

MANAGEMENT OF CONGENITAL ANORECTAL MALFORMATIONS

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Introduction

Anorectal malformations constitute one of the commonest congenital anomalies encountered in the newborn. The enormity of the problem can be judged by the fact that 1 out of every 5000 newborn babies is liable to have this condition. On rough estimates nearly 15000 babies with anorectal malformations are born every year in the densely populated state of Uttar Pradesh, with one of the highest birth rates in the country. With less than ten pediatric surgeons active in the state at present, most of these cases have to be managed at least initially by general surgeons in the next two decades. It is, therefore, imperative that every person aspiring to be a surgeon should have a working knowledge of how to tackle this major but essentially curable birth defect.

Cure in anorectal malformations encompasses anatomical restoration along

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with functional effectiveness. It is a life-long misery for a child to have an incontinent opening in the perineum. It would be better, anytime, to have a permanent colostomy in the anterior abdominal wall where at least a proper colostomy appliance can be fitted. The responsibility of management of this problem is thus great and no surgeon should take it casually.

Classification

Anorectal malformations present with a wide spectrum and, therefore, there have been numerous attempts at classification. To overcome the drawbacks of the various classification systems, some of which were too simple and others too complicated, a group of surgeons from all over the world met at Wingspread in USA in 1984 and formulated a classification which is reproduced in *Table I*.

The essentials of this classification for a preliminary student are as follows:

1. The terms high, intermediate and low are in relation to the terminal end of the bowel remaining above (high), within (intermediate) or below the levator ani muscle (pelvic floor) which is the "main muscle of continence.
2. Most female babies have low or intermediate anorectal malformations while the reverse is true for males.
3. Female babies usually have a fistula from the terminal end of the bowel opening externally while in male this fistula is usually well hidden and, therefore, investigations are required to de-

TABLE I-Wingspread Classification (1984)

Male	Female
<i>High</i>	
Anorectal agenesis	Anorectal agenesis
With	With
rectoprostatic	rectovagin;11
urethral fistula	fistula
Without fistula	Without fistula
Rectal atresia	Rectal atresia
<i>Intermediate</i>	
Rectobulbar	Rectovestibular
urethral fistula	fistula
	Rectovaginal
	fistula
Anal agenesis	Anal agenesis
without fistula	without fistula
<i>Low</i>	
Anocutaneous	Anovestibular
fistula	fistula
Anal stenosis	Anocutaneous
	fistula
	Anal stenosis
<i>Rare malformations</i>	
	Cloacal
	malformations
	Rare
	malformations

termine the level of termination of the bowel.

Clinical Examination

A thorough examination of the baby with anorectal malformation is imperative from the following view points:

1. Examination of the neonate as a whole specially to note the gestational age, weight, temperature, color, cry, respiration, the presence or absence of jaundice, abdominal distension, septicemia, the state of hydration and any oth-

er obvious or occult congenital anomalies.

The points to ponder include:

- (a) What malformation has the baby been born with?
- (b) What effects has the malformation already had on the baby?

2. Examination to determine the type and nature of the anomaly. This is possible clinically in most female and a few male babies. In the female the number of openings in the perineum is highly significant. Three openings means that the problem can be tackled from the perineum while the presence of two or only one opening means staged surgery. Similarly, the detection of a visible fistulous communication in male child means that the anomaly is low or infralevator one.

3. The presence or absence of associated anomalies. The embryological period during which the caudal end of the fetus gets differentiated (5-24 weeks) is also the time during which many other body Systems develop. It is not difficult there fore, to imagine that an embryological defect at this time leading to anorectal malformation would also cause a high incidence of other anomalies. The term 'VACTERL association' has been coined to denote this non-random group of associated anomalies so that the treating surgeon remembers to examine for them:

V-Vetebral, A-Anorectal, C-Cardiac, TE-Tracheo-esophageal, R-Radial and Renal, and L-Limb anomalies.

