Prenatal Airway Management, EXIT & CDH

Gavin Morrison MA FRCS
Consultant Paediatric Otolaryngologist

Guy’s, St Thomas’ & Evelina Children’s Hospitals, London UK

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Co-Workers

- Professor Kypros Nicolaides, MD, MRCOG
  Foetal Medicine, King’s College Hospital, London

- Mr Sean Blaney, PhD FRCS Dept Paediatric ORL,
  Guy’s & St Thomas’ and King’s College Hospitals,
  London

- Mr Gavin Morrison, MA FRCS, Dept Paediatric ORL,
  Guy’s & St Thomas’ and King’s College Hospitals,
  London
Paediatric ENT Services – South London

- 3 hospitals
- 1 medical school
- Two departments of foetal medicine
- 10,000 live births p.a.
- 70 PICU/NICU Beds
- Local Population 5.5 Million
Incidence of prenatal diagnoses

- 4.8% of patients have one or more Congenital Anomalies on antenatal U/S
  - Of these 2.5% involved Head & Neck
Ultrasound Diagnoses Reported in Literature

- Cervical Lymphangioma (Cystic Hygroma)
- Lingual Lymphangioma
- Cervical Teratomas
- Epignathus (Teratoma arising from Basisphenoid)
- Laryngeal & Tracheal Atresia/Stenosis
- Congen. Thyroid Goitre
Prenatal Investigations

- Serial Maternal Ultrasound
- Fetal MRI Scanning
- Maternal Alpha-fetoprotein
- Amniotic Fluid
  - Alphafetoprotein
  - Acetylcholinesterase
Morbidity & Mortality

- **Cystic Hygroma** 1 in 6,000 Pregnancies
  - If diagnosed before 30 wk poor prognosis

- **Cervicofacial Teratomas** *(J Paed Surg 1995;30(2):312)*
  - 1 in 4,000 live births
  - Lifethreatening Airway Obstruction in 35 %
  - 10% Death
  - 10% Hypoxic Cardiac Arrest
  - Pre U/S diagnoses 30 % Mortality

- **Laryngeal Atresia** *(J Paed Surg 1994;29(2):271)*
  - Of 16 Cases worldwide Diagnosed Prenatally between 1988 -1994 only ONE survived.
- A means of providing time to secure an airway while uteroplacental gas exchange preserved

- 1st Described by Harrison et al 1997
EXIT Ex-Utero Intrapartum Treatment
Indications for EXIT

- Mass compressing the Foetal Airway
- Abnormal upper airway anatomy
- CHAOS

- *Prenatal treatment for CDH*
Diagnostic U/S Features

Giant Foetal Head or Neck Mass
Obstructive Lesion Seen in relation to airway
FOETAL MRI

- Rapid sequence MRI
- HASTE (Half-Fourier single shot turbo spin echo)
- Transverse, sagittal and coronal planes
Ultrasound Findings in CHAOS

- Polyhydramnios
- Echogenicity of Lungs
- Dilated Trachea
- Flattened or Inverted Diaphragm
- Abdominal Ascites
- Cardiac Compression
Technique of Ex-utero Intrapartum Airway Surgery

- Multidisciplinary Team
- Caesarian Section approach
- Deep GA to relax Uterus & Maintain Feto-Placental Circulation
Technique of Ex-utero Intrapartum Airway Surgery

- Multidisciplinary Team
  - Obstetrician
  - Anaesthetist
  - Neonatologist
  - ENT Surgeon

- Deliver Head & Torso only through Uterine incision

- Monitor Scalp ECG & Pulse Oxymetry

- Do not ligate Umbilical Cord

- Intubate/perforate or Trachaeostomy
EXIT Technique

- **Deep Maternal GA**
  - Isoflurane & N₂O
  - Discontinue N₂O before Hysterotomy
  - i.v. Vecuronium to maintain muscle relaxation

- **Uterine Stapling**

- **After delivery**
  - Oxytocin

- **Foetal Anaesthesia**
  - i.m. agents
  - 20-60 Mins on Placental Support

- **Surfactant**

- **Establish Airway then Clamp & Ligate Cord**
EXIT Procedure for Cystic Hygroma
Balloon tracheal occlusion for Congenital Diaphragmatic Hernia
CDH - Background

- Incidence: 1 : 3,000 live births
  Higher including late miscarry/TOP

- Most left sided

- 2F : 1M

- Non-syndromic and Syndromic forms (>50)

- Mortality 50 %
CDH – Embryology & Anatomy

- Failure of closure of pleuroperitoneal canal at 8-10 weeks
- 90 % - “Bochdalek Hernia” in Posterolat diaphragm – mostly left sided
- Pulmonary Hypoplasia, pulmonary hypertension
- Associated Anomalies in 70 % - Trisomy 18 (21,23)
- 20 – 25 % Cardiac Defects
CDH - Prognosis

