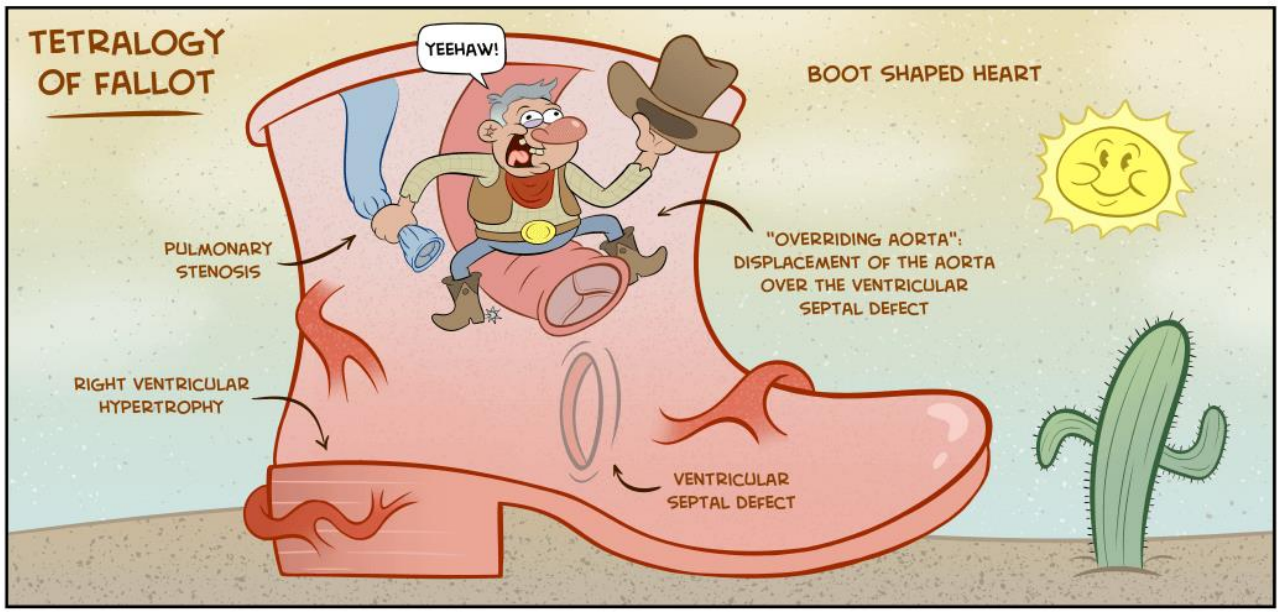


Congenital Heart Disease

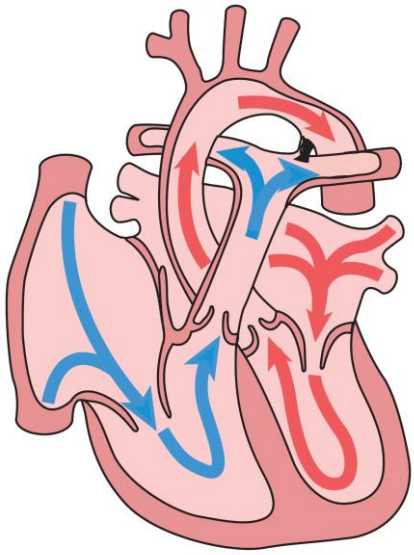
Acyanotic - most common causes

- Ventricular septal defects (VSD) - most common, accounts for 30%
- Atrial septal defect (ASD)
- Patent ductus arteriosus (PDA)
- Coarctation of the aorta
- Aortic valve stenosis

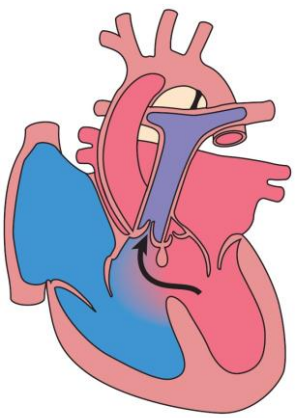
VSDs are more common than ASDs. However, in adult patients ASDs are the more common new diagnosis as they generally present later.

Cyanotic - most common causes

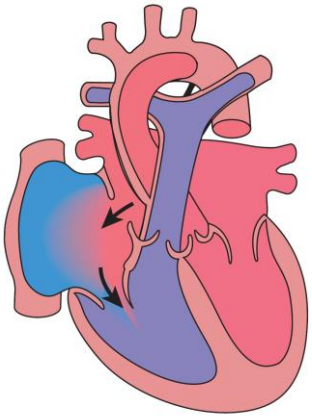
- Tetralogy of Fallot
- Transposition of the great arteries (TGA)
- Tricuspid atresia
- Pulmonary valve stenosis



