NEET SS SURGERY
PAEDIATRIC SURGERY

CONTENT

PAEDIATRIC SURGICAL CONSIDERATIONS

"The farther back you look, the farther forward you are likely to see."
-Winston Churchill.

Introduction

00:00:21

Three questions :

- Whether the operation is mandatory? Can it be avoided?
- If the operation is inevitable, how urgent, can it be deferred for a while?
- The minimum surgical intervention required to correct the anomaly?

Pediatric surgical speciality in India :

- Sushruta-The father of Indian surgery and plastic surgery.
- Only the union of medicine and surgery constitutes the complete doctor.
- The doctor who lacks knowledge of one of these branches is like a bird with only one wing. Thus the need for collaboration between a pediatrician and pediatric surgeon is evident.
- The pediatric surgeon specialty in India, first established in 1965, as a section of association of surgeons of India. Pediatric surgery is new and yet to be recognized for the role of specialty in providing quality care and reducing infant mortality rate.

WOFAPS KYOTO declaration 2002 :

- Children are not just small adults and have medical & surgical problems and needs that are often quiet different from those encountered by adult physicians. Infants and children deserve the very best medical care available.
- Every infant and child who suffers from an illness or disease has the right to be treated in an environment devoted to their care by a pediatric medical and surgical specialist.

"The mere formulation of a problem is far, more essential than its solution, which may be merely a mathematical or experimental skills. To raise new questions, new possibilities, to regard old problems from new angle requires creative imagination and marks real advances in science".

- Albert Einstein.

Why pediatric surgeons ?

- Because of the unique training, they provide a wide range of treatment options and the highest quality care to children.
- To diagnose, treat, manage the surgical needs of the children.
- Different anatomic and physiological parameters.
- Smaller organs need intricate surgical skills.
- Difficult venous access, different dosage schedules.
- Different spectrum of anomalies.

Need of specialiazed centres equipped with :

- Round the clock pediatricians.
- Pediatric intensivist.
- Pediatric radiologist.
- Pediatric anaesthetist.
- Pediatric pathologist.
- Pediatric orthopedician.
- Pediatric oncologist.
- many children hospital across the globe >200 years.

Objectives (Indian scenario) :

1. To provide specialized pediatric surgical services to over 400 million pediatric population at an affordable cost and safety, ethically and professionally.
2. To provide rigorous and uniform training including sub sections, at least in apex and centres of excellence.
3. To utilize the available limited financial resources to conduct only need based research to understand the common diseases in the region.

Pediatric surgery in 19th century :

- Increasing interest in the surgery of children.
- Europe is considered to be the cradle of Pediatric surgery.
- Many children's hospital came up all over Europe mostly during Ist half of 19th century.
- The great ormond street (World famous hospital for sick children) was established in 1852.
- The famous Boston children hospital was opened in 1882.
- Pediatric surgeons of 19th century were engaged in orthopedic surgery.

Modern pediatric surgery :

Three generations of surgeons :

1. William ladd and Robert gross : Father of pediatric surgery.
2. Sir Denis Browne (London) : Founder and first president of the British association of pediatric surgeons.
3. Middle generation.

The Indian scenarion :

- It is just 40 years old.
- Prof UC Chakraborty at Calcutta medical college.
- Dr Raman Nair at Trivendrum medical college.
- Prof O Anjaneyulu at Niloufer hospital college, Hyderabad.
- Dr Arthur E Desa at Bombay.
- Col RD Ayyar at Delhi.

History in India :

- Independent pediatric surgery section in 1964 (Dr R. K Gandhi).
- Postgraduate degree in Pediatric Surgery at University of Madras in 1966.

Scope :

- Wide field and deals with most body parts and organs in newborns, infants and children.
- A general pediatric surgeon is allowed to operate on wide spectrum patients depending on the skill sets in view of lacking well developed exclusive pediatric surgical centers.
- In the interest of patient care, it is important to develop the subspecialities as the distinct discipline.

Scope	
Pediatric gastrosurgery :	
• Anorectal malformation • Malrotation • Intestinal atresia • Duodenal atresia • Hirschprungs disease • Necrotising enterocolitis • Septic ileus • Meconium ileus • Infantile hypertrophic pyloric stenosis • Pyloric atresia	• Achalasia cardia • Biliary atresia • Choledochal cyst • Pseudopancreatic cyst • Omentalcyst • Mesenteric cyst • Congenital duplication cyst • Hernia • Splenomegaly • Abdominnal trauma
Pediatric urology :	
• Phimosis • Labial synechia • UTI • Undescended testis • Torsion testis • Epididymo-orchitis • Congenital hydrocele • Pelviureteric junction obstruction	• Posterior urethral valve • Vesicouretyeric reflux • Antenatal hydronephrosis • Hypospadias • Exstrophy epispadias complex • Non funcxtioning Kidney
Pediatric thoracic surgery :	
• Esophageal atresia and tra-cheo-esophageal fistula • Congenital diaphragmatic hernia • Congenital cystic adenomatoid malformation • Congenital lobar emphysema • Diaphragmatic eventration	• Diphragmatic trauma • Lung cyst • Empyema thoracis • Esophageal replacement surgery
Pediatric neurosurgery :	
• Spina bifida spectrum : Menin-gocele • Meningomyeocele • Lipomeningomyelocele • Diastometamyelia • Tethered cord	• Craniosynostosis • Rachischisis diluraji10@gmail.com • Hydrocephalus : Congenital and acquired
Pediatric oncosurgery :	
• Wilms tumor • Neuroblastoma • Sacrococcygeal teratoma • Soft tissue tumors	• Teratoma • Testicular tumors • Rhabdomyosarcoma
Pediatric minimal invasive surgery :	
• Laproscopy • Cystoscopy • Bronchoscopy : Rigid Foreign Body removal	• Endoscopic guided procedures • USG guided drainage procedures • Recent advances : Robotics

Age group : 12-18 years.

Ethical considerations :

For many congenital condition :

- ARM.
- NTD.
- DSD.
- HD.
- Exstrophy bladder.
- Urological anomalies.
- Vascular malformations.

Fetal surgery :

- Congenital malformations occur in 2-4% globally.
- Prevalence in India is 6-7%.
- Fetal malformations account for 30% perinatal deaths & considerable infant morbidity.
- Advances in imaging sciences, molecular genetics & in utero surgical techniques.

