INTRODUCTION

Hirschsprung's Disease (HSD) or congenital aganglionic mega colon is characterized by absence of ganglion cells in the distal bowel and extending proximally for varying distances ***(Ziegler et al., 2010)***. The condition may be severe enough to be recognized during the neonatal period, but in other children it may not be diagnosed until later infancy or early childhood ***(Marks, 2010).***

Hirschsprung's Disease is caused by an abnormal innervation of the bowel beginning in the internal anal sphincter and extending proximally to involve a variable length of gut. The HSD is the most common cause of lower intestinal obstruction in the neonate, with an overall world wide incidence of 1:5,000 live births. Males are affected more than females (4:1) and there is an increased familial incidence in long segment disease. The HSD may be associated with other congenital defects including Down syndrome and cardiovascular abnormalities ***(Keneel et al., 2011).***

Signs and symptoms of HSD were highly variable, on one side of the edge, it may manifest in the newborn by acute signs of abdominal distension, vomiting and failure to pass meconium. On the other edge of the spectrum, the disease may not be discovered until adult life yet they have symptoms since birth ***(Coran et al., 2008).***

Complications of HSD include fluid and electrolyte imbalances, Enterocollitis and possible perforation of the distended bowel**.** Intestinal disorders are common and may cause problems for the children through life if it is not treated promptly and completely. Even with early treatment, many children require ostomies for treatment of large and small bowel disorders ***(Black and Matassarin-Jacobs, 2007).***

Hirschsprung's Disease is treated by surgerywhere the impaired part of the colon is removed and anastomosis of the intestine is performed. In newborns a temporary colostomy may be necessary and more extensive repair may follow at about age 12 - 18 months. Closure of the colostomy follows in a few months ***(Thompson, 2007).***

Outcome of surgically treated HSD is generally satisfactory with the great majority of children achieving fecal continence. Postoperative problems include recurrent Enterocollitis, stricture, prolapse, prineal abscess and fecal soiling ***(Behrman et al., 2007).***

Child with a lower gastrointestinal tract (GIT) alteration has many special needs, because of the regular necessity of eating and elimination. Lower GIT Problems affect many aspects of daily life, medications, surgery, diet alterations and special elimination needs become a way of life for many children. Activity levels, growth and development and even life span can be affected ***(Montague et al., 2008).***

Nursing care is age dependentwhere in the newborn, detection is high priority. As the child grows older, careful attention to a history of constipation and diarrhea is important. Pre operative care depends on the age and clinical condition. A child who is malnourished may not be able to withstand surgery until the physical status improve often ,this is after symptomatic treatment with enemas, a low fiber, high calorie and high proteins diet and in severe situations, the use of total parental nutrition (TPN) ***(Wong, 2006)***.

Child is cared at home, therefor it is nessesary to help the parents learn about a minimal-residue diet (i.e. one that is low in indigestible fiber, connective fiber and residue),milk, fiber foods and highly seasonal foods are omitted to eliminate chemical irritants from the intestinal tract ***(Pillitteri, 2009).***

Nurse plays vital roles with these children; providing care during surgery and acute care of the illness is only the beginning. Because of the chronic nature of this condition, the nurse must coordinate care with other specialties and arrange long-term follow-up for children and their families. Community and home-based care are essential components of care and the nurse's role must include teaching, nutritional support, emotional support, stomal management, and integration with all health care professionals involved in the child's care ***(Montague et al., 2008).***

Significance of the Study

Hirschsprung's Disease is a life threatening disorder affecting children. The incidence of Hirschsprung's Disease is one in 1000 live births, or almost 20 millions per year all over the world. In Egypt, the estimated percentage of children's HSD was varied from 5-7%; most of them were boys and more common among pre-mature children and children who have another congenital anomaly. The HSD affects both the child health status and parent's psychological status. So it is considered a big problem for both child and his/her parent ***(National cancer institute, 2011).***

So that, Preoperative preparation of children suffering from HSD is essential for the achievement of better postoperative outcomes and decrease short and long term health problems. It is important that nurses and other health professionals be knowledgeable about the care given during the preoperative and postoperative period. It is anticipated that data will be useful for nurses and all health personnel, as well as parents of affected children ***(Montague et al., 2008).***

AIM OF THE STUDY

The apresent study aimed to investigate the effect of nursing intervention on short and long term health problems of children suffering from hirschsprung's disease. This aim was attained through the following objectives:

- Assess nurses' knowledge and practice regarding short and long term health problems of children suffering from Hirshsprung’s Disease.
- Design, implement and evaluate the effectiveness of the nursing intervention regarding short and long term health problems of children suffering from Hirshsprung’s Disease.

