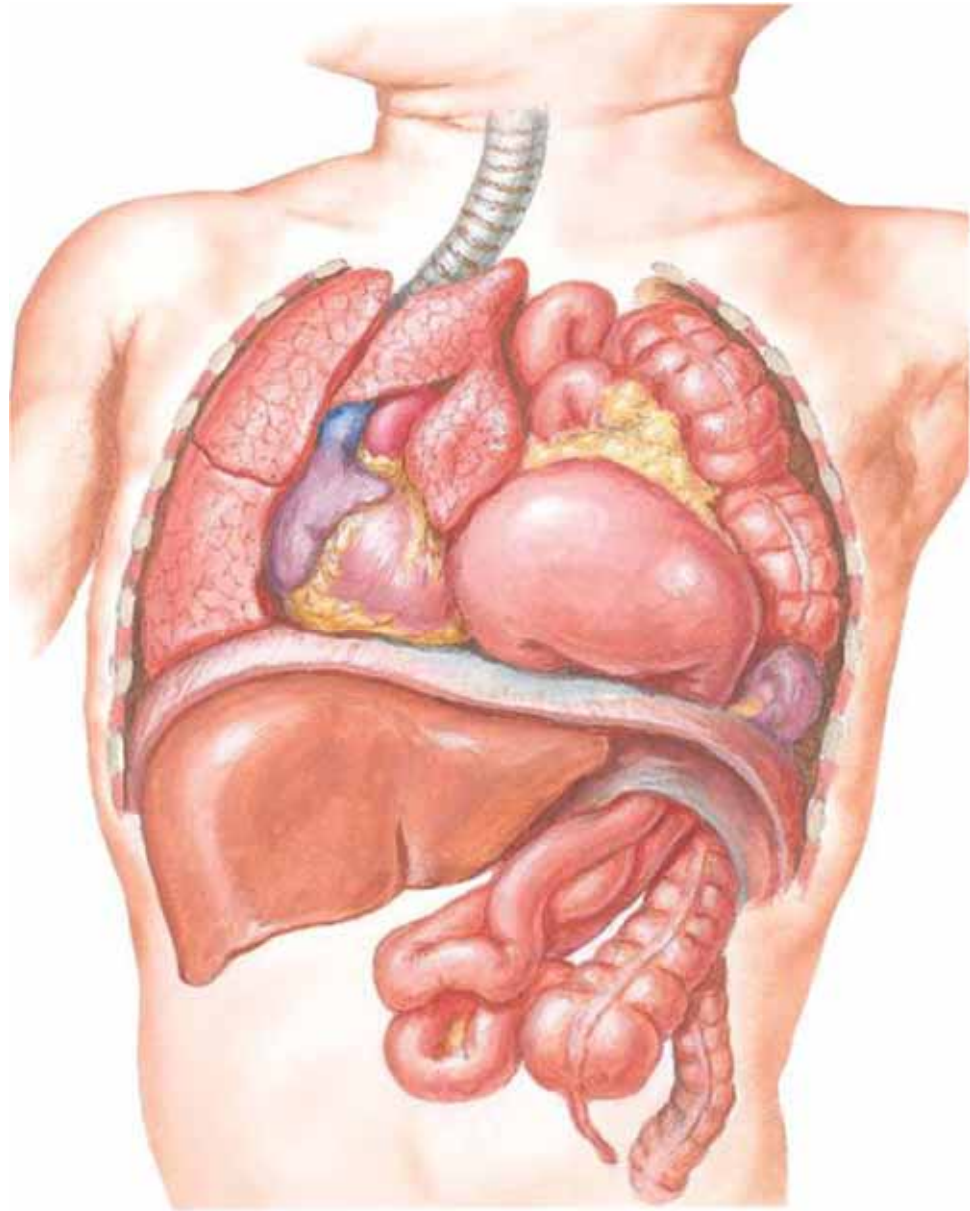


**A recent case of a
Congenital Diaphragmatic Hernia
diagnosed prenatally**

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MEDICAL UPDATE UOM – 11-09-2013



Congenital Diaphragmatic Hernias

- One of the most common severe congenital abnormalities
- Incidence 1 in 3000 births
- Combination of CDH and abnormal karyotype is associated with poor outcome
- Sporadic developmental anomaly, although familial cases have been reported

Bochdalek Hernia

- Postero-lateral diaphragmatic hernia
- Most common manifestation of CDH, accounting for more than 95% of cases
- Majority of Bochdalek hernias (80-85%) occur on the left side of the diaphragm
 - A failure of the diaphragm to completely close during development.
 - Herniation of the abdominal contents into the chest
 - Pulmonary hypoplasia

Pathophysiology

- Pulmonary parenchymal compression by herniated organs and its effect on growth and maturation of lung
- Unilateral hernia is associated with bilateral abnormal pulmonary development (severe on ipsilateral side)
- Lung is smaller, pulmonary vascular bed is abnormal on affected side in pts with CDH

