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ORIGINAL ARTICLE

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Evaluation of dysfunction following reconstruction of an anorectal anomaly

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Abstract To evaluate the utility of anorectal manometry (ARM) and magnetic resonance imaging (MRI) with an endocoil in the assessment of dysfunction in children with repaired anorectal anomalies (ARA), 15 patients aged 1 to 15 years with repaired ARAs and chronic faecal incontinence or constipation were prospectively recruited. They underwent clinical assessment using a modified Wingfield score (MWS). ARM and MRI with an endocoil and conventional external coil were carried out. The results of ARM alone, MRI alone, and a combination of ARM and MRI were correlated with the MWS. Manometric internal anal sphincter (IAS) scores determined from sphincter length and activity correlated with MWS ($r = 0.56$, $P = 0.02$); manometric scores of rectal peristaltic activity did not. Overall manometric score (IAS and rectal scores combined) showed a correlation with MWS ($r = 0.55$, $P = 0.02$). Endoanal MRI sphincter scores did not correlate with MWS, but the presence of a megarectum on MRI did ($r = 0.44$, $P = 0.05$). Overall MRI score did not correlate with MWS. Minor neurosacral anomalies were shown on MRI in 3 children who had poor functional scores. Combined manometric and MRI scores showed a correlation with MWS ($r = 0.58$, $P = 0.01$). ARM and MRI are potentially useful in the assessment of dysfunction of children with repaired ARAs. Both modalities require refinement and further assessment in the context of directing management.

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Introduction

Incontinence and constipation occur in a high proportion of children with anorectal anomalies (ARA) of all grades after reconstructive surgery [1–3]. This leads to physical, psychological, and social morbidity [4]. Anorectal manometry (ARM) is a promising modality for assessing dysfunction in this context [5]. Magnetic resonance imaging (MRI) with an endoanal coil provides exquisite anatomical detail of the anal canal and pelvic floor and may offer some benefit over endoanal ultrasound [6–8]. Endoanal MRI can be combined with conventional MRI to assess associated neurosacral anomalies that also require further evaluation [9].

The aim of this study was to assess ARM and MRI separately and together in children with repaired ARAs by correlating them with clinical status. Clarity as to the causes of dysfunction may lead to better management.

Patients and methods

Fifteen children with chronic incontinence and constipation, median age 2.5 years (range 1 to 15), were recruited. Six had high, 5 had intermediate, and 4 had low anomalies. A variety of techniques had been used for reconstruction by a number of surgeons. Associated problems included genitourinary anomalies, chromosomal abnormalities (partial trisomy 22 in 2, not associated with learning difficulties), and 2 cases of compensated or corrected congenital heart disease. No child had a neuropathy of the lower limbs, but 3 had minor sacroneural abnormalities, 2 of which were clinically evident as deformities of the sacrum.

Children were assessed clinically prior to investigation using the Wingfield score, modified to include constipation (MWS) (Table 1), as this was the major problem in some cases. A simple scoring system was chosen because of the wide age range. Subsequent scoring of ARM and MRI was done blinded to the results of MWS.

ARM was carried out during ketamine anesthesia, which has been validated in children and allows the internal anal sphincter (IAS) to be assessed in detail [10]. The manometry probe had four

Table 1. Modified Wingfield score

0 =	Normal anorectal function (none in this study)
1 =	Constipated
2 =	Intermittent passage of stool, some soiling
3 =	Continuous soiling

water-filled anal chambers, each 1 cm long, with a rectal balloon set 5 cm above the highest chamber, which was inflated with air during the study with 25- or 50-ml aliquots until complete inhibition of the IAS occurred or discomfort was apparent, or a maximal volume of 300 ml was reached. In normal patients individual inflation of the rectal balloon produces a sharp spike followed by a simple wave of rectal peristalsis and a progressive fall in IAS pressure, with complete inhibition at 100 to 150 ml.

A qualitative scale not involving critical volume was constructed for rectal function (Table 2), because not all children with repaired ARAs have a rectoanal reflex and because of the large variation in size of the children. In addition, studies were carried out on unprepared bowel because of the impossibility of evacuating many of the children by non-invasive measures. The IAS score was constructed on the basis of reflex activity and length (Table 3).

