

Case study

A Case Report On Hirschprung's Disease

Abstract:

Introduction: Hirschsprung's disease is the most common cause of large intestinal obstructing in neonates. Hirschsprung's disease is a congenital anomaly caused by migratory failure of neural crest cells leading to abnormal innervations of the bowel. The defect begins in the **internal and sphincter** and extends proximally for a variable length of gut. Hirschsprung's disease is a disorder of the gut caused due to congenital absence of ganglion cells in the sub-mucosal and myenteric plexus of intestine. This disease is also known as megacolon or congenital Aganglionic megacolon. Case presentation: A 2 years old male child, known case of Hirschsprung's disease, was brought to AVBRH for further management. As narrated by the mother, the child was not passing stool since birth. There was no history of abdominal distention, vomiting or fever. The child pass the stool with the help of enema which was given to the child in the morning and evening. Mast. Devansh has past history of NICU stay. In View of not passing stools, USG of abdomen and pelvis done. Endorectal pull through surgery was done on 4/02/2021 under general anesthesia. **No any past surgical** history. There is a past history of rectal biopsy done in 1 1/2 month of age. Conclusion: The patient was admitted in AVBRH for further management. Then the report mainly focused on surgical management and quality nursing care due to which patient was discharged without any further complication and satisfaction. Key words: Hirschsprung's disease, Aganglionic megacolon, sphincter, myenteric plexus, crest cells, intestine.

Introduction:

Hirschsprung infection is a digestive problem described by the shortfall of nerves in pieces of the digestive tract. This condition happens when the nerves in the digestive system (intestinal nerves) don't shape as expected during improvement before birth (undeveloped turn of

events).¹ This condition is generally distinguished in the initial two months of life, albeit less serious cases might be analyzed later in adolescence. Intestinal nerves trigger the muscle constrictions that move stool through the digestive tract. Without these nerves in pieces of the digestive tract, the material can't be pushed through, causing extreme stoppage or complete blockage of the digestive tract in individuals with Hirschsprung illness.² Different signs and indications of this condition incorporate heaving, stomach torment or enlarging, looseness of the bowels, helpless taking care of, hunger, and slow development. Individuals with this issue are in danger of growing more genuine conditions like aggravation of the digestive tract (enterocolitis) or an opening in the mass of the digestive tract (gastrointestinal hole), which can cause genuine contamination and might be deadly.³

Case history:

HISTORY OF ILLNESS:

A 2 years old male child, known case of Hirschsprung's disease, was brought to AVBRH for further management. As narrated by the mother, the child was not passing stool since birth. There was no history of abdominal distention, vomiting or fever. The child pass the stool with the help of enema which was given to the child in the morning and evening.

PAST HISTORY:

Mast. Devansh has past history of NICU stay. In View of not passing stools , USG of abdomen and pelvis done.

PRESENT SURGICAL HISTORY:

Endorectal pull through surgery was done on 4/02/2021 under general anesthesia.

PAST SURGICAL HISTORY:

No any past surgical history. There is a past history of rectal biopsy done in 1 1/2 month of age.

FAMILY HISTORY:

Mast. Devansh belongs to middle class family. His father is the breadwinner of the family. No one in the family has history of congenital anomalies, hypertension and diabetes mellitus.

Table 1: Family composition:

Sr. No	Name of family members	Age / Sex	Occupation	Education	Relation with patient	Health status
1.	Mr. Mahesh Chaudhari	33yrs/M	Farmer	10 th pass	Father	Healthy

2.	Mrs. Nita Chaudhari	26yrs/F	Housewife	11 th pass	Mother	Healthy
3.	Mast. Samarth Chaudhari	5yrs/M	-	Nursery	Brother	Healthy
4.	Mast. Devansh Chaudhari	2yrs/M	-	-	Patient	Unhealthy

SOCIO-ECONOMIC HISTORY:

- ⊙ **Class** : Middle class
- ⊙ **Housing** : Own pakka house
- ⊙ **Electric supply** : Present
- ⊙ **Water supply** : Present
- ⊙ **Drainage system** : Closed drainage system present
- ⊙ **Ventilation** : Present
- ⊙ **Type of the family** : Nuclear family
- ⊙ **Occupation of Father** : Farmer
- ⊙ **Occupation of Mother** : Home maker
- ⊙ **Income** : Rs. 10,000 – 12,000/-

month

BIRTH HISTORY:

Prenatal History:

- ⊙ **Nature of marriage:** Non- consanguineous
- ⊙ **Exposure to radiation:** None
- ⊙ **Antenatal check-up:** 3 times antenatal check-up done at 3, 7 and 9 months
- ⊙ **History of any drugs:** No drugs taken besides routine drugs.
- ⊙ **Tetanus toxoid vaccine:** 2 doses taken.