Depends on:

- Cardiac Defects
- Lung Function
- Overall survival 50 – 60 %
Prenatal prediction of Survival

- Lung-to-head circumference ratio
- Liver in thorax
- Size of diaphragmatic defect
- Lung volume
- Peritoneal/pericardial effusions
Post natal prediction of survival

- Bohn's Ventilation Index

- Gases - formula $paO_2 \ [max] - pCO_2 \ [max]$

- Prognosis improves with
  
  “Gentle ventilation” = permissive cypercapnoea & low pressures
CDH - Management

- TOP, Expectant, or fetoscopic surgery

- Prenatal balloon treatment – FETO (fetal endoluminal tracheal occlusion) Procedure

- Ventilation, HFOV or ECMO then -

- Post-natal Repair:
  - Trans-abdominal
  - Excision of sacs & reduction herniated bowel
  - $1^0$ closure or muscle or patch grafting repair diaphragm
Congenital Diaphragmatic Hernia - Principle of antenatal balloon treatment

- Occlude the trachea in utero
- Foetal lungs net producers of fluid, build up leads to lung maturation and expansion compressing herniated contents downwards
- After maturation - Retrieval of balloon to establish natural airway at EXIT birth
- Stabilize and assess lungs on NICU
- Surgical repair CDH at 48 hours if survivor
Congenital Diaphragmatic Hernias

King’s College Hospital:

12 patients

- 7 EXITS for CDH treatment
- 5 Other tracheal balloon placements
Pre-Natal Intrauterine ENT Surgery - Development

Specialised Endoscopic Instrumentation
– 1.2 mm fetoscope

Custom-made Intra-tracheal Balloon
EXIT Retrieval of Balloon Occlusion
Intubation and Delivery
Problems & Evolution of Techniques

- What optimal gestation to insert balloon?
  Trend for balloon insertion earlier in Gestation - 32 weeks → 26 weeks

- Failures due to leak around balloon - full inflation / tracheal growth

- How to safely remove the balloon

- Emergency if spontaneous onset of early labour
2004 - 2007 Developments

- Diagnose CHD @ 20 + weeks
- Fetoscopically placed tracheal balloon at 26 weeks gestation
- *Ultrasound guided trans-uterine trans-fetal-thoracic detachable balloon puncture at 34 weeks*
- *Or Fetoscopic removal at 34 weeks*
- Normal vaginal delivery at term
- Diaphragm repair day 1-4
Patient Selection & Controls

- Only severe most life-threatening CDH cases
- 20 week U/S - Lung area : head circumference ratio < 1.0
- Controls same criteria who elected not to proceed
Results of Foetal Balloon Placement for CDH

- **Collaborative Team:**
  - King’s College Hospital, London UK – K Nicolaides, G Morrison, S Blaney
  - Catholic University of Leuven, Belgium - Prof Jan de Prest et al.
  - Fetal Medicine Unit, Department of O&G, Hospital Clinic, Barcelona, Spain - Prof Anne Debeer et al.

- **Over 30 Balloon Placements**

- **Control Group = Balloon refusers with same severity** >90 % Mortality
Results of Foetal Balloon Placement for CDH


- Increased incidence of premature rupture of membranes from intervention

- 75 % early neonatal survival
- 66 % surgical repair of CDH
- 58 % survival to 28 days
- 50 % long-term survival with no developmental problems so far.
Evolution of a technique

- Constant changes in indications and technique
- Balloon placement earlier in pregnancy
- EXIT delivery for balloon retrieval
- Pre-Natal endoscopic intrauterine Balloon removal at 34 weeks
- Allow natural birth or standard C-section delivery at term

- Strict Ethical Monitoring
- Committee for Innovative Interventional Treatment
CDH – Management pathway

In utero diagnosis < 26 weeks CDH

Rule out associated anomalies

Isolated position liver & measure LHR

Associated: nature of anomalies

Multidisciplinary counseling

“Severe” = LHR < 1.0 & liver “up”

“good” or “intermediate”

Serious anomalies

FETO 26-28 weeks
Unplug 34 weeks

Expectant

TOP

In utero transfer & optimal postnatal therapy

Postnatal counseling
Long-term Outcome

- Lung function often normal
- Neurological defects – developmental delay
- Hearing loss
- GER
- Growth failure (multi-factorial)
Conclusions

- Early Prenatal diagnosis of CDH allows potential for foetal tracheal balloon occlusion & increased survival rates
- Concept of foetal therapy is based on the ability to predict poor outcome
- 90 -100% mortality if liver herniation and LHR < 1.0
- FETO results in expansion of the foetal lungs and healthy survival until discharge in 50% of cases
- Inherent risk for amniorrheaxis and a higher chance for early preterm delivery
- If possible balloon placement and retrieval or puncture should both be pre-natal
- EXIT now reserved for term baby with balloon still obstructing.