

Evolution in the management of Hirschsprung's disease in the UK and Ireland: a national survey of practice revisited

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ABSTRACT

INTRODUCTION The management of Hirschsprung's disease continues to evolve. This questionnaire survey aimed to determine current surgical management strategies for Hirschsprung's disease in Britain.

SUBJECTS AND METHODS The survey was sent electronically to all British paediatric surgeons. Initial questions explored individual experience and regional service provision. Additional questions, reserved for surgeons who perform definitive Hirschsprung's disease surgery, addressed specific clinical scenarios.

RESULTS Surveys were sent to 142 surgeons yielding 85 responses. After exclusions, 64 surveys from 21 centres were analysed. Forty-seven respondents worked in centres with designated 'Hirschsprung's disease surgeons'. Forty respondents perform definitive Hirschsprung's disease surgery. In a well neonate with left-sided Hirschsprung's disease, 34 of 40 surgeons favour primary pull-through following bowel decompression with rectal washouts; 35 of 40 surgeons aim to perform definitive surgery at less than 3 months of age, with 17 favouring laparoscopic-assisted Soave–Boley and 15 favouring an open Duhamel pull-through. Of the 40 surgeons, 36 use a staged approach to right-sided/total colonic Hirschsprung's disease with 23 favouring a Duhamel or Long Duhamel pull-through.

CONCLUSIONS The primary pull-through, using an open Duhamel or laparoscopic-assisted Soave–Boley technique, during the first 3 months of life, has become the operative strategy of choice in rectosigmoid Hirschsprung's disease in Britain. Marked variation in practice remains for right-sided Hirschsprung's disease.

KEYWORDS

Hirschsprung disease – Diagnosis – Operative procedures – Laparoscopy – Surgical stomas

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Since the seminal description of Hirschsprung's disease¹ in 1889, there has been a gradual evolution in the surgical management of this condition. Recognition that Hirschsprung's disease arises from functional obstruction in the distal, aganglionic colon led Swenson to advocate resection of this segment rather than previous techniques concentrating on the proximal dilated colon.² Later, Duhamel described a retrorectal anastomosis³ and Soave an extramucosal dissection,⁴ to minimise risk of neurovascular injury. Primary endorectal pull-through without enterostomy⁵ has gained in popularity since first described in 1980, being further modified to include minimally invasive approaches.⁶

In 1998, Huddart undertook a national survey to determine 'contemporary' management of Hirschsprung's disease.⁷ The most popular operative strategy in a well neonate was to perform surgery over three stages, with preliminary defunction-

ing stoma followed by definitive pull-through at 6–9 months of age, then stoma closure. Wide variations in preferred management strategies lead to recommendations for regional subspecialisation with dedicated 'Hirschsprung's disease surgeons' performing definitive surgery. Recently published series suggest earlier definitive surgery, over fewer stages⁸ with increased utilisation of laparoscopic^{9,10} and transanal approaches.^{11,12} This survey was designed to determine the prevalence of current management strategies for Hirschsprung's disease in the UK and Ireland, a decade after the last survey.

Subjects and Methods

Following approval by the British Association of Paediatric Surgeons (BAPS) Research and Clinical Outcomes

Committee, email addresses for all practising consultant paediatric surgeons in the UK and Ireland were obtained. A questionnaire was constructed to re-assess the findings of the previous survey⁷ and was validated by two independent consultant paediatric surgeons. Initial questions pertained to individual surgeons' clinical experience in Hirschsprung's disease and service provision at their centre. Subsequent questions focused on management of Hirschsprung's disease in specific clinical scenarios: (i) well neonates with left-sided or right sided/total colonic Hirschsprung's disease; and (ii) Hirschsprung's disease complicated by late presentation, Down's syndrome or enterocolitis. Respondents were asked to select answers from a series of multiple-choice options. Free-text boxes were provided to allow alternative responses. Emails were sent to all consultant paediatric surgeons containing an electronic link to an online version of the survey (QuestionPro™; Seattle, WA, USA). A second round of emails was sent 6 weeks later with the questionnaire attached. Data were analysed using descriptive statistics (mean, median, frequency, percentage) in pre-defined subgroups according to the options for each question.

