

**ABSTRACT**

**Introduction:** Meningiomas account for 2.2% to 2.5% of all cerebral tumors, of which only 2% are located in the foramen magnum. Foramen magnum meningiomas (FMMs) are commonly found in women, with a mean age at onset of 52 years old. They generally behave more aggressively than other meningiomas.

**Materials and Methods:** We performed epidemiological, anatomical and surgical analyses of 20 patients diagnosed with FMMs who underwent surgical treatment from 1999 to 2019 at Santa Paula Hospital in Sao Paulo. This case series was compared with previously published ones to better understand this relatively rare disease.

**Results:** Twenty patients were included, with a mean follow-up of 110 months. Their mean age was 37.8 years old. The mean preoperative Karnofsky performance status scale (KPS) was 84%. We found a female (65%) and left hemisphere predominance (50%). Involvement of both hemispheres was found in 25% of patients. FMM locations were anterior, anterolateral, lateral and posterior, in 45%, 35%, 10%, and 10%, respectively. Simpson resection grades I, II, and III were achieved in 25%, 60%, and 15% of cases, respectively. Mean postoperative KPS was 79%. Three patients with anterior and bilateral located meningiomas had a worse postoperative KPS in comparison to the preoperative one.

**Conclusion:** Anterior and bilateral FMMs seem to be related to a worse prognosis. A gross total resection can reduce the recurrence rates. The KPS is worse in patients with recurrence.

**Keywords:** Foramen magnum, meningioma, skull base

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**INTRODUCTION**

Meningiomas are central nervous system (CNS) tumors originated from arachnoid cap cells, representing 14.3% to 20% of all CNS tumors in the adult population. Those tumors have an incidence of 4.4/100.000 per year and a mean age at presentation of 63 years old.[1-3]

The first foramen magnum meningioma (FMM) was described in 1872 after an autopsy at Lariboiisieres Hospital in Paris; however, the first successfully operated case was only described by Frazier and Spiller in 1922. FMMs represent 1.5% to 3.6% of all intracranial tumors; they are commonly found in women with a mean age of 52 years old. Regarding the localization, about 90% of them are anterior and anterolateral, posterior ones are the least common. An en plaques tumor can be found in rare cases.[4-8]
In the pediatric population, meningiomas present at a mean age of 14.4–15.5 years old. The incidence varies between 1% and 4% during the first two decades of life. Meningiomas account for 2.2% to 2.5% of all cerebral tumors, of which only 2% are located in the foramen magnum.

Pediatric FMMs are usually related to genetic diseases, such as neurofibromatosis type 2. The most frequent localizations are the anterior and anterolateral ones. Vertebral artery (VA) involvement is common, as well as lesions of the IX, X, and XII cranial nerves.

To better understand the natural history and presentation of this particular tumor, we retrospectively reviewed the cases operated in our service. Our main goal was to define how we should manage FMMs.

MATERIALS AND METHODS

We performed epidemiological, anatomical, and surgical analyses of 20 patients diagnosed with FMM who underwent surgical treatment from 1999 to 2019 at Santa Paula Hospital, in Sao Paulo. This case series was then compared with articles published during the same period, to identify if our results were compatible with the ones found in the literature. We used PUBMED and LILACS databases for this review. Multiple variations of the following keywords were used: “Meningiomas,” “foramen magnum,” “skull base” and “skull base tumors.” Case reports and other reviews were not included. The inclusion criteria focused on the following parameters: studies about FMMs, complications, and follow-up) were not included, in humans, pre- and post-operative clinical evaluation, surgical outcome, surgical results, with more than 10 patients and with a clear description of results. Studies with <10 patients, without the description of results (including tumor classification.

RESULTS

Twenty patients were analyzed [Table 1], with a mean follow-up period of 110 months (range 0–241). The mean age at presentation was 37.8 years old (range 19–53). We noticed a female (65%) and left side (50%) predominance. Both sides were affected in 25% of cases. Anterior, anterolateral, lateral, and posterior locations were, respectively, found in 45%, 35%, 10%, and 10% of patients.

All patients underwent surgical procedure through the half-sitting position, using the transesophageal Doppler (to monitor air embolism), electrophysiologic monitoring by motor and sensory evoked potentials, and continuous electroneuromyography (for lower cranial nerves monitoring).