Pathophysiology

- Inappropriate and significant medial muscular hyperplasia in arterioles, leading to increased susceptibility to development of pulmonary hypertension
- Factors contributing to Pulm HTN are
 - Hypoxia
 - Stress
 - Acidosis
 - hypothermia

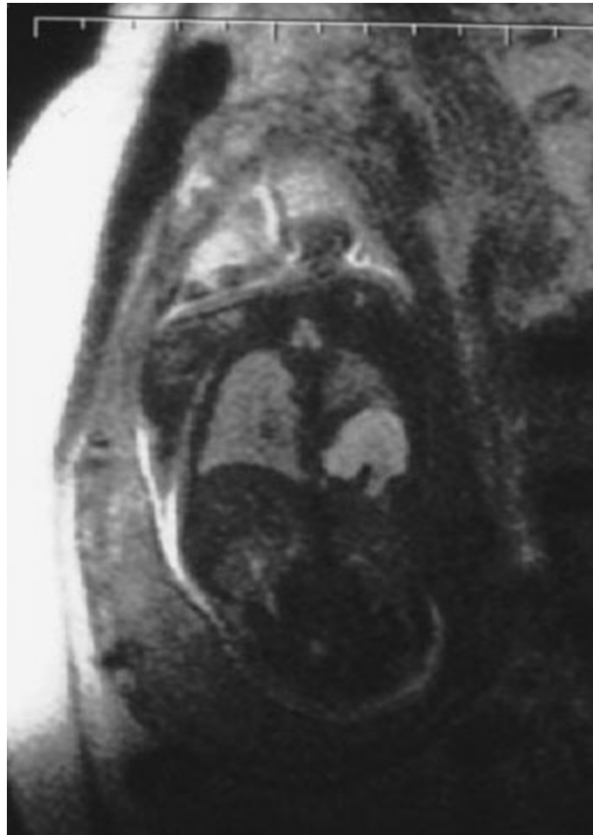
Diagnosis

- Often made on prenatal ultrasound exam
- Prenatal MRI when obstetric US has detected a complex fetal anomaly
- After birth, spectrum of respiratory symptoms depend on degree of pulmonary hypoplasia and pulm HTN
- Respiratory distress at birth within first 24 hours of life

Prenatal diagnosis – foetal US



Prenatal diagnosis – foetal MRI

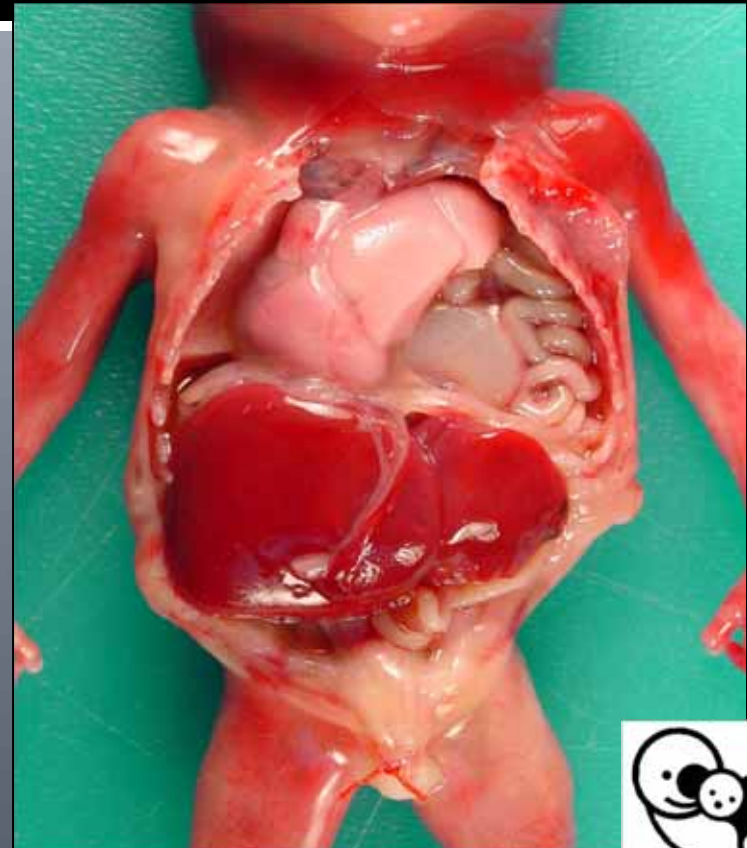
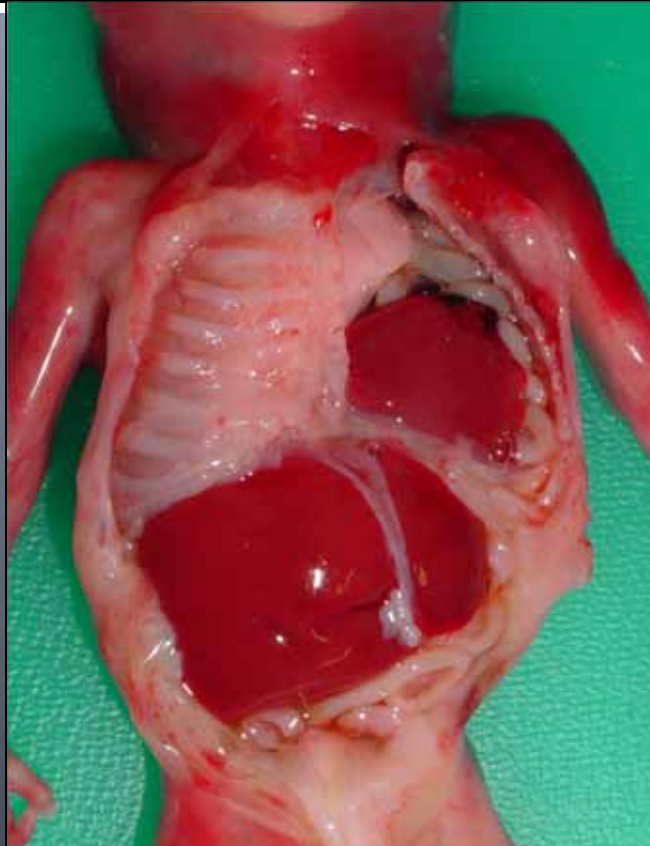


