Title: Review: High Resolution Anorectal Manometry in Children.

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ABSTRACT:

Ano rectal manometry is one of the motility test in children performed by Pediatric gastroenterologist. It evaluates the motility function of anorectal tract. It is helpful for diagnosing children with constipation, rectal hypersensitivity, fecal incontinence, Hirschsprung’s disease, anal achalasia and anorectal malformations. The most common indication to perform ano rectal manometry is to diagnose Hirschsprung’s disease. It is a safe procedure. This paper discusses recent advances and reviews on anorectal motility disorders in children.

Keywords:
Internal anal sphincter Achalasia.
Defecation dys-synergia.
Rectal hypersensitivity.
Non-retentive fecal incontinence.
Hirschsprung’s disease.

Abbreviations:
ARM: Anorectal Manometry.
RAIR: Recto anal inhibitory reflex.
FCSV: first constant sensation volume
DDV: desire to defecate volume
MTV: maximum tolerated volume
NRFI: Non-retentive fecal in continence.
ASRI: Anal sphincter relaxation integral

Introduction:

Ano-rectal manometry is the investigation which can help to diagnose motility issues of anorectal tract in children. This review discusses the role of high resolution ano-rectal manometry in children to diagnose and differentiate between various types of constipation and related fecal incontinence. Constipation in children can be due to defecation dys-synergia or colonic inertia. Improper toilet training and retentive behavior while defecation is responsible for defecation dys-synergia. Colonic inertia can be due to congenital enteric neuropathy e.g. Hirschsprung’s disease or rare enteric myopathies leading to Chronic intestinal pseudo-obstruction in children. Similarly fecal incontinence can be retentive or overflow encopresis and non-retentive fecal incontinence due to neurological or spinal cord problems. All these disorders can be differentiated with the help of anorectal manometry in children.
Anorectal Manometry: Catheters and techniques.

Anorectal manometry can be done as high resolution anorectal manometry or conventional manometry. Conventional manometry catheters have fewer sensors. They are placed at wider intervals. Their output is line plot. High resolution manometry catheters have densely placed sensors. They have output as topographic color contour or line plot. High resolution manometry is more expensive and less time consuming as compared to conventional manometry. Catheters used for anorectal manometry are either solid state or water perfused. In water perfused catheter external transducer detect pressure changes to water flow. In solid state catheters transducers are built inside the catheter to generate electric signal output. Water perfused catheter are cheaper and widely available. Solid state catheters are expensive and are available mostly in research and institute setting.

Pediatric Anorectal Manometry Challenges:

Non cooperative infants and young children remains a challenge to pediatric gastroenterologist performing anorectal manometry. In non-cooperative children sedation can be used. It has limited functional motility analysis. Ano-rectal manometry is best performed in awake state. In neonates one can complete anorectal manometry while breast feeding .But again all parameters cannot be studied. Anorectal resting pressure, squeeze pressure, cough reflex, push pressure, rectal sensation and RAIR (recto anal inhibitory reflex) can be assessed in awake children. Conscious or awake ano-rectal manometry requires child’s understanding and cooperation. Under sedation only resting pressure and RAIR (recto anal inhibitory reflex) can be elicited. Drugs used in anesthesia and level of sedation do affect readings. Muscle relaxant should be avoided while doing ano-rectal manometry. Ketamine do not affect ano-rectal metrics and can be used in children where conscious ano-rectal manometry is not possible.

Indication of Anorectal Manometry:

The important indication of doing ano-rectal manometry is to rule out Hirschsprung’s disease. Table 1 highlights important indications of anorectal manometry. RAIR is absent in Hirschsprung’s disease and internal anal sphincter Achalasia. Anorectal manometry (ARM) can be used in ano-rectal malformation with defecation problems. ARM is used prior and after surgery to assess motor function. In children having fecal in-continenence with or without neurogenic problems ARM is useful. ARM can be used to differentiate types of defecation dys-synergia in patients with chronic constipation. Assessment of rectal sensation is important in chronic constipation, mega-rectum and rectal hypersensitivity. In chronic constipation rectal sensitivity threshold is increased while in rectal hypersensitivity threshold is decreased. Rectal hypersensitivity is seen in patients with inflamed ano-rectum. Rectal hypersensitivity also correlates with visceral hypersensitivity. RAIR is present in patients with spinal cord lesions. Sacral agenesis children have lower external anal squeeze pressure and increased rectal
Children with spinal lesion can have anal spasm. They achieve maximum relaxation of sphincter with small inflation volumes. Patients with internal anal sphincter Achalasia can be assessed with ARM. Selected patients of anal achalasia may benefit from Botulinium toxin injection.

