Original Article

Access this article online
Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.160403
Quick Response Code:

Clinical outcome and bowel function after surgical treatment in Hirschsprung's disease

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ABSTRACT

Background: Bowel function has been reported to be adversely affected following surgery in cases of Hirschsprung. We retrospectively studied both the clinical outcome and bowel function status following surgery in patients diagnosed with Hirschprung's disease (HD). 161 cases, who underwent pull-through operations for HD in Sheikh Pediatric Tertiary Centre, Mashhad, Iran. The specified time bracket spanned between 2006 and 2011. **Materials and Methods:** Data was extracted from Health Information System with the aim of investigating patients for both short and long-term gastrointestinal (GI) complications after surgery bases in addition to the concurrence of any associated anomalies. Three main procedures were analysed in this respect (Swenson, Duhamel and Soave). **Results:** In a study of 96 (59%) boys and 65 (40.3%) girls, mortality rate was reported to be 15.5% (15 males and 10 females). A considerable majority of almost three fourths were detected with both early and late GI complications after surgery. The latter mainly included constipation (30.8%), incontinence (19.8%), enterocolitis (8%), diarrhea (11%) in a declining order of incidence. Down syndrome and others HD-associated anomalies were detected in 3.7% and 24.3% of cases respectively. **Conclusions:** Constipation and foecal incontinence were the most prevalent postoperative complications, which were reported almost as frequent in other studies. Yet, Enterocolitis, was reported slightly less in prevalence. Also mortality rates were considerably higher, compared to developed nations.

Key words: Bowel function, constipation, foecal incontinence, Hirschsprung's disease

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INTRODUCTION

Harold Hirschsprung introduced Hirschsprung's disease (HD) for the first time in 1888.[1] It is a congenital malformation of the colon, characterised by absent parasympathetic intrinsic ganglion cells in the sub-mucosal and myentric plexus.[1] This is the consequence of the premature arrest occurring in the craniocaudal migration of vagal neural crest cells in the colon between the 5th and 12th gestational weeks when the enteric nervous system normally develops.[2] As to the incidence and prevalence of the disease, HD, a complex hereditary sex-dependant condition, occurs in 1 out of 5000 live born infants,[3,4] with a male to female ratio of 4:1-3:1.[5] The disease is commonly associated with a range of congenital anomalies namely nervous, renal and cardiac malformations, of which Down's syndrome is undoubtedly the most noteworthy.[4]

Gastrointestinal (GI) defects are primarily treated by resecting the colonic segments minus ganglion cells.[6] Surgery can entail both early and late GI complications.[7,8] Much as to the favourable outcome in a considerable number of patients, long-term follow-up studies show the contrary,[9] with some developing constipation, incontinence, enterocolitis and diarrhea, which, to a large extent, can affect patients' lifestyle.[8]

While 48.8% of cases were reported to have been inflicted with at least one complication according to Ekenze *et al.*,[10] long-term soiling and constipation after surgery had reached 10.3% and 21.7% in order, claimed another research team.[11]

It is therefore essential to assess both the bowel function as well as the clinical outcome in patients surgically treated for HD as any abnormality in this respect would adversely affect the quality of life and growth. Given the paucity of literature available, we intended to do so in a study focusing on the above-mentioned variables.

MATERIALS AND METHODS

A total of 290 surgically-treated patients diagnosed with HD in Doctor Sheikh Pediatric Centre were registered to our retrospective study. Data was extracted from Health Information System as our chief source. Patients were all referred to the centre in the North-East of the country between 2006 and 2011.

Three major procedures (Swenson, Duhamel and Soave) were mentioned as the mainstay surgery in our centre, with the former as the most popular.

Initial data included basic demographics, the age at which HD was diagnosed and phone numbers. Having contacted by phone, patients, parents or guardians were recruited for follow-up visits after initial explanations regarding the objectives as well as processes involved during the research. Their informed consent was taken prior to study commencing.

They were also dulely reimbursed for their commuting as well as other related costs by the organizational committee of the investigation. Our team of investigators in charge of follow-up visits included a pediatrician, a pediatric gastro-entrologist and an internist, all blind to the research and objectives. They were investigating the occurance of GI complications (Constipation, Incontinence, Enterocolitis, Diarrhea) on both short and long-term bases in addition to the concurrence of any associated anomalies. Physical findings (height and weight) were also duly recorded in comparison with the measurements at birth. Our chief source of data was medical file retrieved from hospital archives. Given certain inevitable limitations namely long distance, loss of contact and reluctance, 129 patients had to be excluded from our registry.

