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AN INTEGRATED APPROACH TOWARDS HIRSCHSPRUNG DISEASE (HD) AND ITS MANAGEMENT.

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ABSTRACT

Hirschsprung disease (HD) or congenital megacolon is a disease of the large intestine that causes severe constipation or intestinal obstruction. It occurs approximately in 1 in 5000-8000 live births & predominates in males in ratio of 4:1. The basic pathophysiological feature of Hirschsprung disease is a functional obstruction caused by a narrowed distal aganglionic colonic segment that prevents the propagation of peristaltic waves. Symptoms in infants include difficult bowel movements, poor feeding, poor weight gain, and progressive abdominal distention. Early diagnosis is important to prevent complications (e.g., Enterocolitis, colonic rupture). The appropriate diagnostic approach may vary, depending on the age of the patient and the presenting clinical picture. A plain abdominal radiograph may show a dilated small bowel or proximal colon. Contrast enema (CE), Anorectal manometry (ARM), and Rectal suction biopsy (RSB) are the most important tests used for the diagnosis. RSB is the most accurate test for diagnosing HD. In Ayurveda there is a disease called *Baddhagudodara* in *Nidanasthan* of *Sushruta samhita* which is remarkably similar to that of Hirschsprung disease. According to *Sushruta*, *Baddhagudodara* is caused by simultaneous derangement of *Vayu* and *Pitta*. As in allopathic medical science there is only surgical method to correct the disease but there is need of repeated surgeries due to chances of spreading the disease over further region of colon, also re-occurrence of the

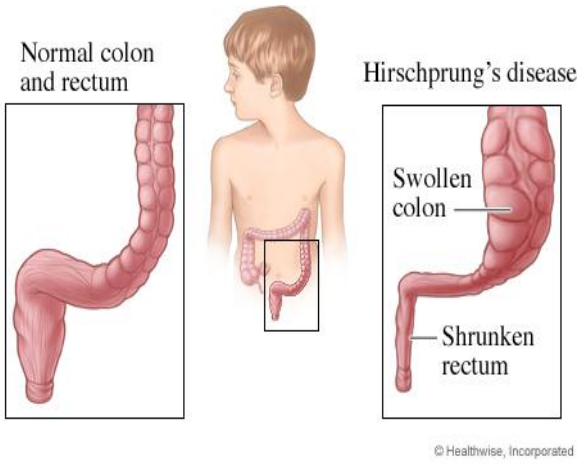
disease is quite common. So to avoid all this, an ayurvedic approach is considered for the management of this disease.

KEYWORDS: Hirschsprung disease, *Baddhagudodara*, Neonatal period, Contrast enema.

INTRODUCTION

Hirschsprung disease also named as congenital megacolon is a disease of large intestine that may cause severe constipation to intestinal obstruction. Hirschsprung's disease is a disease of the large intestine (colon). Stool is normally pushed through the colon by muscles. These muscles are controlled by special nerve cells called ganglion cells. Children with Hirschsprung's disease are born without ganglion cells in the colon. In most cases, only the rectum is affected, but in some cases more of the colon, and even the entire colon, may be affected. Without these ganglion cells, the muscles in that part of the colon cannot push the stool out, which then builds up. *Sushruta samhita* describes a disease called *Baddhagudodara* in *Nidanasthan* is remarkably similar to that of Hirschsprung disease. *Baddhagudodara* means "abdominal distension due to (functionally) blocked rectum" (*Baddha* means obstruction; *Guda* means anus or rectum; *Udara* means abdomen).

Hirschsprung's disease is a deadly birth defect in which the enteric nervous system (ENS) is missing from the end of the bowe.[1,2] Because the ENS controls most aspects of bowel function[3], even a short region of bowel without neurons and glia (i.e., aganglionosis) can be fatal. Down syndrome also predisposes to Hirschsprung's disease. Hirschsprung's disease (Congenital megacolon) is eponymously named after Harold Hirschsprung, who accurately described the clinical features in 1886.