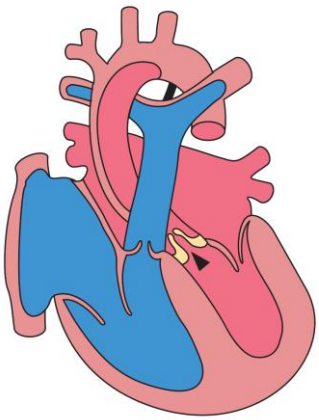
Postnatal circulation



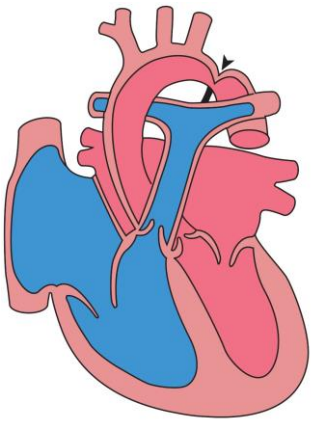
Ventricular septal defect



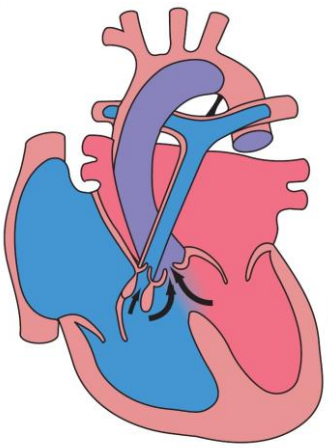
Secundum atrial septal defect



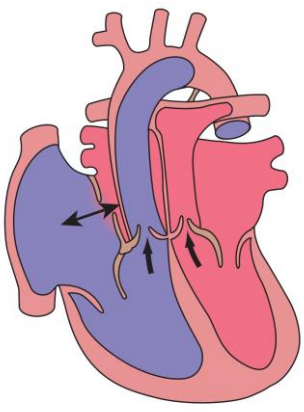
Aortic stenosis



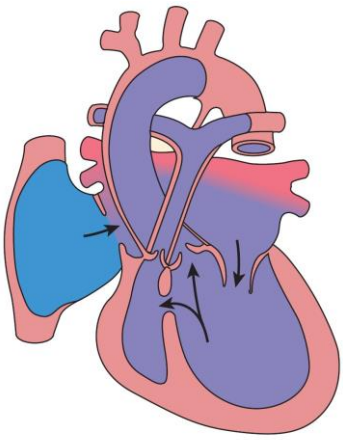
Adult-type coarctation of the aorta



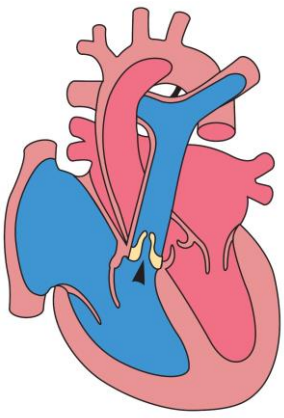
Tetralogy of Fallot



Complete transposition of the great arteries



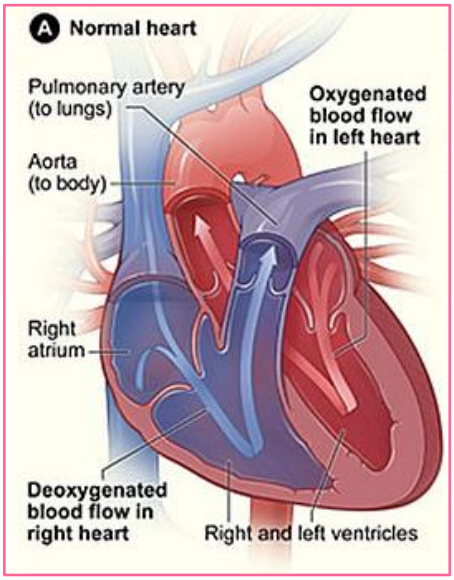
Tricuspid atresia



Pulmonary valve stenosis

Tetralogy of Fallot (TOF)

Tetralogy of Fallot (TOF) is the most common cause of cyanotic congenital heart disease*. It typically presents at around 1-2 months, although may not be picked up until the baby is 6 months old



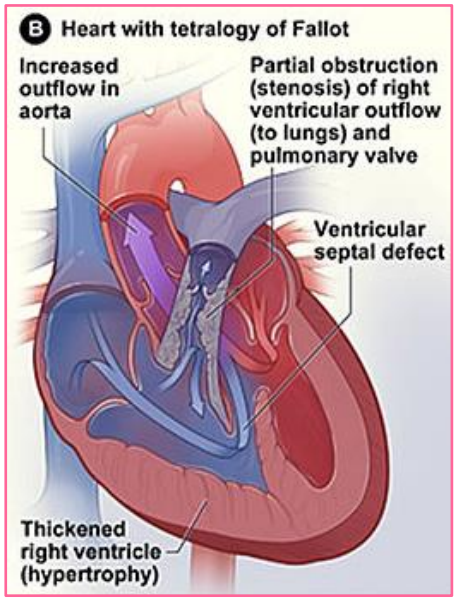
The four characteristic features are:

- Ventricular septal defect (VSD)
- Right ventricular hypertrophy
- Right ventricular outflow tract obstruction, pulmonary stenosis
- Overriding aorta

The severity of the right ventricular outflow tract obstruction determines the degree of cyanosis and clinical severity

Other features

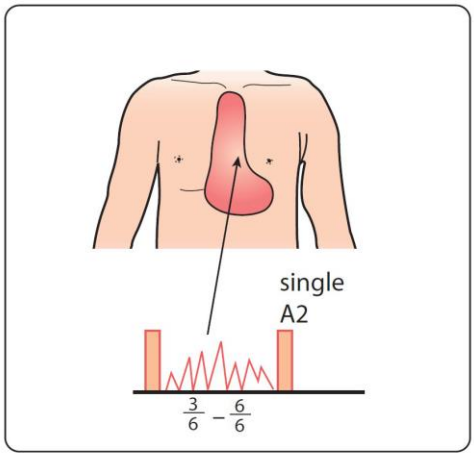
- Cyanosis
- **Right-to-left** shunt
- Ejection systolic murmur due to pulmonary stenosis (the VSD doesn't usually cause a murmur)
- A right-sided aortic arch is seen in 25% of patients
- Chest x-ray shows a 'boot-shaped' heart, ECG shows right ventricular hypertrophy

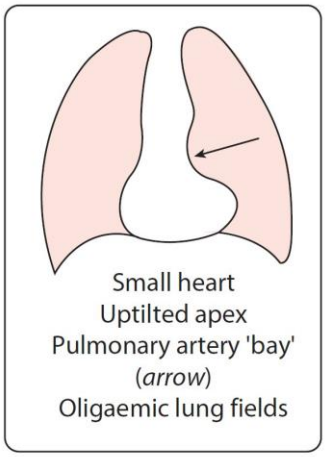


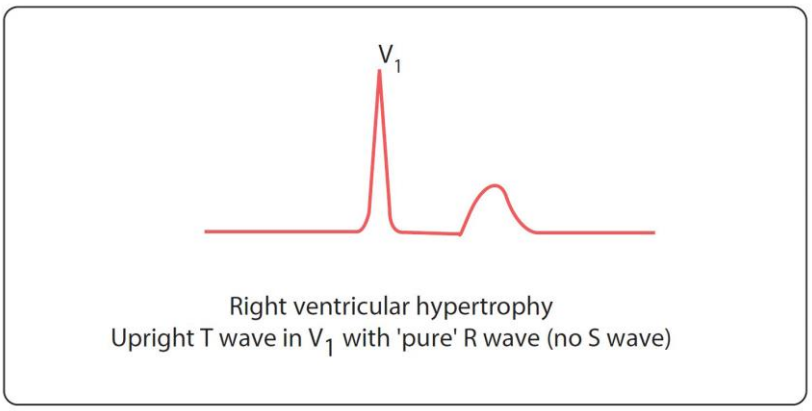
Management

- Surgical repair is often undertaken in two parts
- Cyanotic episodes may be helped by beta-blockers to reduce infundibular spasm

**However, at birth transposition of the great arteries is the more common lesion as patients with TOF generally present at around 1-2 months*







