

Long-term evaluation of fecal continence and quality of life in patients operated for anorectal malformations

ANA CRISTINA AOUN TANNURI¹, MARIANA APARECIDA ELISEI FERREIRA², ARTHUR LOGUETTI MATHIAS³, UENIS TANNURI^{4*}

¹Associate Professor, Faculdade de Medicina, Universidade de São Paulo (FMUSP). Physician, Service of Pediatric Surgery and Liver Transplantation, Instituto da Criança, Hospital das Clínicas, FMUSP, São Paulo, SP, Brazil

²Medical Student, FMUSP São Paulo, SP, Brazil

³Physician, Service of Pediatric Surgery and Liver Transplantation, Instituto da Criança, Hospital das Clínicas, FMUSP São Paulo, SP, Brazil

⁴Full Professor, Department of Pediatric Surgery and Liver Transplantation, FMUSP. Head of the Service of Pediatric Surgery and Liver Transplantation, Instituto da Criança, Hospital das Clínicas, FMUSP, São Paulo, SP, Brazil

SUMMARY

Introduction: Patients operated for correction of anorectal malformations (ARM) can develop fecal incontinence, constipation, and soiling, with loss in quality of life.

Objective: To evaluate, through the use of questionnaires, fecal continence, and quality of life of children in the late postoperative follow-up of ARM correction, both high and low. In addition, the levels of fecal continence and quality of life were compared with those of a control group.

Method: A Fecal Continence Index Questionnaire (ICF) and a Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents (QQVCFCA) were administered to 63 patients with ARM, aged from 7 to 19 years, whose surgical treatment had been completed for at least 6 months. The patients were compared to a control group of 59 children.

Results: In the control group, 25 (42.4%) patients had good continence and 34 (57.6%), normal continence. We found that the quality of life in children with ARM is compromised globally, in all areas and in the ICF questionnaire, compared to controls ($p < 0.001$). There was no difference between patients with high and low defects. Thirty-two (50.8%) patients had other associated anomalies.

Conclusion: In patients operated for ARM correction, quality of life and ICF were compromised, and there was no difference between patients with high-type and low-type of the disease. In about half the cases there are other associated malformations.

Keywords: imperforate anus, anorectal malformation, quality of life, fecal incontinence.

Study conducted at Serviço de Cirurgia Pediátrica e Transplante Hepático, Instituto da Criança, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo (HC-FMUSP), São Paulo, SP, Brazil

Article received: 6/29/2016

Accepted for publication: 7/26/2016

*Correspondence:

Address: Av. Dr. Arnaldo, 455,
4º andar, sala 4109
São Paulo, SP – Brazil
Postal code: 01246-903
uenist@usp.br

<http://dx.doi.org/10.1590/1806-9282.62.06.544>

Financial support: Program of Scientific Initiation, Universidade de São Paulo (PIC/USP), modality Santander

INTRODUCTION

Anorectal malformations (ARM) are pathologies that, despite adequate treatment, may progress to fecal incontinence, constipation, and soiling.¹ Fecal incontinence, defined as the inability to control the emission of flatus and/or feces, can become an obstacle in the occupational, social, emotional, sportive, and sexual spheres of a person's life, leading to psychiatric disorders and even loss of independence.² It is closely related to the concept of quality of life (QoL), defined by the World Health Organization as "the individual's perception of their position in life in the context of culture and the system of values in which they live and in relation to their goals, expectations,

standards, and concerns".³ Thus, QoL has become in the last decade an important indicator of medical outcomes, supporting the follow-up of these patients, which justifies the use of questionnaires for evaluation.

