

Hirschsprung's Disease in Barbados

A 16-year Review

MV Vincent, SU Jackman

ABSTRACT

Objective: To assess the surgical outcome of patients with Hirschsprung's disease (HD) who were treated in Barbados, establish referral patterns and identify factors which can lead to better management.

Design and Methods: All patients with HD who had definitive surgery at the Queen Elizabeth Hospital, Barbados, over a 16-year period (between July 1991 and June 2007) were identified from the hospital records system. Data from patients' notes were collected to establish demographics, including age at referral and definitive surgery, gender, presenting symptoms, surgical intervention(s) and definitive procedure. Long term outcome was assessed by the use of a simple questionnaire which was completed by the authors after personal or telephone contact with patients, their parents or guardians or the referring physician.

Results: Between July 1991 and June 2007, 27 children were identified with HD. There were 20 males and seven females. Fifteen of the 27 children (55 %) were local Barbadians, who were referred earlier when compared to those from the surrounding Caribbean islands.

The most common mode of presentation was chronic constipation (13/27; 48%). The majority (69%) of the children were fully continent in the long-term (mean follow-up of 64 months) and passed a bowel motion on an average of 2–3 times per day without the use of laxatives or enemas.

Conclusion: Children who had definitive surgery for HD in Barbados (using Martin's modified Duhamel procedure) had an overall good long-term outcome comparable to international results. Children referred from the surrounding Caribbean islands were referred at an older age, but their overall outcome was not worse than local Barbadian patients.

La Enfermedad de Hirschsprung en Barbados

Un Estudio de 16 Años

MV Vincent, SU Jackman

RESUMEN

Objetivo: Evaluar el resultado quirúrgico de pacientes con la enfermedad de Hirschsprung (HD) que fueron tratados en Barbados, establecer patrones para las remisiones, e identificar factores que puedan conducir a un mejor tratamiento.

Diseño y Métodos: Todos los pacientes con HD que tuvieron cirugía definitiva en el Hospital Queen Elizabeth, Barbados, por un periodo de 16 años (entre julio de 1991 y junio de 2007) fueron identificados en el sistema de archivos del hospital. Se recogieron datos de las notas de los pacientes para determinar la demografía, incluyendo la edad al momento de la remisión y de la cirugía definitiva, el género, los síntomas que presentaban, las intervenciones (quirúrgicas) y el procedimiento definitivo. El resultado a largo plazo fue evaluado mediante un cuestionario simple respondido por los autores luego de un contacto personal o telefónico con los pacientes, por los padres o guardianes de estos últimos, o el médico que hizo la remisión.

Resultados: Entre julio de 1991 y junio de 2007, se identificaron 27 niños con HD. Veinte eran varones y siete hembras. Quince de los 27 niños (55%) eran Barbadenses locales que fueran remitidos con

anterioridad luego que se les comparara con los que provenían de las islas caribeñas circundantes. El modo más común de presentarse la enfermedad fue el estreñimiento crónico (13/27; 48%). La mayoría (69%) de los niños fueron totalmente continentes a largo plazo (seguimiento promedio de 64 meses) y movían los intestinos un promedio de 2–3 veces al día sin el uso de laxantes o enemas.

Conclusión: Los niños que tuvieron cirugía definitiva para HD en Barbados (usando el procedimiento de Duhamel-Martin) tuvieron una buena evolución clínica general a largo plazo, comparable a los resultados internacionales. Los niños remitidos de las islas caribeñas circundantes fueron remitidos a una edad mayor, pero su resultado global no fue peor que el de los pacientes locales de Barbados.

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INTRODUCTION

Constipation is a common problem in children throughout the world, and the Caribbean is no exception. The challenge for healthcare providers throughout the islands of the Caribbean, especially in the smaller islands, is to identify children who do not have primary constipation, but are constipated from treatable underlying medical or surgical conditions, for example Hirschsprung's disease (HD). It is then imperative that these children be referred as early as possible for specialist care, and once referred appropriately managed. This study outlines the referral patterns to one centre in the Caribbean (The Queen Elizabeth Hospital, Barbados) which offers patients definitive surgery for HD and reports on their long term outcome.

SUBJECTS AND METHODS

All patients with Hirschsprung's disease (HD) who had definitive surgery at the said institution over a 16-year period (between July 1991 and June 2007) were identified from hospital records. Data from patients' notes were collected to establish demographics, including age at referral and at definitive surgery, gender, presenting symptoms, presence of any associated anomalies, family history, surgical intervention(s) and definitive procedure. The long-term outcome of all patients without total colonic HD who had the same definitive procedure (Martin's modified Duhamel procedure) by the same surgeon was then assessed.

Long term outcome was assessed by the use of a simple questionnaire completed by the junior author (MVV) after personal or telephone contact with patients, their parents or guardians or the referring physician. None of the questionnaires were completed by the operating surgeon (SUJ). The questionnaire sought to assess the following: faecal continence [Wingspread classification] (1, 2), presence of constipation, use of laxatives or enemas and school or work attendance.