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<td>6. To diagnose rectal hyposensitivity and hypersensitivity.</td>
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Table 1: Indications of ano-rectal manometry in children.

Anorectal manometry can also be used in biofeedback therapy to teach children toilet training and manage defecation dys-synergia.

**Process of Anorectal Manometry:**

Test should be performed in left lateral position. International Anorectal physiology working group (IAPWG) recommends standardized testing protocol and the London classification for assessment of ano rectal function. It starts with initial stabilization time of 3 min after catheter insertion. Resting pressures are evaluated later over 60 seconds (sec). Short squeeze pressure of 5 sec (3 in numbers) followed by long squeeze pressure of 30 sec are suggested. It is emphasized that there should be 30 sec interval between short squeeze and 60 sec interval after long squeeze to assess cough reflex. After assessment of cough reflex, push for 15 sec (3 in numbers) with interval of 30 sec is assessed. Rectal sensory testing follows push assessment. In assessment of rectal sensation first constant sensation volume (FCSV), desire to defecate volume (DDV) and maximum tolerated volume (MTV) are assessed. RAIR should be elicited at last. If mega rectum is under evaluation more volume to elicit RAIR is required. Balloon expulsion testing is not validated in children. There is no published consensus on type of catheter volume required, balloon expansion with air or water in children.

**Pre procedure assessment for ARM:**

Informed consent needs to be taken. Age appropriate enema should be given. Rectum should be dis-impacted prior ARM.
Anorectal Manometry Metrics: Normal vs. abnormal values.

STEPWISE ANALYSIS OF ANORECTAL MANOMETRY BIOMETRICS:
STEP 1: Resting anal pressure → Anal hypotension / Anal hypertension.
STEP 2: Squeeze pressure
Low anal squeeze pressure with low anal resting pressure → anal hypotension and hypocontractility.
Low resting pressure with normal anal squeeze pressure → anal hypotension with normal contractility.
Normal anal resting pressure with low anal squeeze pressure is anal norm tension with hypocontractility.
STEP 3: Differentiate types of Defecation Dys-synergia.
STEP 4: Establish RAIR.

Figure 1: Stepwise analysis of anorectal biometrics based on London IAPWG\textsuperscript{13} protocol.

Anal resting pressure\textsuperscript{1} is mean maximum anal pressure over 60 sec recording period. It assess baseline anal sphincter integrity. High resting anal pressure suggest voluntary or involuntary muscle spasm, anal stenosis or anal stricture. Low resting anal pressure suggests hypotension, anal sphincter post drug assessment, post trauma or spinal cord disorder. According to London classification\textsuperscript{13} high resting anal pressure is classified as minor anorectal finding. It is called as anal hypertension. While lower resting anal pressure is major anorectal finding called as anal hypotension. Recently published cohort study\textsuperscript{15} in healthy infant’s normal resting pressure values at 2 months and 6 months as 62.9 cm of water(46.26 mm of Hg) and 65.3 cm of water(48.03 mm of Hg) respectively. Banasiuk et al\textsuperscript{16} have published the largest pediatric series of normal children including children from 1 to 18 years without lower gastrointestinal tract symptoms. According to this study normal resting pressure 83+/−23 mm of Hg.

