Rare Neonatal Gastrointestinal Conditions

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Surgical Congenital Anomaly Network for Scotland

• Describe network and key objectives
• Show off some data
• Describe 2 conditions in more detail
  – Gastroschisis and Duodenal Atresia
• Ask for your help

www.scans.scot.nhs.uk
England
Population 55,977,178
Mean density 407 people/km²

Scotland
Population 5,438,100
Mean density 68 people/km²
Commissioned in 2019 (extension of SDHCN)

**Key objectives**

- Improve communication and engagement
- Prioritise discharge planning
- Develop more effective data collection
- Audit the whole pathway (antenatal – transition)
- Improved patient/family engagement
<table>
<thead>
<tr>
<th>Condition</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital Diaphragmatic Hernia</td>
<td>1 in 3000</td>
</tr>
<tr>
<td>Oesophageal Atresia</td>
<td>1 in 4000</td>
</tr>
<tr>
<td>Exomphalos</td>
<td>1 in 2500 - 12000*</td>
</tr>
<tr>
<td>Gastroschisis</td>
<td>1 in 2500</td>
</tr>
<tr>
<td>Duodenal Atresia</td>
<td>1 in 5000</td>
</tr>
</tbody>
</table>
Steering Group
National Audit of Early Outcome of Surgical Congenital Anomalies
Methodology

Five year retrospective audit (2013-2017 inclusive) of liveborn neonates managed in Scotland

Case identification and data collection from Badger.net, discharge coding and Clinical Portal/Notes*
Glasgow – Hannah Hood, Judith Simpson, Gregor Walker
Edinburgh – Lisa Steven, Shona Cowan, Fraser Munro
Aberdeen – Yatin Patel

Cauldicott Guardian approval obtained
Diagnoses by year (n=356)

OA (83)
CDH (85)
DA (51)
Gastroschisis (100)
Exomphalos (37)

Year 2013: 72 cases
Year 2014: 71 cases
Year 2015: 78 cases
Year 2016: 74 cases
Year 2017: 61 cases
When diagnosed
## Other associated anomalies

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>N</th>
<th>Associated anomalies (%)</th>
<th>Co-morbidities (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Genetic</td>
</tr>
<tr>
<td>CDH</td>
<td>85</td>
<td>14%</td>
<td>4%</td>
</tr>
<tr>
<td>Gastroschisis</td>
<td>100</td>
<td>4%</td>
<td>0</td>
</tr>
<tr>
<td>OA / TOF</td>
<td>81</td>
<td>73%</td>
<td>7%</td>
</tr>
<tr>
<td>DA</td>
<td>51</td>
<td>59%</td>
<td>22%</td>
</tr>
<tr>
<td>Exomphalos</td>
<td>37</td>
<td>57%</td>
<td>22%</td>
</tr>
</tbody>
</table>
Survival

OA: 93%
CDH: 73%
DA: 96%
Gastro: 93%
Exomph: 81%

Legend:
- Late Death
- Died on Unit
- Survived
Average* stay in surgical centre

* Median inpatient stay in days for surviving infants

- CDH: 26 days
- OA: 23 days
- DA: 26 days
- Gastrochisis: 35 days
- Exomphalos major: 36 days
- Exomphalos Minor: 13.5 days
Cumulative discharge destination

- Home: 224
- Repatriated: 69

Total: 293
Gastroschisis

- Incidence increasing, reasons unclear
  - 1 in 2,500 live births
- Failure of normal embryological anterior abdominal wall closure
- Residual small (~3cm) defect to the right of umbilicus
Aetiology & diagnosis

- Aetiology unknown
- Well described associations
  - Low socioeconomic status
  - Young maternal age
  - White race
  - Low BMI
  - Smoking / recreational drug use

- Often antenatal diagnosis (>11 weeks gestation)
  - SCANS cohort 100%
Associated anomalies & monitoring

Majority “isolated” defect (96% SCANS)

Bowel complications in ~10% (simple versus complex)
  Atresia (2% SCANS)
  Ischaemia / necrosis (5% SCANS)
  Vanishing (3% SCANS)

Regular in utero monitoring
  Growth - IUGR
  Changes in fetal well being
  Intestinal dilatation
Gestational age & mode of delivery

Median birth weight = 2.44kg (range 1.1 – 3.6kg)