By definition, normal bowel function was claimed when stool was regular, with neither constipation nor soiling. The status was assessed when there had been prescriptions of neither laxatives nor enema. However, entrocolitis was diagnosed on the basis of frequent watery or almost foul-smelling diarrhea, fever, vomiting and abdominal pain.

The time bracket specified to define early and late complications ranged from weeks to months in the former, and months to a full year in the latter case. The retrieved date were finally fed to SPSS version 11 for analysis.

RESULTS

In our study population, there was a male to female ratio of 1.4:1 (81 boys and 55 girls). Ranging from 18 months and 19 years, the median value pertaining to age was 5 years and 6 months [Table 1]. In our study, 91.2% of children were born full-term. The median birth weight was 3.06 kg and median height was 49.42 cm.

Hirschsprung disease was diagnosed immediately at birth in majority of cases (46.3%), 61.7% up to 6 months and 71.1% in their 1st year of life [Figure 1]. The mean postoperative time in our study was 4 years and 6 months.

As to functional complications, 37 patients (28.3%) were reported problem-free while the number of those affected summed up to a considerable 99 (72.7%), with seventy eight (57.3%) and 93 (68.3%) inflicted early and late in the course following surgery respectively and as defined [Table 2 and 3]. Surgical methods do not seem to have affected bowel function, either early or late. Laxative or enemas were required in a total of 22%, 9.1% continuously and 12.9% occasionally.

Despite the higher incidence of functional complications in HD patients with associated anomalies [Table 4] in comparison with those without

Table 1: Demographics data of the 161 patients with typical HD operated between 2006-2011

Age of patients	Number	Gender	
		Male (%) (*N*)	Female (%) (*N*)
18 months-2 year	18	8.8 (12)	4.4 (6)
3-5 years	71	34.5 (47)	17.6 (24)
6-8 years	25	5.8 (8)	12.5 (17)
9-14 years	20	9.5 (13)	5.1 (7)
15-19 years	2	0.7 (1)	0.7 (1)

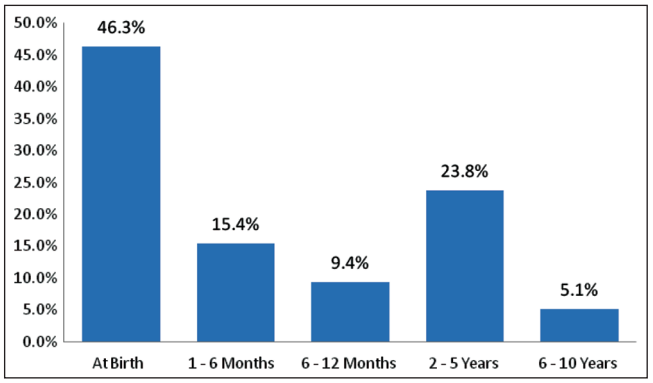

Figure 1: Age of diagnosis Hirschsprung's disease

any accompanying condition, the gap was reported statistically insignificant.

Late bowel function complications were also shown to be independent of the postoperative follow-up time, as there was no statistically substantial disparity between 2 and 5-year incidences of such conditions [Table 5].

Associated anomalies

In our study, HD associated anomalies or chromosomal abnormality was in 24.3% of patients, being 4 (2.9%) congenital heart disease, 12 (8.9%) respiratory disorder, 5 (3.7%) congenital anomalies of the kidney, 4 (2.9%) thyroid disorders, 1 (0.7%) Noonan syndrome and 2 (1.5%) urinary tract infection, also 5 patients (3.7%) (4 male and 1 female) were detected with Down's syndrome or tirosomy 21.

Table 2: Late complications comparison between operative

Complications	Swenson *N* = 86 (63%)	Soave *N* = 31 (22%)	Duhamel *N* = 19 (13%)	Total *N* = 136 (%)
Constipation	42 (48.8)	12 (38.7)	6 (31.5)	60 (44.1)
Incontinence	20 (23.2)	7 (22.5)	4 (21.0)	31 (22.7)
Enterocolitis	6 (6.9)	3 (9.6)	1 (5.2)	10 (7)
Diarrhea	14 (16.2)	4 (12.9)	5 (26.3)	23 (16.9)

Table 3: Early complications comparison between operative

Complications	Swenson *N* = 86 (63%)	Soave *N* = 31 (22%)	Duhamel *N* = 19 (13%)	Total *N* = 136 (%)
Constipation	30 (34.8)	8 (25.8)	4 (21.0)	42 (30.8)
Incontinence	17 (19.7)	6 (19.3)	4 (21.0)	27 (19.7)
Enterocolitis	7 (8.1)	2 (6.4)	2 (10.5)	11 (8)
Diarrhea	9 (10.4)	4 (9.3)	3 (15.7)	16 (11.7)