Most of the reviews show that patients with impaired bowel function also have impaired QoL, and this correlation varies from small to medium or strong.¹ The studies also show that fecal continence is more affected in children than in teenagers, even though the QoL of adolescents is more compromised than that of children.¹

Therefore, questionnaires that allow an objective evaluation and a longitudinal follow-up of patients have been increasingly used. Many questionnaires relate QoL with

fecal continence, and some are specific to the diseases studied. Examples include the Fecal Incontinence Quality of Life (FIQL),⁴ the Hirschsprung's Disease Anorectal Malformation QoL Questionnaire (HAQL)⁵ and the Clinical Evaluation of Continence (Holschneider Criteria).⁶ These questionnaires have been adapted and validated in several countries such as France, Spain, and Sweden. As for the Portuguese language, our group previously validated the Fecal Continence Index (ICF) questionnaire and the Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents (QQVCFCA).⁷

This work was developed with the purpose of applying the ICF and QQVCFCA questionnaires in patients with ARM attending a tertiary center of pediatric surgery. We will thus be able to know more about the profile of patients and their conditions, and to start using these tools in clinical practice. In short, the objectives of this study were:

- To evaluate continence and quality of life of children in the late postoperative follow-up of ARM correction by applying questionnaires designed specifically for our population and the studied age groups.
- To compare the rates of fecal continence and quality of life of children surgically treated for ARM correction with a group of normal children.
- To compare the quality of life of children with high and intermediate ARM with that of children with low-type malformations.
- To document the malformations associated with ARM.

METHOD

The tools used to assess quality of life and fecal continence were: the ICF questionnaire and the QQVCFCA. These instruments were based on questionnaires already established and have gone through a thorough validation process, described in an early work by the same group. The questionnaires were sent to patients and volunteers by mail, and also applied through telephone interviews and after outpatient visits. The research project was approved by the Research Ethics Committee of the Institution.

Fecal Continence Index (ICF)

The ICF questionnaire was based on the Clinical Evaluation of Fecal Continence (Holschneider Criteria). It consists of eight questions easily understood on procedures of everyday life (questions 1-5) and on diarrhea, constipation, and use of auxiliary treatment (questions 6-8), meeting the Holschneider Criteria. The final score ranges from 0 to 16, with 0 to 2 points given to each question. The 0-5 range indicates poor continence, 6-10 indicates

fair continence, 11-15 indicates good continence, and the maximum score of 16 indicates normal fecal continence (Chart 1).

Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents (QQVCFCA)

The QQVCFCA is based on the translation into Portuguese and further validation of the Fecal Incontinence Quality of Life (FIQL) questionnaire. It comprises 24 questions, covering the same domains of the original: lifestyle (questions 2, 4, 7, 10, 16, 19, 20; totaling seven items), behavior (questions 1, 6, 11, 12, 15, 17, 21; seven items), depression (questions 3, 8, 14, 18, 22, 23, 24; seven items), and shame/embarrassment (questions 5, 9 e 13; total of three items). Questions 22 and 23 are related to patient opinion and satisfaction with their own health and bowel function. Each question has a score from 1 to 4, with 1 being the worst situation. The final score is obtained by summing the scores of each domain, and ranges from 4 to 16 (Chart 2).

Patients

The questionnaires were administered to 63 patients with ARM, aged from 7 to 19 years, whose surgical treatment had been completed for at least 6 months. They were all operated by the pediatric surgery team at Instituto da Criança, HC-FMUSP, in São Paulo. Patients were randomly selected, and those with conditions that could affect the sphincter control, such as impaired psychomotor development and neurological disorders, were excluded.

Twenty-eight (44.5%) patients were female and 35 (55.5%) were male, and the mean age was 12.2 years.

Fifty-nine normal children without intestinal functioning complaints and demographically similar to the assessed patients also answered the questionnaires. They were selected from the pediatric surgery outpatient clinic and were suffering from common surgical conditions such as inguinal hernia and phimosis. As done with the study groups, questionnaires were sent by mail or answered by telephone interview or after outpatient visit.

Comparison between patients with ARM and the controls

Data on children with ARM were compared with those of children in the control group and subjected to statistical analysis.

Comparison between patients with low-type ARM and patients with high and intermediate ARM

In this study, the cases of high and intermediate ARM were grouped as "high-type", since they were treated using the same technique. Cases of persistent cloaca were

CHART 1 Fecal Continence Index (ICF) questionnaire.**A – Identification**

Name: _____ Address: _____ District: _____
 City: _____ State: _____ Postal code: _____
 Phone: _____

B – Introduction

1. Who answers?
 - a) Personally.
 - b) With the aid of another person.
 - c) Answered by another person.
2. Current age: _____ Gender: _____
3. What is your main activity?
 - a) Studies. Which year are you in school? _____
 - b) Works.
4. Some time ago you had surgery to correct a malformation of the intestine. In addition to this, did you have other health problems that required medical treatment or surgeries? If so, what were them?
5. Do these problems still cause you any difficulties? If so, which were them?