Paediatric GI Bleeding

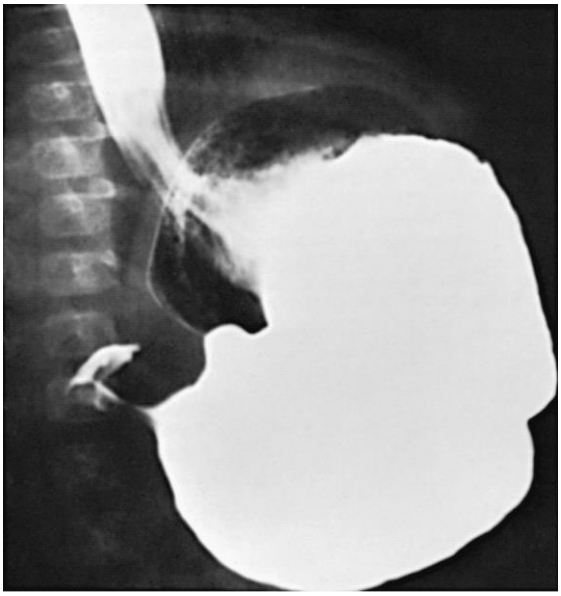
Site	Newborn	1 month to 1 year	1 to 2 years	Older than 2 years
Upper GI tract	• Haemorrhagic disease • Swallowed maternal blood	• Oesophagitis • Gastritis	• Peptic ulcer disease	• Varices
Lower GI tract	• Anal fissure • NEC	• Anal fissure • Intussusception	• Polyps • Meckel's diverticulum	• IBD • Polyps • Intussusception

Paediatric Gastrointestinal Disorders

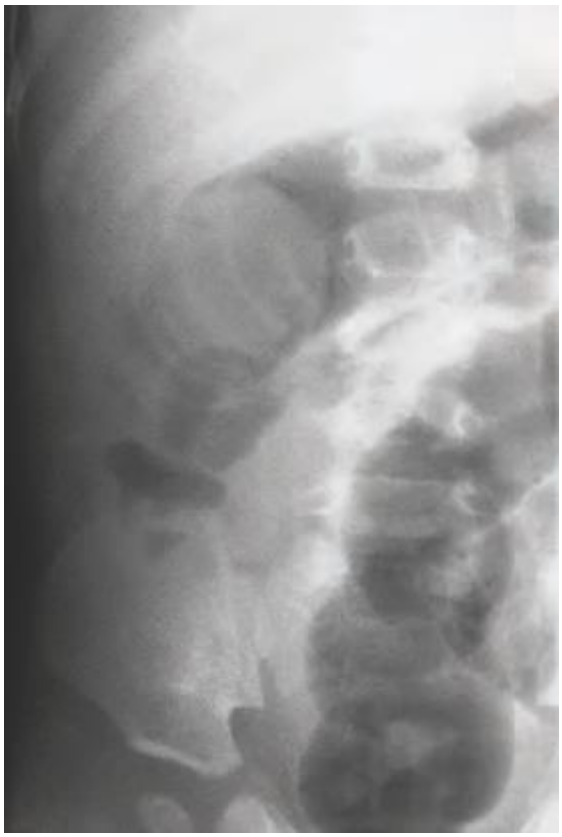
Pyloric stenosis	• M>F • 5-10% Family history in parents • Projectile non bile stained vomiting at 4-6 weeks of life • Diagnosis is made by test feed or USS • Treatment: Ramstedt pyloromyotomy (open or laparoscopic)
Acute appendicitis	• Uncommon under 3 years • When occurs may present atypically
Mesenteric adenitis	• Central abdominal pain and URTI • Conservative management
Intussusception	• Telescoping bowel • Proximal to or at the level of, ileocaecal valve • 6-9 months of age • Colicky pain, diarrhoea and vomiting, sausage shaped mass, red jelly stool. • Treatment: reduction with air insufflation
Malrotation	• High caecum at the midline • Feature in exomphalos, congenital diaphragmatic hernia, intrinsic duodenal atresia • May be complicated by development of volvulus, infant with volvulus may have bile stained vomiting • Diagnosis is made by upper GI contrast study and USS • Treatment is by laparotomy, if volvulus is present (or at high risk of occurring then a Ladd's procedure is performed
Hirschsprung's disease	• Absence of ganglion cells from myenteric and submucosal plexuses • Occurs in 1/5000 births • Full thickness rectal biopsy for diagnosis • Delayed passage of meconium and abdominal distension • Treatment is with rectal washouts initially, thereafter an anorectal pull through procedure
Oesophageal atresia	• Associated with tracheo-oesophageal fistula and polyhydramnios • May present with choking and cyanotic spells following aspiration • VACTERL* associations
Meconium ileus	• Usually delayed passage of meconium and abdominal distension • Majority have cystic fibrosis • X-Rays may not show a fluid level as the meconium is viscid (depends upon feeding), PR contrast studies may dislodge meconium plugs and be therapeutic • Infants who do not respond to PR contrast and NG N-acetyl cysteine will require surgery to remove the plugs
Biliary atresia *(see later)*	• Jaundice > 14 days • Increased conjugated bilirubin • Urgent Kasai procedure
Necrotising enterocolitis	• Prematurity is the main risk factor • Early features include abdominal distension and passage of bloody stools • X-Rays may show pneumatosis intestinalis and evidence of free air • Increased risk when empirical antibiotics are given to infants beyond 5 days • Treatment is with total gut rest and TPN, babies with perforations will require laparotomy

***VACTERL:** *vertebral abnormalities, anal atresia, cardiac abnormalities, tracheoesophageal fistula and/or esophageal atresia, renal agenesis and dysplasia, and limb defects [association]*

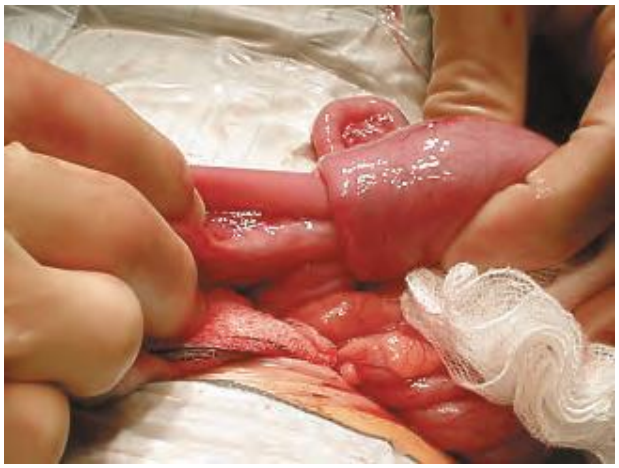
- ***Gastroschisis:*** Isolated abnormality, bowel lies outside abdominal wall through defect located to right of umbilicus.
- ***Exomphalos (Omphalocele):*** Liver and gut remain covered with membranous sac connected to umbilical cord. It is associated with other developmental defects.



