Pediatric Cardiothoracic Surgery

Bjarni Torfason, dósent, yfirlæknir

Háskóli Íslands Læknadeild og
Landspítalinn

Cardiothoracic Surgery

Undirsérgreinar:

- Adult Cardiac Surgery
- General Thoracic Surgery
- Pediatric Cardiothoracic Surgery
- Transplantation

Pediatric Cardiothoracic Surgery

- Thorax
  - Veggur
  - Pleura
  - Lungu
  - Mediastinum
- Hjarta og æðar í thorax
  - Meðfæddir galla
  - Áunnir gallar/ sjúkdómar

Pediatric Cardiothoracic Surgery

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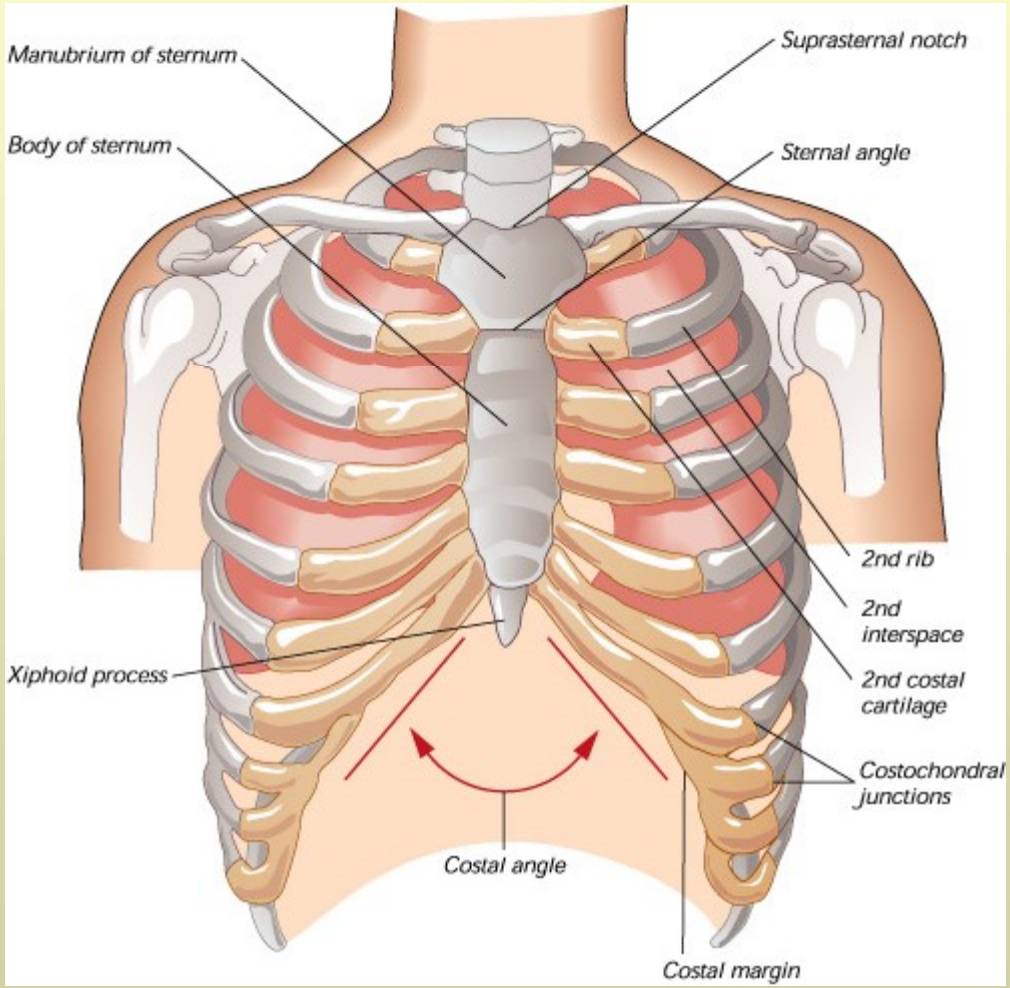
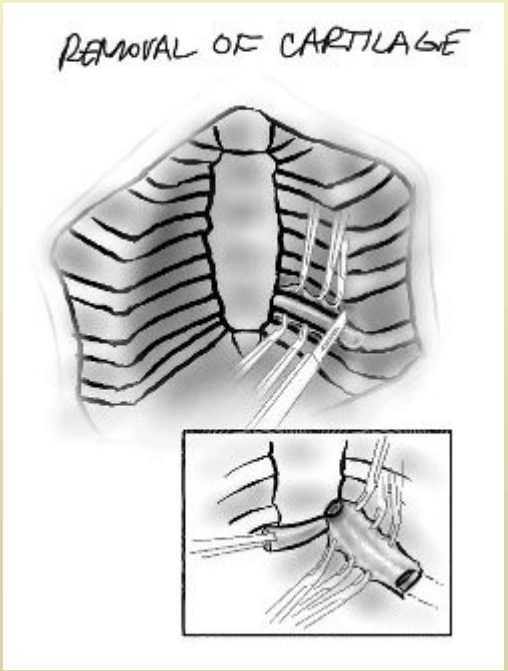
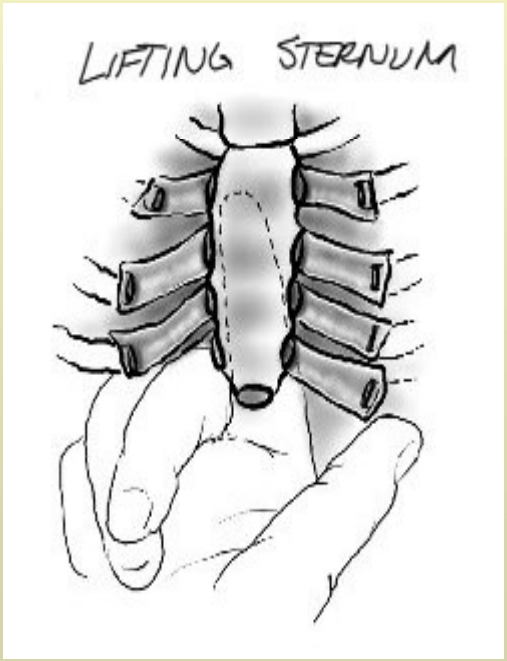

Manubrium of sternum
Body of sternum
Xiphoid process
Suprasternal notch
Sternal angle
2nd rib
2nd interspace
2nd costal cartilage
Costochondral junctions
Costal angle
Costal margin

Ravitch:
Pectus Excavatum (holubringa) &
Pectus Carinatum (bungubringa)

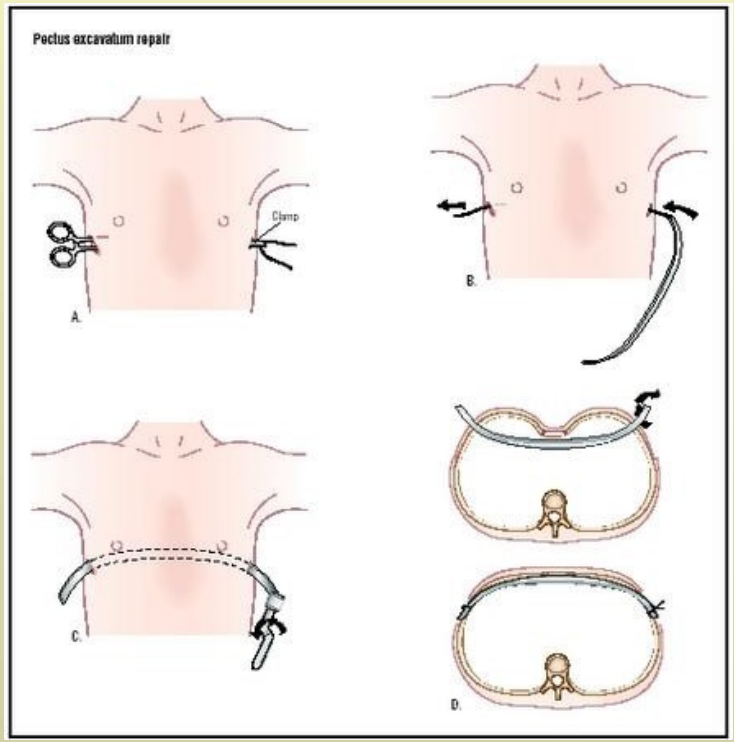
Ravitch Technique (1940's) "opin aðgerð"

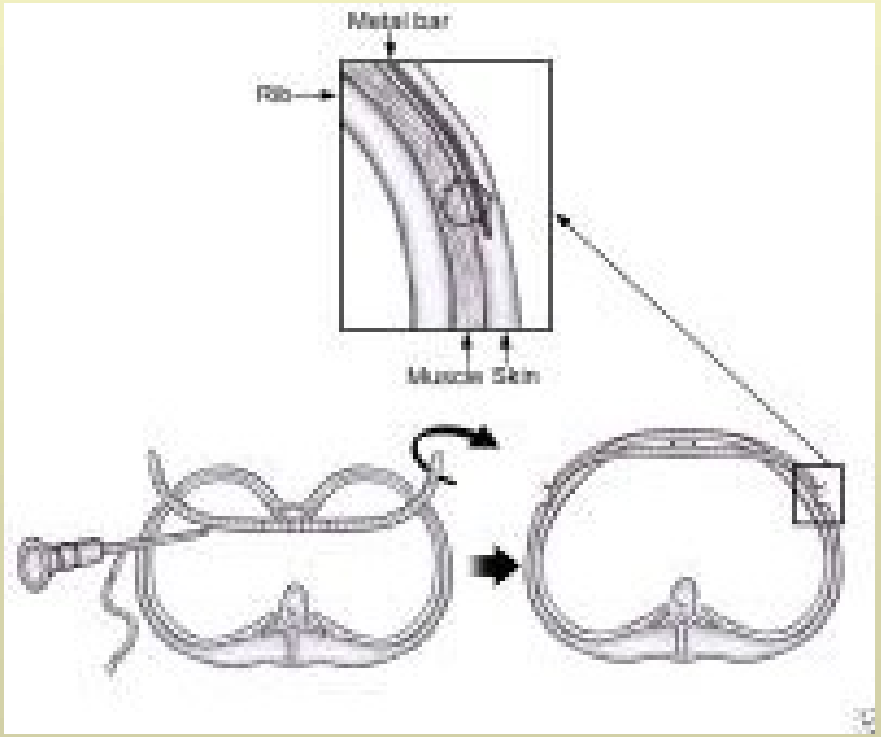




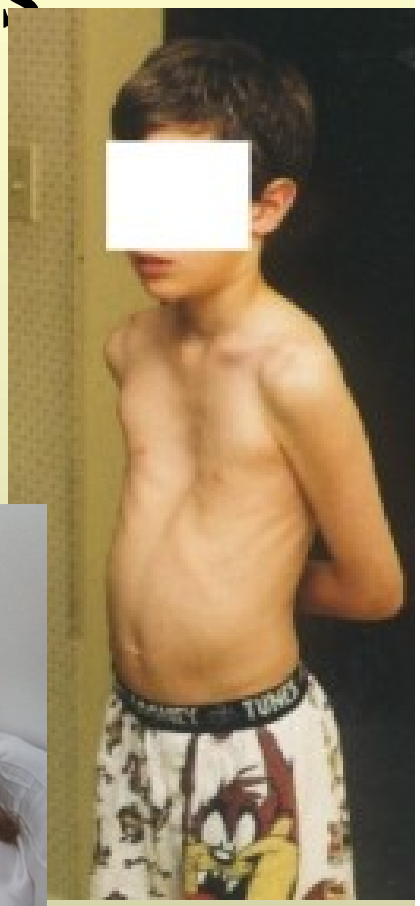
Pectus Excavatum, holubringa

Nuss Technique (1987), “brjóstholsspeglun”





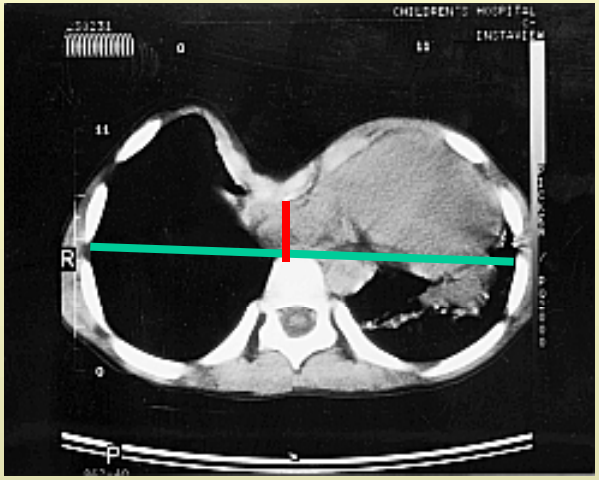
Pectus excavatum - Nuss



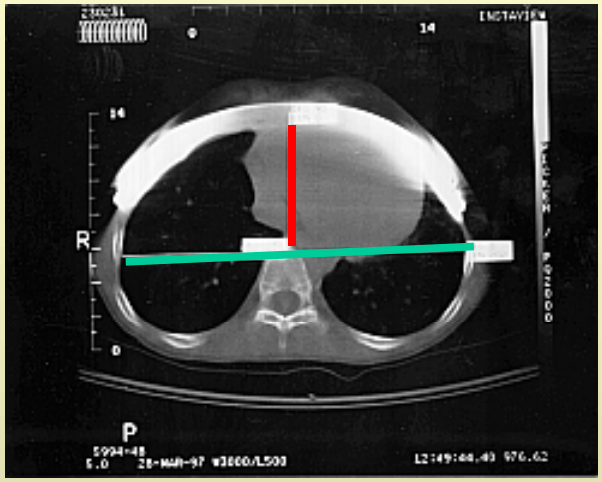
„Haller Index“

HI: úrelt viðmið b/a > 3,2 ?)

Nú: Einkenni + deformitet á hjarta:



HI=8,0

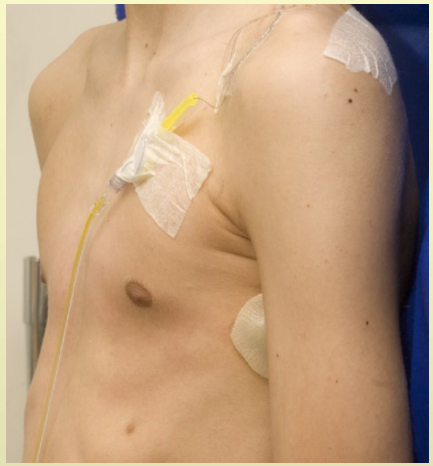


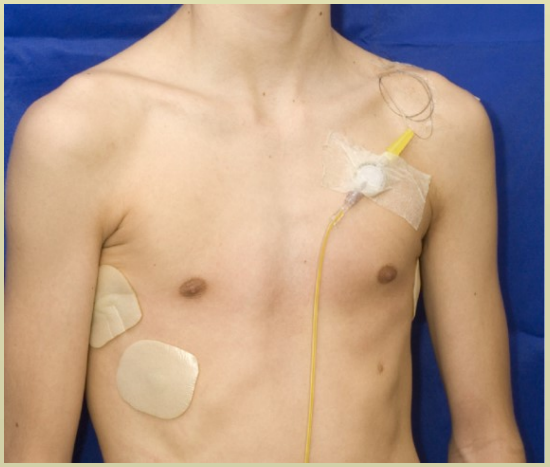
HI= 3,2

Holubringa

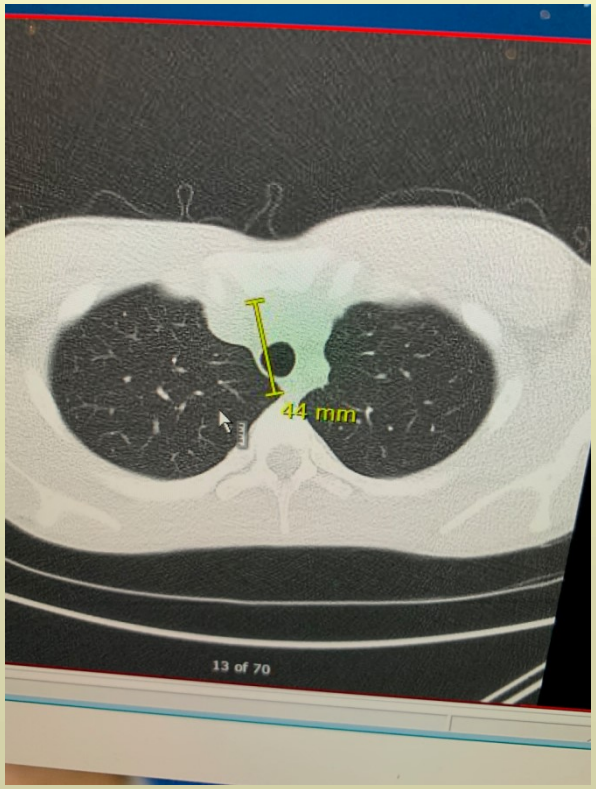
Preop

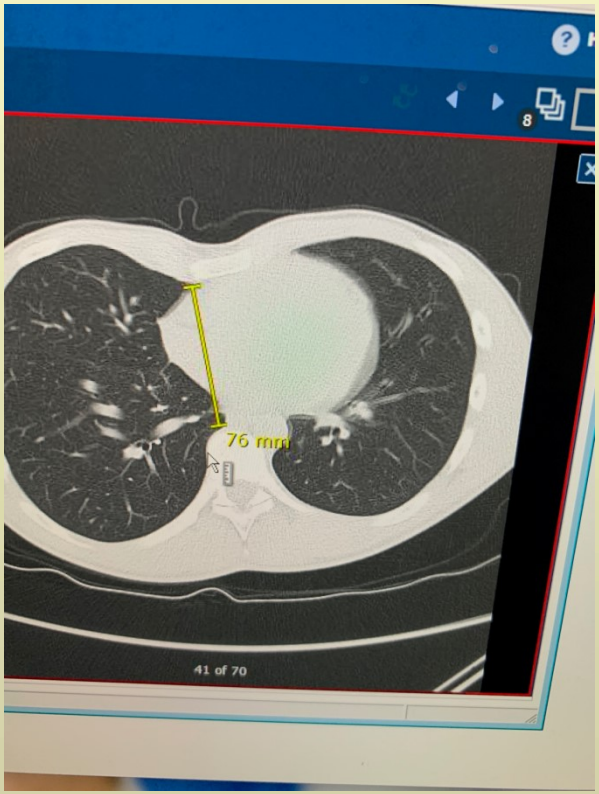
Post op Nuss



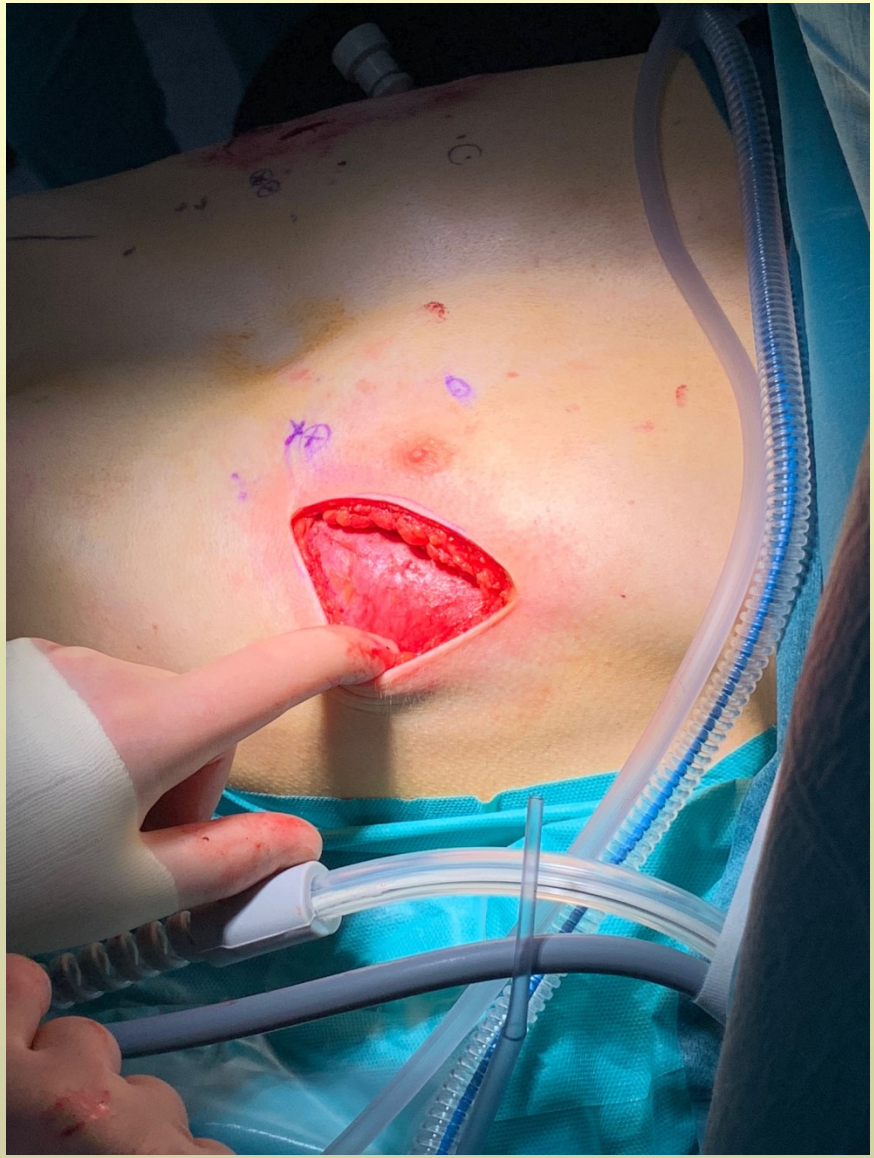


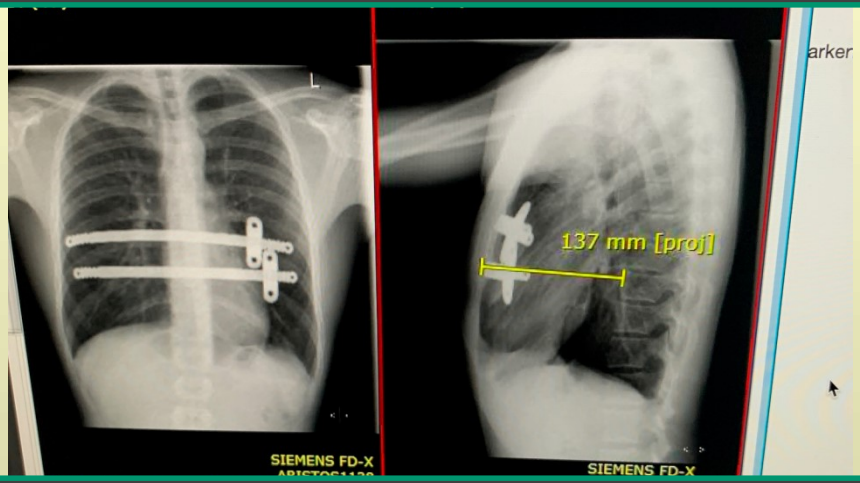
PE sjúkratilfelli Case #1:
Einkenni Þrekleysi og lengi að borða





Intercostal Cryo

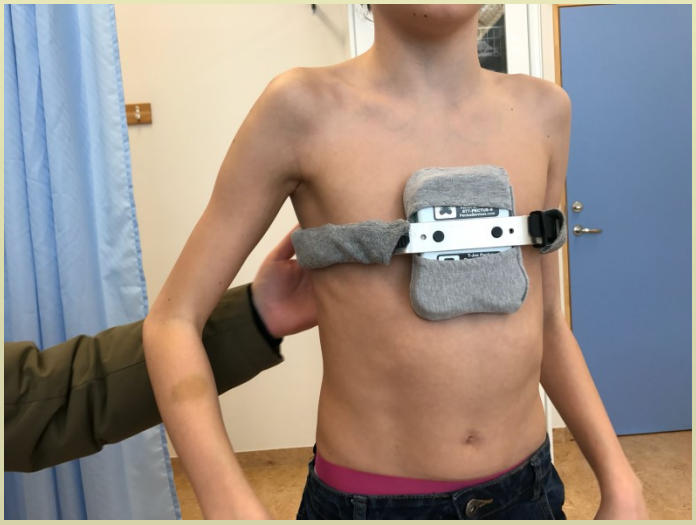



137 mm [proj]
SIEMENS FD-X
SIEMENS FD-X

Pectus Carinatum, bungubringa Case#2

- PC (margar gerðir)
- Einkenni verkir

Bungubringa Pectus Carinatum Beltismeðferð



PC Eftir beltismeðferð 24/7, í ca 9 mánuði



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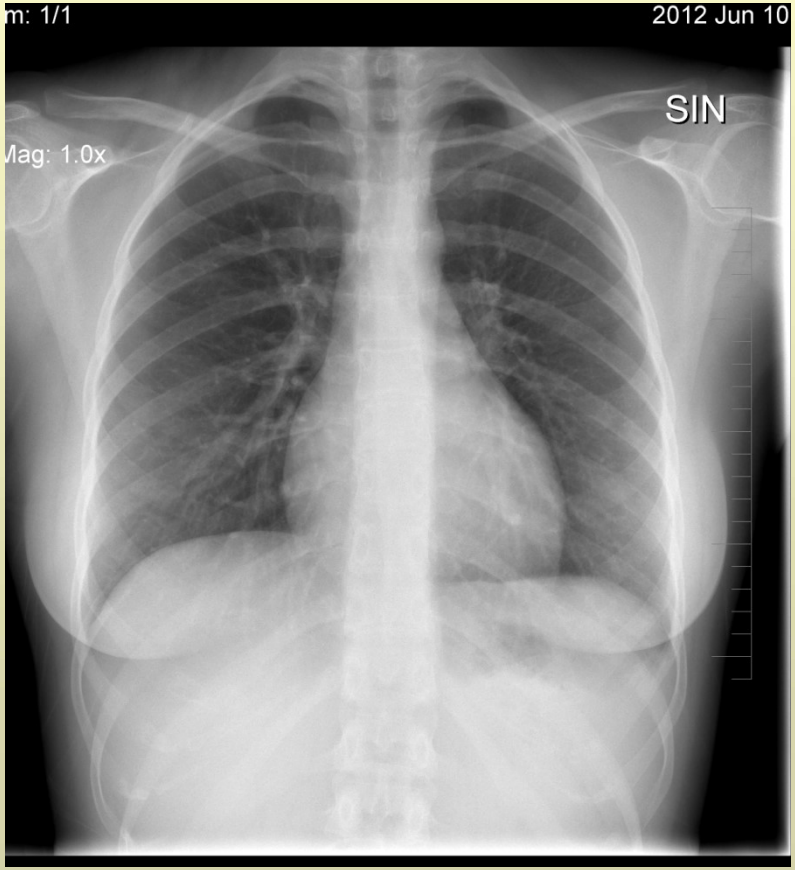
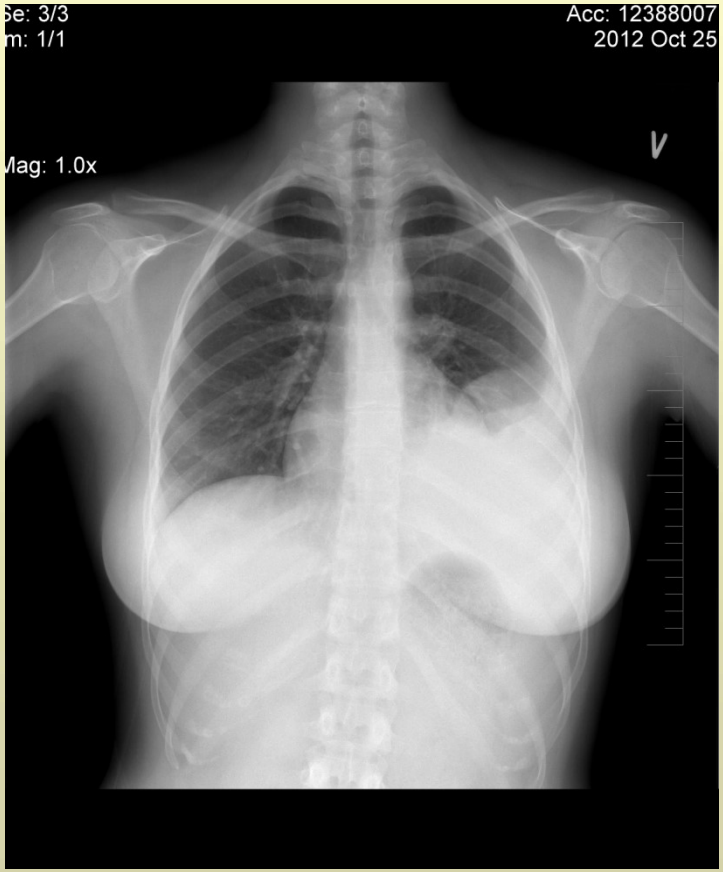
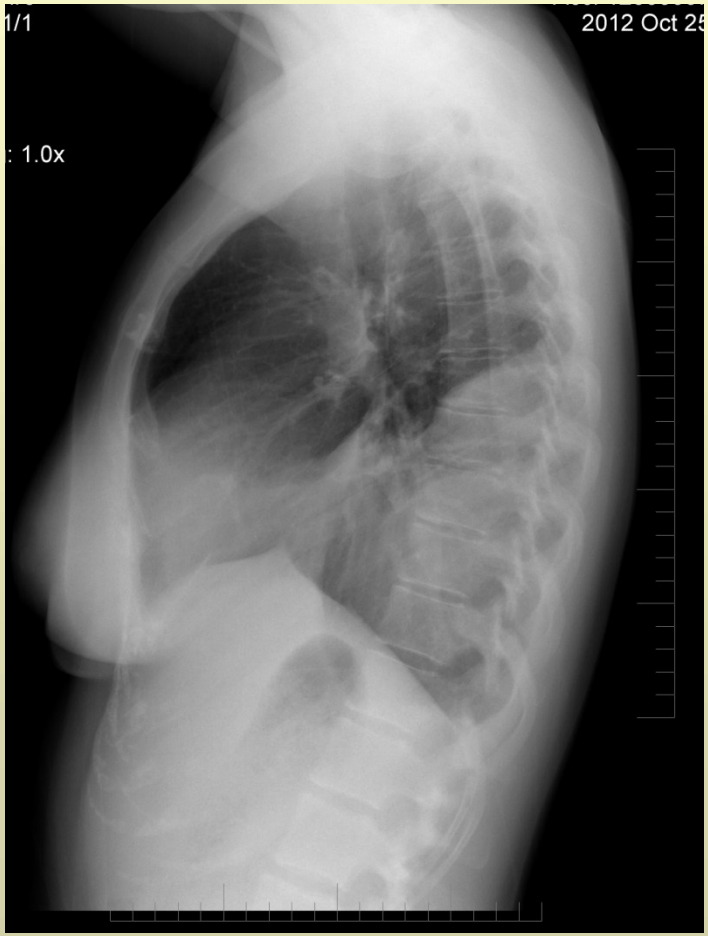
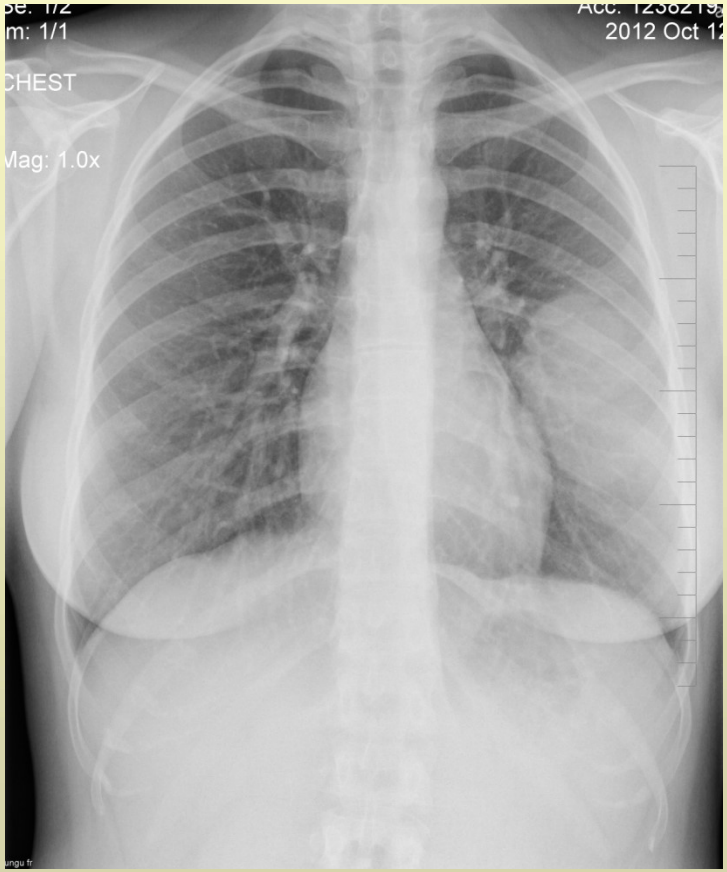
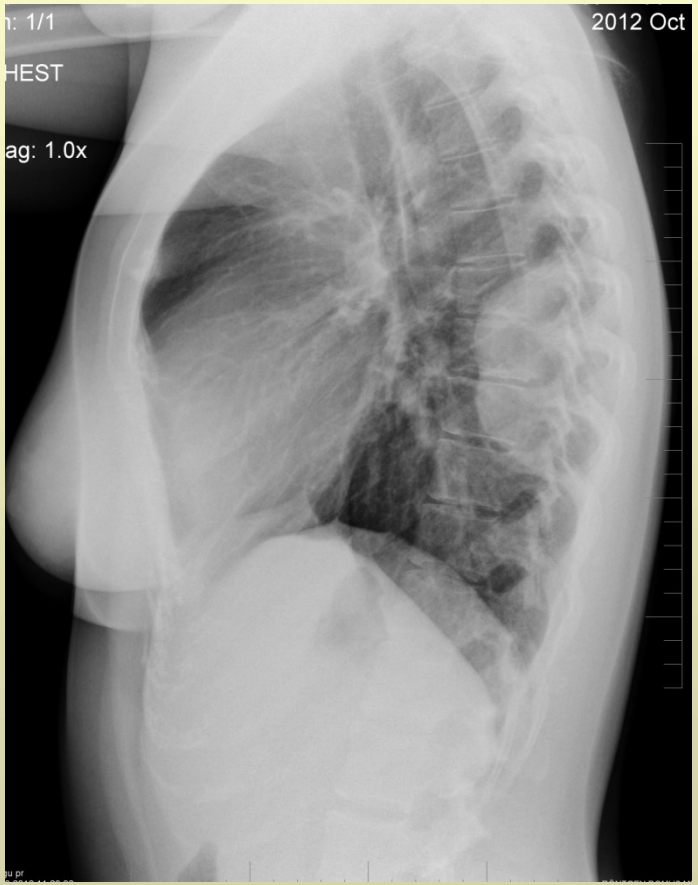
Æxli Case #3

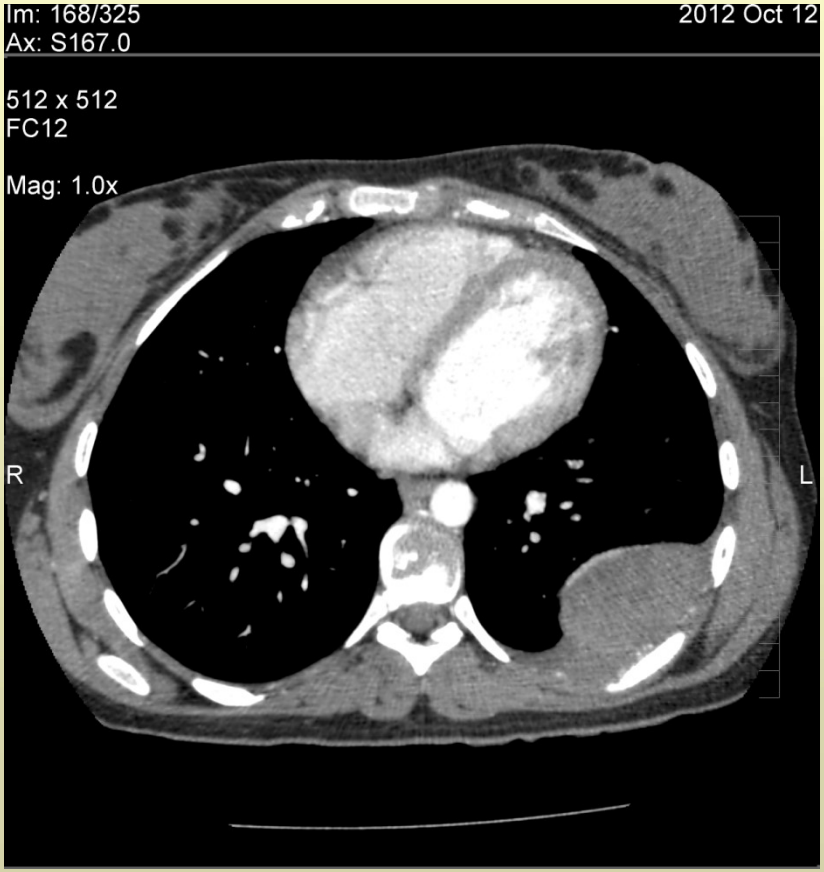



Se: 3/3
m: 1/1
Acc: 12388007
2012 Oct 25
Mag: 1.0x
V


1/1
2012 Oct 25
: 1.0x


m: 1/1
2012 Oct
CHEST
Mag: 1.0x


2012 Oct
HEST
ag: 1.0x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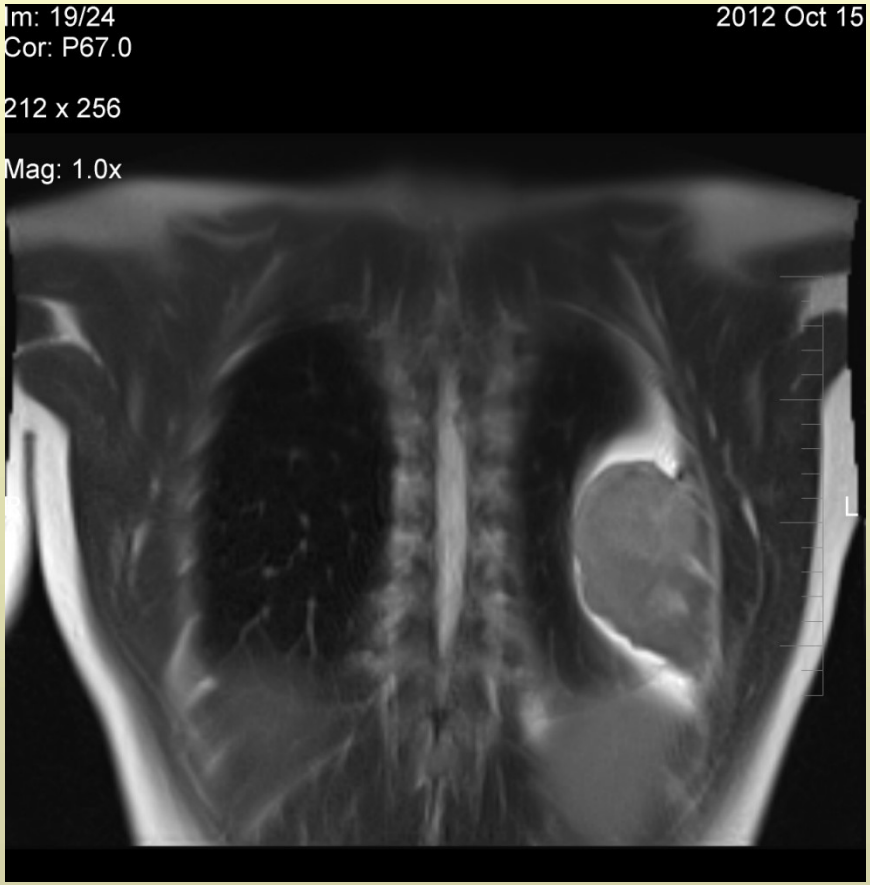
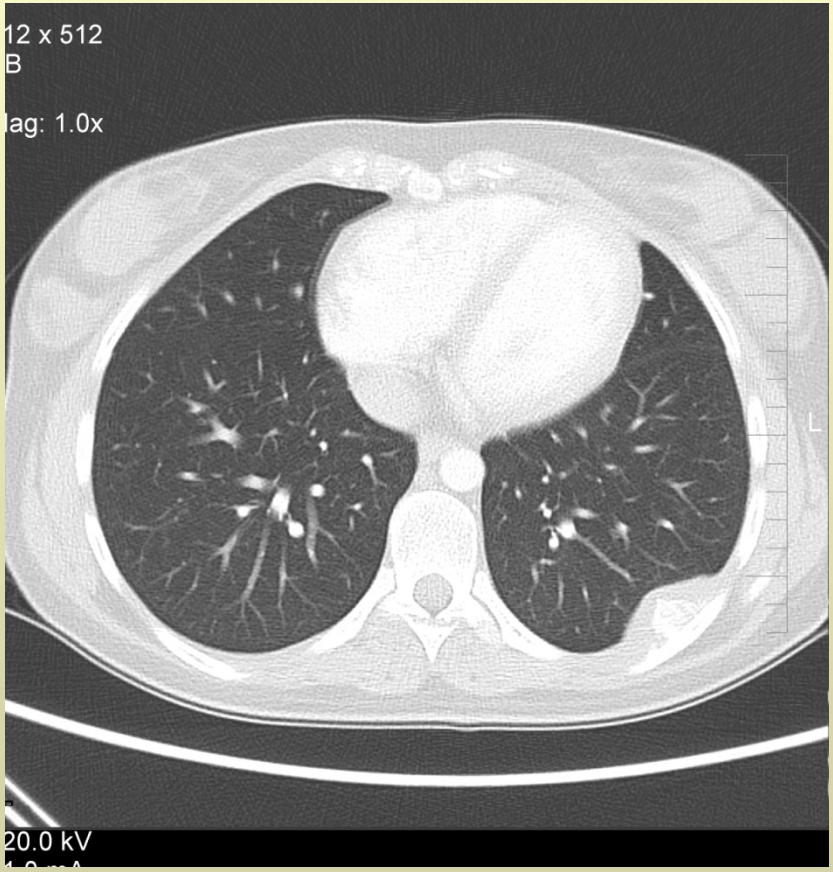
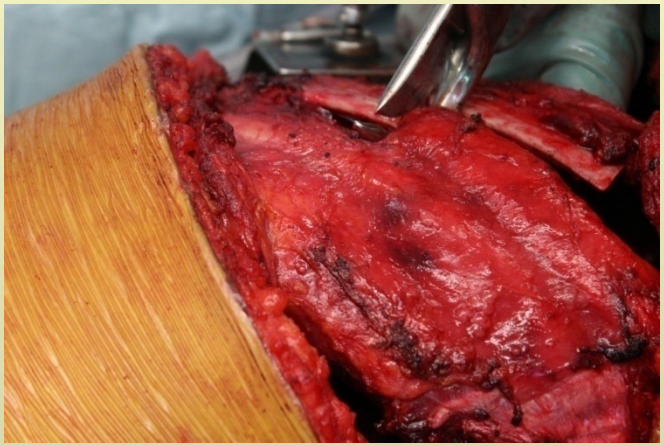
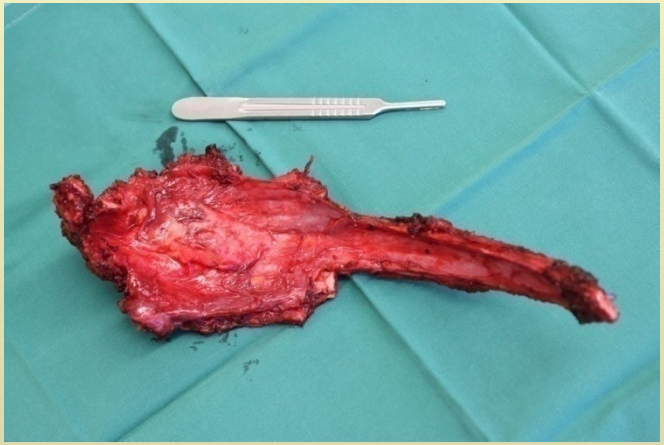
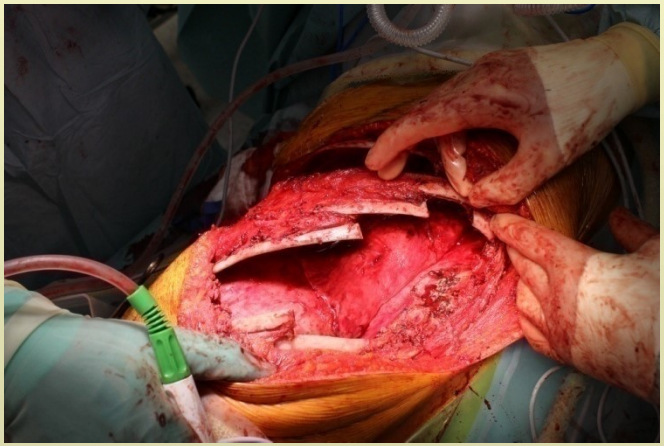
Ax: S167.0
2012 Oct 12
512 x 512
FC12
Mag: 1.0x
R
L

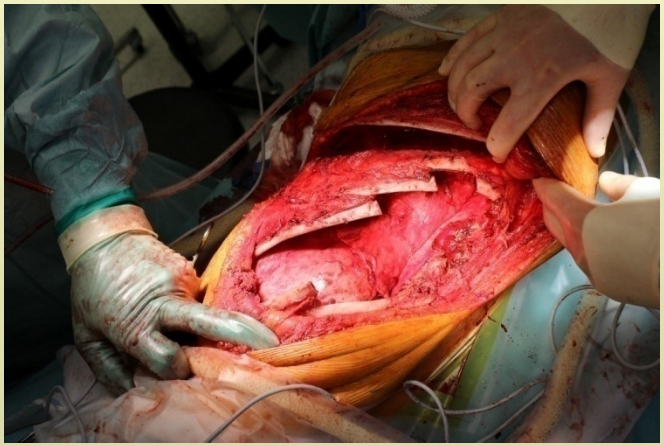

Im: 19/24
Cor: P67.0
2012 Oct 15
212 x 256
Mag: 1.0x
L

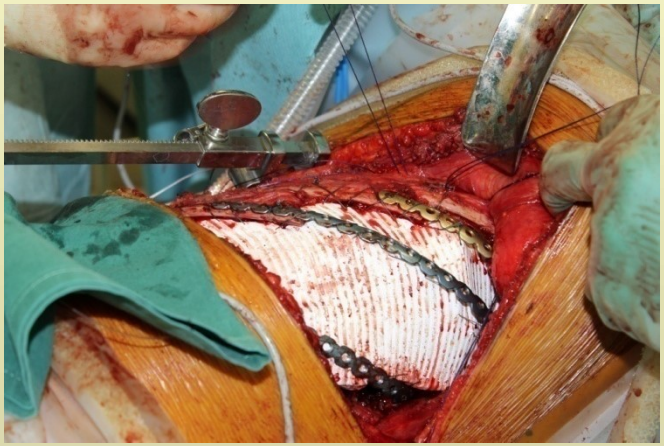

12 x 512
B
lag: 1.0x
20.0 kV

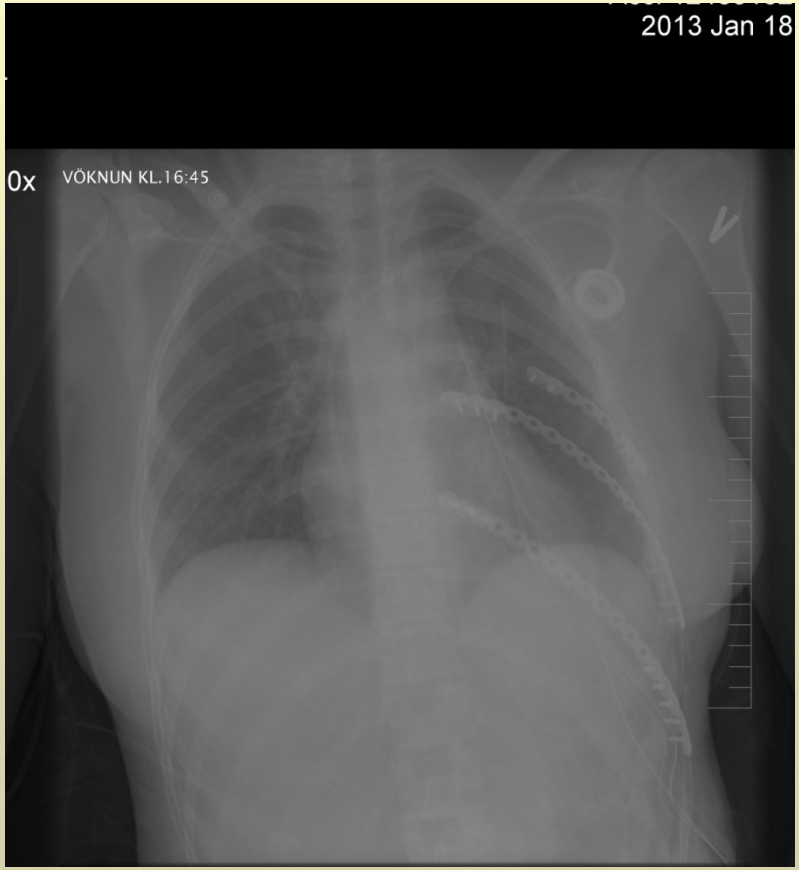


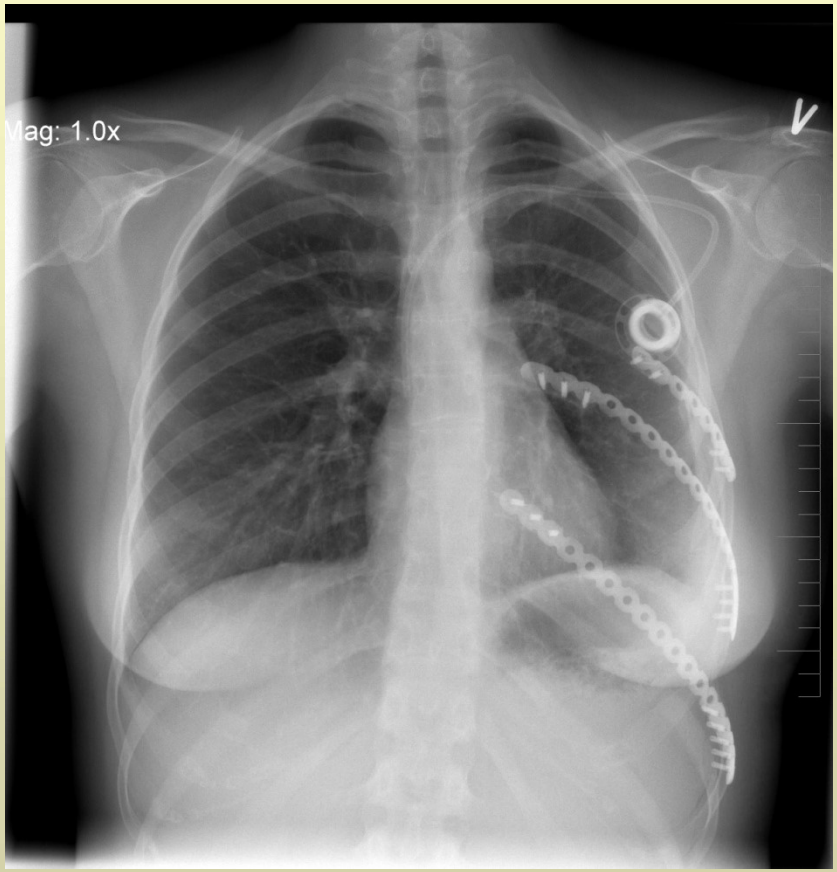


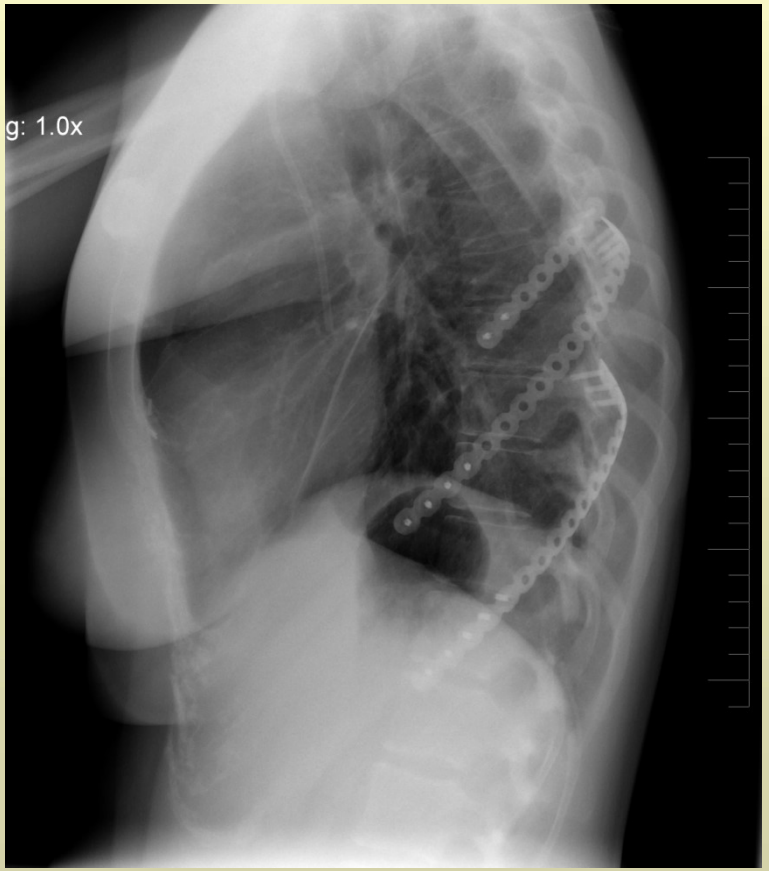







2013 Jan 18
0x
VÖKNUN KL.16:45


Mag: 1.0x


g: 1.0x

Gervirif notað við brottnám á Ewings sarkmeini í brjóstkassa

Bjarni Torfason [1,5] , Halldór Jónsson jr [2,5], Hildur Einarsdóttir [3,5], Helgi Sigurðsson [4,5]

1. Hjarta- og brjóstholsskurðlækningadeild, Landspítali,2. Bæklunarskurðlækningadeild, Landspítali,3. Myndgreiningadeild, Landspítali,4. Krabbameinslækningadeild, Landspítali,5. Læknadeild, Háskóli Íslands

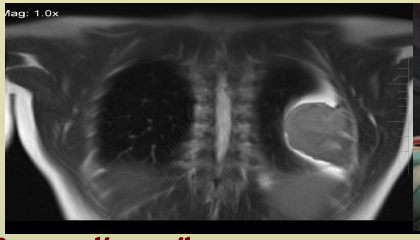
INNGANGUR

Ewings sarkmein(ES) er illkynja stoðkerfisæxli hjá börnum og og unglingum. Um 15% eru upprunnin í brjóstvegg/rifi og eru horfur þá betri. Einkenni eru brjóst-, takverkur og hósti; oft þreifast fyrirferð. Sérstök meðferðarteymi sarkmeina hafa þróast á öllum Norðurlöndum út frá SSG í Lundi 1979; á Íslandi IceSG 2007. Samþætt krabbameinslyfjameðferð og algert brottnám með skurðaðgerð hefur aukið lifun. Við brottnám á brjóstvegg hefur verið notað Gore® Dualmesh®.Tilkoma gervirifja fyrir flekabrjóst og vangróanda á rifbrotum hafa gefið von til að viðhalda betri starfsemi brjóstkassans í kjölfarið.

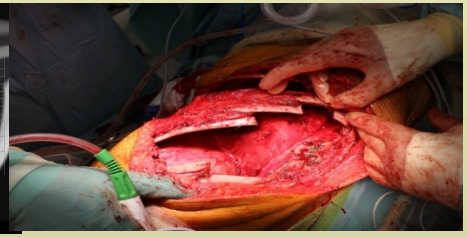
SJÚKRATILFELLI

20 ára stúlka leitaði á bráðamóttöku í júní 2012 vegna bakverkja sem leiddu undir vinstra brjóst og vinstri öxl án sögu um áverka. Röntgenmynd sýndi vökvasamsöfnun í fleiðruholi sem ekki var rannsökuð nánar. Vegna versnandi verkja var tekin ný röntgenmynd í október sem sýndi tilkomu fyrirferðar fyrir miðju vinstra lunga.

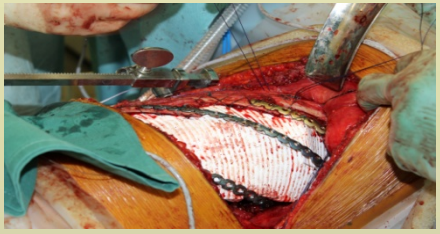
Í samráði við IceSG voru gerðar eftirfarandi rannsóknir og meðferðir: CT og segulómrannsóknir sýndu 8x5cm stórt æxli milli rifja 5 til 9; nálarsýni staðfesti ES og stigun sýndi enga útbreiðslu. Eftir fjóra lyfjakúra minnkaði æxlið um 85%. Sautján dögum síðar var gert brottnám á æxlisrestinni ásamt 3ja sentimetra umhverfi þess; 8.rif var fjarlægt algjörlega og hlutar af 6., 7. og 9. rifi. Enduruppbygging var gerð með Gore® Dualmesh® auk þriggja Synthes® gervirifja. Stúlkan útskrifaðist 10 dögum síðar.



