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Practice Management

Transition of care in patients with anorectal malformations: Consensus by the ARM-net consortium[☆,☆☆,★,★★]

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ABSTRACT

Objectives: To develop the first consensus to standardize the management of patients with Anorectal Malformations (ARMs) transitioning from childhood to adulthood.
Methods: A dedicated task force of experts performed an extensive literature review and multiple meetings to define the most important aspects of transition of care. The findings were discussed with all ARM-net consortium members and a set of practical recommendations agreed upon at the annual meeting in 2016.
Result: We defined seven domains that are essential to provide an effective and practical transition process. Within each domain we have developed a set of key recommendations that are important to be considered for ARM patients entering the age of transition.
Conclusions: It is crucial that transition begins at an early age with regular and well-structured follow-up. Cooperation with a selected multidisciplinary team of pediatric and adult practitioners is required to prepare patients and families for effective transition to adult care and to reduce long term morbidity.
Type of study: Review/Consensus paper.
Level of evidence: III.

Contents

Abbreviations: ARM, anorectal malformation; MDT, multidisciplinary team; QOL, quality of life; ITP, individual transition plan; EUPSA, European Pediatric Surgeons' Association; BAPS, British Association of Pediatric Surgeons.

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[★★] **Table of Contents Summary**This is the first consensus paper to standardize the transition of care in teenagers born with anorectal malformations.

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Anorectal malformations (ARMs) are a complex collection of congenital disorders of the anus, rectum and genitourinary system with a reported incidence of 1 in 1500–5000 live births [1–3]. Despite successful surgical intervention in the neonatal period, more than one third of children with ARMs will experience functional problems in the long term [4].

As recently highlighted by an international survey on transition of care in ARMs, it is the responsibility of the pediatric multidisciplinary team (MDT) to lead a well-organized process of transition of care [5]. In the majority of centers the MDT is composed of pediatric surgeons, continence nurses and pediatric urologists. In addition, the survey showed a lack of international agreement on the following key elements in the care of these patients: structured long-term multidisciplinary follow-up; objective assessment and risk stratification scores; pathways of transition, methods to prepare patients and parents; and role of adult practitioners in the transition process to adult services.

To prepare an effective transition of care it is important to be able to identify active problems early and to have in place a holistic and structured process throughout adolescence. The aim of this paper is to define the minimum standards of transition of care that can be used worldwide by different healthcare providers and in different hospital settings. To achieve this, we will present seven domains with a set of key recommendations that we believe are essential to be considered in each patient approaching the age of transition.

1. Methods

A task force composed by four pediatric surgeons, two specialist nurses, two parents, an adult patient, a psychologist, and two leaders of parents' associations was constituted in June 2015. The task force carried out an extensive literature search on transition of care and anorectal malformations. Briefly, the search was focused on identifying key active problems in ARM patients, methods of transition of care in chronic pediatric conditions, timing and structure of effective transition of care, and patient/parent preparation.

After 2 years of work the task force produced a consensus statement approved by all the Members of the ARM-net Consortium during the Annual General Meeting in Paris (October 2016).

2. Results

2.1. Definition and goals of an effective transition of care in patients with anorectal malformations (domain 1)

A position paper of the Society of Adolescent Medicine clearly defined transition as the *"purposeful planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems"* [6].

Transition to adult services for children with ARMs should not consist of just transfer of care but it should be a well-structured and effective process across adolescence and young adult life [7]. To achieve this, the main goals of the transition process should be: 1) to prepare the patient for the transfer, providing skills and knowledge about the disease; 2) to prepare the parents; 3) to prepare and inform the health provider at the time of the transfer [8].

We believe an effective transition should focus on *"promoting continuity of care, improve or maintain disease control, increase treatment adherence and knowledge of the disease, encourage autonomy management of the disease, increase patient, parent, and healthcare provider satisfaction"* [6].

Since ARM is a spectrum of disease, the burden of transition will vary between patients. It is more likely that those patients who have a complex malformation at birth (i.e. cloaca and bladder neck fistula) or with long standing active problems up to the point of transition will need lifelong support by a team of specialized healthcare practitioners.

For this reason, it is essential to have a comprehensive long-term follow-up from birth to adulthood for every patient with ARMs. This will allow for monitoring of significant chronic medical problems but also the early detection of new physical and psychosocial issues during puberty.

We have defined five separate main 'goals of transitions' (Essential Recommendation 1). To obtain these, the pediatric surgeon should be responsible for coordinating the transition process from onset at early puberty. A coordinated structure, relevant to the type of hospital, is required to deliver an effective long-term follow-up and transition process.

2.1.1. Essential recommendations domain 1

a) Main goals of transition:

- to identify active somatic or psychosocial problems during the transition period
- to prepare the patient, providing skills and knowledge about their ARM and associated anomalies
- to prepare the parents to support their children during the process
- to set up an effective network of a limited number of centers with selected pediatric and adult healthcare providers
- to develop pathways and protocols for a well structured transition of care process between the local pediatric surgery department and few selected national centers able to support the adult patients for life

b) Pediatric practitioners and, in particular, the pediatric surgeon who lead the care during childhood should remain available as long as possible for the adult practitioners and patients

Table 1
Prevalence of active long-term problems in ARMs patients after the first decade of life [9].

	Overall Prevalence in ARMs	High ARMs	Low ARMs
Fecal incontinence	16.7%–76.7%	16.7%–96.4%	43.8%–58.8%
Chronic constipation	22.6%–86.7%	22.2%–61.5%	8.7%–47.1%
Urinary incontinence	2.2%–30.5%	2.2%–28%	6.2%–39.1%
Ejaculatory dysfunction	15.6%–41.2%		
Erectile dysfunction	5.6%–11.8%		