The Anatomically Intact but Electrically Unresponsive Facial Nerve in Vestibular Schwannoma Surgery

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BACKGROUND: Permanent facial nerve (FN) paresis after vestibular schwannoma surgery is distressing to both the patient and surgeon. Intraoperative electrophysiological testing has proven invaluable in reducing the incidence of FN injury and may assist in prognosticating long-term function.

OBJECTIVE: To report definitive FN outcomes among a cohort of patients with an unevokable but anatomically intact seventh nerve after microsurgical vestibular schwannoma resection.

METHODS: All patients undergoing vestibular schwannoma surgery between 2000 and 2010 at a single tertiary academic referral center were identified. Intraoperative FN monitoring data and definitive FN outcomes were reviewed, and all patients with an anatomically intact but electrically unresponsive FN were included.

RESULTS: Eleven patients met the inclusion criteria. The median preoperative and definitive postoperative FN scores were House-Brackmann grades 1 and 3, respectively. The median time to definitive FN recovery was 9.4 months.

CONCLUSION: These data demonstrate that even among this extreme subset, modern electroprognostic testing strategies are incapable of reliably predicting poor outcomes. Therefore, if FN continuity is maintained, attempts at same-surgery FN repair should not be pursued.

KEY WORDS: Acoustic neuroma, Electrophysiological testing, Facial nerve, Vestibular schwannoma

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At the turn of the 20th century, vestibular schwannoma (VS) resection was associated with a high rate of mortality, and surgical intervention was considered a last resort in patients with advanced disease. However, over the last 80 years, synchronous advancements in technology and surgical technique have radically reduced surgical complications. With disease-specific mortality approaching zero, our focus has shifted toward minimizing morbidity and preserving neurological function.[1]

After the introduction of the operating microscope, surgeons were able to preserve the anatomic continuity of the facial nerve (FN) in > 90% of cases; however, only approximately 50% of patients demonstrated normal definitive FN function.[2] In 1979, Delgado and colleagues[3] introduced intraoperative evoked electromyographic (EMG) monitoring, and since that time there has been a substantial improvement in FN outcomes among high-volume VS centers, with > 80% of patients achieving satisfactory long-term function.[4,5] Intraoperative FN monitoring confers several notable benefits with respect to minimizing injury and optimizing long-term FN function: (1) The FN can be accurately identified early during surgery, often before tumor dissection; (2) the course of the FN can be mapped through both tumor and bone; and (3) intraoperative feedback resulting from mechanical and thermal evoked EMG responses may alert the surgeon to unintentional trauma so that the surgical strategy can be modified to minimize neural injury.[6]

Despite these advances, a small group of patients experience disappointing long-term FN outcomes. The large number of studies evaluating

ABBREVIATIONS: EMG, electromyographic; **FN,** facial nerve; **HBG,** House-Brackmann grade; **VS,** vestibular schwannoma

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intraoperative electroprognostic parameters serve as a testament to the vexing nature of the problem.[5,7-9] To the best of our knowledge, no study to date has specifically evaluated outcomes among patients with an anatomically intact but electrically unevokable FN. Here, we report the clinical course and FN outcomes of 11 such patients who were managed at our institution over the last decade to determine whether same-surgery FN repair such as interposition nerve grafting or facial-hypoglossal anastomosis should be considered in this unique subset.

METHODS

Patient Population

A prospectively maintained clinical database including all patients who underwent microsurgical resection of VS between 2000 and 2010 at a tertiary academic referral center was reviewed. Patients with an anatomically intact but electrically unresponsive FN after tumor dissection, as determined by proximal stimulation at the nerve exit zone, were identified. These data were further supplemented via retrospective chart review using the institutional electronic medical record. Initial data collection included neurofibromatosis type 2 status; history of prior tumor treatment; surgical approach; completeness of resection as determined by the intraoperative impression of the neurosurgeon-neurotologist team; intraoperative appearance and course of the FN; intraoperative FN EMG data; preoperative, immediate postoperative, and definitive postoperative FN function; and basic demographic information. Preoperative tumor size and location were reported according to the 1995 American Academy of Otolaryngology–Head and Neck Surgery guidelines.[10] FN function was appraised by use of the House-Brackmann grading (HBG) scale.[11] Gross total resection was designated when no tumor was knowingly left behind; near-total resection was applied when a tumor remnant smaller than 5 × 5 × 2 mm was intentionally left in situ to lessen neurological morbidity; and subtotal removal was used when anything more was left behind.[12] Patients with electrically responsive proximal FN segments, those with anatomic discontinuity of the FN after VS resection, and subjects with < 10 months of postoperative follow-up were excluded. Institutional Review Board approval was obtained before the study was started (No. 10-006841).

