

# 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

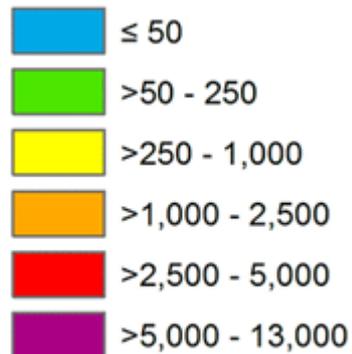
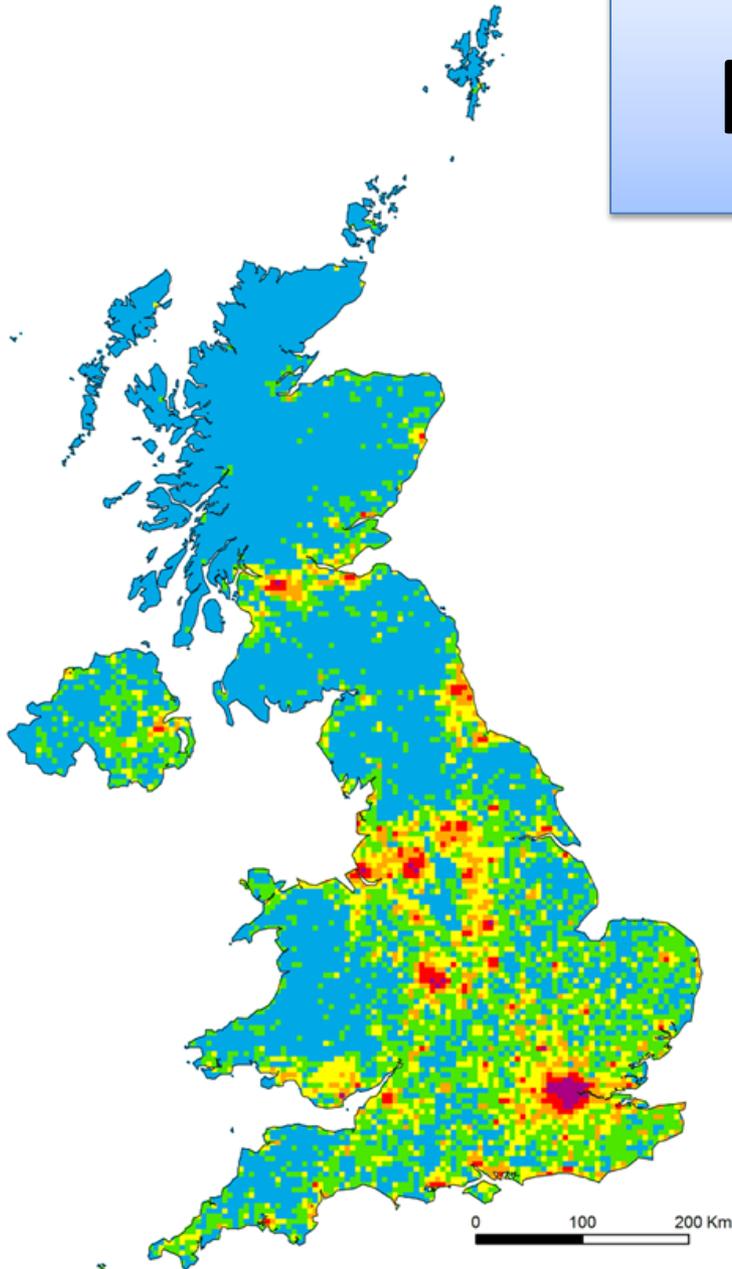
Population 55,977,178

Mean density 407 people/km<sup>2</sup>

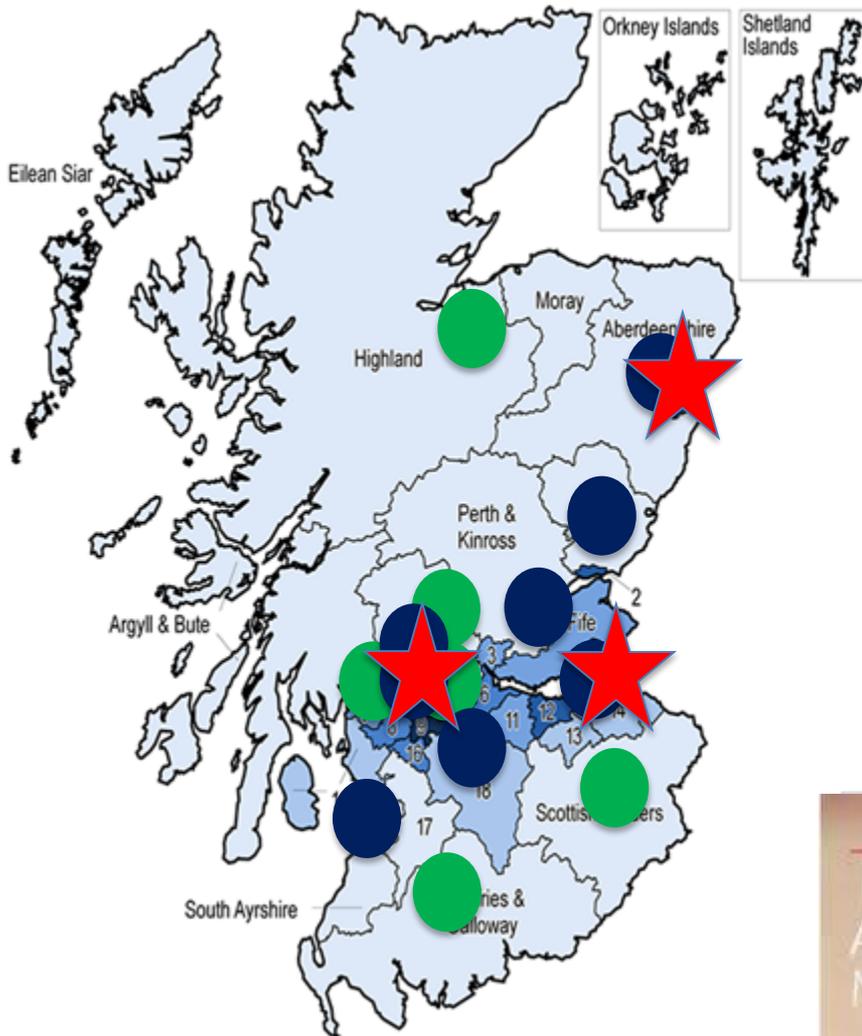
## Scotland

Population 5,438,100

Mean density 68 people/km<sup>2</sup>



# 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

Congenital Diaphragmatic Hernia

1 in 3000

Oesophageal Atresia

1 in 4000

Exomphalos

1 in 2500 -  
12000\*

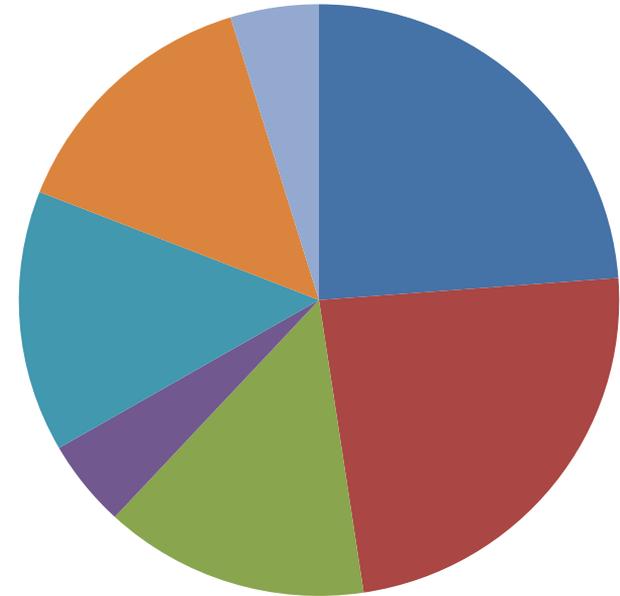
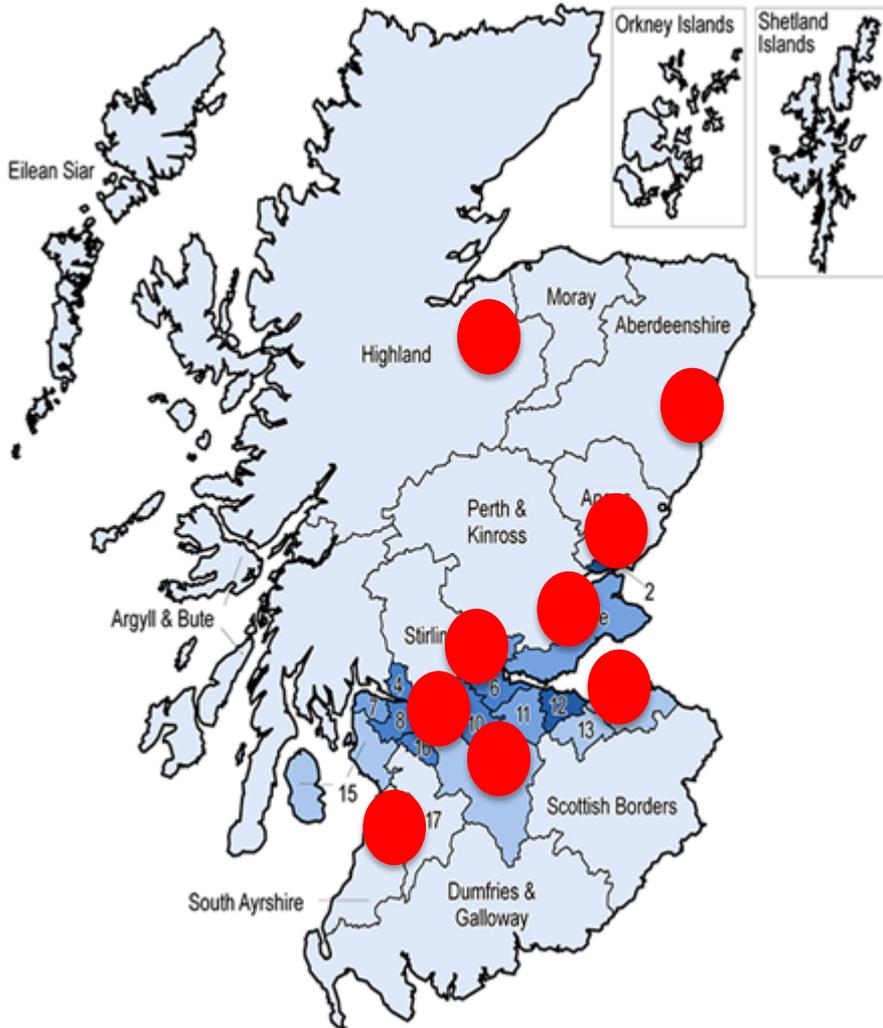
Gastroschisis

