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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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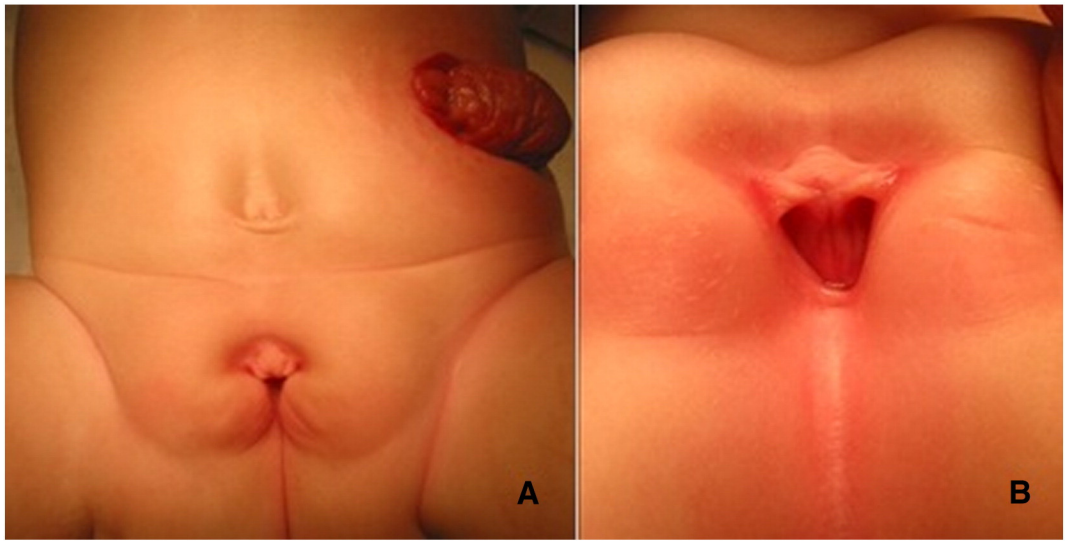

Fig. 1 Classical findings of a covered cloacal exstrophy: low implantation of the umbilicus (A), single large perineal orifice, separation of the pubic bones, and imperforate anus (B).

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.

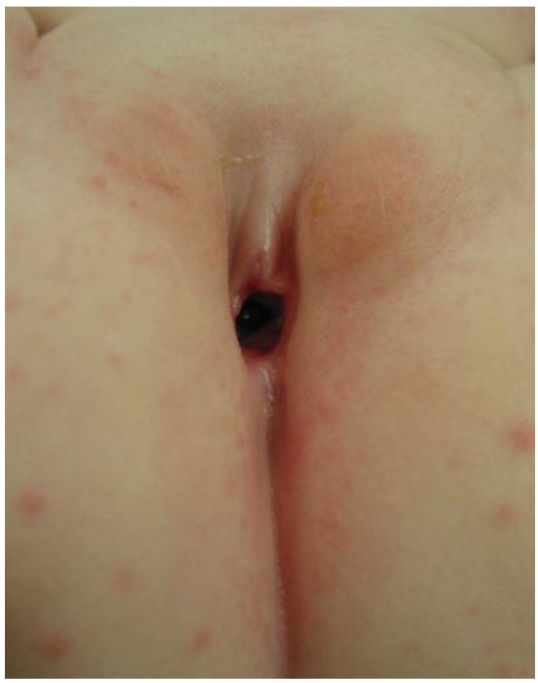

Fig. 2 Single large perineal orifice.

