Surgery for Exophthalmos

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Grand Rounds Presentation
April 7, 2004
Introduction

- Exophthalmos: Discordant relationship between anterior orbit and globe
- Orbital volume fixed = 30 ml
- Graves orbitopathy the most common cause of bilateral exophthalmos; up to 20% of cases are unilateral
Graves Orbitopathy

- Middle-aged women
- Female:male ratio = 5:1
- Peak incidence in 3rd and 4th decades of life
- 6 times more common in Caucasians
- NOT necessarily associated with hyperthyroidism; 20% of Graves orbitopathy patients are euthyroid
Graves Orbitopathy

- Arises from autoimmune dysfunction (primarily T-cell)
- Lymphocyte infiltration and immune complex deposition produce fibroblast proliferation, glycosaminoglycans, collagen, and edema
- Noninfiltrative changes include spastic eyelid retraction, sympathetic hypertonia
Graves Orbitopathy

- **Findings:** Thickening of external ocular muscles, orbital fat herniation, proptosis, descent of eyelid-cheek complex, divergence of gaze, eyelid edema, conjunctivitis, chemosis, lagophthalmos, epiphora, optic neuropathy (stretch vs. compression)

- **Symptoms:** Photophobia, headache, gritty sensation in eye, retrobulbar pain, diplopia (usually involving inferior and medial rectus; check upward and lateral gaze)
Graves Orbitopathy
Graves Orbitopathy

- If left untreated, can result in corneal exposure/injury, and optic neuropathy
- 5% of Graves orbitopathy patients warrant surgical intervention
## Exophthalmos

<table>
<thead>
<tr>
<th>Cause of relative proptosis</th>
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<tbody>
<tr>
<td>Thyroid related orbitopathy</td>
<td>10</td>
<td>44</td>
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<tr>
<td>Relative proptosis, hypoplastic malar eminence</td>
<td>4</td>
<td>17</td>
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<td>Postblepharoplasty lagophthalmos and tearing</td>
<td>3</td>
<td>14</td>
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<tr>
<td>Facial trauma</td>
<td>2</td>
<td>9</td>
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<tr>
<td>Primary exorbitism (shallow orbits)</td>
<td>1</td>
<td>4</td>
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<td>High myopia</td>
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<td>4</td>
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<td>NF-1 (absent sphenoid wing)</td>
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<td>4</td>
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<tr>
<td>Involutional ectropion after orbital tumor removal</td>
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Patient Evaluation

- Physical exam
  - Hyperemia over lateral rectus muscle pathognomonic of Graves orbitopathy
  - Divergence of globes on extreme gaze (usually looking up)
  - Nasal endoscopy
  - Palpation of thyroid gland
Patient Evaluation
Patient Evaluation

- Complete ophthalmologic exam
  - Hertel exophthalmometry
  - Visual acuity, visual fields, color saturation (optic neuropathy)

- Radiographs
  - CT or MRI to include orbits AND sinuses; rule out orbital mass, sinusitis, hypoplastic maxillary sinus
  - T2 relaxation time: If prolonged, suggests good response to steroids

- Labs
  - T3, FT4I, TSH, TRH, TSI
Patient Evaluation
Differential Diagnosis

- Pseudotumor cerebri – second-most common cause of bilateral exophthalmos; improves with steroids
- Meningioma en plaque – lower eyelid edema *without* lid retraction
- Axial myopia – unilateral exophthalmos; dx via retinoscopy and A-scan ultrasound
- Inflammatory pseudotumor – mimics neoplasm; responds to steroids
- Lymphoma – eccentric proptosis
- Shallow orbit (e.g., Apert, Crouzon)
### Treatment

- **Medical:** I-131, levothyroxine to achieve euthyroid state
  - Optic neuropathy: 80-120 mg/day prednisone X 2 weeks; if prolonged use or no response then decompress

- **Radiation:** not appropriate in acute or subacute setting due to early edema; 200 cGy over 2 weeks; resolution is rare; must be certain patient not to undergo orbital surgery

- **Immunomodulation:** unproven
Surgery

- Goal of orbital decompression = enlarge orbit via removal of 1-4 walls with incision of periosteum to allow for prolapse of orbital soft tissues into adjacent spaces
- Up to 15 mm of recession possible
- Indications in acute/subacute phase: optic neuropathy, steroids fail to improve vision or steroids required for long-term maintenance, corneal exposure/keratitis
- Late indications: cosmesis
Surgery

- Ideally performed after orbital findings AND thyroid status have stabilized for 6 months
- Few patients are satisfied with initial procedure
- Optimal order of surgery:
  - Orbital decompression
  - Strabismus surgery (for correction of diplopia)
  - Lid lengthening
- 8-12 mg of Decadron intraoperatively
Surgery

- Superior orbital decompression (Naffziger)
  - Requires frontal craniotomy (neurosurgery)
  - Titanium mesh to prevent transmission of pulsations to globe
  - Very large amount of orbital bone can be removed
  - Uncommon; most frequently used in setting of trauma
Medial Decompression (Sewell)

