Functional Outcome of Hirchsprung's Disease, Post-Surgical Trans-anal Endorectal Pull-through at National Children's Hospital

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ABSTRACT

Background: Hirschprung's Disease, is a congenital illness secondary to lack of ganglion cells in the intestinal tract leading to mechanical obstruction. In the Philippines, Hirschsprung's Disease ranks 9th over the top 10 cases causing morbidity to Filipino children and the mortality rate can reach up to 50%. The treatment is still surgery, such as Soave procedure.

Design: Descriptive cross sectional study

Subjects: The participants of the study were follow up patients at the Surgery Out Patient Department from March 2016 to August 2017, ages 0-7 years old, diagnosed with Hirschsprung's Disease, post Trans-anal endorectal pull-through.

Methodology: Purposive sampling was used to select participants. Sample size was 40 based on the proportion of good functional outcome among patients who underwent trans-anal endorectal pull-through.

Statistical Analysis: Descriptive Analysis using proportion and percentages were used to present the results in all the variables.

Results: Out of the 40, 82 % were diagnosed with the disease as early as the newborn period and 55% of the patients were operated at an age from 1 to 3 years old and about 80% were males. It also showed that post operatively, 60% had normal z scores and 65% of the population still had an abnormal stool. Moreover, the study showed that 95% of the subjects returned to schooling or playing post surgery.

Summary/Conclusion: Comparing it to the 96% result of Dela Merced 2003, this study had 95% of patients who were able to achieve good functional outcome, 4-6 weeks post-surgery.

Hirschsprung's, Endorectal Pull-through, Functional Outcome

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INTRODUCTION

Hirschprung's Disease or aganglionic megacolon, is a congenital illness in which there is a lack of intrinsic nerves (ganglion cells) in the distal segments of the intestinal tract. These abnormal segments produce mechanical obstruction because of failure to relax during peristalsis. (Samuel, 2006).

The incidence of this disease is 1/5000 to 1/10,000 live births, where in males are affected more often (3:1 to 5:1). Based on studies, there is an increased familial incidence in long segment diseases. (Samuel, 2006).

In the Philippines, Hirschsprung's Disease ranks 9th over the top 10 cases causing morbidity to Filipino children and the mortality rate can reach 20 to 50%. (PCMC report, 2013).

Operational intervention is warranted. One of the three surgical procedures (Swenson, Duhamel, Soave (endorectal pull through) is to be performed as standard treatment, the third procedure as the most commonly done.

The use of a primary endorectal pull-through (ERPT) in the management of patients with Hirschsprung's disease represents a significant change from the classic approach to the treatment of this disease. from an article of (Vijaykumar1,2002), because of the simplified nature of this approach and the potential for cost savings, it has been reported that this procedure have shown to be a safe option.

As bowel function is a significant milestone for children to be mastered, studies showed that the functional outcome post pull through is an important determinant, to show the impact of this disease to children physically, socially and psychologically.

As based from Leiri et. Al, (2010), postsurgical treatment of hirschprung's patient creates a great impact and stigma as they continue their development in life. In developed countries, 90% have promising outcomes, regardless of the type of surgery. In developing countries such as in Malaysia and Indonesia, the percent of good outcome is 80%. (Dela merced et. Al, 2003).

The study wishes to determine the functional outcome of National Children's Hospital's patients with Hirschsprung's disease post therapeutic surgery. The researcher deemed it significant to conduct an in-depth investigation regarding the stooling pattern of of patients with Hirschsprung's disease, after surgical end pull through at the said institution, as one of the specialized training hospital in the Philippine setting. Despite its importance and stigma in the lives children, only a few studies on this have been conducted namely, "Hirschsprung's Disease ten year experience at the Philippine Children's Medical Center" (Dela merced et. Al, 2003), which have documented that approximately 75% of their patients post-surgery are doing well and continued living at their fullest. The results of the study will give the institution a baseline information with regards to the said disease and also the current standing of the hospital, to successful rates post surgically.

Statement of the Problem

What is the functional outcome of patients with Hirschprung's Disease, Post Surgical Transanal Enodorectal Pullthrough at National Children's Hospital?

General Objective

To determine the functional outcome of patients with Hirschprung's Disease, Post Surgical Transanal Enodorectal Pullthrough at National Children's Hospital.

