup>1.</sup> Whitlock EL, Kasukurthi R, Yan Y, Tung TH, Mackinnon SE. Fibrin glue mitigates the learning curve of microneurosurgical repair. Microsurgery. 2010;30: 218-222.

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