Screening for structural anomaly :

- Ultrasound screening is recommended at 18-20 weeks of gestation.
- Shift in screening in aneuploidy in the first trimester.
- Anencephaly, open spina bifida, cleft lip, diaphragmatic hernia, gastroschisis, exomphalos, serious cardiac defects, bilateral renal agenesis, lethal skeletal dysplasia, urinary tract anomalies.
- Informed choices.

Prenatal diagnosis and pediatric surgeons :

- Antenatal counselling.
- Significant impact on perinatal management of fetus.
- Modulating the delivery at appropriate centre, to ensure well being.

Common fetal structural anomalies :

- Anterior abdominal wall defect.
- CNS.
- Congenital cardiac defects.
- Congenital lung malformations.
- Fetal genitourinary anomalies.
- Fetal gastrointestinal anomalies.

Surgery

00:31:20

Fetal surgery :

- Ist attempted by Michael R Harrison, at University of California, San Francisco in 1980.
- First successful Vesicoamniotic shunting for a baby with posterior urethral valve (Open/fetoscopic). Techniques, risks (Prematurity, uterine rupture) and outcomes vary.

Anaesthesia :

- Indomethacin.
- Inhalational isoflurane, desflurane.

Types :

- Open.
- Fetoscopy.
- Minimal invasive techniques.

Indication :

- Lethal
- Non-lethal.

Vesicoamniotic shunt placement.

Fetoscopic procedure (Laser ablation for twin twin transfusion, cervical mass).

Open fetal surgery (mmc, moms trial), CDH, sacrococcygeal teratoma.

Exit procedure :

Ex-utero intrapartum treatment.

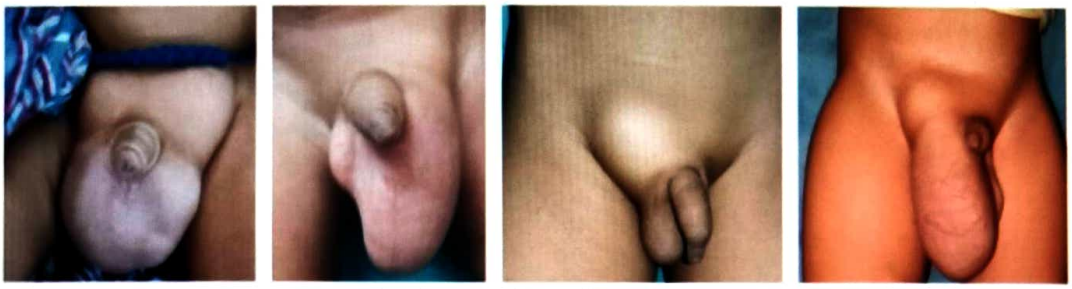
Indication :

- Airway obstruction.
- Large cervical cystic hygroma.
- Tracheal atresia.
- Mediastinal mass.
- Cervical teratoma.

Tracheal occlusion : CDH

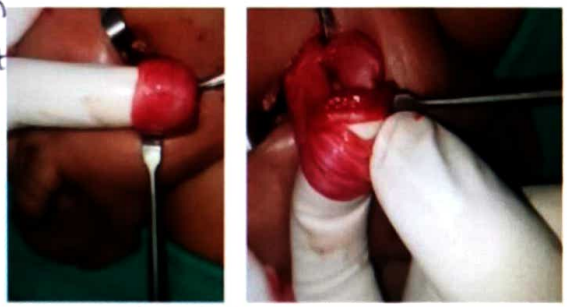
Common paediatric surgical problems & treatment :

Hernia : Hernia to be reparied as soon as it is diagnosed.

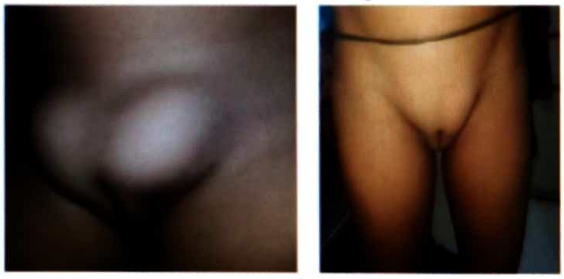


Inguinal scrotal swelling

- Herniotomy should be done as soon as possible as the newborns are at risk of incarceration & strangulation in the first 3 months of life.
- Requirements :
  - Minimum weight : 2 kg.
  - There should be no cardio pulmonary contraindication.
- In females :
  - m/c presentation is bilateral.
  - Sliding component is present.
  - Treatment : Bevans repair.

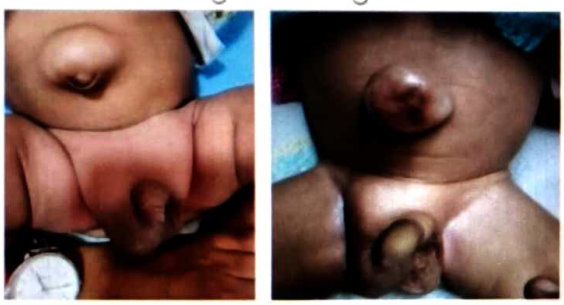


Patent processess vaginalis

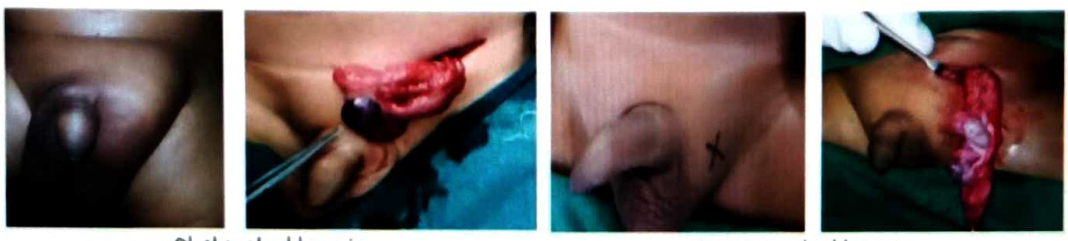


Inguinal swelling

- Umblical hernia can be waited as the umblical ring will close with time. Resolves by 2-3 yrs of age.
- Supra umblical hernia will require surgery as it doesnt heals.