Intraoperative Electrophysiological Testing Protocol

After administration of a short-acting anesthetic induction agent to facilitate intubation, paralytics were strictly avoided throughout the operation. The intraoperative cranial nerve monitoring protocol involved placement of subdermal electroencephalography electrodes providing 4-channel continuous EMG monitoring. Data acquisition used a Viking Select EMG system (Nicolet Biomedical, San Diego, California). Low- and high-frequency filters were set to 30 Hz and 10 kHz, respectively. Responses were conveyed on a digital oscilloscope and by a loudspeaker projected across the operating room to the surgeon.

At the conclusion of tumor resection, the FN was stimulated near the root exit zone at the pontomedullary junction of the brainstem with an insulated flush-tip Prass monopolar stimulator probe (Medtronic, Minneapolis, Minnesota). Cerebrospinal fluid and blood were removed from the stimulation site before measurements were taken to minimize current leak. Current was delivered using a pulse duration of 50 microseconds and pulse frequency of 1 per second. Beginning at 0.1 mA, current was increased by 0.1-mA increments until a reliable compound muscle action potential was obtained or 5 mA was reached. The lowest amperage at which a reliable EMG response could be elicited was considered the minimum stimulation threshold. Electrical stimulation of the FN at a segment distal to tumor dissection was used as a positive control to ensure that proximal silence was not due to technical monitoring error. In cases when the tumor affected the fundus and stimulation of an unadulterated intrameatal nerve segment could not be performed, transcutaneous bipolar electrodes placed near the extratemporal main trunk in the region of the stylomastoid foramen FN were used to obtain a distal response, recorded from a fifth subdermal electroencephalography electrode placed in the nasalis muscle. Therefore, an absent proximal response was designated when low-threshold distal stimulation could be achieved but proximal stimulation of 5 mA was unyielding.

RESULTS

Over the last decade, 350 patients underwent microsurgery for VS at the our institution; of these, 11 patients (7 women, 4 men) met study criteria. The median age at VS resection was 53.5 years (mean, 49.1 years; range, 18.5-66.3 years), and the median duration of follow-up was 81.8 months (mean, 73.7 months; range, 10.1-143.7 months; Table 1). Four patients were excluded secondary to FN transection; 28 had inadequate EMG data available for review; and the remainder of excluded subjects had detectable EMG responses during proximal FN stimulation at the conclusion of tumor dissection.

All 11 patients presented with unilateral sporadic VS. One patient was evaluated for recurrent tumor after microsurgical resection 9 years earlier at an outside facility; after her first operation, she experienced immediate complete FN paralysis that recovered to an HBG 3. Before presenting to our institution, she progressed to an HBG 5 after rapid growth of a 3.5-cm VS macrocyst. The remaining 10 patients presented for primary evaluation and treatment. The most common presenting symptom was hearing loss (9, 81.8%); 2 patients had preoperative hydrocephalus; and aside from the patient presenting after tumor recurrence, all patients denied preoperative FN symptomatology and had normal motor function on examination.

Ten tumors involved the cerebellopontine angle, whereas 1 was purely intracanalicular. The median cisternal diameter was 2.5 cm (mean, 2.7 cm; range, 1.0-4.5 cm); 3 tumors demonstrated macrocystic features. The median distance between the radiographic lateral tumor margin and the fundus (fundal cap) was 3 mm (mean, 3 mm; range, 0-9 mm).

