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Pathological Overview of Constipation in Children - Institutional Study

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Abstract: *Introduction: Constipation is a common problem in the pediatric population. HIRSCHPRUNG'S DISEASE is a congenital condition, also known as congenital aganglionic megacolon. A newborn with Hirschsprung's disease is usually unable to have a bowel movement in the first days after birth. AIMS: To classify the types of Hirschprung's disease and incidence of total colonic aganglionosis among them and to analyze the sex incidence and age distribution of Hirschprung's disease in patients with constipation. Materials and methods: It was a retrospective study of 3 years duration. 52 cases were presented with different pathological diseases which were subjected to surgical biopsy whose chief complaint was constipation. Results: Out of 52 cases of constipation, Hisrchprung's disease constituted 36cases with predominant short segment type. Conclusion: In neonates and young children, constipation due to Hisrchprung disease and other pathological disease should be excluded with rectal biopsy which was a gold standard.*

Key words*: Hirschsprung's disease, aganglionosis, Constipation*

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I. Introduction

Constipation is a common problem in the pediatric population. Chronic constipation with soiling has been reported in up to 3% of the patients referred to teaching hospitals and clinics. Children with marked alterations in frequency size, consistency and ease of defecation should to be considered to be constipated. Patient with idiopathic constipation classically have soiling and dilated ampulla filled with stool. Both these findings are unusual in patients with typical Hirschprung's disease.

HIRSCHPRUNG'S DISEASE is a congenital condition characterized by the absence of ganglion cells in the submucosal and myentric plexus of the distal bowel. It is believed to result from the failure of ganglion cells to migrate fully cadal during the embryonic life. The loss of ganglion cells extends for a variable distance above the anorectal junction. The classical HIRSCHPRUNG'S DISEASE was found restricted to rectosigmoid junction in 75% of cases, long segment in 15% of cases, ultra short segment disease in 5% of cases and variable length was found in 5% of cases. Histologically in HIRSCHPRUNG'S DISEASE there is absence of ganglion cells in normal plexus of all layers of gut together with hypertrophied nerve bundles.

Clinical presentations: The usual presentation is that of a neonate with delayed passage of meconium, abdominal distention, biliary vomiting and intestinal obstruction. In nearly half of patients the diagnosis is established by the first 3 months of life and in another 20% by the first year of life. Up to 15% of cases may be diagnosed as late as 5 years of age.

TABLE NO-1: Clinical comparison between idiopathic constipation and HIRSCHPRUNG'S DISEASE.

Sings, symptoms and diagnostic studies	Idiopathic constipation	Hirschprung"s disease
1.Soiling	common	unusual
2.Stool In Ampulla	common	unusual
3.Obstuctive Symptoms	rare	common
4.Stool Retentive Behavior	common	Rare
5. Enterocolitis	Never	Possible
6.Anorectal Examination Findings	Dilated ampulla	Narrow
7.Contrast Enema Findings	Dilated ampulla	Narrowed distal segment

II. Materials And Methods

The present study was 3 year study. The cases that present with various causes of chronic constipation & intestinal obstruction such as HIRSCHPRUNG'S DISEASE, meconium illeus, illeal atresia, intestinal neuronal atresia, intestinal neuronal dysplasia & hypoganglionosis were examined by surgical biopsies and specimens. Age group varied from a day old child to 5 years. The study comprised colorectal and appendicectomy specimens, various level biopsies and myomectomy specimens. The study included both ganglionic and aganglionic segments of intestine. Several sections were taken from these cases; the blocks were routinely fixed, processed and stained by H&E. The study showed a male: female ratio of approximately 3:1.

TABLE NO-2: HIRSCHPRUNG'S DISEASE and other causes of constipation in pediatric age group.

Disease	**Total no of cases**	**Percentage (%)**
HIRSCHPRUNG'S DISEASE	36	69
Meconium illeus	2	3.8
Intestinal atresia	4	7.69
Intestinal neuronal dysplasia	1	1.9
Hypoganglionosis	3	5.76
Normal	4	7.69
Inadequate biopsies	2	3.8
Total	**52**	

Among 52 cases studied , Hirschprung's disease constituted 36 cases (69%), and other conditions of intestinal obstruction comprised of 31%, that is meconium illeus 2 cases(3.8%), Intestinal atresia 4 cases(7.69%), intestinal neuronal dysplasia 1 case (1.9%), hypoganglionosis 3 cases (5.76%), normal 4 cases (7.69%) and rest are inadequate biopsies.

