

Rare Neonatal Gastrointestinal Conditions

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Plan/Disclaimer



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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

Population Density

England

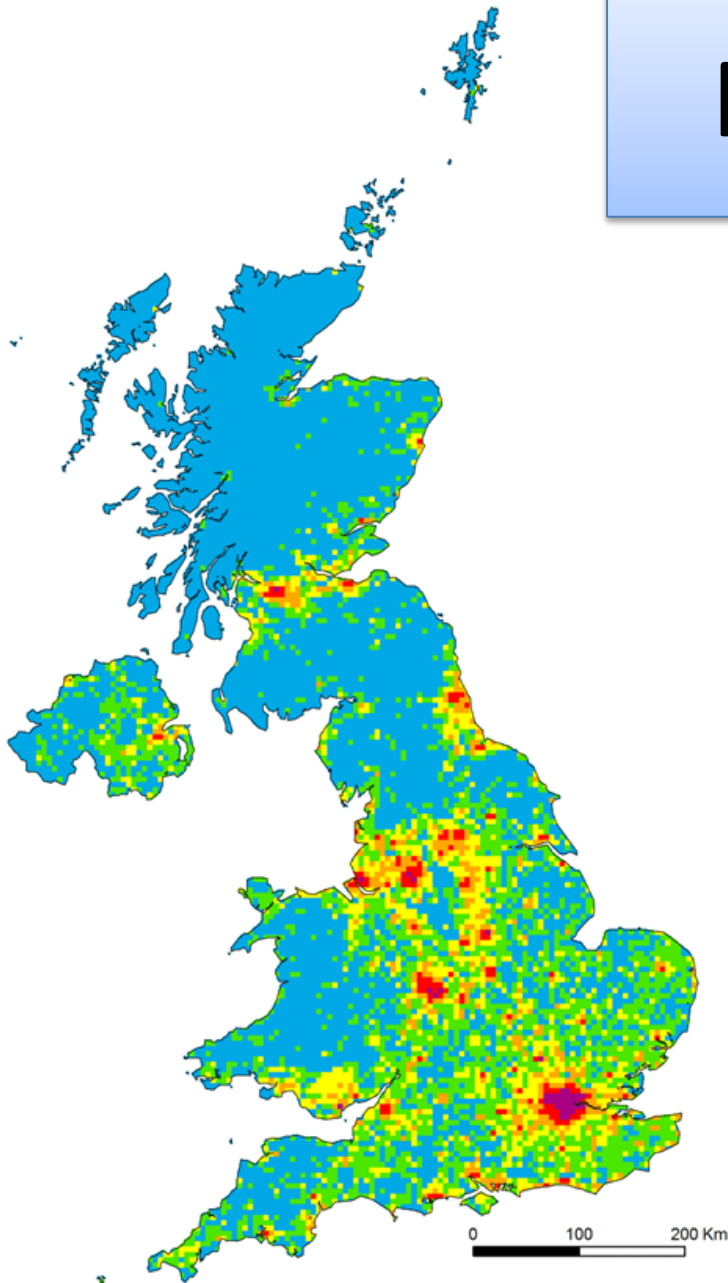
Population 55,977,178

Mean density 407 people/km²

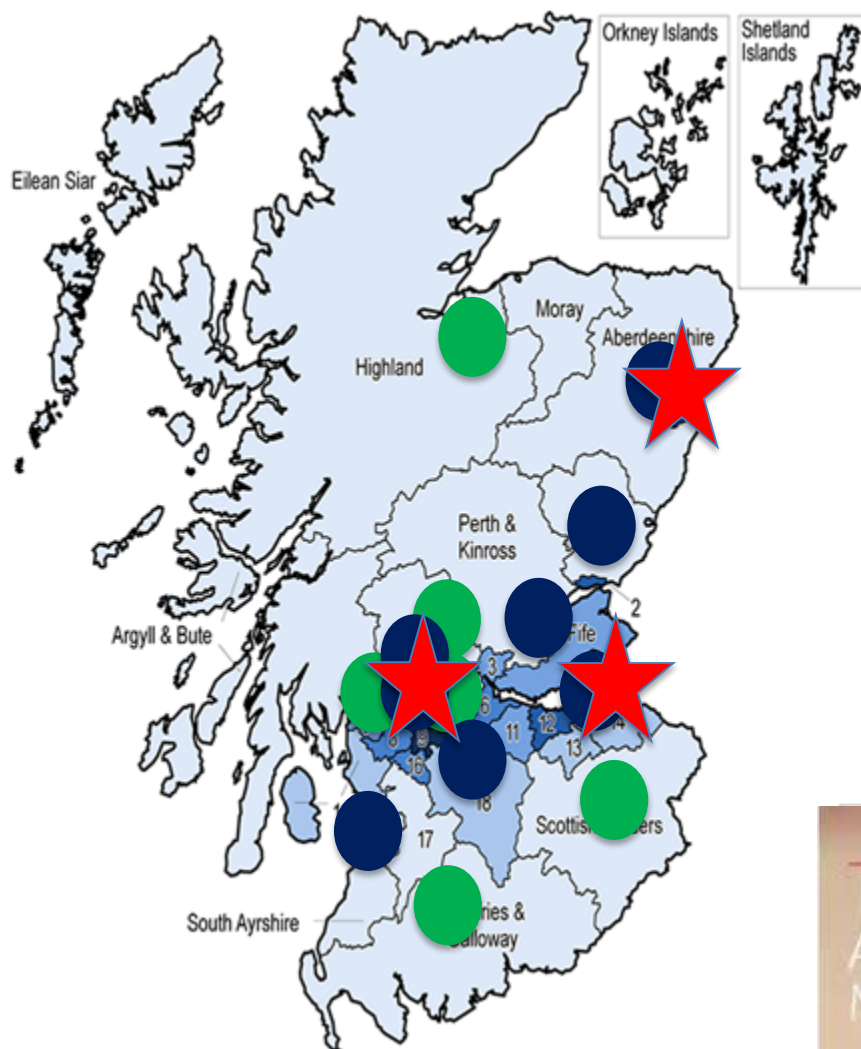
Scotland

Population 5,438,100

Mean density 68 people/km²



Neonatalology/Neonatal Surgery



Level 2 NICU

● 6 centres

Level 3 NICU

● 8 centres

Neonatal surgery

★ 3 Centres

THE BEST START

A Five-Year Forward Plan for
Maternity and Neonatal Care in Scotland



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

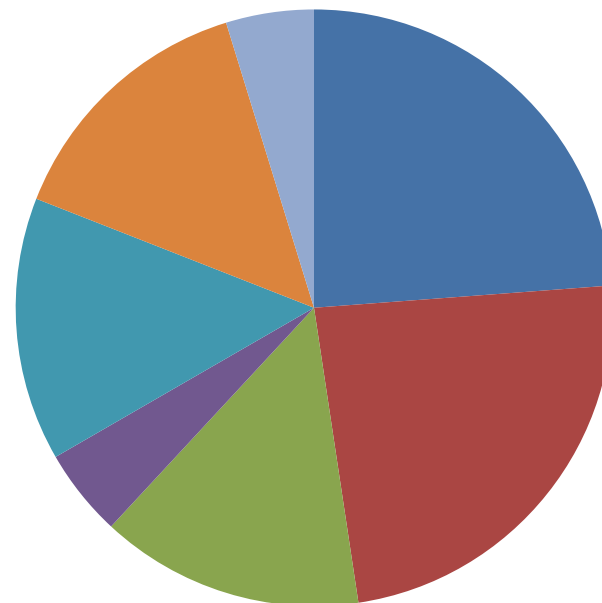
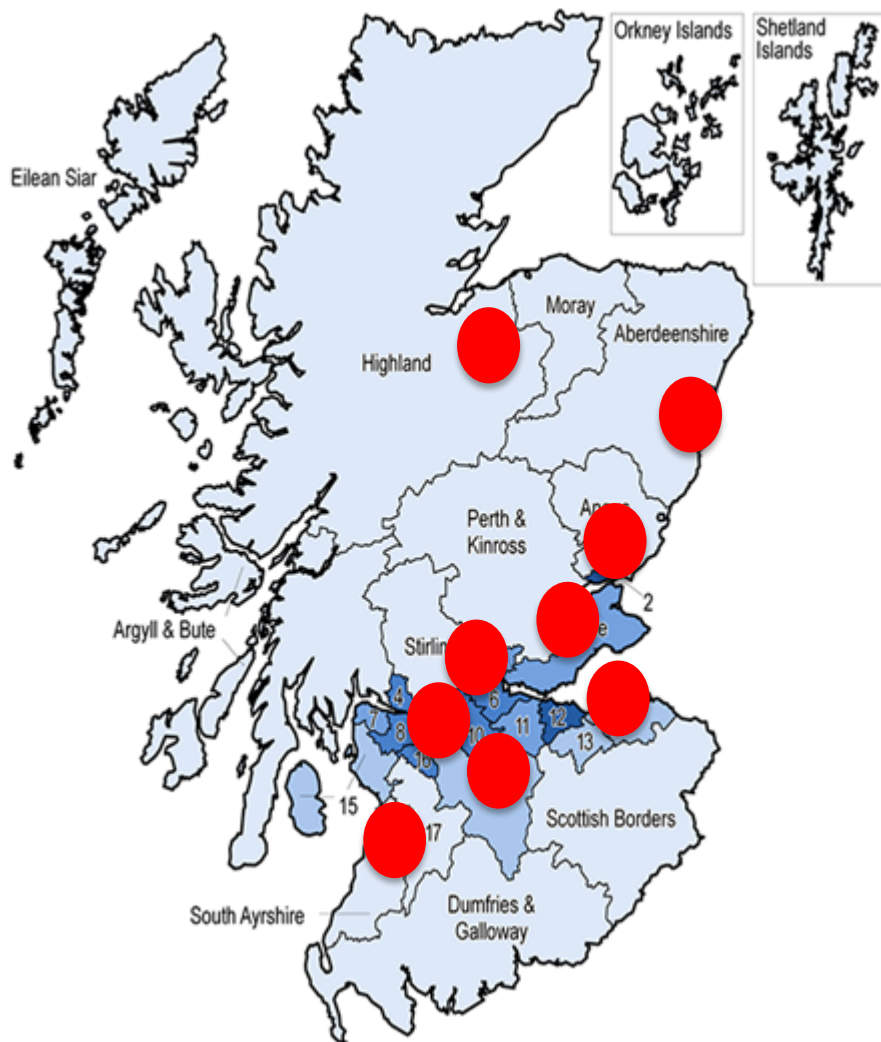