C – Control of bowel movements (stools)

Some people who had intestinal malformation, even after their surgical correction, may have difficulty in controlling bowel movements (stools). Our goal is to know through this questionnaire if the functioning of your gut causes you any annoyances, and how often. If you have the problem mentioned in the question, but not because of the difficulty with bowel movements, leave the question blank.

1. How often do you pass stools into the toilet in a full day (24 hours)? _____
2. At night, after going to bed, how often do you need to get up to evacuate your bowel? _____
3. In a full day (24 hours), how often do you need to change clothes, underwear or panties, because of soiling? _____ If you do not soil your clothes every day, how many times per week or per month does this happen? _____
4. If you need to go out or attend any venue, which of the following alternatives do you choose to keep yourself neat?
 - a) Uses a liner, diaper or carries extra clothes to change.
 - b) Does a bowel cleansing and/or avoids eating before leaving.
 - c) Goes to the toilet before leaving.
 - d) No preparation.
5. If you have time and are willing to do all the preparation before leaving, what is the result?
 - a) Stays clean for over 4 hours.
 - b) Stays clean for up to 4 hours.
 - c) It does not work, soiling occurs unintentionally.
6. Do you usually have diarrhea? How often does it occur?
 - a) Every week.
 - b) Once or twice a month.
 - c) Never.
7. Do you get constipated, going days without passing stool, with stools getting dry and forming round hard lumps in your belly that need to be removed using rectal maneuvers?
 - a) Every week.
 - b) Once a month.
 - c) Never.
8. Mark the care taken regularly, as a routine, to improve bowel function:
 - a) Does not eat certain foods because they make you feel unwell.
 - b) Eats at set hours to go to the toilet after meals.
 - c) Performs bowel cleansing.
 - d) Takes some medication. Which one? _____

CHART 2 Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents (QQVCFCA).

A – Personal and social questions

Below, we will ask other questions as to whether the functioning of your bowel causes you any annoyances, and how often. If this complaint exists, but not due to poor bowel function, leave the question blank.

	almost always	sometimes	rarely	never
1 – When I am away from home I try to stay close to a toilet				
2 – I avoid visiting my friends				
3 – I am invited to parties and to hang out less often				
4 – I avoid spending the night away from home				
5 – I fear that people will notice the smell				
6 – I would rather stay home than going out				
7 – I avoid eating out				
8 – I am not able to participate in activities with my friends				
9 – I avoid talking about the issue with others				
10 – I need to plan my activities based on my bowel function				
11 – It impairs my school performance				
12 – It hinders my professional life				
13 – I pass stools without noticing				
14 – I prefer that people do not know about my problem				
15 – It makes it difficult to play sports				
16 – I avoid travelling				
17 – I have difficulty making friends				
18 – I worry about accidentally passing stool				
19 – Leaving home worries me				
20 – I ceased to do the things I like				
21 – I feel that I do not control my gut				
	poor	regular	good	excellent
22 – Generally you think your health is:				
	poor	regular	good	excellent
23 – What do you think about your bowel function?				

also considered high ARM, for the same reason. Thus, high anomalies in males were the cases of recto-urethral fistula or rectovesical fistula, and in females, rectovestibular fistula or cloaca. Low-type anomalies included cases of perineal fistula, anterior ectopic anus, anal stenosis, anal atresia, and scrotal fistula in males. In cases of anomaly without fistula, Wangersteen-Rice or inverted-position radiographs (Figure 1) were carried out. Thus, when the distance between the air column in the distal rectum and the anus site was greater than 1 cm, the anomaly was considered high, while distances of less than 1 cm were considered low.

All cases of high-type anomaly were treated with double-barrel colostomies in the neonatal period and, later, with posterior sagittal anorectoplasty according to Pe-

ña’s technique. Low-type anomalies were treated with proctoplasty in the neonatal period.

Based on these data, we were able to compare the results of children with high ARM with those of children with low ARM.