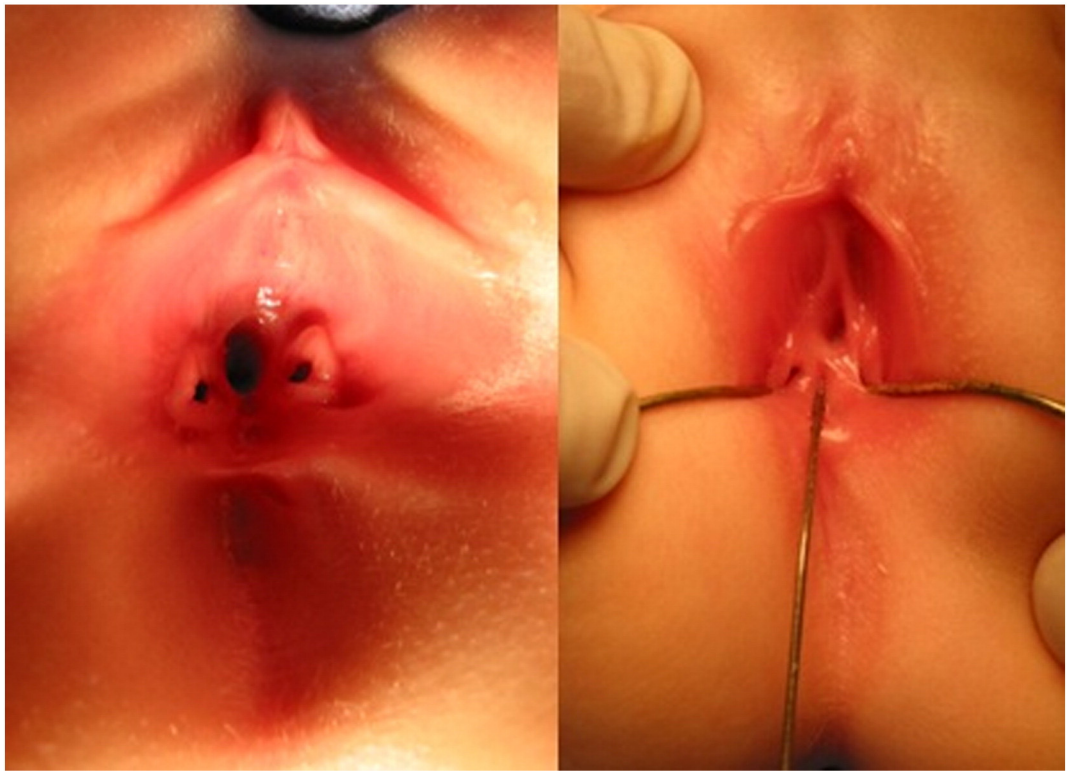

Fig. 3 Two different patients showing four perineal orifices close to each other representing anterior urethra, hemivaginas on each side and posterior rectum.

4. Skeletal anatomic findings:
   - 29 patients had separation of the pubic bones;
   - The sacrum was normal in 18 patients, the sacral ratio was <0.4 in 8 patients, and no sacral information was available in 5 patients.
5. Neurospinal anatomic findings:
   - 8 patients had tethered cord, 2 patients had no tethered cord and in 21 patients the status of the cord was unknown.

Follow up information was available in 27 patients:

Twenty-four patients had undergone a pullthrough procedure, 13 are clean with bowel management, 5 are fecally continent, 3 have a temporary colostomy, 2 have permanent stomas owing to an extremely short colon and incapacity to form solid stool, and 2 patients died.
Thirteen patients underwent urologic reconstruction, 11 patients are dry on intermittent catheterization (8 of them had bladder augmentation), 1 patients is urinary continent, and 1 patient leaks urine. One patient is urinary continent and did not require urological reconstruction. Thirteen patients are waiting for urological reconstruction and there was no information for 2 patients.

3. Discussion

Not many articles have been published about covered cloacal exstrophy [1–11], probably because it is a rare disease, and since patients may present with different anatomic variants, it makes it hard to group or classify them. In this series and for the purpose of this review we excluded patients with a bladder exstrophy component.

