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FACE THE EXAMINER

Anorectal Malformations (Part 3)

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

QUESTIONS

1. What is long-term outcome for children with ARM?
2. What is Normal Continence mechanism?
3. What is assessment of fecal incontinence (FI)?
4. What is management of fecal incontinence?

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ANSWERS

Answer 1:

The aim of treatment of anorectal malformations is not just to create a passage for stools in the perineum but also to have a child who can have voluntary bowel movements without any medications and without any associated iatrogenic or congenital abnormality such as urinary incontinence. An assessment and appropriate management of urinary system pathologies (1) is an important aspect of management of a child with anorectal malformations and has been enlisted as one of the criteria for long term assessment by few researchers. (2-6) The quality of life of a child with anorectal malformation is thus dependent on the following factors:

1. Fecal continence
2. Constipation
3. Urinary continence/Urinary pathologies

The global assessment of long-term outcome of children with various types of anorectal malformations as analysed by Lewitt, et al (7) are tabulated in Table 1.

Table 1: Long-term outcome of children with Anorectal malformations

ANOMALY	BOYS			GIRLS		
	Constipation	Urinary incontinence	Fecal incontinence	Constipation	Urinary incontinence	Fecal incontinence
LOW	54	2	4	55	5	15
INTERMEDIATE	55	5	30	20	20	33
HIGH	16	25	80			
CLOACAL				35	45	45
COMPLEX MALFORMATION	19	58	44			

The terminologies used in the outcome analysis are constipation, urinary incontinence and fecal incontinence and must be clearly understood by the students and the researcher before categorizing the patients.

Constipation:

Definition: The North American Society of Gastroenterology, Hepatology, and Nutrition (NASPGHAN) defines constipation as "a delay or difficulty in defecation, present for 2 weeks or more, and sufficient to cause significant distress to the patient." (8)

The Paris Consensus on Childhood Constipation Terminology (PACCT) defines constipation as "a period of 8 weeks with at least 2 of the following symptoms: defecation frequency less than 3 times per week, fecal incontinence frequency greater than once per week, passage of large stools that clog the toilet, palpable abdominal or rectal fecal mass, stool withholding behavior, or painful defecation." (9)

Lewitt and Pena have graded constipation in children with anorectal malformations as follows:

N = Normal (no constipation)

0 = managed with diet restrictions only

1 = managed with laxatives

2 = managed with enemas

3 = severe; not manageable

Fecal incontinence:

Definition: An inability to hold feces in the rectum due to failure of voluntary control over the anal sphincters permitting untimely passage of feces and gas is defined as fecal incontinence.

In a child with anorectal malformation, total continence is only when there is voluntary bowel movement and no soiling. Those children who remain clean/dry on regular bowel management program are pseudo continent.

Grades of fecal incontinence:

A. Voluntary bowel movements or involuntary escape of feces
B. Soiling

   a. Normal: No soiling
   b. 1 = minimal, occasional, < 2times a week; no change of underwear required
   c. 2 = frequent; once a day; frequently requires change of underwear
   d. 3 = constant

Urinary incontinence:

Definition: The inability to hold urine in the bladder due to loss of voluntary control over the urinary sphincters resulting in the involuntary passage of urine is defined as urinary incontinence. A continent child thus must be dry at all times and must void spontaneously. Those who are on CIC and remain dry are termed as pseudocontinent.

Answer 2:

Continence mechanism for feces includes several factors such as –

1. Intact anal sphincters
2. Anorectal sensation
3. Rectal compliance
4. Colon transit time/motility
5. Stool volume and consistency
6. Adequate cognitive function
7. Appropriate bathroom facilities
8. Position of defecation (squatting or sitting to facilitate the straightening of anorectal angle)

The structural and functional integrity of anorectal unit which is composed of first 4 factors is the key to fecal continence, of which normal anal sphincter function – both the external and internal anal sphincter - are critical parts of continence. (Fig. 1)

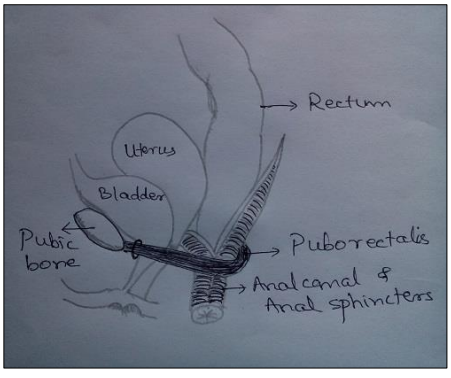

Figure 1: Sketch of anal sphincters.