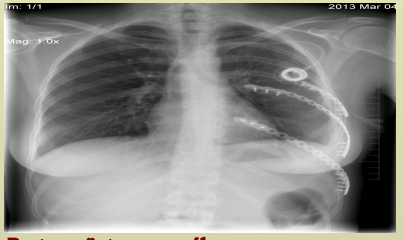
Pre-op segulómrannsókn - Ewing sarkmein í vi. brjóstvegg



Eftir brottnám æxlis og rifbeina



Eftir enduruppbyygingu með Gore Dualmesh og gervirifjum



Post-op röntgenrannsókn – Synthes gervirif í vi. brjóstvegg

ÁLYKTUN

Snemmgreining Ewing sarkmeins, samræmdar rannsóknir og meðferð eins og í þessu tilfelli er lærdómsrík fyrir lækna. Við enduruppbyggingu á stórum líkamshlutum eins og brjóstkassa er nauðsynlegt að geta viðhaldið bæði útliti og sem fullkomnustu starfsemi upp á framtíða lífsgæði. Einnig er mælt með notkun á slíkum gervirifjum við brjóstholsáverka til að stytta megi öndunarvélarmeðferð og gjörgæsluvist þegar um er að ræða umfangsmikið flekabrjóst. Síðkomin spenging þegar rifbrot gróa illa með myndun sársaukafullra gervilíða, kemur einnig sterklega til greina. Notkun okkar á gervirifjum í fyrsta sinn á Íslandi reyndist vel. Útlit og starfsemi brjóstveggsins hélst sem næst óbreytt eins og staðfestist í skjótum bata og stuttri sjúkrahúsvist. Nánari heimildir fást gegnum meðfylgjandi tölvupóst.

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Thorax Feb 2005 Vol 60 Suppl 1

www.brit-thoracic.org.uk

BTS guidelines

for the

Management of Pleural Infection in Children

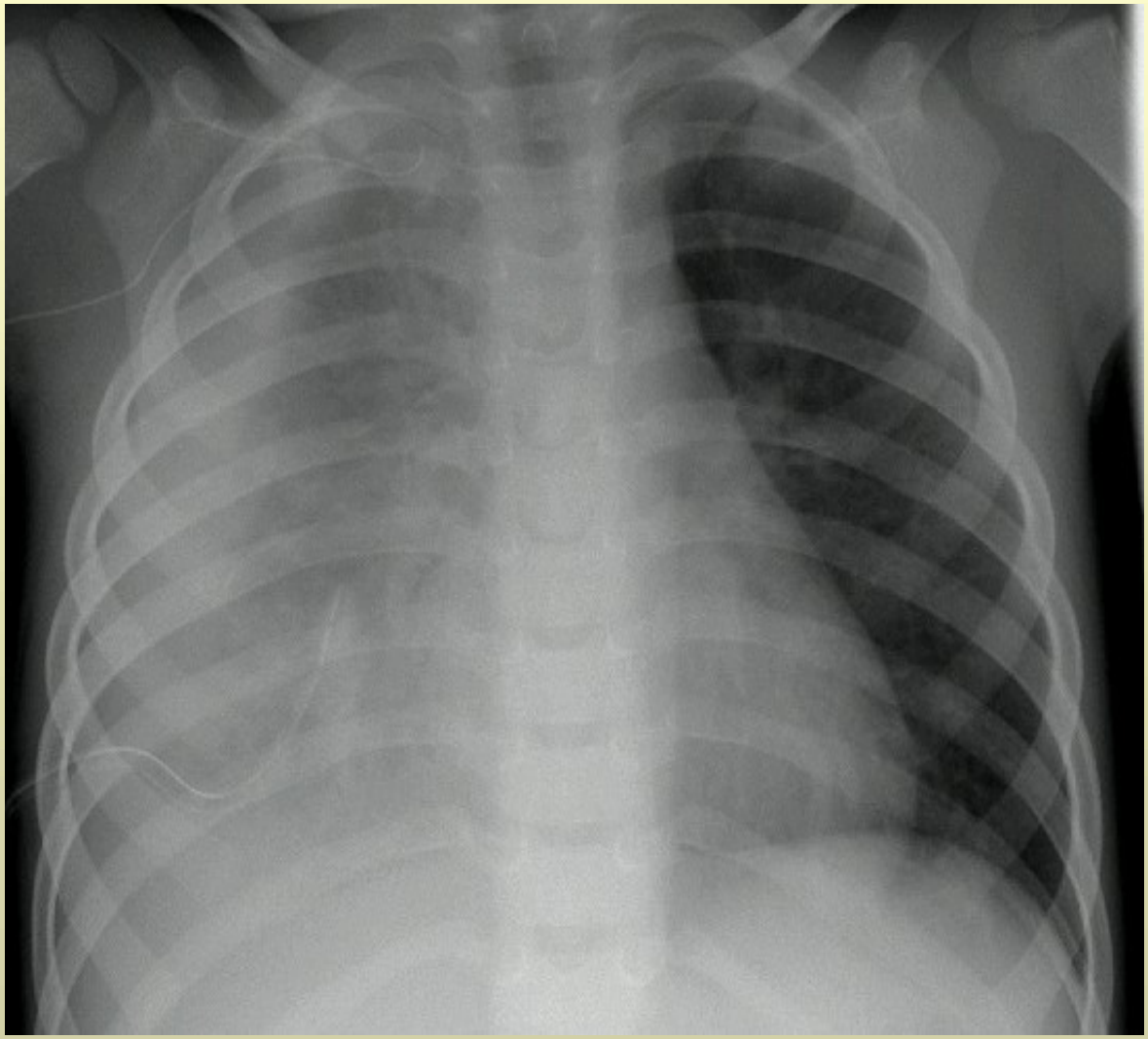
Dr Ian Balfour-Lynn

Royal Brompton Hospital

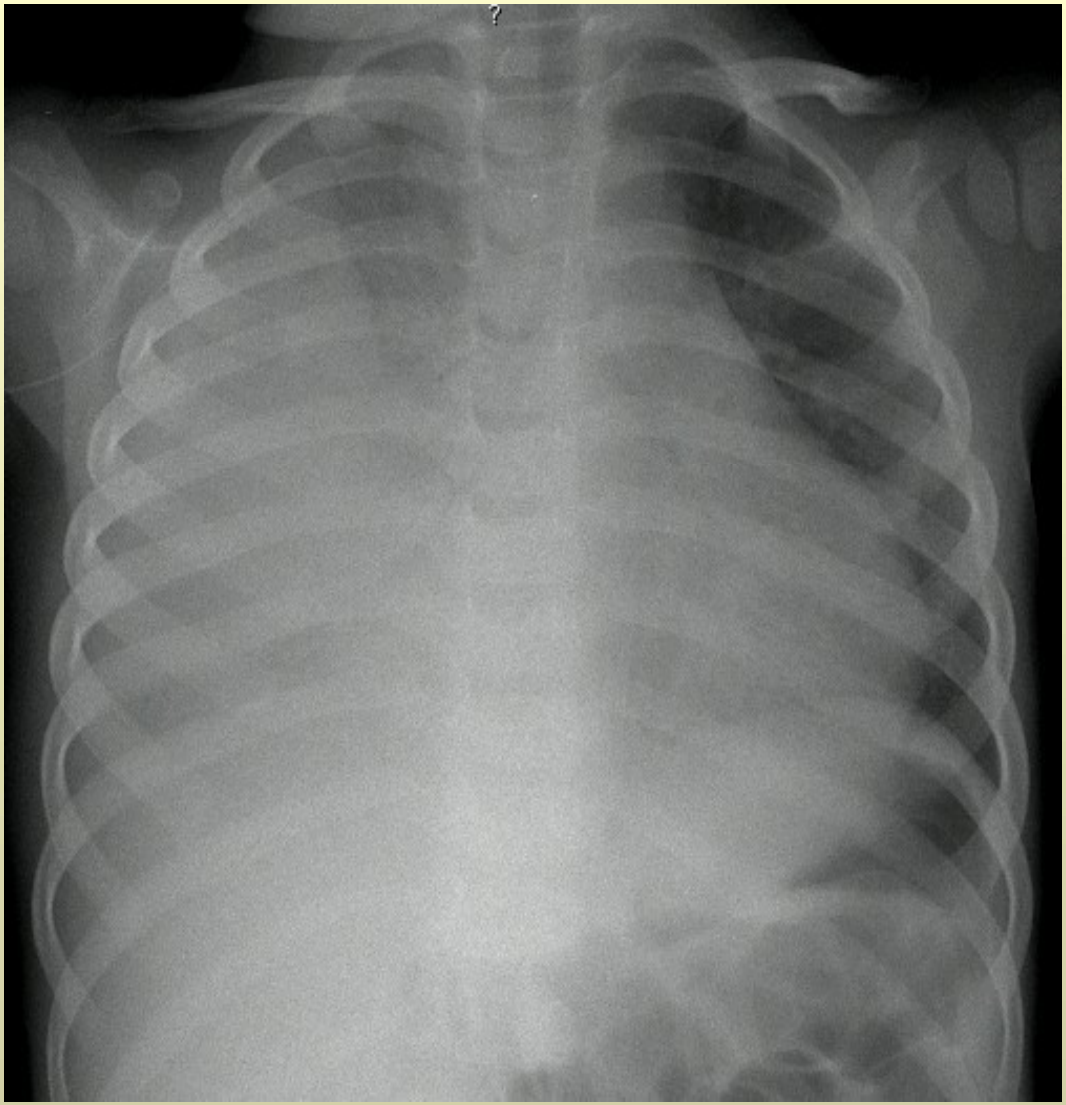
Linkur:

https://thorax.bmj.com/content/60/suppl_1/i1

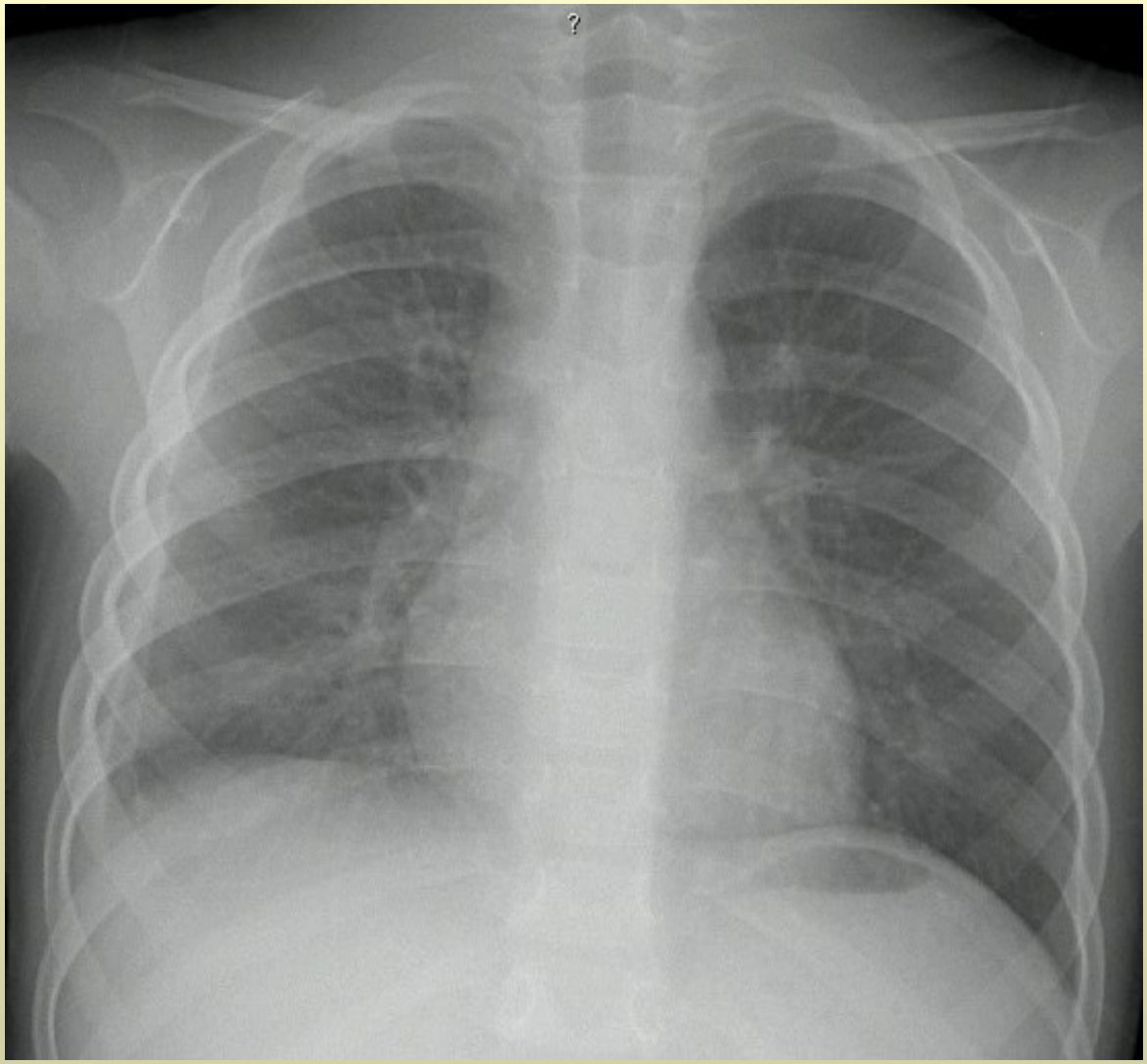
3 year old boy – 1w fever, malaise, cough, DIB
IVABs no improvement so transferred
Drain inserted, urokinase, IV cefuroxime

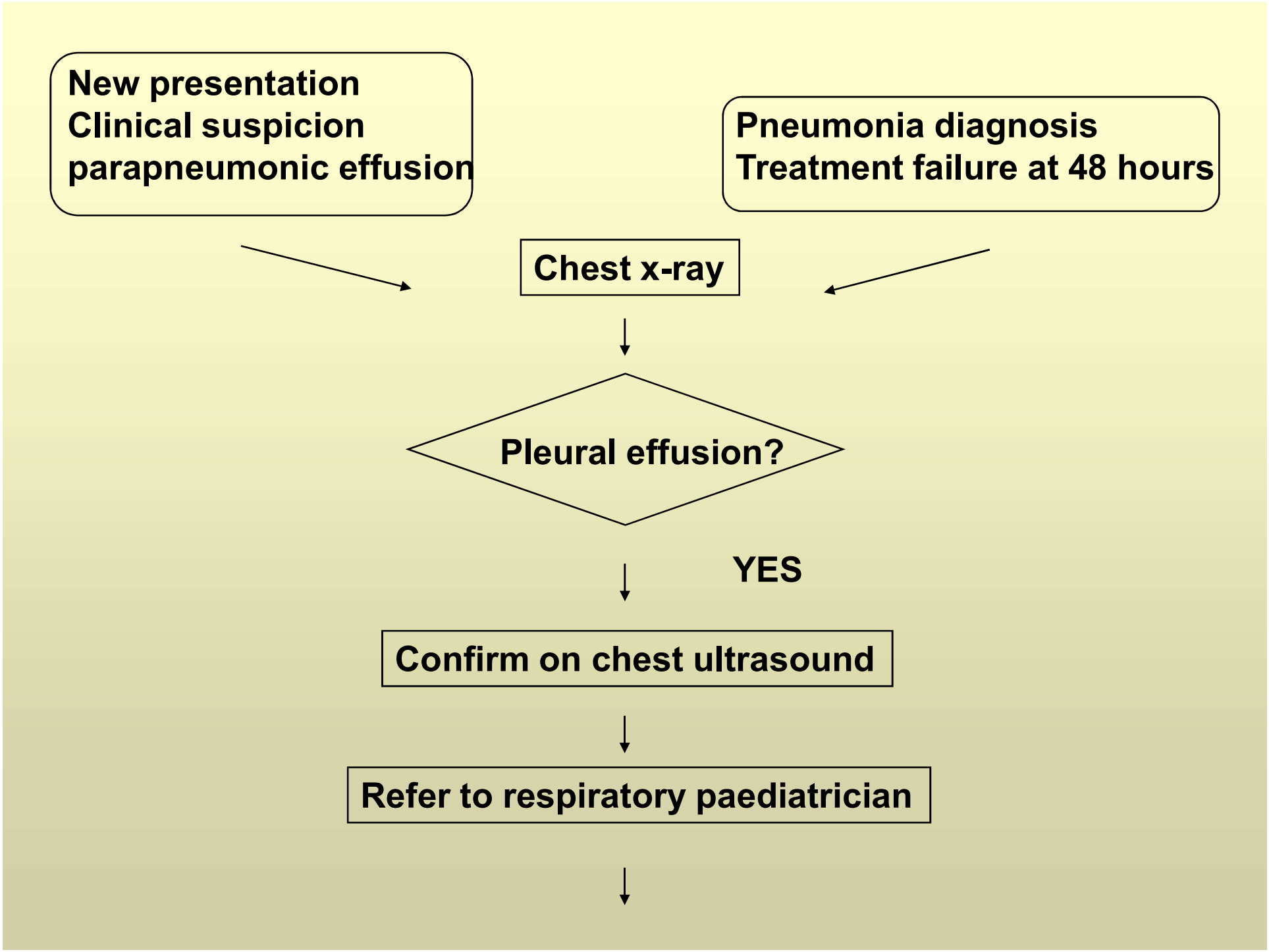


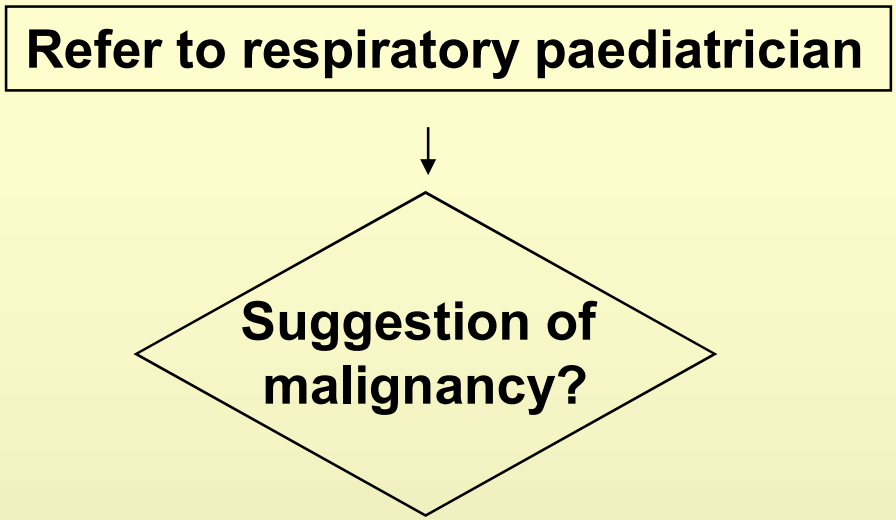
B/C – Pneumococcus. Pleural fluid - sterile
4 days later – well but febrile, drain out & home next day

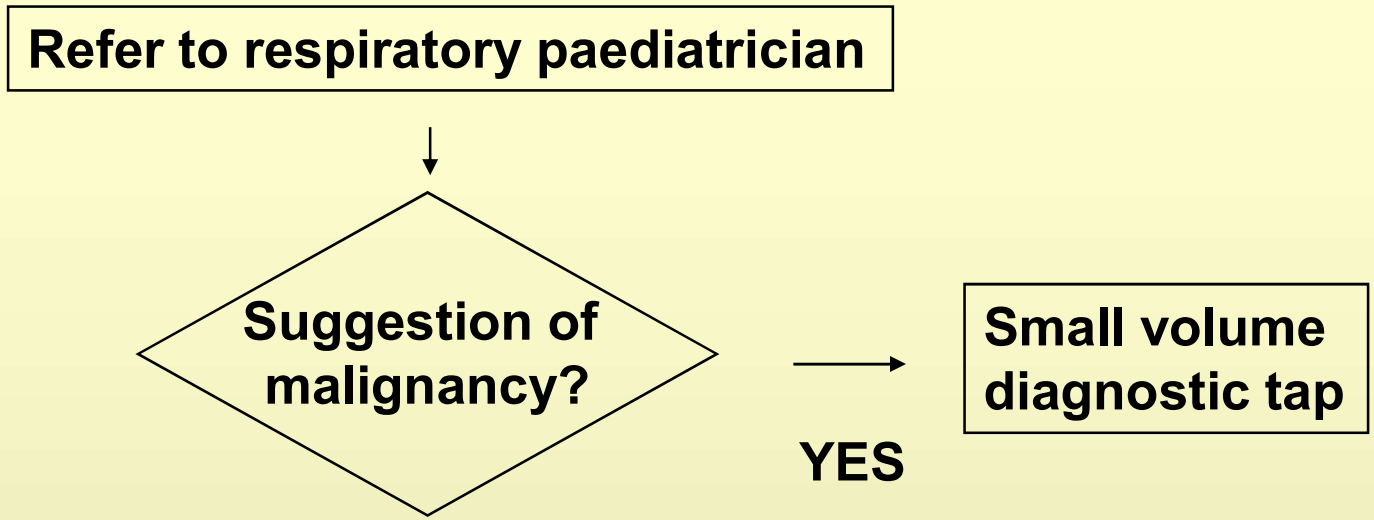


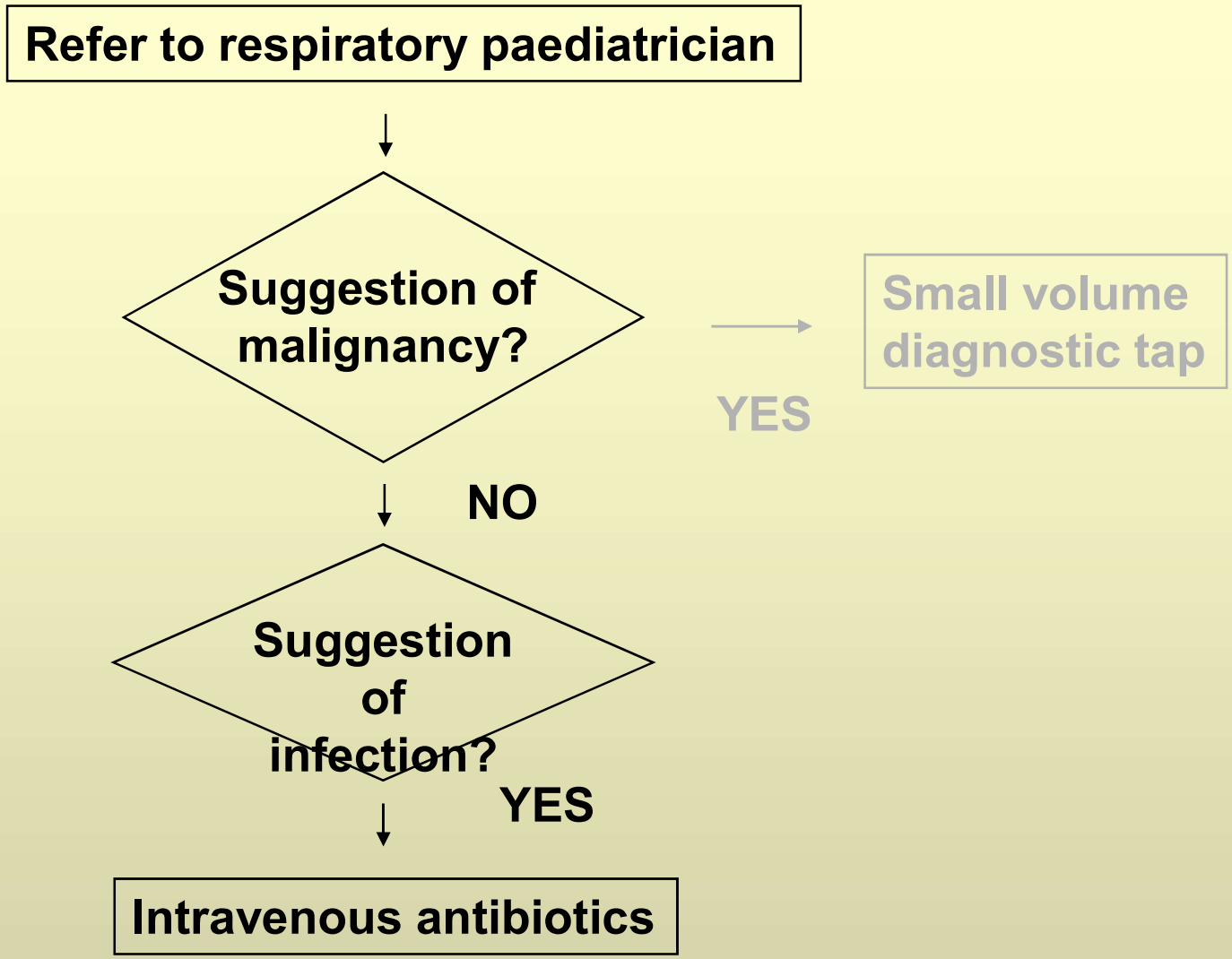
Back to normal by 2 weeks
7 week follow up -

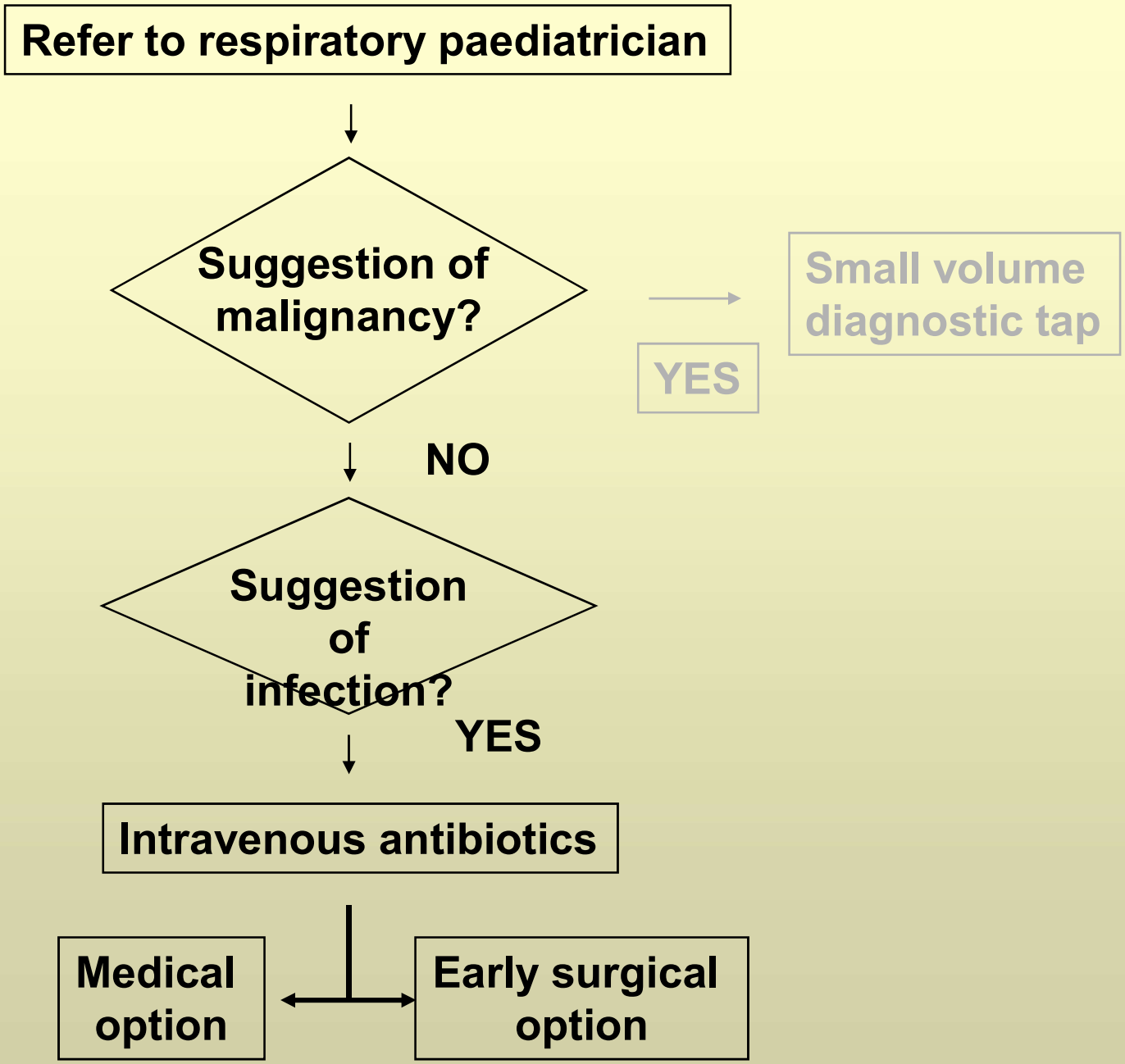


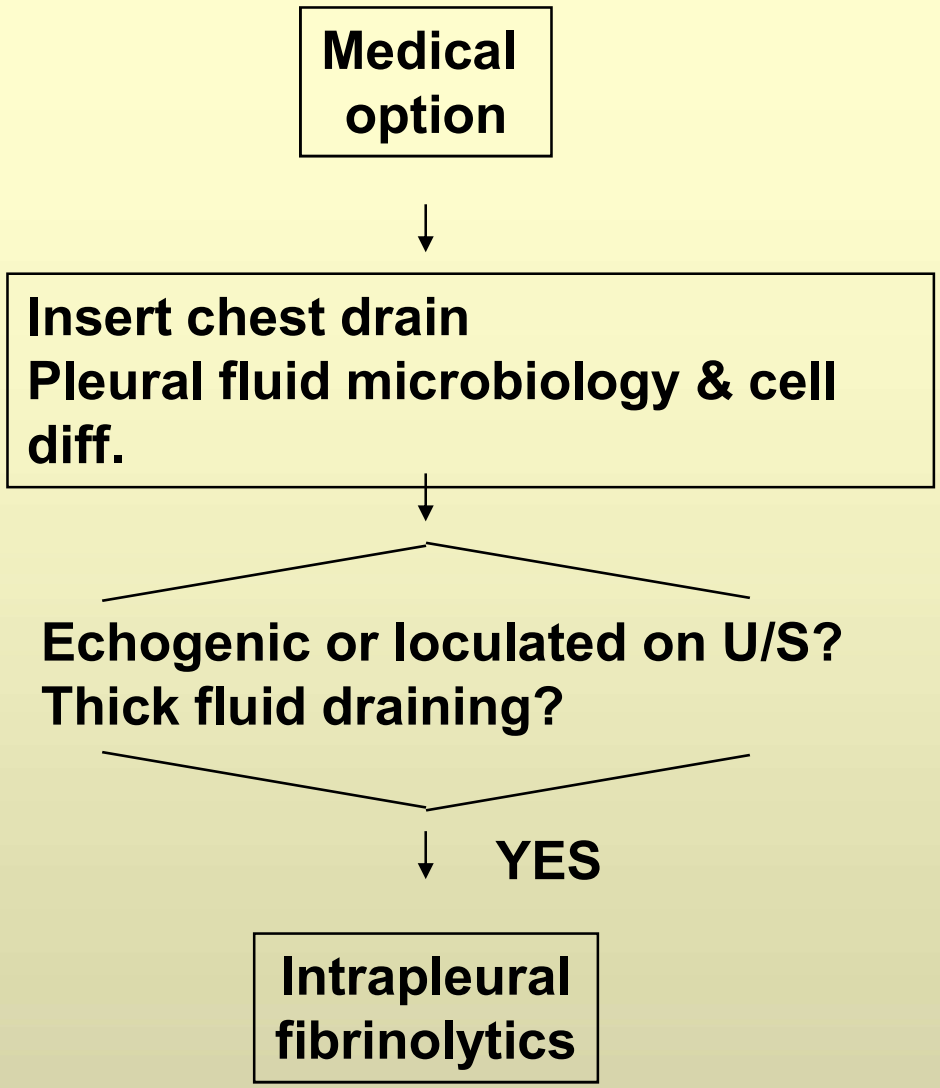

New presentation
Clinical suspicion
parapneumonic effusion
Pneumonia diagnosis
Treatment failure at 48 hours
Chest x-ray
Pleural effusion?
YES
Confirm on chest ultrasound
Refer to respiratory paediatrician

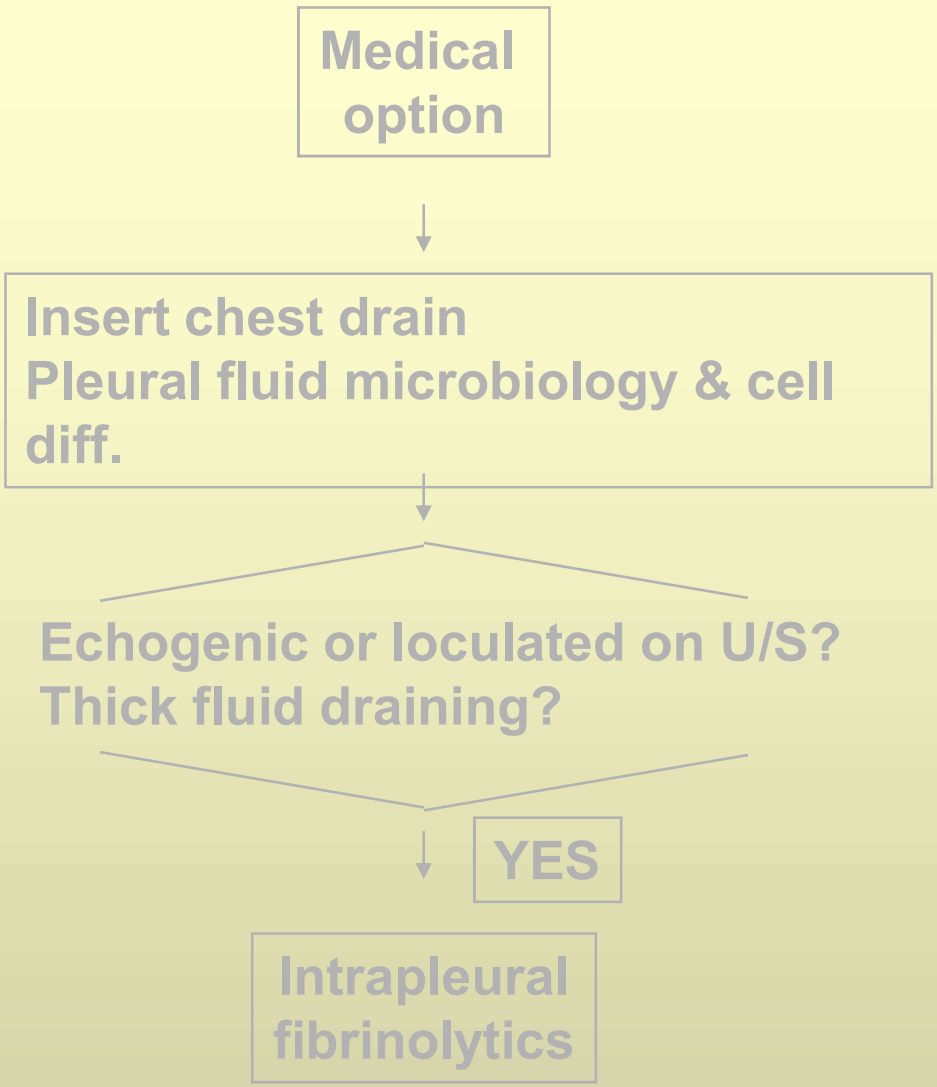

Refer to respiratory paediatrician
Suggestion of malignancy?

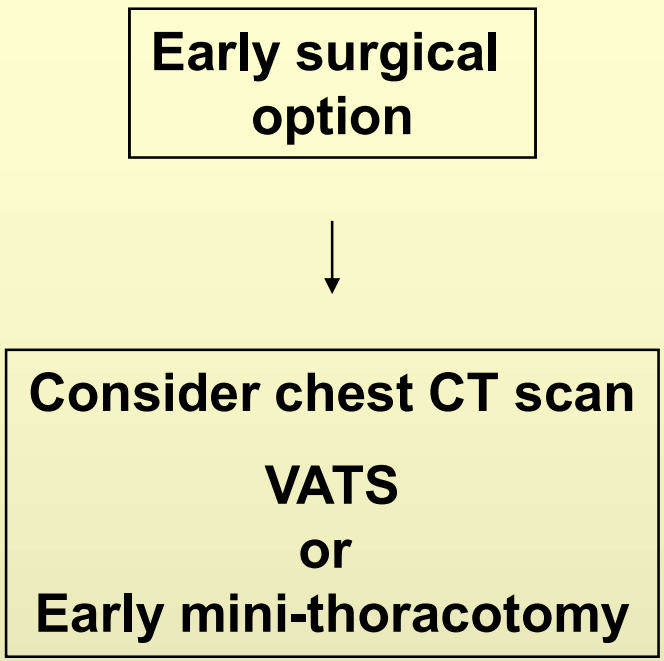

Refer to respiratory paediatrician
Suggestion of malignancy?
YES
Small volume diagnostic tap

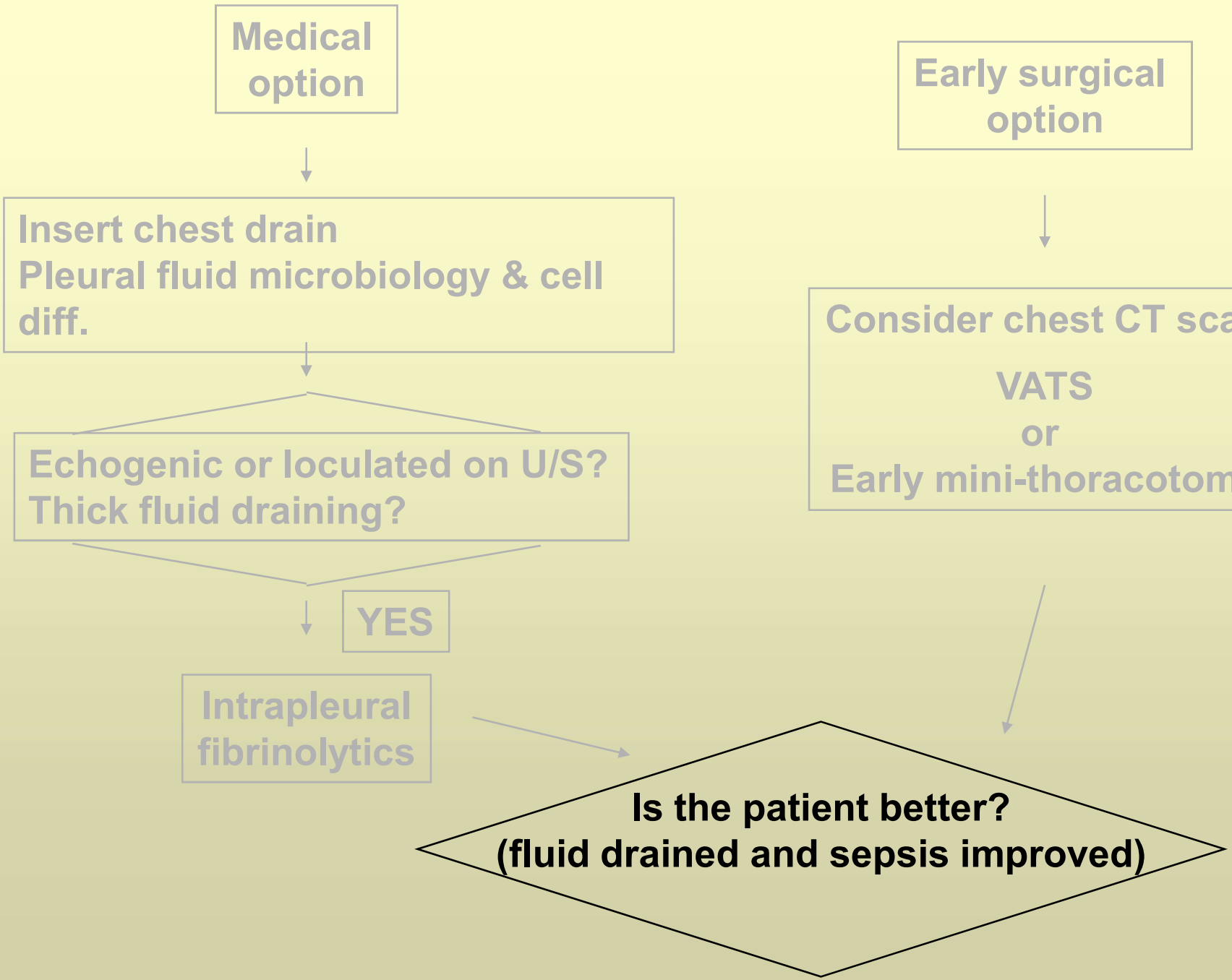

Refer to respiratory paediatrician
Suggestion of malignancy?
YES
Small volume diagnostic tap
NO
Suggestion of infection?
YES
Intravenous antibiotics

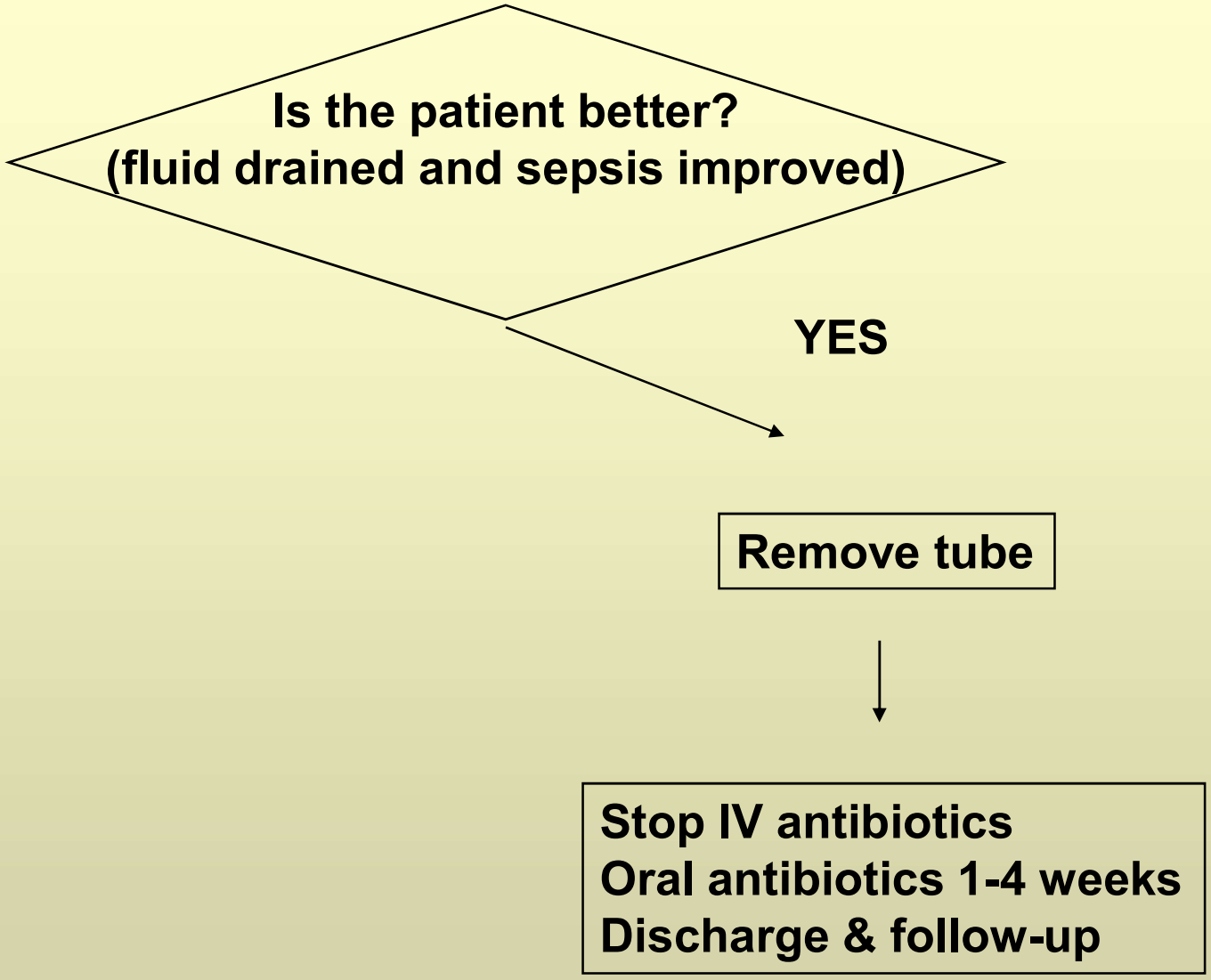

Refer to respiratory paediatrician
Suggestion of malignancy?
YES
Small volume diagnostic tap
NO
Suggestion of infection?
YES
Intravenous antibiotics
Medical option
Early surgical option

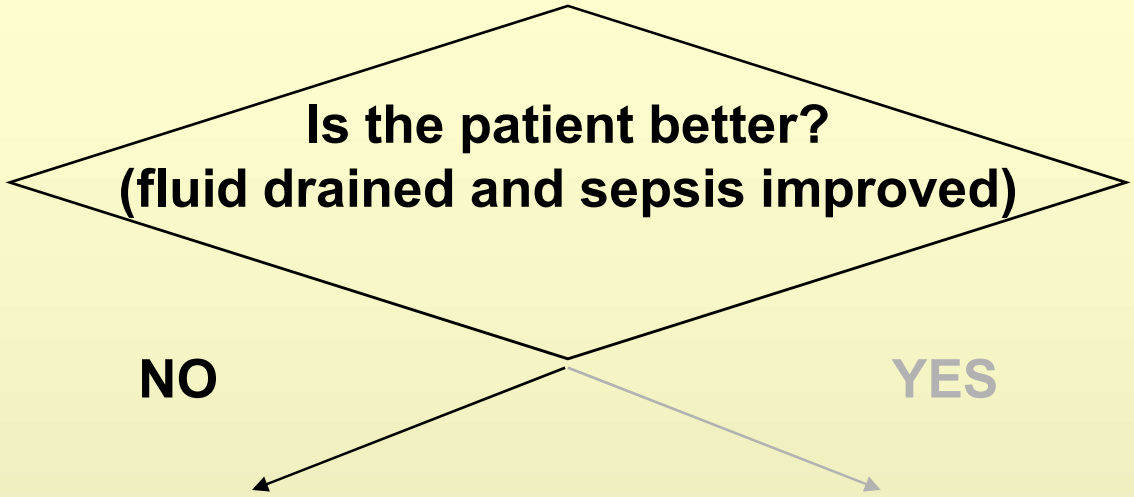

Medical option
Insert chest drain
Pleural fluid microbiology & cell diff.
Echogenic or loculated on U/S?
Thick fluid draining?
YES
Intrapleural fibrinolytics


Medical option
Insert chest drain
Pleural fluid microbiology & cell diff.
Echogenic or loculated on U/S?
Thick fluid draining?
YES
Intrapleural fibrinolytics


Early surgical option
Consider chest CT scan
VATS
or
Early mini-thoracotomy


Medical option
Early surgical option
Insert chest drain
Pleural fluid microbiology & cell diff.
Consider chest CT scan
VATS
or
Early mini-thoracotomy
Echogenic or loculated on U/S?
Thick fluid draining?
YES
Intrapleural fibrinolytics
Is the patient better?
(fluid drained and sepsis improved)


Is the patient better?
(fluid drained and sepsis improved)
YES
Remove tube
Stop IV antibiotics
Oral antibiotics 1-4 weeks
Discharge & follow-up


Is the patient better?
(fluid drained and sepsis improved)
NO
YES
Consult with paediatric thoracic surgeon re. late surgery Consider chest CT scan
Remove tube
Stop IV antibiotics
Oral antibiotics 1-4 weeks
Discharge & follow-up

SIGN levels of evidence

- I – meta-analyses, RCTs (incl. systematic reviews) I^{++}, I^{+}, I^{-}
- II – case-control or cohort studies (incl. systematic reviews) II^{++}, II^{+}, II^{-}
- III – case reports, case studies
- IV – expert opinion

SIGN grades of recommendations

- A – evidence from meta-analysis, systematic review, RCT (I++ or applicable I+)
- B – evidence from applicable II^{++} or extrapolated I^{++}, I^{+}
- C – evidence from applicable II^{+} or extrapolated II^{++}
- D – evidence from III or IV

Clinical picture

- All children with parapneumonic effusion or empyema should be admitted to hospital. [D]
- If a child remains pyrexial or unwell 48 hours after admission for pneumonia, parapneumonic effusion / empyema must be excluded. [D]

Intrapleural fibrinolytics

- Intrapleural fibrinolytics shorten hospital stay and are recommended for any complicated parapneumonic effusion (thick fluid with loculations) or empyema (overt pus). [B]
- Urokinase should be given twice daily for 3 days (6 doses in total) using 40,000 units in 40 mls 0.9% saline for children aged 1 year or above, and 10,000 units in 10 mls 0.9% saline for children aged under 1 year. [B]

Thomson et al Thorax 2002;57 343-7

Surgery

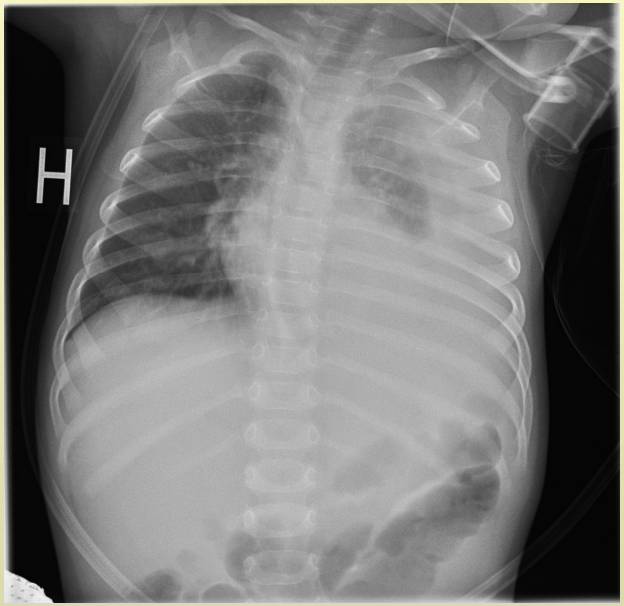
- Failure of chest tube drainage, antibiotics and fibrinolytics should prompt early discussion with a thoracic surgeon. [D]
- Patients should be considered for surgical treatment if they have persisting sepsis in association with a persistent pleural collection, despite chest tube drainage and antibiotics. [D]
- Organised empyema in a symptomatic child requires formal thoracotomy and decortication. [D]

Follow-up

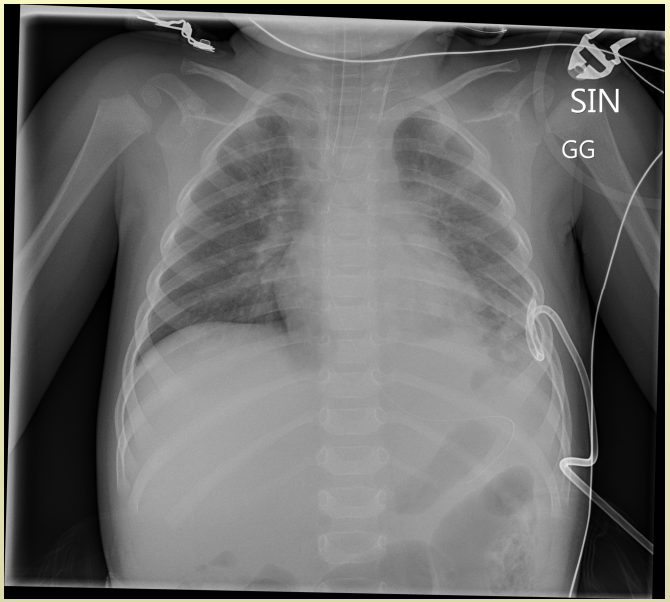
- Children should be followed up after discharge until they have recovered completely and their chest radiograph has returned to near normal. [D]
- Underlying diagnoses – for example, immunodeficiency, cystic fibrosis – may need to be considered. [D]

Case #4

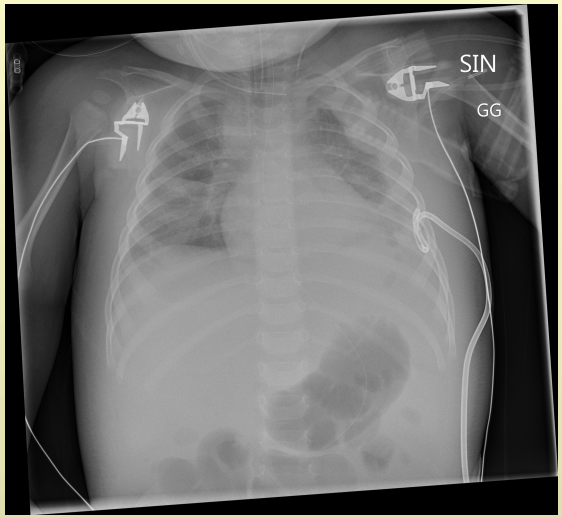
Fyrir dren

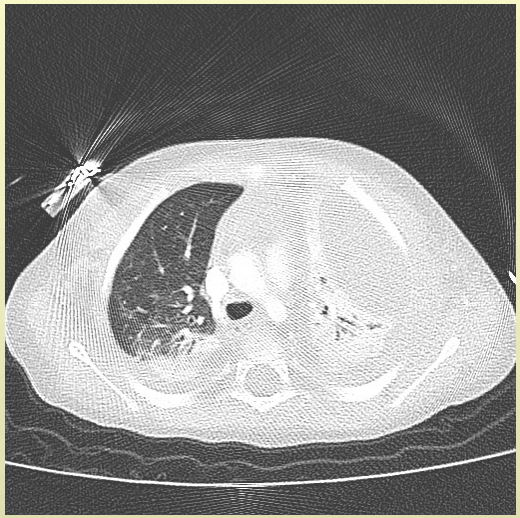


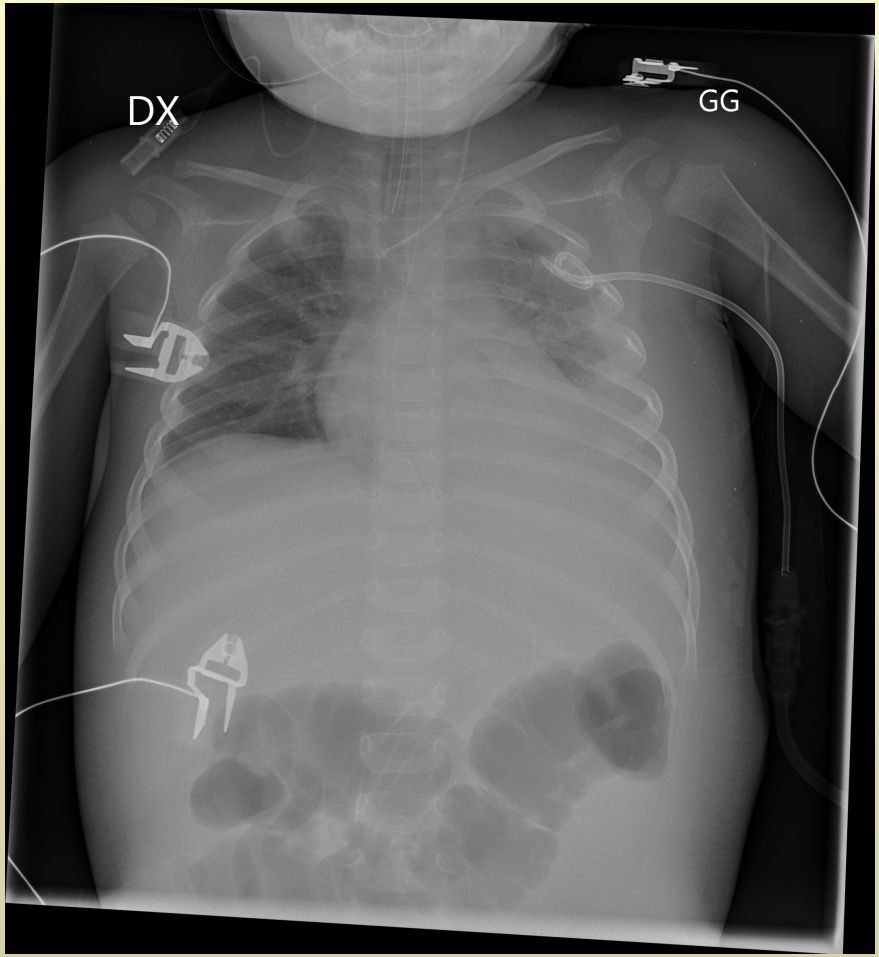
Eftir dren



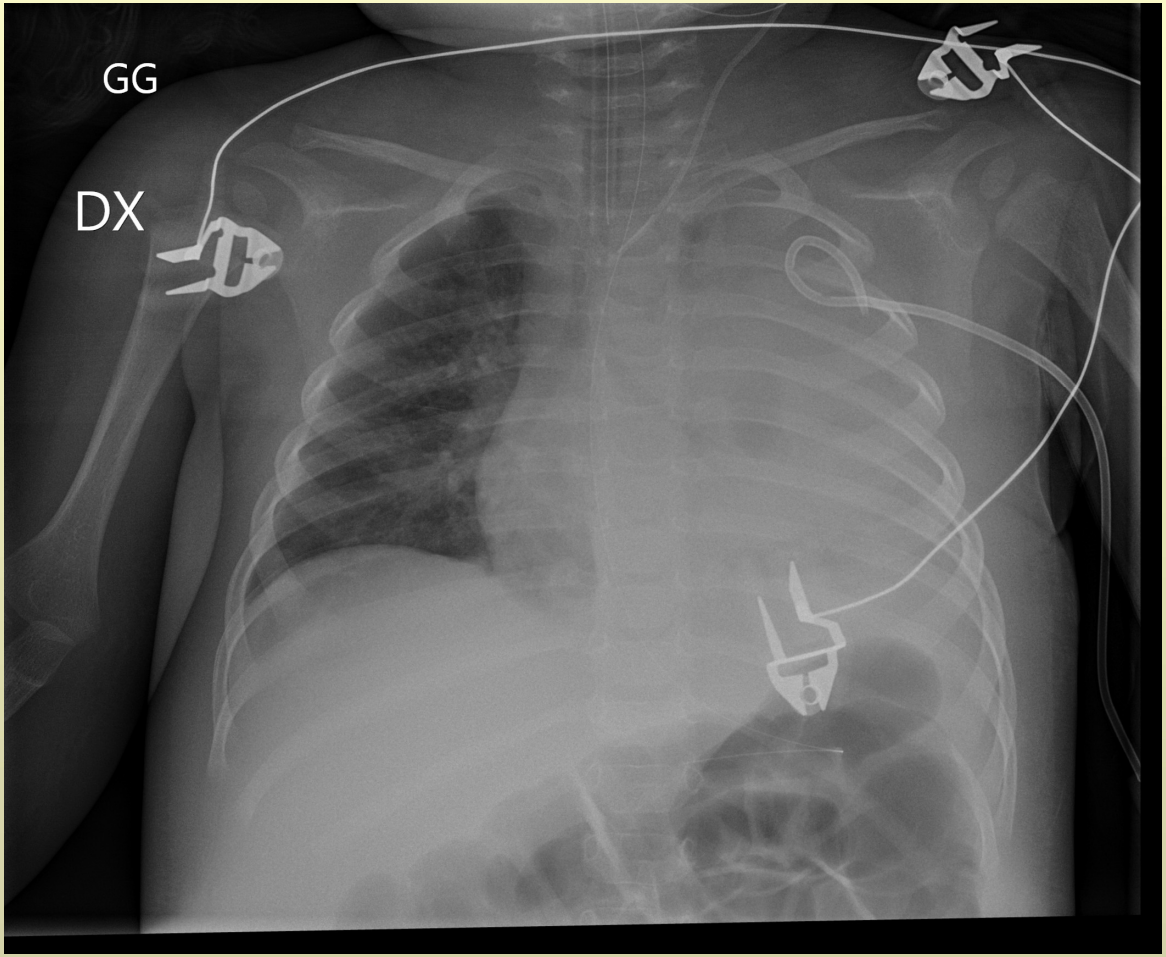
4 dögum síðar:

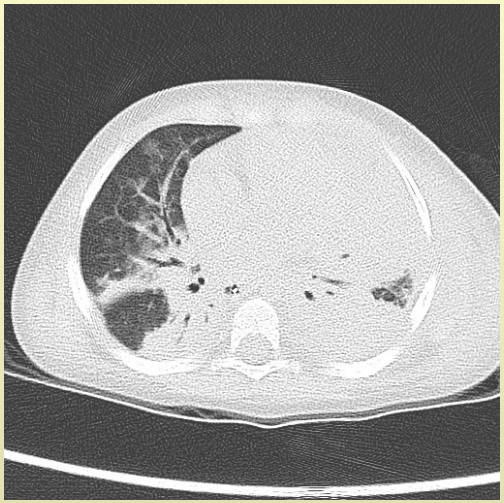




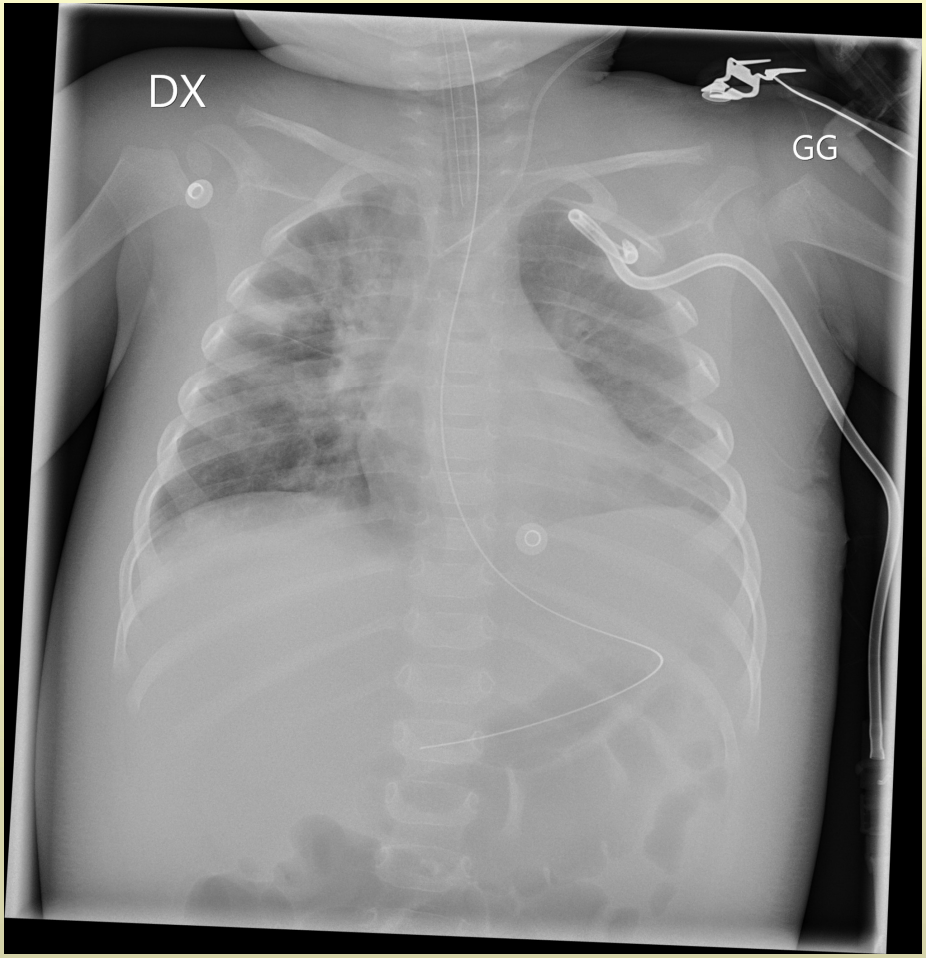

DX
GG

- Gg
- Decortication?
- Fibrinolysis?
- ECMO?