N: Number; *P* value > 0.05

Table 4: Compare of complication after surgery patients with associated anomalies versus patients without anomalies

Gastrointestinal complications	with associated anomalies**N* (%)	without associated anomalies**N* (%)
Constipation	9 (27)	33 (32)
Incontinence	6 (18)	21 (20)
Enterocolitis	3 (9)	8 (7)
Diarrhea	4 (12)	12 (11)

Tables 5: Compare of late complication on the base of postoperative time

Age Groups (year)	Late functional complication (%)
1-2	67.5
5-14	42.3

DISCUSSION

As could be seen in the results, constipation and fecal incontinence were the most prevalent postoperative complications, with no statistically significant difference when accompanied by associated anomalies. Nevertheless, percentages differed in early and late presentations.

Constipation occurred early after operation in %44.1 of our cases whereas it was reported in slightly above a third of patients later throughout our follow-up. Similar studies varied in their percentages, ranging from 8% to 60% of cases.[5-13]

Were not shown to have influenced the incidence of complication surgical methods ($P > 0.05$). This is also corroborated by Menezes *et al.*[11] in the year 2006, who reported complication rates of 29.4% for soave, 8.3% for Swenson and 24% for Duhamel ($P > 0.05$). Niramis *et al.*[14] rates were 8.5%, 7.1% and 10.4% in order.

Mills *et al.*[7] came up with an 8% whereas Rescorla *et al*. claimed the same percentage for those with either constipation or soiling.[15]

Marty *et al*. were slightly lower in their results as 7.5% of cases presented with this complaint late after surgery. Moderate to severe constipation was as low as 12.8% of a total of 107 patients over a 22-year period in a study by Yanchar and Soucy.[16]

Incontinence was almost half as common in our cases, both late and early, while it inflicted 18% and 20% of HD patients with or without associated anomalies in order.

Likewise, incontinence rates fluctuated widely in different study findings (10-80%) yet others reported this complication as high as 58% (Catto-Smith (5) and 49% (Mills *et al.*),[7] or considerably lower (10.3% in Menezes *et al*. study[11] and 12.6% in Marty *et al.*[17]

Our findings included almost twice as many cases affected by enterocolitis (3 and 8 patients with and without associated anomalies). However, this complication was shown substantially broad in incidence owing to differences in definitions by authors. Some reported it as high as 32% and 26% (Hackam *et al.*[18] and Sarioglu *et al.*[19] while Yanchar and Soucy[16] claimed a mere 9.3%). Others, results varied between 20% and 58%[20] whereas Suita *et al.*[21] were more moderate in their findings (20-58%). Niramis *et al*. Another plausible explanation for lower incidence rates

lies in the fact that enterocolitis is often fatal, with a mortality rate of up to 30%.[17-22] We failed to investigate the cause of death in our cases yet Marty *et al*.[17] rates summed up to 13.5%.

Laxative and enema had to be administered in 22% of our cases (9.1% daily and 12.9% occasionally), much akin to Marty *et al*. percentage (21%) and other studies. Yet, Rescorla *et al*.[15] and Sarioglu *et al*.[19] were higher in their figures (27%).

Functional problems often persist years after surgery in HD patients whereas they may improve significantly with age, according to some authors.[1-25]

We investigated them in our research and arrived at a 56.3% incidence in <5 years after surgery, and 56.6% between 1 and 14 years postoperative. Rescorla *et al*.[15] were far higher in their incidence (88% in a 5-year follow-up). Yanker claimed that 58% and 88% of their foecal incontinence cases fully recovered in <5 and 15 years respectively.[23]

This had risen to 100% of cases with full recovery over 15 years of follow-up in Niramis *et al*. study.[14] We could not establish any correlation between recovery and age, possibly due to relatively short follow-up. Extension can probably reveal more in the future.

Hirschsprung disease is currently diagnosed in a vast majority of neonates (90% as reported by an Australian team)[20] thanks to raised public awareness as well as advanced diagnostic methods.[20-24]

Nonetheless, only 47% of our cases were detected during their neonatal period while the rate rose to 71.1% when the time bracket was extended to the 1st year of life. Similar results had been achieved in another study on 420 patients over a 20-year interval (1981-2000), where 50% of patients were diagnosed <1 month of age whereas 73% of them were below 1-year.[26] Japanese were even slightly lower in their rates (40% in neonatal period in a study on 3852 patient between 1978 and 2002).[21] We cannot precisely indentify the cause of delay in diagnosis but a safe assumption could be milder signs and symptoms in older patients, which may alter the presentation of HD. Nevertheless, the precision in diagnosis chiefly depends on parental awareness, diagnostic methods and intensity of signs and symptoms.

