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FACE THE EXAMINER

Anorectal Malformations (Part 2)

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(This section is meant for residents to check their understanding regarding a particular topic)

QUESTIONS

1. What are the pre-operative workup /investigations necessary for a baby born with ARM?
2. Briefly describe the aims and details of surgical management of anorectal malformations.

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ANSWERS

Answer 1:

As discussed in the first part, anorectal malformations represent a wide spectrum of anomalies involving more than one system. Also, the pre-operative work up differs in boys and girls (Tables 1 & 2). The pre-operative work up could be discussed under the following headings:

a) For diagnosis of type of anorectal anomaly
b) For associated anomalies
c) For spinal abnormalities
d) For perineal musculature

Some of the associated anomalies significantly impact the overall outcome in patients with anorectal malformations, so they must be evaluated at birth (Table 3).

Table 1: Investigations for boys with anorectal anomalies

Clinical features	Investigation	Timing	Purpose	Details
Absent anus + Bulge in perineum/anal site especially on crying	-			
Anus present - Perineal fistula/anal stenosis	-			
Absent anus + No bulge in anal site on crying ± meconium stained urine ± meconium at the urethral meatus	Invertogram (not preferred now) Cross table prone lateral (CTPL) X ray (1) (Fig. 1)	18-24 hours after birth	1. Delineate the level of gas filled rectum which gives information of level of atresia. 2. Decisive for type of surgery.	Invertogram Baby held upside down for 3-5 minutes and then lateral X-ray is taken centered at the greater trochanter. CTPL Prone position with hips and knees flexed at 45°, lateral xray centered at greater trochanter INTERPRETATION (Fig. 2)

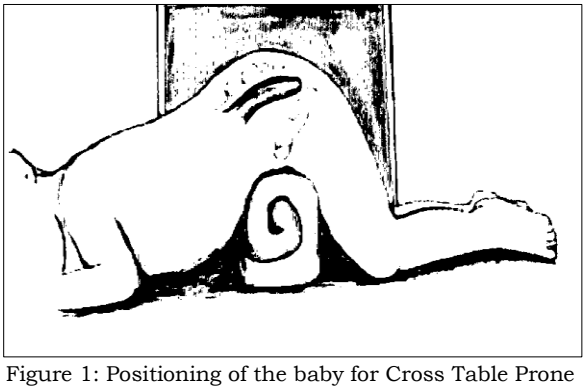

Figure 1: Positioning of the baby for Cross Table Prone Lateral X-Ray.

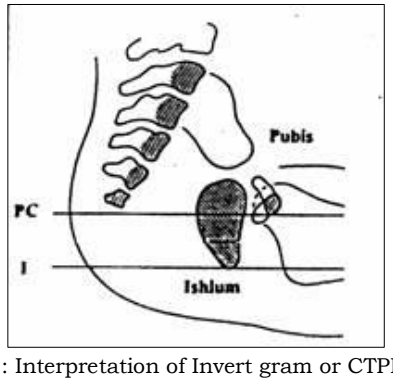

Figure 2: Interpretation of Invert gram or CTPL X-ray.

Table 2: Investigations for Anorectal malformations in girls

Clinical Features	Investigation	Timing	Purpose
Absent anus + 3 openings in vestibule	-	-	-
Absent anus + 2 openings in vestibule (high anomaly rectovaginal fistula most common)	-	-	-
Absent anus + single opening in vestibule (cloaca)	Cloacogram + Cystovaginoscopy	At birth	To plan for type of surgical intervention, either a. Primary repair b. Colostomy
Anteposed anus	-	-	-

Table 3: Investigations for associated anomalies

Investigation	Purpose
Plain X Ray abdomen Erect	To rule out Pouch colon
X-Ray Lumbosacral spine	To rule out Spinal (sacral) anomalies
2-D Echo	Cardiac anomaly
USG Abdomen & Pelvis	Urogenital anomalies
Micturating cystourethrogram (optional at birth, can be done later if needed)	Vesico-ureteric reflux

