Congenital Diaphragmatic Hernia (CDH)

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)
  - 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

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Inadequate closure of the pleuroperitoneal canal allows the abdominal viscera to enter the thoracic cavity when they return from the extraembryonic coelom to herniate into the chest.

As a result, pulmonary hypoplasia develops (decrease in bronchial divisions, bronchioles, and alveoli)

Affects the contralateral lung as well

The pulmonary arteries exhibit a decrease in density as well as an increase in muscularization (**CDH-ASSOCIATED PULMONARY HYPERTENSION**)

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

Male infants more common

90% are posterolateral or “Bochdalek”

- Left sided (80%)
- Right sided (19%)
- Bilateral (1%)

10% are located anteriorly, termed “Morgagni” hernias

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).

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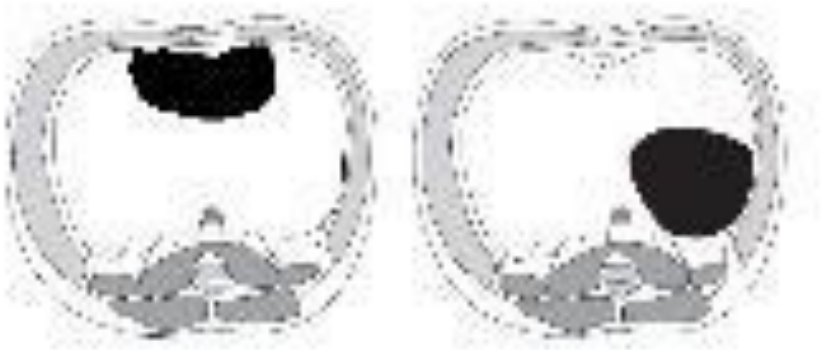

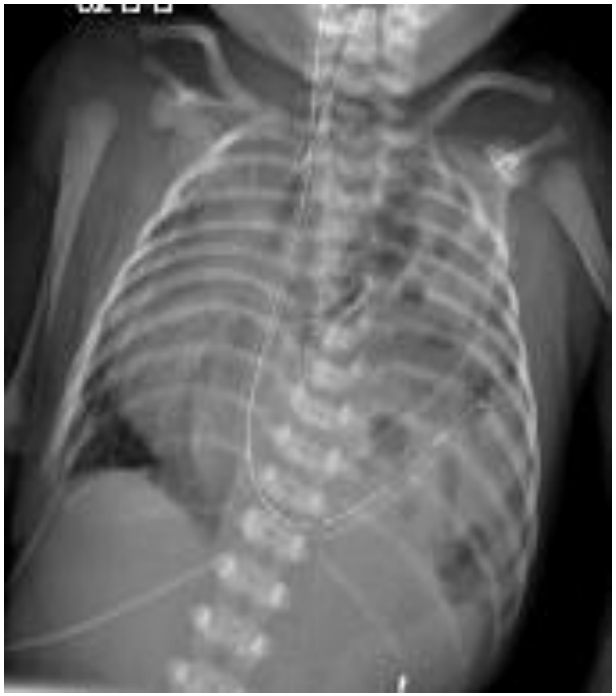

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Diagnosis

PRENATAL DIAGNOSIS:

Approximately 50–70% of infants

Fetal US features include:

Polyhydramnios, intrathoracic fluid-filled bowel loops, mediastinal shift, and/or an intrathoracic stomach, spleen, or liver

The lung-to-head ratio (LHR) is a prenatal US assessment ratio, utilizing the contralateral lung area to the head circumference, which predicts CDH severity.

If the LHR is 1 or less, the prognosis is poor.

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
- Bowel sounds may be auscultated within the thoracic cavity
- Decrease in breath sounds bilaterally

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Postnatal diagnosis

Chest radiograph:
- 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% 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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Treatment

RESUSCITATION AND STABILIZATION

Prompt endotracheal intubation (without bag mask ventilation)

Initiation of conventional mechanical ventilatory support

A nasogastric tube should be inserted to avoid gastric and intestinal distention

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)

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Poor prognosis

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

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Neck anomalies

THYROGLOSSAL DUCT CYST (TGDC):

The foramen cecum is the site of the development of the thyroid diverticulum.

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

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Presentation

Midline painless mass at or just below the hyoid bone, could be lateral

On physical examination, the TGDT is smooth, soft, and nontender.

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 when the tongue protrudes.

This maneuver is more reliable than asking the child to swallow

thyroglossal cysts can become infected with oral flora and present as draining sinus from spontaneous or incisional drainage of an abscess

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- 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 and the small risk (<1%) of cancer (papillary thyroid carcinoma)

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 as described by Sistrunk (Decrease risk of recurrence)

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complications of the Sistrunk procedure

Major complications include recurrence, hematoma or abscess, entry into the airway (tracheotomy), hypoglossal nerve paralysis, hypothyroidism (ectopic thyroid).

