تبييض محاضرة

CDH & neck anomalies

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Done by :

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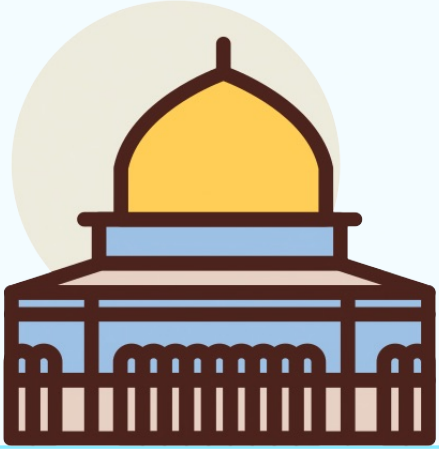


Congenital Diaphragmatic Hernia (CDH) very severe

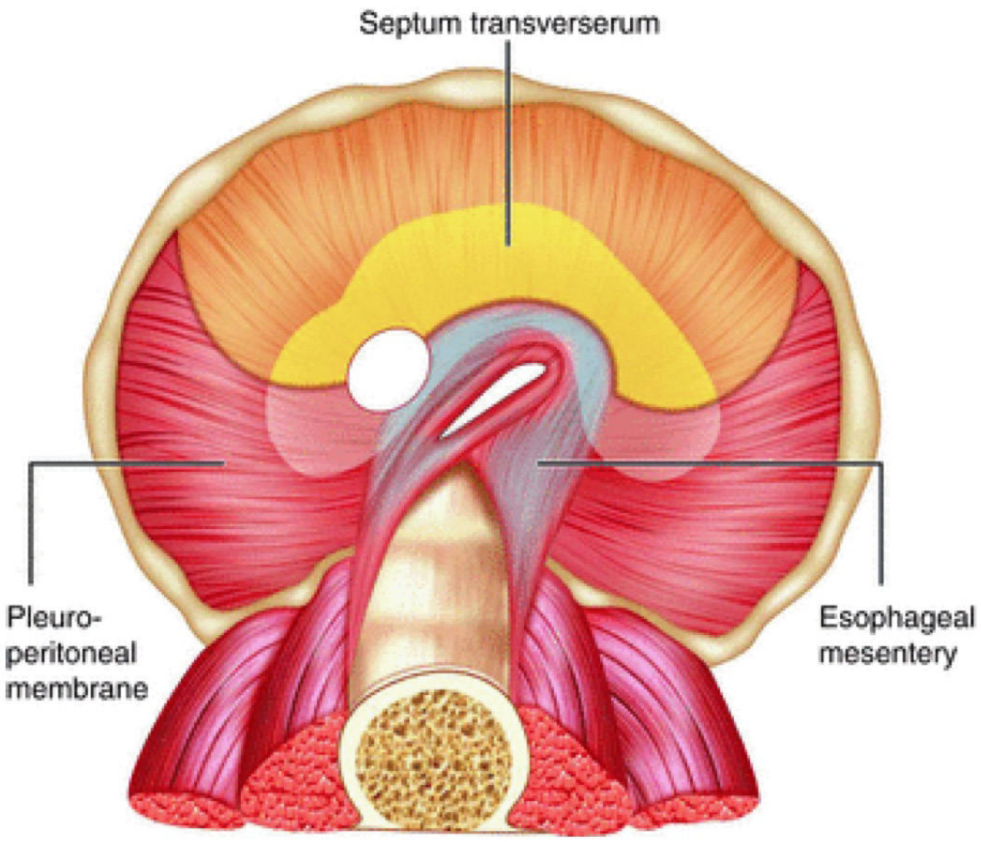
Embryology:

- Precursors of the diaphragm begin to form during the fourth week of gestation.

- The diaphragm develop from the fusion of four embryonic components:
  - Anteriorly by the septum transversum
  - Dorsolaterally by the pleuroperitoneal folds (PPFs),, **the most common component affected by hernia.**
  - Dorsomedially by the crura from the esophageal mesentery
  - Posteriorly by the body wall mesoderm

- The pleuroperitoneal membranes close and separate the pleural and abdominal cavities by the eighth week of gestation.

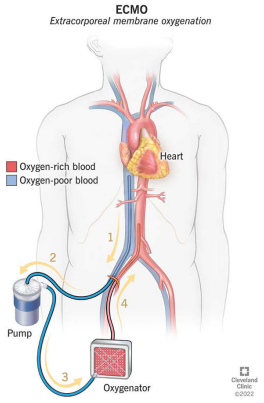
- The right side closes before the left. **(the left is most commonly affected)**



Development of the diaphragm

**Diaphragmatic hernia in adults: the defect in hiatus.
*Two types: sliding, paraesophageal = both with late presentation.

*in neonates: the defect is in the diaphragm itself.
*in neonates: ECMO is helpful. (extracorporeal membrane oxygenation =machine pumps blood from the body and oxygenate it).