The incision was hockey-stick shaped, using the C2 spinous process, external occipital protuberance, and mastoid process as anatomical references, ending 1 cm below the latter. Muscles were dissected to obtain bone exposure, where a trepanation hole was done below the transverse and posterior to the sigmoid sinus. Below the foramen magnum, posterior arch, and lateral sulcus of C1 were identified where the V3 segment of VA lies before entering into the skull. This segment should be transposed carefully to avoid lesions when removing the latero-posterior arch of C1 and, if necessary, to allow removal of condyle's posterior portion for anterior tumor's access. After that, the bulbar nerves were identified, tumor’s debulking and removal were performed. Signs and symptoms at presentation were lower cranial nerves deficits (60%), pyramidal tract syndrome (50%), dizziness (40%), VIII cranial nerve lesion (40%), motor deficits (35%), gait disturbances (25%), dysesthesia (15%), headache (15%), hearing loss (10%), diplopia (10%), dysphagia (10%), hoarseness (10%), Lhermitte’s sign (5%), and VII cranial nerve lesion (5%).

The extreme lateral approach was used in 90% of cases. The other 10%, corresponding to posterior FMMs, were operated through a midline suboccipital approach. Simpson resection grades I, II, and III were achieved in 25%, 60%, and 15% of cases, respectively.

The mean preoperative Karnofsky performance status scale (KPS) was 84%, while postoperative was 79% [Graph 1]. Four patients had a decrease from the preoperative value due to severe complications after surgery. Patient 5 presented meningitis, with later recurrence and death 96 months after the initial surgery. Patient 9 suffered VA lesion and further brainstem ischemia, leading to death during the hospital stay. Patient 12 presented bleeding immediately after surgery, requiring a new surgical procedure. Patient 13 presented...
brainstem ischemia secondary to posterior inferior cerebellar artery lesion.

Postoperative complications were: hydrocephalus (requiring shunt in four patients), gastrostomy (10%), tracheostomy (25%), cerebrospinal fluid (CSF) fistula (5%), meningitis (5%), brainstem ischemia (10%), pulmonary thromboembolism (5%), bleeding on the surgical bed (5%), and death (5%).

We could also observe that three patients with anterior and bilateral located meningiomas had a worse KPS in the follow-up period, as well as the 75% of patients that presented recurrence. All of those patients underwent total resection (one patient Simpson I and three patients Simpson III).

**DISCUSSION**

We performed an analysis of a case series focused on surgical treatment. We intended to provide evidence of several studies and their results, so they could be compared to our own data. Our main goal was to establish an effective surgical treatment for those rare tumors to decrease complications and recurrence rates. In Table 2, we show the studies, year of publication, number of cases, mean age, tumor location, follow-up period, pre- and post-operative clinical statuses, gross total resection rate, and main complications.

About 70% of foramen magnum tumors are meningiomas, followed by schwannomas.\(^{[12]}\) By definition, FMMs are located between the lower third of the clivus and the posterior arch of the C2 vertebra.\(^{[13]}\) They must be anterior, below the lower third of clivus, above the superior border of the axis, lateral to jugular tubercle, and C2 laminae. They are posteriorly limited by the edge of the squamous occipital bone and spinous process.\(^{[14]}\)

Cranial nerves IX, X, and XI arise from postolivary sulcus, pass anteriorly to the foramen of Luschka, posteriorly to the VA and penetrate the jugular foramen. The XII\(^{th}\) nerve originates ventrally from preolivary sulcus, passes anteriorly to VA, and penetrates the hypoglossal canal. Those anatomic aspects explain why all lower cranial nerves can be affected by FMMs.\(^{[8]}\)

In our series, the mean age at presentation was 37.8 years old (range 19–53). This differs from the majority of studies, which showed ages close to the sixth decade of life. However, Goel et al.\(^{[15]}\) showed similar numbers to ours.

