

Anorectal Malformations in Children – A Review

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Anorectal malformations are a wide group of malformations characterized by an abnormal anal opening. Most often detected at birth, this abnormality can range from stenosis of the normally sited opening, to an abnormal position of the anus, to complete absence of the anus. Associated with the apparent problem with the anus, are a range of local abnormalities affecting the rectum and the adjacent genitourinary system. These are most often fistulas between the rectum and the urinary tract in males, and the reproductive tract in females. The most bizarre manifestations are the variations seen in the most severe of the female malformations, the common cloaca, where the lower urinary tract, the uterus and vagina and the anorectum are fused in an array of combinations.

The overall incidence of anorectal malformations is approximately 1 in 4000 live births. The reported sex incidence in the western literature shows a slightly higher incidence in the male.

The origin of anorectal malformations is in early embryonic life. Expectedly, there is much activity in the developing embryo at this time and any insult causing an anorectal malformation may well affect other systems as well. Thus, associated anomalies are common, ranging in incidence from 40-60% in different series. The commonest of these are in the urinary tract (approx 35%), the vertebral system (approx. 18%), and in the

developing heart (approx 10%). A known association of anomalies is known as the VACTERL group (vertebral, anorectal, cardiac, tracheo-oesophageal, renal and limb).

As earlier mentioned, the manifestation of these malformations is over a wide spectrum, ranging from the least severe, with the least chances of associated anomalies, and the best chances of achieving good continence, to the most severe, with a high incidence of associated anomalies, and relatively poorer chances of continence in later life. Surgery in the latter group is predictably more complex and is often done in stages.

The least severe variant in both sexes is anal stenosis, where a stenosed anus opens at the normal position. An ano-cutaneous fistula or perineal fistula is characterized by a small opening on the perineum, anterior to the anus, through which a small bead of meconium may be seen.

More severe variants are described separately for males and females. In both these, an opening is not visible at the normal anal site.

In the male, recto urethral fistulas occur either in the bulbar or in the prostatic urethra. A still more severe type is the recto vesical fistula which opens at the bladder neck.

In the female, the absent anus is associated with a vestibular fistula, in which the anus opens just posterior to the vagina within the vestibule. Recto vaginal fistulas are also described, but very rare. The most complex types in females are the common cloacas, where the urethra, vagina and rectum share a common opening. In this variant, there are

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often other complex situations, like bifid or hemi uteri, with the rectum sometimes opening into one of the uterine components, or into one of the tubes. Associated anomalies are commoner in these severe variants.

Rarely, both males and females may have a blind ending rectal pouch without a fistula. Another uncommon type is known as rectal atresia, in which a normal looking anus is seen, but there is a blind ending rectum above, which may be separated from the anus by a membrane or a thicker septum or longer cord.

Clinical Features

Many anorectal malformations are now diagnosed antenatally, by ultrasound.

At birth, a general examination in any newborn should include the perineum, when the absence or abnormal location of the anus will be apparent.

In the male, besides the absent anus, a note must be made of the anal pit, the site of the absent anus. Sometimes, a "bucket handle" deformity is seen here, which is an indicator of a low variety malformation. At times, a prominent midline raphe is seen anterior to the anal pit. This may be the tract of the no perineal fistula, and may run anteriorly to the scrotal raphe. The fistula may be of small caliber; hence it may take upto 24 hours for it to be apparent. The reason for this is that it takes this long for ingested gas to travel down the gut to the blind rectum. Only when this generates a sufficient pressure to force the meconium into the fistula, will it actually be apparent.

A hypospadias may be present as an association. Additionally, there may sometimes be a bead of meconium at the urethral meatus, indicating a fistula.

In the female child, the diagnosis rests on the appearance of the perineum. Normally, there are three visible openings - the most

anterior being the urethra, followed by the vagina, both of these being within the vestibule. Behind the perineal body is the anus.

The presence of three openings, with the anus not being at its normal site is indicative of either a perineal fistula, formerly called the anterior perineal anus. If the third opening is seen within the vestibule, it is a vestibular fistula. Two openings only, indicate two extremely rare clinical entities, namely a recto vaginal fistula, or a blind ending rectum with no fistula. A single opening indicates the common cloaca.

Investigations

A period of about 24 hours is used to observe the child. As mentioned earlier, this period is required for the meconium to appear at the perineum in case of a fistula. In this period, a clinical examination will indicate if an associated anomaly is suspected, and a 2-D echo and ultrasound examinations will rule out cardiac or renal anomalies. The urine is collected and screened for flakes of meconium. The baby is not offered any feeds, and a Nasogastric tube is inserted to decompress the stomach.

The decision to be made next is whether a colostomy is indicated.

In the male child, if at the end of 24 hours, or sooner, if sufficient abdominal distension has occurred, meconium is seen at the perineum, either surgery or dilatation will relieve the obstruction.