Specific Objectives

1. To determine the demographic profile of patients diagnosed with Hirschsprung's Disease as to:

    a. Age
        a1. age of diagnosis
        a2. age of operation
    b. Sex
    c. Nutritional Status

2. To determine the stooling pattern of patients with Hirschsprung's Disease, post surgical transanal endorectal pull through as to:

    a. Frequency
    b. Type of stool base on Bristol chart
    c. Soiling

3. To describe the quality of life of patients with Hirschsprung's Disease, post surgical transanal endorectal pull through 4-6 weeks after surgery.

Significance of the study

The researcher deemed it significant to conduct an in-depth investigation regarding the stooling pattern of patients with Hirschsprung's disease, after transanal endorectal pull through at the National Children's Hospital, as one of the specialized training hospital in the Philippine setting. Despite its importance and stigma in the lives of children, several retrospective studies from other Asian countries have been conducted with regards to this topic and this can be the first prospective study that will be done, locally only a few studies have been conducted namely, at the Philippine Children's Medical Center, which have documented that approximately 96% of their patients post-surgery are doing well and continued living at their fullest. This study may benefit our society by giving our community a glimpse of understanding on how t hese patients can live a normal life, in spite of their condition. If the community will be able to recognize the capabilities of these patients, discrimination will be lessened and their morale can even be uplifted. By seeing them as an inspiration not as a burden, it can highly create an impact not just to the Filipino community but also to other patients experiencing other kinds of diseases. The outcome of this study may help to further strengthen the prognostication of our future patients and we physicians may further assure them that they can live a better life ahead.

Scope and Delimitation

In this study, the researcher deemed it significant to conduct a baseline data with regards to the functional outcome of patients with Hirschsprung's Disease post surgical transanal endorectal pull through. This was important since our institution still has no study with regards to this topic, and very little data can be found within the Philippine literature. Hirschsprung's disease, in spite of its chronicity that creates great impact with the lives of children, physically as well emotionally and psychologically, has good prognosis and outcome (Dela merced et. Al, 2003). In this study, the researcher limited only her objective to the functional outcome of patients with Hirschsprung's disease post surgical transanal endorectal pull through in terms of the demographilc profile and stooling patterns of patients. The population of this study included those patients that were diagnosed with Hirschsprung's Disease, children of 0 -7 yrs old, and those who underwent post surgical transanal endorectal pull through at the National children's Hospital, who followed up at the Surgery Out Patient Department from March 2016 to August 2017.

Definition of Terms

1. **Stooling patterns** – determined by the frequency of stooling, type of stools based from the Bristol chart, and with or without the presence of incontinence.

    a. Bristol chart: type 1-3 as normal and type 4-7 as abnormal

    b. Stool Frequency: considered as normal base from clinical practice and related literature

        i. School aged: 1-2 stool/day
        ii. Infant: 2-5 stools/day

    c. Soiling- leading to intermittent loss of bowel contents, including flatus (gas), liquid stool elements and mucus, or solid feces.

2. **Quality of Life-** defined as the ability of the child to return to schooling and or ability to play or socialize with other children, 4-6 weeks after surgery.

3. **Bristol Chart-** is a medical aid designed to classify the form of human feces into seven categories. It was developed by Dr. Ken Heaton at the University of Bristol and was first published in the Scandinavian Journal of Gastroenterology in 1997. The authors of that paper concluded that the form of the stool is a useful surrogate measure of colon transit time. It remains in use as a research tool to evaluate the effectiveness of treatments for various diseases of the bowel, as well as a clinical communication aid.

Type 1: Separate hard lumps, like nuts (hard to pass)
Type 2: Sausage-shaped, but lumpy
Type 3: Like a sausage but with cracks on its surface
Type 4: Like a sausage or snake, smooth and soft
Type 5: Soft blobs with clear cut edges (passed easily)
Type 6: Fluffy pieces with ragged edges, a mushy stool
Type 7: Watery, no solid pieces, entirely liquid

Types 1 and 2 indicate constipation, with 3 and 4 being the ideal stools (especially the latter), as they are easy to defecate while not containing excess liquid, and 5, 6 and 7 tending towards diarrhea. (Lewis, 1997) However, in patients with Hirschsprung's disease type 4-7 can be considered as normal.

