8 Anorectal Malformations: Anorectal Manometric and Endosonographic Combined Approach

Tamara Caldaro

8.1 Introduction

Assessment and management of post-operative problems following surgical repair of anorectal malformations (ARMs) represent a specific challenge for paediatric surgeons and paediatric gastroenterologists due to the complexity of the disease.

Despite technical advances and accurate anatomical reconstructions, high prevalence of faecal incontinence (16.7%–76.7%) and chronic constipation (22.6%–86.7%) has been reported in the long-term follow-up, without clear differences in the rate of impaired bowel movements in patients with low or high ARMs [1].

These types of disabilities have a severe impact on patients' quality of life, resulting in many psychologic and social disturbances [2]. Several studies have investigated a wealth of clinical information and various possible prognostic factors that could be related to the outcome [3].

Based on three anatomical elements (type of ARMs, spinal defects and sacral anomalies), an "ARM Continence Predictor Index" has been created to define a personalised potential continence score system prior to surgical intervention, so as to plan a tailored bowel management programme to successfully get patients clean [4].

T. Caldaro (✉)
Digestive Surgery and Endoscopy Unit, Bambino Gesù Children's Hospital-IRCCS, Rome, Italy
e-mail: tamara.caldaro@opbg.net

However, a comprehensive assessment of pelvic floor is necessary in order to better define the appropriate treatment.

Post-operative investigations, that include endoanal ultrasonography (EAUS) and anorectal manometry, provide accurate objective information about the continence status, as they analyse the global anorectal anatomy and functionality [5].

8.2 Anorectal Malformations and Anorectal Dysfunction

ARMs include a wide selection of congenital anomalies that range from the most benign of all defects, the perineal fistula, to the severe anomalies, such as cloaca and cloacal exstrophy [6]; consequently, various anatomical classification systems have been devised to define the pathology of these anorectal anomalies. In 1970, an international classification was proposed for both genders, dividing the anomalies into high, intermediate and low, according to the position of the terminal rectum to the levator ani. In 1984, the Wingspread classification distinguished between high (supra-levator), intermediate (partially trans-levator) and low (trans-levator) ARMs in male and female subjects, with the introduction of special groups for cloacal and rare malformations [7].

Later, Peña proposed a surgically oriented classification based on the presence and position

M. Lima, G. Ruggeri (eds.), *Ano-Rectal Endosonography and Manometry in Paediatrics*,
https://doi.org/10.1007/978-3-030-97668-2_8

of fistula and on the relationship of the terminal colon to the levator sling muscles of the pelvic floor [8].

The location and type of fistula help to guide the operative approach and to determine the extent of mobilisation needed for pulling through the blind pouch. This classification system was also the first one which attempted to define prognosis for each group in terms of functional bowel outcomes. In fact, even after the introduction of the posterior sagittal anorectoplasty (PSARP), the major post-operative problems observed were chronic constipation and overflow incontinence caused by motility disturbances and stool incontinence secondary to true sphincter insufficiency.

In 2005, the Krickenbeck group suggested a new classification to rationalise functional outcome among different studies and to allow more meaningful comparisons. Consequently, the classification comprises three distinct categories: diagnostic, surgical procedure and post-operative functional results [9, 10]. The anorectal anomalies are divided into two groups, the major clinical groups and the rare/regional variants, based on the presence, type and location of fistula, as well as the position of the rectal pouch. For follow-up studies, not only the site of the fistula should be documented but also other additional groupings considering specifically the operative procedure performed and the main post-operative motility disturbances (voluntary bowel movements, soiling and constipation).

Difficulty in finding an exhaustive classification comes from the complexity and variety of the disease, whose prognosis also depends on associated anomalies, which could influence the patients' quality of life.

Normal control of defaecation requires integrity of neuromuscular structures including rectum, internal and external sphincters and pelvic floor muscles; preservation of anorectal sensation; maintenance of neural (both autonomic and somatic) pathways; and normal intestinal transit time [11].

Commonly, one or more of these mechanisms may be altered in patients with ARMs.

As regards the sphincter mechanisms, internal anal sphincter (IAS) and external anal sphincter (EAS) may be practically normal in minor defects but very compromised in the more complex ones, with severe degrees of muscle underdevelopment and atrophy. In addition to these congenital defects, scars of the anorectal sphincters are frequently detected after surgical reconstruction [12].

Megarectum, a condition found in 10–50% of patients with ARMs, is responsible for constipation and overflow incontinence due to accumulation of stool in the dilated segment [13].

Various aetiologies of primary megarectum in ARMs have been proposed, including sacral neuropathy leading to loss of proprioception and denervation [14, 15] or in utero caudal obstruction of the hindgut during development leading to a grossly dilated rectosigmoid. Post-surgical anorectoplasty denervation, chronic constipation and anastomotic anal stricture are the suggested causes of secondary megarectum [16, 17].

The dilated sigmoid and rectal segment are characterised by a delay in the transit time (hypomotility) and a higher sensory threshold, conditions that are more critical in the presence of sacral and/or spinal anomalies.

The anorectal reconstruction is another crucial step in preserving faecal continence.

In reconstructive ARMs surgery, the PSARP procedure is broadly accepted for its potential advantages, but although it provides the most accurate anatomical reconstruction saving the anal sphincters, the outcomes are not in keeping with the technical benefit. The aims of the technique are to place the rectum in the middle of the anal sphincter complex, which is accurately mapped with an electrical stimulator, and to avoid injuries to adjacent sphincter structures [18].

However, mislocation of the anoplasty is a frequently seen complication. The anus can be mislocated anterior or posterior to the muscle complex or in a lateral orientation. Location of the anoplasty outside of the centre of the anal sphincters can impact the ability to voluntarily handle bowel movements and should be corrected if the potential for bowel control is present [6].

Laparoscopic-assisted anorectoplasty (LAARP) may be preferable to the posterior sagittal

method to define the right placement of the anus in high anomalies (recto-bladder neck fistula and complex cloacae), which require an abdominal approach.

LAARP provides direct visualisation of the rectal fistula and surrounding structures, identifies the central portion of the puborectalis from inside reducing the insurgence of injury to nerve plexus and allows accurate placement of the bowel through the anatomical midline and the levator sling with minimal surgical trauma to the continence mechanism [19].

The overall voluntary bowel movement, constipation and faecal incontinence in the series on PSARP by Peña et al. [20] were 75%, 48% and 25%, respectively, while LAARP analysis by Pathak et al. [21] displayed 81% voluntary bowel movement, 9% constipation and 12% faecal incontinence.

In literature, however, the pre-eminence of LAARP over PSARP is not clearly demonstrated, due to lack of homogeneous data and identical criteria regarding the functional outcome assessment and the long-term follow-up [21].

Therefore, accurate evaluation of patients with ARMs remains a challenge in consideration of the complexity of congenital and acquired factors which potentially influence the mechanism of faecal continence, even after proper surgical reconstruction. A directed history and a careful physical examination with particular care for the integrity of the perineum and rectum, a neurologic evaluation and a complete diagnostic assessment are mandatory to define a bowel management programme that can improve the quality of life of patients.

8.3 Anorectal Manometry

Anorectal manometry is a recognised procedure used in the full and proper assessment of defaecatory disorders and also in the pre- and/or postoperative evaluation of the anorectal area.

The ANMS-NASPGHAN consensus document on anorectal and colonic manometry in children recommends to perform anorectal manometry on patients treated surgically for ARMs with persistent defaecation problems [22].

Recent advances in diagnostic techniques have led to an evolution in classical anorectal manometry test with the introduction of both the high-resolution anorectal manometry (HR-ARM) and the three-dimensional high-definition anorectal manometry (3D-HDARM) [23].

Conventional manometry is carried out using a sleeve catheter, a water-perfused and a data acquisition system. The manometry probes have 4–8 side holes that are disposed helicoidally or radially along the catheter and connected to the perfusion apparatus with a pneumohydraulic pump. Data are presented as pressure lines. The catheter is incapable of acquiring the pressures of the entire anal canal simultaneously. Therefore, a pull-through manoeuvre is required to sample the entire area of interest.

In HR-ARM, the recorded data are displayed as highly detailed topographical colour-contoured plots, rather than overlapping line traces. With the improvement of electronics, new manometric water-perfused and solid-state catheters with many miniature pressure sensors have been developed in order to ensure a better interpretation of pressure changes and to allow a simultaneous assessment of the anal sphincters. These new probes are also useful in minimising the motion artefacts and eliminating the pull-through manoeuvres [24].

HDARM allows for three-dimensional physiologic mapping of anal sphincters with the possibility of detailed assessment of pressures and visualisation of symmetry by means of a single introduction of the probe. In fact, thanks to the high number of sensors placed circumferentially on the catheter, 3D pressure models of the organ can be obtained, with a better definition of the different components of the anal canal, including the puborectalis muscle, the EAS and the IAS. Anyway, the anal resting pressure may result overestimated and the canal dynamics upon balloon distension can be misinterpreted with the current catheter, as it has an outer diameter of almost 11 mm and according to the Laplace's law, which states that pressure within a tube is inversely proportional to the radius [25].

Before performing anorectal manometry, especially in patients with ARMs, it is recommended to:

- Classify the type of ARMs and assess associated sacral and/or spinal anomalies
- Inspect carefully the anorectal region and the anal opening to exclude anal stricture or pronounced opening, rectal mucosa prolapse or mislocation of the neo-anus
- Evaluate the presence of a megarectum
- Ensure a complete rectal disimpaction

In fact, any of these factors could modify the results of manometric tests and in some conditions (e.g. anal stricture) the exam could be contraindicated.

8.4 Manometric Parameters and Anorectal Malformations

The manometric test provides comprehensive information regarding (**a**) the pressures of the anal sphincter muscles at rest and during dynamic manoeuvres (squeezing and straining); (**b**) the rectal sensation; (**c**) the innervation of IAS and EAS eliciting the recto-anal inhibitory reflex (RAIR) and the cough reflex; (**d**) the defaecation dynamics (bear-down manoeuvre) by assessing the recto-anal pressure gradient during straining and the balloon expulsion test (BET).

8.4.1 Anal Sphincter Pressures

Function and integrity of the anal sphincters are evaluated by measuring the pressure at different levels of the anal canal. Modifications of pressure values could be useful to determine the location and grade of anal sphincter defects.

A low anal resting pressure (ARP) may identify an underlying problem with the IAS, which supplies between 55% and 85% of resting pressure, whereas a decrease in voluntary anal squeeze pressure (ASP) could be related to a defect of the EAS.