Peri natal history

- ⊙ **Type of delivery** : LSCS (lower segment cesarean section)

- ⊙ **Place of delivery :** Government Hospital, Yavatmal
- ⊙ **Birth weight :** 2.5 kg
- ⊙ **Mother condition following delivery :**

Mrs. Nita did not have any complication following delivery.

Post Natal History

- ⊙ **Child condition at birth:** baby cried immediately after birth.
- ⊙ **Birth weight:** 2.5 kg
- ⊙ History of NICU stay for 5 days. (child not pass the meconium after birth and history of abdominal distention)
- ⊙ **Breastfeeding:** Feeding was given to the child.

GROWTH AND DEVELOPMENT

Table 2. Physical development

Anthropometry	Book picture	In child	Remark
Height	81-82cm	81cm	Normal
Weight	12kg	12kg	Normal
Head circumference	47-48cm	47cm	Normal
Chest circumference	13-14cm	13cm	Normal
Mid-arm circumference	35-38cm	38cm	Normal

Developmental milestones:

Table 3 Gross motor:

Sr. no	Book picture developmental milestones	In child	Book picture	Remark
1.	Steady gait	16month	15-20Month	Achieved
2.	Walks on heel-toe	18month	16-18Month	Achieved
3.	Walks up and down stairs holding wall	23month	24 Month	Achieved

Table 4 Fine motor:

Sr. no	Book picture developmental milestones	In child	Book picture	Remark
1.	Picks up objects from floor	20month	24Month	Achieved

2.	Can build tower of 6-7 cubes	23month	24Month	Achieved
3.	Turn pages, one at a time	22month	24 Month	Achieved
4.	Drinks with glass.	24months	24 Month	Achieved
5.	Pull garments eg socks	22months	24 Month	Achieved
6.	Can brush teeth with help	24months	24 Month	Achieved

Table 5 Language development:

Sr. no	Book picture developmental milestones	In child	Book picture	Remark
1.	Knows at least 4 body parts.	23month	24Month	Achieved
2.	Uses 50+ words	24month	24Month	Achieved
3.	Refers to self by name	24month	24 Month	Achieved

Table 6 Social development:

Sr. no	Book picture developmental milestones	In child	Book picture	Remark
1.	Enjoys parallel play.	24month	24Month	Achieved
2.	Enjoys play with doll.	23month	24Month	Achieved
3.	Will do simple household tasks	24month	24 Month	Achieved

PHYSICAL EXAMINATION

Genitalia

Anal region:

- Redness- present over the surgical area
- Swelling – mild swelling present
- Scar- surgical scar present
- Lesions – absent

Table 7 INVESTIGATION

Investigation	Patient value	Normal values	Justifications
Hb%	11.3 gm%	13-15.5gm%	Decreased (as the nutrients and electrolytes are not absorbed due to large intestinal dysfunction so the nutritional requirements is not fulfilled.)
MCV	80.3cub. micron	80-90 cub. micron	Normal
MCH	24.3pico-gm	26.5-33.5 Pico-gram	Decreased (As the HB level decreased due to interstitial absorption dysfunction the MCH level is decreased)
MCHC	33.2%	30-36.5%	Normal
Total RBC count	4.89millions/c u.mm	4.5-6 millions/cu.mm	Normal
RDW	13.3%	10-15%	Normal
HCT	41.1%	40-50%	Normal
Total WBC Count	4000	4000-11000/cu.mm	Normal
Monocytes	03	4-10%	Decreased (Due to immunological response on colon the monocytes are decreased)
Granulocytes	65	40-50%	Increased (Due to immunological response on colon granulocytes increased)
Lymphocytes	30	17-48%	Normal
Eosinophils	02	0-5%	Normal
Total Platelet count	2.23	1.5-4lac/cu.mm	Normal
Kidney function test: Blood Urea	16mg/dl	10-50 mg/dl	Normal
Serum Creatinine	0.6mg/dl	0.6-1.2mg/dl	Normal

Serum Sodium	140mEq/L	135-155 mEq/L	Normal
Serum Potassium	4.1meq/L	3.5-5.6 mEq/L	Normal

Rectal biopsy done:

Result: section from given biopsy shows unremarkable mucosa with occasional superficial ulceration.

Table 8 Pharmacological management:

Sr.no	Name of drug	Dose	Route	Frequency
1.	Inj. Ceftriaxone	100mg	IV	12hrly
2.	Inj. Amikacine	15 mg	IV	24hrly
3.	Inj. PAN	10mg	IV	8hrly
4.	Inj. Metrogyl	10mg	IV	8hrly
5.	Inj. EMSET	1.5mg	IV	8hrly

Table 9 Medical management:

Book picture	Patient picture	Justification
Administration of isotonic enema	Given to the patient	Trade name: glycerin and sodium chloride enema Action: Distends colon, stimulates peristalsis and softens feces Uses: -To relieve constipation -To treat fecal impaction Adverse effect: Possible sodium retention ⁴
Stool softeners	Not given to the patient	