Prenatal prognosis – ??? interruption of pregnancy

- **Lung-to-head ratio (LHR)**
 - Measures contralateral lung at 24 to 26 weeks gestation
 - <1.0 proposed to identify fetuses at high risk for mortality
- **Liver herniation**
 - Proposed to identify fetuses at high risk for mortality

Post-mortem examination

- One third of infants with CDH are stillborn, usually due to
- associated fatal anomalies like neural tube defects and cardiac defects
 - abnormal karyotype



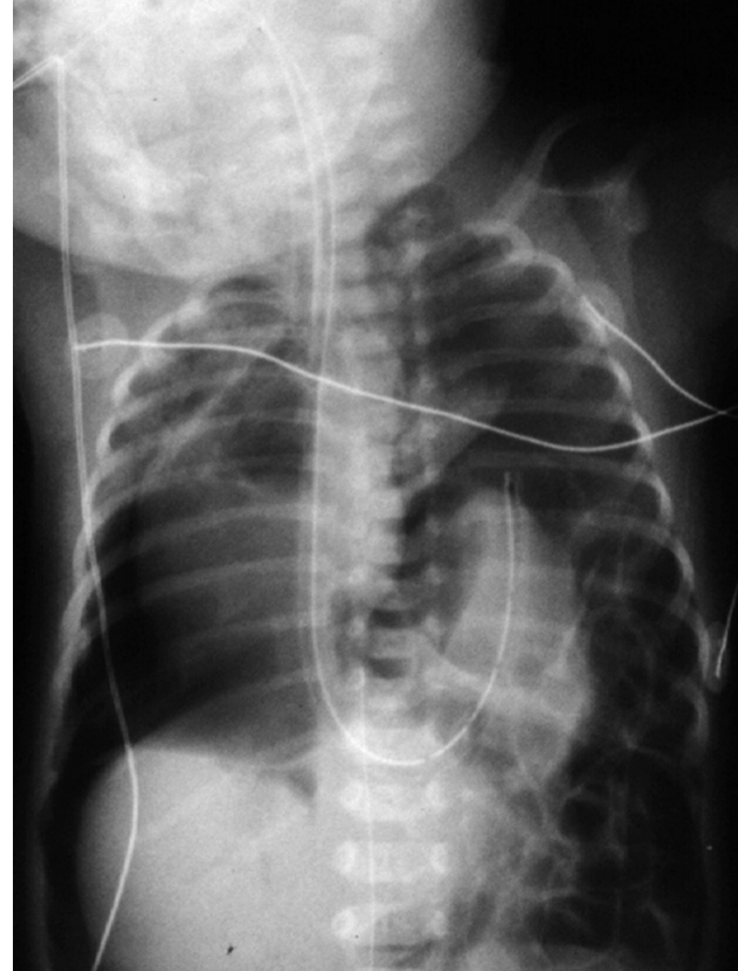
Diagnosis

- Affected infants have a scaphoid abdomen, with heart sounds shifted to the right
- Chest x-ray demonstrates loops of intestines in the chest
- Location of stomach is confirmed by placement of a NGT
- CXR shows shifting of cardiac silhouette into contralateral thorax
- CXR is unreliable for estimating degree of pulmonary hypoplasia

Diagnosis



Diagnosis



Treatment

- CDH is a physiologic emergency and not a surgical emergency
- Pregnant mother should be referred to appropriate tertiary perinatal center where full array of respiratory care strategies (NO, Oscillating ventilators, ECMO) are available
- Endotracheal intubation and NGT insertion