Table 4: Evolution of surgical treatment of Anorectal Malformations

Type of surgery/ Surgeon	Principle	Year published
Stephens surgery	Sacro-perineal approach - Pubo-rectalis sling important for continence, hence must not be damaged	1953 (7)
Rhoads	Perineal and abdominal pull through with blind tunnel with finger	1948 (8)
Rehbein	Endo rectal pull through to avoid damage to pelvic nerves	1967 (9)
Kiesewetter	Sacro-abdomino-perineal approach	1967 (10)
Mollard's anterior perineal surgery	Definition of pubo-rectalis sling, avoidance of extensive pelvic dissection and preserving the anoderm of anal canal	1978 (11)
Iwai	Modified abdomino-perineal pull through and identification of puborectalis and external sphincter with help of electrical stimulator	1988 (12)
Posterior sagittal anorectoplasty (Pena)	Opening up of all the muscles of continence including the external sphincter in a midline plane through posterior approach without damaging the nerve supply	1982 (13)
Georgeson	Laparoscopically assisted anorectal pull through (LAARP)	2000 (14)

Preoperative MRI of the pelvis and perineum is indicated for perineal musculature and the delineation of the sphincter complex in selected cases. MRI is helpful in thorough evaluation of the following (2):

a. Quality and shape of muscles responsible for fecal continence.
b. Location of bowel and its relation to the muscle complex.
c. Level of fistula and posterior urethral diverticulum.
d. Sacral spinal anomalies, if any.
e. Associated genitourinary anomalies.

Apart from pre-operative evaluation, MRI also assists in prognostication of the long-term outcome and the quality of life of the child with anorectal malformation.

Answer 2:

The most important aim of the surgical correction is to create a normal anus with anatomic reconstruction. Surgery should help the child to achieve a socially acceptable bowel function and should ensure avoiding fecal incontinence, urinary incontinence or sexual dysfunction.

The choices of surgical correction are as follows:

a. Primary repair – both boys and girls (3-6)
b. Staged repair – usually 3 stages:

i. Colostomy – most probably high sigmoid loop in left iliac fossa.
ii. Pull through – Posterior sagittal approach, abdomino-perineal approach, abdominal posterior sagittal approach, laparoscopic approach – in boys; anterior sagittal approach, anal transposition, posterior sagittal, abdomino-perineal, abdominal posterior sagittal approach – in girls.
iii. Colostomy closure.

The various surgical techniques that have been used for the management of anorectal malformations have been tabulated below:

To summarize, the management of anorectal malformations, which differs in boys and girls, algorithms are presented for each respectively (Fig. 3 & 4).

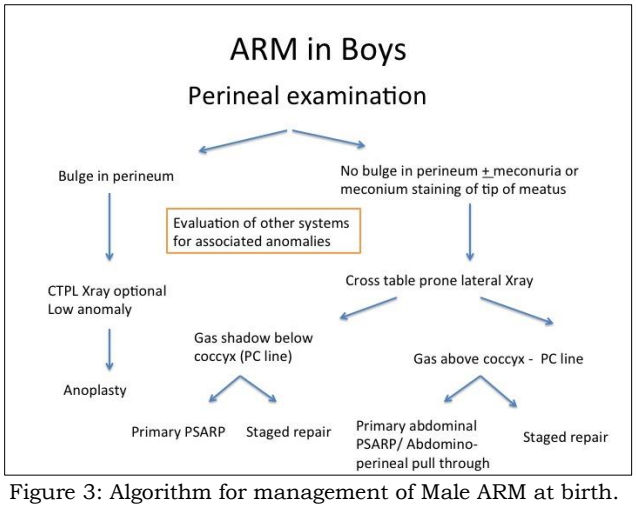

Figure 3: Algorithm for management of Male ARM at birth.

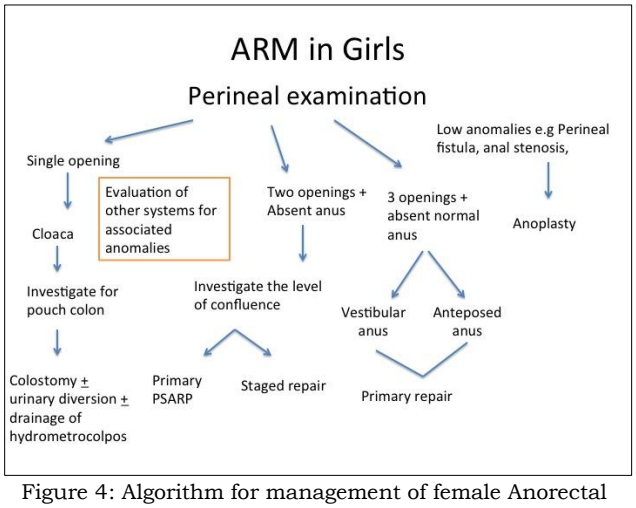

Figure 4: Algorithm for management of female Anorectal malformations at birth.

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