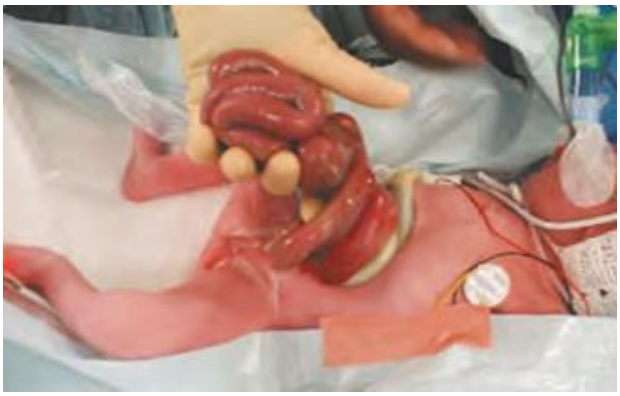
Barium in the stomach of an infant with projectile vomiting. The attenuated pyloric canal is typical of congenital hypertrophic pyloric stenosis.



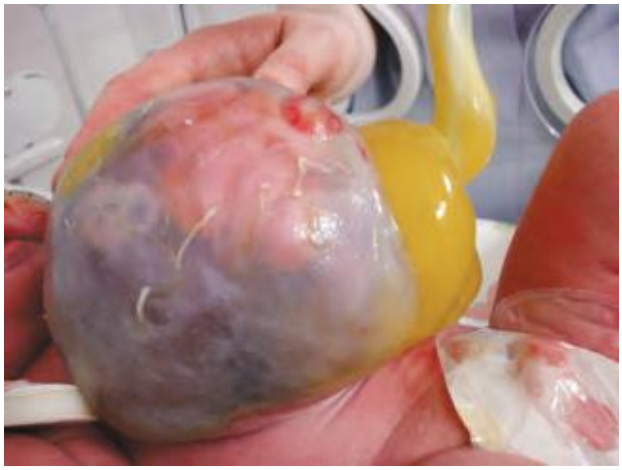
Air enema reduction of an intussusception (the arrows mark the soft tissue shadow of the intussusceptum).



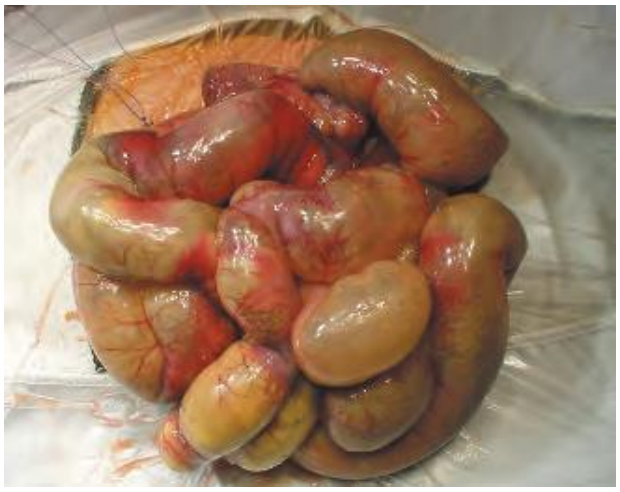
Ileocolic intussusception causing small bowel obstruction.



A newborn infant with gastroschisis.



Exomphalos major.

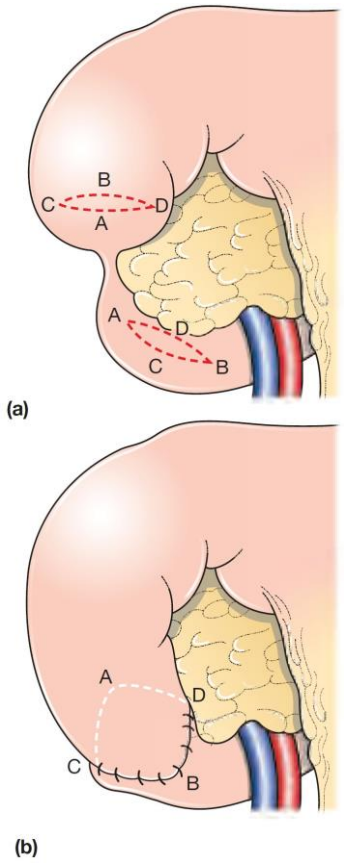


Operative appearance of neonatal necrotising enterocolitis.