As numerous studies have detected, lower ARP and ASP values were found in patients with severe types of ARMs and/or in those with faecal incontinence due to wide sphincter defects or atrophy [26, 27].

8.4.2 Rectal Sensation

Using graded balloon, rectal sensation can be measured by assessing three sensory thresholds: first sensation, urge to evacuate and discomfort.

In case of megarectum, a frequent condition in ARMs, manometry testing shows an impaired defaecatory sensation (rectal hyposensitivity) with an increase of 1 or more of the threshold values, because higher volumes of distention are needed to feel the defaecatory stimulus. Consequently, constipation and soiling appear to be more common among patients with a large rectal volume (>150 mL of air), as Hedlund et al. documented [26].

8.4.3 Reflexes

Innervation of the IAS and the EAS can be studied eliciting the RAIR via the myenteric plexus and the cough reflex via the spinal reflex arc, respectively.

RAIR is a relaxation response in the IAS, namely a pressure drop of at least 25% in the anal canal following rectal distension. It is elicited by rapid insufflation of minimum 20 mL of air into a balloon positioned in the distal rectum at the level of the proximal high-pressure zone.

It is absent in Hirschsprung disease, but also in other conditions such as lower anterior resections and injuries of IAS. Potential technical pitfalls with false-negative results of RAIR could be obtained in the presence of two typical disorders observed in patients operated on ARMs: megarectum, for which a greater volume of rectal distention is required to elicit RAIR, and low ARP due to lesions of the IAS [12, 13].

RAIR is considered an indirect sign of continence. Rintala et al. [28] reported RAIR as positive in all the “good” and in 75% of the “fair”

group patients and negative in the "bad" group with ARMs. Same results were achieved by Senel et al. [27].

The "cough reflex" is a manoeuvre indicated to assess the integrity of spinal reflex pathways in patients with incontinence. It consists of a contraction of the EAS following an intra-abdominal pressure increment (e.g. coughing) [29].

The abnormality of innervation of EAS has been identified as one of the most important factors which affect post-operative anal function in patients with ARMs. An altered response may suggest damages in lumbosacral and motor nerves ending in puborectalis and in EAS, even when the lesion is secondary to a congenital anomaly of lumbosacral vertebrae or to a spinal dysraphism.

William et al. reported that 35% of subjects operated on ARMs had lumbosacral vertebral abnormalities on plain X-ray film, whereas 53% showed abnormalities of spinal cord or vertebrae in magnetic resonance imaging (MRI) study [30].

Capitanucci et al., performing urodynamic evaluations on patients with anorectal anomalies as well as spinal dysraphism, demonstrated that the latter may be asymptomatic in small children [31].

Thus, an accurate functional and morphological evaluation of the lumbosacral spine using neurophysiological tests and MRI is essential to ensure an early diagnosis and treatment of the spinal cord lesion before the clinical symptom appears [32].

8.4.4 Bear-Down Manoeuvre and Balloon Expulsion Test

Normal defaecation dynamics is characterised by an abdominal compression associated with anal relaxation. The bear-down manoeuvre simulates evacuation and it is used to assess anorectal and pelvic floor pressure changes during attempted defaecation. Pelvic floor dyssynergia, defined as failing relaxation and coordination of pelvic floor and abdominal muscles during evacuation, is diagnosed if these coordinated movements do not occur.

BET is a helpful screening test to confirm the presence of dyssynergia because it mimics the stool in the rectal vault that should be expelled in 1 min. It is characterised by high specificity (80% to 90%), although the sensitivity is lower than 50% [33]. There are four manometric patterns of pelvic floor dyssynergia according to Rao classification [34, 35]:

- Type I: Adequate rectal push effort with paradoxical anal sphincter contraction.
- Type II: Inadequate rectal push effort with paradoxical anal sphincter contraction.
- Type III: Adequate rectal push effort but inadequate relaxation (<20%) of anal sphincter pressure.
- Type IV: Inadequate rectal push effort and also inadequate relaxation (<20%) of anal sphincter pressure.

A large proportion of patients with chronic constipation suffer from dyssynergic defaecation and the prevalence ranges from 25% to 50% in adult population and up to 50% in children [36, 37]. The incidence is not defined in patients with ARMs; anyway some studies have documented a manometric dyssynergic pattern also in subjects without constipation [38, 39].

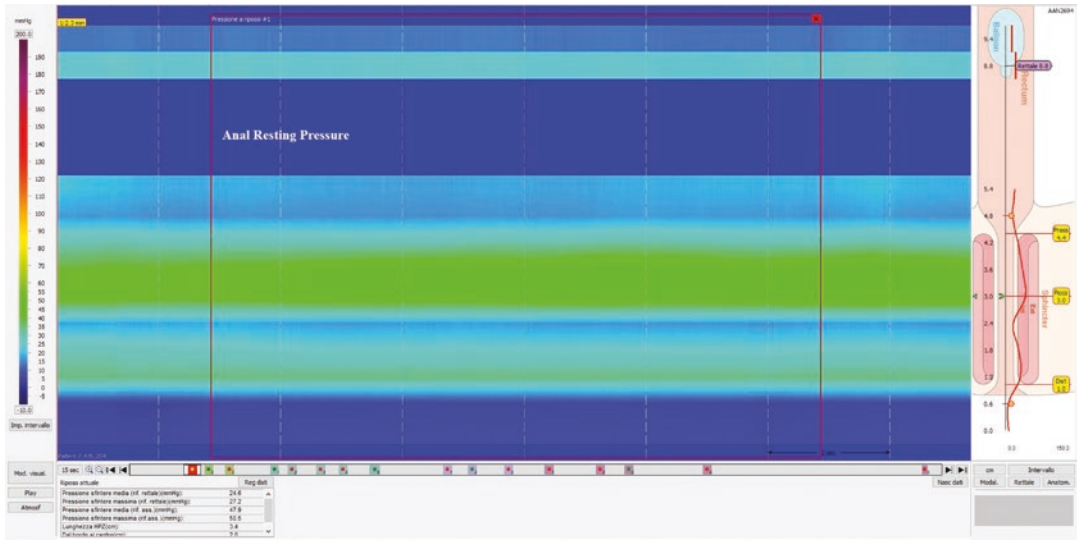
The manometric patterns of a patient surgically treated for ARMs and of a child with sacral agenesis plus non-treated anterior anus are shown in Figs. 8.1 and 8.2, respectively.

8.5 Anorectal Manometric Findings: Data from Literature

Since the late 1970s, some authors have used anorectal manometry to estimate anal pressure in patients operated on ARMs.

One of the first studies on patients with intermediate and high ARMs demonstrated that the anorectal pressure profile had no marked high-pressure zone in the anal canal in subjects suffering from faecal incontinence and that only 20% of the patients with high-type anomalies had a RAIR, 23% of these had poor clinical results [40].

a



b

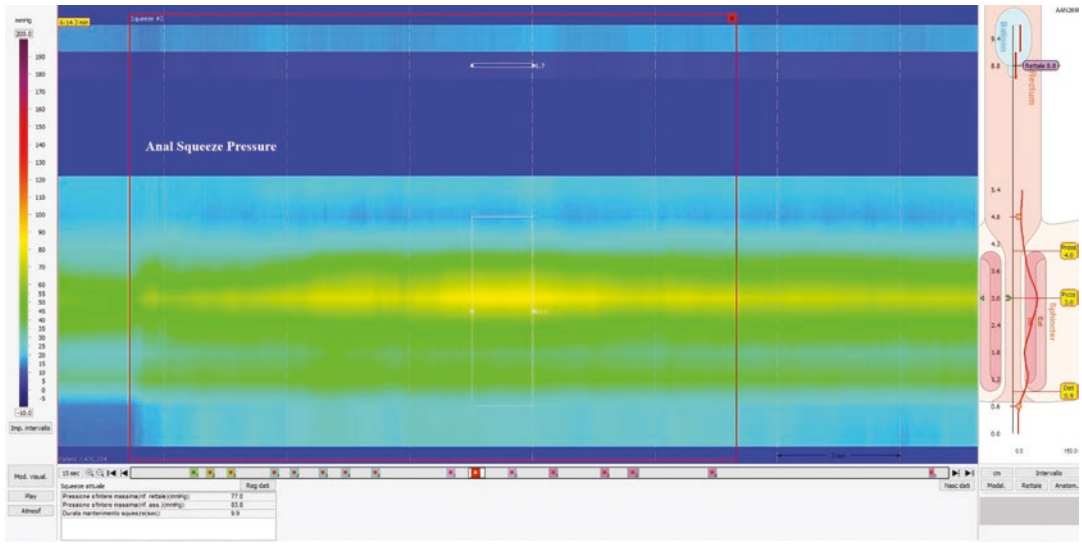


Fig. 8.1 5-year-old female child, anorectal malformation and recto-vestibular fistula, with history of constipation and soiling

HR-ARM documents low ARP (**a**) and ASP (**b**), pelvic floor dyssynergia type I according to Rao's classification (**c**), absence of RAIR (**d**)

HR-ARM High resolution anorectal manometry, *ARP* Anal Resting Pressure, *ASP* Anal Squeeze Pressure, *RAIR* Recto-Anal Inhibitory Reflex

Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

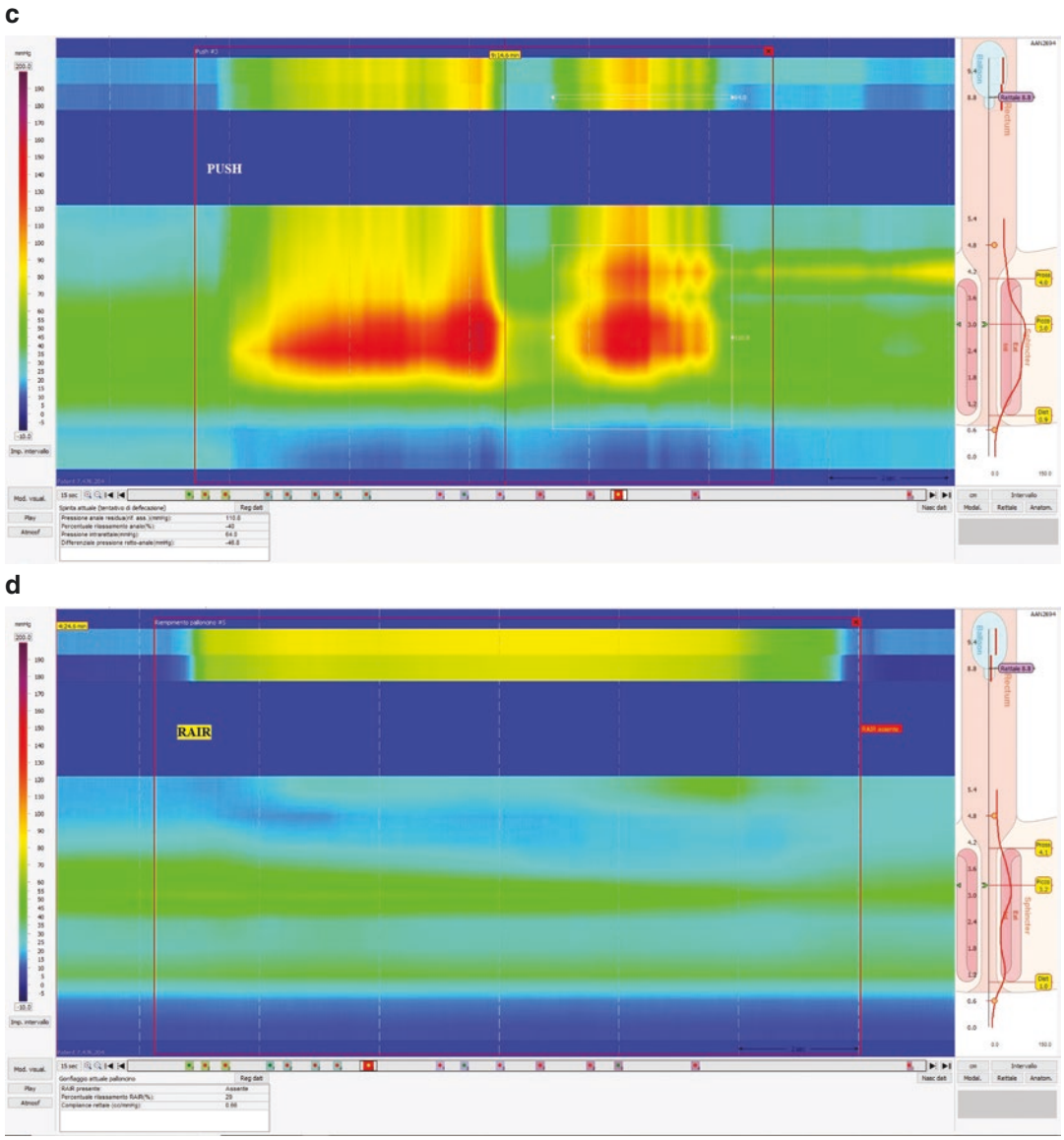
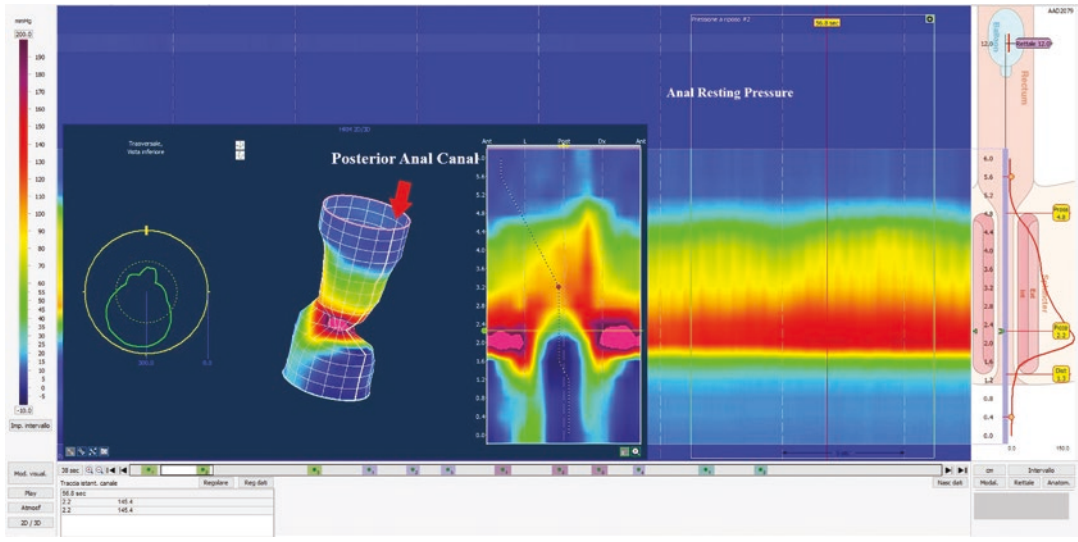


Fig. 8.1 (continued)

In 1998, the same authors reported long-term clinical and manometric data with regard to bowel function in a group of 47 patients treated surgically for ARMs who were 10–30 years.

In comparison to the previous group, poor results were observed in 12% of patients with high-type anomalies, the high-pressure zone was present in 73%, while the RAIR was elicited only in 20% of subjects. Therefore, as a result of these manometric evidences, the presence of RAIR and high-pressure zone was considered to be a chief parameter for objective assessment of anal

a



b

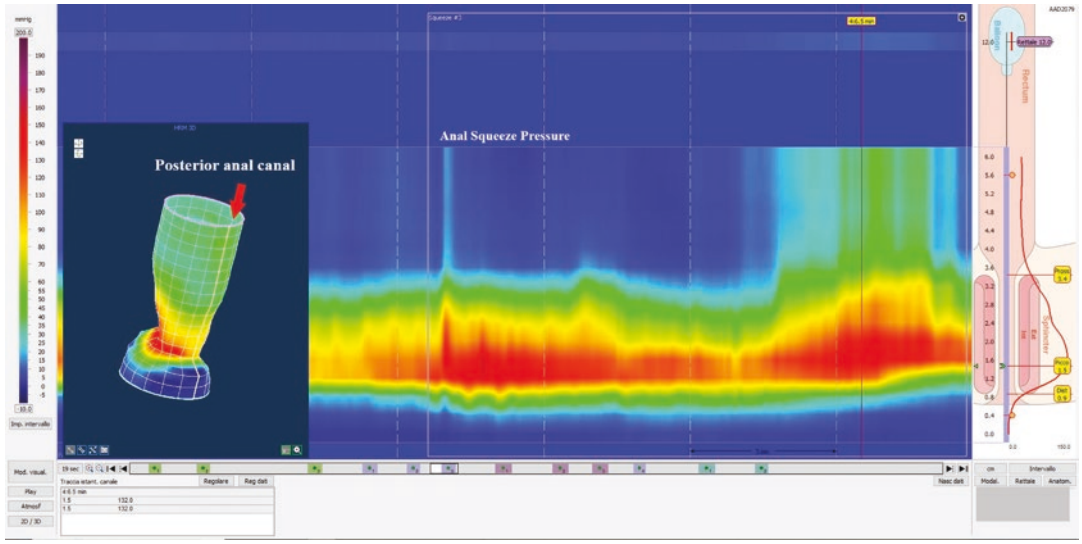


Fig. 8.2 6-year-old female patient with non-treated anterior anus and sacral agenesis, suffering from chronic constipation
HD-ARM detects high ARP (**a**) and ASP (**b**), pelvic floor dyssynergia type I according to Rao's classification (**c**), presence of RAIR (**d**)
HD-ARM High definition anorectal manometry, *ARP* Anal Resting Pressure, *ASP* Anal Squeeze Pressure, *RAIR* Recto-Anal Inhibitory Reflex
Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

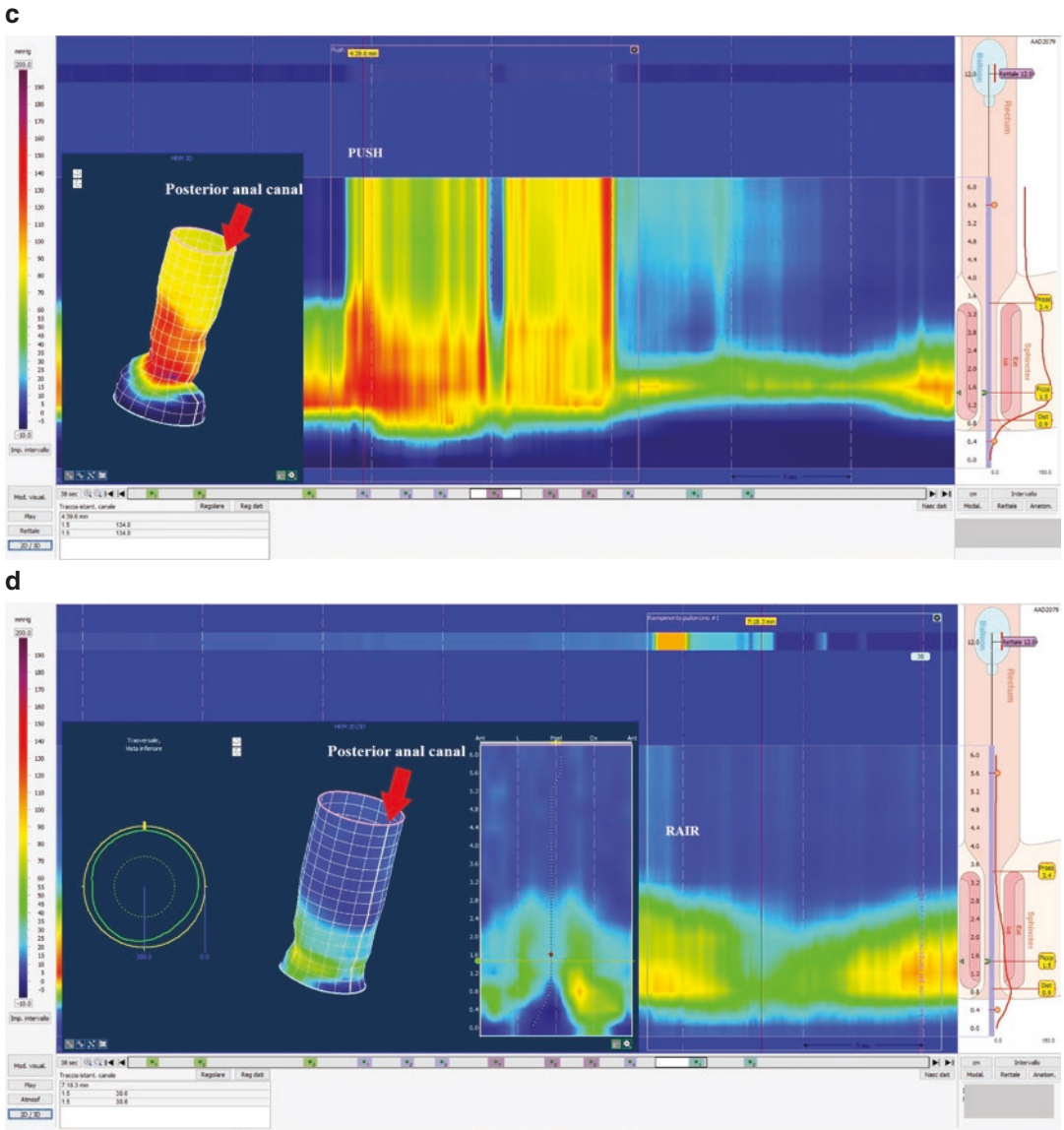


Fig. 8.2 (continued)

sphincter function. The improvement in terms of high-pressure zones in a long-term follow-up was considered secondary to the contractions of the voluntary muscles as intellectual development progresses or as a result of physiotherapeutic training [41].

