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Anorectal malformations with good prognosis: Variables affecting the functional outcome

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ABSTRACT

Backgroud/Purpose: The purpose of this study was to investigate the outcome of patients operated for anorectal malformations (ARMs) with good prognosis.
Methods: Thirty patients underwent clinical evaluation by Rintala score and anorectal manometry recording anal resting pressure (ARP), rectoanal inhibitory reflex (RAIR), and rectal volume (RV). The results were analysed with regard to sex, type of ARM, surgical timing of posterior sagittal anorectoplasty (PSARP), neurospinal cord dysraphism (ND), neonatal colostomy, and institution where they underwent surgery.
Results: 6/30 (20%) presented ND despite normal sacrum. 17/30 (57%) patients had a normal Rintala score. ND and neonatal colostomy were significantly associated with a pathologic score ($p = 0.0029$ and $p = 0.0016$). Patients with ND had significantly lower ARP compared to patients with normal spine (23.5 ± 7.2 mmHg vs 32 ± 7.9 mmHg, $p = 0.023$). ARP was significantly lower in patients with neonatal colostomy compared to patients with primary repair (25.22 ± 10.24 mmHg vs 32.57 ± 6.68 mmHg, $p = 0.026$). RAIR was present in only 2/6 (33%) patients with ND, while in 21/24 (87.5%) without ND ($p = 0.015$) and in 4/9 (44%) patients with neonatal colostomy, while in 19/21 (90.5%) patients submitted to primary repair ($p = 0.014$).
Conclusions: Neurospinal cord dysraphism may be present despite normal sacral ratio. From a clinical point of view, patients with good prognosis ARMs are not completely comparable to healthy children. Neurospinal cord dysraphism and neonatal colostomy seem to worsen the clinical and manometric (ARP and RAIR) outcomes of these patients.

Anorectal malformations (ARMs) are a spectrum of defects ranging from forms that are easier to treat to those that are more complex and frequently associated with other anomalies. In the past it was believed that those traditionally classified as "low" ARMs enjoyed good prognosis but a more complex situation has recently emerged [1]. Thanks to the contribution of Peña and Levitt [2,3], we have come to understand that there are other factors, beyond the level of the rectum, which may influence the prognosis of these patients: the anatomy of the sacrum and the perineal muscle. In other words a "low" ARM does not warrant in itself an excellent functional outcome. The aim of this study was to investigate the outcome of a homogeneous group of patients after corrective surgery for ARMs with good prognosis (which are defined as some types of ARM with prominent midline groove and normal sacrum) through a clinical score and anorectal manometry.

1. Material and methods

Among over 200 ARM patients currently followed-up at the Colorectal Center of our Institution, 84 were affected by forms with good prognosis. Based on Peña's and Levitt's experience [3], we defined as ARMs with good prognosis some anatomical types of ARMs (recto-perineal fistula, recto-vestibular fistula, imperforate anus without fistula, rectal atresia and cloaca with common channel < 3 cm) all with prominent midline groove (good perineal muscle) and normal sacrum. Patients with major sacral malformations (detected through neonatal plain X-rays with anterior–posterior and lateral–lateral sacral ratio) and poor perineal muscles were excluded from this study. Patients who were lost to follow-up and those who were not yet completely toilet-trained were also excluded. The present study included thirty patients (19 females, 11 males) who underwent surgery for ARM and who had completed the toilet training program. Between May and July 2013 they were investigated through a clinical score and anorectal manometry. ARM patients were classified as follows: recto-perineal fistula ($n = 19$), recto-vestibular fistula ($n = 10$) and imperforate anus ($n = 1$) on the basis of operative findings. The age range of patients was from 2.5 years to

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10 years. Mean follow-up at the time of clinical and manometric evaluation was 5 years. Nine out of 30 patients (3 recto-perineal fistula, 5 recto-vestibular fistula and 1 imperforate anus) were treated with a three-stage surgical procedure which included neonatal diverting colostomy (performed between 1 and 6 days of life), posterior sagittal anorectoplasty (PSARP) (between 2 and 11 months of life) and colostomy closure (between 5 and 15 months of life), while in the remaining 21 cases (16 recto-perineal fistula and 5 recto-vestibular fistula) children underwent single-stage repair (PSARP) between the first day of life and 13 months of life (mean time 115 days). Of these 21 patients 6 were newborn, 10 between 1 and 6 months, 4 between 6 and 12 months and 1 was older than 12 months at the time of surgery. Six patients out of 30 underwent PSARP within the first 7 days of life (early surgery). Twenty-one out of 30 patients were operated on at our institution by the two most experienced senior surgeons, while nine out of 30 underwent surgery in other institutions (8 in Italian departments of pediatric surgery and one in a Chinese institution) and then they came to our Colorectal Center to continue follow-up. No significant surgical complication (rectal prolapse, dehiscence and/or mislocation of the neoanus) was registered and no patient required a REDO-procedure. All these patients were studied to detect neurospinal anomalies (such as tethered cord or myelodysplasia) through early lumbosacral ustrasonographic scan (US) within the first 3 months of life and/or magnetic resonance imaging (MRI) [4]. Tethered cord was found in 6 out of 30 patients (20%) despite a normal sacral ratio (normal: anterior–posterior film >0.74, lateral position > 0.77) (Table 1).