GG
DX



Næsta dag



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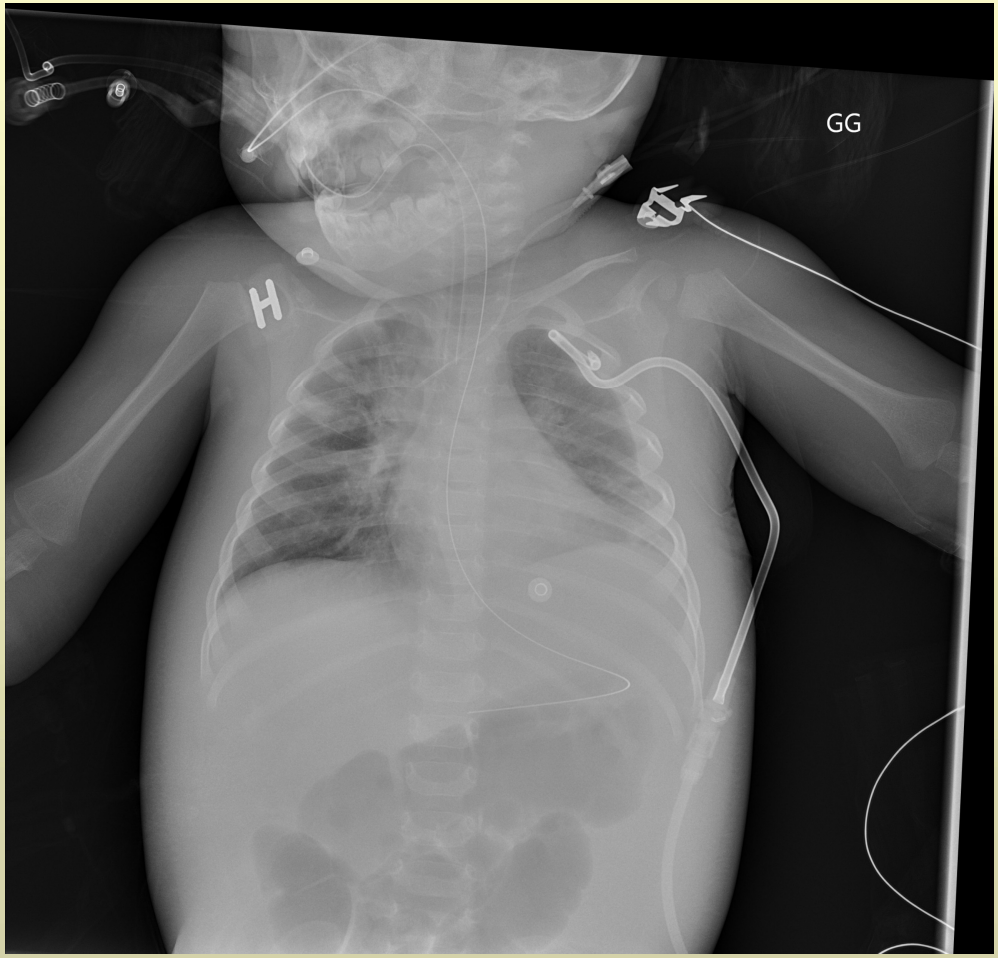

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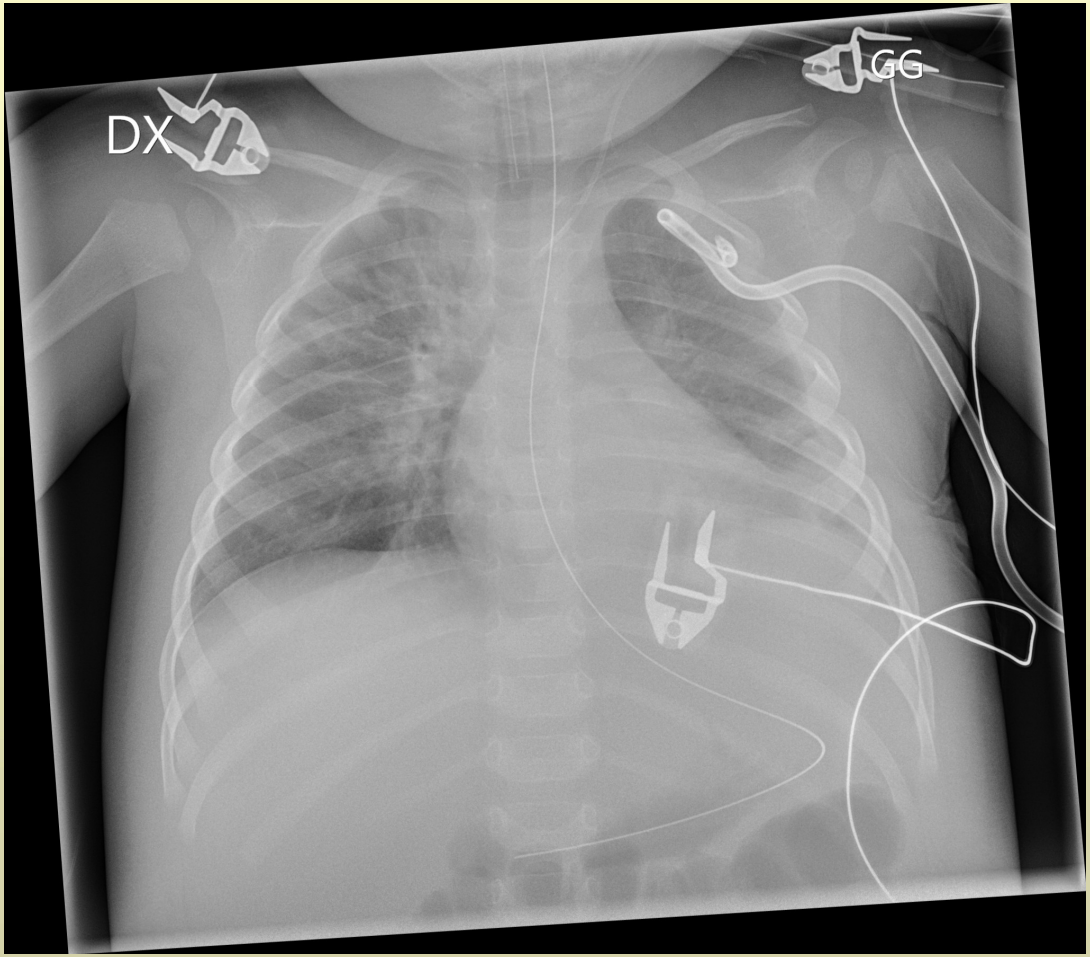

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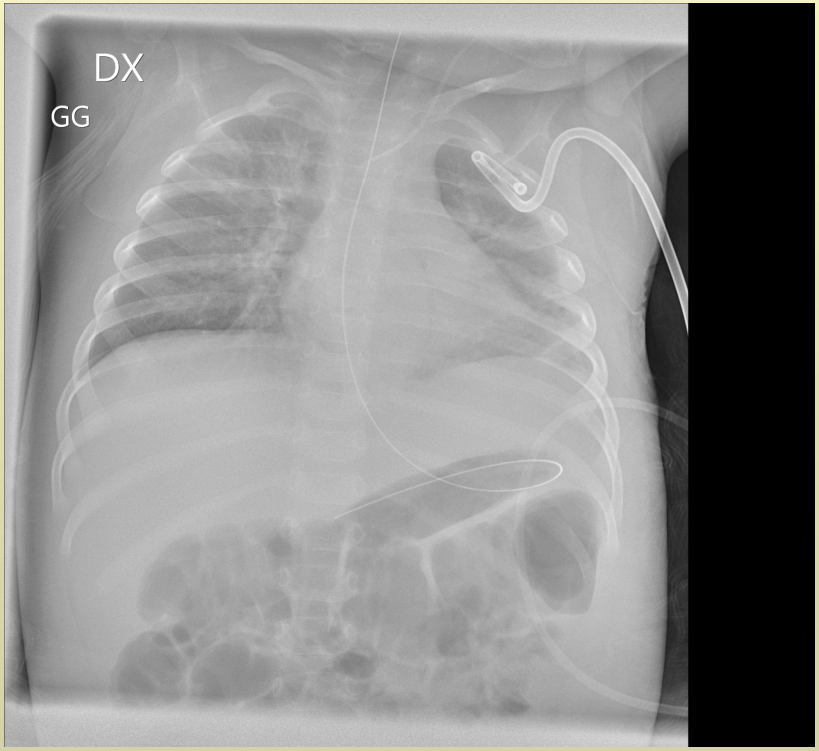
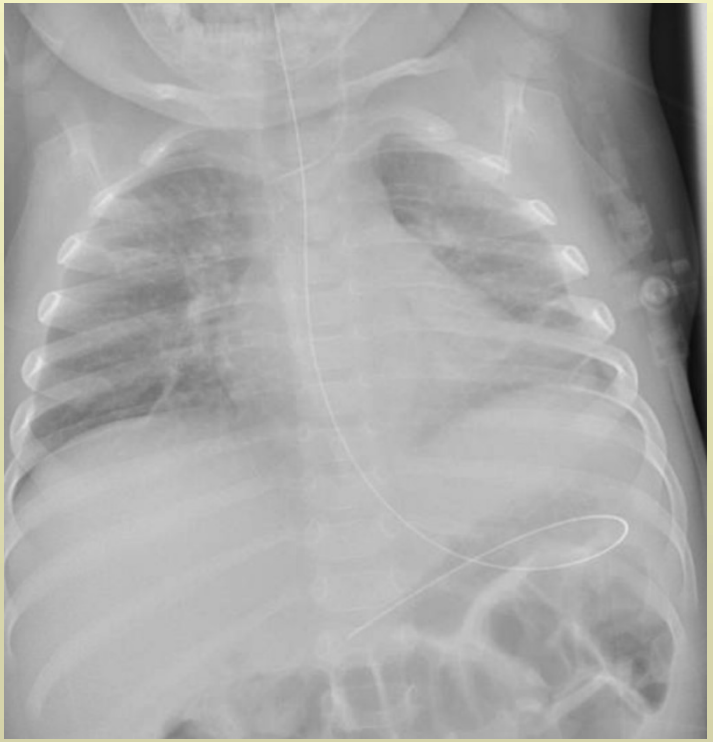
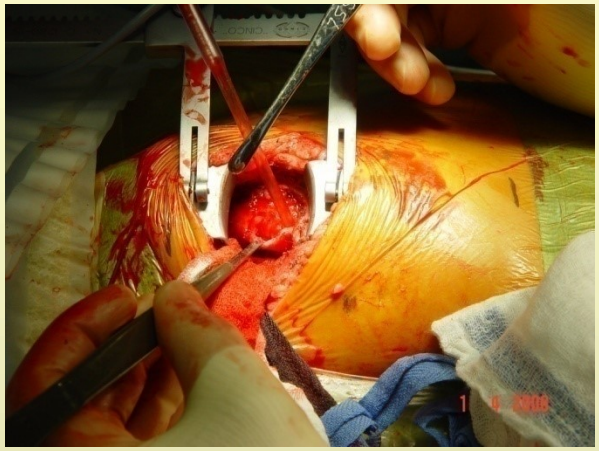
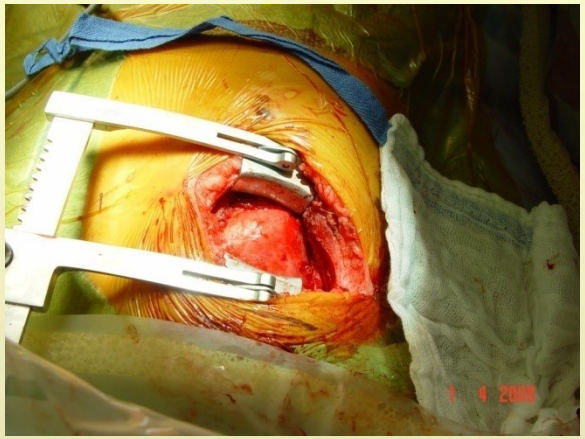
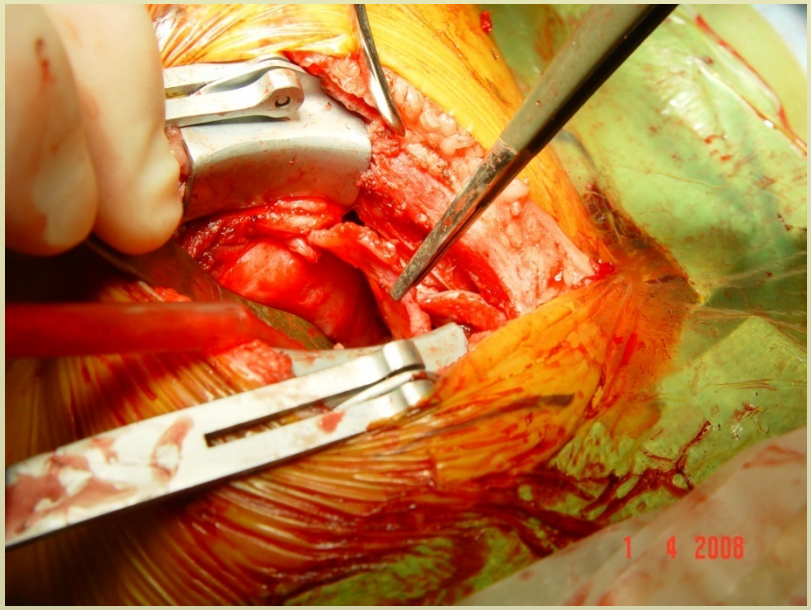


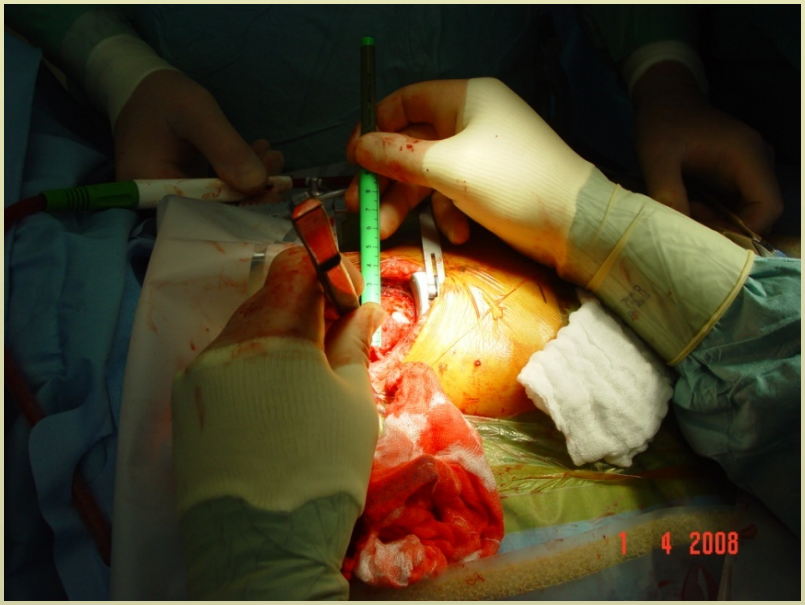


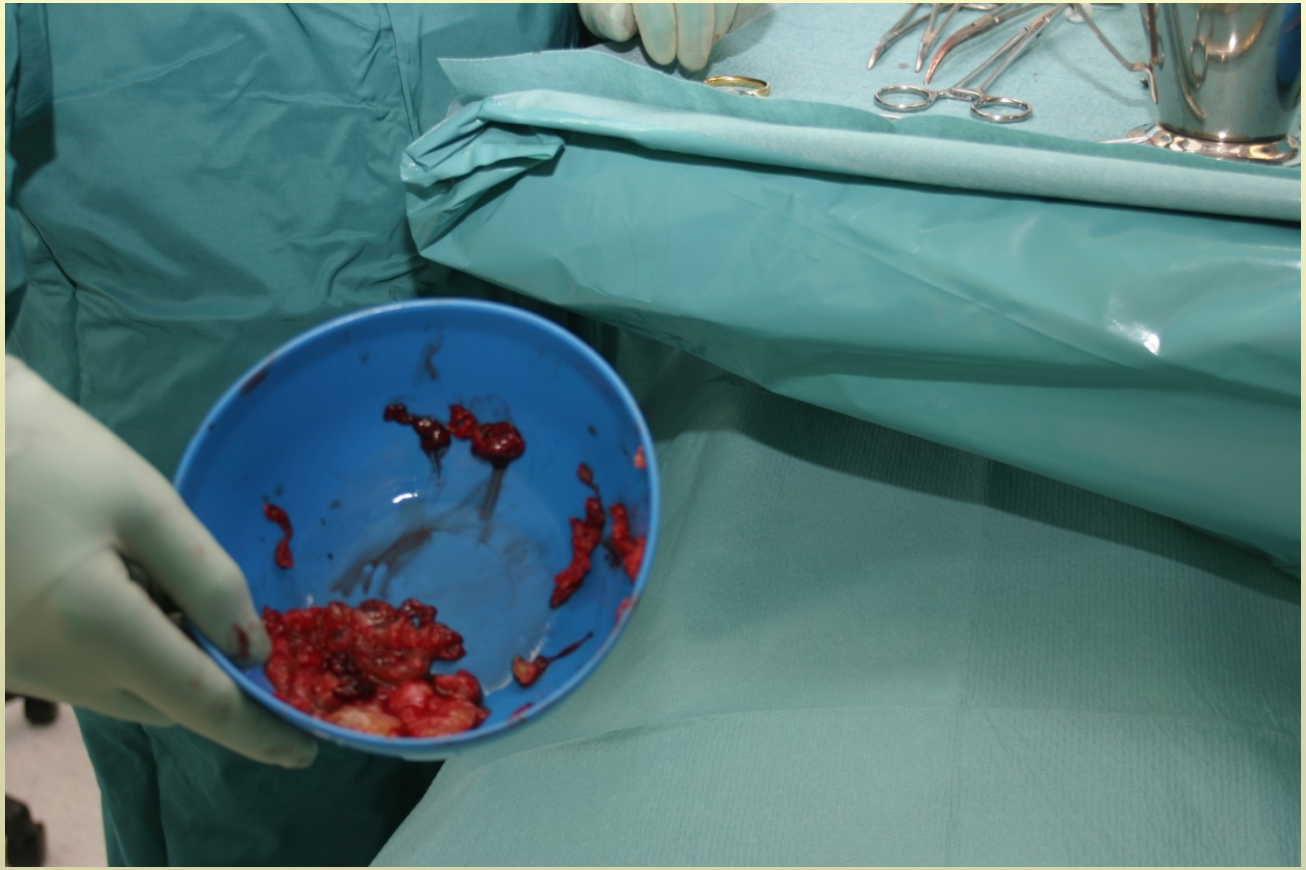
Case # 5
Decortication:











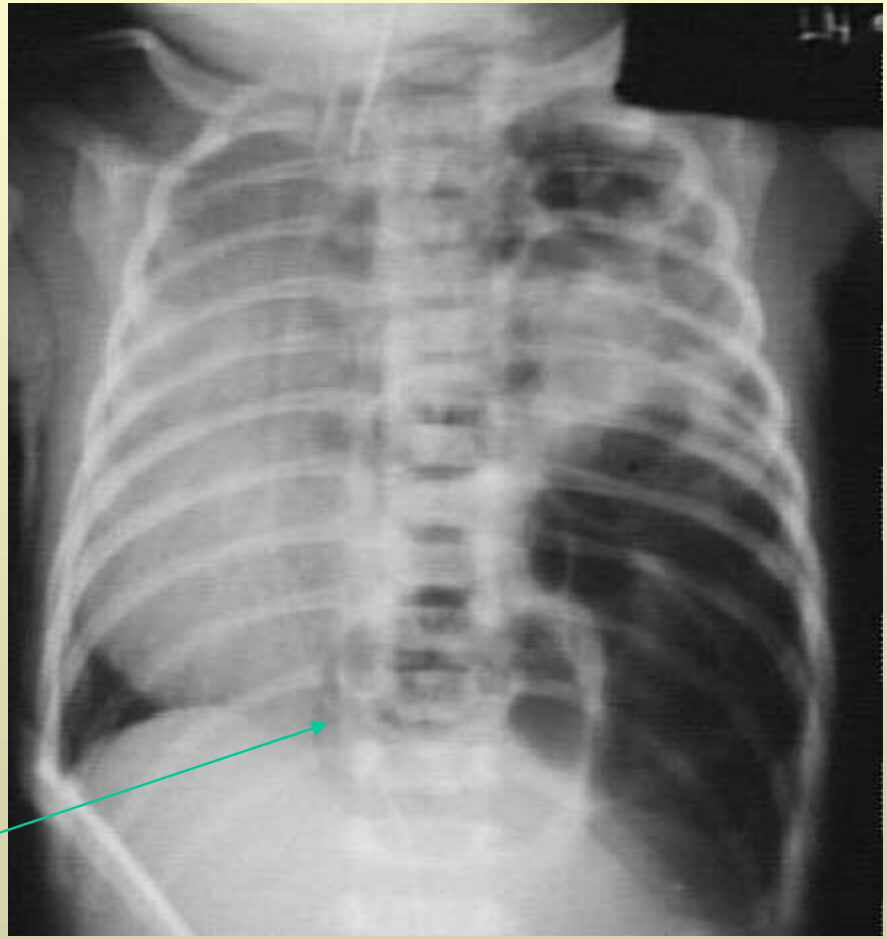
Pediatric Cardiothoracic Surgery

- Thorax
  - Veggur
  - Pleura
  - Lungu
  - Mediastinum
- Hjarta og æðar í thorax
  - Meðfæddir gallar
  - Áunnir gallar/ sjúkdómar

Andnauð við fæðingu
What is your differential diagnosis?

- Congenital diaphragmatic hernia
- Pneumothorax
- Congenital cystic adenomatoid malformation (CCAM)
- Pulmonary sequestration
  - Intrapulmonary
  - Extrapulmonary
- Congenital Lobar Emphysema
- Bronchogenic Cyst

CDH



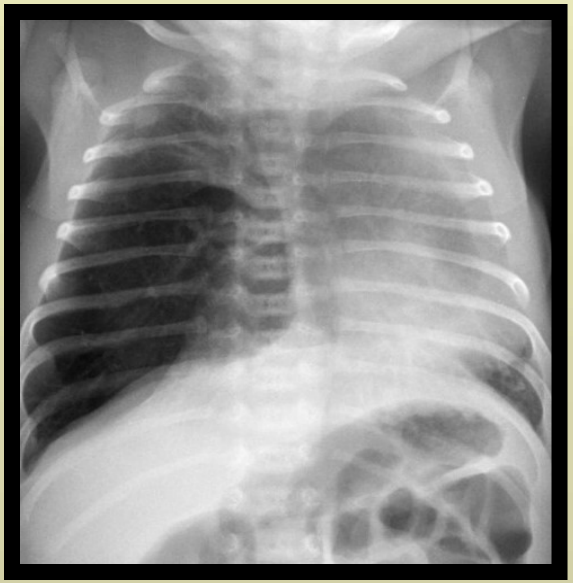
Treatment: CDH

- Delayed surgical approach; *not a surgical emergency*
- Conventional vent, Oscillator, "gentle ventilation", possible ECMO (10-15%)
  - Goal is prevention of barotrauma
- Primary repair; patch sometimes needed
- Overall survival 50-80 %

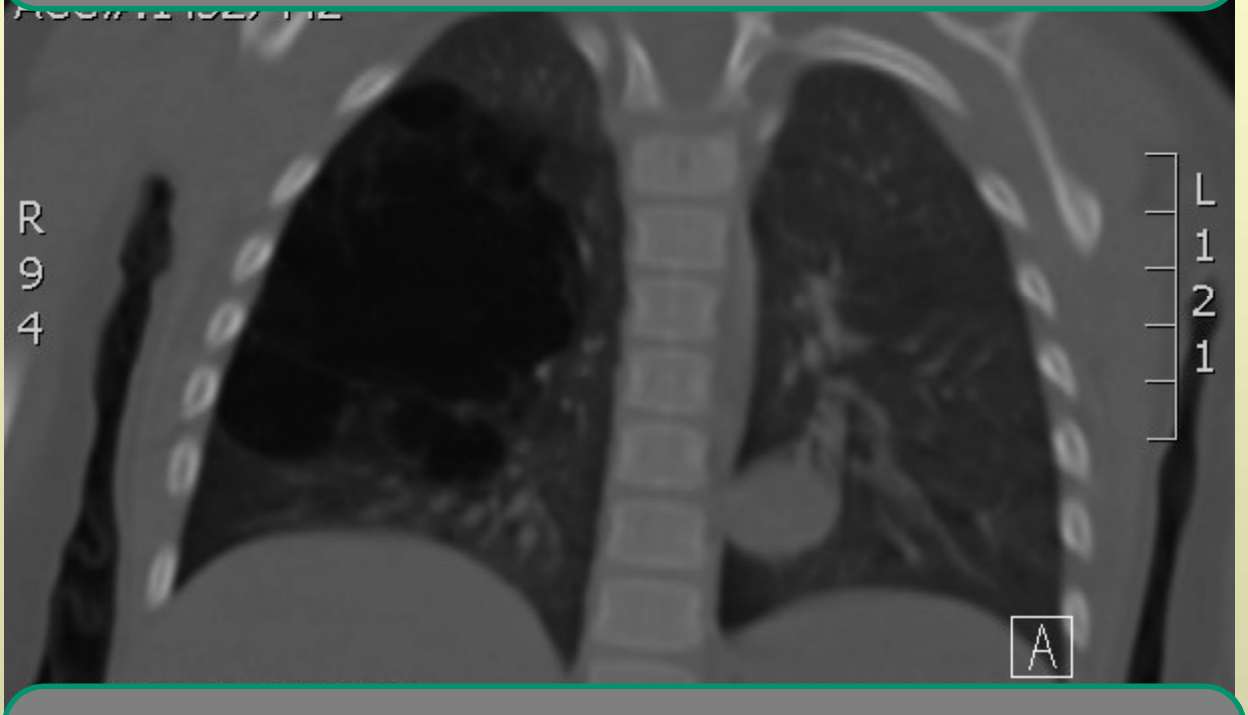
Congenital Lobar Emphysema:

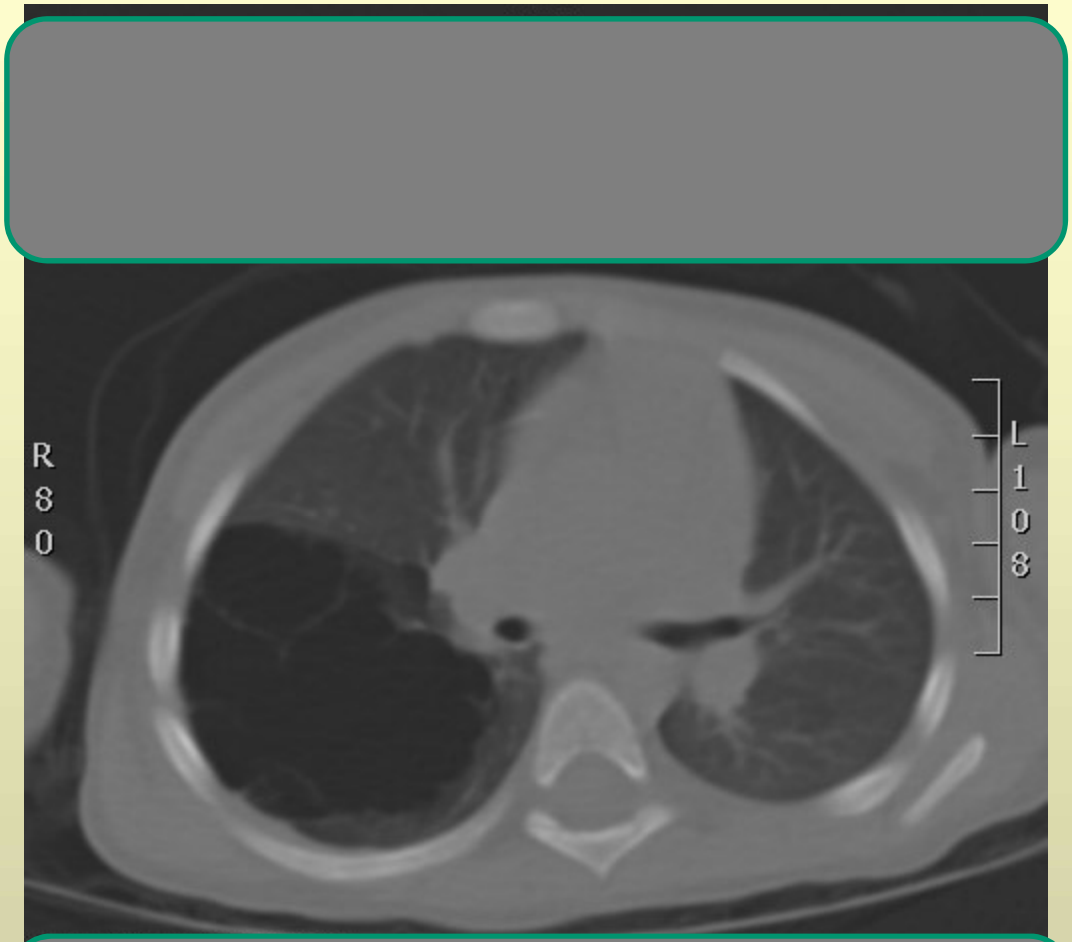
<u>Congenital Lobar Emphysema:</u>

- isolated idiopathic hyperinflation of one lobe; respiratory difficulties often at birth or in infancy; worsens with time by air trapping



Case #6

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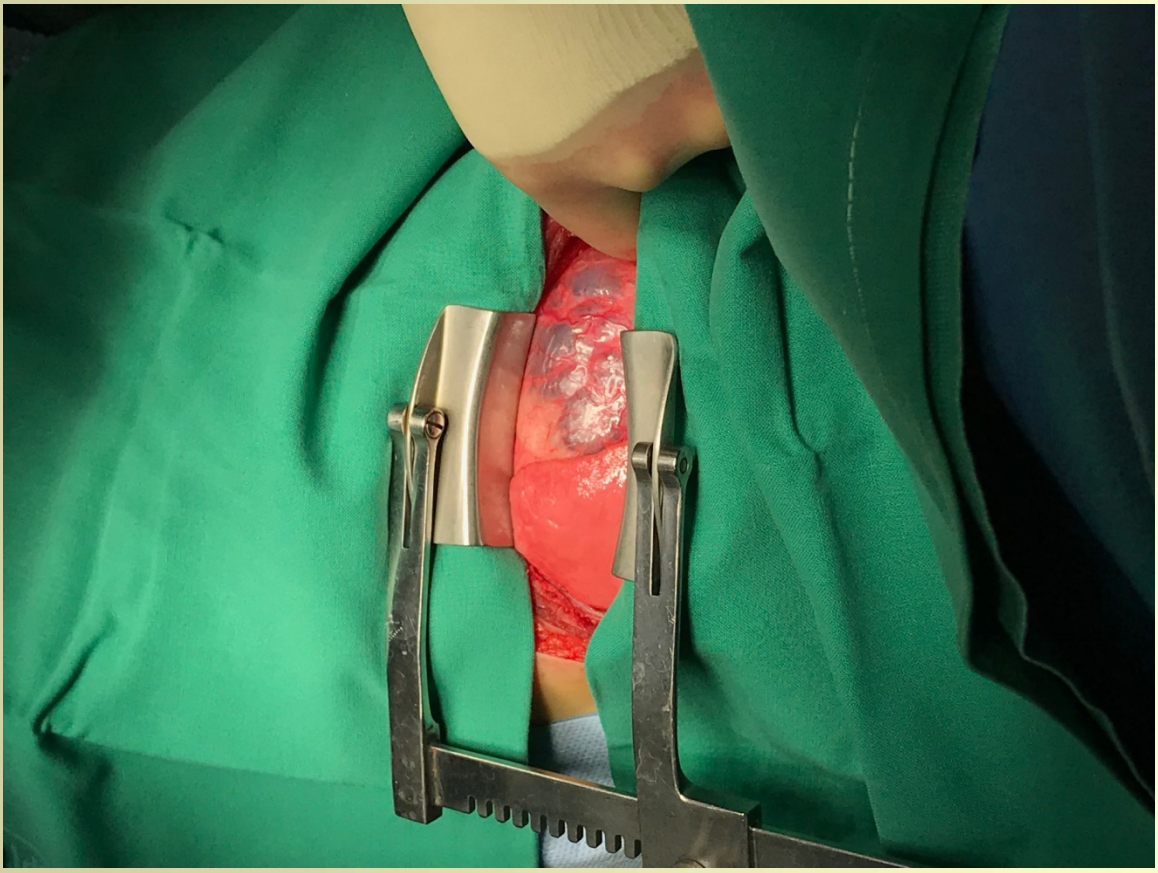
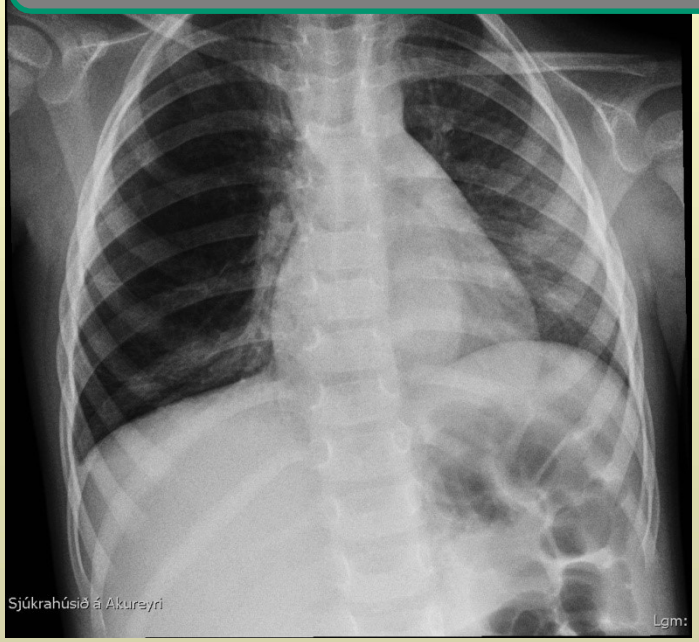



BJARNI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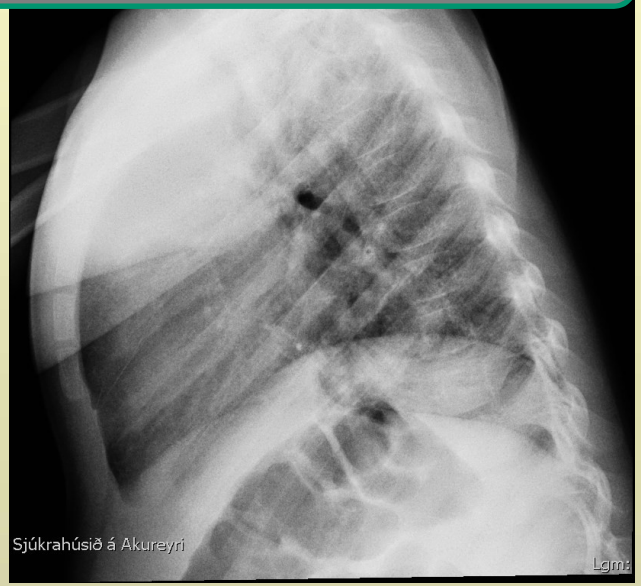
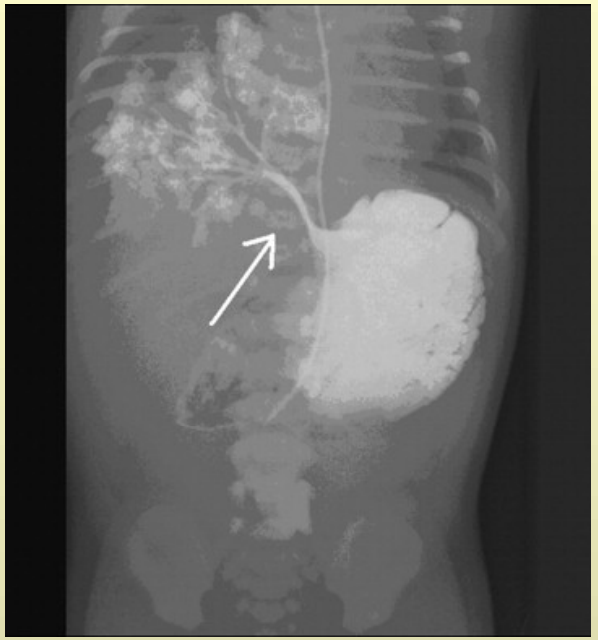
Lgm:

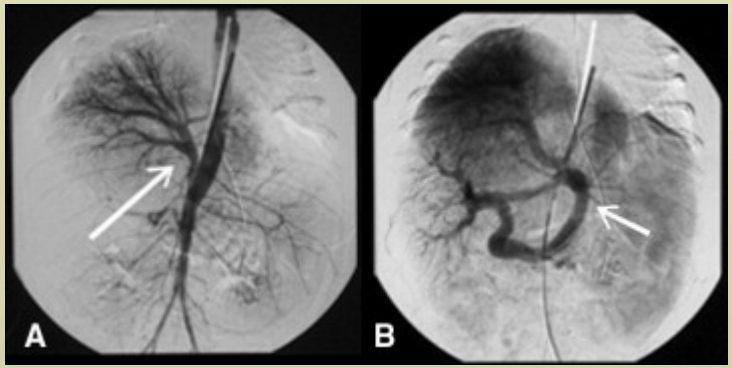

Sjúkrahúsið á Akureyri
Lgm:

Pulmonary Sequestration, Case #7:

Pulmonary Sequestration:

- a segment of lung without anatomic bronchial communication,
- systemic arterial supply from thoracic or abdominal aorta
- "Extralobar": often incidental
- "Intralobar": found within normal lung parenchyma (lower lobes); prone to infection

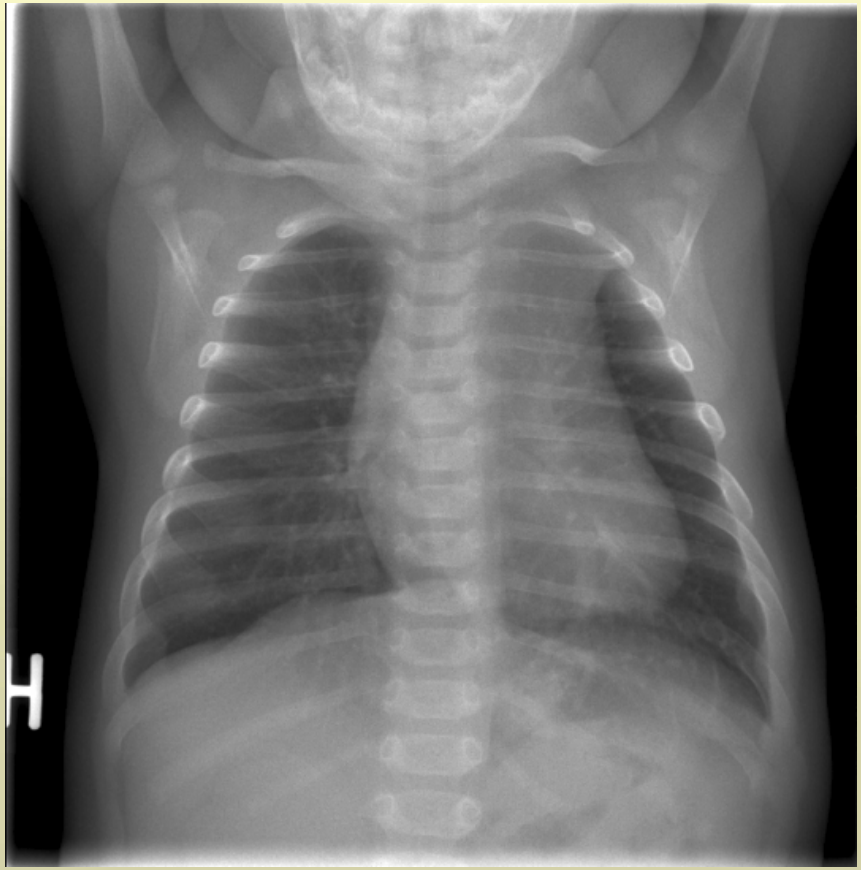


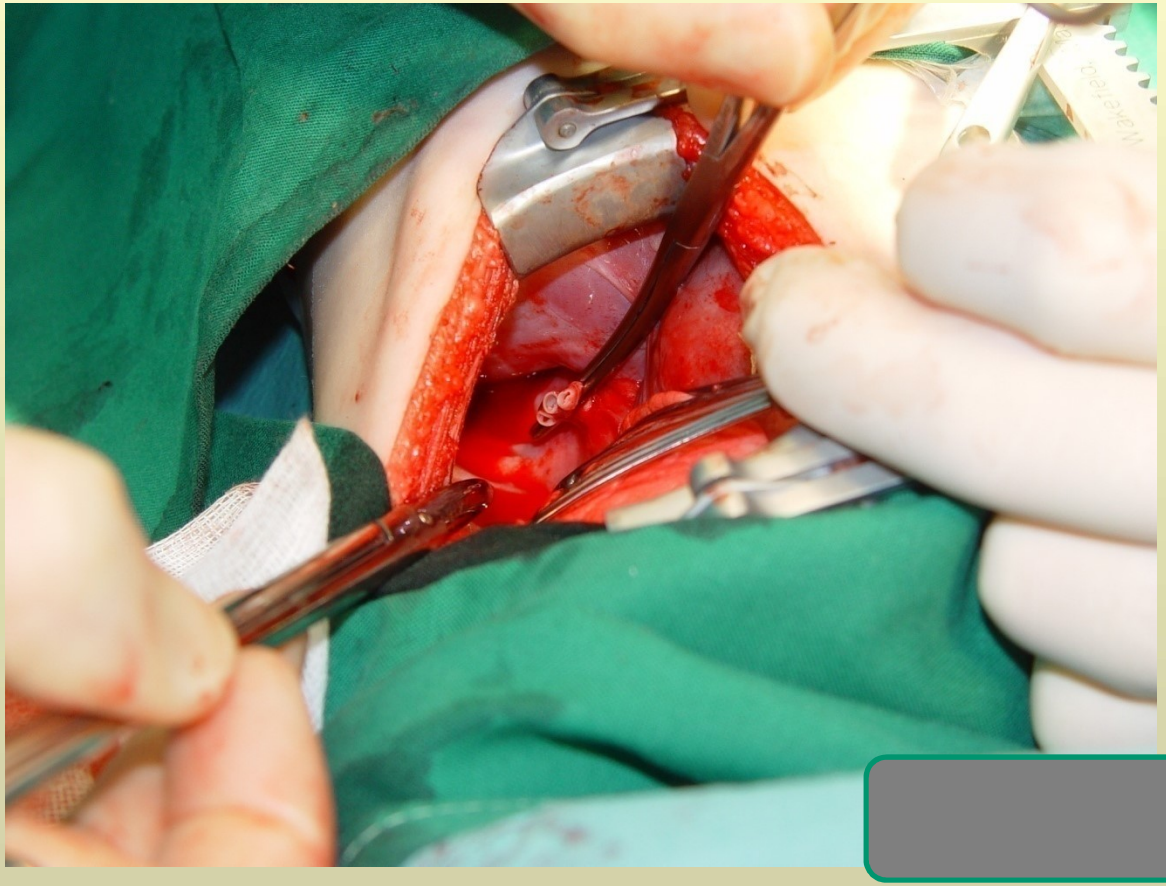


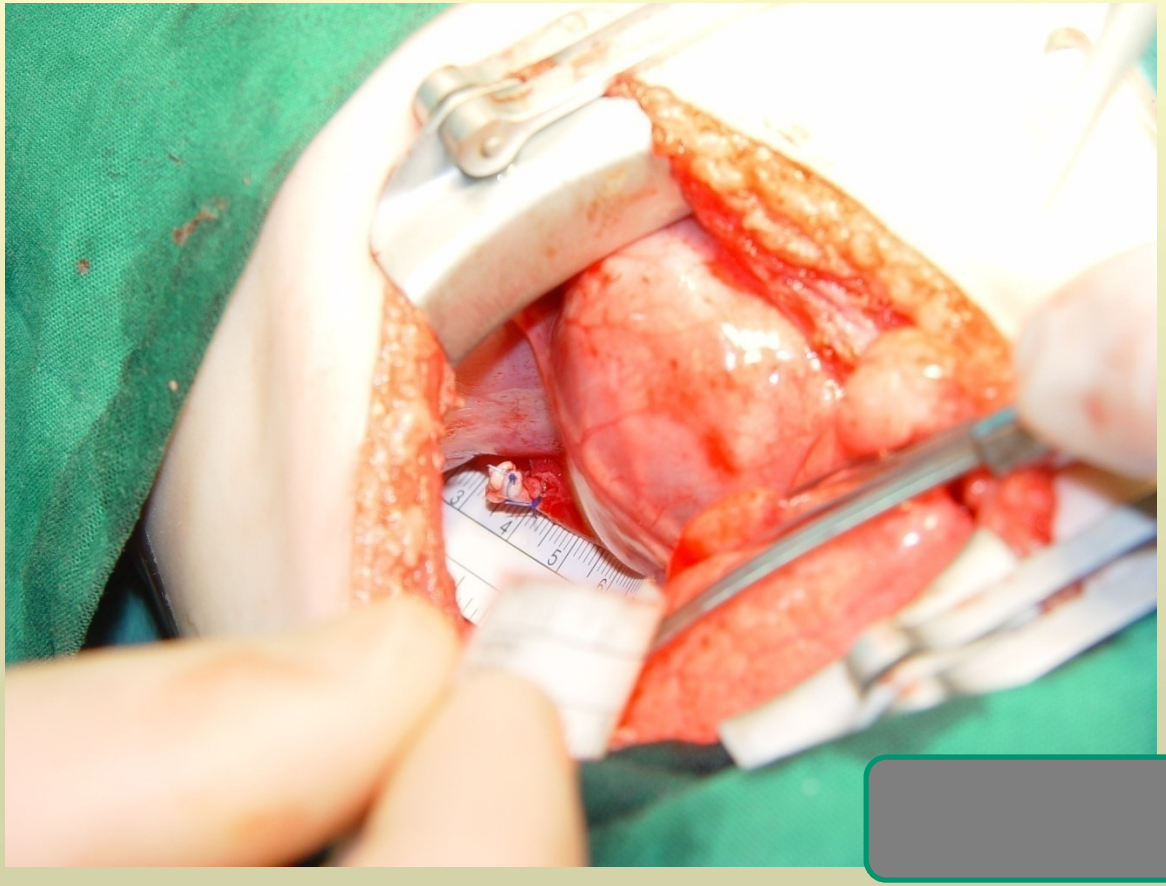
Linkur:

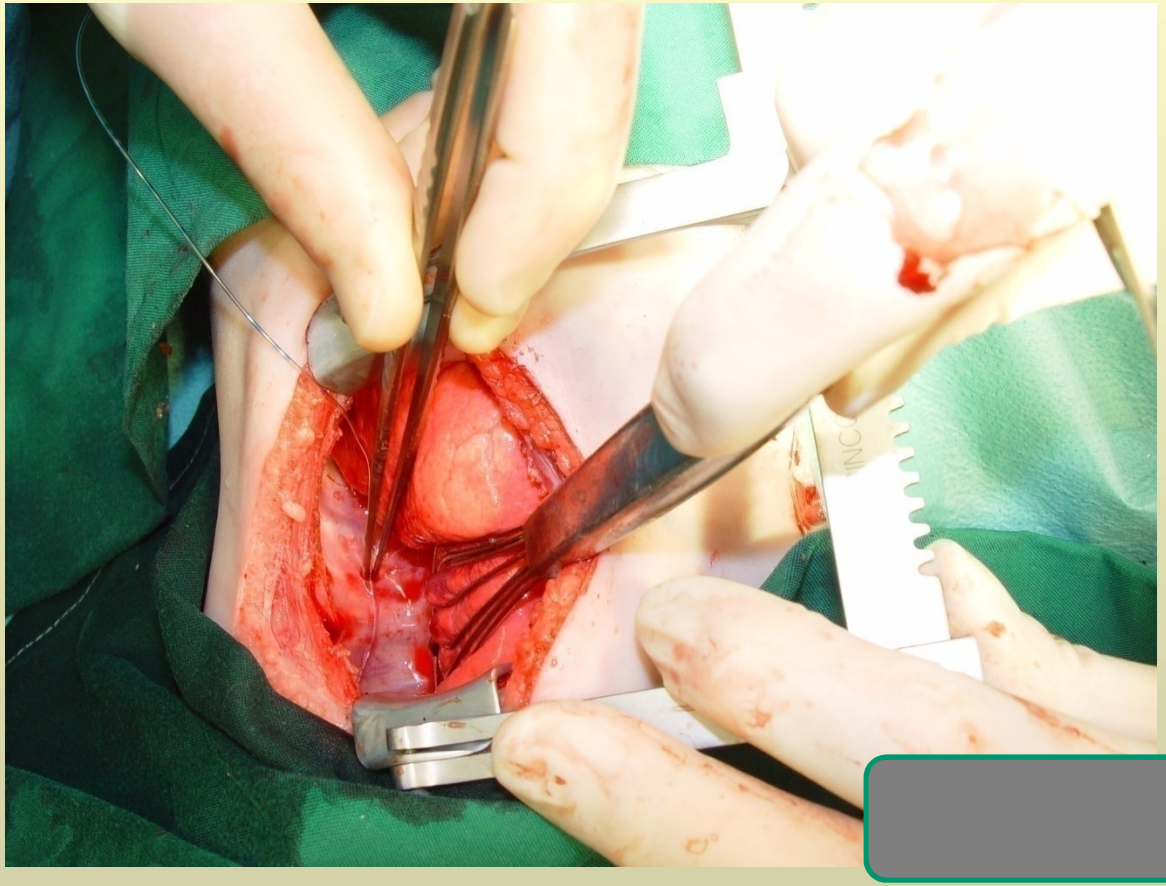
https://www.annalsthoracicsurgery.org/article/S0003-4975(07)00989-7/fulltext