Nine patients underwent a retrosigmoid craniotomy, and 2 received a translabyrinthine approach. The FN most commonly followed a ventral course. The tumor was exceptionally vascular, the tumor-nerve interface was adherent, or the FN was severely attenuated (splayed) in all cases. Six of the 11 patients were treated during the first 3 years of the senior author's experience, whereas only 2 subjects were managed within the last 3 years. In all 11 cases, anatomic continuity of the FN was preserved; however, proximal FN stimulation at the brainstem did not elicit a compound muscle action potential, whereas distal stimulation, lateral to tumor dissection, provided a reliable response at low current levels (Table 2).

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TABLE 1. Preoperative Summary of 11 Study Patients[a]

Patient	Age, y	Sex	Side	Preoperative Symptoms	Preoperative Hearing[b]	Prior Treatment
1	45.2	M	R	HL, T	NR	No
2	66.3	M	R	HL, Hyd, V	D	No
3	65.4	F	L	FN, HL, TN	D	MS
4	18.5	F	L	Hyd	A	No
5	58.3	F	L	HL	C	No
6	51.2	F	R	TN, V	B	No
7	59.8	M	R	HL, TN, V	B	No
8	28.3	F	R	HL, HA, V	B	No
9	53.5	F	L	HL, TN	D	No
10	30.9	M	L	HL, T	D	No
11	63.2	F	L	HL, T	B	No
Summary[c]	53.5	64% F	55% L	81.8% HL	50% A or B	90.9%

[a]FN, facial nerve weakness; HL, hearing loss; Hyd, hydrocephalus; MS, microsurgical resection; T, tinnitus; TN, trigeminal neuropathy; V, vertigo.
[b]American Academy of Otolaryngology–Head and Neck Surgery hearing class reporting guidelines.
[c]Summary presented with most common feature (%) or median value.

All subjects experienced immediate complete postoperative FN paralysis (HBG 6). The median definitive HBG after surgery was 3 (mean, 3.5; range, 2-6), requiring a median time to recovery of 9.4 months (mean, 11.5 months; range, 2.2-24.0 months). Six patients underwent adjuvant procedures, including upper-lid gold weight placement, lateral tarsorrhaphy, and/or temporalis transfer; none of the patients received FN reinnervation surgery (Table 3).

DISCUSSION

Since the dawn of the modern skull base era, much attention has been given to identifying factors that may predict postoperative long-term FN function. Preoperative variables, including large tumor size, cystic features, and history of prior treatment, have been associated with an increased risk of poor definitive FN function and may guide preoperative risk disclosure.[13-17] Intraoperative factors such as degree of tumor adhesion, tumor vascularity, FN "splay," and the microscopic appearance of the FN at the conclusion of tumor dissection may provide a suggestion of long-term function.[5,13,14] Historically, some authors have recommended that if the FN appears intact but injured at the conclusion of tumor dissection, then the damaged segment can be resected and the healthy ends coapted in an attempt to improve long-term function.[18] However most contend that none of the aforementioned factors reliably predicts poor long-term function and that, if the FN remains

TABLE 2. Tumor Characteristics and Operative Detail[a]

Patient	Approach	Extent of Resection	Size,[b] cm	Brainstem Contact	Distance to Fundus, mm	Macrocystic	Course of FN	Notable Intraoperative Findings
1	RS	GTR	IAC	No	0	No	A	Vascular tumor, adherent to FN
2	RS	GTR	4.0	Yes	6	No	A	Vascular, adherent to FN, Splayed FN
3	RS	GTR	2.0	Yes	3	Yes	AS	Adherent to FN
4	RS	GTR	4.5	Yes	0	No	AI	Adherent to FN, splayed FN
5	RS	GTR	2.0	Yes	NR	Yes	AI	Adherent to FN
6	RS	NTR	2.5	Yes	6	Yes	A	Adherent, splayed FN
7	TL	GTR	3.0	Yes	3	No	NR	Vascular tumor, adherent to FN
8	TL	NTR	3.0	Yes	0	No	A	Adherent to FN
9	RS	GTR	2.5	Yes	0	No	AS	Vascular, adherent to FN, splayed FN
10	RS	GTR	2.0	Yes	3	No	A	Adherent to FN, splayed FN
11	RS	NTR	1.0	Yes	9	No	NR	Adherent to FN
Summary[c]	82% RS	73% GTR	2.5	90.9% Yes	3	27.3%	55.5% A	

[a]A, anterior; AI, anterior inferior; AS, anterior superior; FN, facial nerve; GTR, gross total resection; IAC, internal auditory canal; NR, not recorded; NTR, near-total resection; RS, retrosigmoid; STR, subtotal resection; TL, translabyrinthine.
[b]American Academy of Otolaryngology–Head and Neck Surgery reporting guidelines.
[c]Summary presented with most common feature (%) or median value.

