

	PRENATAL	TIMING	FEATURES	AXR	ASSOCIATIONS
Oes Atresia	Polyhydramnios Small stomach	Immediate	Choking/drooling	Empty or gassy stomach	50% VACTERL
Duo Atresia	Polyhydramnios Large stomach	Immediate	Vomiting – non-bilious	Double bubble	30% Down's
Hy Py stenosis	NAD	2-7wks	Vomit – non-bilious	Large stomach	
Congenital intestinal obstruction	Variable		Vomit –bilious Distension Delayed meconium	Dilated bowel loops	HD commonest Assume VN (laparotomy if in doubt)

### Embryology

Foregut develops from YOLK SAC → folds in embryo in 4<sup>th</sup> week

Pharynx, airway, lungs, oesophagus, duodenum to MDP, HPB system

### VOMITING

Forceful ejection of gastric contents (cf possetting = passive regurgitation)

### GASTRO-OESOPHAGEAL REFLUX

<12mths = immature LOS + short intra-abdo oesophagus + liquid food + reclined posture

>12mths = LOS matures + solid food + upright posture

Diff dx: possetting/other causes of vomiting

Problems: aspiration, peptic stricture, anaemia, failure to thrive

Ix= contrast studies (exclude OA/DA etc), OGD, pH studies

Mx= thickening agents, keep child upright, motility agents, PPI

### VOMITING CHILD

O&G: Age/gestation/BWT

HPc:

Vomiting: (i)onset (ii)bilious vs non-bilious + post-vomit hunger

Bowels: (i)meconium passage and other bowel motions (ii)distension and visible peristalsis

Effects: electrolyte deficiencies, failure to thrive, aspiration, UGIB+ anaemia, stricture, opisthotonus

Causes: ambiguous genitalia (CAH), groin or umbilical lumps (hernia), rash (meningitis), urine (UTI)

PMHx:

FHx: Maternal FHx

Prenatal diagnosis: polyhydramnios causes

First-born male

CAH/syndromes

Diff dx: obstructive (HPS, atresias, HD, malrotation, hernia) vs non-obstructive (meningitis, UTI, CAH, overfeeding)

OE: dehydration/ambiguous genitalia, peristalsis, hernia + RUQ olive/LIF sausage

### DEHYDRATED CHILD

	Gen	Neuro	CRT	HR	BP	PULSES	UO	Turgor	Eyes	Mucosa
DEHYDRATION	Unwell	A/V		+	N	N	-	-	Sunken	Dry
SHOCK	Unwell	V/P/U		++	-	Weak	Oliguria	-	Grossly Sunken	Dry

Types:

Isonatraemic: proportional losses of Na<sup>+</sup>/H<sub>2</sub>O

Hyponatraemia: ICFV up → convulsions so avoid free water

Hypernatraemic : diarrhoea, high insensible water losses

hypertonic ECFV depletes/buffered by ICFV → less marked signs → Na<sup>+</sup> <0.5mls/hr

Management:

ORS 50mls/kg over 4hrs + maintenance

IV Fluids NaCl 0.9% 50mls/kg + maintenance

## HYPERTROPHIC PYLORIC STENOSIS

*Hypertrophic pyloric sphincter causes gastric outlet obstruction*

### Epidemiology:

Age: 2-7wks Gender: 80% male Race: whites>blacks>asians<sub>(rare)</sub>  
3 risk factors: maternal FHx/young mother/first borne male

### Pathology:

Hypertrophy of **circular** muscle

### Features:

(i) Vomiting: **non-bilious** (iii) Failure to thrive  
(ii) Constant hunger (iv) **Dehydration**

### Examination:

Test Feed: **visible peristalsis** from left to right  
Mass: **ovoid** mass in **RUQ** (midway between umbilicus and ribcage)

### Investigations:

Bloods: **electrolyte** deficiencies/**uraemia**  
US: pylorus >**16mm** long/**3mm** thick  
Barium: if doubt diagnosis (differentiate from atresias etc)

Paradoxical aciduria

Compensatory phase:

Vomiting with open pylorus → lose H+Cl-Na+K+ in gastric juice/bile/pancreatic juice/intestine

Kidneys excrete HCO<sub>3</sub><sup>-</sup> to compensate → alkaline urine

Decompensatory phase:

Na<sup>+</sup> deficit → H<sup>+</sup> & K<sup>+</sup> preferentially excreted with HCO<sub>3</sub><sup>-</sup> → hypochloraemia, hypokalaemic alkalosis