4. **Nutritional status-** using z score for weight for height, defined as no wasting, with wasting, severe stunting, no stunting, with stunting and with severe stunting.

5. **Transanal endorectal pullthrough -** is the removal of diseased rectal mucosa along with resection of the lower bowel, followed by anastomosis of the proximal stump to the anus, to spare the function of the anus. In this study all hirschsprung's disease patients who underwent the said definitive treatment, resulting to the formation of a "neoanus"(new anus), irregardless of with or without colostomy bag usage prior to the pullthrough. Furthermore, these included patients who had completed series of anal dilatation post surgery.

Review of Related Literature

Hirschsprung's disease also called congenital megacolon or congenital aganglionic megacolon, is a developmental disorder of the enteric nervous system, characterized by an absence of ganglion cells in the distal colon resulting in a functional obstruction (Sultan, 2011).

The pathogenesis of Hirschsprung disease results from the absence of enteric neurons within the myenteric and submucosal plexus of the rectum and/or colon. Enteric neurons are derived from the neural crest and migrate caudally with the vagal nerve fibers along the intestine. These ganglion cells arrive in the proximal colon by 8 weeks' gestation and in the rectum by 12 weeks' gestation. Arrest in migration leads to an aganglionic segment. This results in clinical Hirschsprung disease. According to statistics 75% of aganglionic segment are located at the rectosigmoid area, 10% to which all segment of the colon is affected (Marty TL1et.al, 1995).

The definitive diagnosis of Hirschsprung disease rests on histological review of rectal tissue is obtained by suction rectal biopsy or transanal wedge resection. If a suction biopsy is performed, the biopsy is taken 2-2.5 cm above the dentate line on the posterior wall to minimize the risk of perforation. The biopsied specimen is examined for the presence or absence of ganglion cells in the submucous plexus (suction rectal biopsy) or myenteric plexuses (transanal wedge resection).

In the hands of an experienced pathologist, the resulting biopsy and absence of ganglion cells confirm the diagnosis and allow the initiation of treatment. Skip lesions of aganglionosis have been reported in cases of Hirschsprung disease. Histologic findings include the absence of ganglion cells in the myenteric plexus and hypertrophic extrinsic nerve fibers. Acetylcholinesterase staining of the tissue can be performed to assist with the pathologic assessment. Acetylcholinesterase staining identifies the hypertrophy of extrinsic nerves trunks. In short-segment Hirschsprung disease, the diagnosis can be made with a properly placed rectal suction biopsy alone or in combination with anorectal manometry. Acetylcholinesterase staining and calretinin immunostaining may be helpful in making the diagnosis.

Radiologically, barium enema is requested, wherein a pathognomonic sign will be a transition zone, between the normal dilated proximal colon and a smaller caliber obstructed distal colon caused by non-relaxant of aganglionic bowel. Unprepared single-contrast barium enema is done if perforation and enterocolitis are not suspected, an unprepared single-contrast barium enema may help establish the diagnosis by identifying a transition zone between a narrowed aganglionic segment and a dilated and normally innervated segment. The study may also reveal a nondistensible rectum, which is a classic sign of Hirschsprung disease. A transition zone may not be apparent in neonates, because of insufficient time to develop colonic dilation, or in infants who have undergone rectal washouts, examinations, or enemas.

Nearly all children with Hirschsprung disease are diagnosed during the first 2 years of life. Approximately one half of children affected with this disease are diagnosed before they are aged 1 year (Samuel, 2006). A small number of children with Hirschsprung disease are not recognized until much later in childhood or adulthood. During the newborn period, infants affected with Hirschsprung disease may present with abdominal distention, failure of passage of meconium within the first 48 hours of life, and repeated vomiting. A family history of a similar condition is present in about 30% of cases. Nearly one half of all infants with Hirschsprung disease have a history of delayed first passage of meconium (beyond age 36h), and nearly one half of infants with delayed first passage of meconium have Hirschsprung disease. Unlike children experiencing functional constipation, children with Hirschsprung disease rarely experience soiling and overflow incontinence. Children with Hirschsprung disease may be malnourished. Poor nutrition results from the early satiety, abdominal discomfort, and distention associated with chronic constipation. Older infants and children typically present with chronic

constipation. This constipation often is refractory to usual treatment protocols and may require daily enema therapy.

