Surgical Neck Pathology

Considered here will be

- Branchial pathology
- Thyroglossal cysts
- Cervical node pathology
- Sternomastoid “tumor”
- Lymphatic malformations
- Dermoid and epidermoid cysts

Embryology

Branchial Arch Development

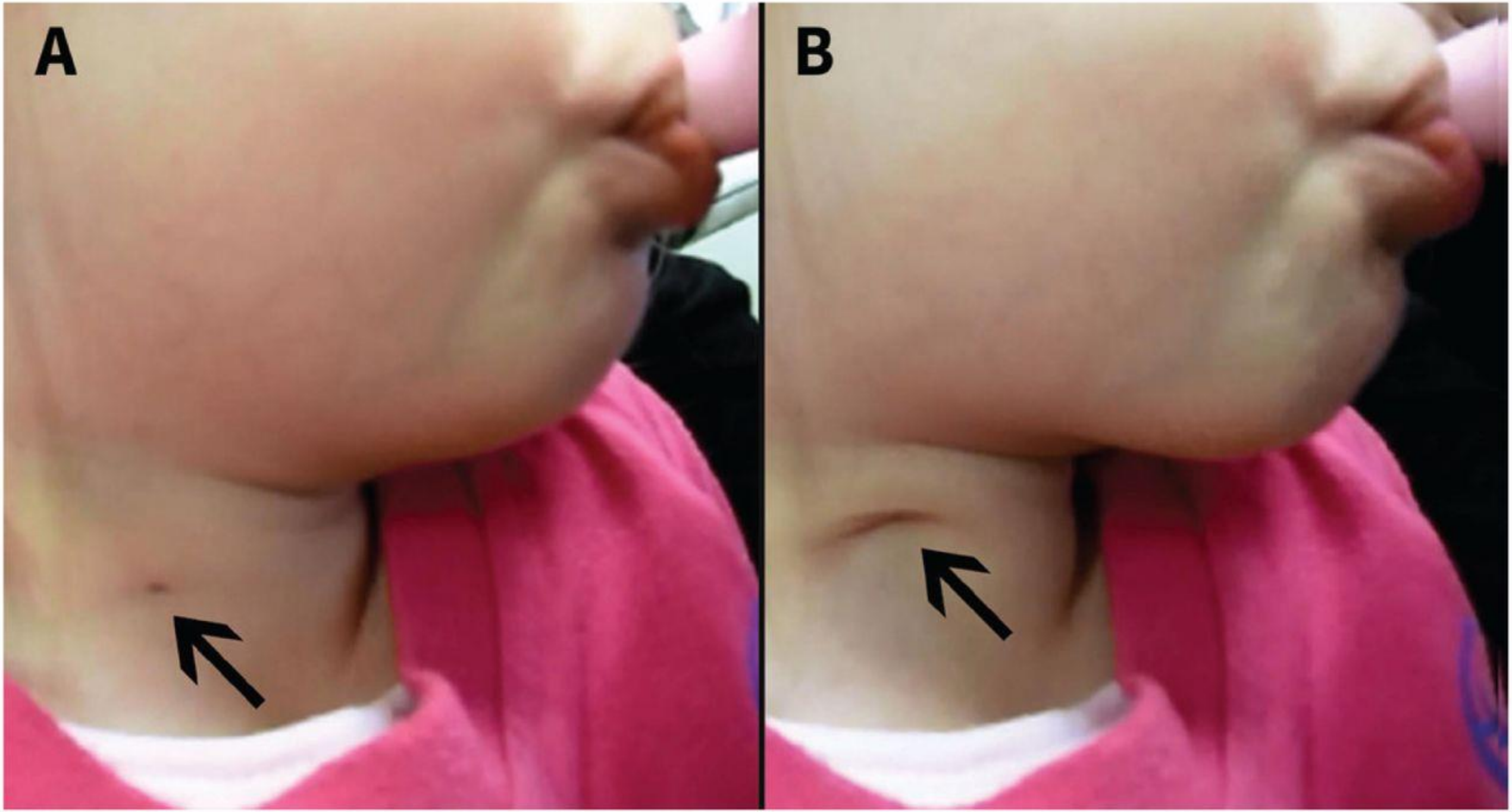
- There are five pairs of branchial (or pharyngeal) arches evident from day 22, numbered I, II, III, IV, VI (V is missing in humans).
- Each has a cartilaginous center, a nerve, and an aortic arch-associated artery.
- They are separated externally by ectodermal clefts and internally by endodermal pouches. Derivatives of the first arch cartilage are the malleus and incus of the middle ear;
- The second arch forms the stapes, stylohyoid, and upper part of hyoid bone, and the third arch the remainder of the hyoid.
- Derivatives of the first pouch include the tympanic cavity and Eustachian tube;
- the second pouch leads to the tonsils, and the third pouch to the thymus gland and inferior parathyroid glands.
- Thyroid Development The thyroid precursor anlagen arises on the back of tongue (foramen caecum) and migrates caudally to lie in front of the proximal tracheal rings, leaving the potential for a track (should obliterate by fifth week) and cyst formation (thyroglossal cyst).