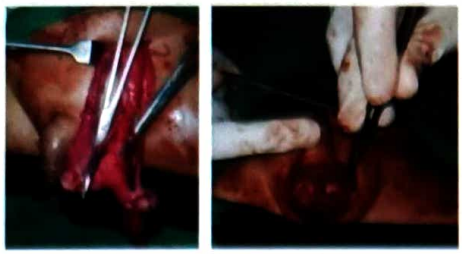


Inguinal, umblical & supra umblical hernia

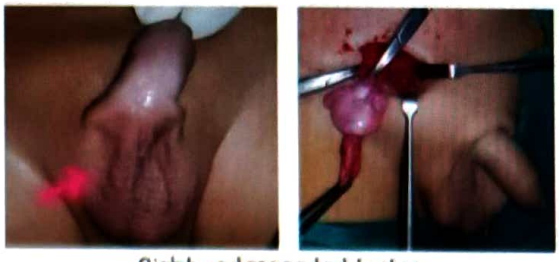


Obstructed hernia

Incarcerated hernia



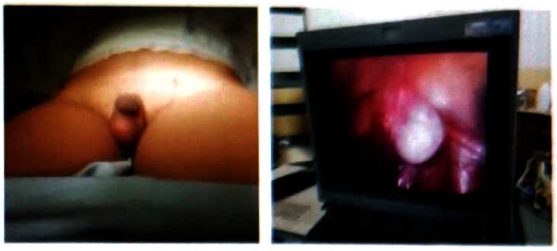
PMDS



Right undescended testes

Undescended testes :

- Age of repair : 6-9 months.
- After 9 months they undergo atrophy due to high temperature in the inhuinal region.
- 80-90% of them are palpable.
- Complication : Torsion.

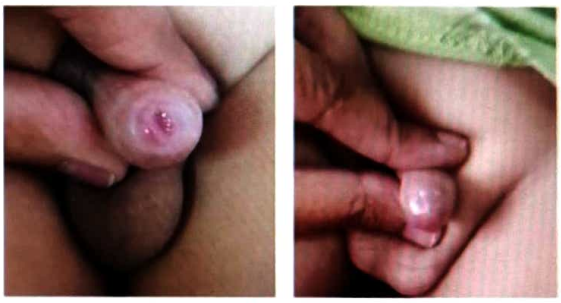


Palpable testes

Phimosis :

Presentation :

- Prepuce covering the meatus.
- UTI, difficulty in micturition.
- Ballooning of prepuceal skin.



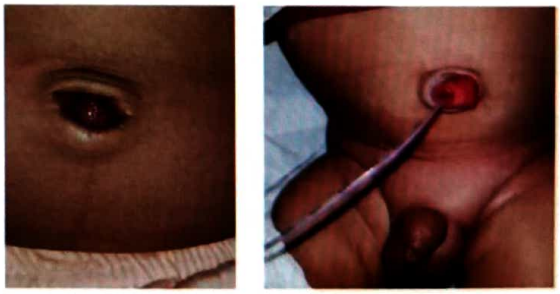
Phimosis

Umblical granuloma & patent urachus :

- Umblical granuloma requires medical
- management. Patent urachus requires surgical correction.

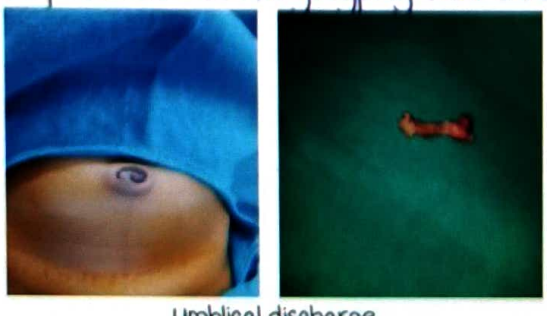
Umblical discharge :

- Cause :
  - Improper umblical hygiene,
  - surgical or infective.

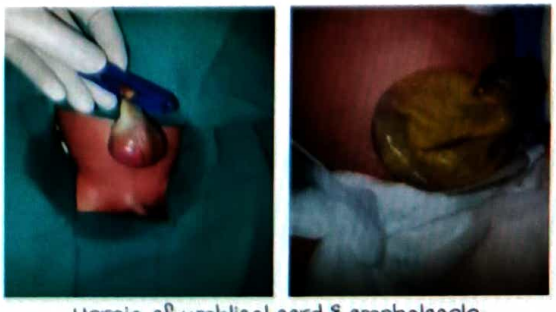


Umblical granuloma and patent urachus

Omphalocele : Do Karyotyping (50% chromosomal anomalies).



Umblical discharge



Hernia of umblical cord & omphalocele

Gastroschisis :

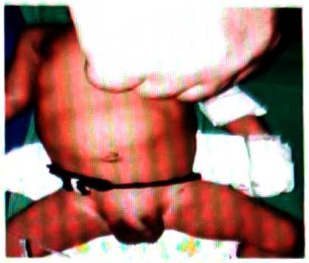
Intestine to be covered to avoid risk of sep
mesentery).


Gastroschisis

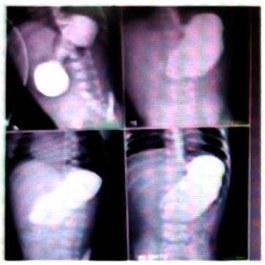
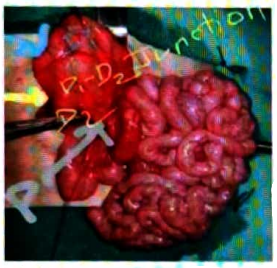
Idiopathic hypertrophic pyloric stenosis :

Treatment : Ramstead pyloromyotomy.

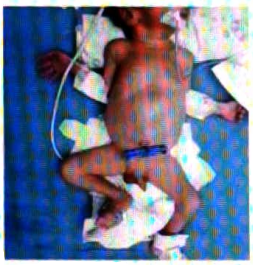
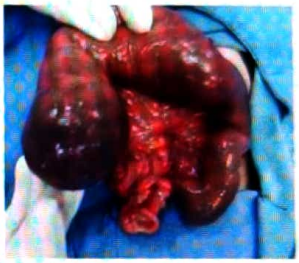
Duodenal atresia : Double bubble sign.




IHPS

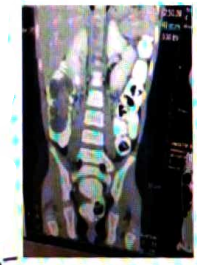
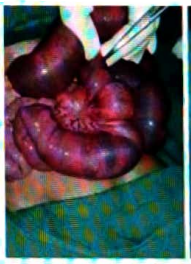
Double bubble sign

Intestinal atresia

Intussception :

- Presentation : Shock, fever.
- Common lead point : meckels diverticulum.
- Treatment : Stoma, resection & anastomosis.