The worldwide frequency is unknown but the represented occurrence is approximately 1:5000-1:7000 per live births, where in males are affected more often (females 4:1). Based on studies there is an increased familial incidence in long segment diseases.

The overall mortality of Hirschsprung enterocolitis is 25-30%, which accounts for almost all of the mortality from Hirschsprung disease. The disease is generally sporadic, although incidence of familial disease has been increasing. Multiple loci appear to be involved, including chromosomes 13q22, 21q22, and 10q. Mutations in the Ret proto-oncogene have been associated with multiple endocrine neoplasia (MEN) 2A or MEN 2B and familial Hirschsprung disease. Other genes associated with Hirschsprung disease include the glial cell-derived neurotrophic factor gene, the endothelin-B receptor gene, and the endothelin-3 gene (Pini, 2011). Hirschsprung disease is strongly associated with Down syndrome; 5-15% of patients with Hirschsprung disease also have trisomy 21. Other associations include Waardenburg syndrome, congenital deafness, malrotation, gastric diverticulum, and intestinal atresia (Padillar, 2003).

Hirschsprung enterocolitis can be a fatal complication of Hirschsprung disease. Enterocolitis typically presents with abdominal pain, fever, foul-smelling and/or bloody diarrhea, as well as vomiting. If not recognized early, enterocolitis may progress to sepsis and transmural intestinal necrosis (Frykman, 2012).

Operational intervention is warranted. In some patients, colostomy is done first for 6 to 12 months then one of the three surgical procedures (Swenson, Duhamel,Soave) is to be performed as standard treatment. The surgical options vary according to the patient's age, mental status, ability to perform activities of daily living, length of the aganglionic segment, degree of colonic dilation, and presence of enterocolitis.

Surgical options include leveling colostomy, which is a colostomy at the level of normal bowel; a staged procedure with placement of a leveled colostomy followed by a pull-through procedure; or a single-stage pull-through procedure. The single-stage pull-through procedure may be performed with laparoscopic, open, or transanal techniques. This procedure can be performed at the time of diagnosis or after the newborn has had rectal irrigations at home and has pass the physiologic nadir. Colostomy followed by pull-through procedure is generally reserved for those patients who present with sepsis due to enterocolitis, massive distention of ganglionic bowel prohibiting pull-through procedure, or are otherwise not medically suitable for the pull-through procedure.

The Soave or endorectal pull-through was introduced by Franco Soave at the Institute G. Gaslini in 1955. Use of this procedure has been conventionally approached with the placement of a decompressing colostomy once the diagnosis is made. This is followed by a definitive pull-through procedure once the child's intestine is decompressed and he or she reaches approximately 10 kg body weight (Langer, 2010). The use of a primary endorectal pull-through (ERPT) in the management of patients with Hirschsprung's disease represents a significant change from the classic approach to the treatment of this disease. The first successful modern report of a primary pullthrough for Hirschsprung disease came from (So et. al, 1980). Subsequently, because of the simplified nature of this approach and the potential for cost savings, it has been reported that this procedure have shown to be a safe option (Vijaykumar1, 2002).

Postoperatively, patients need close follow-up care to assess healing as well as a screen for potential complications (eg, stricture formation). Outpatient dilations may be necessary to alleviate strictures and should be expected in patients who undergo a single-stage pull-through procedure in the newborn period (Zhang, 2005), Postoperative complications may include intermittent fecal soiling and incontinence, anastomotic leak, stricture formation, intestinal obstruction, and enterocolitis.

The outcome in infants and children with Hirschsprung disease is generally quite good. Most children obtain fecal continence and control. However, children with other significant comorbidities, such as major genetic abnormalities, may have lower rates of continence (Vijaykumar et.al, 2002).

Postsurgical treatment of hirschprung's disease in a patient creates a great impact and stigma as they continue their development in life .

From Yanchar NL1, Soucy P. (1999), 64% of patients interpreted having normal stooling habit, 90% of parents were very satisfied with the child's outcome, post therapeutic surgery. According to this journal entitled Long term outcome after Hirschspruung's disease, perspective articles, most children have significant improvement with respect to soiling and that most parents are satisfied with child's outcome and adapt to their functional abilities along with them.

From Terri L Marty et.al (1995), stated that 15 patients over the age of 4 had excellent anorectal function and appeared well adjusted.