Meantime of onset to diagnosis varied between 20 and 27 months and the most common initial symptoms were chronic headache, neck pain, dysesthesia, ataxia, paresis, and difficulty using hands. Most physical findings were hyperreflexia, extremity weakness, Babinski sign, spastic gait, hypoesthesia, occipital neuralgia, cranial nerve XI deficit, C2

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**Table 1: Foramen magnum meningiomas: 20 cases series**

| Age  (years) | Gender | Side | Far lateral | Simpson | WHO grade | Recurrence | KPS (pre/post) | Radiotherapy | Follow-up (months) | Location | Associated diseases |
|-------------|--------|------|-------------|---------|-----------|------------|---------------|--------------|-------------------|----------|---------------------|
| 32          | Male   | Left | Yes         | II      | Clear cell |            | 90/90        | Yes          | 134               | A        |                     |
| 44          | Female | Left | Yes         | I       | I         |            | 60/90        | Yes          | 87               | A        | Neurinoma           |
| 34          | Female | Right| Yes         | II      | I         |            | 90/100       | Yes          | 146              | A        |                     |
| 52          | Female | Bilateral | Yes | II | I | Yes | 90/70 | Yes | 123 | A | ACoA |
| 19          | Male   | Bilateral | Yes | II | I | Yes | 50/10 | No | 96 | A |                     |
| 53          | Female | Left | Yes | III | Atypical | | 90/100 | Yes | 63 | AL |                     |
| 34          | Female | Left | Yes | II | I | | 80/90 | Yes | 144 | A |                     |
| 45          | Male   | Right| Yes | II | I | | 90/100 | Yes | 88 | A |                     |
| 40          | Male   | Left | Yes | III | I | | 90/0 | No | 0 | A |                     |
| 52          | Female | Left | Yes | II | I | | 90/90 | No | 74 | AL |                     |
| 39          | Female | Right| Yes | II | I | | 80/100 | No | 147 | AL |                     |
| 40          | Female | Left | Yes | II | I | | 90/80 | Yes | 241 | AL |                     |
| 48          | Male   | Left | Yes | II | I | Yes | 40/30 | Yes | 74 | AL |                     |
| 40          | Female | Right| Yes | II | I | | 80/90 | No | 144 | AL |                     |
| 39          | Male   | Left | Yes | II | I | | 90/90 | No | 87 | AL |                     |
| 37          | Male   | Bilateral | No | I | I | | 100/100 | No | 100 | P |                     |
| 22          | Female | Bilateral | No | I | I | | 100/100 | No | 120 | P |                     |
| 32          | Female | Right| Yes | I | I | | 100/100 | Yes | 110 | LAT |                     |
| 25          | Female | Left | Yes | I | I | Yes | 90/90 | No | 108 | LAT |                     |
| 30          | Female | Bilateral | Yes | III | I | | 90/60 | No | 123 | A |                     |