Treatment

- **Bag-mask ventilation is contraindicated to avoid distension of stomach and intestines in thoracic cavity – antenatal diagnosis +++**
- Proper temperature regulation, glucose homeostasis, and volume status
- Most infants can be successfully managed with pressure-cycled ventilator
- Goals of respiratory support are $\text{PaO}_2 > 60$ and $\text{PaCO}_2 < 60$

Treatment

- Extremes of hyperventilation (high rates and high pressures) should be avoided
- High frequency technologies like jet ventilator or oscillating ventilator may be required
- Antihypertensives to decrease pulmonary HTN like NO, Viagra
- ECMO if everything fails

Treatment

- **The Past**
- A purely anatomic vision of the disease/ urgent almost heroic surgery / high ventilatory pressures to inflate the ipsilateral lung

- **The Present**
- Physiology >> Anatomy in CDH
- Antenatal diagnosis crucial
- Delay surgery until pulmonary hypertension is controlled

The Past

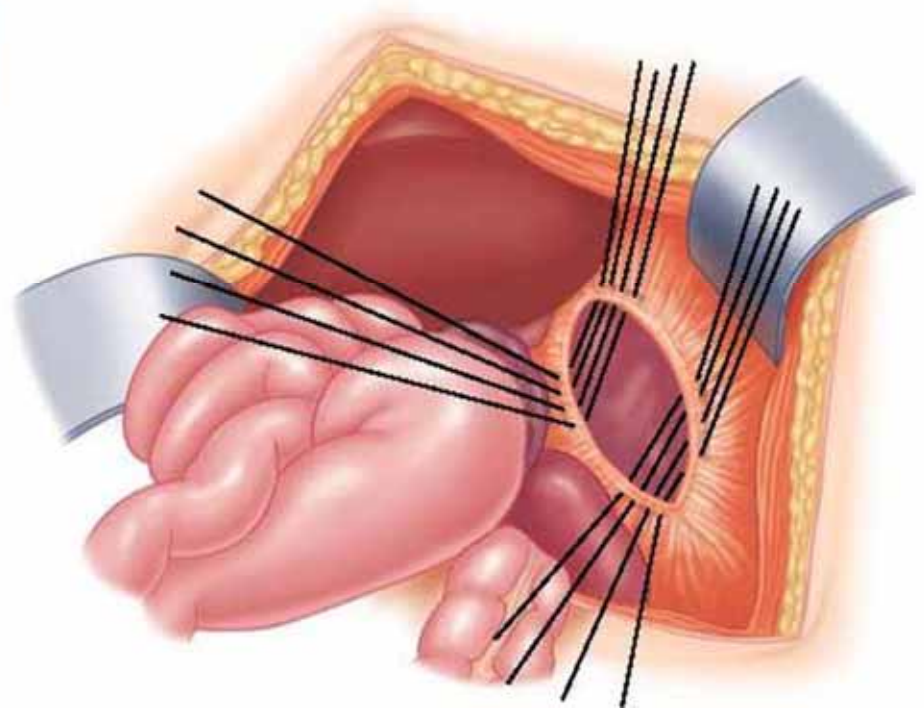
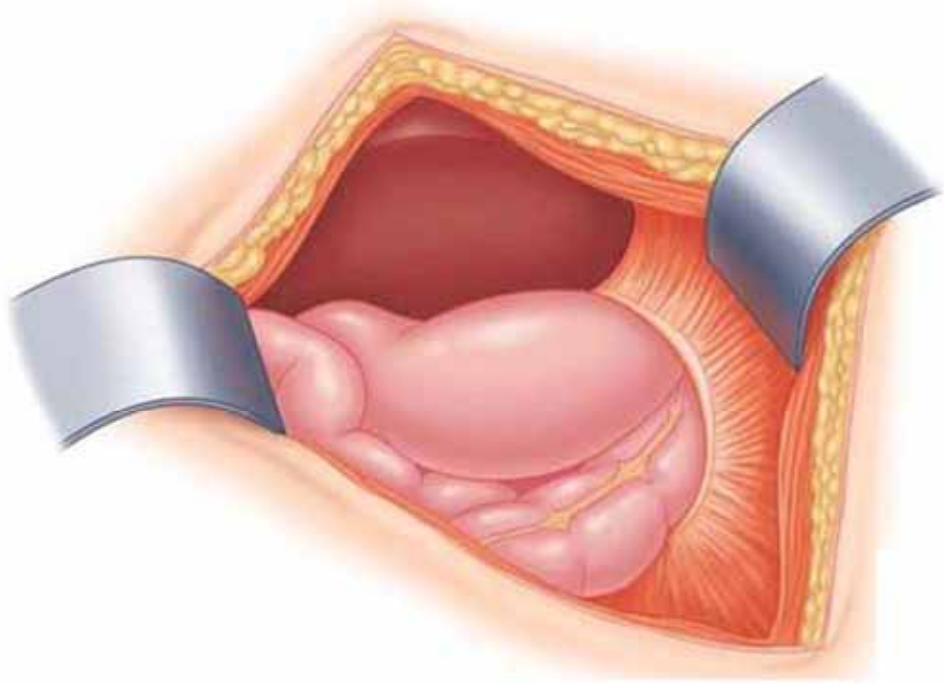
“ I’ll never forget my most spectacular diaphragmatic hernia...I raced eleven blocks to the other hospital, parked my car on the sidewalk, and rushed into the lobby, only to find the elevators were not running.