Research hypothesis:

Nursing intervention will affect nurse's knowledge and practice in relation to children suffering from Hirschsprung's Disease.

REVIEW OF LITERATURE

Study of HSD in the pediatric age group is a relatively new area of scientific interests. It was an almost uniformly fatal disease until the breakthrough in its treatment of HSD which was made by Swenson in 1948, where it was made a sigmoid colostomy in 4 years old child with massive abdominal distension and the child made a spectacular recovery ***(Aboul-Hassan, 2007).***

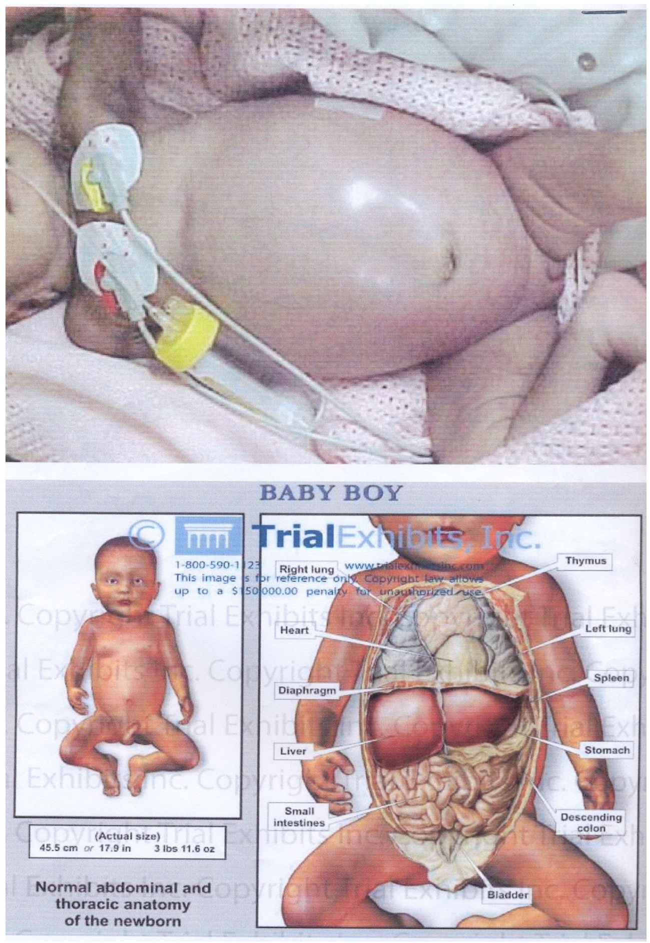
Part I:

Anatomy and Physiology of the Gastrointestinal Tract System:

Gastrointestinal tract (GIT) consists of two components: the alimentary canal and the accessory GIT organs. The alimentary canal, or GIT, consists essentially of a hollow muscular tube that begins in the mouth, pharynx, esophagus, stomach, duodenum, jejunum, small and large, intestines, rectum and anal canal. It is approximately 9m in length and it is controlled by the autonomic nervous system. Accessory organs aiding GIT function include the salivary glands, liver, biliary duct system (gallbladder and bile ducts), pancreas ***(Thibodeau et al., 2010).***

Digestion and absorption of nutrients, as well as the storage and elimination of fecal waste, take place within the the GIT. Digestion starts in the mouth as chew food. Food then passes through the esophagus to be digested in the stomach and is subjected to the action of various digestive fluids and enzymes. Partially digested food moves into the small intestine, where nutrients are absorbed. The remaining undigested protein solidifies as water is reabsorbed in the large intestine or colon, forming solid fecal matter or stool is passed to the rectum, where it is stored until it is excreted through the anus ***(Montague et al., 2008)***.

Fig. (1): Normal abdominal and thoracic anatomy.