Results

Emails were sent to 142 consultant paediatric surgeons, 85 consultants initiated the survey, 21 were excluded (18 incomplete, 2 duplicate responses, 1 consultant working overseas), yielding 64 surveys for analysis. The majority of respondents included an institutional email address confirming representation from at least 21 of 26 tertiary paediatric

centres. Of 64 respondents, 47 work in centres with designated 'Hirschsprung's disease surgeons', 55 are involved in routine management of patients with Hirschsprung's disease but only 40 perform definitive surgery. Further questions targeted the 40 surgeons who routinely perform definitive surgery for Hirschsprung's disease. Median number of pull-through procedures performed per consultant per year was 5 (range, 1.5–16) with 18 surgeons 'often' or 'always' operating with another consultant colleague.

Pre-operative management in the well neonate

Suction rectal biopsy alone is used by 21 of 40 respondents to diagnose Hirschsprung's disease. A further 15 surgeons combine this with contrast enema. Transrectal full-thickness rectal biopsies are used by four surgeons. No respondents use diagnostic anorectal manometry in this age group. Histological diagnosis of aganglionosis is considered mandatory by 39 of 40 consultants before performing definitive surgery in a neonate.

Operative strategies

In a well neonate with left-sided Hirschsprung's disease, 34 of 40 surgeons perform primary pull-through following rectal washouts to decompress the bowel with 28 of 40 allowing wash-outs at home. A staged approach of stoma followed by definitive surgery is utilised by six surgeons. Definitive surgery is planned at less than 3 months of age by 35 of 40 surgeons and 26 of 40 would operate on a neonate weighing less than 4 kg or once birth weight is regained. Nine operative strategies and two operative techniques

Table 1 Management of a well neonate with Hirschsprung's disease: a comparison of contemporary management with the last survey of UK practice in 1998

| Parameter | Huddart ⁷ <i>n</i> (%) | Bradnock <i>n</i> (%) |
|---|--------------------------------------|--------------------------|
| Total number of respondents performing Hirschsprung's disease surgery | 63 (85) | 40 (63) |
| Operate on infant with Hirschsprung's disease at < 3 months of age | 29 (46) | 35 (88) |
| Primary pull-through (well neonate) | 26 (41) | 34 (85) |
| Two-stage pull-through (well neonate) | 20 (32) | 5 (13) |
| Three-stage pull-through (well neonate) | 14 (22) | 1 (3) |
| Primary Soave–Boley pull-through | 12 (19) | 18 (45) |
| Primary Duhamel pull-through | 10 (16) | 15 (38) |
| Primary Swenson pull-through | 3 (5) | 0 |
| Staged Duhamel pull-through | 28 (44) | 3 (8) |
| Staged Soave–Boley pull-through | 3 (5) | 3 (8) |
| Staged Swenson pull-through | 2 (3) | 0 |
| Laparoscopy as preferred approach | 2 (3) | 18 (45) |

were reported. The Soave–Boley and Duhamel techniques were favoured by 22 of 40 and 18 of 40 surgeons, respectively. Laparoscopy was utilised by 18 of 40 surgeons in definitive surgery. The majority of surgeons (17/18) using the Duhamel technique employed an open approach compared to three of 22 surgeons performing the Soave–Boley technique. Pure transanal pull-throughs are performed by three surgeons. Routine postoperative anal calibration is performed by 15 of 40 surgeons at a median time of 18 days (range, 10–42 days).

