Congenital Diaphragmatic Hernia (CDH)

Paediatric Surgery Rounds
Faiz Tuma, PGY2
Sept 26, 2003
Introduction

- Incidence: 1 in 2,000 to 1 in 5,000 births
- Most common congenital abnormalities
- 1/3 of the infants with CDH are stillborn
- 80% are left-sided and 20% right-sided
- Cause is unknown - ? Multifactorial
Smaller Right Lung Pushed Over

Compressed Heart, Pushed Over

Hole in the Left Diaphragm

Small, Compressed Left Lung

Intestines in the Chest Cavity

Spleen

Stomach Herniating into the Chest Cavity
Defect in closure of the pleuroperitoneal canal

Abdominal viscera herniate through the lumbocostal trigone into the ipsilateral thoracic cavity

Compress lungs → pulmonary hypoplasia → determines the C/P long-term outcome
- An emerging school of thought attributes the pulmonary hypoplasia to an early developmental insult to the lung and diaphragm

Contents: SB, spleen, stomach, colon, and liver
Unilateral diaphragmatic hernia ➔ ipsilat and contral abn pulmonary development

Ipsilateral hypoplasia is more severe.

Pulmonary vascular bed is also distinctly abnormal

Increased susceptibility to development of pulmonary hypertension.

Right-to-left shunting
**Diagnosis**

- prenatal U/S, accuracy 40% to 90%
- Polyhydramnios in 80% of cases
  - due to kinking of the GEl junction by herniation of the stomach into the thorax
- C/P depends on degree of pulmonary hypoplasia and reactive pulmonary hypertension
- respiratory distress at birth – milder respiratory symptoms
Diagnosis

- O/E:
  - scaphoid abdomen
  - asymmetrically shaped funnel chest
  - swallowed air $\rightarrow$ distended abd
  - deviation of the trachea $\rightarrow$ obstruction of venous return $\rightarrow$ hemodynamic conseq
DDx

- eventration of the diaphragm
- anterior diaphragmatic hernia of Morgagni
- congenital cystic disease of the pulmonary parenchyma
- unilateral pulmonary effusion
- primary agenesis of the lung
Prognostic Factors

- **Bad Prog. factors:**
  - U/S ratio of lung to head size
  - fetal liver within the thorax

- **Unreliable factors:**
  - Obstetrical factors - polyhydramnios
  - Anatomic factors
    - position of the stomach
    - size of the diaphragmatic defect
Treatment

- was considered a surgical emergency
- Now physiologic emergency rather than a surgical emergency
  - ET intubation
  - GI decompression
  - positive-pressure mechanical ventilation
- Bag-mask ventilation is contraindicated
Treatment

- pulmonary hypoplasia and pulmonary HTN → resp. fail. → needs maximizing medical treatment
- Contralateral pneumothorax is always a risk
- A broad spectrum of drugs and antihypertensive agents have been used in attempts to modify the pulmonary vascular resistance
**Surgical Treatment**

- Delayed surgical repair has resulted in improved survival rates
- Most surgeons approach the defect through a subcostal incision, some prefer thoracotomy incision
- Abdominal contents are reduced
- When diaphragmatic tissue is adequate, a primary repair with nonabsorbable suture material can be performed
Surgical Treatment

- when the defect is too large to be closed in a primary fashion, numerous reconstructive techniques have been tried
- abdominal wall closure may not be possible
  - simple closure of the skin can be accomplished with repair of the ventral wall defect some months later
**Surgical Treatment**

- When ventilatory therapy has failed ECMO, can be used.
- ECMO allows respiratory support without the risks of barotrauma and oxygen toxicity associated with conventional ventilation.
Outcome

- survival rate 39% to 95%, mean 69%
- many surviving infants will have nl PFT
  - total lung capacity,
  - vital capacity,
  - carbon monoxide diffusing capacity
- new gp of survivors is emerging
Outcome

- Long-term problems:
  - chronic lung disease
  - neurologic abnormalities with develop delay
  - skeletal deformities
  - nutritional and growth-related problems.
Evolving Therapies

- CDH still frustrating and diffic clinical problem
- Open fetal surgical repair of CDH has been undertaken → the clinical results have been disappointing
- Observation: fetal tracheal ligation accelerated fetal lung growth and reversed pulmonary hypoplasia
- Partial liquid ventilation was attempted in a small number of CDH infants on ECMO → Improvements in oxygenation and PFT
Evolving Therapies

- Successful neonatal lung transplantation for CDH has been achieved, but there is no long-term experience at this time.
- Pharmacology: involves manipulation of pulmonary vascular tone and parenchymal growth and development.
Fetoscopic temporary occlusion for CDH: Prelude to a randomized, controlled trial

- Michael R. Harrison, Roman M. Sydorak, Jody A. Farrell, Joseph A. Kitterman, Roy A. Filly and Craig T. Albanese

- Journal of Paediatric Surgery

- Volume 38, Issue 7, Pages 997-1129 (July 2003)
Objective

- high postnatal mortality seen in fetuses CDH with liver herniation and low lung-to-head ratio (LHR) appears to be improved in fetuses who undergo fetoscopic temporary tracheal occlusion (TO).
Materials and methods

