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Long-Term Functional Outcomes of an Anorectal Malformation French National Cohort

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ABSTRACT

Objectives: The present study aimed to assess long-term functional outcomes of children with anorectal malformations (ARMs) across a network of expert centers in France.

Methods: Retrospective cross-sectional study of patients ages 6–30 years that had been surgically treated for ARM. Patient and ARM characteristics (eg, level, surgical approach) and functional outcomes were assessed in the different age groups.

Results: Among 367 patients, there were 155 females (42.2%) and 212 males (57.8%), 188 (51.2%) cases with, and 179 (48.8%) higher forms without, perineal fistula. Univariate and multivariate statistical analyses with logistic regression showed correlation between the level of the rectal blind pouch and voluntary bowel movements (odds ratio [OR]=1.84 [1.31–2.57], $P < 0.001$), or soiling (OR = 1.72 [1.31–2.25], $P < 0.001$), which was also associated with the inability to discriminate between stool and gas (OR = 2.45 [1.28–4.67], $P = 0.007$) and the presence of constipation (OR = 2.97 [1.74–5.08], $P < 0.001$). Risk factors for constipation were sacral abnormalities [OR = 2.26 [1.23–4.25], $P = 0.01$) and surgical procedures without an abdominal approach (OR = 2.98 [1.29–6.87], $P = 0.01$). Only the holding of voluntary bowel movements and soiling rates improved with age.

Conclusion: This cross-sectional study confirms a strong association between anatomical status and functional outcomes in patients surgically treated for ARM. It specifically highlights the need for long-term follow-up of all patients to help them with supportive care.

Key Words: age group assessment, clinical functioning, social integration

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What Is Known

- Several studies had already evaluated functional results after surgical treatment of anorectal malformation in children, which mainly depend on the level of the congenital defect, the surgical management, and the follow-up quality.

What Is New

- We assessed the functional status of a nationwide cohort of patients, categorized by age groups. We showed that functional impairments were strongly correlated to the level of the rectal blind pouch and that the holding of voluntary bowel movements and soiling rates improved as age increased.

Despite careful management of children with anorectal malformation (ARM) from birth to adulthood, including surgical procedures in line with the latest international developments and a scheduled multidisciplinary follow-up, some patients still have a functional impairment which seriously impacts their quality of life.

In 2007, the French national rare disease program (*Plan National Maladies Rares*) selected ARM among many other rare conditions as a specific disease to be included in the program, which

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University Hospital, Brest, the ***Pediatric Surgery Department, University Hospital, Bicêtre, the †††Pediatric Surgery Department, General Hospital, St Brieuc, the †††Pediatric Surgery Department, University Hospital, St Denis – La Réunion, the §§§Pediatric Surgery Department, University Hospital, Montpellier, the |||||Pediatric Surgery Department, University Hospital, Tours, the ¶¶¶Pediatric Surgery Department, General Hospital, Colmar, the ###Pediatric Surgery Department, University Hospital, the ****Pediatric Surgery Department, University Hospital, Fort de France – Martinique, the ††††Pediatric Surgery Department, General Hospital, Le Mans, the ††††Pediatric Surgery Department, General Hospital, Orléans, the §§§§Pediatric Surgery Department, University Hospital, Toulouse, the |||||Pediatric Surgery Department, University Hospital, Reims, and the ¶¶¶¶Pediatric Surgery Department, Necker-Enfants Malades, University Hospital, Paris, France.

involved different tasks including keeping records of patients' medical condition via a national database, the establishment of competence centers in France, and the evaluation and improvement of quality of life for patients and their families.

In this context, we conducted the MARQOL (Quality of Life of Patients with Anorectal Malformation) national study. Our aim was to gather and analyze anatomical data from a national database, functional data from medical visits provided by the competence centers belonging to the national network, and data from specific and generic quality of life questionnaires completed by patients and/or their parents. Our goal was to describe, in as much detail as possible, the conditions of French patients with ARM, in order to select those who would specifically require specialist help from our network.

In this study, we described the functional results of our cohort and their relationship to the anatomical status and age of the patient, through correlation, uni- and multivariate analyses. A second study will focus on the association between those results and the patient's quality of life.

