ID Number:

British Association of Paediatric Surgeons Congenital Anomalies Surveillance System (BAPS-CASS)

Meconium Ileus in Association with Cystic Fibrosis

Data Collection Form

Details of treatment up to 28 days following surgery/disimpaction of the ileum

Case Definition:

Any live-born infant presenting between 1st October 2012 and 30th September 2014 with Meconium Ileus in association with Cystic Fibrosis. This is defined as bowel obstruction caused by inspissated meconium in the terminal ileum.

Please return the completed form to:

BAPS-CASS
National Perinatal Epidemiology Unit
University of Oxford
Old Road Campus
Oxford
OX3 7LF
Fax: 01865 617775
Phone: 01865 289714

Case reported in: _____

National Perinatal Epidemiology Unit

Instructions

1. Please do not enter any personally identifiable information (e.g. name, address or hospital number) on this form.
2. Please record the ID number from the front of this form against the infant's name on the Clinician's Section of the blue card retained in the BAPS folder.
3. Fill in the form using the information available in the infant's case notes.
4. If codes or examples are required, some lists (not exhaustive) are included on the back page of the form.
5. Tick the boxes as appropriate. If you require any additional space to answer a question please use the space provided in section 8.
6. Please complete all dates in the format DD/MM/YY, and all times using the 24hr clock e.g. 18:37
7. **If you do not know the answers to some questions, please indicate this in section 8.**
8. If you encounter any problems with completing the form please contact the Study Administrator or use the space in section 8 to describe the problem.

Section 1: Antenatal / Birth Data

1.1 **What was the mother's year of birth?** Y Y Y Y

1.2 **Please give the first alphabetical part of mother's postcode**
(e.g. OX for Oxfordshire, EH for Edinburgh, L for Liverpool) ☐☐

1.3 **Ethnic group[1]*** ☐☐

1.4 **Gestational age at birth** *(completed weeks)* ☐☐

1.5 **Gender** male ☐ female ☐ indeterminate ☐

1.6 **Birthweight** ☐☐☐☐ g

1.7 **Was Meconium Ileus suspected antenatally?** Yes ☐ No ☐

If No, *please go to 1.11*

If Yes, at what gestational age *(completed weeks)*? ☐☐

1.8 **Which feature suggested the diagnosis?** *(tick all that apply)*

- Echogenic bowel ☐
- Pseudocyst ☐
- Fetal Ascites ☐
- Dilated bowel ☐
- Calcification ☐
- Other ☐

If Other, please specify _____

1.9 **Was maternal polyhydramnios present?** Yes ☐ No ☐

1.10 **Is there a family history of cystic fibrosis?** Yes ☐ No ☐

If Yes, please specify which relatives _____

1.11 **Was an antenatal test for cystic fibrosis performed?** Yes ☐ No ☐

If Yes, please specify *(tick all that apply)*

- Maternal DNA ☐
- Paternal DNA ☐
- Fetal DNA ☐

*For guidance please see back cover

Section 2: Presentation

2.1 **Age in days at first presentation to your hospital** ☐☐☐ days

2.2 **What was the date of presentation?** D D / M M / Y Y

2.3 **Was the infant transferred from another hospital?** Yes ☐ No ☐

If Yes, please specify hospital where the infant was born _____

2.4 **Was the infant discharged home after birth and before diagnosis?** Yes ☐ No ☐

2.5 **What features were apparent at presentation?** *(tick all that apply)*

- Bile stained vomiting ☐
- Non-bile stained vomiting ☐
- Abdominal distension ☐
- Failure to pass meconium ☐
- Peritonitis ☐
- Bowel obstruction ☐
- Other ☐

If Other, please specify _____

2.6 **Were there any associated anomalies?** Yes ☐ No ☐

If Yes, please specify _____

Section 3: Initial Investigations and Management

3.1 **Was an abdominal x-ray performed?** Yes ☐ No ☐

If Yes, was there *(tick all that apply)*

Calcification ☐ Free air ☐ Dilated bowel loops ☐ Soap bubble appearance ☐ Other ☐