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\*

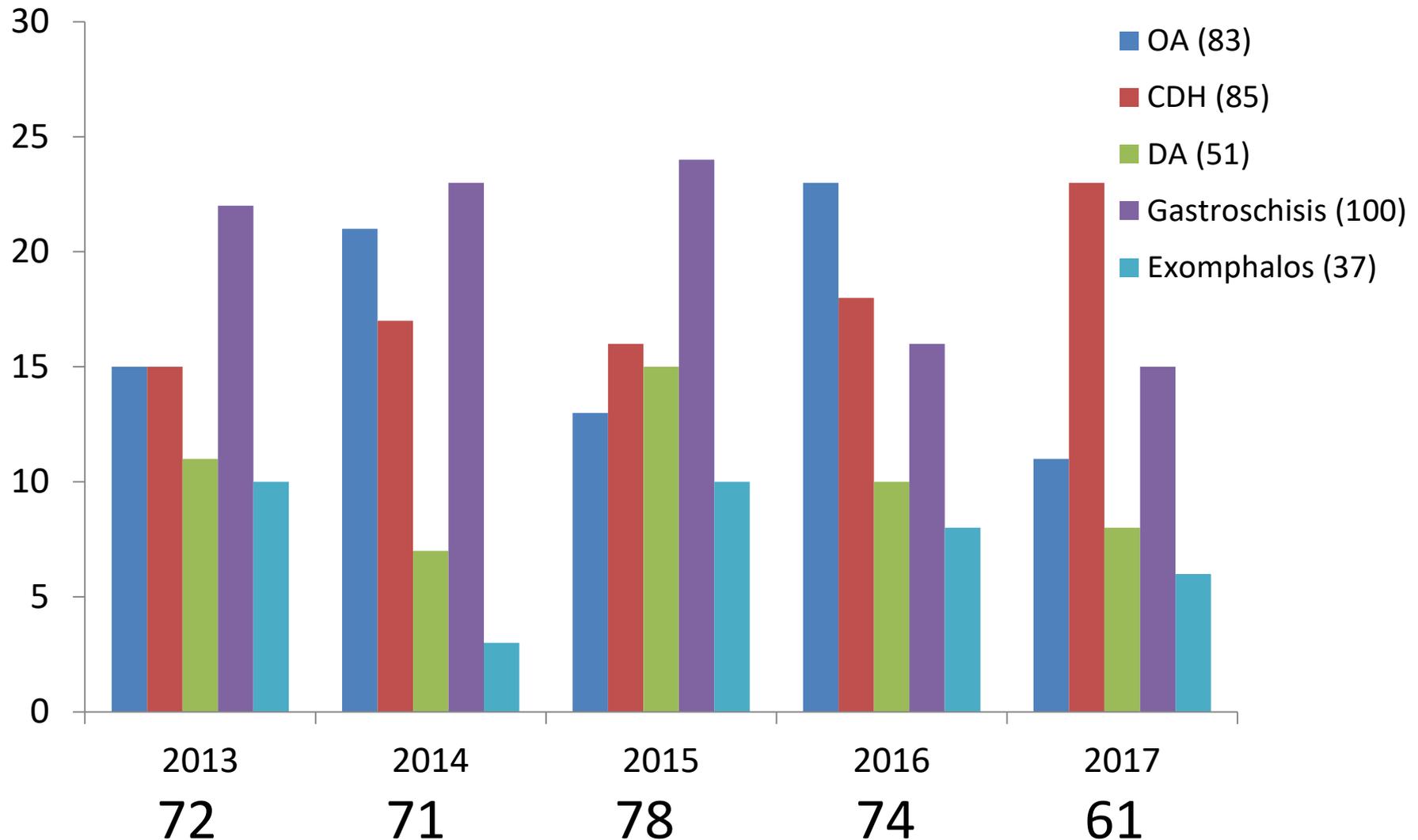
Glasgow – Hannah Hood, Judith Simpson, Gregor Walker