Associated malformations

Given that ARMs are associated with other malformations, a survey of other defects present in these patients was performed. Data were obtained from medical records and questionnaires.

Statistical analysis

The histograms constructed reveal that the data does not have a normal distribution. Thus, in all analyzes, Mann-

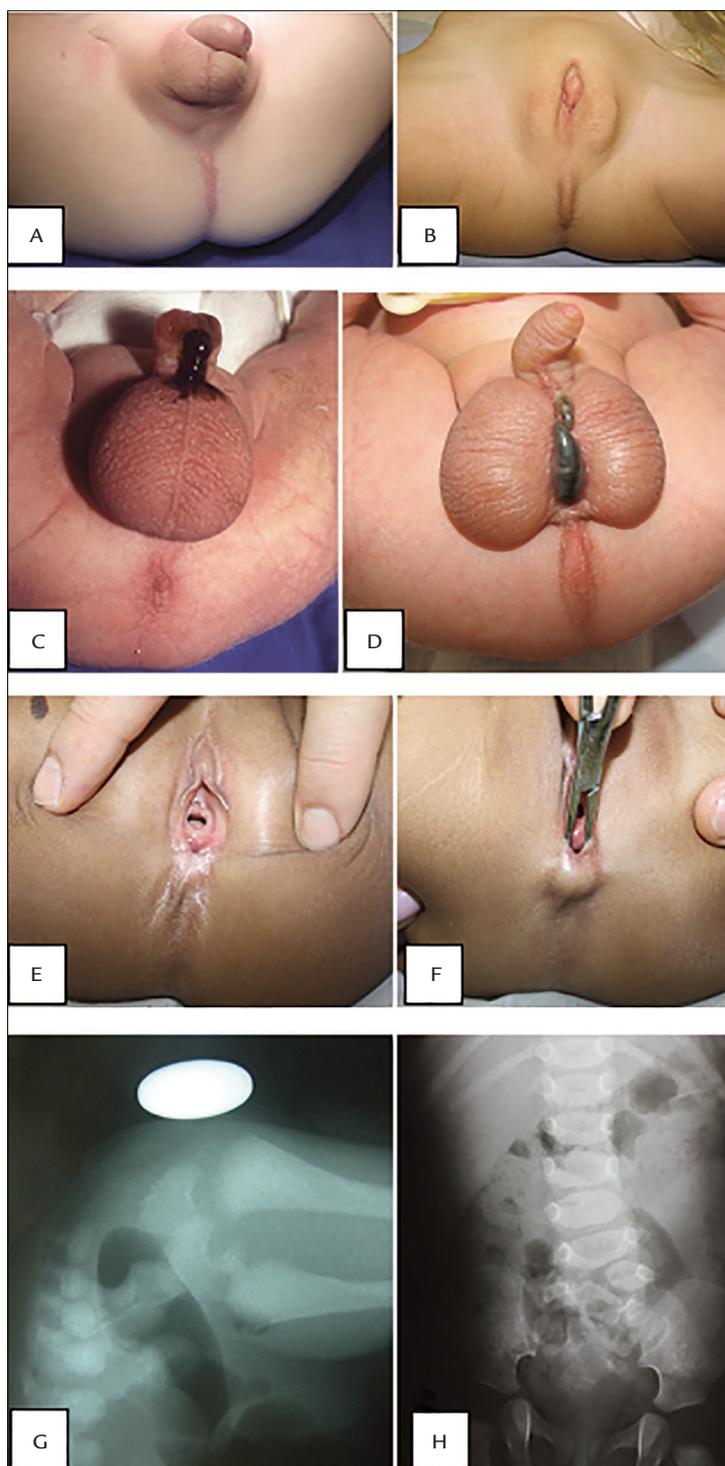


FIGURE 1 A. High anorectal anomaly in a male newborn, 24 hours after birth – note the absence of the anus and any orifices indicating perineal fistula. B. Persistent cloaca in a female newborn – note the absence of the anus and presence of one perineal orifice corresponding to the urinary, genital and digestive tracts. C. High anorectal anomaly in a male newborn, with recto-urethral fistula – note the passing of meconium through the penis. D. Low-type anomaly, with visible perineal fistula, stained with meconium. E. Perineal anus in a girl. F. The placement of a surgical instrument allows the membrane to be sectioned for the correction of the defect to be seen. G. Inverted radiography made in a newborn with anomaly and without perineal fistula – note the distance from the rectal cul-de-sac pouch (filled with gas) up to the anal impression marked by the metallic object (coin). H. Plain radiographs showing abnormality in the lumbar spine (hemivertebra) and partial absence of the sacrum.

