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ORIGINAL ARTICLE

Management of Hirschsprung disease in Australia and New Zealand: a survey of the Australian and New Zealand Association of Paediatric Surgeons (ANZAPS)

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Abstract

Purpose To define the practice of management for Hirschsprung disease (HD) in Australia and New Zealand.
Methods Online survey of Australian and New Zealand Association of Paediatric Surgeons (ANZAPS) members.
Results 56/80 (70%) members from 17 centres responded.
Diagnosis 100% perform suction rectal biopsies; 40% perform a contrast enema.
Histopathological staining H&E (94%), ACHE (70%) and calretinin (75%).
Surgery Primary pull-through (PT) is performed by 88% (100% by < 6/12 months). The Soave–Boley PT is the preferred approach (85%), with laparoscopic assistance (77%) and muscle cuff division (93%). Routine post-operative dilatations are performed by 63% of respondents. If symptoms persist following PT, majority adopt a conservative approach (enemas/laxatives 90%; Botox 74%). If a long-segment is identified at PT, 60% fashion a stoma and delay definitive surgery. If total colonic aganglionosis is identified at PT, 76% fashion a stoma and delay definitive surgery. A dedicated bowel management program is available in 45% of centres with transition to adult services in 29%.
Conclusions A laparoscopic-assisted Soave–Boley PT is the most common technique for recto-sigmoid HD. Differences are noted in both the management of long-segment/total aganglionosis HD and post-operative management/follow-up.

Keywords Hirschsprung disease · Paediatric surgery survey · Diagnosis · Pull-through procedure · Post-operative management

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Abbreviations

HD	Hirschsprung disease
PT	Pull-through
ANZ	Australian and New Zealand
ANZAPS	Australian and New Zealand Association of Paediatric Surgeons

Introduction

Hirschsprung disease (HD), or congenital megacolon, was first reported by Fredericus Ruysch, a Dutch anatomist in Amsterdam in 1691, but it was in 1886 that Harald Hirschsprung presented the first detailed description of the disease at the Society of Pediatrics in Berlin [1]. Although Hirschsprung published his findings in 1888, the pathogenesis of the disease remained unclear for a long time and it was many years later in 1920 that Dalla Valla described the histological finding of absent ganglion cells in biopsies of affected children [2, 3].

Likewise, the optimal surgical management remained uncertain and inadequate for many years until Swenson and Bill in 1948 advocated the resection of the distal non-dilated colonic segment, rather than the proximal dilated colon [4]. Later, Duhamel described a retro-rectal and trans-anal anastomosis, whilst Soave described an extra-mucosal dissection (subsequently modified by Boley) to minimise the possible risks of neurovascular injury associated with the Swenson procedure [5–7]. These techniques, despite having undergone various modifications over the years, are still widely utilised.

However, many controversies remain regarding most aspects of the management of HD, which have been highlighted by surveys conducted in the last 30 years in the USA, UK, Japan and, more recently, Europe [8–14]. These surveys have documented a significant evolution in the surgical management of HD, with a trend towards a one-stage procedure (without the formation of a stoma), performed in the first few weeks of life, using a minimally invasive (laparoscopic-assisted) approach. Nevertheless, they also have highlighted wide differences in practice among different centres, especially in USA and Europe [10, 14]. Singh et al. in their 2003 publication from the Australian Paediatric Surveillance Unit documented that the Soave–Boley procedure was the most common (64%) procedure performed in Australia [12]. However, this study was not specifically focused on the surgical aspects. Besides, a dedicated survey on HD management has never been conducted in Australia and New Zealand (ANZ); therefore, this was the aim of our study.

Methods

Following approval by the Australian and New Zealand Association of Paediatric Surgeons (ANZAPS) Executive Committee, members (qualified paediatric surgeons) were contacted electronically. This email contained a link to an anonymous online survey (Qualtrics™, Provo, Utah, USA) regarding the management of HD. A second round of emails was sent 6 weeks later as a reminder. Members were asked to select answers from a series of multiple-choice options, and free-text boxes were provided for comments.

The questionnaire was developed to assess the current management of HD in ANZ. We focused on the following six domains: (1) individual surgeons' clinical experience in managing HD and the service provision at their centre; (2) diagnosis and pre-operative workup; (3) operative techniques employed; (4) management of HD diagnosed in the neonatal period; (5) management of HD complicated by late presentation and trisomy 21; (6) post-operative management and follow-up protocols. The survey contained 54 questions (full questionnaire is supplied as electronic supplementary material). Data were analysed using descriptive statistics and reported as frequency (percentage) for each question.