Edinburgh – Lisa Steven, Shona Cowan, Fraser Munro

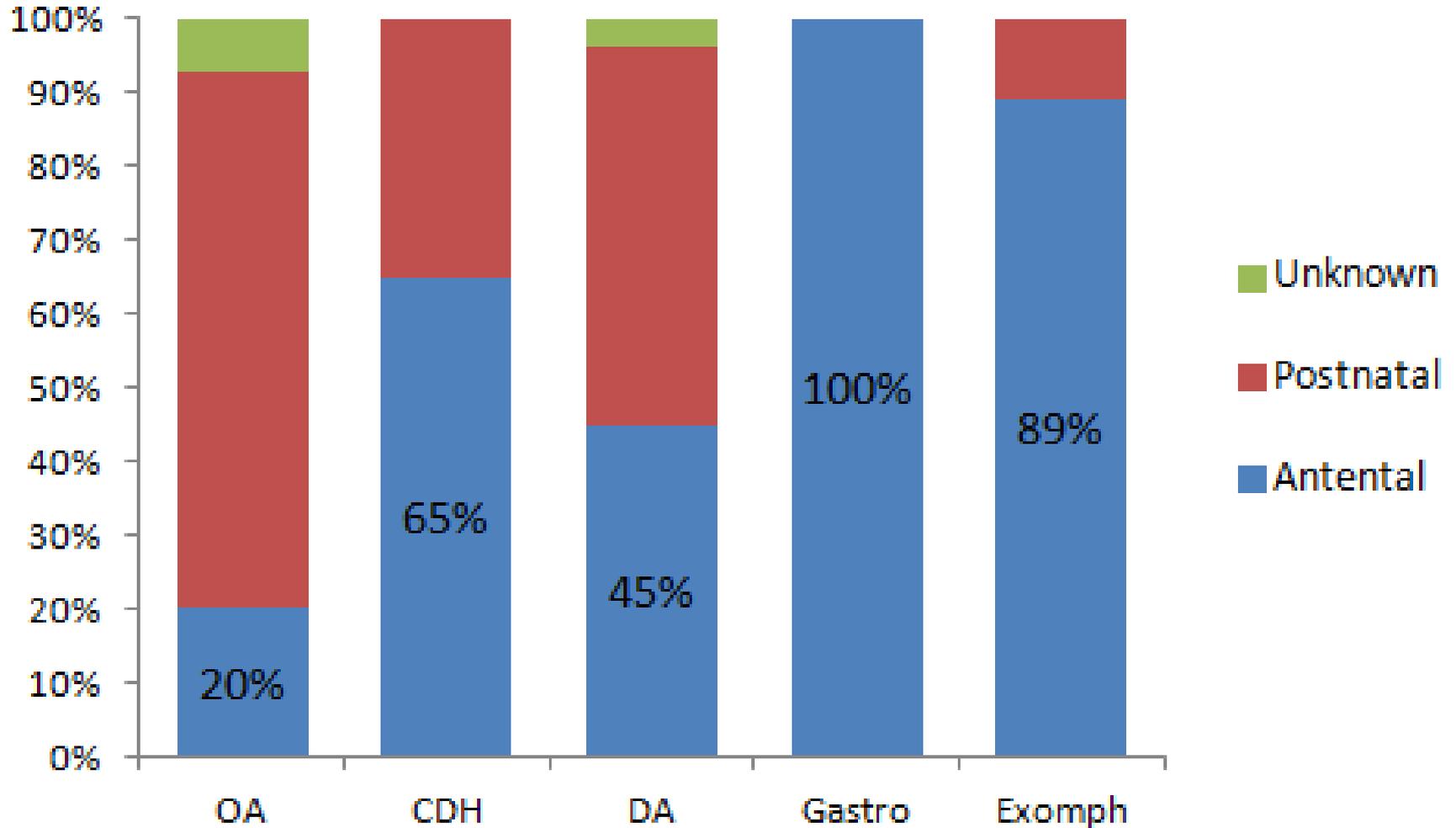
Aberdeen – Yatin Patel

Cauldicott Guardian approval obtained

# Diagnoses by year (n=356)



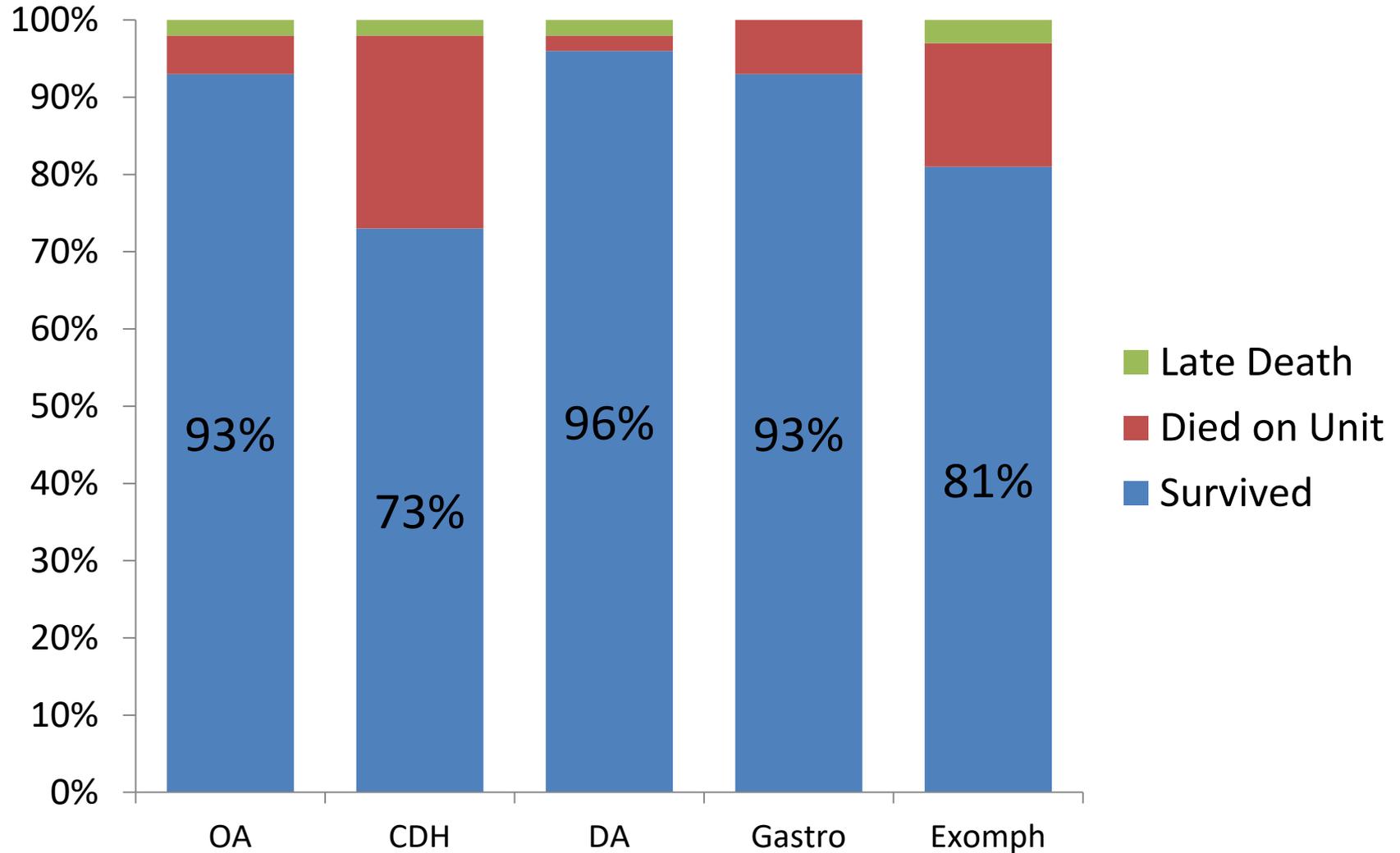
# When diagnosed



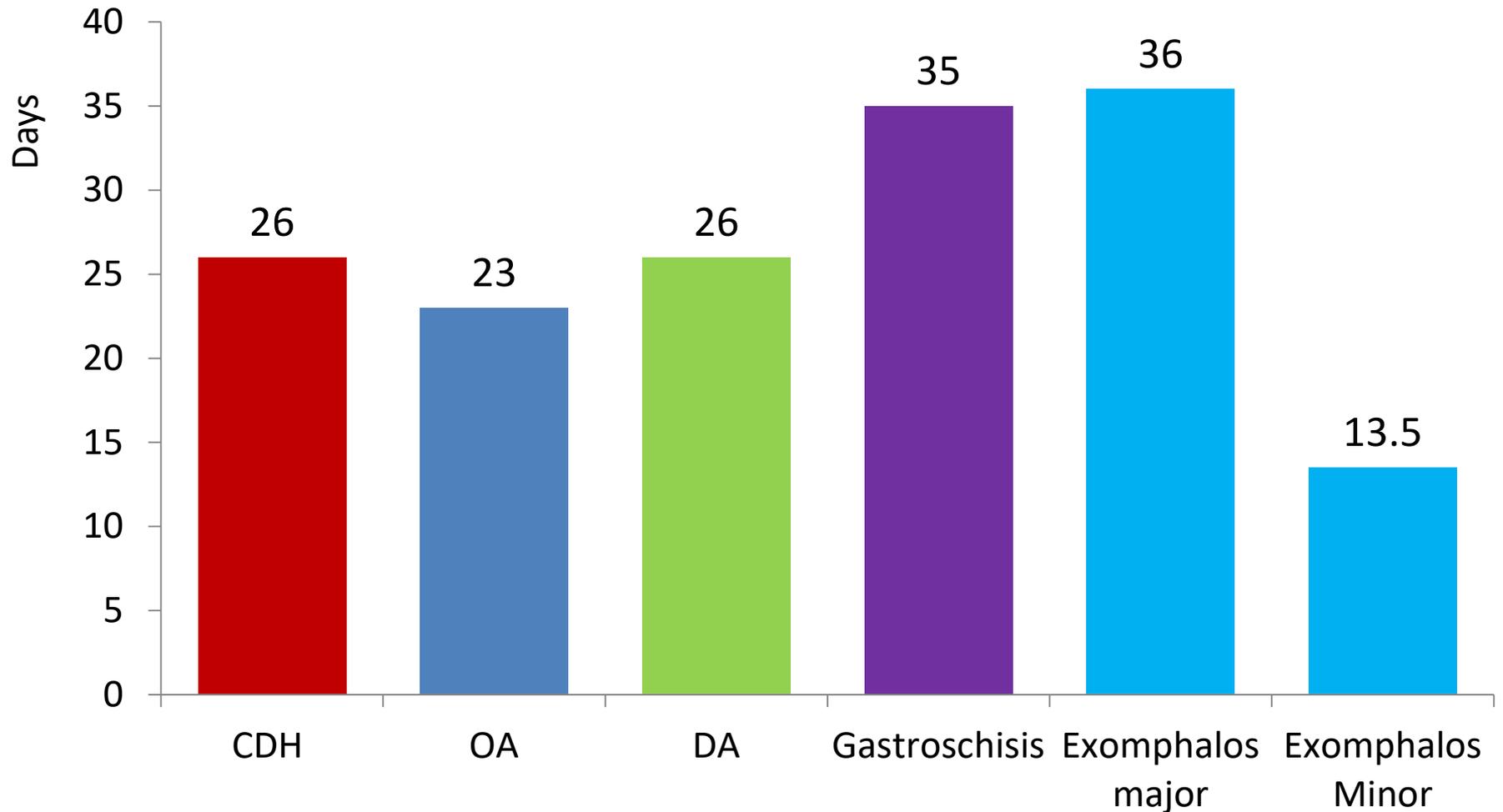
# Other associated anomalies

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

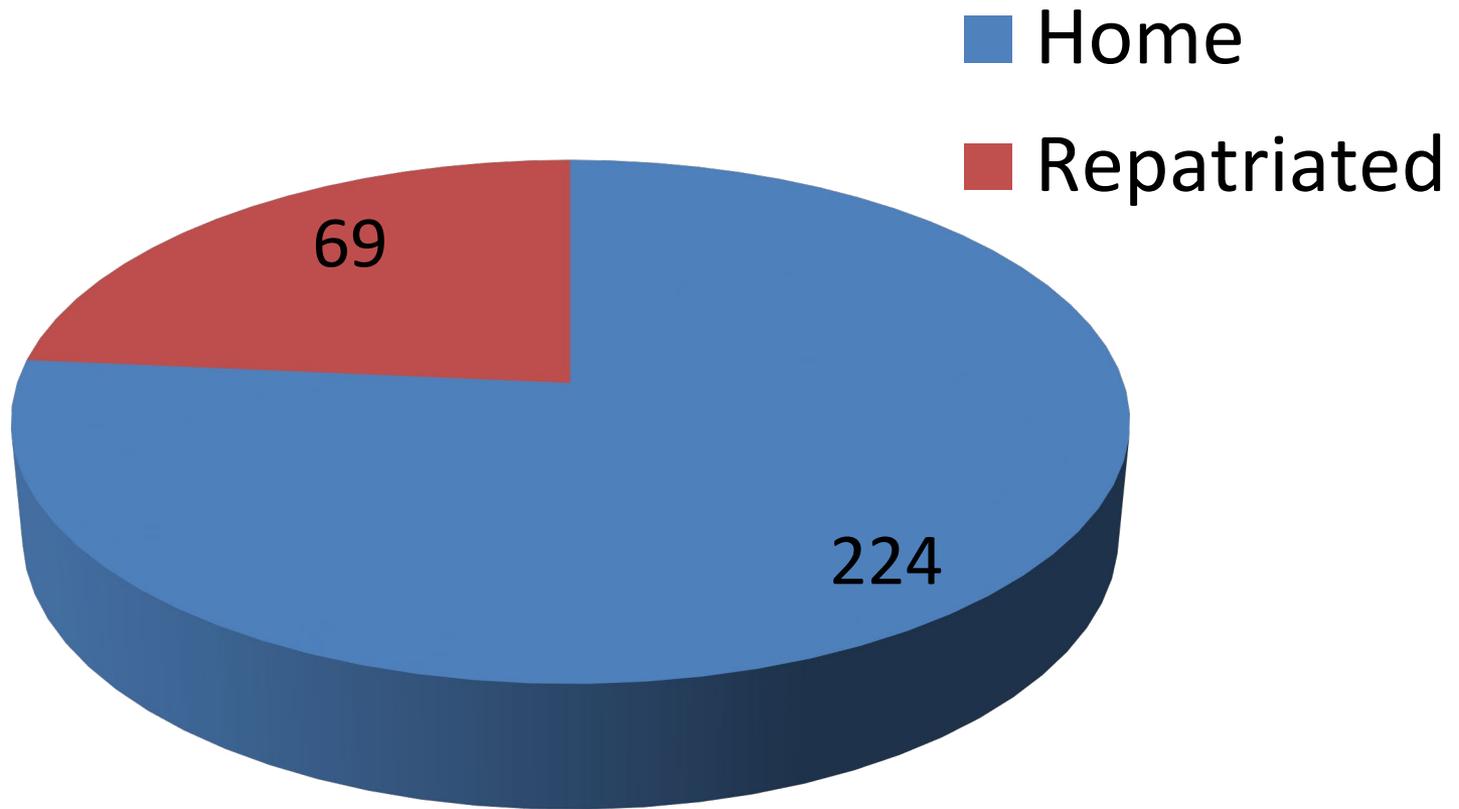


# Average\* stay in surgical centre



\* 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