Branchial Fistula/Sinus/Cyst

Clinical Features

- *Fistula>sinus>cyst*

- Although branchial remnants are congenital and therefore present since birth, there is some debate about the etiology of “branchial cysts,” which is usually found in later childhood.
- Most cysts are located anterior to the sternomastoid muscle, posterior to the submandibular gland and lateral to the carotid sheath.
- Most are usually second arch remnants and their external opening is usually sited near the insertion of the sternomastoid at clavicular level. Sinuses then ascend the neck along the carotid sheath to the level of the hyoid bone turning medially between the internal and external carotid artery, in front of the hypoglossal nerve to end in the region of tonsillar fossa.
- Sometimes, there is mucoid discharge from this opening and occasionally it can become infected.
- Cysts occur deep to the middle 1/3 of sternomastoid, and can be complicated by sepsis.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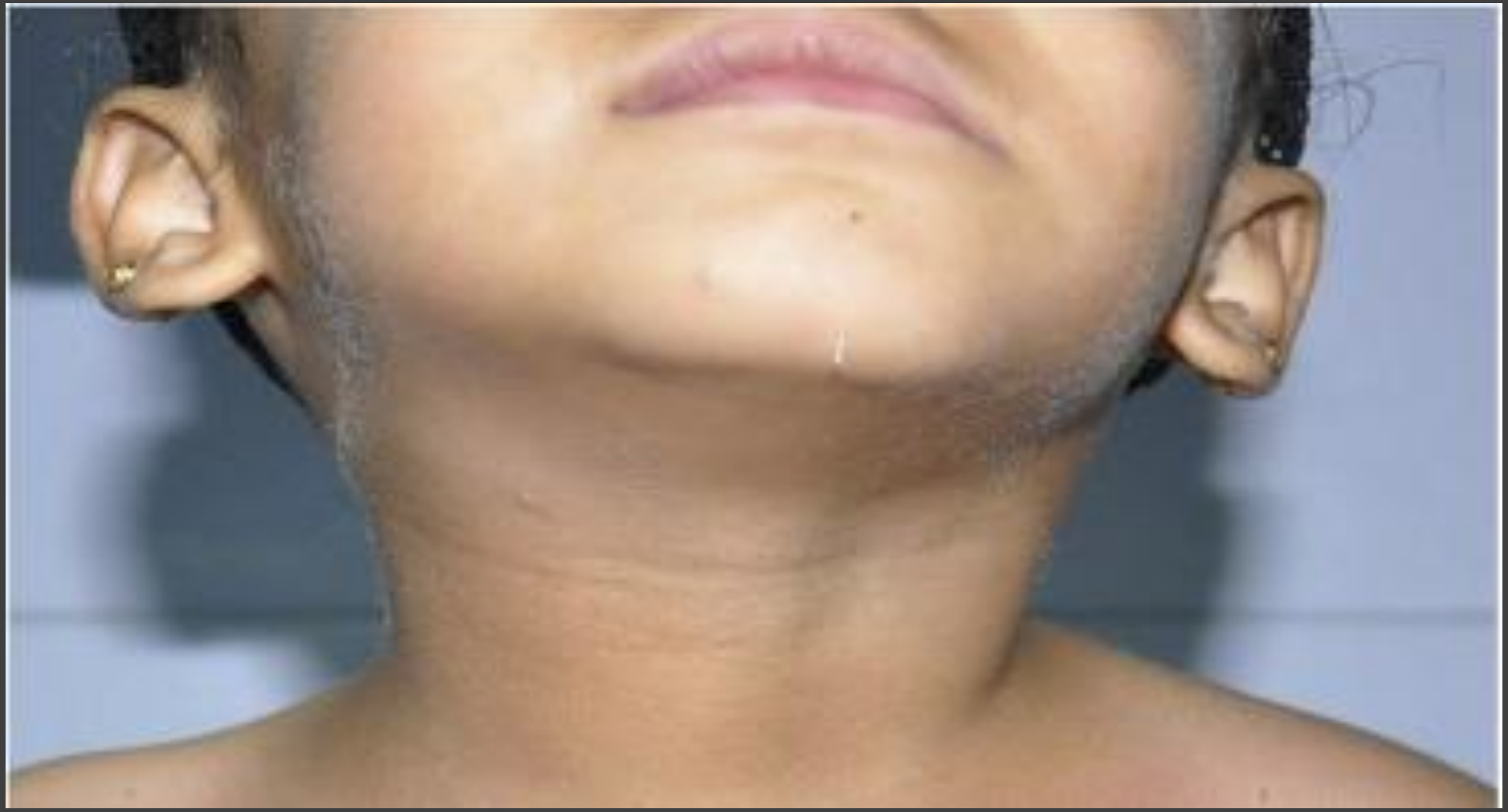


