CARIS

Congenital Anomaly Register and Information Service

25th Annual Meeting 2023

virtual session

Tuesday 28 November 2023

12.00 - 14.00

GIG CYMRU NHS WALES

Iechyd Cyhoeddus Cymru

Public Health Wales

Welcome

Dr Margery Morgan

CARIS Lead Clinician/Consultant Obstetrician,
Singleton Hospital, Swansea

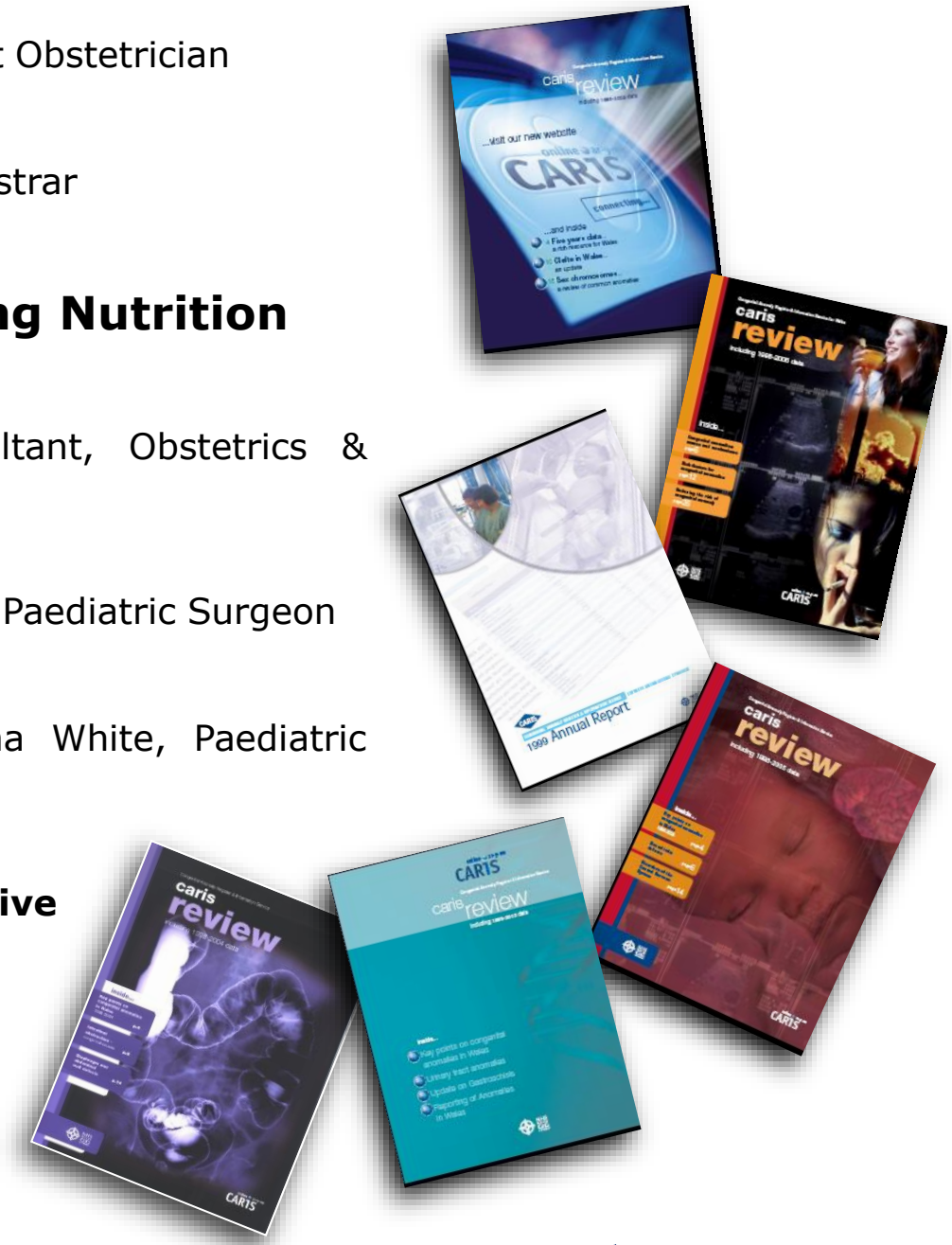
So you have the best experience possible:

- Your microphone should be muted, and your camera turned off
- There may be time for questions after each presentation. Please post yours in the chat bar
- The session will be recorded and will be available on the CARIS website after the event

- **12:00 Welcome** – Margery Morgan, CARIS Lead Clinician, Consultant Obstetrician
- **12:10 CARIS update** – Dr Penelope Cresswell-Jones, Speciality Registrar

Focus session: Abdominal Abnormalities affecting Nutrition

- **12:20 Ultrasound Diagnostic tips**– Armin Vandeperre, Consultant, Obstetrics & Gynaecology
- **12:50 Surgical Challenges** – Oliver Jackson, Consultant Neonatal & Paediatric Surgeon
- **13:15 Post operative nutrition** – Rebecca Seymour and Emma White, Paediatric Advanced Nurse Practitioners
- **13:40 Tracheo-oesophageal fistula (TOF) – a parent's perspective** – Naomi Webborn
- **13:55 Conclusion –** Dr Margery Morgan
- **14:00 Close**



CARIS update

Dr Penelope Cresswell-Jones

Speciality Registrar, Public Health Wales

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CARIS Team Update

Dr Penelope Cresswell-Jones
Specialty Registrar in Public Health

On behalf of Dr **Llion Davies, Consultant in Public Health**

Official Statistics Update: 1998-2022

- Congenital Anomalies
  - >39,000 cases registered
  - 4.8% of all births
  - 84.7% liveborn, with 96.9% surviving to 1 year
- Childhood Rare Diseases
  - >24,000 registered cases
  - >1,250 diseases
- Antenatal Detection rates

CARIS Team Achievements

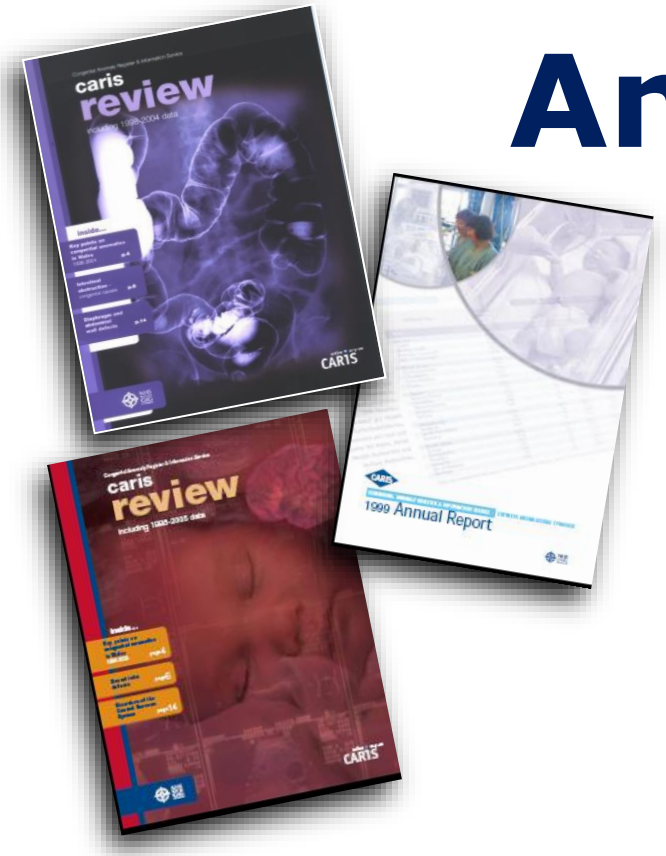
- QI project - (Almost) paper free
- Registries discovery
- Accessibility standards
- Office for Statistics Regulation review
- Data expansion (Syphilis and HIV)

Adult Rare Diseases

- New data officer in December 2023
- Sarcoidosis work progressing
- CARIS team part of data sub-group of the RDIG
- Co-production beginning
- Data to inform action – LHB level

CARIS

Congenital Anomaly Register and Information Service



Annual Meeting 2023

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CARIS *Congenital Anomaly Register and Information Service*

Focus session: Abdominal Abnormalities affecting Nutrition

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Public Health Wales

Ultrasound diagnostic tips

Dr Armin Vandeperre

Consultant in Obstetrics and Fetal Medicine

Abdominal Anomalies affecting Nutrition: Ultrasound diagnostic tips

Dr Armin Vandeperre

Consultant in Obstetrics and Fetal Medicine

University Hospitals of Wales, Cardiff

GI and abdomen

Abdominal wall

- Physiological herniation
- Gastroschisis
- Omphalocele
- Pentalogy of Cantrell
- Body Stalk Anomaly
- Bladder/cloacal exstrophy

Other

- Ascites
- Echogenic bowel

GI obstruction

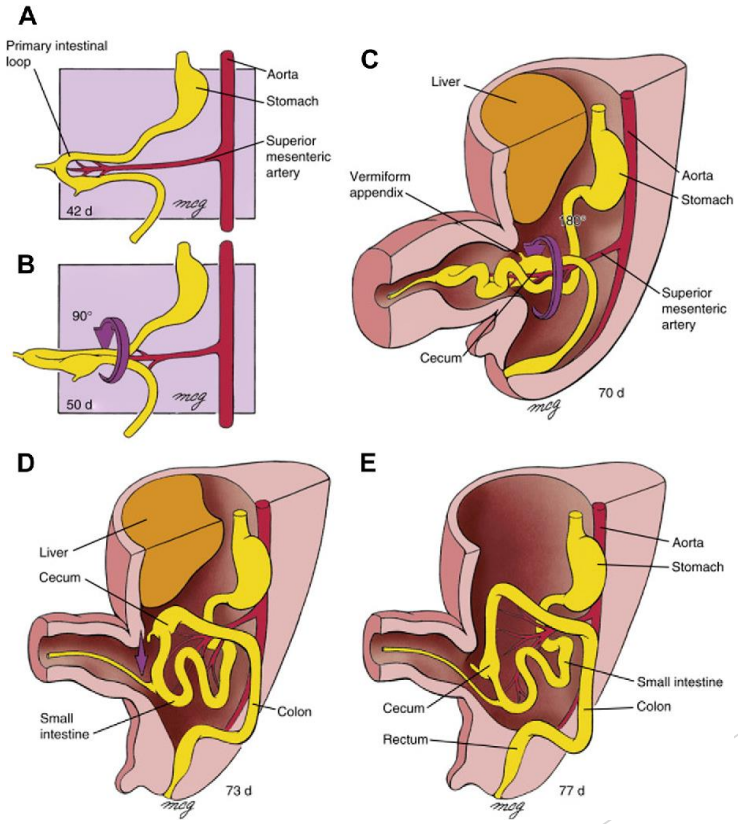
- Esophageal atresia
- Duodenal Atresia
- Jejunal, ileal atresia
- Colonic atresia
- Anal atresia
- Cloacal malformation
- Volvulus

Cyst/Mass

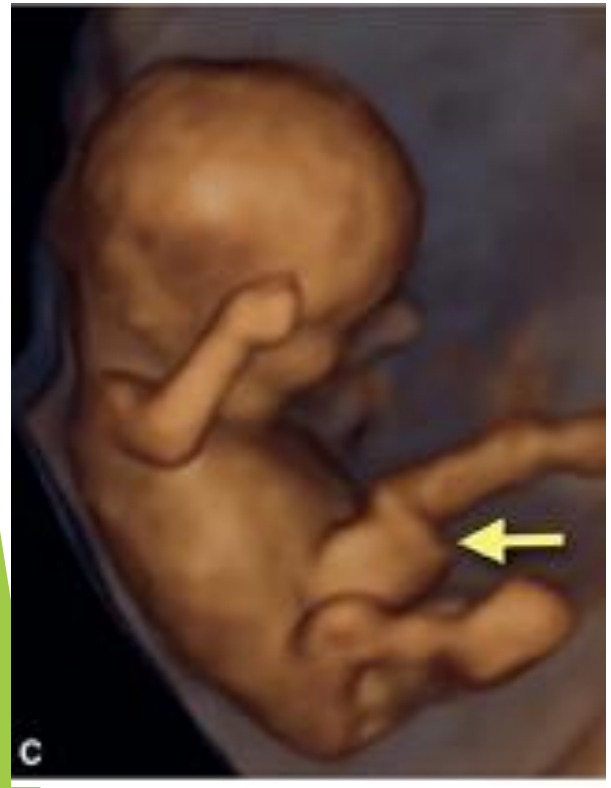
- Duplication Cyst
- Pseudocyst
- Lymphangioma
- Gallstones
- Choledochal cyst
- Hemangioma
- Hamartoma
- Malignant tumors
- Ovarian

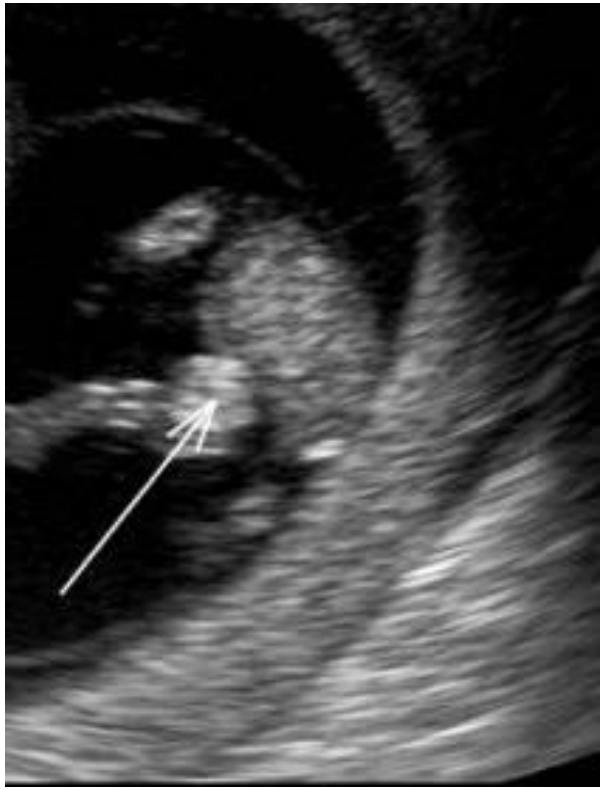
Physiological herniation

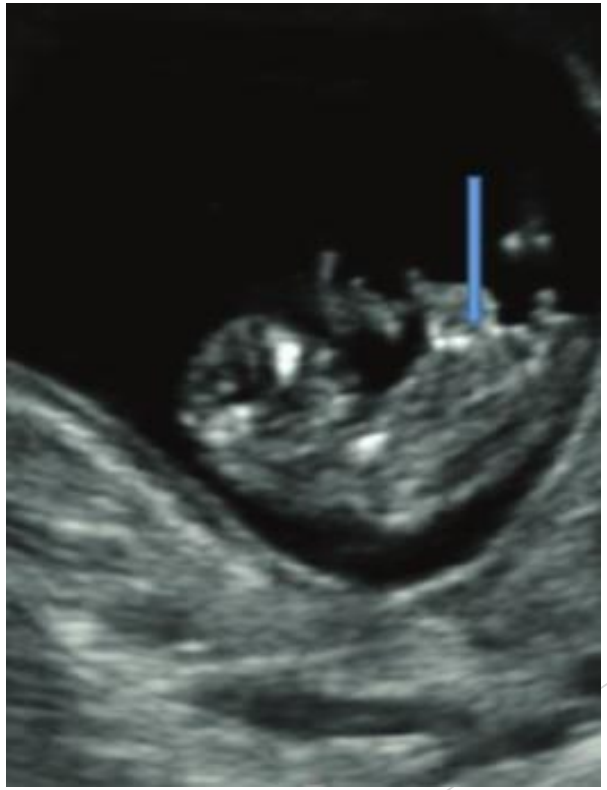
- Before 12 weeks
- temporary physiological midgut herniation.
- Differentiation from small Omphalocele

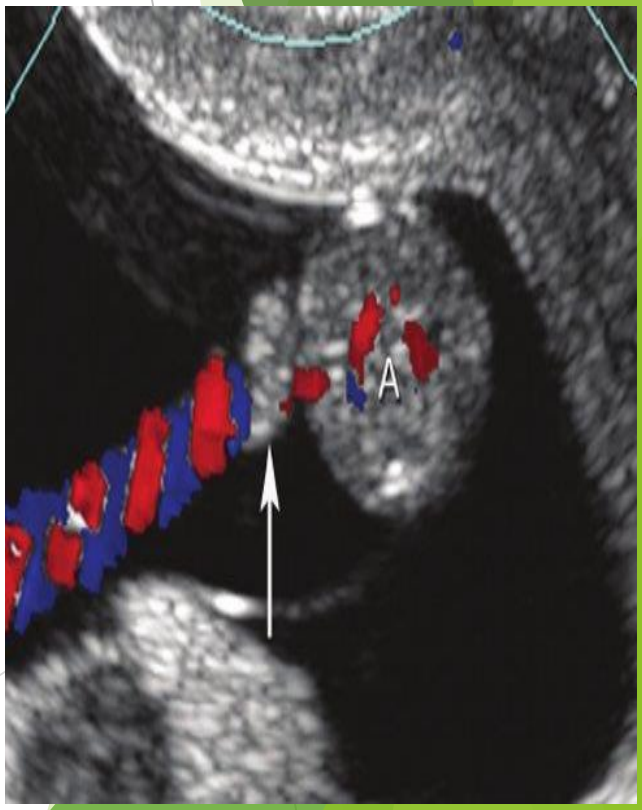


Physiological herniation: ultrasound



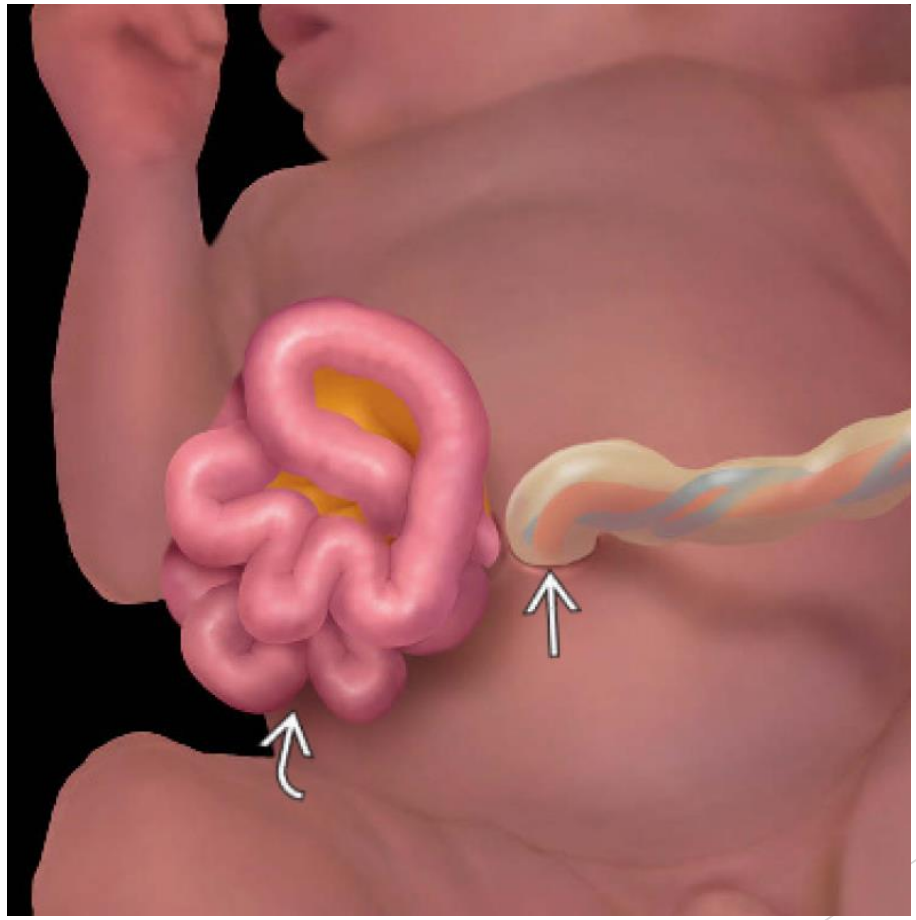




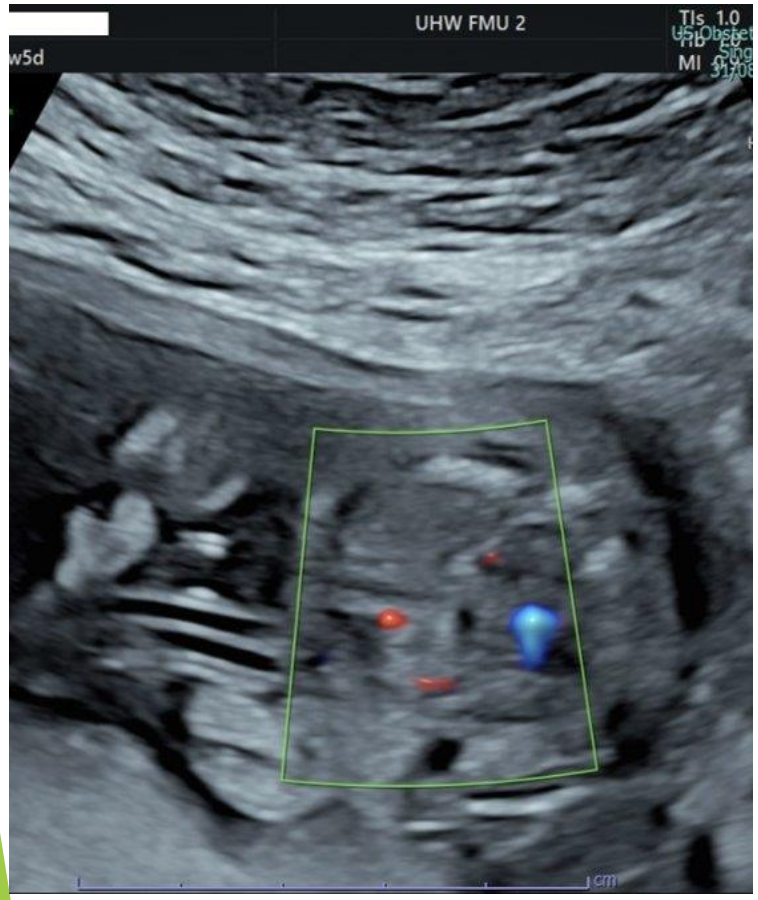


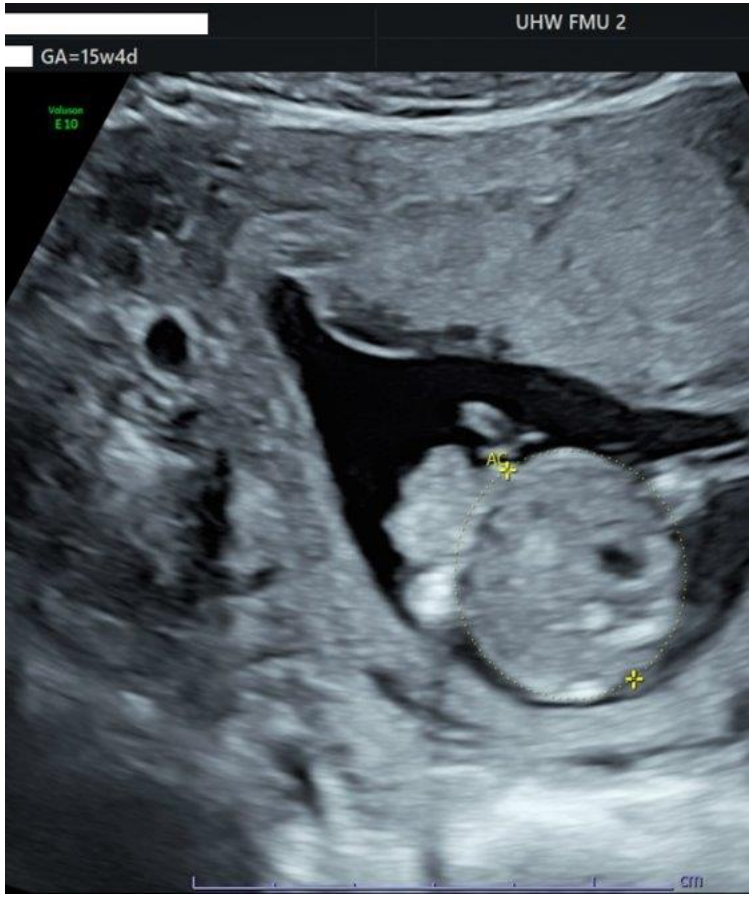
Gastroschisis

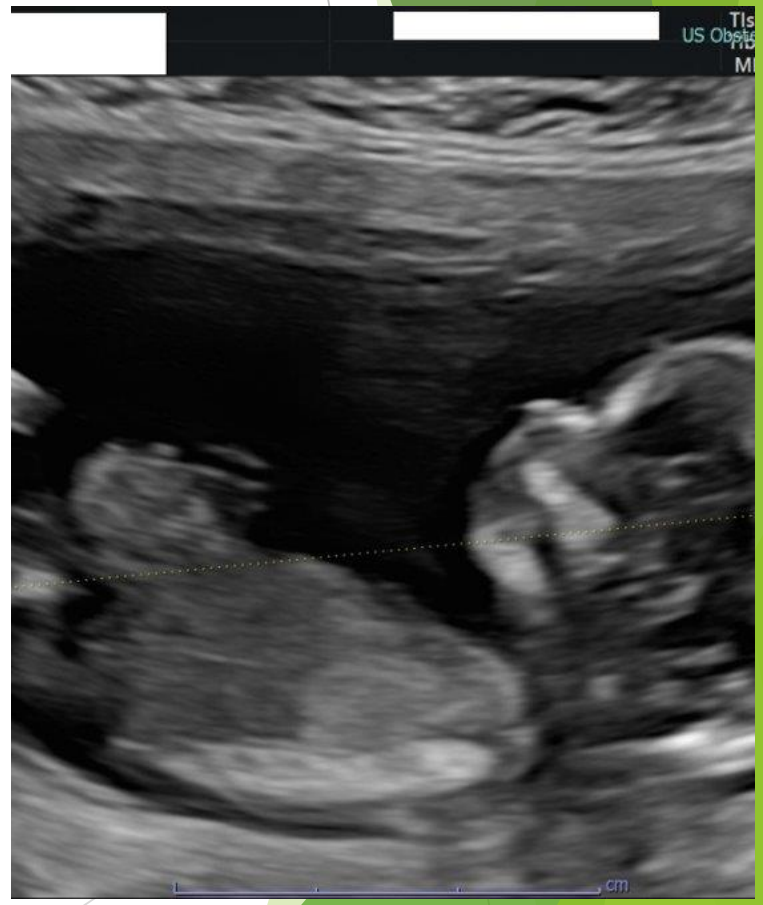
- Paramedian defect
- Almost always right sided
- Complex 12-15%:
  - Dilated bowel
  - Liver
  - Other anomalies
- Stillbirth 4-5%
- FGR 25%
- IABD >14mm
- No genetic association