Intussception

Infantile hemangioma :

Treatment : Propranolol.


Infantile hemangioma

Neck swelling :

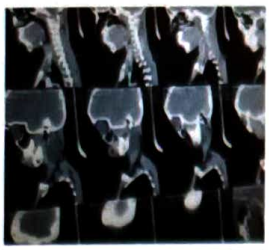
Neck abscess :

- Short duration of history.
- High grade fever, progressive swelling which is indurated, hard and tender.
- Pus on aspiration.
- Treatment : I & D.

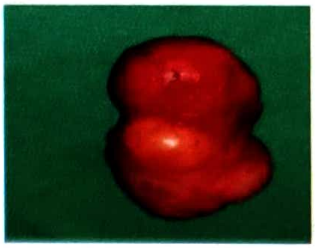
Cystic hygroma :

- Treatment : Intralesional sclerotherapy using bleomycin.


Lymphatic malformation and neck abscess



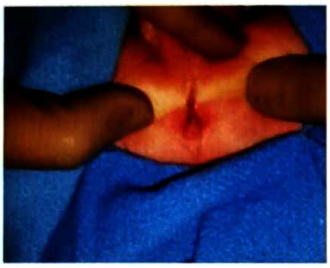
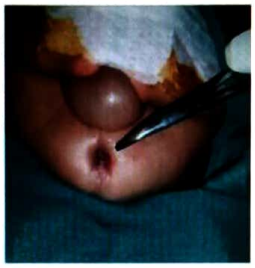

Branchial cyst

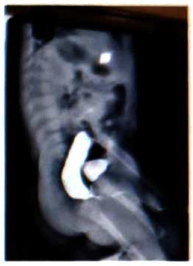



TB lymph node

Anorectal malformation :

- High ARM : Colostomy + definitive pull through.
- Low ARM : One stage surgery.
- Low ARM : meconium pearl.
- Treatment : Anoplasty.
- Anterior ectopic anus is a variant of female ARM.

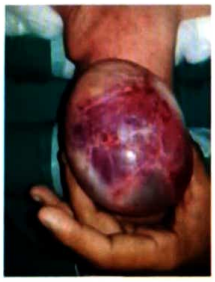
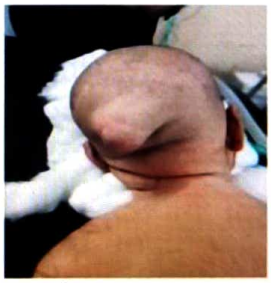
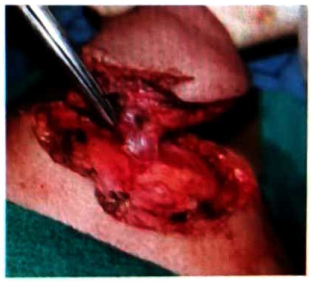


Anorectal malformation

Midline swelling : Spinal defect.

Important consideration :

- Size of the encephalocele.
- Amount of brain tissue.
- Associated hydrocephalus.
- Defect size.

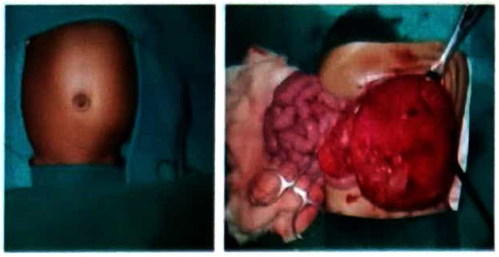
Midline defects

Omental cyst :

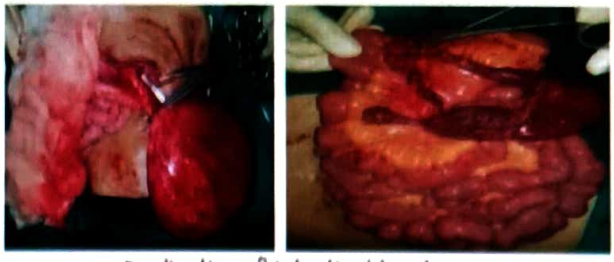
If omental cyst is attched to the tail of trachea make sure theres no pancreatic injury to prevent pancreatic fistula while separating it.

Duplication of intestinal tract :

Contains ectopic gastric mucosa → Secretion of gastric juices → Gastritis → Perforation.



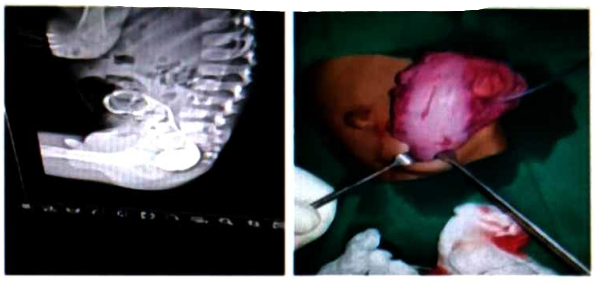
Omental cyst



Duplication of intestinal tract

Hirschprung disease :

Treatment : Colostomy.



Hirschprung disease

Recent advances

01:01:00

Other treatments :

- Stem cell research.
- Minimal invasive surgery.
- Robotic surgery.
- Laser rechnology.
- Hormonic scalpel.
- Tissue engineering.
- Fetal surgery.
- Pediatric tumors.
- Brachy therapy.
- Telemedicine and teleconferencing.

Good news :

- Changing social plethora.
- More antenatal and anomaly scans.
- More precious pregnancies.
- Long dates and flooded government institutions.
- Better understanding of evolution of most diseases.
- Better availability of team and euipments.

Pediatric surgeon and the child :

- Late presentation and referral : Poor outcome and incresed cost.
- Paucity of dedicated pediatric surgical centre & inadequate infrastructure.
- 50% districts in India don't have access to pediatric surgical facility.
- medical facility is ramping up but pediatric surgical care is lagging.
- To ensure 29th december has been marked as "pediatric surgery day" : Childs right to quality surgical care.

CONGENITAL DIAPHRAGMATIC HERNIA

Introduction

00:00:41

Congenital Diaphragmatic Hernia (CDH) is a congenital malformation characterized by a defect in the posterolateral diaphragm, the foramen of bochdalek, through which abdominal viscera migrate into the chest during fetal life.

Common malformation.

Defect in posterolateral diaphragm.

Bochdalek hernia (After the scientist who described it).

Incidence : 1 in 2600 to 1 in 3700.

80% left sided, 5% bilateral.