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

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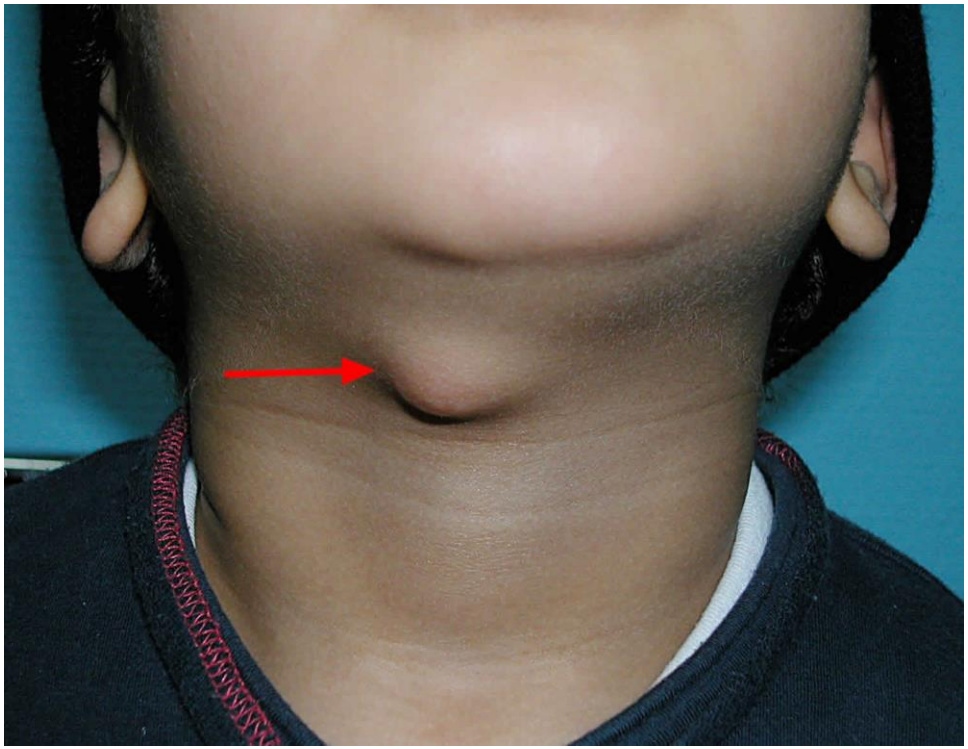

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Branchial Anomalies

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

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

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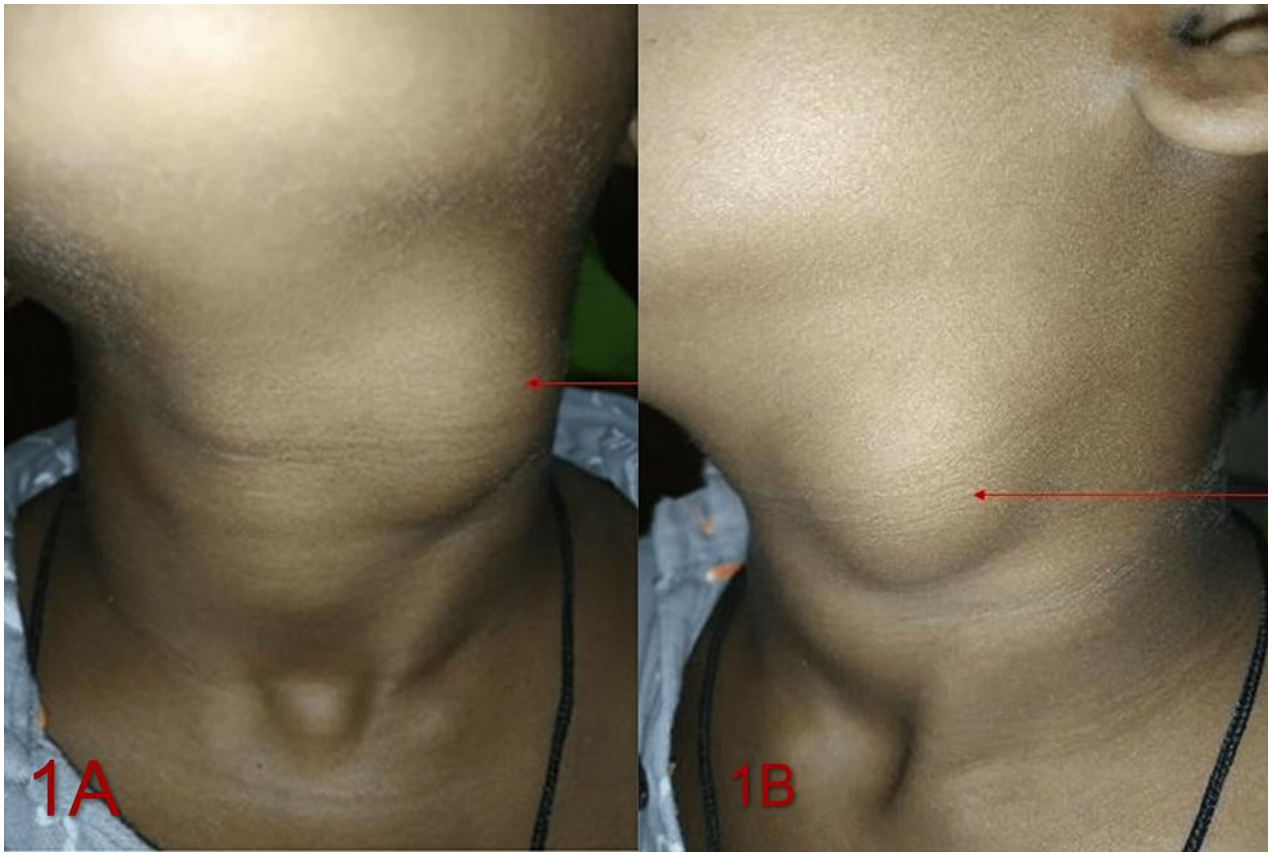

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Branchial arches development

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.

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Second -

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

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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.

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Indications for surgery

Infection
Sinus drainage
Possible malignant transformation.

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Differential diagnosis

Thyroglossal duct cyst
Dermoid
Lymphatic malformation
LNE
Parotid cyst (first branchial lesions).

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