Squeeze pressure is the maximum incremental pressure observed during the assessment. Low squeeze pressure may mean poor understanding by child, neurogenic or myogenic anal sphincter injury. Squeeze pressure assess anal sphincter contractile integrity. The mean squeeze pressure in normal children is 191+/−64 mm of Hg.\textsuperscript{16} Low anal squeeze pressure with low anal resting pressure is classified by London IAPWG\textsuperscript{13} group as major ano rectal finding of combined anal hypotension and hypo-contractility. Low resting pressure with normal anal squeeze pressure is anal hypotension with normal contractility. Normal anal resting pressure with low anal squeeze pressure is anal norm tension with hypo contractility.
Push maneuver assess the coordination in defecation. Adequate push pressure with high anal sphincter pressure is type I dys-synergia. Poor push with high anal sphincter pressure is type II dys-synergia. Adequate pressure with no decrease in anal sphincter pressure is type III dys-synergia. Poor push with no decrease in anal sphincter pressure is type IV dys-synergia. London IAPWG group classified these as disorders of recto anal coordination. Adequate rectal pressure and no decrease in anal pressure is defined as abnormal expulsion with dys-synergia. Poor rectal pressure with decrease in anal pressure is abnormal expulsion with poor propulsion. Poor rectal pressure with no decrease in anal pressure is abnormal expulsion with poor propulsion and dys-synergia. All these are labeled as minor findings as per London IAPWG group.

![Image: Anorectal manometry during push maneuver showing paradoxical raised anal pressure with defecation suggests Type 1 Defecation Dys-synergia.](Images taken post consent from patient and subject to copy right Superspeciality Clinics & Endoscopy Center. Nagpur. Dr Yogesh Waikar)

Defecation dys-synergia is of four types (table 2). All these can present as constipation. It is present in about 81% of children who are diagnosed as functional constipation. While in study of Banasiuk M et al 60% of patients with constipation have defecation by dys-synergia. In both studies type IV defecation dys-synergia is the most common disorder. Adequate intra rectal pressure of more than 40 mm of Hg is normal. Incomplete or absent relaxation of <20% of resting anal sphincter pressure is considered abnormal in push assessment. Study by Banasiuk M et al suggested higher cut off 31% of anal relaxation in children. Defecation dys-synergia is also seen in patient with ano rectal malformations. Isolated puborectalis contraction may contribute to this presentation.
Type 1 Defecation Dys-synergia.  
Increase in intraabdominal pressure (Push) and Paradoxical Increase in anal sphincter pressure (figure 2).

Type 2 Defecation Dys-synergia.  
No increase in intraabdominal pressure (Push) but with Paradoxical increase in anal sphincter pressure.

Type 3 Defecation Dys-synergia.  
Increase in intraabdominal pressure (Push) with incomplete or absent less than 20% anal sphincter relaxation.

Type 4 Defecation Dys-synergia.  
No increase in intraabdominal pressure (Push) & with incomplete or absent less than 20% anal sphincter relaxation.

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| Resting, squeeze & push pressure (figure 4) are important in assessment of rectal incontinence or encopresis. Fecal incontinence can be constipation associated fecal incontinence (CFI) or non-retentive fecal in continence (NRFI). NRFI is more common in boys. They usually have normal formed stools, rarely painful defecation, no nocturnal encopresis and normal colonic transit time \(^{19}\). NRFI children have decreased resting pressure, reduced squeeze pressure and higher thresholds for sensation urge and discomfort \(^{20,21}\). Pressure at rest and squeeze in functional constipation with fecal incontinence are lower than functional constipation without incontinence \(^{22}\). Children with normal ARM with incontinence have lower response to therapy as compared to abnormal ARM \(^{23}\).

Impaired cough reflex suggests damage to sacral arc \(^1\). Cough -anal reflex is described as contraction of external anal sphincter with coughing \(^{24}\). Afferent component of cough reflex comes from muscle spindles and other sensory receptors in pelvic floor muscles, ligament and fascia and bladder \(^{25}\). Afferent is activated by increase in abdominal pressure during cough.

