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Longitudinal study of bowel function in children with anorectal malformations

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Abstract

Purpose: Longitudinal follow-up of changes in bowel function in children with anorectal malformations (ARMs) with or without spinal cord pathology and neurogenic bladder dysfunction (NBD) as they grow. Another purpose was to identify predictors influencing bowel functional outcome.

Material and Methods: The study included 41 patients with ARM, excluding perineal fistula (21 boys and 20 girls). Bowel function was evaluated at ages 5, 10 and 15 years using a structured questionnaire and a three-week registration of number and time of bowel movements, episodes of fecal leakage and soiling. Additional bowel treatment with enemas and stool softeners and use of diapers were recorded. A group of 52 healthy boys and girls was used as control.

Results: A successive improvement in functional outcome with age in children with ARM and normal spinal cord was seen with respect to continence, soiling and constipation. Continence was achieved earlier in girls than in boys (at 10 years: girls 80%, boys 36%). Soiling and constipation decreased with age both in grade and frequency (at 10 years low grade soiling: girls 53%, boys 64%). Boys with spinal cord malformation with NBD in combination with prostatic/bladder neck fistula (PRF/BNF) and sacral agenesis had the worst functional outcome with minimal possibility of improvement over time. Functional outcome in girls with NBD and tethered cord did not differ significantly from those without NBD and with a normal spinal cord.

Psychosocial co-morbidity, neuropsychiatric disorders, developmental delay and megarectosigmoid were also risk factors impeding the functional outcome.

Conclusion: In the present study there was a successive improvement in bowel function during childhood and adolescence in ARM children, but they did not achieve the level of healthy children. NBD, spinal cord malformation, sacral malformation and PRF all were negative predictive factors for bowel score at 5 years.

Abbreviations: ARM, Anorectal malformation; ADHD, Attention-deficit-hyperactivity-disorder; BF, Bulbar fistula; BNF, Bladder neck fistula; CIC, Clean intermittent catheterization; CHARGE, **C**oloboma-**H**eart-**A**tresia-**R**etardation-**G**enital anomalies-**E**ar anomalies; DD, Delayed development; MRS, Megarectosigmoid; NF, Non fistula; NBD, Neurogenic bladder dysfunction; PSARP, Posterior sagittal anorectoplasty; PRF, Prostatic fistula; PSARVUP, Posterior sagittal anorectovaginourethroplasty; PSS, Poor social support; TC, Tethered cord; VF, Rectovestibular fistula.

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Many reports over the years have described the outcome concerning bowel function in children with anorectal malformations (ARM), who have undergone reconstructive surgical procedures. There is great heterogeneity in reported results, probably due to the fact that the criteria used to evaluate long-term outcome have been quite variable. The results are often graded as good, fair or poor, but this does not mean that good results are equivalent to normal bowel function. A patient with ARM can be perfectly socially continent but still need supportive treatment, ie individualized bowel management.

In 2005, The Krickenbeck conference developed a classification based on surgical procedures, and anatomical and functional criteria to better compare functional outcome [1]. The definition of more precise criteria will allow for more uniform assessment and reporting of functional outcome. Hasset et al. are among the first researchers to report according to the Krickenbeck classification for both diagnosis of the presenting anomaly and assessment of long-term outcome [2]. However when comparing functional outcome in an individual over time, one might need to introduce a multivariate scoring method. Rintala and Lindahl established a scoring system as early as 1995 [3]. It enables a quantitative description of the patient's faecal continence and degree of soiling and constipation.

In a recent paper we recognized that ARM children with neurogenic bladder dysfunction (NBD) and spinal cord malformation had the worst outcome concerning both bowel and bladder function [4]. Previous studies, on the other hand, have identified status of the sacrum as a predictor of functional outcome in these patients [3,5,6]. Whether the spinal cord malformation is, in fact, the predictor of functional outcome and the sacral status a part of a more severe malformation, or if the sacral status alone has a predictive value, has not been conclusively shown.

In this study bowel function has been followed longitudinally in a group of children with ARM, excluding perineal fistula. The aim was to determine to what extent faecal continence can be achieved in relation to age and type of malformation, and evaluate risk factors for delay in this maturation process. Risk factors studied were associated NBD and spinal cord malformation, sacral dysplasia, MRS, psychosocial pathology and syndromes.

1. Patients

This material was based on a prospective study of children with ARM, excluding perineal fistulas, referred to the Department of Pediatric Surgery, the Queen Silvia Children's Hospital Gothenburg, Sweden, from 1995 to 2005. The study included 41 patients out of 45 referrals from the region. Seventeen boys had recto-urethral fistulas (12 prostatic (PRF), 5 bulbar (BF)), 1 boy had a bladder-neck fistula (BNF), and 16 girls had vestibular fistulas (VF). Of the remaining patients, 3 boys had no fistula (NF), 1 girl had no fistula and 3 girls had cloacal malformation. Two of the girls with cloacal malformation had a common channel greater than 3 cm (high cloaca), and 1 girl had a channel shorter than 1 cm (low cloaca). For further details see Borg et al. [4].

All patients were treated with a diverting colostomy within 48 h of birth, and anorectal reconstruction (posterior sagittal anorectoplasty, PSARP) was performed at a median age of 7 months (range 3–60).

Investigations were performed in each child in order to rule out associated malformations, vertebral malformations, bladder dysfunction and spinal cord pathology.

For number of patients with associated gastrointestinal, cardiac and urogenital malformations see Borg et al. [4]. Neurodevelopment delay was seen in five children (Down's syndrome 2, CHARGE 1, unspecific 2). Three boys were diagnosed with neuropsychiatric disorders (autism 1, ADHD 2).

Sacral and vertebral anomalies were common, seen in 25 of 41 patients. In 19 of the 25 children the abnormality involved the sacrum.

Thirty-seven out of 41 patients included in the present study were investigated with urodynamics before and after the PSARP procedure, and the results have been reported [4]. Bladder function was evaluated only after surgery in the remaining four. From this earlier study, neurogenic bladder dysfunction was identified in 9 patients (5 boys and 4 girls), 8 of which were found before PSARP. A spinal cord malformation was diagnosed in the eight children with neonatal NBD, whereas one girl with a cloaca had no spinal pathology, and she developed NBD post PSARVUP.

Spinal cord pathological conditions were routinely investigated using ultrasound performed in the neonatal period in 24 patients. The number of patients investigated was determined by the fact that the method was not in clinical use until 1999. Magnetic resonance imaging (MRI) of the spine was added in 14 patients owing to abnormal spinal ultrasound or urodynamics and in all cases of cloaca malformation.

Eight children (5 boys, 3 girls) had clear malformation of the spinal cord. These were the same children shown to have innate NBD according to the urodynamic investigations. In three of the five boys with spinal cord malformation, the distal medulla had a club-shaped abrupt ending (spinal cord regression); in one boy, an anterior lipomyelomeningocele and tethering of the cord were diagnosed; and in the remaining boy, a thick filum with tethering was found. All three girls with spinal cord malformation, had tethering, in one case accompanied by thick filum and fibrolipoma. In addition, one boy with PRF without NBD was found by MRI to have asymmetry of the conus and the nerve roots but without tethering.