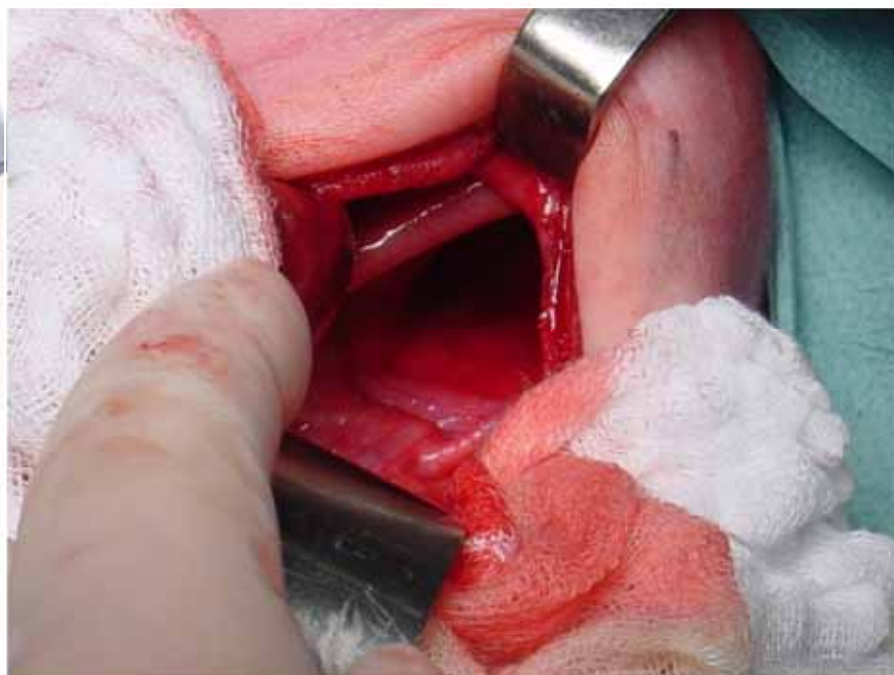
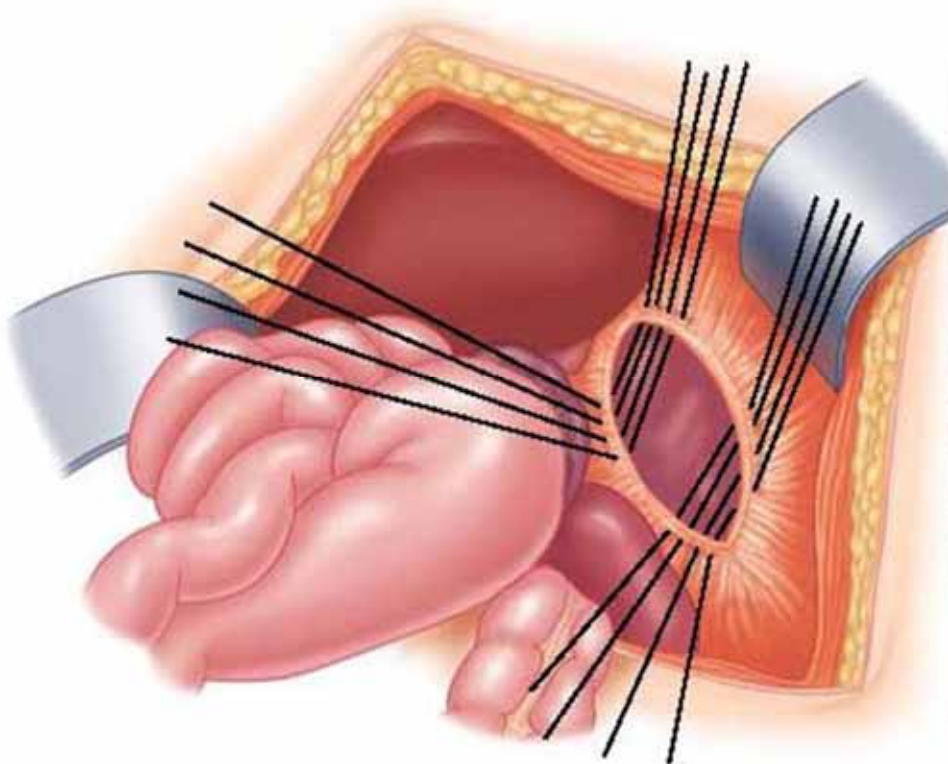
So I ran up to the ninth floor, wrapped the baby in a blanket, ran down the same nine floors and placed the baby on the floor of my car by the heater. Back at the Children’s Hospital, I ran up two flights to the operating room and laid the baby on the operating table. By now the little fellow was dark blue and apparently lifeless. Without taking any sterile precautions, I slashed an incision across the left side of the chest, inserted my fingers, and pulled out the abdominal organs that had made their way up into the chest, thereby relieving the pressure on the lungs and the heart. Then I began to massage his tiny heart with one finger. It began to beat...We...inserted an endotracheal tube into the baby and I completed the operation. About 25 years later, my secretary ushered into my office a young man about 6’4”who stood before me and said “My father thought you’d like to meet me...”