Results

The electronic survey was sent to 80 active ANZAPS members, with a response received by 56 (70%). This represented 17 centres with dedicated paediatric surgeons across ANZ. The majority (80%) of respondents had more than 5 years' experience as a consultant (5–10 years, 20%; 10–20 years, 26%; > 20 years, 34%). 80% were trained in ANZ. Eight (47%) centres managed more than 10 HD cases per year, whilst two (12%) managed more than 20 HD cases per year. Five centres (29%) have surgeons that are sub-specialised in the management of HD.

Diagnosis

All of the surveyed surgeons perform suction rectal biopsies (SRB) for HD diagnosis (three biopsies—90%, two biopsies—10%). Sixteen (29%) of these will consider a SRB adequate for the diagnosis in patients aged over 1 year. A full thickness rectal biopsy is taken at different distances from the anal verge: 1–2 cm (35%), 2–3 cm (53%), and 3–4 cm (12%). Histopathological staining that is available in each centre includes (Fig. 1); haematoxylin and eosin (H&E) (94%), acetylcholinesterase (ACHE) (70%) or calretinin (75%). The majority (96%) of surgeons obtain results within 72 h (30% in < 24 h). Forty percent of surgeons perform a contrast enema.

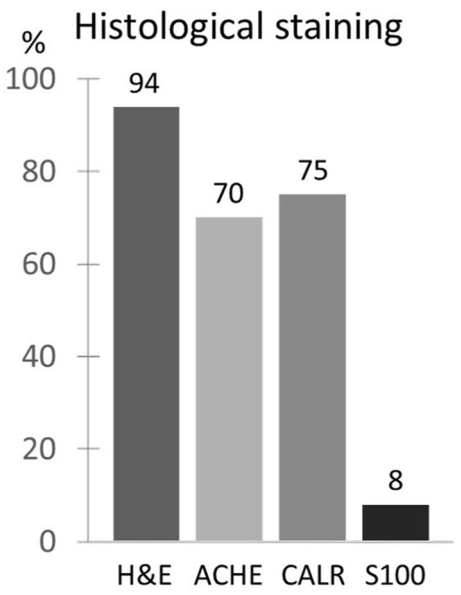

Fig. 1 Histological staining available in different centres. *H&E* hematoxylin and eosin, *ACHE* acetylcholinesterase, *CAL* calretinin

Surgery

In a clinically stable neonate with recto-sigmoid HD, a primary pull-through (PT) is performed by 49/56 (88%). This is performed < 2 months of age in 82% and 100% by 6 months of age. A delayed PT with primary neonatal stoma formation is performed by 7/56 (12%). The Soave–Boley trans-anal PT is the preferred approach (48/56, 85%), with laparoscopic assistance preferred by 37/48 (77%) (Fig. 2A). Muscle cuff division is commonly performed 45/48 (93%). The anastomosis is performed at a variable distance from the dentate line: <1 cm (40%), 1 cm (36%), and 1–2 cm (24%). A total of 27/48 (56%) of surgeons will send a complete circumferential intestinal sample ("donut") for frozen section analysis before performing the final anastomosis.

If a long-segment (more proximal than recto-sigmoid) HD is unexpectedly identified intraoperatively during the PT, 34/56 (61%) of surgeons will fashion a stoma (levelling colostomy or ileostomy) and delay surgery (Fig. 2B). If total colonic aganglionosis is unexpectedly identified intraoperatively during the PT, 43/56 (76%) will fashion a stoma and delay definitive surgery until the patient is at least 3 months old (Fig. 2C). Currently, different operative approaches are utilised in patients with long-segment HD or total colonic aganglionosis: Soave–Boley (56%), Swenson (37%) or Duhamel (7%). A J-pouch is performed by 9/56 (16%) of respondents in patients with total colonic aganglionosis. In patients with trisomy 21, 11/56 (20%) will delay the primary PT with 15/56 (27%) fashioning a stoma in the interim period.