Investigation

- Ultrasound – looking for cystic change

Surgery

Elective excision of the complete tract or cyst. If infected, treat this first prior to definitive procedure.

Transverse skin incision(s) with methylene blue to outline the sinus.

Recurrence ~3% after elective operation, ↑ if previous unsuccessful excision.

First branchial arch sinus – rare, with external opening at the level of angle of jaw, and track ascending theoretically toward the middle ear.

Thyroglossal Cyst

Unknown incidence

Clinical Features

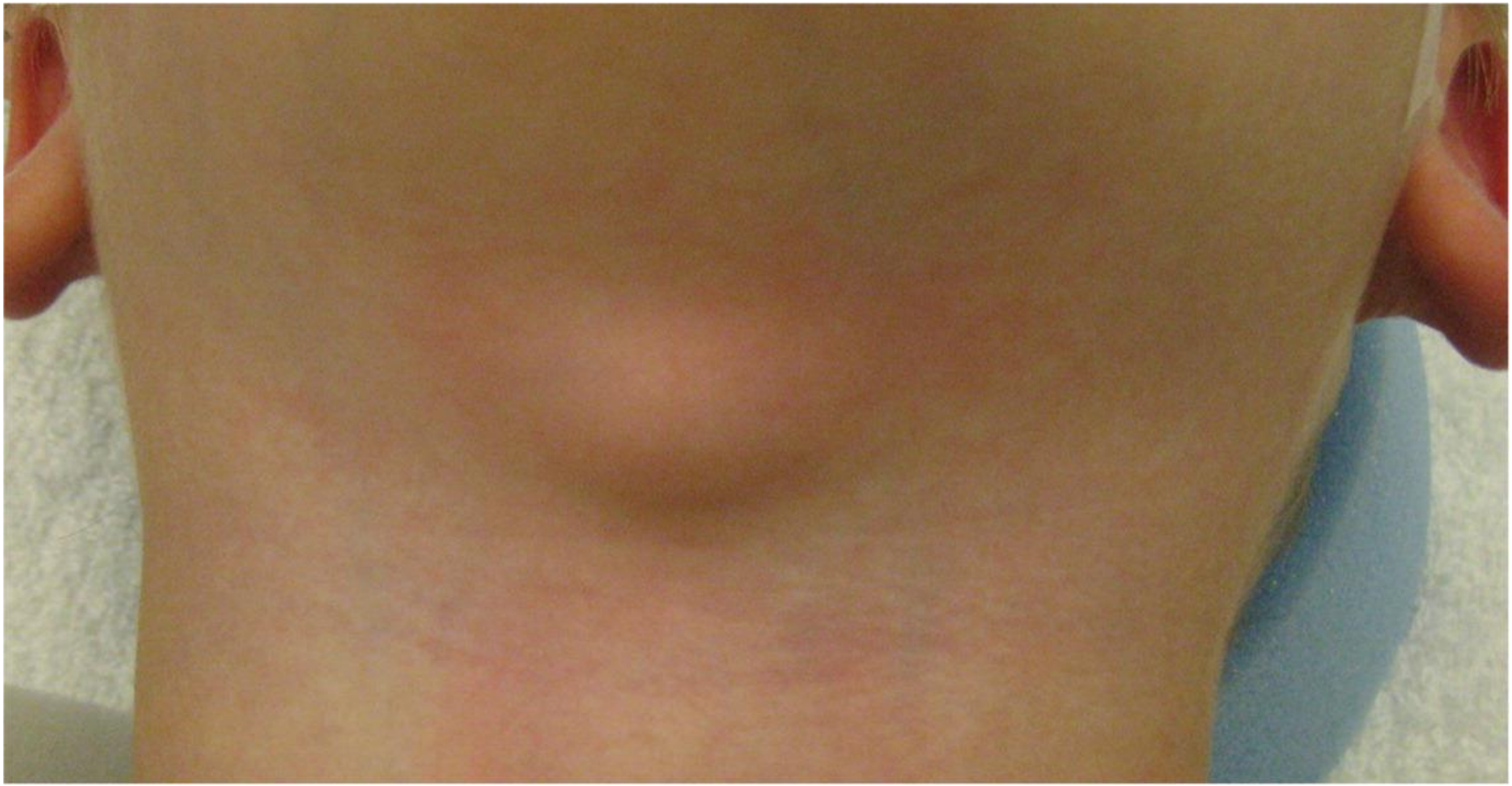
- Most (80%) are related to and in contact with hyoid bone.
- Midline swelling (elevates with swallowing or protrusion of the tongue).

May be complicated by infection and if allowed to discharge a distal midline fistula often results.

Investigation

Cervical US – shows cystic nature and also confirms presence of normal thyroid gland.

Differential diagnosis includes nodal pathology, dermoid cyst, thyroid pathology, and if suprahyoid, a ranula.



Surgery: Sistrunk's Operation

Skin crease incision with development of skin flaps.

1. Mobilize cyst from distal tract (if any).
2. Dissect and isolate the body of hyoid in continuity with the cyst.
3. Identify proximal track to base of tongue.
4. Repair in layers, approximate hyoid remnants.

If everything removed, then recurrence risk is low (~5%).

A very small (<1%) risk of cancer has been described in these cysts.

Cervical Node Infections (Common)