In a well neonate with right-sided or total colonic Hirschsprung's disease, 35 of 40 surgeons adopt a staged approach. Fourteen operative strategies and seven operative techniques were reported. Duhamel pull-through is preferred by 25 of 40 consultants and a further eight of 40 advocate a Lester–Martin (Long Duhamel) technique. Soave–Boley pull-through is employed by two surgeons. A further four operative techniques were reported: one surgeon uses a 'J-pouch' for total colonic aganglionosis and a 'modified Soave' for right-sided disease; one uses a 'Kimura patch'; one performs a 'Shandling–Lester–Martin patch with Soave pull-through'; and one stated that his choice of operation would 'depend on level and progress with a stoma'. A laparoscopic-assisted approach is used by four surgeons, with 36 of 40 adopting an open approach for right-sided or total colonic Hirschsprung's disease.

Special circumstances

When Hirschsprung's disease is complicated pre-operatively by enterocolitis, 19 of 34 surgeons change from a primary to staged procedure. Five of 18 surgeons change from laparoscopic-assisted to open approach. When performing definitive surgery in a neonate with Down's syndrome, seven of 34 surgeons change from primary to staged procedure. Fewer surgeons (4/40) change operative technique. In late-presenting children, 16 of 34 surgeons change from primary to staged procedure. A change in technique is favoured by 10 of 40 surgeons with some stating an alternative operative technique in this scenario depending on the clinical situation and the degree of bowel wall thickening and dilatation.

Trends in the management of Hirschsprung's disease since 1998

A number of changes in management strategy for well neonates with Hirschsprung's disease have occurred over the last 11 years (Table 1). The percentage of surgeons prepared to perform pull-through on infants less than 3 months of age has increased from 46% to 88%. Primary pull-through for left-sided colonic Hirschsprung's disease is routinely performed by 85% of surgeons compared to 41% in 1998. The popularity of the Soave–Boley procedure and use of laparoscopic-assistance have also increased.

Discussion

National surveys of practice provide historical reference points with which to compare contemporary practice. The presented survey, like those conducted in US,^{15,14} Japan^{15,16} and Australia,¹⁷ are limited by an inability to relate practice to clinical outcome, but do identify trends in management, highlight local and national variations in practice, promote discussion regarding the optimal approach to managing Hirschsprung's disease and suggest future areas of research.

In 1998, Huddart recommended subspecialisation within units to maximise individual consultant experience in operative management of Hirschsprung's disease.⁷ In this survey, 47 of 64 (73%) respondents work in centres with designated 'Hirschsprung's disease consultants'. The poor response rate prevents robust conclusions regarding subspecialisation. However, despite an expansion of consultant paediatric surgeon numbers from 90 to 142 over the last decade, the mean number of cases per consultant per year has not fallen providing indirect evidence for increasing subspecialisation. A number of respondents routinely perform definitive surgery with another consultant colleague, further increasing individual consultant exposure. Increasing volume of surgery and specialisation has been shown to improve patient outcomes¹⁸ and there remains a compelling case for further subspecialisation for definitive surgery in Hirschsprung's disease.

Our study highlights changes in the diagnostic modalities for Hirschsprung's disease. In 1975–1976, 70% of US paediatric surgeons that insisted on histological confirmation of aganglionosis used transrectal full-thickness rectal biopsy.¹⁴ In our study, 35 of 40 surgeons use suction rectal biopsy. Suction rectal biopsy allows early definitive diagnosis of Hirschsprung's disease leading to a 90% neonatal diagnosis rate¹⁷ compared to 15% in 1975.¹⁴ Unlike series from other developed countries,¹⁶ anorectal manometry does not play a role in the diagnosis of neonatal Hirschsprung's disease in Britain, perhaps reflecting concerns regarding its accuracy in this age group.¹⁹

In common with other studies,^{15,15,16} a transition from staged approach to wide acceptance of primary pull-through for left-sided Hirschsprung's disease has occurred in the UK over the last decade, with only six of 40 surgeons routinely performing stomas in the well neonate. Surgeons are more comfortable operating on cases at a younger age and smaller body weight and there has been convergence in the management of left-sided colonic Hirschsprung's disease into two predominant operative strategies: primary Soave–Boley and Duhamel techniques. In the US, there has been a move away from the Duhamel pull-through.¹⁵ We cannot ascertain from our data whether the continued popularity of the Duhamel pull-through in the UK is region-

specific or whether surgeons within the same institution routinely employ different approaches. Continued exposure to this technique is potentially useful as it is a popular approach in revisional surgery,²⁰ and most respondents in this survey utilise this technique in right sided/total colonic disease.