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TABLE 3. Facial Nerve Outcomes[a]

Patient	Preoperative HBG	Immediate Postoperative HBG	Definitive Postoperative HBG	Time to Definitive HBG, mo	Total Duration of Follow-up, mo	Adjuvant Procedures
1	1	6	2	2.2	143.7	None
2	1	6	5	3.3	81.8	ULGW, temporalis transfer
3	5	6	2	20.0	59.2	None
4	1	6	4	19.4	91.4	ULGW
5	1	6	3	24.0	84.9	Lateral tarsal strip, ULGW
6	1	6	3	9.5	34.1	ULGW (temporary)
7	1	6	3	7.1	114.4	Lateral tarsal strip, ULGW
8	1	6	3	9.2	53.3	None
9	1	6	4	9.1	10.1	None
10	1	6	3	11.2	20.1	None
11	1	6	6	NA	117.6	ULGW, temporalis transfer
Median	1	6	3	9.4	81.8	45.5% None

[a]HBG, House-Brackmann grade; ULGW, upper-lid gold weight.

intact, any attempt at reinnervation should be delayed for at least 1 year after surgery, even if immediate complete postoperative paralysis ensues.[2,6,15]

The advent of intraoperative evoked EMG provided a new tool for FN outcome prediction, and within the last 20 years, a large number of publications have evaluated optimal cut points designed to best separate patients who are at low and high risk for unfavorable FN outcomes. The 3 common parameters evaluated with direct electrical neural stimulation include stimulation threshold, proximal-to-distal amplitude ratio, and absolute amplitude of the evoked compound muscle action potential. More recently, continuous intraoperative EMG monitoring, specifically A-train activity, has been correlated with early and late postoperative FN outcomes.[19] Results from these studies demonstrate that intraoperative neural testing is generally capable of determining which patients will have satisfactory outcomes, but it is often poor at predicting who will have unsatisfactory long-term function.[5,8,19] All of the previously mentioned intraoperative testing parameters analyze EMG responses. These responses can occur only if at minimum 1 functional axon capable of propagating an action potential to a functioning neuromuscular junction persists. In the present study, we evaluated the most extreme subset of patients with regard to FN testing, understanding that if these subjects did not consistently have poor long-term FN function, then patients with more favorable neurophysiologic test results should conceivably fare better.

The finding of an anatomically intact but electrically unresponsive FN presents the surgeon with a unique and uncommon quandary. Should the compromised nerve segment be left in situ with the hope that the patient will ultimately have a satisfactory long-term outcome? Or should the injured segment, as determined by "electrical mapping" and microscopic inspection, be sacrificed with the anticipation that neurorrhaphy may provide a superior outcome? To support early reinnervation surgery, prognostic testing must be capable of accurately predicting which patients are destined for a definitive HBG 4 or worse because the very best results that can be achieved with FN interposition grafting or facial-hypoglossal nerve anastomosis is HBG 3 function.[2,20]

If intraoperative testing could predict with high certainty that a patient would have poor definitive FN function, then same-surgery FN repair could be considered. This strategy is appealing for several reasons: (1) The patient would be exposed to only a single anesthetic and a single surgery; (2) the surgeon could map the neural segment with severe conduction block, allowing a tailored resection and coaptation; (3) same-surgery repair provides the best opportunity to find the proximal FN stump and eludes scarring and neuroma formation; (4) ipsilateral reinnervation provides the best outcome on the reconstruction ladder with minimal donor site morbidity; (5) the time to definitive function would be much less than if a "wait and see" interval were observed; and (6) early reinnervation would theoretically minimize denervation muscle atrophy and loss of motor end plates.[21]

We found that of 11 patients with anatomically intact but electrically unevokable FNs, all demonstrated immediate HBG 6 paralysis; however, 7 (64%) achieved HBG 3 or better definitive function, with 2 subjects attaining an HBG 2. In addition, 80% of patients achieved definitive FN function in $<$ 1 year from surgery, a much shorter time than commonly seen with primary coaptation, interposition neurorrhaphy, or donor nerve techniques.[21] Similarly, Lalwani and colleagues[5] reported 2 patients with absent EMG responses during proximal high-level

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FN stimulation. Both patients experienced immediate complete paralysis but recovered to an HBG 2 and 3 at the last follow-up.