Conditions



Congenital Diaphragmatic Hernia	1 in 3000
Oesophageal Atresia	1 in 4000
Exomphalos	1 in 2500 - 12000*
Gastroschisis	1 in 2500
Duodenal Atresia	1 in 5000

Steering Group



- Surgeon
- FM/Obs
- Neonatology
- Paeds
- Nurse
- AHP
- Parent



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*

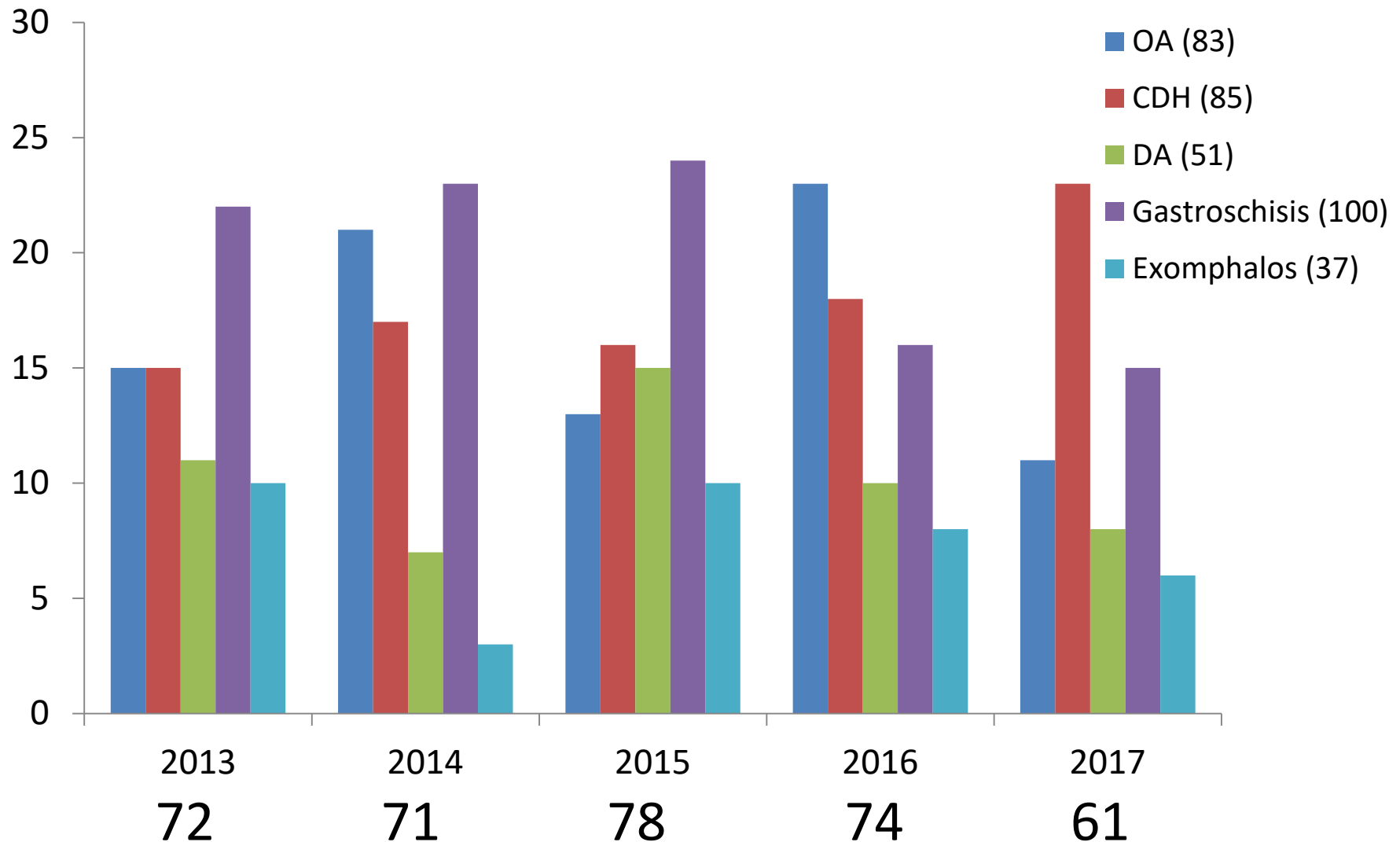
Glasgow – Hannah Hood, Judith Simpson, Gregor Walker

Edinburgh – Lisa Steven, Shona Cowan, Fraser Munro

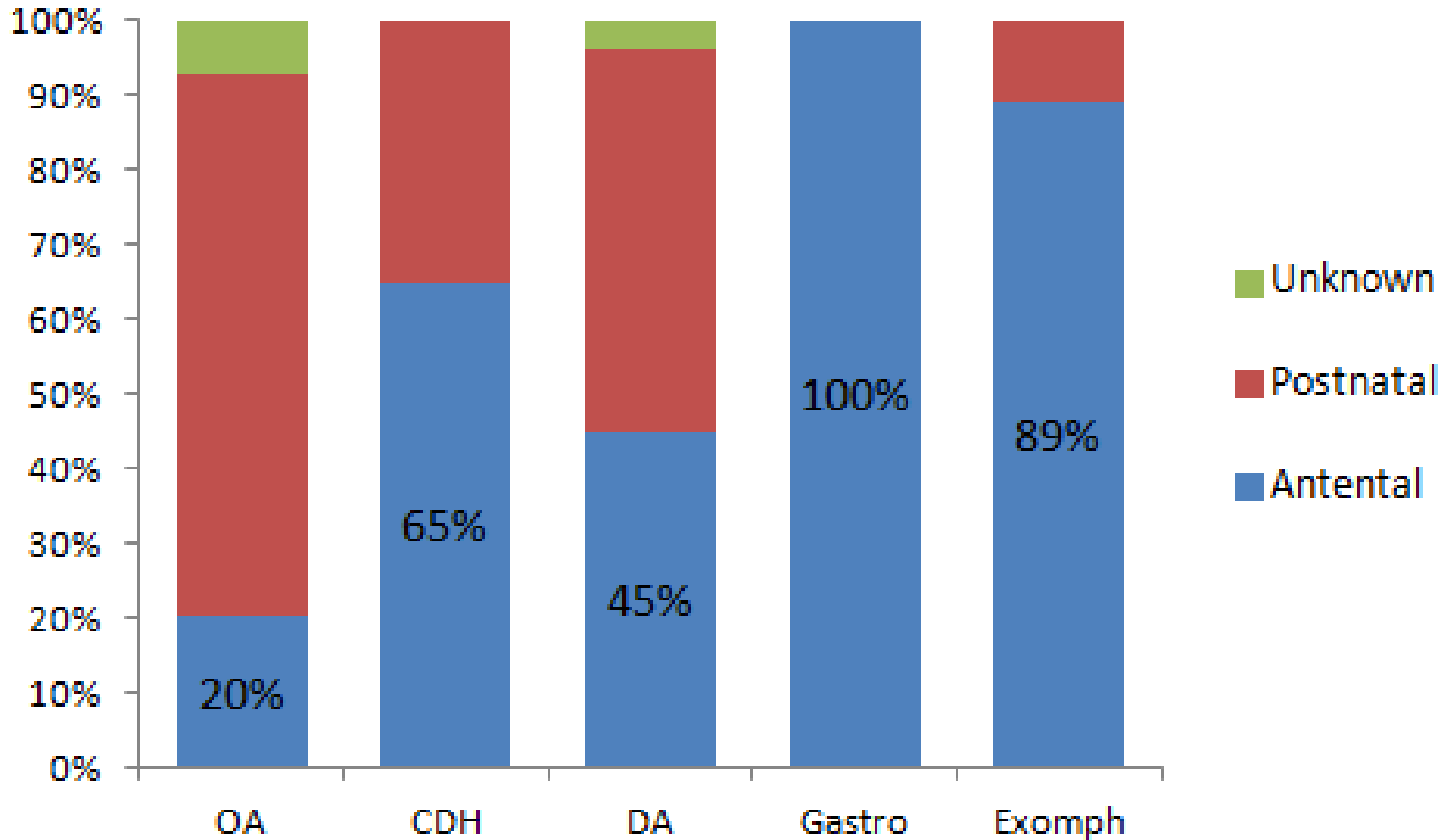
Aberdeen – Yatin Patel

Cauldicott Guardian approval obtained

Diagnoses by year (n=356)