Over 75% of lymphatic tissue is found within the head and neck, and this remains the commonest site of manifestation of lymphatic pathology. The ears, nose, mouth, pharynx, and upper airway remain the commonest portal for bacterial and viral entry and this is reflected in the huge incidence of lymphadenitis seen in childhood. Fortunately, most either goes without comment and subsides spontaneously or is dealt with in general practice by the liberal use of antibiotics.

Age Group	Common pathogens
Neonate	Staphylococcus aureus, late-onset Gp B Strep
<5 years	Gp.A Strep, S. aureus, NTM
5–18 years	EBV, cytomegalovirus, toxoplasmosis, TB, infectious mononucleosis

Clinical Features

Whatever the cause, the usual scenario is to evolve (days) through an active inflammatory cellulitic phase with later central cavitation and pus formation. The disease process then resolves via discharge to the skin or by surgical intervention (I&D). In both NTM and TB, the process is much more prolonged and “cold,” and because of the inadequacy of the host cellular response, it is often not resolved by discharge or I&D.

Miscellaneous Nodal Pathology

Nontuberculous Mycobacterium (NTM)

(Aka Atypical Mycobacterium)

• Mycobacterium avium – intracellulare

Most enters via oropharynx, and is a feature in preschool (<5 years) children. Typical groups affected include cervical and submandibular. There tends to be obvious overlying skin changes, with ultimate discharge but very often fistula formation.