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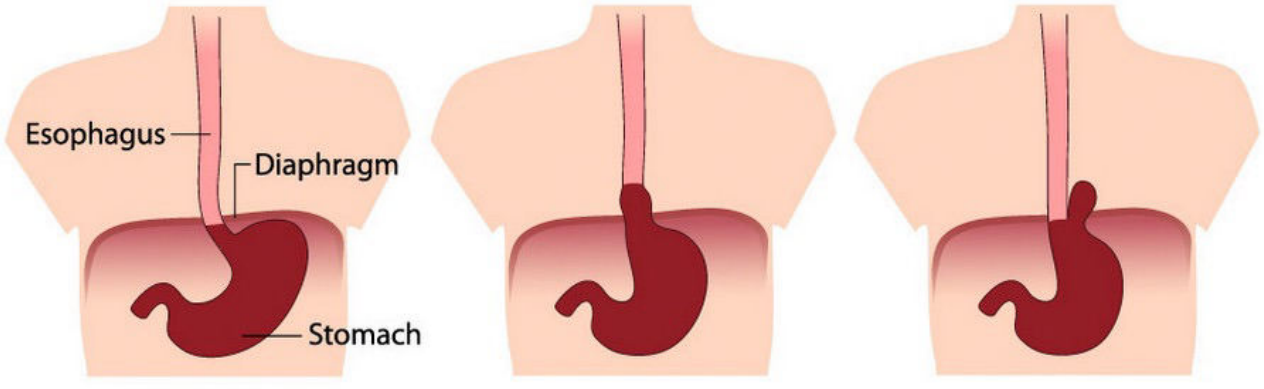
SOUTHLAKE GENERAL SURGERY

Hiatal (Hiatus) Hernia



Normal

Sliding Hiatal Hernia

Paraesophageal Hiatal Hernia

www.southlakegeneralsurgery.com

Inadequate closure of the pleuroperitoneal canal allows the **abdominal viscera(compress the lung for long duration)** to enter the thoracic cavity when they return from the extraembryonic coelom to herniate into the chest.

As a result, **pulmonary hypoplasia *due to chronic compression*** develops (decrease in bronchial divisions, bronchioles, and alveoli)

Affects the contralateral lung as well. **(affect 3/4 of it in severe cases when abdominal viscera compress the mediastinum).**

The pulmonary arteries exhibit a decrease in density as well as an increase in muscularization (**CDH- ASSOCIATED PULMONARY HYPERTENSION**). **Decresad cross section. –very difficult to treat. –very limited medication.**

Prevalence of CDH: 2.3–2.4 per 10,000 live births

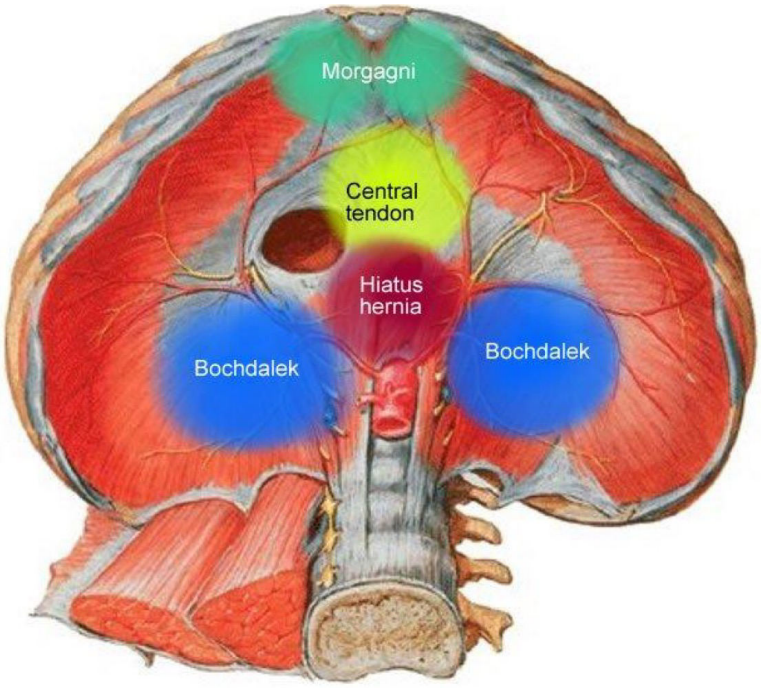
Male infants more common

The most common is left.
90% are posterolateral or "Bochdalek".**The defect is large.**
- -Left sided (80%)
- Right sided (19%)
- Bilateral (1%) **severe**

10% are located anteriorly, termed "Morgagni" hernias. **The defect usually is small.**

60% of CDH cases are isolated, 40% associated with anomalies of the cardiovascular
(27.5%), urogenital (17.7%), musculoskeletal (15.7%), and central nervous (9.8%) (CNS) systems, GI system (atresia).
***If the liver is intrathoracic :: severe.**

Aetiology of CDH



- Bochdalek is the commonest diaphragmatic hernia in infants.
- Sliding H.H is the commonest diaphragmatic hernia in adults.

*morgagni hernia: small defect =content can be seen in chest::
-small bowel
-part of stomach
-no liver
-no spleen

(Large defect)

#liver and spleen can be found in the chest in bochdalek hernia.