Table 10 Surgical management:

Sr.no	Book picture	Patient picture	Justification
1.	TEMPORARY COLOSTOMY	Patient has not undergone this surgery	To decompress intestine divert fecal stream and

			rest the normal bowel. ⁵
2.	SWENSON PROCEDURE	Patient has not undergone this surgery	Abdominoperineal pull through leaving the smallest amount of aganglionic bowel remaining. ⁶
3.	DUHAMEL PROCEDURE	Patient has not undergone this surgery	Retrorectal transanal pull through creating a neorectum with aganglionic anterior wall and ganglionic posterior wall
4.	SOAVES PROCEDURE (Endorectal pull through surgery)	Patient has undergone this surgery	Endorectal pull through in which the ganglionic segment is pulled through the aganglionic muscular cuff, preserving the internal sphincter may be done laparoscopically. ⁷

Nursing diagnosis:

- Acute pain related to presence of surgical incision.
- Risk for infection related to : post- surgical procedure.
- Imbalanced Nutrition, Less Than Body Requirements related to inadequate food intake.
- Risk for fluid / electrolyte volume imbalance related inability absorbs water by the intestinal tract.
- Knowledge Deficit related to disease condition and treatment modalities.

1) Acute pain related to presence of surgical incision.

Goal: Reduce the pain level.

- **Intervention**
- Assess the level of pain by using visual analogue scale
- Assess location, weight and type of pain.
- Provide comfortable position.

- Provide distraction therapies is play therapy music therapy etc
- Administer analgesics as per doctors order

2) Risk for infection related to: post- surgical procedure.

Goal: Reduce the infection.

Intervention:

- Assess the sign and symptoms of infection specially temperature.
- Emphasize the importance of hand washing technique.
- Keep area, around wound clean and dry.

3) Imbalanced Nutrition, Less Than Body Requirements related to inadequate food intake.

Goal: Improve the nutritional status.

Intervention:

- Ascertain understanding of individual needs.
- Discuss eating habit including preference.
- Advice semi-solid or liquid diet.

Discussion:

A 2 years old male child, known case of Hirschprung's disease, was brought to AVBRH for further management. As narrated by the mother, the child was not passing stool since birth. There was no history of abdominal distention, vomiting or fever. The child pass the stool with the help of enema which was given to the child in the morning and evening. Mast. Devansh has past history of NICU stay. In View of not passing stools, USG of abdomen and pelvis done. Endorectal pull through surgery was done on 4/02/2021 under general anesthesia. No any past surgical history. There is a past history of rectal biopsy done in 1 1/2 month of age.

Summary: Mast Devansh admitted in AVBRH for further management of Hirschprung's disease. As narrated by the mother, the child was not passing stool since birth. Medications taken such as Inj. Ceftriaxone, Inj metrogyl, inj pan, inj emset, inj amikacin. Promote diagnosis and appropriate treatment is necessary to achieve better outcome.

Conclusion: In this case result support surgical management including antibiotic treatment. Promote diagnosis and appropriate treatment is necessary to achieve better outcome.

Ethical approval: not applicable

Patient informed consent: while preparing the case report and for publications. Inform consent taken from patients parents.

COMPETING INTERESTS DISCLAIMER:

Authors have declared that no competing interests exist. The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

References:

1. Hirschsprung Disease (for Parents) - Nemours Kidshealth [Internet]. [cited 2021 Sep 22]. Available from: <https://kidshealth.org/en/parents/hirschsprung.html>
2. Hirschsprung's disease - Symptoms and causes [Internet]. Mayo Clinic. [cited 2021 Sep 22]. Available from: <https://www.mayoclinic.org/diseases-conditions/hirschsprungs-disease/symptoms-causes/syc-20351556>
3. Gastrointestinal Perforation: Causes, Symptoms & Diagnosis [Internet]. [cited 2021 Sep 22]. Available from: <https://www.healthline.com/health/gastrointestinal-perforation>
4. Neotomic Enema: Buy bottle of 20 ml Liquid at best price in India [Internet]. 1mg. [cited 2021 Sep 22]. Available from: <https://www.1mg.com/otc/neotomic-enema-otc284210>
5. Types of Colostomies and Pouching Systems | American Cancer Society [Internet]. [cited 2021 Sep 22]. Available from: <https://www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery/ostomies/colostomy/types-of-colostomies.html>
6. Research and Funding | NIDDK [Internet]. National Institute of Diabetes and Digestive and Kidney Diseases. [cited 2021 Sep 22]. Available from: <https://www.niddk.nih.gov/research-funding>
7. What is the Soave procedure for surgical treatment of Hirschsprung disease? [Internet]. [cited 2021 Sep 22]. Available from: <https://www.medscape.com/answers/178493-62825/what-is-the-soave-procedure-for-surgical-treatment-of-hirschsprung-disease>