The position of intraconal tumors relative to the optic nerve dictates the choice of surgical approach. Tumors with their epicenter medial to the optic nerve or “below a plane of resectability” (POR), which represents a plane subtended by the contralateral nostril and the long axis of the optic nerve, are amenable to an endoscopic approach. The feasibility and safety of this approach has been demonstrated in the literature. Tumors located lateral and superior to the POR are not candidates for an exclusively endoscopic resection.

The composition of the surgical team varies depending on the institution. However, a multidisciplinary team including an otolaryngologist, oculoplastic surgeon and in some cases, a neurosurgeon, is invaluable.

The endoscopic approach to intraconal tumors will be described in this chapter using orbital cavernous haemangioma (OCH) as an example, as it is the most common primary orbital tumor in adults, with a reported incidence of 5-15% of all orbital tumors (Figure 1).

Orbital cavernous haemangiomas (OCH)

The International Society for the Study of Vascular Anomalies classifies OCHs as slow-flow cavernous venous malformations. OCHs are more common in women and occur in the 4th and 5th decades of life1. The natural history remains elusive. Progesterone may play a role in the clinical course which may explain sudden growth of OCH seen during pregnancy and a reduction in size or stabilisation in postmenopausal women; however, the exact role of progesterone is yet to be fully elucidated.

A significant number of OCHs present as asymptomatic lesions incidentally found on CT or MRI performed for unrelated reasons. Previous studies have shown that asymptomatic lesions often show no progression 2,3. In many cases if an incidental OCH does not change over several years, it is unlikely to do so over more prolonged periods of followup.4

The majority are located between the optic nerve and the extraocular muscles and are therefore intraconal. It is well documented that OCH have a predilection for the intraconal space. The single most common anatomical location is lateral to the optic nerve, which may reflect the relationship between the optic nerve and the distribution of the ophthalmic vasculature. However, OCH can be found throughout the orbit, including the medial intraconal space, extraconal space, and within the optic canal.5 Rarely, they extend beyond the confines of the orbit into the pterygopalatine fossa,6 cavernous sinus,7 and intracranial space.8
Histologically OCHs appear as cavernous venous malformations and have exclusively venous vasculature. Despite suggestion of arterial flow on imaging studies, histologically there is no evidence of the elastic lamina associated with arterioles. The thicker walled vessels are thought to be the result of thrombosis and recanalisation. If any arterial component is present, it is thought to be inconsequential both histologically and clinically. OCHs are characterised as lesions with mature cellular components and do not tend toward dysplasia or hypercellularity. Although they can incorporate surrounding vessels and nerves, they do not typically infiltrate into surrounding tissue. OCHs tend to be slow growing vascular lesions with a radiological growth rate of 10-15% per year. The growth of OCH is thought to be a cycle of stasis and thrombosis with endothelial cellular proliferation and recanalisation into multiple clefts and vascular channels.

They have a firm fibrous capsule that makes them amendable to endoscopic resection. Surgical resection is indicated for symptomatic lesions whilst smaller asymptomatic lesions can be observed. The goal of surgery is definitive resection. However, given its benign nature, complete resection must be balanced against iatrogenic morbidity. Partial resection of intraconal OCH has been reported, but long term outcomes remain unknown.

Surgical approach

The position of intraconal OCHs within the orbit relative to the optic nerve dictates the choice of approach. OCHs with an epicenter medial to the optic nerve or “below a plane of resectability” (POR), which represents a plane subtended by the contralateral nostril and the long axis of the optic nerve, are amenable to an endoscopic approach. The feasibility and safety of this approach has been consistently demonstrated. OCHs located lateral and superior to the POR are not candidates for an exclusively endoscopic resection.

Surgical anatomy

Knowledge of the neurovascular anatomy within the orbit is crucial and directs safe placement of a ball probe on the medial rectus and the degree of retraction. A branch of the oculomotor nerve penetrates the medial rectus one-third of the distance from the annulus of Zinn to its insertion onto the globe (Figure 2); therefore, direct or prolonged traction here should be avoided.
Operating room set-up

*Figure 3* demonstrates a typical layout of the operating room. This allows for a binarial, four-handed transseptal approach that is commonly used for intraconal OCH resection. With a binarial approach, a posterior septectomy or septal window is required. This has minimal morbidity, allows manoeuvering of endoscopes and other instruments, and may be integrated into elevation of a nasoseptal flap for medial wall reconstruction.