From Dasgupta, Roshni*(2008), during the 6-15 months (average of 8-12 weeks), the follow up period of all of them have had postoperative normal bowel movement.

From Arnold G Corana, b (1997), article titled: Long-term stooling patterns in infants undergoing primary endorectal pull-through for Hirschsprung's disease, concluded that transanal endorectal pullthrough in young infant can yield excellent results including normalization of stool frequency and good to excellent level of continence.

From Hashisha et. Al (2004) Transanal one-stage endorectal pull-through for hirschsprung's disease: a muticenter study, concluded that this technique was associated with excellent clinical functional outcome results.

From Reshma Doodnath, Prem Puri et.al (2010), concluded that majority of patients in whom Hirschprung's disease is diagnosed during childhood have normal bowel function after surgery.

From Jessica L.A. Mills, David E. Konkin, Ruth Milner, et.al (2008). Long-term bowel function and quality of life in children with Hirschsprung's disease, stated that fecal soiling improved significantly with age, and this is the strongest predictor of quality of life of patients. One effect of this chronic disease is the clinical impairment in the psychosocial development of the child.

In our country, little data or information has been gathered and recorded regarding the functional outcome of patients post-surgery. One study at the Philippine Children's Medical Center, by Dela merced et. Al (2003). Hirschsprung's Disease ten year experience at the Philippine Children's Medical Center, have documented that approximately 96% of their patients post-surgery had a good survival rate. Furthermore, basing on this study, the age of diagnosis was younger than 30 days in 50% of patients> constipation was the no. 1 complaint followed by abdominal distention. There was also delayed in passage of meconium in 62% if cases. The average time interval for the onset of symptom and diagnosis was 11.5 months, but 71% of case was confirmed within 6 months from onset.

THEORETICAL CONCEPTUAL FRAMEWORK

Figure 1 shows that genetics plays a big risk in the development of Hirschsprung's Disease. The functional outcome post surgical transanal endorectal pull through may be affected by several factors such as the socio demographic profile, by which this may give this study an overview of the prognosis of the disease based on the factor being described.

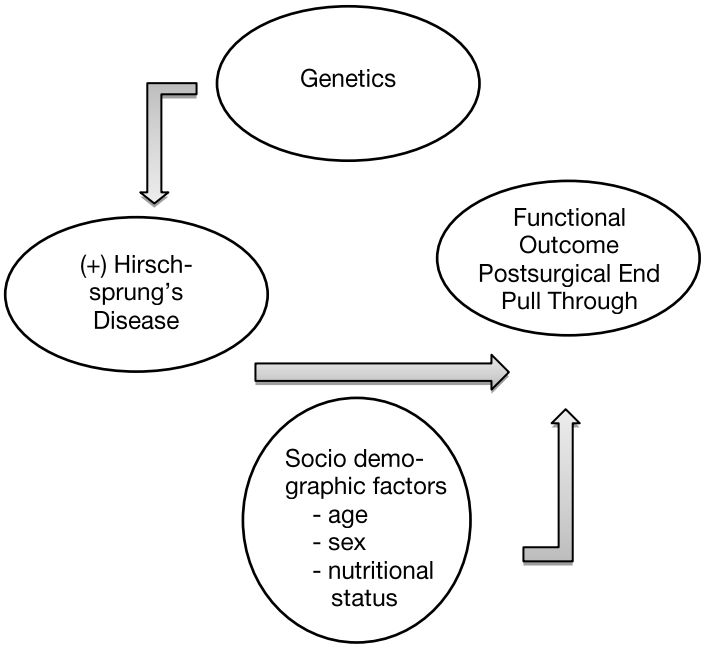

Figure 1: Cause and factors affecting the functional outcome of Hirschsprung's Disease

RESEARCH STUDY DESIGN AND METHODOLOGY

Research Design

A descriptive cross sectional study was conducted by the researchers for this topic.

Sources of Data

The study will be conducted at the Surgery Outpatient Department of the National Children's Hospital, a tertiary pediatric hospital under Department of health.

The participants of the study were patients at the Surgery Out Patient Department from March 2016 to August 2017, who fulfilled the following inclusion criteria:

1. Diagnosed case of Hirschsprung's Disease, s/p Transanal endorectal pullthrough, on follow up.

2. Patients 0-7 y/o who underwent surgical transanal endorectal pull through.

Exclusion Criteria:

1) Those with any associated GI anomalies.

