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The association of the severity of anorectal malformations and intestinal malrotation

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ABSTRACT

Introduction: Intestinal malrotation is a known association of anorectal malformations (ARM). Exact incidence, prognosis and surgical implications related to ARM are unknown. The aim of this study was to identify relevant associations between ARM and the presence of malrotation.
Methods: Records of patients from two referral centers were retrospectively analyzed looking for malrotation associated to ARM and its management, as well as factors for functional prognosis.
Results: 40 patients out of 2572 with ARM (1.6%) were found to have malrotation. Females were more commonly affected, and severe malformations were more frequent (cloaca, covered cloacal exstrophy in females and rectoprostatic and rectobladder neck fistula in males). Factors significantly associated with malrotation included Müllerian or Wolffian duct anomalies ($P < 0.05$), while fecal continence status, presence of constipation, and use of laxatives or enemas were not. Detecting and correcting malrotation early on or at the time of colostomy creation represented a protective factor against additional surgeries for bowel obstruction and volvulus ($P < 0.001$). Removal of the appendix during malrotation treatment required constructing a neoappendicostomy using a cecal flap in 9 out of 14 patients needing antegrade enema administration.
Conclusions: Malrotation presence in patients with ARM has the same frequency as in the general population, but it is more common in severe malformations. Surgeons treating these patients should address the malrotation at the time of colostomy opening if detected. The appendix should be preserved for potential future use as an appendicostomy for antegrade administration of enemas.

Intestinal malrotation is a known association of anorectal malformations (ARM): as high as 8.5% of the patients have been reported in case series in the literature [1–5]. However, exact incidence, prognostic factors, and surgical implications of this association have not been well established. Accepted standard of practice in the management of intestinal malrotation includes surgical intervention if identified on radiologic examinations even if there is absence of symptoms [3,6]. This proactive treatment has been previously described in patients with ARM, but the clinical impact, if any, is not known. The aim of this study was to identify any relevant clinical associations between malrotation and ARM and to describe our experience with potential implications in diagnosis, surgical management and functional prognosis of this cohort of patients.

1. Methods

IRB approval was obtained in the respective collaborator centers (CCHMC IRB #2014–5021, Seattle IRB #15,069). A retrospective review of the patients' clinical charts was performed. Selection was made based on presence of anorectal malformation, diagnosis and type of treatment for malrotation. Data collected included: age, gender, type of ARM, report of intestinal malrotation and its treatment, any diagnostic imaging available for malrotation, surgical reports and time of follow up. Cloaca exstrophy patients were excluded from analysis because this is a very distinct, separate malformation and they have an inherently higher risk of intestinal rotation anomalies if associated omphalocele is

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present. We did include covered cloaca exstrophy since this anomaly is distinctly different from cloaca exstrophy. Statistical analysis utilized Fisher's exact test using the SAS Statistical Software (® SAS Institute Inc., NC).

2. Results

2.1. Anorectal malformations

Among 2572 patients with ARM treated in our centers, 40 were identified to have malrotation (1.6%). There were 24 females and 16 males (1.5:1 female to male ratio). In the female cohort, the most commonly identified malformations were cloaca and the variant known as covered cloacal exstrophy, (18/699) followed by rectovestibular fistula (5/411) and ARM without fistula (1/19). ARM in males included in order of frequency, rectoprostatic fistula (6/309) rectobulbar fistula (4/296), rectobladder neck fistula (4/240), one patient with an unknown type of fistula (his primary surgery was performed at another institution and the location of the fistula was not identified) and covered cloacal exstrophy (1/16) (Fig. 1).

Patients with a more severe or complex malformation, defined as those needing significant mobilization of structures (vagina, rectum, urinary tract) as well as the potential need for extensive separation of structures, showed a statistically significant difference for the presence versus absence of intestinal malrotation ($p \leq 0.01$). This group included such malformations as cloaca, covered cloaca exstrophy, and bladder/bladderneck and prostatic fistula. Of the five patients with rectovestibular fistula and malrotation, two had documented Müllerian structure anomalies, which included vaginal atresia and absence of fallopian tubes in one patient and atretic fallopian tubes and a left streak gonad in the second one. In the remaining 3 patients, there was no identified anomaly aside from the ARM. Any patient with identified Müllerian or Wolffian duct anomalies also showed a statistically significant difference for the presence of intestinal malrotation ($n = 26$ vs $n = 14$, $p < 0.05$).

