

## *Summary*

Anorectal anomalies include a wide spectrum of defects in the development of the lowest portion of the intestinal and urogenital tracts.

These anomalies occur in the fifth to seventh weeks of fetal life during development of the rectum and anal canal. The embryologic basis of such anomalies is failure of descent of the urorectal septum thus failure of partitioning the cloaca. The level to which this septum stop to descend determines the type of anomaly that is present.

Normally, the rectum begins at the level of the sacral promontory. It then descends along the curvature of the sacrum and coccyx and ends by passing through the levator complex, at which level it abruptly turns downward and backward to become the anal canal.

Several mechanisms serve to maintain the function of anal continence, these include internal and external sphincters, rectal and anal sensory perception, neuropathways and reflexes, reservoir function and lastly mechanical factors as anorectal angulation.

In patients with Anorectal anomalies, the rectum may fail to descend through the levator complex. Instead, the rectal pouch ends blindly in the pelvis, above or below the levator ani muscle. In most cases, the blind rectal pouch communicates more distally with the genitourinary system or with the perineum through a fistulous tract.

The origin of anorectal malformations is in early embryonic life. Expectedly, associated anomalies are common, ranging in incidence from 40-60% in different series. The commonest of these are in the urinary tract, the vertebral system, and in the developing heart. A known association of anomalies is known as the "VACTERL syndrome" (vertebral, anorectal, cardiac, tracheo-oesophageal, renal and limb) This suggests that the stimuli that induce abnormal development of anorectum

operate universally in the developing fetus and may cause maldevelopment of several organ systems .

Traditionally, the anatomic description of Anorectal anomalies has characterized it as either "high" or "low" or "Intermediate" depending on whether the rectum ends above or below the levator ani muscle complex or partially descends through this muscle .

A more recent clinical classification "Krickenbeck classification" describing Anorectal anomalies according to the type of fistula present (Perineal, Rectourethral, Rectovesical, ..etc). This classification offers the advantage of better decision making in treatment of Anorectal anomalies according to their types.

Diagnosis of these anomalies is carried out by a large number of diagnostic tools starting prenatally by ultrasonography (dilatation of the rectosigmoid area and Intestinal coprolites).

After delivery diagnosis can be first established clinically by history or by routine physical examination of the newborn revealing any deformity (e.g. Bucket-handle, Forchette.. etc.), or abnormally sited anal opening (fistula) or even its absence.

Investigations can be employed after delivery aiming at exclusion of any associated anomalies as well as identification of type and level of the anorectal anomaly. These investigations constitute a wide spectrum of laboratory and imaging studies as urine analysis, ECHO cardiography, Abdominal and Transperineal ultrasonography, Lateral pelvic radiography, Distal colostography, MRI and more. All these investigations have the goal of individualizing the proper treatment decision for each case of anorectal anomalies.

Generally, There is rarely any urgency in management of newborn with Anorectal anomalies , as most patients tolerate the low bowel obstruction without symptoms for longer than 24 hours as the upper

gastro-intestinal tract is decompressed by a nasogastric tube. In this period, an inapparent opening of a perineal fistula may appear, on the other hand investigations are carried out to decide the appropriate surgical approach.

Early decision making in treatment of anorectal anomalies is crucial. The first step is to decide whether the repair would be carried out in a single, two or three stage repair.

The role of the colostomy in the definitive correction of anorectal malformations is controversial in regard not only to indications for its use but also to its type.

Definitive repair in the form of Anorectoplasty with a posterior sagittal approach (PSARP) has the advantage of intraoperative identification of anatomical relations thus minimizing iatrogenic injury and providing proper placement of the pulled rectum within the sphincter complex.

In anomalies where the rectal pouch is above or merely at the level of the levator complex, an additional abdominal component is needed for proper mobilization of rectum and dividing any fistula if present. This abdominal component is in the form of Laparotomy or, more recent, Laparoscopy (Laparoscopically assisted anorectoplasty).

Follow up of these patients is as much important as surgical repair. This will be in the form of laxatives regular diet to prevent chronic constipation, fecal impaction should be prevented at all costs. Regular anal dilatation with a gradually increasing sets of Hegar's dilators where parents are educated to carry out dilatation without traumatizing the anal verge of their child thus preventing postoperative anal stenosis.

On the other hand, Patients with cloacal anomalies need specialized follow-up care after puberty to assess sexual function and to correct genitourinary problems in addition to special obstetric follow up during pregnancy and delivery.

Anorectal Anomalies still represent a challenging field regarding diagnostic tools, surgical approach and follow up. However, with continuous research and prospective studies we will be able to improve our results and achieve the goal of clearing out the functional and psychological impact of such conditions on the affected children and their parents.