2) Those with any co-morbidities like Down's syndrome, cardiac anomalies, hydrocephalus and other conditions that can alter the quality of living of patients.

3) Severe acute and chronic Malnutrition

Instrument

A patient data collection form was developed based from literature reviews regarding the frequently asked questions post therapeutic surgery, which include: stooling patterns and quality of life of patients.

The variables collected for this study were: age, gender and nutritional status, the frequency of bowel movement, shape and consistency of stools and presence of incontinence.

Furthermore, it also entailed whether the child had returned schooling or at least had the capability to play with other children again.

Procedure

This research protocol was approved by the Institutional Review Board (IRB). The study was conducted at Surgery Outpatient Department of the National Children's Hospital, located at 264 E. Rodriguez Sr. Blvd. Quezon City. The participants who fulfilled the criteria were given a comprehensible consent form, from which they were required to sign.

During the conduct of research, patients' anonymity and confidentiality were ensured. No identified data were out. Each data collection form had a code number, as the basis for data gathering.

Purposive sampling was used to select participants who already underwent end pull through secondary to Hirschsprung's disease. A minimum of 40 participants were considered significant, based on the proportion of good functional outcome among patients with hirschsprung's disease who underwent transanal endorectal pullthrough. Assuming that the proportion of the good functional outcome is 96% (PCMC, Dela Merced et. Al, 2003), with the maximum allowable error of 4% and reliability of 80%.

During follow up, consent form was signed and the physician /researcher used a patient data collection form for data gathering.

Statistical Analysis

Descriptive Analysis using proportion and percentages was used to present the results in all the variables. These included the age, gender and nutritional status, the frequency of bowel movement, shape and consistency of stools and presence of incontinence.

Moreover, it also entailed whether the child has returned schooling or at least had the capability to play with other children again.

Non-probability sample size was used. The sample size computed based on the proportion of postsurgical transanal pull through patients, assuming that the good functional outcome is 96% (from Dela Merced et.al 2003), with maximum allowable error of 4% (plus or minus 4 hence a 92-100% functional outcome for this study) and reliability of 80% (z=1.29), the sample size is 40.

Computation:

N= sample size, z= 80% reliability is 1.29 p= .96 (96% good functional outcome from Dela Merced Study 2003) q=1-p d= maximum allowable error (at 4% or .04)

$N= (z^2 \times p \times q)/ d^2$

N= 40

Description of outcome measurements

The primary outcome was, to describe the stooling patterns of these patients post surgically as well their quality of life.

RESULTS OF THE STUDY

The prevalence rate of patients with Hirschasprungs' Disease post transanal enodrectal pullthorugh at the National Children's Hospital was 31% (40/130) from March 2016 to August 2017. There were a total of 40 patients who fulfilled the inclusion criteria of the study. Out of the 40, 82 % were diagnosed with Hirschsprung's Disease as early as the newborn period, while 18% were diagnosed at the infant phase. Based on the data, 55% of the patients were operated at an age ranging from 1 to 3 years old and about 80% of the population in this study were males. It also showed that patients post operatively had no stunting and no wasting, about 60% of the total number.

However, the study shows that 65% of patients post operatively still had an abnormal type of stool, base on the Bristol chart. The latter, belongs to type 5 which was described as more of watery content of stools. The remaining 35% have type 3 stool, which was described as easy to defecate while not containing excess liquid. The stooling pattern of the included patients basing on its frequency revealed to tally in accordance to their respective age group. Only about 2.5 % had soiling even after 4-6 weeks post pull through.

Furthermore, the study showed that 62% of our patients belonged to the school aged bracket and 95% of those were able to get back to schooling or playing 4-6 weeks post operatively. This goes the same with the 38% of the population who were infants and who were all able to return to their respective quality of living, at the same range of time post operatively.