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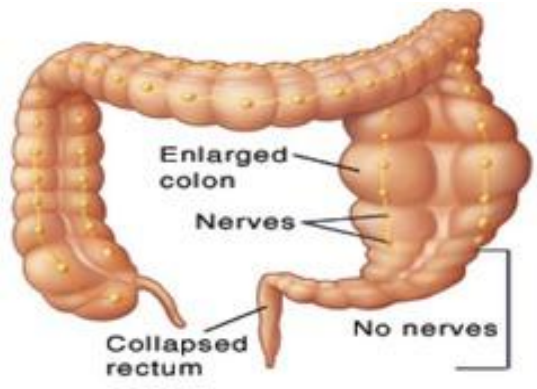

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The disease usually presents in infancy, although some patients present with persistent, severe constipation later in life. Symptoms in infants include difficult bowel movements, poor feeding, poor weight gain, and progressive abdominal distention. If not treated early it may lead to entrocolitis or colonic rupture so early diagnosis is important to prevent complications. Up to one third of patients develop Hirschsprung's-associated enterocolitis, a significant cause of mortality.

Desription of Baddhagudodara in Ayurveda

Ayurveda, the ancient medical system of India, describes a condition called *Baddhagudodara* that closely resembles Hirschsprung disease.[4] Semantically, it means "abdominal distension due to (functionally) blocked rectum" (*Baddha* means obstruction; *Guda* means anus or rectum; *Udara* means abdomen). Etiology of *Baddhagudodara* according to Sushruta is that rectum and distal colon of the affected person are stuffed with gas, stones (fecoliths), hair, and feces. Abdominal distension is characteristically seen between the heart and the umbilicus. Scanty stool is evacuated with greatest difficulty, and the patient may vomit feculent fluid. Hirschsprung disease can be correlated *to* either *Baddhagudodara, Sanniruddha Guda, gudagat*a or *pakwasayagata vata. Similarly Baddhagudodara* may be correlated with at least four different conditions, namely, anorectal strictures, rectal tumors, sigmoid volvulus, and Hirschsprung disease. There is another condition mentioned by Sushruta called as *Sanniruddha Guda* in which the colon is (anatomically) blocked by anorectal constriction. Although both the words *Baddha* and *Niruddha* mean"blockage," Sushruta perhaps used them with a subtle difference. *Baddha* was preferred to mean "functional obstruction," whereas *Niruddha* was frequently used to denote "structural obstruction." However, careful analysis on the basis of symptoms it would reveal that Baddha Gudodaram refers to only Hirschsprung disease and vice versa. *Sushruta* did not mention the age incidence of *Baddhagudodara*.

Aetiology of Hirschsprung's disease

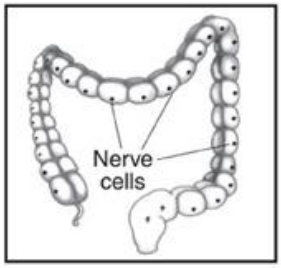
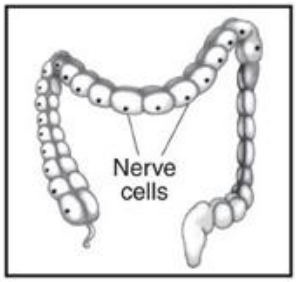
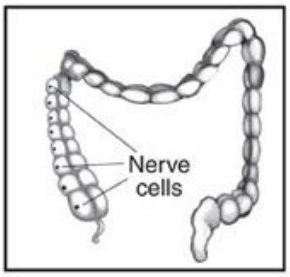
According to modern sciences cause of Hirschsprung's disease is multifactorial, and the disease can be familial or develop spontaneously[5] it is more common in boys than girls.[6] Current research is focusing on the RET proto-oncogene on chromosome 10q11.2.[7] Hirschsprung's disease also can be associated with neurologic, cardiovascular, urologic, and gastrointestinal abnormalities. Down syndrome (trisomy 21) is the most common

chromosomal abnormality associated with the disease, accounting for approximately 10% of patients. Other conditions that have been linked to Hirschsprung's disease include congenital deafness, hydrocephalus, diverticulum of the bladder, Meckel's diverticulum, imperforate anus, ventricular septal defect, renal agenesis and cryptorchidism.[8]

In Ayurveda aetiology of *Baddhagudodara* is mention in *Sushruta samhita* that when the intestine becomes coated inside either with food or other sticky materials ,hair, stone etc then the waste materials of food Aggravate *vata* which in turn gradually accumulate in intestine and after that faeces get obstructed in the rectum and goes out with difficulty in small quality.[9]

Type of Hirschsprung's disease:- People with Hirschsprung's disease have constipation because they lack nerve cells in a part or all of the large intestine. On the basis of nerve cells distribution this disease is two types.

1. **Small segment Hirschsprung's disease:-** Short-segment Hirschsprung's disease means only the last part of the large intestine lacks nerve cells.
2. **Large segment Hirschsprung's disease: -** Long-segment Hirschsprung's disease means most or all of the large intestine, and sometimes the last part of the small intestine lacks nerve cells.