Endoanal MRI was carried out with cylindrical saddle geometry coils 7–12 mm in diameter and 6–10 cm in length [8]. On completion of the endoanal study, the internal coil was removed and T1-weighted spin-echo (720/20 ms) sagittal and coronal images of the pelvic floor and sacrum were obtained using an external quadrature coil appropriate to the size of the child. Children below 2 years required sedation with 50–80 mg chloral hydrate/kg body weight. Children between 2 and 8 years were sedated using up to 5 mg intranasal midazolam and IV propofol titrated to the patient's response. The remaining patients between 11 and 15 years of age were imaged without sedation.

MRI scoring was carried out as follows: 0 = normal; 1 = mild asymmetry; 2 = over 25% reduction in length or thickness of the IAS or external sphincter compared to the contralateral side; and 3 = over 50% reduction in length or thickness of the IAS or external sphincter compared to the contralateral side. In addition, the presence of megarectum was allocated a score of 1 and its absence 0, giving an overall MRI score of 0 to 4.

Table 2. Manometric rectal score

0 =	Normal spike and wave response to each inflation of rectal balloon
1 =	More than two spikes and waves of good quality
2 =	Hyperactive spikes and waves
3 =	Two or less spikes and waves of good quality
4 =	Two or more spikes and waves of poor quality
5 =	Two or less spikes and waves of poor quality
6 =	No defined spikes and waves

Table 3. IAS activity and length scores (sum = total IAS score)

IAS activity score	
0 =	Regular inhibitory response to rectal distension
1 =	Greater than two good inhibitory responses
2 =	Two or less good inhibitory responses
3 =	Greater than two poor inhibitory responses
4 =	Two or less poor inhibitory responses
5 =	No IAS activity
IAS score of length	
0 =	Activity in all four chambers
1 =	Activity in three chambers
2 =	Activity in two chambers
3 =	Activity in one chamber
4 =	No activity

One-tailed Spearman rank-correlation analyses were applied. Permission for the MRI arm of the study was obtained from the Royal Postgraduate Medical School Research Ethics Committee, and written consent was obtained in each case. Manometry is part of the normal clinical evaluation of these problems at Guy's and St Thomas' NHS Trust Hospitals.

Results

Patient details and MWS scores are shown in Table 4. It is of note that children tended to present early with severe morbidity, at a median age of 2.5 years in this study. Manometric and MRI results are shown in Tables 5 and 6, respectively, and summarised in Table 7. There was no correlation between functional score and severity of anomaly in this group, which was to be expected in that they were selected for their problems rather than being representative of an unselected group of children with repaired ARAs. Two children with identifiable neurosacral anomalies had a MWS of 3 and the 3rd 2. The 2 children with MWS of 3 scored poorly

Table 4. Patient details and clinical assessment of function *MWS* (modified Wingfield scale)

Patient no.	Age (years)/Sex	Anomaly	MWS
1	15.7/M	high	1
2	1.8/M	high (+sacral)	2
3	1.4/M	high	1
4	5.8/M	high	3
5	3.1/F	high	3
6	2.4/M	high	3
7	13.6/F	intermediate	3
8	5.7/F	intermediate	3
9	2.9/F	intermediate (+sacral)	3
10	1.0/F	intermediate	1
11	1.8/F	intermediate	3
12	1.3/F	intermediate	3
13	8.2/F	low (+sacral)	3
14	2.0/F	low	2
15	2.5/F	low	3

Table 5. Manometric assessment of internal anal sphincter (IAS) function

Patient no.	Rectal score	IAS activity score	IAS length score	Total IAS score	Overall manometric score
1	1	1	2	3	4
2	4	1	1	2	6
3	1	1	2	3	4
4	4	4	4	8	12
5	4	5	3	8	12
6	4	5	4	9	13
7	5	4	3	7	12
8	6	5	4	9	15
9	6	4	3	7	13
10	4	2	0	2	6
11	2	5	4	9	11
12	5	5	4	9	14
13	4	4	2	6	10
14	1	2	1	3	4
15	2	2	3	5	7