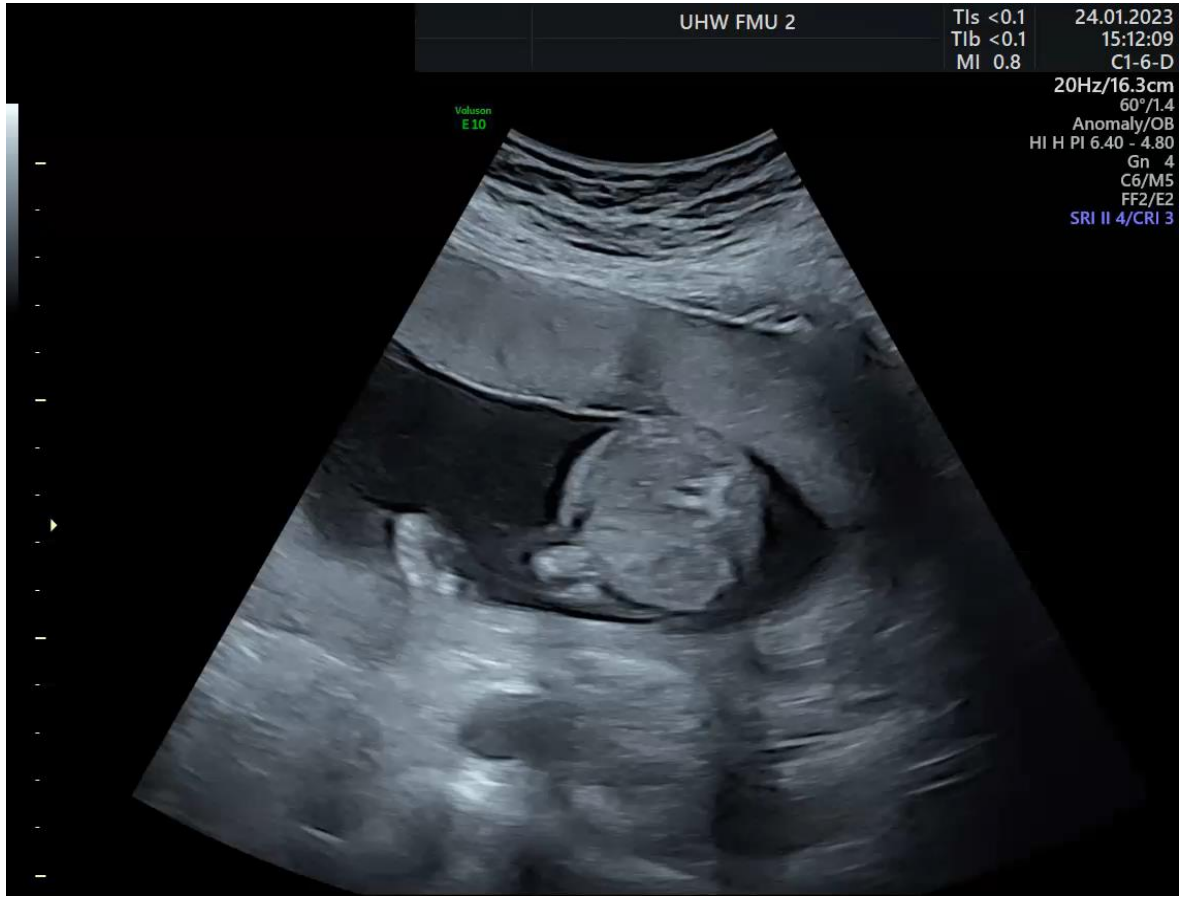
Gastroschisis: Ultrasound

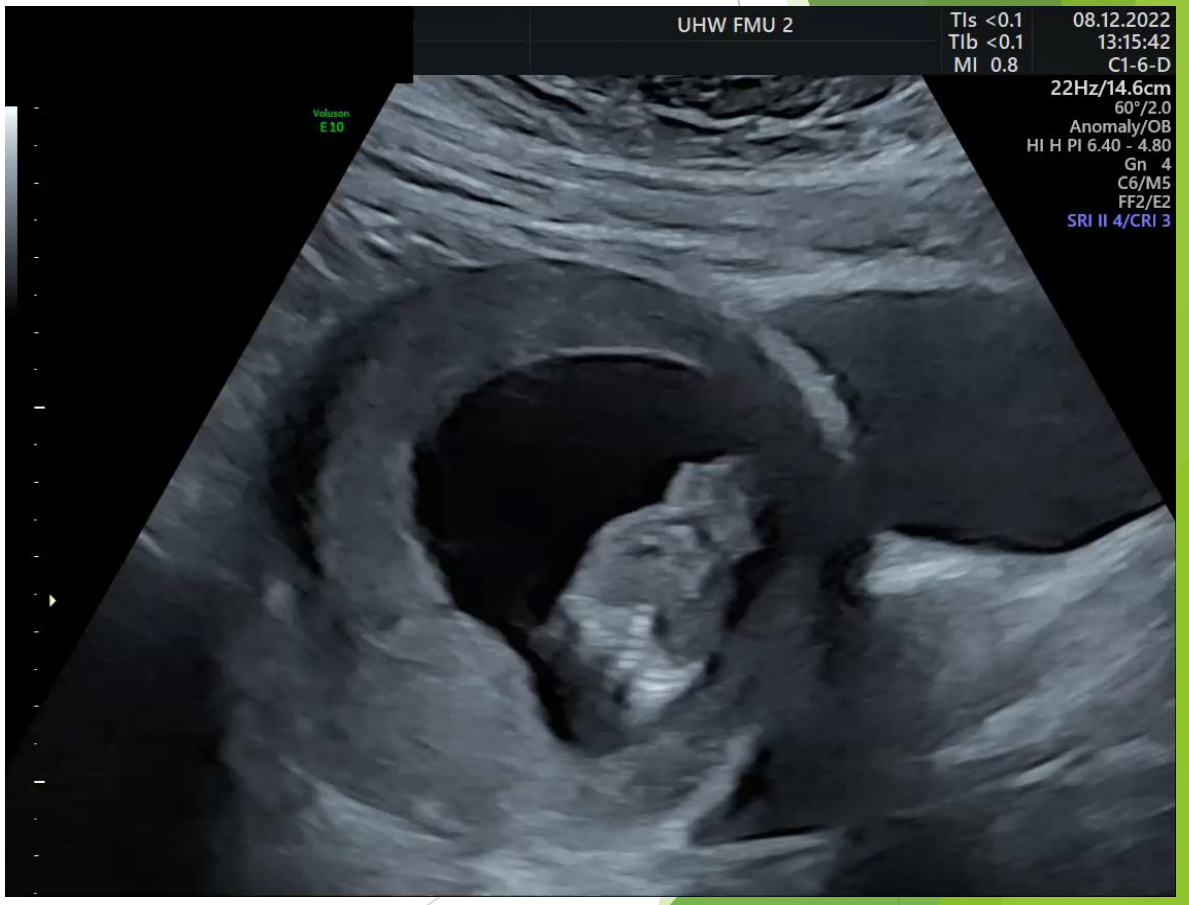




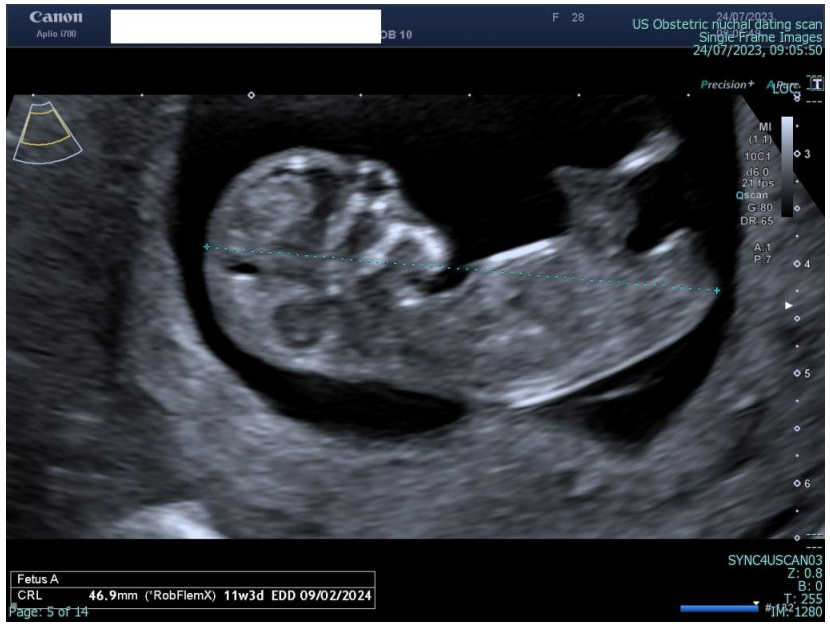


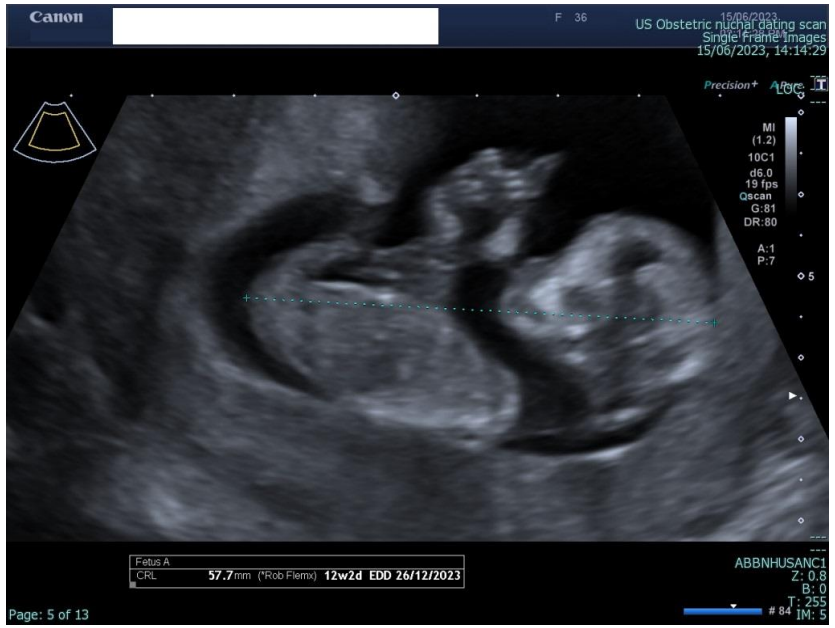
Gastroschisis: Ultrasound

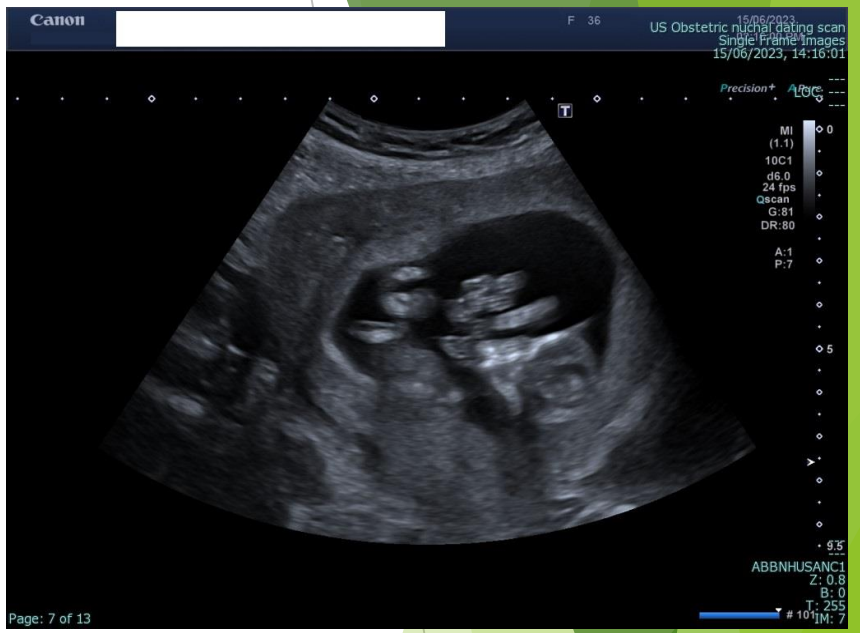


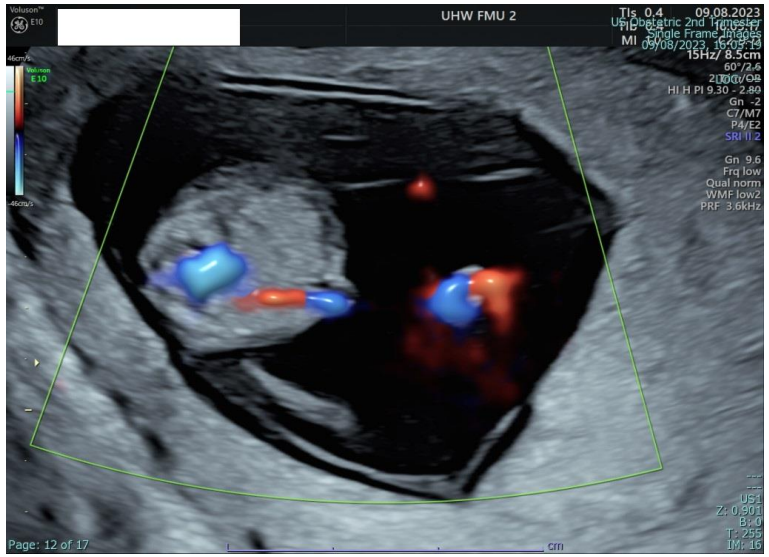


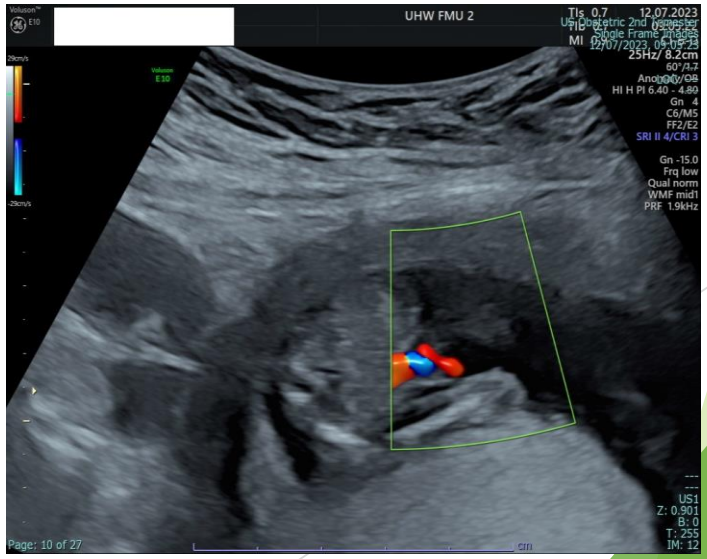
Gastroschisis: pitfalls





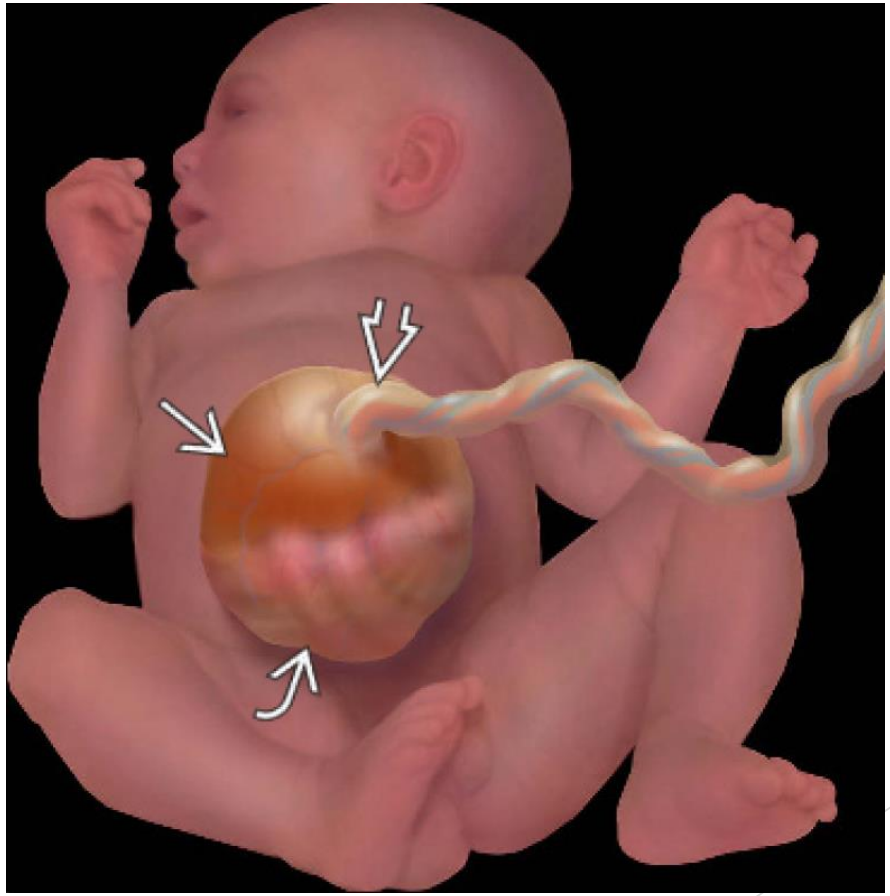




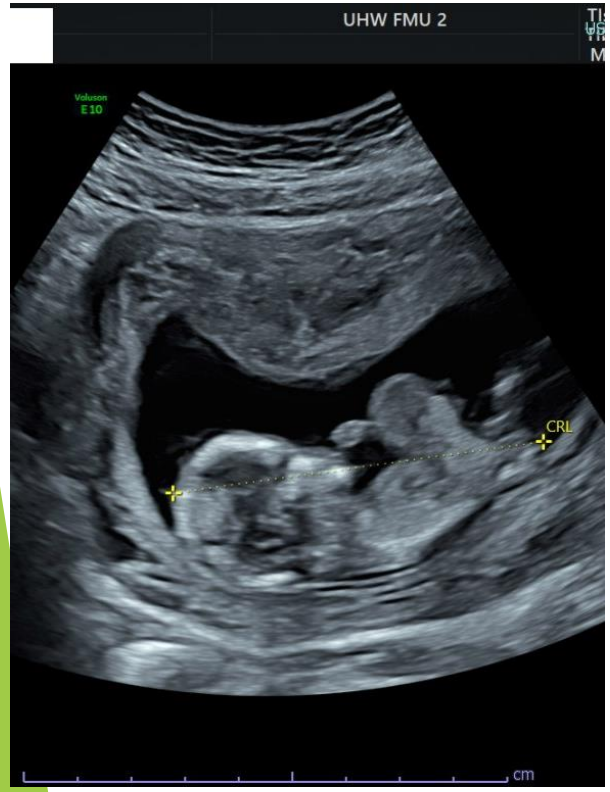


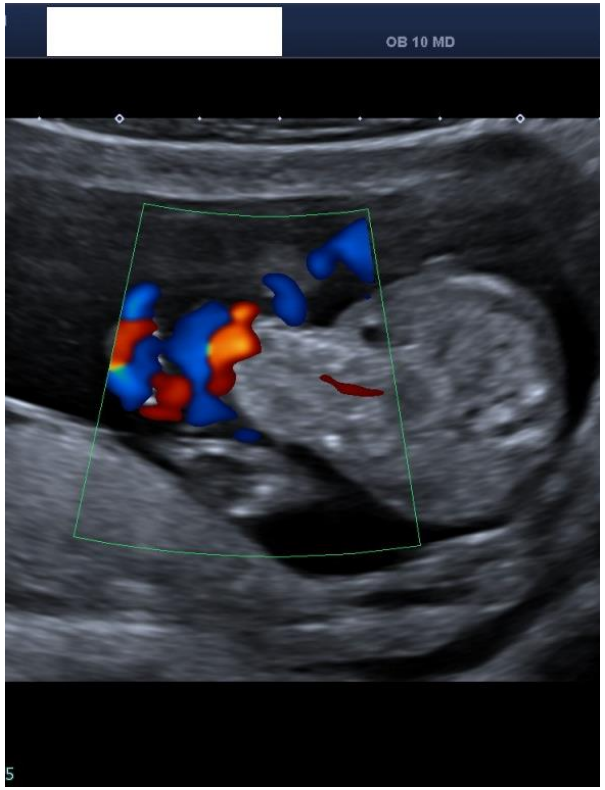
Omphalocele

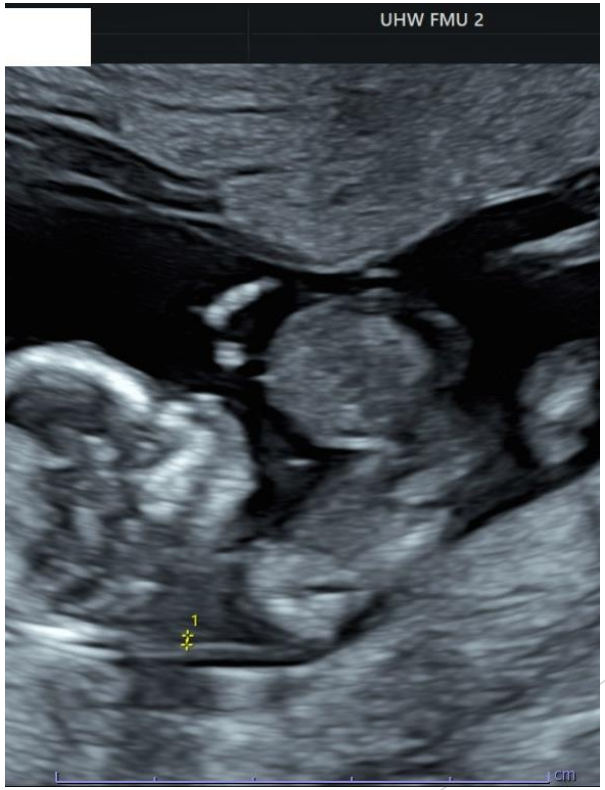
- Membrane covered
- Midline defect
- Umbilical cord inserts onto mass
- Chromosomal conditions 30-40%
- Associated anomalies:
  - GI
  - cardiac
- Giant >6cm

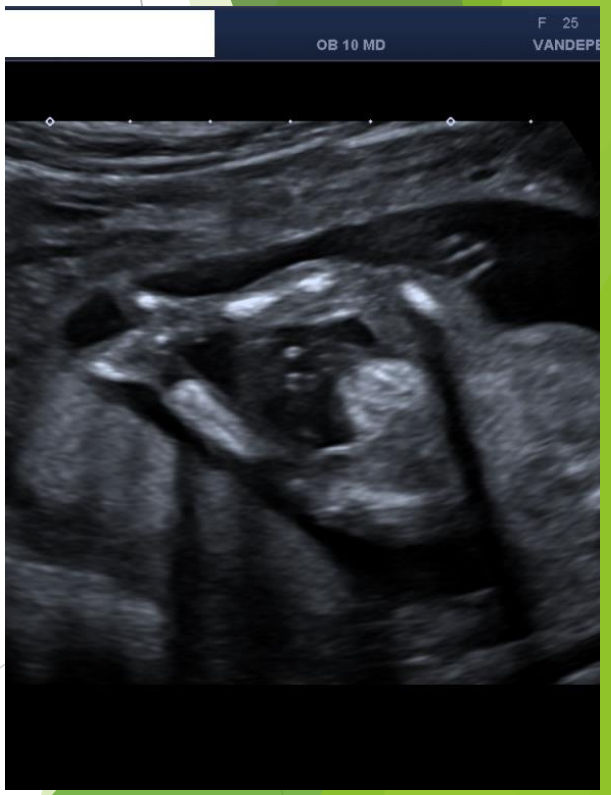


Omphalocele: ultrasound 1st Trim

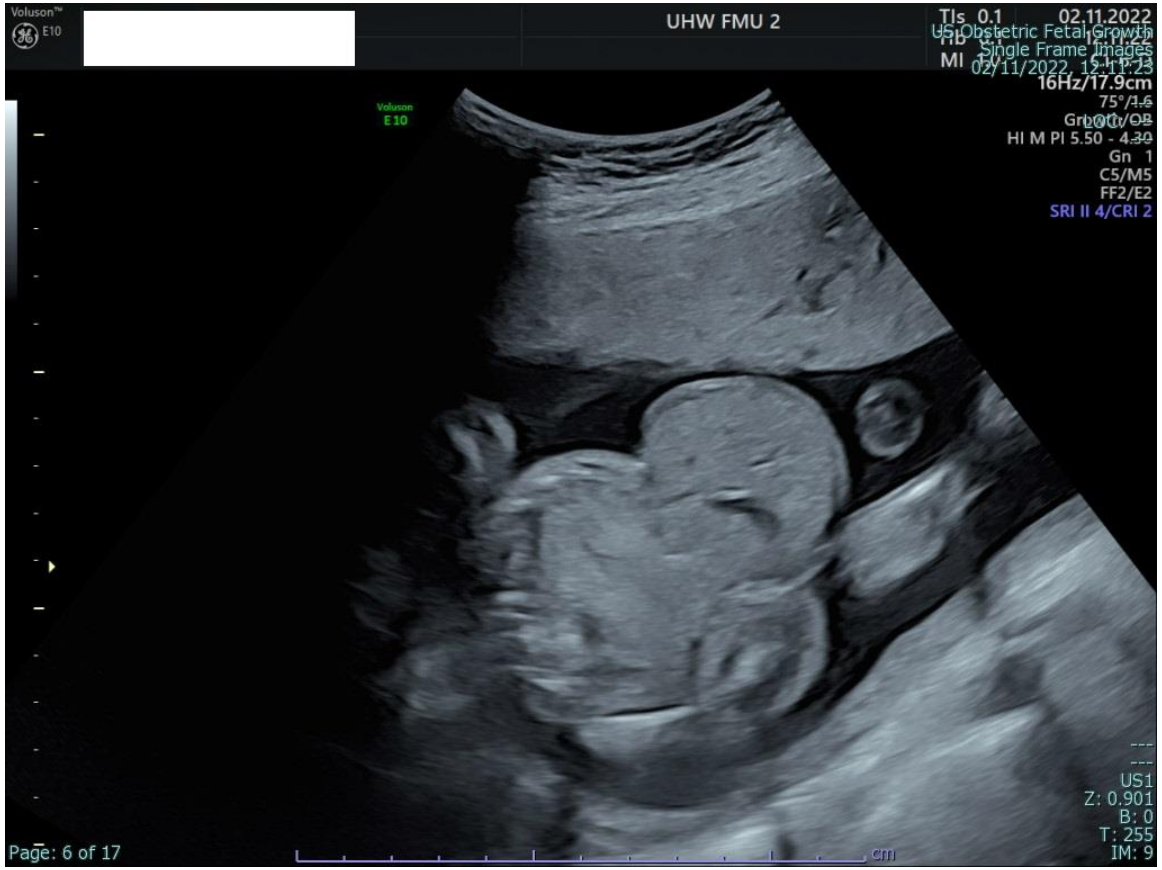


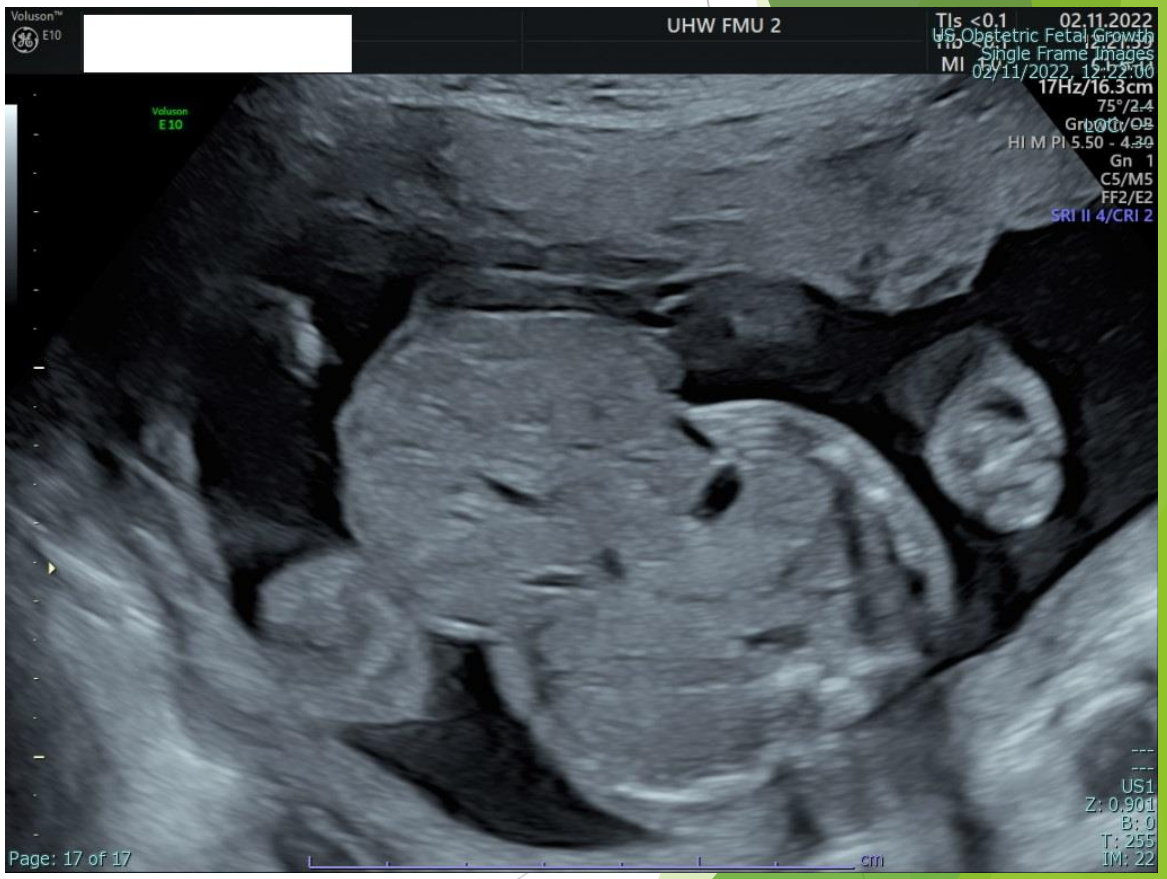






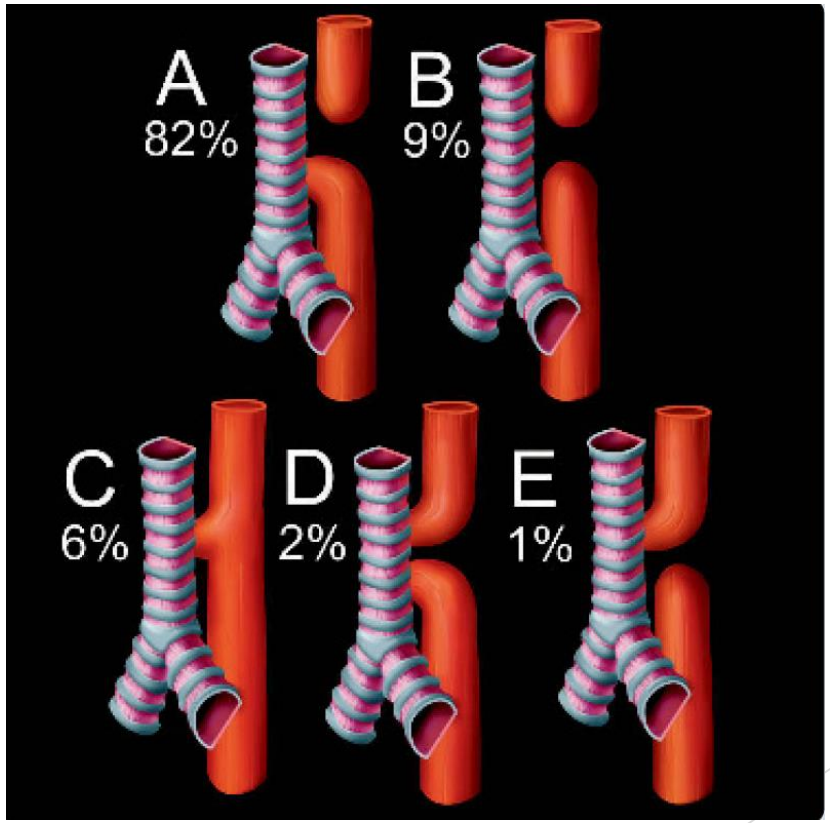
Omphalocele: ultrasound 2nd Trim



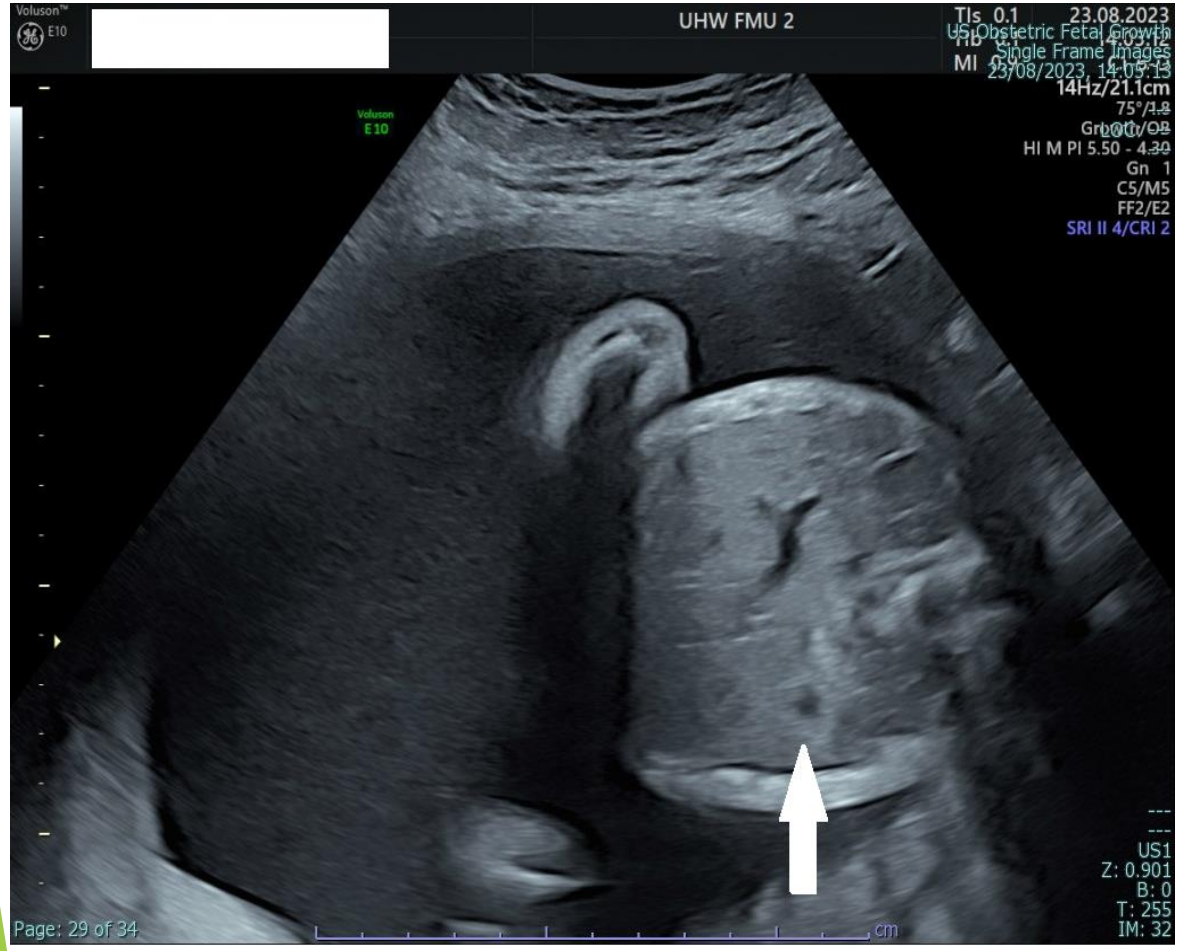


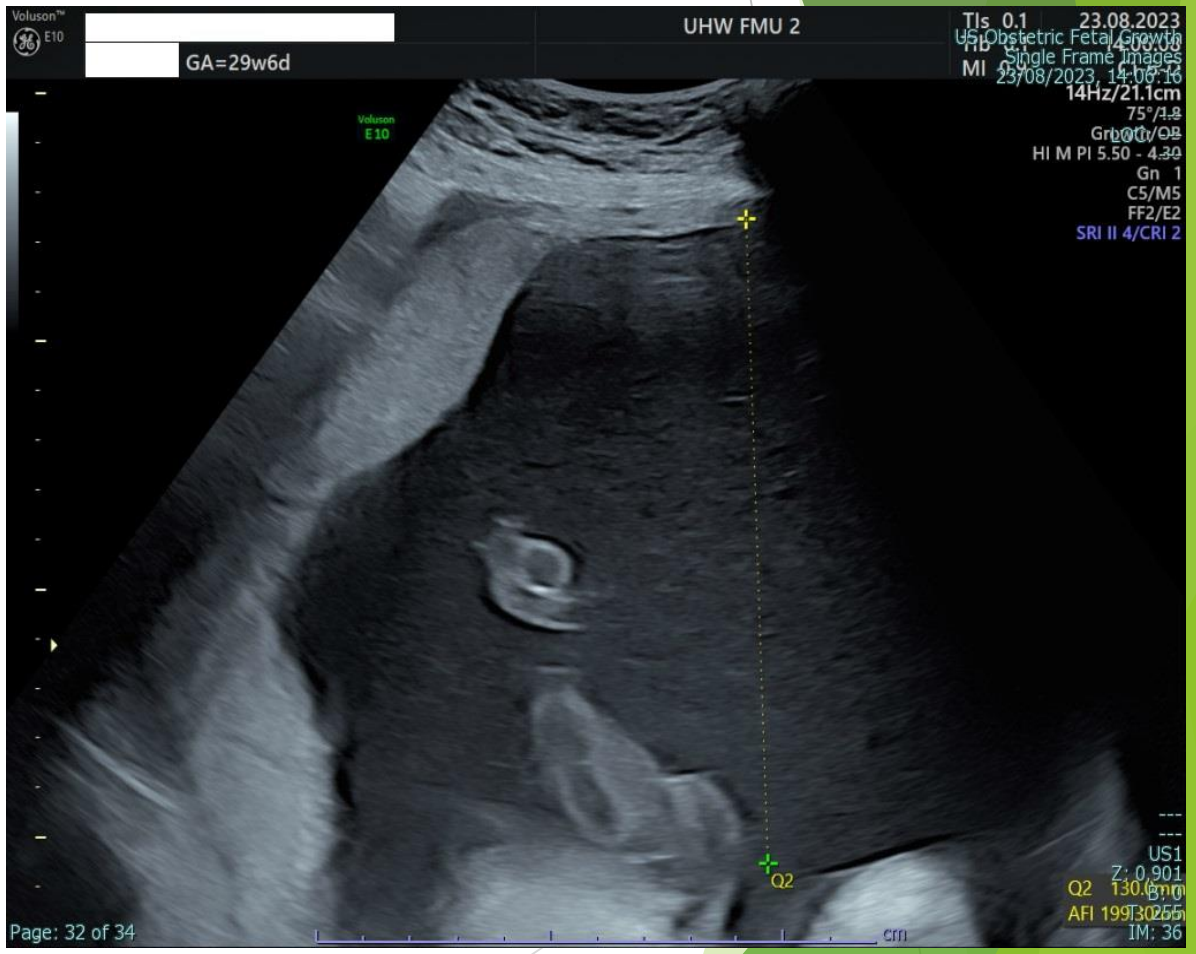
Esophageal atresia
Tracheoesophageal fistula

