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Covered cloacal exstrophy – a poorly recognized condition: Hints for a correct diagnosis

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Abstract

Introduction: Covered cloacal exstrophy requires a high index of suspicion for its diagnosis. Low implantation of the umbilical cord, separated pubic bones, and anorectal malformation are the most common signs.

Methods: Thirty-one patients with this defect were retrospectively analyzed.

Results: Besides the anorectal malformation, the patients had important unique anatomic findings, including a colon shorter than 20 cm (17 patients) and absent bladderneck (27 patients). Twenty-four patients underwent a colonic pullthrough; of those, only 5 of them have voluntary bowel movements. Twelve patients underwent a urinary reconstruction. Eleven of them are dry with catheterization, and one leaks in between catheterization. Two patients are urinary continent.

Conclusions: Covered exstrophy is a serious condition. Externally, the patients may look like having a rather simple malformation. However, the intra-abdominal findings are similar to those seen in cloacal exstrophy. An early correct diagnosis is important to plan a reconstructive strategy and to adjust the parent's expectations concerning bowel and urinary function. In addition to the traditional prognostic factors for bowel and urinary control (sacral ratio, tethered cord, and level of the rectum) these patients have other anatomic defects (absent bladderneck and short colon) that negatively affect the functional prognosis.

Covered cloacal exstrophy is frequently misdiagnosed, leading to negative therapeutic and clinical implications. The correct diagnosis requires a high index of suspicion since the external signs are rather inconspicuous. A low implantation of the umbilical cord in association with separated pubic bones and an anorectal malformation are the most frequent signs (Fig. 1). These signs must alert the surgeon to the fact that the patient has a more serious condition that includes an absent bladderneck (which will require urologic reconstruction) and different degrees of colonic abnormalities (short colon) similar to those seen in cloacal exstrophies, which negatively affect the functional prognosis for bowel control.

1. Methods

A database review was performed and 31 patients were identified with covered cloacal exstrophy. Their anatomic findings were divided into gastrointestinal, urological,

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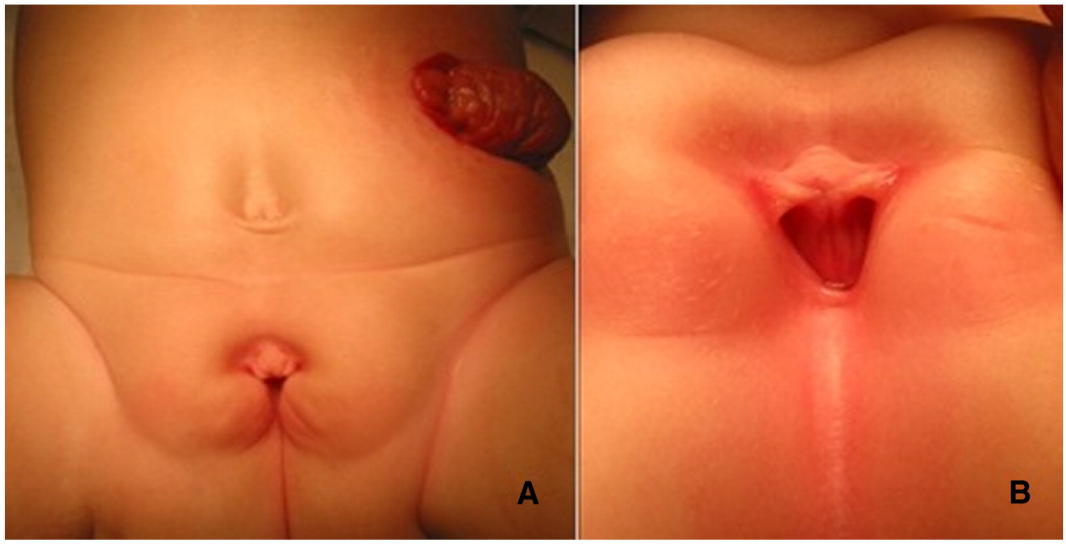

Fig. 1 Classical findings of a covered cloacal exstrophy: low implantation of the umbilicus (A), single large perineal orifice, separation of the pubic bones, and imperforate anus (B).

gynecological, skeletal, and neurospinal. Urological and colorectal follow up were retrospectively analyzed.

2. Results

Thirty patients were female and 1 patient was male. In 30 patients initially seen at other institutions the diagnosis of a covered cloacal exstrophy was missed.

In eight patients a rescue operation had to be performed consisting in rescuing a short piece of colon that was left connected to the urinary tract, to incorporate it into the fecal stream and to create an end colostomy.

Upon inspection of the perineum the two most common findings were: a large single orifice (Figs. 1B and 2) present in 17 patients, and 4 perineal orifices very close to each other (Fig. 3) present in 11 patients.

1. Gastrointestinal anatomic findings:
   - Colonic length: 17 patients had an absent colon or a colonic length shorter than 20 cm, 14 patients had a colonic length greater than 20 cm or normal;
   - Rectal level: 17 patients had a "low" lying rectum (vestibular fistula [10], low implantation in a cloaca [5], perineal fistula [2]), 14 patient had the rectum located at the bladderneck level or above;
   - 10 patients had segmental colonic duplication or duplication of the appendix;
   - Other findings: malrotation (9), omphalocele (8), Meckel's diverticulum (4), ileal atresia (3), esophageal atresia (1), and patent omphalomenseteric duct (1).
2. Urological anatomic findings:
   - 27 patients had an absent bladderneck and 4 patients had a normal bladderneck;
   - 8 patients had a single kidney;
   - 7 patients had vesico-ureteral reflux;
   - Other findings: hypospadias and left undescended testicle in the male patient, bilobulated bladder (3), megacystis (3), absent urethra (2).
3. Gynecological anatomic findings:
   - Normal single clitoris in all female patients (30);
   - 20 patients had 2 hemivaginas and 2 hemiuteri, 3 patients had absent vagina, 3 patients had atresia on one side of the Mullerian structures, 2 patients had a single vagina and single cervix and in 2 patients the status of the vagina was unknown.

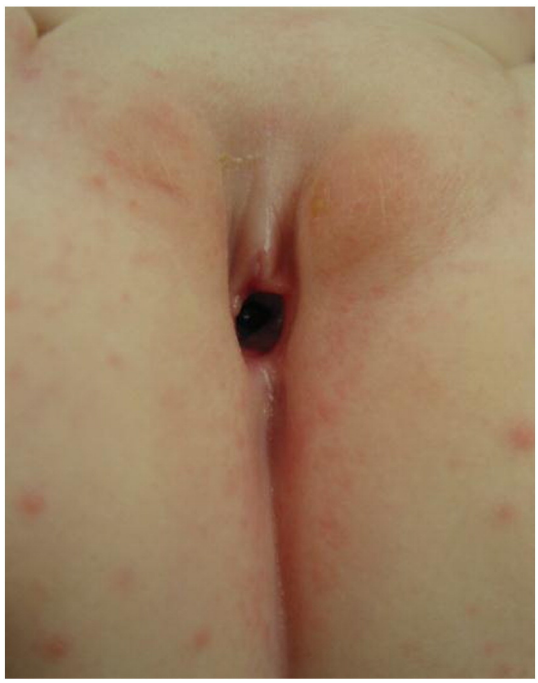

Fig. 2 Single large perineal orifice.