Michael J. Fucci, M.D., Eugene L. Alford, M.D.,
Louis D. Lowry, M.D., William M. Keane, M.D.,
Robert T. Sataloff, M.D., D.M.A.

Endoscopic Management of a Giant Cholesterol Cyst of the Petrous Apex

Abstract—Giant cholesterol cyst (GCC) of the petrous apex is a rare clinical entity. This benign cystic lesion can cause neurologic deficits and vascular compromise by persistent growth and progressive bone destruction. Magnetic resonance imaging studies of GCC show the lesions to be hyperintense on T_1-weighted sequences with progressively lower signal intensities on the first and second echoes of T_2-weighted sequences. These findings are relatively specific for GCC, permitting a narrow differential diagnosis. The goal of surgery is to provide adequate drainage with the creation of a permanent fistula. The classic approaches to these lesions are the posterior fossa craniotomy and the middle fossa extradural craniotomy. The translabyrinthine approach provides wide exposure at the expense of cochlear and vestibular function. The transsphenoidal approach provides adequate drainage with hearing preservation and no craniotomy. The endoscopic, endonasal transsphenoidal approach to a 2.5 cm GCC of the petrous apex accomplished complete drainage with the creation of a fistula. Advances in endoscopic technique and instrumentation facilitated the addition of the approach to the surgeon's armamentarium. In selected cases, this approach provides adequate surgical exposure with minimal morbidity. (*Skull Base Surgery, 4(1):52–58, 1994*)

Giant cholesterol cyst (GCC) is a rare lesion of the temporal bone. Graham et al[1] reported GCC to be a distinct clinical entity with unique histologic and radiographic findings. These lesions must be differentiated from other lesions of the temporal bone such as congenital cholesteatoma, cholesterol granuloma, petrous apex abscess, meningioma, carotid artery aneurysm, and glomus tumor.

The literature is scattered with descriptions of cystic lesions of the temporal bone consistent with GCC. The lesions are believed to originate in well-pneumatized temporal bones, particularly in the petrous apex. The natural history of GCC is to slowly enlarge, destroying bone in its path. Patients generally present with cranial nerve deficits produced by the mass effect of the lesion.[2] Hearing loss and vertigo are the most common presenting symptoms. The abducens nerve and the facial nerve are the next most commonly affected nerves after the cochleovestibular nerve.

The management of GCC consists of drainage and exteriorization of the cyst. The cyst contains a fibrous lining that is occasionally adherent to a skeletonized carotid artery and complete removal of this fibrous lining can cause significant morbidity and mortality.

Several surgical approaches have been used to gain access to these lesions in the petrous apex. The posterior fossa approach has been used to access these lesions.[3] However, the cyst can be drained, but a permanent drainage route cannot be established by this approach. The middle fossa route has been used as well.[4,5] With these approaches, the cysts have been drained into the anterior tympanic cavity and the eustachian tube.[1,2] The transmastoid route has been used successfully, especially when the cyst expands laterally, by creating a permanent fistula

Skull Base Surgery, Volume 4, Number 1, January 1994 Department of Otolaryngology–Head and Neck Surgery, Thomas Jefferson University Hospital, Philadelphia, Pennsylvania, and Department of Otolaryngology and Communicative Sciences (E.L.A.), Baylor College of Medicine, Houston, Texas Presented at the Fourth Annual Meeting of the North American Skull Base Society, Scottsdale, AZ, February 12–14, 1993 Reprint requests: Dr. Fucci, Department of Otolaryngology–Head and Neck Surgery, Thomas Jefferson University Hospital, 909 Walnut Street–Third Floor, Philadelphia, PA 19107

into the middle ear or mastoid cavity.[2] Occasionally, a transcochlear petrous apicoectomy is performed to create a large epithelial lined tract into the middle ear space.[6] Montgomery,[7] in 1977, reported on two cases of petrous apex GCC that were exteriorized by a transsphenoidal approach via an external ethmoidectomy. A pedicled mucosal flap was used to create a permanent fistula into the nasal cavity. The advantages of the approach include preservation of cochlear and vestibular function, drainage into a mucosal lined cavity, and the avoidance of a craniotomy.

Recently, the techniques of endoscopic sinus surgery have expanded to include dacryocystorhinostomy, repair of cerebrospinal fluid leaks, and orbital decompression for dysthyroid orbitopathy.[8–10] These same techniques were used to drain a 2.5 cm GCC of the petrous apex.