Left-sided postero-lateral diaphragmatic defects (Bochdalek hernias) are the most common (90%) and more common on left side , while anterior defects (Morgagni hernias) are rare and more on the right side.

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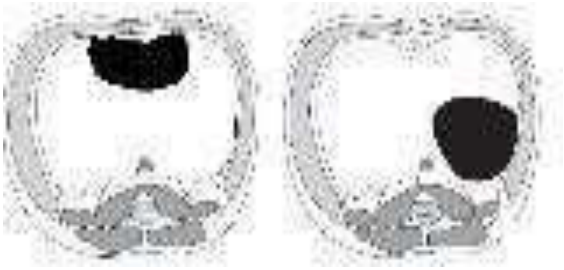
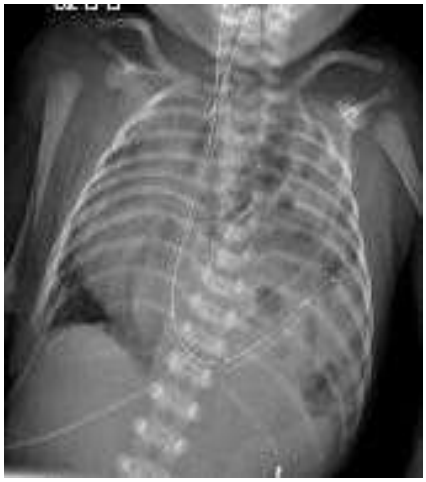
*left picture= on the anterior side :: morgagni.
*right picture= posterior/ laterally :: bochdalek.
** the presentation is usually late (1-2 years)
because it is asyptomatic.



NG tube seen in the chest.



Diagnosis

**severe cases need immediate intervention.
** when there is other congenital anomalies: poor prognosis.
PRENATAL DIAGNOSIS: (antinatal)

Approximately 50–70% of infants . بشخصوهم قبل الولادة

Fetal US features include: **(can asses prognosis through LHR)**

Polyhydramnios **(bec. The featus has problem of swallowing)**, intrathoracic fluid-filled bowel loops, mediastinal shift **(usually to the right)**, and/or an intrathoracic stomach, spleen, or liver.

The lung-to-head ratio (LHR)**:*to asses prognosis*** is a prenatal US assessment ratio, utilizing the contralateral lung area to the head circumference, which predicts CDH severity.
****LHR usually >1.6**
If the LHR is 1 or less, the prognosis is poor.

Lung to head ratio: normal>1 in CDH<1. Very severe cases it can be 0.6
LHR : diameter of the contralateral lung to the circumference of the head

*LHR:: -mild: 1-1.5
-moderate: 0.8-1
-severe: <0.8

**when there is any abnormality in fetal U/S =order fetal MRI (to confirm it by viewing the complete anatomy of fetus).

> ► Any newborn with cyanosis in absence of heart disease and a scaphoid abdomen is considered congenital diaphragmatic hernia until proved otherwise

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CLINICAL PRESENTATION

Respiratory distress:
tachypnea, chest wall retractions, grunting, cyanosis.

On physical examination:
- Scaphoid abdomen
- Increase in thoracic diameter
- The point of maximal cardiac impulse is often displaced (in right side). The heart sounds are better heard on Rt. Side in case of the common left sided hernia.

-Bowel sounds may be auscultated within the thoracic cavity
- Decrease in breath sounds bilaterally.
- **if there is hypoxia in the first (5 to 6 hrs)= you should check CDH.

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It can be antenatal diagnose because it can cause functional obstruction (polyhydroaminos) which means there's defect in GI tract ,
gastric bubble in the chest ,
shifted heart beat in the right side
it's also affecting the other side of the lung (mediastinal compression)
bilateral effect the oxygenation can be normal in the first two mins of delivery then rapidly decrease

Postnatal diagnosis

Chest radiograph:

**** the first choice of studies is X-ray.**

- Intestinal loops within the hemithorax,
- Cephalad displacement of the stomach/orogastric tube
- Mediastinal shift toward the contralateral hemithorax

80% will be diagnosed within the first 24 hours of life
20%(mild cases) may present outside the neonatal period (These patients present with mild respiratory symptoms, chronic pulmonary infections, pleural effusions, pneumonias, feeding intolerance)

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Most common presentation : hypoxia , cyanosis , scaphoid abdomen, menengiocele & characteristic absence of the breath sounds in the ipsilateral chest . The respiratory distress is due to the sever pulmonary hypoplasia , persistent pulmonary hypertension of the new Neonates PPHN due to cross sectional vessels are compressed & poor surfactant production so the vascularity of the lung decrease & the resistance increase

Pulmonary hypoplasia : An underdevelopment of the lungs characterized by a decreased number of alveoli and airways. Result in an impaired gas exchange. Associated with oligohydramnios and Potter sequence.