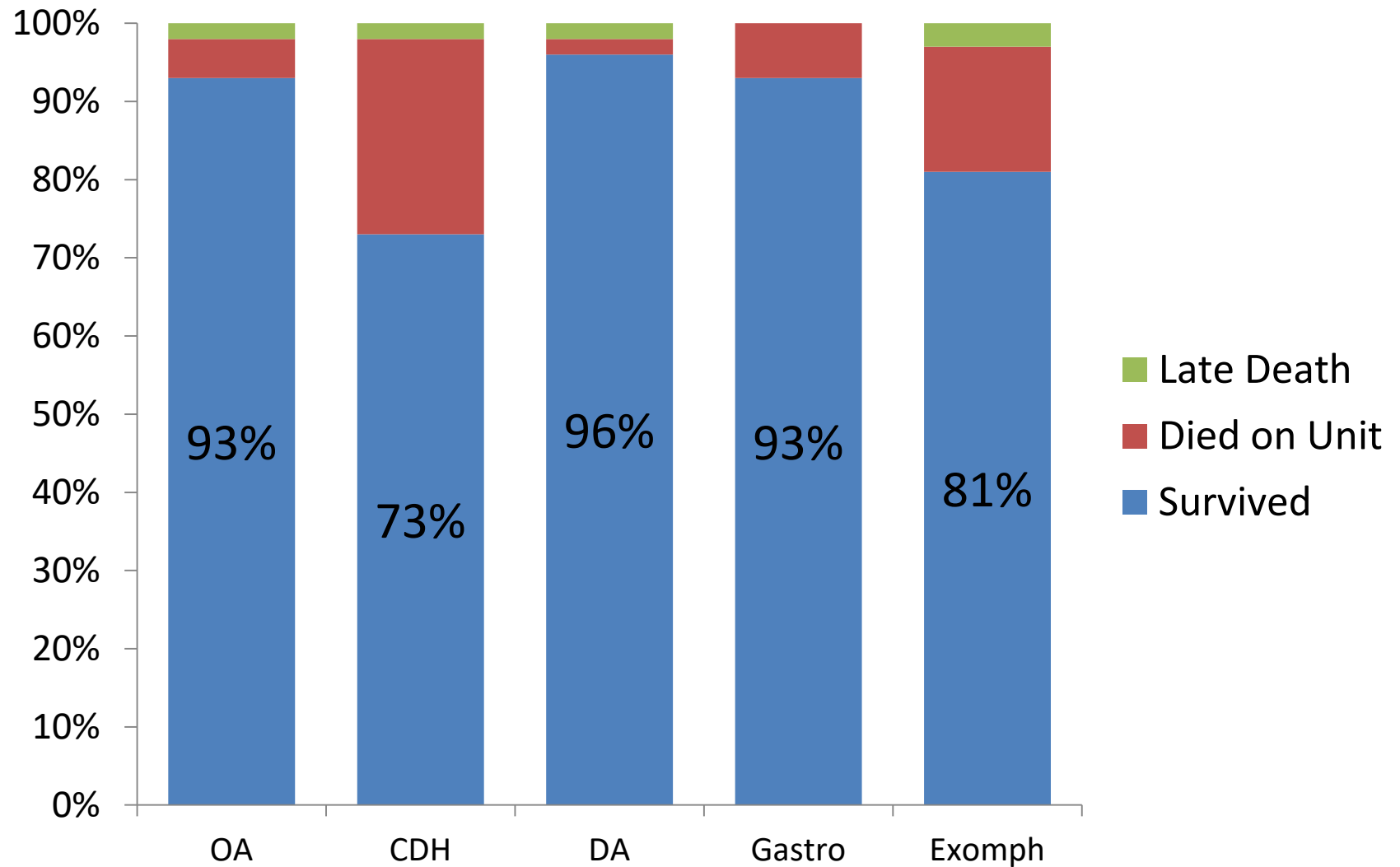
When diagnosed



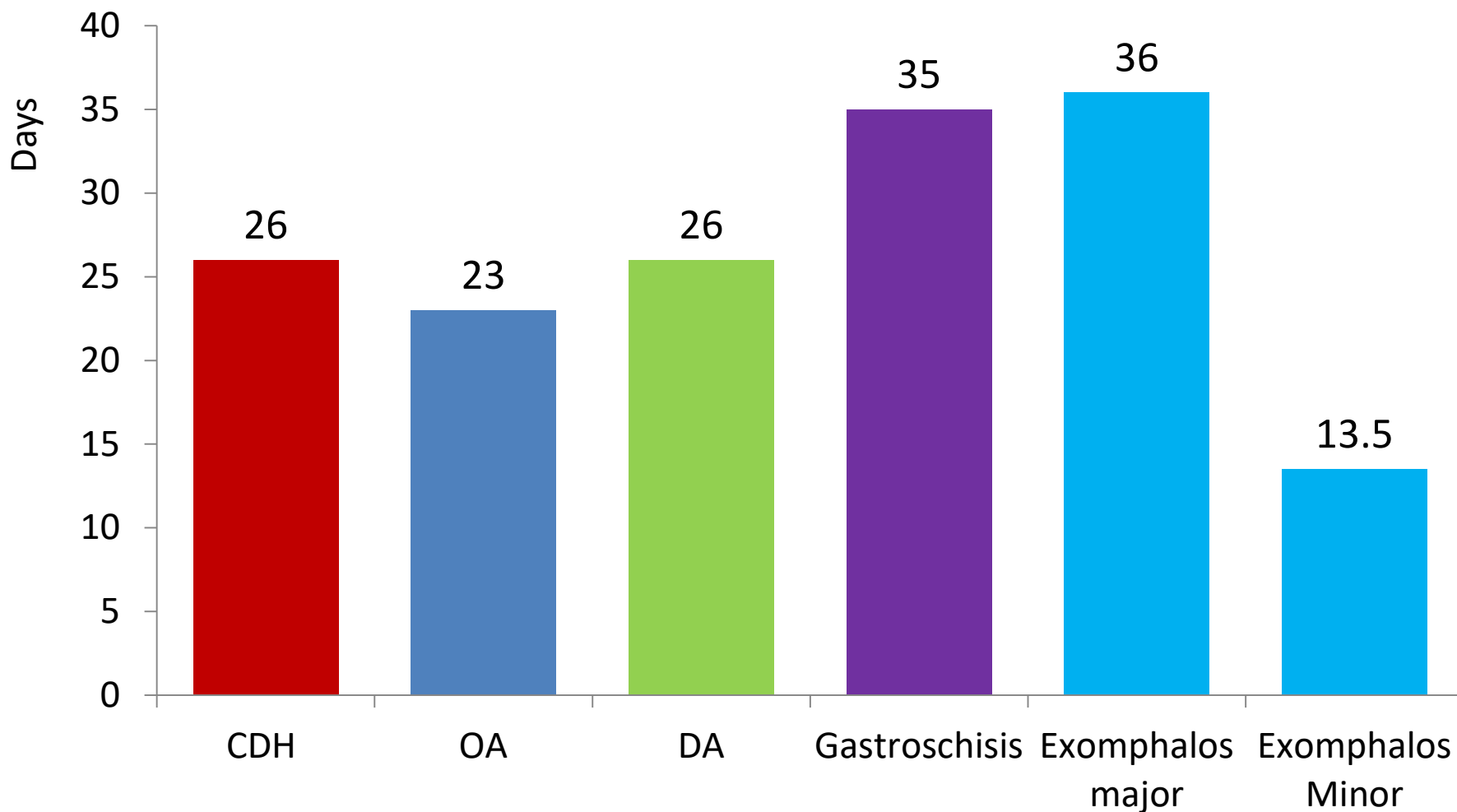
Other associated anomalies

Diagnosis	N	Associated anomalies (%)	Co-morbidities (%)	
			Genetic	Structural
CDH	85	14%	4%	11%
Gastroschisis	100	4%	0	4%
OA / TOF	81	73%	7%	72%
DA	51	59%	22%	57%
Exomphalos	37	57%	22%	40%