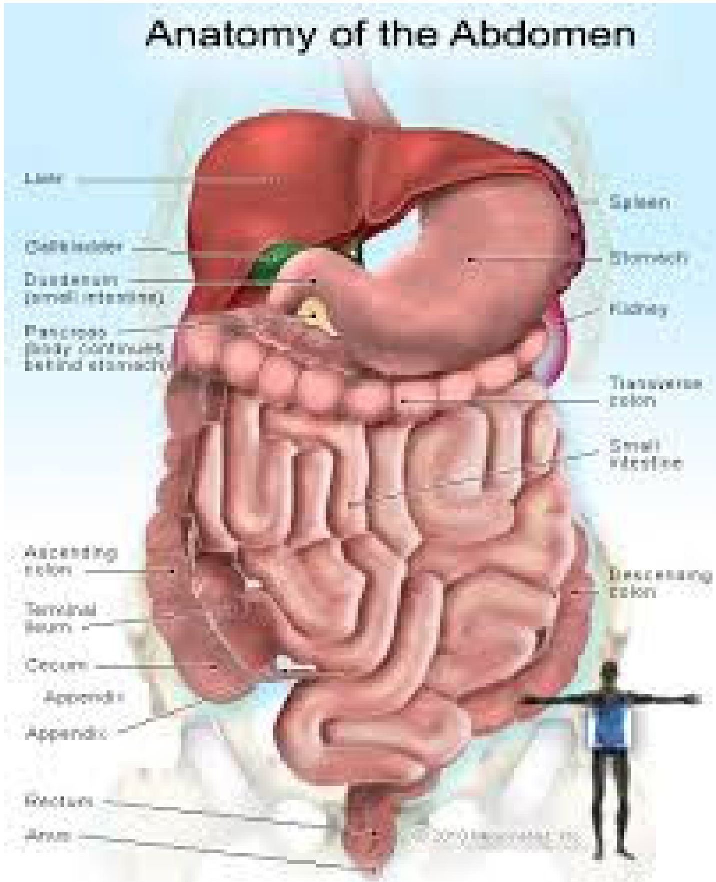


Delormier A, Harrison M and Adzick N (2010). Hirsch sprung disease in current surgical diagnosis and treatment.10th ed. London: Appleton and Laneg Company, pp.1219-1220.

The salivary glands switch into action, as soon as, food enters the mouth, and as the food continues on its journey, enzymes found in the stomach, small intestine, the pancreas and the liver continues the process. These organs deliver their secretion to the duodenum through the hepato pancreatic ampullae, also known as Oddi's sphincter, which helps maintain the function of the tract ***(Tortora and Grabowskin, 2008).***

Lining of the gastro intestinal tract, the walls are made up of mucous membrane, constructed in such a way that the various parts can act independently of each other The walls of the gastro intestinal tract consist of four layers. The adventitia or outer layer consists of a serous membrane composed of connective tissue and epithelium in the abdomen, it is called the visceral peritoneum. The musclaris mostly consist of two layer of smooth muscle, which contract in a wave-like motion, which are made of skeletal muscle that aids swallowing. The two smooth muscle layers consist of longitudinal fibers in the outer layer and circular fibers in the inner layer. The contraction of these two layers of muscle assists in breaking down the food, mixing it with the digestive secretions and propelling it forward. This action is referred to as peristalsis. The sub mucous layer is highly vascular as it houses plexuses of blood vessels, nerves, lymph vessels, and tissue. It consists of connective tissue and elastic fibers. The mucosa is a layer of mucous membrane that forms the inner lining of the GI tract ***(Tucker, 2005).***

Fig. (2): Anatomy of the abdomen



Moral L, Bove M and Karasic R (2011). Pediatric emergency nursing procedures. London: Jones and Bartlett Publishers, Boston Company, p. 219.

Small intestine begins at the pyloric sphincter and coils its way through the central and lower aspects of the abdominal cavity and joins the large intestine (colon) at the ileo caecal valve. The small intestine is divided into three separate segments: the duodenum, jejunum and ileum. The nerve supply for the small bowel is both sympathetic and parasympathetic ***(Thibodeau et al., 2010).***

It is approximately 6.5m long and has a diameter of approximately 2.5cm. The walls of the small intestine consist of the same four layers as the rest of the Gastro intestinal tract; however, both the mucosal and sub mucosal layers are modified. The mucosal layer consists of many glands called intestinal glands. These glands are lined with glandular epithelium and secrete intestinal juice. The sub mucosa in the duodenum contains glands secrete mucus which is alkaline; this is designed to protect the small intestine walls from the acid in chyme and prevent the enzymes from acting on the walls ***(Ellis et al., 2007).***