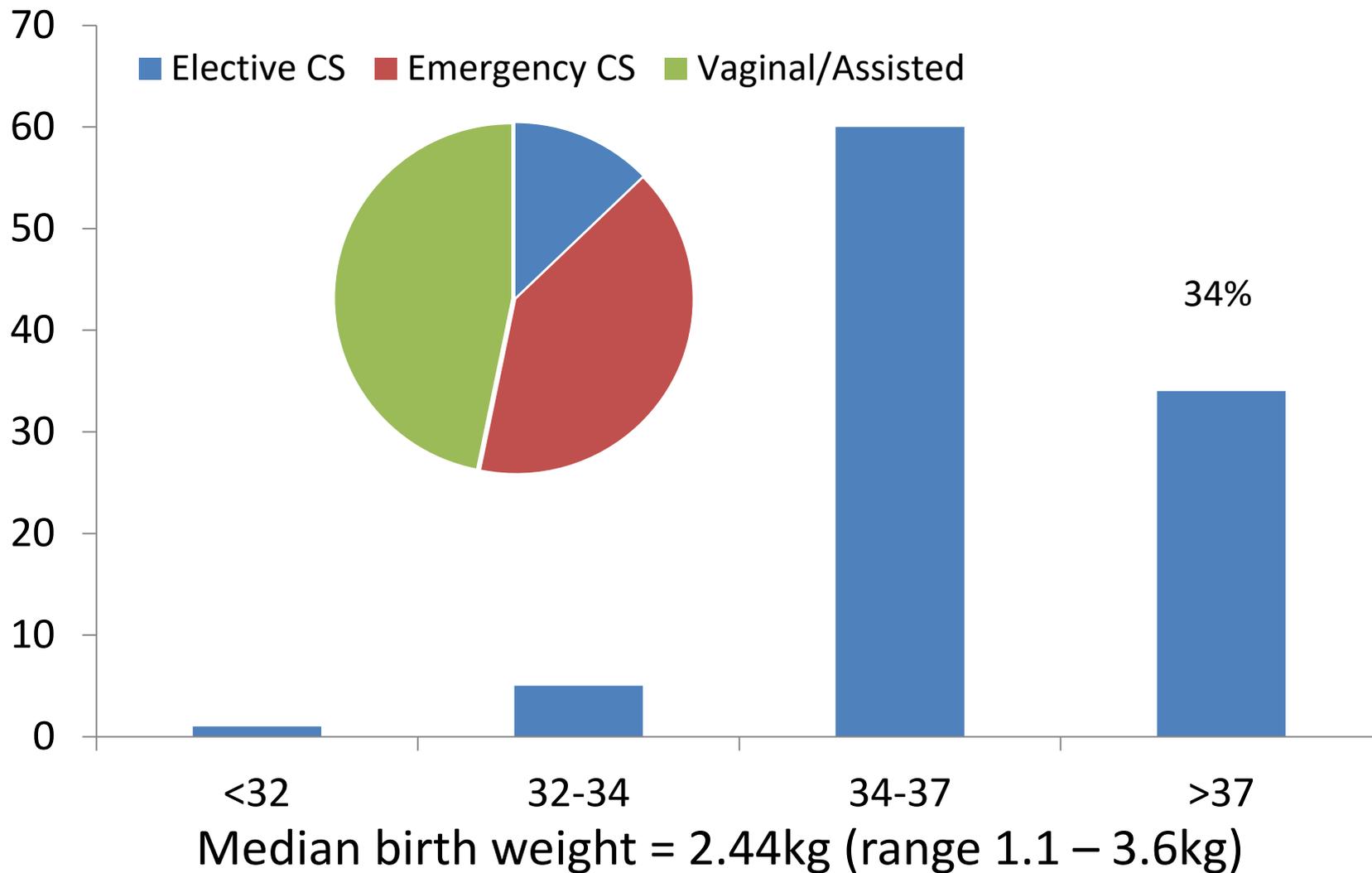
Growth - IUGR

Changes in fetal well being

Intestinal dilatation



# Gestational age & mode of delivery



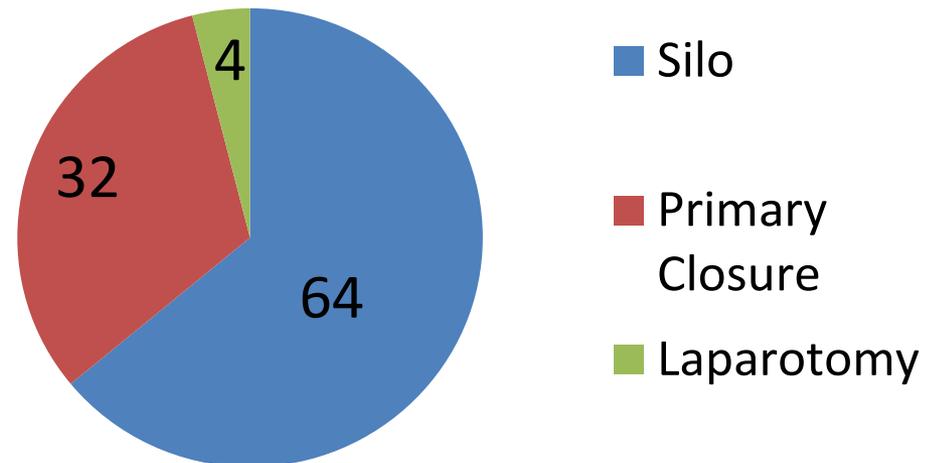
# Delivery management

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



# Surgical management

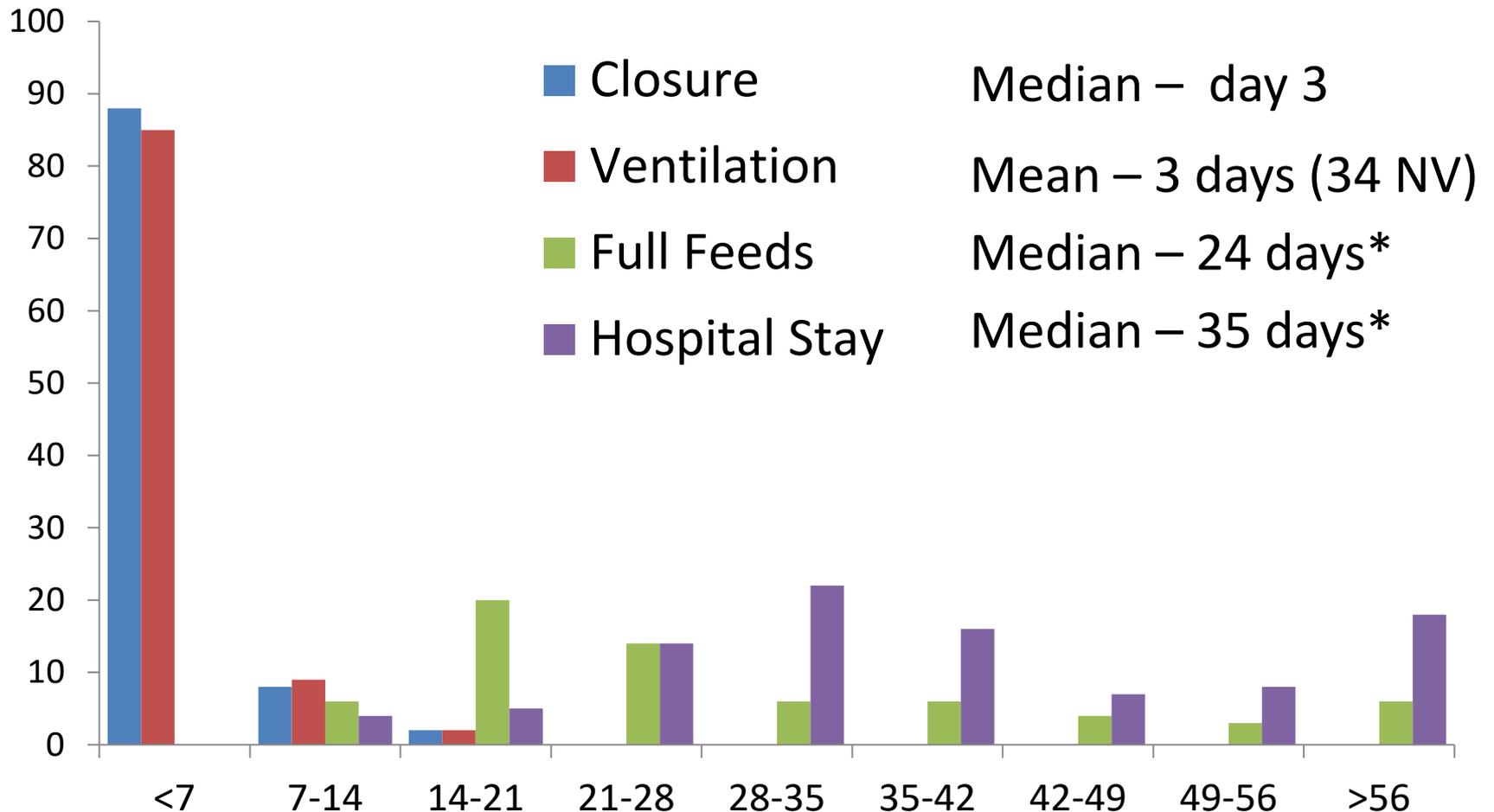
- **Staged (silo) closure**
  - In NNU / theatre
  - Suture / sutureless
- **Primary closure**
  - In NNU / in theatre
  - Suture / sutureless
- **Laparotomy**
  - If bowel complications