CASE REPORT

A 36-year-old woman with a chief complaint of headache had a normal neurologic examination and normal preoperative hearing. She had a computerized tomography (CT) scan that showed a 2.5 cm lytic lesion in the left petrous apex (Figs. 1, 2). The anteromedial extent of the lesion abutted the posterior wall of the sphenoid sinus. There was a complete bony dehiscence between the cyst and the horizontal portion of the carotid artery. There was expansion and deformity of the porus acousticus causing compromise of the internal auditory canal. Magnetic resonance (MR) images showed a 2.5 by 2.0 by 2.0 cm mass expanding the left petrous apex (Figs. 3–5). The mass was hyperintense on T_1-weighted sequences and became progressively lower in signal intensity on the first and second echoes of the T_2-weighted sequences.

Under general anesthesia supplemented with topical and injectable anesthesia and vasoconstrictors, the left middle turbinate was resected to permit more complete posterior exposure. The sphenoid sinus was identified and entered. The anterior wall of the sphenoid sinus was removed using a 90° Kerrison forceps. The posterior wall of the sphenoid sinus was fully exposed and the crest of bone along the posterolateral wall of the sphenoid sinus identified the carotid artery. The posterior wall of the sphenoid sinus was thin and pliable on palpation. A Hardy pituitary dissector was used to remove the bone of the posterior wall of the sphenoid sinus over the GCC. A small amount of brown fluid leaked in the dissected area. The cyst was exposed and opened from medial to lateral using 90° Kerrison forceps, revealing a large, smooth, fluid-filled cavity. The fluid was evacuated and the anterior wall was marsupialized. The posterior wall of the cavity measured 10.5 cm from the anterior nasal spine. A T-shaped silicone stent was placed into the cyst to maintain a tract between the cyst and the nasal cavity. A cross-table fluoroscopy unit was used intraoperatively to help identify and confirm the landmarks. The patient was discharged home on postoperative day 1 with no complications.

Follow-up office examination and nasal endoscopy showed the stent to be open and in place at 3 months after surgery. The headaches had resolved but she did complain of some nasal irritation from the stent.

Figure 1. Axial CT scan shows expansive mass in the left petrous apex. Note well-aerated sphenoid sinus and dehiscence of carotid artery. Middle ear cavity and mastoid air cells are normal.

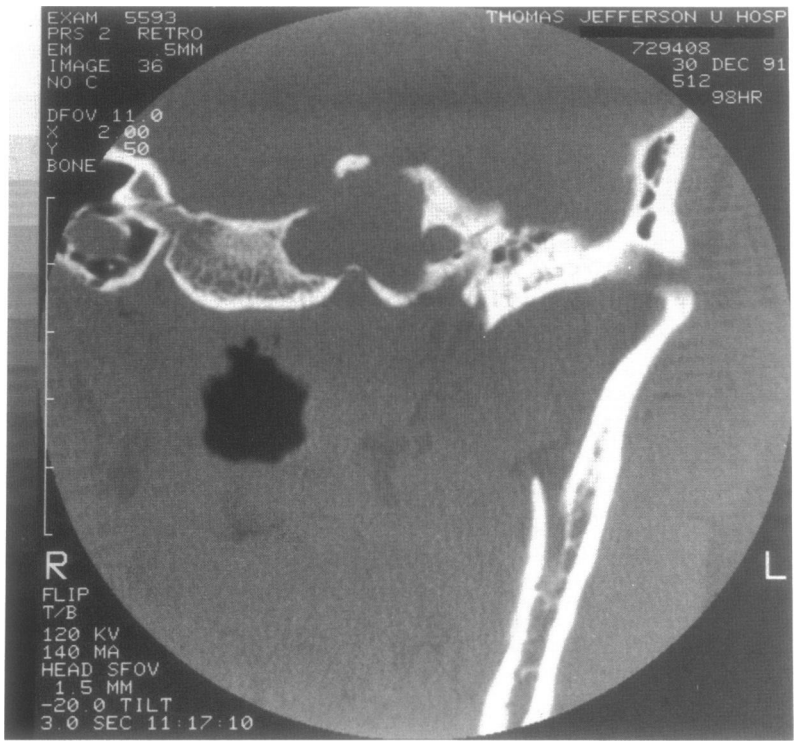

Figure 2. Coronal CT scan shows mass in left petrous apex. Note dehiscent carotid artery at lateral aspect of lesion.