In contrast to the above, Rintala et al. [42] noted no or very little improvement in sphincter function in terms of decreasing quantity of soiling with increasing age of the patients, probably due to the shorter follow-up. Other manometric and clinical factors were introduced in the evaluation

of bowel function: integrity of IAS and presence of sacral and spine congenital anomalies.

Manometrically, poor functional results were associated to low values of anal resting pressure and absence of RAIR which correlate with injuries of IAS. Clinically, patients affected by severe sacral or intraspinal deformities had an unsatisfactory continence outcome, often not related to the concomitant presence of poor sphincter function.

In a comparative manometric study between patients with ARMs and healthy children of different age, lower resting and squeezing pressures with impaired RAIR were demonstrated in all patients of the first group; the length of the anal canal, that contributes to continence, was significantly shorter in operated children who were more than 1 year old [43].

Further studies have underlined the role of anorectal manometry as an objective test compared to the subjective nature of the scoring systems used to evaluate faecal continence in different series of patients affected by ARMs. Based on their own results, clinical continence has been positively correlated with ARP [12, 26, 27, 42, 44, 45], ASP [46], normal rectal sensation [26, 42, 47] and presence of RAIR [26, 42, 44, 48–51], which is considered a good prognostic factor for faecal continence.

Kyrklund et al. [52] reported a good functional outcome with manometric evidence of normal anal pressures and positive RAIR in children with mild type of ARMs (anterior anus in females and perineal fistula in males) after minimally invasive procedures (anal dilatation or conservative follow-up for females with anterior anus and cutback anoplasty for males with perineal fistula).

Results were poorer among patients with more severe ARMs (vestibular and perineal fistula in females, recto-urethral fistula in males), who had lower ARP and ASP at the manometric test. However, RAIR was found in 83% of these patients after IAS-saving bowel mobilisation surgery (PSARP or anterior sagittal anorectoplasty—ASARP), entailing preservation of the distal fistulous bowel termination. These conclusions confirmed that functional IAS tissue may be found in the termination and that its preservation may influence the continence outcomes, as other researchers had previously detected [53, 54].

Data appear to be confirmed by the absence of RAIR after sacro-perineal and sacro-abdominoperineal interventions which involved resection of the terminal fistulous connection [55].

Additional manometric information descends from profilometric evaluation of the anal canal, defined by a computerised analysis of the pressure curves which is obtained using continuous-flow anorectal manometry. The computer programme generates three-dimensional tracings of the anorectal canal, in order to study the total and segmental asymmetry indices, the rectal volume and also the pressure distribution on the anorectal wall. This test was considered capable of providing reliable information concerning the three-dimensional topography of the anorectal canal and even the distribution of the pressures involved in the process of acquiring anorectal faecal continence.

According to Pedro et al., the first results of this technique showed a total and segmental asymmetry index similar between the continent, partially continent and incontinent patients, proving that surgery was technically correct; greater rectal volumes in partially continent subjects, compatible with constipation and soiling (pseudo-incontinence condition); a typical profilometric pattern with predominance of low pressures in the incontinent group [56].

Recently, 3D-HDARM has been used to evaluate patients after the repairing of ARMs in a long-term follow-up. Size and location of functional sphincter defects were determined through the 3D analysis of the anal resting pressure profile. A functional anal sphincter lesion was defined as a pressure area below 25 mmHg and the size was calculated manually through the 3D cylindrical pressure visualisation. About 50% of the participants showed sphincter defects that in 30% of cases affected more than half of the circumference. Soiling was found only in subjects with sphincter lesions.

A statistically significant correlation was documented between the type of ARMs, size of sphincter defect, mean ASP and Wexner incontinence score.

However, if 3D-HDARM is able to correlate sphincter defects to functional outcome, it is not able to make an anatomic distinction between the different elements of the sphincter complex [57].

8.6 Endoanal Ultrasonography

EAUS is currently considered the gold standard for the morphological evaluation of the anal canal in case of faecal incontinence. Most studies revealed a 100% sensitivity of EAUS in identifying sphincter defects, such as discontinuity, localised or generalised scarring, thinning and atrophy. An endosonographic sphincter lesion is described as an interruption in the normal texture of the muscle ring, while scarring is characterised by loss of texture that usually has low reflectiveness (Figs. 8.3 and 8.4).

Atrophic or degenerative sphincters are seen as thin and poorly defined, with heterogeneous increased echogenicity [58, 59].

On 2D images, the different echogenicity and the numerous interfaces of the structures form-

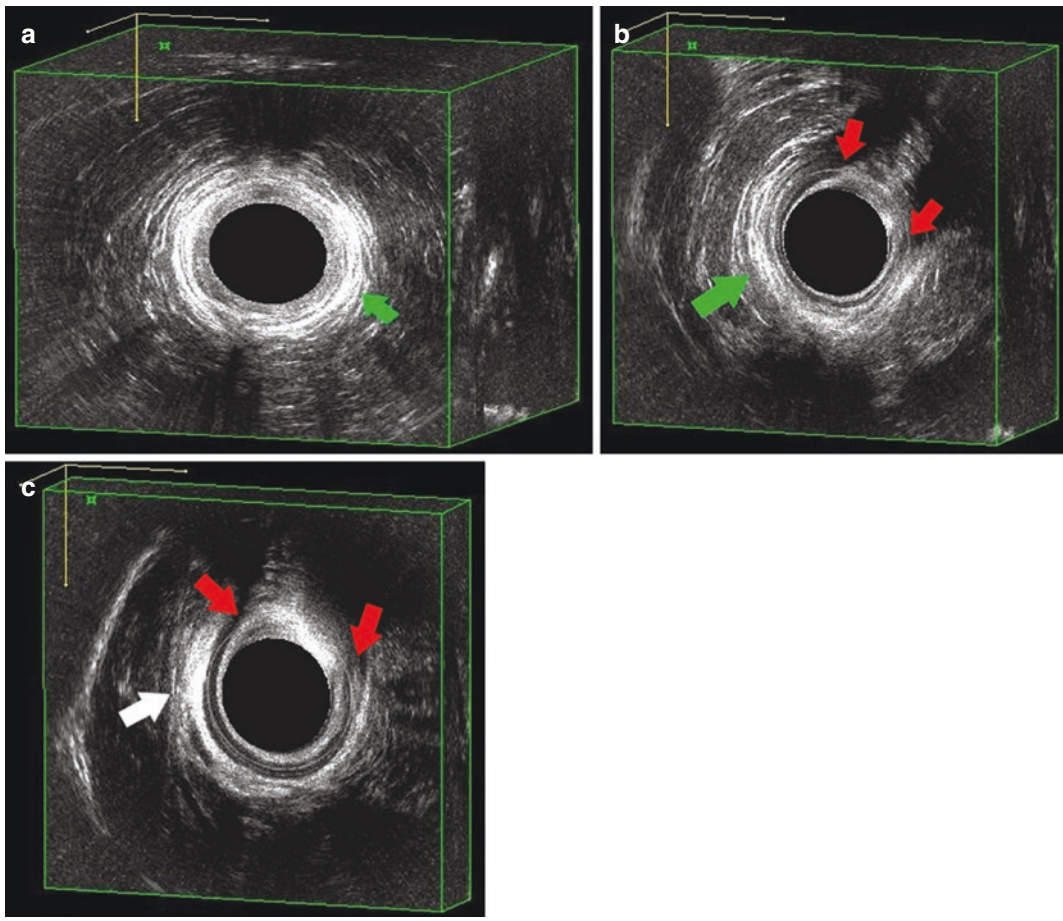


Fig. 8.3 8-year-old male patient with constipation and faecal soiling, surgically treated for anorectal malformation with recto-bulbar fistula (PSARP procedure)
3D-EAUS reveals:
(**a**) Distal anal canal: generalized scarring of EAS (green arrow)
(**b**) Middle anal canal: disruption of IAS (red arrows) and generalized scarring of EAS (green arrow)
(**c**) Proximal anal canal: defect of IAS (red arrows) and generalized scarring of pubo-rectalis muscle (white arrow)
3D-EAUS Three dimensional endoanal ultrasonography, *EAS* External Anal Sphincter, *IAS* Internal Anal Sphincter, *PSARP* Posterior Sagittal AnoRectoPlasty
Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

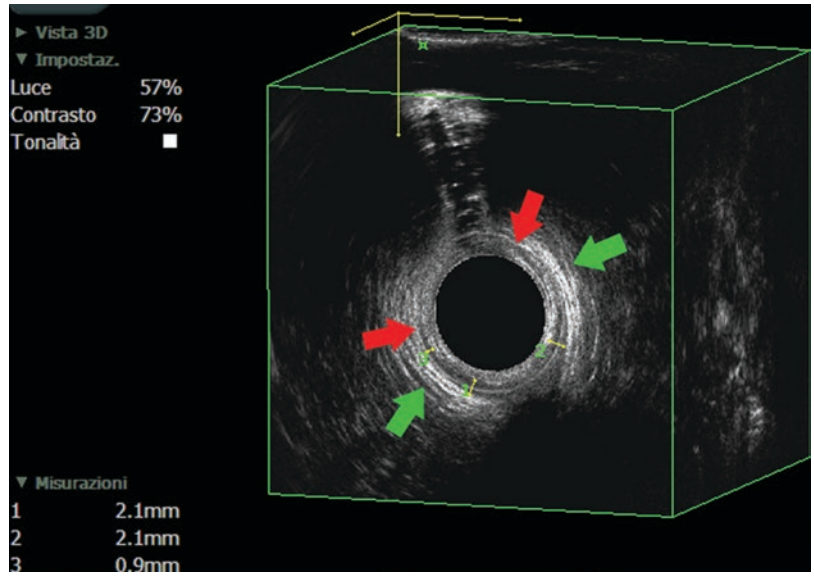


Fig. 8.4 9-year-old female patient with VACTER syndrome and recto-vaginal fistula 3D EAUS shows scars of EAS (green arrows) and lesion of IAS (red arrows); the latter appears thin and disomogeneous in the middle anal canal *3D-EAUS* Three dimensional endoanal ultrasonography, *EAS* External Anal Sphincter, *IAS* Internal Anal Sphincter *Source:* Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

ing the anal canal account for faithful anatomical depiction of the region and the ability to recognise the single muscular layers composing it [60].