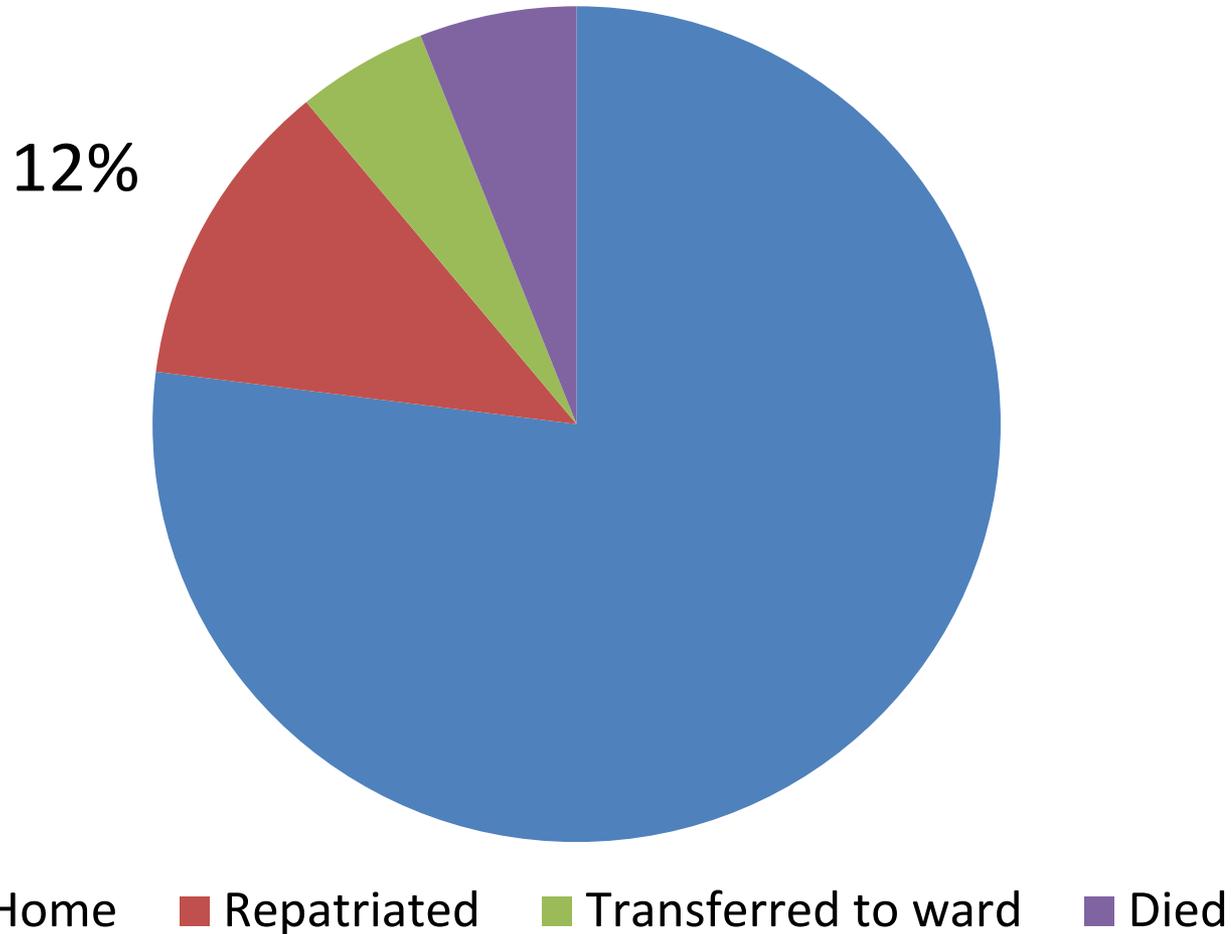
# Post closure management

- 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

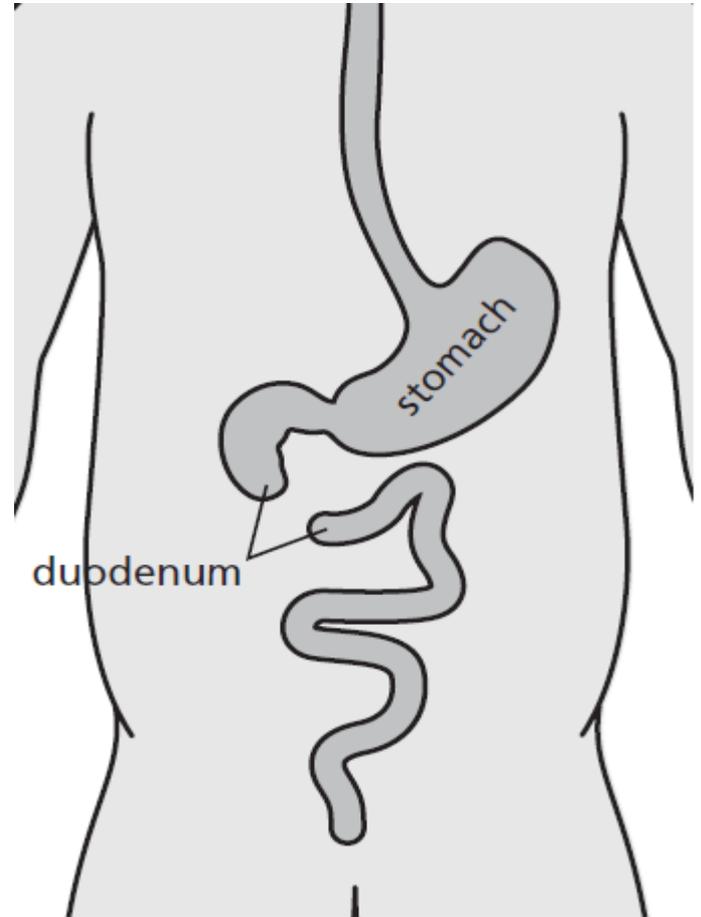
Obstruction in duodenum

Incidence ~1 in 5000 births

~10 births/year in Scotland

Important Associations

- Down Syndrome in 30%
- Malrotation in 30%

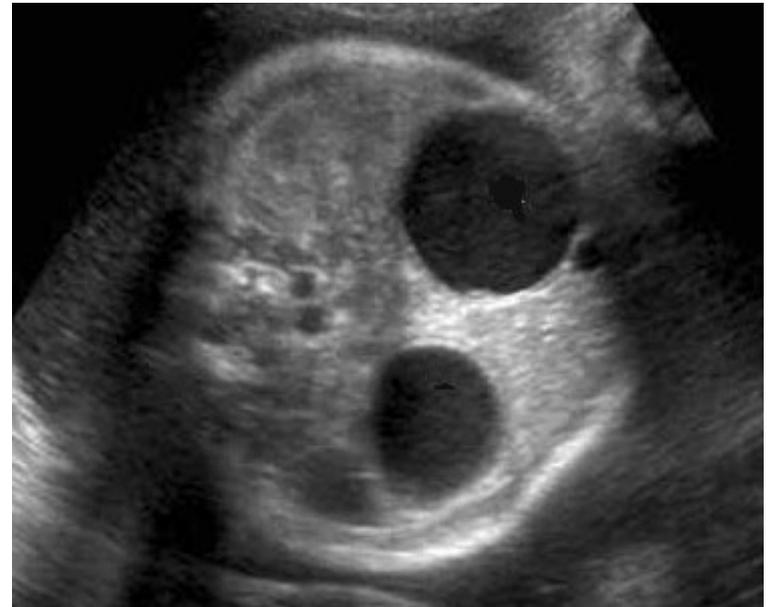


# Antenatal Factors

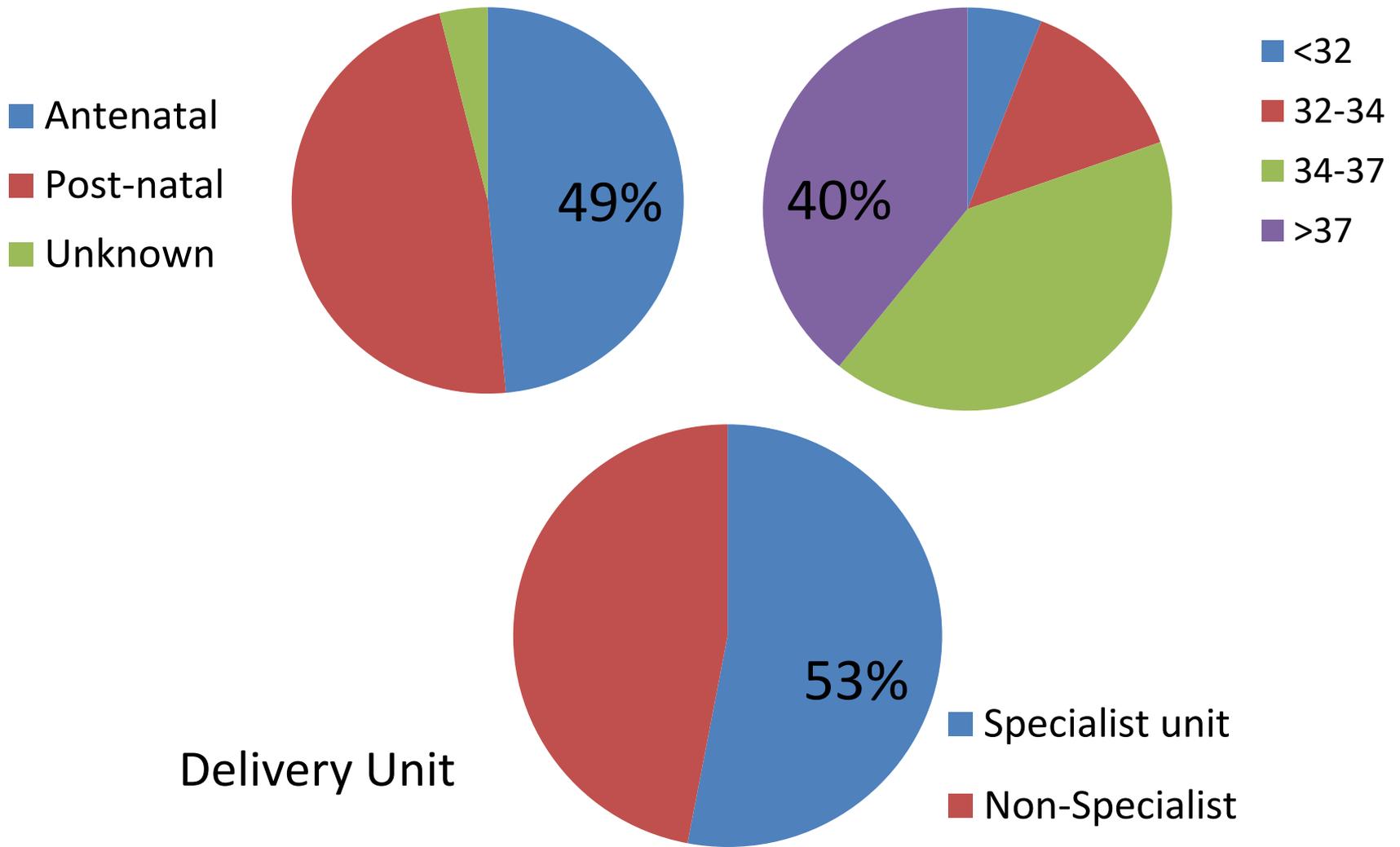