Table 6. MRI assessment of sphincter anatomy

Patient no.	Endoanal MRI sphincter score	Megarectum score	Overall MRI score
1	2	0	2
2	2	0	2
3	2	1	3
4	1	1	2
5	2	1	3
6	3	1	4
7	1	1	2
8	3	0	3
9	3	1	4
10	2	0	2
11	3	1	4
12	2	0	2
13	3	0	3
14	2	0	2
15	1	0	1

Table 7. Correlation between manometric and MRI scores and modified Wingfield score

Parameter	Spearman correlation coefficient	Significance
Rectal score	0.38	0.08
Total IAS score	0.56	0.02[a]
Overall manometric score	0.55	0.02[a]
Endoanal MRI sphincter score	0.20	0.24
Megarectum score	0.44	0.05[a]
Overall MRI score	0.39	0.08
Combined manometric and MRI score	0.58	0.01[a]

[a] $P \leq 0.05$ considered significant

on ARM and MRI and the 3rd patient scored relatively well, but all have clinical and urodynamic evidence of bladder dysfunction. One child with relatively good function (MWS 1) stands out with poor manometry and moderately good results from imaging, but had a low anomaly without evidence of neuropathy.

The overall ARM correlation with MWS (r = 0.55, P = 0.02) (Fig. 1) is largely due to the relatively good performance of the IAS assessment on manometry (r = 0.56, P = 0.02), whereas assessment of the rectum on manometry did not correlate with function (r = 0.38, P = 0.08). The correlation of assessment of the sphincter complex on endoanal MRI with MWS was disappointing (r = 0.20, P = 0. 24), although all components of the sphincters were demonstrated, showing that all repairs were anatomically aligned but with differing patterns of scarring due to surgery [11]. The presence of a megarectum on MRI did correlate with MWS (r = 0.44, P = 0.05), producing overall improvement of conventional plus endoanal MRI (r = 0.39, P = 0.08) (Fig. 2). Overall manometric and MRI results produced a better correlation with MWS (r = 0.58, P = 0.01) (Fig. 3). Figures 4 and 5 illustrate some features of repaired ARAs on endoanal and conventional MRI.

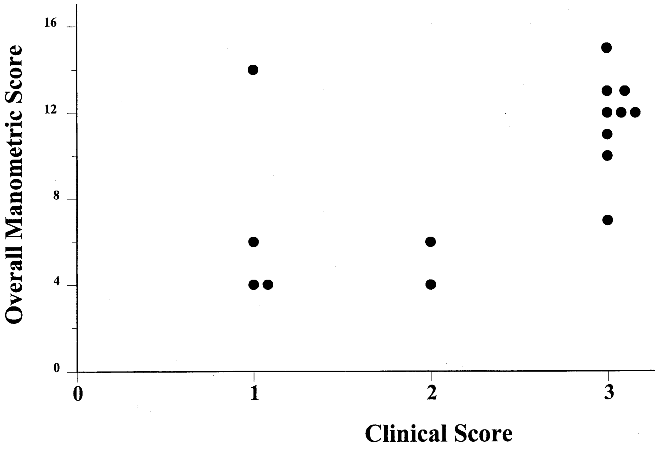

Fig. 1. Correlation between manometric assessment and clinical assessment (MWS)

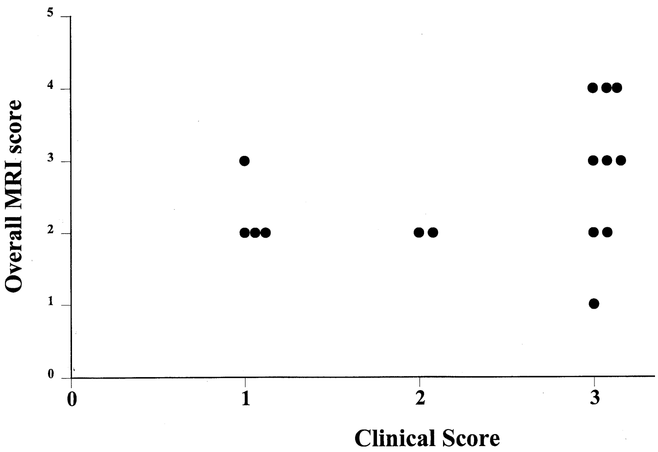

Fig. 2. Relationship between overall MRI assessment and clinical assessment (MWS)