*Figure 3*: The operating room setup for a binarial four-handed 2 surgeon transseptal approach. The endoscope is held by surgeon 2 (in this case for a left-sided tumour; the surgeons can be reversed for right-sided tumours). Surgeon 1 performs the dissection and retraction.

**Essential instrumentation** (*Figure 4*)

- Rigid endoscopes:
  - 0-degree
  - Angulated 30, 45 and 70-degree
- Pledgets soaked in saline
- Suction cannulae
  - Frazier size 9 and 10
  - Curved olive tip suction
- Medial rectus retractor
- Penfield dissector Number 4
- Lusk ball tip probe

**Figure 4**: (A) Penfield dissector number 4; (B) Curved olive tip suction; (C) Frazier suction cannula; (D) Lusk ball tip probe

**Surgical Steps**

1. **Anaesthesia and positioning**

   The patient is placed supine in a head ring with the head of the bed raised 15-30 degrees. Total intravenous anaesthesia is used. The patient is intubated with the tube positioned to the left. The room is set up as seen in *Figure 3*.

2. **Preparation of nasal cavity**

   The nasal cavity is prepared with cottonoid pledgets soaked in 1:1000 adrenaline. This is then changed to saline soaked pledgets once the periorbita are exposed. The nasal cavity is infiltrated with 10mls of lignocaine 1% with 1:100,000 adrenaline.

3. **Create a working space and define orbital wall and orbital axis**

   This is a fundamental part of the surgery and requires the following
   - Complete uncinectomy
   - Wide maxillary antrostomy
   - Sphenoethmoidectomy
   - The middle turbinate may be resected to improve access and visibility
4. Reflect the mucosa from medial to lateral off the posterior wall of the maxillary antrum.

- Use the Lusk ball probe to assist with removal of the maxillary sinus mucosa and place a pledget to hold the mucosa out of the operating field (Figure 5).

4. Posterior septectomy & nasoseptal flap

- The need for a posterior septectomy vs. a septal window with reconstruction is based on the tumour location and extent of the dissection required.
- Raise the nasoseptal flap on the contralateral side of the nose.
- Tailor the size of the flap to the individual case.
- Begin the incision inferiorly, starting at the anterior inferior choana and sweeping anteriorly and inferiorly towards the floor of the nose.
- Carry this incision anteriorly along the floor of the nose, and tailor it to the anticipated defect of the exposed lamina papyracea.

- Raise the anterior flap by blunt dissection with a Cottle elevator to the sphenoid ostium.
- Make the superior incision with cold steel dissection.
- Tuck the flap into the nasopharynx until completion of the resection.
- The septectomy is then made posteriorly in a location that will permit unrestricted passage of instruments.

5. Posterior maxillary wall and orbital process of palatine bone

- The ipsilateral posterior maxillary wall and the orbital process of the palatine bone is then thinned using a 15-degree diamond burr (Figures 7, 8).
- Removing the orbital process provides an additional 0.36+/−0.42 cm$^3$ of exposure in the inferolateral vector.\textsuperscript{23}

\begin{figure}[h]
\centering
\includegraphics[width=0.4\textwidth]{figure5.png}
\caption{Placement of cottonoid to retract left posterior maxillary mucosa. (A) marks the maxillary sinus with the mucosa elevated to expose the bone of the posterior maxillary wall; (B) indicates the cottonoid placed between the mucosa and posterior maxillary wall to assist with the dissection and securing and protecting the mucosa.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=0.4\textwidth]{figure6.png}
\caption{Nasal septal flap (outlined) and posterior septectomy viewed from right (contralateral) nose.}
\end{figure}
6. **Exposing periorbita (Figures 9, 10)**

- Remove the lamina papyracea with blunt dissection to expose the underlying periorbita

- If necessary, the optic strut may be drilled to facilitate safe removal of the bone overlying the optic nerve (*Figure 10*)