METHODS

Subjects

Patients ages between 6 and 30 years, who had been surgically treated in France for ARM, were put forward for inclusion in the MARQOL national multicenter observational cross-sectional study. Patients' informed consent was obtained and a functional evaluation was carried out during a routine visit or by telephone by the pediatric surgeon who ensured follow-up in the competence center. Data collection, which included anatomical, functional and socio-economic items, started in April 2012, was completed in April 2015, and was registered in the CEMARA (*Centre des Maladies Rares*) national database. Patients were categorized into four age groups: 6–7 years, 8–12 years, 12–17 years, and >17 years.

Ethics

The research protocol has been validated by an ethical committee (*Groupement Nantais d'Ethique dans le Domaine de la Santé*; no. 2011-06-01). The CEMARA database has been approved by the French Data Protection Authority (no. DR_2011-377).

Outcome Measures

The primary outcome measure was the functional status measured by the Krickenbeck criteria (1). Soiling was categorized into 3 grades: occasional soiling (grade 1) and constant soiling which did (grade 3), or did not (grade 2), result in social problems. Similarly, constipation was categorized into constipation that is manageable with diet (grade 1), constipation that requires laxatives (grade 2) or constipation that is resistant to laxatives and diet (grade 3). Finally, voluntary bowel movements were assessed using three questions: whether the patient could hold stools, could feel the urge

to defecate and/or could verbalize this feeling. The patient was then scored from 0 (they answered yes to all three questions) to 3 (they answered no to all three questions).

For other clinical and anatomical items recorded, see the CEMARA data set in Supplemental Digital Content 1, <http://links.lww.com/MPG/C763>. Note: the rectal blind pouch was assessed on the colostogram (2), according to levator ani and sacral bones.

Statistical Methodology

Statistical analysis was performed using SPSS 15.0 for Windows software. All tests were two-sided and the statistical level for significance was set at $P < 0.05$. Patients' characteristics were described as percentages for qualitative variables and as means \pm standard deviation for continuous variables. Comparative analysis between two groups of patients was performed using the Student *t*-test for quantitative variables according to the normal distribution assumption and using Fisher exact test for qualitative variables. Multiple groups' comparisons were made using the analysis of variance (ANOVA) for quantitative data and a Pearson chi square test (2) for qualitative variables.

Correlation analyses between the different graded sub-scores of the Krickenbeck scale, namely "voluntary bowel movements," "soiling," and "constipation," and the multinomial qualitative variables (age groups, type of fistula and localization of the rectal blind pouch) were then performed using the non-parametric Spearman test.

Finally, prognostic factors associated with gas/stool discrimination, voluntary bowel movements, soiling and constipation were determined using logistic regression. First, univariate analysis was used to select relevant variables for each of the above-mentioned items; hence, variables with a *P* value $< 20\%$ or already known to interact or to be a potential confounding factor entered in the final model. Multivariate analysis was performed using a backward stepwise likelihood-ratio test and data were expressed as an odds ratio (OR) with 95% confidence intervals (for all results concerning multivariate analysis, see Tables, Supplemental Digital Content 2, <http://links.lww.com/MPG/C764>).

RESULTS

Description of the Population

Figure 1 shows the flow chart. Our cohort dropped from 762 to 367 patients for several reasons, including 99 patients who were excluded because their functional status was not assessed during a medical visit (91 patients) or because they had a stoma (8 patients).

The mean age of the study population was 13.4 ± 6.2 years old [6.0–34.6]. There were 155 females (42.2%) and 212 males (57.8%). Our sex ratios were stable for age groups, except for boys who were less frequent in the 12- to 16-year-old group (female/male: 61/38).

Types of fistula are listed in Figure 2. "Other fistulae" included anal stenosis (21), vaginal fistula (4), rectal atresia (1) and H fistula (1). Sixty-three (17.2%) patients had associated