The endoanal transducer consists of a crystal that rotates (4–6 cycles per second) in order to obtain a 360° image. A 6.0-cm-long image is captured along the proximal-distal axis for up to 55 s by moving the crystals on the extremity of the transducer [61].

High-resolution 2D images are usually recorded at three different levels of the anal canal (proximal, middle and distal), typified by the presence of [62–64]:

- IAS and puborectalis muscle appearing, respectively, as a hypoechoic ring and a hyperechoic horseshoe sling in the proximal anal canal.
- IAS and EAS in the middle anal canal: EAS forms a broad and mixed echogenicity ring tending to hyperechogenicity, which lies immediately outside the IAS.
- EAS in the distal anal canal.

The advent of 3D technology has further improved the understanding of the two-dimensional technique, also solving the downside of being an operator-dependent study.

3D reconstruction provides a multiplanar imaging of the anal canal, allowing length, thickness, area and volume measurement. After a 3D dataset has been acquired, it is possible to select coronal anterior-posterior or posterior-anterior as well as sagittal right–left views, together with any oblique image plane. The 3D image, showed as a "cube", can be rotated, tilted and sliced in any other direction to enable visualisation from different angles. This yields more information on the anal sphincter complex and makes it easier to perform sphincter measurements [63–66].

Several studies have compared EAUS with MRI, concluding that 3D-EAUS is superior in diagnosing IAS injury, equivalent in detecting EAS injury and inferior in identifying EAS atrophy [67].

A paediatric study on patients surgically treated for ARMs confirmed that EAUS has higher accuracy than MRI in recognising slight malposition of the neo-anus within the striated muscle complex and sphincter defects in subjects with abnormal muscle contraction on perineal muscle stimulation [68].

Besides, 3D EAUS has the plus of being easier, quicker, cheaper and better tolerated by patients than MRI; therefore it may be considered the method of choice [69] even in children.

Although a medical technician may also be able to technically perform EAUS, it is recommended that the procedure is carried out by a specialist to ensure the best insight of the awaited anatomy in accord to the underlying disease.

The joint committee of the Italian Society of Colorectal Surgery (SICCR) and the Italian Society of Ultrasonology in Medicine and Biology (SIUMB) defined the endoanal sonographic centres' accreditation requirements and established that physicians can be considered "expert" if they usually perform at least 5–6 studies per week [70].

An accurate inspection of the perineum and a digital rectal examination should be conducted to exclude severe stenosis of the anal opening and to avoid complications before 3D-EAUS, especially after reconstructive surgery of the anus.

Sedation is not indispensable in adulthood, but may be necessary for younger or complaining children.

8.7 Endoanal Ultrasonography and Anorectal Manometry in Anorectal Malformations

EAUS and anorectal manometry are combined investigations that should be used together to perform a complete estimation of anal sphincters in case of evacuatory dysfunctions (Figs. 8.5 and 8.6).

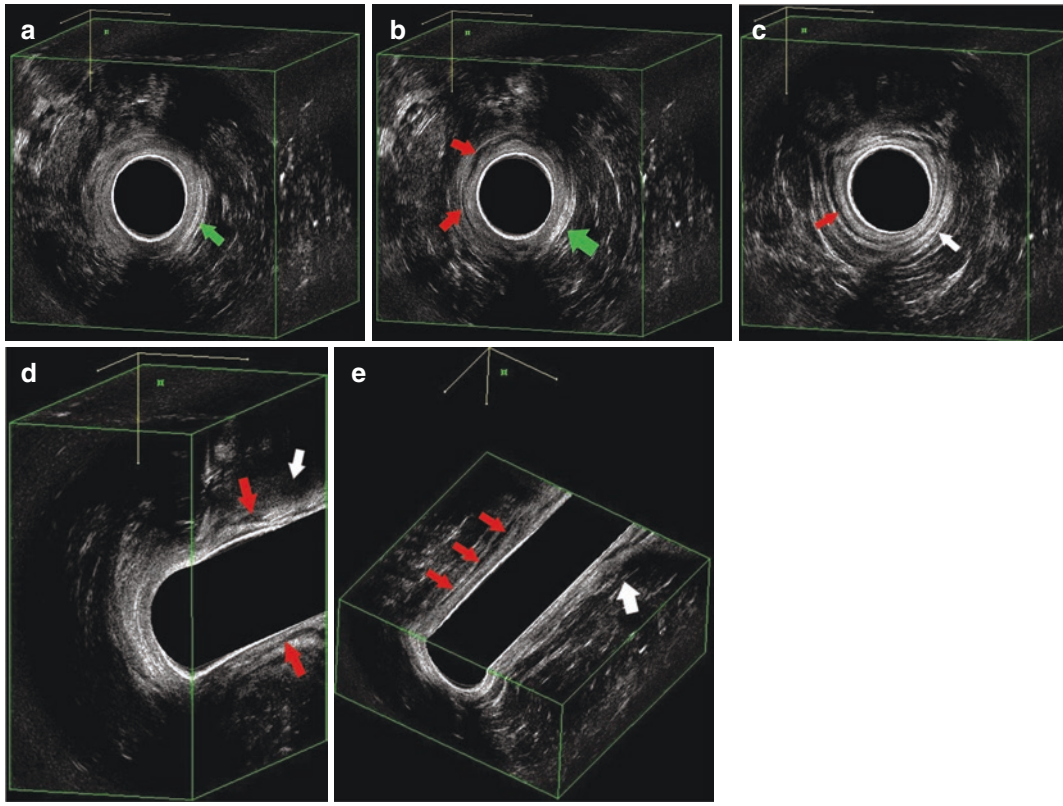


Fig. 8.5 3D endosonographic pattern of a 17-year-old female patient with VACTER syndrome and faecal incontinence

Axial sections:

(**a**) Distal anal canal: localized scarring of EAS (green arrow)

(**b**) Middle anal canal: fragmented IAS with only a few remnants (red arrows); scarring of EAS (green arrow)

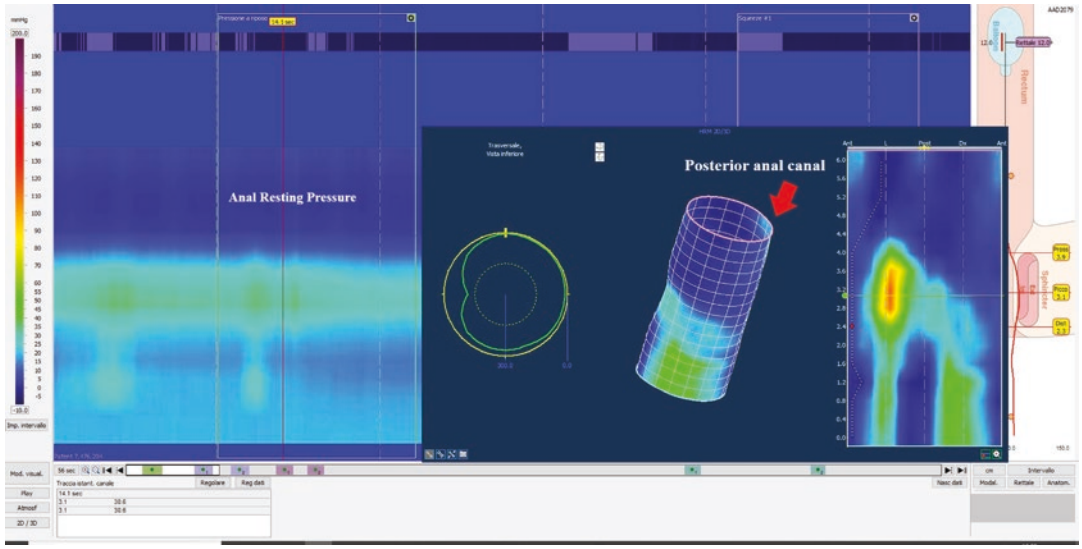
(**c**) Proximal anal canal: scar tissue (white arrow) of puborectalis muscle; fragmentation of IAS (red arrow)

Sagittal (**d**) and coronal (**e**) planes: thin fragments of IAS (red arrows) and atrophy of puborectalis muscle (white arrow)