KPS - Karnofsky Performance Score Scale; A - Anterior FMM; AL - Antero-lateral FMM; P - Posterior FMM; LAT - Lateral FMM; ACoA - Anterior communicating artery aneurysm; WHO - World Health Organization; FMM - Foramen magnum meningiomas
| Study                              | Year | Patients (n) | Gender | Age (years) | A | AL | P | Follow-up (months) | KPS (pre/postoperative) | Simpson I/II (%) | Complications                                                                 |
|-----------------------------------|------|--------------|--------|-------------|---|----|---|-------------------|--------------------------|-----------------|-------------------------------------------------------------------------------|
| George et al.[27]                 | 1997 | 40           | 11 males 29 females | 51.6 | 18 | 21 | 1 | NA                | NA                       | 94/50**          | Death (7.5%) Cranial nerve palsy (IX and X)                                      |
| Arnautovic et al.[16]            | 2000 | 18           | NA     | NA          | 16 | 0  | 0 | 40                | NA                       | 75              | Lower cranial nerves lesion (5.8%)                                             |
| Goel et al.[30]                  | 2001 | 17           | 6 males 11 females | 39.2 | 0  | 17 | 0 | 43                | NA                       | 82.3             |                                                                                 |
| Boulton and Cusimano[17]         | 2003 | 10           | 2 males 8 females | 55   | 0  | 7  | 3 | 33                | NA                       | 90              | CSF fistula (10%) Brown-squared syndrome (10%) Neurocognitive impairment (10%) |
| Pannir et al.[18]               | 2004 | 22           | 4 males 18 females | 47.2 | 0  | 20 | 2 | 40                | 73/94                    | 95.5             | Fistula (18%) Hydrocephalus (4.5%) Lower cranial nerves paralysis (9%) Vascular injury (4.5%) Mortality (4.5%) |
| Bassiouni et al.[26]           | 2006 | 25           | 6 males 19 females | 59.2 | 8  | 14 | 3 | 73                | 79/89                    | 96              | CSF fistula (16%) Lower cranial nerves lesion (4%) Mortality (4%)               |
| Borba et al.[19]               | 2009 | 15           | 1 male 14 females | 55.9 | 8  | 7  | 0 | 23.6              | NA                       | 80              | XII nerve paralysis (6.6%) Fistula (6.6%) Hydrocephalus (6.6%) Transient myelopathy (6.6%) |
| Kandenwein et al.[24]          | 2009 | 16           | 4 males 12 females | 61   | 3  | 12 | 1 | 43.5              | NA                       | 87.5             | Cranial nerve palsy (31%)                                                      |
| Wu et al.[21]                   | 2009 | 114          | 46 males 68 females | 52.3 | 80 | 24 | 10 | 90.3              | 72.5/83.5                | 86              | Dysphagia (55%) Tracheostomy (28.9%) hemianesthesia (2.6%) hydrocephalus (7.8%) CSF fistula (6.1%) Infection (2.6%) hypothyroidism (2.6%) hypopituitarism (3.5%) Hyponatremia (1.8%) Diabetes insipidus (0.08%) Death (1.8%) Neck movement restriction and instability (39%) Hemiparesis (4.3%) Dysphagia (30%) Lower cranial nerves palsy (13%) VI palsy (17%) Hoarseness (13%) |
| Kano et al.[14]                  | 2010 | 23           | 8 males 15 females | 56   | 9  | 14 | 0 | 42.8              | 83.9/89.5                | 62.5             | Pulmonary thromboembolism Death (1.9%)                                          |
| Bruneau and George[12]            | 2010 | 107*         | NA     | NA          | 41 | 57 | 6 | 120               | NA                       | 86              | Pulmonary thromboembolism Death (1.9%)                                          |
| Pirotte et al.[22]              | 2010 | 26           | 9 males 13 females | 53   | 7  | 15 | 0 | 88                | NA                       | 73              | Death (4.5%) Hydrocephalus (4.5%) Fistula Hemiparesis 4.5% Lower cranial nerves paralysis (27.2%) Tracheostomy (4.5%) |
| Talacchi et al.[20]            | 2012 | 64           | 16 males 48 females | 59   | 24 | 40 | 0 | 138               | NA                       | 81              | Cranial nerves palsy IX XII (35%) Dysphagia Temporary gastrostomy               |

Contd...
hyperalgesia, neck rigidity, and Brown-Sequard syndrome. General symptoms can also be found, such as a headache and neck pain; they are explained by nervous innervation of meninges. Forty percent of patients may present a normal neurological examination.

When analyzing location, we observed a predominance of anterolateral tumors on most studies, except for Arnautovic et al. series, in which tumors were exclusively anterior, and Li, et al., and Borba et al., series. In our series, there was no predominance, with 45% anterior and 45% anterolateral.

Surgery is the treatment of choice. The main goal is to achieve complete removal of the lesion, corresponding to Simpson I and II resection grades, which are obtained in 46% to 96% of cases. Meningiomas attached to the brainstem, venous sinuses, VA and cranial nerves or presenting malignancy, high mitotic activity, or loss of 1p36.1-p34 have been associated with incomplete resection. Recurrence is lower in gross total resection cases. Simpson I and II resections were able to reduce recurrence rates. However, surgeons should be aware that complications can be higher in those resections.

We identified three risk factors for a worse prognosis: anterior location, bilaterality, and recurrence. When analyzing location, we observed a predominance of anterolateral tumors on most studies, except for Arnautovic et al. series, in which tumors were exclusively anterior. There was no predominance in our current case series.