If Other, please specify _____

3.2 **Was a contrast enema performed?** Yes ☐ No ☐

What features were noted? *(tick all that apply)*

- Microcolon ☐
- Meconium plugs in colon ☐
- Meconium plugs in ileum ☐
- Contrast reached the ileum ☐
- Contrast reached proximal dilated bowel ☐

Please complete table below: (*continue in Section 8.5 if necessary*)

Date of Contrast Enema	Type of Contrast used	Successful	Complications*[2]
D D / M M / Y Y		Yes ☐ No ☐	
D D / M M / Y Y		Yes ☐ No ☐	
D D / M M / Y Y		Yes ☐ No ☐	

3.3 **Did the infant receive enteral N-acetylcysteine?** Yes ☐ No ☐

If Yes, Was N-acetylcysteine successful in disimpacting the terminal ileum? Yes ☐ No ☐

And please complete table below:

	Proximal gastrointestinal N-acetylcysteine (Nasogastric)	Distal gastrointestinal N-acetylcysteine (Per rectum)
Number of days used		
Volume (mls)		
Dilution (%)		
Frequency (times/day)		

3.4 **What was the date of diagnosis?** D D / M M / Y Y

Section 4: Initial Surgery

4.1 **Did the infant have surgery?** Yes ☐ No ☐

If No, please state reason for **not** having surgery _____

Now go to Section 5

If Yes, please continue.

4.2 **What was the indication for surgery?** *(tick one only)*

- Failure of contrast enema ☐
- Failure of N-acetylcysteine to disimpact the terminal ileum ☐
- Complication of enema ☐
- Complicated Meconium Ileus ☐
- Other ☐

If Other, please specify _____

4.3 **What was the date of initial surgery?** D D / M M / Y Y

4.4 **What was the infant's weight at time of surgery?** ☐☐☐☐ g

4.5 **What were the intra-operative findings?** *(tick all that apply)*

- Meconium obstruction in distal ileum (simple meconium ileus) ☐
- Perforation ☐
- Atresia ☐
- Volvulus ☐
- Meconium peritonitis ☐
- Meconium pseudocyst ☐
- Other ☐

If Other, please specify _____

*For guidance please see back cover

4.6 What surgical procedure was performed? *(tick one only)*

Intra-operative enterotomy and bowel irrigation ☐

If So, what irrigation agent was used? _____

Resection and primary anastomosis ☐

Resection and bowel irrigation ☐

If So, what irrigation agent was used? _____

Resection and Enterostomy

- Mikulicz double barrel ileostomy ☐
- Santuli procedure ☐
- Bishop Koop ileostomy ☐
- T-tube ileostomy ☐
- Terminal ileostomy and mucous fistula ☐
- Loop ileostomy ☐

Other ☐

If Other, please specify _____

4.7 Was any small bowel resected? Yes ☐ No ☐

If Yes, what length of small bowel was resected? ☐☐ cm

What length of small bowel is remaining?

- Proximal ☐☐ cm
- Distal ☐☐ cm
- Remaining bowel length unknown ☐

Is the Ileocaecal valve still present? Yes ☐ No ☐

4.8 Did the infant receive N-acetylcysteine post-operatively? Yes ☐ No ☐

	Proximal gastrointestinal N-acetylcysteine (Nasogastric)	**Distal gastrointestinal N-acetylcysteine (Per rectum)**
Number of days used		
Volume (mls)		
Dilution (%)		
Frequency (times/day)		

Section 5: Feeding at 28 days after surgery/ disimpaction

5.1 **Was the infant still on parenteral feeds at 28 days?** Yes ☐ No ☐

5.2 **Were enteral feeds started in first 28 days?** Yes ☐ No ☐

If Yes, please give date started D D / M M / Y Y

5.3 **Did the infant progress to full enteral feeds in first 28 days?** Yes ☐ No ☐