The fact that 30 patients were previously seen at another institution and the diagnosis was missed is perhaps explained by the fact that the external appearance of the abdomen and perineum of these patients may not be very conspicuous. It is understandable how the diagnosis is missed if the clinician is not looking for specific external, rather subtle abnormalities. The low implantation of the umbilical cord may not be very obvious unless we observe the patient carefully (Fig. 1A).

The separation of the pubic bones can be easily seen on any radiographic film. However, without the help of the radiologic study, one must look for two mild prominences away from the midline in the pubic area (Fig. 1B). Palpation of the area demonstrates a fibrous band in between the separated pubic protuberances.

Finally, the presence of a rather large perineal orifice, through which one can see a constant dribbling of urine (Fig. 1B) represents an important sign to establish the diagnosis.

Interestingly, in spite of the separated pubic bones, all the patients had a single clitoris in our series.

We consider this malformation as part of the cloacal exstrophy spectrum, even when the majority of the patients (23) had an intact abdominal wall; the intraabdominal anatomic abnormalities resemble those observed in cases

of cloacal exstrophies. The absence of bladderneck is usually present together with a very small bladder, therefore the great majority of these patients will require a major urinary reconstruction with bladder augmentation and Mitrofanoff to remain dry of urine.

From the bowel control point of view, it is a fact that an absent colon means incapacity to form solid stool. In patients with normal sphincters and a normal anal canal it is conceivable to do an ileo-anal anastomosis and still achieve bowel control, like in cases of total colonic aganglionosis, familiar polyposis or ulcerative colitis. However, in cases with anorectal malformations, that type of operation is contraindicated; it will provoke fecal incontinence and an extremely severe diaper rash, since these patients do not have anal canal and their sphincter mechanism is underdeveloped or not present. We have also learned that the bowel management with daily use of enemas designed to keep incontinent patients artificially clean is not feasible in patients with liquid stool. Therefore, incapacity to form solid stool (patients with extremely short colon or absent colon) in cases of anorectal malformation is considered as an indication for a permanent stoma. Since the colonic length in patients with cloacal exstrophy and covered cloacal exstrophy represents a spectrum, patients must be judged on an individual basis [12].

A pullthrough is not performed unless we have evidence that the patient is capable of forming solid stool [12], like in those cases with normal colonic length. In the other extreme of the spectrum are patients with no colon; those receive a permanent stoma.

The majority of cases however are in the middle of the spectrum. In other words, they have different degrees of short colons. In those patients we recommend to open an end colostomy, putting special emphasis on not leaving any piece of colon attached to the urinary tract, as we have previously recommended in cases of cloacal exstrophy [13]. We are convinced that little pieces of colon may experiment a significant growth, provided they are incorporated into the fecal stream. Pieces of colon left isolated, defunctionalized or attached to the urinary tract will not grow and may absorb urine producing hyperchloremic acidosis.

When there is doubt concerning the capacity of a short colon to absorb enough water to form solid stool and therefore respond to a bowel management with daily enemas, we implement our enema program through the stoma [12,14–16]. Our goal is to have an empty stoma bag for 24 hours which is considered a successful bowel management. In such cases, we are able to offer the family a pullthrough of the colostomy, provided they understand that the patient will receive the same enema that was successfully used through the stoma but through the rectum or via an antegrade route (appendicostomy or neo-appendicostomy).

Concerning the urinary reconstruction we firmly believe that it must be postponed until we have defined the final bowel control status of the patient. In other words, it is very important not to make decisions to reconstruct the urinary tract, involving pieces of bowel (augmentation, Mitrofanoff, Monti) until we have established if the patient is going to have a pullthrough or is going to have a permanent stoma.

4. Conclusion

Covered exstrophy is a serious condition. An early correct diagnosis is important to plan a reconstructive strategy and to adjust the parent's expectations concerning bowel and urinary function. Traditional prognostic factors for bowel and urinary control (sacral ratio and level of the rectum) do not apply to this anomaly, since these patients have other defects (short colon and absent bladderneck) that can negatively affect the prognosis.

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