~50% diagnosed antenatally

Double bubble

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



# Diagnosis and Delivery



# Making the diagnosis

Bile vomiting/aspirates

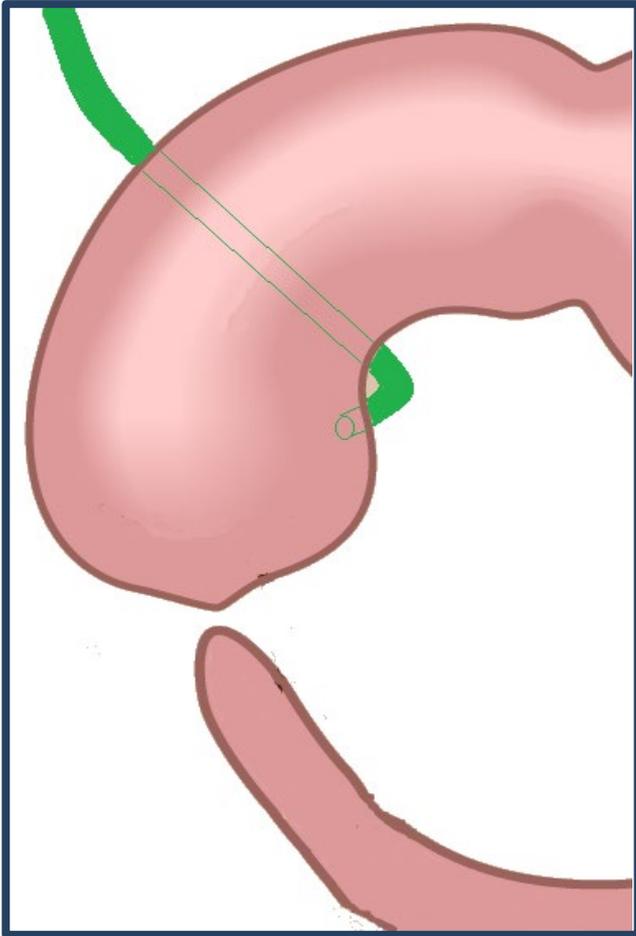
Characteristic x-ray

Look for associations

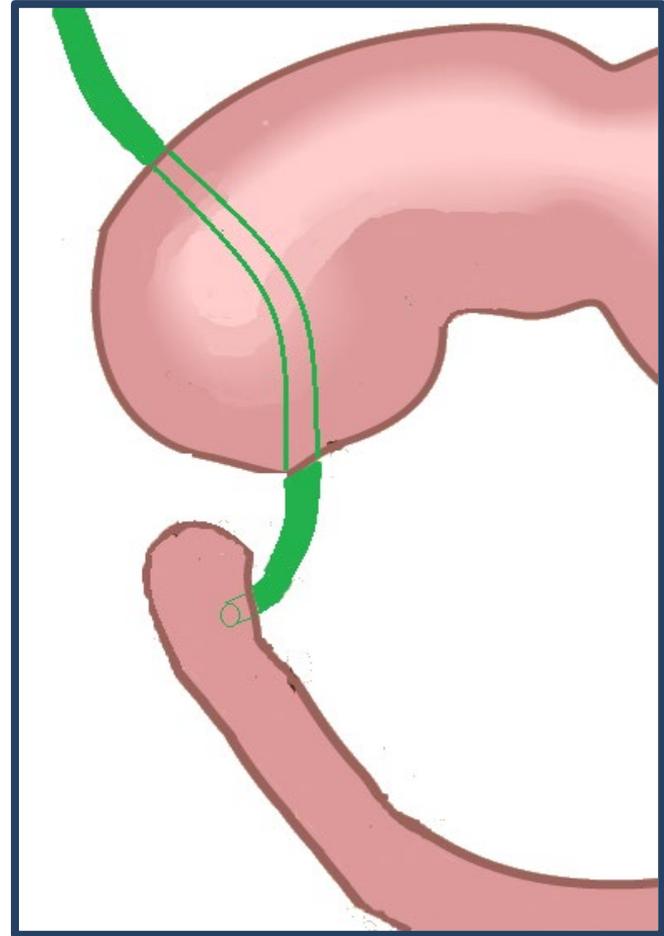
- Down syndrome
- Cardiac anomalies
- Oesophageal atresia
- Anorectal anomalies