If Yes, please give date started D D / M M / Y Y

5.4 **Did the infant progress to full oral feeds in first 28 days?** Yes ☐ No ☐

If Yes, please give date started D D / M M / Y Y

5.5 **What type of milk was being given?** _____

5.6 **Were any enteral nutritional supplements given?** Yes ☐ No ☐

If Yes, please specify, e.g. carbohydrate, protein, medium chain triglycerides

_____

5.7 **Is the weight at 28 days known?** Yes ☐ No ☐

If Yes, please give weight ☐☐☐☐ g

Section 6: Early Morbidity (28 days post initial surgery /disimpaction)

6.1 **Did the infant develop a wound infection requiring antibiotics?**

Yes ☐ No ☐ Not applicable

6.2 **Did the infant develop an anastomotic leak?** Yes ☐ No ☐ Not applicable

6.3 **Did the infant develop an adhesive bowel obstruction?** Yes ☐ No ☐

6.4 **Did the infant develop a stoma complication?** Yes ☐ No ☐ Not applicable

If Yes, please tick all that apply

- Necrosis ☐
- Retraction ☐
- Prolapse ☐
- Parastomal skin breakdown ☐
- Parastomal hernia ☐
- Other ☐

If Other, please specify _____

6.5 **Did the infant develop a further meconium obstruction?** Yes ☐ No ☐

6.6 **Were there any other early (up to 28 days) complications?** Yes ☐ No ☐

If Yes, please specify _____

6.7 **Were any further surgical procedures required in the first 28 days post disimpaction/post surgery?** Yes ☐ No ☐

If Yes, please specify

*For guidance please see back cover

Date of surgery	Details of Further Surgical Procedure
D D / M M / Y Y	
D D / M M / Y Y	
D D / M M / Y Y	

Section 7: Tests for Cystic Fibrosis

7.1 **Did the infant have newborn IRT (Immunoreactive Trypsin) screening for cystic fibrosis?** Yes ☐ No ☐

If Yes, did it confirm cystic fibrosis? Yes ☐ No ☐

7.2 **Did the infant have DNA testing for common CFTR (cystic fibrosis transmembrane conductance regulator) gene mutations?** Yes ☐ No ☐

If Yes, was a recognised CFTR gene mutation identified? Yes ☐ No ☐

If Yes, what was the genotype? *(tick one only)*

- F508/F508 ☐
- F508/other ☐
- other/other ☐

7.3 **Did the infant have a sweat test performed?** Yes ☐ No ☐

If Yes, did it confirm cystic fibrosis? Yes ☐ No ☐

Section 8: Other information

8.1 **Has the infant been discharged home?** Yes ☐ No ☐

If Yes, specify date of discharge D D / M M / Y Y

8.2 **Has the infant been discharged to another hospital?** Yes ☐ No ☐

If Yes, please give name of hospital _____

Name of responsible consultant _____

Date of transfer D D / M M / Y Y

8.3 **Did this infant die?** Yes ☐ No ☐

If Yes, please give date of death D D / M M / Y Y

Cause of death as stated on the death certificate *(please state if not known)*

_____

8.4 **Were the parents given any of the following?** *(tick all that apply)*

- In-hospital produced information leaflets ☐
- Contact details for support groups ☐
- Information leaflets for support groups ☐
- Offer of Genetic Counselling appointment ☐

8.5 Please add other relevant information below

Section 9:

Name of person completing the form _____

Designation _____

Today's date D D / M M / Y Y

You may find it useful in the case of queries to keep a copy of this form.

Definitions

1. UK Census Coding for ethnic group

WHITE
- 01. British
- 02. Irish
- 03. Any other white background

MIXED
- 04. White and black Caribbean
- 05. White and black African
- 06. White and Asian
- 07. Any other mixed background

ASIAN OR ASIAN BRITISH
- 08. Indian
- 09. Pakistani
- 10. Bangladeshi
- 11. Any other Asian background

BLACK OR BLACK BRITISH
- 12. Caribbean
- 13. African
- 14. Any other black background

CHINESE OR OTHER ETHNIC GROUP
- 15. Chinese
- 16. Any other ethnic group

2. Complications:

Perforation

Hepatotoxicity

Hypovolemic shock