Investigation

1. CXR (usually negative)
2. Mantoux (i.e., PPD) test (usually negative), now specific NTM antigen skin test
3. Ziehl–Neilsen (ZN) stain and culture (takes up to 12 weeks)
4. (rapid) PCR (looking for mycobacterial RNA)

Management

• Surgical excision (if possible)

– Beware of proximity to mandibular branch of Facial (VII) nerve and spinal Accessory (XI) nerve.

• Clarithromicin/azithromicin

Tuberculosis

This is now rare in developed countries but may occur in the Immunosuppressed (e.g., posttransplant).

Surgery is limited to diagnostic biopsy (or fine-needle aspiration cytology), or I&D of abscesses.

Cat Scratch Disease

- Bartonella henselae – found in cats .

Clinical Features

Usually causes regional lymphadenopathy (typical site is axillary or epitrochlear nodes) but can cause pre-auricular lymphadenopathy (and granulomatous conjunctivitis – Parinaud's syndrome).

Investigations

- ELISA possible.
- Biopsy – granulomatous reaction. Visible on Warthin–Starry stain.
- PCR on sampled material possible.
- Most are self-limiting in immunocompetent children, but azithromycin for severe symptoms.

Kawasaki's Disease

This is a vasculitic disease of unknown etiology, which principally affects

- Heart – coronary artery aneurysm.
- Skin and mucous membranes – "strawberry" tongue, red palms and soles with later desquamation.
- Lymph nodes – cervical adenopathy.

Investigation

No specific test – diagnosed on clinical criteria (5-day fever, erythema of lips, etc.)

Management

- High-dose immunoglobulin and salicylates ± steroids

Sternomastoid "Tumor" (Rare)

Uncertain etiology and unknown incidence

There has sometimes been a history of birth trauma or breech delivery suggesting that there has been hemorrhage within muscle, followed by fibrosis and muscle shortening.

Clinical Features

Typically presents with painless lump within sternomastoid muscle at 2–4 weeks of age with or without torticollis (head rotated and tilted away from the affected side). Initially, any abnormal head positioning can be corrected, but with time this becomes fixed. There is then secondary soft tissue changes and facial asymmetry.

Investigation

1. US may show focal or diffuse enlargement of the sternocleidomastoid muscle.
2. Cervical radiography – exclude vertebral anomalies (e.g., hemivertebra).
3. MRI – precise definition of cervical vertebral anomalies.

Management

- Cervical Physiotherapy – successful in >90% of cases.
  – Early diagnosis and intervention.
  – Passive neck movement and stretching exercises .
- Surgery – division of sternomastoid muscle – achieves ↑ length.

Lymphatic Malformations (Rare)

- ~50% of these malformations are diagnosed at birth
- ~90% present before 2 years of age.
- Uncertain incidence – 1 in 6,000 live-births

Associations

- Chromosomal anomalies (e.g., Turner's (XO), Noonan's syndrome (single gene defect on Ch12q24.1), Down's syndrome (Trisomy 21)).

Lymphatic malformations are vascular malformations composed of primitive embryonic lymph sacs of varying sizes. Mostly found in relation to head and neck, but can be ubiquitous.

Cystic hygroma – term used to describe congenital neck lesions. Some are huge and can cause polyhydramnios, obstructed labor, and failure to establish an airway at birth.

They can be divided into three clinical types:

1. Microcystic
2. Macrocystic
3. Mixed lesions

They may also be described as unilocular, multilocular; focal, or diffuse/infiltrative.

