



## **A Rare Case Report on Hirschsprung's Disease**

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### **Authors' contributions**

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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**Case Study**

### **ABSTRACT**

**Introduction:** Hirschsprung's disease is a condition that affects the large intestine and causes problems passing stool. It was first identified by "Ruysch" in 1961 and popularized by Hirschsprung's in 1886. A missing nerve cell in the baby's colon muscles causes the condition, which is present at birth.

**Clinical Findings:** Difficulty in passing stool, Abdominal pain, fever, (Temperature 100-degree f) Vomiting, Failure to thrive, poor feeding.

**Diagnostic Evaluation:** CBC Infestation, Blood test, Hb -10.4 gm%, Total RBC Count -4.37 million /cu mm, RDW-13.3%, HCT-30.5%, Total WBC Cont-4000 /cu mm, Monocytes- 13%, Granulocytes-65%, Lymphocytes-30%, AST (SGOT)-28 U/L.

**Therapeutic Intervention:** Blood Transfusion, Inj. Aminoven 200 mg BD, Cefotaxime 400 mg BD IV, Inj. Amikacin 130 mg OD, Inj. Metrogyl 90 mg BD, Inj. Pantocid 10 mg OD.

**Outcome:** After Treatment the child show improvement in his passing stool frequently and relived Vomiting, fever, and increases feeding pattern.

**Conclusion:** My patient was admitted to pediatric ward no 22 at AVBRH with a known case of Hirschsprung's Disease, and he had complained of difficulty in passing stool, at birth after getting appropriate treatment and surgery his condition was improving.

**Keywords:** *Hirschsprung's disease; congenital megacolon; intestinal aganglionosis; enteric nerves system; congenital aganglionosis; megacolon.*

## 1. INTRODUCTION

Hirschsprung's Disease is a rare disease that affects the nerve cells in the intestine, causing bowel obstruction. A section of the bowel is missing nerve cells in HD. These cells usually control the bowel muscles, which push the contents along. The contents of the bowel continue to move until they reach the section of the bowel where the cells are missing. As a result, there is a blockade. As a result, the poo passage becomes blocked or slowed. Hirschsprung's disease is a condition that affects the rectum and a part of the bowel above it. It may affect the rectum and a portion of the bowel above it. It usually only measures a few centimeters in length, but it can occasionally affect the entire bowel [1].

While HD can run in households, no cause has been identified. Hirschsprung's disease is characterized by a partial or complete lack of enteric nervous system ganglion cells in the distal bowel. This is the most common congenital bowel motility defect. As a result, the poo passage becomes blocked or slowed. This is usually only a few centimeters long, but it can affect the entire bowel. and affected neonates usually have a distal intestinal obstruction within a few hours of birth [2].

Resection of the ananglionic bowel and a 'pull-through' operation to carry the usually innervated bowel down to the anal margin<sup>2</sup> are the current treatments It's worth noting that knowledge of congenital megacolon was known the goal of this review is to give a general overview of Hirschsprung's disease, including the etiology and treatment of HSCR in children. Hirschsprung's disease is a complex enteric nervous system disorder that results in functional intestinal obstruction. In children aged 1 to 5 HD is the most common cause of distal intestinal obstruction [3].

Nearly 4000 years before Harold Hirschsprungs, prehistoric Indians diagnosed the disease as a result of nerve defects and proposed sigmoid colostomy as a treatment. The length of the diseased bowel has an impact on the outcome. which always starts distally at the internal

sphincter and extends proximally to varying distances, is classified by the M: F ratio until it becomes almost the same 1.5:1 in total colonic aganglionosis HD [4].

Short segment aganglionosis (which affects 80% of HD patients), total colonic aganglionosis (which affects at least the ileocecal valve but not more than 50 cm of the small bowel), long-segment aganglionosis (which falls between the previous two categories), and finally Zuezer syndrome (which affects 20% of HD patients). The internal anal sphincter achalasia, which is more precise for this purpose, has replaced the ultra-short segment [5].

### 1.1 Patient Identification

A male child of 2 years from Nanda Dist. Yavatmal was admitted to pediatric ward no 22 in AVBRH on 1<sup>st</sup> Feb. 2021 with a known case of Hirschsprung's disease, his weight is 8.4kg and his height is 80 cm.

### 1.2 Present Medical History

A male child of 2 years old was admitted to AVBRH on 1<sup>st</sup> February 2021 by his parents with a complaint of the child was not passing stool since birth, and fever, vomiting, abdominal pain, failure of thriving, poor feeding and he was admitted to the pediatric ward he is a known case of Hirschsprung's disease, and child is weak and inactive on admission time.

