SUMMARY

Anorectal anomalies are one of the most common congenital anomalies, with overall incidence 1:5000 live births. They provoke a great interest due to their variability and association with neonatal intestinal obstruction and anal continence.

Anatomically, the classification of the anal sphincter into 3 parts other than the puborectalis sling has been a point of maximum controversy, but de Vries and Pena found that, there is no separation between one group of muscles and the other. The vertical fibres (muscle complex and parasagittal fibres) create two corners where they meet and that is the anterior and posterior limit of the anus.

Proper continence is achieved by many factors including external sphincter, puborectalis, skin, rectal mucosa, intract pathway and good intestinal function.

Embryologically, the main phases of anorectal region development are, formation of the cloaca, division of the cloaca into the urogenital sinus and rectum, and development of the anal canal. The deviation from normal embryology lead to cloacal malformations, which are similar in both sexes until the Müllerian ducts develop in the female, but undergo atrophy in the male. Faulty partitioning of the internal or external cloaca results usually in rectal or anal fistula opening in the urinary or genital tracts or onto the perineal skin. Less commonly the rectum or anus ends blindly. The different types of anorectal anomalies
have an intimate relationship to their embryogenesis and the way of their formation and development, which are described in detail.

Classification of these anomalies is a difficult process. They are classified in many ways, starting early in 1904 by Wood and in 1908 by Keith, passing by Ladd and Gross, (1934) and ending by 3 famous classifications. An international classification (1970), based on anatomical configuration of visceral deformities in relation to the principle muscle of continence (puborectalis and levator ani). The clinical classification (1974) depends on the clinical presentation of the neobron infant with ileus or without, and the level of these anomalies in relation to levator ani muscle. Recently, the Wingspread classification is developed in 1984. It defines these malformations according to sex and level at which arrest of rectal descent occurred.

Diagnosis and accurate determination of the exact type of the anomaly is an urgent problem. Depending on such information, a decision can then be made whether to perform a primary perineal operation in the neonatal period or to perform colostomy only, postponing definitive operation until a later period.

Clinical presentation of the neobron carry many important symptoms and signs up to 24 hours. After this, if none of the characteristic features become evident, a radiographic evaluation is indicated.

In inverteography, the relation of the gas shadow to the PC line, I point, I line, anal pit line, PCI triangle, U line, M point and M line is
of great diagnostic value. Sometimes a radioopaque dye studies are needed. Fistulography, needle aspiration and injection of the rectal pouch and micturition cystography may reveal the exact passage of the anomalies.

US, CT and MRI are valuable in documentation of all perineal structures including sphincter defects and assemtery and measure the thickness of the internal sphincter. Also, they are useful in assessment of the perianal structures.

In addition, US may be valuable intra-operatively during sacroperineal anorectoplasty in tunneling the center of sphincter muscle from the identified entrance to the outlet with visual confirmation on the US screen.

Postoperatively, CT can clearly demonstrate the anatomic location of the pulled-through intestine in relation to the levator sling.

MRI clearly reveals the extent of the pelvic musculature even in patients with severe sacral agenesis. It is a useful tool in planning operative strategy and predicting the outcome by providing information about pelvic musculature in relation to the size of the patient.

EMG can determine the exact site of the external anal sphincter by detecting the contractile activity of striated muscles and by obtaining separate recording from the external sphincter and from puborectalis muscle.

Anorectal anomalies is frequently associated with anomalies of other viscera or limbs and unfortunately most of them are serious. This
association is due to the occurrence of major organ differentiation at the same time of cloacal subdivision. The genitourinary system is the most serious and frequent site of associated defects in anorectal malformation. So each case must be evaluated early endoscopically and radiologically to guide immediate and later treatment.

Our subject is a kind of multiple complex syndrome, which need an urgent and accurate management. The first attempt to free the bowel and its pull down and suturing it to the skin was described by Amussat in 1935. The low type anomalies have long been successfully treated by physicians, midwives, and laymen by rupturing the anal membrane by a finger or an instrument. The simple form of anoplasty is described by Nixon in 1967, but is not widely known. It is done in cases of covered anus. Anal cutback is favoured for anocutaneous fistula, covered anus complete and covered anal stenosis, imperforate anal membrane and anal membrane stenosis. As an alternative to cutback, many surgeons do anal transplant to establish the bowel amid the voluntary external sphincter.