2.2. Other associated malformations

38 out of 40 patients had, at least, one VACTERL association in addition to the anorectal malformation. Urologic anomalies were the most commonly observed in 23 patients, followed by cardiac defects in 20.

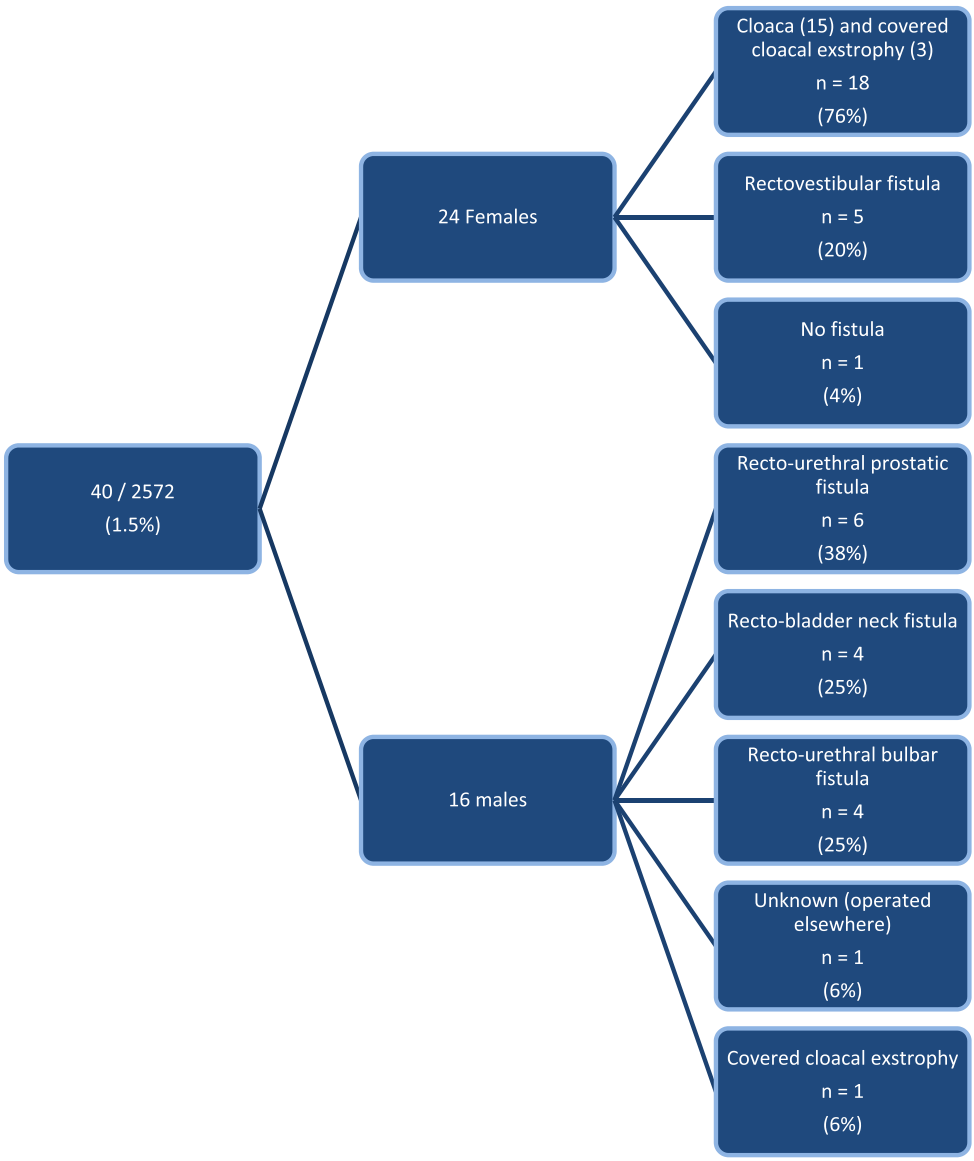

Fig. 1. Distribution of anorectal malformations with associated intestinal malrotation.