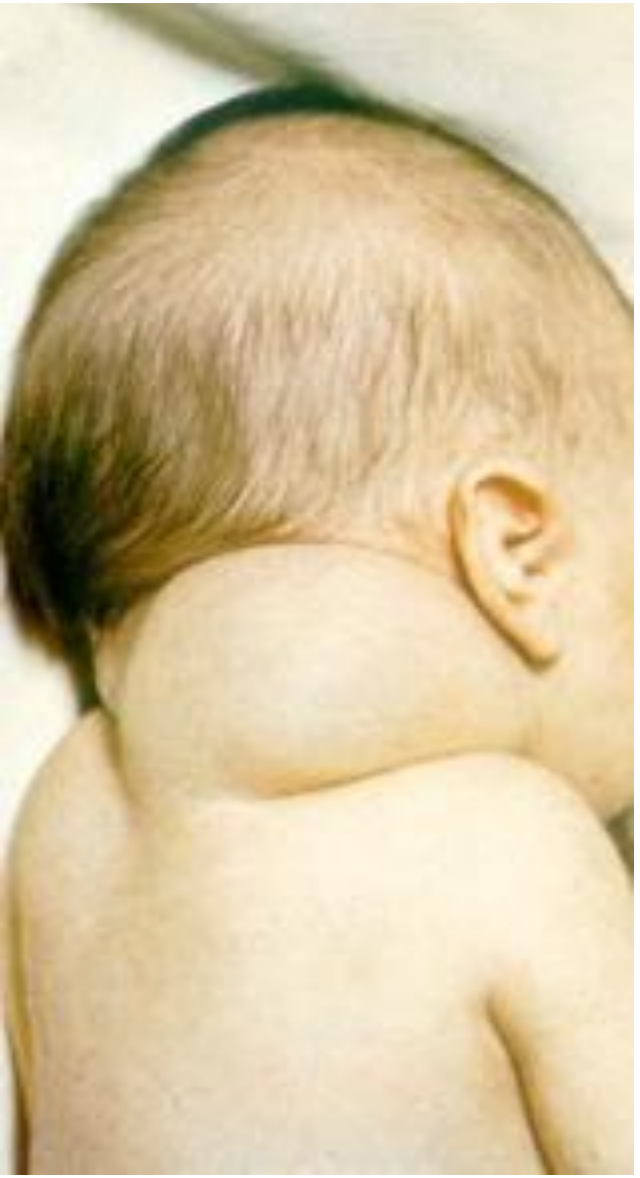
Clinical Features

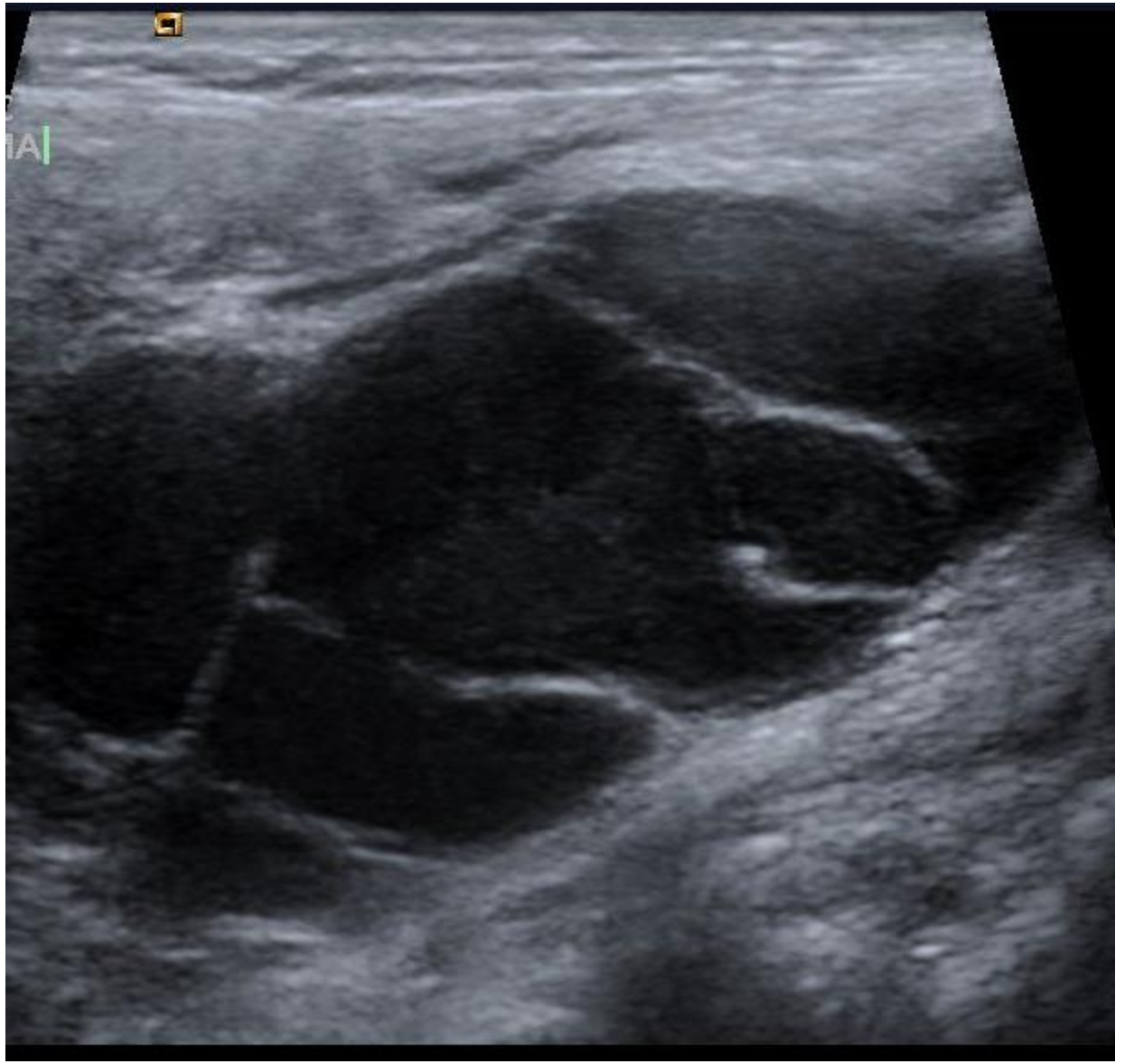
Lymphatic malformations typically increase in size as the child grows, and they may shownrapid increase in size in association with upper respiratory tract infection or intralesional hemorrhage, causing compromise of airway if present in suprahyoid region.