Treatment

RESUSCITATION AND STABILIZATION
(reoxygenation)
Prompt endotracheal intubation= **first intervention.**
(without bag mask ventilation) ***bec It is fatal*.**
Initiation of conventional mechanical ventilatory support
(machine that gives O2)
A nasogastric tube *or orogastric* should be inserted to avoid gastric and intestinal distention =**for decompression.**
Operative Repair
CDH is no longer considered an emergency procedure.
surgery is performed after cardiopulmonary stability(Low ventilator support with evidence of resolving pulmonary hypertension by echocardiogram) كميات قليلة من o2

If there's any finding in the antenatal ultrasound we refer to foetal MRI . First step after de sat = Endotracheal tube + orogastric tube with mechanical ventilation comes with high setting (100% & pressure above 25) & low setting (40% with pressure of 16 or 14) The repair of CDH is not an emergency we should stabilise the patient on low setting ventilation

Poor prognosis

- Large defect size
- Major congenital heart disease
- prematurity
- liver herniation
- LHR less than 1
- Right more than left
- Bilateral herniation

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Neck anomalies

THYROGLOSSAL DUCT CYST (TGDC):

The foramen cecum is the site of the development of the thyroid diverticulum**. ((at the base of tongue)).**

***fluid filled sac at midline of neck.**

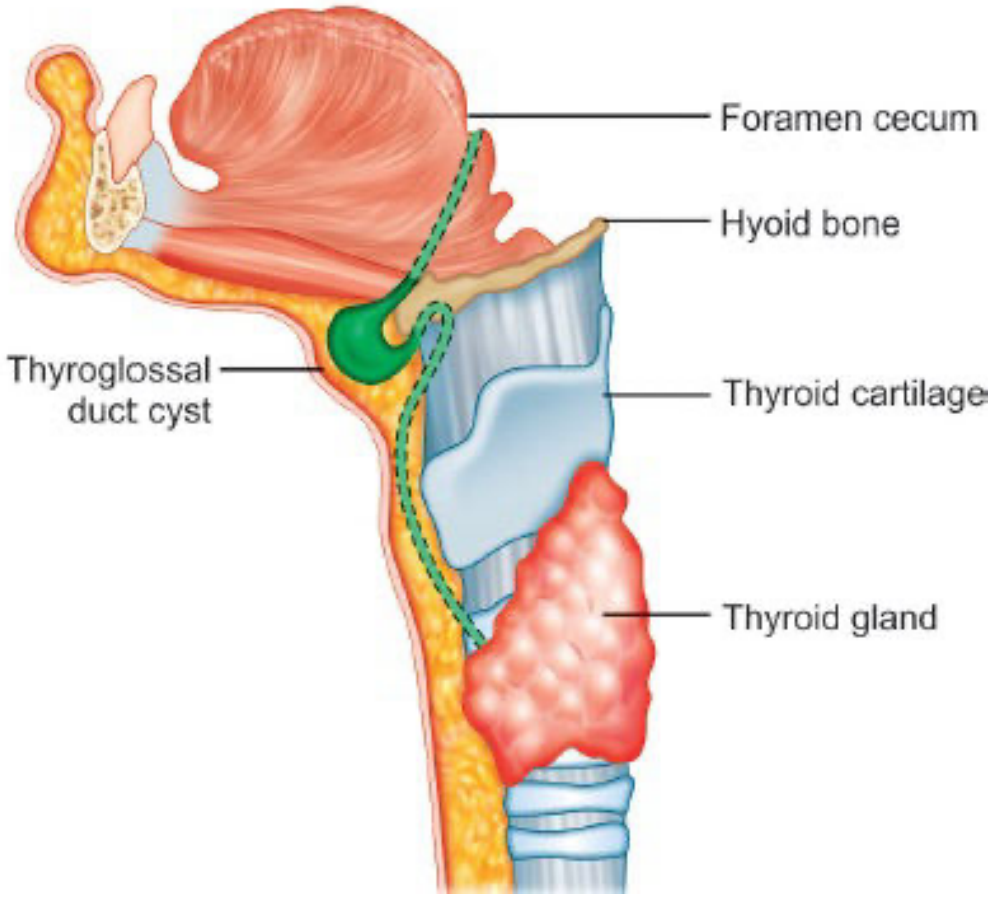
***most in children <5 years.**

Thyroid diverticulum descends into the neck, maintaining its connection to the foramen cecum.

the thyroglossal duct may pass in front of or behind the hyoid bone, but most commonly, it passes through it.

Normally, the duct disappears by the time the thyroid reaches its appropriate position by 5–8 weeks of gestation.

A cyst can be located anywhere along the migratory course of the thyroglossal tract if it fails to become obliterated



Presentation

Midline painless mass at or just below the hyoid bone, could be lateral
On physical examination, the TGDT is smooth, soft, and nontender.
**at midline anterior round mass in neck,, it is small, fluctuant, painless, movable (تتحرك لفوق وتحت عن طريق اخراج اللسان لبرا لتمييزها) - protrusion of tongue.
To distinguish this lesion from the more superficial dermoid cyst, one should palpate the lesion while the child sticks out his or her tongue. Owing to its attachment to the foramen cecum, the thyroglossal duct cyst usually moves cephalad (upward) when the tongue protrudes.
A thyroglossal duct cyst moves superiorly with tongue protrusion whereas a dermoid cyst may not.
This maneuver is more reliable than asking the child to swallow

thyroglossal cysts can become infected with oral flora (not skin flora) and present as draining sinus from spontaneous or incisional drainage of an abscess
from flora

Other DDX:: -dermoid cyst
-ectopic thyroid
-lymphadenopathy (submental LN) most common.
-sebaceous cyst
-lipoma.