EAS External Anal Sphincter; *IAS* Internal Anal Sphincter

Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

a



b

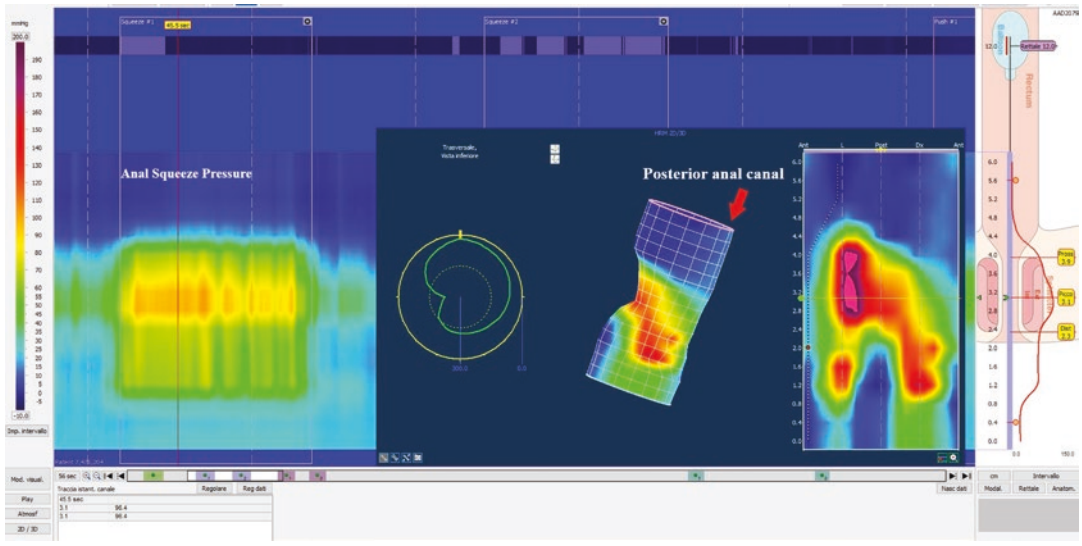


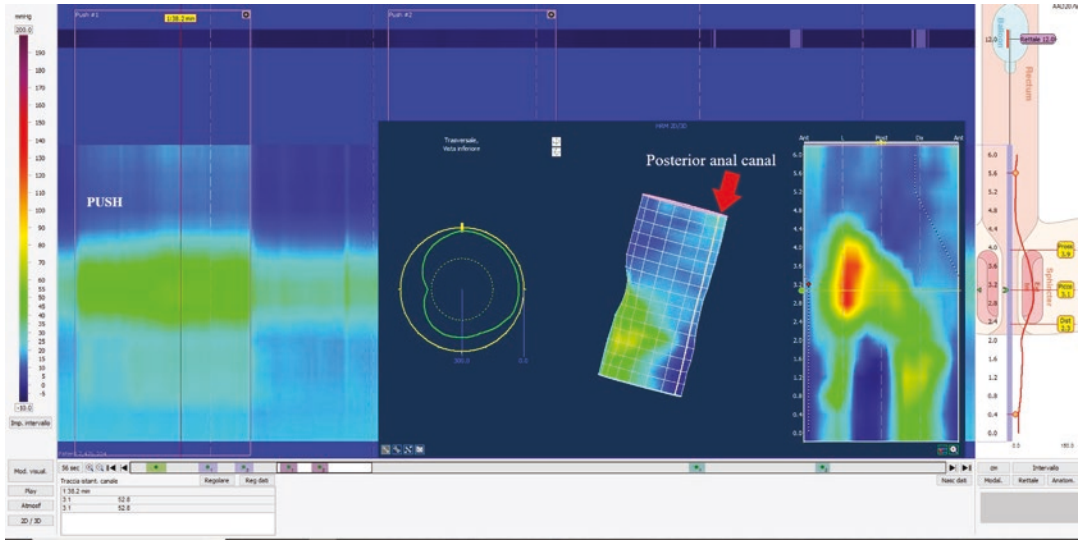
Fig. 8.6 High definition manometric pattern of a 17-year-old female patient with VACTER syndrome and faecal incontinence

HD-ARM shows low ARP (**a**) and ASP (**b**), no signs of pelvic floor dyssynergia (**c**) and absence of RAIR (**d**). Data of HD-ARM are complementary to the endosonographic findings (Fig. 8.5)

HD-ARM High definition anorectal manometry, *ARP* Anal Resting Pressure, *ASP* Anal Squeeze Pressure, *RAIR* Recto-Anal Inhibitory Reflex

Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

c



d

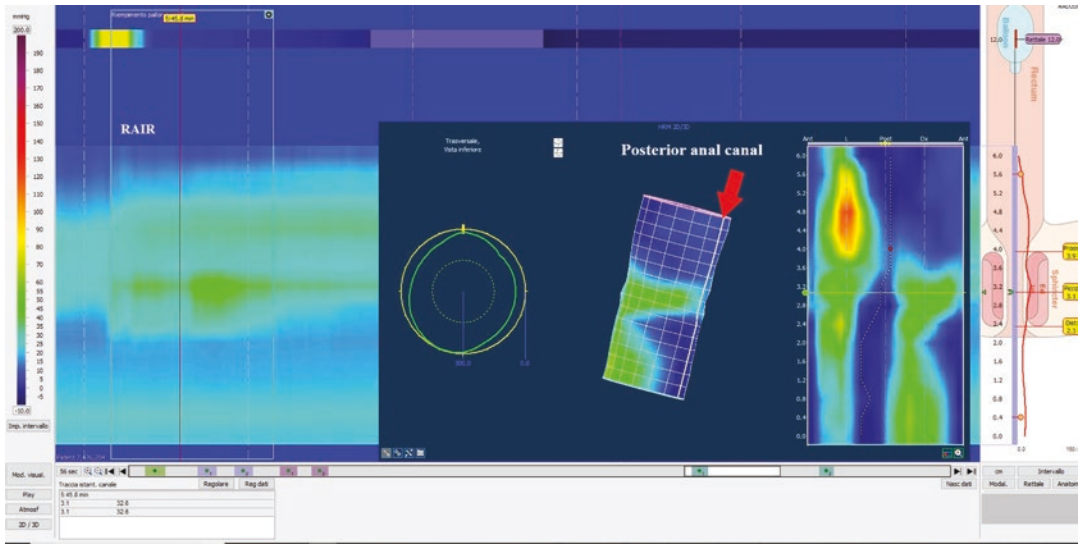


Fig. 8.6 (continued)

Although faecal incontinence may be the result of several causes, anal sphincter injury is highly prevalent, particularly after anorectal surgery.

The two procedures offer an objective baseline assessment of anal sphincter, and the results obtained may help to define the therapeutic medical or surgical programme for the improvement of faecal incontinence and constipation.

Until now, only a few studies have evaluated patients who had undergone reconstructive sur-

gery for anorectal anomalies by EAUS, but very interesting data have been derived from these series.

Emblem et al. found a strong correlation between clinical, manometric and endosonographic findings in patients with ARMs and a control group [71]. Specifically, children with high/intermediate ARMs had a poorer continence outcome, lower ARP and ASP and sphincter defects consisting of scars of IAS and EAS, some remnants of EAS and/or absence of IAS. In addition, the sphincter muscle complex and its relation to the anal opening were visualised by anal endosonography and varying degrees of eccentrically placed anal canal in the EAS were identified.

In a series of 54 children with ARMs, the anatomical integrity of the IAS correlated well with ARP, RAIR and type of ARMs (low-ARM group had superior quality of the IAS and better faecal continence than the one with high ARMs). Nevertheless, megarectum and and/or neuropathy are confirmed as unfavourable prognostic factors for faecal dynamics because their occurrence outweighs the benefit of good IAS and causes incontinence [48].

3D-EAUS could be superior to anorectal manometry in identifying mild and modest sphincter defects (Fig. 8.7).

As Caldaro et al. [12] reported, 3D-EAUS showed small/moderate disruptions of the IAS in a high percentage of cases with low ARMs, notwithstanding normal ARP and presence of RAIR.

In severe types of ARMs, correspondence between symptoms, manometric data and ultrasound findings was more significant; in these subjects, generalised scarring of the EAS, width defects and/or absence of the IAS were found (Figs. 8.8 and 8.9).

EAUS and anorectal manometry have been utilised even to document the effectiveness of PSARP to preserve the IAS and the post-operative anal functions in children with intermediate and high defects. In fact, if major differences in the thickness of IAS were discovered between patients with ARMs and healthy controls, no substantial dissimilarities were found between the PSARP group and transperineal anorectoplasty one, which components were affected by low lesions [47].

Both procedures are also useful exams for monitoring patients in the follow-up period and to plan therapeutic options.

Interesting clinical and endosonographic findings have been documented in a small group of adults affected by persistent faecal incontinence secondary to congenital imperforate anus [72].

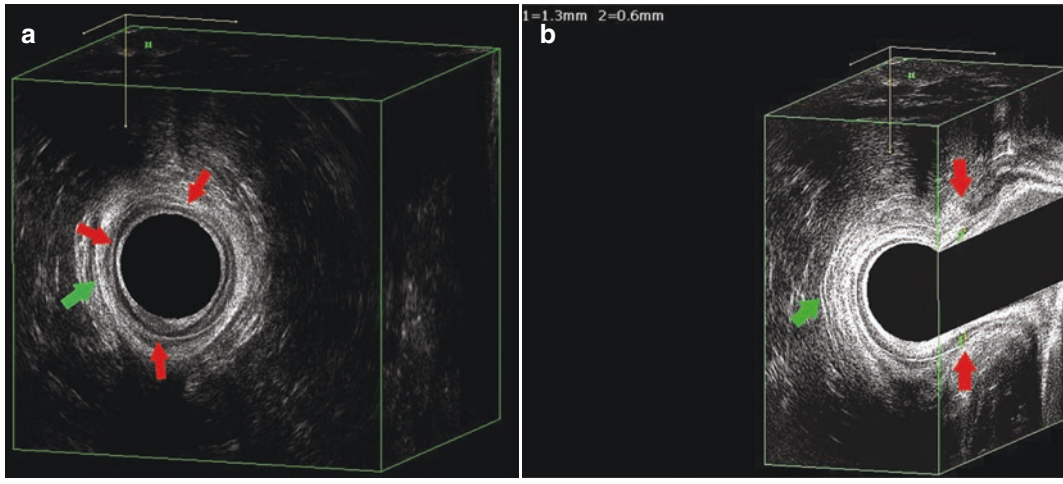


Fig. 8.7 3D EAUS in 5-year-old female patient operated on vestibular fistula, with chronic constipation
Axial (**a**) and sagittal (**b**) planes: Scarring of EAS (green arrow) and irregular thickness of IAS (red arrows)

3D-EAUS Three dimensional endoanal ultrasonography, *EAS* External Anal Sphincter, *IAS* Internal Anal Sphincter
Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

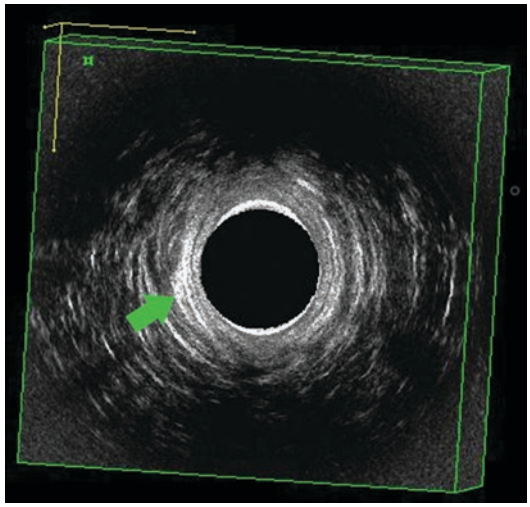


Fig. 8.8 9-year-old male patient with anorectal malformations and recto-bladder neck fistula. Clinical history of severe faecal incontinence
EAUS detects abundant scar tissue of the EAS (green arrow) and absence of the IAS in middle anal canal
EAUS Endoanal ultrasonography, *EAS* External Anal Sphincter, *IAS* Internal Anal Sphincter
Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

