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Long term follow-up and transition of care in anorectal malformations: An international survey

Stefano Giuliani [a],*, Emily Decker [a], Ernesto Leva [b], Giovanna Riccipetitoni [c], Pietro Bagolan [d]

[a] Department of Pediatric and Neonatal Surgery, St. George's University Hospitals NHS Foundation Trust, Blackshaw Rd, SW17 0QT London, United Kingdom
[b] Department of Pediatric Surgery, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Via Francesco Sforza 35, 20122 Milan, Italy
[c] Department of Pediatric Surgery, Buzzi Children's Hospital, Via Lodovico Castelvetro 32, 20154 Milan, Italy
[d] Department of Medical and Surgical Neonatology, Bambino Gesu' Research Children's Hospital, Piazza Sant'Onofrio 4, 00165 Rome, Italy

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ABSTRACT

Background: The aim of the study was to assess current international practice in the long term follow-up, in managing active problems and transition of care for teenagers born with anorectal malformations (ARM).
Methods: An original survey was administered to delegates attending two large colorectal surgical meetings in 2015. The 21 questions covered long term follow-up, specific issues for teenagers and transition of care.
Results: 96/236 delegates completed the survey. Follow-up was routinely suspended before 10 year of age by 33% of respondents. 90% of them did not use a scoring system to assess or risks stratify patients, despite 81% stating that an objective score would be beneficial. 40% of respondents felt that >30% of their teenagers had ongoing active medical or psychosocial issues. 42% thought their patients were not ready to be transitioned. The process of transition should start around 13–16 years according to 54% of respondents. 72% had no protocol for transition and 82% did not hold multidisciplinary meetings with adult practitioners before transition.
Conclusions: International consensus on the following aspects of the care in ARM is needed: structured long term follow-up, objective assessment and risk stratification scores, pathways of transition and methods to prepare patients, parents and adult practitioners.

Anorectal malformations (ARM) are a spectrum of rare disorders of the rectum and anus, with variable anatomy and unpredictable long-term outcomes [1]. ARM have an incidence of 1 in 2000–5000 live births [2].

Associated anomalies (including genito-urinary, skeletal and spinal malformations) may be present in up to 60–70% of cases [3]. Cloaca and high ARM have the highest incidence of associated anomalies and long term active problems [1,4]. The complexity of the original malformation and the associated anomalies plays a role, not only in the long-term fecal and urinary continence, but also in the psychosocial development and quality of life at different ages [5].

There is no international consensus on follow-up, objective assessment of active problems or well-structured transition pathways for children with ARM [6]. The aim of this study was to evaluate international practice and to identify areas that require further research in the long term follow-up, management of active problems during adolescence, and transition of care to adult services for children with ARM.

1. Methods

The original questionnaire was designed to investigate the following three topics: long term follow-up, active issues present in teenagers (12–16 years old) and transition of care (Appendix 1 – full survey).

The survey was administered to delegates attending the UK-Pediatric Colorectal Group (UKPCG) meeting held in May 2015 in Nottingham, UK and the 22nd International Meeting of the Pediatric Colorectal Club (IMPCC) held in June 2015 in Milan, Italy. Approval was obtained from the Scientific Committee of the International Colorectal Club. Those attending the UKPCG meeting (31 participants) received hard copies of the survey. IMPCC meeting delegates (205) were circulated an electronic survey (SurveyMonkey) after the conference; they had 2 months to respond and received a maximum of 4 electronic reminders.

2. Results

2.1. Demographics of respondents

136 out of 236 (58%) respondents completed the questionnaire; 40 partially completed the survey and, therefore, were excluded from the

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Abbreviations: ARM, anorectal malformations; MDT, multiple-disciplinary team.

* Corresponding author at: Department of Pediatric and Neonatal Surgery, St. George's University Hospitals, NHS Foundation Trust, Blackshaw Road, London, SW17 0QT, United Kingdom. Tel.: +44 208 67252926; fax: +44 208 7250711.
E-mail address: stefano.giuliani@nhs.net (S. Giuliani).

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final analysis of 96 completed surveys. 73/96 (76%) of respondents were consultants, 12 (13%) trainees and 11 (11%) identified as other (3 chiefs of surgery, 2 psychologists, 3 surgical fellows and 3 staff grade surgeons). Respondents largely worked in European (EU) countries (76%) and 23 (24%) worked in non-EU countries. The most represented countries were: Italy, United Kingdom, Scandinavia, Germany, The Netherlands, USA, Russia, South Africa, India, Australia, Saudi Arabia and Brazil.

2.2. Long term follow-up

A well-structured multidisciplinary team (MDT) follow-up was used routinely from birth by 73% (70/96) of the respondents. The most common specialists involved in the MDT are shown in Fig. 1. Almost one-third of respondents (25/96, 26%) did not provide an MDT follow-up from birth for their patients.

