SURGICAL OUTCOME OF HIRSCHSPRUNG'S DISEASE IN MALE CHILD: A RARE CASE REPORT

ABSTRACT

INTRODUCTION: This disorder characterized by absence of particular nerve cells (ganglions) in a segment of the bowel in an infant. Ganglion cell absence allows the muscles of the intestines to lose their capacity to pass faeces across the peristalsis of the intestine. This condition affects the large intestine and causes problem with passing stool. The condition is congenital. In around 1 out of 5000 live births, Hirschsprung disease occurs and is three times more frequent in boys than girls. About 12% of cases are due to genetic disorders.
Patient history: A 2 years old male was admitted in A.V.B.R. Hospital in paediatric ward. His chief complaint was inability to pass stools on his own , fever , vomiting , constipation **Paediatric history :** This male child was born by Lower segment caesarean section. At the age of 17 months, the complaint started with inability to pass stool on his own . After that his parents approached A.V.B.R. Hospital and Doctor advised for further medical and surgical management.

Main symptoms and importance of clinical findings: The patient had undergone various investigations like blood tests, USG, Physical examination and. rectal biopsy and per abdominal x-ray.

Medical Management: Patient was treated with trans-anal endorectal pull through surgery under general anaesthesia

Nursing management: Administered fluid replacement i.e. DNS and RL, monitored all vital signs checked 8 hourly.

Conclusion: The 2 years old male was admitted in A.V.B.R. Hospital in paediatric ward. His chief complaint was inability to pass stools on his own, fever, vomiting, constipation etc. after undergoing investigation he was diagnosed as Hirschsprung's disease.

Keywords: Hirschsprung's disease, Intervention, management,

Introduction

The Hirschsprung disease is an intestinal disorder characterized by the lack of nerves in area of the stomach. This disorder occurs when the intestinal nerves fail to form properly during pre-birth development. This disease condition is usually identified in the first two months of life, but later in infancy, less serious cases may be diagnosed.[1] When some nerve cells in the wall of the colon do not shape the correct direction when the foetus develops, Hirschsprung disease (aganglionic megacolon) occurs.[2]
Hirschsprung disease occurs in around one in 5,000 newborns. There is a higher risk for children with Down syndrome and other medical conditions, such as congenital heart defects.[3] Hirschsprung's disease can pass it on to their children in their genes, particularly mothers. Boys get more out of it than girl children.[4] In the first week of life, eight percent of

children with Hirschsprung disease exhibit symptoms. In the first 24-48 hours of life, the signs are most frequently observed.[5]

Patient information –
A 2 year male patient was admitted in A.V.B.R. Hospital in paediatric ward no 22. That time his chief compliant was inability to pass stools on his own, fever, vomiting, constipation, abdominal distention, swelling over the belly since last 17 month of age . Patient's parents first came to A.V.B.R. Hospital and they approached the paediatric department. The concerned paediatric doctor advised medical and surgical management of Hirschsprung's disease.

Primary concern & symptoms of the patient: Included constipation, abdominal distension, fever, swelling over the belly. Child was very irritable and sleeping pattern had also changed since last 17 months.

Medical, family, and psycho-social history:
Present case reported no medical morbidity history. He belonged to nuclear family and his father, mother had no any medical history i.e. DM. Hypertension etc. he was mentally stable, conscious and oriented. He had maintained the good inter personal relationship with doctors and nurses as well as other patients.

Relevant past interventions with outcomes:
Present case was not able to pass stools on his own, had abdominal distension, vomiting, constipation since last 17 months.

Clinical findings:
General examination
State of health: Unhealthy
State of consciousness: Conscious
Body built: Moderate
Breath order: Present
Hygiene: Good
General Parameters:
Height: 82 cm
Weight: 9 kg
Vital parameter:
Blood pressure: 130/90 mm Hg
Temperature: 98.6^{o} F
Pulse: 92 beats/min.
Respiration: 24 breath/ min.

Diagnostic Assessment:
Abdominal x-ray showed a bowel obstruction, rectal biopsy for confirm the diagnosis of Hirschsprung's disease
HBsAg – Non reactive

FBS- glucose plasma (fasting) – normal, post meal- normal

Diagnostic testing:

Kidney function test

Potassium (k+) – serum = normal
Creatine – serum = normal
Urea – serum = slightly decreased
Sodium (Na+) = Normal
Complete blood count
Hb% = slightly decreased
Total RBC count = Normal
Total platelet count = Normal
Total WBC count =Increased
Prognosis: Good

Therapeutic intervention: General measure: To check the vital sign (temperature ,pulse , respiration and BP) airway , fluid and electrolyte balance and prevention of complication .

Medical Management: Inj. Ceftriaxone IV stat, Inj. Amikacin 130 mg ,OD for 3 days ,Inj. KCL 4ml OD, Inj. Neomol15 ml BD for 3 days ,InjEmset1 mg SOS,InjMetrogel90 mg BD for 4 days .

Collaborative Management:- Some patients have long –segment Hirschsprung disease , they need sodium supplements .

Surgical Management: - Pull-through surgery - This procedure is a definitive operation of Hirschsprung's disease. In most of cases, this procedure can be done with minimally invasive technique in a single operation . The goal of this surgery is to remove the diseased section of intestine and then pull the healthy portion of this organ down to the anus [6.]

Nursing Management:

1. Administered IV fluid as per physician order.
2. Administered medication as per physician order.
3. Maintained I/O chart
4. Provided patient & relative with psychological support.
5. Established good interpersonal relationship.
6. Explained about surgical procedure to parents
7. Explained about the prognosis of disease.

Follow-up and outcomes: Patient was planned for follow up regularly on basis of advice given by physician. The patient symptoms improved after surgery.

Discussion:
Hirschsprung disease, also known as congenital megacolon or intestinal aganglionosis. This disease is a congenital that causes an obstruction of the intestine.[7]. The recto-sigmoid area of the colon is most frequently involved , but may affect the entire colon .Some patients exhibit recurrent , serious constipation later in life , the condition usually occurs in infancy . Infant signs include difficult bowel movement, nausea , vomiting , weight loss and distension of the abdomen .[8] About 80% of patients with Hirschsprung's disease are diagnosed in the first few months of life.[9] It happen in 1/5000 live births and normally has the inability to pass meconium , along with abdominal distension and pain that generally involves aganglionic bowel surgical resection.[10]

The diagnosis is usually made using Abdominal X-ray, Contrast enema and Anorectal Manometry. Rectal biopsy can be performed if Hirschsprung's disease. A pathologist taking a sample of rectum to view under a microscope .[11] The severity of this disease is differ from case to case because it is a rare case of Hirschsprung disease. This disease can be treated with prompt medical and surgical management. In Hirschsprung's disease the surgeon can perform the procedure of pull through. This surgery usually done in minimally invasive technique. After this procedure physician recommended a balanced diet, laxatives and other medication to help regulate bowel function [23].

Most children who have treatment and surgery do very well and go on to have normal, healthy bowel function.Complications are seen inflammation and infection of the intestines, swelling of the abdomen, Diarrhea, nausea , vomiting , Perforation of the intestine . Few of the rare case reports were reported [12-17]. Relevant literature on child health was reviewed[18-22.]

Conclusion:

A 2 years old male was admitted in A.V.B.R. Hospital in paediatric ward. His chief complaint was inability to pass stools on his own , fever , vomiting , constipation etc. after undergoing investigation he was diagnosed as Hirschsprung's disease**.**

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.

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