- Small or absent stomach
- > 90% fistula
- Pouch sign
- FGR 40%
- Polyhydramnios
- Associations:
  - Diabetes
  - VACTERL
  - Other anomalies >50%

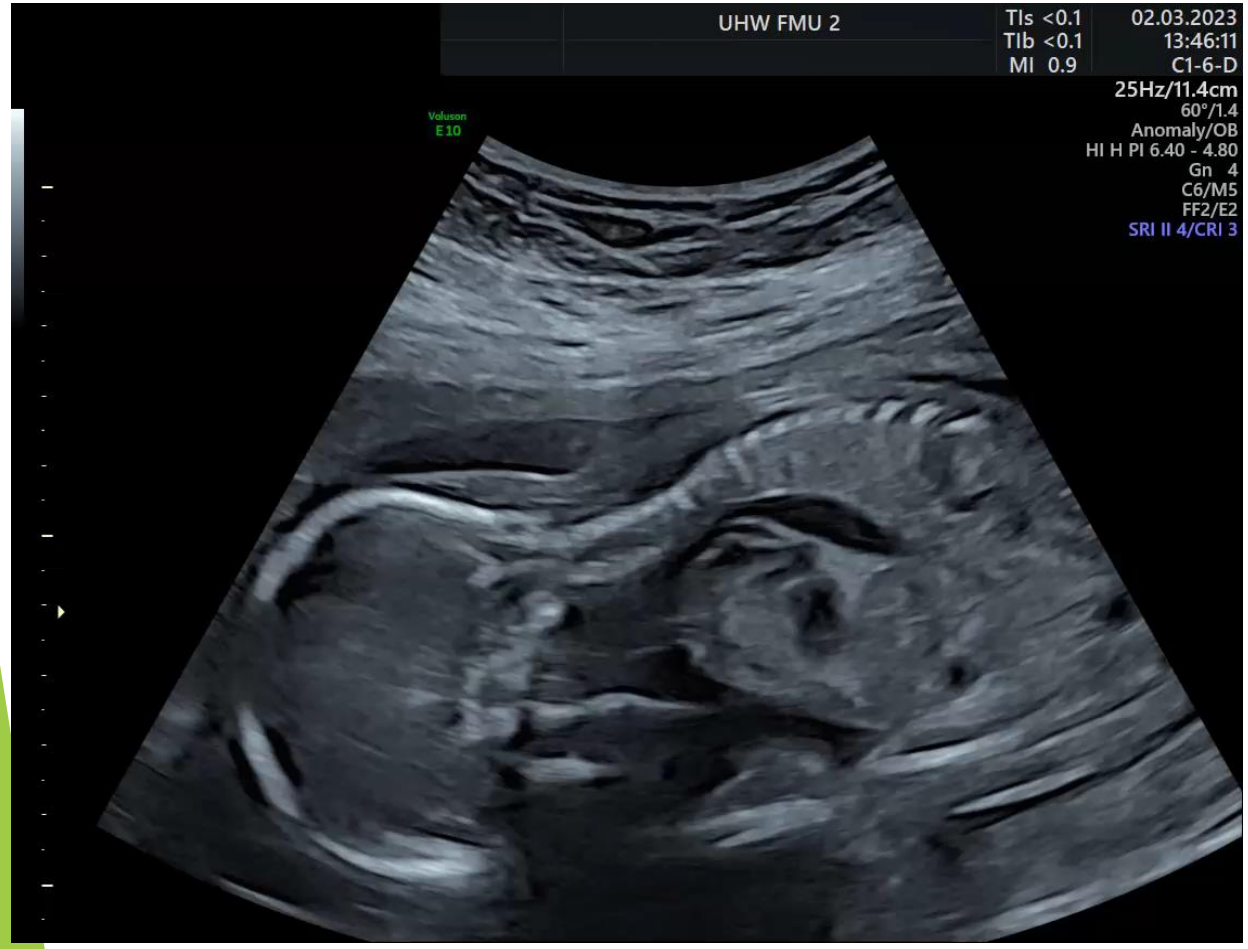


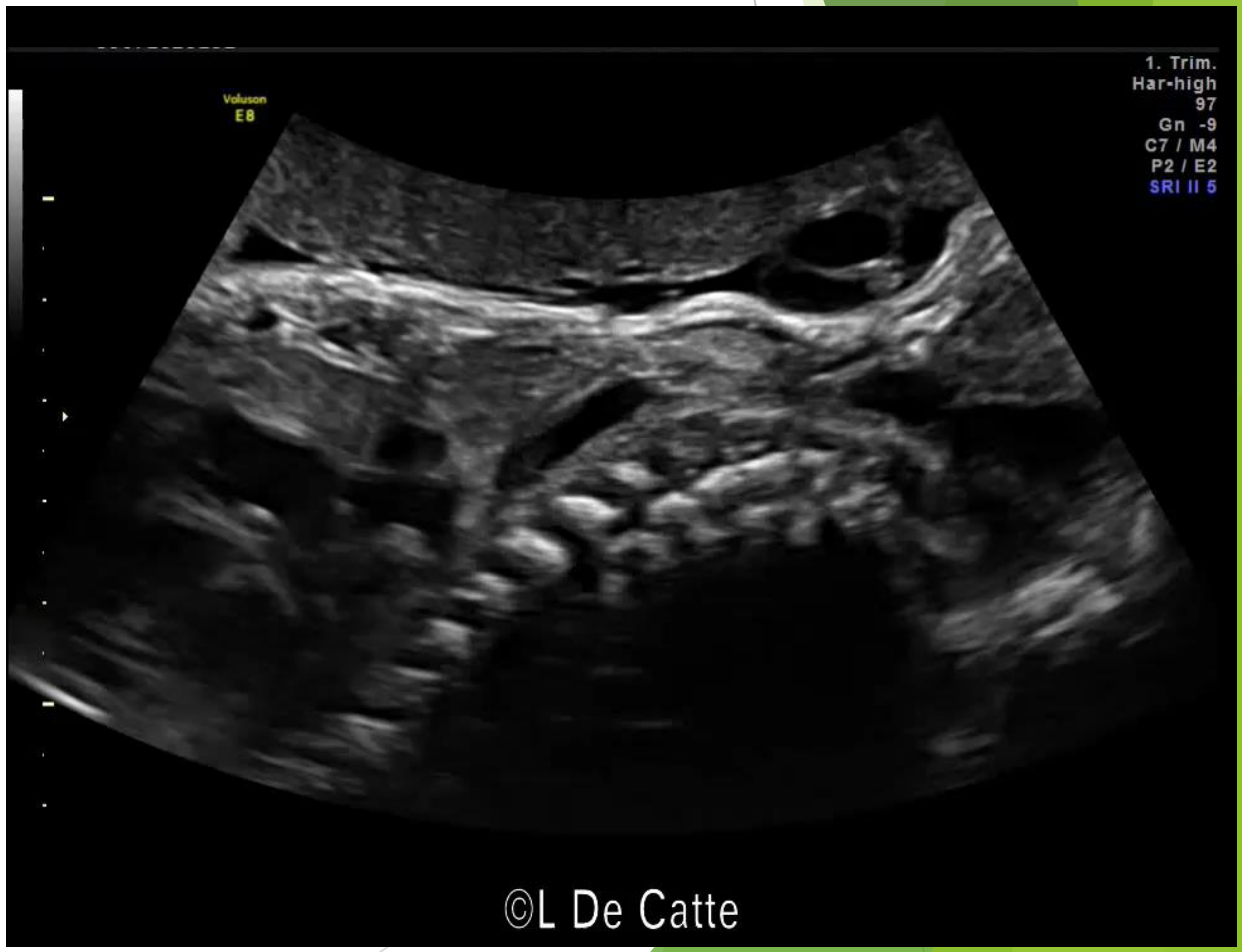
EA / TOF: ultrasound





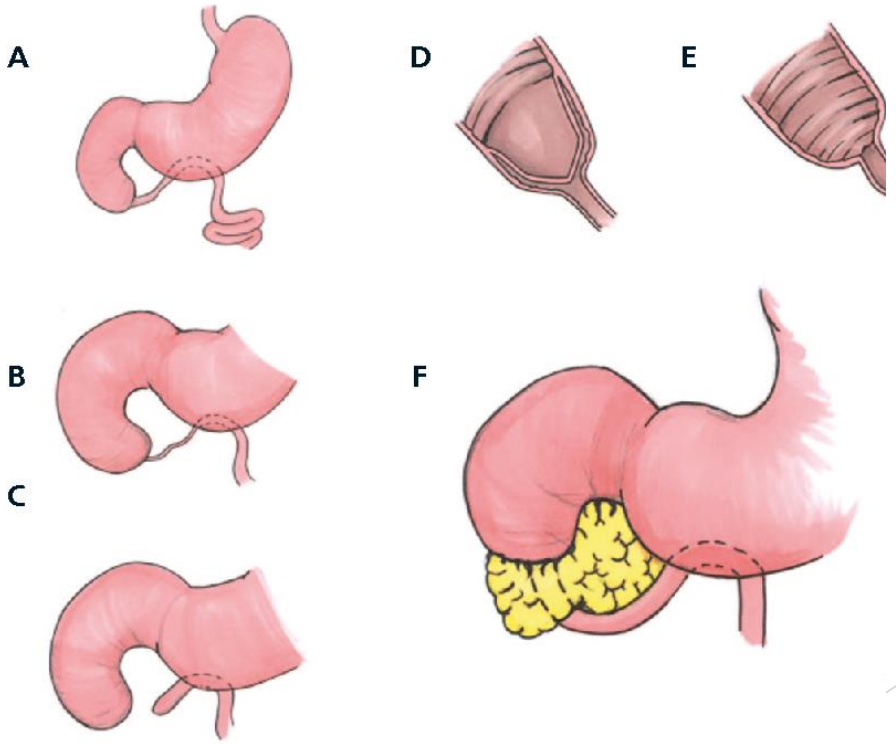
EA / TOF: ultrasound Pouch sign





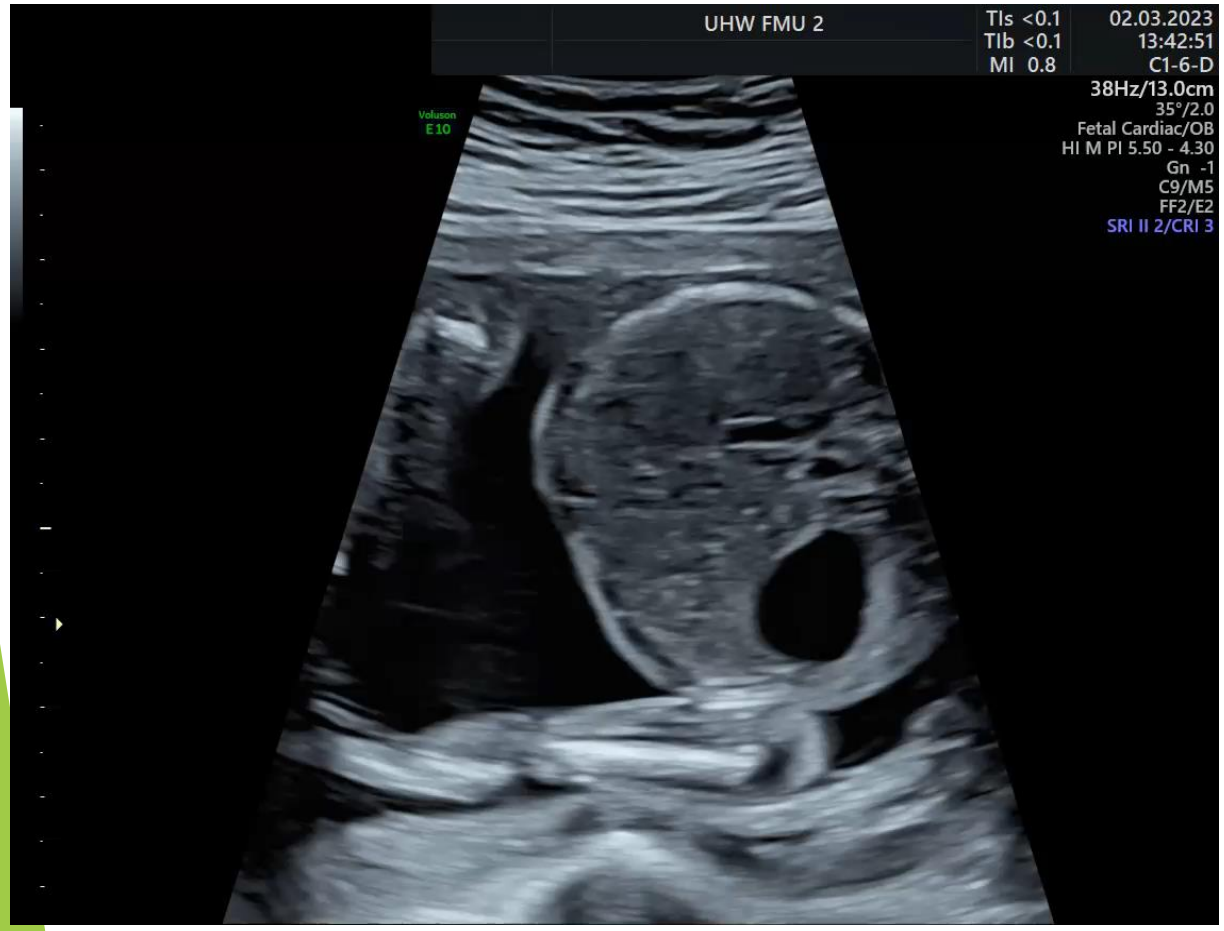
Duodenal Atresia

- Partial or complete:
  - Atresia
  - Web
  - Stenosis
  - Annular pancreas
- Double bubble
- Polyhydramnios (>24w)
- Most common place for obstruction
- 30% T21
- 50-70% other anomalies

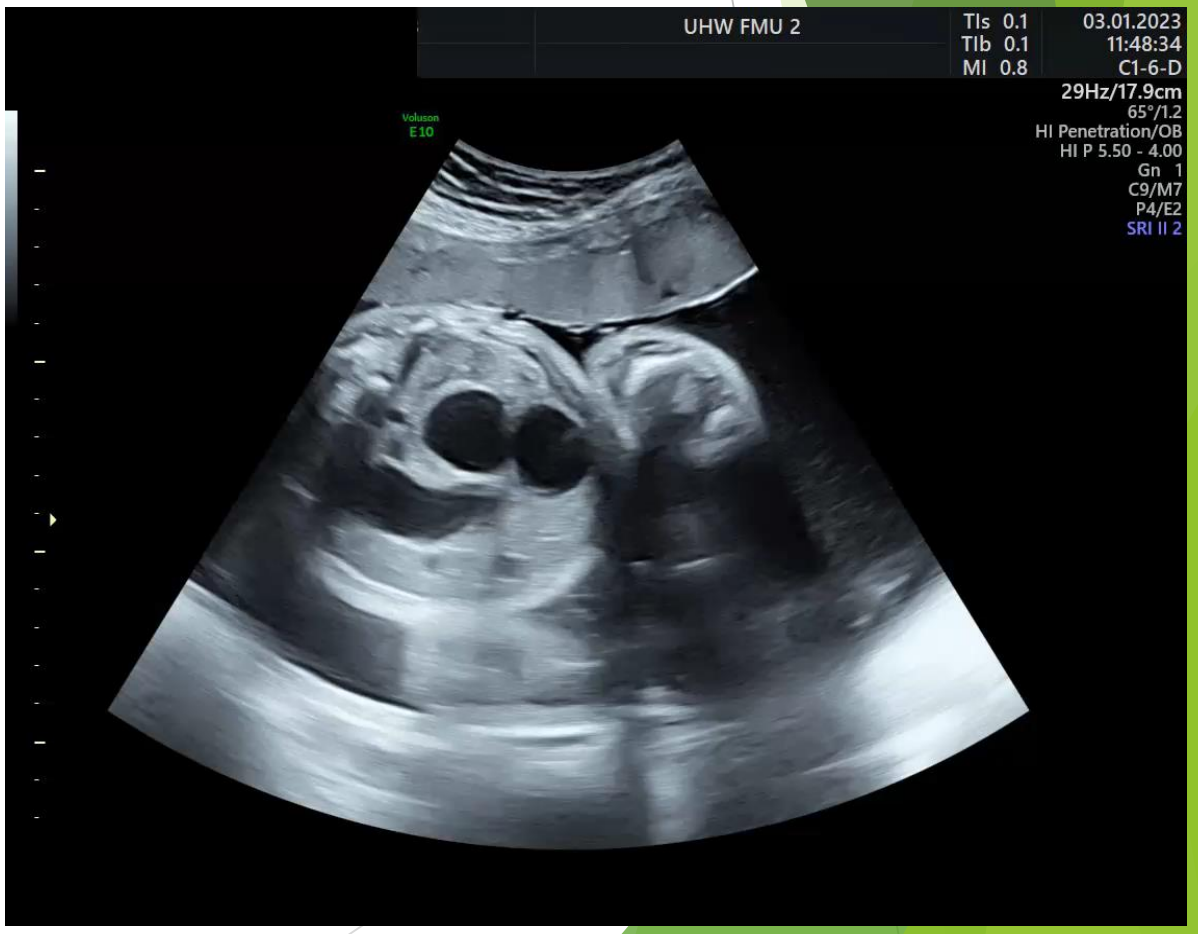


Duodenal Atresia: ultrasound

Double bubble: Duodenal



Triple bubble: Jejunal



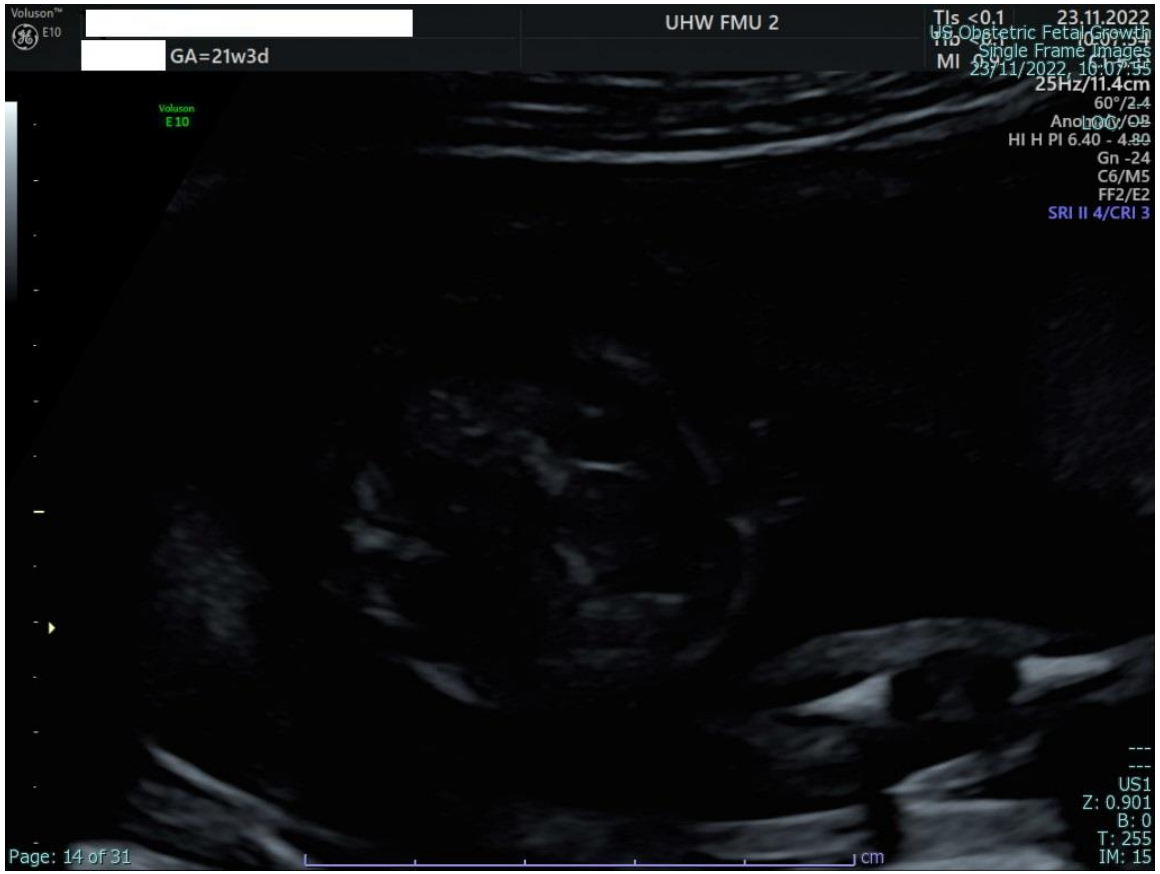
Echogenic bowel

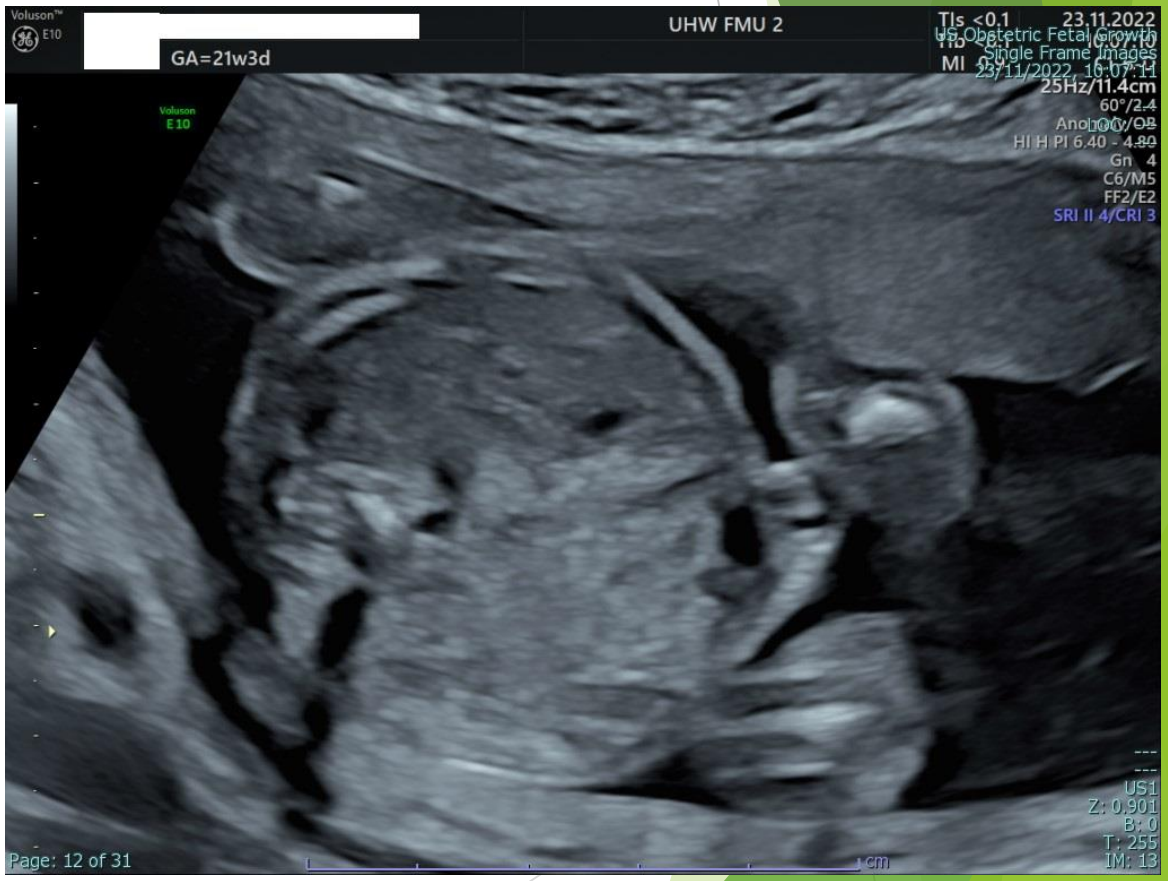
- 0.4-2%
- Causes:
  - Normal/ blood > 80%
  - Aneuploidy
  - Infection: Parvo, CMV, Toxo
  - CF
  - FGR
  - GI:
    - Meconium peritonitis
    - Ischemia

- Ultrasound:
  - Bright as bone!
  - Focal vs diffuse
  - Probe 3.5-5 MHz
  - Low gain

