Summary and coclusion

Hirschsprung's disease: is a congenital disorder characterized by the absence of enteric neurons in the distal colon and rectum. The aganglionic gut loses its tonic neural inhibition and thus remains contracted, obstructing the passage of food residue. The most accepted theory Of Hirschsprung's disease is due to a defect in the craniocaudal migration of neuroblasts originating from the neural crest that occurs during the first 12 weeks of gestation (colonic development).

Hirschsprung's disease is a worldwide accepted entity whose diagnosis is based on rectal suction biopsy. This technique allows sampling of a mucosal-submucosal specimen required for proper ENS evaluation. The most important diagnostic features of HD are the combination of hypertrophic nerve trunks and aganglionosis in an adequate specimen. Acetylcholinesterase staining the best diagnostic technique to demonstrate hypertrophic nerve trunks in lamina propria mucosae, but many pathologists from different centers still use H&E staining effectively. Newly developed diagnostic kits for enzymatic-histochemistry will likely increase the use of AChE for the diagnosis of HD. The new industrial kits lyophilize the components of the medium that can be sent at room temperature anywhere in the world. Although barium enema is not essential to confirm the diagnosis of HD; in many cases, it is useful in evaluating the level of aganglionosis and aids in the decision regarding the surgical approach (ie,transanal, transabdomin -al laparoscopic,or open). Although anorectal manometry is frequently used in association with rectal biopsy and barium enema in evaluating patients for HD, it is not routinely necessary. Intraoperative pathological evaluation of the extent of aganglionosis is

mandatory to be sure that normal aganglionated bowel is used for a colostomy or pull through procedure. In some instances, intraoperative biopsies may also demonst- rate the possible presence of associated proximal hypoganglionosis or IND ***(Hisayoshi et al., 2007)***.

The LATEP and the TOSEPT have become the two most popular methods for primary repair of HD in infants and children. The LATEP is preferred in neonates and children undergoing a primary operation for two reasons. First, laparoscopic biopsies allow confirmation of the extent of the aganglionosis before any division of the colonic mesentery or rectal ablation occurs. This is especially advantageous in the neonate in whom the contrast enema can be somewhat unreliable in predicting the level of the transition zone. If a long segment of aganglionosis is identified, delaying the definitive operation until the histopathologic results are available from the permanent sections may prevent unnecessary resection of long segments of colon due to errors in frozen section analysis. Second, the use of laparoscopy helps in performing a tensionfree coloanal anastomosis by releasing the restraining ligaments to the descending colon and ensuring that there is no twisting of the pull-through colon during the anastomosis. In a neonate, unless contraindications are present, LATEP is performed as soon as the diagnosis is confirmed by suction rectal biopsy

Transanal one-stage endorectal pull-through (TOSEPT) offers an improved approach to the child with Hirschsprung disease during neonatal period. Data suggested that TOSEPT is efficacious and safe in the management of newborn with HD. The technique is not difficult, and associated with acceptable short-term outcomes. Further studies

documenting the long-term results of this approach, particularly with respect to continence and stool frequency will be needed ***(Obermayr et al ., 2008)***

In case of long aganglionosis segment or of impossibility to mobilize the proximal colon, a 2–3 cm mini-laparotomy was used to mobilize the splenic flexure in order to gain an anastomosis without tension**.**

Hirschsprung's disease is a neurogenic intestinal obstruction with potential for chronic illness. A wide spectrum of complications has been reported following definitive repair of Hirschsprung's disease . Enterocolitis remains the most serious late complication following definitive repair. Continued advances in our understanding of the disturbances in bowel motility and the immunological and neurohormonal forces involved in this disorder will result in an improving prognosis. Traditional multistage procedures still have a role,especially in the very small, critically ill child. The longterm complications of one-stage and laparoscopically assisted procedures are currently not clearly known. Fortunately, the majority of patients with Hirschsprung's disease do quite well following definitive operation regardless of the technique employed. The great majority (94%) of children will become well-adjusted members of society. Early development milestone deficiencies appear to improve over time. Appropriate preoperative conference with family members must include a candid discussion of the importance of realistic expectations and the needfor close parental surveillance for late complications ***(moore et al.,1994)***.