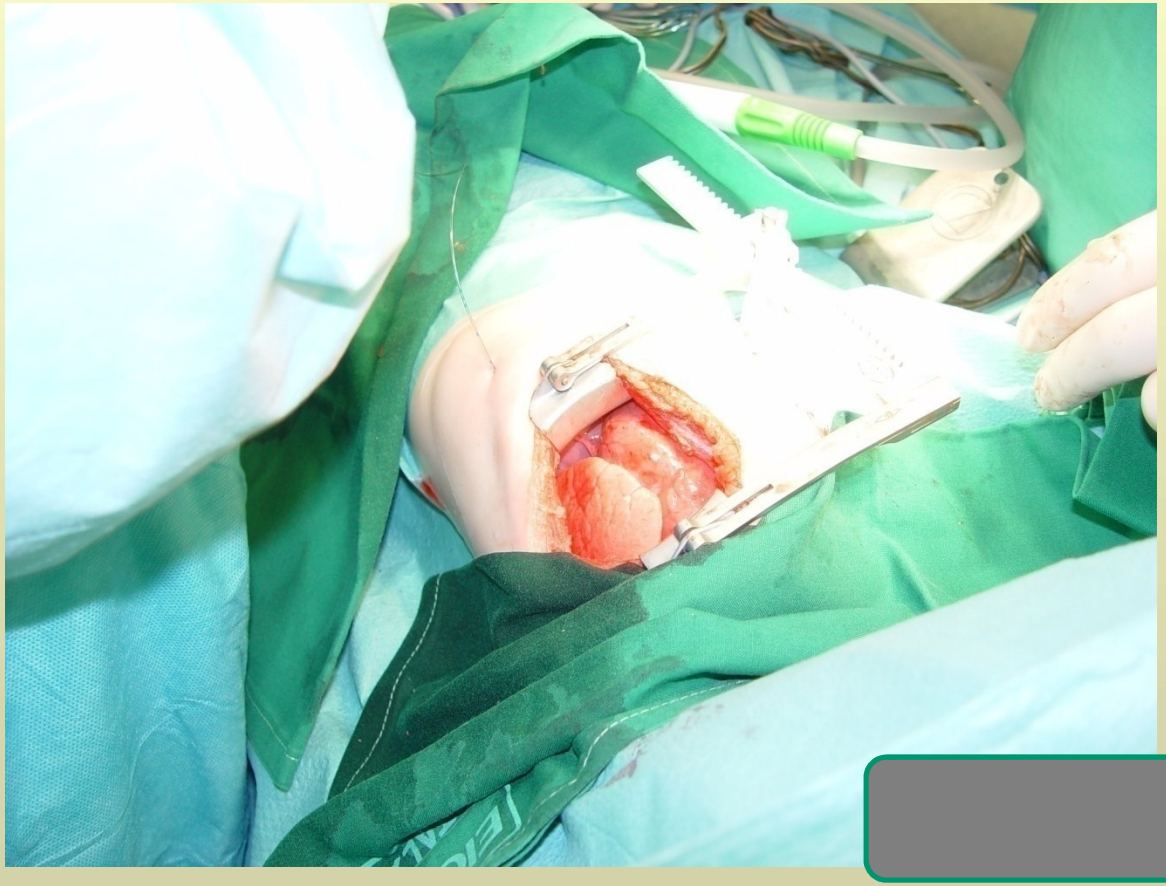
Case #8

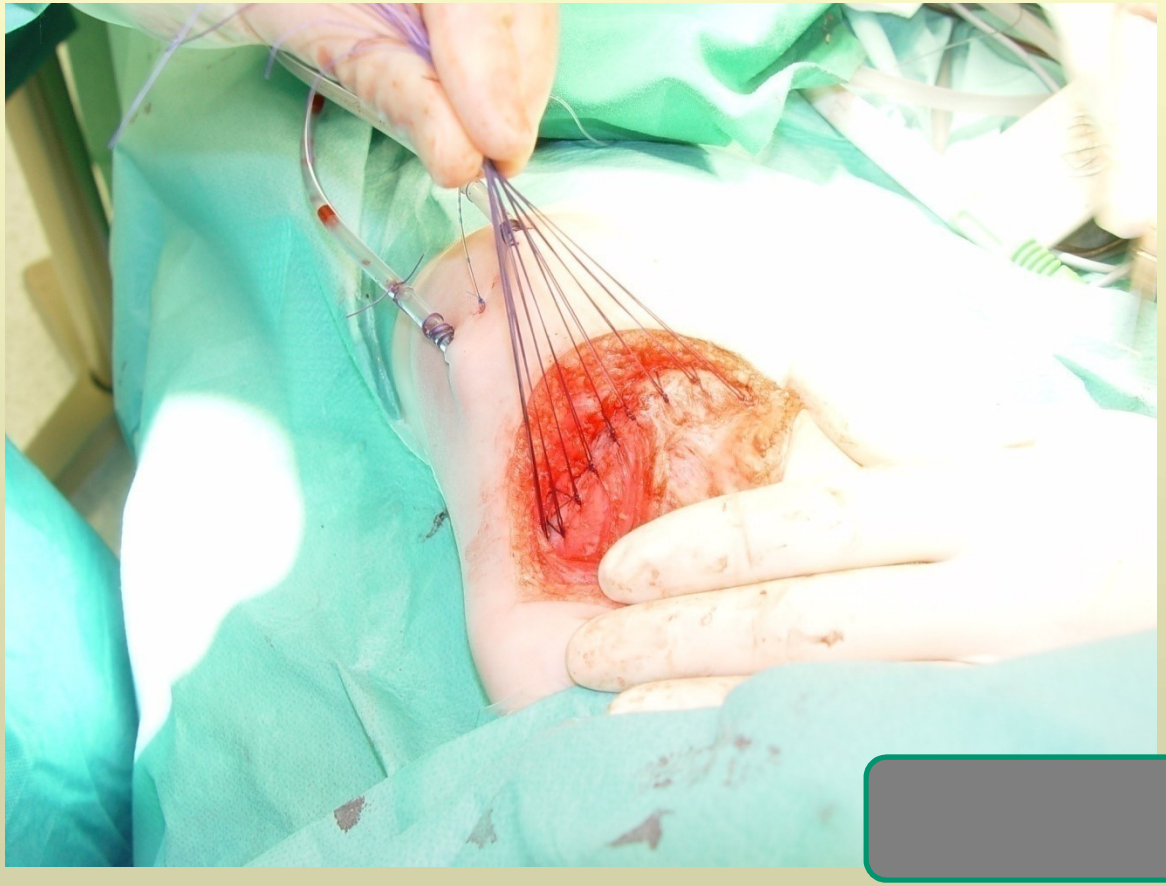


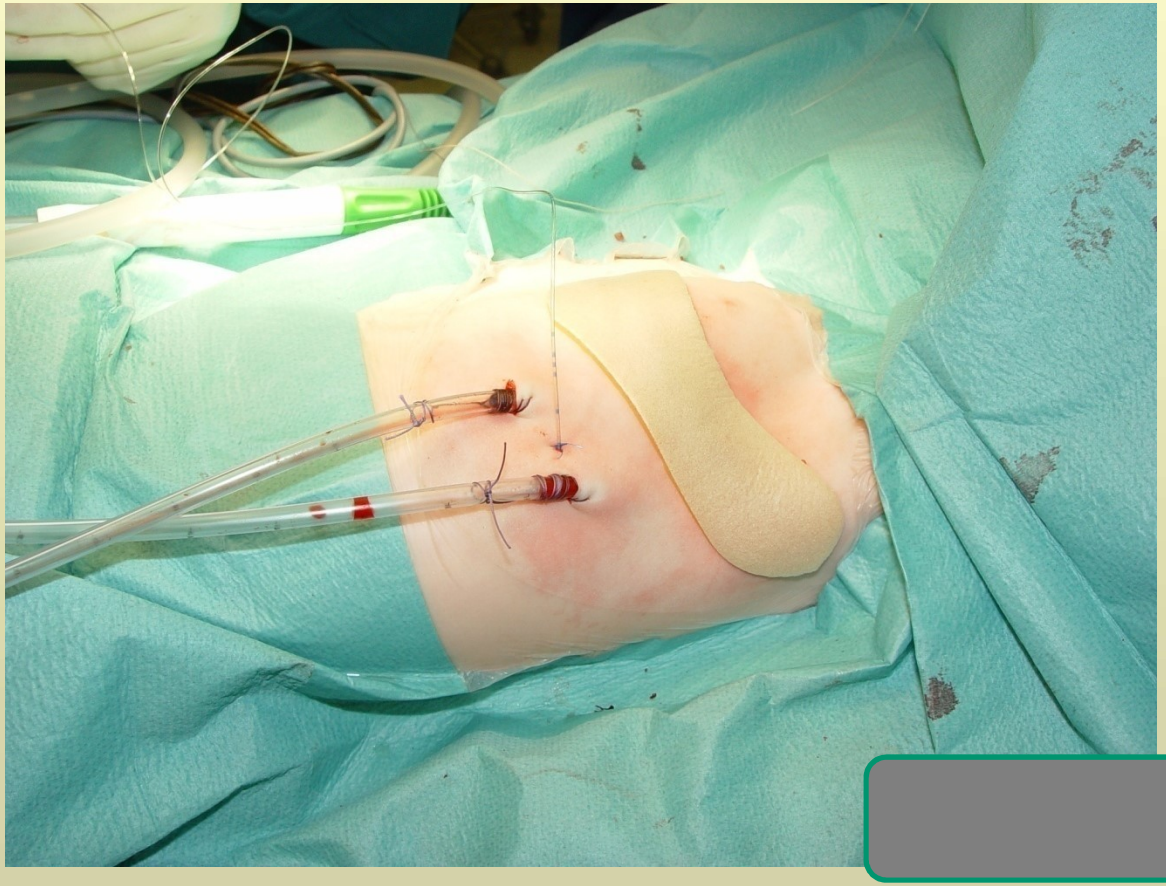


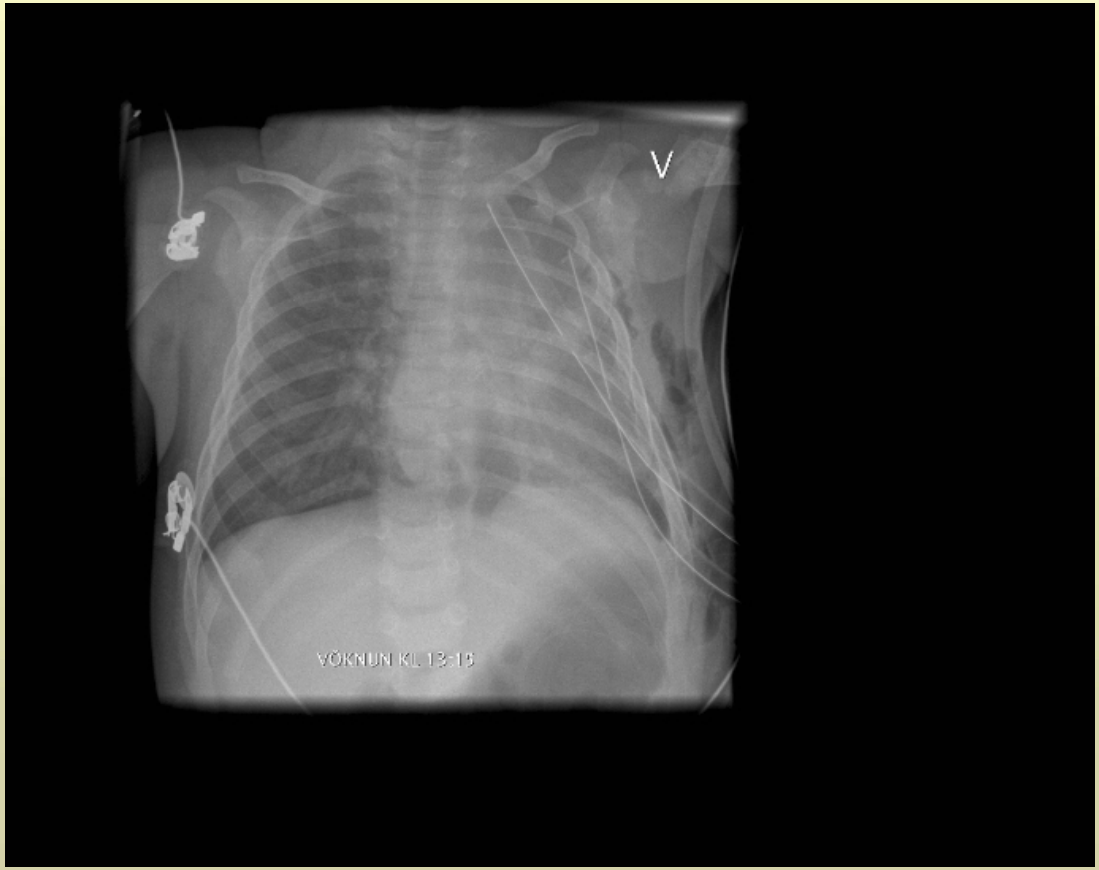










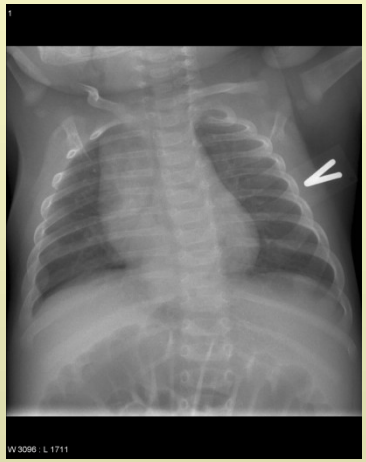

V
VÖKNUN KL 13:19

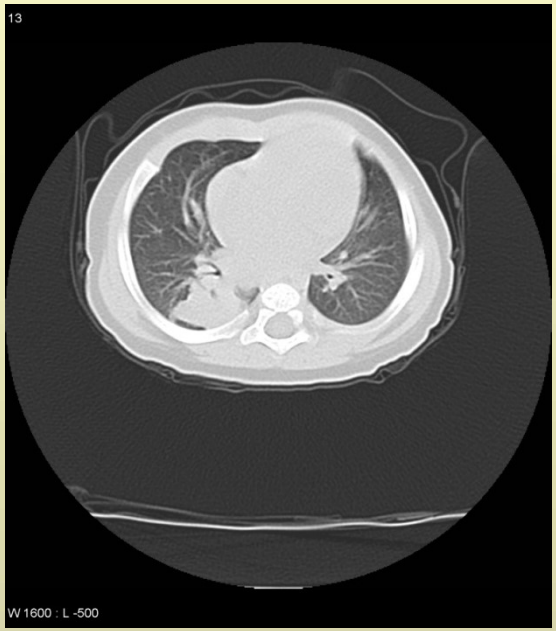
CCAM

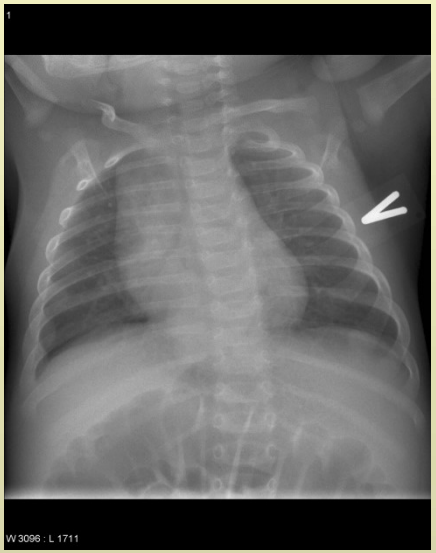
Congenital Cystic Adenomatoid Malformation (CCAM)

- solid/cystic lung malformation
- Can present at prenatal U/S or resp distress at birth or with infection in first few years of life. If large, can cause fetal hydrops.

Case # 9



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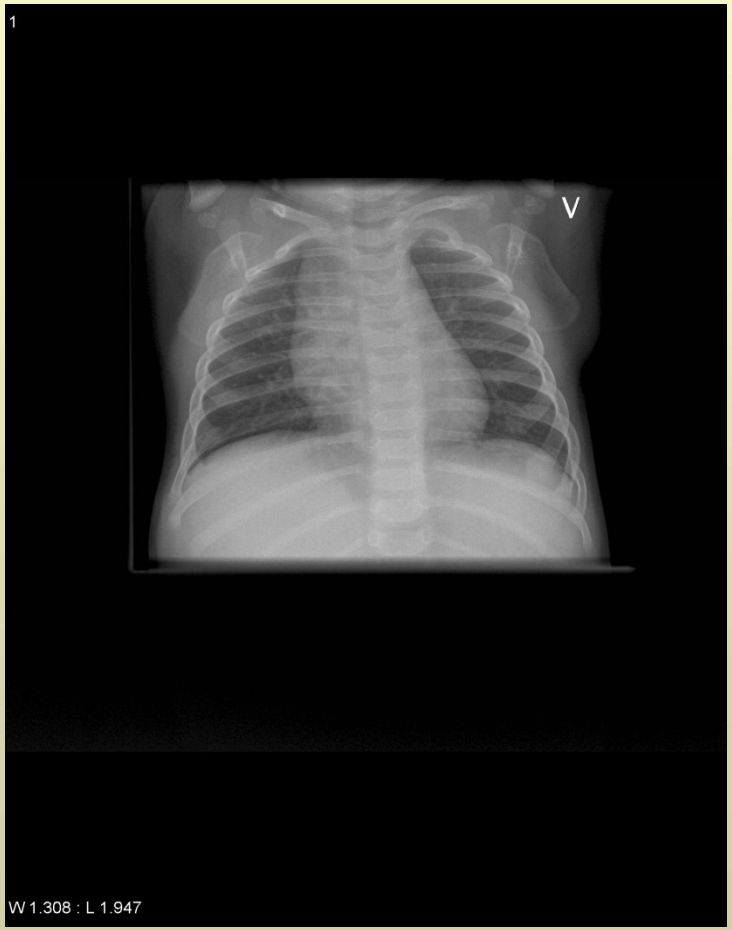
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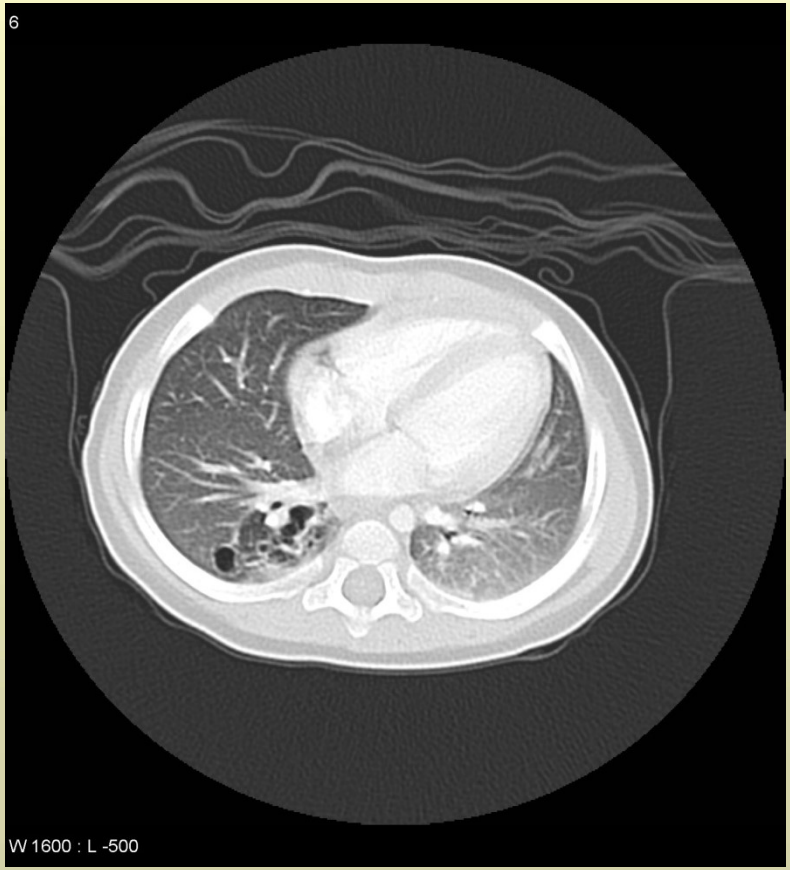
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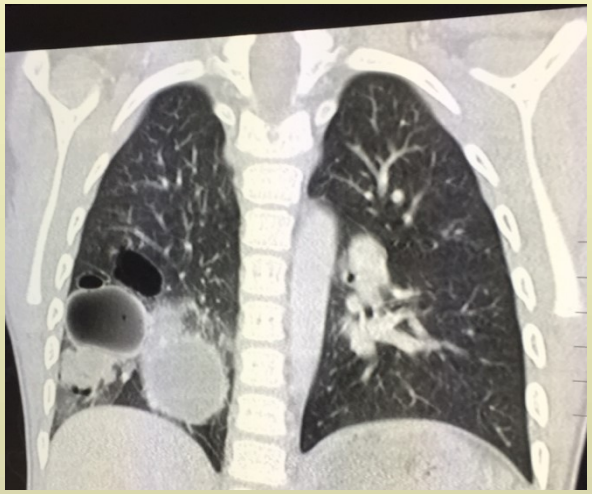
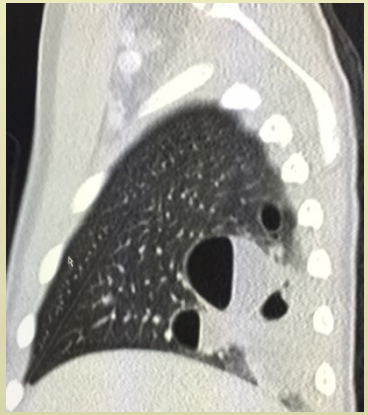
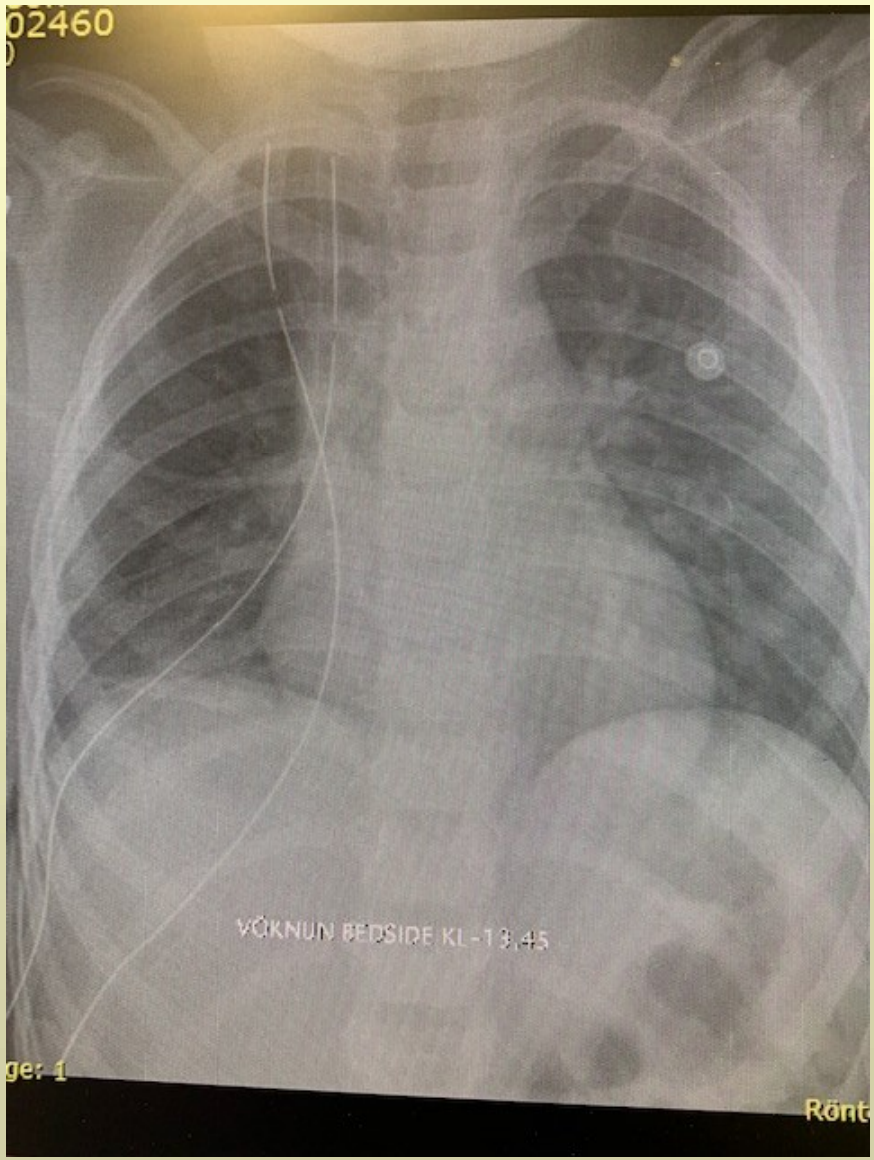
CCAM? Case #10






Help
11
27 of 52


11
A
29 of 40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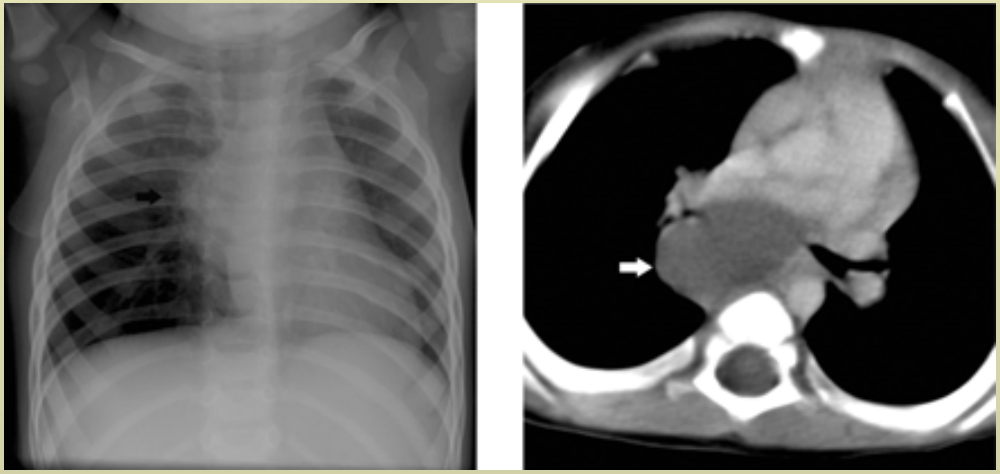
VÖKNUN BEDSIDE KL - 13.45
ge: 1
Rönt

Bronchogenic Cyst

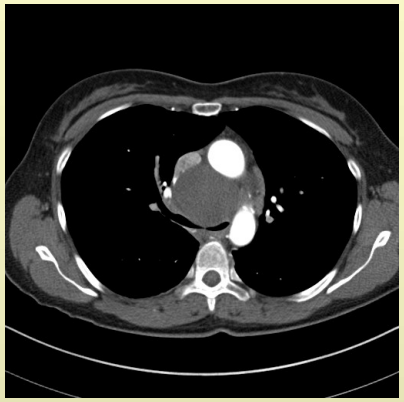
Bronchogenic Cyst

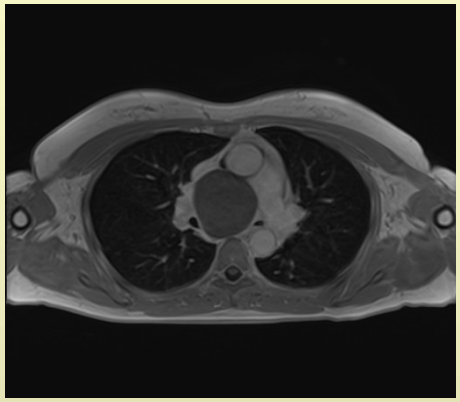
- Cyst found in hilum, mediastinum, or within lung parenchyma.
- Can compress airway and cause atelectasis, pneumonia, air trapping.

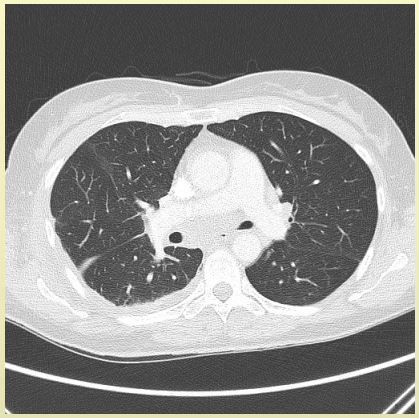


Case # 11

Pre op og post op

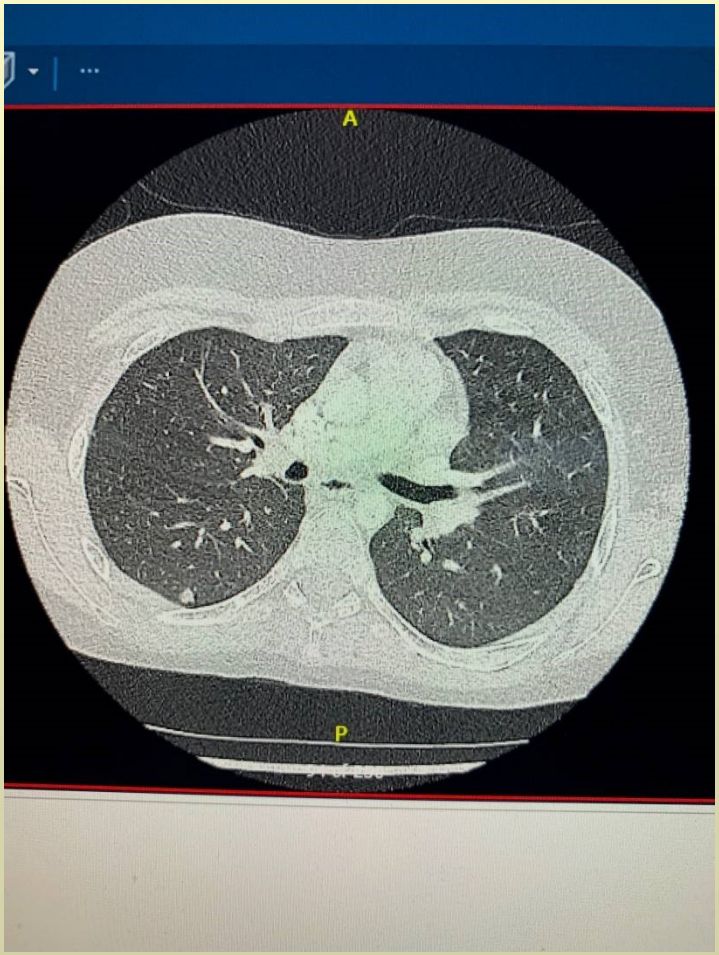






Æxli-Metastasis

Hepatoblastoma Case #12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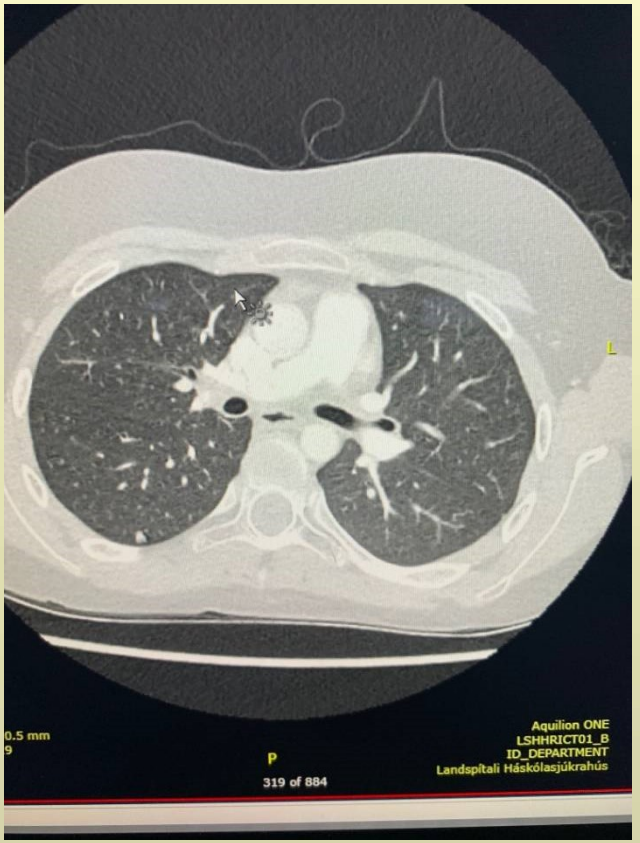
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Aquilion ONE
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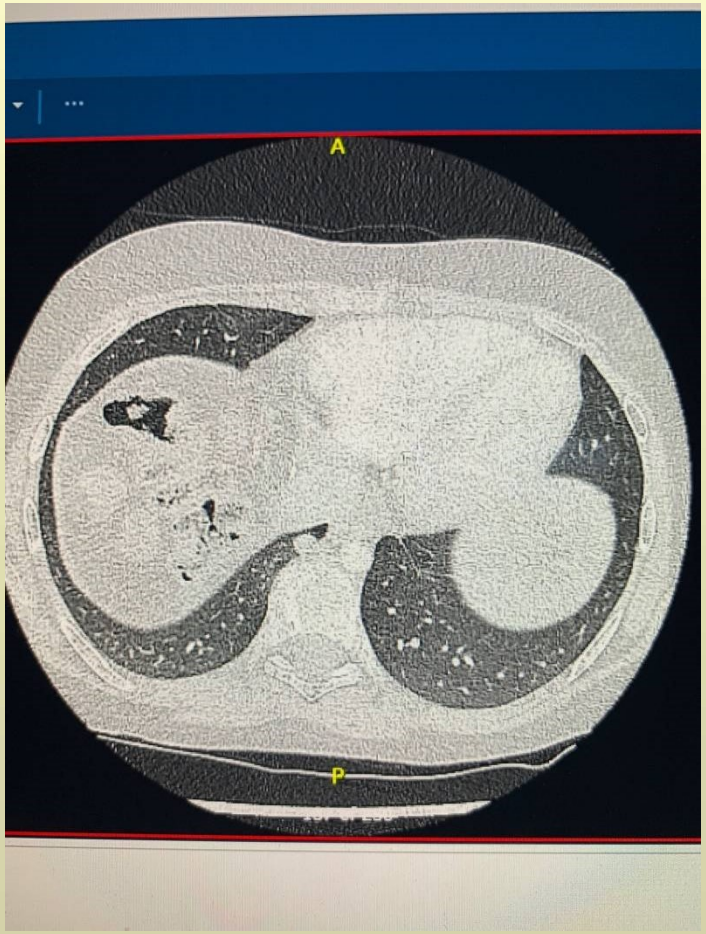
ID_DEPARTMENT
Landspítali Háskólasjúkrahús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546 of 884
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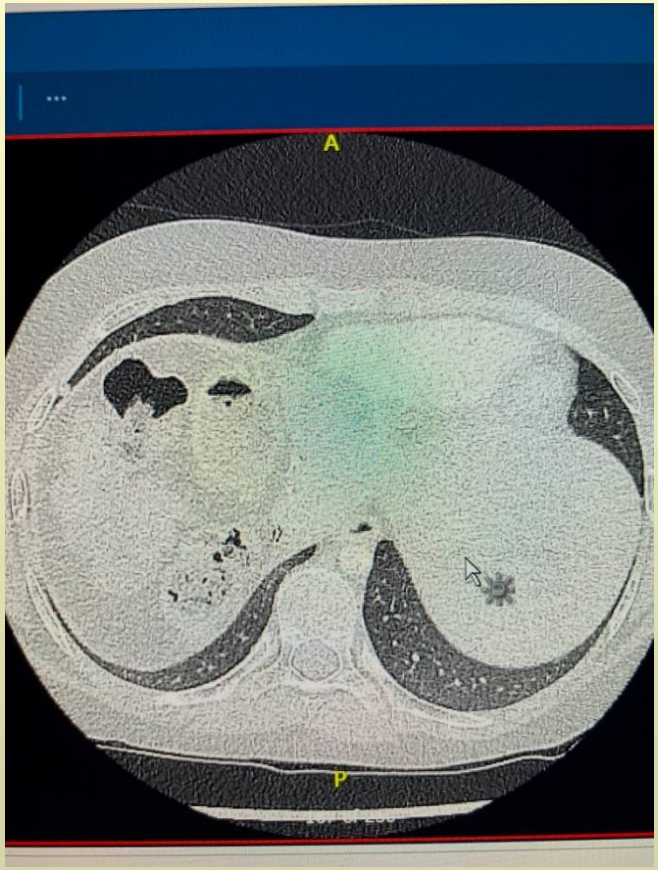
LSHHRICT01_B
ID_DEPARTMENT
Landspítali Háskólasjúkrahús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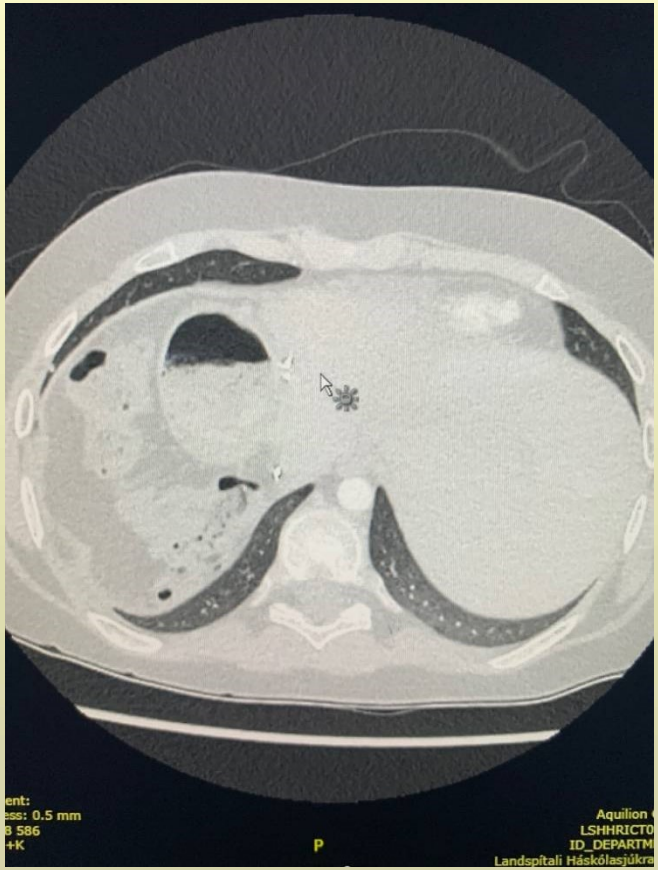
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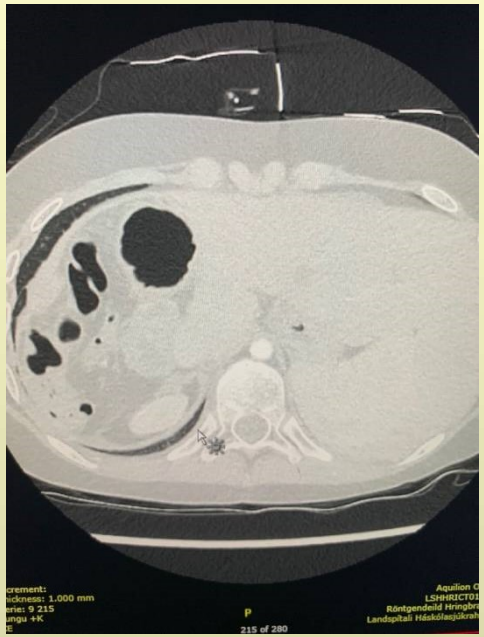
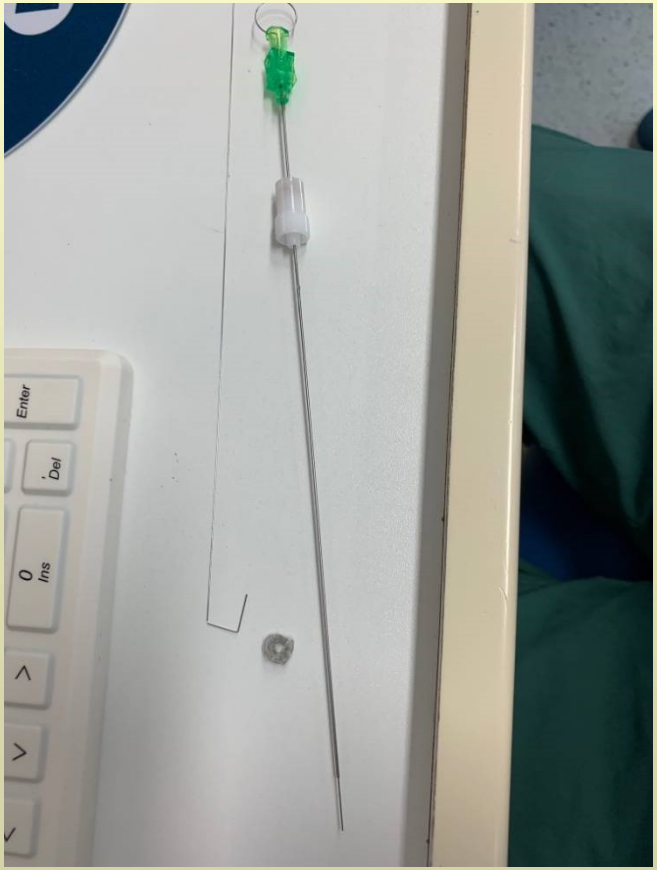
Þindarslit og hernia

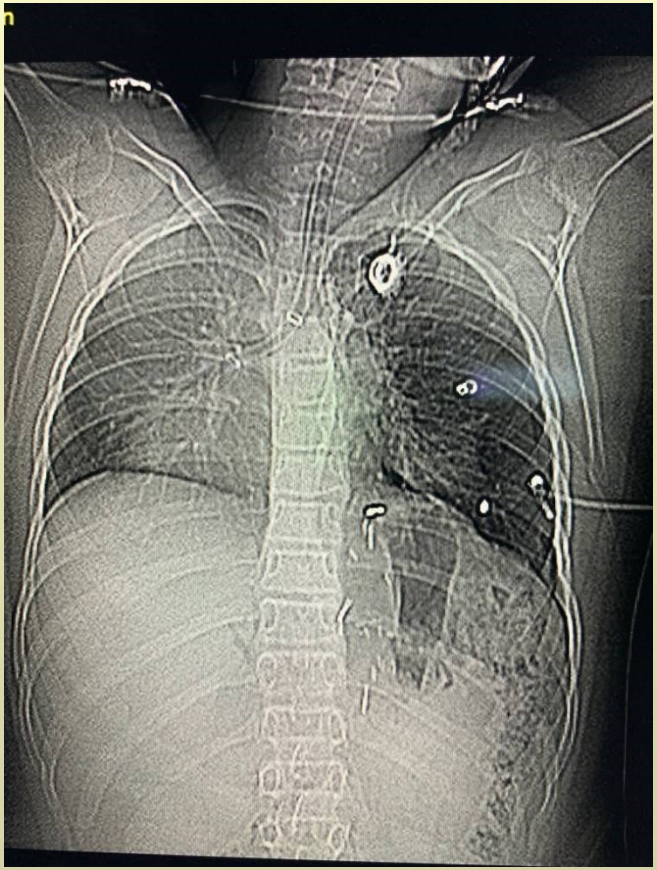
Preop



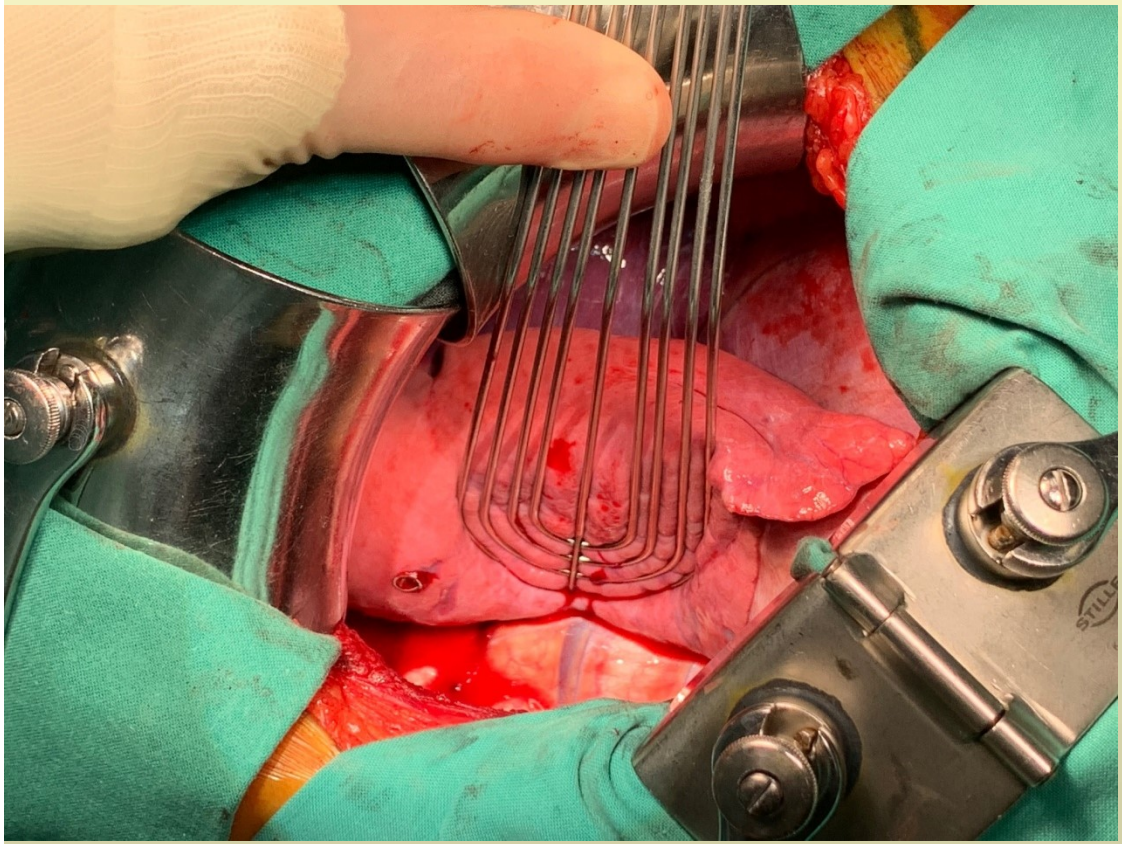
Coil til merkinga



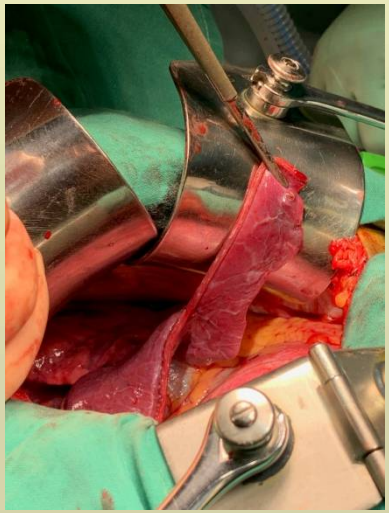
CT coil merking

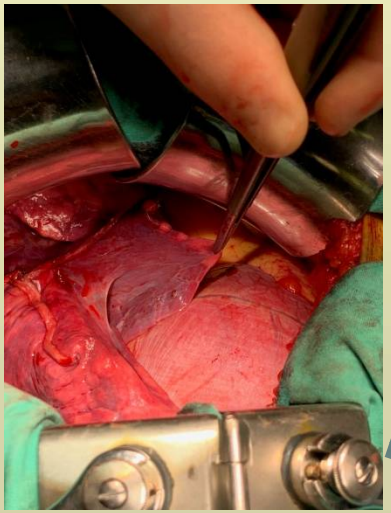


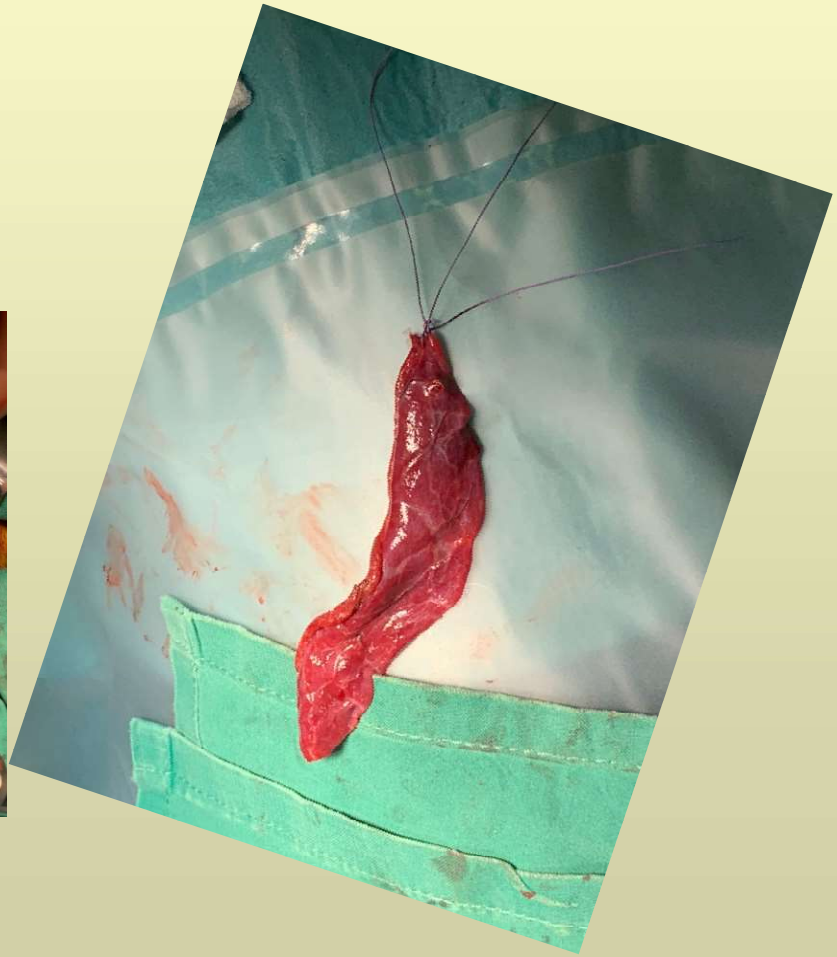
Op
Coil í lobus inf dxt posteriort



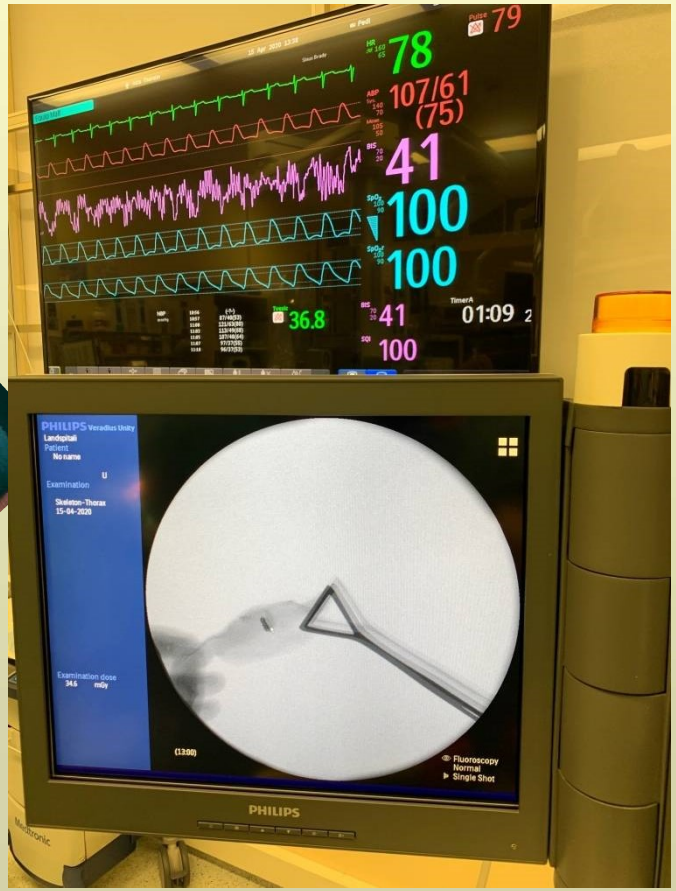
Coil í lobus Med inferiort



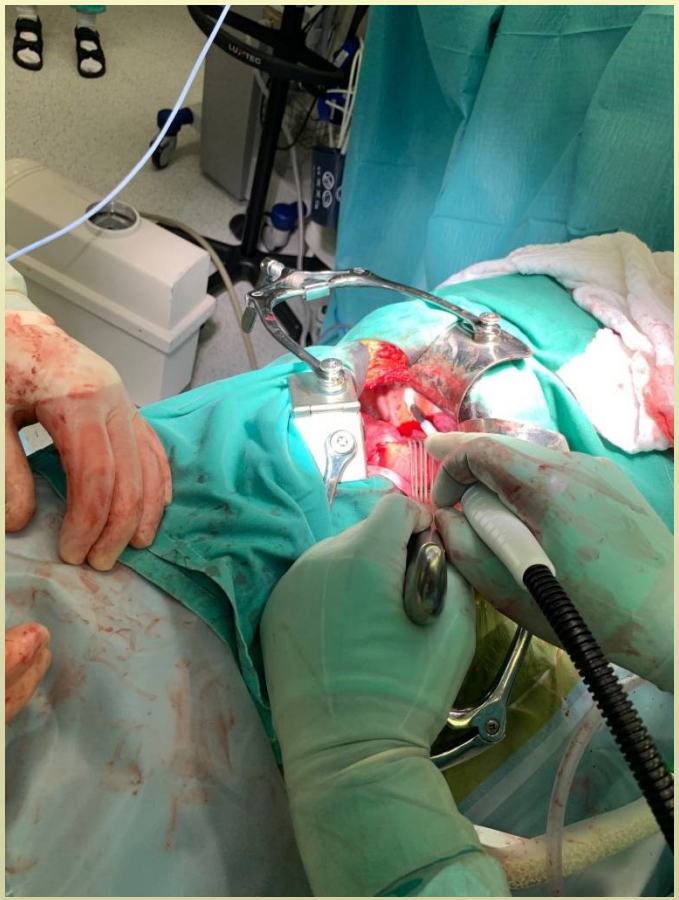




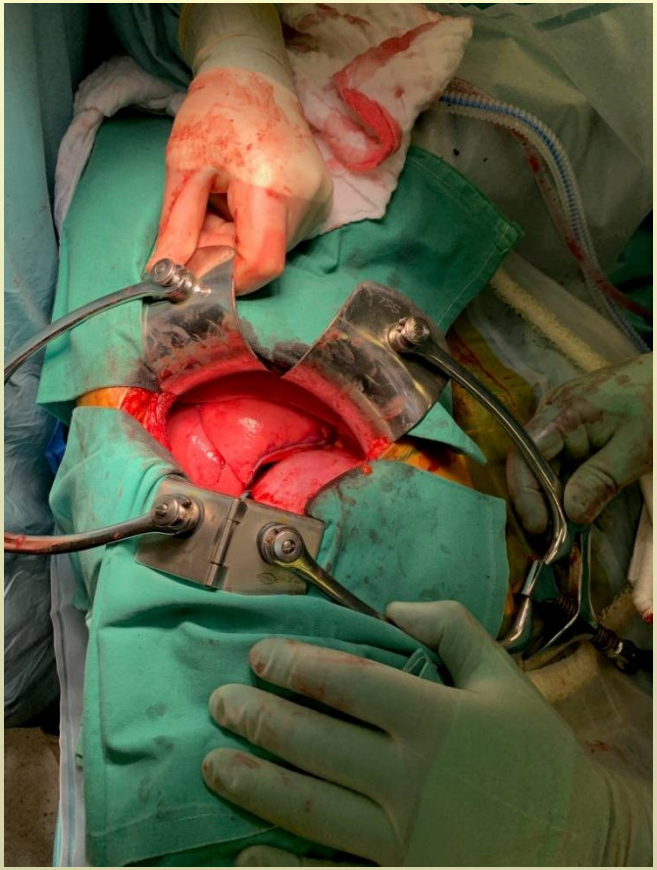
Coil í lobus inf dxt medialt staðsett með rtg skyggningu



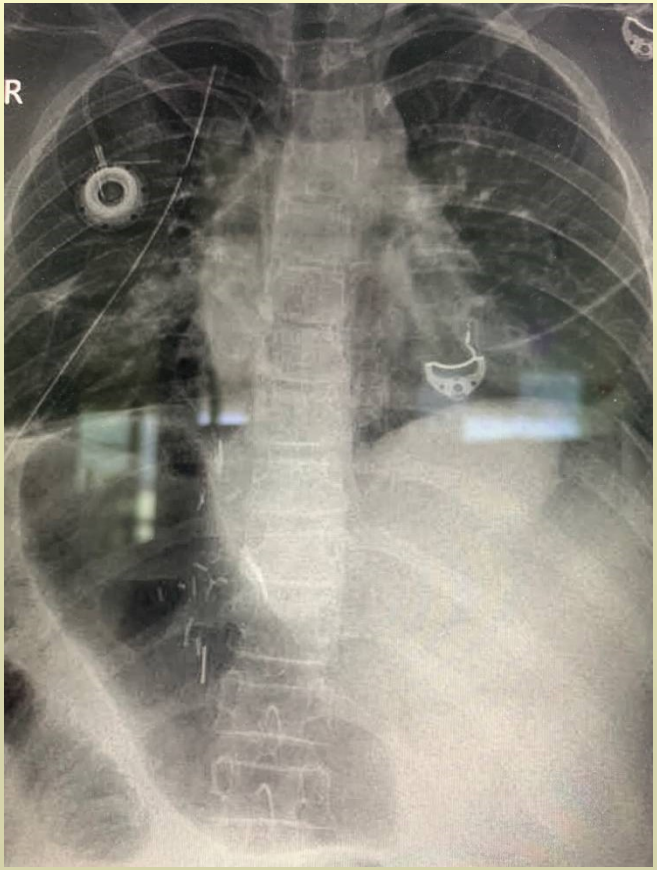
Verkjastilling:
Frysting intercostal tauga 4.-8.