Advanced alkalosis with acid urine → hypoca<sup>2+</sup> and tetany

### Management:

1. Correct **fluid/electrolyte** imbalances (Hartmann's 20ml/kg then 9ml/kg/hr)

2. Ramsted's **pyloromyotomy**: cut circular muscle but not mucosa of pylorus  
stay 1mm short of Mayo's vein at pylorus/D1 serosal demarcation

\*feed next day and discharge\*

## OESOPHAGUS

### OESOPHAGEAL ATRESIA

*Definition: proximal and distal oesophagus fail to communicate*

Upper end = dilated pouch with hypertrophic muscle wall || Lower end= atretic pouch with thin muscle wall

#### Pathology:

EA + DISTAL TOF 84%

ISOLATED EA 8%

ISOLATED TOF 4%

EA + PROX + DISTAL TOF 3%

EA AND PROX TOF 1%

#### Gross' Classification

A= ISOLATED EA

B= EA + PROX TOF

C= EA + DISTAL TOF

D= EA + PROX + DISTAL TOF

E= ISOLATED TOF

#### Features:

Pre-natal: **polyhydramnios + absent stomach**

Neonatal: **choking/drooling** (aspirates)

TOF: air enters stomach when baby coughs/cries and **aspirates** gastric secretions into lungs

Associations: 50% VACTERL; tracheomalacia → seal-bark cough

#### Investigations:

Pre-natal= **polyhydramnios + small stomach**

Neonatal: CXR/AXR= site of **NG hold-up + stomach gas** presence<sub>(TOF)</sub>/absence<sub>(isolated EA)</sub>

Contrast= fistula/gapogram

Endoscopy= OGD/bronchoscopy

#### Management:

Pre-op: NBM, keep upright, O<sub>2</sub>, iv fluids, abx, vit K, TPN

Temporising: **Stamm gastrostomy**: allow early enteral feeding

Definitive: **ligate TEF/EA** primary **anastomosis** or **interposition**

### DUODENAL ATRESIA

#### Aetiology:

Most sporadic; 1/3 have Trisomy 21

#### Pathology:

**Failed vacuolation** of duodenum at D1/2 → proximal hypertrophy/distal narrowing

Associated with biliary/pancreatic abnormalities eg annular pancreas

Week 4: **epithelial tube** of caudal foregut and cranial midgut surrounded by **mesenchyme**

Week 6: epithelium proliferates to **obliterate** lumen

Week 10: epithelial apoptosis → **vacuolation** → recanalization

#### Classification:

1. **Membrane** traverses lumen 2. Atretic ends connected by **fibrous cord** 3. Complete **separation** of atretic ends

#### Features:

Pre-natal: polyhydramnios + dilated stomach

Neonatal: bilious vomiting

#### Investigations:

Pre-natal: amniocentesis for **Trisomy 21**; US= **polyhydramnios** and **dilated stomach**

Neonatal: AXR: **double-bubble** stomach with no air distally

Others: contrast to differentiate from other atresias; rectal biopsy to exclude Hirschsprung's

Diff Dx: oesophageal atresia (choking/drooling, stomach size but both have polyhydramnios)

HPS (non-bilious vomiting without polyhydramnios, later onset, palpable mass)

Congenital intestinal obstructions (dilated bowel loops on AXR)

#### Management:

WEB: duodenotomy or endoscopic excision

ATRESIAS: bypass (**duodenoduodenostomy** or duodenojejeunal bypass; former has earlier recovery of function)

## DIAPHRAGMATIC HERNIA

### Normal development:

Formed by fusion of several structures in week 5-7

Septum transversum → central tendon (head folds → ventrally/caudally → anterior diaphragm (long phrenic nerve course))

Pleuroperitoneal membranes from somites in C3-5 segments close primitive communication between cavities

Lateral muscular ingrowth from body wall completes diaphragm muscle

### Congenital Defects:

Foramen of Morgagni: between xiphoid and costal origins → present later as small hernia with little effect

Foramen of Bochdalek: defect in pleuroperitoneal segment (T18,21; posterior and left; pulmonary hypoplasia)

Central tendon defect:

Oesophageal hiatus enlarged:

### Acquired:

Trauma: blunt force → left > right (liver protective)

Sliding/rolling

## GASTROSCHISIS

Isolated anomaly to right of midline

## EXOMPHALOS

Via congenital umbilical hernia