- TGDC is the most common neck mass in children
- Most patients present in first five years of life.

Elective surgical excision of a thyroglossal duct cyst is advised to avoid the complications of infection (abscess, sinus) and the small risk (<1%) of cancer (papillary thyroid carcinoma- the most common)

The operation includes complete excision of the cyst and its tract upward to the base of the tongue, and resection of the central portion of the hyoid bone (to avoid recurrence) as described by Sistrunk (Decrease risk of recurrence).

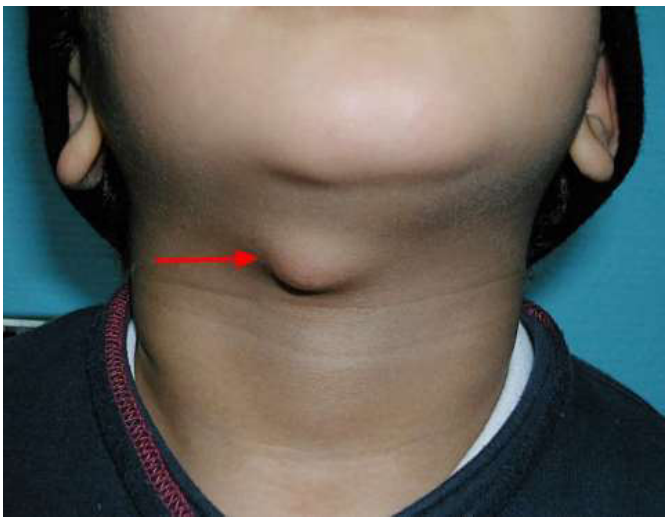
**before doing the surgery:: do thyroid function test then US or isotope scan to rule out ectopic thyroid حتى ما نشيلها مغرين انها TGDC.(ectopic thyroid usually presented with hypothyroidism)=

complications of the Sistrunk procedure (removal thyroglossal cyst only –not ducts ++ mid portion of hyoid bone (نشيلها معها حتى لا تتكرر

Major complications include recurrence **(when part from tract not removed)**, hematoma or abscess, entry into the airway- **injury to trachea** (tracheotomy), hypoglossal nerve paralysis, hypothyroidism (ectopic thyroid).

High TSH
thyroxine بنعالجه نعطيه

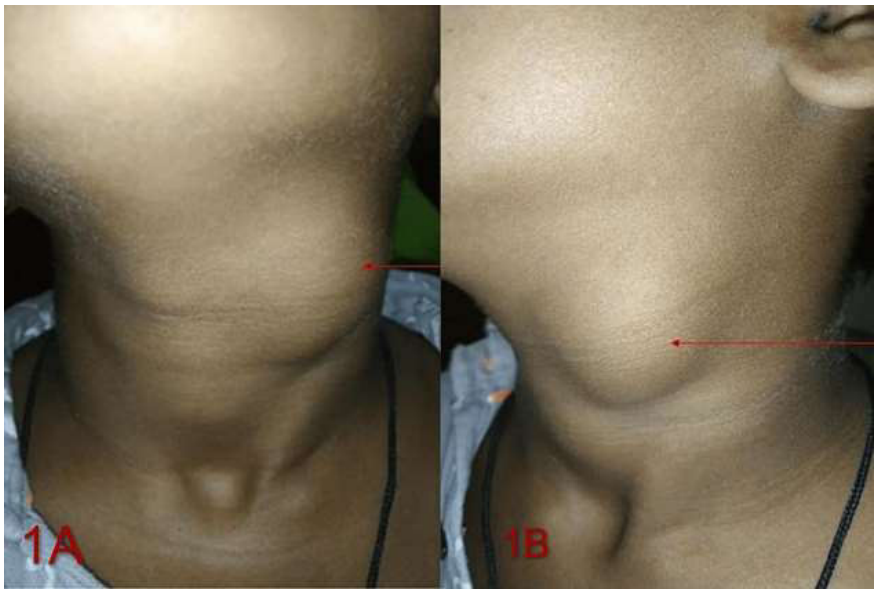
Minor complications include Seroma formation, wound dehiscence, local wound infection, and stitch abscesses