Aðgerð lokið: Ekki loftleki



Postop rtg á Vöknun



Pediatric Cardiothoracic Surgery

- Thorax
  - Veggur
  - Pleura
  - Lungu
  - Mediastinum
- Hjarta og æðar í thorax
  - Meðfæddir gallar
  - Áunnir gallar/ sjúkdómar

Pediatric Thorax Trauma
+ see last years lectures Tx Trauma

Organ Weight During Violent Impact at Various Velocities

Weight (kg)	40 km/h	72 km/h	105 km/h
HEART (0.35)	3.5 kg	14 kg	31.5 kg
BRAIN (1.5)	15 kg	60 kg	135 kg
Whole	700 kg	2800 kg	6300 kg

Prevalence of Pediatric Trauma

- Trauma is the leading cause of death in infants and children
- Trauma is the cause of 50% of deaths in people between 5 and 34 years of age
- Motor vehicle related accidents account for 50% of pediatric trauma cases
- $16 billion is spent annually caring for injuries to children less than 16 years of age

Anatomic Characteristics of the Pediatric Patient and Significance to Trauma Care

Variable	Significance
•Large volume of blood in head	•Cerebral edema develops rapidly
•Poor muscular support in neck	•Flexion/extension injuries occur
•Decreased alveolar surface area •Increased metabolic rate	•Injury leads to rapid compromise
•Decreased airway caliber	•Increased airway resistance
•Heart higher in chest, •Small pericardial sack	•Prone to injury and cardiac tamponade
•Thin walled, small abdomen	•Organs not well protected

Anatomic and Physiologic Differences Child/Adult

- Pliable chest wall
  - *Transmission of force within the thoracic skeleton to the pulmonary parenchyma*
- Mobility of the mediastinal structures
  - *Vulnerability to tension pneumothorax and vascular disruptions*

Chest Trauma

- 5 - 10 % of all injuries involve the chest
- 2/3 chest injured children have injury in another organ system
- thoracic injuries account for ¼ of pediatric trauma related deaths
- Rib fractures are a marker of significant force and occur with < 50 % of chest trauma

Pediatric Cardiothoracic Surgery

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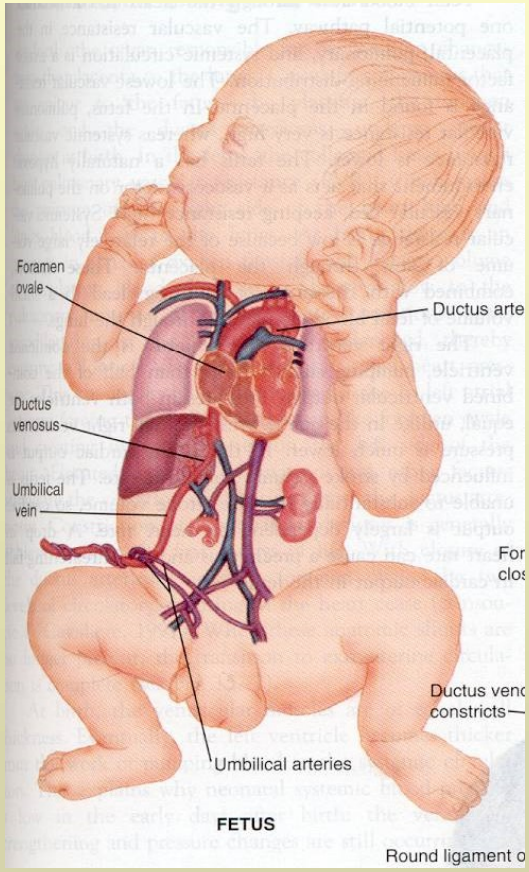
Cardiovascular Surgery



Nokkrir helstu frumkvöðlar

- 1952 Robert Edward Gross
  - Atrial trekt "Open heart surgery" ASD 0/3
- 1952 Floyd John Lewis
  - Hypothermia "Open heart surgery" A
- 1953 Dr. John Gibbon
  - CPB support ASD 1/5
- 1954 Dr. Walt Lillehei
  - Cross circulation VSD
  - Tetralogy of Fallot

Atrial Septal Defect repaired
with the Gross Atrial Well

Hiti og kuldi

- Hiti er merki um líf og bruna
- Metabolisk vörn í náttúrunni:
  - Hibernation: dýr liggja í dvala yfir veturinn.
  - Animals seek cool environment when ischemic
- ” kæling matvæla fyrirbyggir skemmdir.
- Reynslan hefur kennt okkur að:
  - ”Ice on injuries reduces swelling and tissue damage”
- Physiological and biochemical rationale
  - Tissue metabolic rates decrease as body temperature decreases

Surface Cooling Limitations

- Surface cooling used in COOL AID* feasibility trial demonstrated limitations of surface cooling
- Target temperature of 32°C
- Actual temperatures reached as low as 28°C
- Wide range in actual temperatures and slow cooling times
- Intensive cooling regime including blankets, alcohol wipes with paralyzed and intubated patients

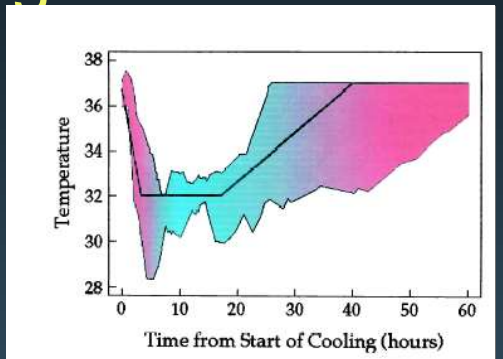
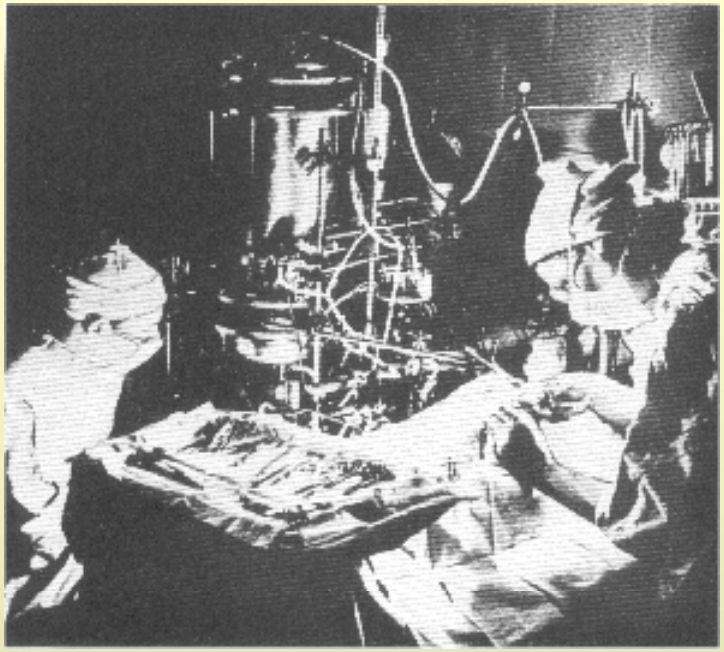
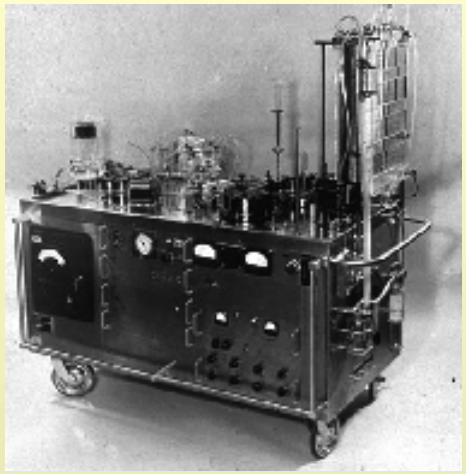


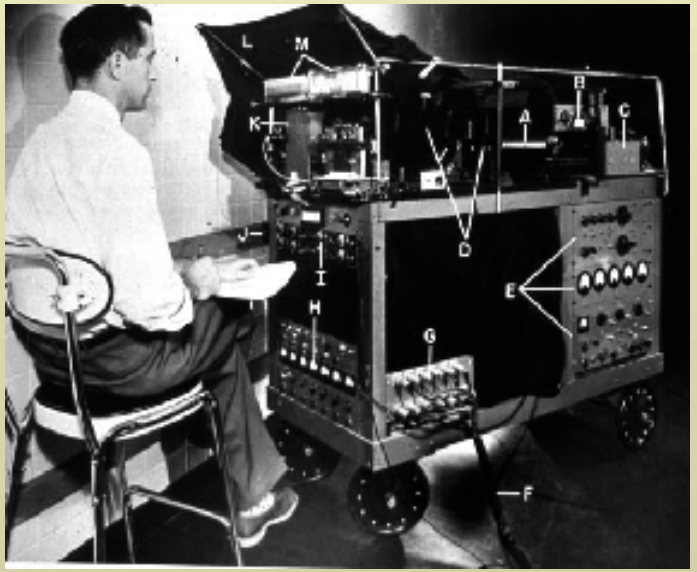
Figure 1. Representation of bladder temperatures obtained during initiation, maintenance, and termination of moderate hypothermia.

[redacted]

* Stroke 2001; 32: 1847-54



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Frumkvöðlar

- 1952 Robert Edward Gross
  - Atrial well “Open heart surgery” ASD 0/3
- 1952 Floyd John Lewis
  - Hypothermia “Open heart surgery” ASD 9/11
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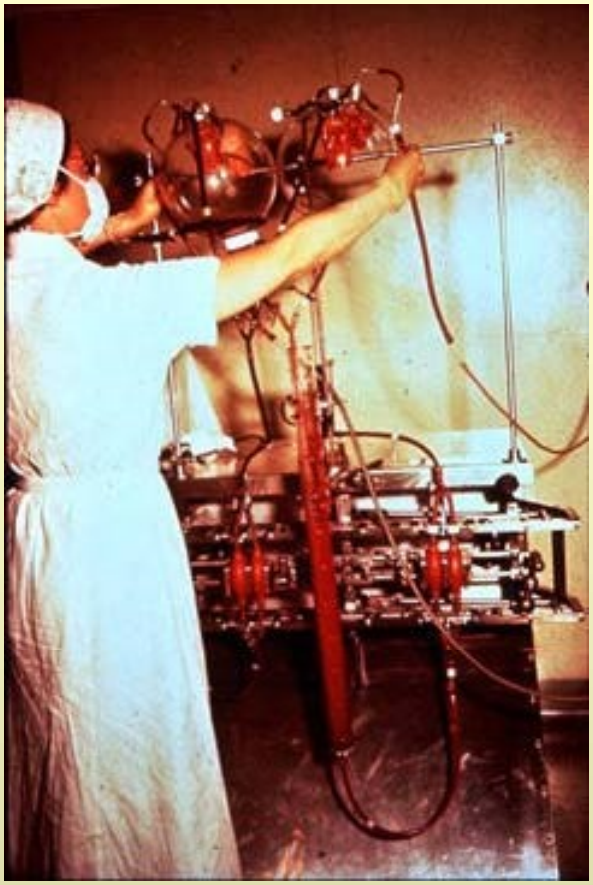
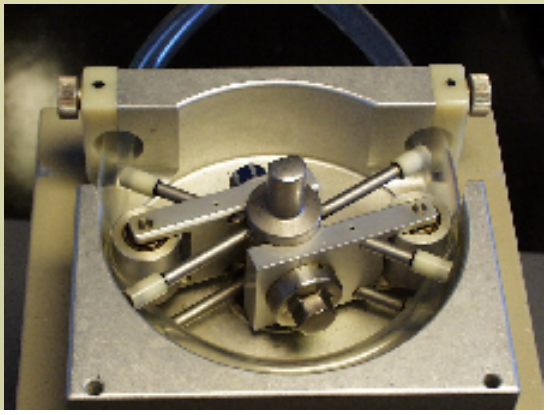
  - CPB support ASD 1/5
- 1954 Dr. Walt Lillehei
  - Cross circulation VSD
  - Cross circulation Tetralogy of Fallot

The first father/son cross circulation
Lyman and Gregory Glidden
for a ventricular septal defect

Howard Holtz cross-circulation donor for Mark Shaw

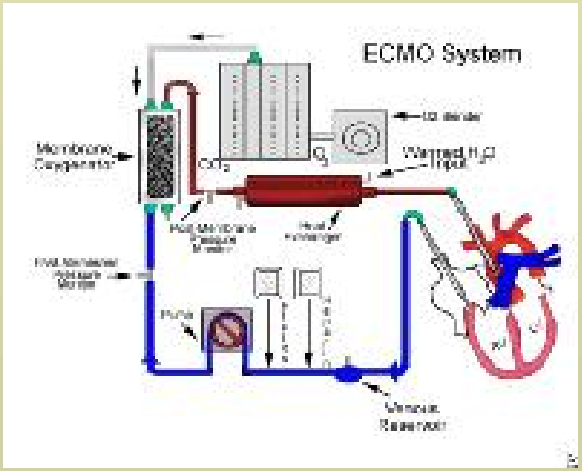
- CPB
  - svæfingalækningar
  - Blóðflokkar
  - EKG 1903
  - Positive pressure ventilation
  - Rtg.1895 Contrast 1923
  - Hjartaþræðing
  - Heparin 1915
  - Protamin
  - Oral anticoagulants 1940
  - Roller pump





- 1955 John W. Kirklin
  - teamwork
- “Cardiac team”
  - Cardiac Surgery
  - Anesthesiology assoc. 1972
  - Perfusion technology
  - Cardiac intensive care medicin

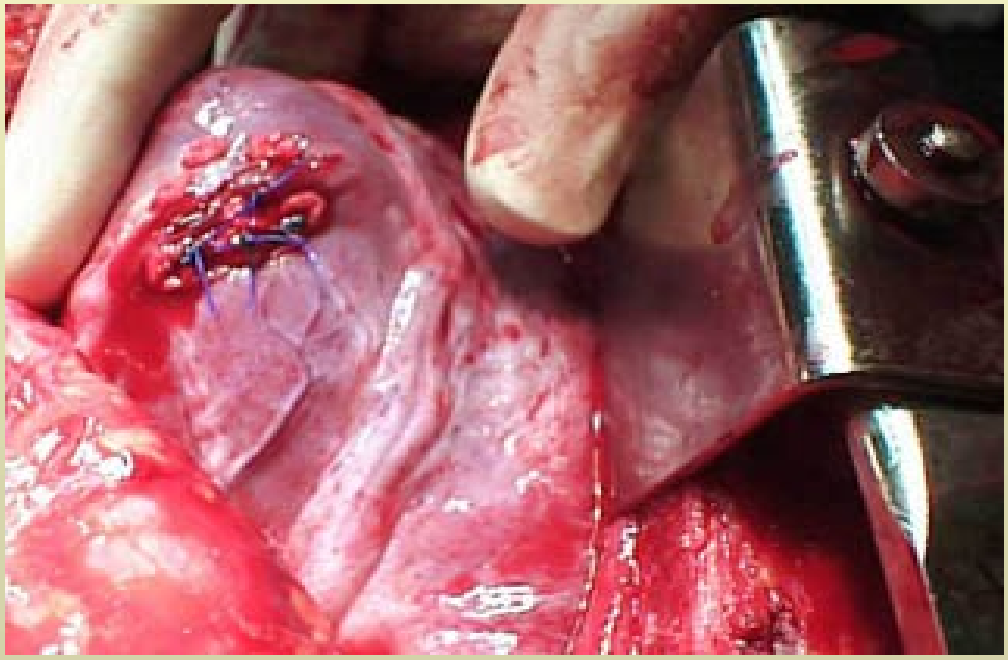
- 1955 Dr. DeWall Bubble oxygenator
- Nszih Zuhdi Hemodilution techniques
- 1960 combined pump and hypothermia
  - Ca. 1.000.000 / Y / US
    - CABG,VALVES, CONG.
    - NEUROSURG..
    - HEART/LUNG TRANSPL.,
    - THORAC.AORTA
    - ECMO



1883

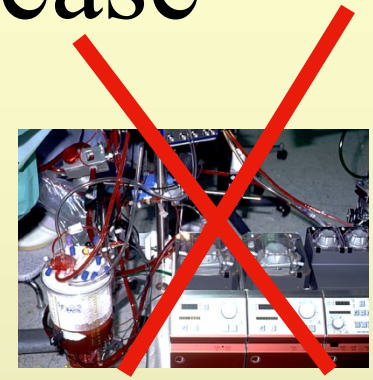
Theodore Bilroth

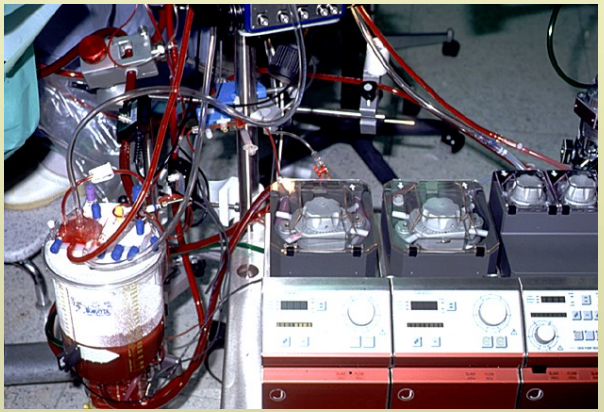
' *The Surgeon who should attempt to suture a wound of the heart would lose the respect of his colleagues.*



Congenital Disease

- Patent Ductus Arteriosus
- Aortic Coarctation
- Atrial Septal Defect
- Ventricular Septal Defect
- Valvular Heart Disease
  - Ross Procedure





Open Heart Surgery

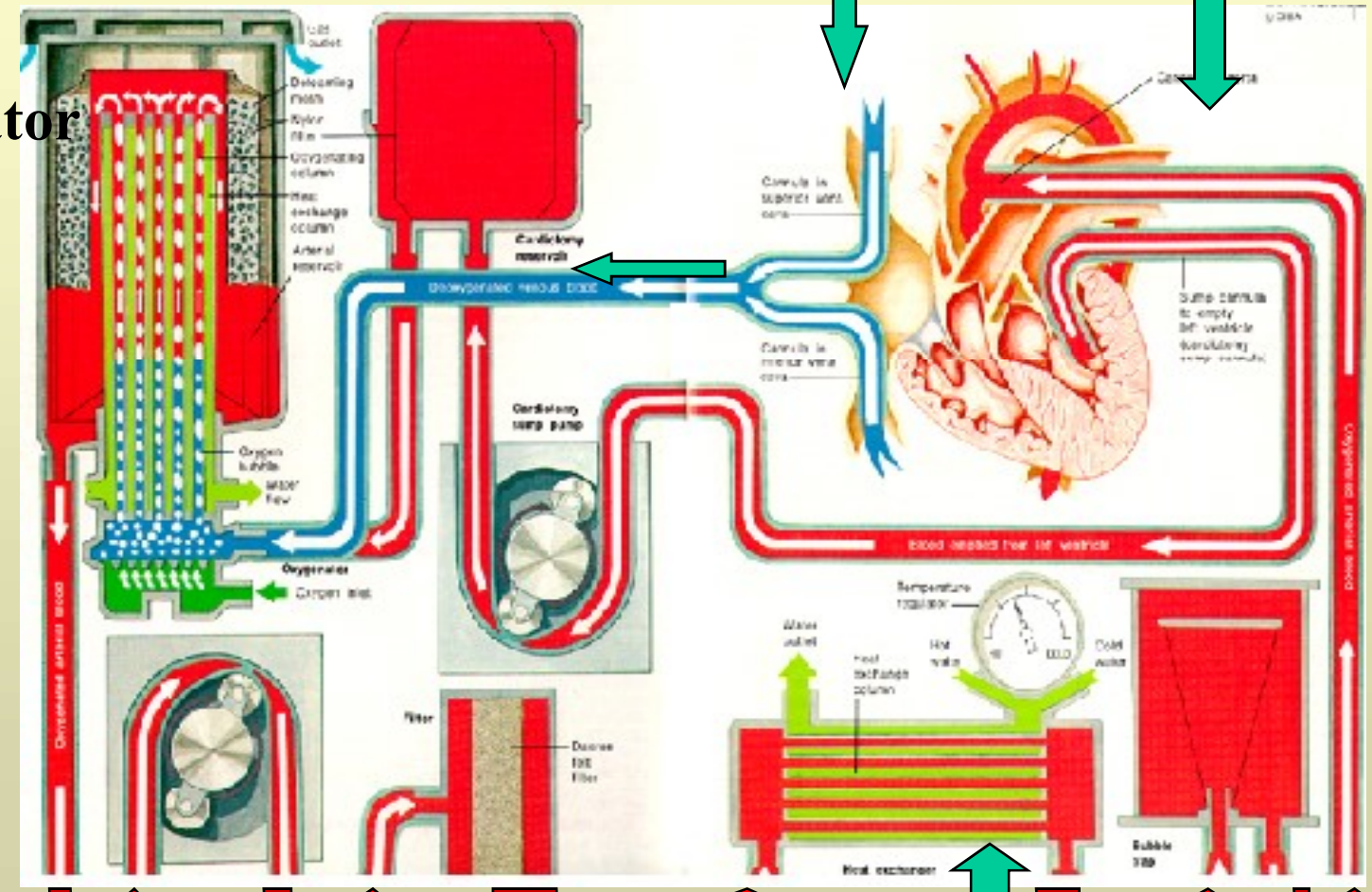
- Management of chest /thoracotomy tubes
- Risk for Infection
- Risk for Hypo/Hyperthermia
- Risk for Fluid Volume Overload
- Complications
  - Hemmorhage
  - Shock
  - Heart Block
  - CHF
  - Post Cardiac Surgery Syndrome
  - Post Perfusion Syndrome
  - Infection
  - Atelectasis

ECC :

Venous (IVC and SVC) Cannulae

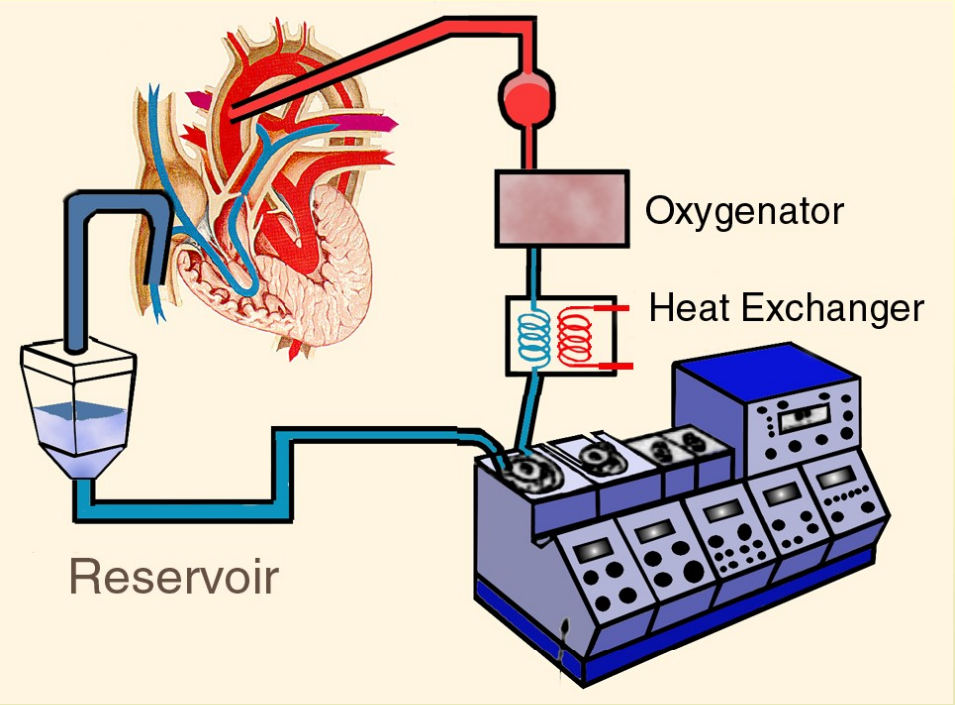
Aortic Cannula

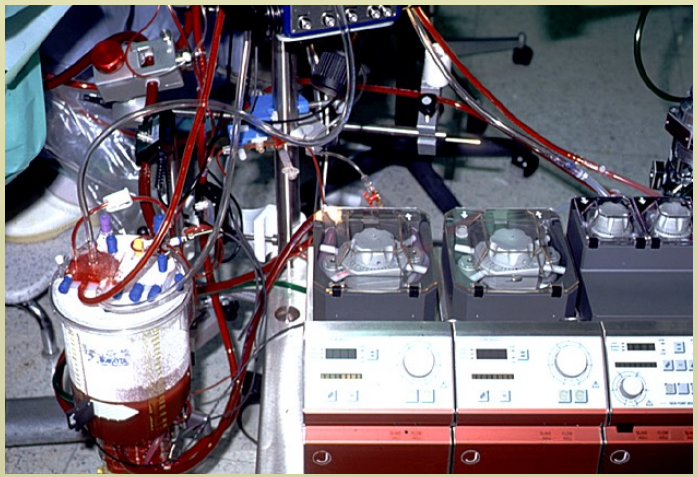
Oxygenator



Pump

Heater/Cooler

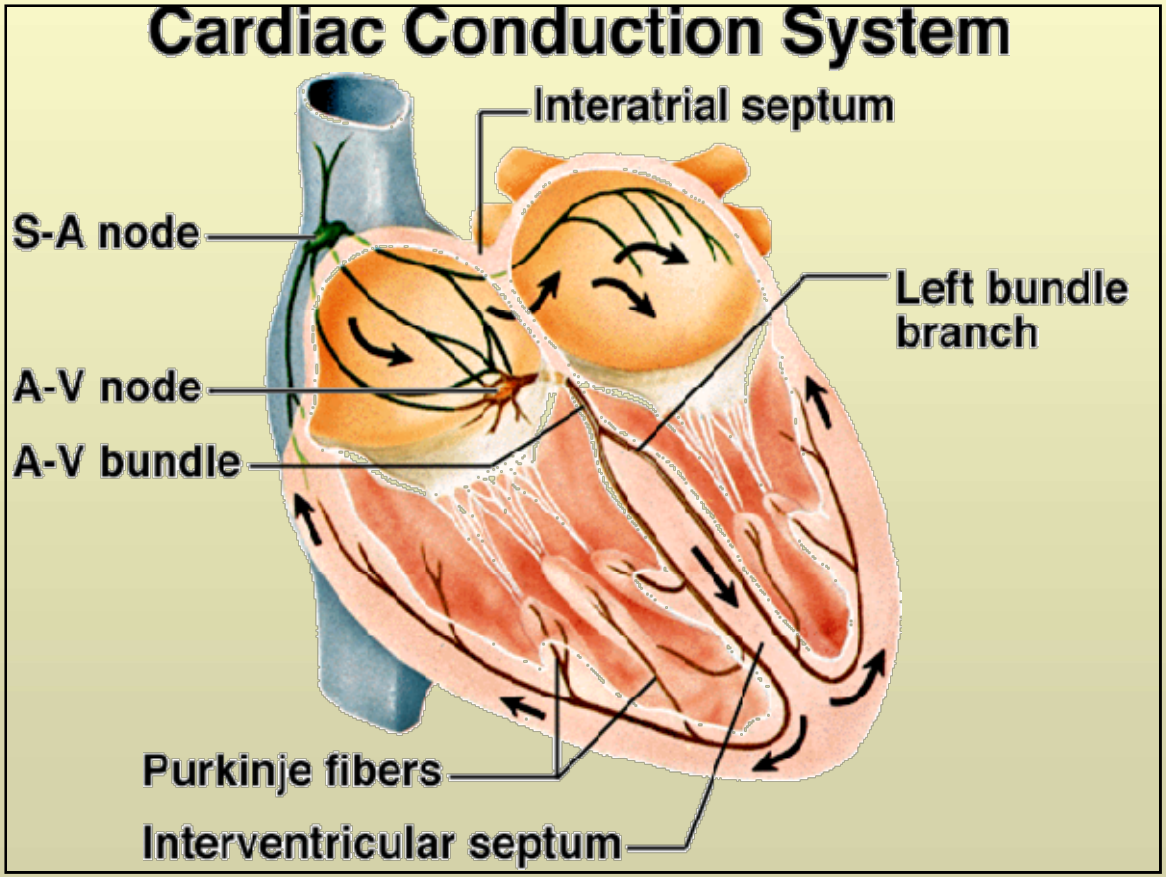

Oxygenator
Heat Exchanger
Reservoir



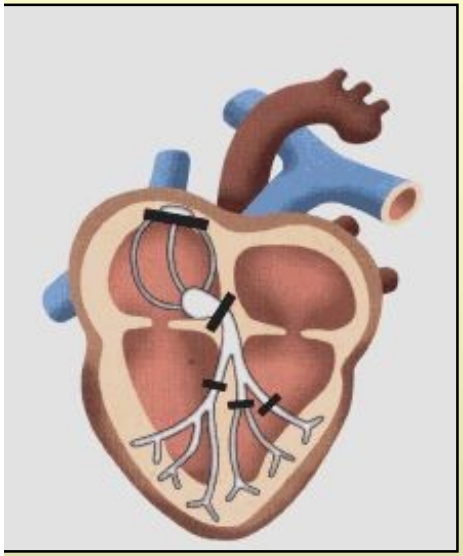
CardioPulmonary Bypass
Hjartalungnavél

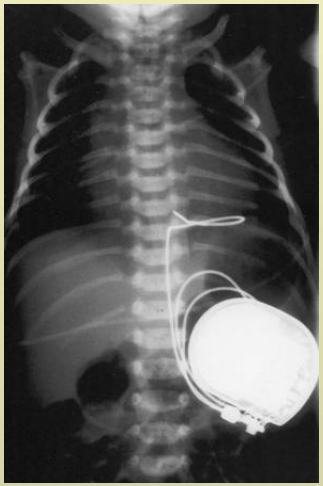
Quantum Spectrum/ Medtronic

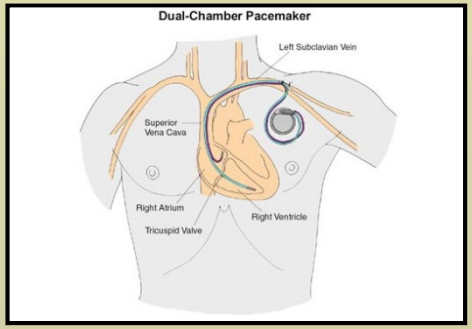
Samhæfing


Cardiac Conduction System
Interatrial septum
S-A node
Left bundle branch
A-V node
A-V bundle
Purkinje fibers
Interventricular septum

Paceaker Case #13





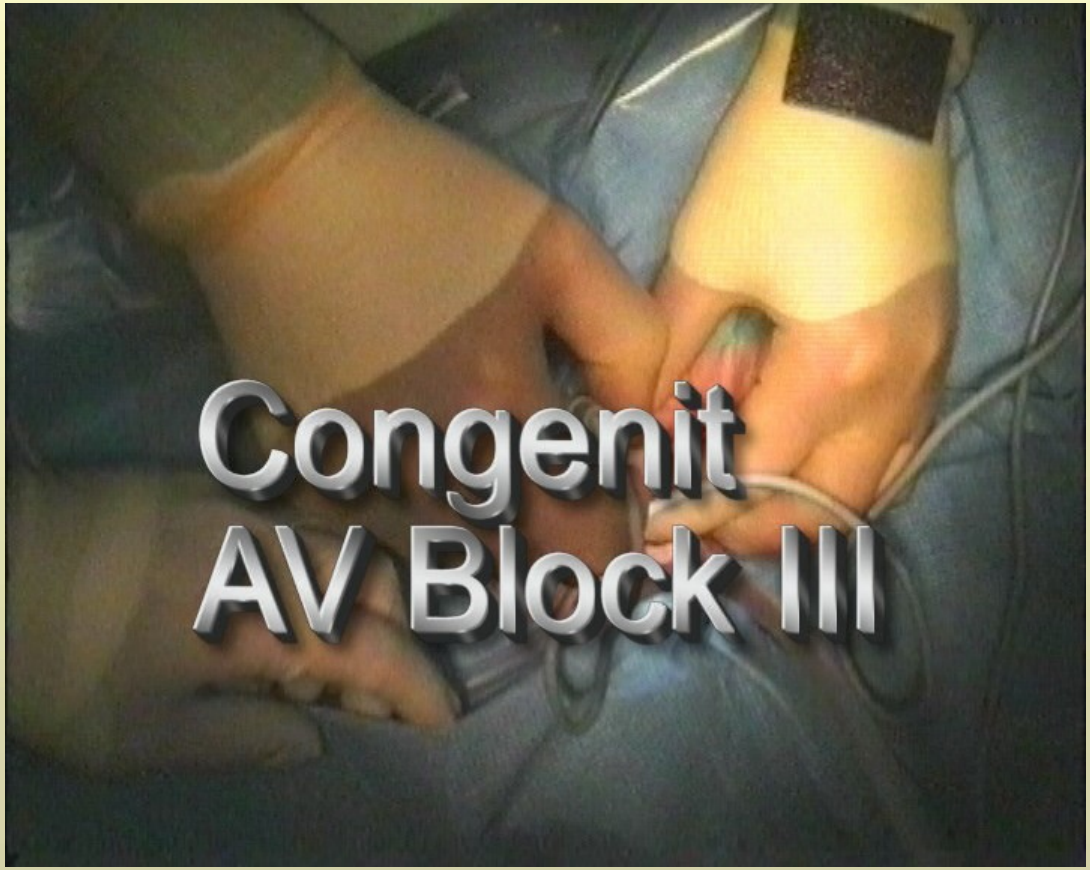


Indian Pacing Electrophysiol. J. 2003;3(1):23

Video: Pacemaker

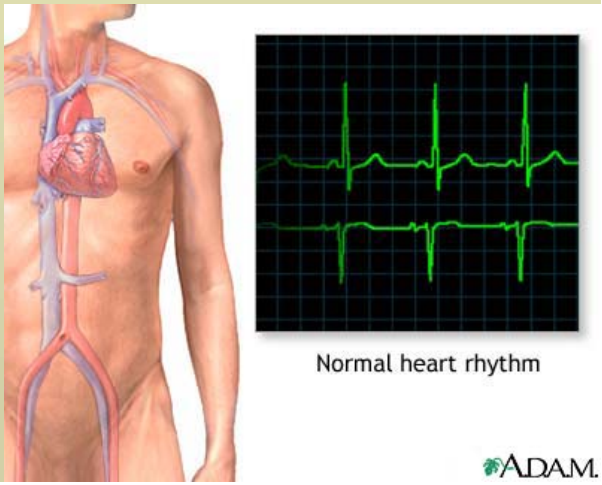
1:20

Video



Pacemaker

- Usually placed sub-xyphoid or mid to lower abd
- Teach parents how to take child's pulse daily
- Batteries can last up to15 years



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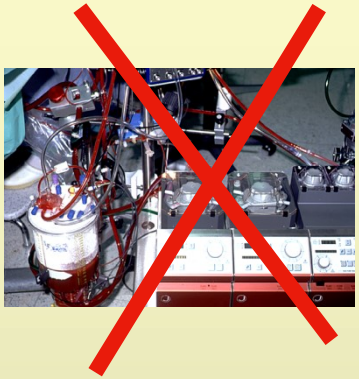
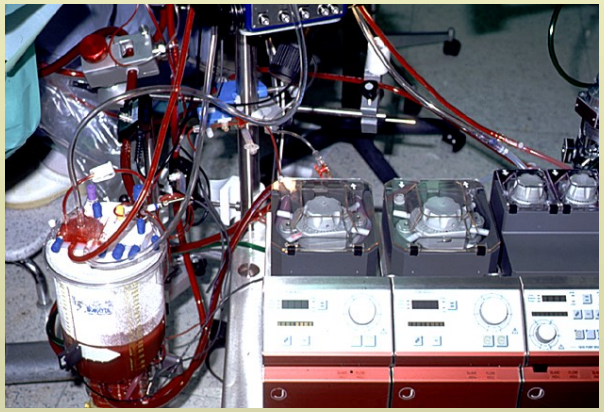
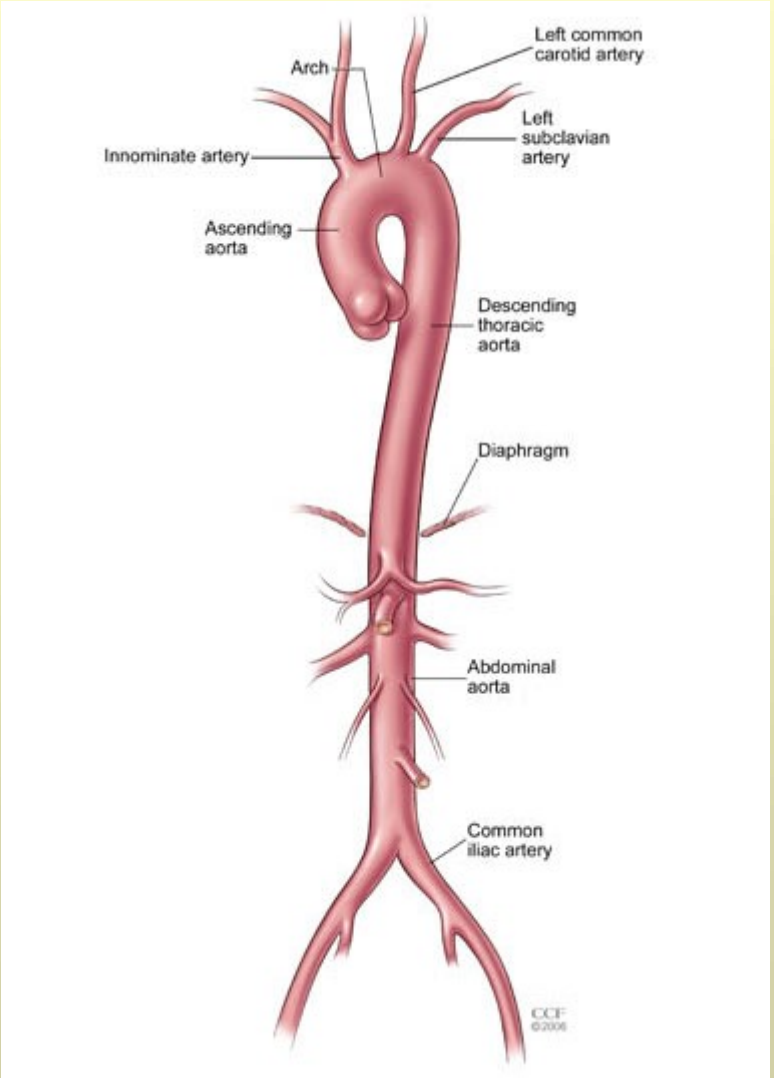




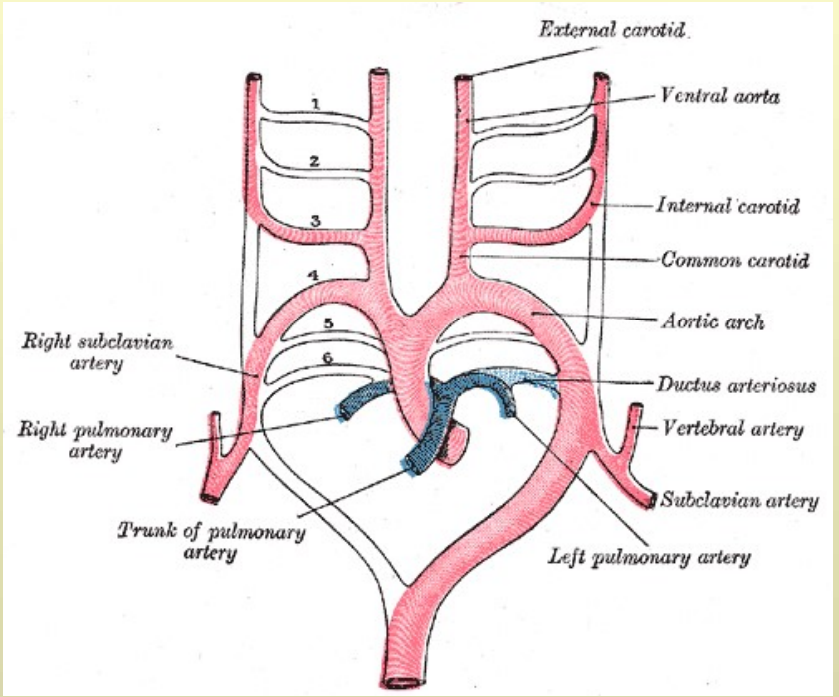
Skipting ósæðar

- Ósæðarrót
- Rishluti
- Ósæðarbogi
- Fallhluti



Fósturfræði

- Pharyngeal arches
- Arch 4
  - Hægri
    - Right subclavian
  - Vinstri
    - Arcus
- Dorsal aorta
  - Descending

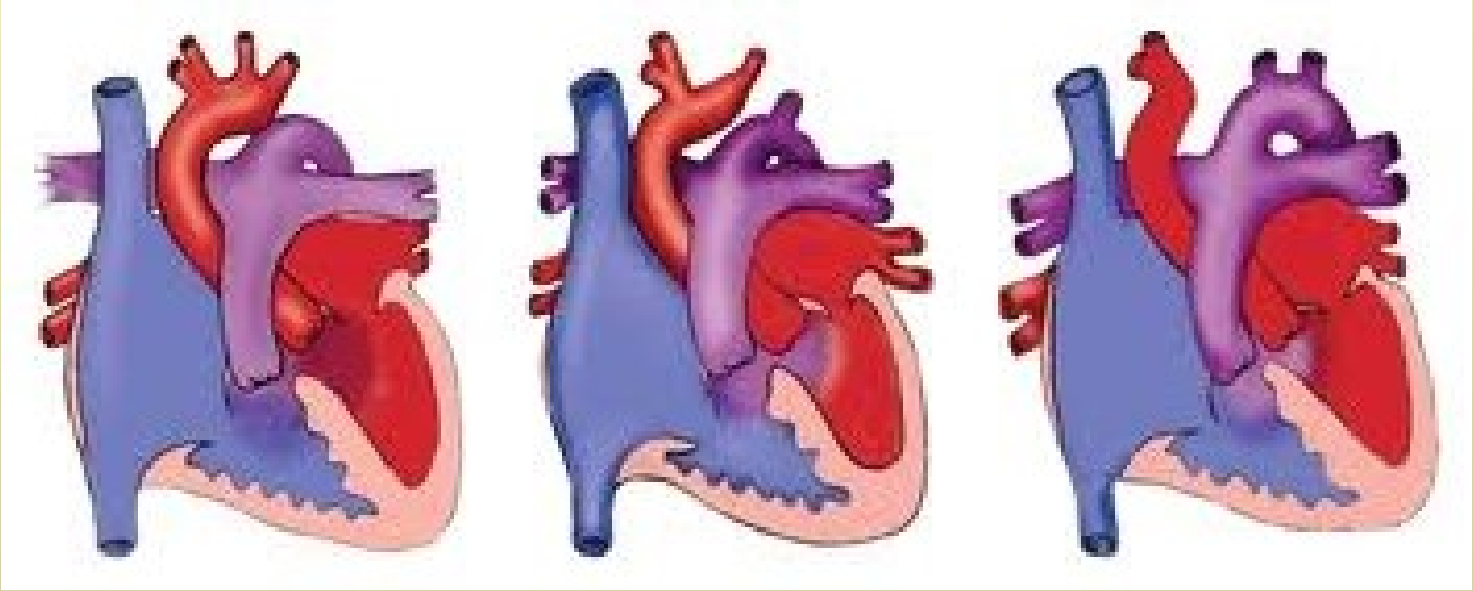


Congenital aortic syndrome

- Aortic Coarctation
- Interrupted Aortic Arch
- Vascular rings and slings

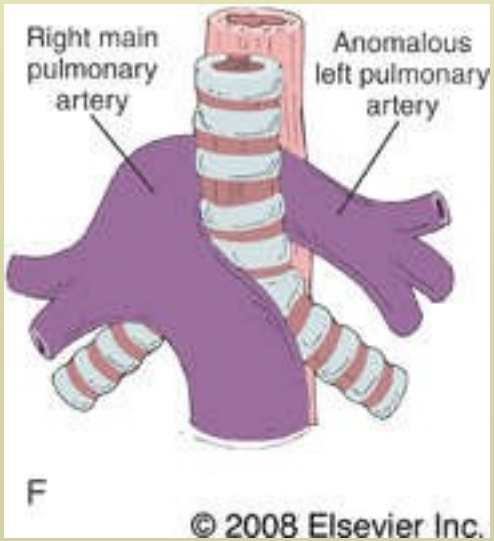
Interrupted Aortic Arch

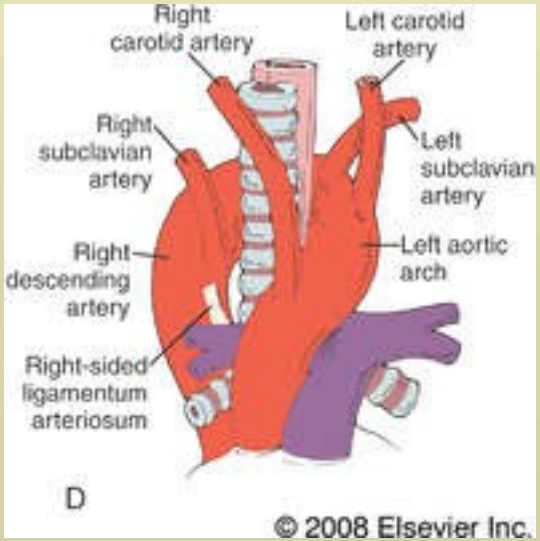
- Nær alltaf tengsl við
  - ventricular septal defect
  - Patent ductus arteriosus

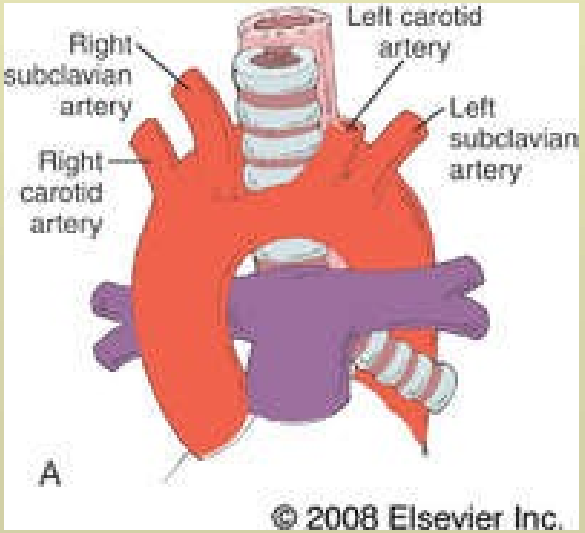


Vascular rings and slings

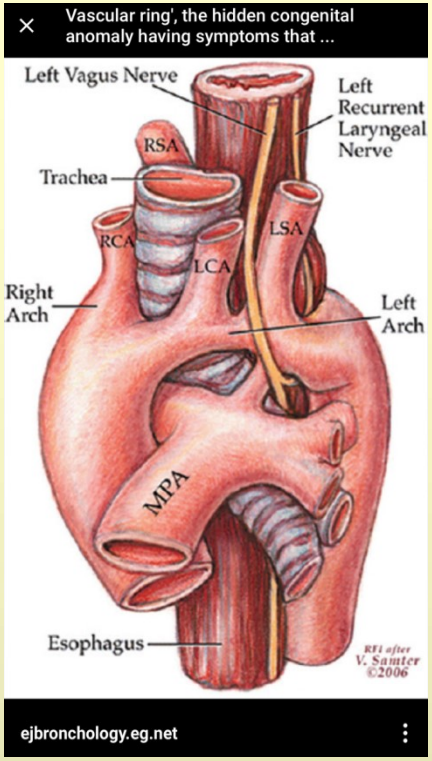
- Anomaliur í greinum aortic arch eða pulmonary artery
- Local þrenging á
  - Trachea
  - Esophagus

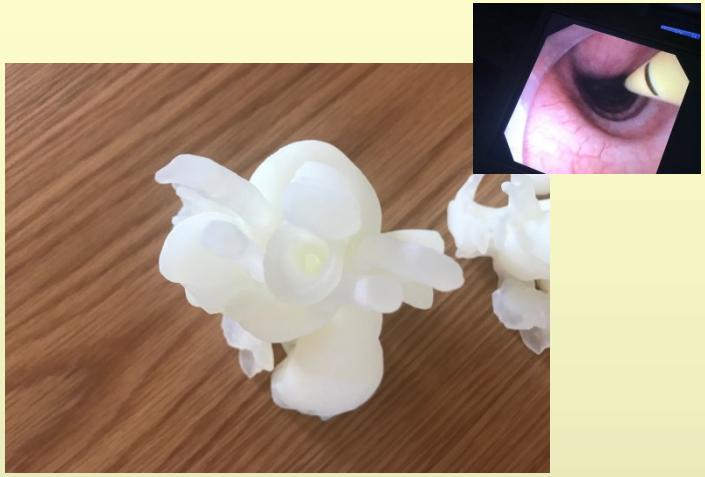


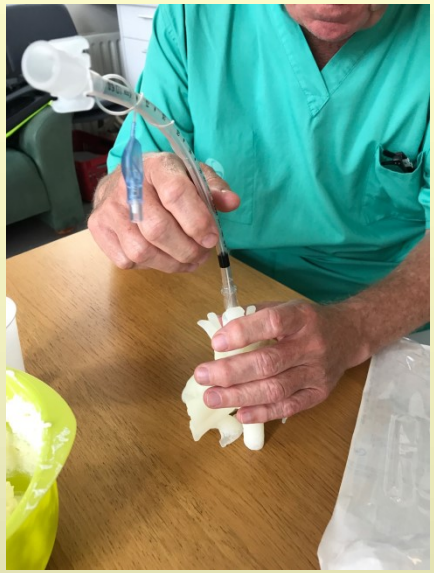


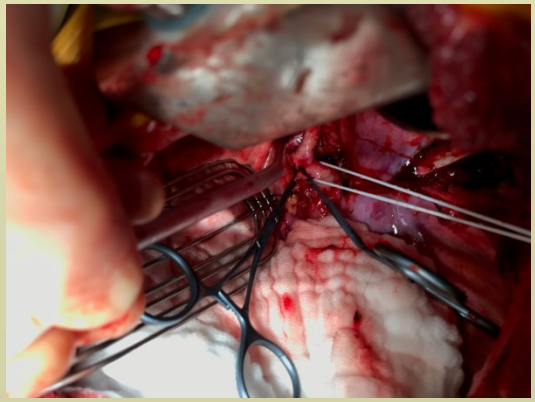


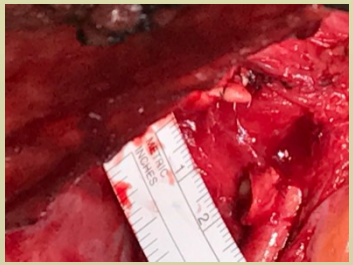
Case # 14


Vascular ring', the hidden congenital anomaly having symptoms that ...
Left Vagus Nerve
Left Recurrent Laryngeal Nerve
RSA
Trachea
RCA
LCA
LSA
Right Arch
Left Arch
MPA
Esophagus
RS after V. Samter ©2006
ejbronchology.eg.net







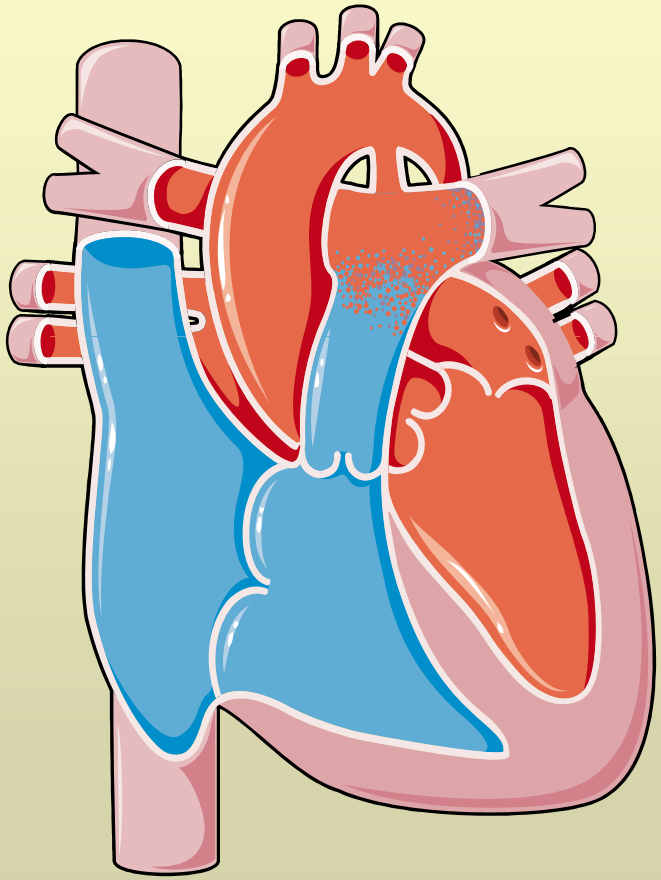


Patent Ductus Arteriosus

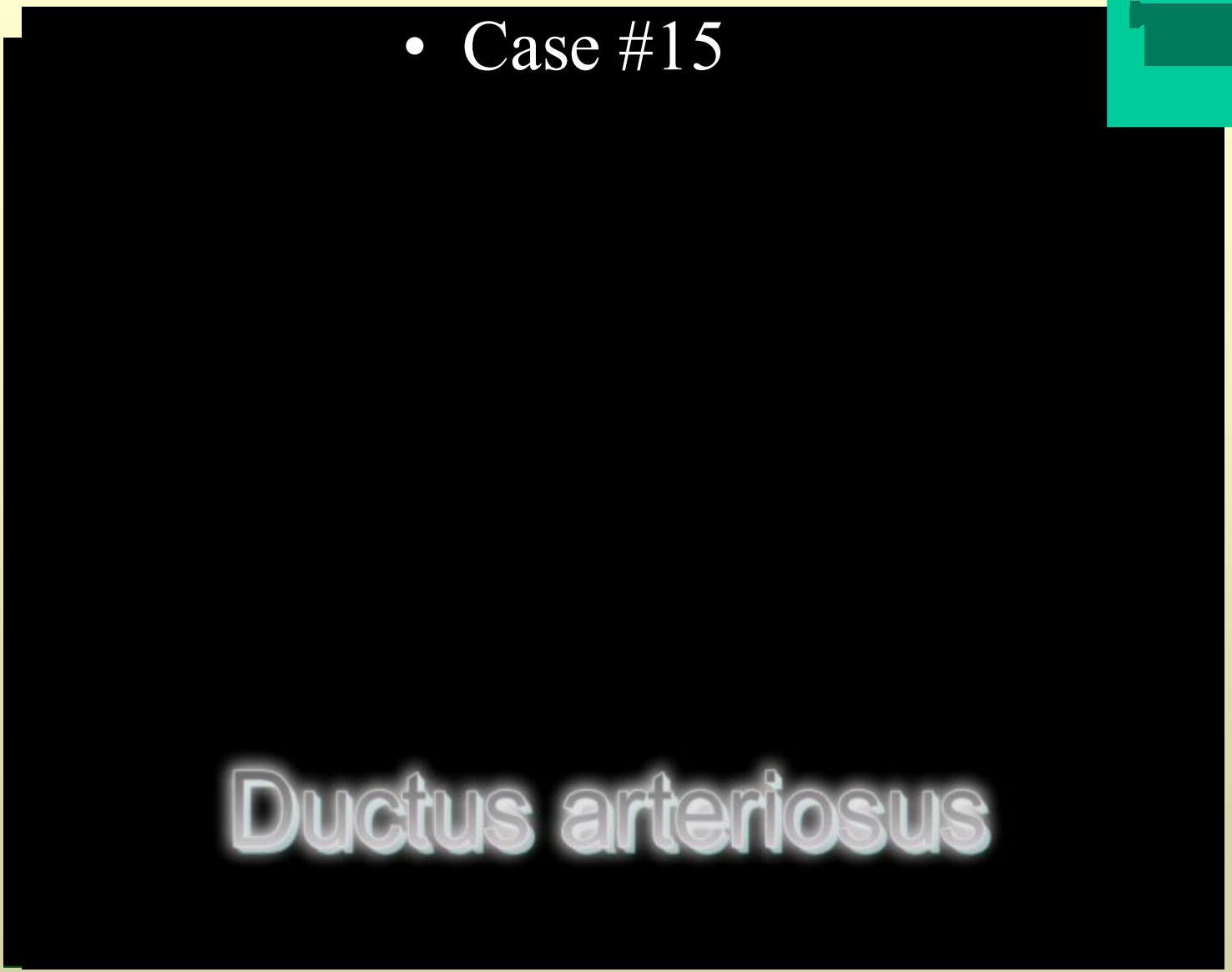
- Small defect no symptoms.
- Large defect:
  - Wide pulse pressure
  - Enlarged heart
  - Thrill in L second IS
  - Continuous murmur
  - X-ray: prominent pulmonary artery with increased vascular markings.