- Coronal incision or Lynch incision
- Medial canthal tendon (unless coronal approach) and anterior ethmoid a. divided; posterior ethmoid a. identified
Surgery

- Inferior Decompression (Hisch and Urbanek)
  - Creation of orbital blowout fx with sparing of infraorbital nerve
  - Transconjunctival or subciliary incision + Caldwell-Luc antrostomy
  - Total of 3 cm of bone from anterior to posterior adequate and safe
  - Should perform forced duction at conclusion to ensure medial rectus not entrapped.
Surgery

- Lateral decompression (Kronlein)
  - First technique to be described (1911)
  - Approached via coronal incision, direct rim incision, lateral extension of subciliary/transconjunctival incision, extended lateral canthotomy, or upper lid crease incision with extension along laugh line
  - If combined procedure, best performed last
  - Can retract orbital contents medially
  - CSF leak common complication
Surgery

- Combined medial and inferior decompression (Walsh-Ogura)
  - Necessitates Caldwell-Luc antrostomy
  - High incidence of OAF
  - Technique of choice for orbital decompression until 1990’s with endoscopic technique
Endoscopic Orbital Decompression

- Advantages: no external incision, limited morbidity, excellent access to optic nerve and orbital apex when needed, possible to do under local, no risk of oroantral fistula
- Eyes must be included in surgical field; usually protect with corneal shields
- Uncinectomy, large middle meatal antrostomy, total ethmoidectomy, sphenoidotomy
- Identify infraorbital nerve in roof of maxillary sinus
- Resection of middle turbinate controversial
Endoscopic Orbital Decompression

- Retain small piece of lamina papyracea in frontal recess area to prevent prolapsing fat from obstructing frontal sinus.

- Leave periorbita intact until all bone is removed.

- Remove bone from orbital floor MEDIAL to infraorbital nerve (bone typically fractures naturally at canal).

- Incise periorbita with sickle knife; must keep blade superficial.

- Gentle pressure on orbit to extrude fat.
Endoscopic Orbital Decompression
Endoscopic Orbital Decompression

- Capable of correcting 3.5 mm of exophthalmos
- Can correct 5.4 mm if combined with lateral decompression
- Postoperatively check visual acuity and EOM
- Avoid nasal packing
- Avoid nose blowing for 2 weeks
- Sinonasal exam/cleanings per routine
- Bilateral orbital decompressions at 1 week intervals
Surgery: Other Options

- Orbital fat removal
  - Preoperative CT to locate orbital fat pockets
  - Upper lid crease + subciliary/transconjunctival incision
  - Open orbital septum in BOTH upper and lower lids longitudinally
  - Up to 6 mm of proptosis correctable
  - NOT adequate in setting of optic neuropathy
  - Suboptimal with significant EOM involvement
Surgery: Other Options

- Orbital rim advancement (Goldberg)
  - Onlay grafts (porous polyethylene) to inferolateral orbital rim
  - Transconjunctival approach
  - Suspension of suborbital orbicularis oculi fat (SOOF) from implant; corrects descent of eyelid-cheek complex
  - Best for woody orbits, shallow orbits
  - Frequently in combination with decompression
Surgery: Other Options
Results and Complications
Results and Complications

- 75% of patients stabilize or improve with surgery
- Corneal abrasion, retrobulbar hematoma, temporary V2 numbness, retinal hemorrhage (diabetics), orbital cellulitis, retinal vascular occlusion, DIPLOPIA
- Retrobulbar hematoma
  - Avoid with meticulous hemostasis w/ bipolar and Penrose drain
  - If it occurs open skin incision, evacuate hematoma, irrigate, bipolar for hemostasis, place Penrose
Results and Complications

Retinal vascular occlusion – heralded by pain in eye and/or decreased vision; an emergency; ophthalmology consult required; must warn patient to seek immediate help for pain in eye/decreased vision on d/c
Diplopia

- Up to 50% of patients with some degree of post-op diplopia
- Usually medial and inferior rectus mm.
- Many patients experience resolution of diplopia with resolution of inflammation, or diplopia only on peripheral gaze
- If diplopia persists 6-8 months after decompression, must be evaluated and treated by ophthalmologist
Diplopia

- Options to prevent diplopia
  - Orbital lipectomy rather than decompression
  - Preservation of bony strut in lamina (technically difficult)
  - Lateral decompression only
  - "Balanced" decompression with endoscopic and lateral approaches
  - Preservation of periorbital sling (Metson)
Diplopia
Conclusion

- Graves Orbitopathy does not correlate with thyroid status
- Though an autoimmune process, treatment remains primarily surgical
- Treatment multidisciplinary: endocrinologist, ophthalmologist, otolaryngologist
- Preoperative imaging to include sinuses
- Endoscopic orbital decompression and/or lateral decompression
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