Hidden/antenatal mortality is high.

History :

- Earliest gross anatomy and pathophysiology : mccauley in 1754.
- Bowditch : Bedside clinical diagnosis in 1847.
- Bochdaleck : Hernia resulted from posterolateral rupture of membrane.
- Heidenhain : In 1905, reported first successful repair.
- Gross : In 1946, reported 100% survival (Immediate surgery).
- Standard care : Immediate neonatal surgery followed by postoperative resuscitative therapy uptil 1980.

Current trend :

Current survival rate is 55-70%.

mortality is attributed to pulmonary hypoplasia and persistent pulmonary hypertension.

Epidemiology and genetics :

Incidence : 1 in 2000/5000

1/3rd infants are stillborn.

Thin and underweight mothers.

Premature, macrosomic, male.

mostly sporadic.

CDH with abnormal karyotype is associated with poor outcome.

Epidemiology :

- Cause unknown.
- Exposure to pharmacological agents
- Drugs : Phenmetrazine, thalidomoide, quinine, cadmium, lead, nirofen (Herbicide).
- Vitamin A deficiency : Retinoid regulated target genes.

Associated malformations :

- 10-50%.
- Skeletal defects : Limb reduction and costovertebral defects.
- Cardiac anomalies 33%. (VSD is m/c).
- Neural tube defects (m/c : meningoceles, anencephaly).
- Abdominal wall defect : Omphalocle, gastroschisis, pentalogy of Cantrell.
- Urinary tract anomalies.
- Malrotation.
- Trisomies (21, 18, 13).
- Frey, Beckwith-wiedemann, Goldenhar, Coffin-Siris.

Associated syndromes :

- Pallister-Killian.
- Fryns.
- Ghersoni-Baruch.
- WAGR.
- Denys-Drash.
- Wolf-Hirschhorn syndrome.

Embryogenesis 00:11:30

Sporadic.

Failure of pleuroperitoneal canal to fuse.

Pulmonary hypoplasia : D/t mechanical compression, ipsilateral, later bilateral.

Diaphragmatic development :

- Anterior central tendon : Septum transversum.
- Dorsolateral : Pleuroperitoneal membranes.
- Dorsal crura : Esophageal mesentery.
- Muscular portion : Thoracic intercoastal muscle groups.
- 4th week of gestation : Peritoneal fold from lateral mesenchyma and septum

tranversum.

- 8th week : Formation of pleuroperitoneal membrane, right side >left.
- muscularization : Posthepatic mesenchymal plate.

Lung development :

Foregut derivative.

Phases of development :

- Embryonic (3rd – 6th).
- Pseudoglandular (7th – 16th).
- Canalicular (16th–24th) : Appearance of pneumocytes I, II.
- Saccular (24th week – Term) : Formation of respiratory unit.
- Alveolar (Shortly after birth – 2 years).

Pulmonary vascular development :

- Follows airway and alveolar growth.
- Pre-acinar : 16 weeks.
- Homebox genes, nuclear transcription factor, hormones and growth factors.

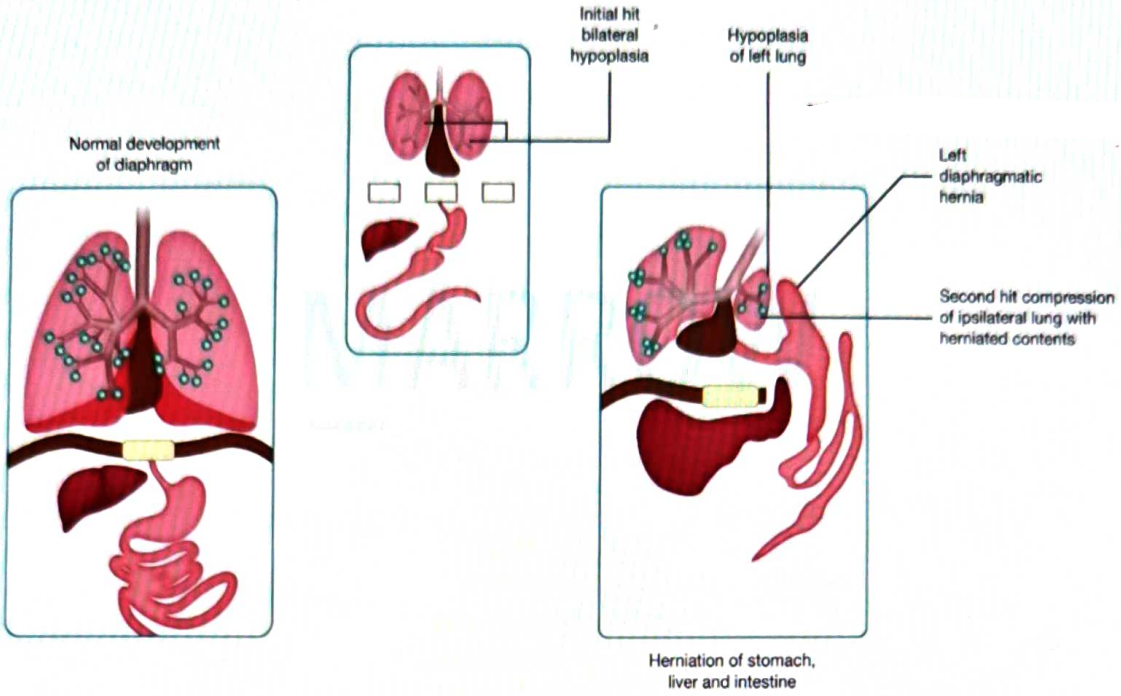
Pulmonary hypoplasia :

- Fewer alveoli.
- Thickened alveolar walls.
- Increased interstitial tissue.
- markedly diminished alveolar air space and gas exchange area.
- Pulmonary vasculature is abnormal : Reduced vessels, adventitial thickening, medial hyperplasia.

Embryological explanation of CDH :

1. Theory of mechanical compression.
2. Dual hit theory : Pharmacological agent → Pulmonary hypoplasia.
3. Exposure to teratogen.
4. Theory of low airway pressure antenatally.