DISCUSSION

Diagnosis

Patients with GCC of the petrous apex commonly present with cranial nerve deficits from the compressive effect of the lesion.[2] Cranial nerves V through XII can be affected, depending on the location and size of the GCC. Audiometry, tympanometry, electronystagmography, and auditory brainstem evoked potentials reveal variable and nonspecific findings based on the lesion's location and its effect on surrounding structures.

CT scans of GCC show an expansile lytic lesion usually in the petrous apex of a well-aerated temporal bone. The lesion's margins are often sclerotic and the petrous apices are usually well-pneumatized. MR scanning shows that the defect is hyperintense on T_1-weighted sequences and becomes progressively lower in signal intensity on the first and second echoes of the T_2-weighted

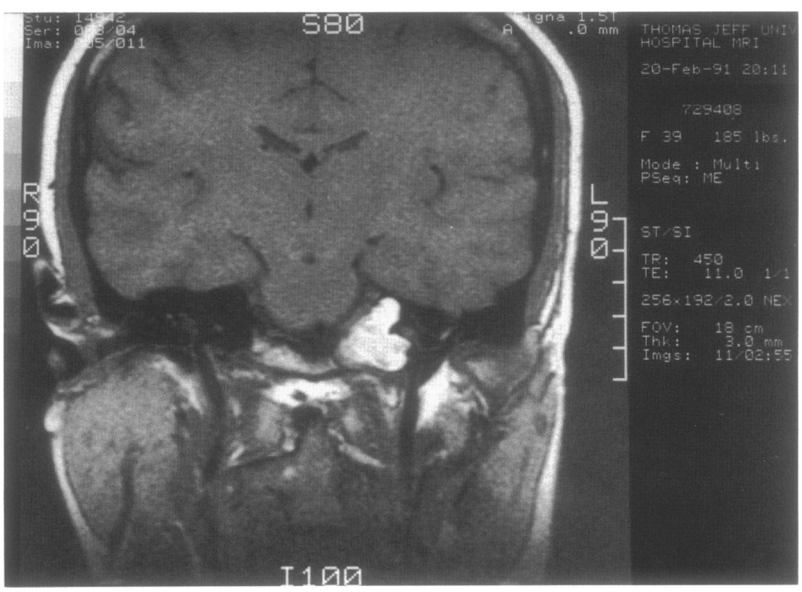

Figure 3. Coronal T_1-weighted MR image (TR = 450 msec, TE = 11 msec) shows slightly inhomogeneous mass with high intensity signal.

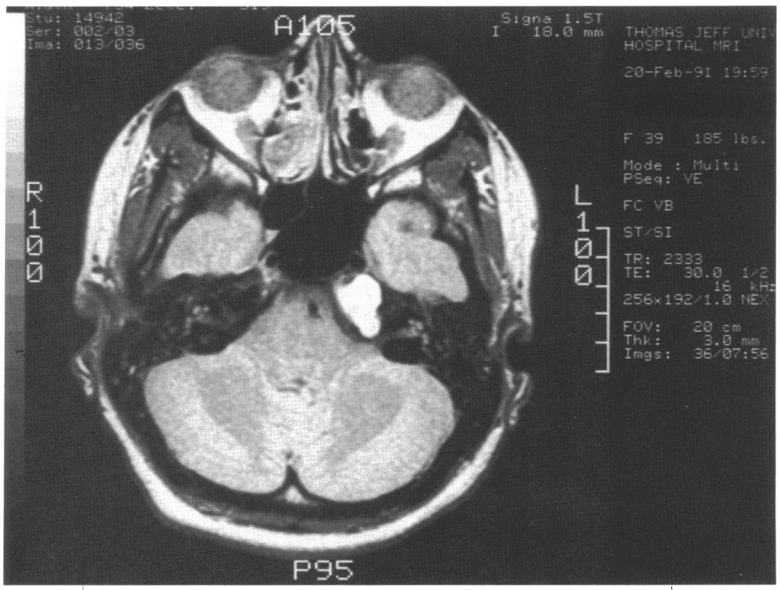

Figure 4. Axial double-echo proton density T_2-weighted image (TR = 2333 msec, TE = 30 msec) shows the mass to have high signal intensity and slight inhomogeneity.