In high and intermediate lesions the aim of surgical treatment is to have a maximal continence of the newly fashioned anal canal. The long history of attempts at surgical corrections over the past 150 years revealed many surgical procedures with many modifications.

In spite of the various arguments about one-stage or staged programme surgery, under certain conditions all surgeons advise a preliminary colostomy and later rectoplasty.
A colostomy with two stomas was obviously superior to any other form of colostomy. It is a problem to place it distal in the sigmoid and not to leave enough length for the pull-through procedures. Facing this the surgeon may be obliged to refashion the colostomy again prior to rectoplasty.

In the time of its introduction, Stephens procedure revolutionized the treatment of these anomalies and has formed the basis of every advance then.

Swenson and Donnellan appreciating the concepts of Stephens, endeavored to define the correct plane through the puborectalis sling from the abdominal route and fed the neorectum through this sling to the perineum.

Kiesewetter and Rehbien, utilizing the concepts of Romuldi, avoided any dissection outside the rectum by bringing the neorectum down inside a demucosed sleeve of the original rectum, in addition to Stephens sacral approach. However, the puborectalis definition is blind.

Mollard et al., approached the sphincter complex by a perineal incision anterior to the expected neoanus. A plane to the fistula and puborectalis is opened up immediately behind the urethra with positive visualization of both fistula and levator, but limited recognition of the external sphincter component.

De Vries and Penna reintroduced the perineal approach to the rectum, aided by electrostimulation of all muscle fibres, commencing with precise definition of the maximum confluence of external sphincter
components at the proposed anal site. Dividing the muscles in the sagittal plane affording a wide access to divide a fistula under vision, to mobilize and taper the terminal rectum, and then to reconstitute all muscle elements accurately around the neorectum in precisely the correct anatomic position. Through this approach the majority of lesions (even - high level anomalies) can be dealt with, without abdominal exposure.

Yokoyama and his colleagues using electrostimulation and perineal exposure, combine some elements of the de Vries - Pena PSARP with the Rehbein abdominal submucosal dissection. The approach to the fistula is via the abdomen and essentially recognizes a potential internal sphincter in the terminal rectum at the fistula, through which the neorectum is tunneled. The external sphincter is identified but not divided. It is not clear how the pathway through the center of the external sphincter is determined, and the procedure always requires abdominal laparotomy.

Durham SImth utilizes all the contributions of these outstanding surgeons, and add another outstanding contribution to the actual skin anastomosis - the skin lined anus of the Nixon anoplasty. Perineal rectoplasty is utilized in all intermediate lesions and can be used in many high lesions. Only the very high lesions, or in complex anomalies, it is sometimes necessary to open the abdomen.

The functional status of the patients are sequentially evaluated using clinical criteria, Kelly clinical score, thorough bowel irrigation or rectal stimulation to evaluate motility, Kelly score and radiologic
correlation and manametry, Kiesewetter's criteria or multifactorial assessment including change in anal canal force, rectal distinsion, palpable nature and position of puborectalis sling, rectal sensation, smooth muscle activity and rectal pressure following distinsion.

These evaluations revealed that, most if not all of those individuals who are truely continent following surgery for supravelator defects have had the full thickness of their terminal bowel preserved. In all likelihood, they have also had at least a portion of their internal sphincter preserved. Similiarly, the findings in favour of carrying out a definitive repair in early infancy are becoming persuasive. It is clear that the further proximally the dissection around the terminal bowel is extended, the greater the likelihood of ablating from the rectum those sympathetic and parasypathetic, extrinsic nerves necessary for its optimal function.

Complications of treatment may be any of that encountered generally in surgery, specially anal and rectal stricture, prolapse of rectal muscosa, rectal inertia, mucous discharge, megasigmoid and megarectum and other rare complications.

So, the child with anorectal anomalies may be safed a life time of misery and social seclusions by the surgeon who is skill, deligence and judgement performs the first operation on the malformed rectum.