1.1. Clinical outcome assessment

Bowel function was assessed by Rintala score [5,6]: results were classified as normal (score ≥ 18), good (12–17), fair (7–11) and poor (score ≤ 6) considering the ability to hold back defecation, to feel the urge to defecate, frequency of defecation, soiling, accidents, constipation and social problems. Only patients with a score of at least 18 were considered as normal and completely comparable to healthy children while scores below 18 were classified as pathological.

1.2. Anorectal manometry evaluation

Anorectal manometry was performed using a water perfused 4-channel anorectal motility catheter (4.5 mm outer diameter) with 4 side openings placed spirally and the catheter tip was at 4.5 cm from the first side hole. All side holes were perfused with saline and pressure was measured by pressure transducers connected to a PC. The measured signals were converted in pressure tracings. The calibration of the machine was performed before each evaluation. The examination was performed without sedative and with the patient in the supine position. Bowel preparation was done through an enema performed the evening before the day of the study. The lubrificated catheter was gently introduced up to about 10 cm until a decrease in the pressure was registered, as this meant that the catheter was inside the rectum. The catheter was then gently retracted from the rectum to the anal canal. The following parameters were evaluated during the examination:

- anal resting pressure (ARP): measured as the mean pressure that was registered during the passage of the catheter through the anal canal;
- rectoanal inhibitory reflex (RAIR) elicited when the catheter was inside the anal canal with a rapid inflation of the rectal balloon with an air volume ranging from 10 to 50 ml considering the age of the patient. The reflex was considered as present when the anal pressure presented a drop of at least 5 mmHg;
- rectal volume (RV) measured in the rectum by inflating the balloon with air and asking the patient to communicate the feeling of needing defecation

The anorectal manometry parameters in healthy adults are known but they are not applicable in children. Normal values in children, even after a literature review, are very difficult to find. We decided to compare our results with age-matched controls published by Kumar et al. [7]. Specifically, these authors reported that healthy children over 18 months of age have an anal resting pressure ranging between 30 and 60 mmHg. They also set that a positive rectoanal inhibitory reflex could be demonstrated in all healthy children but the volume of air needed to evocate that reflex varied with age: in children over 18 months the reflex is elicited between 10 and 50 cc of air injection in the rectal balloon. Regarding the rectal volume we referred to the article published by Hedlund and Peña in 1992 [8] that considered normal rectal values when lower than 150 cc of air injection in the rectal balloon.

The clinical and manometric results were analyzed with regard to sex, type of ARMs, surgical timing of PSARP (early surgery if anoplasty was done within the first 7 days of life or late surgery if it was done later), presence of neurospinal cord dysraphism (ND), neonatal colostomy (for patients treated in 3-stages) and the fact of being operated in another institution. A specific consent of the parents and the children (when applicable) was obtained before every procedure.

1.3. Statistical analysis

T-test was used to compare quantitative variables. Qualitative variables were compared using Fisher's test. A p-value under 0.05 was taken as level of significance.

2. Results

2.1. Clinical results

Seventeen out of 30 (57%) patients who underwent clinical evaluation had a normal Rintala score while 13 patients (43%) had a pathological score (scores ranging from 12 to 17). No one had fair or poor score. Significant correlations were noted between a normal score and lack of ND ($p = 0.0029$) and between a normal score and

Table 1
Clinical features of patient with Neurospinal dysraphism (ND: neurospinal dysraphism, ARM: anorectal malformation, RV: rectovestibular, IA: imperforate anus, ARP: anal resting pressure, RAIR: rectoanal inhibitory reflex, RV: rectal volume).

Type of ND	Sex	Type of ARM	Surgical timing	Colostomy	Clinical Score	ARP	RAIR	RV	Neurological symptoms
Tethered cord	F	RV	Late	Yes	Pathologic	35	Absent	160	None
Tethered cord	F	RV	Late	No	Pathologic	27	Present	60	None
Tethered cord	F	RV	Late	Yes	Pathologic	26	Present	75	None
Tethered cord	F	RV	Late	Yes	Pathologic	15	Absent	200	None
Tethered cord	F	RV	Late	Yes	Pathologic	20	Present	100	None
Tethered cord	M	IA	Late	Yes	Pathologic	18	Absent	360	None