-Whitney test was used. The confidence interval was 95%, thus with an alpha error of 0.05.

RESULTS

Comparison between patients with ARM and the controls

We found that the quality of life of children with ARM is globally compromised in every domain and in the ICF questionnaire compared with the control group ($p < 0.001$ for all comparisons) (Table 1).

Twenty-five (42.4%) of the control children showed good continence, and 34 (57.6%), normal continence. Conversely, among patients with ARM, 10 (15.9%) had poor continence; 20 (31.7%), regular; 24 (38.1%), good; and 9 (14.3%) had normal continence.

Comparison between patients with low-type ARM and patients with high and intermediate ARM

Forty of 63 patients with ARM had high-type anomalies and 15 had low-type. As for the eight remaining patients, there was no information regarding type of anomaly. Patients with high-type ARM had a mean age of 12.9 years, and included 18 females and two males. With respect to continence, six (15%) showed poor; 13 (32.5%), regular; 15 (37.5%), good; and six (15%), normal continence. Patients with low-type ARM, however, had a mean age of 9.93 years, and six were female, while nine were male. Two (13.3%) patients had poor continence; six (40%), regular; four, (26.7%) good; and three (20%), normal continence. Statistical comparisons produced the following p-values: ICF ($p = 0.95$), QQVCFA ($p = 0.87$), lifestyle ($p = 0.68$), behavior ($p = 0.71$), depression ($p = 0.50$), and shame/embar-

rassment ($p = 0.40$). Therefore, both types of ARM presented similar performances (Table 2).

Associated malformations

Thirty-two patients with ARM had other associated malformations. Of these, 13 had defects in more than two systems. We found 14 patients with urinary tract malformations, of which: one cake kidney, three neurogenic bladders, two cases of crossed renal ectopia, two of single kidney, three of vesicoureteral reflux, one of two urethral meatuses, one of penoscrotal transposition, one of urethral stenosis, one of bilateral hydronephrosis, one of horseshoe kidney, one of contracted kidney, one of double bladder, and two of malformations not specified in the medical records. Six patients had cardiac abnormalities, including one with persistent arterial duct, one with permeable foramen ovale; two with interatrial communication; two with interventricular communication; and three with defects not defined in the medical records. Three patients had esophageal atresia and one intestinal atresia. As for the skeletal system, five had involvement of the sacrum (agenesis or hypoplasia), two hypoplastic thumbs, six vertebral malformations, one radius agenesis, and one unspecified. One patient had meningocele. Two patients had vaginal agenesis, and two had Rokitansky syndrome. One patient had unspecified larynx malformation.

DISCUSSION

Fecal incontinence can occur with some degree of intensity in approximately 30 to 50% of the patients with ARM,^{8,9} similarly to that observed in the present study.

TABLE 1 Comparison of ICF and QQVCFA questionnaires, and their domains between the children with ARM and the control group.

		Mean	Median	Standard deviation	Percentile 25	Percentile 75
ICF	ARM	10.76	11.00	4.01	8.00	14.00
	Control	15.37	16.00	1.00	15.0	16.00
QQVCFA	ARM	11.17	11.79	3.27	8.99	14.03
	Control	15.23	15.57	0.95	15.07	15.71
LS	ARM	3.11	3.28	0.90	2.40	4.00
	Control	3.88	4.00	0.30	4.00	4.00
BEH	ARM	3.08	3.28	0.81	2.57	3.85
	Control	3.88	4.00	0.27	3.85	4.00
DEP	ARM	2.63	2.71	0.81	2.00	3.28
	Control	3.58	3.66	0.31	3.42	3.85
EMB	ARM	2.33	2.30	1.07	1.33	3.33
	Control	3.89	4.00	0.27	4.00	4.00

ICF: Fecal Continence Index; QQVCFA: Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents; ARM: anorectal malformations; LS: lifestyle; BEH: behavior; DEP: depression; EMB: shame/embarrassment.