Survival

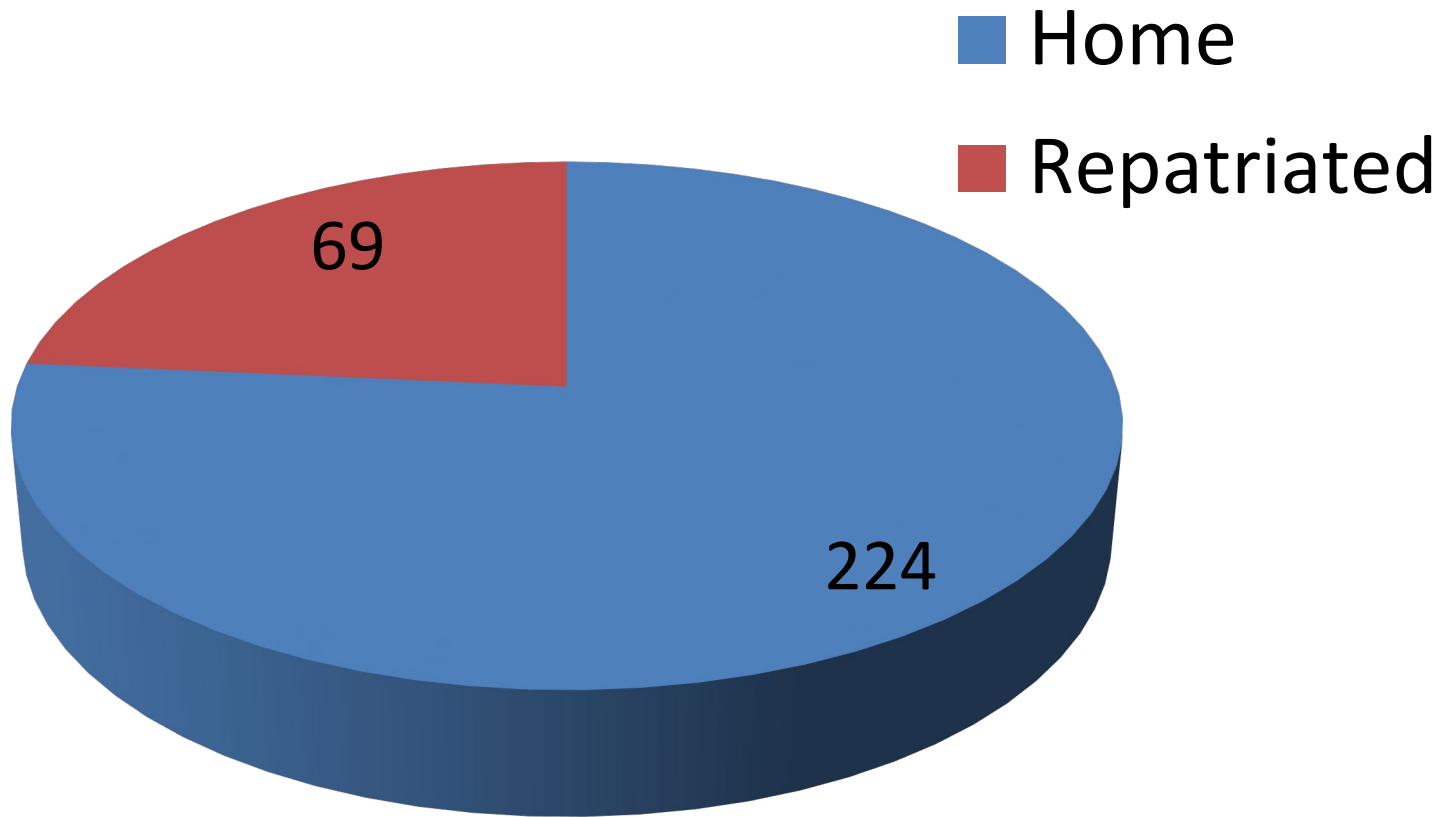


Average* stay in surgical centre



* Median inpatient stay in days for surviving infants

Cumulative discharge destination



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

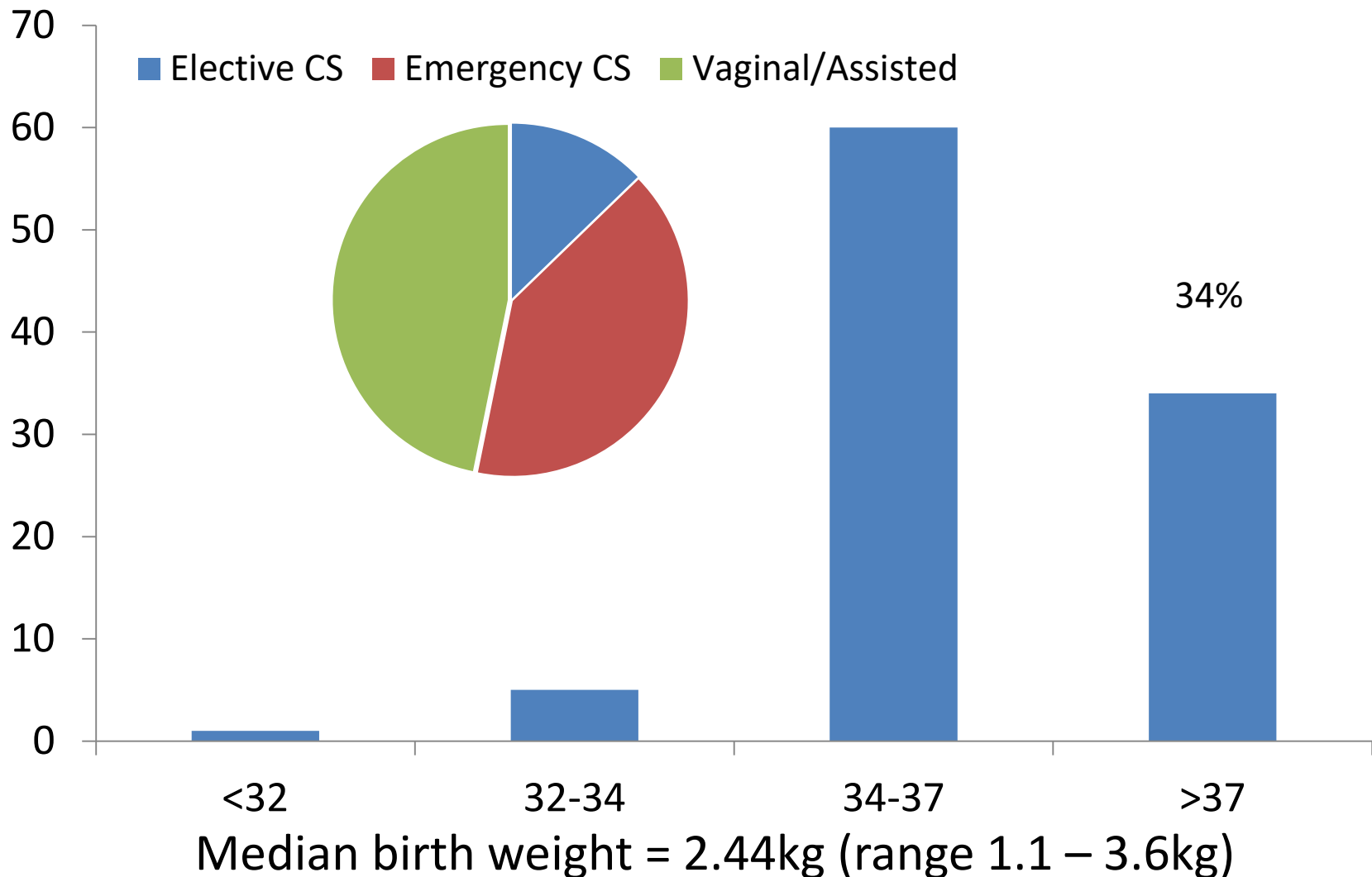
- Growth - IUGR

- Changes in fetal well being

- Intestinal dilatation



Gestational age & mode of delivery



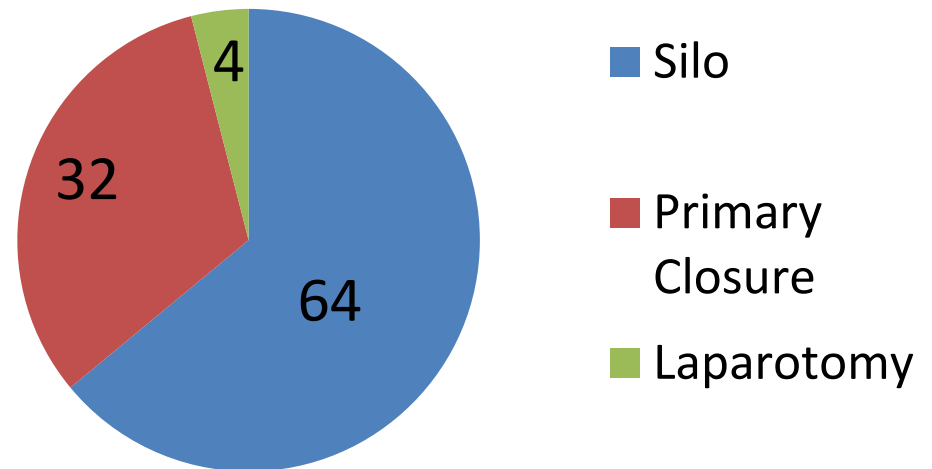
Delivery management

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



Surgical management

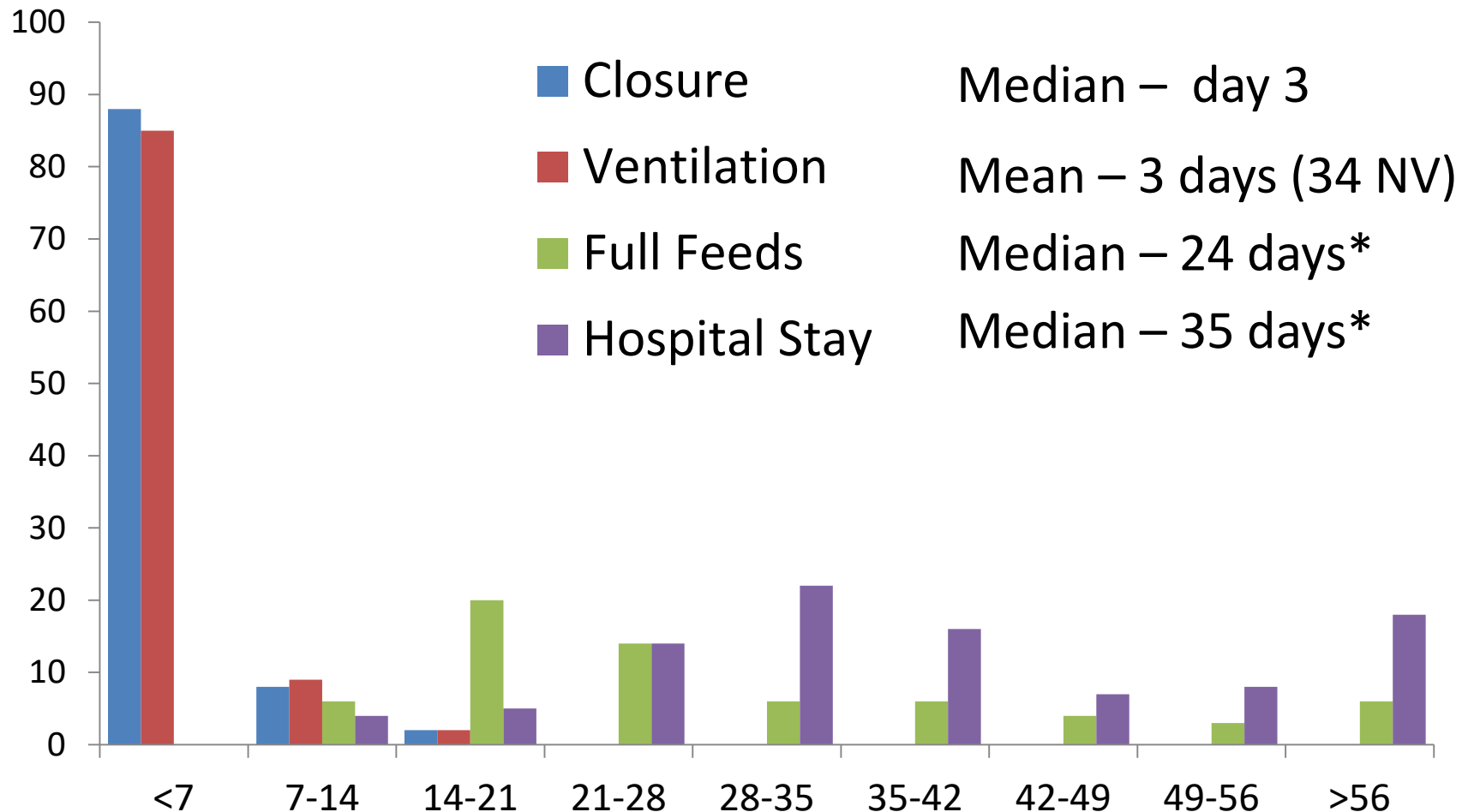
- **Staged (silo) closure**
 - In NNU / theatre
 - Suture / sutureless
- **Primary closure**
 - In NNU / in theatre
 - Suture / sutureless
- **Laparotomy**
 - If bowel complications