The importance of this association may be judged from the fact that 30% of the patients with anorectal malformations have urinary tract anomalies. Ten

per cent have cardiac anomalies, five per cent have esophageal atresia and tracheoesophageal fistula. Sacral agenesis, which has a direct bearing on the continence in these cases is present in 4% cases. The aims of looking for these anomalies are: (a) Priority of management, *e.g.*, esophageal atresia has priority of treatment over anorectal malformation; (b) A severe cardiac anomaly may preclude successful treatment; and (c) Associated anomalies may themselves have a bearing on the ultimate outcome, *e.g.*, sacral agenesis and meningo-myelocele.

Detection of some anomalies, *e.g.*, urological may not be possible on clinical examination and investigations are then electively undertaken at a later date.

Investigative Procedures

The aims of investigative procedures are:

1. To determine the nature of the anomaly and the level of termination of the bowel whether low, intermediate or high.
2. To detect a fistulous communication.
3. To determine the presence of associated anomalies having a direct bearing on the immediate outcome of treatment, *e.g.*, gastrointestinal anomalies like duodenal and intestinal atresias, midgut volvulus, short colon and others.
4. Biochemical and bacteriological studies to check the effects of the anorectal malformation on the body.
5. Later radiological studies to further determine the bowel anatomy (dis-

tal cologram) and assess the urinary system (micturating cystourethrogram, intravenous urography and ultrasonography, *etc.*).

6. Invertogram in a dead lateral position with the hips slightly flexed gives accurate information regarding the nature of the anomaly. The radiological landmarks used for this purpose are the PC line (pubis to coccyx line) and the I point (tip of the ischium) to which the dark air shadow of the terminal end of the bowel is correlated. If the shadow crosses the I point it is a low anomaly whereas if it crosses the PC line but stops short of the I point it is an intermediate anomaly. The shadow of the terminal end of the bowel stopping above the PC line denotes a high anomaly.

Treatment

Having determined whether the anomaly is low, intermediate or high the treatment is as follows:

1. In males, low anomalies are treated with single stage perineal surgery while intermediate and high anomalies require a preliminary colostomy.
2. In females low and intermediate anomalies can be treated through the perineal route without a colostomy while high anomalies require staging.
3. When in doubt as to the nature of the anomaly in spite of all possible investigations it is always better to do a colostomy rather than explore the perineum.
4. Pelvic colostomy is physiologically more sound compared to transverse colostomy for the following reasons: (a) more solid stool consistency, (b) less area for the absorption of urine refluxing from the colourinary fistula, if

present, and (c) in the event of the all too common diarrhea, the pelvic colostomy is safer than the transverse.

Definitive Treatment of High Anomalies

At the age of 4-6 months and usually when the baby is weighing around 5 kg definitive surgery is undertaken. A distal cologram showing the terminal end of the bowel extending below the second sacral vertebra indicates that posterior sagittal anorectoplasty can be undertaken. Alternatively in higher lesions one can combine the abdominal and the posterior sagittal approaches, tackling either one first and turning the baby over. Exposure through the posterior sagittal approach enables the accurate placement of the bowel through the levator ani and the striated muscle complex leading to better chances of continence. Besides this, the delineation and closure of a fistula is greatly facilitated. For the complex cloacal anomalies in the female and rectal atresia in either sex it is probably the best approach.

Colostomy Closure

Six to eight weeks after definitive surgery the diverting colostomy (pelvic or transverse as the case may be) is

closed. This is preceded by daily dilatation of the neorectum beginning two weeks after definitive surgery. This schedule of dilatation results in adequate calibre of the anal canal.

Post-operative Care and Follow up

Daily dilatations are continued for a few months in all treated cases of anorectal malformations.

Toilet training is essential, especially in operated cases of supralelevator anorectal malformation. This includes attention to diet, development of a gastrocolic reflex, proper timing and training for defecation, sibling demonstration and rectal washouts if needed. In no case should the neorectum be allowed to dilate due to constipation.

Results

Most cases of low and intermediate anorectal malformations will have problem free anorectal function. In supra-levator malformations also acceptable social continence can be achieved if the baby does not have associated anomalies and if the surgery and postoperative care and follow up have been meticulous.