Echogenic bowel: ultrasound

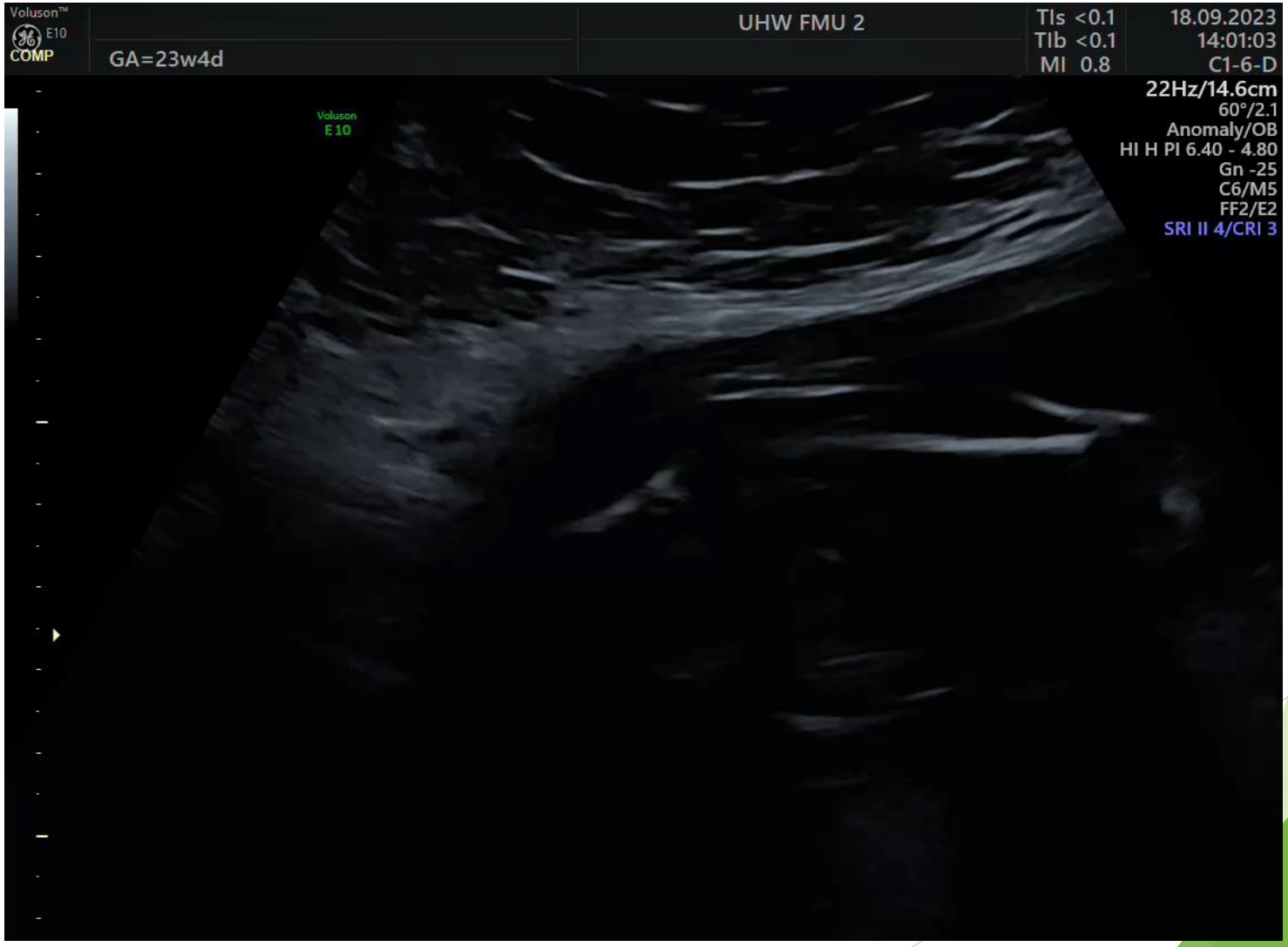
No





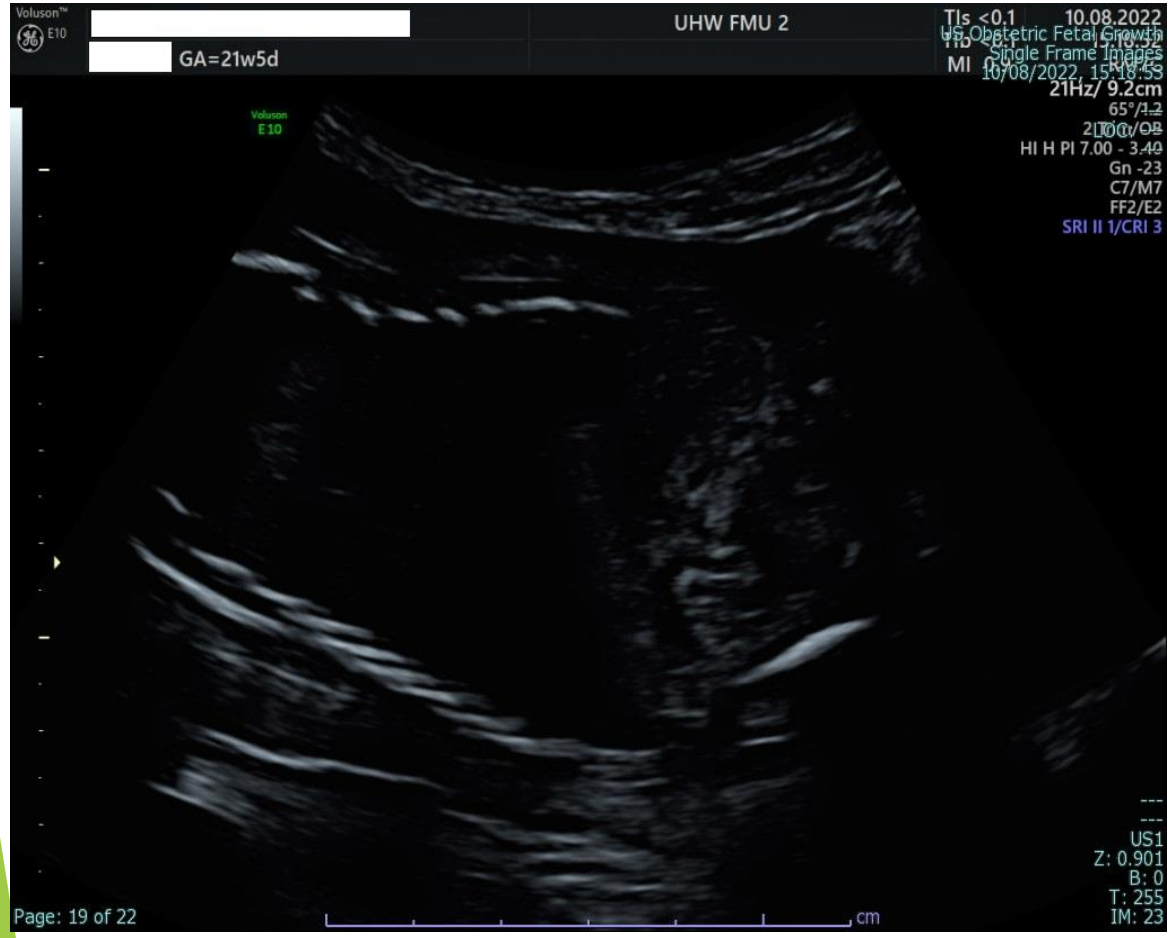
Echogenic bowel: ultrasound

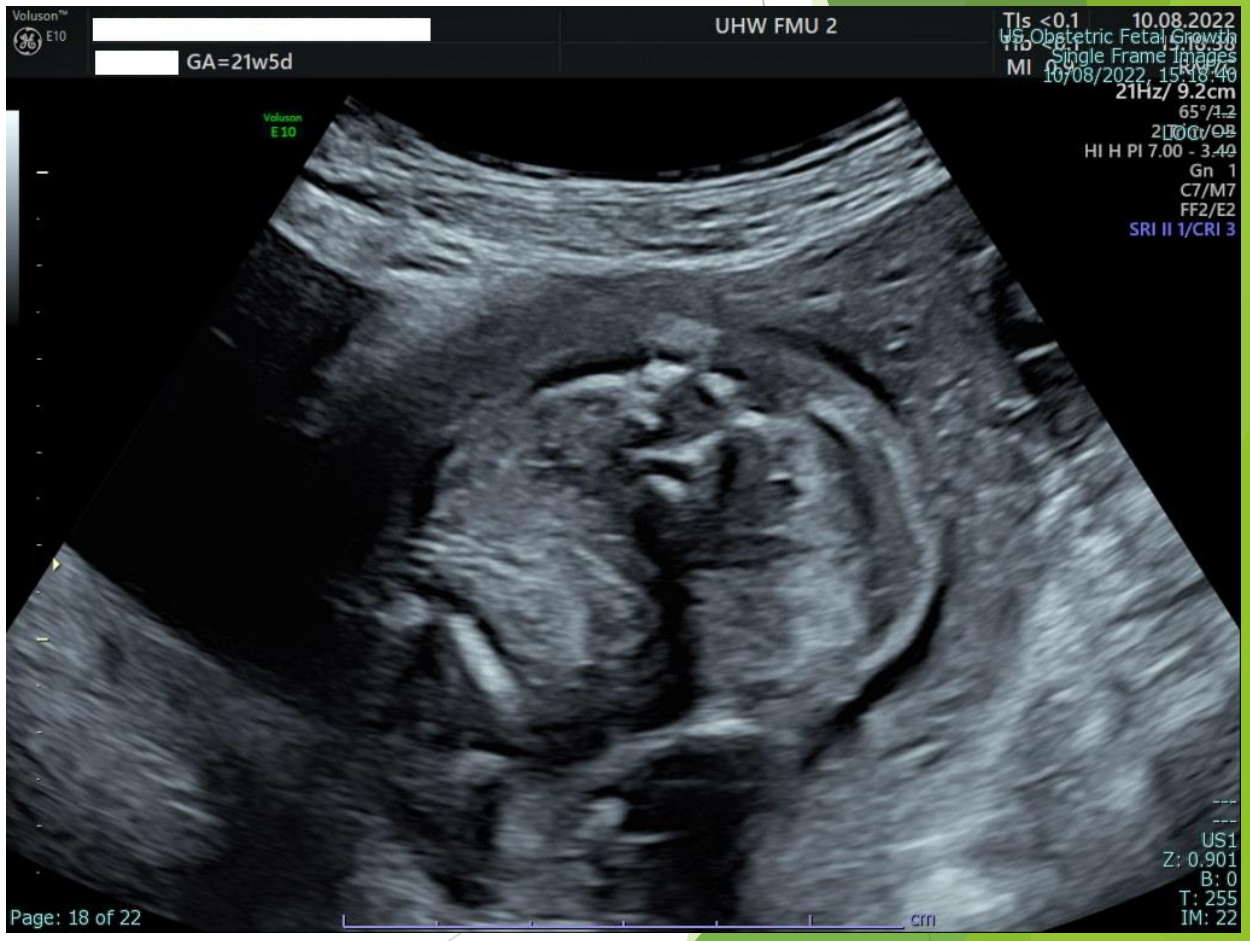
No



Echogenic bowel: ultrasound

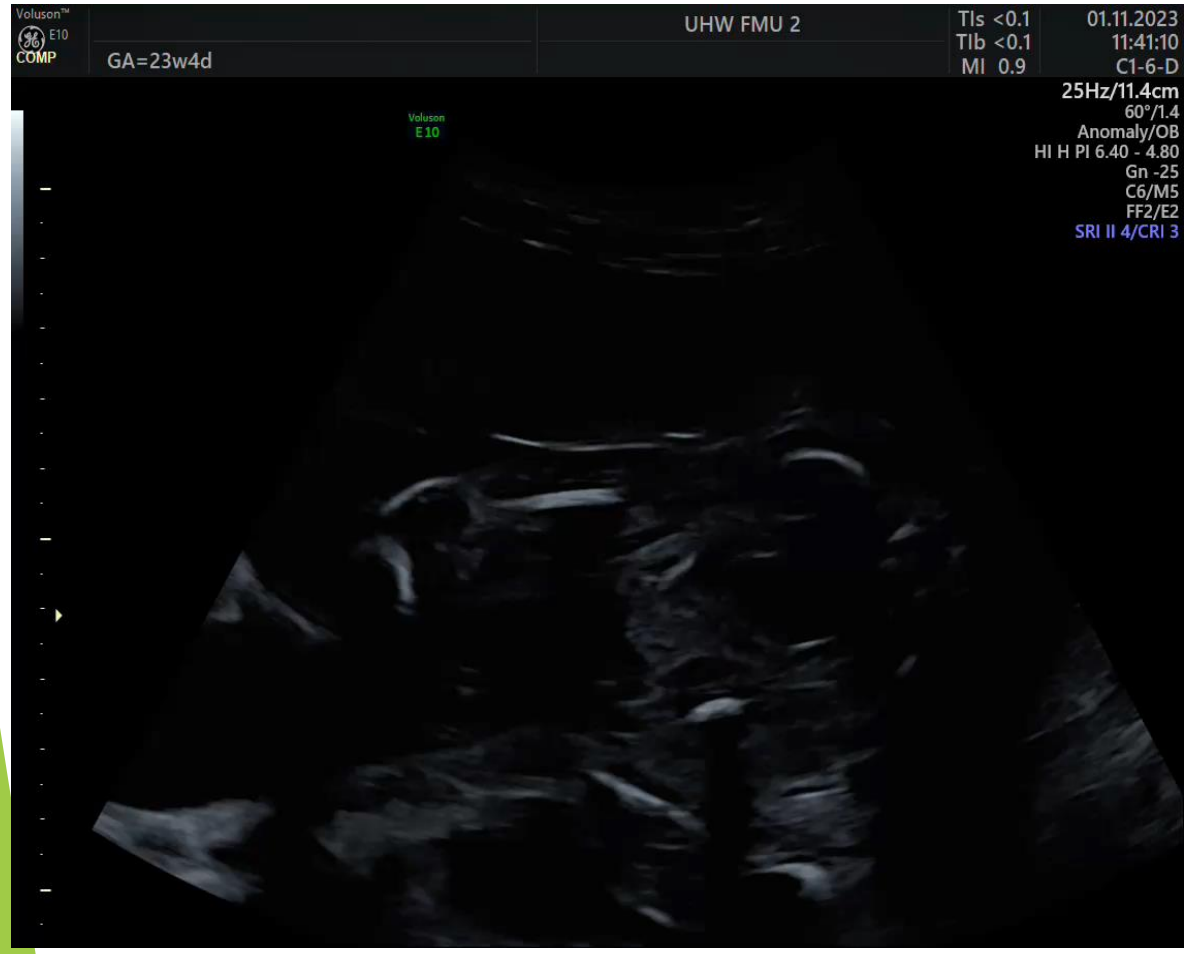
Mild

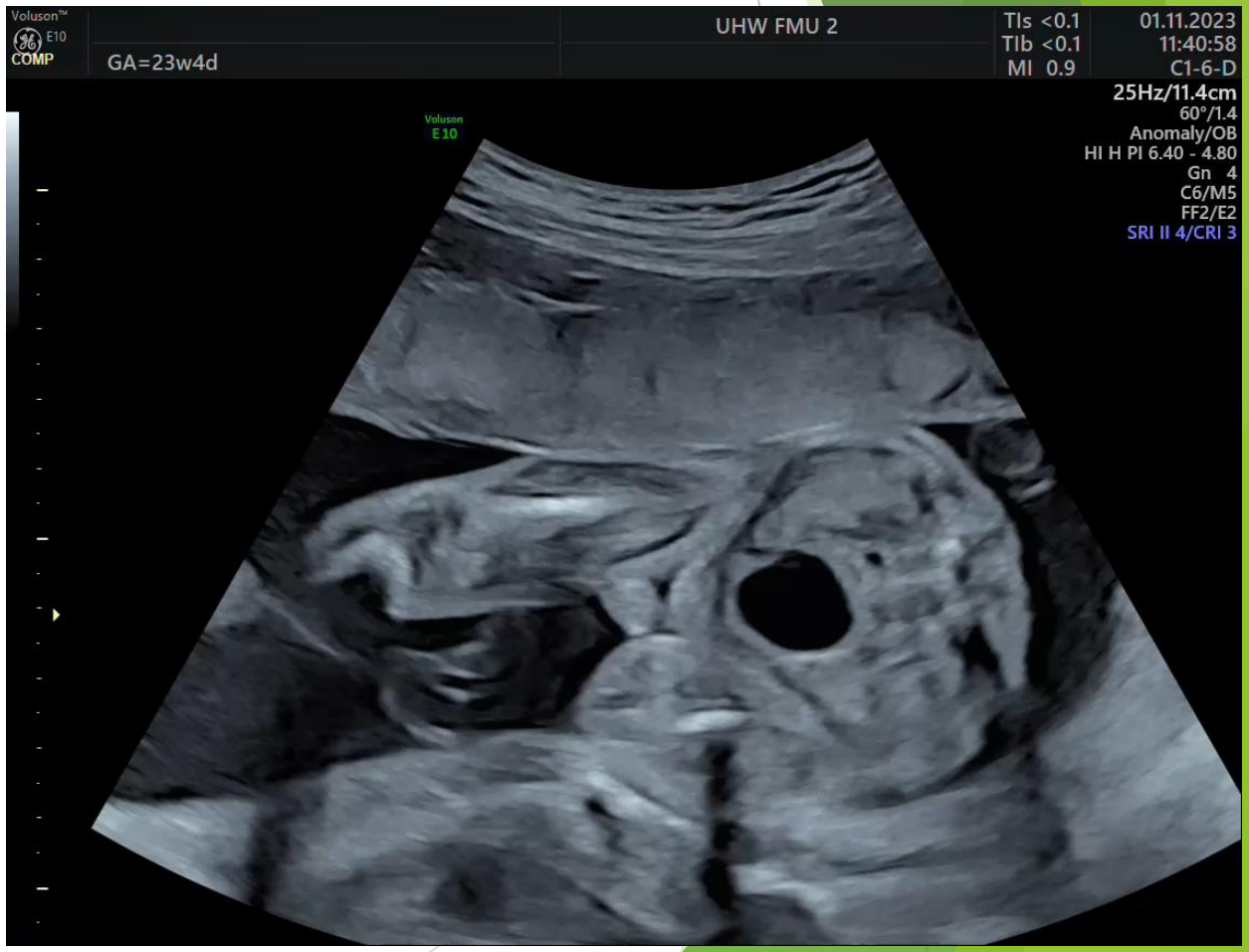




Echogenic bowel: ultrasound

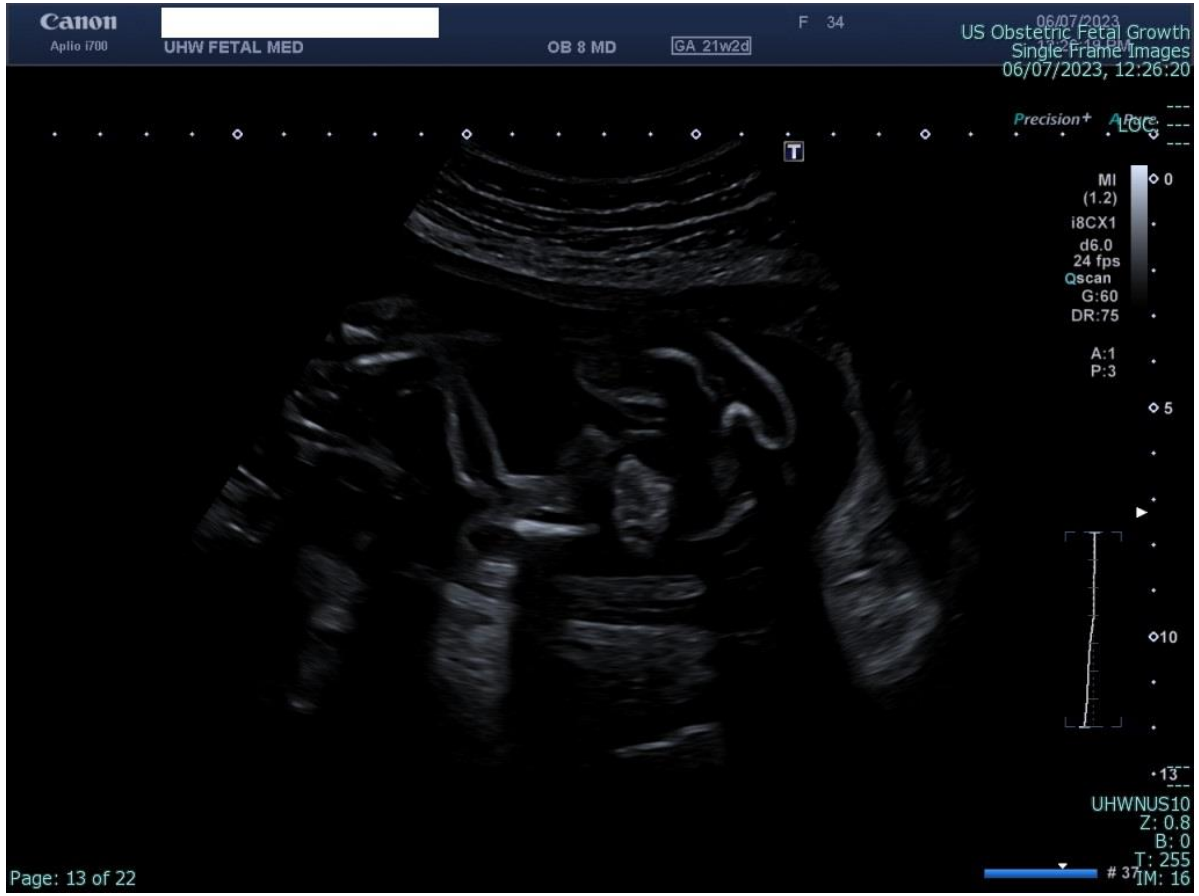
Mild

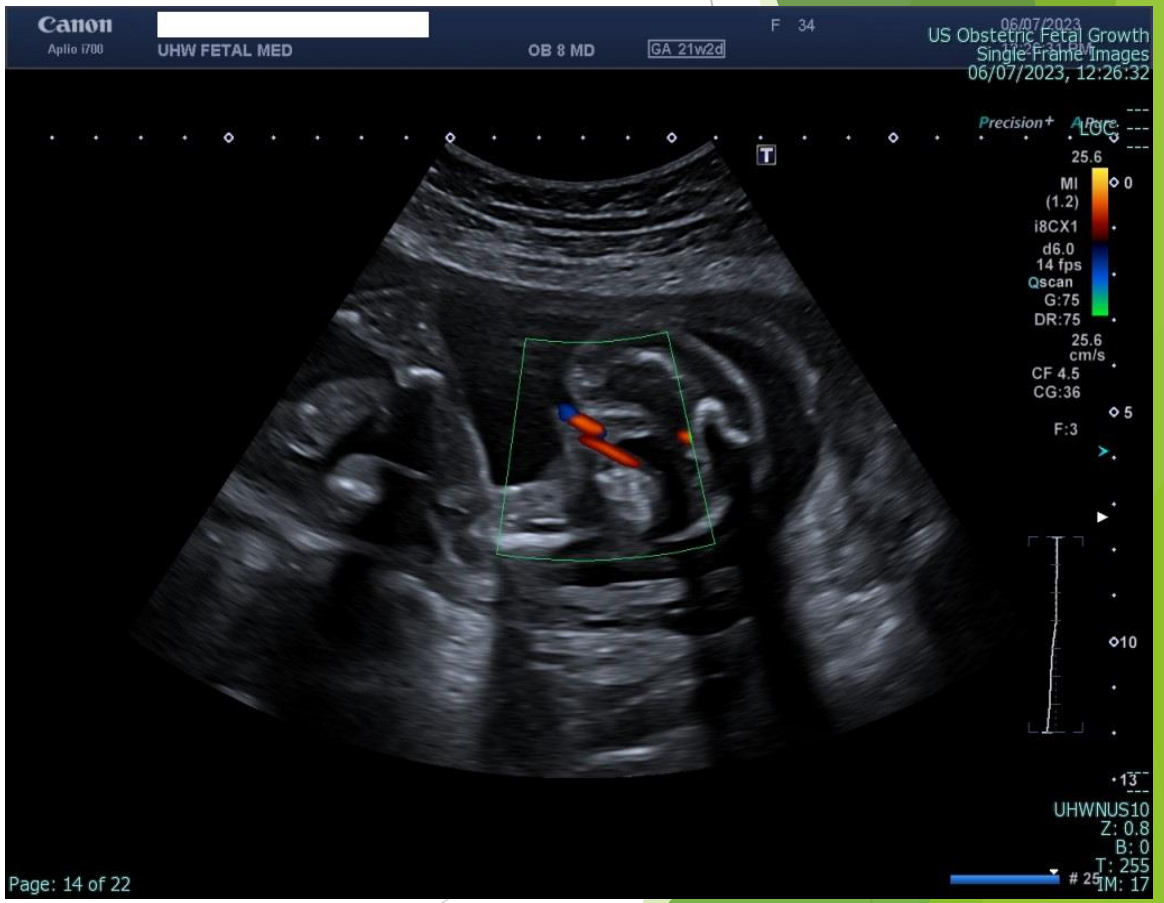




Echogenic bowel: ultrasound

Moderate - Severe





Sources:

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Annual Meeting 2023

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Public Health Wales

Surgical challenges

Mr Olly Jackson

Consultant Paediatric and Neonatal Surgeon

CARIS annual meeting 2023

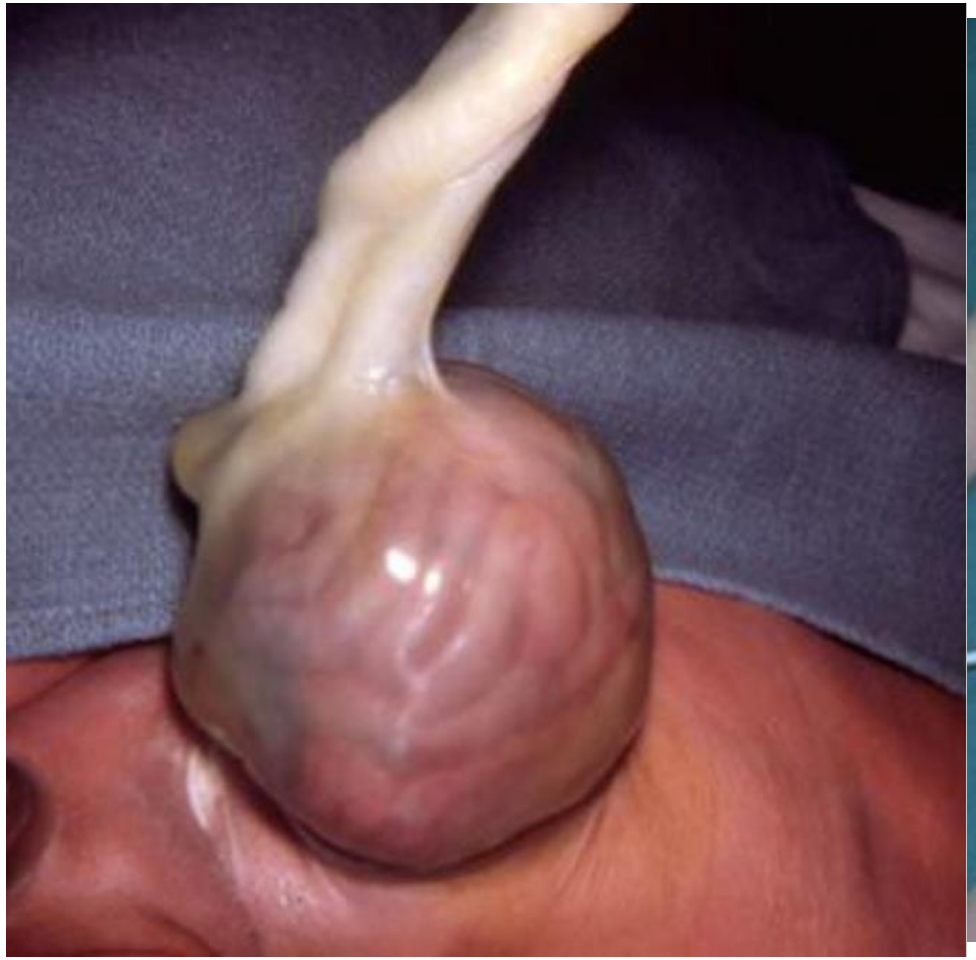
Surgical Challenges

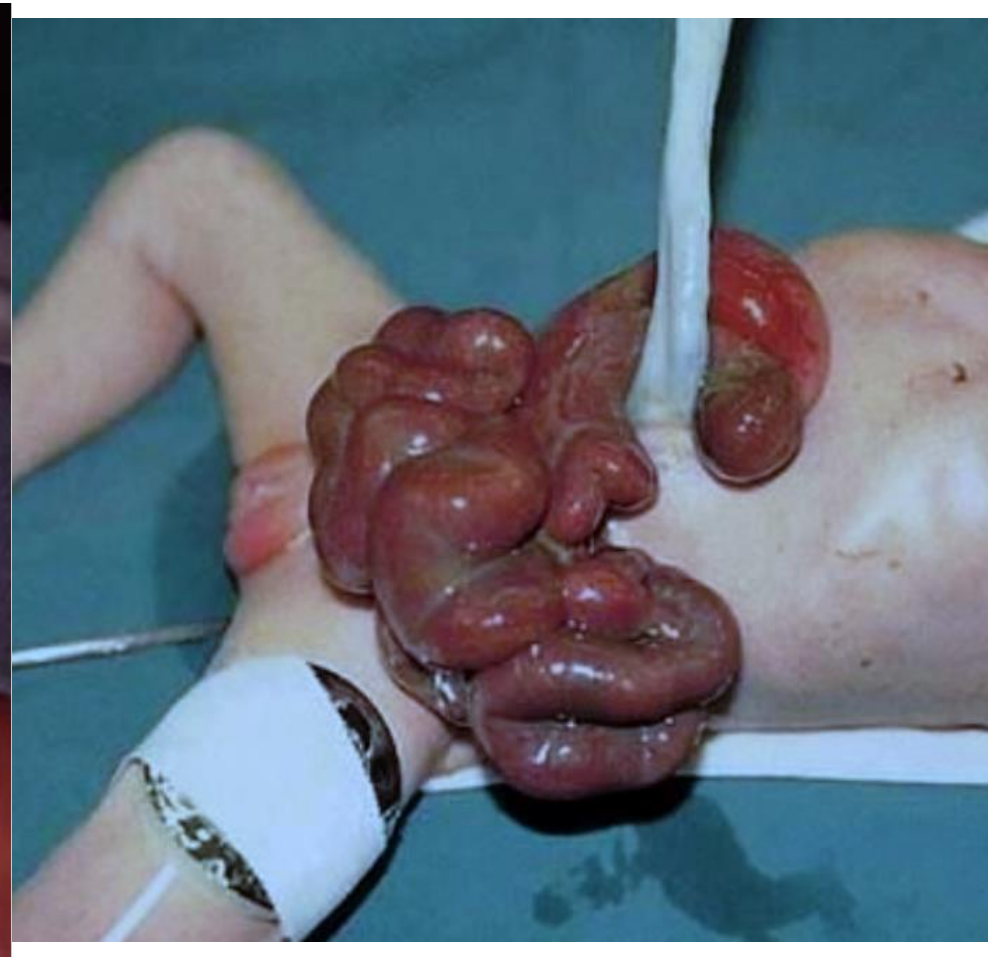
Mr Olly Jackson,
Consultant Paediatric and Neonatal Surgeon.

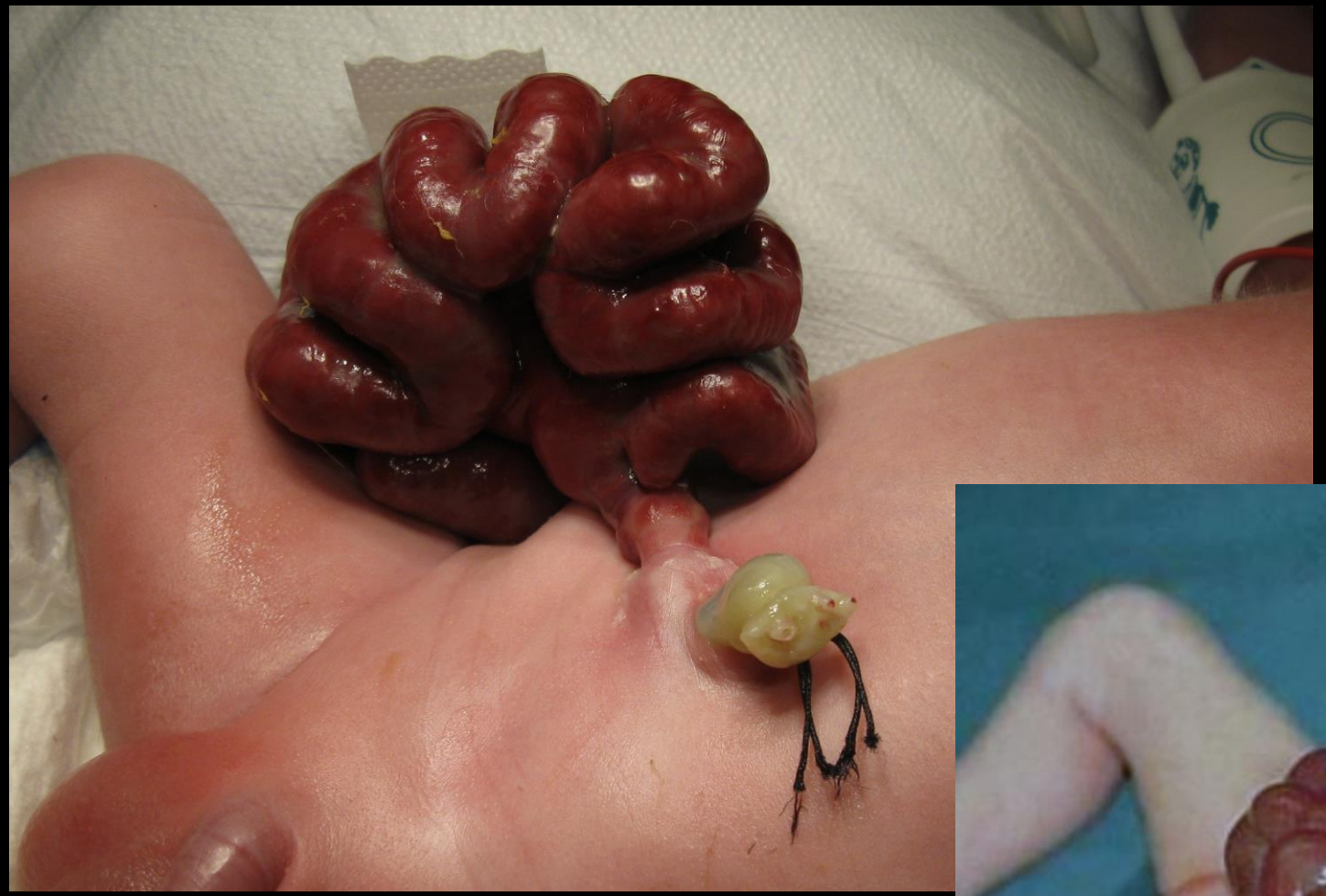
Aims

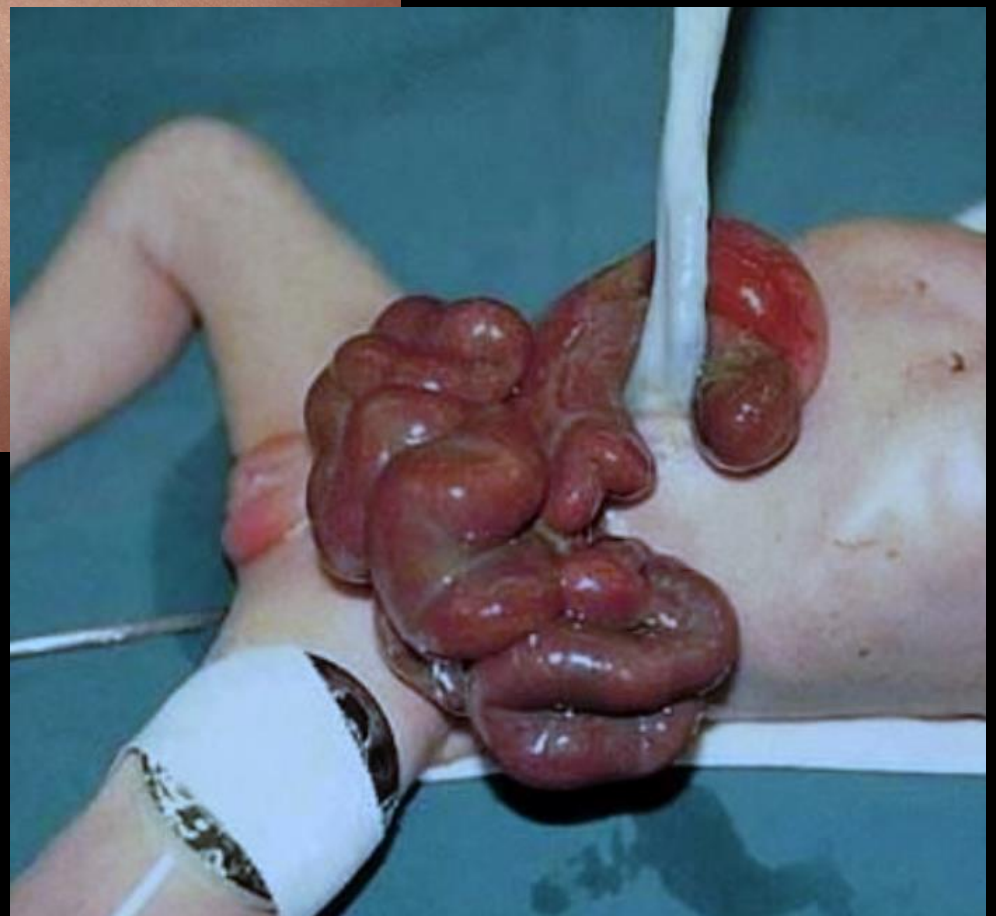
- To provide the best care possible to every baby and family we look after.
- Overview of conditions that we see in paeds surgery.
- Understand more about a baby’s surgical journey.

