# A RESEARCH ON HIRSCHSPRUNG'S DISEASE IN AMIRKABIR AND BAHRAMI CHILDREN HOSPITALS

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***Abstract:*** ***Hirschsprung's disease is a developmental disorder of the enteric nervous system, characterized by absence of ganglion cell in the myenteric and submucosal plexuses along a variable portion of the distal intestine.***

***The purpose of this study is the presentation of a new procedure for reduction of the postoperative complications of Hirschsprung's disease (HD) by offering a modified Swenson's technique. This was achieved through a 10 years study of 157 patients with HD, of whom 74% were male and 26% were female. In 100 cases the disease was diagnosed in the early neonatal period, in 129 cases the involved segments were rectosigmoid or shortsegment. Our choice of operation was a new modification of Swenson's type of pull through (Swenson's operation +posterior rectal wall myectomy). Pathological examination has confirmed the diagnosis of HD in all patients. The overall results of surgery were good in 81% and there was no mortality.***

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***Key words:*** ***Hirschsprung's disease, new modification of Swenson's technique, results of treatment.***

## INTRODUCTION

The incidence of congenital aganglionosis or Hirschsprung's disease is one per 4400-7000 live births (1,2). The disease occurs in all races, although its incidence is lower in blacks. The incidence of the disease follows a familial pattern. It afflicts 1.5% to 17.6% of the children in families already involved with a disease which is respectively 130 and 360 folds higher for males and females in comparison to the incidence in general population (1,2,3). In general the incidence ratio of males to females is 4/1. To date the etiopathology of the disease is unknown. The old hypothesis which states that, migration blockage of primary neuroblasts in a part of GIT, responsible for impairing the function of intestine from that point on, has not been ruled out.

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Nevertheless just recently some histochemical disorder, and genetic (RET proto-oncogen, EDN3…) and micro-environmental factors have been proposed as possible causes in the pathogenesis of aganglionosis (1-7).

Hirshsprung's disease has been seen from newborn to adolescence ages although diagnosis is established in newborn in 90% of cases. The chief complaint is defecation disorder, which in newborns is manifested by delayed passage of meconium for more than 48 hours, intestinal obstruction, or even perforation of colon (1-3). Intestinal perforation occurs mostly in cecum and then in transverse portion of splenic flexure. In older patients (infants and children) the chief complaint is chronic constipation and its related problems. In some cases with history of chronic constipation the patient comes down with severe and explosive diarrhea, abdominal distention, fever, and bad general condition. These symptoms indicate that the worst consequence of the disease, enterocolitis, has occurred which leads to death in 30-40% of cases unless the appropriate treatment is performed immediately (1,2,8), the most prevalent area involved is the recto-sigmoid region (75-80%), although more extensive forms, involving even the entire colon, have also been reported.

Between the region containing ganglions and the aganglionic, there exists an intermediate region of hypoganglionsis termed as transitional zone (TZ), determining that TZ is of critical importance for performing colostomy and definitive surgery. The aganglionic region sometimes is very short and includes just the terminal portion of the large intestine (2-3,5,9). Definitive diagnosis is established by patients' symptoms, plain abdominal film, barium enema, rectal manometry, suction rectal biopsy, full-thickness rectal biopsy 2cm above the dentate line, and histochemical staining. Once the diagnosis is established, colostomy is performed just proximal to TZ, and then, depending on the growth of the infant, in 6-9 months of age the definitve operation is performed to eliminate the aganglionic region and with the establishment of anastomosis between the ganglionic colon and the point 1.5-2 cm from the end of colon, the treatment comes to its end. Swenson, Soave, and Duhamel techniques are among the most

popular and routinely utilized surgical techniques all over the world. Various surgeons have used these techniques with over 80% desirable results. In ultra-short cases Lynn technique is indicated. Today, some centers recommend laparoscopy or surgical operation on newborn for definitive treatment of Hirschsprung's disease (1-2).

Postoperative complications include: anastomotic leak, anastomotic stenosis, persistent constipation, fecal incontinence, fecal soiling, anastomostic disruption, peritonitis, intra abdominal abscess, enterocutaneous fistula, wound infection, etc. According to various statistics, these complications occur in 5 to 20% of cases (1-3,5,10). In the 10 years experience, the author has assessed the treatment results obtained by him and three other attending surgeons. In this research, the authors studied the records of hospitalized and operated cases at Amirkabir hospital (first 7 years) and Bahrami hospitals (last 3 years) during the 1990s.

A considerable number of patients over 2 years of age with chronic constipation who had undergone Lynn operation and also other patients after colostomy but with negative pathological result for HD have been excluded from the study. The inclusion cases were 157 patients, out of whom 116 (74%) were male and 41 (26%) female. This study was conducted upon the results gained from the contents of the files as well as post operative follow-ups from 6 month to 4 years (2.5 years average) of the patients. All of the patients had undergone definitive surgery.