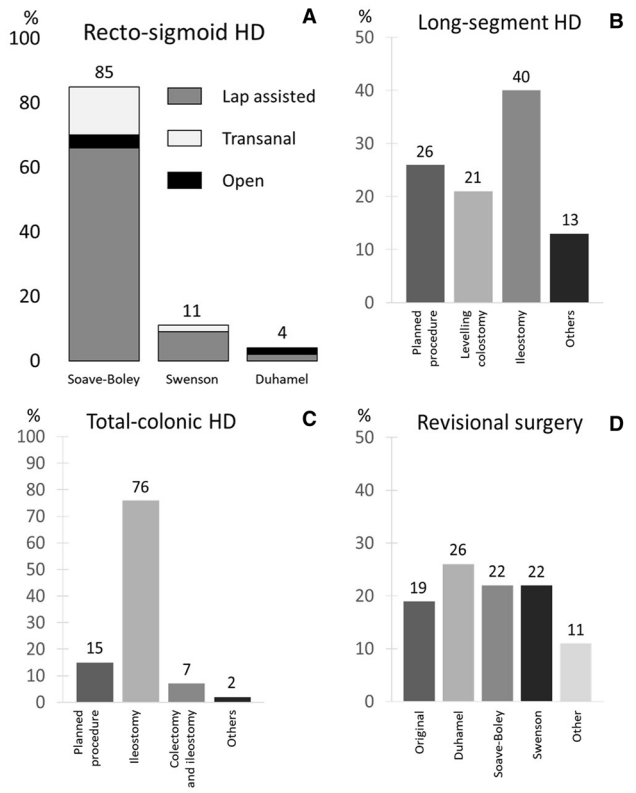

Fig. 2 Distribution of responses on the preferred surgical procedure for different segment length and for revisional surgery. *HD* Hirschsprung disease

Revisional surgery

In those patients requiring revisional surgery, the following procedures are performed: Duhamel (26%), Soave–Boley (22%), Swenson (22%), same as the original intervention (19%) and others (11%) (Fig. 2D).

Late presentation

For patients presenting with symptoms at an older age (beyond the neonatal period), surgeons will utilise the following diagnostic investigations: full thickness rectal biopsy (90%), SRB (10%), contrast enema (31%) and anorectal manometry (5%).

In the absence of a megarectum, 40% of surgeons will perform a primary PT and 60% fashion a stoma. Of those that fashion a stoma: 56% form a colostomy and perform a delayed PT, 17% a colostomy and concurrent PT, 17% an ileostomy and delayed PT, and 10% an ileostomy with concurrent PT.

In the presence of a megarectum, 91% will perform a stoma (85% colostomy, 15% ileostomy) with a delayed PT. Types of these delayed PTs include either a Soave–Boley (76%), Swenson (14%), or a Duhamel (10%).

Post-operative management and follow-up

A routine post-operative digital rectal examination is performed by 64% of surgeons following surgery (70% at 2 weeks, 30% at 3 weeks). Routine post-operative anastomotic dilatations are utilised by 63% of surgeons, with 95% initiating these in the outpatient with parents continuing at home. The frequency of these dilatations varies, with 54% of surgeons performing once-daily and 28% twice-daily dilatations. Routine dilatations are continued for a variable time period: < 1 month (5%), 1–2 months (16%), 2–6 months (44%), or 6–12 months (17%). However, 18% of surgeons will choose the duration of the dilatations based on the individual patient response. If symptoms persist following a PT, the majority will initially adopt a conservative approach with minimal interventions (enemas/laxatives 90%; botulinum toxin injection 74%). In patients with recurrent Hirschsprung-associated enterocolitis following surgery, the preferred management options include regular washouts (21%), regular dilatations (19%), botulinum toxin injection (17%), oral probiotics (15%), prophylactic metronidazole

(13%), prophylactic oral vancomycin (3%), cycling oral metronidazole and vancomycin (7%), or a combination of these different managements (5%).

A dedicated bowel management program is available in 8/17 (47%) centres. Scheduled follow-up is in place in 9/17 (53%) centres, whilst standardised transition to adult services occurs in 5/17 (29%) centres. A multi-disciplinary post-operative approach is routinely utilised by 18/56 (32%) of surgeon respondents.

Discussion

The results of our survey show that the laparoscopic-assisted Soave–Boley PT, performed in the neonatal period, is the most common technique utilised in ANZ. This highlights a main difference to other published surveys that have demonstrated wide variations in practice among different centres, especially in USA and Europe [10, 14]. One possible explanation is that 80% of respondents in our survey were trained in ANZ. Therefore, the surgeon population is significantly homogenous in our setting. Furthermore, we only included consultant/attending paediatric surgeons, with 80% of these having had more than 5 years of clinical experience. One additional strength of our survey is that we had a high response rate (70%).

We found relative standardisation in the way rectal biopsies are obtained and processed. There has been an increase in the usage of calretinin staining in ANZ, as it is now available in 75% of centres. This is compared to a calretinin staining rate of 31% in Europe [14]. Only 40% of the surgeons who responded will perform a contrast enema, this is similar to the UK-reported practice (38%) [8], but significantly less common when compared to Europe (96%) [14].