TABLE NO-3: Site of origin of HIRSCHPRUNG'S DISEASE

Types of Hirschprung's disease	**Total no of cases**	**%**
Ultra short segment	2	5.5
short segment	28	77.7
Long segment	5	13.8
Total colonic aganglionosis	1	2.77
Total	**36**	

In our study out of 36 cases of confirmed Hirschprung's disease , short segment was the most commonly occurring type constituted 28 cases(77.7%). The least common was Total colonic aganglionosis constituting 1 case (2.77%) while long segment was 5 cases (13.8%)

Fig -1: Intestinal segment showing absence of ganglion cells in submucosa(10X H&E)

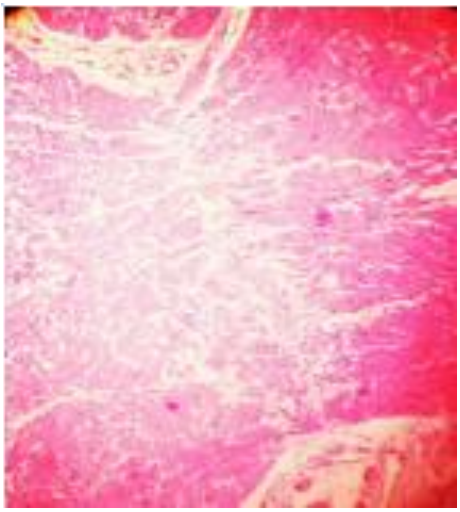

Fig -2: Intestine showing hypertrophied nerve bundles

TABLE NO -4: Age incidence:

Age	Total no of cases	%
0-3 months	36	69.2
3-6"	3	5.76
6-12	4	7.69
1-2 years	6	11.53
2-4	2	3.84
4-6	1	1.92
Total	52	

Age group ranged from neonate to 5 years. Youngest patient recorded was one day old boy with long segment Hirschprung's disease. Oldest patient recorded was 5 years old boy with short segment Hirschprung's disease. Majority of the cases occurred in age group of first 3 months of life and next commonly from 1-2 years, after 5 years the incidence decreased.

TABLE NO-5: Sex distribution

Total no of cases	Male	Female	M:Fratio
52	**39**	**13**	**3:1**

According to literature the incidence of Hirschprung's disease is higher in males. The M:F ratio being 4:1 ratio given according to American academy of pediatrics survey , collective Japanese report gave 3:1 ratio (M.Schiller, P.Levy et al).In our study of 57 cases 39 were male children and 13were female children. The male, female ratio 3:1

III. Discussion

Study of Hirschprung's disease in pediatric age group was undertaken to observe the age, sex incidence and to study the various types of Hirschprung's disease.In the present study we have recorded 4 types of Hirschprung's disease- ultra short segment, short segment, long segment and colonic aganglionosis. In our study we found that there is no absence of ganglion cells, but the clinical presentation of chronic constipation was taken into account .Ultra short segment and short segment Hirschprung's disease involved the rectum and rectosegmoid colon. This was the most common type encountered in the present study occupying 64.5% of cases. Long segment involve the colon but not including illeocecal valve, occupying 21.5% of cases.

Total colonic agangliosis : It is a severe form of Hirschprung's disease.Incidence varies from 5-8% (Harjai Metal)) more commonly recognized in neonatal period and more likely to be associated with enterocolitis. In this study total colonic aganglionosis occupying 2.77% of cases. It is usually associated with significant morbidity and mortality. To diagnose this entity a high index of suspicion is required in any neonate with signs of intestinal obstruction.

TABLE NO-6: Extent of agangliosis in Hirschprung's disease

	Polley et al	Kleinhaus et al	Klient&Phillipart et al	Vane &Grosfield et al	Ikeda et al	Swenson et al	In our study
Transition zone	23	27	32	23	23	23	4
Rectosigmoid colon	74	74	58	81	79	79	25
Long segment of intestine	11	18	26	10	13	24	5
Total colon	12	8	12	9	05	03	1
Small intestine	3	*	0	*	3	0	1

Polley, kleinhaus,klein and phillipart , vane &gross field, ikeda, Swenson et al in their studies found the involvement of rectosigmoid colon(short segment Hirschprung's disease) occupying the highest incidence. Least incidence was that of total colonic aganglionosis when small intestine was included.

Polley&ikeda in their study observed that the small intestine involvement was the least common type. In our study short segment Hirschprung's disease is the most common type involving 77.7% cases, lowest incidence is occupied by total colonic aganglionosis that is 2.77%.

AGE: The age incidence as observed in these Hirschprung's disease at our centre ranged from birth to 5 years. In our study , almost 1/3rd of cases were established by the first 3 months of life, only 8% by the first year, from 1-2 years they were almost 15%.

SEX:Rajashekaran, koteswaran, baba Krishnan et al studies on Hirschprung's disease was found in age group between 1 month-6 years among which 17 were males and 6 were females with ratio of 3.3:1. In our study of 3 years, comprising 39 males and 13females gives a male: female ratio of 3:1 .Over all males are most commonly affected than females.

IV. Conclusion

Though constipation accounts for many visits to the pediatrician, a careful history and focused physical examination should reveal those patients with a history of delayed passage of meconium, abdominal distention, or vomiting who require rectal biopsy to exclude HD and other pathological causes for constipation. The present study carried out to analyze the types of Hirschprung's disease & incidence of total colonic aganglionosis among the various causes causes of constipation.. HD is still a disease with many unclear aspects. Rectal biopsy remains the gold standard and definitive in most cases. It is a safe procedure with a low complication rate when performed by an experienced pediatric surgeon or gastroenterologist. In doubt, ancillary techniques may be applied, but a re-biopsy is the best choice. Despite genes and pathways identified in patients at the beginning of the 21st century there is still a lot to do for investigating etiology, pathogenesis, and treatment modalities of this frightful disease.

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