Orbita

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Anatomy
Anatomy

- Orbita is a pear-shaped cavity, the stalk of this pear is the optic canal

- Within the orbita the length of the optic nerve (25 mm) is longer than the distance between the globe and the optic canal (18 mm)

- This allows for significant forward displacement of the globe (proptosis) without excessive stretching of the optic nerve
Clinical Signs in Orbital Diseases
Clinical Signs in Orbital Diseases

- Soft Tissue Involvement
- Proptosis
- Enophthalmos
- Dystopia
- Ophthalmoplegia
- Dynamic Properties
- Fundus Changes
Clinical Signs in Orbital Diseases

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Soft Tissue Involvement

**Signs**
- lid and periorbital oedema
- ptosis
- chemosis (oedema of the conjunctiva and caruncle)
- epibulbar injection

**Common Causes**
- thyroid eye disease
- orbital inflammatory diseases
- obstruction to venous drainage
Clinical Signs in Orbital Diseases

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Proptosis

Abnormal protrusion of the globe (which may be caused by retrobulbar lesions)

• Axial proptosis

• Eccentric proptosis

• Asymmetrical proptosis
  – is best detected by looking down at the patient from above and behind
Proptosis

Hertel exophthalmometer

- Corneal apexes are visualized in mirrors and degree of ocular protrusion is read from a scale

- Reading greater than 20 mm are indicative of proptosis

- Difference of 2 mm between the two eyes is suspicious
Proptosis

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Proptosis

Pseudo-proptosis (false impression of proptosis)

- Facial asymmetry
- Severe ipsilateral enlargement of the globe
  - high myopia
  - buphthalmos
- Ipsilateral lid retraction
- Contralateral enophthalmos
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Enophthalmos

- Implies recession of the globe within the orbit:
  - Structural abnormalities of the orbital walls
    - post-traumatic, such as blow-out fractures of the orbital floor, congenital, or inflammatory, silent sinus syndrome
  - Atrophy of the orbital contents
    - radiotherapy, scleroderma, or eye poking (oculodigital sign) in blind infants
  - Sclerosing lesions of the orbita
    - schirrous carcinoma, chronic inflammatory orbital disease

- Pseudo-enophthalmos may be caused by phthisis bulbi or micro-phthalmos
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Dystopia

• Globe displacement in the **coronal plane**
  – usually extra-conal orbital mass (lacrimal gland tumor)

• Horizontal dystopia
  – distance from midline (nose) to the nasal limbus

• Vertical dystopia
  – distance along vertical scale perpendicular to a horizontal line placed over the bridge of the nose
Dystopia

• Globe displacement in the coronal (frontal) plane
  – usually extra-conal orbital mass (lacrimal gland tumor)

• Horizontal dystopia
  – distance from midline (nose) to the nasal limbus

• Vertical dystopia
  – distance along vertical scale perpendicular to a horizontal line placed over the bridge of the nose
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Ophthalmoplegia

- Is a paralysis or weakness of one or more of the muscles that control the eyes movements

Common Causes
- *An orbital mass*
- *A restrictive myopathy*
  - thyroid eye disease
  - orbital myositis
Ophthalmoplegia

- **Oculomotor nerve (third cranial nerve) involvement in the:**
  - cavernous sinus
    - carotid-cavernous fisutla
  - superior orbital fissure
    - Tolosa-Hunt syndrome
  - posterior orbit
    - malignant lacrimal gland tumor

- **Incarceration** of extraocular muscles or fascia in a blowout fracture
Ophthalmoplegia

Restrictive versus Neurological

• **Forced duction test**
  – the insertion muscle is grasped with forceps and the globe rotated in the direction of limited mobility
  – the test is repeated in the unaffected eye
  – it can be painful, therefore a cotton pledged soaked with anesthetic should be applied on muscles to be tested (5 minutes)

• **Positive result:**
  – difficulty to move the eye globe indicates a restrictive problem

• **Negative result:**
  – no resistance suggests a neurological cause
Ophthalmoplegia

Right eye is in primary adduction deviation

In dextroversion, there is limited abduction

Mechanical restriction: Test is positive

No mechanical restriction: Test is negative
Ophthalmoplegia

**Restrictive versus Neurological**

- **Differential intraocular pressure test**
  - IOP in the primary position of gaze
  - IOP when patient looks into the direction of limited mobility
  - less discomfort and end-point more objective than forced duction

- **Positive result:**
  - ≥ 6 mm Hg increase, compression transmitted by muscle, suggests restriction