- 1996 -1999
- University of California at San Francisco (UCSF) Fetal Treatment Center
- 19 fetuses → fetoscopic temporary TO (Fetendo clip) for CDH
  - Previous 8 Lt sided CDH
  - 11 new: 8 Lt sided + 3 Rt sided
Materials and methods

- Choosing Criteria:
  - (1) diagnosis of CDH made before 25 wk gest,
  - (2) a major portion of the liver herniated into the hemithorax,
  - (3) lung-to-head ratio (LHR) < 1.4 for left CDHs
  - (4) normal karyotype and echocardiogram
  - (5) no other anomalies detected by prenatal US
Materials and methods

- multidisciplinary team:
  - pediatric surgeon,
  - perinatologist,
  - neonatologist,
  - psychiatric social worker,
  - anesthesiologist
Materials and methods

- Offers to Families:
  - (1) standard postnatal care,
  - (2) termination (if < 24 wks gest)
  - or (3) fetal temporary TO

- During the period of this study,
  - 45 families fulfilled all criteria.
  - Of these,
    - 19 families chose fetal intervention,
    - 7 decided to terminate the pregnancy,
    - 19 opted for conventional postnatal care.
Materials and methods

- a fetoscopic approach (the Fetendo clip procedure) was attempted first, and, only if unsuccessful → an open hysterotomy

Procedure:
- maternal laparotomy with exposure of the uterus.
- Using US guidance, the fetus is fixed in neck extension position and stabilized by placing a temporary transuterine chin suture
**Materials and methods**

T-fastener and suture are placed in the fetal trachea to aid in locating the midline fetal neck. After anterior tracheal dissection, a tracheal "screw" can be placed in the anterior tracheal wall to facilitate safe posterolateral dissection, if necessary.

GA via maternoplacental circulation + fetus is relaxed with an i.m. Pancuronium and Fentanyl injection.
Materials and methods

Fetoscopic instruments → dissect the trachea, identify the recurrent laryngeal nerves, and apply 2 titanium transtracheal occluding clips

warmed irrigation and suction → maintaining a homeostatic environment, facilitates visualization

Once the clips have been applied, the T-bar and chin stitch are removed, the port sites are closed via instillation of a fibrin glue sealant followed by a single figure of 8 medium-sized absorbable suture.
**Materials and methods**

Betamethasone to mature the fetal lungs pre-, intra- and post-operative tocolysis to relax uterus

Mothers discharged when no evidence of membrane rupture or separation, and preterm labor could be controlled with orally administered tocolytics.

After discharge, bedrest, biweekly exam including fetal US
Materials and methods

Delivery: The fetus, while still on placental support, is delivered partially. The fetal neck is exposed and the clips removed with rigid bronchoscopic visualization. Lung fluid is aspirated, and surfactant is given immediately after endotracheal intubation.

Repair of the diaphragmatic hernia was performed electively when the baby was stable, from 2 to 8 days after birth.

Survival was defined as alive at 90 days of age. Long-term survivors were all infants surviving beyond this time.
Results

- Operation:
  - Average operating time was 221 minutes ± 69 minutes
  - Average blood loss was 221 mL ± 127 mL with none of the mothers requiring a transfusion.
  - 5- or 10-mm balloon tip trocars used in 1st 5 cases, then 3 5-mm radially expanding trocars has been used
Fetal outcome and factors affecting survival

- 13 of 19 fetuses (68%) survived 90 days after delivery
- 1 fetus died in utero on the 2nd PO day after prolonged postoperative bradycardia with no determinable cause at autopsy.
- The other 18 infants were all liveborn
Stratified by LHR, the 12 patients with LHR less than 1.0 had a survival rate of 63%, with only one infant requiring ECMO support.

The 7 patients with LHR greater than 1.0 had a survival rate of 86% also with only one requiring ECMO support.
<table>
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<tr>
<th>LHR</th>
<th>Days of Tracheal Occlusion</th>
<th>Nissen</th>
<th>Redo CDH</th>
<th>Chylo</th>
<th>Tracheal Injury</th>
<th>Comment</th>
<th>Outcome</th>
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<td>VC paresis, tracheostomy, Cotton procedure</td>
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Statistically, there was no difference between the 13 survivors and 6 nonsurvivors in:

- maternal age
- gestational age at prenatal diagnosis
- LHR
- gestational age at Fetendo clip
- interval between the fetal and delivery procedures or tracheal clip days
- birth weight and gender.
5 liveborn infants did not survive the neonatal period.