### Table 2: Contd...

| Study         | Year | Patients (n) | Gender | Age (years) | A | AL | P | Follow-up (months) | KPS (pre/postoperative) | Simpson I/II (%) | Complications                                      |
|---------------|------|--------------|--------|-------------|---|----|---|--------------------|------------------------|----------------|---------------------------------------------------|
| Colli et al.  | 2014 | 13           | 2 males, 11 females | 54.15 | 4  | 9  | 0  | 47.3               | >80**                  | 69.2          | CSF fistula (30.8%), difficulty breathing (7.7%), Transient lower cranial nerves palsy (38.5%), Permanent lower cranial nerves palsy (7.7%) |
| Dobrowolski et al. | 2016 | 24           | 6 males, 18 females | 52   | 3  | 19 | 2  | 45.6               | 85/-                   | 83.3          | Hydrocephalus (4.1%), CSF fistula 4.1%, Pneumonia 4.1%, hemorhaghe 4.1% |
| Li et al.     | 2017 | 185          | 61 males, 124 females | 49.4 | 122| 49 | 14  | 110.3              | 80/>80                 | 83.2          | Sinus thrombosis 4.1%, CSF fistula 8.3%, dysesthesia 4.1% |
| Current case series | 2018 | 20           | 7 males, 13 females | 37.85 | 9 | 9  | 2  | 110                | 84/79                  | 85           | Hydrocephalus (20%), CSF fistula (5%), meningitis (5%), Brainstem ischemia 10%, pulmonary thromboembolism 5%, Rebleeding (5%) |

A - Anterior FMM; AL - Antero-lateral FMM; P - Posterior FMM; CSF - Cerebrospinal fluid; KPS - Karnofsky performance score scale; NA - Not available; FMM - Foramen magnum meningiomas; VI - Abducens nerve; IX - Glossopharyngeal nerve; X - Vagus nerve; XII - Hypoglossal nerve; **Intradural/extradural
CONCLUSION