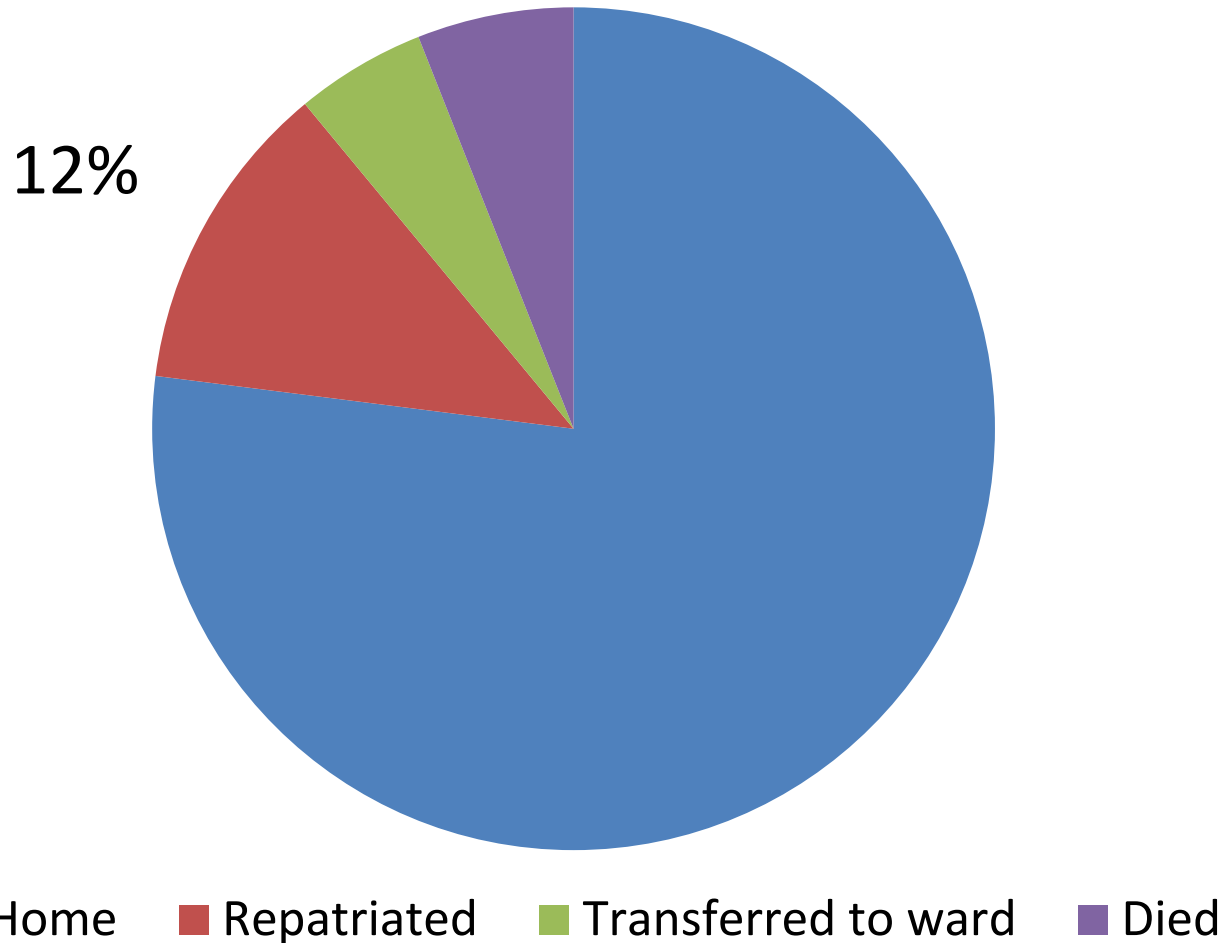
Post closure management

- 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



Discharge destination



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

Duodenal Atresia

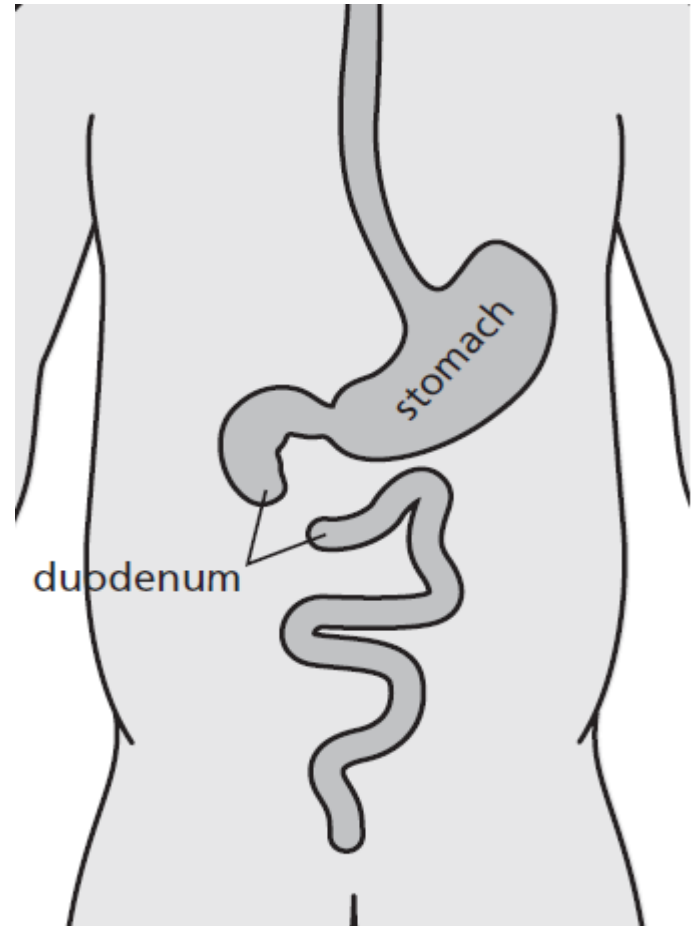
Obstruction in duodenum

Incidence ~1 in 5000 births

~10 births/year in Scotland

Important Associations

- Down Syndrome in 30%
- Malrotation in 30%

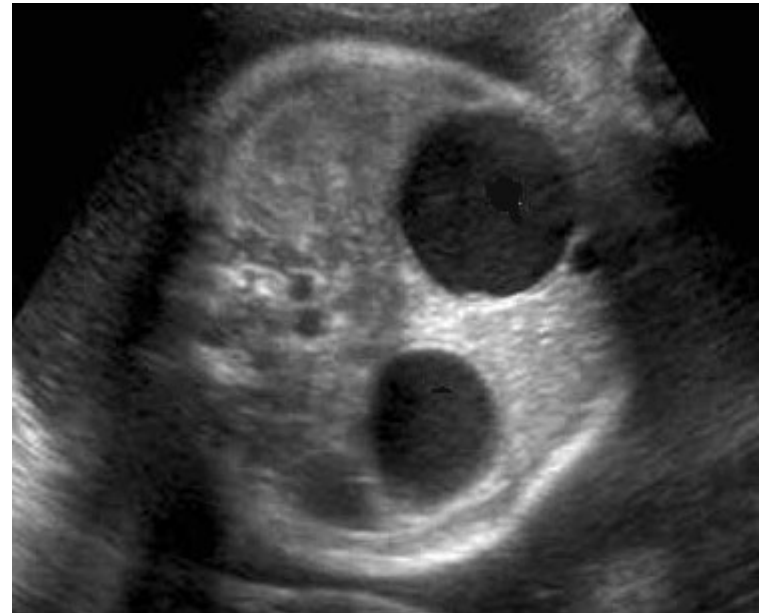


Antenatal Factors