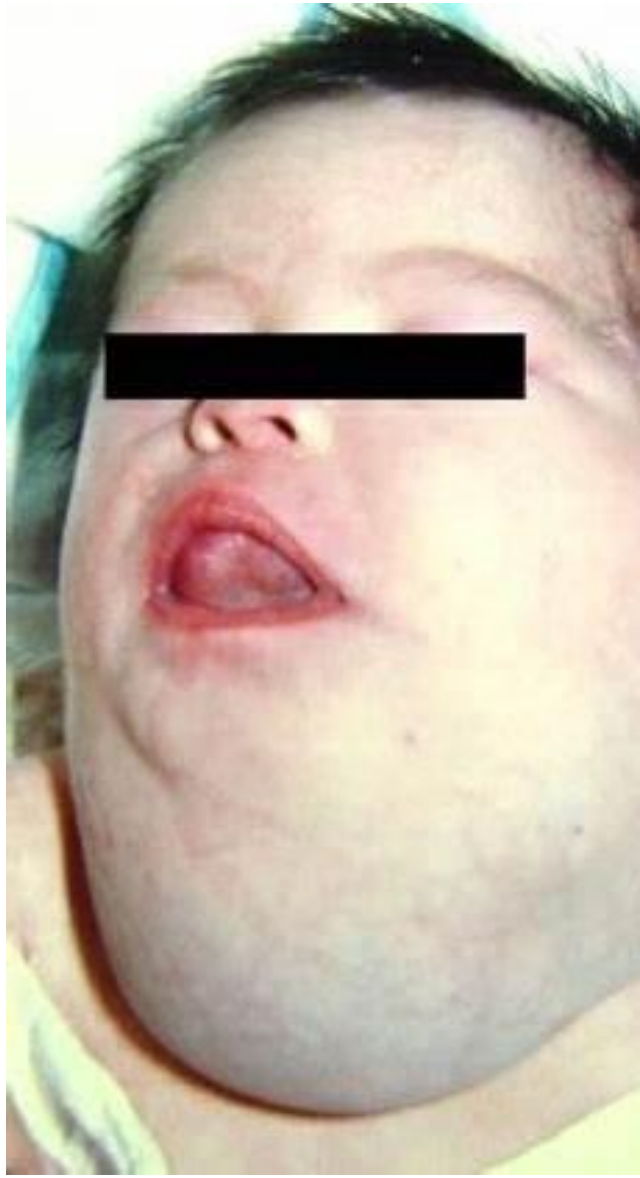
Investigation

1. US – to determine cyst size and number.
2. MRI – defines anatomical relationship to cervical vessels and trachea.

Sometimes, lymphatic malformations may be a part of mixed vascular malformations(typically venous)







Management

Contentious, confusing, and contradictory!

