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Case Report

Ayurvedic management of chronic constipation in Hirschsprung disease – A case study

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ABSTRACT

Hirschsprung disease (HSCR) or congenital intestinal aganglionosis is characterized by complete absence of neuronal ganglion cells from a portion of the intestinal tract, most commonly in the large intestine. The main sign or symptom of HSCR is constipation usually appearing shortly after birth. This constipation is chronic in nature and usually not relieved with laxatives. The present case is of a patient having HSCR which was successfully managed with Ayurvedic treatment. A four year old boy with complaint of severe constipation, abdominal pain, abdominal distension and occasional vomiting was treated with *Panchakarma* procedures and Ayurvedic oral drugs. The Ayurvedic diagnosis of the case was *Pakvasayagata vata*. *Shashtikashali pinda swedana* (sudation with medicated cooked bolus of rice) and *Matra basti* (enema with medicated oil) with *Ashwagandha taila* (Ayurvedic medicated oil) was given for first 16 days. From the 2nd month of treatment, *Matra basti* was administered daily for 3 months in the dose of 25 ml. In 5th and 6th month *Matra basti* was administered on alternate days in the dose of 25 ml. From the 7th month *Matra basti* was administered once weekly in the dose of 25 ml. In 14th month *Shashtikashali pinda swedana* and *Erandmooladi yapna basti* (medicated enema) was given for 16 days. Eight scales based Medical outcome study (MOS) – 36 item short form - health surveys was periodically assessed for outcome which shows good improvement. Experience of this case showed that HSCR may satisfactory be managed with Ayurvedic treatment.

1. Introduction

Hirschsprung disease (HSCR), or congenital intestinal aganglionosis, is a birth defect characterized by complete absence of neuronal ganglion cells from a portion of the intestinal tract most commonly large intestine [1]. In a child suffering from HSCR, stool moves normally up to the part lacking nerve cells, then in that portion, the stool moves slowly or becomes stagnant. The main sign or symptom of HSCR is constipation or intestinal obstruction, usually appearing shortly after birth. The affected infants frequently present this symptom in the first two months of life and the early symptoms of impaired intestinal motility such as failure to pass meconium within the first 48 h of life. The late symptoms are constipation, emesis, abdominal pain or distention, and occasionally diarrhea. Most often, an infant or a child with HSCR will also have other symptoms, including growth failure or unexplained fever.

The incidence of HSCR is approximately one in 5000 live births [2]. However, the incidence varies significantly among ethnic groups (1.5, 2.1, and 2.8 per 10,000 live births in Caucasians, African-Americans, and Asians, respectively) [3]. The cause of HSCR is most commonly attributed to defective craniocaudal migration of neuroblasts originating from the neural crest during the first twelve weeks of gestation, resulting in functional intestinal obstruction [4]. Genetic defects can increase the chance of a child developing HSCR. Children with Down syndrome and other medical problems, such as congenital heart defects, are at much greater risk. No testing is available that can diagnose a child while the mother is pregnant.

HSCR is a life-threatening illness, and treatment requires surgery. Generally, patients can manage this condition through the use of cathartic agents. However, at some point, the dilated proximal colonic segment may decompensate secondary to the distal obstruction and patients may experience rapidly worsening constipation or even acute obstruction. Here is a case report of a child who suffered from HSCR and was effectively managed with Ayurvedic medications and *Panchakarma* procedures.

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2. Patient information

A four year old boy with complaint of severe constipation, abdominal pain, abdominal distension and occasional vomiting visited in O.P.D. of department of *Panchakarma*, National Institute of Ayurveda, Jaipur. He had history of delayed passage of meconium at birth and was managed successfully with soap and water enema. Since then patient was not being able to pass stools without enemas. Patient's developmental milestones, chest and vital signs were normal but he had distended abdomen. The height of patient was 100.2 centimeters (cm) and the weight of patient was 14.5 kilograms (kg). His appetite was normal. Patient had normal micturation. The patient had undergone for consultations in All India Institute of Medical Sciences (AIIMS), New Delhi two months before, where he was diagnosed as a case of HSCR and surgical management was recommended. He was only child of his parents and none of his parent was affected with these symptoms.

2.1. Clinical findings

On physical examination, patient was anxious and his tongue was uncoated. Patient had *Vata-pitta prakriti* with *Avara samhanana* (lowermost body constitution), *Avara sara* (~lowermost purest body tissue), *Avara satmya* (lowermost homologation), *Avara satva* (lowermost mental strength), *Madhyam vyayamshakti* (middlemost capability to carry on physical activities), *Madhyam aharshakti* and *Jaranshakti* (middlemost food intake and digestive power). The patient demonstrated normal gait. On neurological examination higher mental function and speech were normal. All cranial nerves were intact. On motor examination, bulk, tone, power and coordination of arms and legs were normal bilaterally. Joint position sense and vibration sensation was normal bilaterally. Upon abdominal examination, patient demonstrated marked abdominal distention with palpable dilated loops of colon. Rectal examination revealed an empty rectal vault and resulted in the forceful expulsion of fecal material upon completion of examination. Complete blood count (CBC), Thyroid profile and tissue transglutaminase-IGA (TTG) were within limits.

