Review of the Surgical Neonate

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The speaker has signed a disclosure form and indicated she has no significant financial interest or relationship with the companies or the manufacturer(s) of any commercial product and/or service that will be discussed as part of this presentation.

Session Summary

This presentation will review common and uncommon surgical anomalies of the neonate, including embryology, pathophysiology, current treatment options, emerging evidence and best practices, and outcomes.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- describe current national quality improvement efforts related to neonatal surgery;
- describe potential areas for future research to improve outcomes for surgical neonates.

References


**Session Outline**

See presentation handout on the following pages.
Quality Improvement Using Evidence Based Practices for Neonates

- Children’s Hospital Neonatal Database (CHND) (www.childrenshospitals.org)
- Vermont Oxford Network (VON) (https://public.vtoxford.org)
- American College of Surgeons National Quality Improvement Program-Pediatric (ACS NQIP-P) (www.facs.org/quality-programs/pediatric/overview)
- NICHD Neonatal Research Network (www.neonatal.rti.org)
- Cochrane Library (www.thecochranelibrary.com)
- Pediatrix (www.pediatrix.com)
- ELSO Registry (http://dwww.elso.org/registry/statistics/limited)

Children’s Hospitals Neonatal Database (CHND)

- Launched in 2010 (Murthy et. al., 2014)
  - 27 level 3C regional NICUs
  - ≥25 beds
  - >50% outborn
  - 100% offer ped surgery
  - 93% offer ECMO
  - 89% offer cardiac surgery
  - 24% admits for surgical evaluation/management

2012 Data

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>N</th>
<th>% survival</th>
<th>LOS Non-survivors</th>
<th>LOS survivors</th>
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</thead>
<tbody>
<tr>
<td>Gastroschisis</td>
<td>879</td>
<td>98%</td>
<td>10</td>
<td>34</td>
</tr>
<tr>
<td>Imperforate anus</td>
<td>691</td>
<td>93%</td>
<td>6</td>
<td>11</td>
</tr>
<tr>
<td>CDH</td>
<td>501</td>
<td>68%</td>
<td>16</td>
<td>37</td>
</tr>
<tr>
<td>Omphalocele</td>
<td>261</td>
<td>79%</td>
<td>16</td>
<td>20</td>
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<tr>
<td>CPAM</td>
<td>201</td>
<td>98%</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Midgut volvulus</td>
<td>183</td>
<td>97%</td>
<td>2</td>
<td>20</td>
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<tr>
<td>SCT</td>
<td>64</td>
<td>94%</td>
<td>12</td>
<td>11</td>
</tr>
<tr>
<td>TC Hirschbrung's</td>
<td>40</td>
<td>95%</td>
<td>25</td>
<td>16</td>
</tr>
</tbody>
</table>

Wide Inter-Center Variability

CHND Publications

American College of Surgeons National Surgical Quality Improvement Program-Pediatric

- ACS NSQIP
  - Developed in 1990s in VA hospitals
  - Validated assessment of surgical outcomes in adults
  - Led to improved outcomes, expanded to private sector

- ACS NSQIP-P:
  - Collaboration between ACS and American Pediatric Surgical Association
  - N= 3315 pediatric patients from 2008-2009 in 4 hospitals
  - 121 data points (dem, preop, intraop, postop, 30 day)
  - Prospective data collection
  - Beginning stages (Raval et. al., 2011)

ACS NSQIP-P

- 2010: 29 centers, 37,141 patients
- Outcomes:
  - Overall mortality= 0.25% (2.39% for neonatal cases)
  - Overall morbidity= 7.9% (18.7% for neonatal cases)
  - SSI= 1.8% (3% for neonatal cases) (Drury et. al., 2013)
- Current goal to develop procedure-specific outcomes
  - Now collects 94 data points, total of 40 beta testing sites
  - For more info: http://pediatric.acsnsqip.org/about.jsp

Review of Selected Neonatal Surgical Conditions

- Embryology
- Pathophysiology
- Initial Management
- General Outcomes
- Future research, innovations

Congenital Diaphragmatic Hernia (CDH)

First reported by Lazarus Riverius in 1679 in a 24 yr male (postmortem)

Definition: migration of abdominal viscera into the thoracic cavity through a developmental defect of the diaphragm.