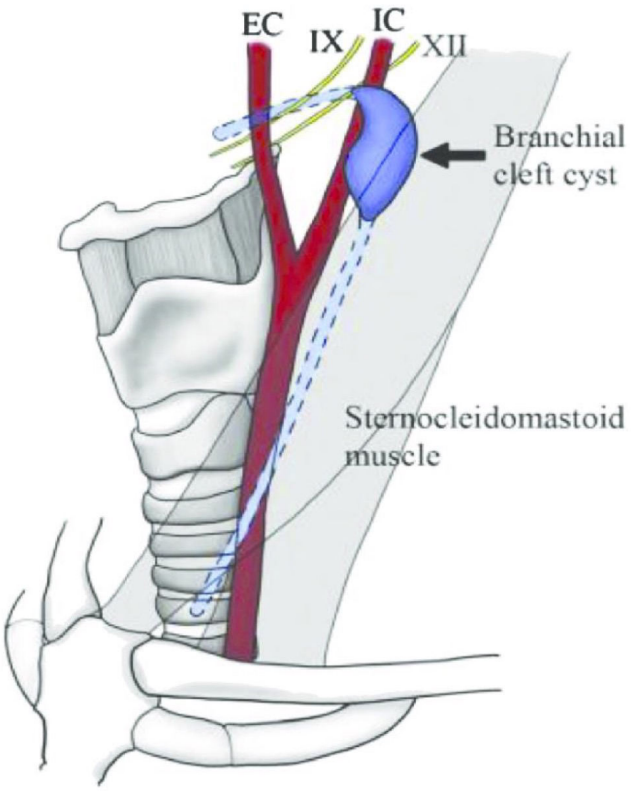


Branchial Anomalies

Incomplete regression of branchial structures results in cysts, sinuses, or fistulae.

Second branchial anomalies being the most common (approximately 95%).





Anatomical course of branchial fistula

Branchial arches development

First - ear, eustachian tube, mastoid air cells, tympanic membrane, auditory canal, maxillary artery, muscles of mastication, trigeminal nerve, Meckel cartilage (stapes, styloid process, portion of hyoid bone)

Second - facial nerve, muscle of facial expression, stapedial artery, tonsillar fossa, palatine tonsil, Reichert cartilage (stapes, styloid process, hyoid bone)

Third - inferior parathyroid gland, thymus, carotid artery, stylopharyngeus muscle, glossopharyngeal nerve, hyoid

Fourth - superior parathyroid, thyroid c-cells, cricothyroid muscle, most of the pharynx and palate, vagus, superior laryngeal nerve, thyroid and epiglottic cartilages

Fifth - regresses

Sixth- pulmonary artery, muscle of larynx except cricopharygeus, vagus and recurrent laryngeal nerve, cricoid cartilage, arytenoid complex

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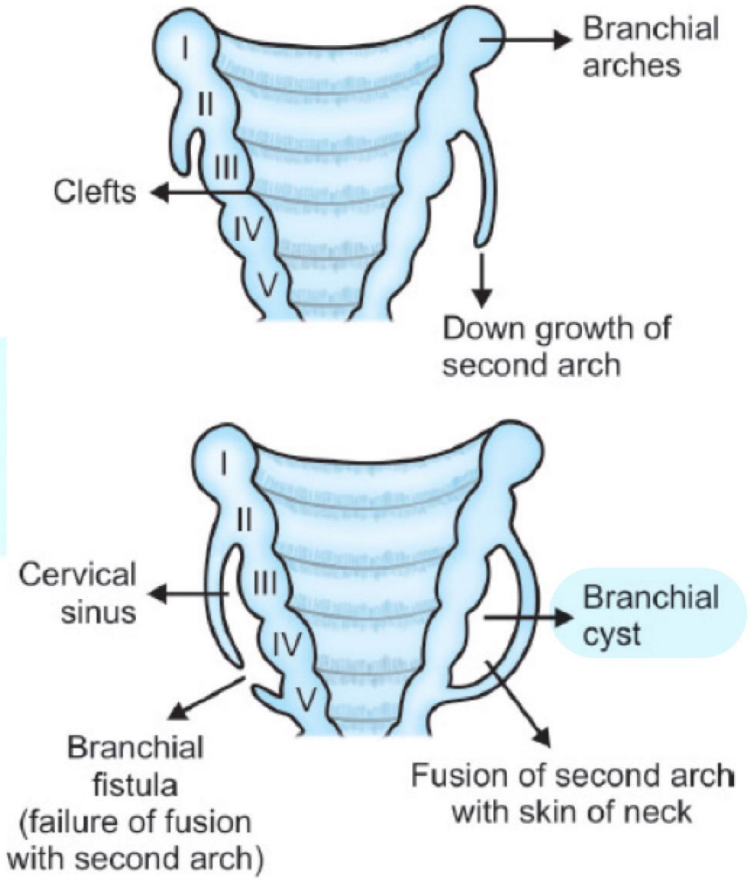


Presentation

First -

- Duplications of the external auditory canal

- Cyst, sinuses and fistula between the external auditory canal and angle of the mandible.

Second - **Most common in second**

They present as a cyst (branchial cyst), sinus or fistula anterior to the SCM in the lower neck.

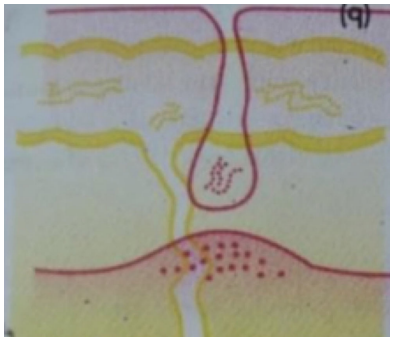
****branchial cyst:: seen in lateral side of neck or in upper SCM muscle.**

Third and fourth - These may present with recurrent neck infections or abscess, suppurative thyroiditis and enlarging cyst or abscess may cause airway compromise, or dysphagia.