- 1 infant with a normal karyotype was diagnosed as having multiple pterygium syndrome immediately after birth.
- 2 fetuses showed no response to tracheal occlusion and died shortly after delivery.
  - 1 infant had left pulm agenesis at autopsy.
  - The other had no lung expansion before birth → severe fetal distress with cardiac arrest during the delivery, and died on day of life 3.
One infant suffered a pulmonary hge during CDH repair on day of life 3.
The last infant died at 78 D of life from sepsis caused by obstructed isch. Bowel.
Average number of hosp days for liveborn infants was 66 D ± 49 D and 90 D ± 35 days for long-term survivors.
Neonatal outcome

- 15 / 18 liveborn infants were stabilized after delivery and underwent diaph. repair using a Gore-Tex patch at the age of 4.9 ± 2.5 days.
- postnatal imaging and subjective operative appearance showed that the lungs were markedly larger than other CDH babies without fetal intervention.
In the 13 long-term survivors:
- there have been 3 major types of morbidity:
  - tracheal injuries
  - gastroesophageal reflux
  - recurrent diaphragmatic herniation
- 7 had some form of tracheal problem (e.g. bilateral recurrent laryngeal nerve injury tracheomalacia)
  - 4 had bilateral nerve injury leading to vocal cord paresis
  - 2 died from tracheostomy dislodgement at home
  - 2 had tracheomalacia with collapse of the airway on inspiration
- 5/18 liveborn infants had tracheal injuries that required treatment
Neonatal outcome

*Significant GER has necessitated a Nissen fundoplication in 7 patients;
*SBO and NEC with sepsis developed in one other patient.
*6 patients have had symptomatic separation of the Gore-Tex patch.
*Other complications:
  - pleural or lung infections in 2 patients
  - ipsilateral chylothoraces after CDH repair in 4 infants
Late mortality:
Of the 13 infants that survived 90 days:
- 3 have since died.
  - 2 of these died at home at 11 and 15 months of age, secondary to tracheostomy accidents.
  - The other late death at 9 months was from sepsis associated with a case of meningitis.

As of June 2001, the average follow-up for long-term survivors is 41 months (range, 26 to 63 months).
Ten infants currently are alive and well for a survival rate of 53%.
Discussion

- Without prenatal intervention, high-risk infants do poorly even with optimal postnatal care with mortality rates of 60% for LHRs less than 1.4 and nearly 100% for LHRs less than 1.0.
- In this experience with 19 patients, all with LHRs less than 1.4 and most less than 1.0, suggests that the Fetendo clip procedure may improve survival rate when compared with fetuses treated after birth with standard care.
**Discussion**

- Evolution in the technical aspects of in utero treatment of CDH:
  - from hysterotomy and complete repair to
  - hysterotomy and internal tracheal occlusion, to
  - hysterotomy and tracheal clip, and finally to
  - fetoscopic tracheal clip (Fetendo clip)

- further evolution in techniques, endotracheal occluding device (balloon) through a single, small port.
Discussion

- Striking evolution in technique:
  - Ports from four 10-mm ports to three 5-mm ports, by miniaturization of ports, irrigating scope, continuous perfusion pump, and methods of fetal positioning and fixation.

- 2 persistent problems needs resolution:
  - Maternal morbidity related to fetal intervention
  - Fetal tracheal complications related to the neck dissection and clip occlusion
**Discussion**

- **Significant Problem:**
  - 7 of 13 long-term survivors had tracheal injury; 4 of them needed a permanent tracheostomy secondary to bilateral recurrent laryngeal nerve paralysis.
  - Tracheal stenosis and tracheomalacia after tracheal clipping also have been troublesome.
- **These difficulties leaded to development of endotracheal balloons**
Duration of TO:

- Optimal amount of time tracheal occlusion needs to be present to achieve maximal effect is a major area of controversy.
- Short-term tracheal occlusion (2-3/52) can result in significant functional improvement in lung function even in the absence of lung growth, and at the same time preserve the number of type I and II cells.
- Balloon will be easier to deflate.
Discussion

- Failures and complications:
  - postoperative in utero demise of one fetus
  - the lack of physiologic response to TO in 2 other fetuses who died shortly after birth
    - ? Due to failure of occlusion, bec even tiny leak will negate the physiologic response
  - associated lethal anomalies – 2 fetuses died
**Discussion**

- **long-term outcome:**
  - only 1 of the neonates had no complications.
  - Most required interventions:
    - GERD
    - recurrent diaphragmatic hernia
    - tracheal injury
    - chylothorax.

- However, given that many of these survivors were in the highest risk category with LHRs less than 1.0, a status that predicts nearly 100% mortality in historical controls, the simple fact that they are alive is significant.
Discussion

- Fetendo clip technique, is technically difficult with significant complications and should be superceded by simpler techniques such as Fetendo balloon
**conclusions**

- The strategy of temporary TO enlarge the hypoplastic lung, must be evaluated in a proper prospective, randomized trial.
- The timing of any proposed trial is crucial because further positive experience with this new technique will surely lead to increasing clinical application.
**conclusions**

- Women carrying fetuses with a left-sided CDH with the liver herniated, an LHR less than 1.4, and no other detectable fetal anomalies are assigned randomly to endoscopic fetal occlusion or optimal postnatal care including ECMO support, inhaled nitric oxide, and high-frequency ventilation.
- Outcome variables include neonatal mortality as well as long-term morbidity and cost.
- Enrollment has been surprisingly good, and the results will be reported when available.
Thank you for listening...
Thank you for all what you did for me in my rotation...