Patent Ductus Arteriosus
Acyanotic Heart Defect

- PDA
  - Incidence 5-10% CHD
  - Hemodynamic
  - Diagnosis
  - Management
    - Med
    - **Surgical** Mort <1%



- Case #15

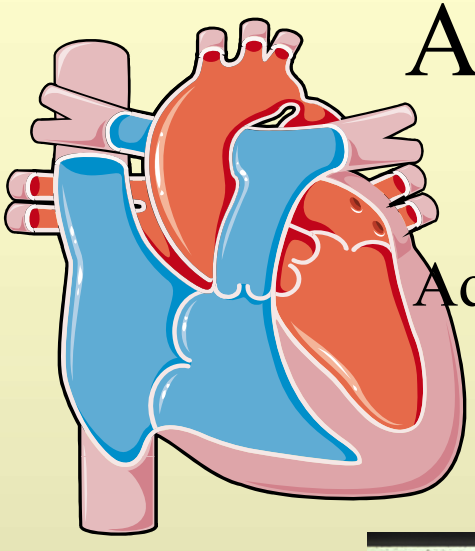


Video: Ductus 2:43

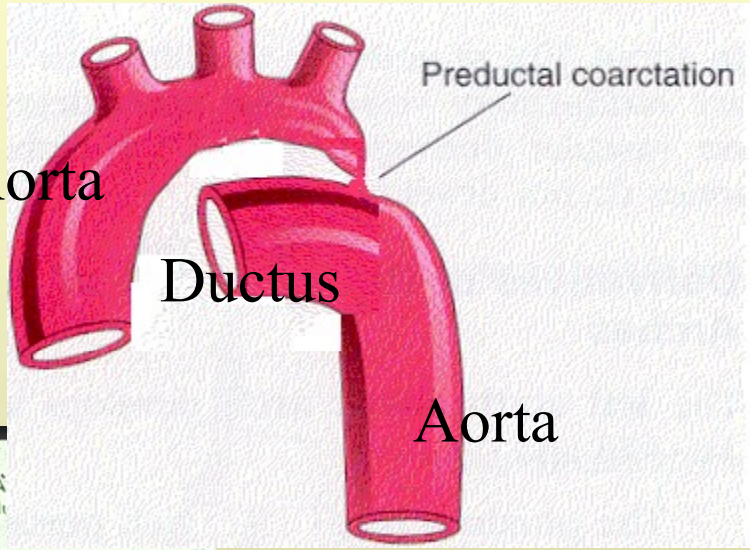
Aortic Coarctation

- Tíðni 40/100,000
- Þrengsli í descending aorta
- Á mörkum þar sem ductus arteriosus tengist aortu
- Börnum
  - Hjartabilun
- Unglingar og fullorðnir
  - Háþrýstingur
  - Heilablæðing

Aortic Coarctation
Acyanotic Heart Defect



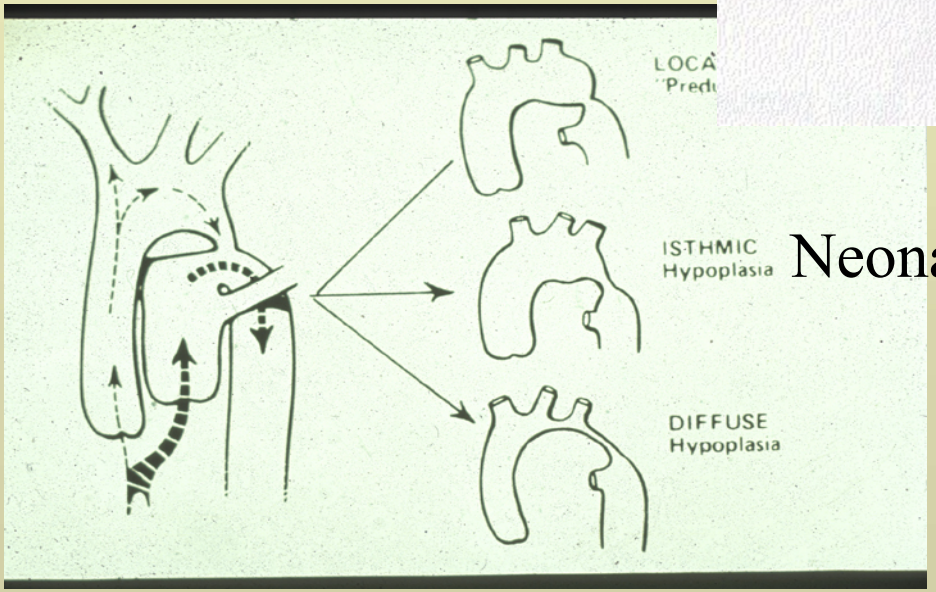
Adult Týpa



Aorta

Ductus

Aorta



Neonatal Týpa

Coarctation of Aorta

- COA
- 7 % of defects
- Congenital narrowing of the descending aorta
- 80% have aortic-valve anomalies (Bicusp)
- Difference in BP in arms and legs (severe obstruction)

Diagnosis and Treatment

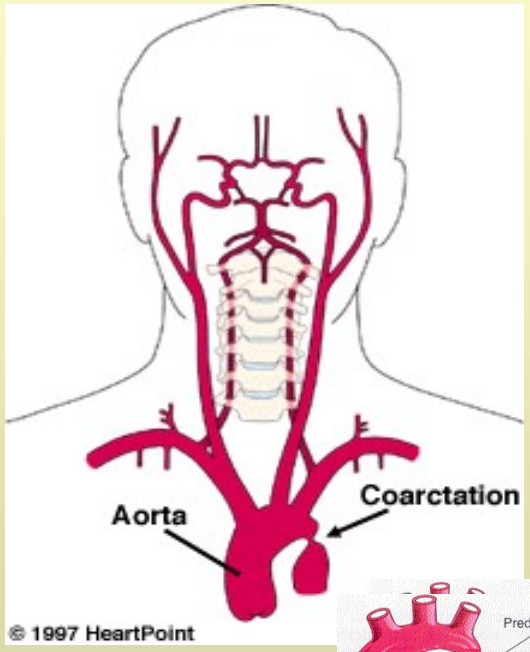
- In 50% the narrowing is not severe enough to cause symptoms in the first days of life.
- When the PDA closes a higher resistance develops and heart failure can develop.
- Pulses in the groin and leg will be diminished
- Echocardiogram will show the defect in the aorta

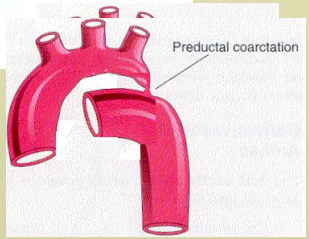
Treatment

- Prostaglandin may be given to keep the PDA open to reduce the pressure changes
- The most common repair is resection of the narrowed area with re-anastomosis of the two ends
- Surgical complications – kidney/spinal damage due to clamping off of blood flow during surgery
- High blood pressure post surgery – may need to be on antihypertensives
- Antibiotic prophylactic need due to possible aortic valve abnormalities.

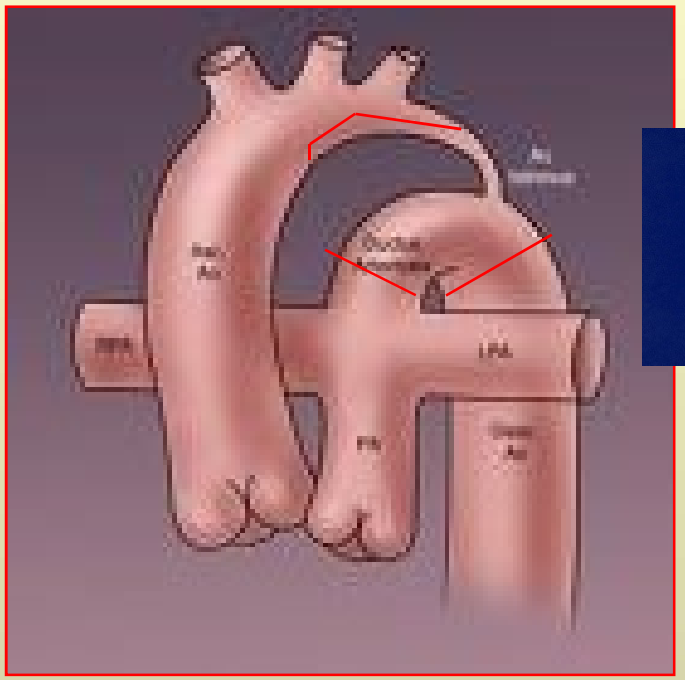
Coarctation of the Aorta
Acyanotic Heart Defect

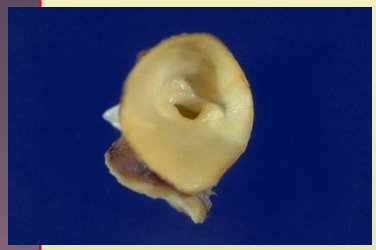
- Incidence 8% CHD
- Hemodynamic
- Diagnosis
- Management
  - Med
  - **Surgical** Mort <5%
    - » Ath mun á nýbura- og fullorðinspresentation

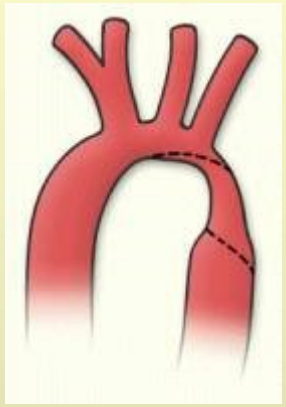


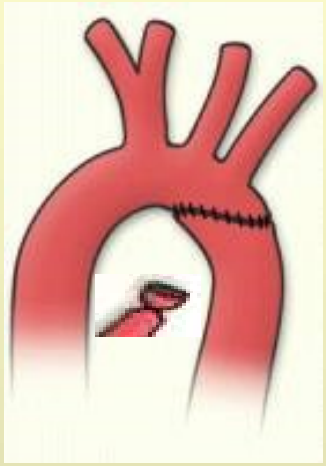


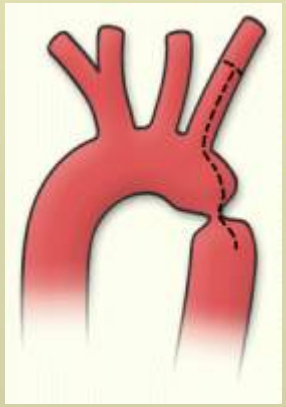
Extended end to end eða subclavian flap reconstruction

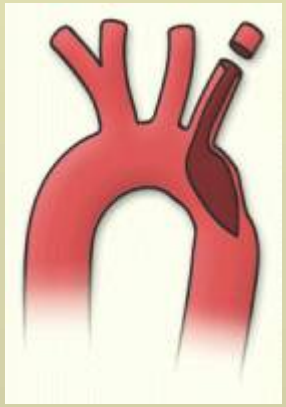


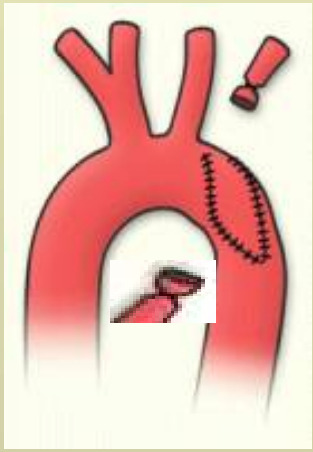




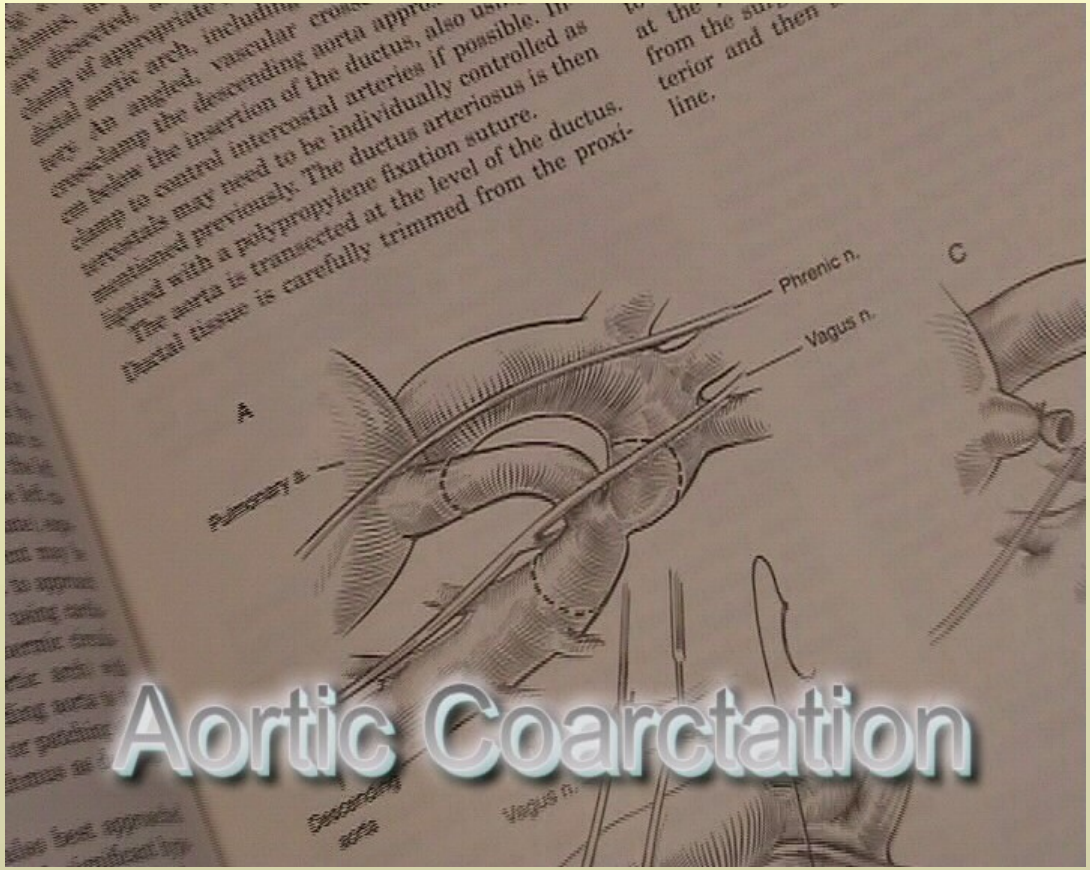








Aortic Coarctation Case #16

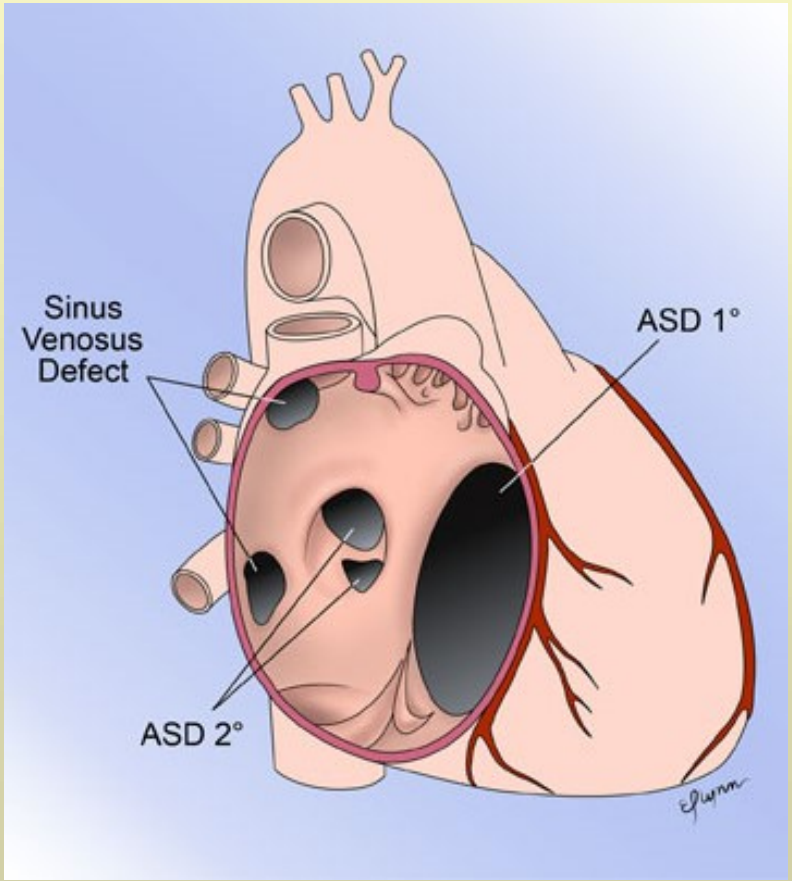


◆Video 2

4:51

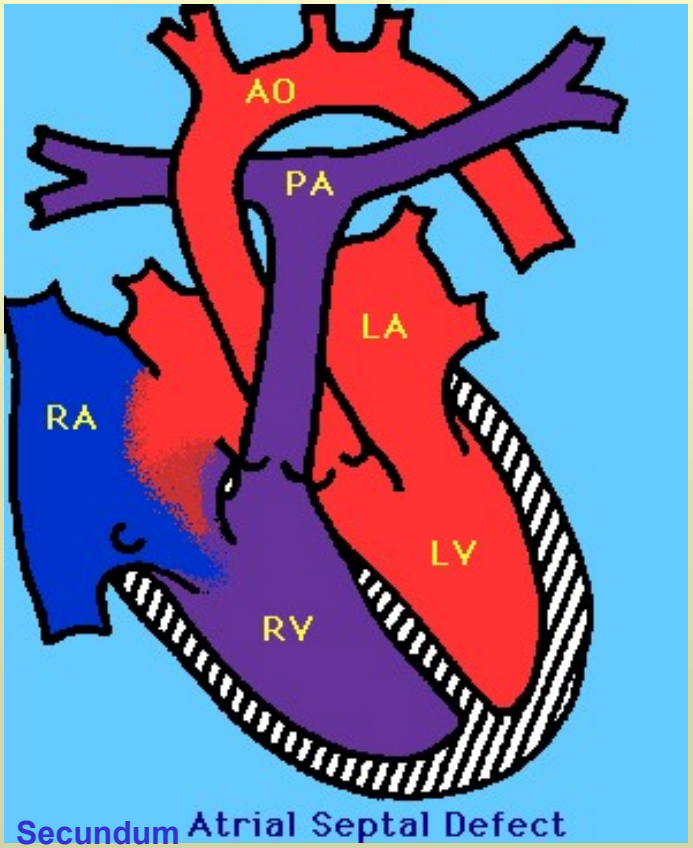
Atrial Septal Defects
Acyanotic Heart Defects

- 3 Types of ASD:



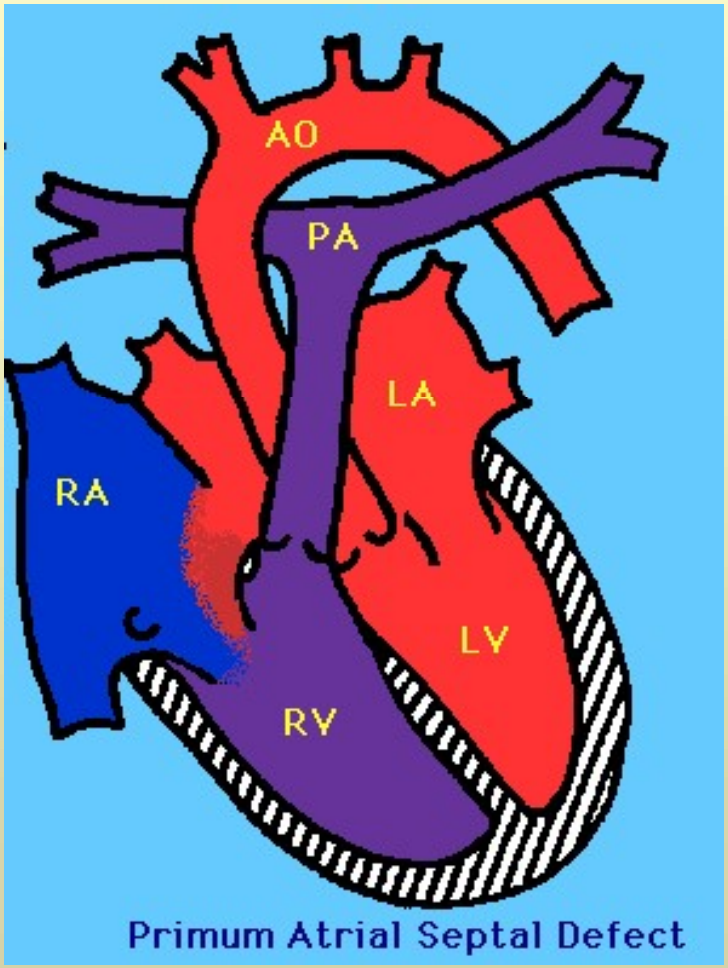
Atrial Septal Defects: secundum

- Most common form of ASD (fossa ovalis)
- In large defects, a considerable shunt of oxygenated blood flows from the left to the right atrium.
- Mostly asymptomatic
- The 2nd heart sound is characteristically widely split and fixed.



Atrial Septal Defects:primum

- Situated in the lower portion of the atrial septum and overlies the mitral and tricuspid valves. In most instances, a cleft in the anterior leaflet of the mitral valve is also noted.
- Combination of a left-to-right shunt across the atrial defect and mitral insufficiency
- C/F similar to that of an ostium secundum ASD



Atrial Septal Defect

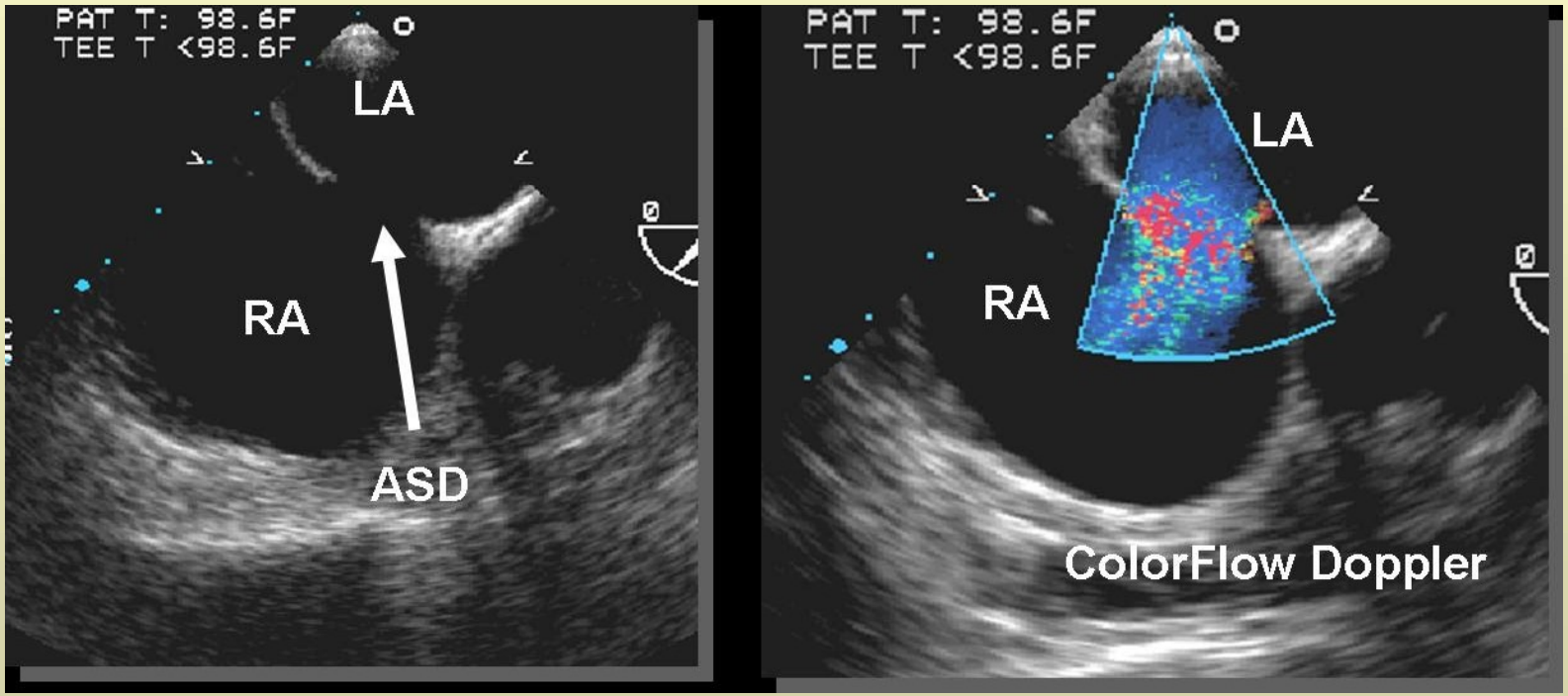
- ASD
- 6-10% of defects
- Blood in left atrium flows into right atrium
- Pulmonary hypertension
- Reduced blood volume in systemic circulation
- If left untreated may lead to pulmonary hypertension, congestive heart failure or stroke as an adult.

Treatment

- Surgical closure of the atrial septal defect
- After closure in childhood the heart size will return to normal over a period of four to six months.
- No restrictions to physical activity post closure

Atrial Septal Defect

- Enlargement of the right ventricle
- Enlargement of atrium
- Large pulmonary artery
- increased pulmonary vascularity


PAT T: 98.6F
TEE T <98.6F
LA
RA
ASD
PAT T: 98.6F
TEE T <98.6F
LA
RA
ColorFlow Doppler

Atrial Septal Defects

- Secundum ASDs are well tolerated during childhood.
- Antibiotic prophylaxis for isolated secundum ASDs is not recommended.
- Surgery closure is advised for all symptomatic patients and also for asymptomatic patients with a Qp:Qs ratio of at least 2:1.
- Secundum defects are approached surgically but some secundum defects can be closed by transcatheter device
- Ostium primum defects are approached surgically
- Sinus venosum defects are approached surgically

Atrial Septal Defect Case #17

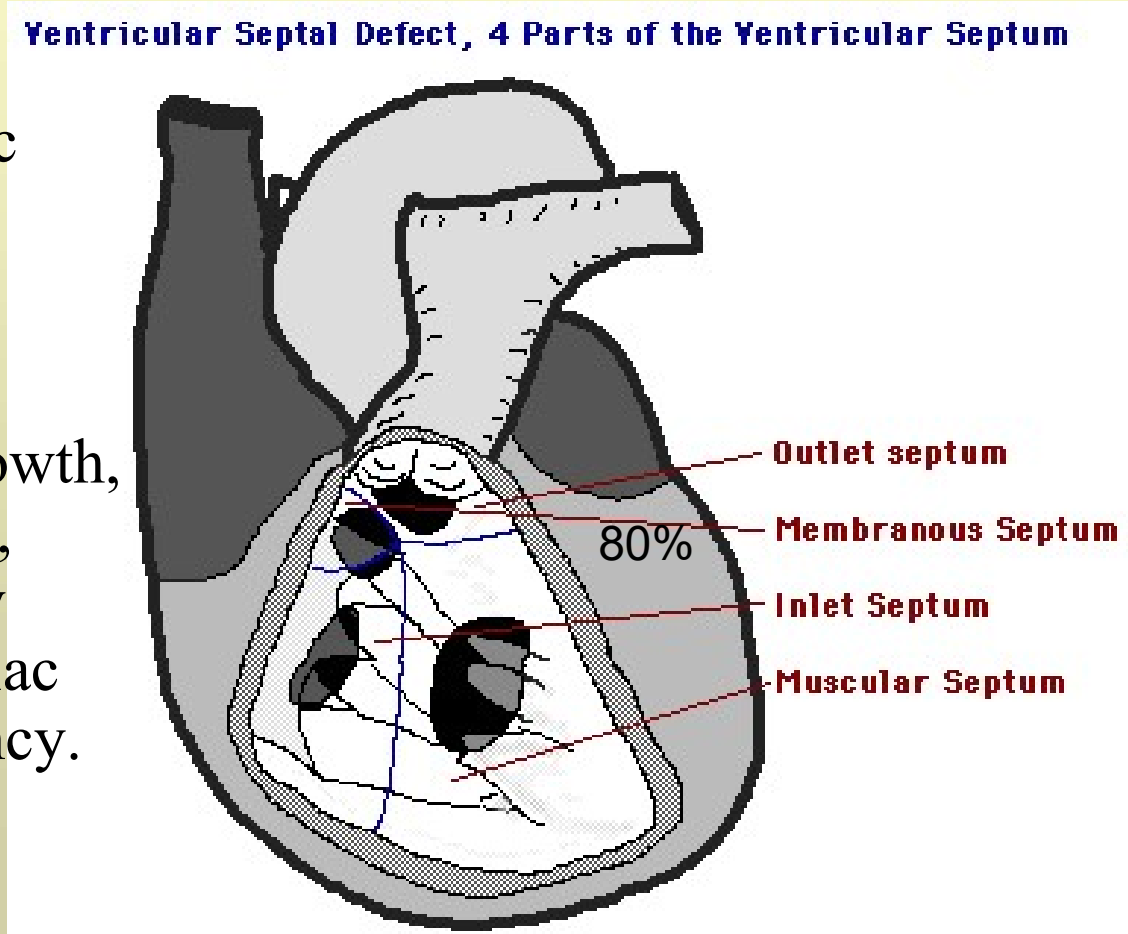
ASDHjartagallar videoVASD.AVI

Video:

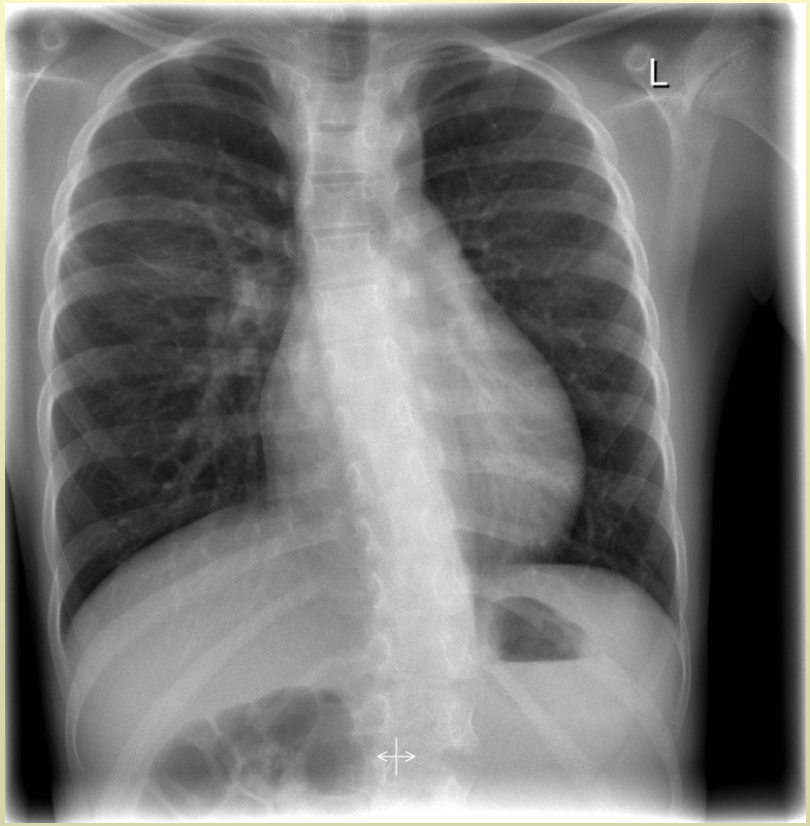
6:44

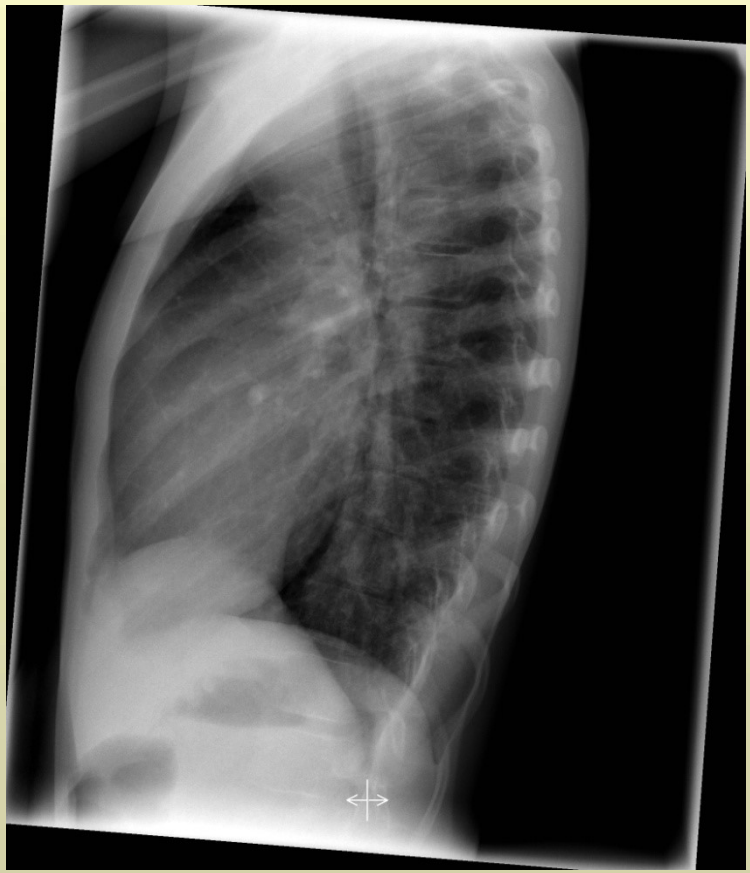
Ventricular septal Defects

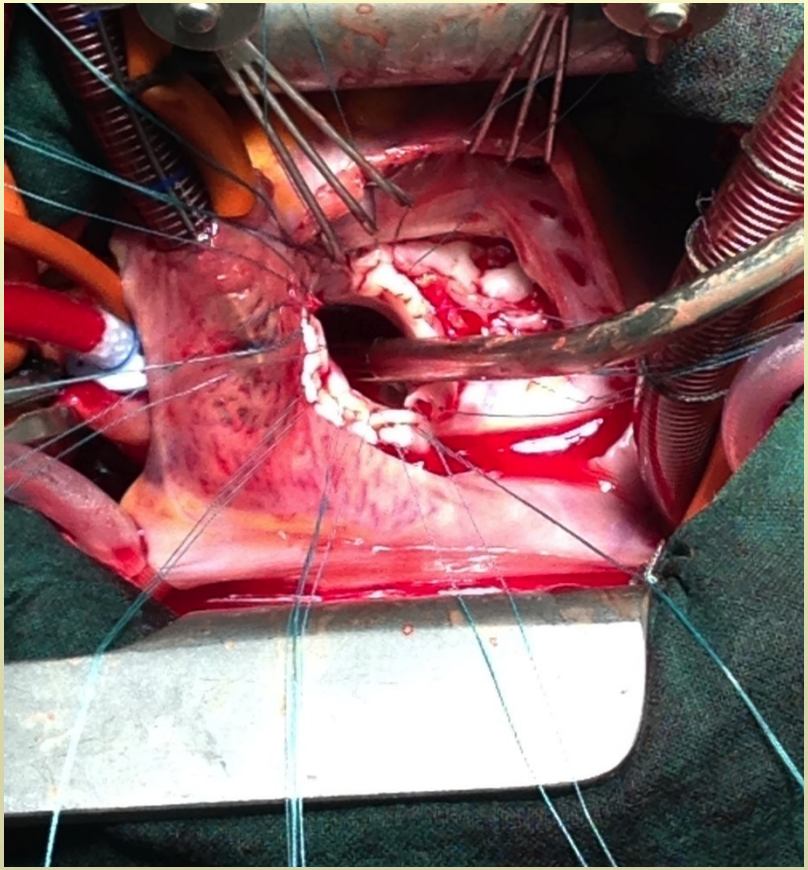
- **Small VSD**
  - Asymptomatic
  - A loud, harsh, or blowing holosystolic murmur.
- **Large VSD**
  - dyspnea, feeding difficulties, poor growth, profuse perspiration, recurrent pulmonary infections, and cardiac failure in early infancy.

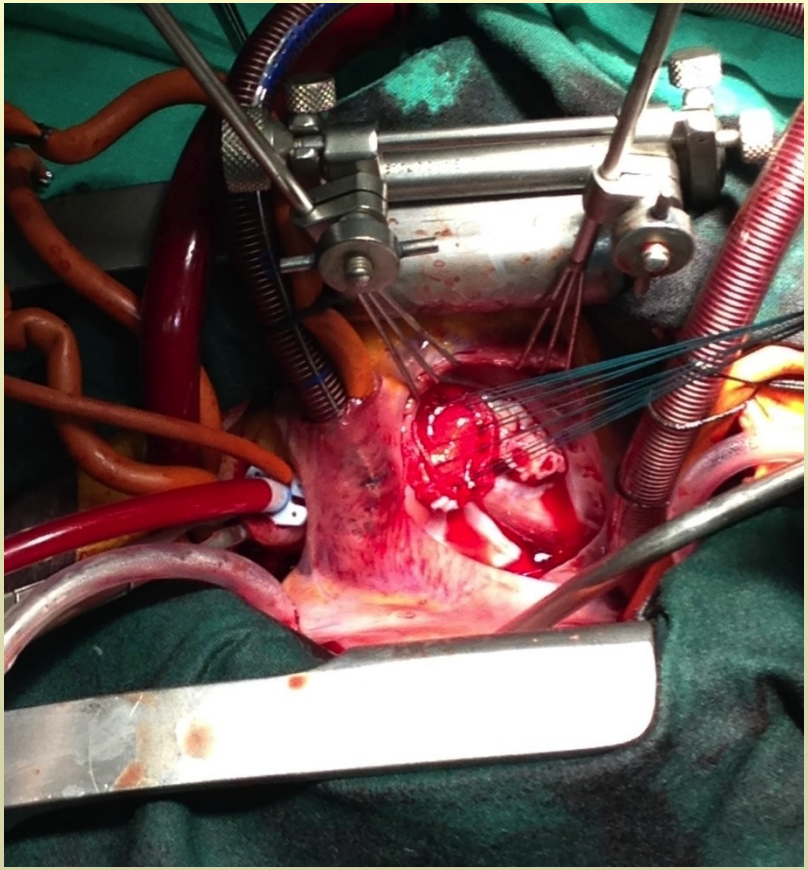


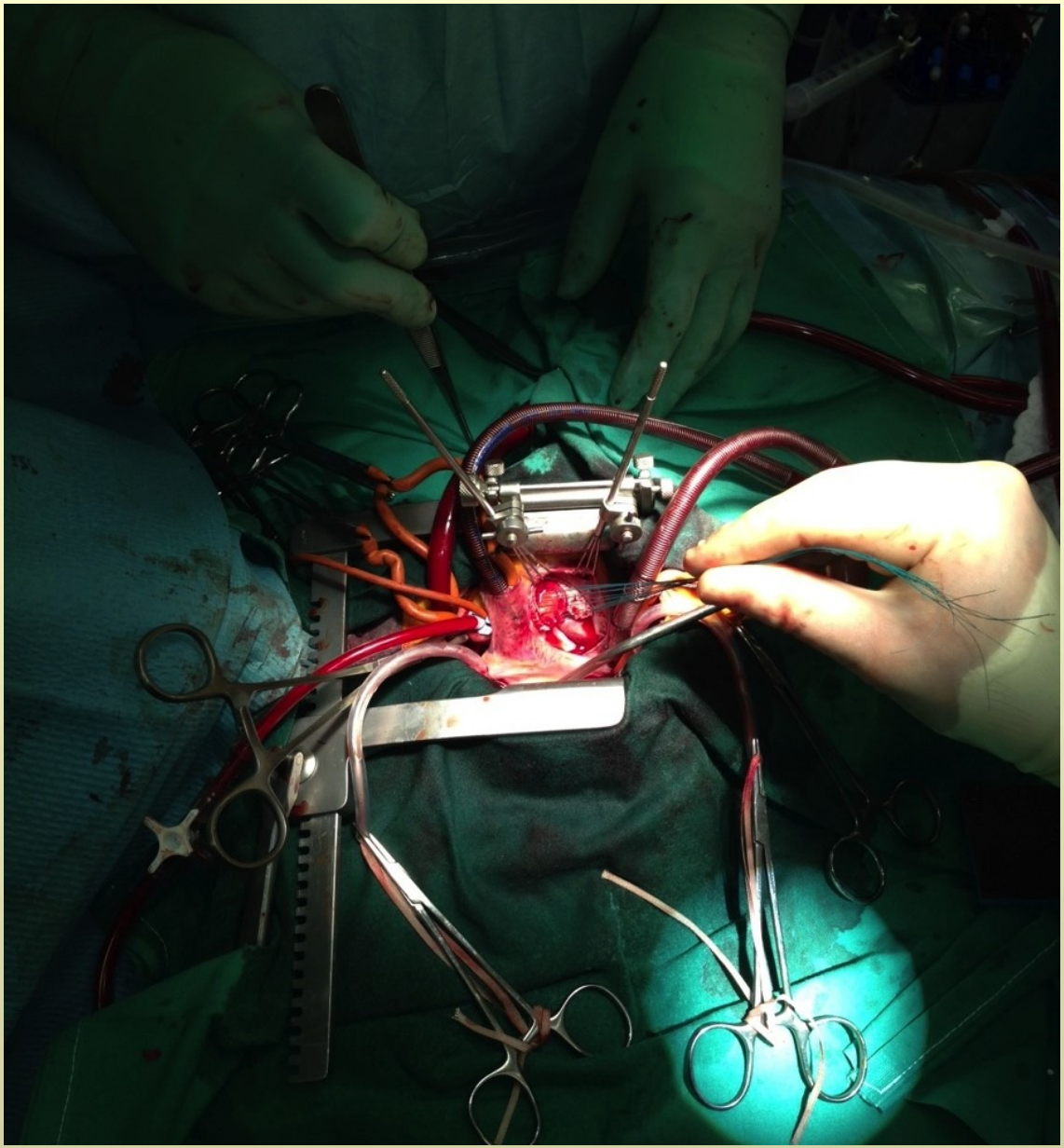
Case # 18












V
GG KL-15,50


Ventricular Septal Defect

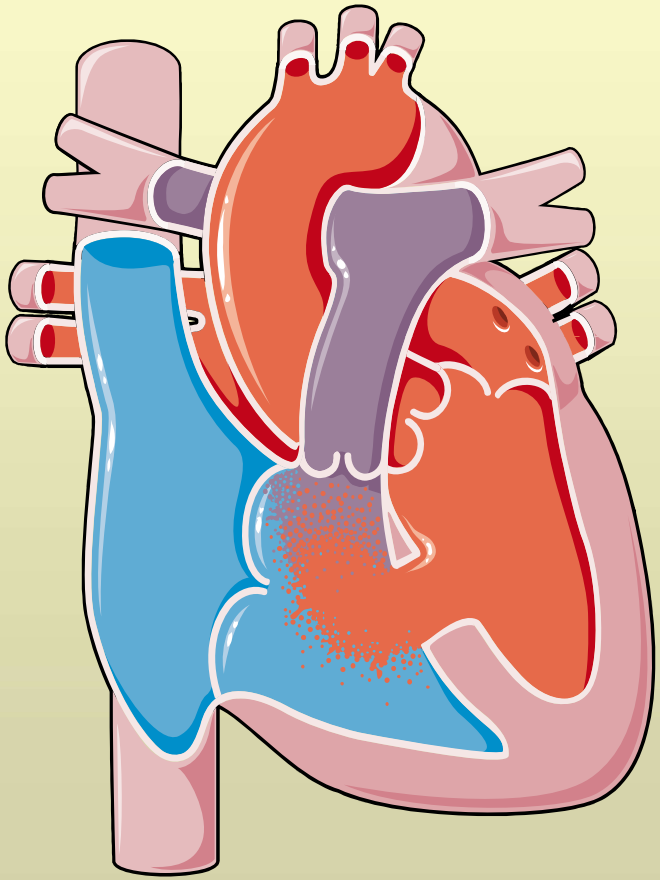
- VSD
- 25-30% of defects
- Opening in the ventricular septum
- Left-to-right shunt
- Right ventricular hypertrophy
- Deficient systemic blood flow

Surgical Repair

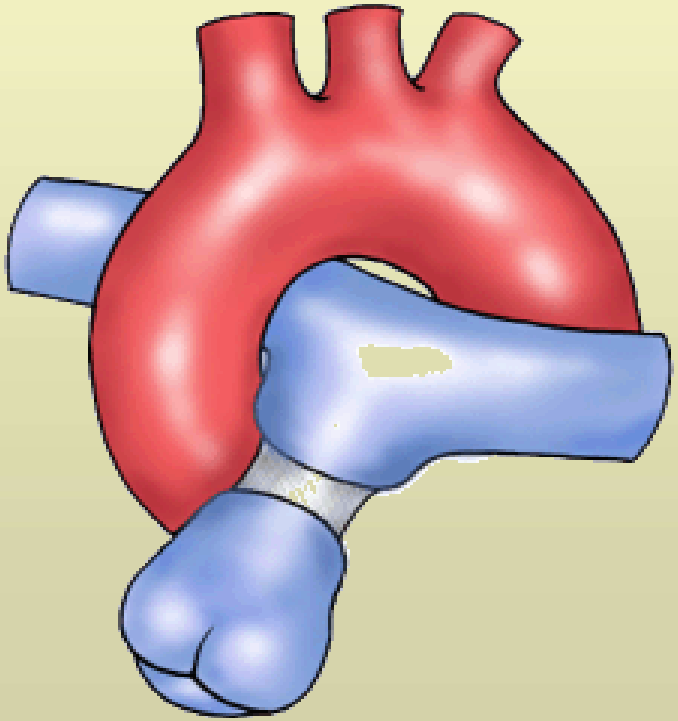
- Over a period of years the vessels in the lungs will develop thicker walls – the pressure in the lungs will increase and pulmonary vascular disease
- If pressure in the lungs becomes too high the un-oxygenated blood with cross over to the left side of the heart and un-oxygenated blood with enter the circulatory system.
- If the large VSD is repaired these changes will not occur.

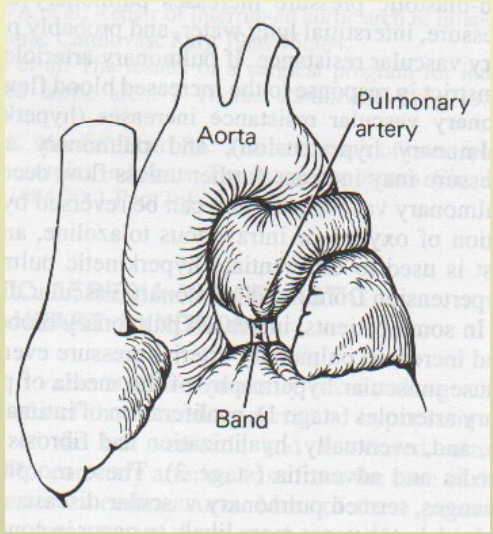
Acyanotic Heart Defects

- VSD
  - Incidence 20-25% CHD
  - Hemodynamic
  - Diagnosis
  - Management
    - Med
    - **Surgical** Mort <5%



Pulmonary artery banding stundum sem palliative aðgerð og definitive viðgerð gerð síðar á bernskuskeiði

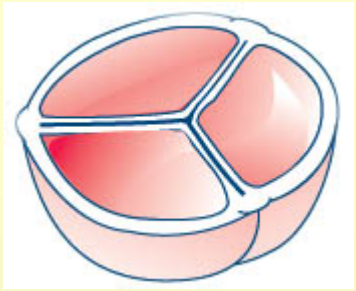




Ventricular Septal Defect, Case 19

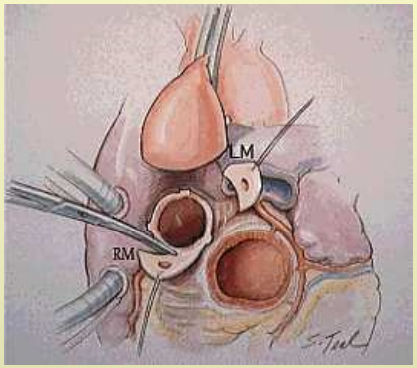
Video: VSD

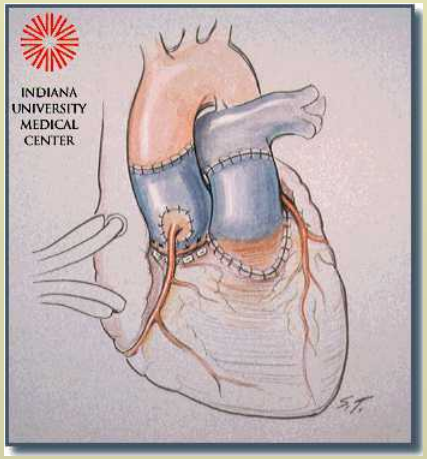
2:34

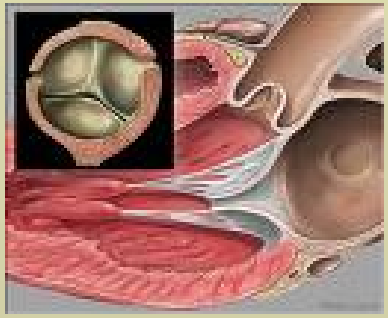


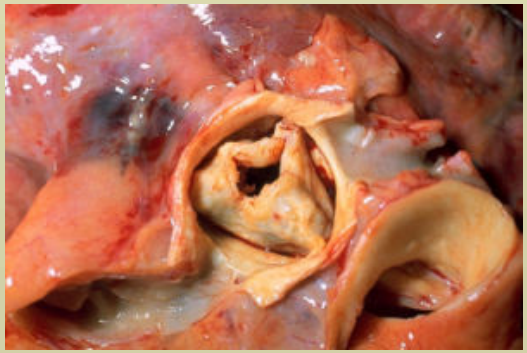
Aortic Valve disease
Acyanotic Heart Defects

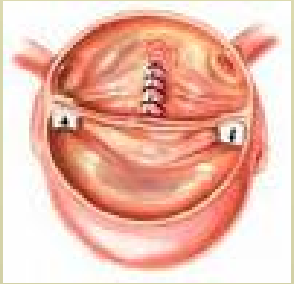
- AS
  - Incidence 5% CHD
  - Hemodynamic
  - Diagnosis
  - Management
    - Med
    - **Surgical** Mort 1-2%%, infant 15-20%





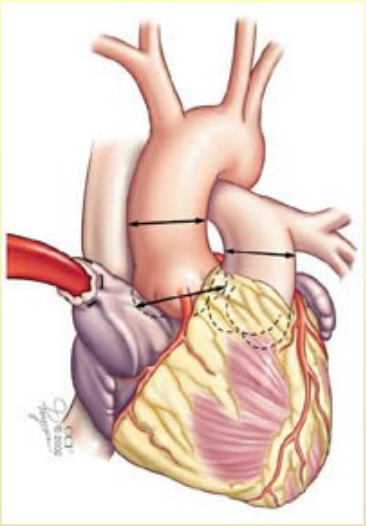
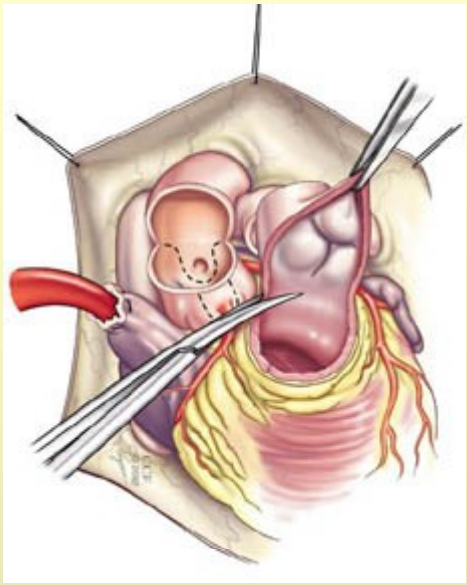
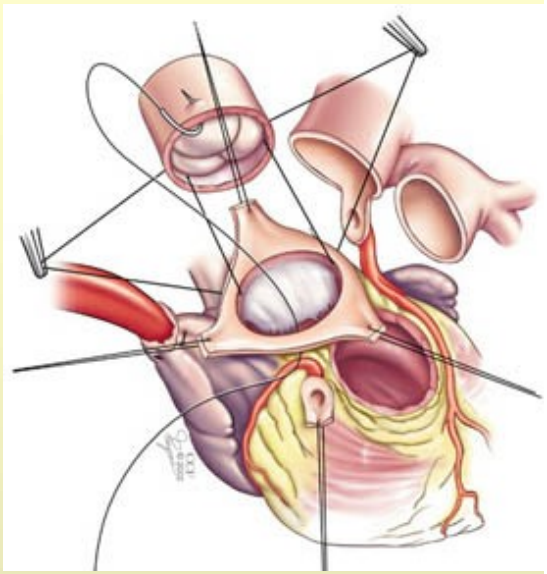
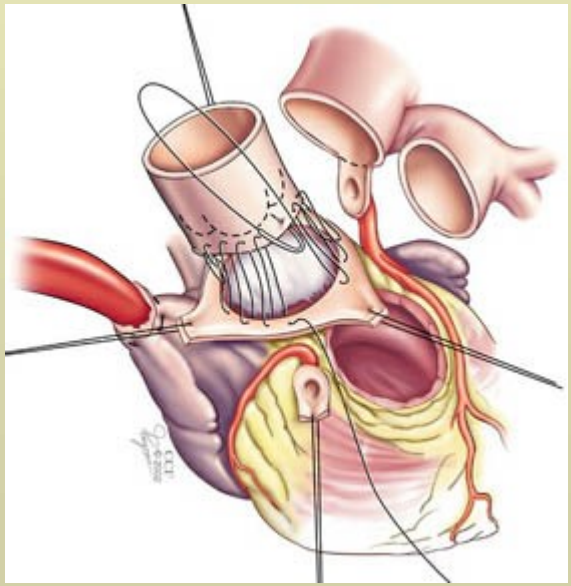
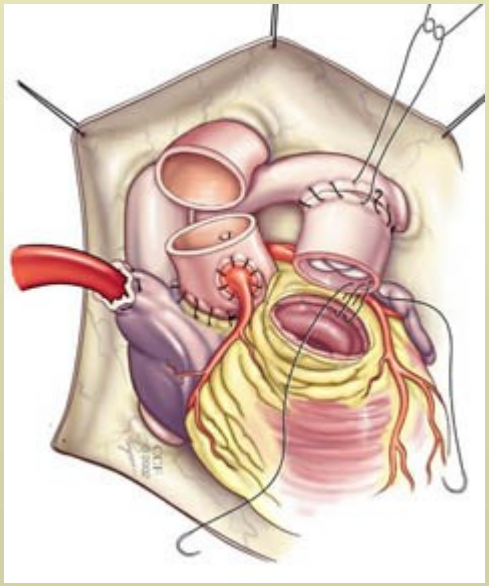
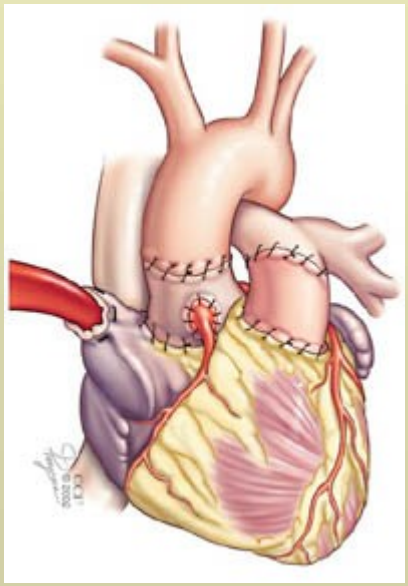