Small intestine is further modified in that throughout its length, the epithelium covering the lining and the mucosa is made up of simple columnar epithelium. This contains both absorptive and goblet cells. The absorptive cells actually contain projections described as "finger-like" the projections are known as microvilli and allow the small intestine to deal with larger amounts of digested nutrients, having simply increased the surface area for digestion ***(Thibodeau et al., 2010).***

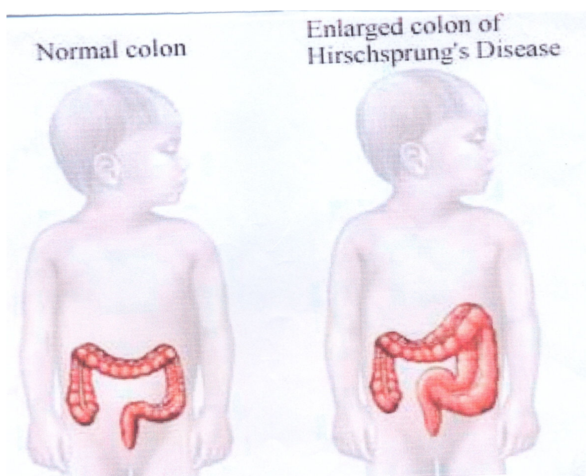
The main function of the small intestine is digestion and absorption and its make up is designed to help this process. The chyme is broken into small molecules that can be transported across the epithelium and into the blood stream. This occurs in the presence of pancreatic enzymes and bile, which are important in the digestive process. The small intestine absorbs most of the water, electrolytes and glucose from the chyme. The small intestine not only provides nutrients to the body but also plays a critical role in water and acid-base balance ***(Tortora and Grabowski, 2008).***

The chyme from the stomach moves along the small intestine at approximately 1cm, it can remain in the small intestine for up to eight hours. The chyme is moved along by peristaltic movements, which are controlled by the autonomic nervous system. Digestion is completed in the small intestine with the aid of juices from the liver and pancreas. Waste is then transported to the large intestine for disposal. The superior mesenteric artery supplies the whole of the small intestine and the venous blood is drained by the superior mesenteric vein that links with other veins to form the hepatic portal vein ***(Ross et al., 2007)***.

Function of Colon:

There are differences in the large intestine between different organs. The large intestine is mainly responsible for storing waste, reclaiming water, maintaining the water balance, absorbing some vitamins, such as vitamin k, and providing a location for flora-aided fermentation ***(Barklin, 2008).***

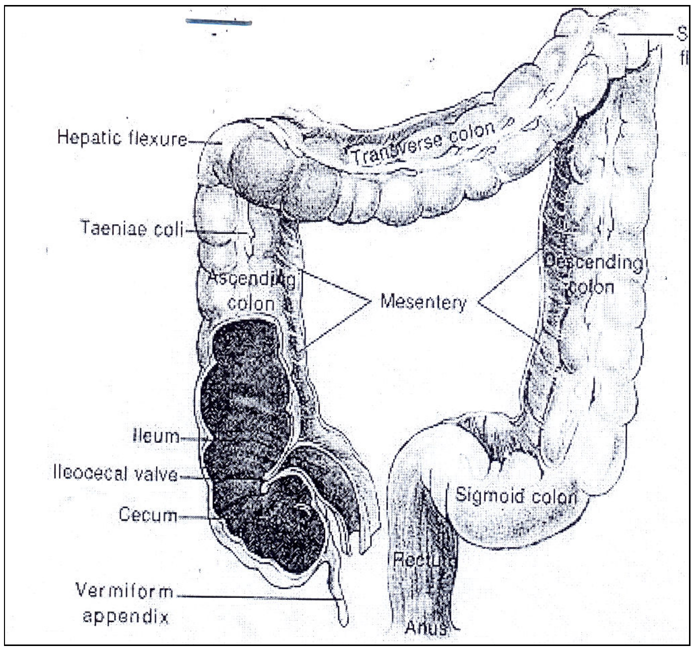
Fig. (3): Difference between normal colon and mega colon



Curley M, Smith J and Moloney-Harmon B (2011). Critical care nursing of infants and children. Tokyo: W.B. Saunders Company, pp.524.