TABLE 2 ICF and QQVCFCA questionnaires, and their domains in patients with high-type ARM and patients with low-type ARM.

		Mean	Median	Standard deviation	Percentile 25	Percentile 75
ICF	High-type ARM	10.89	11.5	3.90	8.00	14.00
	Low-type ARM	10.86	10.00	4.22	7.00	1.005
LS	High-type ARM	3.19	3.42	0.83	2.60	4.00
	Low-type ARM	2.99	3.27	0.97	2.16	4.00
BEH	High-type ARM	3.14	3.28	0.73	2.57	3.71
	Low-type ARM	2.99	3.00	0.92	2.57	3.85
DEP	High-type ARM	2.61	2.49	0.74	2.03	3.10
	Low-type ARM	2.71	2.85	0.92	2.00	3.28
EMB	High-type ARM	2.28	2.15	1.04	1.33	3.00
	Low-type ARM	2.55	2.66	1.15	1.33	4.00
QQVCFCA	High-type ARM	11.22	11.72	3.01	9.06	13.24
	Low-type ARM	11.26	11.79	3.73	7.42	15.28

ICF: Fecal Continence Index; QQVCFCA: Questionnaire for Assessment of Quality of Life Related to Fecal Continence in Children and Adolescents; ARM: anorectal malformations; LS: lifestyle; BEH: behavior; DEP: depression; EMB: shame/embarrassment.

The importance of fecal continence and its relation with QoL has been much explored through questionnaires, allowing assessments that are useful for clinical practice and crucial for clinical trials.

In addition to traditional questionnaires such as the Fecal Incontinence Questionnaire,¹⁰ The Fecal Incontinence Severity Index,¹¹ Fecal Incontinence Quality of Life (FIQL),¹¹ Hirschsprung's Disease Anorectal Malformation QoL Questionnaire (HAQL),⁵ Medical Outcomes Study 36 – Item Short – Form Health Survey,¹² and the Clinical Evaluation of Continence (Holschneider Criteria),⁶ many translated versions have been created. Minguez et al. validated the FIQL into Spanish, applying it to patients with fecal incontinence caused by different etiologies;¹³ Rullier et al. validated the FIQL into French,¹⁴ and in Sweden, Wigander et al. translated and culturally adapted the HAQL.¹⁵ In Brazil, Yusuf et al. validated the FIQL into Portuguese, but did not adapt it to children and adolescents since the mean age of their patients was 52.8 years.¹⁶

In keeping with this trend, our group created and validated the QQVCFCA and the ICF questionnaires, since the population seen in our service has specific characteristics that were not covered by other questionnaires. The QQVCFCA and the ICF questionnaires are in Portuguese, they address issues relevant to children and adolescents, use language that is easy to understand, without overly technical terms or complex alternative words that are difficult to comprehend, they can be applied in interviews or answered by the patient or guardian with-

out the need for an interviewer, and excluded issues considered unsuitable for children, involving sexuality and situations of severe depression. The steps of translation, cultural adaptation, reproducibility in time, construct validity and discriminant validity were carried out judiciously. Once finalized, these instruments could be applied to a larger number of patients and this work shows the data obtained.

The patients with ARM presented higher rates of fecal incontinence compared with the controls. The diseases themselves may present with malformations of perineal structures, which are fundamental for fecal continence, and surgical treatment of anatomic area often can damage microstructures, such as nerves and muscle fibers. Consequently, a reduction in quality of life of these patients was identified. QQVCFCA domains most affected were: shame/embarrassment (median of 4.0 for the controls and 2.33 for the ARM group) and depression (medians of 3.66 and 2.71 for ARMs). Thirty patients (47.6%) were considered incontinent (poor or regular continence), while none of the children in the control group had such problem.