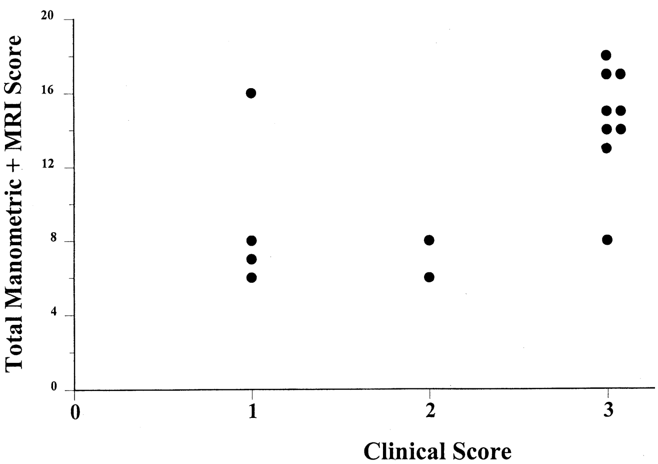

Fig. 3. Correlation between combined manometric and MRI scores with modified wingfield score

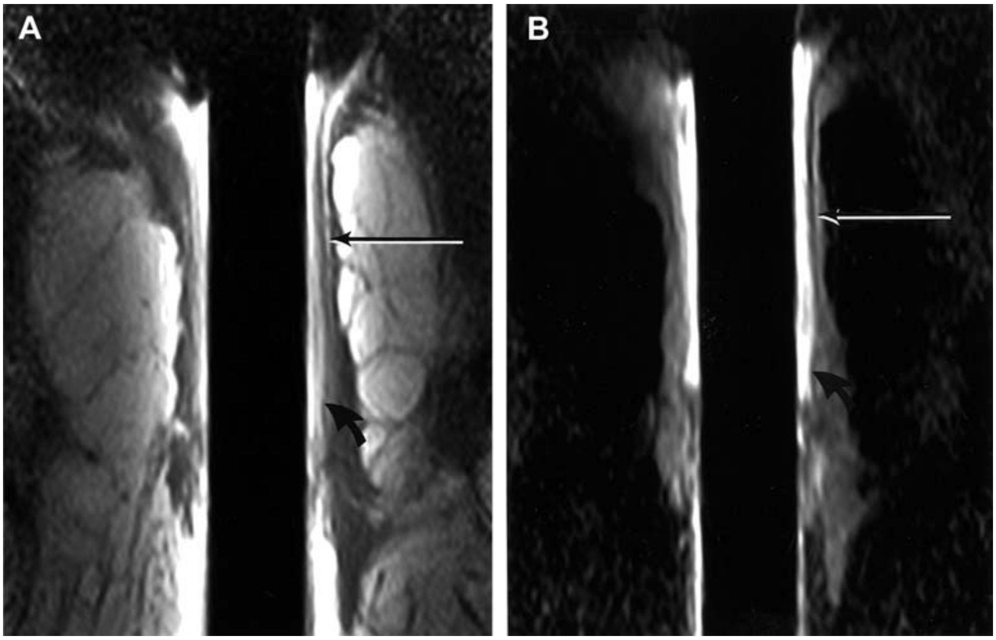

Fig. 4A, B. Endoanal MRI of patient with intermediate anomaly. **A** Coronal T1 image. **B** Short inversion time inversion recovery at same level. External sphincter is hypoplastic (*straight arrows*). Internal sphincter is somewhat short and hypoplastic (*curved arrows*)