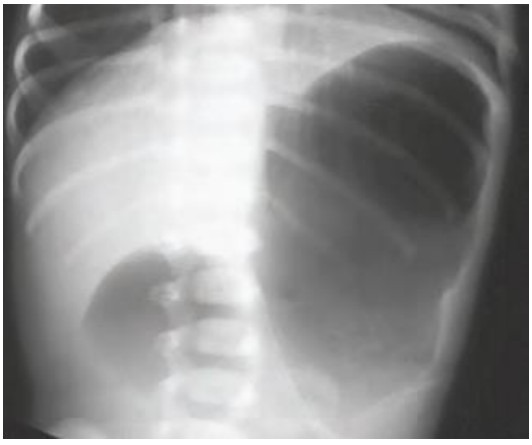
Bilious Vomiting in Neonates

Causes of intestinal obstruction with bilious vomiting in neonates

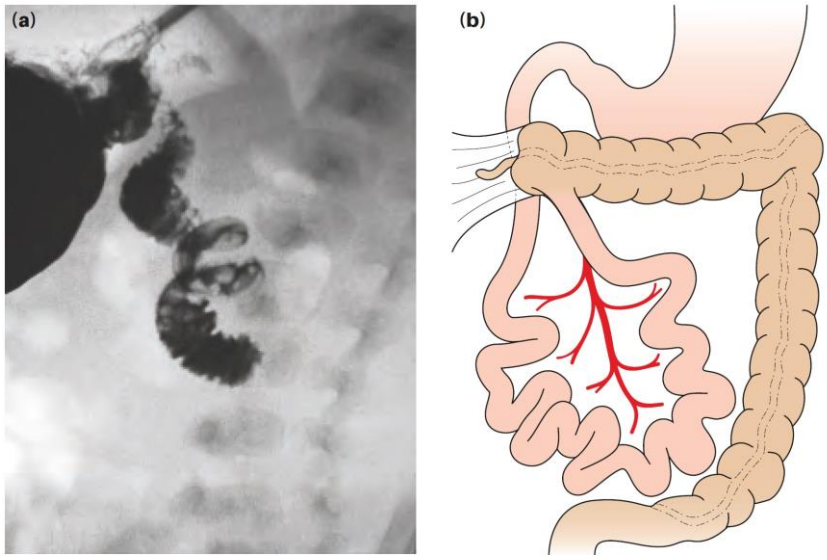
Disorder	*Incidence & causation*	*Age at presentation*	*Diagnosis*	*Treatment*
Duodenal atresia	1 in 5000 (higher in Downs syndrome)	Few hours after birth	AXR shows "**double bubble"** sign, contrast study may confirm	Duodenoduodenostomy
Malrotation with volvulus	Usually cause by incomplete rotation during embryogenesis	Usually 3-7 days after birth, volvulus with compromised circulation may result in peritoneal signs and haemodynamic instability	Upper GI contrast study may show DJ flexure is more medially placed, USS may show abnormal orientation of SMA and SMV	Ladd's procedure
Jejunal/ ileal atresia	Usually caused by vascular insufficiency in utero, usually 1 in 3000	Usually within 24 hours of birth	AXR will show air-fluid levels	Laparotomy with primary resection and anastomosis
Meconium ileus	Occurs in between 15 and20% of those babies with cystic fibrosis, otherwise 1 in 5000	Typically in first 24-48 hours of life with abdominal distension and bilious vomiting	Air - fluid levels on AXR, sweat test to confirm cystic fibrosis	Surgical decompression, serosal damage may require segmental resection
Necrotising enterocolitis	Up to 2.4 per 1000 births, risks increased in prematurity and inter-current illness	Usually second week of life	Dilated bowel loops on AXR, pneumatosis and portal venous air	Conservative and supportive for non-perforated cases, laparotomy and resection in cases of perforation of ongoing clinical deterioration



(a, b) Duodenal atresia and the incisions used to repair it: a diamond anastomosis is shown



Neonatal abdominal x-ray showing the 'double bubble' of duodenal atresia.



(a) Contrast showing malrotation with a volvulus.
(b) The narrow origin of the small bowel mesentery predisposes to midgut volvulus.

Causes of vomiting

Infants

Gastro-oesophageal reflux
Feeding problems
Infection:
- Gastroenteritis
- Respiratory tract/otitis media
- Whooping cough (pertussis)
- Urinary tract
- Meningitis

Food allergy and food intolerance
Eosinophilic oesophagitis
Intestinal obstruction:
- Plyoric stenosis
- Atresia – duodenal, other sites
- Intussusception
- Malrotation
- Volvulus
- Duplication cysts
- Strangulated inguinal hernia
- Hirschsprung disease

Inborn errors of metabolism
Congenital adrenal hyperplasia
Renal failure

Preschool children

Gastroenteritis
Infection:
- Respiratory tract/otitis media
- Urinary tract
- Meningitis
- Whooping cough (pertussis)

Appendicitis
Intestinal obstruction:
- Intussusception
- Malrotation
- Volvulus
- Adhesions
- Foreign body – bezoar

Raised intracranial pressure
Coeliac disease
Renal failure
Inborn errors of metabolism
Torsion of the testis

School age and adolescents

Gastroenteritis
Infection – including pyelonephritis, septicaemia, meningitis
Peptic ulceration and *H. pylori* infection
Appendicitis
Migraine
Raised intracranial pressure
Coeliac disease
Renal failure
Diabetic ketoacidosis
Alcohol/drug ingestion or medications
Cyclical vomiting syndrome
Bulimia/anorexia nervosa
Pregnancy
Torsion of the testis

Causes of acute abdominal pain

Surgical

Acute appendicitis
Intestinal obstruction including intussusception
Inguinal hernia
Peritonitis
Inflamed Meckel diverticulum
Pancreatitis
Trauma

Intra-abdominal

Medical

Non-specific abdominal pain
Gastroenteritis
Urinary tract:
- urinary tract infection
- acute pyelonephritis
- hydronephrosis
- renal calculus

Henoch–Schönlein purpura
Diabetic ketoacidosis
Sickle cell disease
Hepatitis
Inflammatory bowel disease
Constipation
Recurrent abdominal pain of childhood
Gynaecological in pubertal females
Psychological
Lead poisoning
Acute porphyria (rare)
Unknown

Extra-abdominal

Upper respiratory tract infection
Lower lobe pneumonia
Torsion of the testis
Hip and spine

Biliary Atresia

- 1 in 17,000 affected
- Biliary tree lumen is obliterated by an inflammatory cholangiopathy causing progressive liver damage

Clinical features

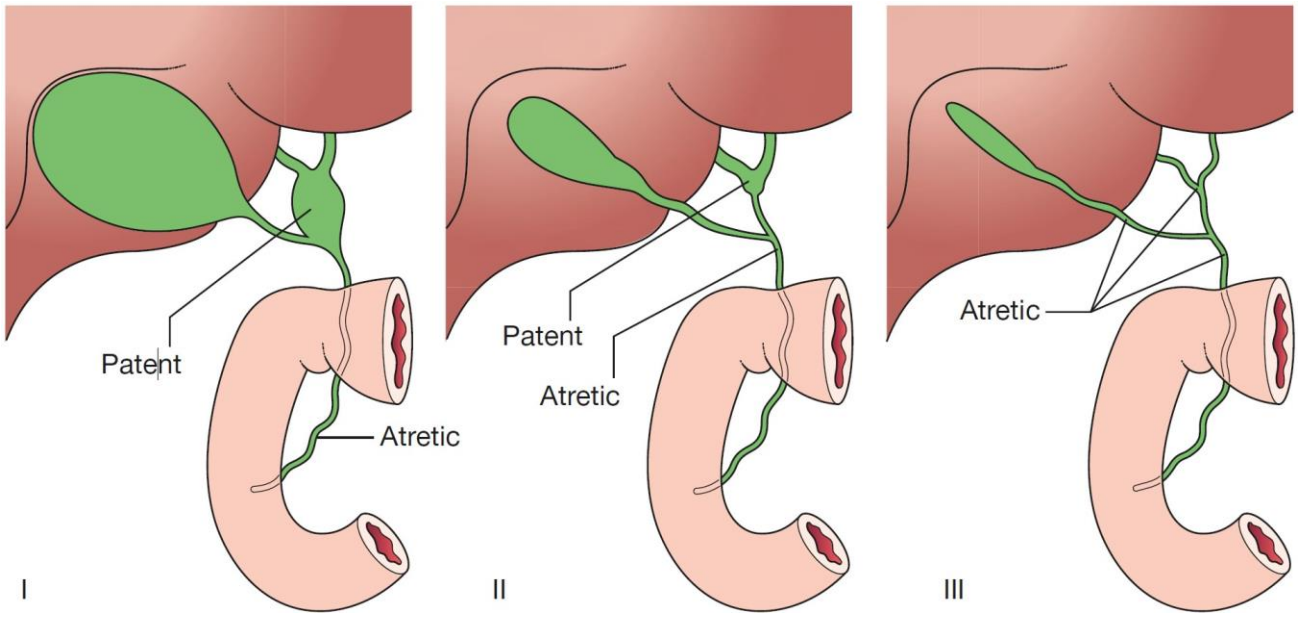
- Infant well in 1st few weeks of life
- No family history of liver disease
- Jaundice in infants > 14 days in term infants (>21 days in pre term infants)
- Pale stool, yellow urine (colourless in babies)
- Associated with cardiac malformations, polysplenia, situs inversus