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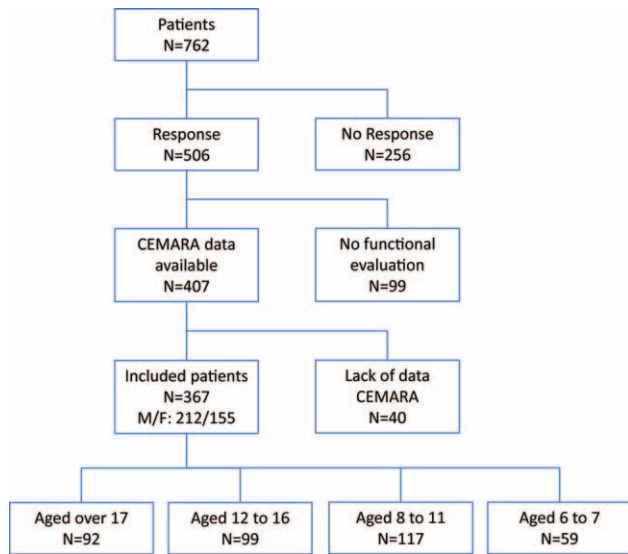


FIGURE 1. Flow chart showing a decrease in our cohort from 762 to 367 patients, including 99 patients excluded because their functional status was not assessed.

malformations (VACTER (3) 29/Currarino 6/caudal regression 5/ other 23) and 79 (23%) had sacral malformations. Surgical correction was performed by fistula transposition or proctoplasty in 151 (41.4%) cases, through posterior sagittal anorectoplasty (PSARP) in 160 (43.6%) cases, or through an open or laparoscopic abdominoperineal approach in 53 (14.4%) cases (3 not recorded).

Global Analysis of Functional Outcomes

On average, patients had 6.1 ± 3.5 stools per week, 76.5% were able to discriminate between stools and gas and 78.6% had voluntary bowel movements. Soiling was present in 46.8% of the patients and was mainly occasional. Conversely, constipation occurred in 40.9% of patients, but the three grades of constipation were found in rather similar proportions. High-volume enemas were required in 21.3% of cases and were more often retrograde enemas (79.5%) than antegrade enemas through a Malone conduit (20.5%).

Concerning social integration, 8% required adapted schooling, 25% had a partial or total dependency, but 69.6% enjoyed extracurricular leisure or sport, of which 123 of 199 were members of a sports club.

Correlation Between Functional Outcomes and Anatomical Status (Fig. 2)

Rates of soiling, constipation, voluntary bowel movement and enemas, according to levels of fistula in males and females, were displayed in Figure 2. The rates of soiling and constipation increased overall with the level of the fistula. The lack of voluntary bowel movement concerned forms other than ARM with perineal fistula in both sexes and vestibular fistula in girls. Enemas were mainly required for recto-vesical fistula cases in males or cloaca cases in females (Fig. 2).

Statistical analyses (see Supplemental Digital Content 2, <http://links.lww.com/MPG/C764> describing multivariate analysis for gas/stool discrimination, voluntary bowel movements, soiling and constipation) also showed that the score of voluntary bowel movements was directly correlated to the type of fistula ($r=0.21$,

$P < 0.001$) and the level of the rectal blind pouch ($r=0.33$, $P < 0.001$). After logistic regression, the presence of voluntary bowel movements was mostly associated with a low level of the rectal blind pouch (OR = 1.84 [1.31–2.57], $P < 0.001$). Stool/gas discrimination was impaired by the presence of a sacral abnormality (OR = 3.24 [1.69–6.18], $P < 0.001$).

The grade of soiling was correlated with the type of fistula ($r=0.17$, $P=0.002$) and the level of the rectal blind pouch ($r=0.28$, $P < 0.001$), and the latter was the only anatomical factor that appeared to impact soiling after logistic regression (OR = 1.72 [1.31–2.25], $P < 0.001$). The other factors statistically associated with soiling were the absence of stool/gas discrimination (OR = 2.45 [1.28–4.67], $P=0.007$) and the presence of constipation (OR = 2.97 [1.74–5.08], $P < 0.001$).

The severity of constipation was correlated to the level of the rectal blind pouch ($r=0.15$, $P=0.006$). After logistic regression, risk factors for constipation were the presence of a sacral abnormality [OR = 2.26 [1.23–4.25], $P=0.01$] and an exclusively perineal surgical approach (ie, without laparotomy or laparoscopy) (OR = 2.98 [1.29–6.87], $P=0.01$).