RESULTS

Over a 16-year period, between July 1991 and June 2007, 27 children were identified with Hirschsprung's disease (HD), including twenty males and seven females – [male:female = 3:1]. Fifteen of the 27 children (55 %) were local Barbadians, the others were referred from St Kitts (3), Antigua and Barbuda (3), St Vincent (2) and one each from the British Virgin Islands (BVI), Grenada, St Lucia and St Maarten.

The most common mode of presentation was chronic constipation [14/27; 52%] (Table 1). The mean age at pre-

Table 1: Mode of presentation

Mode of presentation	Number of patients (%)
Chronic constipation	14 (52%)
Bilious vomiting	6 (22%)
Non-bilious vomiting and abdominal distention	6 (22%)
Abdominal pain	1 (4%)

sentation was 30 months (ranging from day 1 of life to 24 years). Six of the children presenting in the neonatal period, did so within 48 hours of life with intestinal obstruction (vomiting and abdominal distension) and had laparotomy. At laparotomy, these children had frozen section confirmation of aganglionosis prior to stoma formation + sigmoidectomy or descending colectomy. The majority of children presented in the neonatal period (10), the rest being referred during infancy (7), childhood (8) and adulthood (1). All the children who presented in the neonatal period with one exception were local Barbadians, with children referred from outside Barbados tending to be referred at later ages (Table 2). The

Table 2: Age at referral, showing referring country

Country of origin/referral	Age at referral
Barbados (15)	Day 1(4), Day 2 (4), Day 3 (1), 2 months, 3 months, 4 months, 7 months, 13 months, 32 months
Antigua & Barbuda (3)	Day 1/ 44 months/ 17 years
St Kitts (3)	22 months/ 25 months/ 24 years
St Vincent (2)	9 months/ 33 months
BVI (1)	8 yrs
Grenada (1)	5 months
St Lucia (1)	9 months
St Maarten (1)	31 months

*BVI = British Virgin Islands

mean age at presentation for children in Barbados was four months (range from day one of life to 32 months) compared with a mean of 62 months (ranging from day one of life to 24 years) for referrals outside Barbados. A history of delayed passage of meconium was not documented in nine of the

children but was noted to be present in 13 children (13/27, 48%) and absent in five.

There were no associated chromosomal anomalies, however one child had congenital auricular atresia. Three of the children had a positive family history of HD. Two children had cousins with HD and one child had an older sibling with the condition.

Based on the inclusion criteria outlined above, children were excluded from long-term follow-up, leaving 16 (16/27, 59%) for assessment. The mean time to follow-up was 64 months (ranging from two to 163 months).

The 11 children excluded from long-term follow-up included two with total colonic HD – one of whom had definitive surgery elsewhere, three who were lost to follow-up, one child whose parents declined to take part in the study, one child awaiting definitive surgery, one child who had a stoma at the time of study and three who did not have a Martin's modified Duhamel procedure as definitive surgery for HD. These latter children had the following procedures:

- One child had a Soave pull-through procedure for presumed total colonic Hirschsprung's disease on the basis of frozen section analysis (which was later not substantiated on paraffin section)
- One patient who presented from St Kitts at the age of 24 years had a rectal myotomy
- One child who had an anorectal myectomy

Of the 16 children assessed for long term outcome for continence using Wingspread classification (1, 2) 11 (69%) were fully continent (Grade 1), two experienced staining (Grade 11), one soiled at times (Grade 111) and two soiled all the time [Grade 1V] (Table 3).

Table 3: Long-term continence assessment using Wingspread classification (1, 2)

Wingspread classification	Number of children of 16 assessed (%)
Grade 1 (fully continent)	11 (69%)
Grade 11 (staining)	2 (12.5%)
Grade 111 (soils at times)	1 (6%)
Grade 1V (soils all the time)	2 (12.5%)

The child who required a stoma was one of three children in the series who suffered from chronic constipation following that pull-through procedure. This child had persistent constipation and abdominal distention unresponsive to medical management, and would most likely necessitate a repeat pull-through procedure in the future. The other two children were able to maintain regular bowel habits with the use of regular laxatives (1) and enemas (1). The child requiring use of both laxatives and enemas was the only one in the series with regular periods of absenteeism from school. These two children were 26 months and 38 months at the time of assessment and were 38 months and 22 months post definitive surgery.

The two children with Grade 1V continence were both developmentally delayed, one child having cerebral palsy with a perinatal history of maternal pregnancy-induced hypertension and fetal distress at birth and the other having congenital hydrocephalus. None of the children experienced diarrhoea on long term follow-up.

DISCUSSION

The majority of patients with Hirschsprung's disease (HD) have a satisfactory outcome after definitive surgery (3). However, despite advances in surgical procedures, some patients continue to have signs of persistent bowel dysfunction even in the presence of adequate resection of the aganglionic segment. The aetiology of the persistent bowel dysfunction in these patients remains controversial, but is noted to improve with age, in particular fecal incontinence (4). In the study group of patients, the majority were fully continent (Wingspread Group 1) at a mean follow-up of 64 months. Only two children of the 16 assessed soiled all the time [Wingspread Group 1V] (Table 3). These two children were well within the mean time to follow-up for this group of patients (64 months) and in addition both were developmentally delayed and so the reason for their incontinence may be multi-fold.