Table 1: Demographic Profile of Patients with Hirschsprung's Disease Post Surgical Transanal Endorectal Pull Through at National Children's Hospital (March 2016 to August 2017)

Profile	**N (%)**
A. Age of Diagnosis	
Newborn	33(82)
Infant	7 (18)
B. Age of Operation	
<1 y/o	15 (38)
1-3y/o	22 (55)
4-7y/o	3 (7)
C. Sex	
Male	32 (80)
Female	7 (20)
D. Z score (wt for ht)	
No wasting	19 (47.5)
With wasting	14 (35)
Severe wasting	0 (0)
No stunting	5 (12.5)
With stunting	2 (5)
Severely stunting	0 (0)

Table 2: Stooling Patterns of Patients Post Surgical Transanal Endorectal Pullthrough with Hirschprung's Disease at National Children's Hospital (March 2016 to August 2017) based on Bristol chart, 4-6 weeks post surgery

Criteria	**N (%)**
Normal	19 (35)
Abnormal	21 (65)

After transanal endorectal pullthrough, 95% of school-aged children all had normal stool frequency (1-2 stools per day), while 100% of infant patients had normal frequency (2-5 tools per day).

In the study, only one patient had soiling after surgery. Further, 95% of the population (n=38) were able to play, socialize and go back to school post-operatively.

DISCUSSION

A cross-sectional descriptive study of patients who underwent transanal endorectal pull through secondary to Hirschsprung's disease was conducted in our institution.

Base on this study, Hirschsprungs's Disease was more common in males, affecting around 30 patients (80%), as what most literatures reported such as at Granéli 2017. According to this journal, the said disease had a skewed gender distribution, with a female to male ratio of 1 : 4.

From other literatures, children with Hirschsprung's disease post surgically were malnourished as to the study of Kressmann 2006. According to this journal, 1.5% developed severe malnutrition due to early satiety, abdominal discomfort, and distention associated with or without chronic constipation. Our study, described 24 patients (60%) of the population had a normal z score on weight for height and 35 % were wasted. No patients were described as severely malnourished despite that 69% of the total patients had abnormal type of stool (type 5) based on the Bristol Chart.

1.5% developed severe malnutrition due to early satiety, abdominal discomfort, and distention associated with or without chronic constipation. Our study, described 24 patients (60%) of the population had a normal z score on weight for height and 35 % were wasted. No patients were described as severely malnourished despite that 69% of the total patients had abnormal type of stool (type 5) based on the Bristol Chart.

This study signified that comparing it to the result Nakatsuji T. Et. al, 2010, their percent of good outcome was 80% in developing countries such as in Malaysia and Japan, while in developed countries, 90% had promising outcomes. The previous Philippine study from Dela Merced et, al 2003 showed 96% survival rate, our institution's data has almost the same percentage of survival rate (95%) 4-6 weeks post-surgery, and this was 80% reliable. Furthermore, this gave us an idea that, in spite of the disease' chronicity, patients can be able to live their life to its maximum

Despite of the stigma that this disease can brought about, both physically and psychologically, as pediatricians, this study can support and strengthen our prognosis towards the said disease and help re-assure the parents of the condition of their children. This will eventually create a positive view as to how mother and child can cope with the impact of the disease. It has been noted that 2.5 % (1) of the patients had soiling and another 2.5% (1) developed enterocolitis, signifying that the surgery had 5 % chance of developing its complications and would further pave us a way for exploring these factors.

Lastly, this study gave us a baseline data of the functional outcome of patients with Hirschsprung's Disease 4-6 weeks post endorectal pull through, at National Children's Hospital, Philippines.

SUMMARY

The prevalence rate of patients with Hirschasprungs' Disease post transanal enodrectal pullthorugh at the National Children's Hospital was 31% (40/130) from March 2016 to August 2017. Base on this study, from the total of 130 patients who followed up at the Surgery Outpatient Department, there were 40 patients who were diagnosed with Hirschsprung's disease, and underwent trans anal endorectal pull through, were included as study subjects. Among those, 82 % were diagnosed at the newborn period and predominantly were males. The most common age group to which surgery was done, belonged to age 1-3 years old, at around 55%. The study also showed that 60% about 47.5% of the patients had no wasting and 12.5% had no stunting. However, 69% of these patients presented with abnormal stool type based on the Bristol Chart (type 5), with 2.5 % of the total population who still had soiling post operatively. In our study it showed that 95% of our patients were able to go back to playing and schooling, about 4-6 weeks post-surgery. The remaining 2 patients (5%) as follows; one of whom still had soiling and the other developed enterocolitis, readmitted and treated.