- C. Everett Koop, MD: “Koop” 1991

Surgery

- *Delayed surgical repair is now widely accepted*
- Most institutions perform surgery in NICU
- Laparotomy, thoracotomy, laparoscopy and thoracoscopy
- When diaphragmatic tissue is adequate, primary repair with nonabsorbable suture can be done





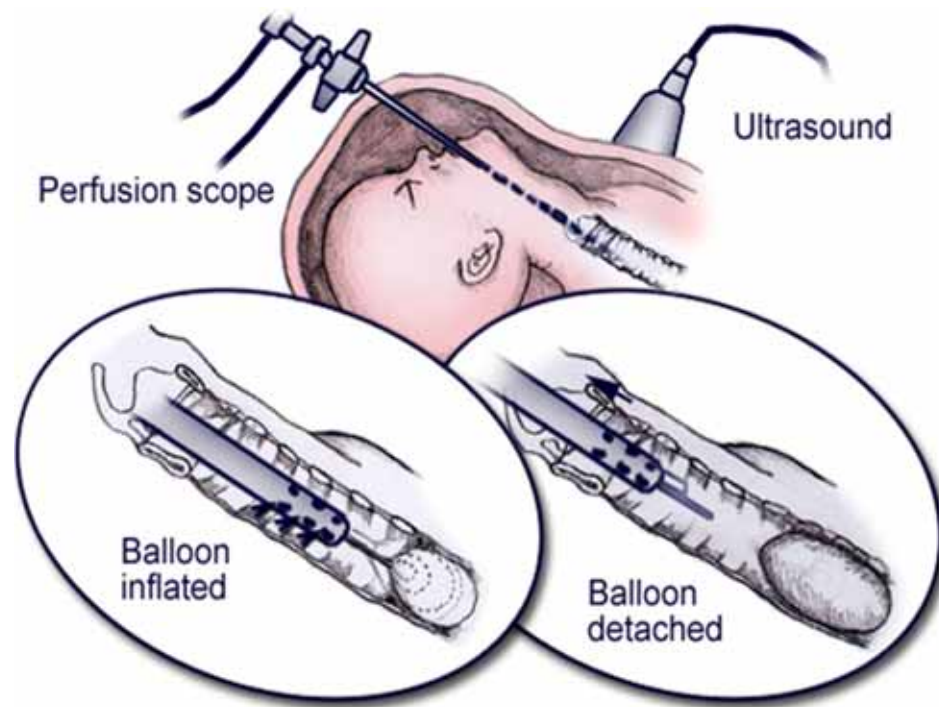


Outcome

- Survival rates for isolated CDH have improved dramatically (>90%)
- Major predictor of survival is resolution of pulmonary HTN
- In the survivors, long term problems include chronic lung disease, developmental delay, nutritional and growth related problems