Cause: failure of posterolateral retrosternal portion of diaphragm to develop normally prior to week 12

Incidence: 1:2000-5000 live births

Presentation: 85% posterolateral= Bochdalek (left) 2-15% retrosternal= Morgagni (right) Males > females; avg GA 38 weeks; 3 Kg 28% cardiac anomalies Survival varies; 28% mortality in CDHG

CDH

I. Development of diaphragm (4-12 weeks)
  - Septum transversum membrane
  - Pleuroperitoneal membrane
  - Dorsal mesentery of esophagus
  - Body wall

II. Development of pulmonary system (4-24 weeks)
  - Pseudoglandular period (5-17 weeks)
  - Canalicular period (16-25 weeks)
  - Terminal sac period (24 weeks to birth)
  - Alveolar period (late fetal period to 8 years)
CDH

III. Development of gastrointestinal tract

- Physiologic umbilical herniation (7 weeks)
- Rotation of mid gut loop (7-10 weeks)
- Reduction of mid gut hernia (10 weeks)
- Fixation of intestines (10 weeks)

Pathophysiology: Intestinal tract in thorax = space occupying lesion → maturational arrest > decreased number of terminal airways, alveoli and pulmonary vasculature = Pulmonary hypoplasia
Pulmonary hypertension

Clinical Presentation: Barrel chest
Peri in right chest - mediastinal shift
Respiratory distress, failure
Scaphoid abdomen
Altered cardiac output

Predicting Outcomes

Congenital Diaphragmatic Hernia Study Group (Brindle et al., 2014)
A Clinical Prediction Rule for the Severity of Congenital Diaphragmatic Hernias in Newborns:
- N=2200, born 2007-2011
- range 0-8
- low birth weight (1)
- absent (2) or low 5-minute Apgar score (1)
- chromosomal anomaly (1)
- major cardiac anomaly (2)
- severe pulmonary hypertension (2)
- risk of death: high (~50%): ≥3
  intermediate risk (~20%): 1-2
  low risk (<10%): 0

Prenatal O/E LHR
- (observed/expected lung to head ratio)
  - Fetal MRI
  - significant correlation for survival or death at 45 days of life (Sebastià et al., 2014)

Use of iNO

Inhaled Nitric Oxide Use in Neonates With Congenital Diaphragmatic Hernia: (Campbell et al., 2004)
- N=1713 born 2003-2011
- Wide inter-center (33) variability of use (34%-92%)
- $81 million dollars in iNO costs
- No statistically significant change in ECMO use or mortality
- Routine use not recommended

CDH: Management

NTE
Respiratory
Avoid PPV with bag and mask
Mild hypercarbia
Consider surfactant replacement
Decompress GI tract

Cardiac/Fluids
- Limit fluid boluses
- ECHO
- Fluids
  - IV Access
  - Monitor glucose, ion calcium
Sedation
- Minimally invasive care
- Procedural pain treatment
Survival and Morbidities

3 year pulmonary function: abnormal forced expiratory flow, elevated FRC and residual volume; normal total lung capacity (Panitch et al., 2014)

Study of 18 survivors (Leeuwen et al., 2014)
Neurodevelopmental outcomes of CDH patients were no different to healthy matched controls at 12 and 36 months using Bayley
Risk of sensorineural and conductive hearing loss (30% in one study) (Partridge et al., 2014)

Innovations in CDH Repair

- Autologous tissue engineering
  - 51% of CDH cannot be repaired primarily
  - Meta-analysis of MIS vs Open Repair of L CDH with patch (Chan et al., 2014)
    - MIS: higher recurrence, longer length of surgery
    - Open Repair: longer ventilation time, higher mortality

Abdominal Wall Defects: Gastroscisis and Omphalocele

Described in the 16th century, but categorized as separate entities in 1953 by Moore and Stokes

Gastroscisis: Herniation of midgut through a defect in the abdominal wall, usually located to the right of the umbilical cord.

Omphalocele: Herniation of abdominal viscera into the umbilical cord. Viscera is generally covered by intact membrane. Presentation varies.

GI Tract Embryology

3 weeks: embryo is trilaminar 2-D disk
3-4 weeks: embryo folds laterally, cephalically and caudally

Gastroscisis

Cause: fetal vascular accident> occlusion of mesenteric artery> leading to weakness of abdominal wall to the right of the umbilical cord.