The usual duration of the follow-up for patients with ARM is shown in Fig. 2.

A published scoring system to objectively assess and compare progresses was not routinely used by the majority of respondents (68/96, 71%). The scoring systems used by the remaining 28% (27/96) were: the Krickenbeck classification for fecal continence (9/27); Holshneider score (2/27); Baylor continence scale (1/27); Modified Wingfield score (1/27); Rome III criteria (1/27), Wingspread classification (1/27); Rintala score of continence (1/27). Three were not able to specify the scale they used and 9 used their own unpublished score.

2.3. Active issues in teenagers (12–16 years old)

The majority of the respondents (85/96, 89%) acknowledged that a significant proportion of ARM patients develop new problems requiring a larger MDT in their teenage years.

There was wide variation in the incidence of active medical or psychosocial issues in teenagers with ARM (Fig. 3). To identify the most common active issues, we asked respondents to rank the following issues: fecal control, urinary continence, sexual function, psychosocial issues and quality of life (with 1 representing the most common issue and 5 the least). Fecal continence was considered the most common active problem in teenagers (median 1), followed by psychosocial issues (median 2) and quality of life (median 3). Least common problems for the respondents were jointly urinary continence and sexual function, both median 4. Notably, each issue was ranked both most and least common by several respondents.

An objective scoring system to risk stratify teenagers, who may need additional support during adolescence, was not used by the majority of the respondents (86/96, 90%). Conversely, when asked if they felt a practical assessment tool may be useful: 78 (81%) responded yes, 6 (7%) no and 2 (2%) were undecided. Among 10 respondents who risk stratified patients for transition, 5 used unpublished scoring systems, 2 used a quality of life score and 3 did not specify (Table 1).

Assessing the patient's preparedness for transitioning, respondents were divided about if their patients were fully aware and understood their anorectal condition and associated anomalies (58% yes versus 42% no). Optimum age for initiating the transition care process was variable among the respondents: 13–16 years for 52/96 (54%), 17–18 years for 26/96 (27%) and, as early as, 9–12 years for 18/96 (19%).

2.4. Transition of care

The majority of the respondents (82/96, 86%) acknowledged that it is responsibility of the pediatric surgeon to transition patients properly and effectively to the adult services. Small numbers felt that the general practitioner (5/96, 5%), pediatric urologist (2/96, 2%) pediatrician (2/96, 2%), continence nurse (1/96, 1%) or other healthcare professionals (4/96, 4%) should have this responsibility.

Relatively small numbers were transferred to adult services every year: 33 (35%) of the respondents transferred 3–5 patients, 32 (33%) 1–2 patients, 14 (15%) 5–10 patients, 9 (9%) >10 patients and 7 (7%) no patients. One person was undecided (1%). There was not a defined protocol for transition to adult services for 72% of respondents (69/96) and there were no dedicated transition MDT meetings held between pediatric and adult practitioners in 82% (79/96) (Fig. 4).

Regarding parental preparation and participation to transition, the respondents (53%) acknowledged that the main role for parents was to promote their child's independence during this transitional period. Secondary roles were to educate their child about the condition and to help managing medical care (e.g. appointments). Table 2 demonstrates the main barriers to transition of care, as perceived by the respondents.

The following topics, with the percentage of respondents, were discussed directly with the teenagers without their parents present in

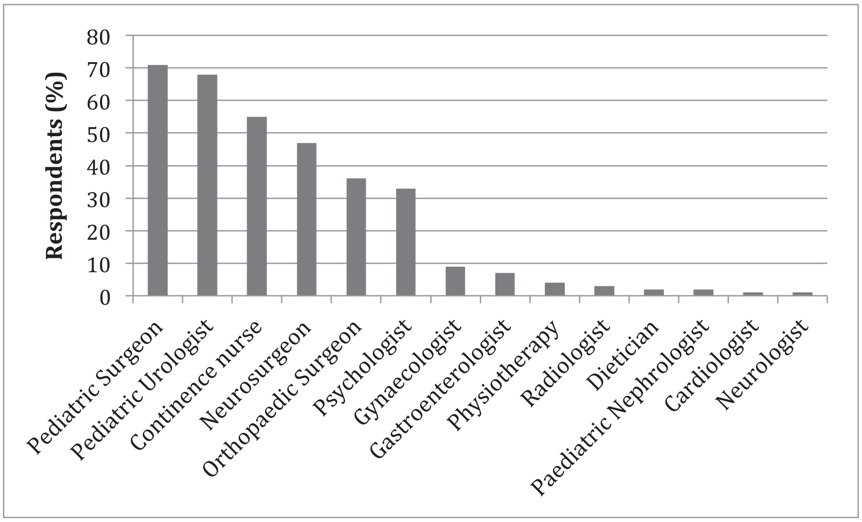

Fig. 1. Composition of the multidisciplinary team (MDT) involved in the follow-up of patients with ARM for the respondents.