Functional Outcomes in Different age Groups (Table 1)

Gas/stool discrimination, feeling of urge and capacity to verbalize bowel movements were quite stable between the different groups of age. Yet, the score for the holding of voluntary bowel movements appeared to be correlated with the patients' age ($r=0.10$, $P=0.04$). Only grade 2 soiling progressively decreased from 12.1% of the 6- to 7-year-old children to 2.3% of adults ($P=0.01$). Constipation more often affected older children and teenagers (47.2% of the 8- to 16-year-old patients) than young children and adults (20.2% at 6–7 and >17 years old; $P=0.005$).

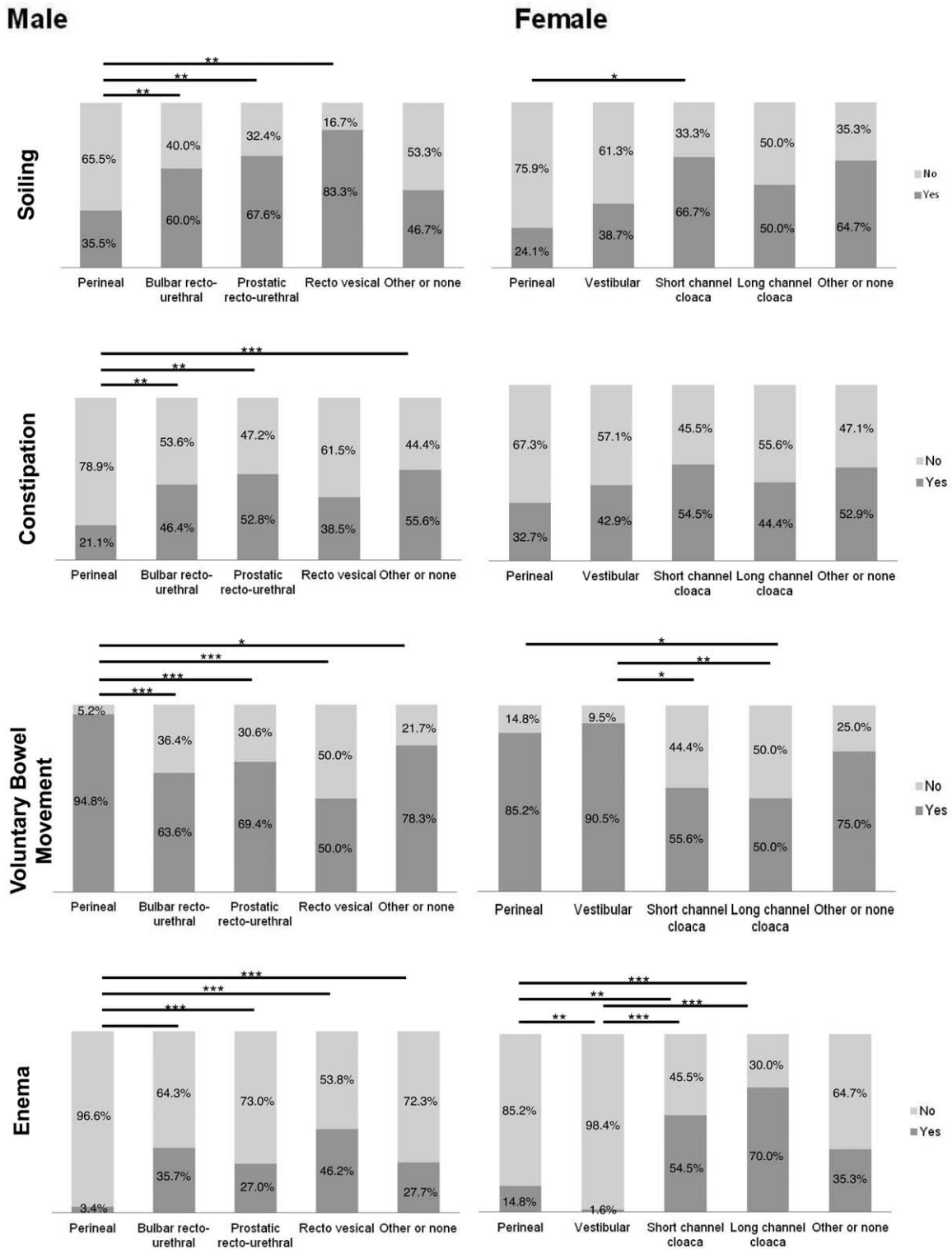
The use of enemas did not significantly differ in different age groups, but the use of antegrade enemas through a Malone conduit almost exclusively concerned adolescents and adults (32.5% vs 7.7% before 12 years old; $P=0.003$).