The percentages of patients with fecal incontinence in the groups of high and low ARM were similar (47.5 and 53.3%, respectively). We expected that continence in cases of high ARM would be worse than in low-type anomalies. We noted that the average age of the patients with high ARM was 12.9 years and of those with low ARM, 9.9 years. Since, classically, children with anorectal anomalies take longer than normal children to acquire fecal continence (often only in adolescence), it is possible that this

difference of three years between cases of high and low anomalies may have influenced the fact that the two groups have shown similar results.¹⁷⁻¹⁹

Last, the combination of ARM with other congenital malformations is a consensus in the scientific literature.²⁰ The most common are those in the urinary tract, musculoskeletal system and cardiovascular. Therefore, patients often undergo several corrective surgical and monitoring procedures in different specialized outpatient services, increasing treatment complexity. In addition, the association of ARMs and genetic diseases, such as Down syndrome, is common and affects a significant percentage of patients. Due to such variation regarding conditions and associated genetic diseases, the fact that the patients can present other factors interfering with their assessments of QoL and fecal continence, which is a cause for bias, must be taken into account. There is a great need for the creation and validation of specific methods to assess such children, both in terms of fecal continence and quality of life.

As the questionnaires were validated for children and adolescents aged 7-19 years, children under 7 years of age were not studied. Creating different questionnaires and play evaluations is fundamental due to difficulties related to understanding and abstraction required for the answers.

CONCLUSION

Patients with ARM must learn to live with a variety of symptoms and difficulties, including fecal incontinence. The data presented in this study show the need for comprehensive and multidisciplinary care of children with ARM in the long term, aimed at improving the quality of life and to overcoming the psychological and social difficulties caused by changes in fecal continence. A better understanding and acceptance of limitations, leading to self-awareness and the adoption of more appropriate nutritional strategies, can improve both fecal continence and quality of life.

RESUMO

Avaliação tardia da continência fecal e da qualidade de vida em pacientes operados por malformações anorretais

Introdução: os pacientes operados para correção de malformações anorretais (MAR) podem evoluir com incontinência fecal, constipação e *soiling*, com prejuízo na qualidade de vida.

Objetivo: avaliar pela aplicação de questionários a continência fecal e a qualidade de vida de crianças no seguimento pós-operatório tardio de correção de MAR, formas altas e baixas. Também foram comparados os índices

de continência fecal e qualidade de vida com um grupo controle de crianças.

Método: foram utilizados o Questionário para o Índice de Continência Fecal (ICF) e o Questionário para Avaliar a Qualidade de Vida Relativa à Continência Fecal em Crianças e Adolescentes (QQVCFCA). Os questionários foram aplicados em 63 pacientes de 7 a 19 anos com MAR, com tratamento cirúrgico finalizado há pelo menos 6 meses. Os pacientes foram comparados com um grupo controle de 59 crianças.

Resultados: nos pacientes do grupo controle, 25 (42,4%) apresentaram boa continência e 34 (57,6%), normal. A qualidade de vida nas crianças com MAR está comprometida globalmente, em todos os domínios e no ICF, quando comparada com a dos controles ($p < 0,001$). Não houve diferença entre os pacientes com anomalias altas e anomalias baixas. Trinta e dois (50,8%) pacientes apresentaram outras anomalias associadas.

Conclusão: nos pacientes operados para correção de MAR, qualidade de vida e ICF foram comprometidos, não havendo diferença entre os pacientes com as formas altas ou baixas da doença. Em cerca de metade dos casos existem outras malformações associadas.

Palavras-chave: ânus imperfurado, malformação anorretal, qualidade de vida, incontinência fecal.

