Chest wall and thoracic organs

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Chest wall deformities

Three categories of deformities:

I. There is rib overgrowth causing either depression (Pectus excavatum) or protrusion (Pectus carinatum) of the anterior chest wall.

II. Failure of normal development (aplasia or dysplasia) - bifid sternum, (with ectopia cordis, defects of heart, pericardium, diaphragm, abdominal wall = Pentalogy of Cantrell) – Poland’s syndrome (unilateral absence of ribs, pectoralis muscles and breast tissue)

III. Deformities due to trauma or pressure effects (iatrogenic after ribs resections in pectus excavatum - *acquired asphyxiating chondrodystrophy* )

Frequency increased in familial connective tissue disorders such as Marfan`s Syndrome and Ehler`s-Danlos Syndrome, systemic weakness of connective tissue and poor muscle development = scoliosis in 20% of patients.
Pectus excavatum

*Pectus excavatum*- depression of the anterior chest wall of variable severity and configuration (localised and deep = cup-shaped, diffused = saucer-shaped, eccentric=grand canyon, or mixed excavatum/carinatum = horseshoe, pouter pigeon)

Incidence: 1:300-1:1000 (more common in European and Asian than African)
Etiology- genetic predisposition, abnormal distribution of colagen types in ribs cartilages.

40% have a positive family history.
Boys / Girls = 4 / 1

Patients usually tall – basketball players posture, 18% have scoliosis, 16% features of Marfan`s Syndrome, 1%- Ehlers-Danlos Syndrome
Pectus excavatum
Pectus excavatum – clinical features

- Symptoms: exercise intolerance (80%), chest pain (65%), lack of endurance (65%), shortness of breath (45%), asthma-like symptoms (30%), frequent URI (25%), scoliosis (18%).

- Teenagers: poor body image, stop in team sports = isolation and feelings of worthlessness (even suicidal thoughts)!

- On physical exam: „old age” or „pectus posture”
The Haller CT index: objective measurement of deformity severity
It divides the internal transverse diameter of the chest by the anteroposterior diameter. In normal patients the index is less than 2.

The sternum is frequently twisted so that it is at a 45% angle to the anterior chest wall.
Chest computed tomography (CT) scan with Haller index greater than 3.25 = indicative of a severe depression
Indications for operation

- Effects of cardiac compression (decreased cardiac output, impaired valve function and arrhythmias)
- Pulmonary effects (restrictive lung disease, atelectasis, paradoxical respiration)
- CT index of 3.25 or greater
- Failed previous repair

Ideal age: just before puberty
Postoperative chest X-ray after Ravitch method
Minimally invasive pectus excavatum repair (Nuss method)
95% of patients have good to excellent long-term effect after minimally invasive pectus repair (Nuss method)
Pectus carinatum

• protrusion deformity of the chest
• much less common than the excavatum deformity
• the protrusion may involve: upper chest – manubrium, lower chest – gladiolus, unilateral, bilateral or mixed
• more common in boys (4:1), 25% have a positive family history
• appears in late childhood and progresses rapidly during puberty
• complains of chest pain when pressure applied on anterior chest, no cardiac or pulmonary compression
• application of a pressure brace or surgical resection (depending on PCI)
Pectus carinatum
Pectus carinatum
Poland`s Syndrome

Described by Alfred Poland, may include absence of all or some of:
• ribs
• pectoralis major
• pectoralis minor
• serratus anterior
• rectus abdominis
• latissimus dorsi
• nipple deformities
• limb deformities (syndactyly, brachydactyly)
• absent axillary hair
• limited subcutaneous fat

Indications for surgical intervention: large rib defects causing lung hernia, concerns regarding injury to the heart or lungs. Adolescent girls require breast reconstruction in case of amastia.
Pleural Effusion and Empyema

Pleural effusion means the accumulation of liquid in the pleural space. It may be:

- Blood
- Chyle (Lymph)
- Transsudate
- Exudates
- Pus

- Transsudates: protein content less than 30g/L and total leukocyte count less than 2000/mm3
- Chyle: rich in lymphocytes and contains chylomicrons
- Empyema = accumulation of pus in a cavity (...thoracic)
Primary fetal hydrothorax in 1:10000 – 15000 pregnancies

- Effusion may be secondary to mediastinal tumor, adenomatoid tumor, pulmonary sequestration, infection, chromosomal anomaly.

- Congenital primary chylothorax – debatable

- Rarely effusions may arise in the context of lymphangiomatosis (present at birth or manifest later)
Aquired Forms

- Hemothorax – usually traumatic, mostly associated with pneumothorax, may occur as a complication of the central venous line catheterisation.

- Chylothorax – damage of thoracic duct, and increased pressure in the systemic venous system, incidence is rising (from 1% to 5%).