Investigation

- Conjugated bilirubin (prolonged physiological jaundice or breast milk jaundice will cause a rise in unconjugated bilirubin, whereas those with obstructive liver disease will have a rise in conjugated bilirubin)
- Ultrasound of the liver (excludes extrahepatic causes, in biliary atresia infant may have tiny or invisible gallbladder)
- Hepato-iminodiacetic acid radionuclide scan (good uptake but no excretion usually seen)

Management

- Early recognition is important to prevent liver transplantation.
- Nutritional support.
- Roux-en-Y portojejunostomy (Kasai procedure).
- If Kasai procedure fails or late recognition, a liver transplant becomes the only option.



Classification of biliary atresia. Gallbladder filling provides a clue to the type of atresia.

- *type I: atresia restricted to the common bile duct;*
- *type II: atresia of the common hepatic duct;*
- *type III: atresia of the right and left hepatic ducts.*

Paediatric Umbilical Disorders

Embryology

During development the umbilicus has two umbilical arteries and one umbilical vein. The arteries are continuous with the internal iliac arteries and the vein is continuous with the falciform ligament (ductus venosus). After birth the cord dessicates and separates and the umbilical ring closes.

Umbilical hernia

Up to 20% of neonates may have an umbilical hernia, it is more common in premature infants. The majority of these hernias will close spontaneously (may take between 12 months and three years). Strangulation is rare.

Paraumbilical hernia

These are due to defects in the linea alba that are in close proximity to the umbilicus. The edges of a paraumbilical hernia are more clearly defined than those of an umbilical hernia. They are less likely to resolve spontaneously than an umbilical hernia.

Omphalitis

This condition consists of infection of the umbilicus. Infection with *Staphylococcus aureus* is the commonest cause. The condition is potentially serious as infection may spread rapidly through the umbilical vessels in neonates with a risk of portal pyaemia, and portal vein thrombosis. Treatment is usually with a combination of topical and systemic antibiotics.

Umbilical granuloma

These consist of cherry red lesions surrounding the umbilicus, they may bleed on contact and be a site of seropurulent discharge. Infection is unusual and they will often respond favorably to chemical cautery with topically applied silver nitrate.

Persistent urachus

This is characterised by urinary discharge from the umbilicus. It is caused by persistence of the urachus which attaches to the bladder. They are associated with other urogenital abnormalities.

Persistent vitello-intestinal duct

This will typically present as an umbilical discharge that discharges small bowel content. Complete persistence of the duct is a rare condition. Much more common is the persistence of part of the duct (Meckels diverticulum). Persistent vitello-intestinal ducts are best imaged using a contrast study to delineate the anatomy and are managed by laparotomy and surgical closure.

Paediatric Inguinal Hernia

Inguinal hernias are a common disorder in children. They are commoner in males as the testis migrates from its location on the posterior abdominal wall, down through the inguinal canal. A patent processus vaginalis may persist and be the site of subsequent hernia development.

- Children presenting in the first few months of life are at the highest risk of strangulation and the hernia should be repaired urgently.
- Children over 1 year of age are at lower risk and surgery may be performed electively.
- For paediatric hernias a herniotomy without implantation of mesh is sufficient.
- Most cases are performed as day cases, neonates and premature infants are kept in hospital overnight as there is a recognised increased risk of post-operative apnoea.

Meckel's Diverticulum

- Congenital abnormality resulting in incomplete obliteration of the **vitello-intestinal duct**
- Normally, in the foetus, there is an attachment between the vitello-intestinal duct and the yolk sac.This disappears at 6 weeks gestation.
- The tip is free in majority of cases.
- Associated with enterocystomas, umbilical sinuses, and omphaloileal fistulas.
- Arterial supply: **omphalomesenteric (vitelline)** artery.
- 2% of population, 2 inches long, 2 feet from the ileocaecal valve.
- Typically lined by ileal mucosa but ectopic gastric mucosa can occur, with the risk of peptic ulceration. Pancreatic and jejunal mucosa can also occur.

Clinical

- Normally asymptomatic and an incidental finding.
- Complications are the result of obstruction, ectopic tissue, or inflammation.
- Removal if narrow neck or symptomatic. Options are between wedge excision or formal small bowel resection and anastomosis.

Paediatric Fluid Management

Since 2000 there have been at least 4 reported deaths from fluid induced hyponatraemia in children. This led to the National Patient Safety Agency introducing revised guidelines in 2007.
Indications for IV fluids include:

- Resuscitation and circulatory support
- Replacing on-going fluid losses
- Maintenance fluids for children for whom oral fluids are not appropriate
- Correction of electrolyte disturbances

Fluids to be avoided

Outside the neonatal period saline / glucose solutions should not be given. The greatest risk is with saline 0.18 / glucose 4% solutions. The report states that 0.45% saline / 5% glucose may be used. But preference should be given to isotonic solutions and few indications exist for this solution either.

Fluids to be used

- 0.9% saline
- 5% glucose (though only with saline for maintenance and not to replace losses)
- Hartmann's solution

Potassium should be added to maintenance fluids according patients plasma potassium levels (which should be monitored).

Intraoperative fluid management

Neonates should receive glucose 10% during surgery.
Other children should receive isotonic crystalloid.

Maintenance fluids

Weight	*Water requirement/kg/day*	*Na mmol/kg/day*	*K mmol/kg/day*
First 10Kg body weight	100ml	2-4	1.5-2.5
Second 10Kg body weight	50ml	1-2	0.5-1.5
Subsequent Kg	20ml	0.5-1.0	0.2-0.7

Glucose will need to be given to neonates- usually 10% at a rate of 60ml/Kg/day.