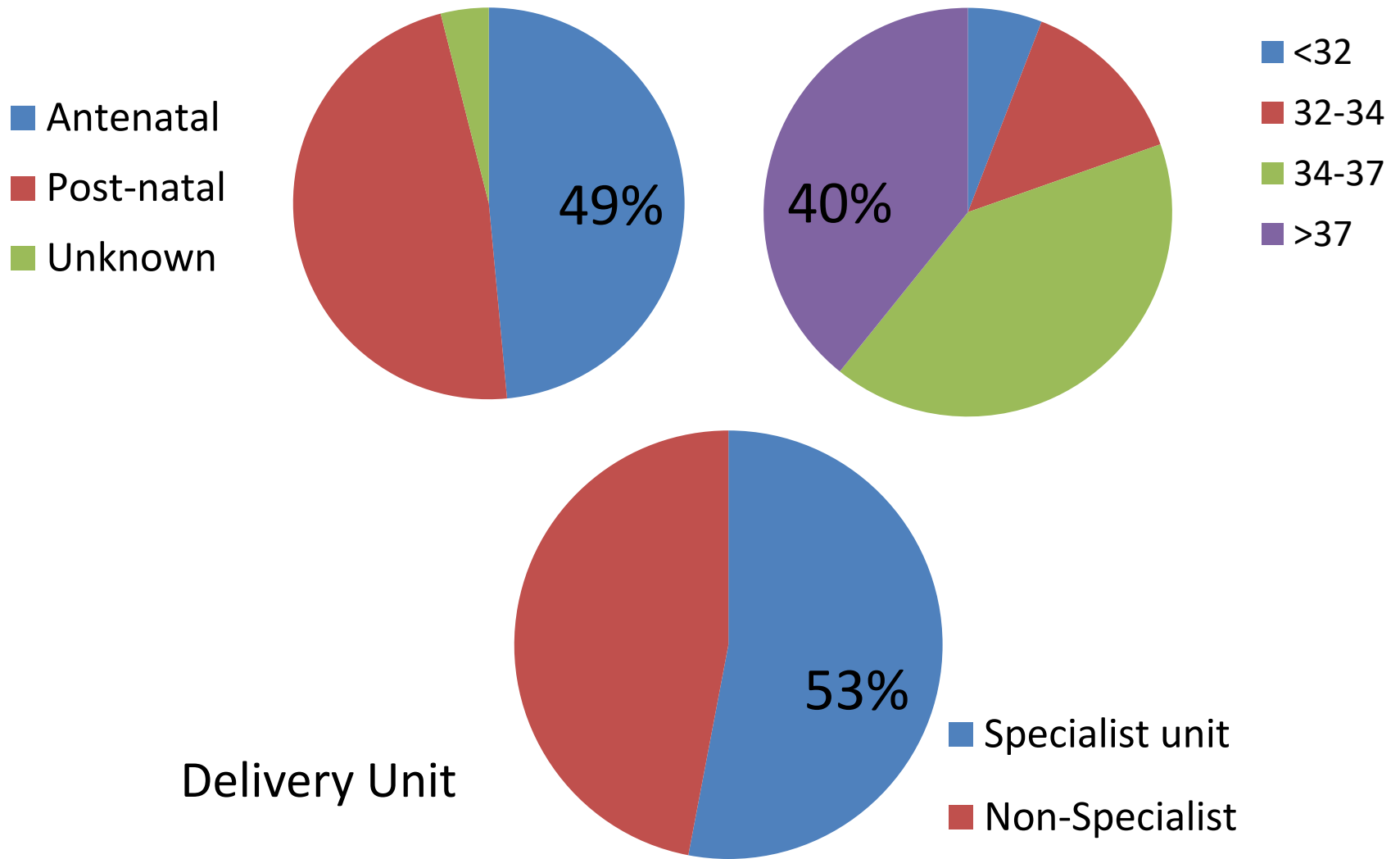
~50% diagnosed antenatally

Double bubble

- Amniocentesis
- Look for cardiac anomaly
- Plan for delivery (prem)



Diagnosis and Delivery



Making the diagnosis

Bile vomiting/aspirates

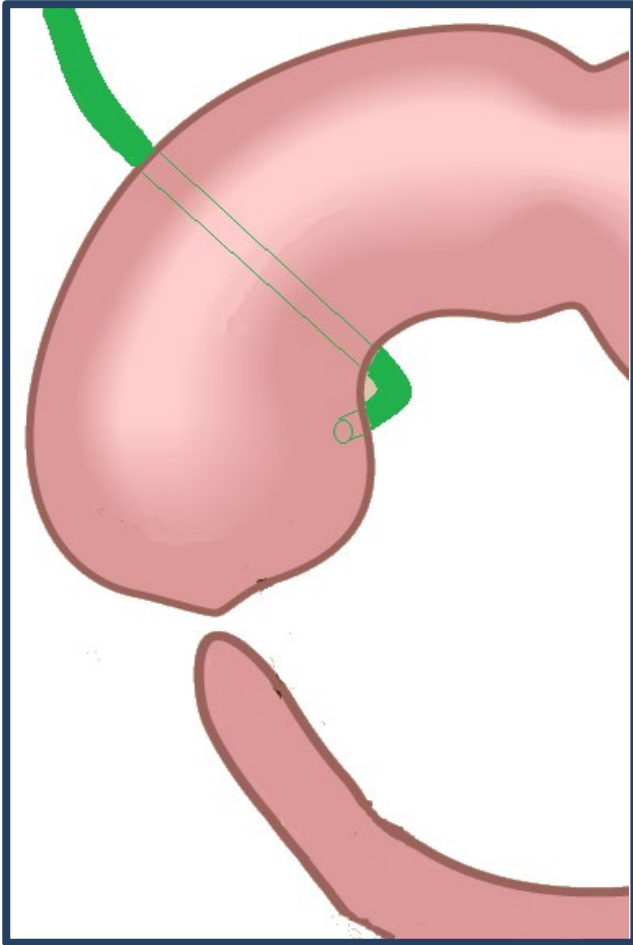
Characteristic x-ray

Look for associations

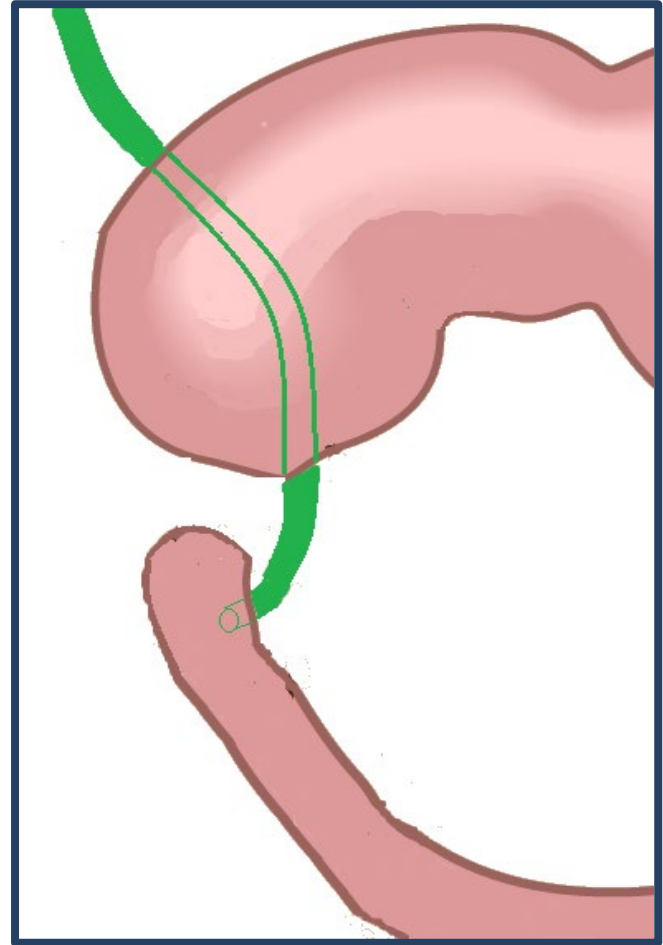
- Down syndrome
- Cardiac anomalies
- Oesophageal atresia
- Anorectal anomalies



You don't need bile...