- **Negative result:**
  - < 6 mm Hg suggests a neurological lesion
Clinical Signs in Orbital Diseases

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Dynamic Properties

- **Increasing venous pressure** may induce or exacerbate proptosis in patients who have orbital venous anomalies or in infants with capillary orbital haemangiomas
  - dependent head position
  - Valsalva maneuver
  - jugular compression

- **Pulsation** is caused either by an arterio-venous communication (with bruit) or a defect in the orbital roof (no bruit)
  - mild pulsation is best detected on the slit-lamp, particularly when performing applanation tonometry

- **Bruit**, a sign of carotid-cavernous fistula, can be abolished by compressing the ipsilateral carotid artery on the neck
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Fundus Changes

- **Optic nerve swelling**
  - may be initial feature of compressive optic neuropathy

- **Optic atrophy**, a feature of severe compressive optic neuropathy
  - thyroid eye disease and optic nerve tumors
**Fundus Changes**

- **Optociliary collaterals** are large tortuous vessels (pre-existing capillaries) diverting blood from the central retinal venous to the peripapillary choroidal circulation
  
  - May be associated with intraorbital optic nerve compression most commonly optic nerve sheath meningioma
  
  - Also associated with optic nerve glioma, cavernous haemangioma, central retinal vein occlusion, idiopathic intracranial hypertension
Fundus Changes

- **Choroidal folds** are a series of roughly parallel alternating light and dark delicate lines of striae at the posterior pole.

- Can occur in a wide variety of orbital lesions including tumors, dysthyroid ophthalmopathy, inflammatory conditions and mucoceles.

- Usually asymptomatic although some patients develop an increase in hypermetropia.
Special Investigations
CT Scan

- **CT-Scan** is useful to illustrate bony structures and the location and size of space-occupying lesions.

- **CT-Scan** is useful in orbital trauma as it can detect small fractures, foreign bodies, blood, extraocular muscle herniation, and emphysema.

- **CT-Scan** is, however, unable to distinguish different pathological soft tissue masses which are radiologically isodense.
MRI & Fine Needle Biopsy

- **MRI** can image orbital apex lesions and intracranial extension of orbital tumors.

- **MRI** can assess some inflammatory activity in thyroid eye disease.

- **Fine needle biopsy**, performed under CT guidance using a 23-gauge needle can be of value in patients with suspected orbital metastases and in those with orbital invasion by neoplasms from contiguous structures.
Causes of Orbital Disease
### Cause of Orbital Disease

(1041 Patients at Moorfields Eye Hospital)

<table>
<thead>
<tr>
<th>Cause</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thyroid eye disease (TED)</td>
<td>178</td>
</tr>
<tr>
<td>Vascular anomalies</td>
<td>148</td>
</tr>
<tr>
<td>Idiopathic orbital inflammatory disease (pseudotumors)</td>
<td>87</td>
</tr>
<tr>
<td>Neoplasm (excluding lacrimal gland)</td>
<td>82</td>
</tr>
<tr>
<td>Infections</td>
<td>82</td>
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<td>ENT problems</td>
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<tr>
<td>Fracture and trauma</td>
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<tr>
<td>Lacrimal gland tumors</td>
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<td>Dermoid and other cysts</td>
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<td>Normal</td>
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<td>Meningiomas</td>
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<td>Neurofibromatosis</td>
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<tr>
<td>Optic nerve glioma</td>
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<tr>
<td>Neurilemomas</td>
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<tr>
<td>Bone change</td>
<td>10</td>
</tr>
<tr>
<td>Others</td>
<td>100</td>
</tr>
</tbody>
</table>
Thyroid Eye Disease
Thyrotoxicosis

- Thyrotoxicosis (Graves’ disease) is an autoimmune disorder, women > men (5/1), 3rd - 4th decade

- Thyroid eye disease (TED) affects 25-50% of patients with Graves’ disease, of which 5% severely

- TED may precede, coincide with, or follow hyperthyroidism

- TED can vary from being merely a nuisance to blindness
Risk Factors

• Once a patient has Graves’ disease, the major risk factor for developing TED is **smoking**

• The greater the number of cigarettes smoked per day, the greater the risk, and giving up smoking seems to reduce the risk

• Women are five times more likely to be affected by TED than men

• Radioactive iodine used to treat hyperthyroidism can worsen TED
Pathogenesis

• TED is an organ-specific autoimmune reaction in which a humoral agent (IgG) produces cellular inflammation with glycosaminoglycan and edema. This in the:

• **A) Extra-ocular muscles**
  – Enlarged muscles can compress the optic nerve, or with time, enlarged muscles can develop fibrosis that may lead to restrictive myopathy and diplopia

• **B) Intra-orbital content** (interstitial tissue, fat, lacrimal gland)
  – The increase in the volume of the intra-orbital content may lead to a secondary elevation of *INTRA-ORBITAL* pressure
Main Clinical Manifestations

Two Clinical Stages

- **A) Congestive stage** (inflammatory), in which the eyes are red and painful. This stage tends to resolve within 3 years and only 10% of patients develop serious long-term ocular problems

- **B) Fibrotic stage** (quiescent) in which the eyes are white but where a painless motility defect may be present
Main Clinical Signs in TED

- Soft Tissue Involvement and Lid Retraction
- Proptosis
- Enophthalmos
- Dystopia
- Ophthalmoplegia (Restrictive Myopathy)
- Dynamic Properties
- Fundus Changes (Optic Neuropathy)
Soft Tissue Involvement

- **Symptoms**
  - Grittiness
  - Photophobia
  - Lacrimation
  - Retrobulbar discomfort

- **Signs**
  - Epibulbar hyperaemia is a sensitive sign of inflammatory activity
  - Intensive focal hyperaemia may outline the insertions of the horizontal recti
Soft Tissue Involvement

- **Signs** (continued)
  - Periorbital swelling behind orbital septum which may be associated with chemosis and prolapse of retro-spetal fat into the eyelids
  - Superior limbic keratoconjunctivitis
  - Keratoconjunctivitis sicca secondary to infiltration of the lacrimal gland
Soft Tissue Involvement

Treatment

• **Lubricants** for superior limbic keratokonjunctivitis, corneal exposure, and dryness

• **Head elevation** with three pillows during sleep to reduce periorbital oedema

• **Eyelid taping** during sleep may alleviate mild exposure keratopathy
Lid Retraction

Reported in up to 50% of patients with Graves’ disease as a result of different postulated mechanisms:

• Fibrotic contracture of the levator

• Secondary over-action of the levator-superior rectus complex

• Humorally-induced over-action of Müller’s muscle
Lid Retraction

Signs

- **Dairample sign** is lid retraction in primary gaze

- **Kocher sign** describes a staring and frightening appearance of eyes which is particularly marked on attentive fixation

- **Von Graefe sign** signifies retarded descent of the upper lid on down-gaze
Lid Retraction

Management

• Mild lid retraction does not require treatment because it frequently improves spontaneously

• Surgery may be considered in patients with significant but stable lid retraction after addressing proptosis and strabismus

• Recession/desinsertion of the levator aponeurosis or recession of lower lid retractors ± hard palate graft
Proptosis

Signs

- Proptosis is axial
- Unilateral or bilateral
- Symmetrical or asymmetrical
- Frequently permanent

- May compromise lid closure which can result in:
  - an exposure keratopathy
  - a corneal ulceration
  - an infection

- Can lead to blindness
Proptosis

Management

• Management is controversial. Some favor early surgical decompression whereas others consider surgery only when non-invasive methods have failed or are inappropriate.

• Systemic steroids
  – **Oral prednisolone** 80-100 mg/day (response in 48 hours), maximal response in 2-8 weeks, in principle not more than 3 months.
  – **Intravenous methylprednisolone** 500 mg repeated for 3 days usually reserved for compressive optic neuropathy.
Proptosis

Management

• **Radiotherapy**
  – In addition to steroids or when steroids are contraindicated. A positive response is usually evident within 6 weeks with a maximal improvement by 4 months

• **Combined Therapy**
  – With irradiation, azathioprine (antimetabolite), and low-dose prednisolone may be more effective than steroids or radiotherapy alone
Proptosis

- **Surgical decompression** may be considered either as primary treatment or when non-invasive methods are ineffective, such as for cosmetically unacceptable proptosis in the quiescent phase

  - **Two-walls** (anthral-ethmoidal), involves removal of the floor and the posterior portion of the medial wall (3-6 mm)
  - **Three-walls**, involves an anthral-ethmoidal decompression and removal of the lateral wall (6-10 mm)
  - **Four-walls**, involves a three-walls decompression, removal of the lateral half of the orbital roof and a large portion of the sphenoidal at the apex of the orbit (10-16 mm)
Restrictive Myopathy

- Up to 30-50% of patients with TED develop ophthalmoplegia
- Initially due to inflammatory oedema and later by fibrosis
- In order of decreasing frequency the four ocular motility defects are:
  - Elevation
  - Abduction
  - Depression
  - Adduction
Restrictive Myopathy

Surgical Treatment

- The indication is diplopia in the primary gaze or reading positions of gaze, provided the disease is quiescent and the angle of deviation has been stable for at least 6 months.