Hirschsprung disease is said to occur rarely in both premature as well as full-term infants. Suita *et al*. reported a varying incidence rate of between 5.5% and 10.4% in neonates under 2500 g[21] whereas Japanese did not exceed the bottom rate in this respect.[22] Our study included 90.9% full-term patients, with one-fifth of cases below 2500 g of weight, which may be accounted for by mothers' low socioeconomic status.

Mortality rates also differed widely in investigations, ranging from 1% to 10% in developed and developing nations. Menezes *et al*. rates (3.5%) were half as much as Natalies' (6.6%) whereas Wildhabers' rose to 12%.[16-27] Rates even rose to as high as 23.8% and 16% in Nigeria and Bandre respectively.[28] In comparison, we were close to the latter in our rates, though this can be due to surgical techniques, postoperative care and associated anomalies. Death has chiefly been attributed to enterocolitis by authors. Nevertheless, we investigated neither the cause of death nor surgical techniques in our study, which can be further probed in the future. There were only few patients who died in the hospital after the operation.

Our findings were consistent with other studies in the rates pertaining to associated anomalies. They were reported between 5% and 32%, with Down syndrome being on top of the list (2%-10% of HD cases). We detected such conditions in 24.3% of our cases; with Trisomy 21 comprising 3.7% of patients. Japanese HD patients who had diagnosed with associated anomalies were almost half as many (11.1%)[22] whereas Australian was slightly lower (26.2%).[20] In as much as the fact that there is no consistent pattern pertaining to anomalies, cardiac and renal defects were reported to have comprised 5% and 44% of HD cases. Ours were lower, with 2.9% and 3.7% in order.

Despite HD following a multi-factorial inheritance with sex-dependent penetration (reported by a study with a male/female ratio of 4:1 or 3:1), we found an M/F ratio of 1.4/1 (59.6% male and 40.3% female). Mills showed a male predominance of 4-1.[7] So did Single (3.8-1) and Tehran (3-1). The almost equal ratio in our cases should be investigated in terms of genetic factors and other influential parameters.

Our study was restricted in that the average follow-up summed up to 5 years and 6 month whereas long-term complications resolution may take years further. We also failed to investigate the cause and the time of death. Given our high mortality rate, this will be of paramount significance.

Given our findings as compared to others, surgery as the only treatment modality in Hirshprung's disease is inevitably followed by early and late complications. Aside from enterocolitis, which was reported to be slightly lower, other after-effects were almost within the average range claimed by other research teams. Nevertheless, our mortality rates were considerably higher, compared to developed nations. Thus, it is suggested that future research should focus on the underlying causes in this respect as this had not been an objective in this study, yet a number of potential causes, seem more likely to have raised mortality rates in our region.