sequences. The first and second echo images are generated by increasing the echo time (TE) from 30 to 90 msec while keeping the pulse repetition time long at greater than 2000 msec. This is different from the MRI characteristics of a cholesteatoma, where the squamous debris slows low intensity on T-weighted images and high intensity on T_2-weighted images.[11] Meningiomas show immediate intensity on T_1-weighted MR imaging with intense enhancement after gadolinium administration. Linearly enhancing lesions adjacent to the mass represent the dural tail sign frequently seen with meningiomas. Schwannomas have MR characteristics similar to meningiomas, but they have no tail sign. Cerebral angiography or MR angiography is most helpful in studying vascular lesions such as carotid aneurysms and glomus tumors. They can also provide a "road map" of the vessels supplying and surrounding the lesion (Table 1).

CT scan, MR imaging, and angiography along with history and physical examination frequently narrows the differential diagnosis to permit the exclusion of congenital cholesteatomas, intratemporal schwannomas, meningiomas, apical petrous abscesses, carotid aneurysms, glomus tumors, metastasis, lymphomas, chordomas, and histiocytosis.

Histologic examination shows GCC to have a fibrous wall with no endothelial or epithelium lining. The cyst contains a thin, brownish fluid with cholesterol crystals. The etiology of these unusual lesions is unknown. No

Figure 5. Axial double-echo T_2-weighted image (TR = 2333 msec, TE = 90 msec) shows inhomogeneous mass with high, but decreasing signal intensity.

Table 1. Differential Diagnosis

Giant cholesterol cyst	Petrous apex abscess
Cholesterol granuloma	Lymphoma
Cholesteatoma	Chordoma
Schwannoma	Metastasis
Meningioma	Carotid aneurysms
Glomus neoplasm	

theories regarding their origin appear in the literature. GCC do appear to originate only in a well-pneumatized petrous apex.

Graham et al[1] made the distinction between GCC and cholesterol granuloma. On histologic examination, cholesterol granuloma has cholesterol crystals surrounded by multinucleated giant cells, macrophages, and round cells. The fluid is thick and the mucosa adjacent to the granuloma undergoes metaplasia to ciliated columnar respiratory epithelium. Cholesterol granulomas most commonly occur within the middle ear or mastoid.

Treatment

Surgical management of GCC consists of drainage and exteriorization of the cyst. The fibrous lining of the cyst is occasionally adherent to the carotid artery. Consequently, most authors recommend draining the cyst without complete removal of the fibrous lining, which can cause significant morbidity and mortality.

The literature reports five approaches to GCC of the petrous apex: middle fossa, posterior fossa, transmastoid, translabyrinthine, and external ethmoid-transsphenoidal. The choice of approach should be based on the following factors:

1. Status of hearing in both ears
2. Pneumatization of the mastoid cavity and petrous apex
3. Location of the internal carotid artery relative to the GCC
4. Size and pneumatization of the sphenoid sinus
5. Ability to achieve a permanent drainage

The middle fossa approach is an extradural temporal craniotomy that provides access to lesions that extend anteriorly to the cochlea and to the internal carotid artery in the petrous apex. This approach is excellent for hearing preservation and allows the diagnosis and drainage of GCC. Permanent drainage is more challenging and requires the removal of the tegmen tympani and placement of tube or stent from the cyst into the middle ear or mastoid cavity. This fistula has a high rate of stenosis due to the small size of the stent, nondependent drainage, and long route. Sataloff et al[2] performed an eustachian tuboplasty that removes the superior wall of the eustachian tube anteriorly. A stent is placed from the cyst into the eustachian tube. The stent length is shorter and the drainage route is gravity dependent, theoretically decreasing the chance of obstruction and stenosis.

The posterior fossa approach (suboccipital craniotomy) permits biopsy and drainage of GCC with hearing preservation. This approach is limited to posterior lesions and a permanent drainage route is difficult to establish. Access to the anterior petrous apex is limited and this approach can be combined with other approaches to increase exposure.

The classic approach to the petrous apex is through the mastoid cavity. Lindsay[12,13] in two landmark articles described the anatomy of the petrous pyramid and the surgical management of petrous apicitis. Hearing is preserved and drainage of the petrous apex is accomplished. However, biopsy of the cyst wall is difficult and creating a permanent tract from the cyst to the mastoid cavity is challenging. Obviously, draining purulent material filling a well-pneumatized apex is much different from creating a permanent drainage tract to the mastoid from a discrete cyst within the apex.