80%

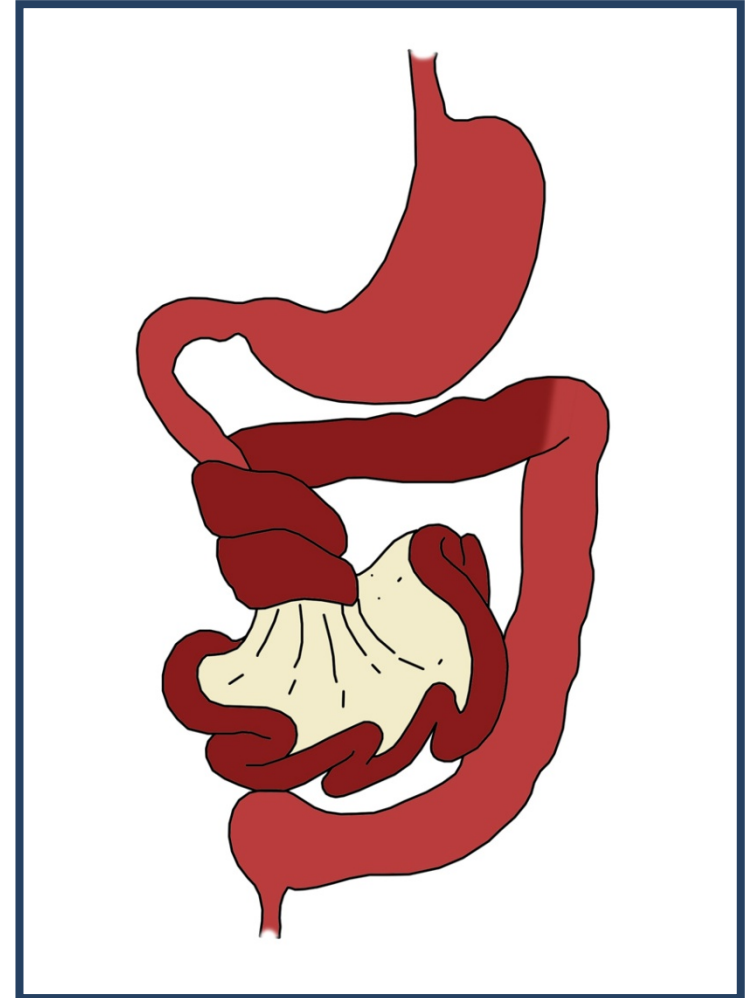
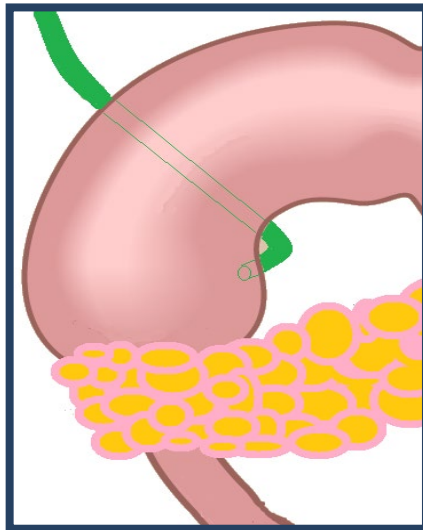
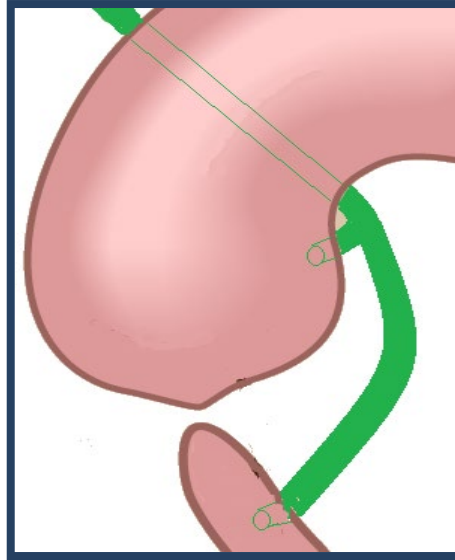
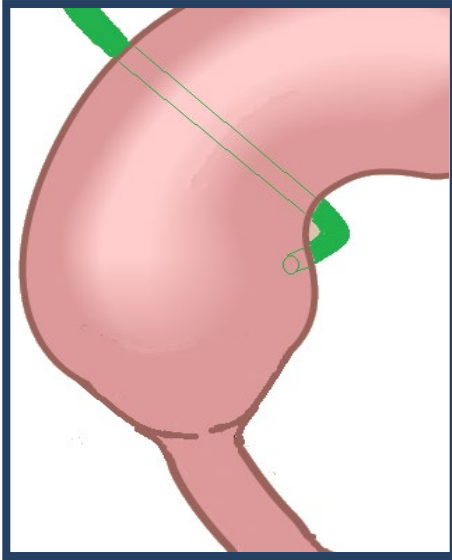


20%

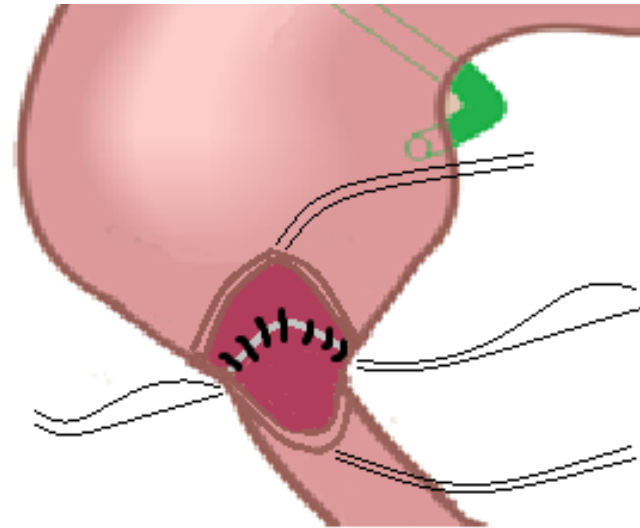
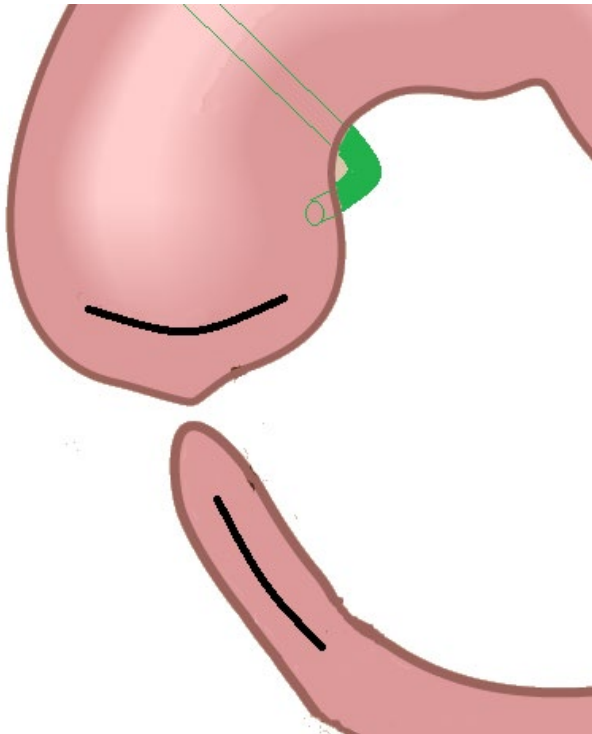
If there is gas beyond...



If there is gas beyond...



Surgery



Duodenoduodenostomy
Diamond configuration

Can be done laparoscopically (in Edinburgh)

Post-operative Management

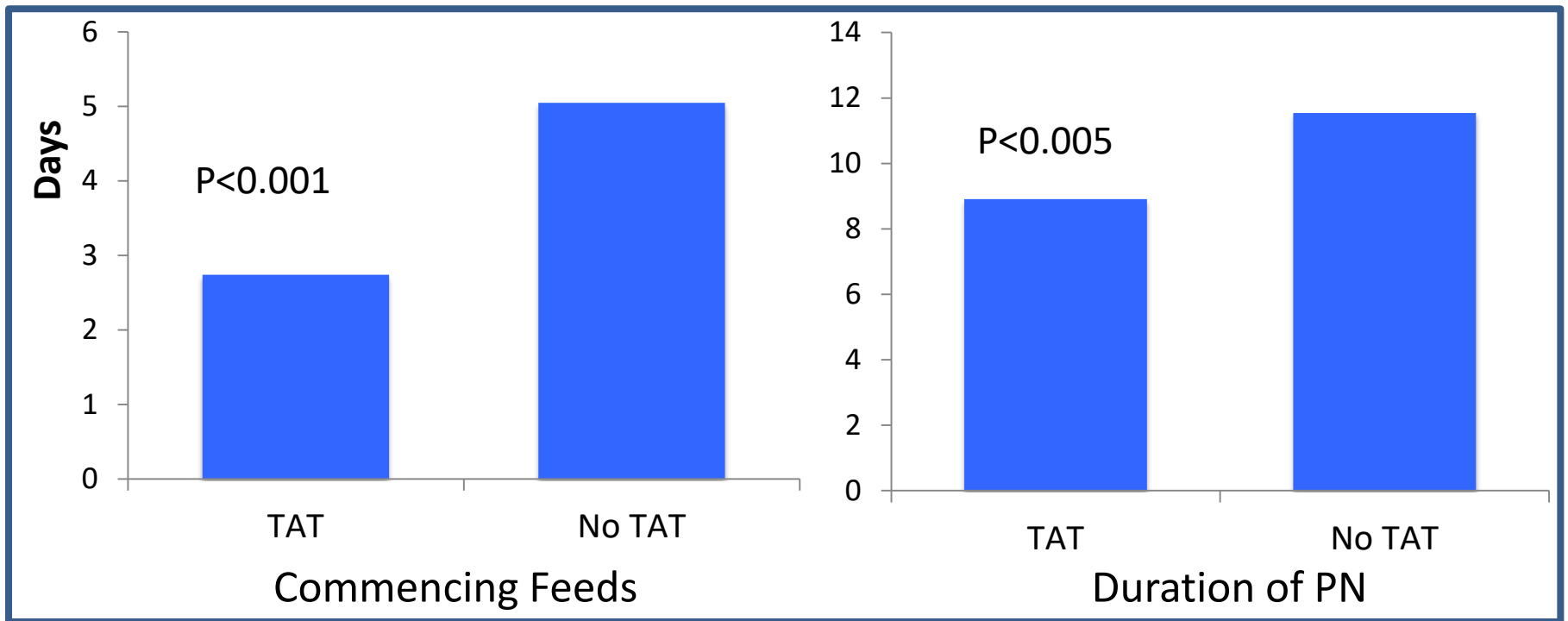
- Issues of Prematurity
- Associated anomalies
- Nutrition
 - Dilated duodenum
 - Slow to empty
 - Parenteral nutrition
 - Variable duration