- Hydrothorax – iatrogenic (CVC, right-sided: transdiaphragmatic- ventriculoperitoneal shunting or peritoneal dialysis, from urinary ascites)
  - noniatrogenic (in cases of hepatic ascites, congestive heart disease, nephrotic syndrome, malignancy: especially non-Hodgkin lymphoma, metastatic disease)
Pleural Exsudate and Empyema

• May be result of: trauma of the chest, esophageal foreign body perforation, after surgery- leaking bronchial stump and esophageal anastomotic leak. Also infradiaphragmatic pathology: retained gallstones after cholecystectomy, pancreatitis, appendiceal perforation with peritonitis.

• Increasingly common – parapneumonic effusion (due to pneumonic disease)- 14 in 100000 (Streptococcus pneumoniae, Haemophilus influenzae, Ebstein-Barr virus). In third world countries: Staphylococcus aureus. In endemic areas: tuberculosis, echinococcus.
Pleural Empyema

Classically, three stages are distinguished:

- Exudative phase (few cells in the fluid) - 24-72 hours
- Fibrinopurulent phase (accumulation of fibrinous material, loss of lung mobility) - 7-10 days
- Organizing phase (formation of a pleural peal)

- Clinical features: respiratory symptoms (tachypnea, dyspnea, ortopnoe); dullness on the affected side with diminished breath sounds, signs of inflammation (fever, tachycardia, lethargia, pain during respiration)
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Pleural Empyema – Imaging:

- plain chest X-ray in AP position
- ultrasound
- CT and MR scanning
Pleural Empyema – Imaging:
Pleural Empyema – management:

- Antibiotics (adjusted according to culture results)
- Thoracocentesis
- Tube thoracostomy
- Intrapleural fibrinolytics
- Decortication (thoracoscopically or through thoracotomy)

Loculated empyema should be treated surgically!
Pleural Empyema – management:

Decortication - thoracoscopically
Pleural Empyema – management:

Decortication through thoracotomy
Pneumothorax

• Air trapped in the hemithorax

• Spontaneous or posttraumatic (due to rib fracture and laceration, deceleration injury, crush injury or due to increased intrathoracic pressure causing pulmonary rupture)

• In newborns usually caused by barotrauma

• In younger children: search for underlying congenital broncho-pulmonary malformations. Other causes: advanced pulmonary infections, after forced cough

• In older children: rupture of the pulmonary alveolar wall – causing formation of subpleural blebs.
TENSION PNEUMOTHORAX
- Can quickly result in cardiovascular demise if tension within hemothorax develops
- Findings: hyperresonance, diminished breath sounds, tracheal deviation, hemodynamic instability
- Potentially lethal – chest x-ray not required, may waste time!
- Needle decompression followed by placement of chest tube
- Large bore angiocath in the second intercostal space along mid-clavicular line
- Alternative decompression site! – anterior axillary line just above the sixth rib
- Reexpansion of the lung, evacuation of the air and fluid, sealing of the injured lung, resolving of the air leak...
Pneumothorax- treatment:

- Tension pneumothorax – immediate decompression

- Small pneumothorax – conservative treatment

- Most often - chest tube drainage

- In case of persistent air leakage – consider operative treatment

- Primary spontaneous pneumothorax- in case of second episode: thoracoscopic pulmonary apical resection with partial pleurodesis
VATS – in refractory spontaneous pneumothorax

zestaw trójbutlowy

zastawka wodna
kontrola ciśnienia ssania
zbiornik płynu

od pacjenta do atmosfery do ssania

2 cm 20 cm
Pulmonary abscess

- Accumulation of purulent content in the cavity formed as a result of pulmonary parenchyma necrosis and destruction.

- Most common pathogens: aerobic (Staphylococcus aureus, Streptococcus pn, Streptococcus pyogenes, Klebsiella pn, Escherichia coli, Pseudomonas aer), anaerobic (Fusobacterium, Bacteroides, Peptococcus, Bacillus fragilis).

- Pathogenesis of pulmonary abscess development: aspiration of naso-pharyngeal or gastric secretions; retention of purulent mucus in bronchioles, bacteraemia, diffusion through continuity, complication of the other pulmonary conditions: infections, bronchial dilatations, cysts.