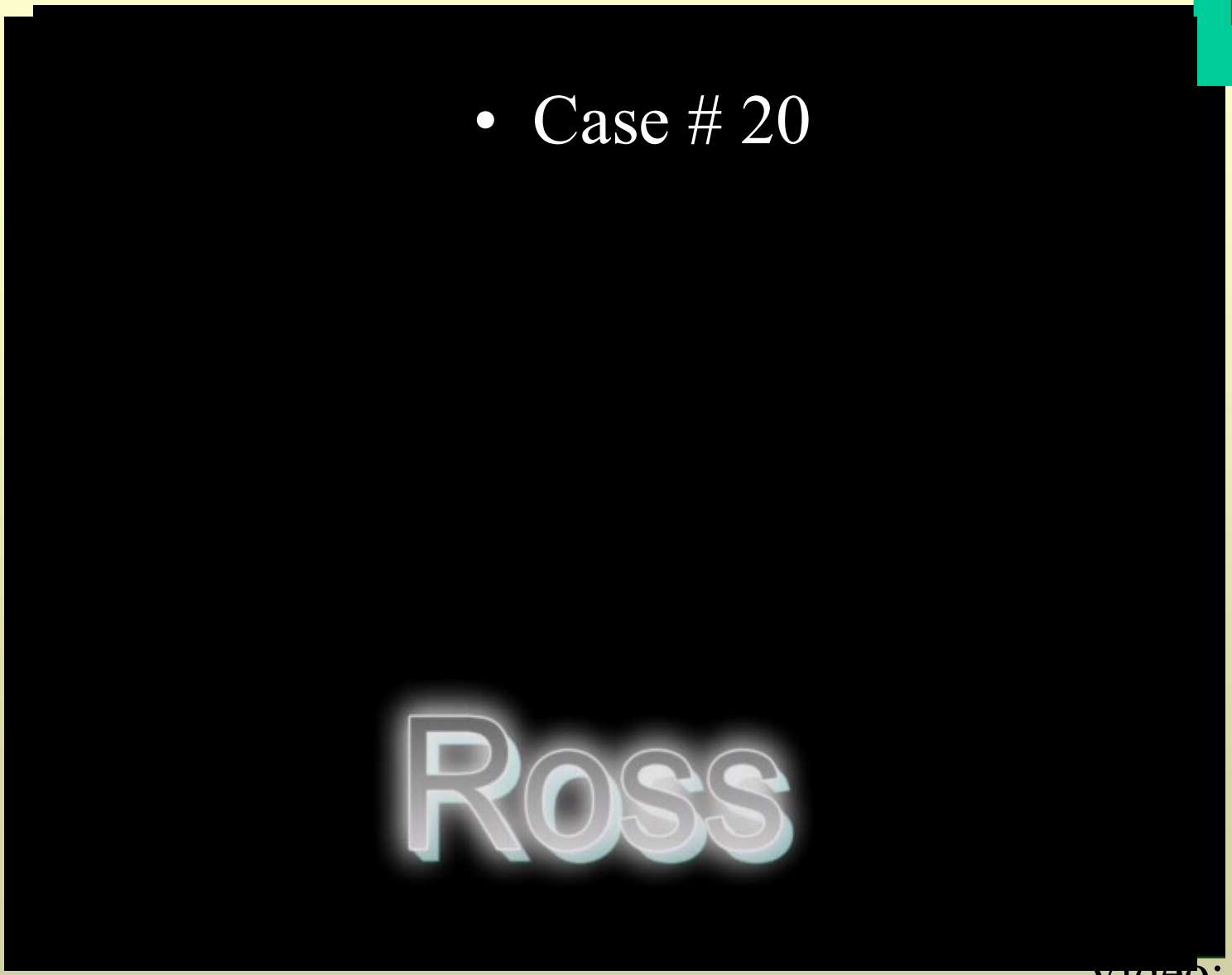


http://www.hsforum.com/stories/storyReader$1469

- Ross

- Case # 20

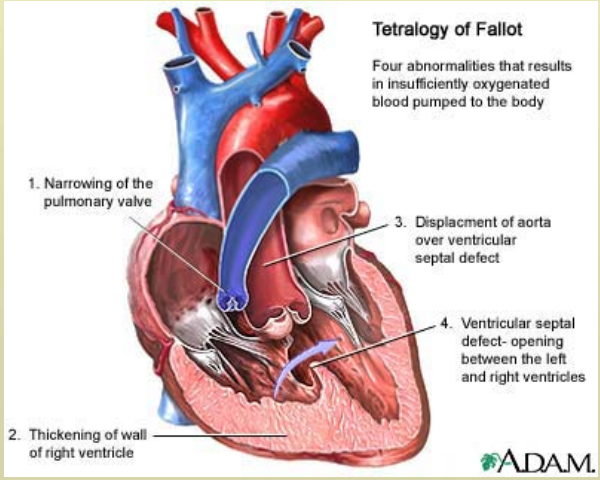


Video: Ross

10:22

Tetralogy of Fallot (TOF)

- 5-6% of defects
- Most common cardiac malformation responsible for cyanosis in a child over 1 year



TOF

- Four Components
  - VSD
  - Pulmonary stenosis – narrowing of pulmonary valve
  - Overriding of the aorta – aortic valve is enlarged and appears to arise from both the left and right ventricles instead of the left ventricle
  - Hypertrophy of right ventricle – thickening of the muscular walls because of the right ventricle pumping at high pressure

Diagnosis

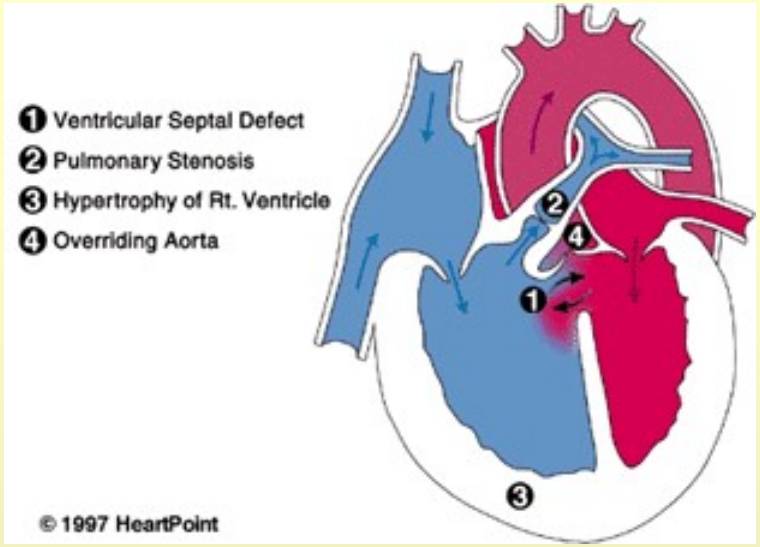
- Cyanosis
- Oxygen will have little effect on the cyanosis
- Rtg
- Loud heart murmur
- Echocardiogram – demonstrates the four defects characteristic of tetralogy

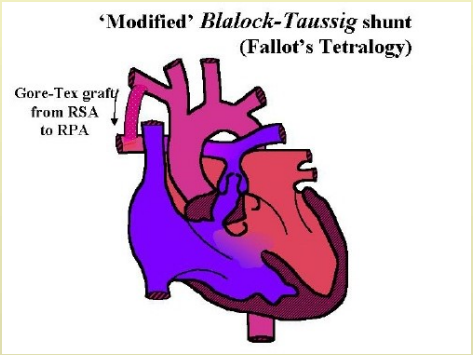
Treatment

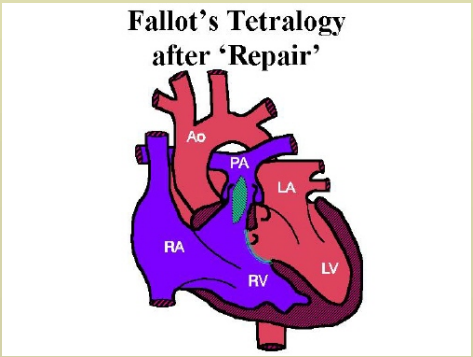
- If oxygen levels are extremely low prostaglandins may be administered IV to keep the PDA open
- Complete repair is done when the infant is about 6 months of age
- Palliation with BT shunt may be needed
- Correction includes
  - Closure of the VSD with dacron patch
  - The narrowed pulmonary valve is enlarge
  - Hypertrophy of right heart should remodel within a few months when pressure in right side is reduced

TOF

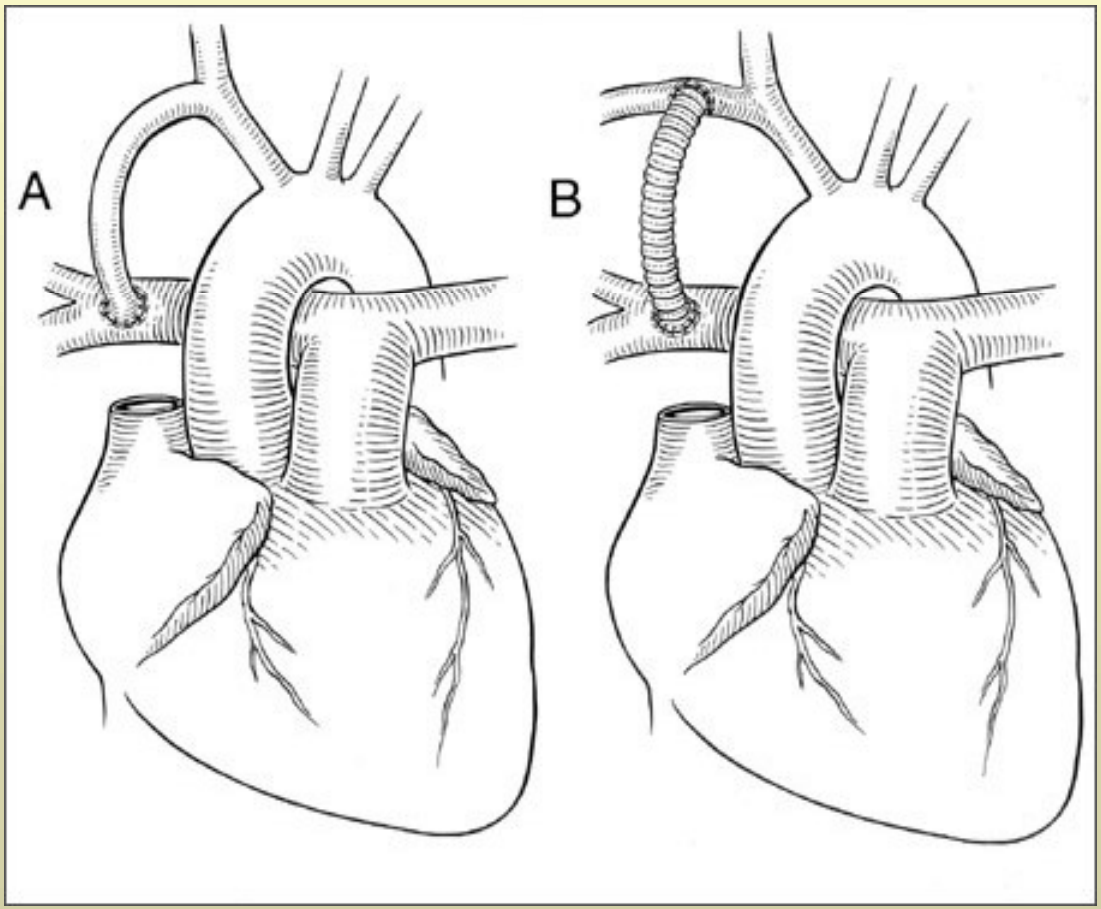
- TOF
  - Incidence 5-6% CHD
  - Hemodynamic
  - Diagnosis
  - Management
    - Med
    - **Surgical** Mort varies + 5-10/2POY





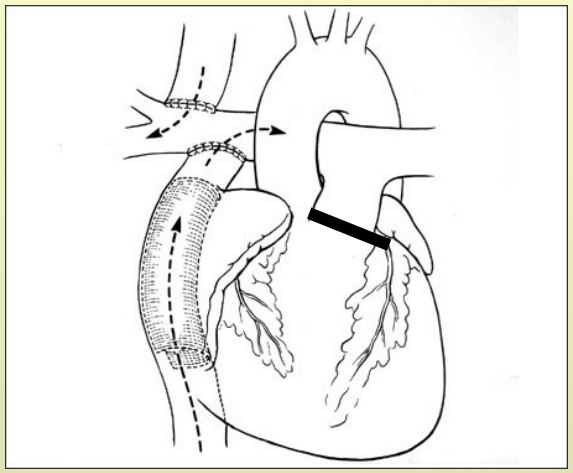


BT, Modified BT

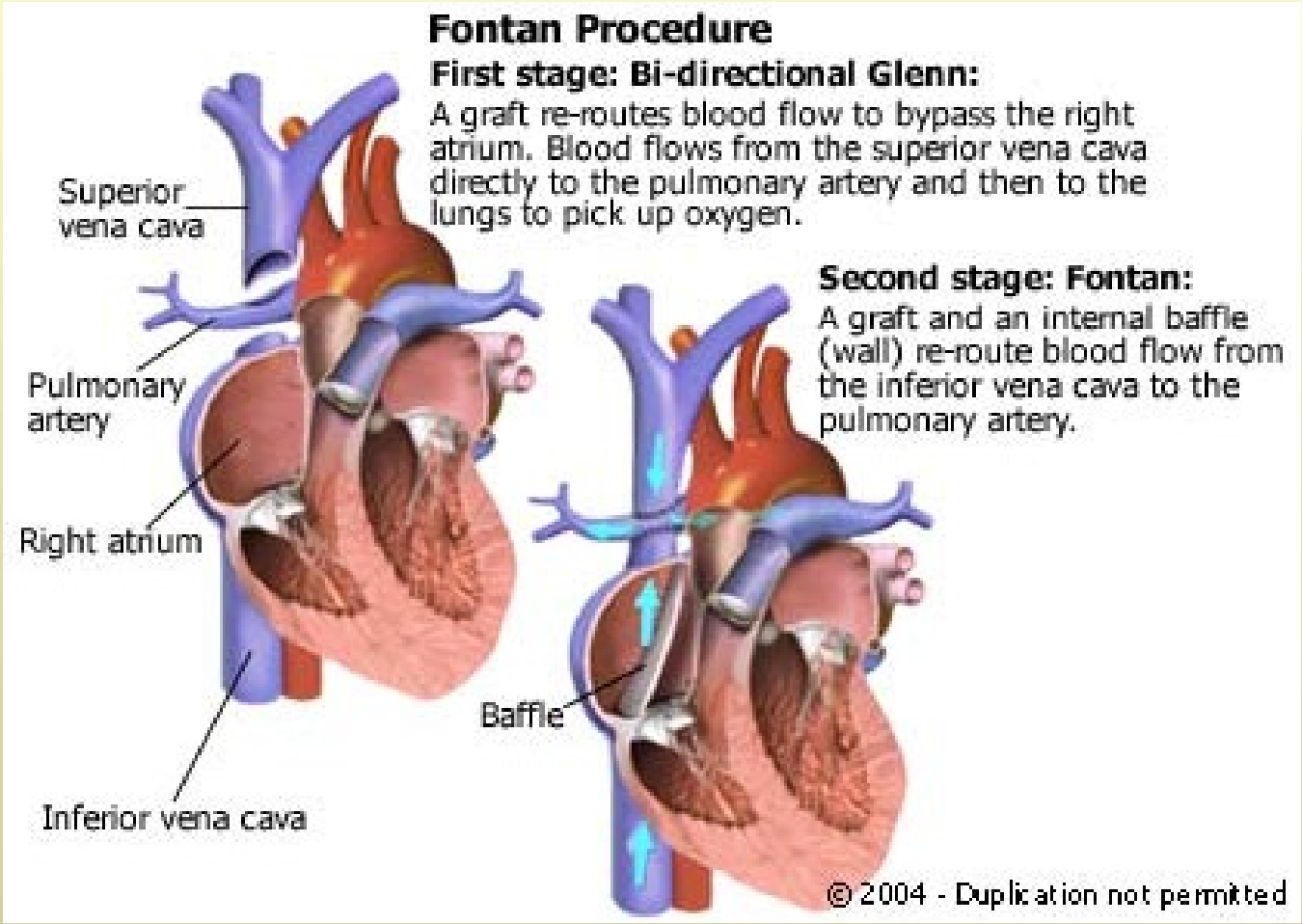


Cyanotic Heart Defects

- PA
  - Incidence <1% CHD
  - Hemodynamic
  - Manifestation
  - Diagnosis
  - Management
    - Med
    - **Surgical** septostomy,Blalock-Taussig,valvotomy:
      - » Mort: 10-25%, OutflTractReconstr 25%
      - » Fontan <40%

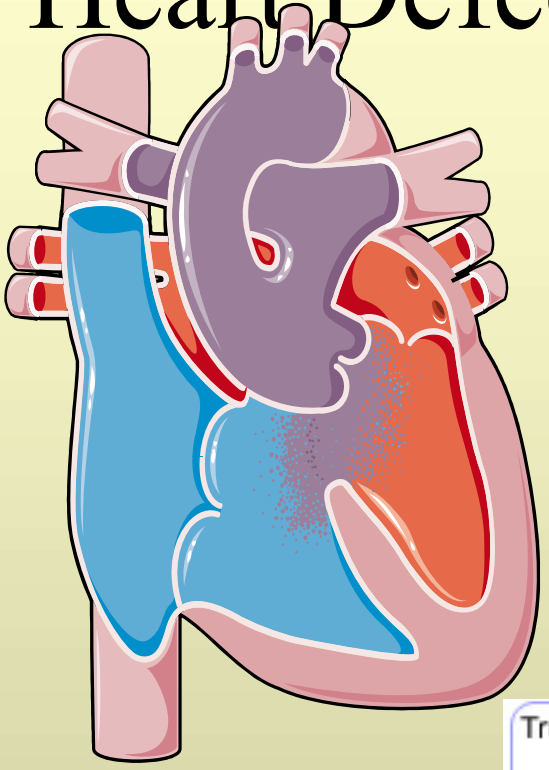


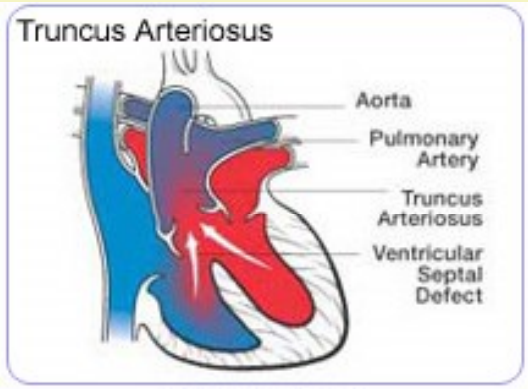
FONTAN


Fontan Procedure
First stage: Bi-directional Glenn:
A graft re-routes blood flow to bypass the right atrium. Blood flows from the superior vena cava directly to the pulmonary artery and then to the lungs to pick up oxygen.
Superior vena cava
Pulmonary artery
Right atrium
Inferior vena cava
Second stage: Fontan:
A graft and an internal baffle (wall) re-route blood flow from the inferior vena cava to the pulmonary artery.
Baffle
© 2004 - Duplication not permitted

Cyanotic Heart Defects

- TA
  - Incidence <1% CHD
  - Hemodynamic
  - Manifestation
  - Diagnosis
  - Management
    - Med
    - **Surgical**,
      - » Mort 30%,
      - » Rastelli´s Mort20-60%





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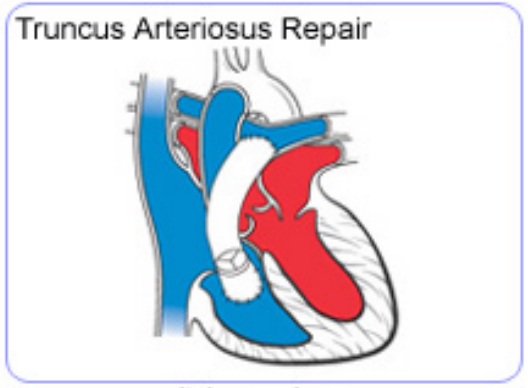
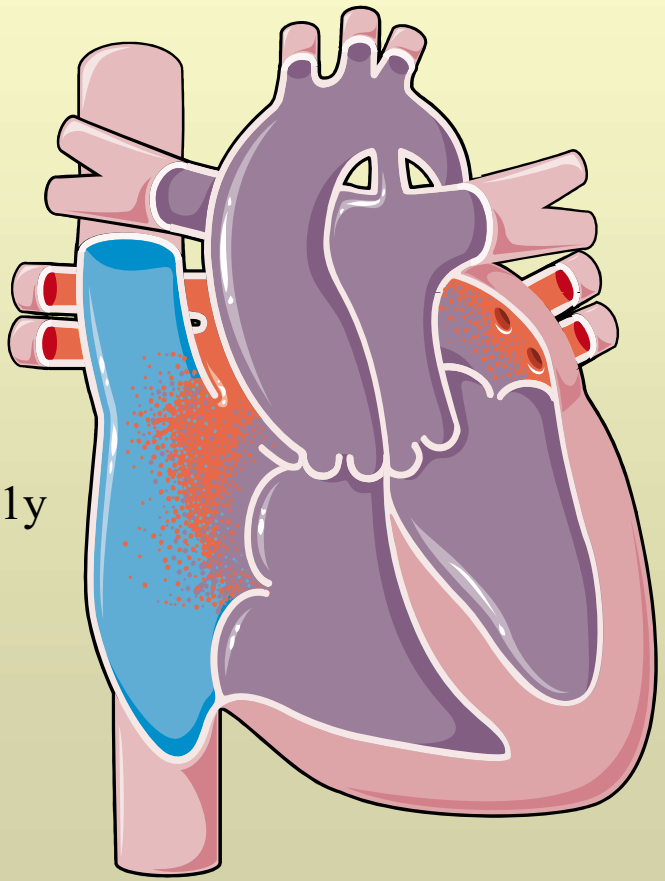


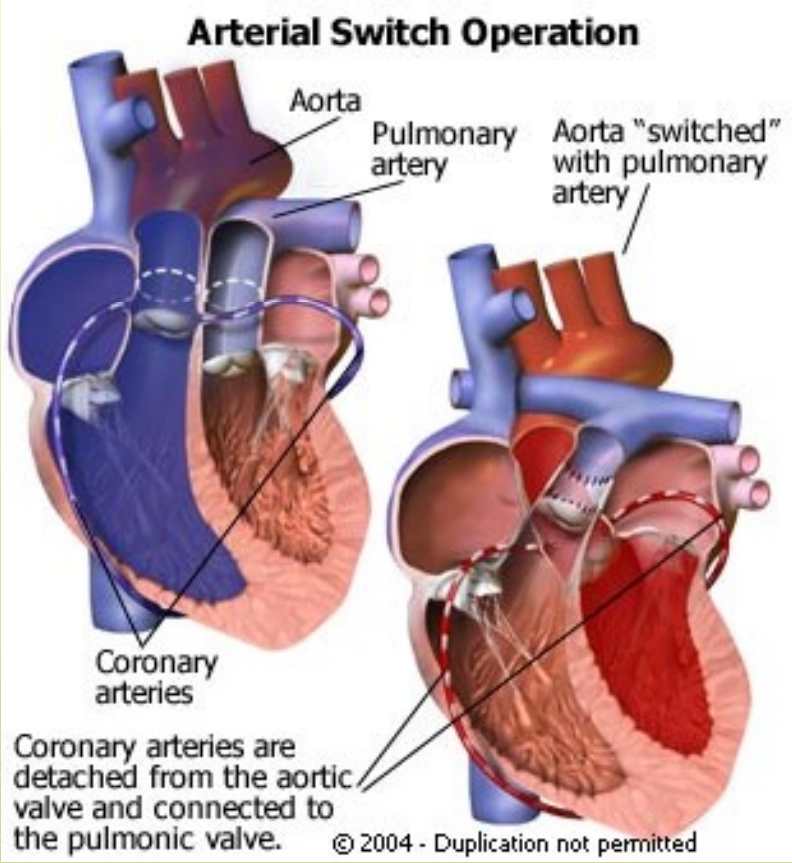
click to enlarge

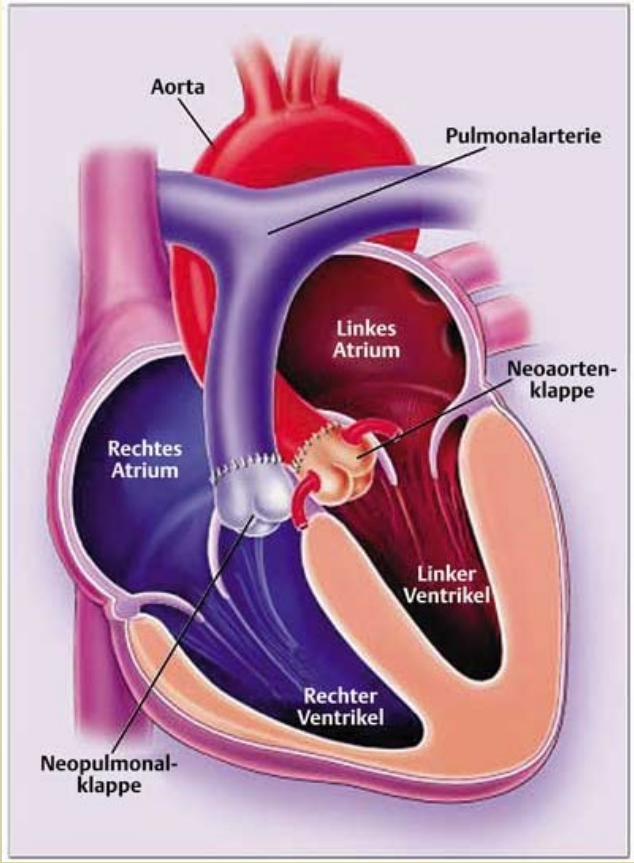
Cyanotic Heart Defects

- TGA-Complete Transposition of the GA
  - Incidence 5% CHD
  - Hemodynamic D-TGA
  - Manifestation
  - Diagnosis
  - Management - Cardiac emergency
    - Med septostomycatlab/open Mort 10-25% 90%/1y
      - » Surgical
      - » Ventricular-level: Rastelli, or
      - » Arterial-level Jatene´s, or
      - » atrial Senning, Mustard´s



TGA: Arterial-level Jatene´s





Pediatric Cardiothoracic Surgery

- Thorax
  - Veggur
  - Pleura
  - Lungu
  - Mediastinum
- Hjarta og æðar í thorax
  - Meðfæddir gallar
  - Áunnir gallar/ sjúkdómar

Pericardial Effusion

- Presenting complaint
  - Precordial pain
  - Cough
  - Dyspnoea
  - Abdominal pain
  - Vomiting
  - Fever
  - Other organs involvement
- Signs:
  - Position: leaning forward.
  - Puffy face
  - Friction rub
  - Absent apical impulse
  - Muffled heart sounds
  - Pulsus paradoxus
  - Distended neck veins
  - Low QRS complex, T inversion

Pericardial Effusion Case # 21

- A relatively large pericardial effusion must be present to cause an enlarged cardiac shadow with the usual "water bottle" configuration on a *chest roentgenogram*

Video: Chilothorax
8:56

Infective Endocarditis

Presenting symptoms and clinical features include:

- Fever
- Malaise
- Fatigue
- Anorexia
- Weight loss
- Splenomegaly
- Cardiac murmur
- Petechiae
- Roth spots
- Janeway lesions
- Osler nodes

Some of the more diagnostic symptoms (the latter half of the above list) are occurring less frequently in patients with subacute IE, making diagnosis a greater challenge.

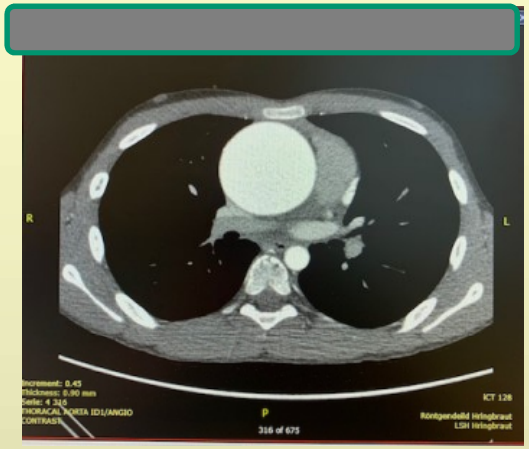
Class I Indications for Surgery

- Acute AR or MR with heart failure.
- Acute AR with tachycardia and early closure of the MV.
- Fungal endocarditis.
- Annular or aortic abscess.
- Sinus or aortic aneurysm.
- Persistent bacteremia and valve dysfunction
  - After 7-10 days of appropriate antibiotics.

Circulation. 98(18):1949-1984, 1998

Case # 22

- Brottnám á aorta aneurysma og reconstruction á aortarót með total rótarskiptum með lokusparandi viðgerð á aortalokunni og ígræðslu kransæða, svokölluð David aðgerð. FMSD40
- Central plicering á aortaloku, bicuspid loku FMSC 10
- Resuspension á aortalokunni í 28 mm Vascutek æðagrafti FMSC 96
- Reconstruction á aorta ascendens með Vascutek æðagrafti FCSB08
- Correction á pectus excavatum með 9" Nuss spöng GASF0313
- Wrapping á aorta ascendens arcus mótum með Prolene neti FCSA04
- Extracorporel circulation með ante- og retrograd kaldri blóðcardioplegiu og moderativ hypothermia *FZSA00*



Aortic diameter/BSA

Table 5. Risk of Complications by Aortic Diameter and Body Surface Area With Aortic Size Index Given Within Chart

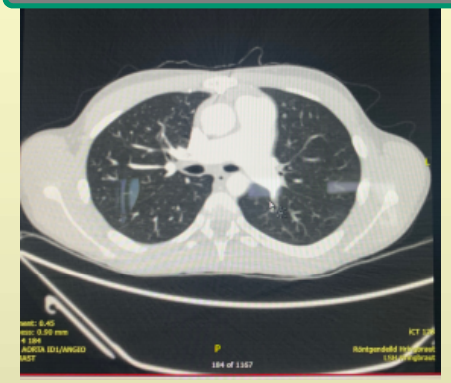
	Aortic Size (cm)									
	3.5	4.0	4.5	5.0	5.5	6.0	6.5	7.0	7.5	8.0
BSA										
1.30	2.69	3.08	3.46	3.85	4.23	4.62	5.00	5.38	5.77	6.15
1.40	2.50	2.86	3.21	3.57	3.93	4.29	4.64	5.00	5.36	5.71
1.50	2.33	2.67	3.00	3.33	3.67	4.00	4.33	4.67	5.00	5.33
1.60	2.19	2.50	2.80	3.13	3.44	3.75	4.06	4.38	4.69	5.00
1.70	2.05	2.35	2.65	2.94	3.24	3.53	3.82	4.12	4.41	4.71
1.80	1.94	2.22	2.50	2.78	3.06	3.33	3.61	3.89	4.17	4.44
1.90	1.84	2.11	2.37	2.63	2.89	3.16	3.42	3.68	3.95	4.22
2.00	1.75	2.00	2.25	2.50	2.75	3.00	3.25	3.50	3.75	4.00
2.10	1.67	1.90	2.14	2.38	2.62	2.86	3.10	3.33	3.57	3.80
2.20	1.59	1.82	2.05	2.27	2.50	2.72	2.95	3.18	3.41	3.64
2.30	1.52	1.74	1.96	2.17	2.39	2.61	2.83	3.04	3.26	3.48
2.40	1.46	1.67	1.88	2.08	2.29	2.50	2.71	2.92	3.13	3.33
2.50	1.40	1.60	1.80	2.00	2.20	2.40	2.60	2.80	3.00	3.20

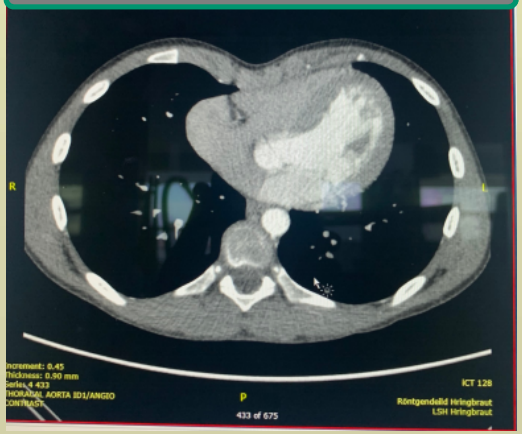
□ = low risk (~1% per yr); ▒ = moderate risk (~8% per yr); ■ = severe risk (~20% per yr).
White area indicates low risk, light gray area indicates moderate risk, and dark gray area indicates severe risk.
BSA = body surface area.

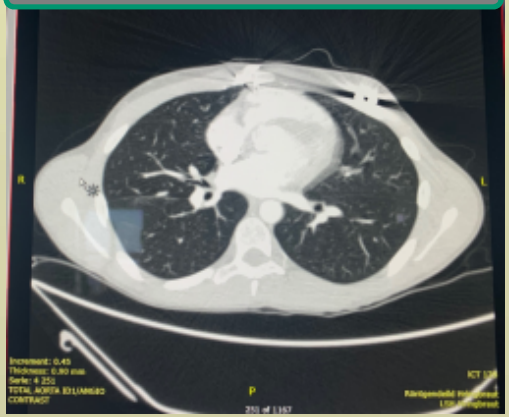
Operate when ASI >2.75 cm/m^2

Ann Thorac Surg 2006;81:169

BJT 2020







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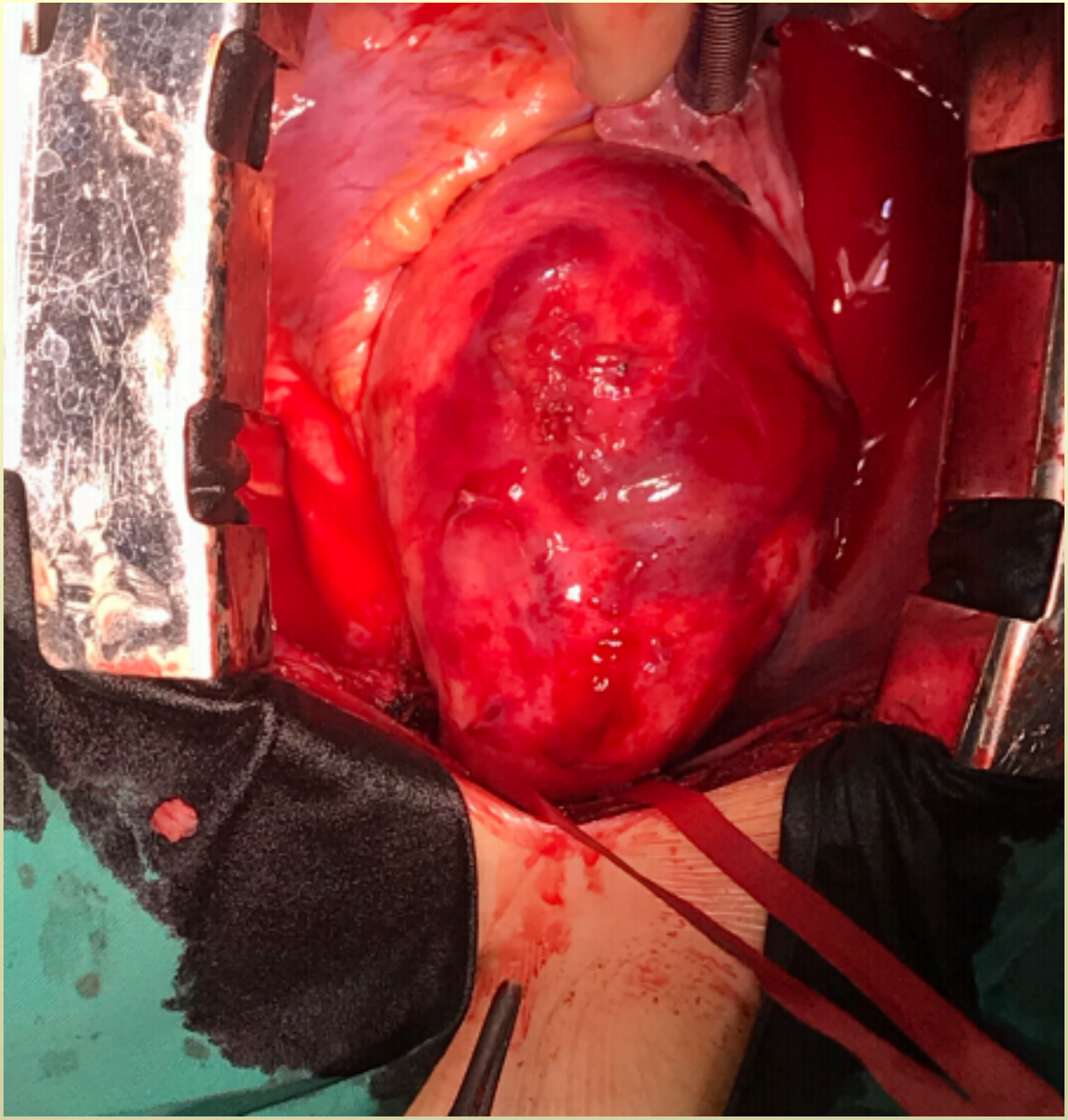

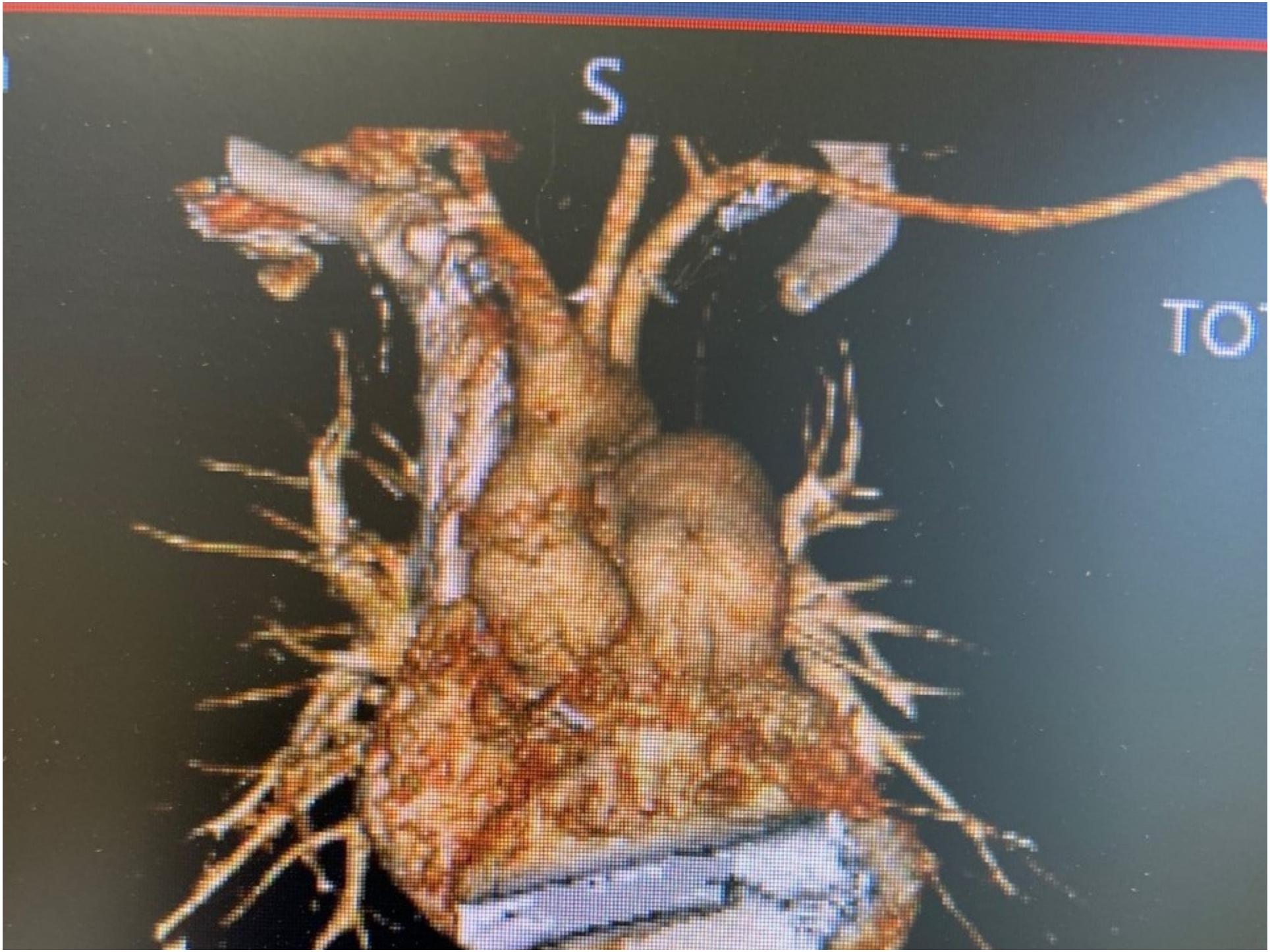
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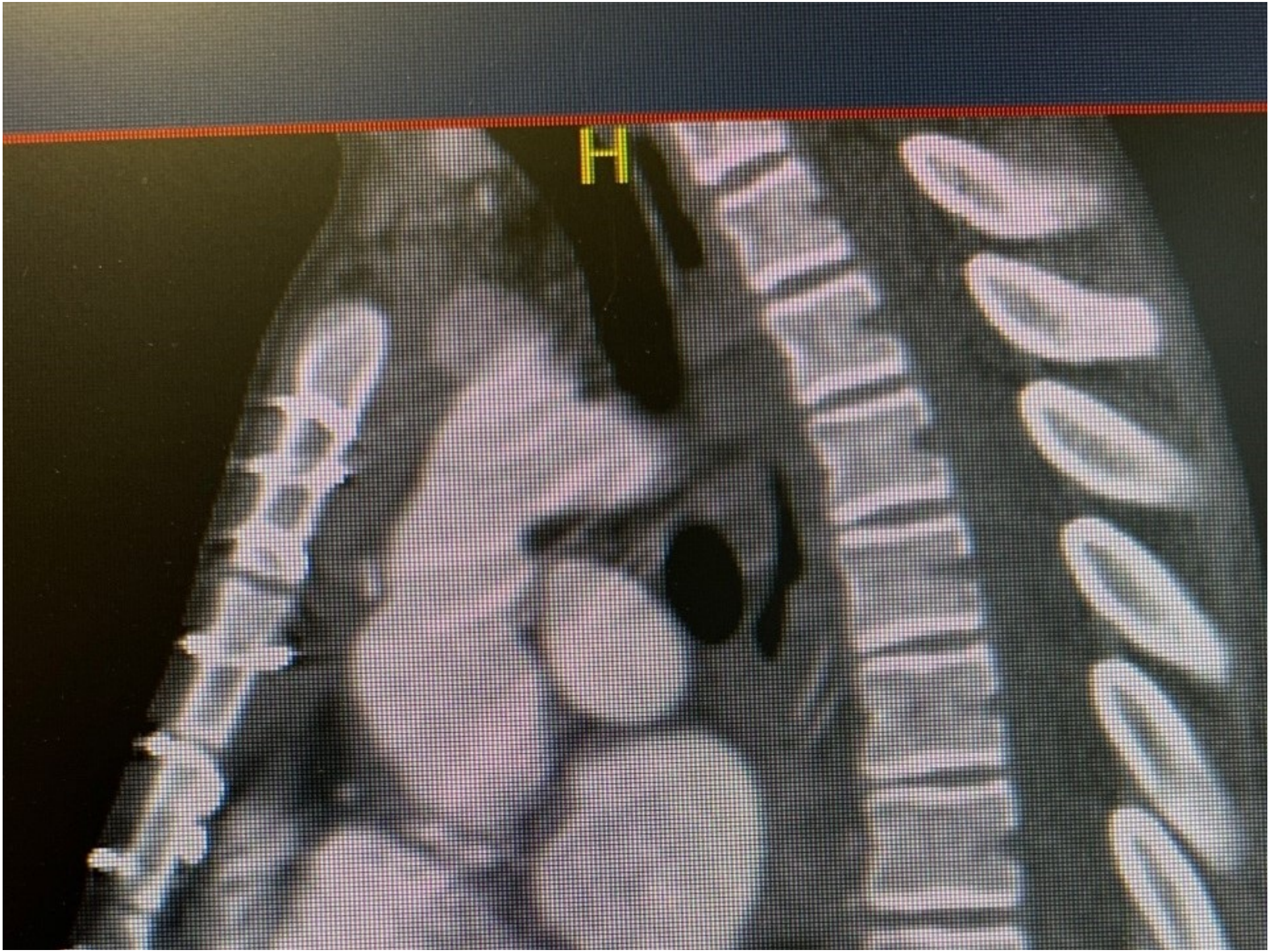

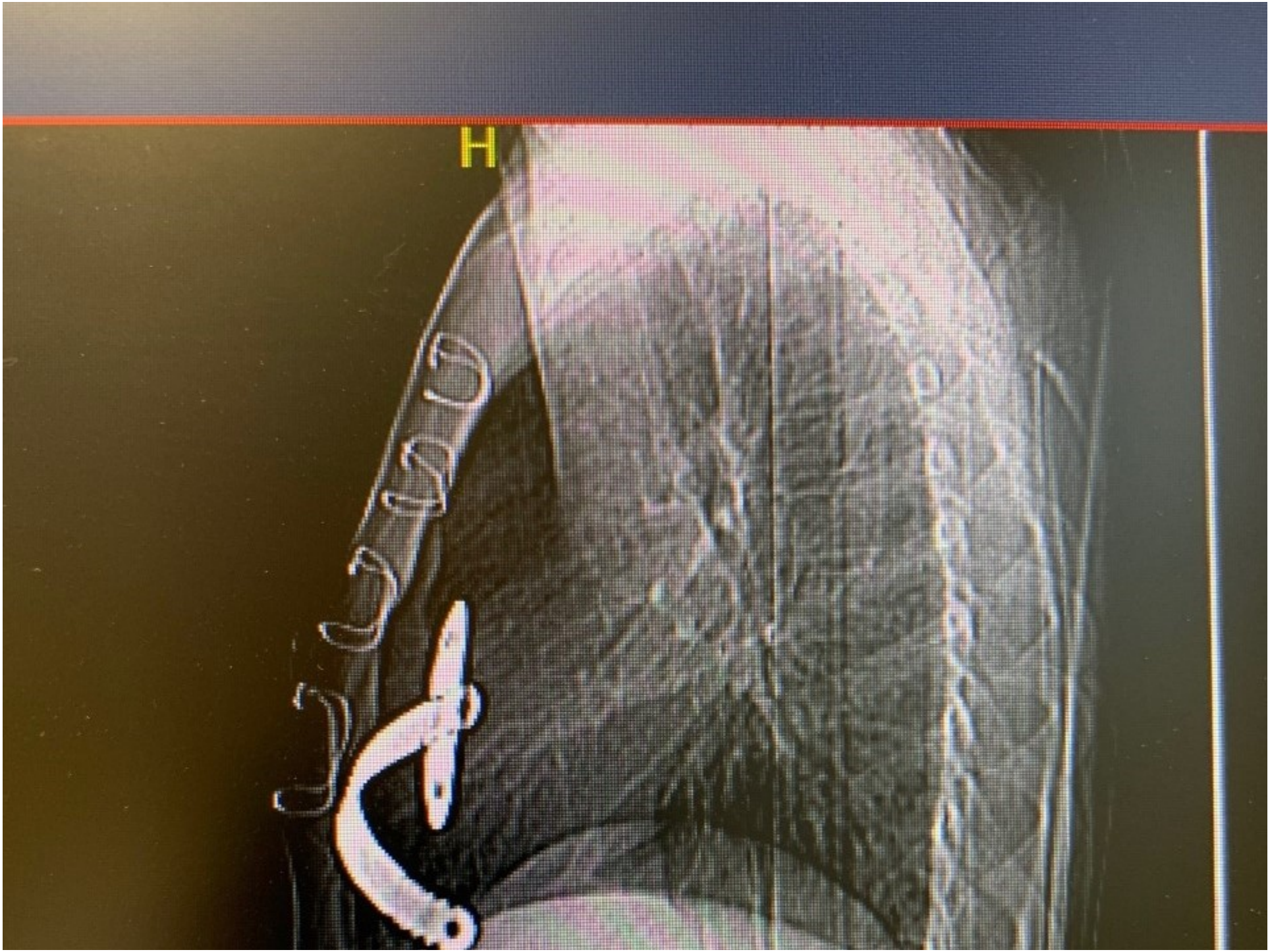
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Case 23

ECMO

1:53

Video: ECMO

DEFINITION

- Prolonged extracorporeal cardiopulmonary bypass in patients with acute reversible respiratory or cardiac failure

TYPES OF ECMO

- 2 Types:
  1. Veno-arterial
  2. Veno-venous

INDICATIONS

Neonatal Cardiopulmonary failure:

- Meconium Aspiration
- Primary Pulmonary Hypertension
- Respiratory Distress Syndrome
- Pneumonia
- Massive Air Leak

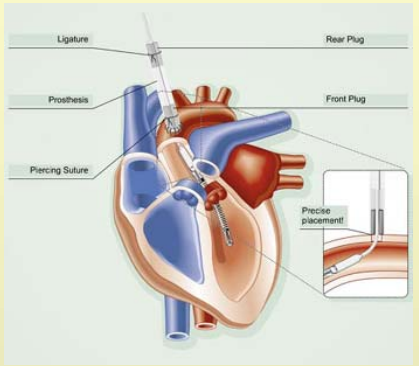
Survival > 90%

- Congenital Diaphragmatic Hernia
- Sepsis

Survival 60 %

Heart Failure

- Bridge to improvement
- Bridge to transplantation
- Destination therapy



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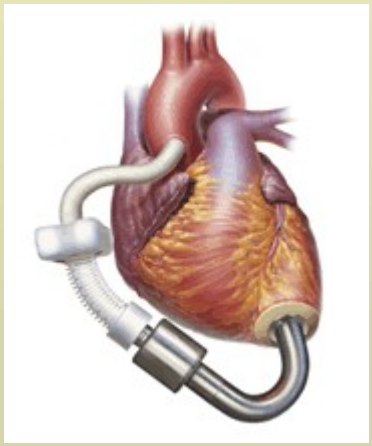
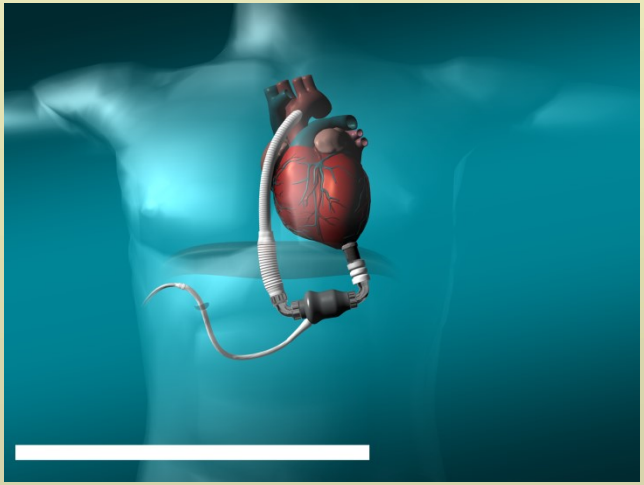


Animation Impella inn



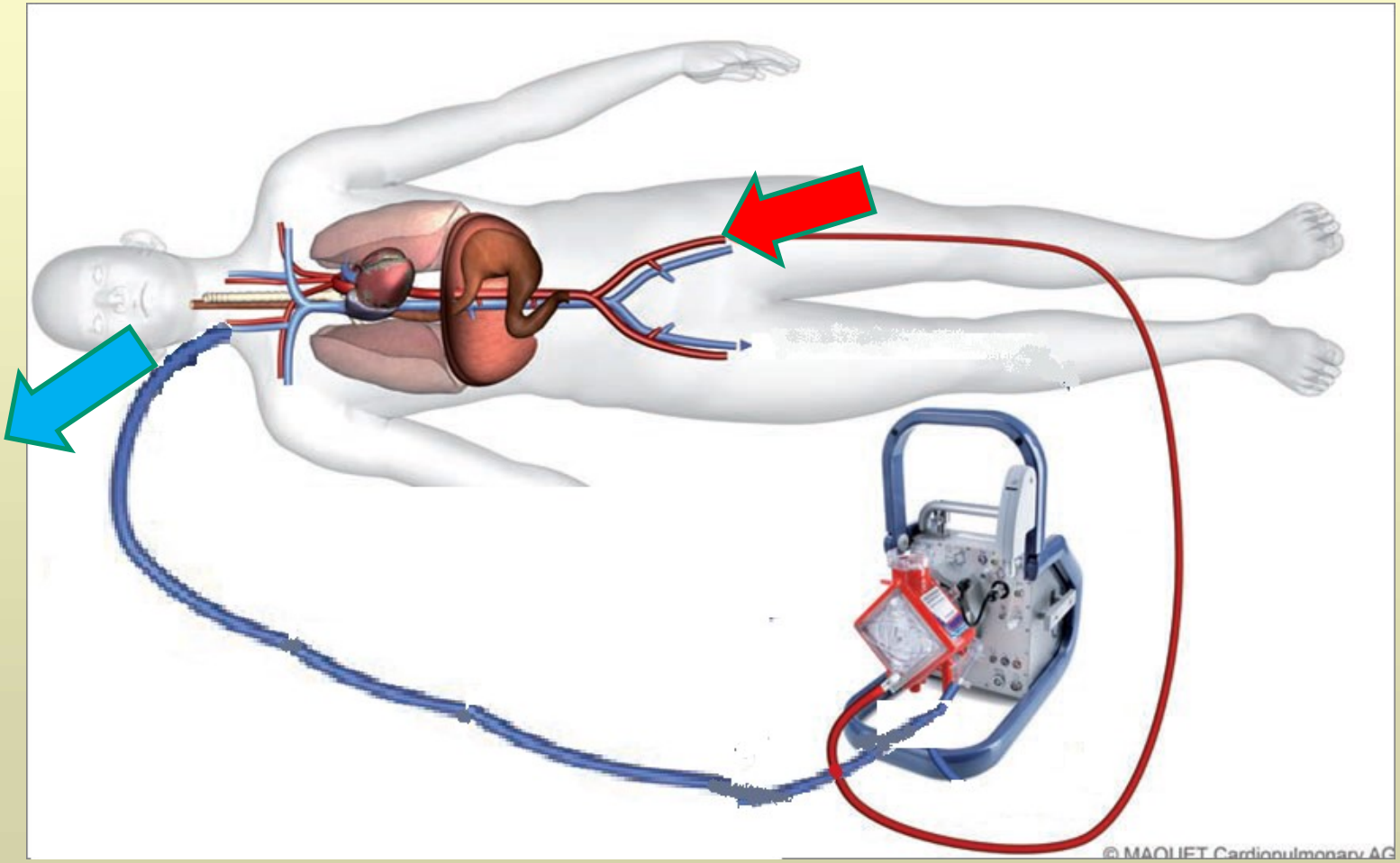
HMII nám

http://www.maquet-cardiohelp.com/index.php?id=100&L=1

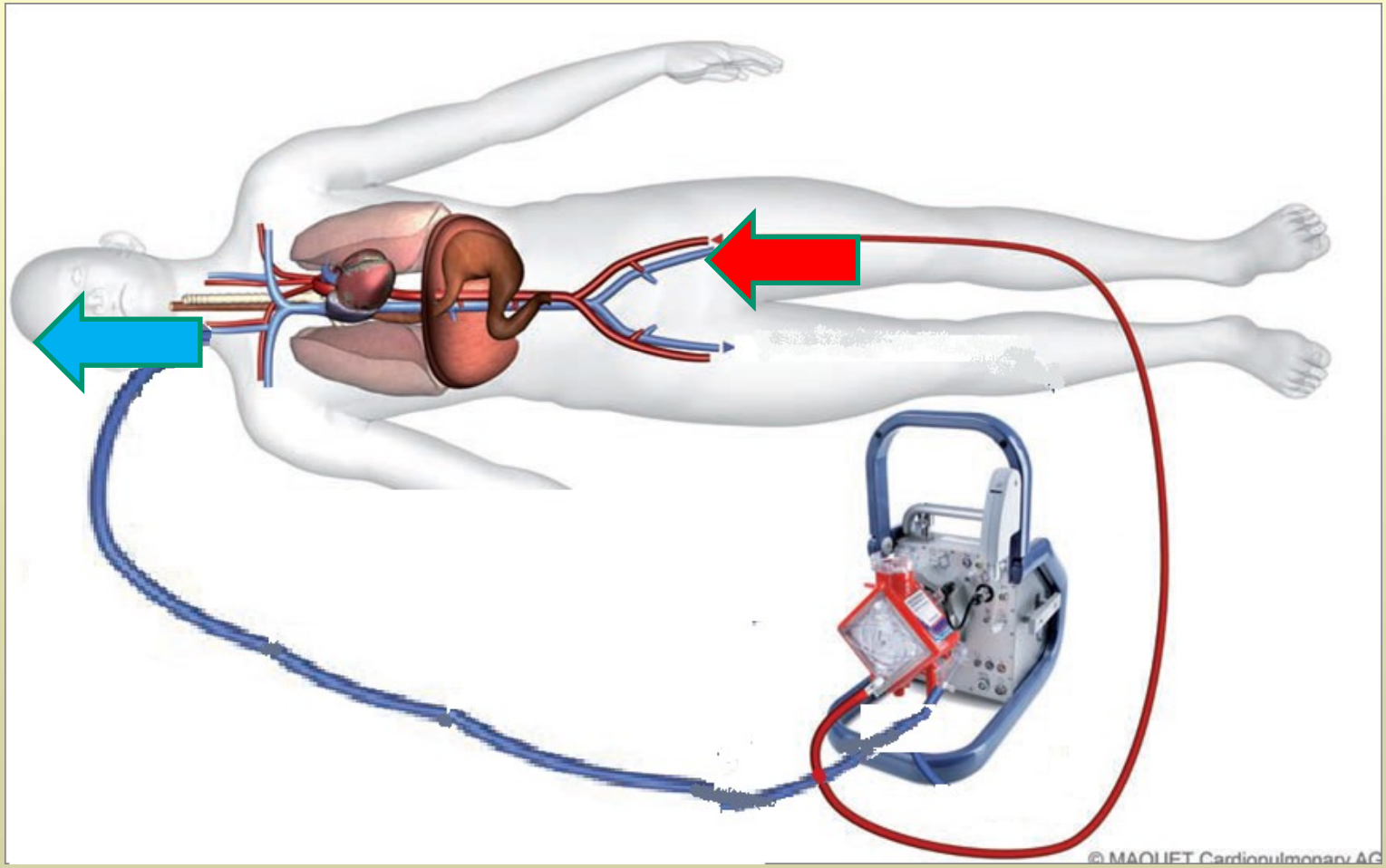
http://www.maquet-cardiohelp.com/index.php?id=17&L=1