A primary laparoscopic-assisted trans-anal Soave–Boley PT, with muscle cuff division, is performed by the majority of surgeons. Only 11% of surgeons will perform a Swenson procedure and 4% of surgeons a Duhamel procedure. Because of the anonymous nature of the questionnaire, we were not able to clarify if the surgeons that perform the Swenson or Duhamel procedure are among the 20% trained outside ANZ. Although speculative, it is debateable if the selection of a specific technique over others is simply related to geographical aspects or could be somehow related to the fact that it provides a better outcome. It will be, therefore, interesting to clarify if this standardisation of the surgical technique in ANZ has also led to similar outcomes across different paediatric surgical institutions. We do acknowledge that without objective data this is difficult to prove. Our survey also revealed differences in the management of patients with a long-segment/total colonic aganglionosis. The majority of surgeons will fashion a stoma and delay the definitive surgery. Although a Swenson PT is advocated by more surgeons (37%) in this setting, the Duhamel PT remains rarely utilised (7%). However, 33% of surgeons will consider the Duhamel PT appropriate for revisional surgery.

Unique to our survey is that we also investigated aspects related to the post-operative follow-up and the management of patients with a late presentation of HD. Although a full thickness rectal biopsy is advocated by most of surgeons (90%) for HD diagnosis, we identified differences in the management of children in relation to stoma formation before the definitive PT. However, the majority of surgeons will fashion a stoma in patients presenting beyond the neonatal period with a megarectum. The rational for this practice is to reduce the complications related to anastomosis dehiscence, as these have been reported to be higher in children with late diagnosis of HD [15]. Also, many of these patients may have significant co-morbidities such as malnutrition or chronic anaemia, which influence the surgical intervention decision pathway [16]. Data regarding the management of patient with HD disease presenting after childhood are limited in the literature. In their systematic review, Doodnath and Puri reported that 47.2% were treated with a Duhamel PT [17]. However, this review did not provide details regarding the use of a stoma or the geographical distribution of the surgeons. We have found that in ANZ, the laparoscopic-assisted trans-anal Soave–Boley PT remains a popular procedure, even in children with late presentation HD and megarectum. Large series from low- and middle-income countries have confirmed that the Soave–Boley PT remains feasible in late-presenting HD children, providing that the megarectum has been treated with adequate decompression [16].

We do acknowledge that, like other surveys, ours provides quantitative and qualitative data which offer insights into the attitudes and opinions of the current management of HD in ANZ, rather than objective data. One interesting aspect of surveys in general is the ability to identify changes in practice when repeated at different points in time. Repeat surveys conducted in either the USA (1979 [11] and 2009 [10]) or UK (1998 [9] and 2011 [8]) have documented a significant evolution in the surgical management of HD, with a trend towards a primary PT (without a stoma formation), performed in the first few weeks of life, with a minimally invasive approach. Singh et al. in their 2003 publication from the Australian Paediatric Surveillance Unit, collected data regarding the surgical management of HD, but their study was not specifically focused on surgically relevant aspects [12]. For example, it did investigate aspects of the diagnosis or specific management of these patients and did not document the role of laparoscopy, although the latter is not surpassing as the technique was only described few years before the survey [18].

Comparison between the data from Singh et al. and our survey has revealed that there has also been an evolution in

the management of HD in the last 15 years in ANZ. There is an increased number of cases being performed as a primary PT in the neonatal period (50% vs 88%) with a Soave–Boley PT (64% vs 85%). We hope that the results of our survey, when compared to the results from other Countries, will stimulate future discussion and possible multicentre collaborations to standardise the management of HD across different geographical areas.

Finally, we found differences among surgeons in the postoperative management and follow-up of HD patients. Particularly, the management of patients with persisting symptoms following surgery, such as Hirschsprung-associated enterocolitis, remains a controversial topic. This is likely related to the fact that the evidence surrounding strategies, including regular anal dilatations, botulin toxin injection, and oral probiotics, remains anecdotal [19].

Conclusions

We report the first survey dedicated to the management of HD in ANZ. The laparoscopic-assisted Soave-Boley transanal PT is the most common technique performed for both recto-sigmoid HD diagnosed in the neonatal period, and for children presenting later. Differences are noted in the management of patients with long-segment/total colonic HD, patients requiring revisional surgery, as well as postoperative management and follow-up.

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Compliance with ethical standards

Conflict of interest None of the authors has a conflict of interest to declare.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

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