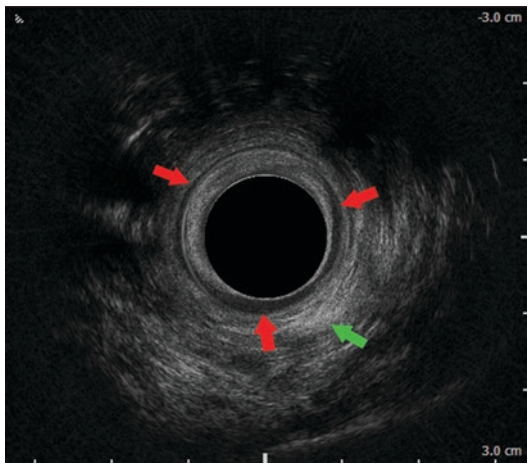


Fig. 8.9 17-year-old male patient with faecal incontinence secondary to anorectal malformation and recto-bulbar fistula
2D image of middle anal canal: EAUS identifies irregular thickness of the IAS (red arrows) and scarring of the EAS (green arrow)
EAUS Endoanal ultrasonography, *EAS* External Anal Sphincter, *IAS* Internal Anal Sphincter
Source: Digestive Surgery and Endoscopy Unit; Bambino Gesù Children's Hospital-IRCCS-Rome, Italy

Abnormal clinical evidences (e.g. anal stricture, prolapse, misplaced neo-anus) and EAS injuries were found in more than 90% and in 50% of cases, respectively. Conservative or surgical therapeutic choices have been proposed according to an algorithm of recommended management in relation to the causes of incontinence. Therefore, medical treatment was administered or optimised when absent or incongruous; biofeedback training was suggested when a patient with an intact sphincter was unable to contract it on demand and surgery was indicated in case of prolapse or anal stricture. Other minimally invasive therapies, as injection of bulking agents or sacral nerve stimulation (SNS), were advocated in non-responder subjects.

Injectable bulking therapy (non-animal stabilised hyaluronic acid with dextranomer-NASHA/Dx) has been used with a significant effect on the number of incontinence episodes in adults with rectobulbar or vestibular fistula [73]. All patients were assessed preoperatively and in follow-up by 3D-EAUS to evaluate migration of the implants.

SNS seems to be another promising alternative treatment [74]. Data are partial and heterogeneous, but encouraging results have been achieved in subjects with intact IAS, low ARP and ASP and even in the presence of partial sacral agenesis, which could on the other hand complicate the placement of stimulator leads in the foramina of the sacrum [75, 76].

Patients suffering from such severe incontinence that has not been possible to amend with other less invasive measures could be candidates to surgical correction. In fact, although the outcome of dynamic graciloplasty in cases of ARMs was proved inferior compared to the ones with other underlying aetiologies, it may be indicated in selected occurrences in which forming a stoma is the next step [77].

8.8 Conclusions

Faecal incontinence and chronic constipation are disabling conditions due to congenital, anatomical and post-surgical factors in patients operated on ARMs, with impact on the quality of life [78].

In adulthood, anorectal manometry and EAUS are validated tests in the management of anorectal disorders. EAUS has a high degree of sensitivity and specificity and correlates well with manometric findings, so both investigations are considered complementary in assessing the morphology and function of anal sphincters [79].

In paediatrics, instead, controlled prospective studies aiming to evaluate the impact of these exams on treatments and long-term outcomes are still limited.

However, the results achieved so far have provided useful information to define the anorectal pathophysiology, even in patients with AMRs; consequently it should be mandatory to consider EAUS and manometry in the diagnostic algorithm of evacuatory dysfunction.

References

1. Rigueros Springford L, Connor MJ, Jones K, Kapetanakis VV, Giuliani S. Prevalence of active long-term problems in patients with anorectal malformations: a systematic review. Dis Colon Rectum. 2016;59:570–80. https://doi.org/10.1097/DCR.0000000000000576.
2. Ludman L, Spitz L, Kiely EM. Social and emotional impact of fecal incontinence after surgery for anorectal abnormalities. Arch Dis Child. 1994;71:194–200.
3. Minneci PC, Kabre RS, Mak GZ, Halleran DR, Cooper JN, Afrazi A, et al. Can fecal continence be predicted in patients born with anorectal malformations? J Pediatr Surg. 2019;54:1159–63. https://doi.org/10.1016/j.jpedsurg.2019.02.035.
4. Wood RJ, Marc A, Levitt MA. Anorectal malformations. Clin Colon Rectal Surg. 2018;31:61–70.
5. Tantiphlachiva K. Comprehensive clinical approach to fecal incontinence. Book chapter doi: https://doi.org/10.5772/intechopen.86346.
6. Frischer JS, Rymeski B. Complications in colorectal surgery. Semin Pediatr Surg. 2016;25:380–7. https://doi.org/10.1053/j.sempedsurg.2016.10.008.
7. Stephens FD, Smith ED. Classification, identification, and assessment of surgical treatment of anorectal anomalies. Pediatr Surg Int. 1986;1:200–5.
8. Peña A. Anorectal malformations. Semin Pediatr Surg. 1995;4:35–47.
9. Holschneider A, Hutson J, Peña A, Beket E, Chatterjee S, Coran A, et al. Preliminary report on the international conference for the development of standards for the treatment of anorectal malformations. J Pediatr Surg. 2005;40:1521–6. https://doi.org/10.1016/j.jpedsurg.2005.08.002.
10. Gupta DK. Anorectal malformations—wingspread to Krickenbeck. J Indian Assoc of Pediatr Surg. 2005;10:75–7.
11. Guillaume A, Salem AE, Garcia P, Chander RB. Pathophysiology and therapeutic options for fecal incontinence. J Clin Gastroenterol. 2017;51:324–30. https://doi.org/10.1097/MCG.0000000000000797.
12. Caldaro T, Romeo E, De Angelis P, Gambitta RA, Rea F, Torroni F, et al. Three-dimensional endoanal ultrasound and anorectal manometry in children with anorectal malformations: new discoveries. J Pediatr Surg. 2012;47:956–63.
13. Borg H, Bachelard M, Sillén U. Megarectosigmoid in children with anorectal malformations: long term outcome after surgical or conservative treatment. J Pediatr Surg. 2014;49:564–9. https://doi.org/10.1016/j.jpedsurg.2013.08.003.
14. Burjonrappa S, Youssef S, Lapierre S, Bensoussan A, Bouchard S. Megarectum after surgery for anorectal malformations. J Pediatr Surg. 2010;45:762–8. https://doi.org/10.1016/j.jpedsurg.2009.10.043.
15. Miyahara K, Kato Y, Seki T, Arakawa A, Lane GJ, Yamataka A. Neuronal immaturity in normoganglionic colon from cases of Hirschsprung disease, anorectal malformation, and idiopathic constipation. J Pediatr Surg. 2009;44:2364–8. https://doi.org/10.1016/j.jpedsurg.2009.07.066.
16. Holbrook C, Misra D, Zaparackaite I, Cleeve S. Postoperative strictures in anorectal malformation: trends over 15 years. Pediatr Surg Int. 2017;33:869–73. https://doi.org/10.1007/s00383-017-4111-6.
17. Kyrklund K, Pakarinen MP, Rintala RJ. Long-term bowel function, quality of life and sexual function in patients with anorectal malformations treated during the PSARP era. Semin Pediatr Surg. 2017;26:336–42. https://doi.org/10.1053/j.sempedsurg.2017.09.010.
18. Peña A, Bischoff A. Surgical treatment of colorectal problems in children. Springer; 2015. https://doi.org/10.1007/978-3-319-14989-9.
19. Gurusamy R, Raj SV, Maniam R, Regunandan SR. Laparoscopic-assisted anorectoplasty: a single-center experience. J Indian Assoc Pediatr Surg. 2017;22:114–8.
20. Peña A, Hong A. Advances in the management of anorectal malformations. Am J Surg. 2000;180:370–6. https://doi.org/10.1016/s0002-9610(00)00491-8.
21. Pathak M, Saxena AK. Postoperative "complications" following laparoscopic-assisted anorectoplasty: a systematic review. Pediatr Surg Int. 2020;36:1299–307. https://doi.org/10.1007/s00383-020-04748-3.
22. Rodriguez L, Sood M, Di Lorenzo C, Saps M. An ANMS-NASPGHAN consensus document on anorectal and colonic manometry in children. Neurogastroenterol Motil. 2017;29:10.1111.
23. Rasijeff AMP, Withers M, Burke JM, Jackson W, S. Scott SM. High-resolution anorectal manometry: a comparison of solid-state and water-perfused catheters. Neurogastroenterol Motil. 2017;29:e13124.
24. Scott SM, Emma V, Carrington EV. The London classification: improving characterization and classification of anorectal function with anorectal manometry. Curr Gastroenterol Rep. 2020;22:–55. https://doi.org/10.1007/s11894-020-00793-z.