If meconium is seen at the urethral meatus, it is indicative of a recto urinary fistula, which will need a colostomy. If the picture is unclear, an X-ray is indicated to clarify the further course. The traditional approach was to perform an invertogram, i.e. an X-ray taken with the baby suspended upside-down, to allow gas to rise upto the blind

rectum, and delineate its distance from the perineum. An alternative that is now popular is an X-ray taken in the knee chest position with the baby being held prone, and shot from the lateral side. The results of either are the same. The information available from this X-ray helps in determining whether the anus will be safely accessible from the perineum, or whether a colostomy will be indicated.

In the female, a perineal examination is usually sufficient to make this decision. If three openings are seen, and meconium is seen at the perineum and the orifice is sufficient to decompress the rectum, the baby may be observed. If the orifice is small, dilatation may be done to enlarge it and to buy time. If two openings are seen, without any meconium, an X-ray is indicated to demonstrate if there is a blind ending rectum that may be approached perineally. If not, a colostomy will be needed. In case of a common cloaca, there may be associated genitourinary anomalies which will be seen on ultrasound. Besides a colostomy, a vesicostomy or a vaginostomy may be needed. The management of the common cloaca is complex and should be done by a paediatric surgeon experienced in this subject.

The definitive surgery of an anorectal malformation is today done most often through the posterior sagittal trans sphincteric approach. This approach is made through a midline incision from the mid-sacrum to the anal pit. The muscles of continence are split in the midline to access the blind ending rectum, isolate the fistula and close it, and to mobilize the rectum and bring it down ensuring that it lies within the confines of the muscles of continence.

In case of a recto vesical fistula, an abdominal exploration is additionally needed to close the fistula and mobilize the rectum. Laparoscopy is also being used in these

situations.

In females with a vestibular anus, a perineal approach may be used to separate the rectum from the, often closely adherent, vagina. In case of a common cloaca, a complex reconstruction is needed.

Following surgery, a period of anal dilatation is needed, following which the colostomy can be closed. For a period, sometimes lasting over a year, there may be problems related to bowel movements. Frequent stools may cause perineal excoriation. In children with low anomalies, and with vestibular fistulae, constipation following surgery is often severe.

After the age at which stool control is achieved, generally at the age of three years, a child can be assessed for results based on continence. Various parameters have been used to define continence, and a normal stool pattern. A general definition would be the ability to have a voluntary bowel movement, without staining of the underclothes.

An overall incidence of 40% continence is reported. This includes the whole spectrum of anorectal malformations, including the low and intermediate types (the perineal, urethral and vestibular fistulae), where the continence is expected to be better than in the vesical fistulae and common cloacas. Some children will lose their ability to control their stools during an episode of diarrhoea, while others may be constipated, and may be encopretic, and thus soil their underclothes. The results depend on the inherent capacity of the muscles of continence, as well as on the surgical technique used.

Children who persist in being incontinent should be evaluated by an experienced paediatric surgeon. Possible solutions are a repeat operation, a bowel management programme (which includes dietary management and regular enemas),

performing a continent appendicostomy to aid bowel management, or doing a sling operation for improving continence.

A continent appendicostomy is based on the principle that enemas given per anum are difficult to administer, and will clean the rectum and colon in a retrograde fashion, leading to incomplete cleansing, and thus early soiling due to fresh stools reaching the rectum. Through an appendicostomy, a tube can be introduced, which can clean the colon in an antegrade fashion, leading to better continence.

Conclusion

Anorectal malformations are a spectrum of malformations which need surgical treatment in the infancy. The aim is to provide the child with a normally situated anus, and normal bowel function. This is dependant on the severity of the malformation, and on the expertise while performing the surgery. The overall

continence rate after surgery is approximately 40%. Incontinent children should be evaluated by a specialist to consider conservative and surgical options to achieve social continence.

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ALLERGIC RHINITIS : COMMON, COSTLY, AND NEGLECTED

Some 22 million Americans visited their doctor because of allergy symptoms, with doctors' bills accounting for about a third of allergy costs. The remainder mostly went on prescription drugs, such as the antihistamines loratadine and cetirizine, with an average annual spend of \$ 520 per person.

Studies to date have found a high prevalence of seasonal allergic rhinitis across all of western Europe. However, the seasonal and perennial nature of this condition often thwarts epidemiological surveys. To improve the monitoring of the condition, WHO has proposed that allergic rhinitis be classified by whether symptoms are intermittent or persistent.

In other words, squeaky-clean modern life could be a contributing factor, and may indeed be harmful to children. Another commonly cited contributing factor is environmental pollution.

More specialists would of course be a welcome start, just to add to the confusion, the evidence base for effective treatment and management is scarce.

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