The surgical treatment of HD has evolved over the past 20 years from traditional multi-stage procedures to one-stage pull-through techniques (5–8), and more recently to laparoscopic-assisted approaches (9, 10). There is ample evidence that the primary pull-through procedure for HD is safe and effective in most situations, including the neonatal period (8). The rate of complications for single stage procedures is comparable to that seen in patients having a preliminary stoma but without the associated added complications of stomas. Hence it is claimed that the single-stage repair is ideal for use in developing countries where access to medical care may be hampered by poor transportation and communication systems (8). Since the first report of totally trans-anal surgery by de La Torre *et al* (7) however, there has been few published reports to assess the feasibility and/or safety of these newer techniques in developing countries (11). But how safe and feasible are these newer techniques for developing countries like the Caribbean?

While transportation and poor communication systems may indeed be a challenge for the Caribbean, especially in the presence of multiple small islands, a number of factors make routine use of the primary pull-through procedure inappropriate for the Caribbean, if not impossible. These include the limited number of paediatric surgical specialists and support staff and lack of facilities and equipment for the handling of frozen section specimens as well as paediatric pathologists trained to read and interpret frozen section specimens. False-positive intra-operative frozen sections reported in centres where experienced pathologists are unavailable can have serious consequences (12). Our insti-

tution had one experience of having had a patient as outlined above who had a total colectomy (and Soave pull-through procedure) for presumed total colonic HD based on frozen section analysis which was later not substantiated on paraffin section. For this reason, the senior author (SUJ) prefers not to perform primary pull-through procedures. In addition, as evident in this study, a significant number of children were diagnosed with HD outside the neonatal period, and by the time of referral have well-established megarectum or megacolon, which are contraindications to the primary pull-through procedure (9). None of the children in this series had a primary pull-through procedure.

Somme (8) has claimed that the following factors have all contributed to earlier diagnosis of HD, which has resulted in increasing interest in performing a one-stage procedure over the last several decades: greater awareness of HD, improved neonatal nursing care with specialized neonatal intensive care units, the use of suction rectal biopsy and a larger number of paediatric pathologists. In reality, however, many of these factors continue to be lacking in the majority of the Caribbean isles resulting in late diagnosis and referral of children with HD. The mere fact, for example, that for a third of patients in this study, there was no clear documentation of whether or not the child had delayed passage of meconium, a factor present in up to 94% of patients, especially newborns to pre-schoolers with HD (13–15) is testament to the need for greater education among healthcare professionals about constipation in children and HD. However, many factors outlined by Somme (8) which will affect earlier diagnosis and referral have improved and will continue to improve throughout the Caribbean. For example, awareness of HD will improve with increased educational and training opportunities created both locally and abroad. With respect to Somme's (8) finding of using a rectal suction biopsy as a factor contributing to earlier diagnosis, we note the delay in diagnosis of some of the children who presented in the neonatal period (Table 4). The recently acquired rectal

Table 4: Patients presenting in the neonatal period, showing country of origin/referral and age at diagnosis

Referring country	Age at presentation	Age at diagnosis
Barbados	Day 1	Day 2
Barbados	Day 1	Day 3
Barbados	Day 1	Day 8
Antigua	Day 1	Day 18
Barbados	Day 1	4 months
Barbados	Day 2	Day 2
Barbados	Day 2	Day 3
Barbados	Day 2	Day 6
Barbados	Day 2	35 months
Barbados	Day 3	Day 17

suction biopsy at the Queen Elizabeth Hospital ought to help in early diagnosis, but also eliminates the need for an open rectal biopsy, which has to be performed under a general

anaesthesia unlike a rectal suction biopsy which can be performed without general anaesthesia.

Bruce *et al* (16) have reported on their preliminary experience with the use of the rectal suction biopsy some 10 years ago and have recommended its use in the Caribbean setting. During the review of patients' records in this study, it was clear that a number of children who initially presented in the neonatal period and re-presented later in childhood were being treated for necrotizing enterocolitis (NEC) in the neonatal period when most likely they were experiencing Hirschsprung's enterocolitis. It is a known fact that the clinical features of NEC – foul-smelling, explosive diarrhoea, often bloody in nature, abdominal distention, hypovolaemic shock, vomiting, fever and lethargy may mimic that of Hirschsprung's enterocolitis. Hence NEC in the term infant should raise one's suspicion and a rectal suction biopsy performed to exclude HD (17).

In conclusion, Hirschsprung's disease is a rare cause of constipation and can be successfully managed in Barbados with long-term results comparable to that seen both regionally (18) and internationally (4, 19, 20) despite the availability of limited resources and specialists personnel. However, use of the more modern definitive surgical procedures, most of which are laparoscopically-assisted, single-stage procedures cannot be safely adapted in Barbados at present.

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