25. Tambucci R, Quitadamo P, Thapar N, Zenzeri L, Caldaro T, Staiano A, Verrotti A, Borrelli O. Diagnostic tests in pediatric constipation. J Pediatr Gastroenterol Nutr. 2018;66(4):e89–98. https://doi.org/10.1097/MPG.0000000000001874.
26. Hedlund H, Peña A, Rodriguez G, Maza J. Long-term anorectal function in imperforate anus treated by a posterior sagittal anorectoplasty: manometric investigation. J Pediatr Surg. 1992;27:906–9.
27. Senel E, Demirbag S, Tiryaki T, Erdogan D, Cetinkursun S, Cakmak O. Postoperative anorectal manometric evaluation of patients with anorectal malformation. Pediatr Int. 2007;49:210–4. https://doi.org/10.1111/j.1442-200X.2007.02342.x.
28. Rintala RJ. Fecal incontinence in anorectal malformations, neuropathy, and miscellaneous conditions. Semin Pediatr Surg. 2002;11:75–82.
29. Lee TH, Bharucha AE. How to perform and interpret a high-resolution anorectal manometry test. J Neurogastroenterol Motil. 2016;31(22):46–59. https://doi.org/10.5056/jnm15168.
30. William PT, John CA, Patrick DB. Neuroradiologic evaluation of sacral abnormalities in imperforate anus complex. J Pediatr Surg. 1987;22:58–61.
31. Capitanucci ML, Rivosecchi M, Silveri M, Lucchetti MC, Mosiello G, De Gennaro M. Neurovesical dysfunction due to spinal dysraphism in anorectal anomalies. Eur J Pediatr Surg. 1996;6:159–62.
32. Yuan Z, Bai Y, Zhang Z, Ji S, Li Z, Wang W. Neural electrophysiological studies on the external anal sphincter in children with anorectal malformation. J Pediatr Surg. 2000;35:1052–7. https://doi.org/10.1053/jpsu.2000.7770.
33. Chiarioni G, Kim SM, Vantini I, Whitehead WE. Validation of the balloon evacuation test: reproducibility and agreement with findings from anorectal manometry and electromyography. Clin Gastroenterol Hepatol. 2014;12:2049–54.
34. Rao SS. Dyssynergic defecation and biofeedback therapy. Gastroenterol Clin N Am. 2008;37:569–86. viii. https://doi.org/10.1016/j.gtc.2008.06.011.
35. Rao SS, Ozturk R, Laine L. Clinical utility of diagnostic tests for constipation in adults: a systematic review. Am J Gastroenterol. 2005;100:1605–15.
36. Palsson OS, Heymen S, Whitehead WE. Biofeedback treatment for functional anorectal disorders: a comprehensive efficacy review. Appl Psychophysiol Biofeedback. 2004;29:153–74.
37. Rao SS, Tuteja AK, Vellema T, et al. Dyssynergic defecation: demographics, symptoms, stool patterns, and quality of life. J Clin Gastroenterol. 2004;38:680–5.
38. van Meegdenburg MM, Heineman E, Broens PMA. Dyssynergic defecation may aggravate constipation: results of mostly pediatric cases with congenital anorectal malformation. Am J Surg. 2015;210:357–64.
39. Bjørsum-Meyer T, Christensen P, Baatrup G, Jakobsen MS, Asmussen J, QvistN. Dyssynergic patterns of defecation in constipated adolescents and young adults with anorectal malformations. Sci Rep. 2020;10(19673) https://doi.org/10.1038/s41598-020-76841-5.
40. Iwai N, Ogita S, Kida M, Fujita Y, Majima S. A clinical and manometric correlation for assessment of postoperative continence in imperforate anus. J Pediatr Surg. 1979;14:538–43.
41. Iwai N, Yanagihara J, Tokiwa K, Deguchi E, Takahashi T. Results of surgical correction of anorectal malformations. A 10-30 year follow-up. Ann Surg. 1988;207:219–22.
42. Rintala RJ, Lindahl H. Is Normal bowel function possible after repair of intermediate and high anorectal malformations? J Pediatr Surg. 1995;30:491–4.
43. Kumar S, Ramadan SA, Gupta V, Helmy S, Debnath P, Alkholy A. Use of anorectal manometry for evaluation of postoperative results of patients with anorectal malformation: a study from Kuwait. J Pediatr Surg. 2010;45:1843–8.
44. Iwai N, Hashimoto K, Goto Y, Majima S. Long-term results after surgical correction of anorectal malformations. Z Kinderchir. 1984;39:35–9.
45. Heikenen JB, Werlin SL, Di Lorenzo C, Hyman PE, Cocjin J, Flores AF, et al. Colonic motility in children with repaired imperforate anus. Dig Dis Sci. 1999;44:1288–92.
46. Molander ML, Frenckner B. Anal sphincter function after surgery for high imperforate anus—a long term follow-up investigation. Z Kinderchir. 1985;40:91–6.
47. Wang Z, Hu L, Jin X, Li X, Xu L. Evaluation of postoperative anal functions using endoanal ultrasonography and anorectal manometry in children with congenital anorectal malformations. J Pediatr Surg. 2016;51:416–20.
48. Keshtgar AS, Athanasakos E, Clayden GS, Ward HC. Evaluation of outcome of anorectal anomaly in childhood: the role of anorectal manometry and endosonography. Pediatr Surg Int. 2008;24:885–92.
49. Aspirot A. Anorectal malformations. In: Faure C, Thapar N, Di Lorenzo C, editors. Pediatric neurogastroenterology. Cham: Springer; 2017. https://doi.org/10.1007/978-3-319-43268-7_29.
50. Kelly JH. The clinical and radiological assessment of anal continence in childhood. Aust NZJ Surg. 1972;42:62–3.
51. Kiesewetter WB, Chang JH. Imperforate anus: a five to thirty-year follow-up perspective. Prog Pediatr Surg. 1977;10:111–20.
52. Kyrklund K, Pakarinen MP, Rintala RJ. Manometric findings in relation to functional outcomes in different types of anorectal malformations. J Pediatr Surg. 2017;52:563–8.
53. Kluth D, Hillen M, Lamprecht W. The principles of normal and abnormal hindgut development. J Pediatr Surg. 1995;30:1143–7.
54. Ruttenstock EM, Zani A, Huber-Zeyringer A, Höllwarth ME. Pre- and postoperative rectal manometric assessment of patients with anorectal malformations: should we preserve the fistula? Dis Colon Rectum. 2013;56:499–504. https://doi.org/10.1097/DCR.0b013e31826e4a38.

55. Rintala RJ, Lindahl HG. Posterior sagittal anorectoplasty is superior to sacroperineal sacroabdominoperineal pull-through: a long-term follow-up study in boys with high anorectal anomalies. J Pediatr Surg. 1999;34:334–7.
56. Júnior PFV, Martins JL, Peterlini FL. Posterior sagittal anorectoplasty in anorectal anomalies: clinical, manometric and profilometric evaluation. Sao Paulo Med J. 2007;125:163–9.
57. Bjørsum-Meyer T, Christensen P, Jakobsen MS, Baatrup G, Qvist N. Correlation of anorectal manometry measures to severity of fecal incontinence in patients with anorectal malformations—a cross sectional study. Sci Rep. 2020;10:19673. https://doi.org/10.1038/s41598-020-76841-5.
58. Sultan AH, Kamm MA, Talbot IC, Nicholls RJ, Bartram CI. Anal endosonography for identifying external sphincter defects confirmed histologically. Br J Surg. 1994;81:463–5.
59. Nichols CM, Lamb EH, Ramakrishnan V. Differences in outcomes after third-versus fourth-degree perineal laceration repair: a prospective study. Am J Obstet Gynecol. 2005;193:530–4.
60. Reginelli A, Mandato Y, Cavaliere C, Pizza NL, Russo A, Cappabianca S, et al. Three-dimensional anal endosonography in depicting anal-canal anatomy. Radiol Med. 2012;117:759–71.
61. Murad Regadas SM, Regadas FS, Rodrigues LV, Holanda EC, Barreto RG, et al. The role of 3-dimensional anorectal ultrasonography in the assessment of anterior trans-sphincteric fistula. Dis Colon Rectum. 2010;53:1035–40.
62. Hussain SM, Stoker J, Laméris JS. Anal sphincter complex: endoanal MR imaging of normal anatomy. Radiology. 1995;197:671–7.
63. Abdool Z, Sultan AH, Thakar R. Ultrasound imaging of the anal sphincter complex: a review. Br J Radiol. 2012;85:865–75.
64. Villanueva Herrero JA, Hansen MR, Bernes LA, Bobadill J. Endoanal ultrasound for anorectal diseases. In: Gastrointestinal diseases. SM Group. p. 1–23.
65. Albuquerque A. Endoanal ultrasonography in fecal incontinence: current and future perspectives. World J Gastrointest Endosc. 2015;10:575–81.
66. Gosselink MP, West RL, Kuipers EJ, Hansen BE, Schouten WR. Integrity of the anal sphincters after pouch-anal anastomosis: evaluation with three-dimensional endoanal ultrasonography. Dis Colon Rectum. 2005;48:1728–35.
67. Nuernberg D, Saftoiu A, Barreiros AP, Burmester E, Ivan ET, Clevert DA, et al. EFSUMB recommendations for gastrointestinal ultrasound part 3: endorectal. Endoanal Perineal Ultrasound Int Open. 2019;5:E34–51.
68. Athanasakos EP, Ward HC, Williams NS, Scott SM. The value of anal endosonography compared with magnetic resonance imaging following the repair of anorectal malformations. Pediatr Radiol. 2003;33:183–5.
69. Emblem R, Morkrid L, Bjornland K. Anal endosonography is useful for postoperative assessment of anorectal malformations. J Pediatr Surg. 2007;42:1549–54.
70. Dal Corso HM, D'Elia A, De Nardi P, Cavallari F, Favetta U, et al. Anal endosonography. A survey of equipment, technique and diagnostic criteria adopted in nine Italian centers. Tech Coloproctol. 2007;11:26–33.
71. Emblem R, Diseth T, Morkrid L. Anorectal anomalies: anorectal manometric function and anal endosonography in relation to functional outcome. Pediatr Surg Int. 1997;12:516–9.
72. Danielson J, Karlbom U, Graf W, Wester T. Persistent fecal incontinence into adulthood after repair of anorectal malformations. Int J Color Dis. 2019;34(3):551–4. https://doi.org/10.1007/s00384-018-3220-6.
73. Danielson J, Karlbom U, Wester T, Graf W. Injectable bulking treatment of persistent fecal incontinence in adult patients after anorectal malformations. J Pediatr Surg. 2020;55:397–402.
74. Dewberry L, Trecartin A, Peña A, Pierre MS, Bischoff A. Systematic review: sacral nerve stimulation in the treatment of constipation and fecal incontinence in children with emphasis in anorectal malformation. Pediatr Surg Int. 2019;35:1009–12. https://doi.org/10.1007/s00383-019-04515-z.
75. Thomas GP, Nicholls RJ, Vaizey CJ. Sacral nerve stimulation for faecal incontinence secondary to congenital imperforate anus. Tech Coloproctol. 2013;17:227–9.
76. Eftaiha SM, Melich G, Pai A, Marecik SJ, Prasad LM, Park JJ. Sacral nerve stimulation in the treatment of bowel dysfunction from imperforate anus: a case report. Int J Surg Case Rep. 2016;24:115–8. https://doi.org/10.1016/j.ijscr.2016.05.018.
77. Danielson J, Karlbom U, Wester T, Graf W. Long-term outcome after dynamic graciloplasty for treatment of persistent fecal incontinence in patients with anorectal malformations. Eur J Pediatr Surg. 2019;29(3):276–81. https://doi.org/10.1055/s-0038-1641599.
78. Reddymasu SC, Singh S, Waheed S, Oropeza-Vail M, McCallum RW, Olyaee M. Comparison of anorectal manometry to endoanal ultrasound in the evaluation of fecal incontinence. Am J Med Sci. 2009;337:336–9.
79. Bordeianou L, Lee K, Rockwood T, Baxter N, Lowry A, Mellgren A, et al. Anal resting pressures at manometry correlate with the fecal incontinence severity index and with presence of sphincter defects on ultrasound. Dis Colon Rectum. 2018;51:1010–4.