Our surgical choice of approach was proximal to TZ colostomy and then definitive surgery mostly with modified Swenson technique in classical cases, other techniques were utilized when necessary. The preferable age for definitive operation was 6 to 9 months.

# RESULTS

Of our 157 cases, 92 patients were hospitalized with symptoms of intestinal obstruction and 8 intestinal perforation. Colostomy was performed for all of them.

Twenty seven patients were diagnosed under the age of 6 months and in 30 cases diagnosis and treatment were established between 6 month and 12 years of age. Clinical manifestation, the length of aganglionic segment and treatment results are shown in tables 1-5.

It bears mentioning that for these 157 patients documented positive pathological report for Hirschsprung's disease exists.

**Table 1.** Clinical manifestations

| From of manifestation | Number |
|---|---|
| Neonatal intestinal obstruction | 92 |
| Colon perforation | 8 |
| **Chronic constipation (enterocolitis) 6 m. to 12y** | 57 |

**Table 2.** Diagnostic basis

| Diagnostic assay | Number |
|---|---|
| Clinical presentation, barium enema, and open biopsy | 100 |
| Clinical presentation, and barium enema | 22 |
| **Clinical presentation, barium enema and rectal biopsy** | 35 |

**Table 3.** Surgical techniques

| Utilized surgical technique | Number |
|---|---|
| Modified Swenson technique | 109 |
| Lynn operation | 32 |
| Soave's technique | 7 |
| Kimura's technique | 5 |
| **Martin's technique** | 3 |

**Table 4.** Length of the involved segment

| The involved segment | Number |
|---|---|
| Recto-sigmoid | 99 |
| Short segment | 37 |
| Long segment | 20 |
| **Total colonic aganglionosis** | 8 |

**Table 5.** Treatment results

| Obtained result | number |
|---|---|
| Good | 127 |
| Relatively good | 20 |
| Poor | 10 |
| **Mortality** | 0 |

# DISUSSION

Hirschsprung's disease is a relatively common disease in pediatric surgery. In spite of significant developments in embryology, etiology, and pathology of the disease, little is known about Hirschsprung's disease. The disease is a developmental disorder of neural system of the intestine and is determind by the lack of ganglion cells in myenteric and sub-mucosal plexus in a variable length of the lower portions of the intestine. The most agreeable etio-pathological hypothesis for its onset is blockage of neuroblasts' cranio-caudal migration from crest, which starts from vagus nerve at 5th week of embryonic life and reachs the rectum at 12th week (1-3). Treatment techniques of Hisrschs-prung's disease have been vastly modified in the scond half of 20th century and utilized for curing the disease. This multiplicity of treatment

techniques indicates that neither of these procedures can rule out the others and skillful and experienced surgeon utilizing the available technique can achieve optimal results. Also it must be kept in mind that with all of the approaches utilized, there is a percentage (5-20%) of undesirable results such as persistent constipation, fecal incontinence, fecal soiling and the most rankling possibility: enterocolitis (1-2,10). In this study the authors investigated manifestations of the disease, extent of aganglionic area, surgical technique utilized, pathological results and the overall outcome together with consideration of the diagnostic procedures. The results of treatment has been classified in 3 groups.

1. Good: reestablishment of normal defecation, normal growth on follow-ups, with no adverse consequences.

2. Fair: relatively good intestinal defecation, subsequent enterocolitis and intestinal obstruction due to adhesion bands, with repeated operation, or wound infection in some cases, eventually re-establishment of normal stature.

3. Poor: persistent constipation, fecal incontinence, fecal soiling, recurrent enterocolitis leading to repeated surgical operations.

For 10 years the four surgeons at our centers used same surgical technique cases of classical Hirschsprung's disease.

Our technique of choice was added to Swenson technique to minimize its post-operative constipation and other undesirable consequence. In short or ultra-short cases, we used Lynn technique. Also other techniques such as Soave, Kimura, and Martin were utilized when necessary. In 109 patients out of all 157 cases, the chosen technique was modified Swenson. The result of its use was excellent.

Most of the patients were diagnosed at infancy and under 6 months of age and the diagnosis was based on clinical symptoms, Barium enema and definitive surgery were performed at the age of 6-9 months. Pathological proof of HD for all 157 cases exists.

Average postoperative follow up was 2.5 years. The results obtained were good in 81%, fair in 12.5%, and poor in 6.5%, which are consistent with world-wide statistics, (1-3,10). Observing the considerable post operative complications of different surgical techniques in treatment of Hirschsprung's disease the authors recommend their modified technique as a procedure of choice.

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