Congenital bowel disorders

- Anatomical abnormality
  - Obstructing
    - Atresia
    - Stenosis
  - Not obstructing
    - Cysts
    - Diverticula
    - Fistulae

- Disturbance of Function
  - Digestion and absorption
    - Congenital enzyme deficiencies
    - Congenital abnormalities of gut mucosa
    - Congenital pancreatic dysfunction
      - Cystic Fibrosis
  - Neuro-endocrine
    - Motility
      - Hirschsprung and related conditions
Congenital abnormalities of the gut

• With other malformations
  Chromosominal
  Syndromic

• Without other malformations
  Teratogenic
  Vascular
Congenital abnormalities of the gut

- Overt
  - Visible at birth
  - Progressive lesion
- Covert
  - Not visible at birth
  - Recurrent or intermittent
  - Manifest by disturbance of function
Congenital abnormalities of the gut

• When to suspect
  Evident malformations
  Symptoms of intestinal obstruction
    Pre- ; peri- ; postnatal
  Unexplained gut malfunction
  Motility
  Digestion/Absorption
Congenital malformations

• In all cases
  Identify anatomical pattern
  Appropriate imaging studies
  Identify biochemical status
  Appropriate resuscitation
  Refer paediatric surgeon
Congenital disturbance of gut function

- Congenital diarrhoea
  - Enzyme deficiency
  - Transport defect across mucosa
  - Dysplasia and enteropathy
- Pancreatic malabsorption
  - Cystic Fibrosis