CONCLUSION

The prevalence rate of the said disease is 31 % (40/130). Majority of our patients with Hirschsprung's Disease (82%) were diagnosed during the newborn period. The prevalence of this disease is more common to the male gender and the common age group from which trans anal endorectal pull through was done, was at 1-3 years of age. Postoperatively, most of the patients improved with more than half of the study population had normal z scores. In spite the fact that 1 patient had soiling and more than half of the population had an abnormal stool type based on the Bristol Chart (type 5), 95% of our patients were able to go back to playing, socializing and schooling, at about 4-6 weeks post-surgery, signifying a great impact with the prognosis of the disease. Furthermore, this provided the National Children's Hospital a baseline data with regards to Hirschsprung's disease, a chronic disease which created a stigmata in the lives of children as a whole; physically, emotionally and psychologically.

RECOMMENDATION

There are other factors that can affect the functional outcome of Hischsprung's disease post operatively. These include the length of the segment to be removed. Some studies noted that if a shorter segment is affected, it has better prognosis. Other factors include other comorbidities; such as, Down's syndrome, malnutrition and complications; which include enterocolitis and gastroenteritis. These topics can be further explored and correlated with the functional outcome of the said disease. Furthermore, a comparison of the prognosis between a single-stage versus a multi-stage pull through procedure in the Philippine set-up can be a good area to be investigated as part of this topic.

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ANNEX

Patient Data Collection Form 1

Code: Age: Sex: Wt: Ht:

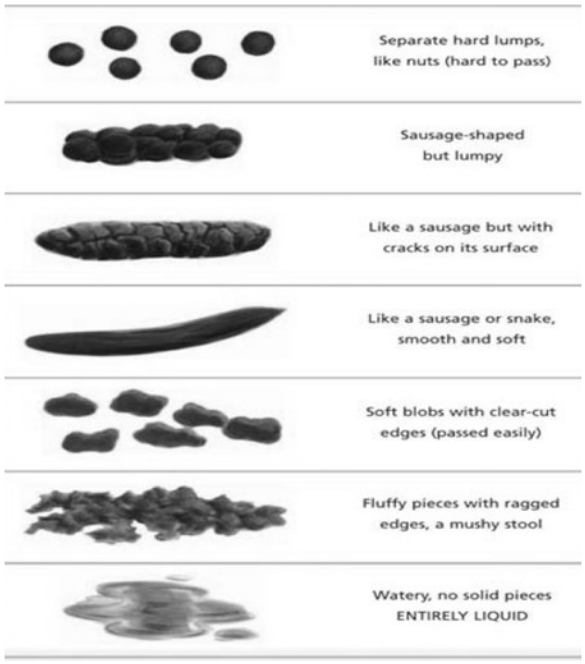
wt/ht z score:

***Operational Definition of Stooling Pattern**

1. How many bowel movements does your child have each day?

☐ Once ☐ Twice ☐ Thrice ☐ ≥ Four

2. what is the type of the stool?



3. Is there any incontinence?
☐ Yes ☐ No

***Operational Definition of Quality of Life**

1. (if 4-7 yrs. old) Does the child attends/returned to schooling? Or became active again?

☐ Yes ☐ No

2. (if 0-3 yrs. old) Did the child regain his/her playfulness?

☐ Yes ☐ No

If Child is 0-3 yrs old:

a. Neonatal period
   - ☐ Visual preference to human face

b. 1 months
   - ☐ Body movements in cadence with voice of other in social contact

c. 2 mo
   - ☐ Smiles on social contact

d. 3 months
   - ☐ Listens to music says "aah, ngah", sustained social contact

e. 4 months
   - ☐ Laughs out load

f. 7 months
   - ☐ Babbles

g. 10 months
   - ☐ Plays peek a boo, waves bye-bye

h. 15 months
   - ☐ Indicates needs by pointing

i. 18 months
   - ☐ feeds self, kisses parent pucker

j. 1 year old
   - ☐ Plays simple ball game

k. 30 months
   - ☐ pretends in play

l. 36 months
   - ☐ Plays simple games in parallel with other children

If Child 4-7 yrs. Old

a. 4-5 yrs. Old
   - ☐ role playing, creation of scenarios that are imaginary, recognition of rules

     OR

   - ☐ knows basic color, counts 1-10 penies, prints first name (underline if needed to emphasized)

b. 6-7 yrs. Old
   - ☐ dresses completely, abides with rules

     OR

   - ☐ formation of group of friends