Embryological Explanation of CDH



Dual hit theory of abnormal lung development in CDH

Pathogenesis

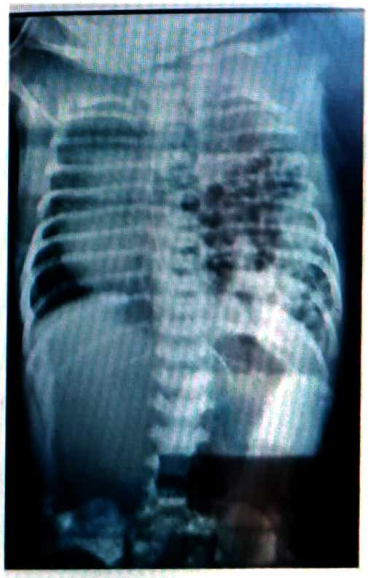
00:26:14

Pathophysiology :

- Onset and severity : Amount of abdominal viscera.
- Degree of pulmonary hypoplasia.
- Hypoxia, hypercarbia : Pulmonary vasoconstriction and pulmonary hypertension.
- Reversal of right - left shunt.
- Abnormal pulmonary vascular bed.
- Increased endothelin 1.
- Apparent surfactant deficiency : Secondary to respiratory failure.

Pathology :

- 2 to 4 cm defect in posterolateral diaphragm.
- Herniation.
- Both ipsilateral and contralateral lungs are affected.
- Abnormal pulmonary bed.
- Significant adventitial and medial wall thickening.



Congenital diaphragmatic hernia.

Diagnosis

00:33:10

40 – 90% cases can be diagnosed antenatally.

mean gestational age : 24 weeks.

Clinical clues :

- Polyhydramnios : 80% pregnancies.
- Stomach in fetal thorax at same cross section of heart.
- Absence of stomach in abdomen.

1. Prenatal ultrasound :
    - At 20 weeks.
    - Absence of diaphragm.
    - Intrathoracic presence of abdominal viscera.

2. Amniocentesis : To pick up chromosomal anomalies.
3. LHR (Lung head ratio) :
    - Prognostic marker of fetal lung hypoplasia.
    - Calculated suing ultrasound.
    - Area of contralateral lung/fetal head circumference.
    - LHR > 1 : Risk of severe lung hypoplasia is low.
    - LHR < 0.85 : 100% mortality.

4. Lung/thorax in transverse axis.
5. Prenatal MRI for lung volume.
6. Presence of liver.
7. Stomach position.
8. Mc goon index, pulmonary artery index

Antenatal diagnosis :

Increased awareness and availability of antenatal ultrasound and screening.

Chromosomal aberrations and syndromes : Termination of gestation advised.

FETENDO : Fetal Endoscopic Balloon Occlusion, aids in lung growth.

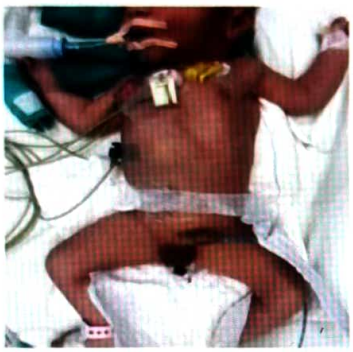
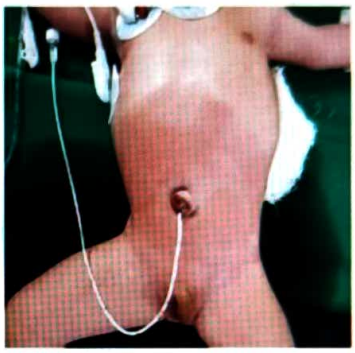
Prenatal care :

- Properly inform and prepare parents.
- Fetus and mother should be referred to appropriate tertiary care centres. (NO, oscillating ventilators, ECMO).
- Mode of delivery depends on obstetric issues.

- Fetal tracheal occlusion (with Fogarty balloon, FETENDO).
- EXIT procedure : Ex Partum Intrauterine procedure to remove tracheal occlusion.

Postnatal diagnosis :

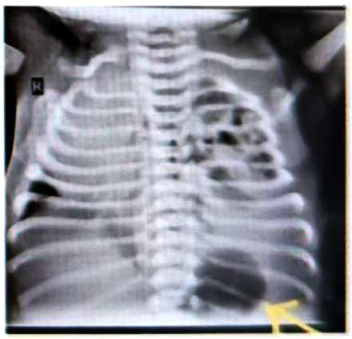
- Severe respiratory distress at birth.
- Shifting of apical cardiac impulse to opposite side.
- Auscultation of bowel sound to affected side.
- Scaphoid abdomen.
- Asymmetric distended chest.

Scaphoid abdomen, hyperinflated chest.

Delayed presentation :

- Patent pulmonary circulation.
- Honeymoon period : D/t persistance of fetal circulation.
- 10-20% : Recurrent lower respiratory tract infection, respiratory distress.



CDH : Left side.

Confirmatory : Chest X-ray PA view.

Assessment :

Arterial blood gas.

2D ECHO :

- Assessment of right to left shunt.
- Degree of pulmonary hypertension.
- Rules out associated cardiac malformation.
- mc goon index (MGI) :

  RPA diameter + LPA diameter/Descending aorta diameter < 1.31
- Pulmonary artery index (PAI) :

  RPA area + LPA area/ BSA.

Physiological parameters :

- VI =RRX (PIP-PEEP)
- MVI.
- OI (<6 and >17.5).

Treatment continues to be guided best by clinical judgement.

Differential diagnosis :

- Eventration of diaphragm.
- Anterior diaphragmatic hernia of morgagni.
- Congenital esophageal hiatal hernia.
- Congenital cystic disease of lung.

Prognostic factors :

Anatomic factors :

- Antenatal diagnosis is associated with worse prognosis.
- Polyhydramnios.
- Herniation of liver.
- Position of stomach.
- Right side has worse prognosis.
- Imaging parameters : LHR <0.85

Respiratory distress :

- Uncorrectable pulmonary hypoplasia.
- Potentially reversible pulmonary hypertension.

Management

00:59:33

Pre-operative care :

- Stabilize the cardiorespiratory system.
- Physiological emergency, not surgical.
- Respiratory distress : Endotracheal intubation.
- Mask ventilation contraindicated.
- Nasogastric tube.
- Preoperative stabilization (Stabilization of pulmonary circulation).
- Optimization of blood gases prior to surgery for best surgical outcome.
- Arterial and venous access.
- Hemodynamic stability, thermoregulation and glucose hemostasis.
- Proper sedation.

- Systemic hypotension and inadequate tissue perfusion.
- Correction of metabolic acid-base disturbance.