ECMO:
Börn sérstaklega

V-A



V-V

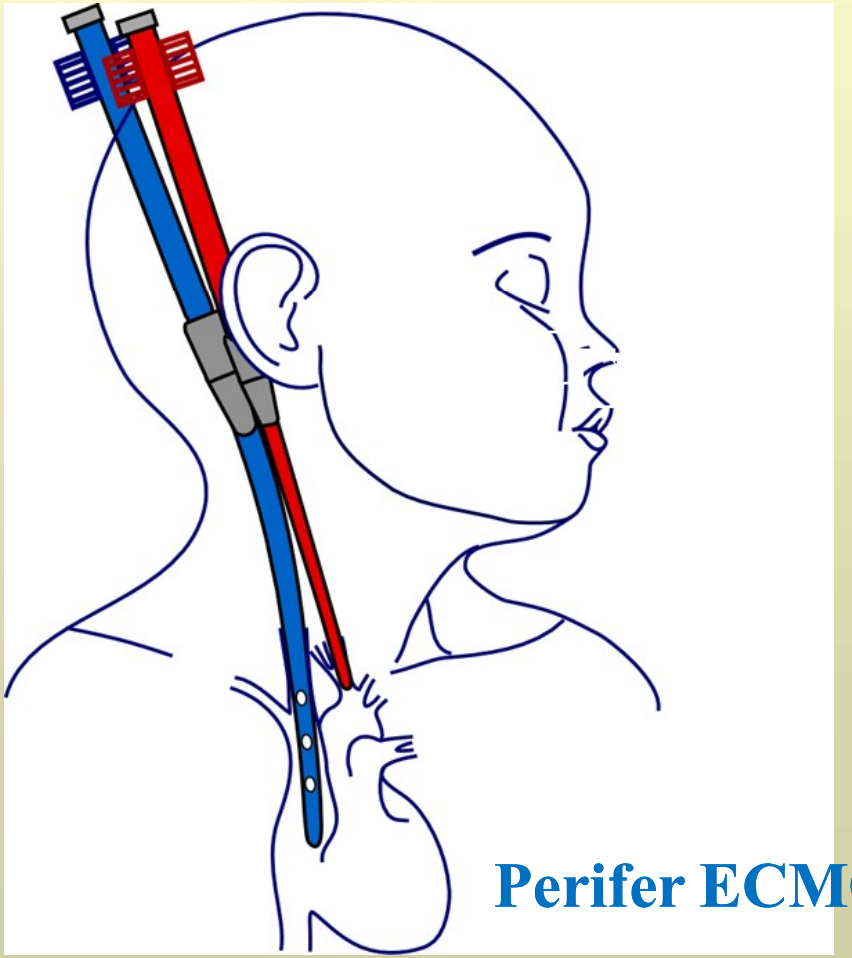


Perifer ECMO

Case #24, nýfætt barn

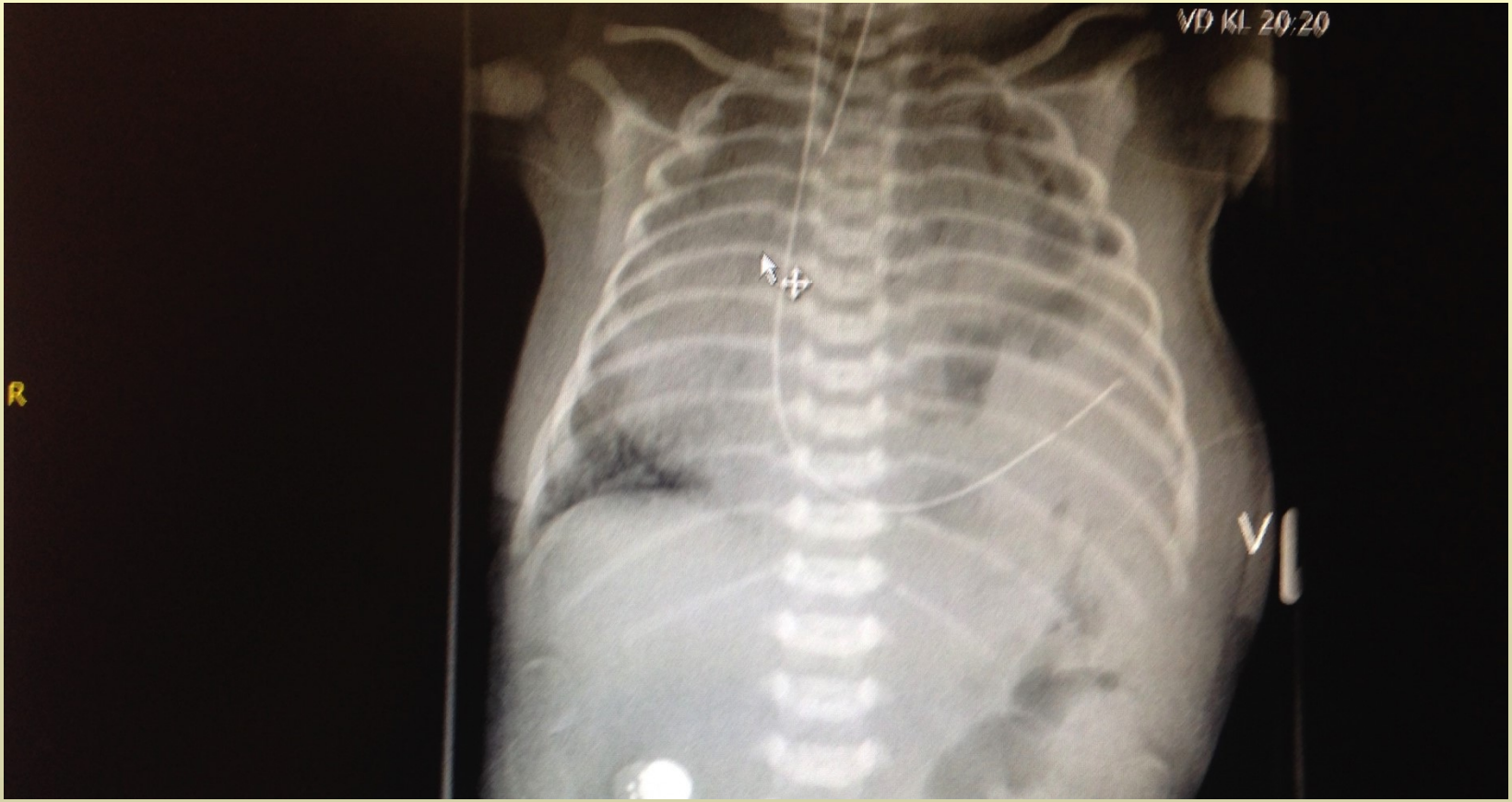
- CDH
- Landspítalinn – Karolinska Stockholm

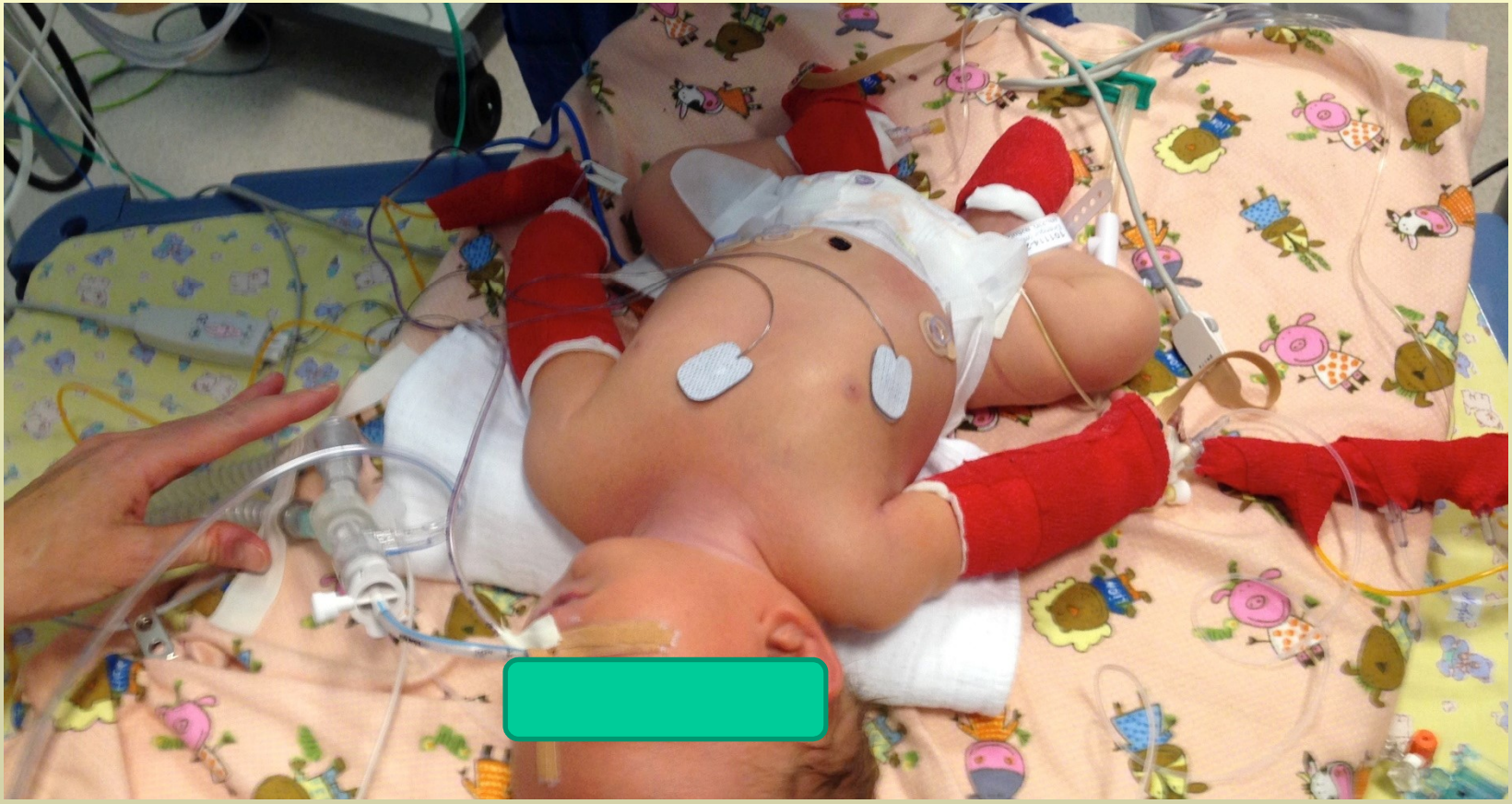
V-A

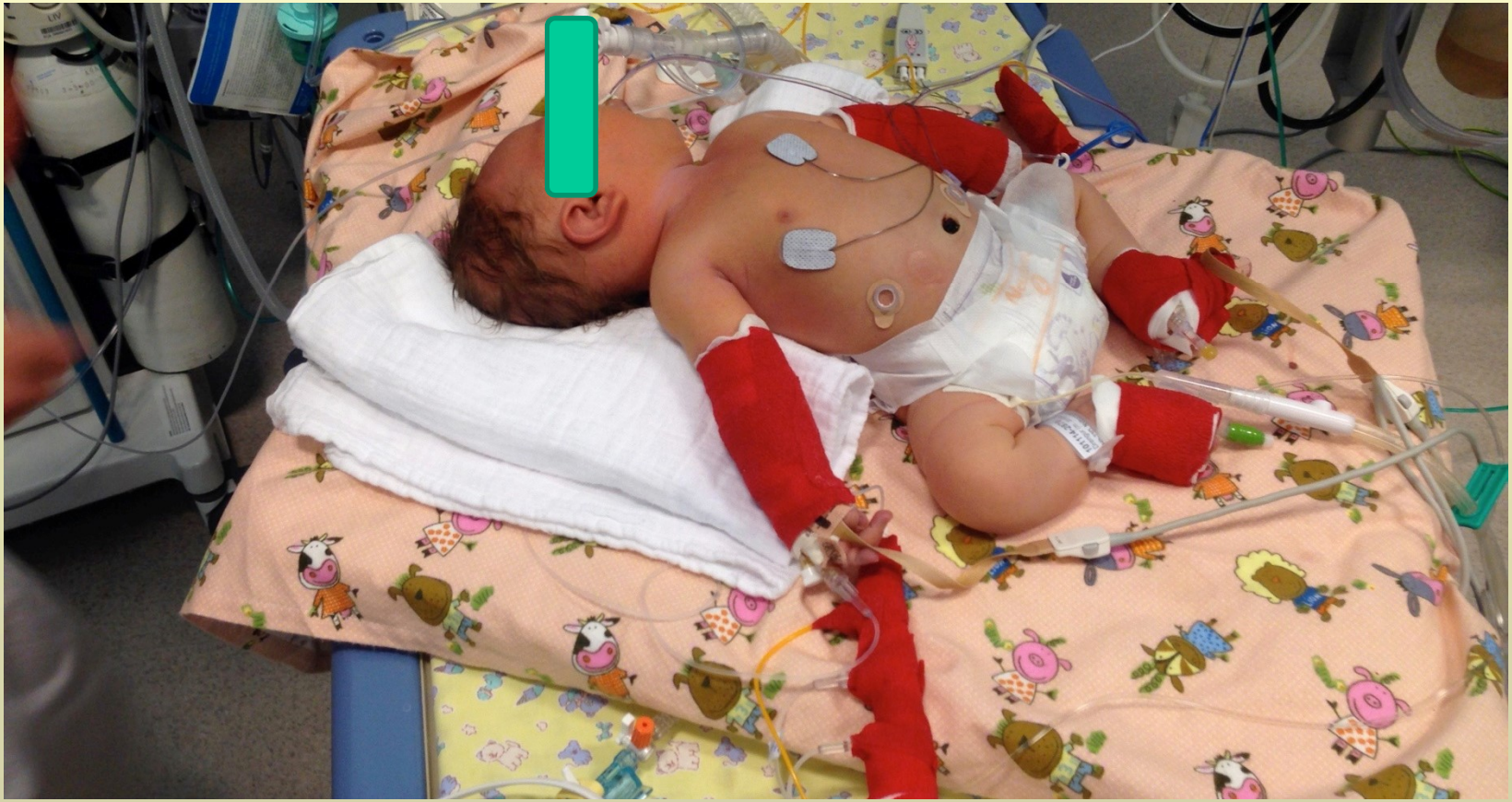


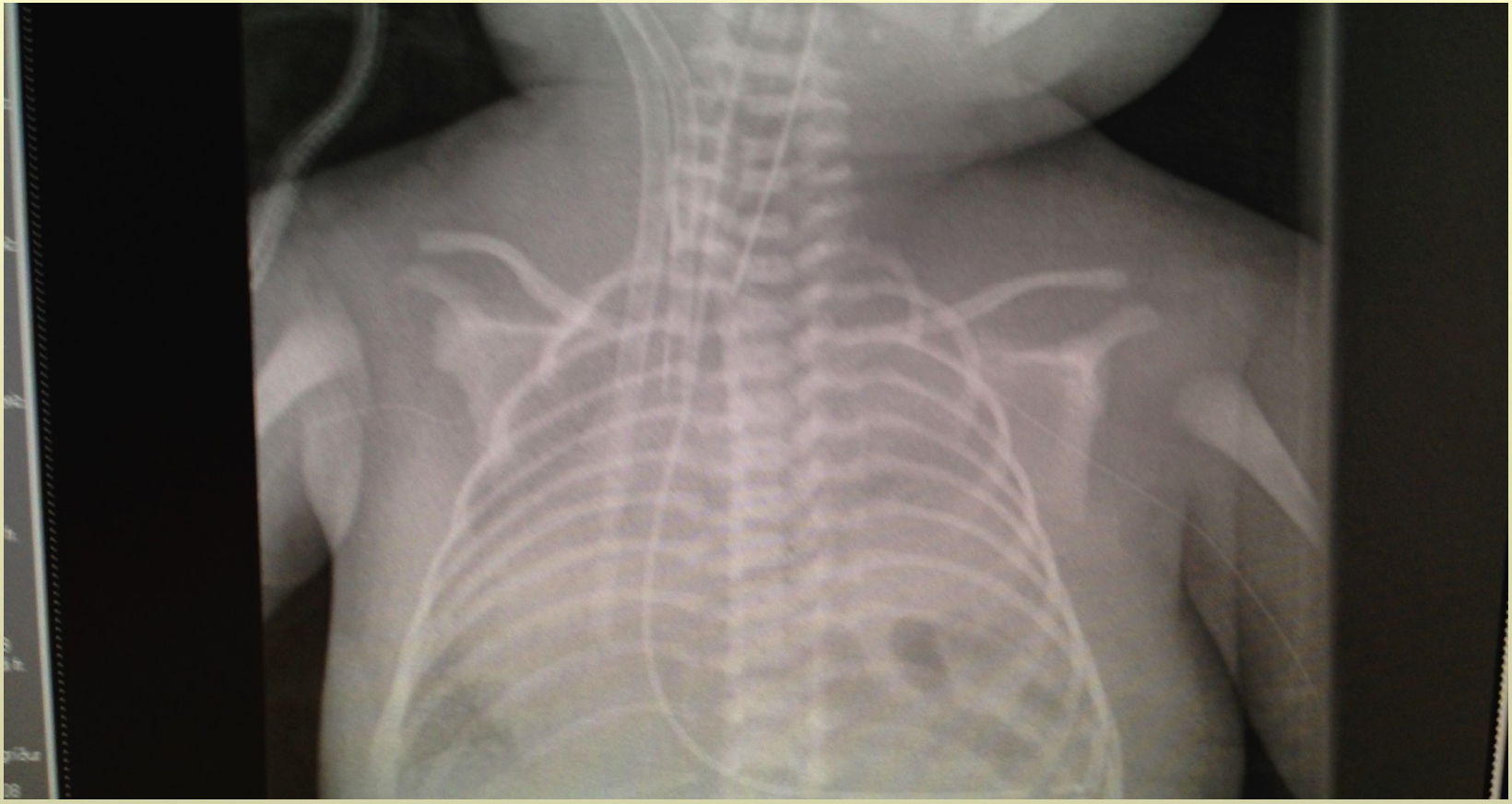
Perifer ECMO

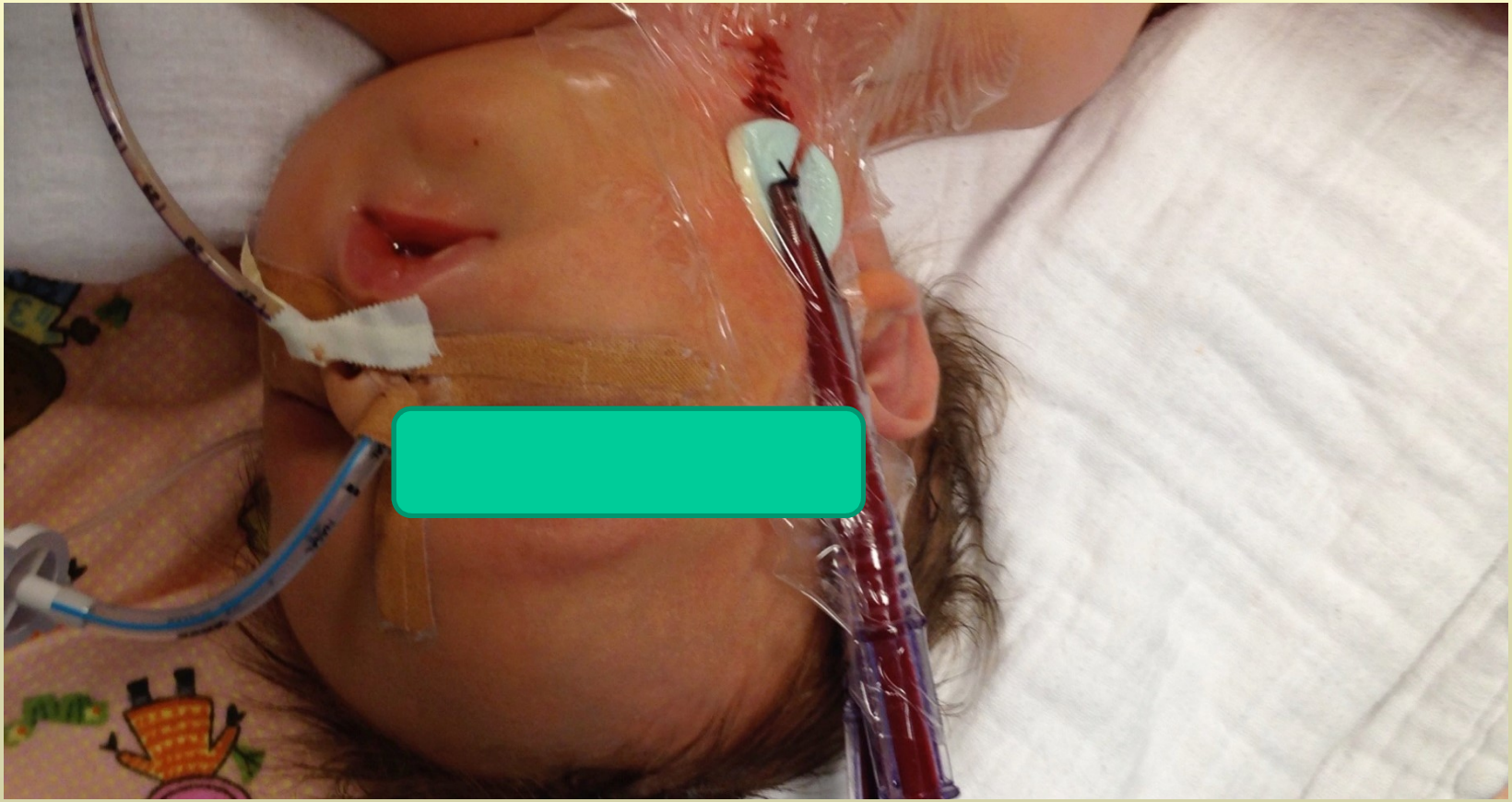
Case # 24

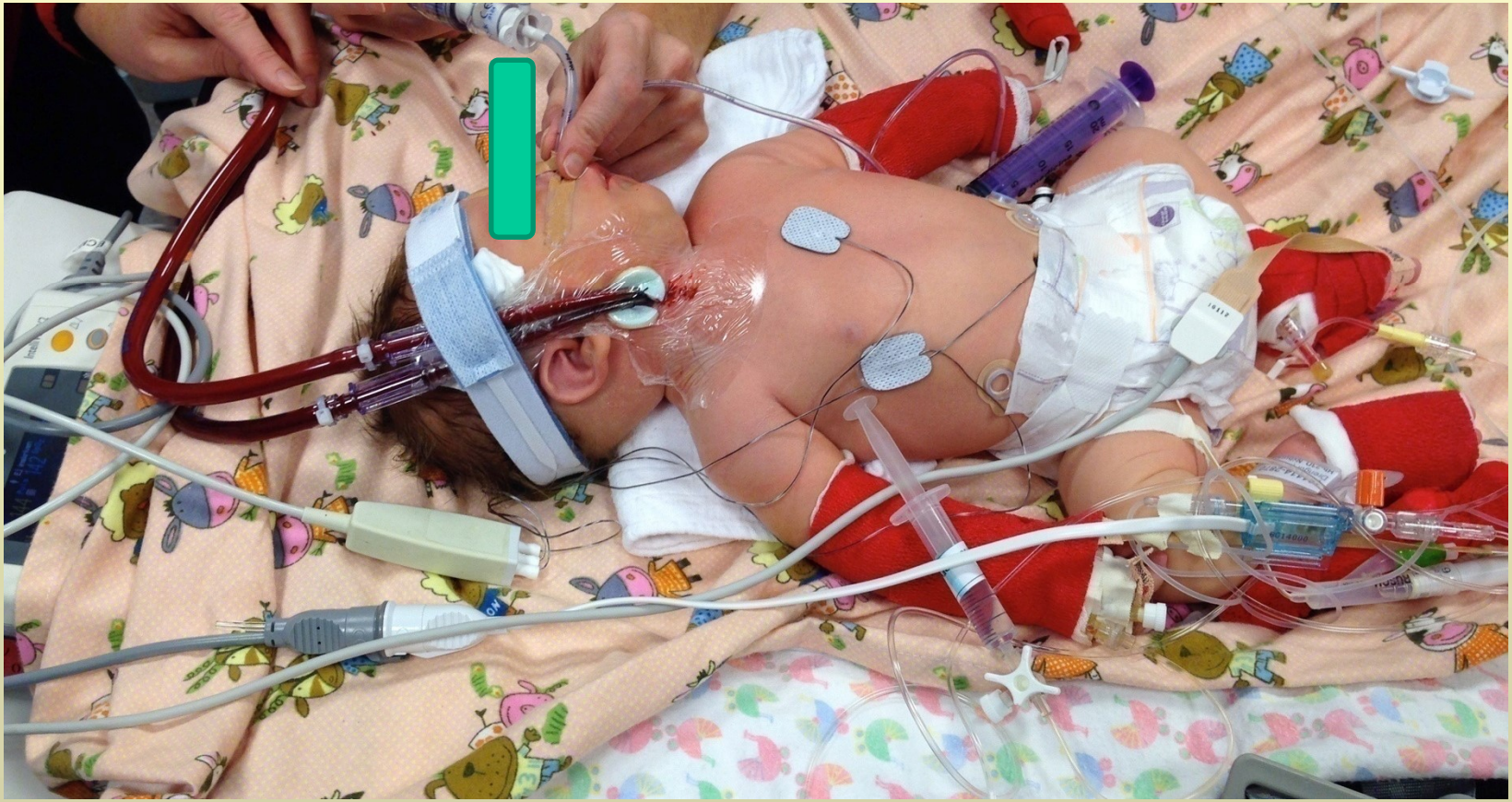










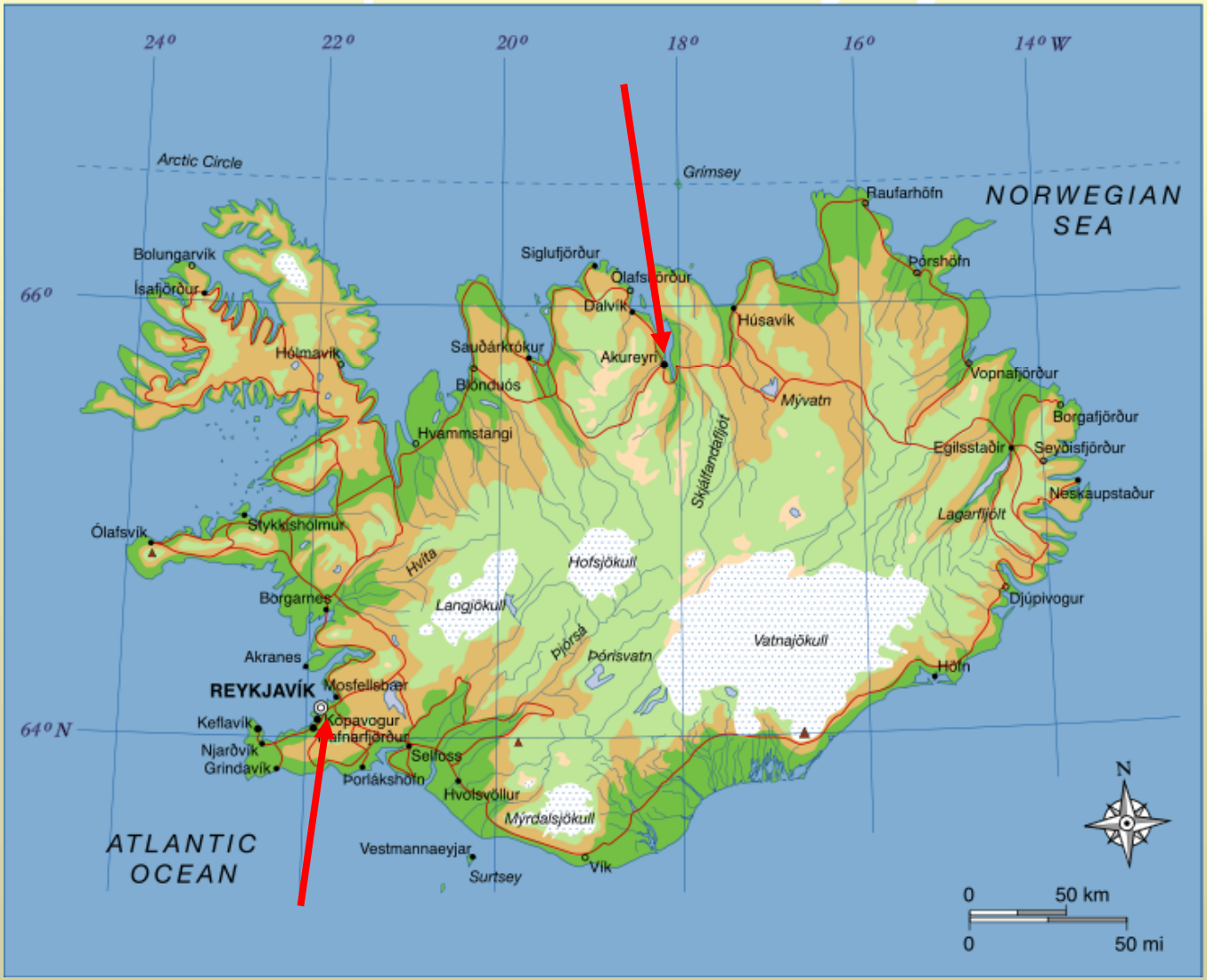


Case # 25, tilfelli, 12 ára stúlka

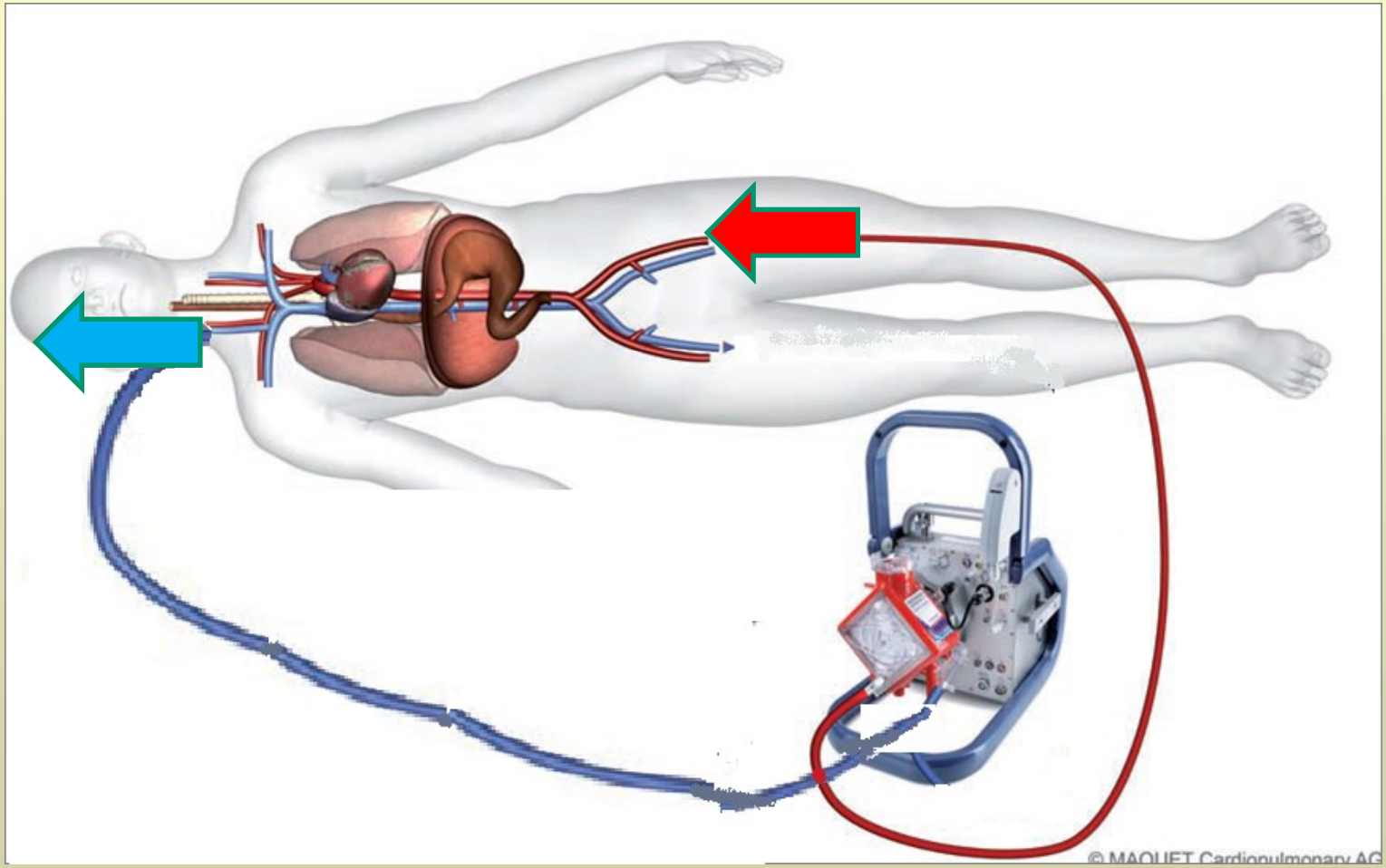
- Kransæðastífla hjartastopp
- FSA – Landspítali Hringbraut

Akuna vi Nov ember 24th

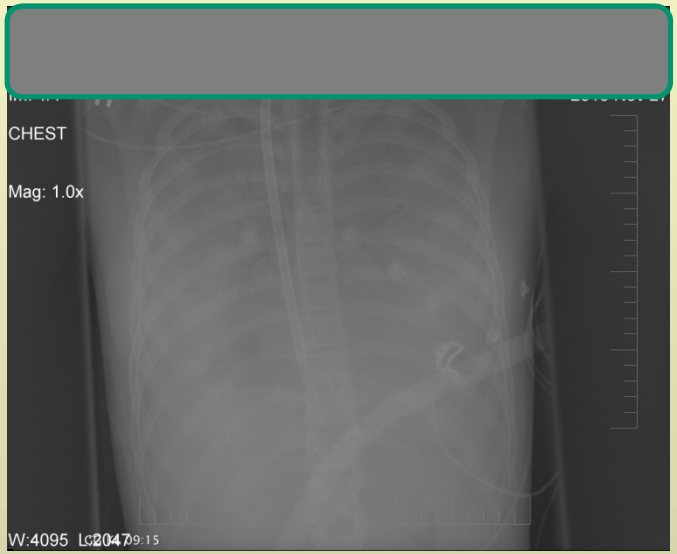
Transport to Reykjavik



V-A

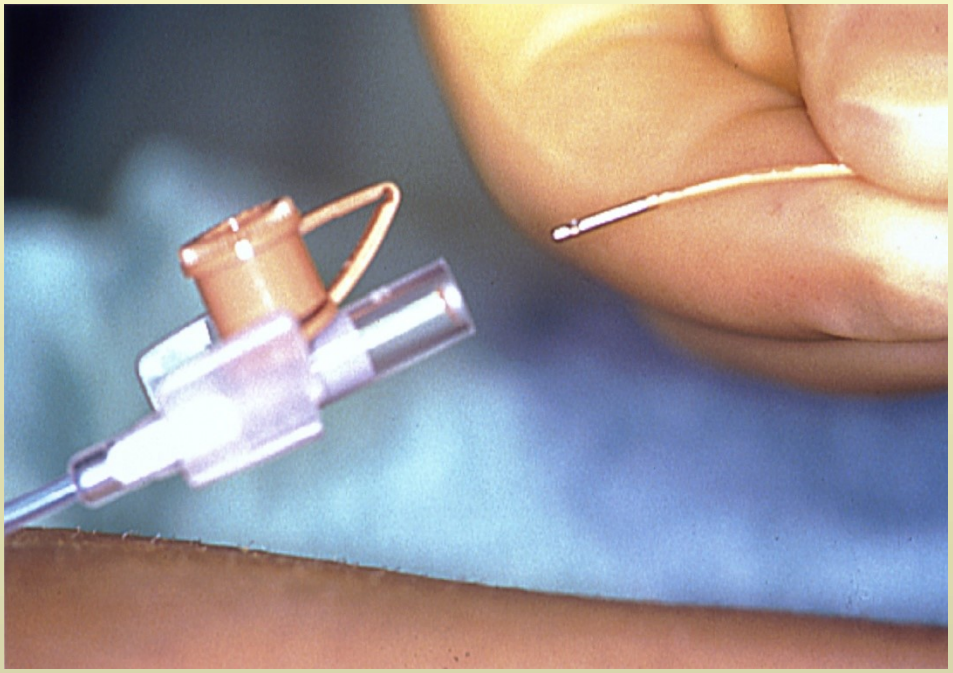


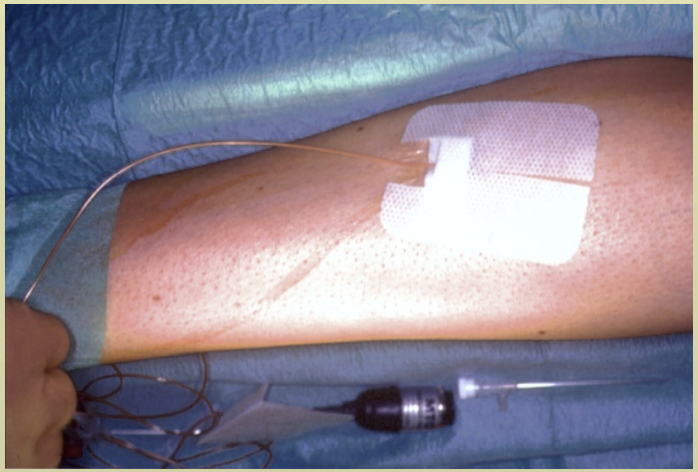
ECMO




PRESSURE DISPLAY BOX - 96000
Medtronic
MAQUET
26 11 2010

Compartment syndrome



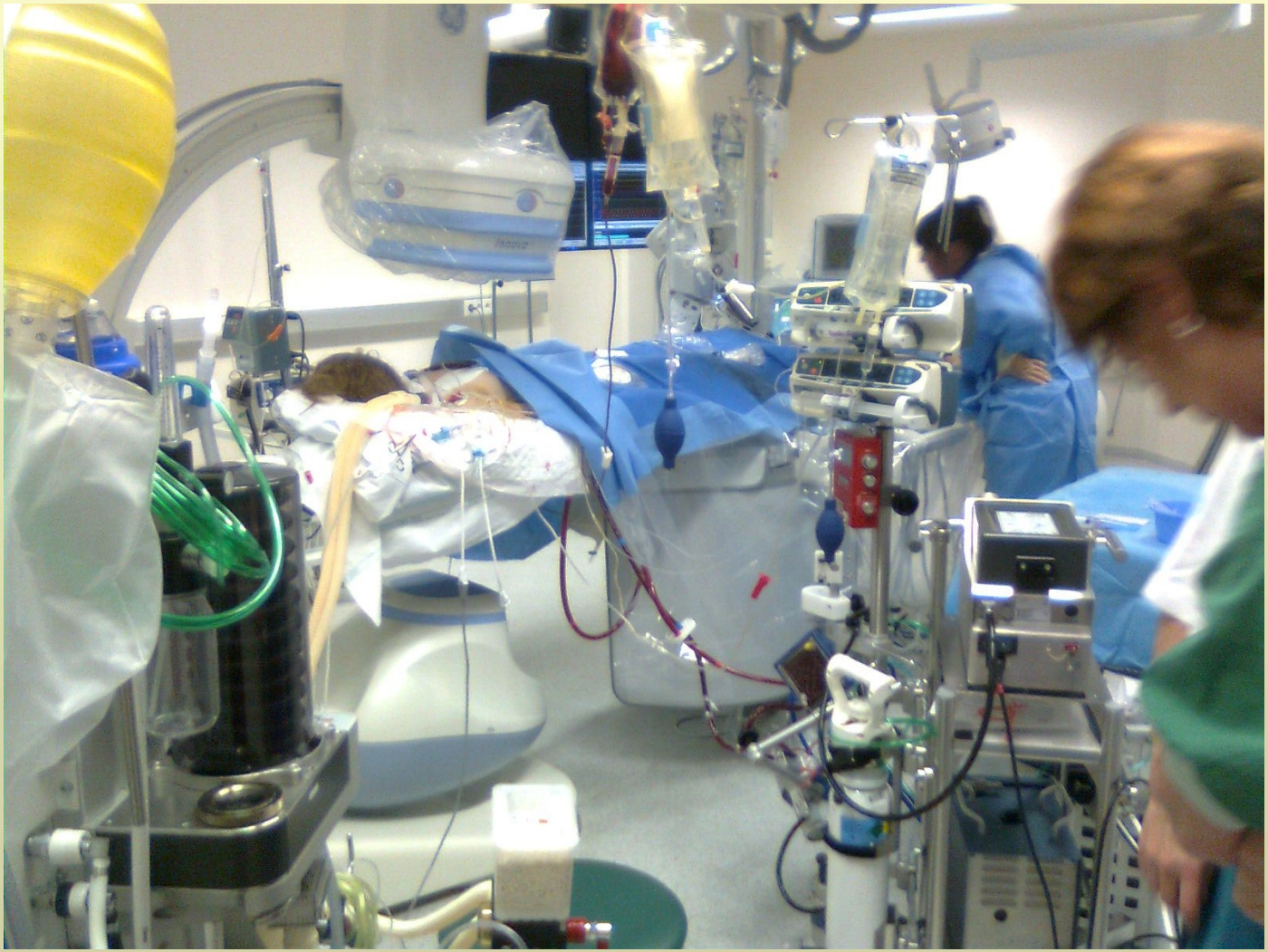


MTC® micro transducer

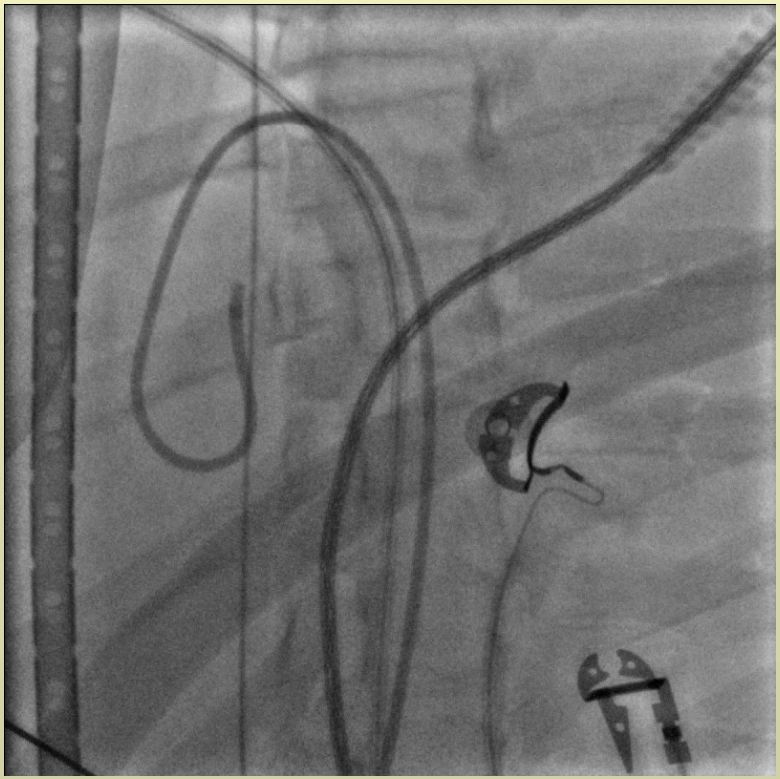
Fasciotomy, ECMO day 1

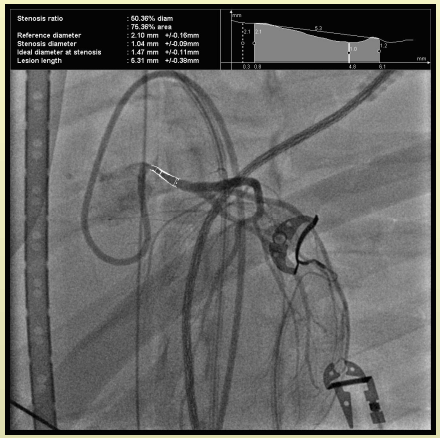
Fasciotomy: compartments of the lower leg

The Cath Lab ECMO day 2



Stenting left main

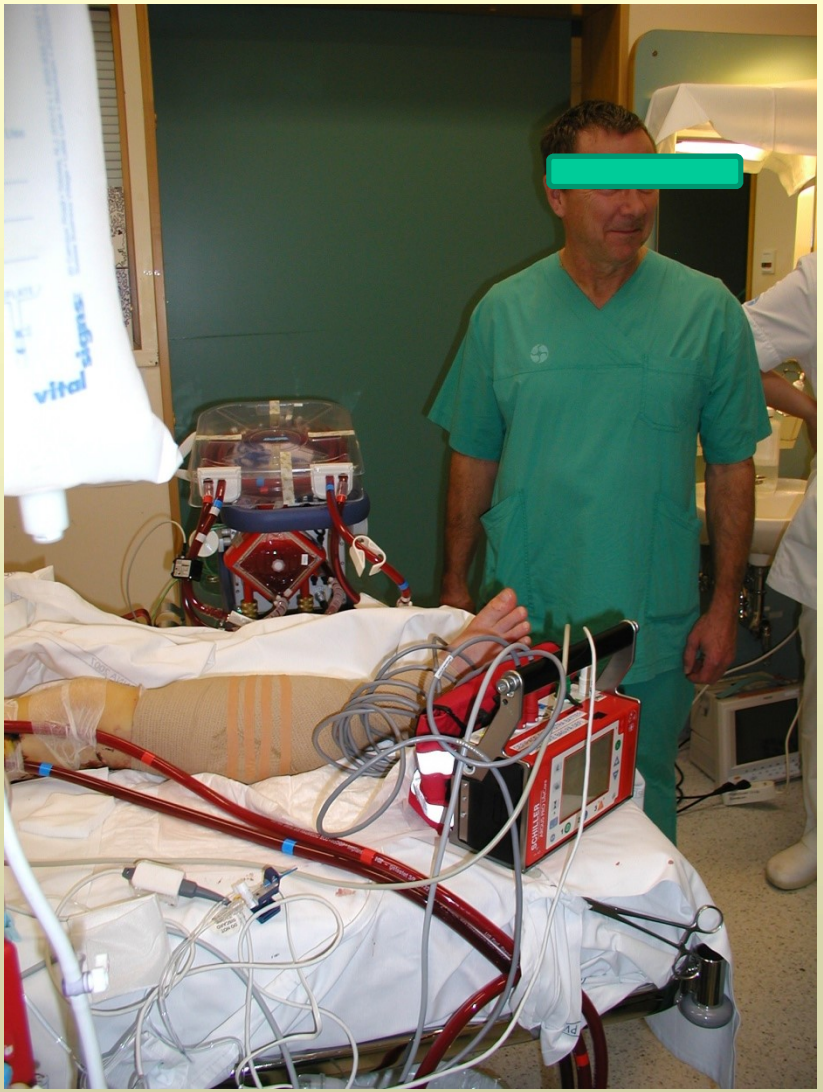




ECMO day 3 (27/11)

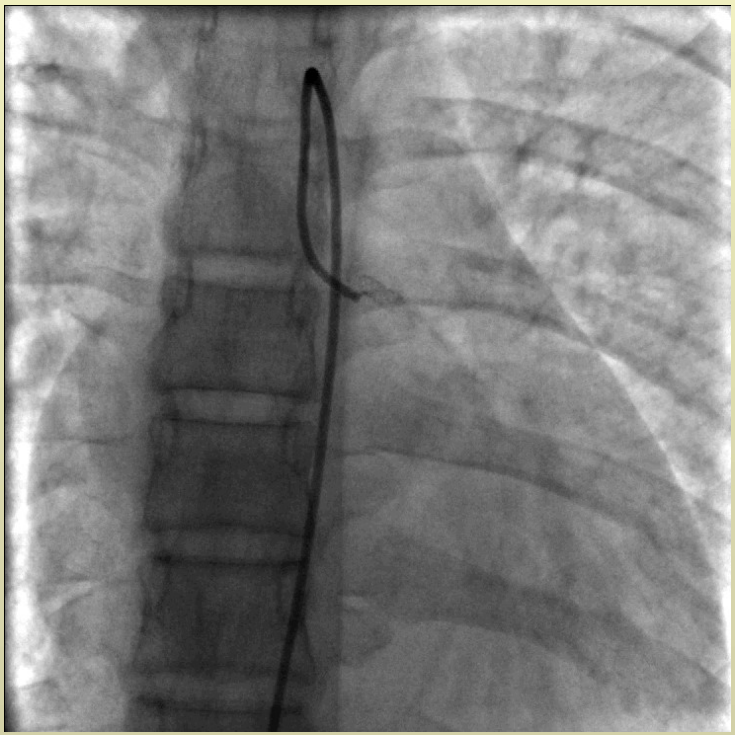
Reperfusion

In the lower leg


vital signs

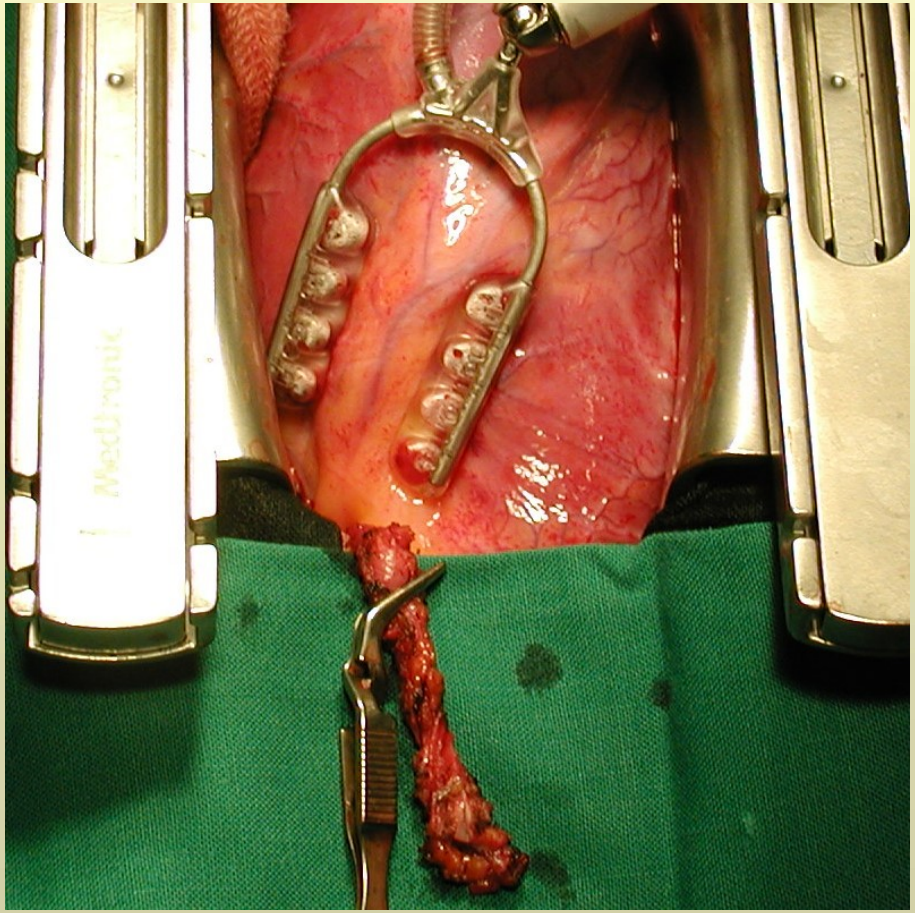

14
112
RABÍLL
112

Cardiac follow up at 3 months post PTCA : “in stent stenosis”

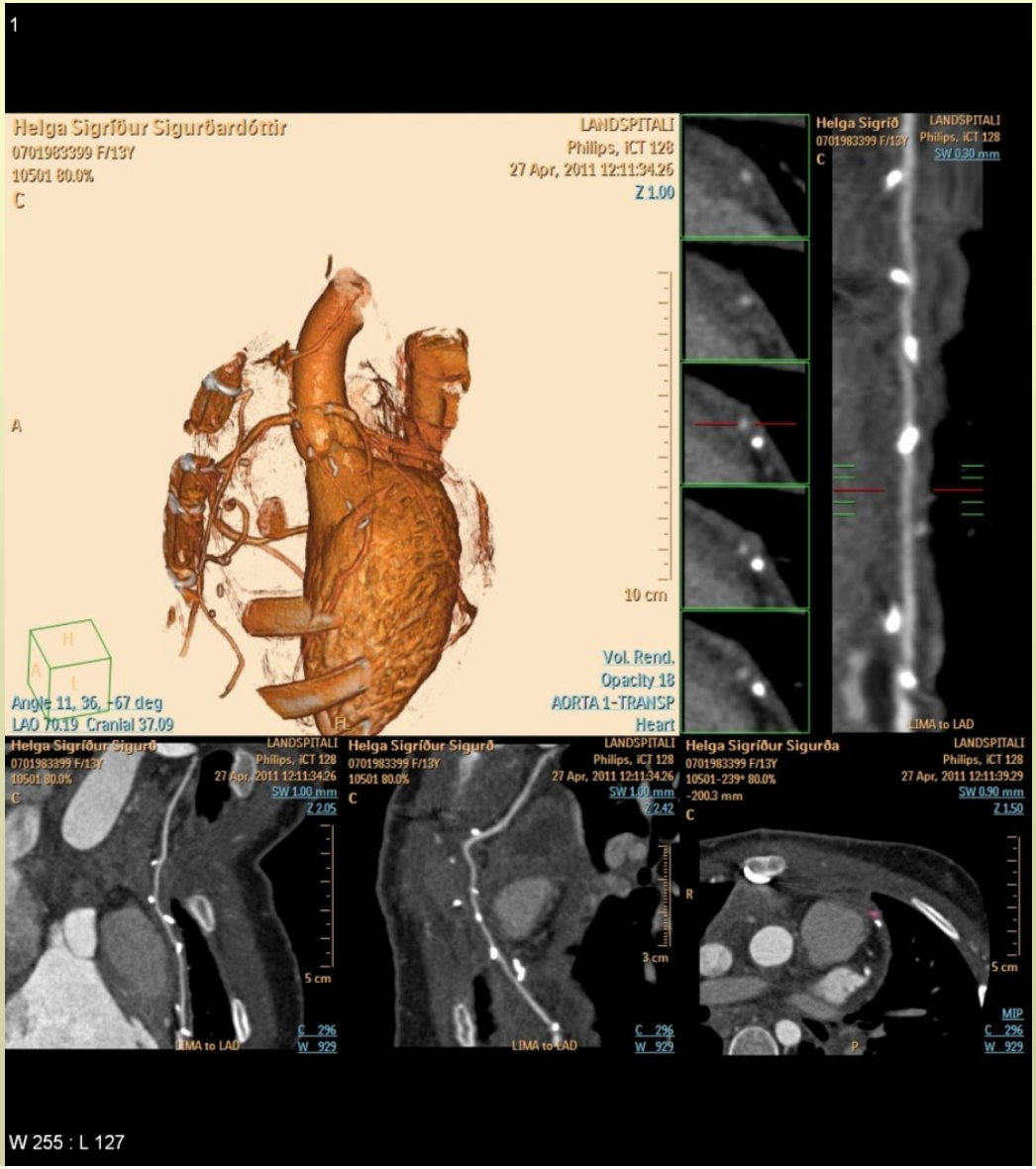


- Healed fashiotomies

OPCAB (LIMA to LAD):
Off Pump Coronary Artery Bypass



LIMA to LAD open on postoperative coronary CT angiography




GYMNOVA

- ECMO in this case showed to be a safe bridge to further treatment, a coronary bypass grafting in a child with a congenital life threatening coronary anomaly

Takk

References

- Text
  - Rogers: Textbook of Pediatric Intensive Care
  - Critical Cardiac Disease of Infants and Children
- On-line
  - Picubook.net
  - Pedi-heart web-site
  - http://www.cincinnatichildrens.org/health/heart-encyclopedia/default.htm

Surgical Terminology in Adult Congenital Heart Disease

Surgical Shunts in CHD

Shunt	Procedure
Classic Blalock-Taussig	Subclavian artery to pulmonary artery as end to side anastomosis.
Modified Blalock-Taussig	Gore-Tex tube graft from subclavian artery to pulmonary artery.
Bidirectional Glenn	SVC to RPA. End to side. Second stage to fontan.
Fontan Procedure	IVC to RPA to complete systemic venous circulation to PAs. Gore-Tex tube that may be in the heart (lateral tunnel) or outside the heart (extracardiac).
Waterson	Ascending aorta to RPA. A punch hole between the vessels.
Potts	Descending aorta to LPA. Punch hole between the vessels.

Surgical Procedures

Procedure	Surgery
Rastelli Procedure	Transposition with a VSD whereby the VSD is closed baffling LV to Ao and a RV to PA conduit is placed.
Norwood Procedure	Hypoplastic LV syndrome. The pulmonary valve and artery are used to create neo-aorta and a BT shunt is placed to provide pulmonary blood flow.
Mustard Procedure	Transposition of the great vessels. Baffle the SVC/IVC to the MV - LV -- PA and baffle the pulmonary veins to TV -- RV -- Ao.
Arterial Switch	TGV where the aorta and MPA are switched and coronary arteries re-implanted.

Congenital Heart Disease. ACCSAP V