RAIR (recto anal instability reflex) is tested by rapidly inflating rectal balloon with incremental volume ranging from 5-10ml to 60ml. The reflex is considered present when decrease in resting pressure reaches 25%. The mean rectal balloon volume required to elicit RAIR \(^{16}\) in children is 15.7 +/- 10.9 cm\(^3\). BSPGHAN, British society of pediatric gastroenterology, hepatology and nutrition \(^1\) suggest to start with 5ml in infants and 10ml in older children. There is usually incremental degree of anal sphincter relaxation with increasing balloon volume. Anti-cholinergic, glycopyrrolate may result in false positive absence of RAIR \(^{26}\). Ketamine does not affect the RAIR. Sevoflurane and chloral hydrate do not affect RAIR \(^{26}\). Positive RAIR excludes Hirschsprung’s disease. (Figure 3). Negative RAIR suggest
Hirschsprung’s disease or internal anal sphincter Achalasia. Partial RAIR is seen in ano rectal inflammatory condition.

Figure 3: RAIR present rules out Hirschsprung’s disease. Reflex marked area shows reduction in anal pressure post rectal balloon inflation (Images taken post consent from patient and subject to copy right Superspeciality Clinics & Endoscopy Center. Nagpur. Dr Yogesh Waikar)

The sensitivity of ARM and rectal suction biopsy is equivalent. The specificity of ARM is lower than rectal biopsy. The negative predictive value of ARM is 100% while positive predictive value of ARM is only 56%. Hence ARM can be used to exclude Hirschsprung’s disease. It may reduce the number of rectal suction biopsy. Functioning RAIR is decrease in anal sphincter pressure of at least 20 mm of Hg following balloon dilatation. Post-operative RAIR is not seen till approximately 32 months in Hirschsprung’s disease and is associated with variable outcomes.

Anal sphincter relaxation integral at 10 sec (ASRI 10) can be used as supportive metrics to RAIR to diagnose Hirschsprung’s disease. ASRI 10 < 7 suggest Hirschsprung’s disease. In London IAPWG group absent RAIR is defined as recto anal areflexia. It is the major finding in ARM.

Rectal sensation are assessed by inflating air at 1 ml/sec. According to London IAPWG first constant sensation volume, desire to defecate volume and maximum tolerable volume are calculated. More than 2 out of 3 abnormal test (more than normal volume required to sense) is classified under major finding of rectal hyposensitivity. 1 out of three abnormal rectal sensory test of more than normal volume required to trigger the sensation is classified as borderline rectal hyposensitivity. Even one abnormal rectal sensory test of less than normal volume required to trigger sensation is classified as rectal hypersensitivity. The first constant sensation volume in normal children 24.4+/-23.98 cm³; desire to defecate volume is 45.9+/-34.55 cm³;
maximum tolerate volume 91.6+/−50.17 cm$^3$. Common cutoff values are described in table 3 for diagnosing Anorectal dysmotility.

Age appropriate mean first sensation volume $^{16}$ are 34, 25, 14.7 and 22.1 for <5 years, 5-8 years, 9-12 years and >12 years respectively. Age appropriate mean desire to defeicate volume $^{16}$ are 36, 37.2, 36 & 55 for <5 years, 5-8 years, 9-12 years and >12 years respectively. Age appropriate means maximum tolerable volume are 48, 75, 88, and 127 for <5 years, 5-8 years, 9-12 years and >12 years respectively. For children >12 years adults values can be used $^1$.

<table>
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**Table 3: Common cut off used in Anorectal biometrics.**

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**Figure 4: Anorectal manometry images during Rest, Squeeze & Push.** It depicts normal topographical graph while performing anorectal manometry in three states viz. rest, squeeze and push. In squeeze maneuver anal pressure increases (red-purple color in the middle). In push maneuver rectal pressure increases showed by yellow and red lines in upper part of topographical chart. (Images taken post consent from patient and subject to copy right Superspeciality Clinics & Endoscopy Center. Nagpur. Dr Yogesh Waikar)

**Conclusion:**

Anorectal manometry is one of the most commonly asked motility test in children. The most common indication of anorectal manometry is to establish RAIR to rule out Hirschsprung’s disease. It is a safe procedure. Normal values for children are now available. Both solid state and water perfused catheter can be used for ARM. ARM can help in diagnosis of anorectal motor dysfunction. It can also be used in bio-feedback therapy for treatment of defecation dys-synergia in children. More studies are needed before using advanced ARM – metrics in children. London IAPWG protocol should be followed in future studies for comparative analysis.

**REFERENCES:**


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