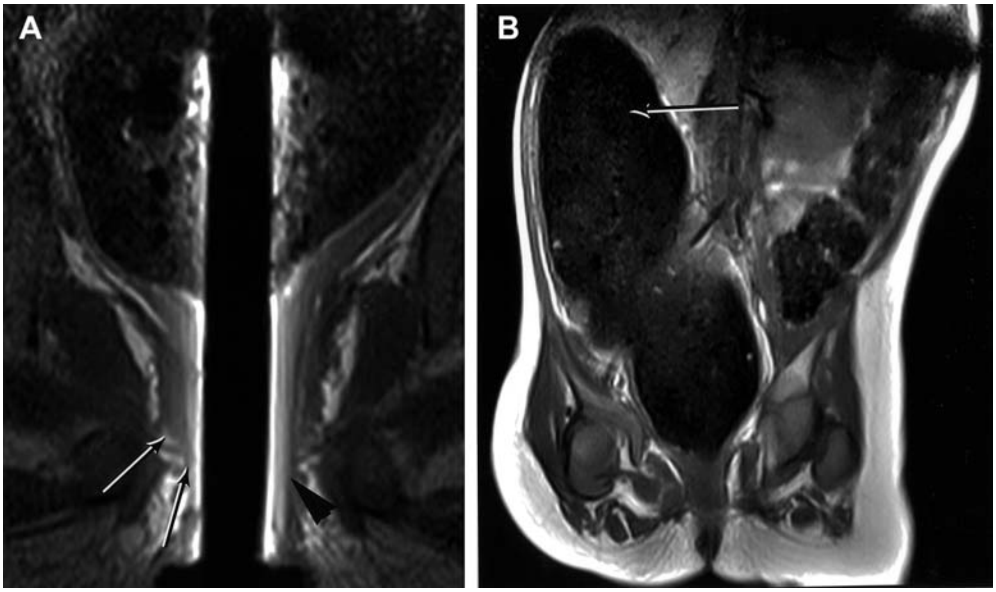

Fig. 5A, B. Endoanal and conventional MRI. **A** Coronal T1 image. **B** T1 image of whole abdomen: focal deficiency of external sphincter on right, but smooth muscle extends to inferior margin of complex. Gross megarectum is shown

Discussion

This study illustrates the utility of a combined physiological and anatomical assessment of the results of surgery for ARAs as evidenced by the moderately good correlation between the overall ARM and MRI assessment and MWS. There are many details within the study that reduce it's power: the numbers are small, while the age range and variations in severity of anomaly are wide. Scoring systems for analysing the results of ARM and MRI were drawn up by the authors empirically and require re-evaluation. For instance, assessment of a megarectum on manometry usually involves measurement of the critical volume at which inhibition of the IAS is complete. In the context of a repaired ARA, the sphincter may be dysfunctional or absent, and so the scoring in this study depended on a short, subjective assessment of peristalsis in unprepared bowel.

The quality of anatomical repairs in this study was good in that the gross anatomy had been correctly established in all cases, possibly diminishing the value of endoanal MRI. In addition, we did not do an evaluation of the impact of scarring due to surgery on outcome, and weighting for features of neuropathy may improve the value of evaluation on MRI [9, 11]. However, this study confirms that the integrity of the IAS is of major importance in the prognosis of repaired ARAs [12, 13]. It appeared to be assessed better by manometry than by imaging in this study. The correlation between the presence of a megarectum on MRI and clinical score is supported by experience from resection of the megarectum, which may produce dramatic clinical improvement [14].

Developments in the techniques of manometry, endoanal MRI, and endosonography need to be evaluated in the context of repaired ARAs. These include three-dimensional sonographic imaging of the sphincter in combination with manometry and improved imaging with endoanal MRI with new parameters for scoring the images, particularly features that indicate the presence of a neuropathy [15, 16].

The application of these studies is to identify the causes of dysfunction and to remedy them. An antegrade continence enema (ACE) stoma will be applicable to those patients with poor anorectal function, especially when combined with a neuropathy [17]. Reduction of a megarectum can be beneficial as an addition to an ACE stoma in the presence of poor sphincter function, and may result in more normal function in the presence of a good sphincter without a neuropathy [14]. Achalasia of the IAS can be a feature of a repaired ARA [1]. Anal dilatation may help this situation, and perhaps a trial of botulinum toxin is indicated in this context [18]. When sphincter pressure is very poor, an anal tampon or artificial sphincter may be applicable [19].

Looking to the future, artificial means of continence will become more sophisticated and paediatric surgeons will be expected to apply them ever earlier. The means to assess the potential for continence in infancy and the preschool years is becoming a necessity so that the appropriate measures can be implemented at an early stage.

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