# You don't need bile...



80%

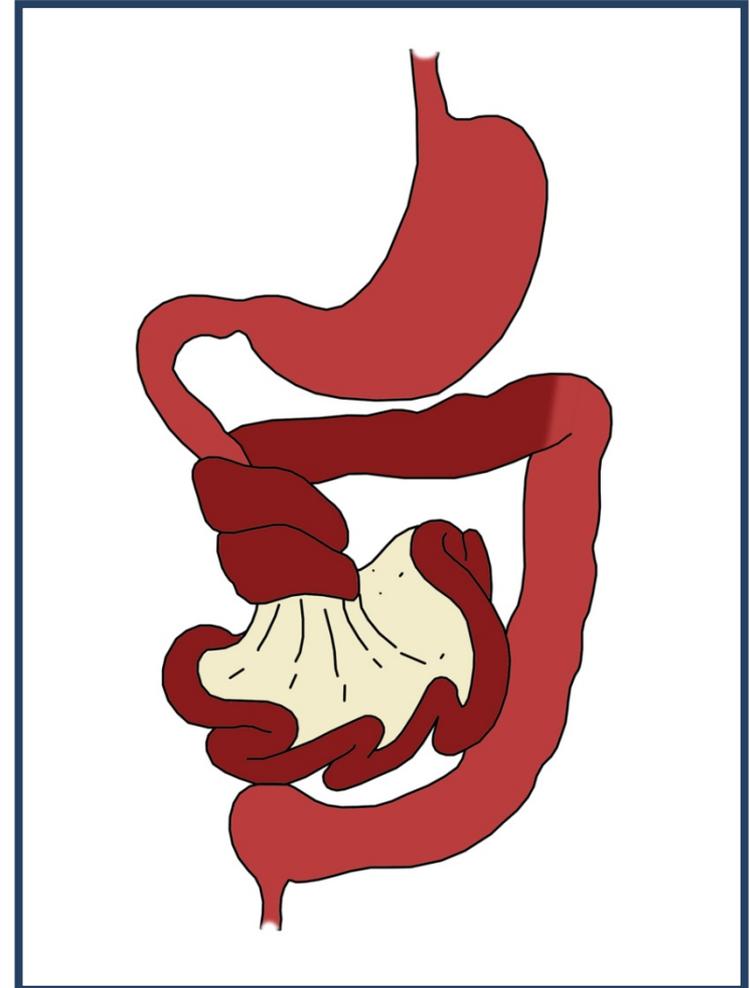
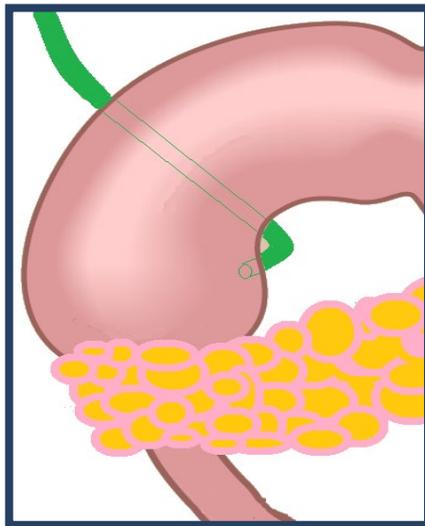
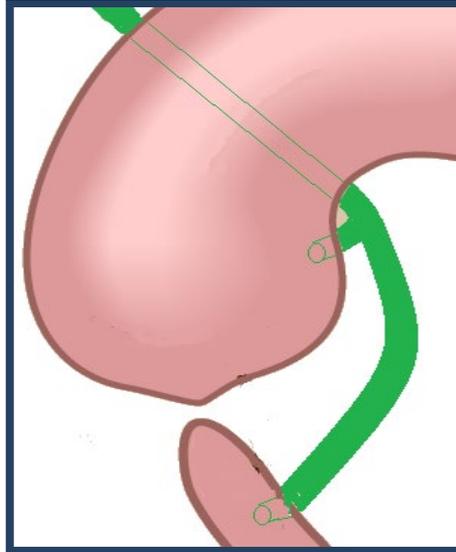
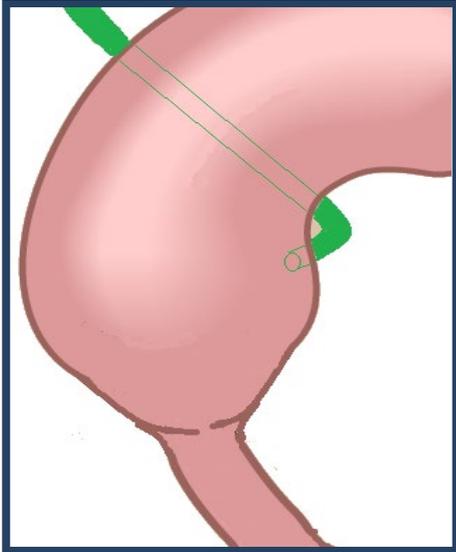


20%

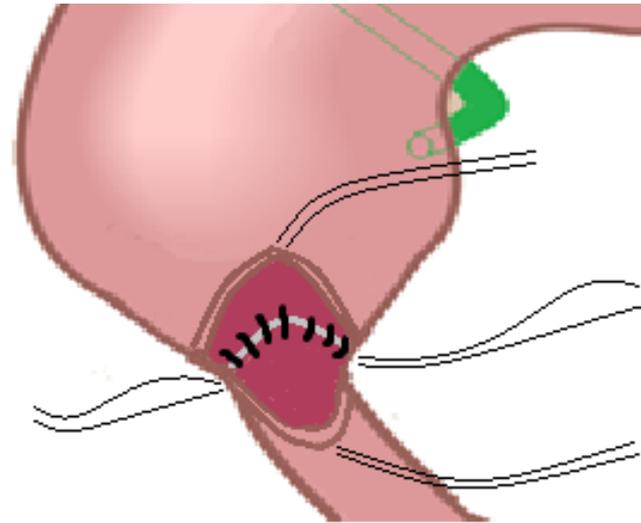
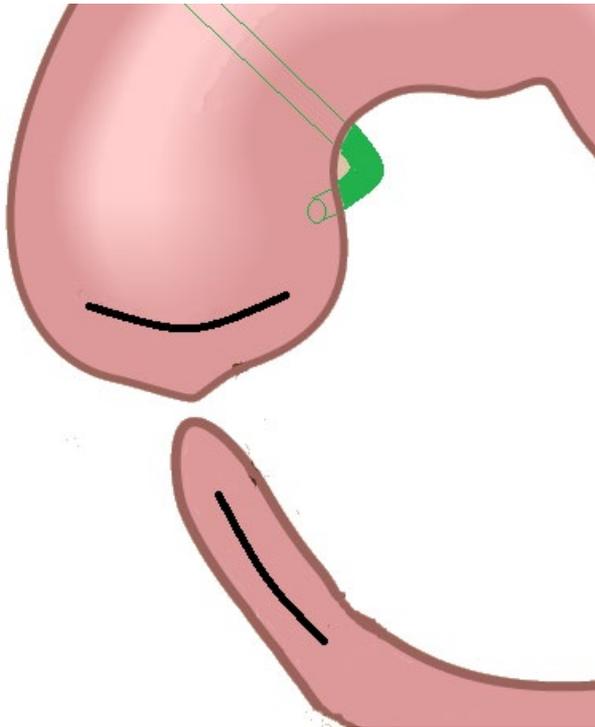
If there is gas beyond...



If there is gas beyond...



# Surgery

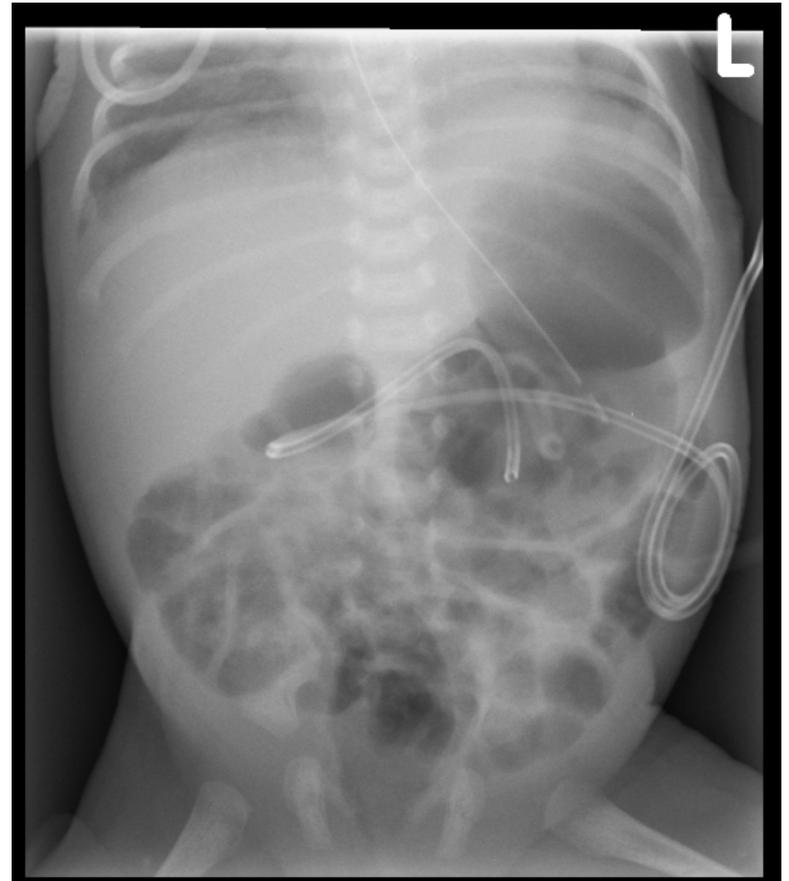


Duodenoduodenostomy  
Diamond configuration

Can be done laparoscopically (in Edinburgh)

# Post-operative Management

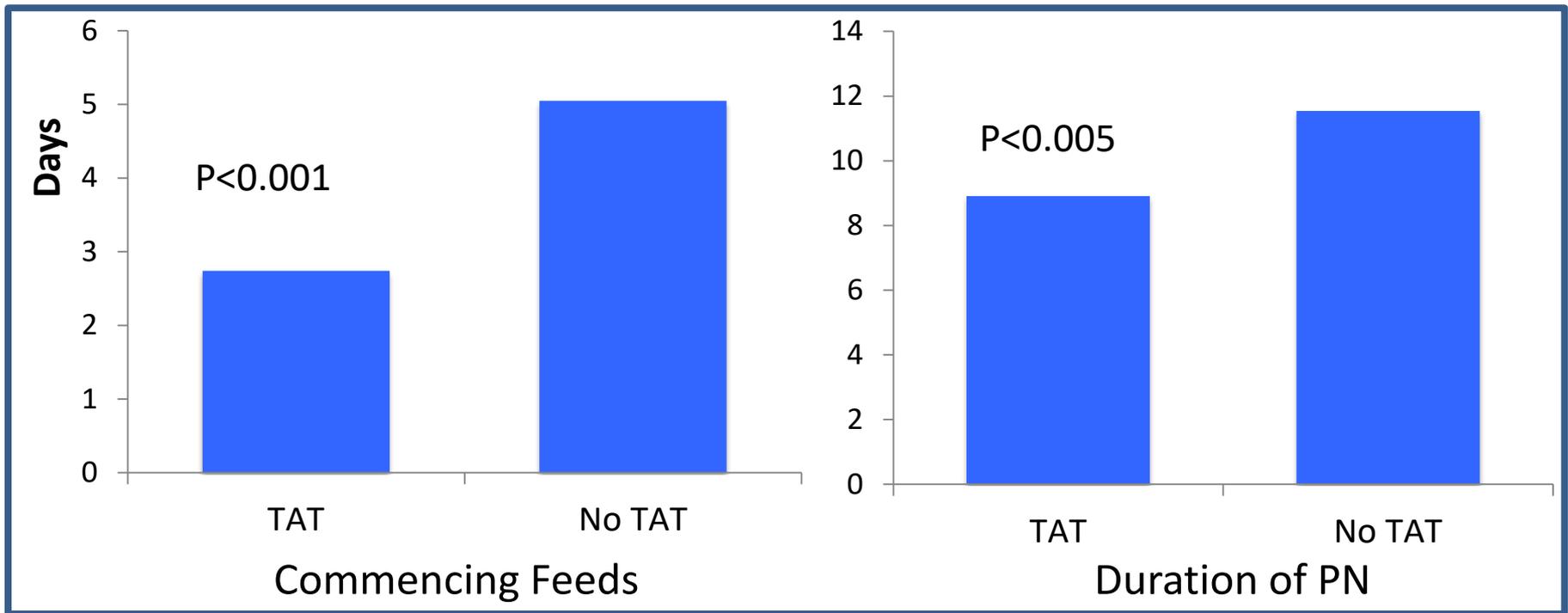
- Issues of Prematurity
- Associated anomalies
- Nutrition
  - Dilated duodenum
  - Slow to empty
  - Parenteral nutrition
  - Variable duration