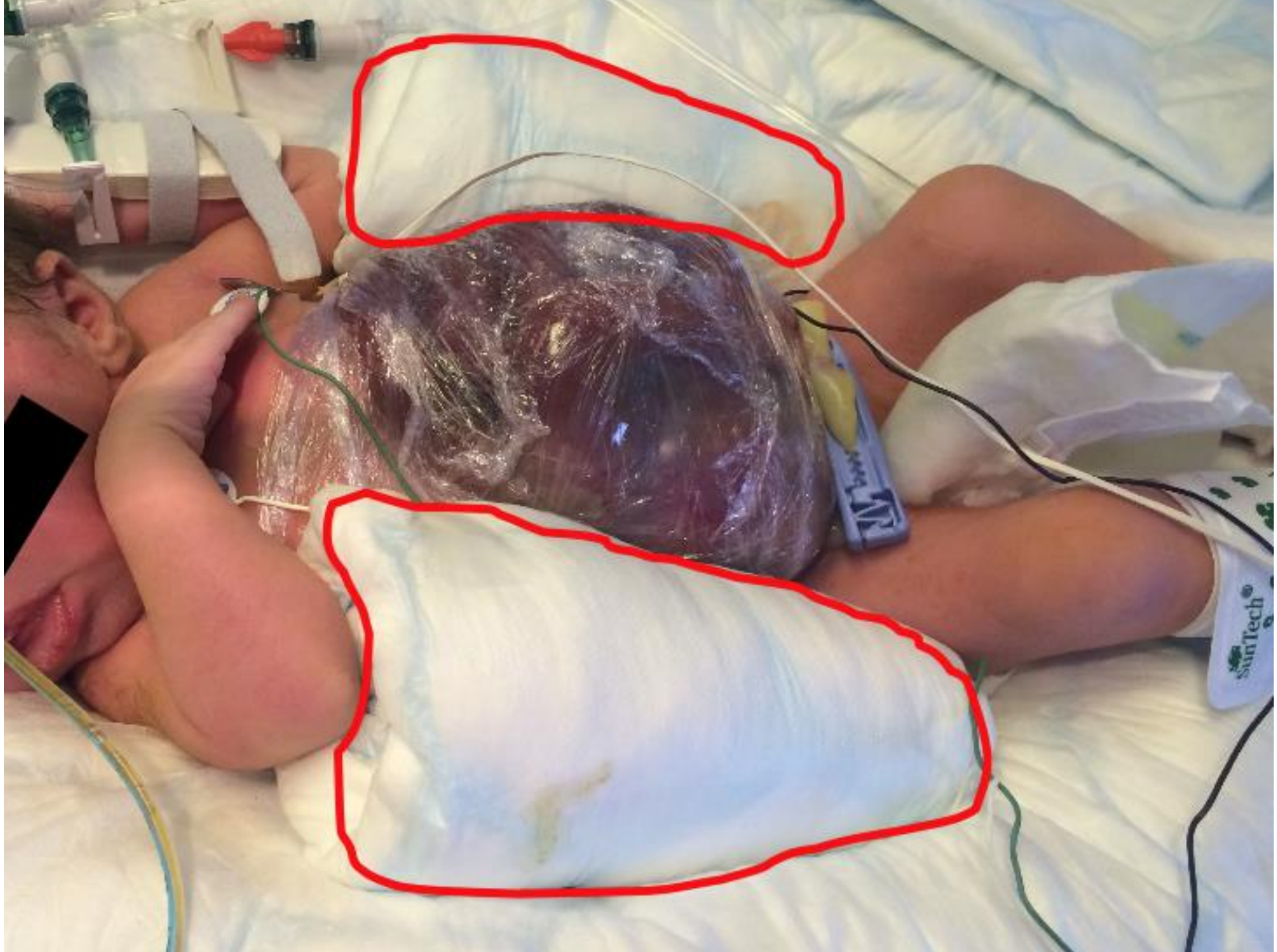
- Abdominal wall defects = gastroschisis and exomphalos.
- OA/TOF (oesophageal atresia and tracheo-oesophageal fistula).
- VACTERL.
- Pyloric stenosis.
- CDH (congenital diaphragmatic hernia).

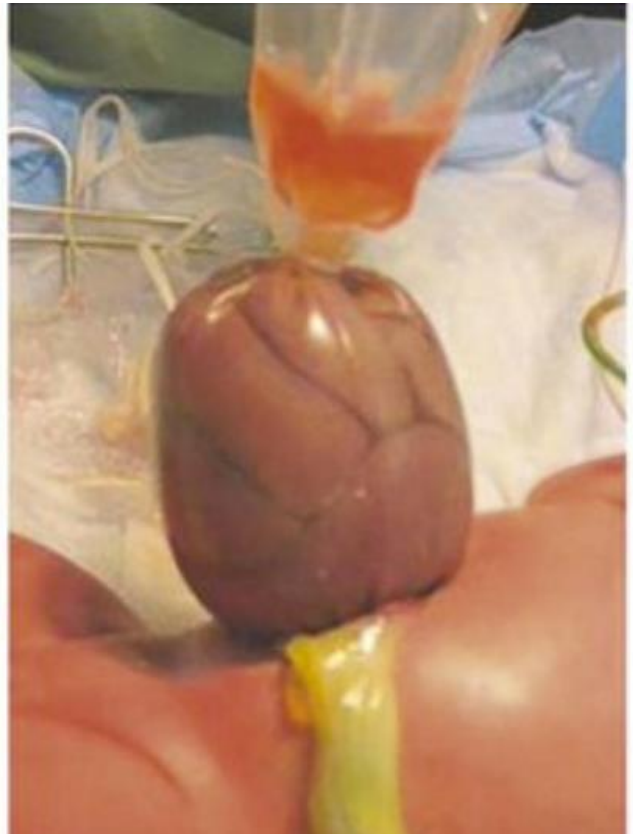


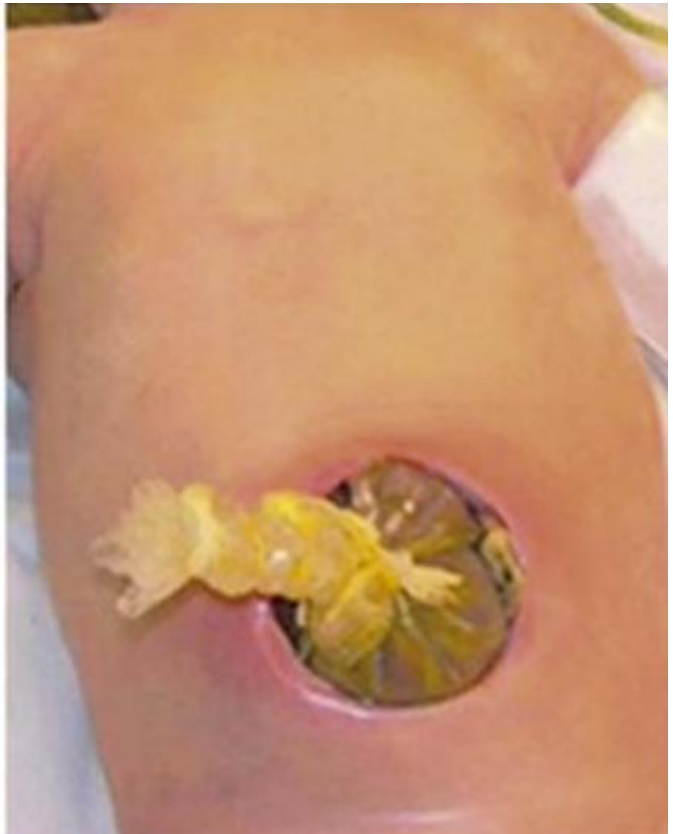


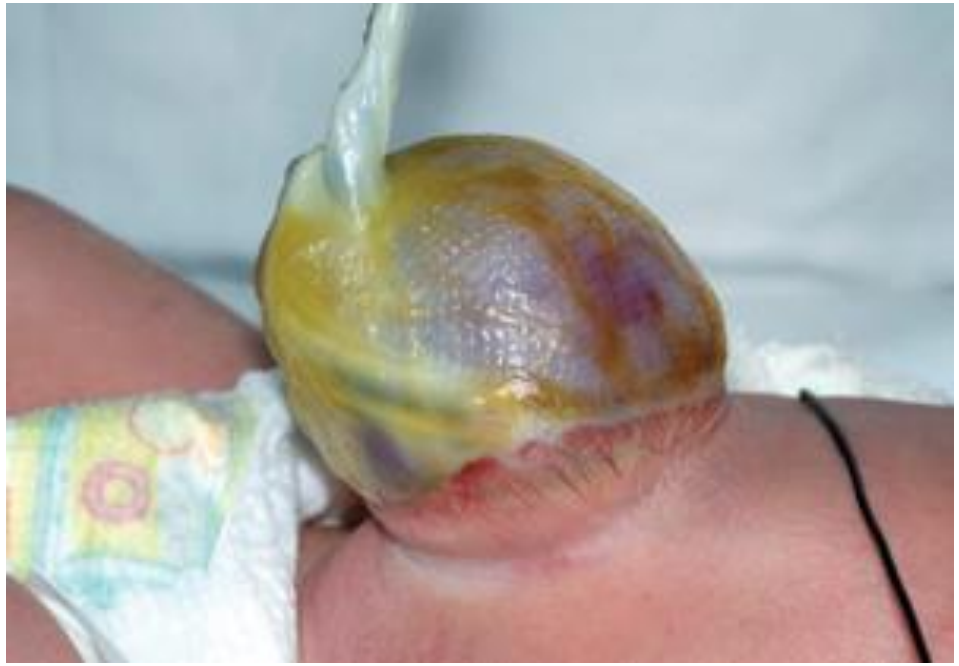


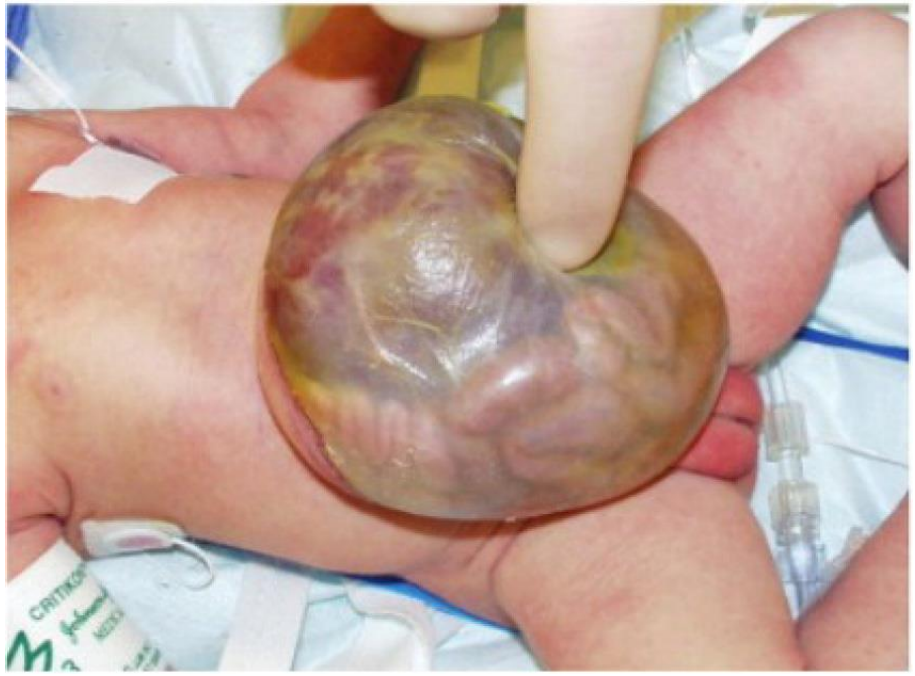


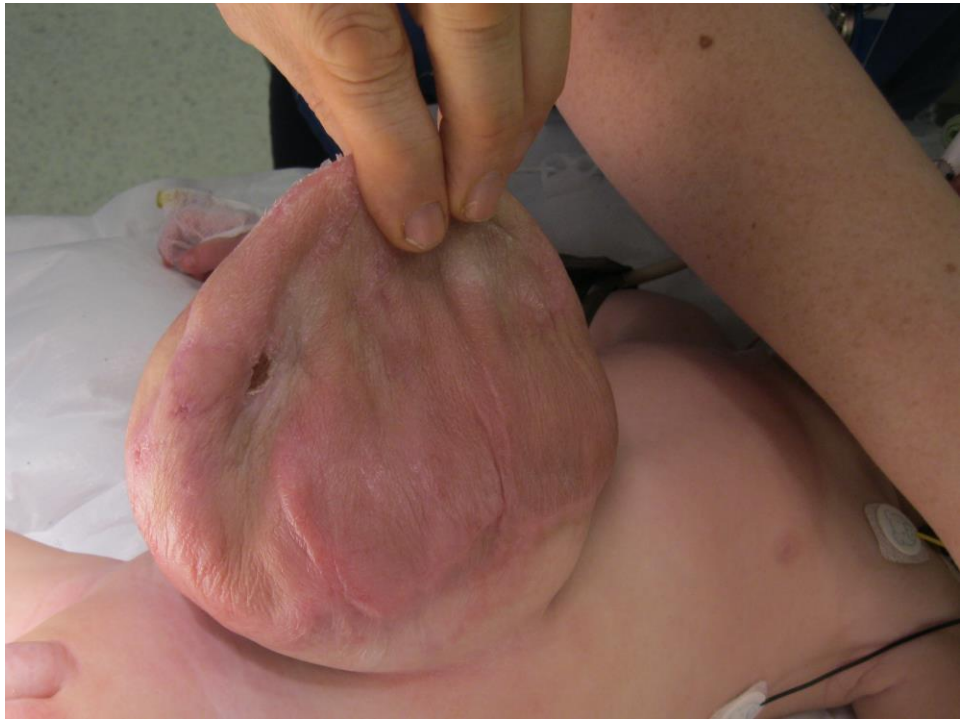

SunTech

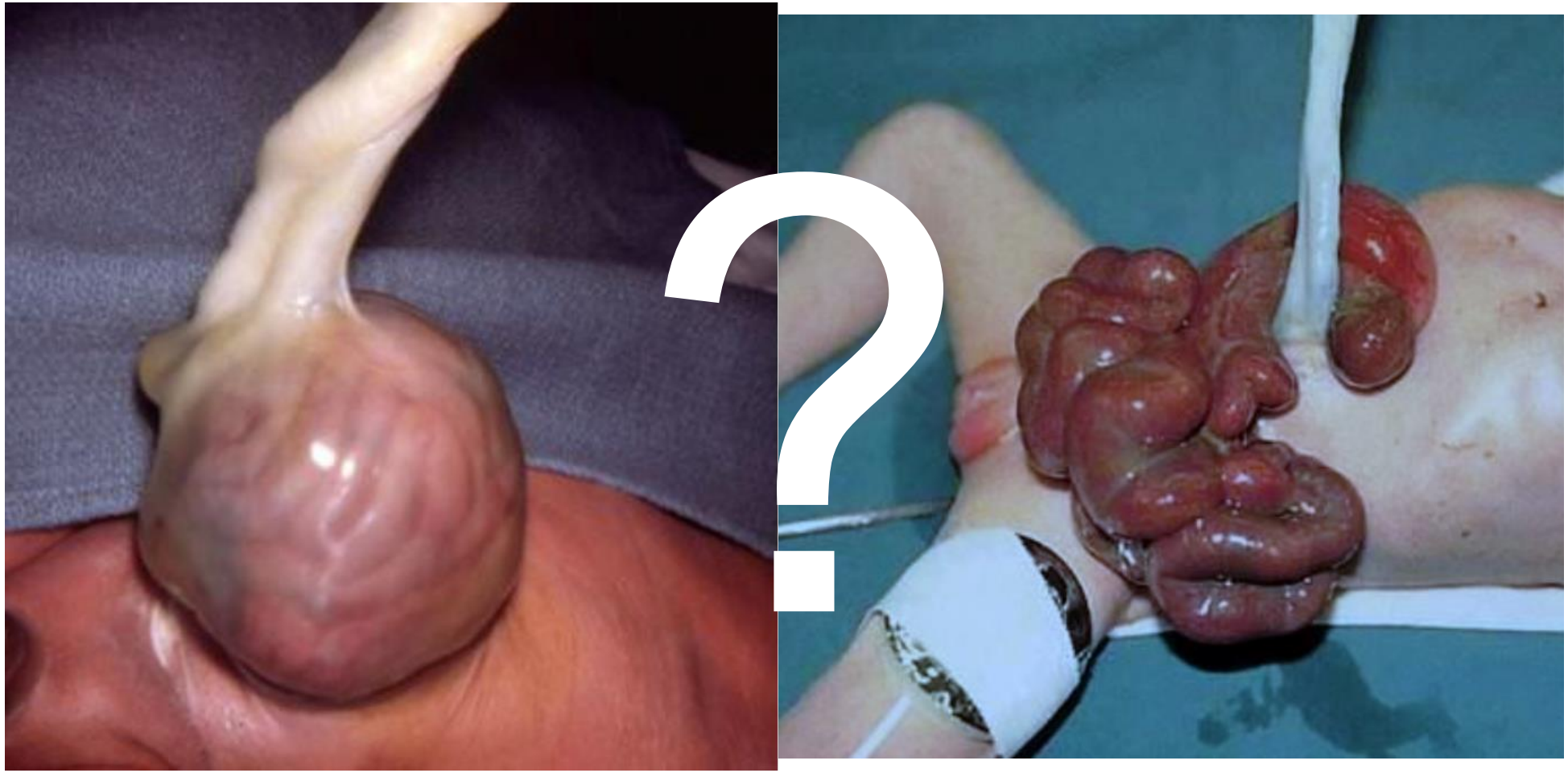




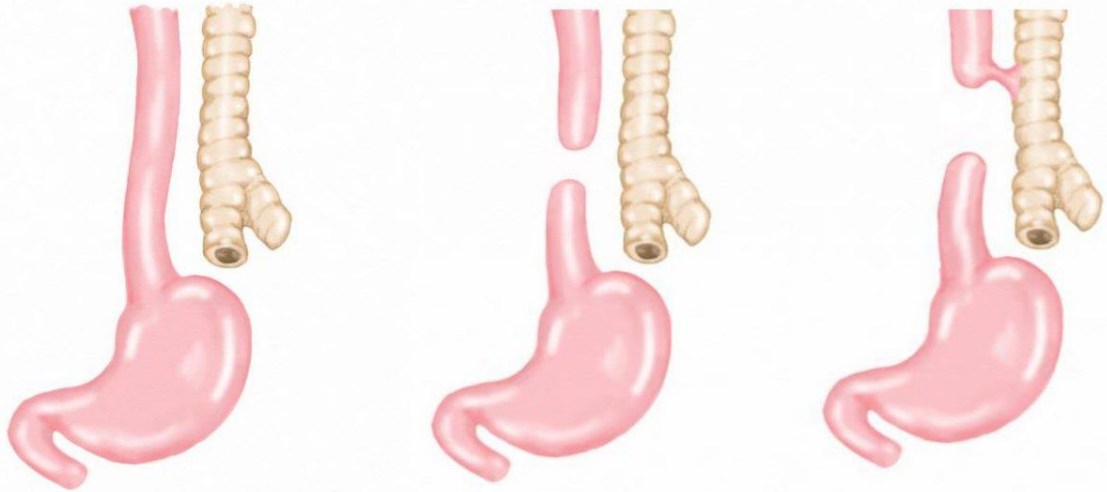








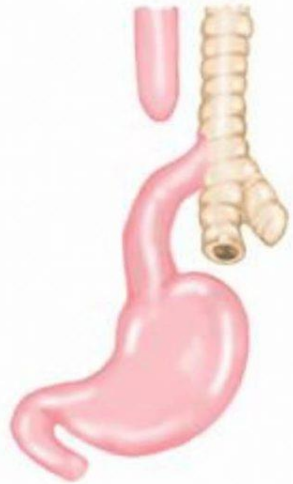
Different Types of Oesophageal Atresia and Tracheo-Oesophageal Fistula



"Normal"