Paediatric Fluid Management

Since 2000 there have been at least 4 reported deaths from fluid induced hyponatraemia in children. This led to the National Patient Safety Agency introducing revised guidelines in 2007.
Indications for IV fluids include:

- Resuscitation and circulatory support
- Replacing on-going fluid losses
- Maintenance fluids for children for whom oral fluids are not appropriate
- Correction of electrolyte disturbances

Fluids to be avoided

Outside the neonatal period saline / glucose solutions should not be given. The greatest risk is with saline 0.18 / glucose 4% solutions. The report states that 0.45% saline / 5% glucose may be used. But preference should be given to isotonic solutions and few indications exist for this solution either.

Fluids to be used

- 0.9% saline
- 5% glucose (though only with saline for maintenance and not to replace losses)
- Hartmann's solution

Potassium should be added to maintenance fluids according patients plasma potassium levels (which should be monitored).

Intraoperative fluid management

Neonates should receive glucose 10% during surgery.
Other children should receive isotonic crystalloid.

Maintenance fluids

Weight	*Water requirement/kg/day*	*Na mmol/kg/day*	*K mmol/kg/day*
First 10Kg body weight	100ml	2-4	1.5-2.5
Second 10Kg body weight	50ml	1-2	0.5-1.5
Subsequent Kg	20ml	0.5-1.0	0.2-0.7

Glucose will need to be given to neonates- usually 10% at a rate of 60ml/Kg/day.

Meckel's Diverticulum

- Congenital abnormality resulting in incomplete obliteration of the **vitello-intestinal duct**
- Normally, in the foetus, there is an attachment between the vitello-intestinal duct and the yolk sac.This disappears at 6 weeks gestation.
- The tip is free in majority of cases.
- Associated with enterocystomas, umbilical sinuses, and omphaloileal fistulas.
- Arterial supply: **omphalomesenteric (vitelline)** artery.
- 2% of population, 2 inches long, 2 feet from the ileocaecal valve.
- Typically lined by ileal mucosa but ectopic gastric mucosa can occur, with the risk of peptic ulceration. Pancreatic and jejunal mucosa can also occur.

Clinical

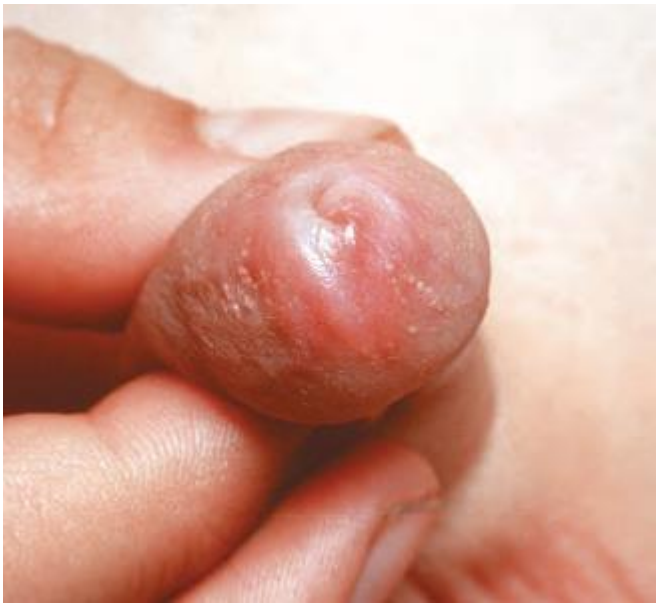
- Normally asymptomatic and an incidental finding.
- Complications are the result of obstruction, ectopic tissue, or inflammation.
- Removal if narrow neck or symptomatic. Options are between wedge excision or formal small bowel resection and anastomosis.

Paediatric Urology - Foreskin disorders

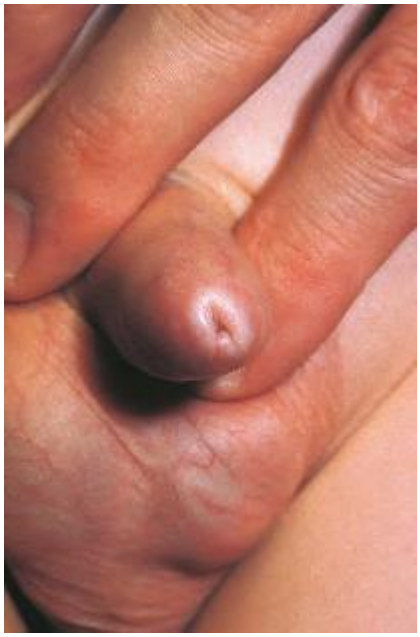
Disorders of the foreskin

At birth and in the neonatal period the normal foreskin is non retractile due to the presence of adhesions between the foreskin and glans. In most cases these will separate spontaneously. By the end of puberty 95% of foreskins can be retracted. In some children the non-retractile foreskin may balloon during micturition. This is a normal variant and requires no specific treatment.

Balanitis	This is **inflammation of the glans penis**. It may occur in both circumcised and non-circumcised individuals.
Balanitis xerotica obliterans	This is a dermatological condition in which **scarring** of the foreskin occurs leading to phimosis. It is rare below the age of 5 years. Treatment is usually with circumcision.
Posthitis	This is **inflammation of the foreskin**. It may occur as a result of infections such as gonorrhoea and other STD's. It may also complicate diabetes. Posthitis may progress to phimosis and as this may make cleaning of the glans difficult and allow progression to balanoposthitis.
Phimosis	This is **inability to retract the foreskin** and may be partial or complete. It may occur secondary to balanoposthitis or balanitis xerotica obliterans. Depending upon the severity and symptoms treatment with circumcision may be required.
Paraphimosis	**Prolonged retraction** of the foreskin proximal to the glans may allow **oedema** to occur. This may then make foreskin manipulation difficult. It can usually be managed by compression to reduce the oedema and replacement of the foreskin. Where this fails a dorsal slit may be required and this followed by delayed circumcision.



Phimosis: *Non-retractile foreskin. The patient presented with a recent history of symptoms. Examination reveals thickening and scarring with a true phimosis*



Balanitis xerotica obliterans *causing true phimosis*

Bronchogenic Cysts

Overview

Bronchogenic cysts most commonly arise as a result of anomalous development of the ventral foregut. They are most commonly single, although multiple cysts are described.

They often lie near the midline and most frequently occur in the region of the carina. They may be attached to the tracheobronchial tree, although they are seldom in direct connection with it.

Cases may be asymptomatic or present with respiratory symptoms early in the neonatal period.

They are the second most common type of foregut cysts (after enterogenous cysts) in the middle mediastinum. Up to 50% of cases are diagnosed prior to 15 years of age.