- The goal is to achieve single vision in the primary and reading positions.

- The technique most commonly involves recession of the inferior and/or medial recti.
Optic Neuropathy

- Is a serious complication but uncommon

- Compression of the optic nerve or its blood supply at the orbital apex by the congested and enlarged recti

- May occur in the absence of significant proptosis

- May lead to severe, permanent visual impairment (blindness)
Optic Neuropathy

- **Presentation** is usually with impairment of central vision
- Patients should *daily* monitor their own visual function
- **Visual acuity** is usually reduced, but not invariably, and is associated with
  - relative afferent pupillary defect
  - colour desaturation
  - diminished light brightness appreciation
Optic Neuropathy

- **Visual field defects**
  - may be central or paracentral and combined with nerve fiber bundle defects (DD: POAG)

- **Optic disc**
  - usually normal, occasionally swollen and rarely atrophic

- **Treatment**
  - methyl-prednisolone i.v.

- **Orbital decompression**
  - may be considered if steroids are ineffective or inappropriate
Infections

- Preseptal Cellulitis
- Orbital Cellulitis
- Rhino-Orbital Mucormycosis
Preseptal Cellulitis

- Subcutaneous tissues infection anterior to the orbital septum
- Can progress to orbital cellulitis
- **Causes:**
  - **skin trauma**
    - laceration, insect bites (*S. aureus* or *S. pyogenes*)
  - **spread of local infection**
    - from a hordeolum or dacryocystitis
  - **from remote infection**
    - hematogenous spread from the respiratory tract or middle ear infection
Preseptal Cellulitis

- **Signs** are unilateral, tender and red periorbital oedema
- **CT** shows opacifications anterior to the orbital septum
- Unlike orbital cellulitis, proptosis and chemosis are absent, visual acuity, pupillary reactions and ocular motility are **unimpaired!!!**
- **Treatment** is usually with oral co-amoxicilline
Bacterial Orbital Cellulitis

- Bacterial orbital cellulitis is *life-threatening* infection of the soft tissues behind the orbital septum

- Can occur any age but is more common in children
  - S. pneumoniae
  - S. aureus
  - S. pyogenes
  - H. influenzae
Bacterial Orbital Cellulitis

**Pathogenesis**
- **Sinus-related**
  - most commonly ethmoidal in children and young adults
- **Extension of preseptal cellulitis**
- **Local spread**
  - dacryocystitis, midfacial, or dental infection
- **Haematogenous spread**
- **Post-traumatic**
  - within 72 hours of an injury that penetrates the orbital septum
  - may be masked by associated laceration or haematoma
- **Post-surgical**
  - retinal, lacrimal, or orbital surgery
Bacterial Orbital Cellulitis

- **Presentation**
  - rapid onset of severe malaise, fever, pain, and visual decrease

- **Signs**
  - unilateral tender, warm and red periorbital oedema
  - **proptosis** which is often obscured by lid swelling, is most frequently lateral and downwards
  - painful **ophthalmoplegia**
  - optic nerve dysfunction

- **CT** shows pre-setptal and orbital opacifications
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Bacterial Orbital Cellulitis

- **Ocular complications**
  - includes exposure keratopathy, raised IOP, occlusion of the central retinal artery or vein, endophthalmitis, and optical neuropathy
- **Intracranial complications** (rare)
  - include meningitis, brain abscess, and cavernous sinus thrombosis
- **Sub-periosteal abscess**
  - is most frequently located along the medial orbital wall
- **Orbital abscess**
  - usually occurs in post-traumatic or postoperative cases
Rhino-Orbital Mucormycosis

- **Mucormycosis** is a very rare infection caused by fungi (Mucoraceae), usually seen
  - by diabetic ketoacidosis, or
  - by immunosuppression

- This aggressive and **potentially lethal** infection is acquired by the inhalation of spores, giving rise to an upper respiratory infection
Rhino-Orbital Mucormycosis

- The infection then spreads to the contiguous sinuses and subsequently to the orbit and brain

