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? 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*

1.4 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 ☐
   24-48 hrs ☐
   > 48 hrs ☐
   Not known ☐

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

<table>
<thead>
<tr>
<th>Date</th>
<th>Method (please tick)</th>
<th>Result (please tick)</th>
</tr>
</thead>
<tbody>
<tr>
<td>DD/MM/YY</td>
<td>Suction</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Abnormal</td>
</tr>
<tr>
<td></td>
<td>Punch</td>
<td>Suspicious</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Inadequate</td>
</tr>
<tr>
<td></td>
<td>Full-thickness</td>
<td></td>
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</tbody>
</table>

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 h:mm

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

*For guidance please see back cover
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?
   - D D / M M / Y Y

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

5.2 What was the date of definitive surgery?
   - D D / M M / Y Y

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?

5.9 How was the distal rectum dissected (i.e. operative technique)?
   - Submucosal dissection (i.e. Soave-Boley)
   - Perirectal dissection (i.e. Swenson)
   - Posterior dissection (i.e. Duhamel)

   If Yes, estimated length of anterior aganglionic rectum/colon
   - cm

   Other

   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
   - D D / M M / Y Y
Section 6: Results from Pathology Report

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

<table>
<thead>
<tr>
<th>Date of Surgery</th>
<th>Details of Further Surgical Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>DD/MM/YYYY</td>
<td></td>
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<tr>
<td>DD/MM/YYYY</td>
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</tbody>
</table>

Section 8: Other information

8.1 Has the infant been discharged home? Yes No

If Yes, please specify date of discharge DD/MM/YYYY

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 DD/MM/YYYY
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? (please tick all that apply)  
   Yes ☐ No ☐  
   In-hospital produced information leaflets ☐  
   Contact details for support groups ☐  
   Information leaflets from 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. 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