Investigation

Many cases are diagnosed on antenatal ultrasound. Others may be detected on conventional chest radiography as a midline spherical mass or cystic structure. Once the diagnosis is suspected a CT scan should be performed.

Treatment

Thorascopic resection is the ideal treatment. Very young babies can be operated on once they reach six weeks of age.

Urinary Tract Infection - Paediatric

- UTI's may occur in 5% of 0young girls and 1-2% males. The incidence is higher in premature infants.
- E-Coli accounts for 80% cases.
- In children with UTI it is important to establish whether there is underlying urinary stasis or vesico-ureteric reflux (or both).
- Pyelonephritis in children carries the risk of renal scarring 10% and this translates into a 10% risk of developing end stage renal disease.

Diagnosis

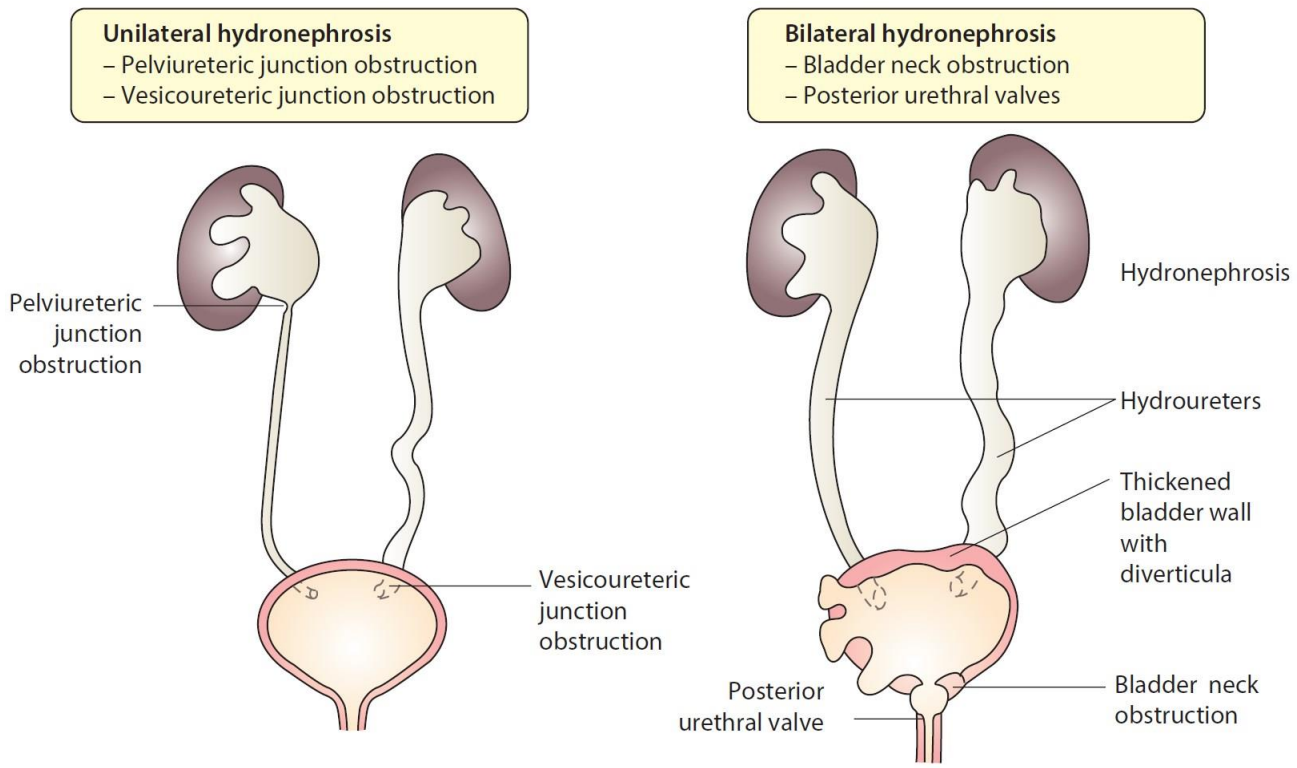
- Pyrexia lasting for more than 3 days mandates urine testing.
- Samples may be taken from mid-stream urine samples or supra pubic aspiration. Urine collected from nappies usually have faecal contaminants. In samples showing mixed growth contamination of the sample has usually occurred.
- As in adults $>10^5$ colony forming units of a single organism are usually indicative of a UTI.

Management

- A single isolated UTI (in girls) may be managed expectantly.
- > 2 UTI's (or 1 in males) in a 6-month period should prompt further testing.
- Voiding cystourethrogram (VCUG) show the greatest anatomical detail and is the ideal first line test in males; isotope cystography has a lower radiation dose and is the first line test in girls.
- USS should also be performed. Renal cortical scintigraphy should be performed when renal scarring is suspected.

Urethral Valves

Posterior urethral valves are the commonest cause of infravesical outflow obstruction in males. They may be diagnosed on ante natal ultrasonography. Because the bladder has to develop high emptying pressures in utero, the child may develop renal parenchymal damage. This translates to renal impairment noted in 70% of boys at presentation. Treatment is with bladder catheterization. Endoscopic valvotomy is the definitive treatment of choice with cystoscopic and renal follow up.



Urinary tract obstruction
Obstruction to urine flow results in dilatation of the urinary tract proximal to the site of obstruction. Obstruction may be at the pelviureteric or vesicoureteric junction (left), the bladder neck, or urethra (right).

Vesicoureteric Reflux

Vesicoureteric reflux (VUR) is the abnormal backflow of urine from the bladder into the ureter and kidney. It is relatively common abnormality of the urinary tract in children and predisposes to urinary tract infection (UTI), being found in around 30% of children who present with a UTI. As around 35% of children develop renal scarring it is important to investigate for VUR in children following a UTI

Pathophysiology of VUR

- Ureters are displaced laterally, entering the bladder in a more perpendicular fashion than at an angle
- Therefore, shortened intramural course of ureter
- Vesicoureteric junction cannot therefore function adequately

The table below summarizes the grading of VUR

{Grade}	
I	Reflux into the ureter only, no dilatation
II	Reflux into the renal pelvis on micturition, no dilatation
III	Mild/moderate dilatation of the ureter, renal pelvis and calyces
IV	Dilation of the renal pelvis and calyces with moderate ureteral tortuosity
V	Gross dilatation of the ureter, pelvis and calyces with ureteral tortuosity

Investigation

- VUR is normally diagnosed following a voiding (micturating) cystourethrogram (MCUG)
- DMSA scan may also be performed to look for renal scarring