Neurotonic discharges, characterized by asynchronous trains of motor unit potentials, have been associated with impending or ongoing neural damage. In 2007 and subsequently in 2010, Prell and colleagues[19,22] demonstrated the prognostic importance of "train time," a quantifiable value derived from A-train activity. In the former study, they found that a train time of $>$ 10 seconds was statistically significantly associated with an increased risk for long-term FN paresis. Of the 13 patients with $>$ 10-second train time, 4 acquired HBG 4 or worse long-term function; however, it should be noted that 5 of the 13 returned to HBG 1 function. In the 2010 study, they demonstrated that among 30 patients undergoing VS microsurgery, a train time of $>$ 2.5 seconds correlated with poorer long-term FN function compared with a train time $<$ 2.5 seconds. Specifically, 94% (15 of 16) of subjects with a train time of $<$ 2.5 seconds had HBG 1 to 2 function at the last follow-up. Unfortunately, a train time of even $>$ 10 seconds was associated with a high false-positive rate when long-term FN function was prognosticated because 60% (3 of 5) of such patients recovered HBG 2 function.

In 2002, Nakao et al[23] correlated spontaneous and mechanically evoked EMG patterns during the last steps of VS resection with long-term FN function. They found that among 49 patients, 3 demonstrated a "stray" pattern characterized by persistent train responses up to 20 minutes in duration, and 11 subjects demonstrated a "silent" pattern with little or no EMG activity. All patients with stray activity and 73% (8 of 11) of those with a silent pattern returned to HBG 3 or better function. Similar to direct electrical neural stimulation testing, these studies demonstrated that continuous intraoperative EMG analysis appears to excel at identifying which patients will have satisfactory long-term FN outcomes but cannot consistently predict who will have poor long-term function.

These data support the concept that if the anatomic continuity of the FN can be maintained, it should be left intact regardless of unfavorable electrophysiological test results. In theory, the present study tests the limits of modern electrodiagnostic testing capability. Our results demonstrate that contemporary direct electrical neural stimulation testing strategies are incapable of reliably determining who will have poor long-term FN function (HBG 4-6) and that alternative methods are needed if we are to answer this important but difficult question. These findings should not be surprising, however, because proximal "electrical silence" only infers that all tested axons have a complete conduction block and does not provide any data regarding the type of axonal injury incurred.[6,8] Without serial testing and visual inspection, there is no way to differentiate between simple neuropraxia and complete nerve transection. Furthermore, intraoperative electrophysiological measures do not estimate neuroregenerative capacity.

The present study carries several notable strengths. First, we used high-level monopolar stimulation to interrogate FN conduction. Low stimulation levels ($<$ 1 mA) or bipolar probes may overlook splayed bundles of functioning axons, thereby only testing a portion of the FN.[5,9] Second, the FN was routinely stimulated at the proximal nerve root exit zone of the pontomedullary junction. This is critical because stimulation at the porus or internal auditory canal may miss the injured FN segment in tumors with large cisternal extension.[9] Third, an internal positive control was used when the distal undissected FN was also tested to ensure that the lack of a proximal EMG response was not secondary to faulty monitoring technique. In the present series, all 11 patients experienced immediate complete FN paresis, as would be expected if the entire nerve was tested and was unresponsive to high-level proximal stimulation. The above issues may explain reports in which patients experienced only partial FN paralysis immediately after surgery despite unevokable FN segments.[24]

Finally, several limitations deserve mention. First, the data were collected retrospectively. Therefore, the authors relied on the accuracy and completeness of the existing patient medical record. Second, statistical conclusions are limited by small patient numbers inherent to the rarity of the studied condition.