- Invasion of blood vessels by the hyphae results in **occlusive vasculitis** with **ischemic infarction** of orbital tissues
Rhino-Orbital Mucormycosis

- **Presentation** is with gradual onset of facial and periorbital swelling, diplopia and visual loss

- **Signs**
  - ischemic infarction (**black eschar**) which may develop on the palate, turbinate, nasal septum, skin and eyelids
  - ophthalmoplegia

- **Complications**
  - retinal vascular occlusion
  - multiple cranial nerve palsies
  - cerebrovascular occlusion
Rhino-Orbital Mucormycosis

Treatment

- Amphoterecin i.v.
- Amphoterecin irrigation of the involved areas
- Wide excision of devitalized and necrotic tissues
- Adjunctive hyperbaric oxygen may be helpful
- Correction of the underlying metabolic defect, if possible
- Exenteration may be required in severe unresponsive cases
Inflammation

- Idiopathic Orbital Inflammatory Disease
  “Pseudo-tumor”

- Acute Dacryoadenitis

- Orbital Myositis
Idiopathic Orbital Inflammatory Disease

- **IOID** previously referred as *orbital pseudotumor* is an uncommon disorder characterized by non-neoplastic, non-infectious, space-occupying, orbital lesion.

- The inflammatory process may involve any or all of the orbital soft tissues, resulting for example, in myositis, dacryoadenitis, optic perineuritis, or scleritis.
Idiopathic Orbital Inflammatory Disease

- Histopathological analysis reveals pleomorphic cellular inflammatory infiltration followed by reactive fibrosis

- No correlation between histology and the disease course
Idiopathic Orbital Inflammatory Disease

- **Presentation** is in the 3rd to 6th decades with acute periorbital redness, swelling and pains

- **Signs**
  - congestive proptosis and
  - ophthalmoplegia may occur in severe courses
  - optic nerve dysfunction if the inflammation involves the posterior orbit
Idiopathic Orbital Inflammatory Disease

- CT shows ill-defined orbital opacification and loss of definition of contents
Idiopathic Orbital Inflammatory Disease

Course

• Spontaneous remission after a few weeks without sequels
• Prolonged intermittent activity episodes with remission
• Frozen orbit characterized
  – by ophthalmoplegia,
  – which may be associated with ptosis, and
  – visual impairment caused by optic nerve involvement
Idiopathic Orbital Inflammatory Disease

Treatment

- **Observation**, for relatively mild disease, in anticipation of spontaneous remission

- **Biopsy** in persistent cases to confirm the diagnosis and to rule out neoplasia

- **NSAIDs** are often effective and should precede steroid therapy
Idiopathic Orbital Inflammatory Disease

Treatment

- **Systemic steroids** only after the diagnosis has been confirmed
  - they can mask an infection or Wegner granulomatosis

- **Oral prednisolone**, initially 80-100 mg/day
Idiopathic Orbital Inflammatory Disease

Treatment

• **Radiotherapy** may be considered if there has been no improvement after 2 weeks of adequate steroid therapy.

• **Antimetabolites** such as methotrexate or mycophenolate mofetil may be necessary in the context of resistance to both steroids and radiotherapy.
Idiopathic Orbital Inflammatory Disease

Differential Diagnosis

- **Bacterial orbital cellulitis**
  (antibiotic trial)
- **Severe acute TED** (often bilateral)
- **Systemic disorders** (Wegner granulomatosis, polyarteritis nodosa, and Waldenström macroglobulinaemia)
- **Malignant orbital tumors**
  (particularly metastatic)
- **Ruptured dermoid cyst**
  (secondary painful granulomatous inflammation reaction)
Acute Dacryoadenitis

- Acute dacryoadenitis commonly occurs in isolation, resolves spontaneously and does not require treatment
- **Presentation** is with acute discomfort in the region of the lacrimal gland
Acute Dacryoadenitis

Signs

- S-shaped ptosis and mild downward and inward dystopia
- Tenderness over the lacrimal gland fossa
- Injection of the palpebral portion of the lacrimal gland and adjacent conjunctiva
- Lacrimal secretion may be reduced
Acute Dacryoadenitis

- Is most commonly due to a **viral lacrimal gland infection** caused by mumps, infectious mononucleosis, and less commonly, by a bacteria

**Differential Diagnosis**

- **Ruptured dermoid cyst** may cause localized inflammation in the region of the lacrimal gland

- **Malignant lacrimal gland tumors** may cause pain but the onset is not usually acute