# Previous Scottish Audit (1997-2011)

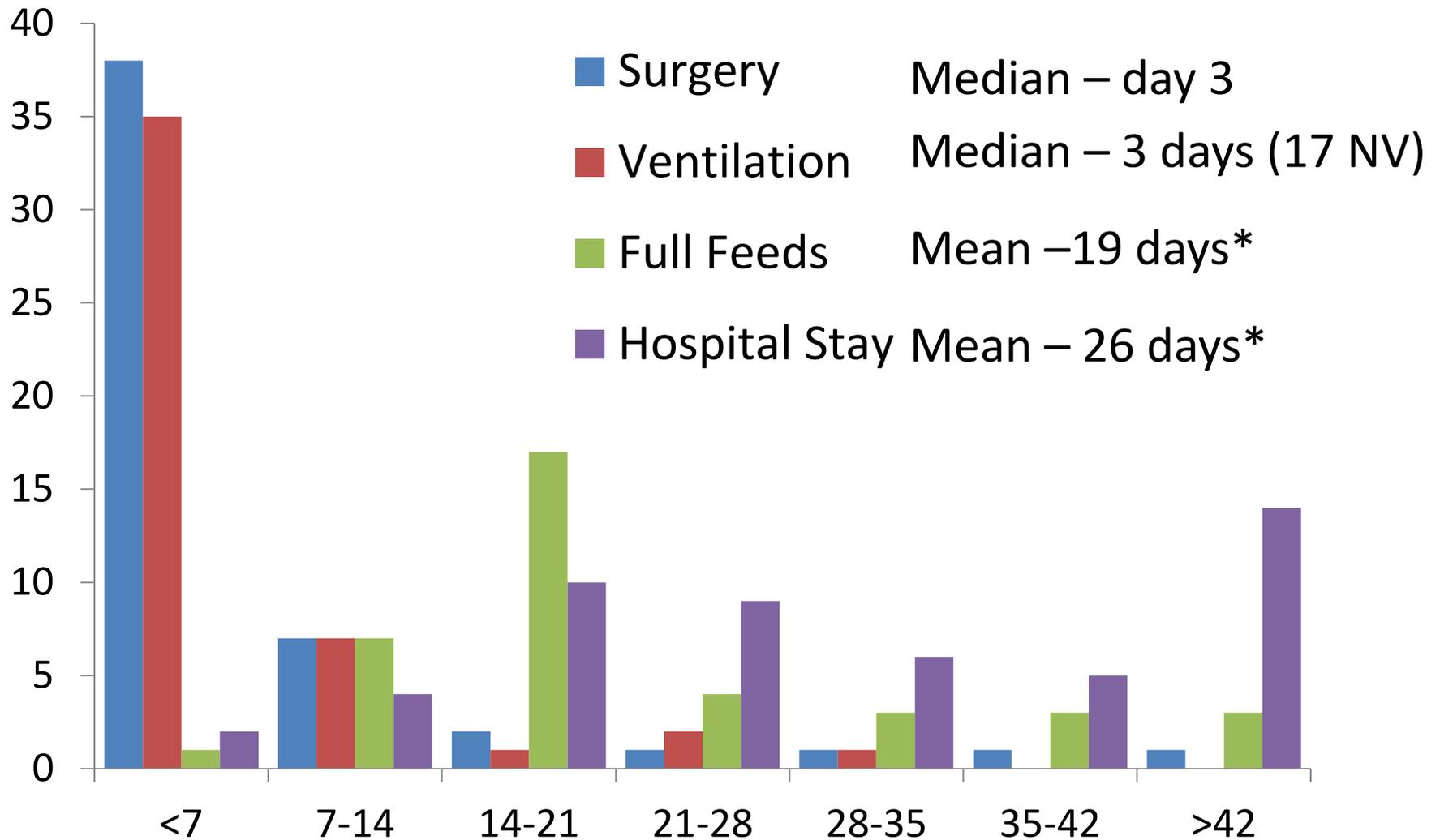
97 patients

TAT used in 68 (73%)

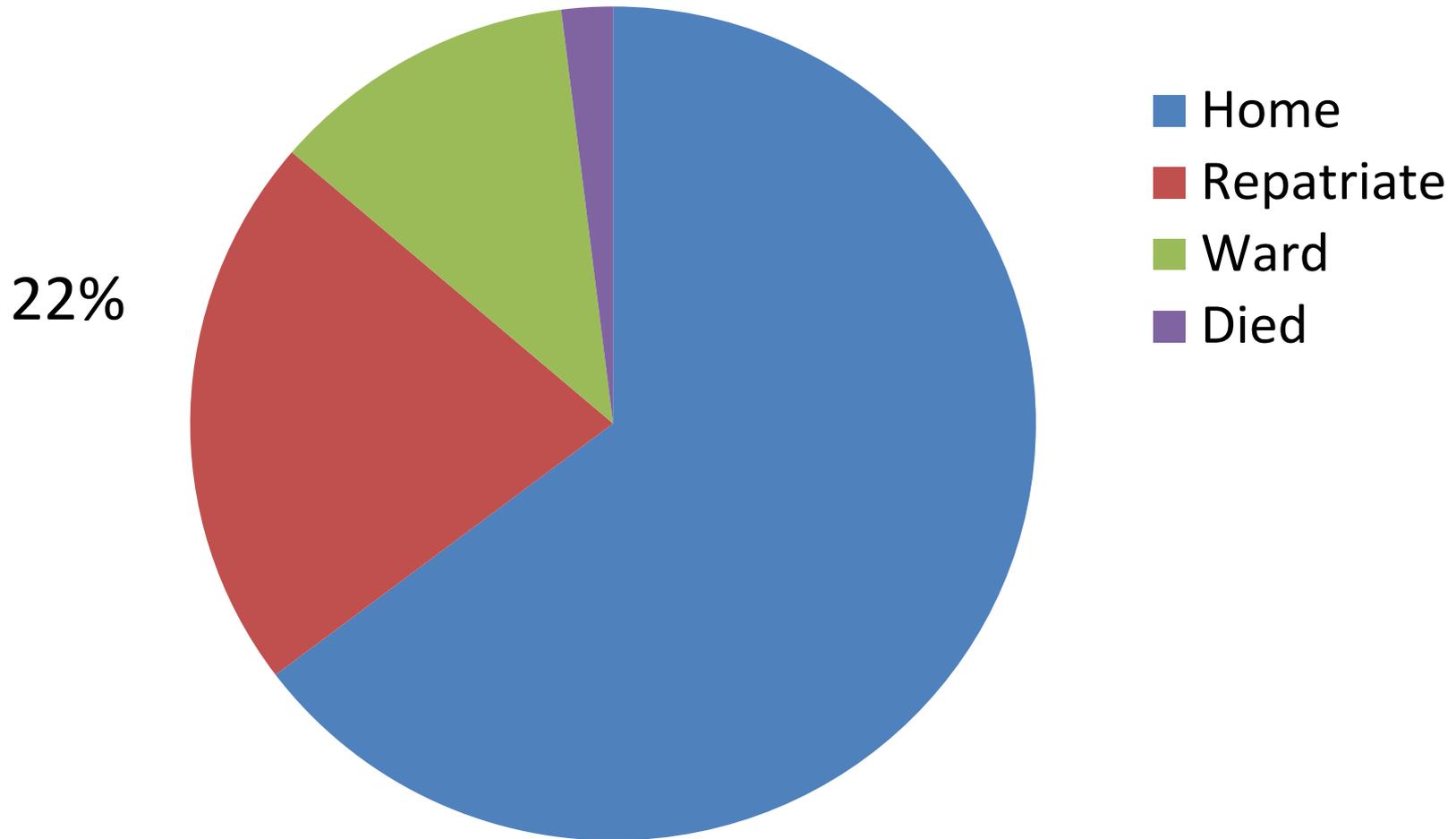


Use of TAT tube associated with longer inpatient stay ( $p=0.03$ )  
**-27 days** (95% CI 30.3-52) vs **20.5 days** (95%CI 17.2-27.5)

# 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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