Incidence: 1: 2000-2500 births- increasing globally; Males=Females
15% SGA, preterm (CHND= avg 36 GA)
1300-1500 US children per year; $200-240 million per 40 day hospitalization (Nichol et al., 2008)

Risk factors: maternal age, parity and exposure, UTI in first trimester in young mothers (Yazdy et al., 2014 and Shaw et al., 2014)

Presentation: Midgut foreshortened, edematous, peel; 8-11% associated anomalies (32% in Texas), cryptorchidism; 15% complex

Simple vs complex different disease/conditions? (Corey et al., 2014)
Gastroschisis: Management

No benefit for elective C section over vaginal delivery in the absence of obstetric indications (Carnaghan et. al., 2014)

NTE: risk for hypothermia
Respiratory
Cardiovascular: hypovolemia r/t evaporative losses
Infection
GI: protect from injury, decompress stomach
Fluids: minimize fluid losses; aggressive fluid resuscitation
Family

Gastroschisis: Surgical Management

Primary Repair
Staged Repair

Operative Repair

CHND study 2010-2012: (Murthy et. al., 2014)
N=442, ≥34 GA, no other morbidities.
68% staged repair with increased LOS (37 vs 28 days)

Canadian study (Stanger et. al., 2014)
N=679
55% attempted primary repair, 81% successful
45% silo (staged repair) placed intentionally.
Patients with attempted primary repair were more likely to be inborn, have daytime admissions, and higher SNAP-II scores.
Successful primary repair was predicted by low risk GPS (bowel injury severity) and high volume center.
With the exception of higher rates of SSI in the planned silo group, outcomes in the successful primary repair and planned silo groups were comparable

Gastroschisis

Outcomes:
Survival= 95-99%
Quality of life rated high (Rankin et. al., 2014)
Prolonged ileus, GI dysfunction
Prolonged TPN
Undescended testes (15-30%)
Risk for:
-Infusions
-NEC (2-4%)
-GI compromise
-Short Bowel Syndrome
-psychological stress related to abnormal umbilicus

Future Research

- Prenatal intervention with patch coverage?
- Amniotic fluid manipulation
- Causes…..prevention

Omphalocele

Cause: Embryo folds incorrectly at weeks 3-4, leads to incomplete closure of abdominal wall and umbilical ring

Incidence: 1:1100 fetuses, 1:4-6000 live births (Islam, 2012)
Males=Female

Risk factors: maternal age

Presentation: Generally covered with sac, Highly associated with other anomalies (80%) and genetic syndromes (49%); small, central defects= higher incidence (Islam, 2012)
No standard definition of giant omphalocele
### Omphalocele: Management

**NTE:** risk of hypothermia  
**Respiratory:** loss of abdominal support in respirations, prematurity, relative hypoplasia  
**Cardiac:** assess for anomalies  
**GI:** protect from injury; decompress stomach  
**Fluids:** monitor fluid balance, higher fluid needs than avg  
**Anomalies:** careful assessment  
**Family**

### Treatment Options:

- Primary closure  
- Closure with patch  
- Delayed/Eschar  
- No standard for topical agents

### Outcomes:

- Survival 80-85%  
- Dependent upon size of defect and associated anomalies

### Predicting Outcomes

- Prenatal Omphalocele/Head Circumference ratio (O/HC): greater than 0.21 = 84% sensitive and 58% specific for respiratory insufficiency, need for staged closure (retrospective study) (Monteiro, 2011)

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### VACTERL ASSOCIATION (Solomon, 2014)

First described in 1970s  
1:10,000-40,000 live births  
Presence of 3 or more:
- Vertebra: 60-90% occurrence  
- Anorectal Malformations: 55-90% occurrence  
- Cardiac and Cardiovascular: 40-80%  
- TEF: 50-80%  
- Esophageal  
- Renal: 50-80%  
- Limb: 40-55%  
- Other: Hydrocephalus

### Management Plan

- Meticulous physical exam  
- Detailed family history, pedigree  
- Genetics testing, other blood tests  
- Radiographs, ultrasounds, MRI, etc  
- Regular developmental and physical follow up
Tracheoesophageal Fistula (TEF) and Esophageal Atresia (EA)

Described in 1697 by Thomas Gibson

**Cause:** incomplete separation of the lung bud from the foregut at 4-5 weeks, creating a fistulous connection.

- Esophageal atresia results from vascular accident.