Previous Scottish Audit (1997-2011)

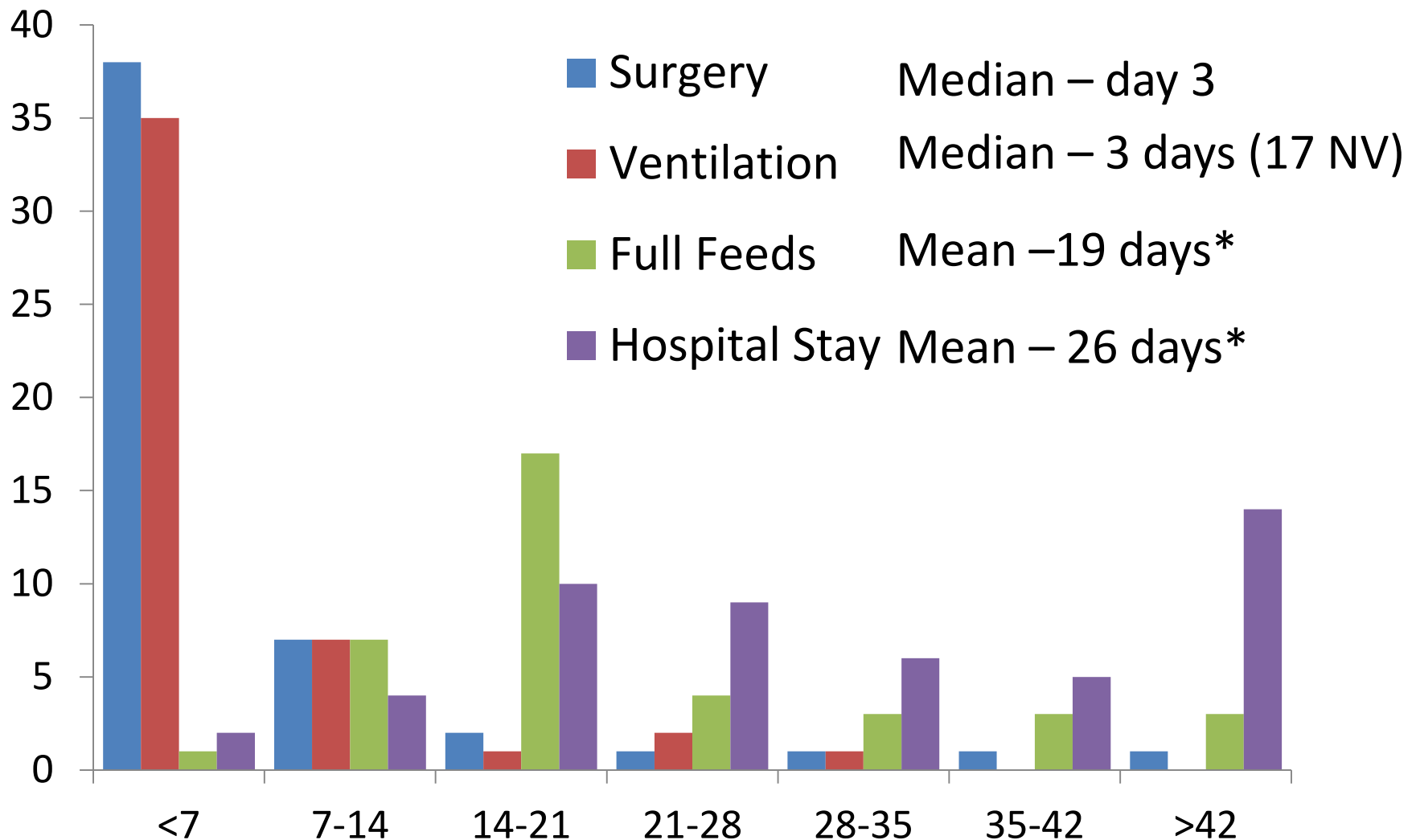
97 patients

TAT used in 68 (73%)

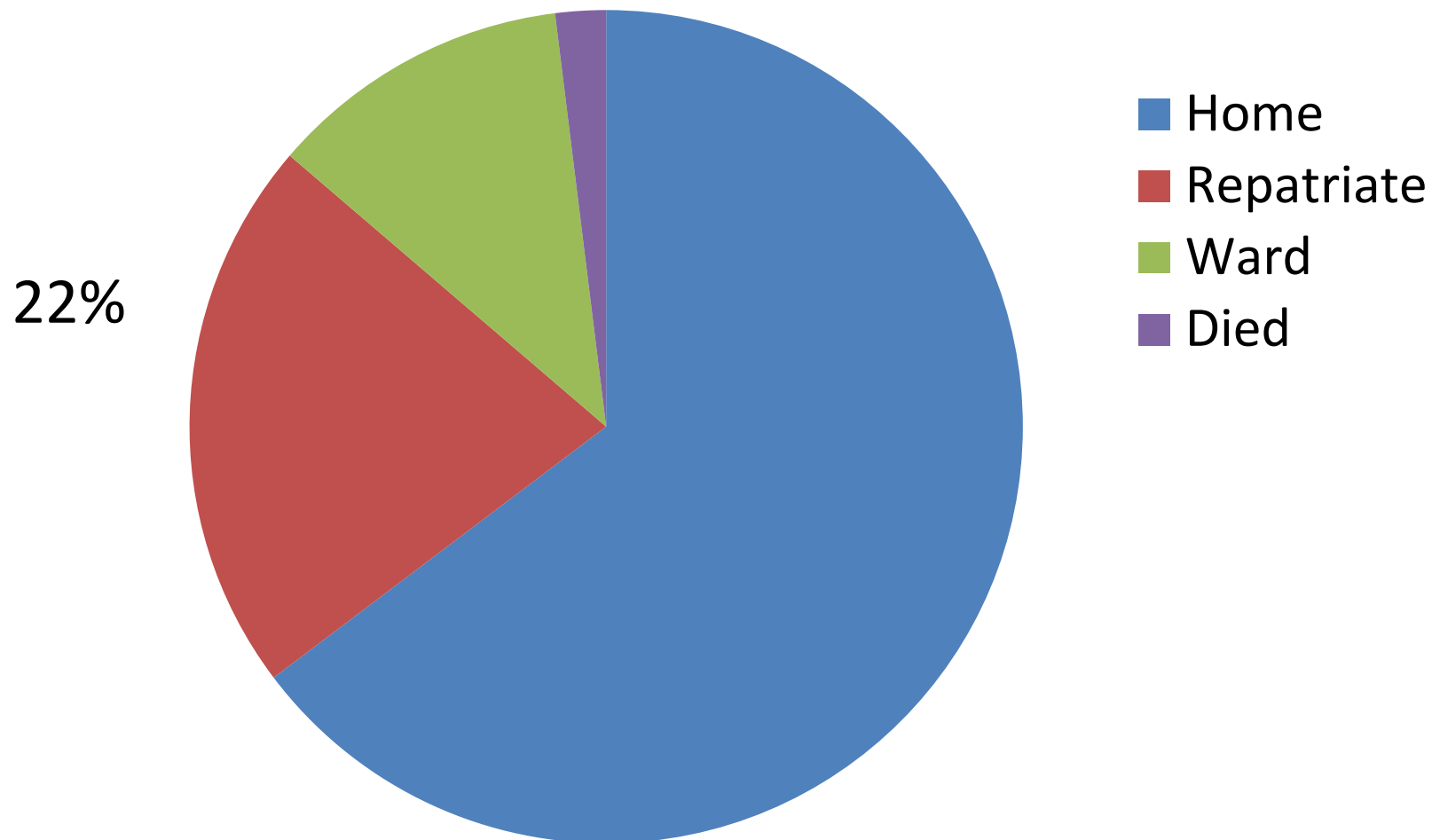


Use of TAT tube associated with longer inpatient stay ($p=0.03$)
-27 days (95% CI 30.3-52) vs **20.5 days** (95%CI 17.2-27.5)

SCANS Inpatient Data



Discharge destination



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

	SCANS Audit	ISD
CDH	85	79 (93%)
Gastroschisis	100	92 (92%)
Exomphalos	37	34 (92%)
OA/TOF	83	79 (95%)
DA	51	42 (82%)

- We think we have a solution in Badger Alert...

Health Board	Maternity	Neonatal
Ayrshire & Arran		
Borders	Yes	
Dumfries & Galloway		
Fife	Yes	Yes
Forth Valley	Yes	
GGC	Yes	Yes
Grampian		Yes
Highland		
Lanarkshire	Yes	
Lothian		
Orkney		Yes
Shetland		Yes
Tayside		
Western Isles	Yes	



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