### 1.3 Past Medical History

My patient was diagnosed to have Hirschsprung's disease, at the age of 17 months when he was admitted to hospital due to fever, and vomiting, that time child rectal biopsy is done now he has come for further surgical intervention.

### 1.4 Present Surgical History

My patient was diagnosed to have Hirschsprung's disease, now the endorectal pull-through surgery is done.



**Fig. 1. Hirschsprung's disease**

### 1.5 Family History

There are four members in the family my patient was diagnosed to have Hirschsprung's disease, and his parents are healthy and he belongs to the nuclear family type of marriage parent is nonconsanguineous marriage all other members of the family were not having complaints in their health except my patients who were being admitted in the hospital.

### 1.6 Past Intervention and Outcome

My patient was diagnosed with a Hirschsprung disease, when he was 17-month-old from that time onwards he was admitted to the hospital from time to time for further treatment of the disease it was found effective as the patient does not develop complications till them.

### 1.7 Clinical Finding

Difficulty in passing stool, abdominal pain, fever (temperature 100-degree f), vomiting, Failure of thriving, poor feeding.

## 2. ETIOLOGY

The cause of Hirschsprung's disease is unknown; The enteric nervous system is in

charge of the intestine's normal peristalsis movement., which is made up of a network of nerves, regulates the gastrointestinal tract's physiology in a largely autonomous manner.

The myenteric and submucosal ganglia in the bowel have more neurons and glial cells than the spinal cord [4]. The absence of ganglion cells in the affected area resulted in chronic spasm, which manifested clinically as functional intestinal obstruction.

### 2.1 Physical Examination

There are not any abnormalities found in head-to-toe examination except abdomen in the abdomen it's found in abdominal distention the child is then and having dull look he is weak and not so cooperative and his irritable.

### 2.2 Diagnosis Assessment

CBC Investigation, Blood test, Hb -10.4gm%, Total RBC Count -4.37million /cu mm, RDW-13.3%, HCT-30.5%, Total WBC Count-4000 /cu mm, Monocytes- 13%, Granulocytes-65%, Lymphocytes-30%, AST (SGOT)-28 U/L, Platelet Count 2.23 micro litter, MCV -69.8%, MCH -32.9%, MCHC -34.2%.

Eosinophil -02%, Basophil 0%, KFT-Urea 15%, Creatinine – 0.4 %, Sodium – 138%, Potassium-43%.

### 2.3 X-ray

In my patient's abdominal x-ray is done there are not many abnormalities found in the abdomen. Some abnormalities are seen in air-fluid levels can be seen in erect abdominal views.

### 2.4 Rectal Biopsy

In my patient's rectal biopsy is done. in a rectal biopsy, the same abnormalities are seen it is used to determine the cause of blood or pus in the stool.

### 2.5 Pull Through Surgery

To treat Hirschsprung's disease, which causes intestinal obstruction. Pull-through surgery aims to remove the diseased portion of your child's intestine and then pull the healthy portion down to the anus. In most cases, this procedure can be completed in a single operation using minimally invasive techniques [6].

### 2.6 Therapeutic Intervention

Blood Transfusion, Inj. Aminoven 200 mg BD, Cefotaxime 400 mg BD IV, Inj. Amikacin 130 mg OD, Inj. Metrogl 90 mg BD, Inj. Pantop 10mg OD, Inj. KCL 2ml BD, Inj. MVI 2ml BD, Inj Emset 1.5mg 8 hourly.

### 2.7 Nursing Management

To offer a comprehensive evaluation and history of the newborn, to monitor the kid's vital signs and abdominal girth, to give the infant in a semi-fowlers position, to feed the child a low residue diet, and to provide patient and family education. To keep the electrolyte equilibrium in the fluid. And to give proper health education about the family members.

## 3. DISCUSSION

A male child of 2 years old from Nanda Dist. Yavatmal was admitted to pediatric ward no 22 on AVBRH on 1<sup>st</sup> February 2021 with a complaint of difficulty in passing stool, abdominal pain, fever, vomiting, failure of thriving, poor feeding, he is a known case of Hirschsprung's disease

which was diagnosed when he was 17 months old as soon as he was admitted to hospital investigation were done and appropriate treatment was started and surgery is done after getting the treatment he is showing great improvement and the treatment was still going on till my last date of care.

## 4. CONCLUSION

Hirschsprung's Disease is one of the most common cases found among children it varies importance to diagnose in early stage so that the child will not develop a complication from the disease successful surgery for Hirschsprung's disease is required through pre and perioperative planning assess to acquire reliable histopathology experience and pull through operative technique.

## CONSENT

The patient's parents informed consent was taken and signed by the patient before writing a case report.

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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