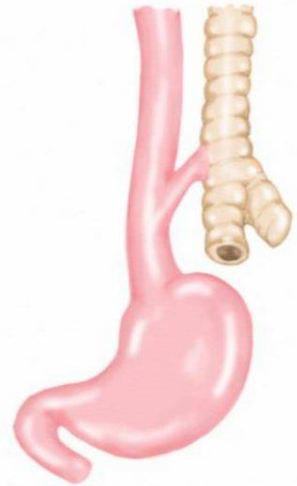
Type A

Type B

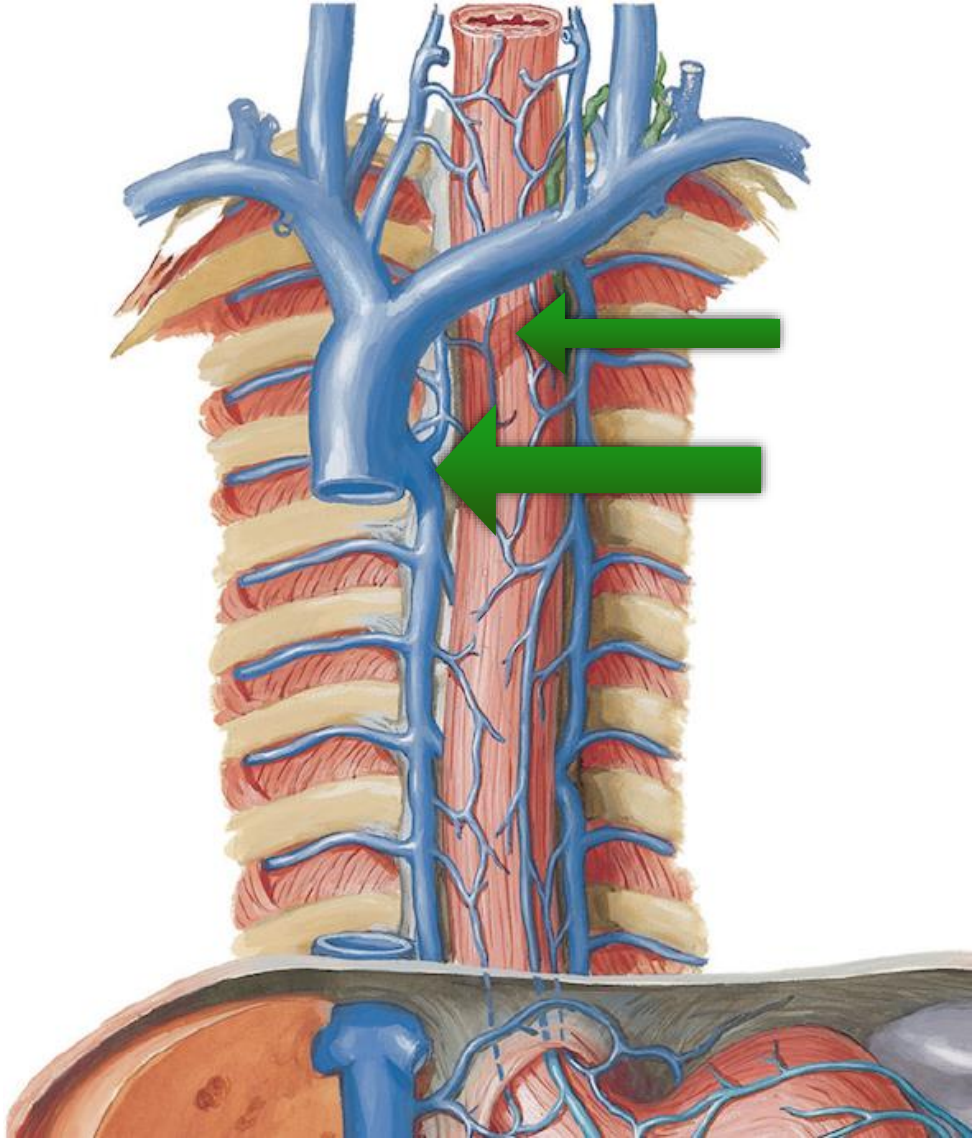


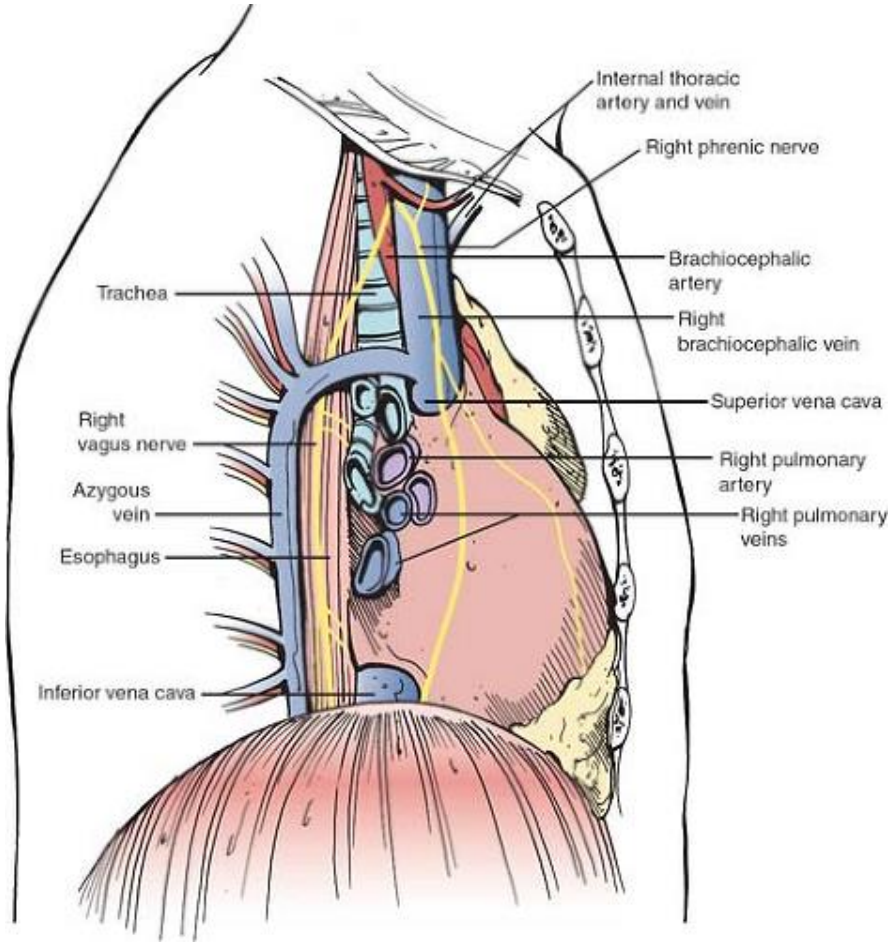
Type C

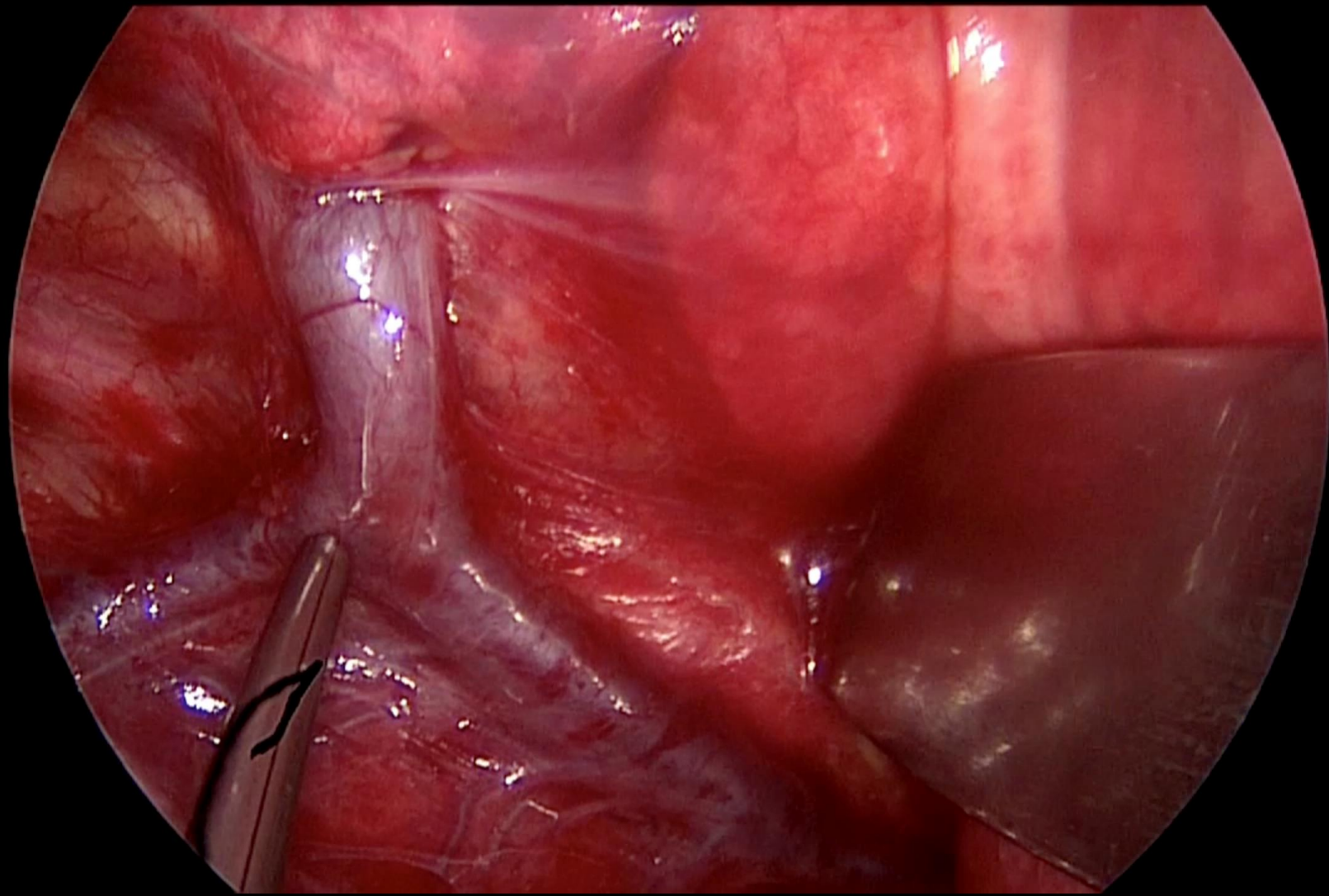
Type D

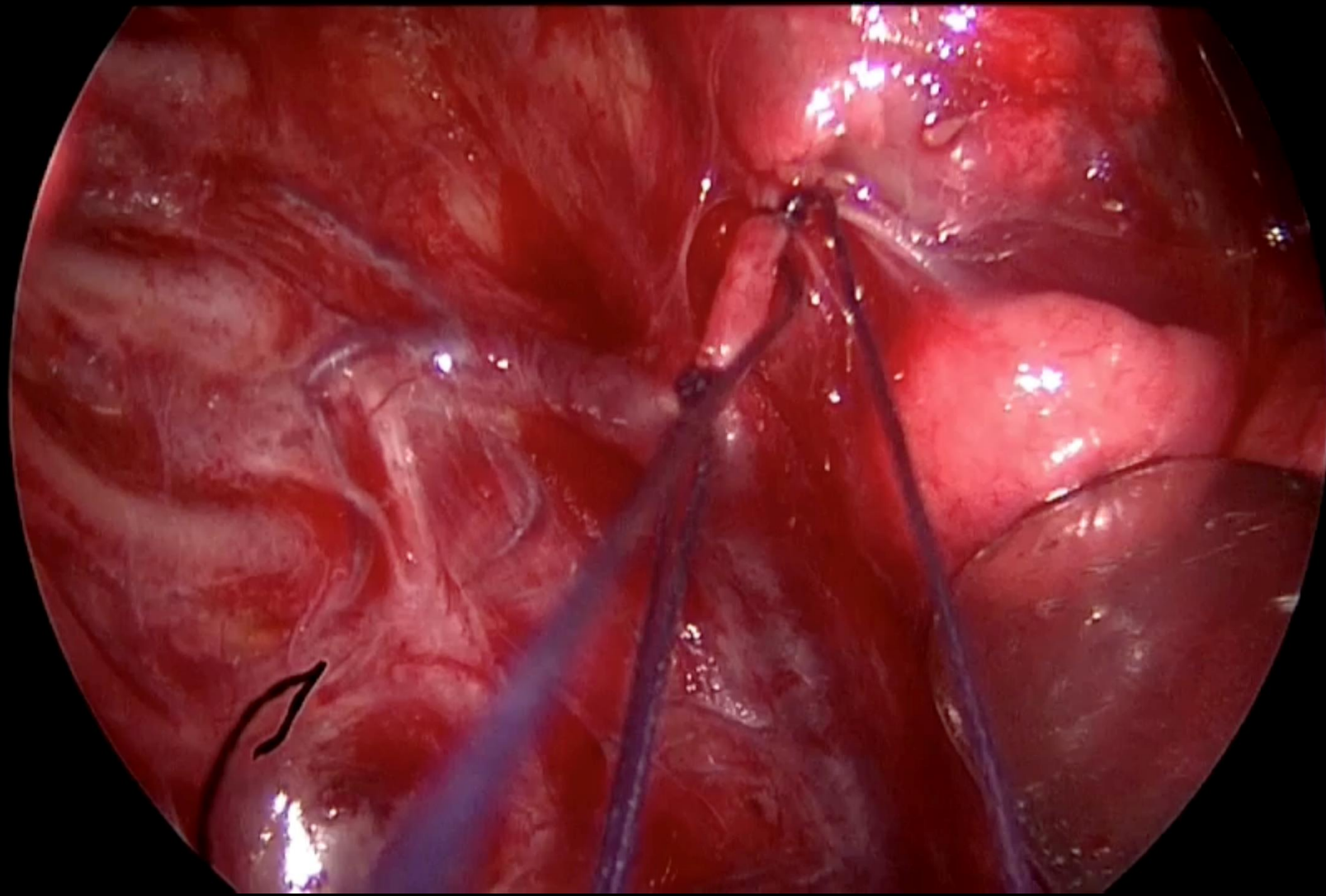


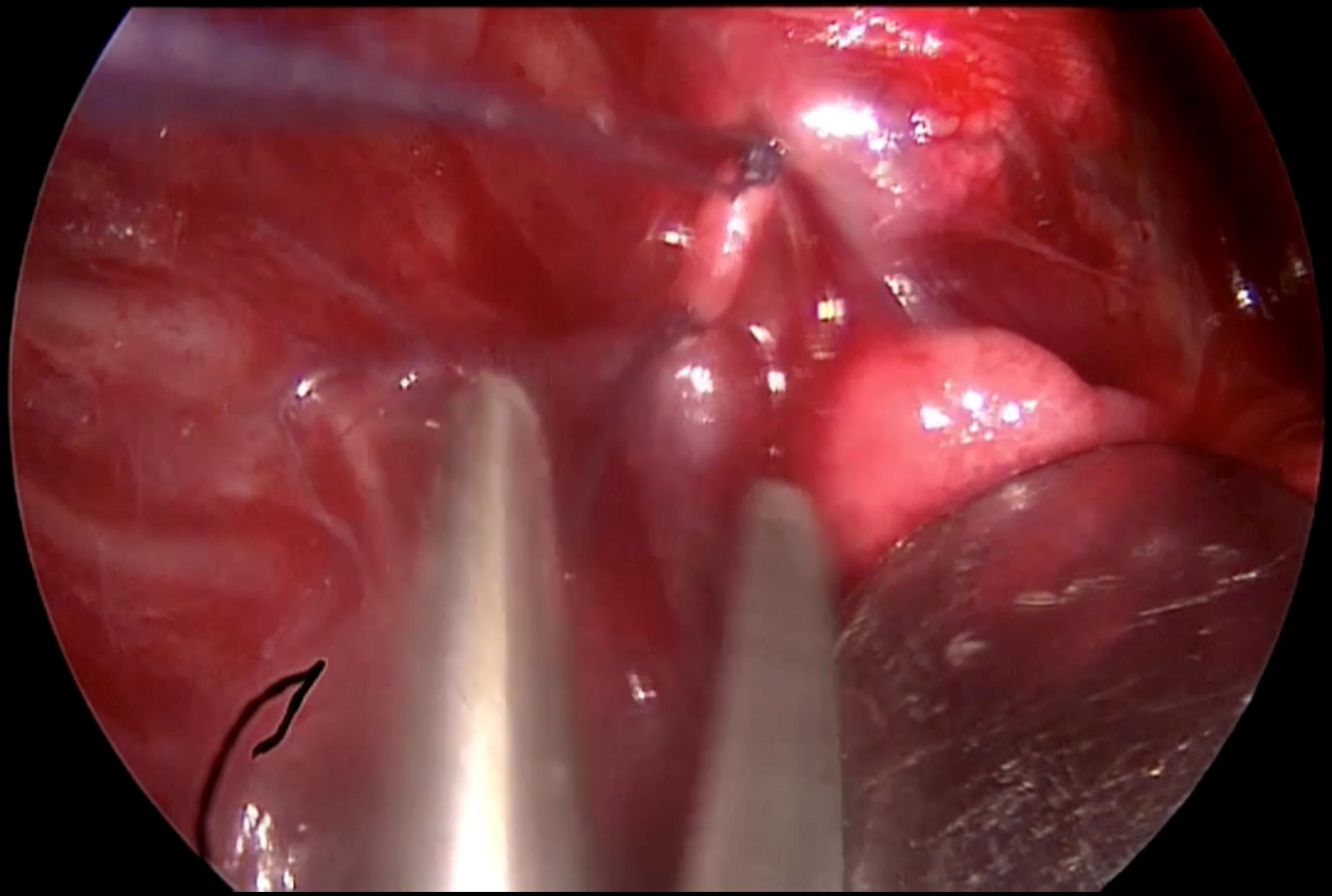
Type E

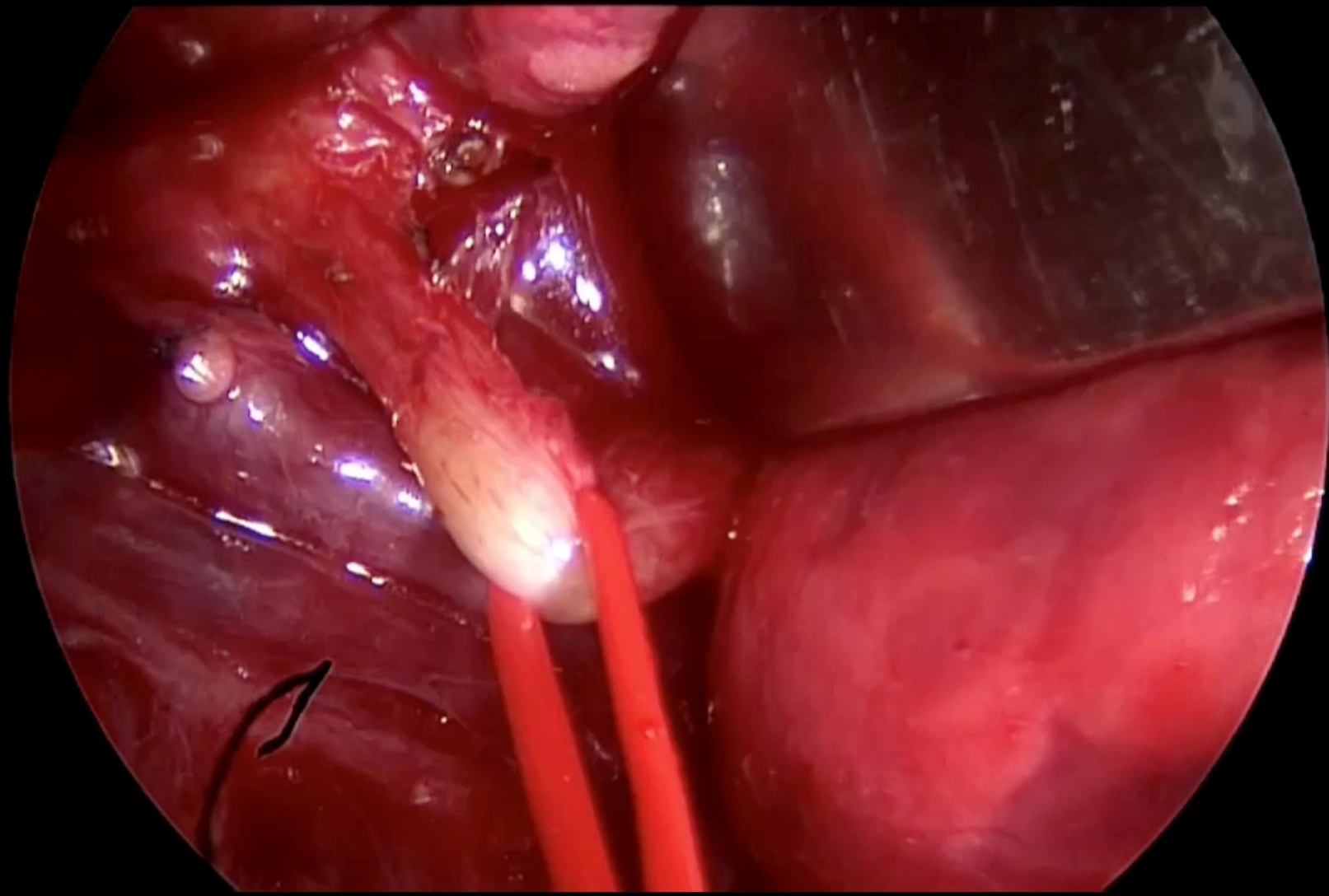


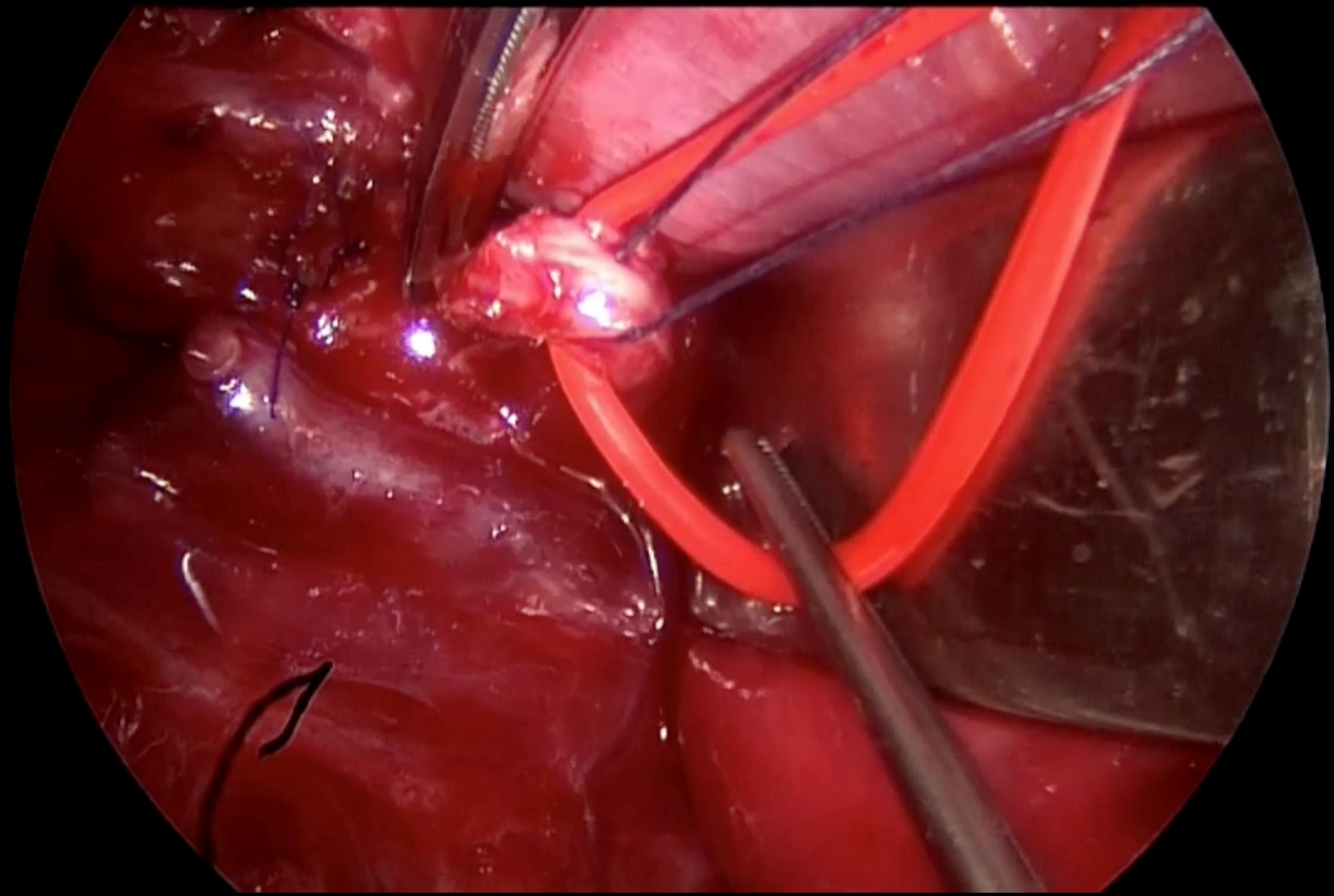

Internal thoracic artery and vein
Right phrenic nerve
Brachiocephalic artery
Trachea
Right brachiocephalic vein
Superior vena cava
Right vagus nerve
Right pulmonary artery
Azygous vein
Right pulmonary veins
Esophagus
Inferior vena cava