The translabyrinthine, transcochlear approach allows for wide exposure and drainage of GCC with the sacrifice of hearing and vestibular function. Preoperative hearing in both ears is obviously important.

Montgomery[7] described the external ethmoid, transsphenoidal approach to cystic lesions of the petrous apex. He described a contralateral external ethmoidectomy and an ipsilateral sphenoidotomy with drainage of the cyst into the nasal cavity. Early stenosis of the drainage tract prompted the addition of septal mucosal flaps and a silicone stent, decreasing the incidence of stenosis.

The role of endoscopic surgery has expanded dramatically in the past several years. Messerklinger is credited with pioneering endoscopy in sinonasal disease.[14] Endoscopic ethmoidectomy and sphenoidotomy have become routine surgical procedures.[15] Advancements in light sources, light transmission, endoscopes, and television monitors coupled with an increase in experience have made the use of endoscopes routine in the treatment of inflammatory and neoplastic disorders of the nose and paranasal sinuses. The uses have expanded as the techniques are refined and the anatomy elucidated. Stankiewicz[10] reported on the use of endoscopic techniques to close cerebrospinal fluid leaks of the anterior skull base. Metson[8] reported on the efficacy of endoscopic dacryocystorhinostomy. Goodstein and Kennedy[9] reported on using endoscopic techniques for orbital decompression in dysthyroid orbitopathy.

This article reports the use of endoscopes to treat cystic lesions of the petrous apex transnasally. This technique can be used to treat lesions that meet specific anatomic criteria. These criteria include:

1. Lesion is located immediately adjacent to the sphenoid or posterior ethmoid sinus
2. Well-developed sphenoid sinus
3. Safe location of neurovascular structures such as the internal carotid artery and the optic nerve relative to the lesion

The use of this technique can be fraught with danger. There is significant potential for hemorrhage, cerebrospinal fluid leak, and damage to vital neurologic structures.[16–18] The indentation of the carotid artery in the wall of the sphenoid sinus must be identified before the posterior wall of the sinus is removed.[19] The carotid artery is the lateral border of the dissection and its wall in the temporal bone is on 0.15 mm thick or one seventh of its wall thickness within the neck.[20] Cross-table radiography with positioning probes should be used to check and recheck where the cyst will be entered.[21] A clear operative field is mandatory. The cyst should be opened widely and stented because the tract has a high rate of stenosis. The position of the stent can be checked by cross-table radiography and followed in the office by x-rays.

SUMMARY

GCC is a rare lesion of the temporal bone that can cause neurologic deficits and vascular compromise by continued growth and bone destruction. Surgical management of GCC consists of drainage and exteriorization. Middle fossa, posterior fossa, transmastoid, translabyrinthine, and external ethmoid/transsphenoidal approaches to these lesions have been described. We report the endoscopic, endonasal transsphenoidal approach for the treatment of a 2.5 cm GCC of the petrous apex. In carefully selected cases, this approach can provide adequate surgical exposure with minimal morbidity.