Branchial sinuses

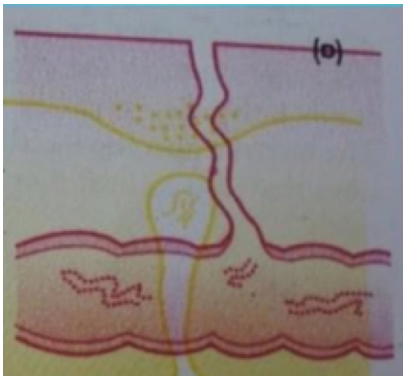
* Opening at the skin over the anterior border of the sternomastoid

- Arising from the 2nd branchial cleft
- More common than fistula
- Mucoid or purulent discharge



Branchial fistula

* One end open into the tonsillar fossa ,and the other in the skin over the anterior border of the lower third of sternomastoid muscle



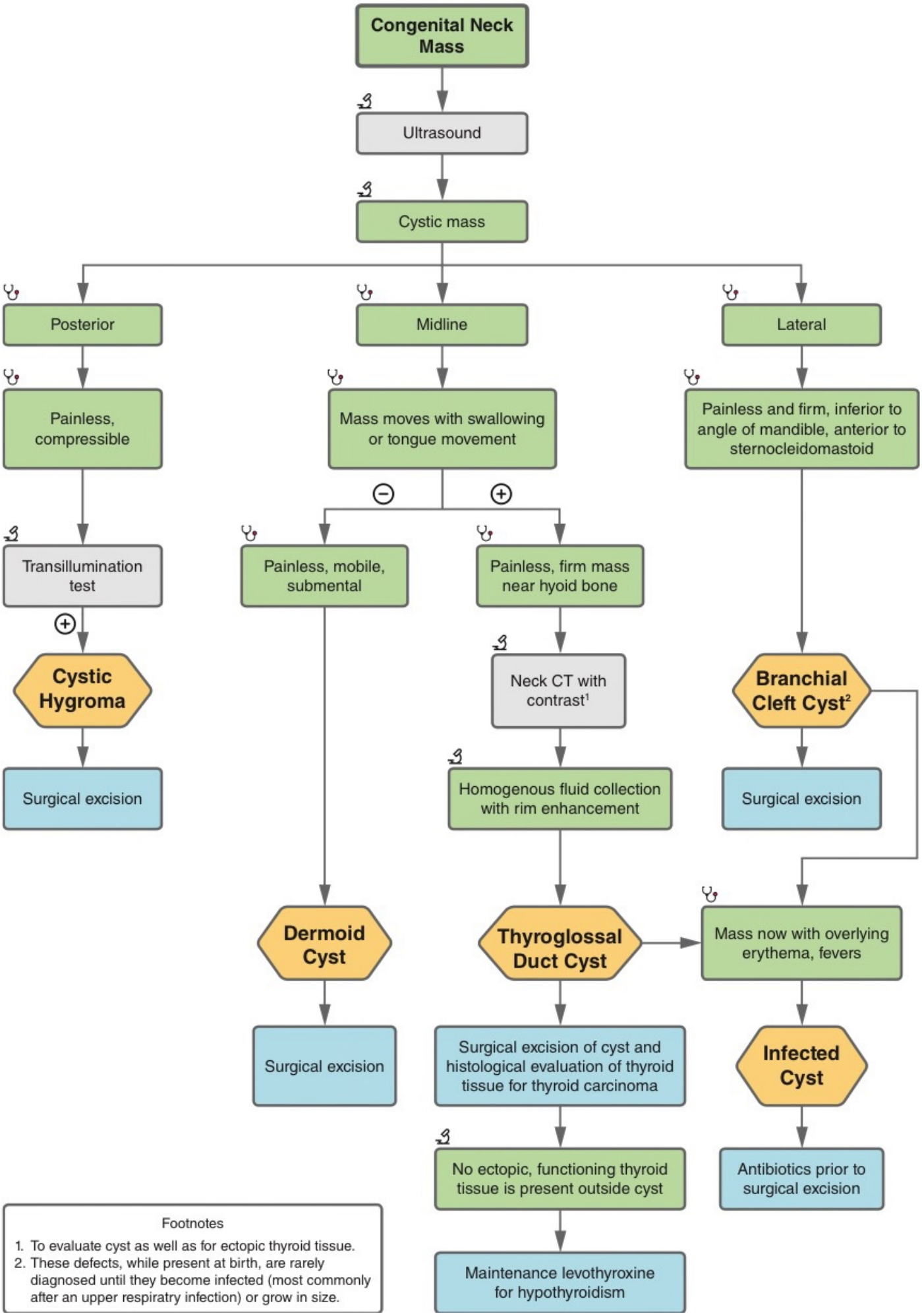
Indications for surgery

Infection
Sinus drainage
Possible malignant transformation.