**Incidence:** 1:3000–5000 births

**Presentation:**
- Males > Females
- 64% caucasian
- 80% congenital anomalies
- 37% preterm

**VACTERL Association:**
- V- vertebral
- A- anal/GI
- T- trachea
- E- esophagus
- C- cardiac
- L- limb
- N- neurological
- G- genetic

**NTE**
- Respiratory: Keep esophageal pouch decompressed, consider HOB elevated, may require oxygen, ventilation
- Cardiac: assess for anomalies
- Infection: at risk for aspiration pneumonia
- GI: NPO, IV for fluids, glucose, assess for anomalies

**Family**
- Treatment:
  - Primary repair laparoscopic
  - Staged repair
  - Esophagostomy
  - Gastric transposition (better perfusion, less leaks and strictures)
  - Colon or jejunum interposition

**Future Research**
- Tissue engineering for esophageal replacement (Maghsoudlou et al., 2014)
- non-surgical/minimally invasive, magnetic compression repair? (Zaitzky et al., 2014)
TEF/EA Outcomes (Sulkowski et al., 2014)

Outcome:
- Related to birth weight/GA, presence of anomalies (esp cardiac), RDS
- In-hosp mortality rates 5-9%
- Avg LOS= 27 days
- 17% readmitted within 30 days, 55% within 2 years (pneumonia)
- Survival lower for African American babies compared to Caucasian
  (Sulkowski et al., 2014 and Wang et al., 2014)

Morbidities:
- Anastomotic leak
- Anastomotic stricture
- Double fistula, recurrent fistula (5%)
- GERD (57% home on antireflux meds, 4-11% fundoplication)
- FTT
- Prematurity, anomalies, pneumonia

Hirschsprung’s Disease

Described in 1888 by Harald Hirschsprung

Cause:
- Failure of migration of neural crest cells of a portion of intestine
- 80% limited to recto-sigmoid area
- 70% isolated defect
- Usually noted in the neonatal period, but can be diagnosed later in life.

Incidences: 1:5000 births
Males>Females (4:1)

Risk Factors:
- Familial history
- Down Syndrome

Clinical Presentation:
- Failure to pass stool in first 24-48 hours
- Abdominal distention
- Emesis- becoming bilious
- Enterocolitis, perforation

Hirschsprung’s Disease: Management

NTE
Respiratory
Cardiac
ID: nec for colitis
GI/fluids:
- Decompress stomach
- X-rays
- IV, fluids
- Monitor electrolytes

Diagnosis:
- Contrast enema
  - Dilated rectum
  - Transition zone
  - Microcolon
- Suction rectal biopsy
  - Presence of ganglion cells
- Full thickness biopsy in the OR

Treatment:
- Primary pull through
  - Laparoscopic vs. open
- Exploratory laparotomy with stoma

Outcomes

- Morbidity increases with length of affected intestine
- Strictures, adhesions, stomas
- Enterocolitis
- Constipation
- Incontinence (10-30%)
- Poor growth, FTT
- Prolonged TPN
- hearing and visual disturbances
- congenital anomalies- kidneys and urinary tract

(Pini Prato et al., 2013)
Future Research

Transplantation of neural stem/progenitor cells (McKeown et al., 2013)

Anorectal Malformations

Aristotle described anorectal malformation in the 3rd century BC

Cause: failure of the anorectal septum to divide the cloaca at 4-6 weeks and/or the anus to perforate the perineum and recanalize by 9 weeks. Genetic cause

Incidence: 1:4-5000 births

Clinical Presentation:
50-67% associated anomalies - VACTERL
10-30% cardiac
30-50% vertebral/spinal
20-30% tethered cord;
5-10% genetic disorders
15% family history for long seg

Imperforate Anus: Clinical Presentation

Imperforate Anus Clinical Presentation

Imperforate Anus Clinical Presentation

Imperforate Anus: Management

NTE
Cardioresp: abdominal distention could impact respirations
GI
OG to LIWS
X-rays (AP, “inverted”), US, Fluor
Fluids
IV fluids, glucose, electrolytes
Genetics
VACTERL, syndromes
Imperforate Anus: Operative Repair

Continence

“Good” degree of continence
“Low” defects

Poor or absence continence
High defects
tethered cord
absent sacral vertebrae

Genitourinary

- 33-50% related genitourinary anomalies
- kidney anomalies resulting in CRF (2-6%)
- spinal cord or LS anomalies: 43% lower urinary tract
dysfunction (?neuropathic)
- VUR
- 17% gynecologic malformations in females, including
  vaginal atresia
  □ may not be recognized until puberty

Imperforate Anus

Outcomes:
Constipation
Incontinence
Strictures
Stomas
Prolapse
Tethered spinal cord
Sacral anomalies, agenesis
Management of urologic system, gynecologic concerns

Conclusion

- Congenital anomalies have been described for many
decades/centuries
- Approaches to treatment remain perplexing
- Wide inter-center variability exists
- Newly established databases will help to establish
  benchmarks to evaluate and improve quality
- Research is needed to understand prevention, treatment
  and attainment of optimal outcomes