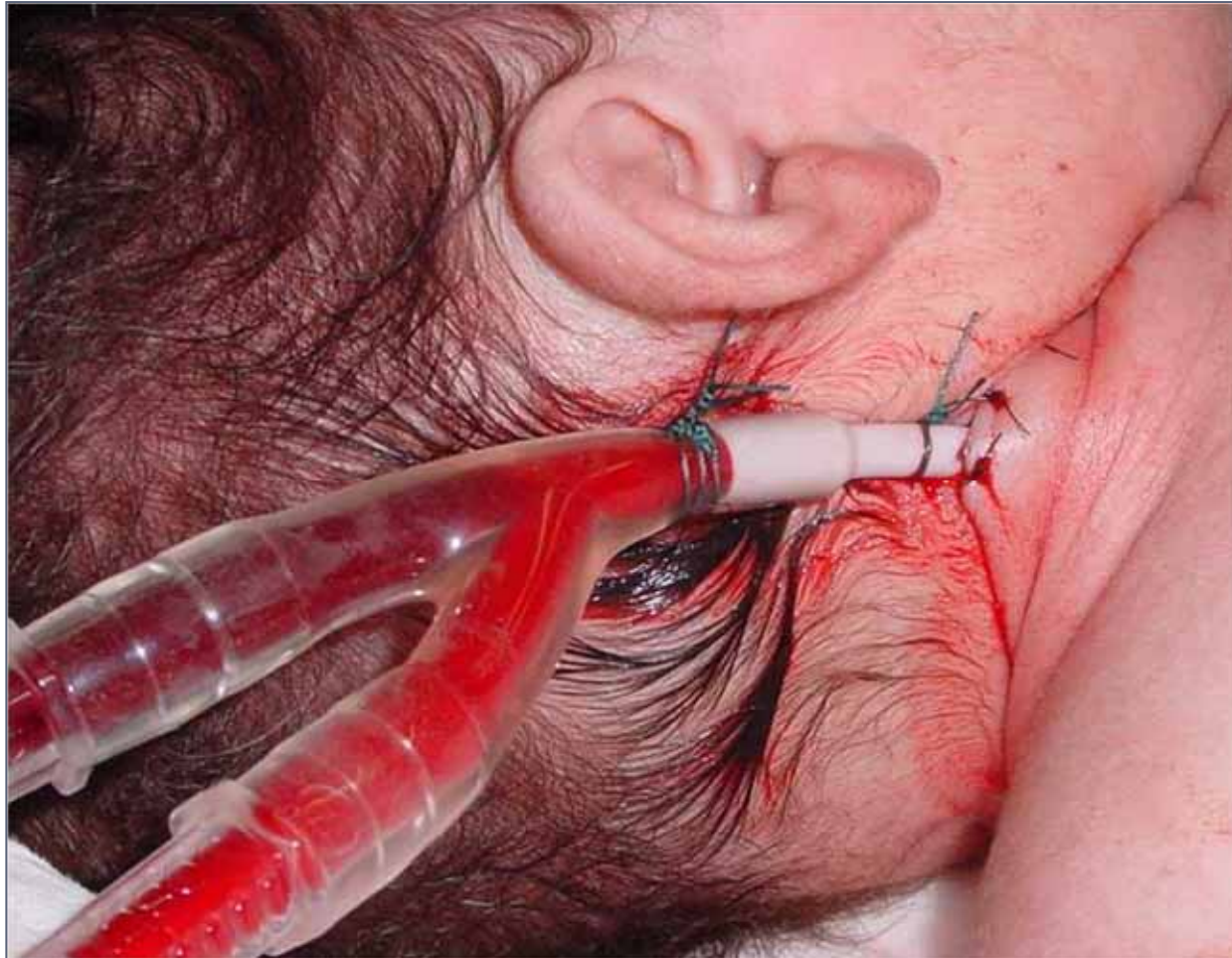
Evolving Therapies

- In utero surgery – Fetal Endoscopic Tracheal Occlusion for Severe Fetal CDH



The benefits of improved lung growth due to fetal tracheal occlusion may be outweighed by the adverse effects of premature delivery

Evolving Therapies - ECMO



Evolving Therapies - ECMO



Baby S. – CDH with antenatal diagnosis and managed with a “modern” attitude

- Antenatal diagnosis of CDH at 30 weeks of pregnancy by a private obstetrician
- Referred to Victoria Hospital on 18/08/2013 with uterine contractions

by date : term

by USG : 32/52

Steroids given to mother to help maturation of foetal lungs

Neonatal period

- Elective CS done on 20/08/2013
- BWt 2.0 kg
- Apgar at 1" : 8/10, at 3" : 4/10

- Heart sounds shifted to the right
- Even before the diagnosis was confirmed by a chest X-Ray, the baby was intubated and ventilated and a naso-gastric tube inserted
- Surfactant given via endotracheal tube

Delayed repair

- Baby kept on ventilator with gentle ventilation to prevent barotrauma / spontaneous ventilation with permissive hypercapnia / no muscle relaxants
- Baby was stabilised and surgery performed only 4 days later, to give time to the pulmonary vascular resistance to fall

Congenital Diaphragmatic Hernia

Permissive Hypercapnea/Spontaneous Respiration

How is it done?

- nasal intubation
- time cycle/pressure control infant ventilator
 - no muscle relaxants
 - no hyperventilation
 - no induced alkalosis
- to keep *pre-ductal* SaO₂ > 90%
 - like to keep PCO₂ < 60
 - Kinder, gentler ventilation

Surgery

- No anesthetic inhalation agents were used, particularly no nitrous oxide
- Left posterolateral diaphragmatic hernia containing: stomach, 2/3 small bowel, spleen, left lobe of the liver
- Hernia reduced carefully and defect closed without using any prosthetic material
- No chest drain inserted in left thorax