Differential diagnosis

Thyroglossal duct cyst (the most common)
Dermoid
Lymphatic malformation
LNE (lymphnode enlargement)
Parotid cyst (first branchial lesions).

	1)Thyroglossal cyst and fistula	2)Branchial cyst and fistula
Aetiology	• The thyroglossal duct develops from the foramen caecum and descends into the neck to give rise to the thyroid isthmus and adjoining parts of lateral lobes. Then it is obliterated and disappears. • Thyroglossal cyst is tubulo-dermoid cyst develops from unobliterated portion of this duct.	• The neck develops from 5 pharyngeal arches. A process descends from the 2nd arch and fuses with the 5th arch. • The space enclosed is called cervical sinus which obliterate later on. • Branchial cyst is a congenital cyst due to persistent potency of cervical sinus. • Failure of this fusion → congenital branchial fistula
Pathology	1- Outer fibrous layer & the wall is rich in lymphoid tissues.	
	2. The cyst and fistula are lined by columnar epithelium.	2. The cyst and fistula are lined by stratified squamous epithelium.
	3. The content of the cyst or the dischage of fistula are mucoid fluid or pus if infected	
		• The mucoid content is rich in cholesterol crystals (appears microscopically as square minus a corner).
	4. A fibrous cord connect the cyst to the formen caecum at the base of the tongue. It may pass in front, behind or through the body of the hyoid bone.	4. A fibrous cord or rarely open duct, passing between bifurcation of common carotid artery connect the cyst to supra-tonsillar fossa (fossa of Rosenmuller).


Congenital Neck Mass
Ultrasound
Cystic mass
Posterior
Midline
Lateral
Painless, compressible
Mass moves with swallowing or tongue movement
Painless and firm, inferior to angle of mandible, anterior to sternocleidomastoid
Transillumination test
Painless, mobile, submental
Painless, firm mass near hyoid bone
Cystic Hygroma
Neck CT with contrast[1]
Branchial Cleft Cyst[2]
Surgical excision
Homogenous fluid collection with rim enhancement
Surgical excision
Dermoid Cyst
Thyroglossal Duct Cyst
Mass now with overlying erythema, fevers
Surgical excision
Surgical excision of cyst and histological evaluation of thyroid tissue for thyroid carcinoma
Infected Cyst
No ectopic, functioning thyroid tissue is present outside cyst
Antibiotics prior to surgical excision
Maintenance levothyroxine for hypothyroidism
Footnotes
1. To evaluate cyst as well as for ectopic thyroid tissue.
2. These defects, while present at birth, are rarely diagnosed until they become infected (most commonly after an upper respiratry infection) or grow in size.

USMLE step 2 Q :

A 7-year-old F with PMH of tonsillitis presents with throat pain. Exam shows a non-tender, mid-line mass that elevates when she swallows. Labs reveal high TSH and low free T4 levels.

→ TGDC

An 11-year-old F with PMH of asthma presents with cough and sore throat. Exam shows a painless, firm mass anterior to the sternocleidomastoid that does not move when she swallows. On imaging, US shows a mass with homogeneous low echogenicity.

→ BRANCHIAL CLEFT CYST

Archive

1. **Regard branchial cyst, all true except.**
   a. May be confused with nodal metastasis.
   b. Is treated by aspiration.
   c. Is derived from ectoderm of the second branchial pouch.
   d. Is lined with squamous epithelium and contains cholesterol crystals.
   e. May complicated with branchial fistula.

2. A 13 years old boy presented with a rounded painless swelling in the midline of the anterior aspect of his neck that moves with deglutition and with protrusion of the tongue is mostly having:

A. A solitary thyroid isthmus nodule
B. Sublingual dermoid cyst
C. Thyro-glossal cyst
D. Carotid body tumor
E. Chondroma of the thyroid cartilage

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3. Regardless of age or sex the most common neck swelling is:

A. Lymph node enlargement
B. Goitre
C. Thyroglossal cyst
D. Aneurysm of carotid artery
E. Salivary gland tumor

4. The following are differential diagnosis of midline neck swelling.

a. Sub mental lymph node.
b. Laryngocele.
c. Branchial cyst.
d. Submandibular cystic hygroma.
e. Carotid body tumor.

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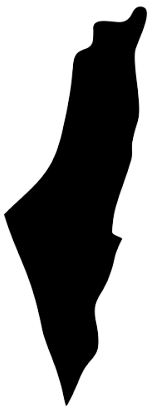

اللهُمَّ انصُر أهل غَزَّة وثبِّت أقدامهم.
اللهُمَّ احرُس أهل غَزَّة بِعينك التي لاتَنام.
اللهُمَّ كُن لأهل غَزَّة عوناً ونصيراً، وبدِّل خوفهم أمناً وأماناً.

اللهُمَّ اجبُر كسرهم، واشفِ مَرضاهم، وتقبَّل شُهداءَهم بِرحمتكَ.
اللهم سخر لهُم ملائكة السماءِ وجُنودَ الأرض...
اللهم انصرهم على من عاداهم وافتح لهم فتحًا قريبًا.