Preoperative treatment :

- Endotracheal intubation : Gentle ventilation and permissive hypercarbia.
- HFOV (High frequency oscillatory ventilation) :
  Non conclusive.
- The constant distending airway pressure may be more detrimental than helpful.
- use of inhaled nitric oxide which provides selective pulmonary vasodilatation without systemic hypotension seem a promising therapy, adjunct to mechanical ventilation.
- ECMO (Extra Corporeal membrane Oxygenation) : Conflicting.
- Partial liquid ventilation.
- Intratracheal pulmonary ventilation.
- Appropriate fluid management with ionotropes is crucial.
- Surgery to be performed when the infant is stable.

Guidelines :

Use of spontaneous mechanical ventilation when possible.

Bag and mask ventilation is contraindicated.

Use of low pressure (<25 cm of H_2O and high frequency ventilation).

Avoiding use of muscle relaxants.

Maintaining mild alkalosis.

Goal :

- Preductal oxygen saturation of 80-95%.
- Paoz 55-60 mm of Hg.
- Pacoz of 60 mm Hg (Permissive hypercapnia).

Pharmacology :

To improve pulmonary artery hypertension.

- Nitroprusside
- Isoproterenol
- Nitroglycerin
- Captopril
- Calcium channel blockers
- Prostacyclin derivatives
- Endothelin receptor antagonists

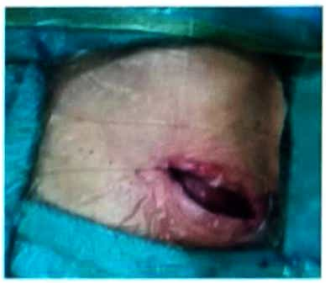
- Phosphodiesterase inhibitors
- Inconclusive : Surfactant, nitric oxide.

ECMO :

- Offered to 10-20% cases of CDH with severe hypoplasia.
- For preoperative surgical stabilization.
- Lung is bypassed and ECMO allows gaseous exchange through arteriovenous and venovenous cannulation connected to membrane gas exchange, thus protecting lung from barotrauma.
- Level of evidence to support ECMO is very weak.

Operative repair :

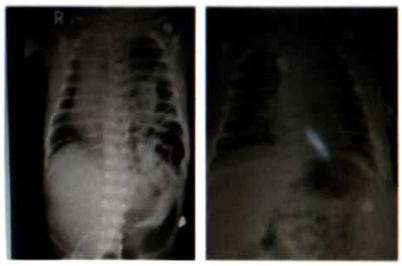
- Timing : Immediate vs delayed.
- Most commonly preferred approach : Abdominal approach.
- Use of intercostal chest tube : Controversial.
- 10-15% cases of hernia have a sac.
- Defect repair : Use of mesh, latissimus dorsi muscle flap.



Abdominal approach for CDH.

Role of minimal invasive surgery :
Thoracoscopic/ laparoscopic.
Indicated in :

- Stable patients.
- Delayed presentation/less severe symptoms.



Pre and post operative chest X ray in CDH.

Prognosis :

- Survival : 70-80%.
- Actual survival rate : 50-60%.
- Risk of pneumonia in CDH survivors : 35% by 12 years.
- First 3 years : Respiratory syncytial virus/viral bronchiolitis.
- Gastro-esophageal reflux : Prophylactic fundoplication.
- Neurodevelopmental delay.

Conclusion :

- CDH is a spectrum which ranges from severe pulmonary hypoplasia and pulmonary hypertension non-responsive to conventional management at one end to those with benign course and better outcome at the other end.

- Change in approach from pure surgical emergency to elective surgery after a period of medical management (To improve ventilation and prevention/ reduction of pulmonary hypertension).
- CDH management remains a complex unsolved mystery for both pediatric surgeons and neonatologists.

CONGENITAL LUNG LESIONS

Introduction 00:00:21

Definition :

Spectrum of lung abnormalities :

- Diverse etiologies
- Overlap in presentation, anatomic variation and radiographic appearance of chest lesions.

Dramatic increase and documentation with widespread prenatal scan.

Advancement in fetal intervention.

History :

- Surgical treatment first used in 1930 and 1940.
- Reinhoff successfully excised a unilocular cyst from right upper lobe in 1933.
- Fischer : Removal of right upper and middle lobe for cystic disease in 1 month.

Features :

- Developmental in origin.
- Prognosis is usually good.
- Asymptomatic or critically ill with respiratory insufficiency.
- Better understanding of natural history. } Excellent outcome
- Efficient and organized approach for evaluation. }
- Treatment is necessary as they cause :
    1. Possibility of infection
    2. Respiratory difficulty
    3. Airway obstruction

Anatomy 00:04:02

Normal anatomy :

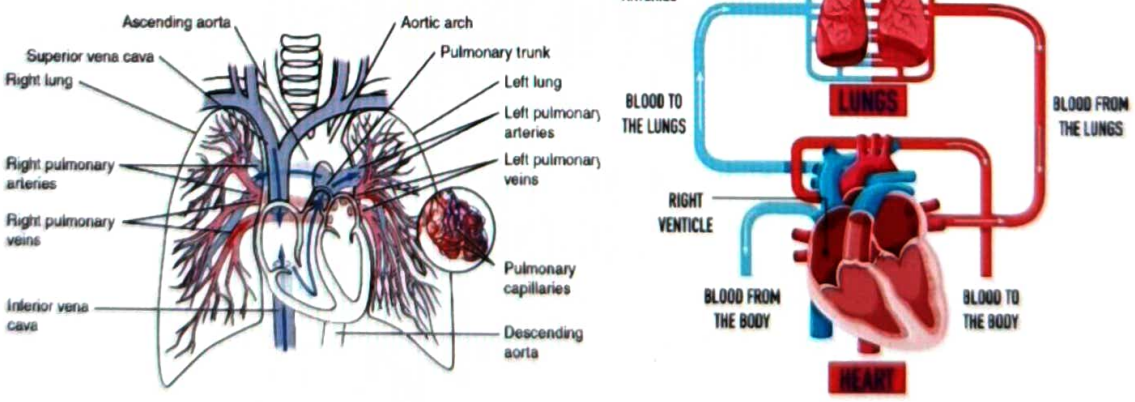
- Right lung : Upper, middle and Lower lobes.
- Left lung : Upper lobe, Lower lobe and Lingua.
- Tracheal bifurcation : Fourth or Fifth vertebral body.
- Right mainstem bronchi is larger and more vertical (Preferential drainage of

endobronchial material or foreign body into right lung.).

Lobes of lung.