- Primary vs secondary abscess.
- Chronic abscess – duration >6 weeks.
Pulmonary abscess
Pneumatocele

• Usually in the course of stapylococcal pneumonitis

• thin-wall spaces filled with air, as a result of the purulent process and staphylococcal toxins

• varies in size and localization

• Complications: pneumothorax, bronchio-pleural fistulae, pulmonary abscess, pleural empyema
Pneumatocele - complete resolution possible but remember about complications!
Pathologies of mediastinum and thymus, esophagus, diaphragm
Compartments of the mediastinum
Anterior mediastinum:

<2 Years: Benign teratoma, Thymic hyperplasia, Cystic hygroma

>2 Years: Malignant germ cell tumor, Thymoma, Lymphoma
Middle mediastinum:

<2 Years: Bronchogenic cyst

>2 Years: Lymphoma, Granuloma
Posterior mediastinum:

<2 Years: Neuroblastoma, Enterogenous cyst

>2 Years: Ganglioneuroma, Sarcoma
Esophageal Atresia and Tracheoesophageal Fistula (EA TEF)
EA TEF:

- Incidence: 1 in 3000 to 4500 births (male predominance 3:2)

- most cases sporadically, can be genetically determined (trisomy 13 and 18 in 6-10% cases)

- etiology: major role of Sonic hedgehog signalling pathway (involved in the normal morphogenesis of vertebra, differentiation of the trachea and esophagus and other organs)
EA TEF associated malformations:

- in range of 50-80%
- 20-70% musculoskeletal malformations
- 20-50% cardiovascular (VSD, ASD, FT, CoA)
- 15-25% genitourinary
- 15-25% gastrointestinal (ARM, DA, malrotation, intestinal atresia)
- 5-10% chromosomal anomalies
Anatomical classification (Vogt/Gross)

2/A (7%)

3a/B (2%)

3b/C 85%

3c/D (3%)

4/E (3%)
EA TEF:

• Preoperative Management

• Diagnosis (Clinical features and Radiological diagnosis)

• Operative Management
Operative treatment of EA TEF
Operative treatment of EA TEF

Livaditis myotomy
Long-gap esophageal atresia

Kimura`s multi-staged elongation of proximal esophagus
EA TEF:

- Postoperative treatment

- Early complications (recurrence of TEF, anastomotic leak, anastomotic stenosis, tracheomalacia)

- Late complications (GERD, microaspirations, abnormal peristaltic problems of the lower esophageal segment)
EA TEF risk classification:

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<th>Birthweight</th>
<th>Waterstone</th>
<th>Survival (%)</th>
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<tr>
<td>Birthweight &gt;2500g</td>
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<td>100</td>
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<tr>
<td>Birthweight 2000-2500g, mild pneumonia or moderate cardiac anomalies</td>
<td>B</td>
<td>85</td>
</tr>
<tr>
<td>Birthweight &lt;2000g, severe pneumonia or severe cardiac anomalies</td>
<td>C</td>
<td>65</td>
</tr>
</tbody>
</table>
**EA TEF risk classification:**

<table>
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<tr>
<th>Birthweight</th>
<th>Spitz</th>
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<tbody>
<tr>
<td>Birthweight &gt;1500g</td>
<td>I</td>
<td>97</td>
</tr>
<tr>
<td>Birthweight &lt;1500g or major cardiac anomaly</td>
<td>II</td>
<td>59</td>
</tr>
<tr>
<td>Birthweight &lt;1500g And major cardiac anomaly</td>
<td>III</td>
<td>22</td>
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Congenital Diaphragmatic Hernia (CDH)

- defect in postero-lateral diaphragm (foramen of Bochdaleck), through which the abdominal viscera migrate into the chest during fetal life
- incidence: 1 in 2500-5000 births
- 80% of posterolateral occurs on the left side
- Size of defect varies
- Incidence of associated anomalies – 40% (cardiovascular, skeletal, CNS, genitourinary, gastrointestinal, craniofacial, abdominal wall defects, chromosomal defects)
- Despite advances have high mortality attributed to pulmonary hypoplasia and persistent pulmonary hypertension
Congenital Diaphragmatic Hernia (CDH)

- **Etiology:** generally sporadic, multifactorial, syndromic or nongenetic factors (thalidomide, nitrofen, vitamin A deficient diet)
- **Embryogenesis:** failure of closure of the pleuroperitoneal canal (…have to be revisited…)
- **Diagnosis:** antenatal, at birth- first 24 hours of life, 10-20% later presentation)
Congenital Diaphragmatic Hernia (CDH)

- Prognostic factors of poor prognosis: polyhydramnios, intrathoracic stomach and liver, lung to head ratio (LHR <1.0; >1.4)
- Management: preoperative- paralysed and sedated, gentle ventilation and permissive hypercapnia to minimize barotrauma, inhaled nitric oxide, High-frequency oscillatory ventilation (HFOV), extracorporeal membrane oxygenation (ECMO)
Congenital Diaphragmatic Hernia (CDH)