Table 2: Predictors of prognosis in patients with ARM

INDICATORS OF GOOD PROGNOSIS	INDICATORS OF POOR PROGNOSIS
Normal sacrum	Abnormal sacrum
Prominent midline groove	Flat perineum
Type of ARM Rectal atresia Vestibular fistula Imperforate anus without a fistula Cloacas with common channel < 3 cm Less complex malformations: perineal fistula	Type of ARM Recto-bladder neck fistula, cloacas with common channel > 3 cm Complex malformations

Table 3: Prognostic signs for patients with ARM

GOOD PROGNOSIS SIGNS	POOR PROGNOSIS SIGNS
Good bowel movement pattern- 1-2 movement per day – no soiling	Constant soiling and passing of stool
Sensation of passing stools	No sensations
Urinary control	Urinary incontinence, dribbling of urine

Normal colonic motility propels stools in the rectum. Distension of rectum causes rectal contraction and pelvic floor and internal anal sphincter relaxation for defecation. If conditions are suitable, external anal sphincter relaxation occurs voluntarily causing defecation process to be completed. A normal sensory innervation

at all levels, i.e. spinal cord, brain stem, thalamus and cortex is mandatory for the normal defecation process to occur and hence those children with sacral spinal abnormalities could have a neurological cause of fecal incontinence wherein they are unable to appreciate the fecal consistency, differentiate the sense of feces from rectal gas, quantity of feces, and co-ordination with other actions of perineal and abdominal muscles.

The clinical parameters of the child with anorectal malformations can predict and prognosticate the long-term outcome of these children which is tabulated in Table 2 and Table 3.

Answer 3:

Several scoring systems exist and the pediatric surgeon can choose any one scoring systems. Globally, there is still no consensus as to the best scoring system and also due to wide variations in extent of the anomaly and an inability to categorise the anomalies, the comparative evaluation is extremely difficult. Table 4 gives an overview of the existing scoring systems and the components assessed in these children.

Table 4: Overview of the scoring systems

Scoring system	Continence components	Scores	Maximum score
Kelly's score (1972) (10)	Voluntary bowel movements	2	6
	No soiling	2	
	Strong anal squeeze	2	
Templeton score (1985) (11)	Toilet trained	1	5
	No Accidents	1	
	No Soiling	1	
	No Social problems (fecal odour)	1	
	No restriction in activity	0.5	
	No current problems	0.5	
Holschneider score (1994) (12)	Normal frequency of stools (1-2)	2	14
	Normal consistency of stools	2	
	No Soiling	2	
	Normal rectal sensation	2	
	Ability to hold defecation	2	
	Discrimination between formed, loose or gaseous stools	2	
	No therapy (enemas/drugs)	2	
Rintala (1995) (13)	Always able to hold back defecation	3	20
	Feels urge to defecate	3	
	Normal frequency of stools	2	
	No Soiling	3	
	No Accidents	3	
	No constipation	3	
	No social problems	3	
Pena (1995) (14)	No Soiling	N (Normal) Grade 1-3	-
	No constipation	N (Normal) Grade 1-3	
	No urinary incontinence	N (Normal) Grade 1-2	
Bai (2000) (15)	Never unhappy or anxious	2	6
	No food restriction	2	
	No peer rejection	2	
Krickenbeck (2005)	Voluntary bowel movements	Yes/No	-
	Soiling	Yes/No Grade 1-3	
	Constipation	Yes/No Grades 1-3	

Answer 4:

Once a clinical evaluation is done and the severity of the fecal incontinence is assessed by utilizing the scoring system, further investigations are needed to ascertain the exact etiology of fecal incontinence. Depending on the cause of incontinence, treatment in the form of conservative or medical or surgical intervention is planned. Table 5 provides the sequence of diagnostic tests and the management thereof.