Large intestine is also called distended intestine because of its ability to distend. It forms a three-sided frame around the small intestine leaving its inferior area open to the pelvic. It is designed to absorb water from the contents of the small intestine that pass into it ***(Lancaster, 2010).***

Fig. (4): Anatomy of the large Intestines



Adopted from Sloan, E (2009). Anatomy and Physiology of the large intestine; An Esau Learner, (2nd ed.) London and Bartlett Publishers Con. P. 296

The large intestine is approximately 1.5m in length and extends from the ileum to the anus. Its size decreases gradually from the caecum, where it is approximately 7cm in diameter to the sigmoid, where it is approximately 2.5cm in diameter. The large intestine has four segments: the caecum, colon, rectum, and canal. The colon is divided into four sections: the ascending colon, transverse colon, descending colon and sigmoid colon ***(Keshav, 2006).***

The large intestine also houses a variety of bacteria. These bacteria, known as commensals, live happily in the bowel and generally do not cause any problem. In fact, they play an important part in digestion; they ferment carbohydrates and release hydrogen, carbon dioxide and methane gas. The bacteria also synthesize a number of vitamins such as vitamin K and some different B vitamins. They are also responsible for break down the bilirubin into urobilinogen, which gives the faeces its characteristic brown color. However, outside the bowel the bacteria can cause illness and even death. Two substances arising from bacterial decomposition give the characteristic odour of feces; these are indole and skatole ***(Siegfried, 2008).***

The blood supply to the large intestine is mainly by the superior and inferior mesenteric arteries. The internal iliac arteries supply the rectum and anus. Venous drainage is mainly by the superior and inferior mesenteric veins and the rectum and anus are drained by the internal iliac veins. The nerves supplying the large intestine are via the sympathetic and parasympathetic nerves ***(Townsend et al., 2009).***

The following constitutes the main functions of the large intestine: (a) absorbs about 80% to 90% of the water and electrolytes from the remaining chyme and reduces the chyme from fluid to a semi-solid mass, (b) produces only mucus, its secretions contain no digestive enzymes or hormones, (c) digests small amounts of cellulose, and produce a few calories of nutrients to the body each day. The bacteria present in the colon produce vitamins such as vitamin K, riboflavin, and thiamin; and various gases ***(Cole, 2010)***. The average person expels about 500ml of flatus (gas) per day. Flatus is composed of Nitrogen (N2), Carbon dioxide (CO_2), hydrogen (H2) and two amines ***(Saladine, 2009)***.

Elimination of faeces or flatus via the rectum is normally voluntarily controlled by using sphincter muscles. However, a stoma does not have sphincter muscles. Patients are therefore not able to control elimination of faeces or flatus via their stoma. Some people with a sigmoid colostomy report their stoma as 'controlled' to act at particular times. This routine is actually due to regular mass movement of faeces after certain meals ***(Guyton and Hall, 2009)***.

Gases can also enter the gastrointestinal tract from swallowed air or by diffusion from blood. Most swallowed air is usually eelled by belching and only small amounts of gas are presenting the small intestine. The amount of gases entering or forming ill the large intestine daily averages 7 to 10 liters the majority of this being absorbed through the intestinal mucosa and only about 500 ml being expelled as flatus ***(Guyton and Hall, 2007).***

Feaces are 75% to 80% water. The solid material is about one third bacteria and the rest consists of 2 to 3 percent nitrogen, and organic and inorganic residues from digestive secretions which includes mucus and fat. Feces also contain variable amounts of roughage, or indigestible fibbers and cellulose. The brown colour is from bile pigments, the odor from bacterial actions ***(Solomon et al., 2003)***. Each day about 500 ml of food residue enters the large intestine. It undergoes no further chemical digestion, but its volume is reduced over the next 12 to 24 hours as the colon absorbs water and electrolytes (especially NaCI) from it. The average adult voids about 150ml of feces per day, consisting of 75% water and 25% solid matter. The latter is about 30% bacteria, 30% undigested dietary fibre, 10% to 20% fat, and smaller amounts of protein, sloughed epithelial cells, salts, mucus, and other digestive secretions ***(Cohen and Guillem, 2012)***.

The most common Anomalies of the Gastrointestinal Tract according to Montague et al. (2008) Include:

- Hirschsprung's disease,
- Pyloric and duodenal obstruction,
- Meconium aspiration, Meconium peritonitis, and the meconium plug syndrome,
- Colon Artesia,
- Uncommon forms of neonatal bowel obstruction,
- Short Gut syndrome and
- Duplication of the alimentary tract.

Embryology of Gastrointestinal System (GITS):

Gastrointestinal tract is formed in the first 4 weeks of embryological development, so congenital defects can be traced to this period. Development of GIT is anatomically complete at birth but physiologically immature, affecting enzymes,