- Timing of Surgery
- Surgical Technique
- Postoperative treatment
- Long term follow-up: pulmonary hypoplasia, bronchopulmonary dysplasia, persistent pulmonary hypertension, reactive airway disease, neurodevelopmental delay (motor and cognitive skill), GERD< sensorineural hearing loss, musculoskeletal: chest asymmetry, pectus deformity, recurrence)
Congenital Diaphragmatic Hernia (CDH)
Congenital eventeration of the diaphragm

- abnormally high position of the hemidiaphragm
- congenital or acquired (result of phrenic nerve palsy)
- in congenital: muscular aplasia secondary to nerve damage
- clinical feature: asymptomatic to severe respiratory distress (repeated attacks of pneumonia, bronchitis or bronchiectasis)
- diagnosis: Fluoroscopy X-ray (paradoxical movement of diaphragm, ultrasonography and CT most useful in the diagnosis.
Congenital eventration of the diaphragm
Eventeration repaired by plication of the diaphragm
Bronchopulmonary sequestrations

- one-third of congenital pulmonary malformations
- either an intrathoracic or subdiaphragmatic mass of nonfunctional pulmonary tissue that lacks communication with the tracheo-bronchial tree
- Extralobar or intralobar
- arterial blood supply arising from descending aorta, venous drainage of intralobar via the pulmonary veins, of extralobar- via either the azygous or hemiazygous veins, IVC or directly into atrium (in 20% blood supply from infradiaphragmatic source)
- Diagnosis: CT or MRI
- 90% of Extralobar – left hemithorax, 60% of Intralobar – left lower lobe, 10-15% in upper lobe, bilateral infrequent
Bronchopulmonary sequestrations

Clinical features:
- extralobar often diagnosed prenatally or in infancy (during surgery for CDH..)
- intralobar often in later childhood – recurrent pulmonary infections or haemorrhage,
- both can present in the newborn period with respiratory distress due to mass effect or congestive heart failure because of arteriovenous shunting

Management:
- Resection (straight-forward in extrapulmonary, for intralobar often lobar resection)
Congenital Cystic Adenomatoid Malformation of the lung (CCAM)

- benign hamartomatous or dysplastic tumors
- morphologically: overgrowth of terminal bronchioles in a glandular or adenomatoid pattern – composed of disorganized cysts lined with ciliated cuboidal or columnar epithelium (grossly: have both cystic and solid components)
- CCAM constitute 10-30% of all congenital lung malformations with slight male predominance
- Normal pulmonary arterial and venous blood supply
- Diagnosis: relatively common prenatally by sonography (echogenic pulmonary mass), often with associated polyhydramnios, mediastinal shift, pleural effusions and fetal hydrops…
- macrocystic or microcystic ( > 5mm<)
- differential diagnosis: CDH, pulmonary sequestration, bronchogenic cyst…
- high accuracy MRI (prenatally), contrast enhanced CT (preoperatively: anatomy, aberrant systemic blood supply…)
Congenital Cystic Adenomatoid Malformation of the lung (CCAM)

- Clinical features: - newborn (respiratory distress, asymptomatic), - childhood or adolescents (infectious complications, pneumothorax, bronchiectasis) – adulthood (malignancy: bronchioalveolar cancer, sacoma, pulmonary blastoma, mesenchymoma)

Management
- Fetal therapy (EXIT procedure in third trymester, ultrasound guided thoracoamniotic shunting for a macrostystic CCAM)
- Postnatal therapy (complete spontaneous resolution in 4%, resection should be performed in any persistent CCAM to prevent complications)
- Resection performed through open thoracotomty or minially invasive
Endoscopic lobectomy (CCAM type II)
Congenital lobar emphysema (CLE)

- Congenital lobar overinflation – characterized by expiratory air trapping within the affected lobe
- Overdistension leads to compression of adjacent lung and mediastinal shift
- Etiology: focal absence of cartilaginous components, endobronchial obstructions with secretions, granulation tissue, ingested foreign bodies or endobronchial tumors (intrinsic causes)
- Extrinsic causes of compression (mediastinal lymphadenopathy or aberrant pulmonary artery, mediastinal cyst or tumor)
- CLE most commonly in full term infants, but in prematures acquired form (barotrauma, oxygen toxicity, lung immaturity)
- Lobar resection is curative, in 10-15% life-saving emergency thoracotomy
Aquired emphysema in premature infant:
Brochogenic cyst (BC)

- Result from abnormal budding of the bronchial tree in which a portion of the lung bud develops independently.
- The cyst walls frequently contain cartilage and are lined with ciliated columnar epithelium.
- These lesions tend to enlarge causing airway obstruction.
- Plain chest X-ray may suggest the presence of BC, but CT scan confirms the diagnosis.
- Resection is curative.
Bronchogenic cyst resection
Endoscopic removal of cystic mediastial mass:
The end...