CONCLUSION

Optimizing FN outcomes after VS microsurgery remains a priority. Although the great majority of patients in all modern series demonstrate satisfactory long-term FN function, a subset of subjects develop substantial long-term FN dysfunction despite intraoperative FN EMG monitoring and less-than-complete resection.

We report the outcomes of 11 patients who were found to have anatomically intact but electrically unresponsive FNs at the conclusion of tumor dissection to determine whether same-surgery FN repair should be considered. Even among this extreme subset, the majority of patients recovered to HBG 3 or worse function in $<$ 1 year. Although somewhat unpredictable, these outcomes are on par with or better than what could be expected with primary neurorrhaphy or interposition grafting. These data demonstrate that current electrophysiological interrogation strategies are incapable of reliably predicting who will do poorly and that if the anatomic continuity of the FN is maintained, attempts at same-surgery FN repair should not be pursued.

Disclosures

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COMMENTS

The authors retrospectively reviewed a prospectively collected internal database for patients undergoing vestibular schwannoma resection in which facial nerve stimulation at 5 mA was unyielding despite anatomic continuity. They reported 11 of 350 patients between 2000 and 2010 who met these criteria. One patient presented with a preoperative House-Brackmann score of 5; the remaining patients had no preoperative facial weakness. Nine patients (81.8%) presented with unilateral hearing loss and underwent a retrosigmoid craniotomy for tumor resection; the rest underwent a translabyrinthine approach. The mean tumor diameter was 2.5 cm, with many tumors being vascular or firmly adherent to or splaying the facial nerve. All patients had either a gross total or near-total resection. The median duration of follow-up was 81.8 months.

Intraoperative stimulation was 100% predictive of immediate postoperative facial function with all patients developing a complete facial palsy (House-Brackmann grade 6). However, it was incapable of predicting long-term outcome, and 80% of patients recovered to a House-Brackmann grade 3 or better. The definitive recovery period took on average 9.4 months, after which no further return of facial function was identified.

A significant body of literature describes facial nerve outcome prediction in relation to different electromyographic parameters. However, little has been published on the extreme subset reported in this article. Waiting for the recovery of an anatomically intact facial nerve for up to 1 postoperative year is the more common practice. However, concerns regarding denervation atrophy highlight the need for a more definitive way of predicting facial nerve recovery. In our practice, we use the A-train neurotonic discharges, stimulus threshold, response amplitude, and proximal-to-distal amplitude to predict functional outcome. We still wait a 1 full year before considering facial reanimation.

Although the cohort reported in this article is limited in number, it provides some evidence that in this rare situation, facial nerve reinnervation strategies should be withheld during the first postoperative year. We congratulate the authors on their valuable contribution to the literature.

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This is an interesting and well-written article focusing on a particular issue that has not yet been explored systematically. The authors evaluated the facial nerve functional outcome in patients with an electrically unresponsive but anatomically intact seventh nerve after microsurgical vestibular schwannoma resection. They aimed to determine whether same-surgery facial nerve repair should be considered in this unique subset. Eleven patients met the inclusion criteria of this retrospective study. In 7 patients (63.6%), the facial nerve at follow-up recovered to a House-Brackmann grade 3; in 2 patients (18.2%), it recovered to grade 4; in 1 (9%), it recovered to grade 5; and in 1 patient (9%), complete facial palsy persisted. These results correspond to our experience at the International Neuroscience Institute in Hannover: Some patients with electrically unevokable but anatomically intact nerve recover spontaneously to an acceptable level. Facial nerve reconstruction in this subgroup should not be pursued. Other patients, however, do not recover satisfactorily.

We base the decision of whether to undertake same-surgery nerve repair on 2 criteria: the response to electrical stimulation and the morphological condition of the nerve at the end of surgery. Anatomically intact but unresponsive nerves do not make up a homogeneous group; they may be of almost preserved morphology, splayed, atrophic to a variable extent, or partially interrupted. In case of very severe atrophy, immediate nerve reconstruction should be considered. Alternatively, the facial recovery is observed closely, both clinically and electrophysiologically, and if complete facial palsy persists within 10 to 12 months, facial nerve reanimation procedure is performed. Our favorite technique is the side-to-end hypoglosso-facial anastomosis.

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