Vascular supply :

- Circulation for respiratory gas exchange: Pulmonary artery and its branches.
- Circulation for nutritive support : Systemic.
- Left 2 bronchial arteries from descending thoracic aorta.
- Right single bronchial artery from third intercoastal artery.
- Collateral supply comes from inferior thyroid artery.
- Venous drainage : Azygous and hemiazygous systems.



Vascular supply of lungs.

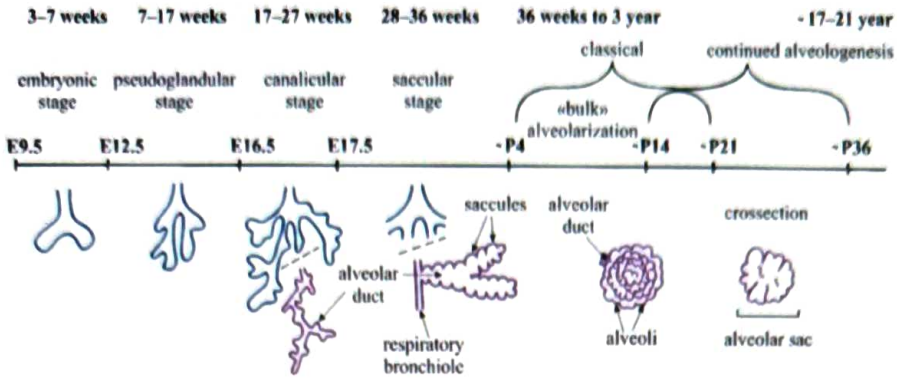
Embryology :

Fetal lung development is divided into Five stages.

1. Embryonic.
2. Pseudoglandular.
3. Canalicular.

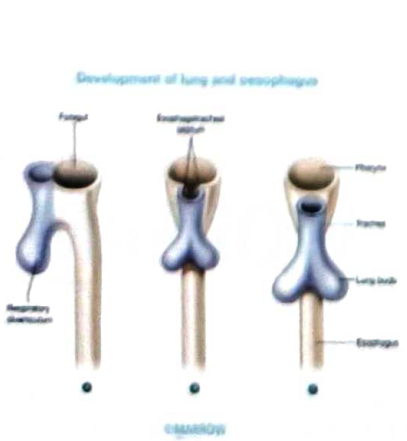
4. Saccular.

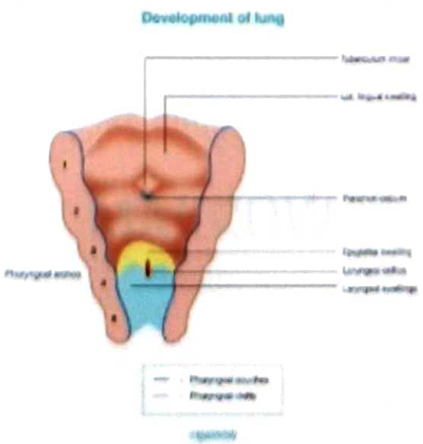
5. Alveolar.

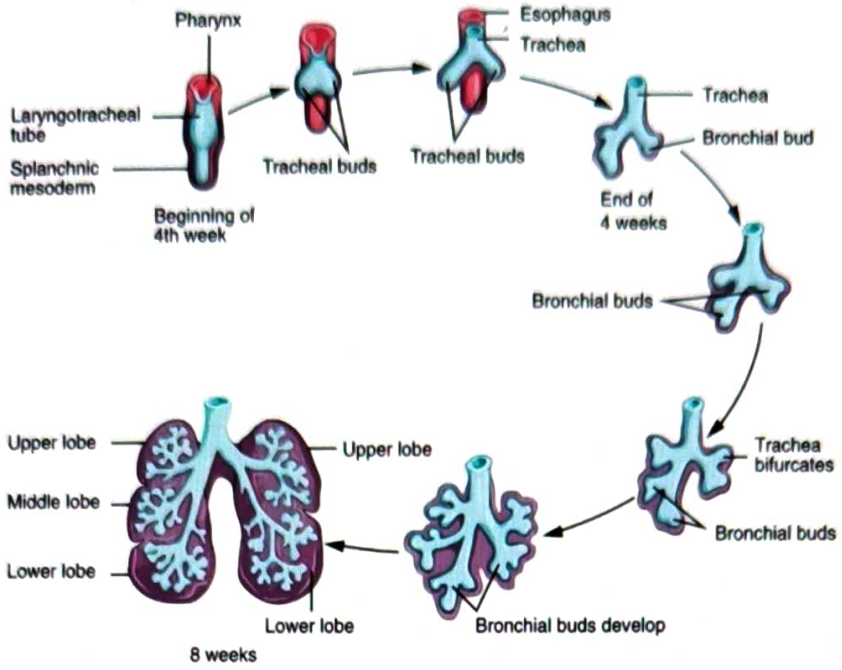


Phases of development of lung.

- Foregut derivative.
- Diverticulum at the caudal end of laryngotracheal groove.
- Trachea and Two lung buds by 4th week of gestation.
- Lobar structure: 6th week.
- Pseudoglandular phase 7th to 16th weeks.
- Bronchial airways develop.
- Canalicular phase: 16-24 weeks.
- Crude alveolar air sacs : Type I pneumocyte begin to differentiate, Type 2 pneumocytes responsible for gas exchange appear.
- Gas exchange process starts appearing from canalicular phase.
- Saccular Phase : 24 weeks of gestation to term.
- Remodelling of airspace dimensions and maturation of surfactant synthesis.
- Alveolar : Adult like alveoli soon after birth.
- Extensive alveolar maturation and multiplication takes place from birth upto 8 years of age with 10 fold increase in the number of functioning alveoli.
- Complete Alveolar formation by 2 years of age.







Development of lung.

Development of different parts of lung :

Pulmonary vasculature : 16 weeks

Preacinar :

- Trachea.
- Major bronchi.
- Lobar bronchi upto terminal bronchioles.

Acinar :

- >24 weeks.
- Respiratory bronchiole.
- Alveolar ducts.
- Alveoli.
- Gene expression, Growth factor interactions, Hormones.
- Homebox genes, Nuclear transcription factor (Hepatocyte nuclear factor 3 beta, Thyroid transcription factor 1).
- Hormones(Thyroid, Glucocorticoides and Retinoic acid).

Congenital lung lesions :

Prenatal lung disease :

- Bronchogenic cyst.
- Congenital cystic adenomatoid malformation.
- Bronchopulmonary sequestration.