		
Healthy large intestine. Nerve cells are found throughout the large intestine.	*Short-segment HD. Nerve cells are missing from the last segment of the large intestine.*	*Long-segment HD. Nerve cells are missing from most or all of the large intestine.*

How severe HD is depends on how much of the large intestine is affected.

Signs and symptoms:- Typically, Hirschsprung's disease is diagnosed shortly after birth, although it may develop well into adulthood, because of the presence of mega colon, or because the baby fails to pass the first stool (meconium) within 48 hours of delivery.[10] Normally, 90% of babies pass their first meconium within 24 hours, and 99% within 48

hours. The child may experience fecal retention, constipation, or abdominal distention. With an incidence of one in 5,000 births[11,12] the most cited feature is absence of ganglion cells. Overall symptom of Hirschsprung's disease can be different in infants and older children.[13, 14, 15]

Symptoms of Hirschsprung's Disease in infants

1. Tight anal sphincter with an empty rectum
2. Failure to Progressive abdominal distention
3. Donot pass meconium in the first 24 hours of life
4. Infrequent, explosive bowel movements; difficult bowel
5. Bilious vomiting
6. Jaundice and poor feeding

Symptoms of Hirschsprung's Disease in infants

1. Not being able to pass stools without laxatives or enemas. A laxative is medicine that loosens stool and increases bowel movements. An enema is performed by flushing water, or sometimes a mild soap solution, into the anus using a special wash bottle.
2. Progressive abdominal distention
3. Malnutrition
4. Bloody diarrhea.
5. Slow growth or development.
6. Lack of energy because of a shortage of red blood cells, called anemia.

Baddhagudodara, according to *Sushruta* is a type of tympanitis caused by functional blockage of guda. Due to *Vata* accumulation faeces get obstructed in rectum and goes out with difficulty in small quantity, abdomen get distended greatly in between area of the heart and the umbilicus patients vomits material having the smell of faeces (**16**).

Diagnostic examinations

The diagnosis is established in 15% within the first month of life, in 40-50% in the first 3 months, in 60% at the end of the first year of age, and in 85% by 4 years Diagnostic aids include barium enema on unprepared bowel, suction biopsy for acetyl cholinesterase staining. Anorectal manometry; as a preliminary screening test also has a place (**17**).

i. **Contrast enema (CE)**:- The test is most useful in new-borns when the baby cannot push the stool out and an obstruction to the lower end of the colon is suspected. Children with

Hirschsprung's disease will have a narrow colon where the ganglion cells are missing. The diagnosis of Hirschsprung's disease is made by rectal biopsy, but the enema X-ray may have a role in determining examining the length of the affected colon.

ii. **Ano Rectal Manometry (ARM)**:- In ano-rectal manometry, the doctor places a small balloon into the rectum and measures how well the muscles around the anus respond to inflations of the balloon. Children with Hirschsprung's disease will not relax the muscles in the anus. If there is no relaxation, then this suggests that there are other parts of the large intestine that may not be able to push stool through. Manometry may not be very reliable in infants less than 12 days of age.

iii. **Rectal Biopsy Specimen (RBS):-** This is the most accurate test for Hirschsprung's disease. The doctor takes a very small piece of the rectum to look at under the microscope. Children with Hirschsprung's disease will not have any ganglion cells in the sample taken.

Treatments

The treatment of Hirschsprung's disease is surgical. Initial medical management is important; this includes the correction of fluid and electrolyte imbalances, antibiotic therapy if enterocolitis is present and rectal decompression with the use of rectal irrigations and rectal tubes until the time of surgery.

Surgery is done to remove that part of the colon that lacks the ganglion cells and then to connect the healthy colon above this to the anus. The operation can be done in two stages:-

1. In the first stage the surgeon separates the healthy colon from the affected colon. Then the healthy colon is brought out to the skin as a colostomy (opening of the colon to the abdomen) which then empties into a special bag that the parents can manage.
2. In second stage (Several months later) the surgeon removes the affected colon and takes the healthy colon from the colostomy and connects this to the rectum just above the anus.

Generally it is agreed that the interval between colostomy and definite surgery should be six months to one year or when the infant gains weight up to 10 Kg. however the interval depends on the age at presentation for colostomy. The recent trend to perform definite surgery i.e. pull through at an early age and even in the neonatal period (**18, 19, 20**). In some babies the surgery can be done as a single operation without a colostomy. This can only be done safely if the baby is healthy and the colon is not full of stool.

