

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

Hirschsprung's Disease

Data Collection Form

Details Of Treatment Until 28 Days
Following Definitive Surgery

Case Definition:

Any live-born infant, **up to 6 months of age DIAGNOSED** between 1st October 2010 and 31st March 2012 with Hirschsprung's Disease. This is defined as an absence of ganglia in the enteric nervous system of the distal bowel (aganglionosis).

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: _____

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?
- 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** Is there a family history of HD? Yes No
If Yes, please specify which relatives and indicate if maternal or paternal?

- 1.8** Age at first spontaneous meconium passage (not stained liquor)
- | | |
|-----------|--------------------------|
| < 24 hrs | <input type="checkbox"/> |
| 24-48 hrs | <input type="checkbox"/> |
| > 48 hrs | <input type="checkbox"/> |
| Not known | <input type="checkbox"/> |

*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?

/ /

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)

Abdominal distension

Bile vomiting

Non-bile vomiting

Suspected enterocolitis

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 What was the date of definitive diagnosis?

/ /

3.2 Was a contrast enema performed?

Yes No

If Yes, were the following performed < 24 hrs prior to contrast study?

PR examination

Yes No Don't know

Washout/enema

Yes No Don't know

Was a Transition Zone reported by radiologist?

Yes No

If Yes, please select site

Rectosigmoid

Descending colon

Transverse colon

Ascending colon

Small bowel

3.3 Were rectal biopsies performed?

Yes No

(Please give details from **separate** episodes i.e. on different dates)

Date	Method (please tick)	Result (please tick)
DD / MM / YY	Suction <input type="checkbox"/> Punch <input type="checkbox"/> Full-thickness <input type="checkbox"/>	Normal <input type="checkbox"/> Abnormal <input type="checkbox"/> Suspicious <input type="checkbox"/> Inadequate <input type="checkbox"/>
DD / MM / YY	Suction <input type="checkbox"/> Punch <input type="checkbox"/> Full-thickness <input type="checkbox"/>	Normal <input type="checkbox"/> Abnormal <input type="checkbox"/> Suspicious <input type="checkbox"/> Inadequate <input type="checkbox"/>
DD / MM / YY	Suction <input type="checkbox"/> Punch <input type="checkbox"/> Full-thickness <input type="checkbox"/>	Normal <input type="checkbox"/> Abnormal <input type="checkbox"/> Suspicious <input type="checkbox"/> Inadequate <input type="checkbox"/>

3.4 Was anorectal manometry performed?

Yes No

3.5 Was the infant allowed home after diagnosis and before definitive surgery?

Yes No

Section 4: Management before Definitive Surgery

4.1 Was the infant managed with washouts?

Yes No

If Yes, were any washouts performed at home?

Yes No

4.2 Was a de-functioning stoma performed before definitive surgery?

Yes No

If Yes, please continue

If No, please go to section 5

4.3 Date and time of stoma formation

DD / MM / YY hh : mm
24hr

4.4 What was the body weight at stoma formation? (please state if not known)

g

4.5 What was the reason for stoma formation?*

4.6 Were biopsies taken?

Yes No

If Yes, how were biopsies obtained?

Laparoscopically Open

Was the biopsy result available intra-operatively?

Yes No

4.7 Please indicate the site of the stoma

- Sigmoid
- Descending colon
- Transverse colon
- Ascending colon
- Small bowel

4.8 Were there any stoma-related complications requiring revision?

Yes No

4.9 Was the infant discharged home after stoma formation? (before pull-through)

Yes No

If Yes, please give date of discharge

DD / MM / YY

Section 5: Definitive Surgery

5.1 Has the infant had definitive surgery?

Yes No

If No, has a date for definitive surgery been planned?

Yes No

If Yes, what is the planned date for definitive surgery?

/ /

If the infant has NOT had surgery, please go to section 8

5.2 What was the date of definitive surgery?

/ /

5.3 How many consultants were involved (operating and/or assisting)?

5.4 What was the body weight at definitive procedure? (please state if not known)

g

5.5 Were intra-operative biopsies obtained?

Yes No

If Yes, how were biopsies obtained?

Laparoscopically Open Transanal

5.6 Please indicate site of pathological transition zone diagnosed at surgery

Rectosigmoid

Descending colon

Transverse colon

Ascending colon

Small bowel

5.7 What distance above "normal" biopsy was the pull-through carried out?

cm

5.8 How was the colonic mobilisation performed?

Procedure	Attempted	Successful	Please indicate the order in which procedures were attempted (1,2,3 etc)
Open (abdominal incision)	<input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/>	<input type="checkbox"/>
Entirely laparoscopically	<input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/>	<input type="checkbox"/>
Entirely transanally	<input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/>	<input type="checkbox"/>
Combined laparoscopic/transanal	<input type="checkbox"/>	Yes <input type="checkbox"/> No <input type="checkbox"/>	<input type="checkbox"/>

5.9 How was the distal rectum dissected (i.e. operative technique)?

Submucosal dissection (i.e. Soave-Boley)

Yes No

If Yes, was the muscle cuff split?

Yes No

Perirectal dissection (i.e. Swenson)

Yes No

Posterior dissection (i.e. Duhamel)

Yes No

If Yes, estimated length of anterior aganglionic rectum/colon

cm

Other

Yes No

If Yes, please specify _____

5.10 What distance was the anastomosis above dentate line?

. cm

5.11 Was the distal bowel everted to suture the anastomosis?

Yes No

5.12 Has the stoma (if present) been closed?

Yes No N/A

If Yes, what date was it closed

/ /

Section 6: Results from Pathology Report *(please state if not known)*

- 6.1 What was the total length of bowel resected (after fixation)? cm
- 6.2 What was the minimum length of ganglionic bowel resected (after fixation)? . cm
- 6.3 Were thickened nerve trunks reported at the proximal resection margin? Yes No
- 6.4 Were features of Intestinal Neuronal Dysplasia reported? Yes No
- 6.5 Were there features of enterocolitis in the specimen? Yes No
- 6.6 If the Soave-Boley technique was used, what was the length of the mucosal sleeve (after fixation)? . cm

Section 7: Post-surgery Morbidity (<28 days post definitive procedure)

- 7.1 Did an anastomotic leak occur at site of pull-through? Yes No
- 7.2 Were anastomotic dilatations performed post-operatively? Yes No
If Yes, why were they employed? Electively Diagnosed stricture
- 7.3 Did the infant have a wound infection requiring antibiotics? Yes No
- 7.4 Did the infant have a pelvic/cuff abscess? Yes No
- 7.5 Did perianal excoriation delay discharge or require re-admission? Yes No
- 7.6 Was post-operative enterocolitis suspected? Yes No
- 7.7 Were there any other early complications? Yes No
If Yes, please specify _____
- 7.8 Were any further surgical procedures required in the first 28 post-op days? Yes No

If Yes, please specify

Date of Surgery	Details of Further Surgical Procedure
<input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/>	
<input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/>	

Section 8: Other information

- 8.1 Has the infant been discharged home? Yes No
If Yes, please specify date of discharge / /
- 8.2 Has the infant been transferred to another hospital? Yes No
If Yes, please give name of hospital _____
Name of responsible consultant _____
Date of transfer / /

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. Reason for stoma formation, for example:

Consultant preference for stage approach in all cases

Long segment disease

Emergency laparotomy, e.g. for perforation

Failure to decompress

Enterocolitis

Co-morbidity