REFERENCES

- Hartman EE, Oort FJ, Aronson DC, Sprangers MA. Quality of life and disease-specific functioning of patients with anorectal malformations or Hirschsprung's disease: a review. *Arch Dis Child.* 2011; 96(4):398-406.
- Herman RS, Teitelbaum DH. Anorectal malformations. *Clin Perinatol.* 2012; 39(2):403-22.
- Witvliet MJ, Slaar A, Heij HA, van der Steeg AF. Qualitative analysis of studies concerning quality of life in children and adults with anorectal malformations. *J Pediatr Surg.* 2013; 48(2):372-9.
- Rockwood TH, Church JM, Fleshman JW, Kane RL, Mavrantonis C, Thorson AG, et al. Fecal incontinence quality of life scale: quality of life instrument for patients with fecal incontinence. *Dis Colon Rectum.* 2000; 43(1):9-17.
- Hanneman MJ, Sprangers MA, De Mik EL, Ernest van Heurn LW, De Langen ZJ, Looyaard N, et al. Quality of life in patients with anorectal malformation or Hirschsprung's disease: development of a disease-specific questionnaire. *Dis Colon Rectum.* 2001; 44(11):1650-60.
- Holschneider AM, Metzger EM. Elektromanometrische Untersuchungen der Kontinenzleistung nach rektotoanalen Fehlbildungen. *Z Kinderchir.* 1974; 14:405-12.
- Mathias AL, Tannuri AC, Ferreira MA, Santos MM, Tannuri U. Validation of questionnaires to assess quality of life related to fecal incontinence in children with anorectal malformations and Hirschsprung's disease. *Rev Paul Pediatr.* 2016; 34(1):99-105.
- Borg HC, Holmdahl G, Gustavsson K, Doroszkiewicz M, Sillén U. Longitudinal study of bowel function in children with anorectal malformations. *J Pediatr Surg.* 2013; 48(3):597-606.
- Julià V, Tarrado X, Prat J, Saura L, Montaner A, Castañón M, et al. Fifteen years of experience in the treatment of anorectal malformations. *Pediatr Surg Int.* 2010; 26(2):145-9.
- Reilly WT, Talley NJ, Pemberton JH, Zinsmeister R. Validation of a questionnaire to assess fecal incontinence and associated risk factors. *Dis Colon Rectum.* 2000; 43(2):146-54.

11. Rockwood TH. Incontinence severity and QOL scales for fecal incontinence. *Gastroenterology*. 2004; 126(1 Suppl 1):S106-13.
12. Ciconelli RM, Ferraz MB, Santos W, Meinão I, Quaresma MR. Brazilian-Portuguese version of the SF-36. A reliable and valid quality of life outcome measure. *Rev Bras Reumatol*. 1998; 39(3):143-50.
13. Minguez M, Garrigues V, Soria MJ, Andreu M, Mearin F, Clave P. Adaptation to Spanish language and validation of the fecal incontinence quality of life scale. *Dis Colon Rectum*. 2006; 49(4):490-9.
14. Rullier E, Zerbib F, Marrel A, Amouretti M, Lehur PA. Validation of the French version of the Fecal Incontinence Quality-of-Life (FIQL) scale. *Gastroenterol Clin Biol*. 2004; 28(6-7 Pt 1):562-8.
15. Wigander H, Frenckner B, Wester T, Nisell M, Öjmyr-Joelsson M. Translation and cultural adaptation of the Hirschsprung's Disease/Anorectal Malformation Quality of life Questionnaire (HAQL) into Swedish. *Pediatr Surg Int*. 2014; 30(4):401-6.
16. Yusuf SA, Jorge JM, Habr-Gama A, Kiss DR, Gama Rodrigues J. [Evaluation of quality of life in anal incontinence: validation of the questionnaire FIQL (Fecal Incontinence Quality of Life)]. *Arq Gastroenterol*. 2004; 41(3):202-8.
17. Kyrklund K, Pakarinen MP, Taskinen S, Rintala RJ. Bowel function and lower urinary tract symptoms in males with low anorectal malformations: an update of controlled, long-term outcomes. *Int J Colorectal Dis*. 2015; 30(2):221-8.
18. Kyrklund K, Pakarinen MP, Koivusalo A, Rintala RJ. Long-term bowel functional outcomes in rectourethral fistula treated with PSARP: controlled results after 4-29 years of follow-up: a single-institution, cross-sectional study. *J Pediatr Surg*. 2014; 49(11):1635-42.
19. Neuvonen MI, Kyrklund K, Rintala RJ, Pakarinen MP. Bowel function and quality of life after transanal endorectal pull-through for Hirschsprung disease: controlled outcomes up to adulthood. *Ann Surg*. 2016. [Epub ahead of print]
20. Stoll C, Alembik Y, Dott B, Roth MP. Associated malformations in patients with anorectal anomalies. *Eur J Med Genet*. 2007; 50(4):281-90.