Table 5: Management of fecal incontinence

Scenario	Investigation	Result	Treatment
FI with suspected malpositioned rectum and anal canal	MRI of pelvis	Normal	-
		Displaced	Surgical Relocation
Fecal incontinence with tendency to constipation (colonic hypomotility)	>Barium enema >Rectal and colonic manometry >Scintigraphy (to assess colonic motility)	Normal rectum and sigmoid	Bowel management program
		Megarectum	Rectosigmoidectomy
Fecal incontinence with loose stools – suspected sphincteric incompetence	MRI Pelvis	Normal	>Bowel management program (BMP)+ ↓ Electromyography of external anal sphincter (EMG)++ ↓ Anal re-education therapy (ART)+++
		Deficient/atrophic sphincteric muscles	BMP EMG Gracilis transposition with ART
		Discontinuity in pelvic diaphragm	Levatoroplasty

+ Bowel management program – many programs exist , choice is as per the severity and feasibility for the parents to carry out the program effectively.

++EMG – Author's experience – electromyography of the external anal sphincters done by physiotherapist and severity of sphincteric incompetence assessed, both pre-therapy and post-therapy

+++ ART – Anal re-education therapy – Author's experience - which includes strengthening the pelvic musculature and sphincters with regular and monitored exercise regimen coupled with Faradic stimulation of the sphincteric muscles with an individualized protocol depending on the need of the child.

REFERENCES

1. Senel E, Akbiyik F, Atayurt H, Tiryaki HT. Urological problems or fecal continence during long-term follow-up of patients with anorectal malformation. Pediatr Surg Int. 2010; 26:683-9.
2. Hashish MS, Dawoud HH, Hirschl RB, Bruch SW, El Batarny AM, Mychaliska GB, et al. Long-term functional outcome and quality of life in patients with high imperforate anus. J Pediatr Surg. 2010; 45:224–30.
3. Ojmyr-Joelsson M, Nisell M, Frenckner B, Rydelius PA, Christensson K. High and intermediate imperforate anus: psy- chosocial consequences among school-aged children. J Pediatr Surg. 2006; 41:1272–8.
4. Goyal A, Williams JM, Kenny SE, Lwin R, Baillie CT, Lamont GL, et al. Functional outcome and quality of life in anorectal malformations. J Pediatr Surg. 2006; 41:318–22.
5. Winter S, Schmidt D, Lenz K, Lehmkuhl U, Jenetzky E, Mau H, et al. Prospective evaluation of comorbidity and psychosocial need in children and adolescents with anorectal malformations. Part 2: evaluation of psychosocial need. Pediatr Surg Int. 2009;25: 895–900.
6. Iwai N, Deguchi E, Kimura O, Kubota Y, Ono S, Shimadera S. Social quality of life for adult

patients with anorectal malformations. J Pediatr Surg. 2007; 42:313-7.
7. Levitt MA, Pena A, Sutcliffe JR, Williams AR. Management of Anorectal malformations – Course companion, Cincinnati Children's Hospital, Cincinnati, 2006, pg. 24-5.
8. North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Evaluation and treatment of constipation in children: summary of updated recommendations of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. J Pediatr Gastroenterol Nutr. 2006; 43:405-7.
9. Benninga M, Candy DC, Catto-Smith AG, Clayden G, Loening-Baucke V, Di Lorenzo, et al. The Paris Consensus on Childhood Constipation Terminology (PACCT) Group. J Pediatr Gastroenterol Nutr. 2005; 40:273-5.
10. Kelly JH. The clinical and radiological assessment of anal continence in childhood. Aust N Z J Surg. 1972; 42:62-3.
11. Templeton JM Jr, Ditesheim JA. High imperforate anus--quantitative results of long- term fecal continence. J Pediatr Surg. 1985; 20:645-52.
12. Holschneider AM, Pfrommer W, Gerresheim B. Results in the treatment of anorectal malformations with special regard to the histology of the rectal pouch. Eur J Pediatr Surg. 1994; 4:303-9.
13. Rintala R, Lindahl H. Is normal bowel function possible after repair of intermediate and high anorectal malformations? J Pediatr Surg. 1995; 30:491-4.
14. Pena A. Anorectal malformations. Semin Pediatr Surg. 1995; 4:35-47.
15. Bai Y, Yuan Z, Wang W, Zhao Y, Wang H, Wang W. Quality of life for children with fecal incontinence after surgically corrected anorectal malformation. J Pediatr Surg. 2000; 35:462-4.

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