- Elective CS
- Emergency CS
- Vaginal/Assisted

34%
Delivery management

- Cover/Protect intestine
- Occlusive wrapping
- Place baby on right side
  - Place large bore NGT
  - IV access/fluids/antibiotics
  - Wait for the surgeons...
Surgical management

• **Staged (silo) closure**
  – In NNU / theatre
  – Suture / sutureless

• **Primary closure**
  – In NNU / in theatre
  – Suture / sutureless

• **Laparotomy**
  – If bowel complications
Post closure management

• Ventilatory support as required
• Central venous access & parenteral nutrition
  – Median duration of PN for simple 22 days (range 6-83) versus 44 days (5-119) for complex.
• Enteral feeds as tolerated with slow increases
  – Oral colostrum if available
  – Trophic fresh maternal milk/donor milk if unavailable
  – Time to full enteral feeds 23.5 days (range 9-68) for simple versus 87 (52-135) for complex.
Post delivery timings in Scotland

- Closure: Median – day 3
- Ventilation: Mean – 3 days (34 NV)
- Full Feeds: Median – 24 days*
- Hospital Stay: Median – 35 days*

Median – 35 days*
Mean – 3 days (34 NV)
Discharge destination

- Home
- Repatriated
- Transferred to ward
- Died

12%
Outcome

• Neonatal death
  – SCANS 7%
  – Intestinal ischaemia (early), NEC (later)

2 Year Follow-Up Data Collection

• Intestinal failure and long term PN (IFALD)
  – 5-10%; complex >> simple
• Adhesive bowel obstruction / segmental volvulus
• Undescended testicles
Obstruction in duodenum
Incidence ~1 in 5000 births
~10 births/year in Scotland
Important Associations
- Down Syndrome in 30%
- Malrotation in 30%
~50% diagnosed antenatally

- Double bubble
- Amniocentesis
- Look for cardiac anomaly
- Plan for delivery (prem)
Making the diagnosis

Bile vomiting/aspirates
Characteristic x-ray
Look for associations
- Down syndrome
- Cardiac anomalies
- Oesophageal atresia
- Anorectal anomalies
You don’t need bile...
If there is gas beyond...
If there is gas beyond...
Surgery

Duodenoduodenostomy
Diamond configuration
Can be done laparoscopically (in Edinburgh)
Post-operative Management

• Issues of Prematurity
• Associated anomalies
• Nutrition
  – Dilated duodenum
  – Slow to empty
  – Parenteral nutrition
  – Variable duration
Previous Scottish Audit (1997-2011)

97 patients  TAT used in 68 (73%)

Use of TAT tube associated with longer inpatient stay (p=0.03)
- **27 days** (95% CI 30.3-52) vs **20.5 days** (95% CI 17.2-27.5)
SCANS Inpatient Data

- Surgery: Median – day 3
- Ventilation: Median – 3 days (17 NV)
- Full Feeds: Mean –19 days*
- Hospital Stay: Mean – 26 days*
Discharge destination

- Home
- Repatriate
- Ward
- Died

22%
Outcome

- Most have no issues at all...
  - Dilated, dysmotile duodenum
  - Anastomotic problems
  - Gastro-oesophageal reflux
  - Adhesional problems
- Outcome usually relates to associations

2 Year Follow-Up Data Collection
Why would we ask for your help?

- Work together on repatriation pathways
- Help identify affected pregnancy/baby/family

<table>
<thead>
<tr>
<th>Condition</th>
<th>SCANS Audit</th>
<th>ISD</th>
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</thead>
<tbody>
<tr>
<td>CDH</td>
<td>85</td>
<td>79 (93%)</td>
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<tr>
<td>Gastroschisis</td>
<td>100</td>
<td>92 (92%)</td>
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<tr>
<td>Exomphalos</td>
<td>37</td>
<td>34 (92%)</td>
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<tr>
<td>OA/TOF</td>
<td>83</td>
<td>79 (95%)</td>
</tr>
<tr>
<td>DA</td>
<td>51</td>
<td>42 (82%)</td>
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- We think we have a solution in Badger Alert...
<table>
<thead>
<tr>
<th>Health Board</th>
<th>Maternity</th>
<th>Neonatal</th>
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</thead>
<tbody>
<tr>
<td>Ayrshire &amp; Arran</td>
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<td>Borders</td>
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<td>Dumfries &amp; Galloway</td>
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<td>Fife</td>
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