3. Timeline

A detail of the case study and follow up is given in [Table1].

4. Diagnostic focus and assessment

The patient was a known case of HSCR. It was confirmed by previously done anorectal manometry and barium enema that show a dilated proximal colon with empty rectum and barium enema study that demonstrate delayed emptying time and a funnel-like transition zone between proximal dilated and distal constricted bowel (Figs. 1 and 2) [5]. History of delayed passage of meconium at birth and empty rectal vault and forceful expulsion of fecal material just after completion of rectal examination in the case were suggestive of HSCR. *Pakvasayagata vata* was considered as Ayurvedic diagnosis which is included in *Vatavyadhi* [~neurological, rheumatic and musculoskeletal diseases]. *Antrakoojana* (bowel sounds), abdominal pain, *Atoop* (flatulence), difficulties in passing urine and stool, *Anaaha* (~abdominal distension) and *Trikvedana* (pain around sacral region) are the symptoms of *Pakvasayagata vata.* [6] Acute mega colon, chronic mega colon, constipation, hypothyroidism, intestinal motility disorders, celiac disease, irritable bowel syndrome and toxic mega colon were the differential diagnosis for the case [7]. Thyroid profile and tissue transglutaminase-IGA (TTG) were within normal limit which excluded the diagnosis of hypothyroidism and celiac disease.

5. Therapeutic intervention

Line of treatment for *Pakvasayagata vata* is similar to general line of management of *Udavarta* (~abdominal distention due to constipation and other causes). *Abhyanga* (massage) with *Shita jwarokta* oil (oil used for *Shita jwara*), *Svedana* (sudation), *Varti* (suppository), *Niruha basti, Snehana*, *Virechana* (mild purgation), use of *Anulomak* (carminative) foods are indicated for the treatment of *Udavarta* [8]. Since the patient was in his childhood, mild form of different *Panchakarma* procedures was used. Patient was treated with *Mridu abhyanga* (mild massage) and *Mridu swedana* (mild sudation) and *Matra basti.*

Abhyanga (massage) with *Chandanabalalakshadi Taila* and *Mridu svedana* with *Shashtikashali pinda swedana* were done for 16 days. *Matra basti* with *Aswagandha taila* were also prescribed for 16 days [Table 2]. After completion of these *Panchakarma* procedures a rest of 14 days from all these procedures was given to the patient. From the 2nd month of treatment, *Matra basti*

Table 1
Timeline.

Year	Incidence/intervention
2012	Patient had severe constipation since childhood.
2014–2015	Patient was consulted for chronic constipation in J.K. Loan hospital and medical college Jaipur and Imperial hospital and research center Jaipur. Patient was suspected to suffer from HSCR. Patient was advised conservative treatment.
February–March-2016	Patient was consulted in Pediatric surgery department of AIIMS New Delhi. Diagnosis of HSCR was confirmed in AIIMS New Delhi. Patient was advised to go for biopsy and colostomy.
April-2016	Patient was unwilling for surgery. Patient visited O.P.D. of National Institute of Ayurveda Jaipur for these problems and was advised for administration of *Panchakarma* Procedures.
12/04/2016–27/04/2016	*Shashtikashali pinda svedana* was done for 16 days along with *Matra Basti* for 16 days. Selected Ayurvedic oral drugs- *Vrihatavatachintamani ras* and *Drakshaveleha* twice a day were also prescribed along with these *Panchakarma* procedures. There was clinical improvement in patient condition after one month of therapy. Same oral medication was provided to the patient.
12/05/2016–11/08/2016	*Vrihatavatachintamani ras* was discontinued from 12/07/2016. *Matra basti* with *Ashwagandha taila* once in a day was continued for three months.
12/08/2016–11/10/2016	*Drakshaveleha* twice a day was continued as oral medicine. *Matra basti* with *Ashwagandha taila* was given on alternate days for two months.
12/10/2016–11/05/2017	*Drakshaveleha* twice a day was continued as oral medicine. *Matra basti* with *Ashwagandha taila* was given once a week.
14/12/2016	Serum bilirubin both direct and indirect, serum glutamic oxaloacetic transaminase (SGOT), Serum glutamic pyruvic transaminase (SGPT), serum creatinine ESR,CBC, serum albumin, serum calcium, serum electrolytes, serum phosphorus and total lipid profile were investigated. These were within limit. Barium enema study demonstrates delayed emptying time and a funnel-like transition zone between proximal dilated and distal constricted bowel –suggestive of Hirschsprung disease.
17/05/2017	Above hematological and biochemical parameters were reinvestigated which were within limit.
12/05/2017–28/05/2017	*Shashtikashali pinda svedana* was done for 16 days along with *Erandmooladi yapana basti* for 16 days. *Drakshaveleha* twice a day was continued as oral medicine.