Treatment of Hirschsprung disease (Baddhagudodara) in Ayurveda

Sushruta described the surgical treatment of *Baddhagudodara* in great details. In the opening verses of this chapter, he declares that it is incurable but subsequently narrates a surgical technique —probably as palliative measure. According to *Sushruta,* the patient should initially be anointed with *sneha* (antiseptic oil). An abdominal incision should be made below the umbilicus and 4 fingers' breadth (approximately 7-8 cm) to the left of median hairline. Through this incision, a loop of the large intestine to the length of 4 fingers' breadth should be gently drawn out. After careful examination, the intestine is opened to remove inspissated feces, hair, and stones. Then, the intestinal wound is soaked with honey and clarified butter. It should then be gently replaced into the abdomen, and the mouth of the incision is sewn up at the selected place.[21] Intestinal anastomosis has been done using a technique similar to surgical staplers. Large black ants were applied to the grip intestinal edges. The ants fasten them by gripping with their claws. The ants were then decapitated leaving their heads. Approximation of edges was retained intact by rigor mortis of claws. According to some author Hirschsprung's disease can be correlated with *gudagat*a and *pakwasayagata vata,* the line of treatment is *Udavarthar chikitsa*.

DISCUSSION

Hirschsprung's disease is a congenital disorder of large intestine due to absence of certain ganglionic cells in a particular segment of colon which results in absence of peristalsis. The symptoms of the disease indicate that there is vitiation of *Udarvata* leading to *Baddagudoda.* The condition can also be correlated to vitiation of *Apan vayu* in *Guda* and *Pakwashaya*. According to *Sushruta samhita* symptoms of *Baddhagudodara* states that due to *Vata* accumulation faeces get obstructed in rectum and goes out with difficulty in small quantity, abdomen get enlarges greatly in between area of the heart and the umbilicus patients vomits material having the smell of faeces. *Baddagudodar* according to *Sushruta*, is a type of tympanities caused by (functional) blockage of anorectum due to derangement of *Vata* and *Pitta*. The rectum and distal colon of the affected person are stuffed with gas, stones (fecoliths), hair and faces which results in abdominal distension between the heart and the umbilicus.

There may be four different conditions, namely Anorectal strictures, rectal tumors, sigmoid volvulus and Hirschspung disease which can be discussed with *Baddagudodara* but with careful analysis result can be yielded. Sigmoid volvulus, an acute emergency can be

excluded. Huge gaseous distention of transverse colon in *Baddagudodara and occurance of fecoliths is more symmetrical to* Hirschspung disease. *Sushruta* mentioned another condition called *Sanniruddha Guda* in which the colon is (anatomically) blocked by anorectal constriction. Although both the words *Baddha* and *Niruddha* mean "blockage," Sushruta perhaps used them with a subtle difference. *Baddha* was preferred to mean "functional obstruction," whereas *Niruddha* was frequently used to denote "structural obstruction." Occurrence of fecoliths in *Baddagudodara* suggests that it is a chronic ailment, thus can be revealed that *Baddagudodara* refers to Hirschspung disease.

Thus line of treatment can be planned separately or collectively as mentioned in our classics with reference to *Baddagudodara, Gudagata vata* and *Pakwashayagata vata.* The treatment of *Baddagudodara, Gudgat vata* and *Pakwashayagat vata* is *Udarwart har kriya* and surgery.

CONCLUSION

Hirschsprung disease (HD) is a disease of the large intestine that causes severe constipation or intestinal obstruction. People with HD are born with it. The large intestine, which includes the colon and rectum, is the last part of the digestive tract. The cause of HD is unclear. HD is not caused by anything a mother did while pregnant. The main symptoms of HD are constipation or intestinal obstruction, usually appearing shortly after birth. Newborns with HD almost always fail to have their first bowel movement within 48 hours after birth. HD is diagnosed based on symptoms and test results. HD is treated with surgery called a pull-through procedure. The principle of management is the removal of the aganglionic portion and a "pull-through" of the proximal ganglionated bowel. These 3 procedures have been widely used in the surgical management of the condition and the outcome and prognosis has been very good.

Sushruta have knowledge about Hirschsprung disease. They, perhaps, called it by the name *Baddhagudodara*. It was etiologically linked to deranged *vayu* (nerves) within the intestine. Although it was considered incurable, palliative surgery was done. Descriptions of the surgical operation resemble that of a sigmoid colostomy.

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