Despite evidence of improved consensus regarding the management of left-sided Hirschsprung's disease, the operative strategies employed in right-sided and total colonic Hirschsprung's disease remain variable. The most popular strategies were staged, open Duhamel and Lester–Martin (Long Duhamel) procedures, but a further five operative techniques and 15 alternative operative strategies were described. There is little comparative data in the literature regarding the management of right-sided or total colonic aganglionosis, as previous studies have not analysed operative strategies according to the level of aganglionosis.^{15–17}

In 1998, 10 of 65 surgeons stated they would 'consider' using laparoscopy in Hirschsprung's disease.⁷ Eighteen of 40 surgeons now use laparoscopy for colonic mobilisation or biopsies. The increased popularity of the Soave–Boley technique may be related to an acceptance of laparoscopic (and transanal) approaches. Similar trends toward avoiding a laparotomy wound have been reported elsewhere.^{15,16} Where these studies differ, however, is in the proportion of surgeons utilising a purely transanal approach to rectosigmoid Hirschsprung's disease, with 29% of surgeons in Japan¹⁶ and 38% in the US¹⁵ employing an exclusively transanal approach, compared to three of 40 surgeons in the UK.

The relative impact of complicating factors such as late-presentation, history of Hirschsprung's enterocolitis or Down's syndrome in determining operative strategy was difficult to assess, as respondents stated that the degree of bowel wall thickening or dilatation would ultimately determine the technique they would adopt. Despite this, it appears that a history of Hirschsprung's enterocolitis would cause many surgeons to change from primary pull-through to staged procedure and nearly a third to change from laparoscopic to open approach.

Study limitations

The limitations of this study are those inherent in all questionnaire-based surveys. Mindful of the risk of 'survey fatigue' and poor response rate, we used an online survey system to streamline the process of responding. Overall, we attained a 45% (64/142) response rate, which is equivalent to the response rates obtained by two major US surveys.^{15,14} It is possible that surgeons who chose not to respond do not have a specific interest in this subject because they are not involved in definitive surgery. The respondents in our study perform a cumulative total of 212 pull-through procedures per year although the actual number is likely to be lower

given that 18 of 40 responding surgeons 'often' or 'always' perform the surgery with another consultant colleague resulting in potential duplication. Using an estimated incidence for Hirschsprung's disease of 1 in 5000 live births,²¹ approximately 175 neonates with Hirschsprung's disease are born each year in the UK. Accordingly, we feel that the findings of this study are representative of the current management of Hirschsprung's disease in the UK and Ireland.

Conclusions

Primary pull-through, performed either with an open or laparoscopic-assisted approach during the first 3 months of life, has become the operative strategy of choice for rectosigmoid Hirschsprung's disease in the UK. The use of laparoscopy for colonic mobilisation and biopsies is increasing and the Soave–Boley procedure has doubled in popularity. An open, staged Duhamel procedure is now the procedure of choice for the management of right-sided and total colonic Hirschsprung's disease, although wide variations in practice persist within this group. Although there is some evidence of subspecialisation with designated Hirschsprung's disease surgeons in the majority of units, a further reduction in the number of surgeons performing the definitive surgery in Hirschsprung's disease is needed to maximise the number of cases performed by 'Hirschsprung's disease specialists'. There is a need for prospective, long-term, follow-up studies to assess whether these changes in practice lead to improved outcomes.

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