- Chemotherapy – percutaneous injection/inflitration

– OK-432 (Picabanil®) is a lyophilized mixture of Streptococcus spp.

– Absolute alcohol solution.

– Ethibloc® – combination of alcohol, mixed irritant proteins, and a radio-opaque marker.

– Cyclophosphamide – IV, reserved for life-threatening examples.

– Bleomycin – intralesional injection (total dose 5 mg/kg)

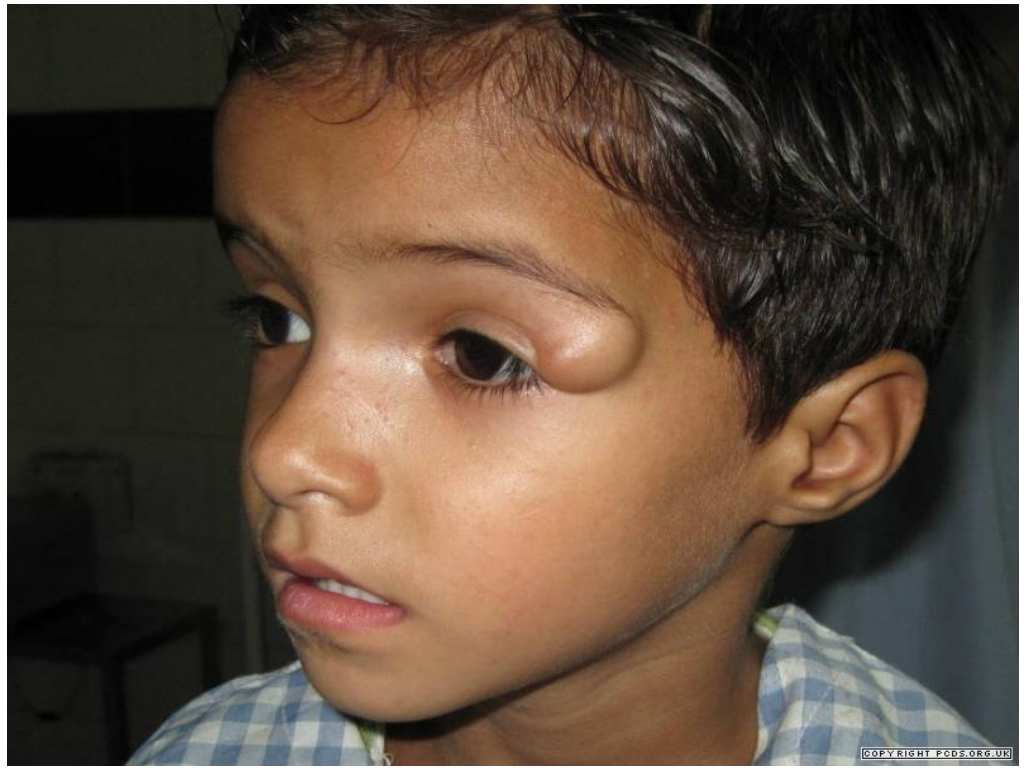
- Surgery

– Aim for complete excision – but this is easier said than done! May be difficult owing to nerve proximity, etc.

Dermoid Cysts and Epidermoid Cysts (Common)

Dermoid cysts represent superficial ectodermal elements, which have become trapped beneath the skin and occur at sites of ectodermal fusion. Typical sites include anywhere along the body's midline, or along the line joining upper ear to upper outer part of eyebrow (external angular dermoid).

- Dermoid cysts contain squamous epithelium and skin appendages such as hair follicles and sebaceous glands.
- Epidermoid cysts contain only squamous epithelium.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