REFERENCES

1. Graham MD, Kemink JL, Latack JT, Kartush JM: The giant cholesterol cyst of the petrous apex: A distinct clinical entity. Laryngoscope 95:1401–1406, 1985
2. Sataloff RT, Myers DL, Roberts BR, et al: Giant cholesterol cysts of the petrous apex. Arch Otolaryngol Head Neck Surg 114:451–453, 1988
3. Waltner JG, Karatay S: Cysts of the mastoid bone. Arch Otolaryngol 46:398–404, 1947
4. Gacek RR: Diagnosis and management of primary tumors of the petrous apex. Ann Otol Rhinol Laryngol 84 (Suppl):18, 1975
5. Hendershot EL, Wood JW, Bennhoff D: The middle cranial fossa approach to the petrous apex. Laryngoscope 86:658–663, 1976
6. DeLozier HL, Parkins CW, Gacek RR: Mucocele of the petrous apex. J Laryngol Otol 93:177–180, 1979
7. Montgomery WW: Cystic lesions of the petrous apex: Transsphenoid approach. Ann Otol 86:429–435, 1977
8. Metson R: Endoscopic surgery for lacrimal obstruction. Otolaryngol Head Neck Surg 104:473–479, 1991
9. Goodstein ML, Kennedy DW: An endoscopic approach for orbital decompression in dysthyroid orbitopathy. Operative Techniques Otolaryngol Head Neck Surg 1:117–125, 1990
10. Stankiewicz J: Cerebrospinal fluid fistula and endoscopic sinus surgery. Laryngoscope 101:250–256, 1991
11. Rao VM, Flanders AE, Tom BM: MRI and CT Atlas of Correlative Imaging in Otolaryngology. Norwalk, CT: Martin Dunitz, 1992
12. Lindsay JR: Petrous pyramid of temporal bone: Pneumatization and roentgenologic appearance. Arch Otolaryngol 31:231–235, 1940
13. Lindsay JR: Suppuration in the petrous pyramid. Ann Otol Rhinol Laryngol 47:3–36, 1938
14. Stammberger H: History of nasal endoscopy. In: Stammberger H: Functional Endoscopic Sinus Surgery. Philadelphia: BC Decker, 1991, pp 1–15
15. Schaefer SD: Endoscopic total sphenoethmoidectomy. Otolaryngol Clin North Am 22:727–732, 1989
16. Stankiewicz JA: Complications in endoscopic intranasal ethmoidectomy. Laryngoscope 97:1270–1273, 1987
17. Stankiewicz JA: Complication in endoscopic intranasal ethmoidectomy: An update. Laryngoscope 99:686–690, 1989
18. Stammberger H: Results, problems, and complications. In Stammberger H: Functional Endoscopic Sinus Surgery. Philadelphia: BC Decker, 1991, pp 459–477
19. Kennedy D, Zinreich S, Hassab M: The internal carotid artery as it relates to endonasal sphenoethmoidectomy. Am J Rhinol 4:7–12, 1990
20. Chole RA: Petrous apicitis: Surgical anatomy. Ann Otol Rhinol Laryngol 94:251–257, 1985
21. May M, Master SJ, O'Daniel TG, Curtin HD: Decreasing the risks of endonasal endoscopic sinus surgery by imaging techniques. Operative Techniques Otolaryngol Head Neck Surg 1:89–91, 1990

REVIEWER'S COMMENTS

It was a pleasure to review the manuscript "Endoscopic Management of a Giant Cholesterol Cyst of the Petrous Apex." The authors are to be congratulated on their thoughtful application of endoscopic technology to this challenging problem in the petrous apex, which can be difficult to reach regardless of which approach is used.

The concept of draining cholesterol cysts (and potentially, other lesions of the petrous apex as well) into the sinonasal tract through the sphenoid sinus is logical and attractive for a number of reasons. The proximity of the petrous apex to the well-pneumatized sphenoid sinus makes the transsphenoidal approach the most direct avenue for the surgeon to reach the pathology in process. The possibility for eventual or immediate mucosal lining of the endoscopically created fistula tract with respiratory epithelium may enhance long-term control of such lesions. The ease of inspection and cleansing of the postoperative cavity (using endoscopes in the outpatient setting) is certainly an advantage.

The authors rightly point out the risks of the endoscopic approach to the petrous apex, and these are not to be taken lightly. Carotid artery injury within the sphenoid sinus or petrous apex constitutes a major disaster that must obviously be avoided, since it is unlikely that such an injury could be successfully managed by endoscopic means using current technology. The potential is

also very real for cerebrospinal fluid fistula, meningitis, and injury to the brain, adjacent cranial nerves, and pituitary gland if the endoscopic instruments are misplaced or misdirected.

Because of these safety concerns, a number of precautions must be recommended. Without question, this kind of endoscopic surgery should be done only by surgeons who have mastered the technology and who are also familiar with alternative approaches to the petrous apex. As presented here, highly detailed preoperative imaging studies play a critical role in the selection of appropriate candidates for this approach. They also serve to provide a "road map" to the unique anatomic and pathologic variations in each case and, as such, are indispensable. Maintenance of a clear operative field is vital, since even small amounts of bleeding can ruin the visualization—and surgical orientation—through the endoscope. The lateral wall of the sphenoid sinus has to be manipulated with extreme caution to prevent uncontrolled fracture and injury to nearby structures. Finally, the surgeon must be willing to abandon the endoscopic approach and use another technique if the exposure is not satisfactory.

These precautions acknowledged, the endoscopic approach described here offers a very valuable alternative for managing selected lesions of the petrous apex, with potentially much less morbidity than some of the traditional approaches.

Daniel W. Nuss, M.D.