7. **Retraction of medial rectus**

- Create a periorbital window to identify the extraconal fat and muscles
• Incise the periorbita in a reverse hockey stick fashion using a sickle knife (Figure 11)
• Place the incision just anterior to the border of the tumour to prevent unnecessary fat prolapse into the anterior surgical field

![Figure 11: Incision line of the periorbita in a reverse hockey stick fashion. This can be performed with a sickle knife](image)

- The **intraconal dissection corridor** is bordered by the medial rectus muscle above and the inferior rectus muscle below (Figure 12)
- Use a dissector e.g. Penfield or Lusk probe to bluntly dissect between the medial and inferior rectus muscles and to visualise the intraconal space
- The medial rectus serves as a landmark of the medial orbit and must be retracted to access intraconal OCHs
- Retract the medial rectus with care using a right-angled instrument under the inferior border of the muscle and gently and dynamically retract the muscle in a superomedial direction to prevent postoperative diplopia

![Figure 12: Medial rectus as a landmark of the medial orbit retracted to enter the intraconal space](image)

8. Haemostasis

Judicious and precise use of bipolar cautery, saline soaked cottonoid pledges, and warm water irrigation are relatively safe. Avoid monopolar cautery within the orbit due to the significant risk of thermal injury to important neural structures.

9. Orbital fat

Fat herniation can be troublesome and keeping the intraconal dissection corridor open is challenging. Preserving extraconal fat not only helps to preserve orbital volume, but also minimises the risk of medial rectus scarring and entrapment. Proper placement of the periorbital incision as previously described and the use of cottonoid pledges to separate and retract the orbital fat can assist with visualisation and should avert the need to remove extraconal fat (Figure 13).
10. Resection techniques

- The fibrous capsule permits dissection in an extracapsular plane with preservation of the capsule
- A process of gentle traction, use of cottonoids, and blunt dissection is used

- Sharp cutting instruments have also been used but do increase the risk of bleeding from inadvertent arteriolar injury
- Incomplete resection has been documented in cases where the lesion is adherent to the optic nerve, although long-term follow-up results are unknown

11. Reconstructing medial orbital wall

- Placement of orbital fat over the exposed extraocular muscles should be considered to prevent scarring (Figure 15)

- Reconstructing the medial orbital wall to preserve orbital volume should be strongly considered following removal of intraconal or large extraconal lesions
- There is no clear consensus on the best method of reconstruction
- A pedicled nasoseptal flap technique is preferred by the authors as it provides the opportunity for delayed contraction thereby reducing the risk of diplopia and enophthalmos (Figure 16)
- Immediate rigid reconstruction does place the orbit at risk for compartment
syndrome secondary to oedema and postoperative bleeding (Figure 17)

Figure 16: The pedicled nasoseptal flap draped along the medial orbital wall

Figure 17: A left compartment syndrome with an IOP of 55mmHg

12. Nasal packing

There is lack of consensus on the use of post-surgery nasal packing. Nasal packing is not recommended by some studies, citing possible exertion of pressure on the globe or optic nerve.\textsuperscript{12,21} Alternatively, absorbable haemostatic packing has been used\textsuperscript{27} and the use of a polyvinyl acetate sponge placed in the nasal cavity to assist with adherence and positioning of a nasoseptal flap for 7 days has been reported. However, packing should not be placed in a position which could risk exerting direct pressure on the exposed orbit.

Figure 18a: A T2-weighted MRI coronal scan demonstrating a left orbital haemangioma

Figure 18b: A T2-weighted MRI coronal scan demonstrating postoperative situation with a left nasoseptal flap positioned along the medial orbital wall

References


**Authors**

Catherine Banks MBChB FRACS

Department of Otolaryngology Head & Neck Surgery.

Rhinology Fellow

Massachusetts Eye and Ear Infirmary/ Harvard, Boston MA 02114 USA
catherinebanks@MEEI.HARVARD.edu

Benjamin Bleier MD FACS

Associate Professor of Otolaryngology

Massachusetts Eye & Ear Infirmary/ Harvard, Boston MA 02114 USA
Benjamin_Bleier@MEEI.Harvard.edu

**Editor**

Johan Fagan MBChB, FCS (ORL), MMed

Professor and Chairman

Division of Otolaryngology

University of Cape Town

Cape Town, South Africa

johannes.fagan@uct.ac.za

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