Even though we had a small number of patients, we believe that anterior location, bilaterality, and recurrence are indicators of a worse prognosis. Gross total resections can reduce the recurrence rates, consisting of the main surgical goal. More aggressive tumors can be found in children, anaplastic tumors are more common in this population. The analysis of cases and studies described in this article makes us emphasize the necessity of even more studies about FMMs, with greater numbers of patients and a better exposition of pre- and post-operative KPS.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Flores BC, Boudreaux BP, Klinger DR, Mickey BE, Barnett SL. The far-lateral approach for foramen magnum meningiomas. Neurosurg Focus 2013;35:E12.
2. George B, Lot G, Boissonnet H. Meningioma of the foramen magnum: A series of 40 cases. Surg Neurol 1997;47:371-9.
3. Mawrin C, Perry A. Pathological classification and molecular genetics of meningiomas. J Neurooncol 2010;99:379-91.
4. Bassiouni H, Ntoukas V, Asgari S, Sandalcioglu EI, Stolke D, Seifert V. Foramen magnum meningiomas: Clinical outcome after microsurgical resection via a posterolateral suboccipital retrocondylar approach. Neurosurgery 2006;59:177-85.
5. Bruneau M, George B. Foramen magnum meningiomas: Detailed surgical approaches and technical aspects at Lariboisière Hospital and review of the literature. Neurosurg Rev 2008;31:19-32.
6. Dobrowolski S, Ebner F, Lepski G, Tatagiba M. Foramen magnum meningioma: The midline suboccipital subtonsillar approach. Clin Neurol Neurosurg 2016;145:28-34.
7. Mohindra S, Savardekar A, Tripathi M, Rane S. En-plaque foramen magnum meningiomas: Rare presentations. Br J Neurosurg 2012;26:899-901.
8. Mostofi K. Foramen magnum meningioma: Some anatomical and surgical remarks through five cases. Asian Spine J 2015;9:54-8.
9. Athanasiou A, Magras I, Sarlis P, Spyridopoulos E, Polyzioudis K. Anterolateral meningioma of the foramen magnum and high cervical spine presenting intradural and extradural growth in a child: Case report and literature review. Childs Nerv Syst 2015;31:2345-51.
10. Gump WC. Meningiomas of the pediatric skull base: A review. J Neurol Surg B Skull Base 2015;76:66-73.
11. Menezes AH. Craniovertebral junction neoplasms in the pediatric population. Childs Nerv Syst 2008;24:1173-86.
12. Bruneau M, George B. Classification of foramen magnum meningiomas. J Craniovertebr Junction Spine 2010;1:10-7.
13. Colli BO, Carlotti Junior CG, Assirati Junior JA, Borba LA, Coelho-Junior Vde P, Neder L. Foramen magnum meningiomas: Surgical treatment in a single public institution in a developing country. Arq Neuropsiquiatr 2014;72:528-37.
14. Kano T, Kawase T, Horiguchi T, Yoshida K. Meningiomas of the ventral foramen magnum and lower clivus: Factors influencing surgical morbidity, the extent of tumour resection, and tumour recurrence. Acta Neurochir 2010;152:79-86.
15. Goel A, Ch M, Desai K, Muzumdar D. Surgery on anterior foramen magnum meningiomas using conventional posterior sub-occipital approach: A report on an experience with 17 cases. Neurosurgery 2001;49:102-6.
16. Arnaoutovic KI, Al-Mefty O, Husain M. Ventral foramen magnum meningiomas. J Neurosurg 2000;92:71-80.
17. Boulton MR, Cusimano MD. Foramen magnum meningiomas: Concepts, classifications, and nuances. Neurosurg Focus 2003;14:e10.
18. Pamir MN, Kiliç T, Özdeman K, Türe U. Experience of a single institution treating foramen magnum meningiomas. J Clin Neurosci 2004;11:863-7.
19. Borba LA, de Oliveira JG, Giudicissi-Filho M, Colli BO. Surgical management of foramen magnum meningiomas. Neurosurg Rev 2009;32:49-58.
20. Kandenwein JA, Richter HP, Antoniadi G. Foramen magnum meningiomas – Experience with the posterior suboccipital approach. Br J Neurosurg 2009;23:33-9.
21. Wu Z, Hao S, Zhang J, Zhang L, Jia G, Tang J, et al. Foramen magnum meningiomas: experiences in 114 patients at a single institute over 15 years. Surg Neurol 2009. https://doi.org/10.1016/j.surneu.2009.05.006.
22. Pirotte BJ, Brocht J, DeWitte O. Management of anterolateral foramen magnum meningiomas: Surgical vs conservative decision making. Neurosurgery 2010;67:s58-70.
23. Talacchi A, Birolli A, Soda C, Masotto B, Bricolo A. Surgical management of ventral and ventrolateral foramen magnum meningiomas: Report on a 64-case series and review of the literature. Neurosurg Rev 2012. https://doi.org/10.1007/s10143-012-0381-6
24. Li D, Wu Z, Ren C, Hao SY, Wang L, Xiao XR, et al. Foramen magnum meningiomas: surgical results and risks predicting poor outcomes based on a modified classification. J Neurosurg 2017;126:661-76. Epub 2016 May 13. Erratum in: J Neurosurg 2017;126:1017.
25. Aguiar PH, Neto MA, Junior OL. Princípios Técnicos de Neurocirurgia: Atlas e Texto. Meningioma de Forame Magno. Tratamento Cirúrgico, 1ª ed. São Paulo: Di livros; 2016. p. 445-52.
26. Kim NH, Yang SY, Koo JB, Jeong SW. Occipital neuralgia as the only presenting symptom of foramen magnum meningioma. J Clin Neurosurg 2009;5:198-200.
27. Marin Sanabria EA, Ebara K, Tamaki N. Surgical experience with skull base approaches for foramen magnum meningioma. Neurrol Med Chir. 2002;42:472-8.
28. Meyer FB, Ebersold MJ, Reese DF. Benign tumors of the foramen magnum. J Neurol 1984;61:136-42.
29. Polyzoïdes S, Koletsia T, Panagiotidou S, Ashkan K, Theoharides TC. Mast cells in meningiomas and brain inflammation. J Neuroinflammation. 2015;12:170.
30. Sheehan JP, Starke RM, Kano H, Barnett GH, Mathieu D, Chiang V, et al. Gamma knife radiosurgery for posterior fossa meningiomas: A multicenter study. J Neurosurg 2015;122:1479-89.
31. Starke RM, Nguyen JH, Reames DL, Rainey J, Sheehan JP. Gamma knife radiosurgery of meningiomas involving the foramen magnum. J Craniotube Junction Spine 2010;1:23-8.
32. Matsushima T, Kawashima M, Masuoka J, Mineta T, Inoue T. Transcendental fossa (supracondylar transjugular tubercle) approach: Anatomic basis for the approach, surgical procedures, and surgical experience. Skull Base 2010;20:83-91.