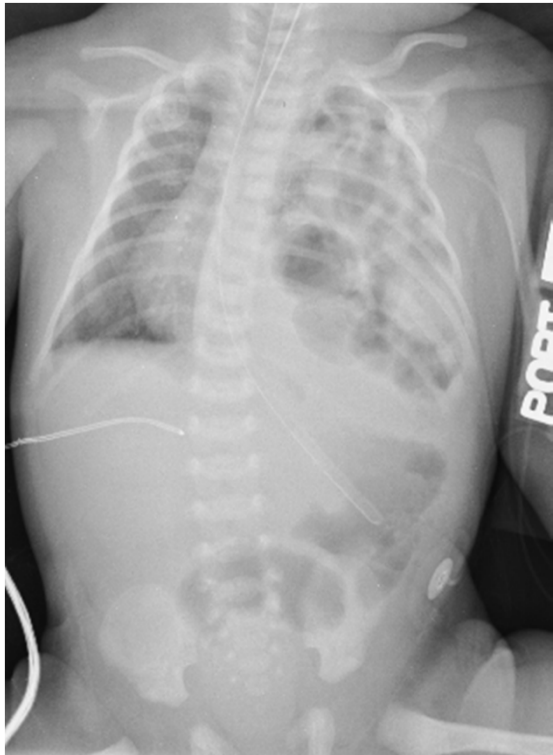
WITH CHEST TUBE



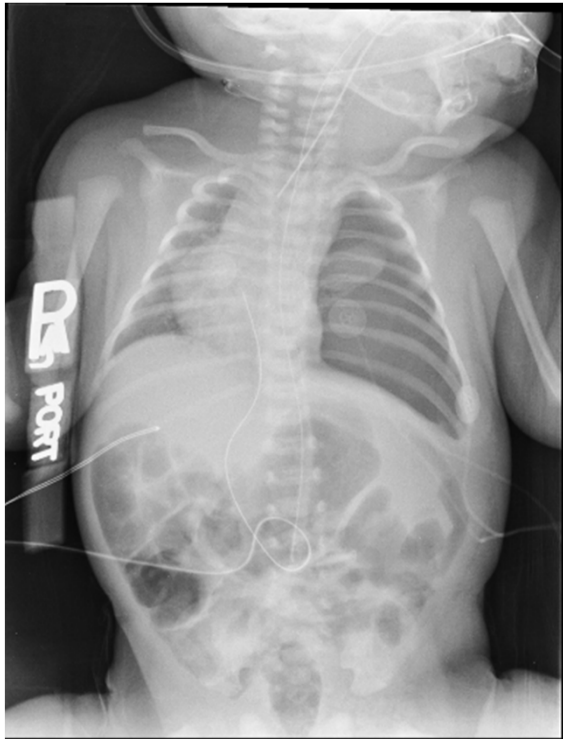
WITHOUT CHEST TUBE

Post-op care

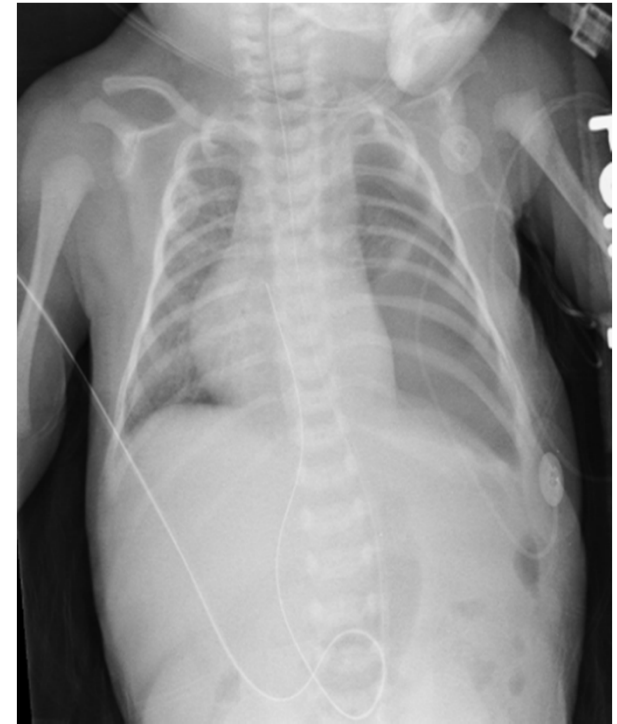
- Gentle ventilation/ to prevent a sudden mediastinal shift towards the left
- Chest Xrays showed progressive improvement
- Baby extubated on 27/08/2013, 4 days postop
- Oral feeds increased progressively to achieve exclusive breastfeeding 2 weeks after surgery
- Baby doing well/ chest Xray shows a well inflated right lung/ discharged from hospital



pre-op



4 days post-op



10 days post-op

CONCLUSION

CDH is not necessarily a grim diagnosis!

- CDH is a physiological, not an anatomical emergency.
- Surgical repair can be delayed without fear.
- Treat the baby, not the PaO₂ and PaCO₂.
- High airway pressure and FiO₂ can ruin the lungs.
- Iatrogenic disease can be more prevalent than intrinsic disease.
- Most infants with CDH can be saved with a simple ventilator, few drugs, and without the latest device or technique
- Antenatal diagnosis +++