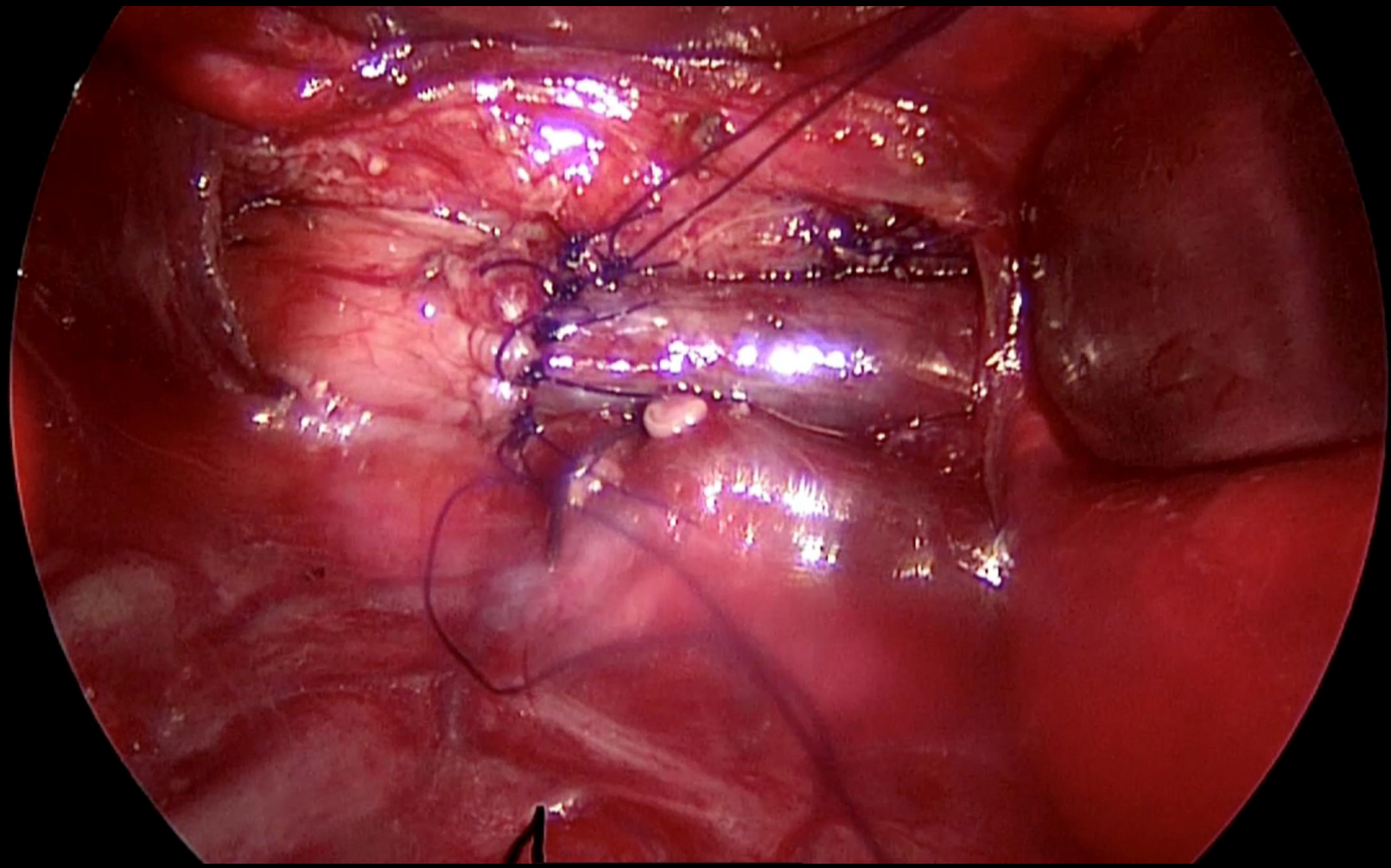


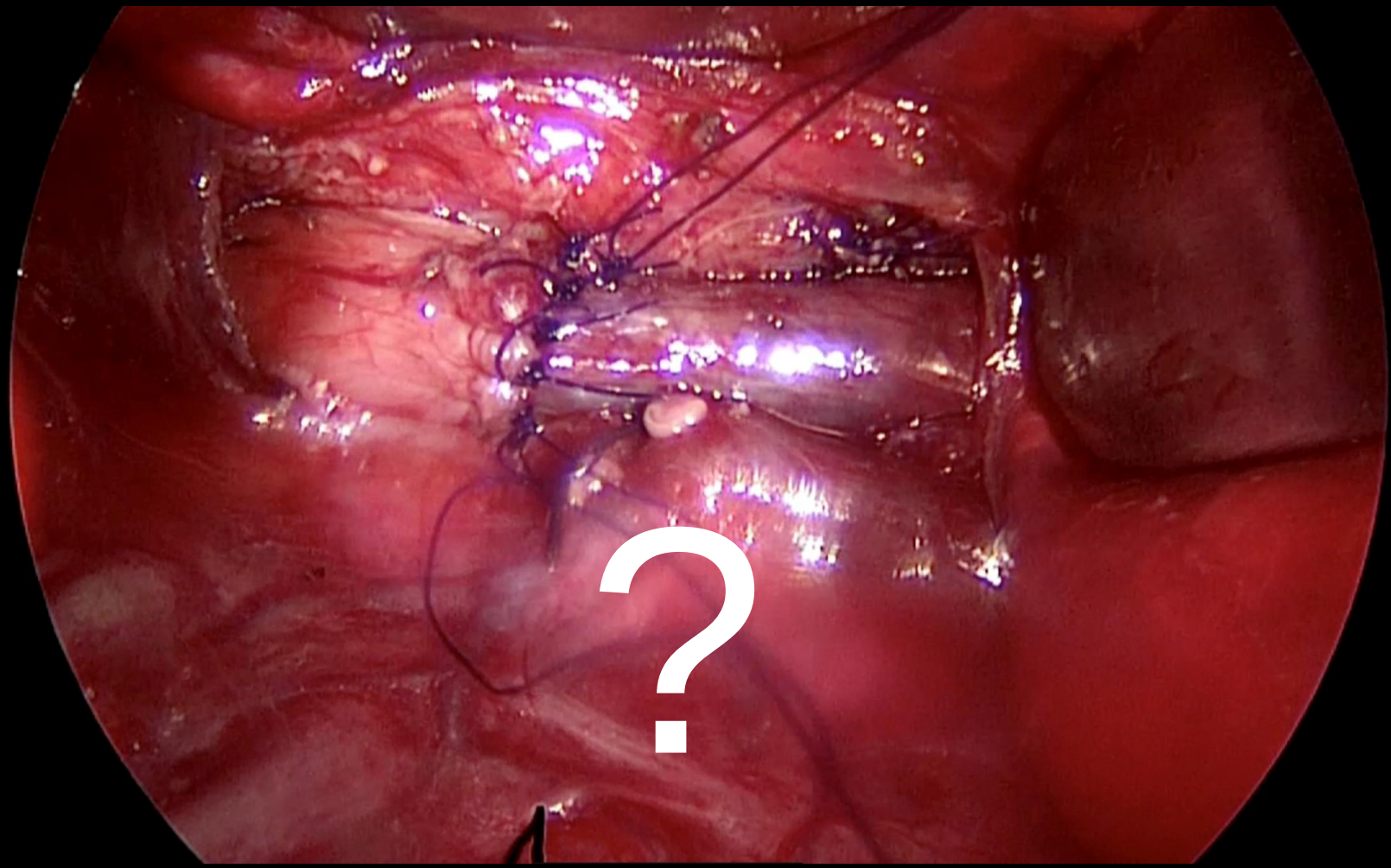










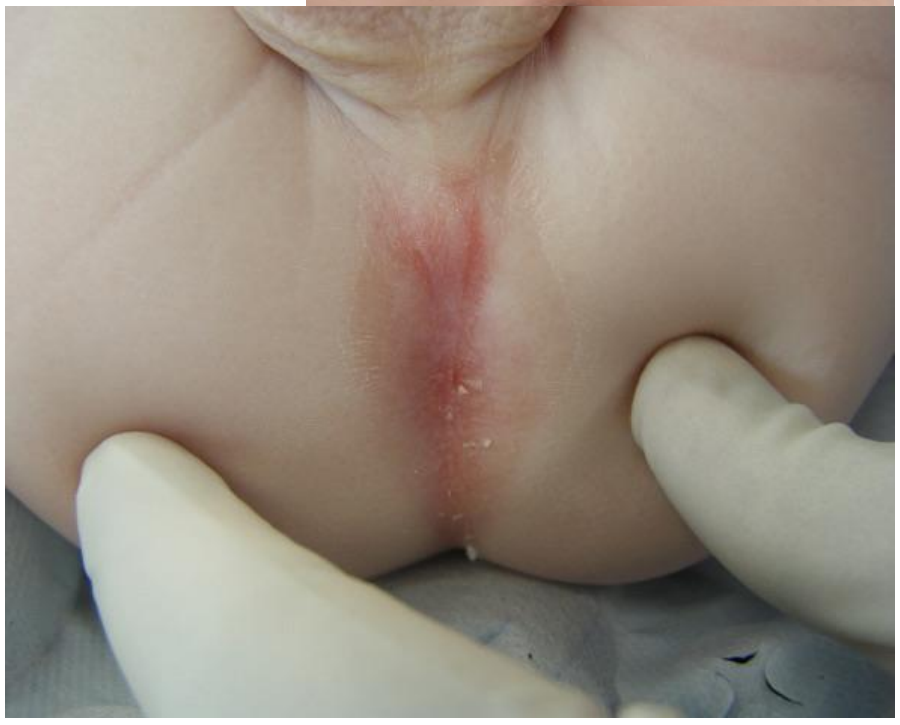


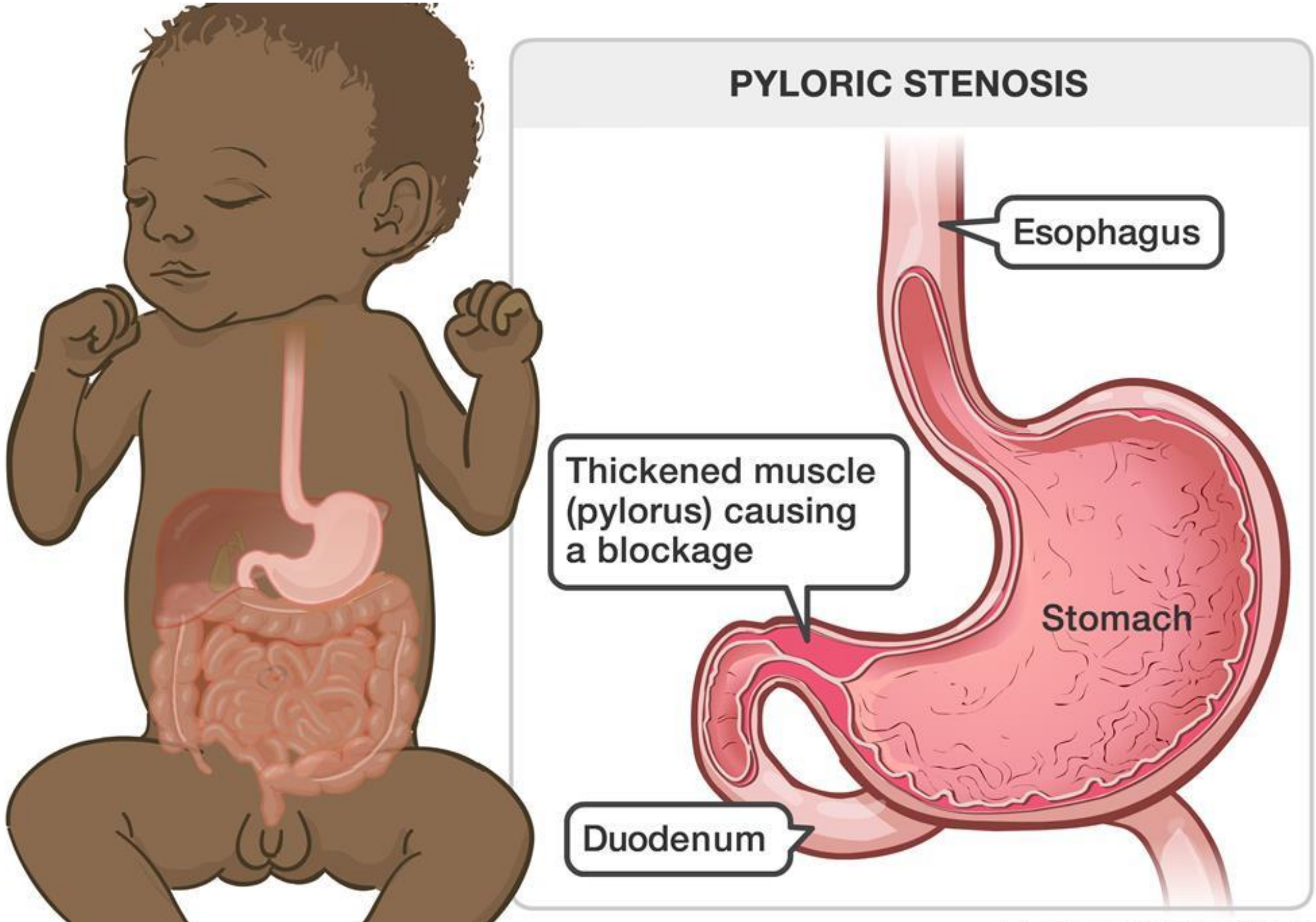
VACTERL Association

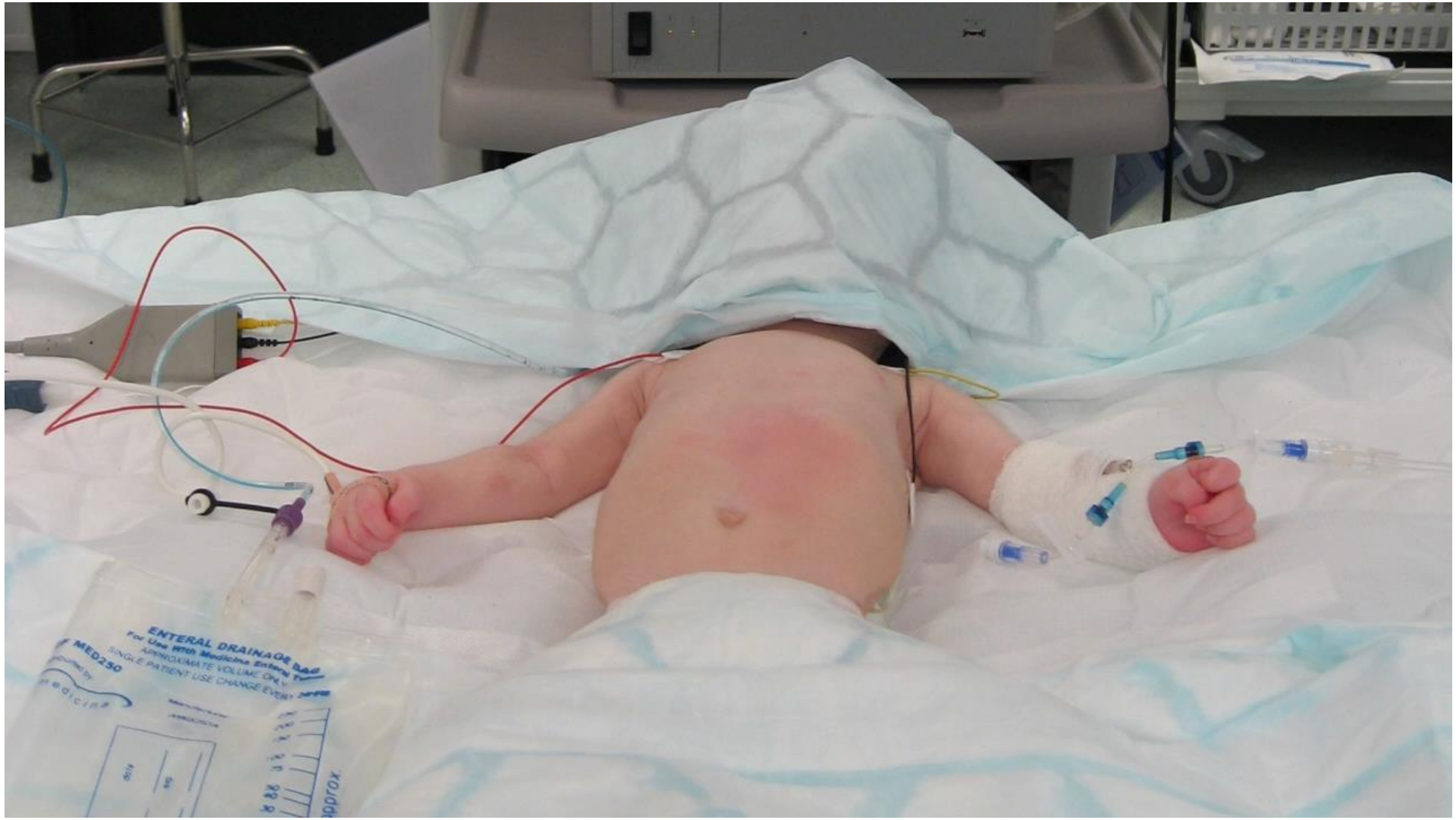
Vertebral
Anorectal

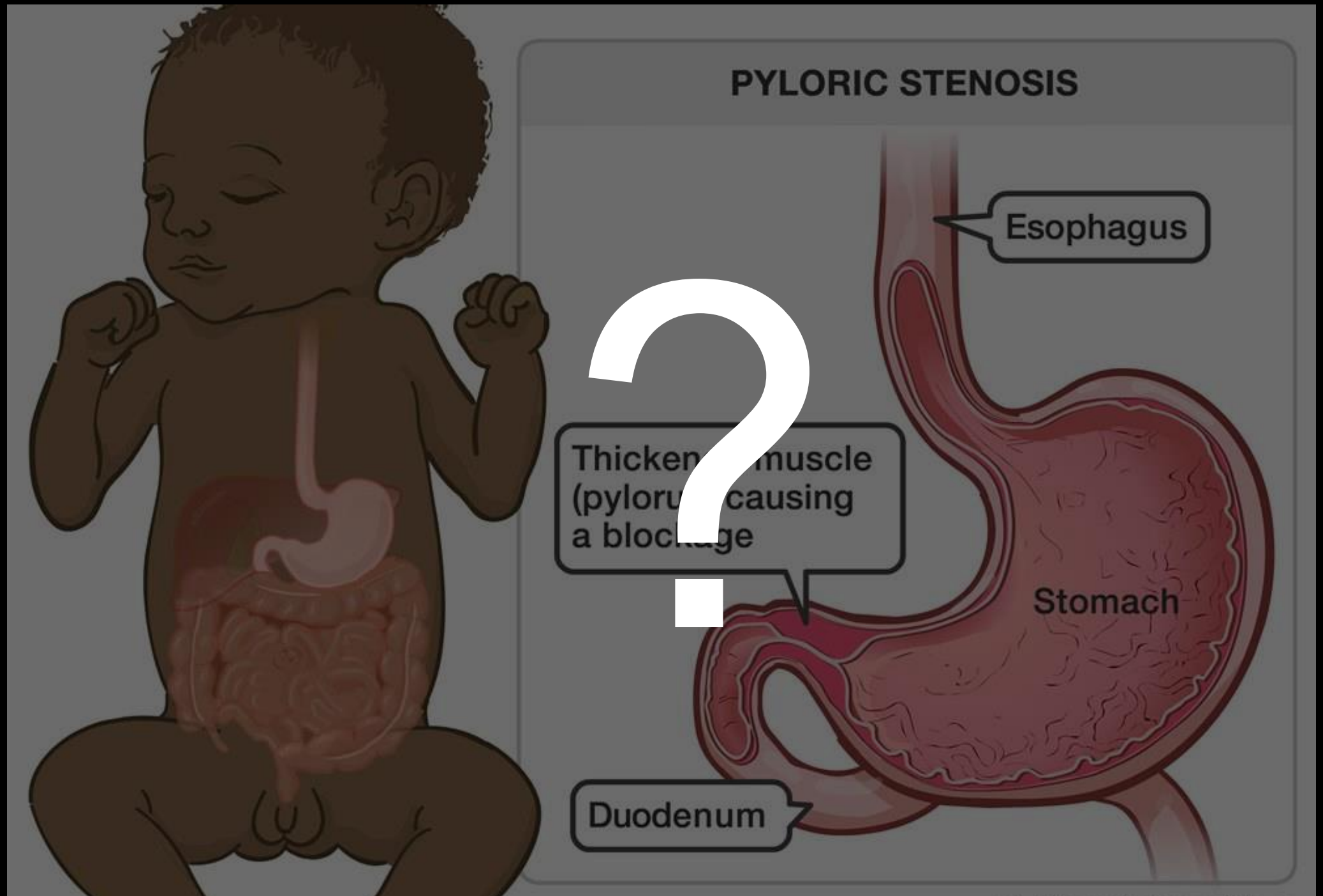
Cardiac

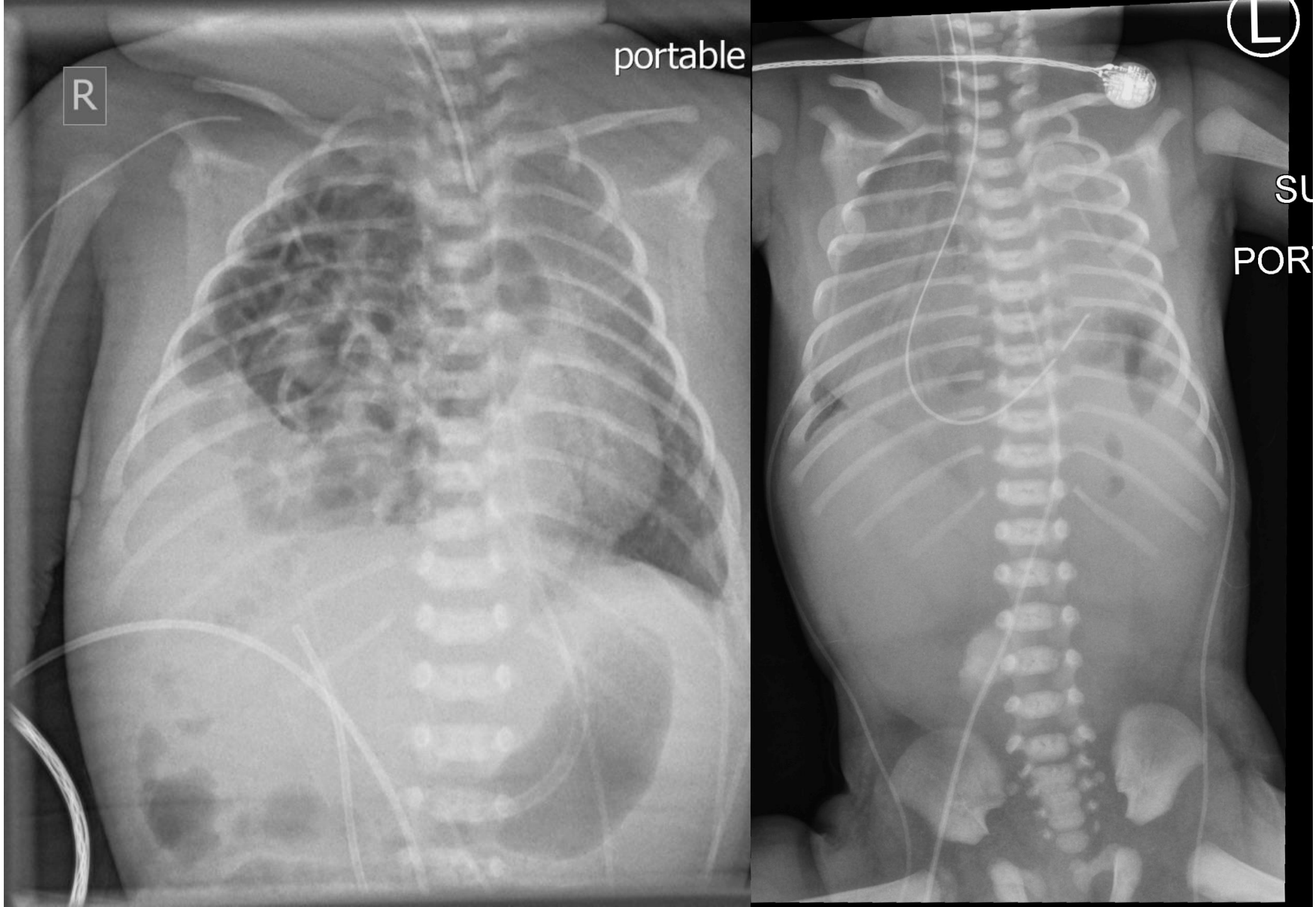
Tracheo**E**sophageal (T-E) fistula/esophageal atresia
Renal
Limb




PYLORIC STENOSIS
Esophagus
Thickened muscle (pylorus) causing a blockage
Stomach
Duodenum


ENTERAL DRAINAGE
MED230


PYLORIC STENOSIS
Esophagus
Thicken muscle
(pyloru causing
a bloc ge
Stomach
Duodenum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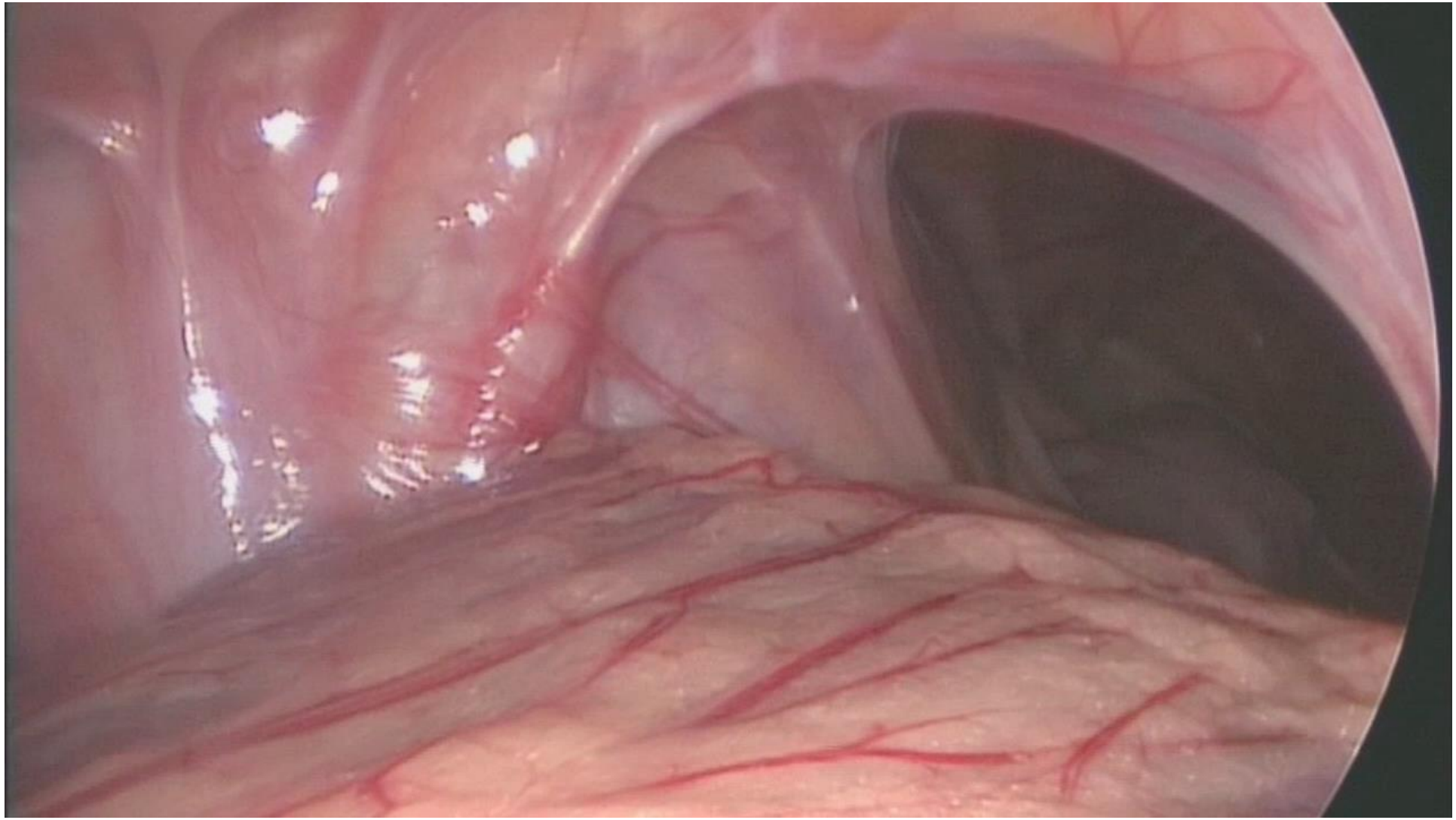

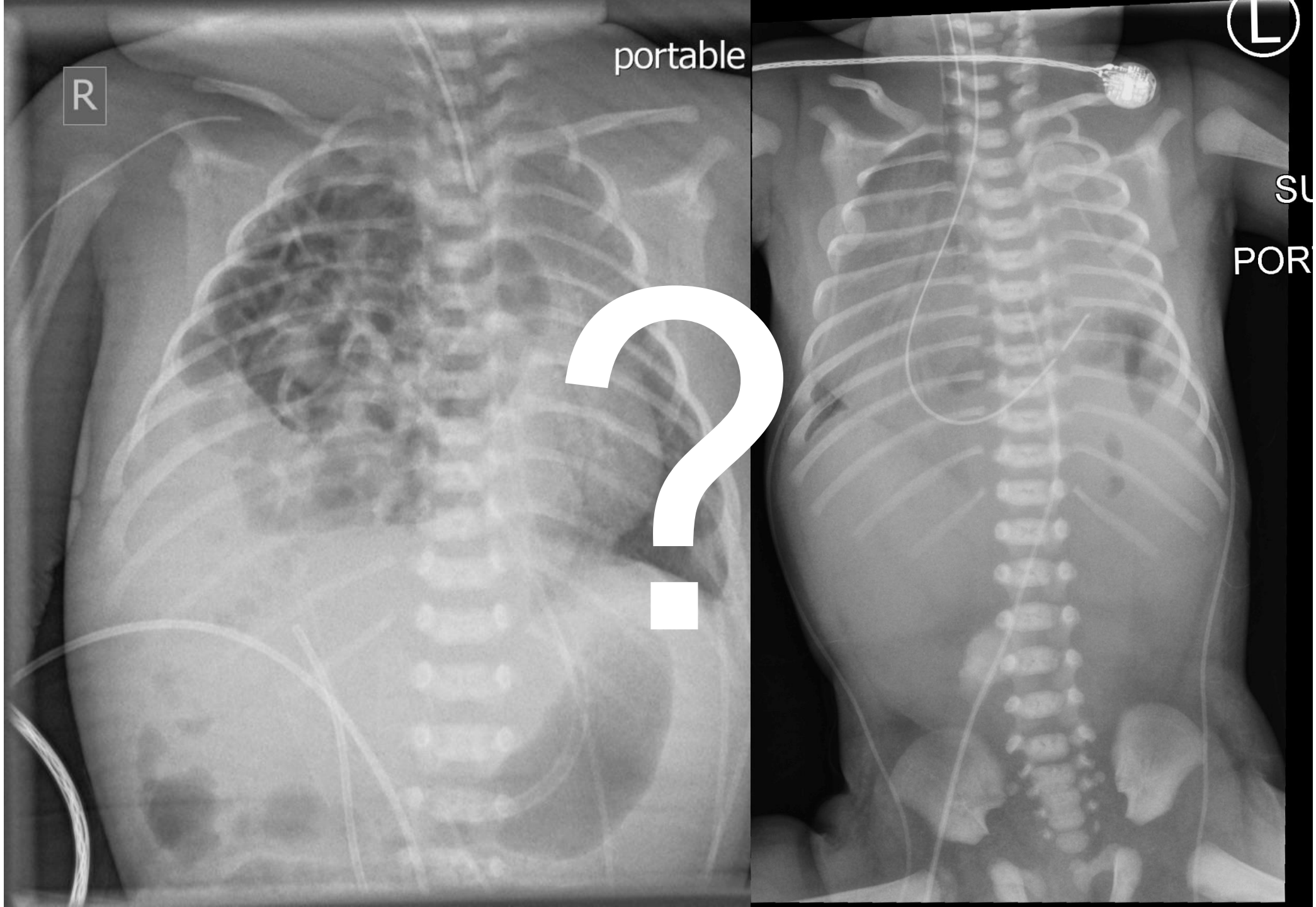
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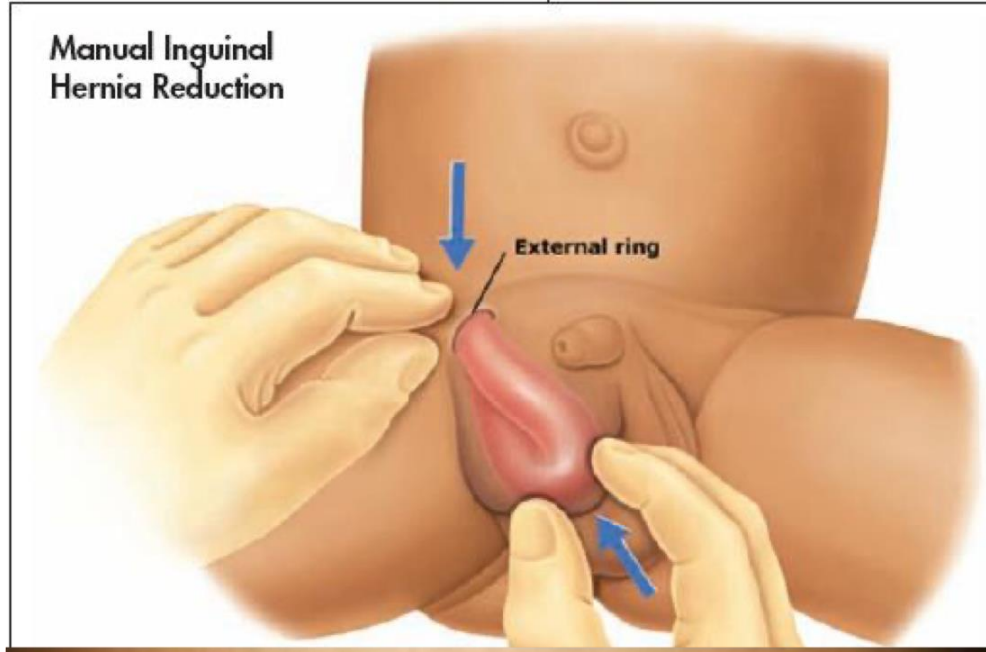
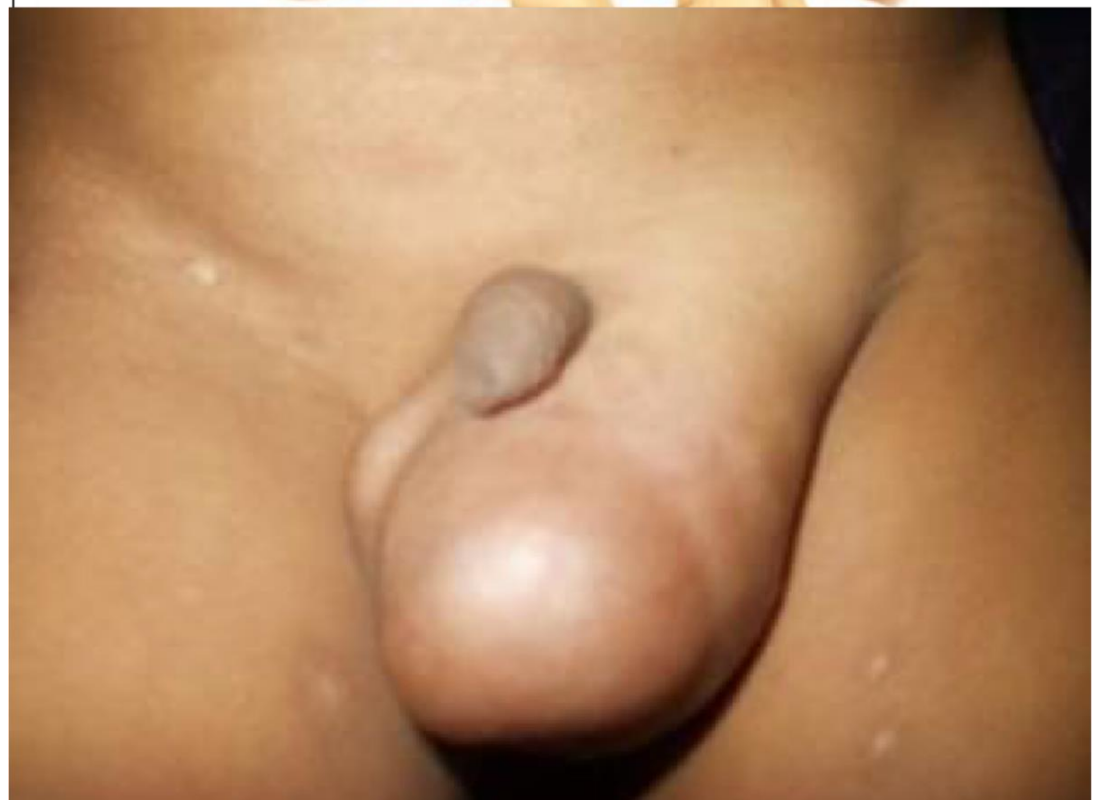

Manual Inguinal
Hernia Reduction
External ring

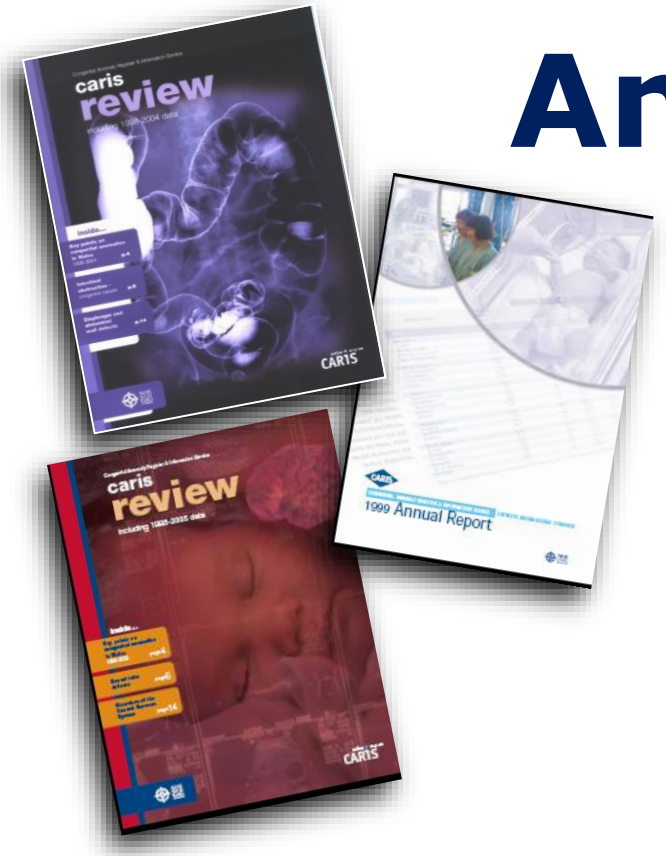


Summary

- To provide the best care possible to every baby and family we look after.
- Overview of gastroschisis, exomphalos, OA/TOF, Py, CDH.
- Understand more about a baby’s surgical journey.

CARIS

Congenital Anomaly Register and Information Service



Annual Meeting 2023

GIG CYMRU NHS WALES

Iechyd Cyhoeddus Cymru

Public Health Wales

Post-operative nutrition

Rebecca Seymour & Emma White

Paediatric Advanced Nurse Practitioners

Nutritional assessment of the infant surgical patient

Erica Thomas

Paediatric Surgical Advanced Nurse Practitioner

Noah's Ark Children's Hospital for Wales.

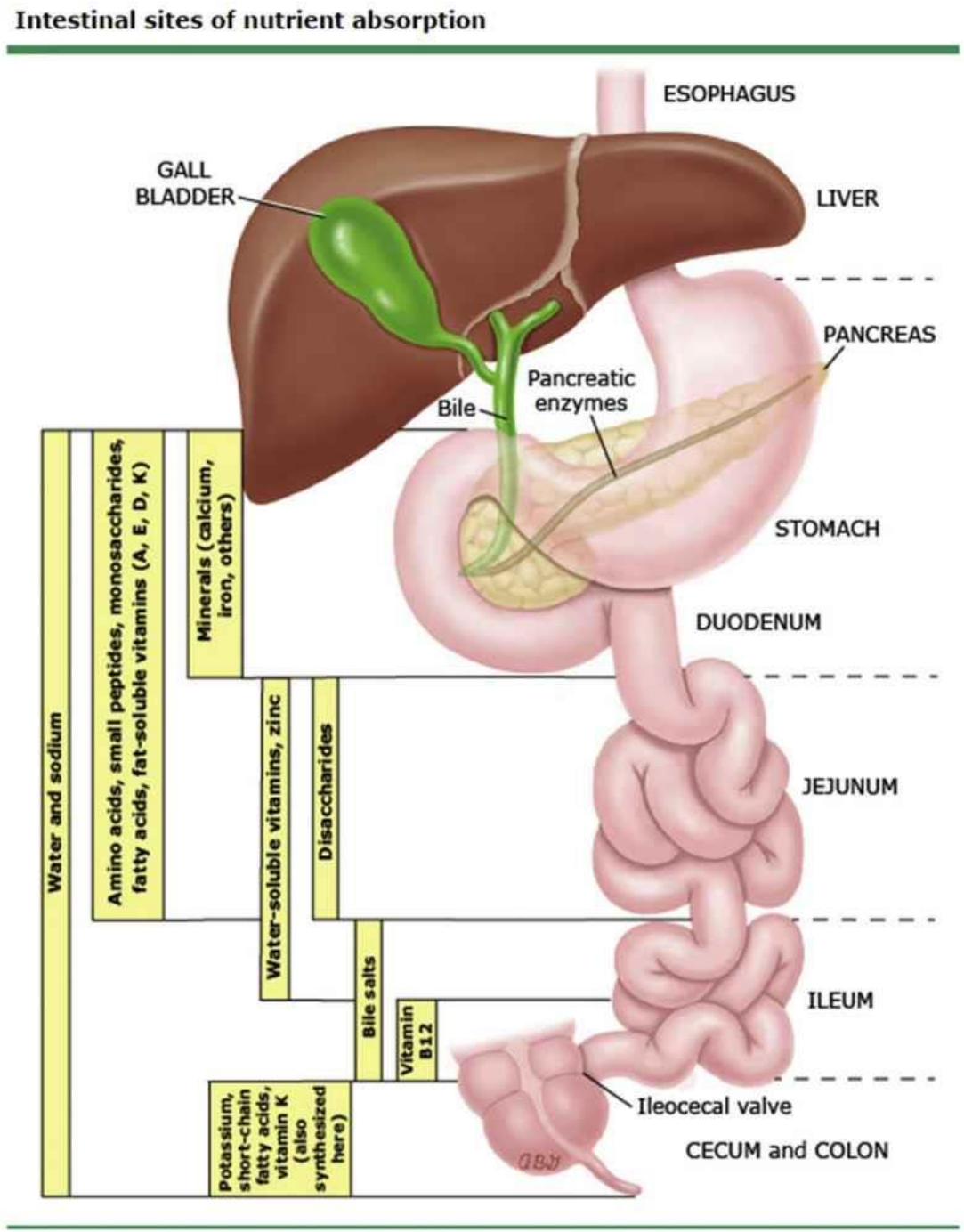
Parents questions?

- Where can I park the car?

- How long will my baby be in hospital?

Congenital malformations

- Points for consideration
  - The type of feed and the method by which it is delivered will be determined by
    - ❖ the area of GI tract affected
    - ❖ the surgery performed to correct the defect
    - ❖ the condition and function of the remaining gut



Ref: Mayer, O. and J. Kerner. (2017) Management of short bowel syndrome In postoperative very low birth weight infants. Seminars in fetal &neonatal medicine (22), 49-56.

Nutritional monitoring

Age of child	male	female
preterm	110-120	110-120
0-1 month	113	107
1-3 month	100	97
3 months – 1 year	80	80
1 -4 years	82	78

Target caloric intake (Kcal/kg/day)

- Good health requires good nutrition
- Calorific requirements in enterally fed infants
- Energy storage is limited
- Early growth deficits which reflect inadequate nutrition have long lasting effects
  - short stature
  - neurodevelopmental delay

Choice of feed

- Breastmilk is recommended when feeds first introduced
- Infants weighing <2kgs pre-term formula
- Protein hydrolysate feed with 50% fat as MCT (Pepti-junior)

- Feed changes should be guided by stool/stoma output, quantity, reducing sugars

Methods of nutrition administration

- Oral
- Nasogastric tube
- Transanostomotic tube
- Gastrostomy
- Naso-jejenal feeding
- Parenteral nutrition

Enteral feeding the benefits!

- Enteral route is the preferred option as it has
  - Fewer infection rates
  - Preserves the gastrointestinal mucosa and immunity
  - Offers better metabolic control
  - Has better long term outcomes
  - Cost

- Which route of delivery?

Oral feeding

- When is a baby ready to feed?
  - Suck-swallow-breath pattern
- Cue-based feeding patterns

ref: Spagnoli (2023), NIHR (2021)

- Gestation – medical intervention
- Oral anatomy – micrognathia, cleft palate, Pierre Robin syndrome
- Oesophagus abnormalities
- Intestinal obstruction

Nasogastric / orogastric tube

- NG tubes are used for gastric decompression following surgery
- Gastric and oesophageal perforation 0.4-0.5% in pre-terms
- Stimulation of naso-oropharynx causing relaxation of oesophageal sphincter and worsening GOR
- Aspiration pneumonia
- Easy displacement of tube

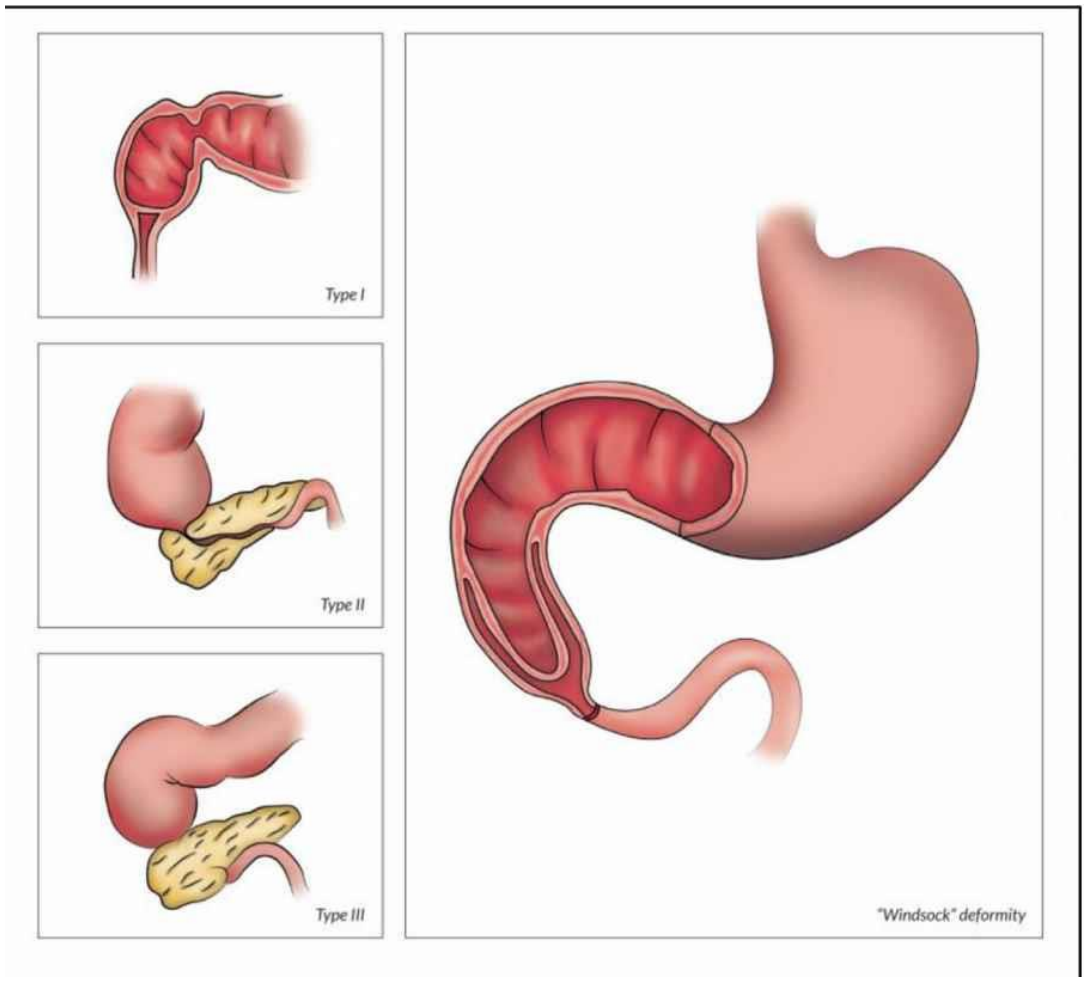
When to start feeds?