REFERENCES

1. Amiel J, Lyonnet S. Hirschsprung disease, associated syndromes, and genetics: A review. J Med Genet 2001;38:729-39.
2. Benish BM. Letter: "The neurocristopathies: A unifying concept of disease arising in neural crest development". Hum Pathol 1975;6:128.
3. Dasgupta R, Langer JC. Evaluation and management of persistent problems after surgery for Hirschsprung disease in a child. J Pediatr Gastroenterol Nutr 2008;46:13-9.
4. Askarpour S, Samimi K. Epidemiologic and clinical specifications of patients with Hirschsprung's disease in Khouzestan province Iran. J Surg Pak (Int) 2008;13: 75.
5. Haricharan RN, Georgeson KE, editors. Hirschsprung Disease. Seminars in Pediatric Surgery.University of Alabama at Birmingham, Alabama, Elsevier; 2008.
6. Amiel J, Sproat-Emison E, Garcia-Barcelo M, Lantieri F, Burzynski G, Borrego S, *et al*. Hirschsprung disease, associated syndromes and genetics: A review. J Med Genet 2008;45:1-14.
7. Mills JL, Konkin DE, Milner R, Penner JG, Langer M, Webber EM. Long-term bowel function and quality of life in children with Hirschsprung's disease. J Pediatr Surg 2008;43:899-905.
8. Baillie CT, Kenny SE, Rintala RJ, Booth JM, Lloyd DA. Long-term outcome and colonic motility after the Duhamel procedure for Hirschsprung's disease. J Pediatr Surg 1999;34:325-9.
9. Engum SA, Grosfeld JL, editors. Long-term results of treatment of Hirschsprung's disease. Seminars in Pediatric Surgery. Indiana University Medical Center. Elsevier; 2004.
10. Ekenze SO, Ngaikedi C, Obasi AA. Problems and outcome of Hirschsprung's disease presenting after 1 year of age in a developing country. World J Surg 2011;35:22-6.
11. Menezes M, Corbally M, Puri P. Long-term results of bowel function after treatment for Hirschsprung's disease: A 29-year review. Pediatr Surg Int 2006;22:987-90.
12. Diseth TH, Bjørnland K, Nøvik TS, Emblem R. Bowel function, mental health, and psychosocial function in adolescents with Hirschsprung's disease. Arch Dis Child 1997;76:100-6.
13. Athanasakos E, Starling J, Ross F, Nunn K, Cass D. An example of psychological adjustment in chronic illness: Hirschsprung's disease. Pediatr Surg Int 2006;22:319-25.
14. Niramis R, Watanatittan S, Anuntkosol M, Buranakijcharoen V, Rattanasuwan T, Tongsin A, *et al*. Quality of life of patients with Hirschsprung's disease at 5-20 years post pull-through operations. Eur J Pediatr Surg 2008;18:38-43.
15. Rescorla FJ, Morrison AM, Engles D, West KW, Grosfeld JL. Hirschsprung's disease. Evaluation of mortality and long-term function in 260 cases. Arch Surg 1992;127:934-41.
16. Yanchar NL, Soucy P. Long-term outcome after Hirschsprung's disease: patients' perspectives. J Pediatr Surg 1999;34:1152-60.
17. Marty TL, Seo T, Matlak ME, Sullivan JJ, Black RE, Johnson DG. Gastrointestinal function after surgical correction of Hirschsprung's disease: long-term follow-up in 135 patients. J Pediatr Surg 1995;30:655-8.
18. Hackam DJ, Filler RM, Pearl RH. Enterocolitis after the surgical treatment of Hirschsprung's disease: Risk factors and financial impact. J Pediatr Surg 1998;33:830-3.
19. Sarioglu A, Tanyel FC, Büyükpamukçu N, Hiçsönmez A. Clinical risk factors of hirschsprung-associated enterocolitis. II: Postoperative enterocolitis. Turk J Pediatr 1997;39:91-8.
20. Singh SJ, Croaker GD, Manglick P, Wong CL, Athanasakos H, Elliott E, *et al*. Hirschsprung's disease: The Australian Paediatric Surveillance Unit's experience. Pediatr Surg Int 2003;19:247-50.
21. Suita S, Taguchi T, Ieiri S, Nakatsuji T. Hirschsprung's disease in Japan: analysis of 3852 patients based on a nationwide survey in 30 years. J Pediatr Surg 2005;40:197-201; discussion 201.
22. Ikeda K, Goto S. Diagnosis and treatment of Hirschsprung's disease in Japan. An analysis of 1628 patients. Ann Surg 1984;199:400-5.
23. Catto-Smith AG, Coffey CM, Nolan TM, Hutson JM. Fecal incontinence after the surgical treatment of Hirschsprung disease. J Pediatr 1995;127:954-7.
24. Puri P, Montedonico S. Hirschsprung's Disease: clinical Features. Hirschsprung's Disease and Allied Disorders. Springer; 2008.
25. Hackam DJ, Reblock KK, Redlinger RE, Barksdale EM Jr. Diagnosis and outcome of Hirschsprung's disease: Does age really matter? Pediatr Surg Int 2004;20:319-22.
26. Kouranloo J, Sadeghian N, Monfared MK. Treatment and postoperative complication of 420 patients with congenital megacolon. Saudi Med J 2003;24 Suppl:S25-8.
27. Wildhaber BE, Teitelbaum DH, Coran AG. Total colonic Hirschsprung's disease: A 28-year experience. J Pediatr Surg 2005;40:203-6.
28. Nasir A, Adeniran J, Abdur-Rahman L. Hirschsprung's disease: 8 years experience in a Nigerian teaching hospital. J Indian Assoc Pediatr Surg 2007;12: 68-71.

Cite this article as: Khazdouz M, Sezavar M, Imani B, Akhavan H, Babapour A, Khademi G. Clinical outcome and bowel function after surgical treatment in Hirschsprung's disease. Afr J Paediatr Surg 2015;12:143-7.

Source of Support: Nil. **Conflict of Interest:** None declared.