DISCUSSION

Thanks to the French national rare disease program, the national CEMARA database provided us with a tool for standardized assessment of the anatomical status of our patients which were treated for anorectal malformation. In order to correlate anatomical abnormalities and functional status, we consensually chose to apply the Krickbeck score because it has been chosen by an international consortium (1), offers clear definitions of anatomical groups which we used in our CEMARA database and has already been used by many other teams (4–9), despite the fact that it is descriptive and not validated.

The first limitation of our study was its multicenter status with heterogeneity in the size of the 37 centers and its cross-sectional nature. Second was the loss of inclusion with >250 patients or parents who ultimately did not complete the quality of life (QOL) questionnaire (for the second part of the study), totaling a hundred patients with missing functional data. Furthermore, the stratification in age group came from the design of the QOL questionnaires, and was not a cohort follow-up, but a national picture taken at one t-time. Finally, one could argue in regard to data collection that the patients/parents may not have given a reliable picture of the functional outcome to the operating surgeon who was in charge of the follow-up of the patient throughout their childhood.

As reported in other publications (10), we first confirmed that the higher the level of the fistula or the rectal blind pouch, the worse the functional results, demonstrated here with statistical analysis



* p<0.05; ** p<0.01; *** p<0.001.

FIGURE 2. Level of fistula in males and females, with rate of soiling, constipation, voluntary bowel movement and enemas. "Other or none" includes anal stenosis, vaginal fistula, rectal atresia and H fistula.

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TABLE 1. Comparison of functional outcomes for different age groups of patients with anorectal malformation

Age (y)	6–7		8–11		12–16		≥17		Total		Missing data		P value*
	n	%	n	%	n	%	n	%	n	%	n	%	
No. of patients	59/367	16.1	117/367	31.9	99/367	26.9	92/367	25.1	367	100	0	0	
No. males	33/59	55.9	69/117	59.0	38/99	38.4	49/92	53.3	212/367	57.8	0	0	0.7
No. females	26/59	44.1	48/117	41.0	61/99	61.6	43/92	46.7	155/367	42.2	0	0	
Gas/stool discrimination	37/53	69.8	82/106	77.4	68/86	79.1	67/87	77.0	254/332	76.5	35	9.5	0.6
Voluntary bowel movement	44/58	75.9	90/117	76.9	77/95	81.1	71/89	79.8	282/359	78.6	8	2.2	0.8
Feeling of urge	30/59	50.8	71/117	60.7	57/99	57.6	62/92	67.4	220/267	59.9	0	0	0.2
Capacity to verbalize	27/59	45.8	67/117	57.3	49/99	49.5	58/92	63.0	201/337	54.8	0	0	0.1
Hold the bowel movement	24/59	40.7	61/117	52.1	51/99	51.5	56/92	60.9	192/367	52.3	0	0	0.1
Soiling	28/59	47.5	56/113	49.6	51/96	53.1	33/91	36.3	168/359	46.8	8	2.2	0.1
Grade 1, occasionally (once or twice per week)	17/58	29.3	38/112	33.9	34/94	36.2	22/88	25.0	111/352	31.5			
Grade 2, every day, no social problem	7/58	12.1	12/112	10.7	6/94	6.4	2/88	2.3	27/352	7.7	7	1.9	0.1
Grade 3, constant, social problem	3/58	5.2	5/112	4.5	9/94	9.6	6/88	6.8	23/352	6.5			
Constipation	20/59	33.9	53/117	45.3	49/99	49.5	28/92	30.4	150/367	40.9	0	0	0.03
Grade 1, manageable by changes in diet	5/59	8.5	10/117	8.5	18/99	18.2	8/92	8.7	41/367	11.2			
Grade 2, requires laxatives	7/59	11.9	21/117	17.9	11/99	11.1	7/92	7.6	46/367	12.5	0	0	0.1
Grade 3, resistant to diet and laxatives	6/59	10.2	18/117	15.4	15/99	15.2	12/92	13.0	51/367	13.9			
Enema	11/59	18.6	28/117	23.9	24/99 [†]	24.2	15/92	16.3	78/367 [†]	21.3	0	0	0.4
Retrograde enema	11/59	18.6	25/117	21.4	20/99	20.2	7/92	7.6	63/367	17.2	0	0	0.04
Anterograde enema (Malone)	0	0	3/117	2.6	5/99	5.1	8/92	8.7	16/367	4.4	0	0	0.04 [‡]
Autonomy	25/49	51.0	75/109	68.8	72/93	77.4	83/89	93.3	255/340	75.0			<0.001
Partial dependency	20/49	40.8	31/109	28.4	18/93	19.4	6/89	6.7	75/340	22.1	27	7.4	<0.001
Total dependency	4/49	8.2	3/109	2.8	3/93	3.2	0/89	0	10/340	2.9			0.06