- Evaluation of enteral feeding tolerance
- Includes
  - Abdominal distention, vomiting,
  - NG aspirate colour
  - Gastric aspirate volume
  - Stooling patterns and frequency

Naso-jejunal feeding

- In patients that cannot tolerate gastric feeds – what next?
- Indications include significant foregut dysmotility and microgastria
  - Oesophageal atresia
  - GORD
  - Delayed gastric emptying
- Reduced the need for surgery – fundoplication

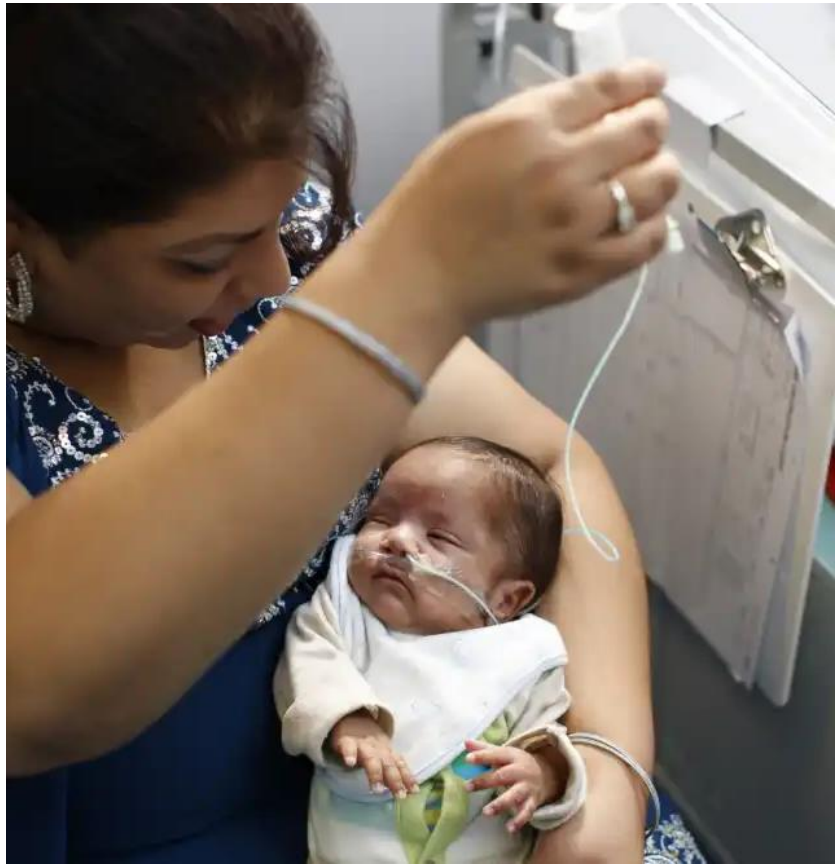
- Problems encountered –
  - requires continuous feeding,
  - tube displacement requires fluoroscopy to confirm placement



Upper Intestinal atresia

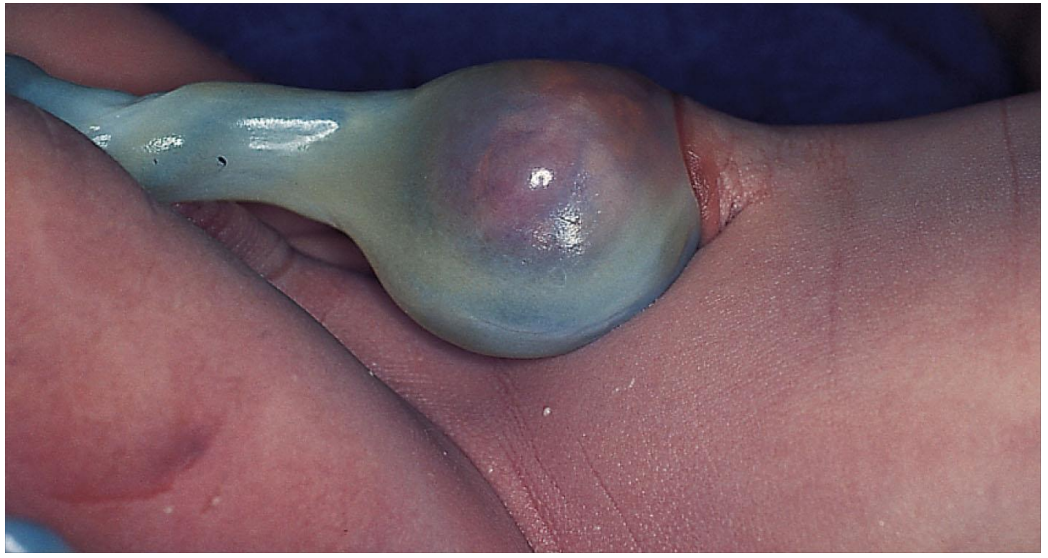
Transanastmotic tube

- TAT is placed at the time of surgery
- The end of the tube is placed past the anastomosis to allow early feeding
  - Oesophageal atresia
  - Duodenal atresia
- In utero duodenum proximal to the atresia is stretched resulting in a baggy segment which can delay feeding
- Can also be used as a stent for tight anastomosis – OA

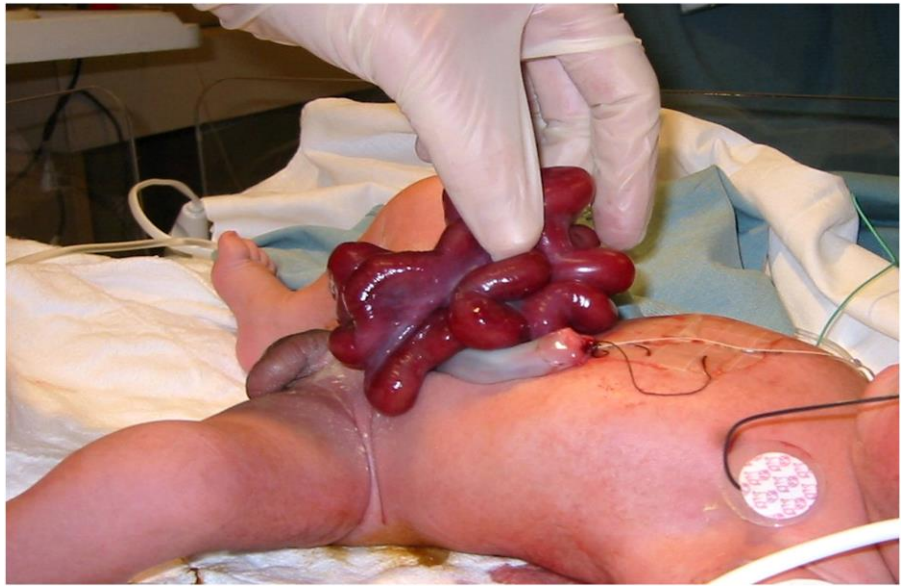


Abdominal wall defects

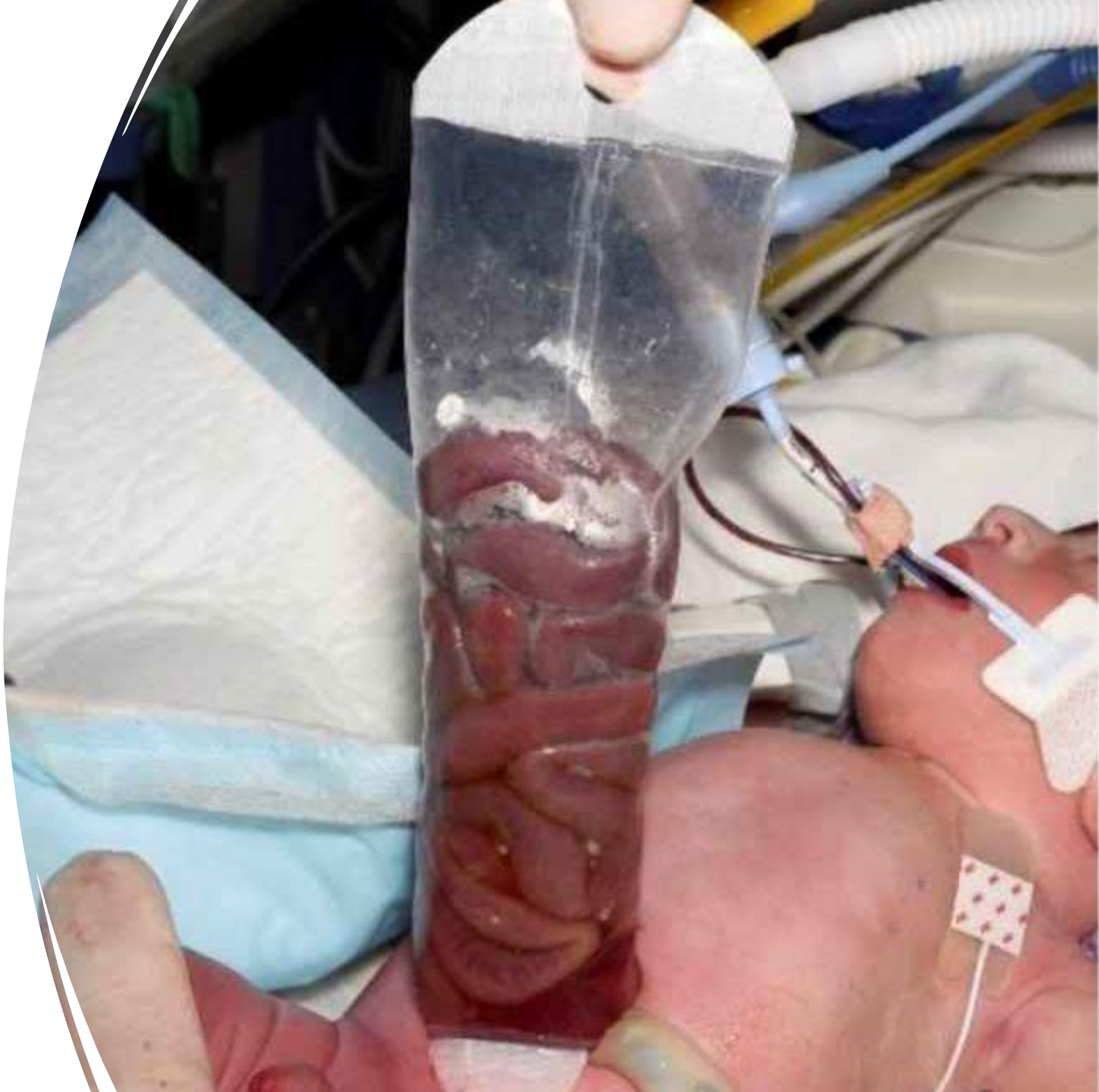
Exomphalos



Gastroschisis



Silo – staged reduction



Special requirements for feeding

80% of Gastroschisis can be closed in a single operation - *simple*

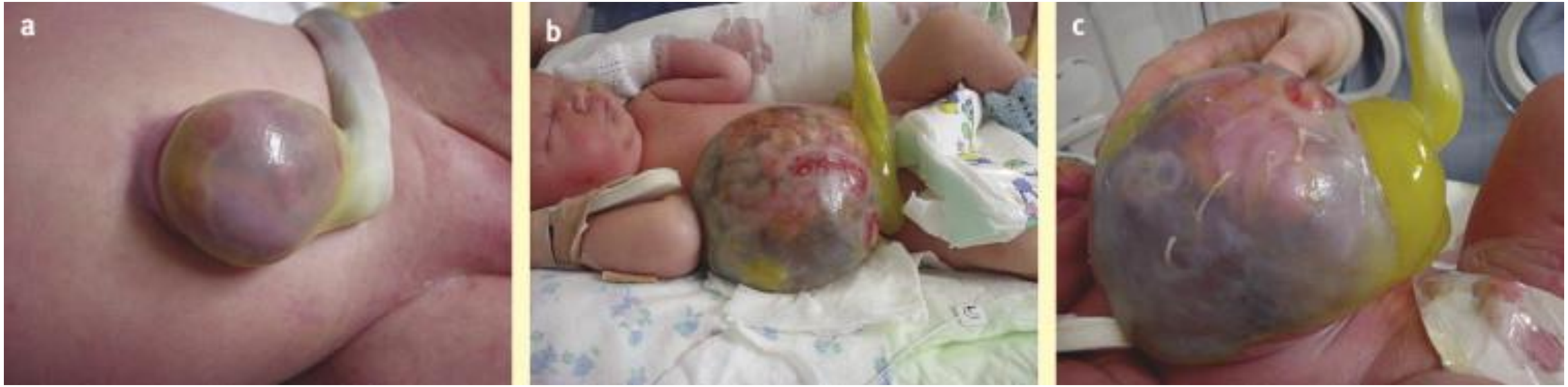
Sluggish motility of the bowel due to exposure to the amniotic fluid and prolonged pressure results in functional ileus

10% have associated intestinal atresia – which may not be evident until multiple failures to introduce enteral nutrition - *complex*

Liver problems related to PN

Enteral feeding is introduced when bowel function returns

Exomphalos



Prognostic factors

- Chromosomal abnormalities – 32%, cardiac defects, trisomy 13 and 18.
- Beckwith-Wiedemann syndrome – macroglossia and hypoglycaemia

- Size of defect –
  - exomphalos minor facial defect <5cm, only intestine
  - exomphalos major fascial defect > 5cm potentially containing liver, midgut, gonads and spleen

Long term morbidity includes gastroesophageal reflux, pulmonary insufficiency and feeding difficulties.

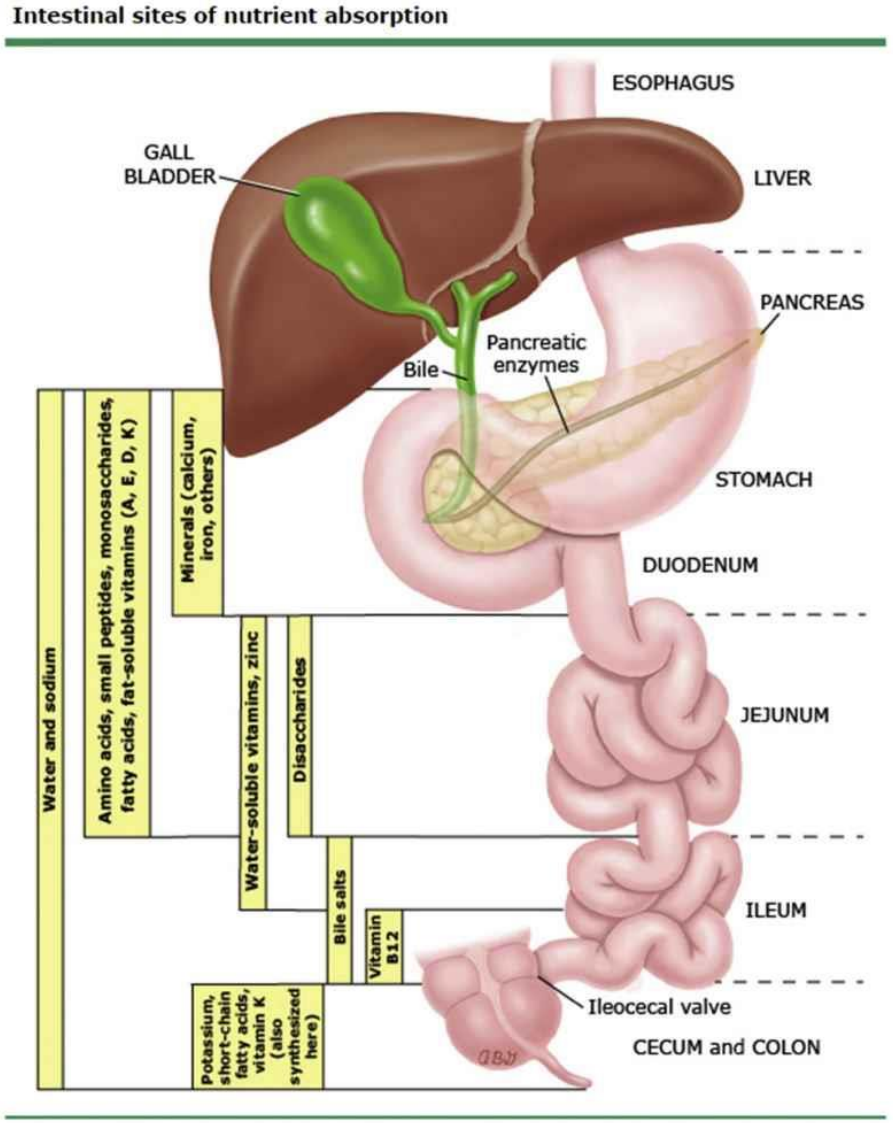
Parenteral Nutrition

- Delivered by PIC line or central
- Parental nutrition (PN) provides nutrition to bridge the gap from placental transfer of nutrients to enteral nutrition
- First used in neonates in 1968
- Biggest influence on the increase in survival rate
- Prolonged PN usage results in significant decrease in intestinal mass, a decrease in mucosal enzyme activity, and increase in gut permeability

Short gut – the consequences

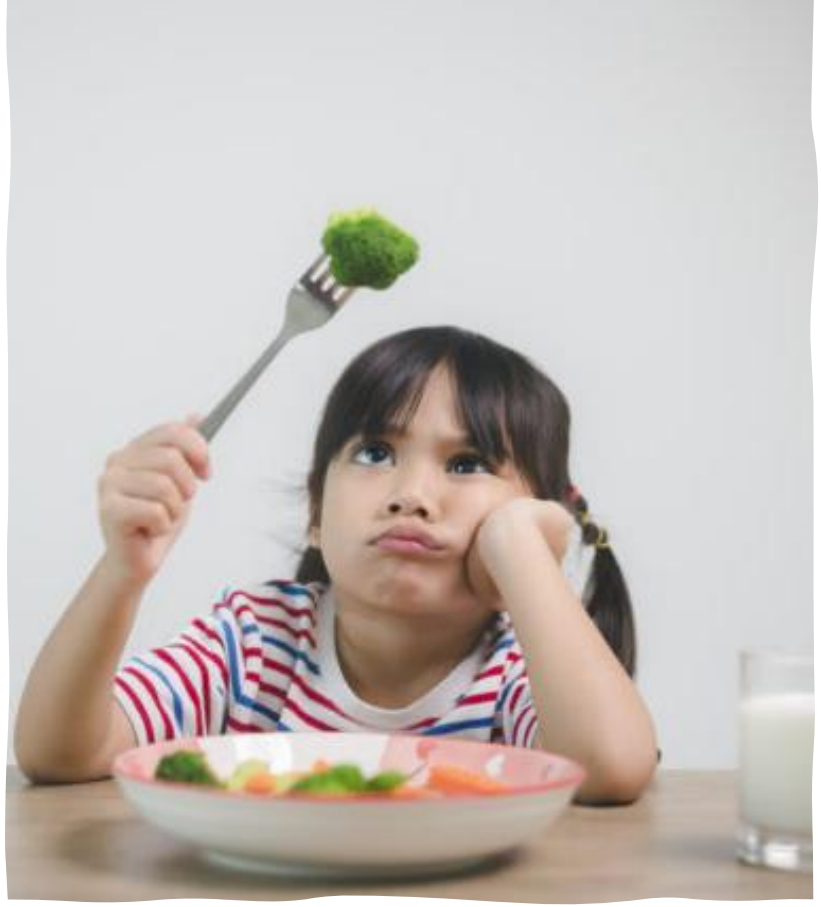
- Collection of disorders where loss of intestinal length that compromises the ability to digest and absorb nutrients
- Pre-term infants have physiological advantage – small bowel doubles in length in last 15 weeks.
- Term infants
  - Length of bowel 250cm +/- 10%
  - >15cm small bowel with IC valve

  Or
  - 40cm without intact IC valve



Factors determining the outcome of short bowel syndrome

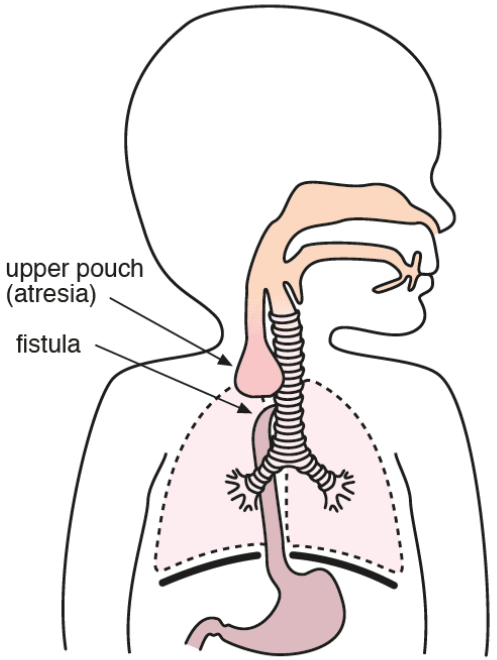
- Length of bowel
- Quality of bowel
- Jejunum versus ileal resection
- Presence or absence of colon
- Complications- liver, sepsis, line access
- Translocation of gut bacteria due to gut stasis



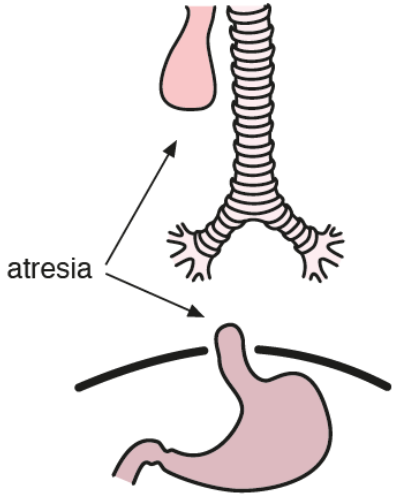
Oesophageal atresia

- A life time of feeding challenges

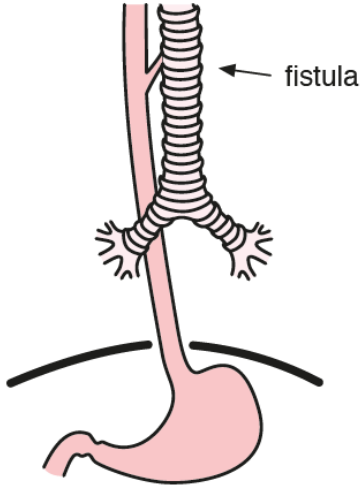
Oesophageal atresia
with lower pouch fistula
(most common type - 85%)



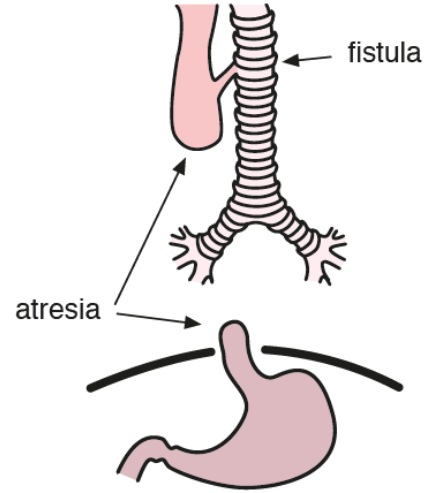
Oesophageal atresia
without fistula
(approximately 8%)



Fistula without atresia
often called H fistula
(approximately 5%)



Oesophageal atresia
with upper pouch fistula
(less than 2%)

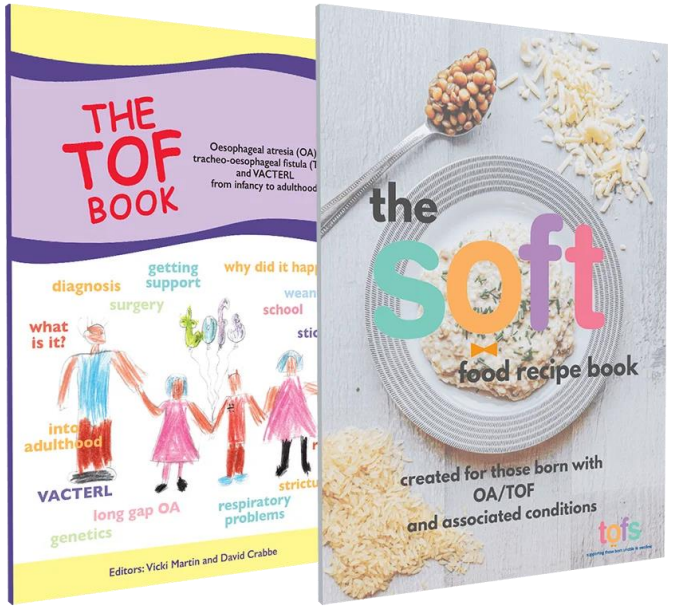


Oeosphageal atresia and Tracheo-oesophageal fistula

- Feeding challenges – related to the surgical option
- Primary anastomosis
- Delayed or staged repair
- Cervical oesophagostomies
  - Sham feeding
- Desensitisation to oral aversion

Oesophageal function after repair

- Gastro-oesophageal reflux
  - Incidence reported 22% to 45%
  - Small stomach – small volume frequent feeds
  - Upright position when feeding and a following feeds
  - Management includes PPI’s
  - Thickening agents – constipation!
  - Nissen fundoplication
- Eosinophilic oesophagitis
- Strictures



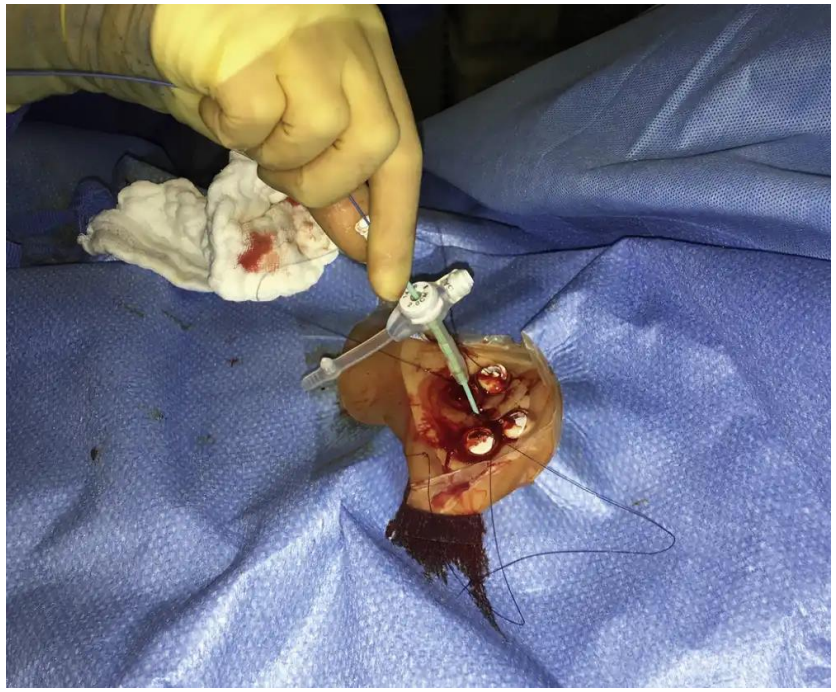
tofs lifelong support for those born unable to swallow

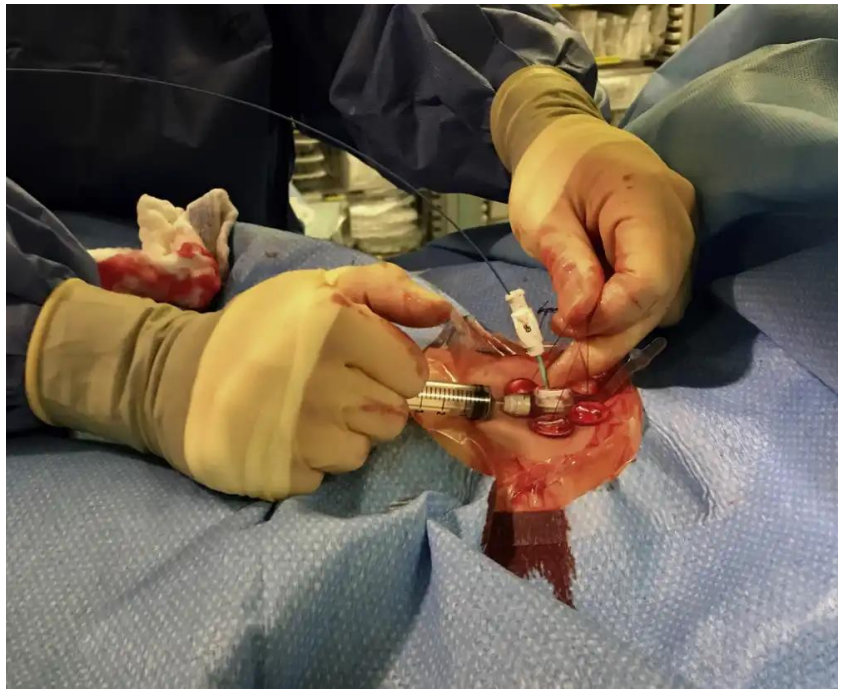
Feeding problems after surgical repair

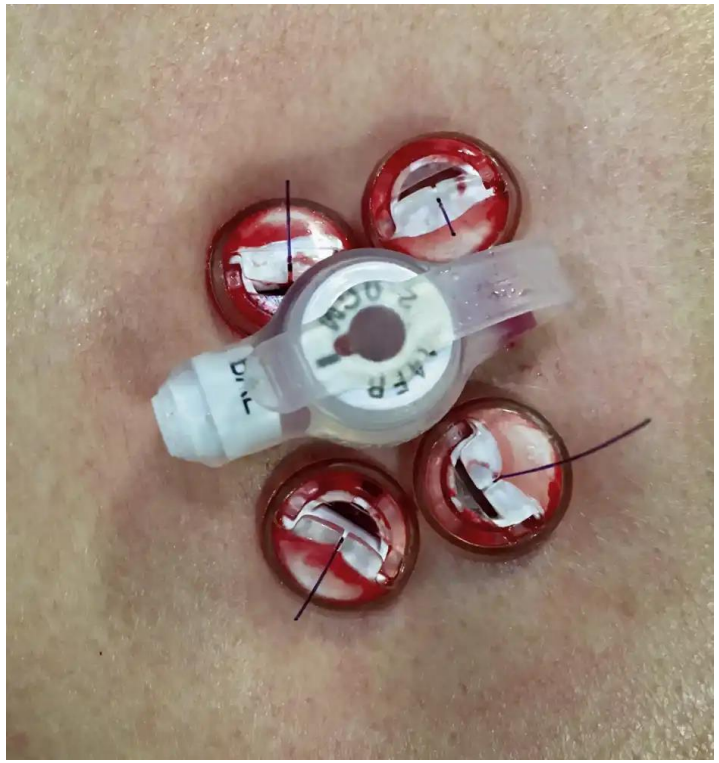
- Meal times are not necessarily pleasurable
  - Chew food – avoid lumps of meat and bread
  - Small frequent meals
  - Drink fluids
- Gastrostomy combination feeding

- Feeding difficulties include
  - Dysphagia
  - Coughing
  - Choking
  - Aspiration – recurrent chest infections
  - Slow feeding
  - Oral food aversion

Laparoscopic Gastrostomy





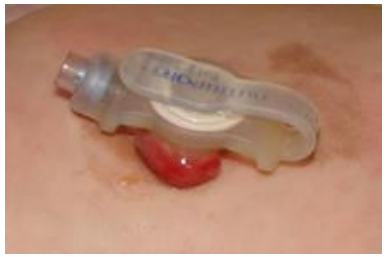


Gastrostomy – button device

Gastrostomy

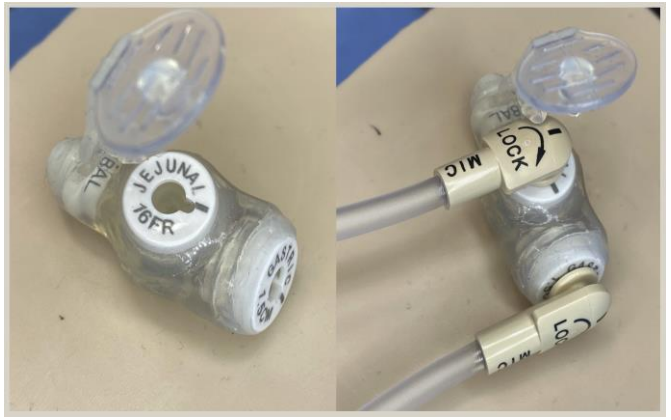
- Indications
- Congenital abnormalities of mouth, oesophagus, or stomach
- Swallowing dysfunction
- Medication administration – metabolic conditions

- Troubleshooting
  - Leaking from stoma site
    - Delayed emptying of stomach
    - Tube displacement
    - Balloon burst
  - Granulation tissue
  - Appropriate length
  - Blockage
    - Inadequate water flush
    - Medication



AMT – Gastro-jejunal feeding tube

Table 1			
FR Size	Min Volume	Recommended Volume	Max Volume
14 F	3 ML	4 ML	5 ML



Learning points in nutritional support

- Nutritional support needs to be tailored to the individual child
- Nutritional support requires the skills of a multidisciplinary nutrition team
- PN is essential to maintaining hydration and nutritional status
- Enteral feeds are the single most important factor in promoting adaptation and should be started early, even if trophic in nature


THANK YOU

CARIS

Congenital Anomaly Register and Information Service

Annual Meeting 2023

GIG CYMRU NHS WALES

Iechyd Cyhoeddus Cymru

Public Health Wales

Please remember to:

Continue reporting cases

Minimum of: NHS Number, date of birth & postcode

- **By email to:** Caris.**Safehavenmailbox**@wales.nhs.uk
- **By internet:** nww2.nphs.wales.nhs.uk:8080/CARISWarningCard.nsf/WarningCardForm?OpenForm
- **By CARIS Cards or Data Forms**

Visit our website for prevalence data:

https://phw.nhs.wales/services-and-teams/caris/

Contacts:

Dr Margery Morgan
margery.morgan@wales.nhs.uk

Dr Llion Davies
llion.davies2@wales.nhs.uk

David Tucker:
david.tucker2@wales.nhs.uk

Email CARIS: CARIS@wales.nhs.uk

- https://phw.nhs.wales/services-and-teams/caris/

Thank you for attending

Please remember to complete the short feedback form that will be dropped in the chat shortly

A certificate of attendance will be issued on request upon completion of this form

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