*Statistical tests compared age groups, and grade distribution for soiling, constipation and autonomy.

[†]One patient uses both retrograde and anterograde enema.

[‡]This result is not valid as some cells have expected count <5.

through logistic regression. Secondly, the constipation was not statistically correlated with ARMs with perineal fistula. Patients with severe forms without bowel control also often suffer from constipation and must be treated (8), which helps for the management of their incontinence (11). We have shown that constipation was the only non-anatomical statistical factor involved in triggering soiling.

Since the first publication by Peña and Devries (12), the PSARP has been popularized in France, and most of our patients also benefited from other rules promoted by Alberto Peña, such as the performance of a colostomy for all cases without a perineal fistula, or the implementation of bowel management for patients with incontinence. As a result, our functional data were very similar to those previously published by Peña and Levitt (8), despite the multicentric character of our work compared to that expert center. Conversely, our functional results showed some improvement in comparison to historical data from before the PSARP period. We have pooled the results published in two available studies (13,14) of adult patients with severe ARMs. Our results showed a slight improvement when comparing their series for true incontinence rate (12/84 (14.3%) vs ours 4/45 (8.9%); NS) and the use of bowel enemas (28/84 (33.3%) vs 8/32 (25%); NS). This may mean that patients with high-level anorectal malformations and impairment of their pelvic innervation would not be the group which benefited most from surgical improvements; however, bowel management strategies provided to such patients became less invasive and more efficient, with a lower rate of stoma in our more recent series (9/91 (9.9%) vs 2/57 (3.5%) (data not shown); $P = 0.015$) and a significantly better rate of patients without any soiling (13/84 (15.5%) vs 22/47 (46.8%); $P < 0.001$).

Concerning enemas, the antegrade or retrograde route clearly depended on patient age, since the use of an antegrade enema

through a Malone conduit almost exclusively concerned adolescents and adults. The first explanation for this age distribution is probably the complications of a Malone procedure, which are now well-known by our teams and in literature, with 44% reporting stenosis and 15% ceasing use for Tiryaki et al (15), 30% of whose patients complained about the duration of the enema being longer than an hour. We also know that some patients experienced the return of spontaneous and regular bowel movements during childhood or adolescence, responsible for 17.5% of reversed Malone for Peeraully et al (16). Over the last two decades, we have therefore proposed the retrograde approach, started by parents when the child is between 4 and 6 years of age, and then taught to the child when they are older. This approach has been shown to be well tolerated with a high adherence rate (17) and is efficient in treating either incontinence or constipation (5,18). Then, as done by others (19), we may discuss with the teenager and his/her parents the possibility of a Malone procedure for definitive incontinence cases, mainly in forms with a recto-vesical fistula or long channel cloaca. We confirmed the assessment that bowel management must be associated with pragmatic, personalized and repeated training to be efficient, particularly to prevent persistent soiling (15,20).

The results we have provided in different age groups allowed us to highlight some specific features. The holding of voluntary bowel movements improved with patient age and the soiling rate decreased from childhood into adulthood, features we would have to compare with the natural evolution among the general pediatric population (21). We have no specific explanation for our higher rate of constipation during adolescence, unless our lack of follow-up in male teenagers, who were doing well, would have artificially changed the denominator. We were pleased to see the gain in autonomy of our patients between early childhood and adulthood (data not shown). It has already been shown that this resilience is

expressed in adult patients through a higher level of education and a preference for office-based work, as compared to the general population (22); however, we have still to explore in a further publication the impact of such functional results on quality of life in all its dimensions for our patients from different age groups and their parents.

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