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(RESEARCH ARTICLE)

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Cloacal malformations in the pediatric surgery department of Fez

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Abstract

Introduction: Cloacal malformations, which exclusively occur in females. These anomalies are rare. The immediate prognosis depends mainly on the severity of the associated malformations and relies on the quality of the surgical repair.

Patients and methods: This is a retrospective descriptive study of 5 patients managed in the pediatric surgery department of the Hassan II University Hospital of Fez, during a 13-year period from January 2008 to January 2021.

Results: The diagnosis is made postnatally following systematic clinical examination in the delivery room, with only one case presenting with intestinal obstruction. The malformative assessment revealed: One case with cardiopathy, abnormal renal ultrasound findings in 3 cases (predominantly uretero-pyelocaliceal dilation), and spinal anomaly found in 2 cases, and abnormal MRI findings in 3 cases, while not performed in 2 cases.

Four contrast studies were performed after colostomy, revealing 2 rectovesical fistulas and 2 bilateral VUR (vesicoureteral reflux).

Immediate postoperative course was uneventful, with no dehiscence, stenosis, or obstruction noted. One patient died due to cardiac decompensation at 3 months of age. Long-term evolution was characterized by the presence of anal incontinence.

Conclusion: Cloacal malformation is a serious pathology, it can frequently lead to functional and vital prognosis without forgetting the psychological one. The prognosis depends mainly on the quality of management and the presence of associated malformations.

Keywords: Cloacal malformation; Surgery; Management; VACTERL; Anal incontinence

1. Introduction

Cloacal malformations, which exclusively occur in females, represent the most complex anomaly within the spectrum of anorectal malformations. These malformations are rare, with an estimated frequency of 1 in 20,000 live births. The immediate prognosis depends mainly on the severity of associated malformations, while the long-term functional prognosis depends much more on the length of the common channel and the quality of surgical repair.

In this present study, we aimed to share our experience in the management of this affection.

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2. Material and methods

This is a retrospective descriptive over 13-years (January 2008 to January 2021). Were included all the children admitted and managed for cloacal malformation during our period of study. We record and analyzed clinical aspects, imaging finding and complications. These parameters was evaluated in the short mid and long term.

3. Results

The diagnosis is made postnatally following systematic clinical examination in the delivery room, with only one case presenting with intestinal obstruction. All of our patients underwent colostomy, with four on the right side and only one on the left side. One patient underwent vesicostomy after 8 days. After performing cardiac ultrasound on all of our patients as part of the malformative assessment, one case presented with a large ventricular septal defect and minimal coarctation of the aorta with agenesis of the coccyx.

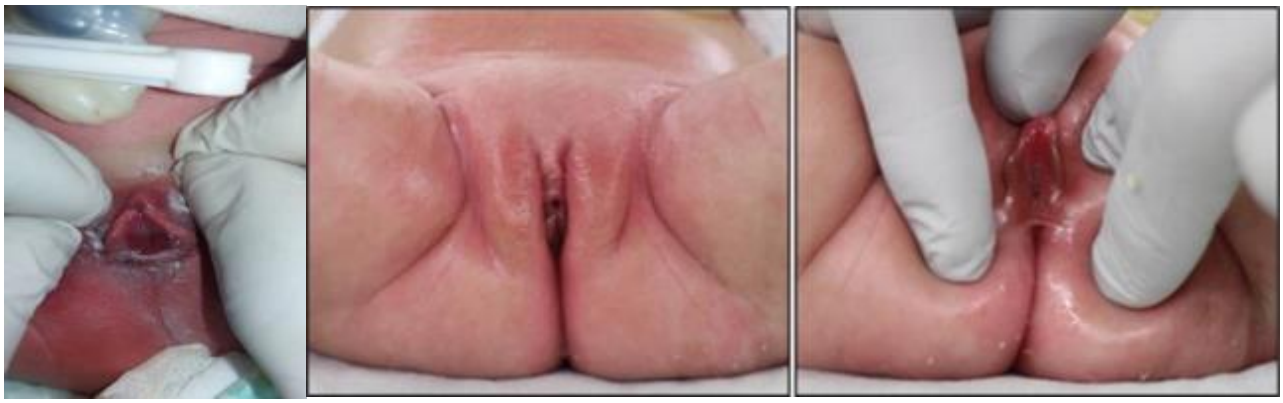


Figure 1, 2 and 3 Clinical images showing a single orifice

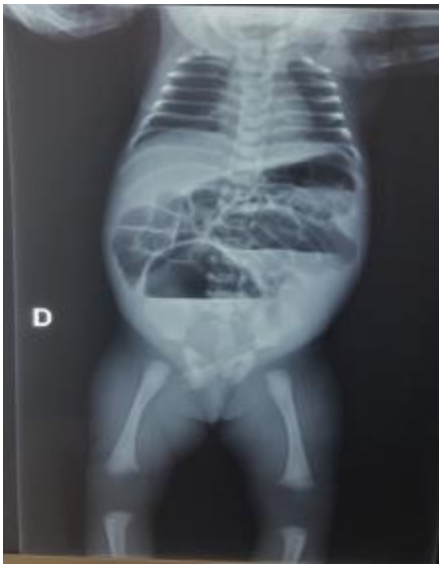


Figure 4 X-ray showing air-fluid levels suggestive of intestinal obstruction

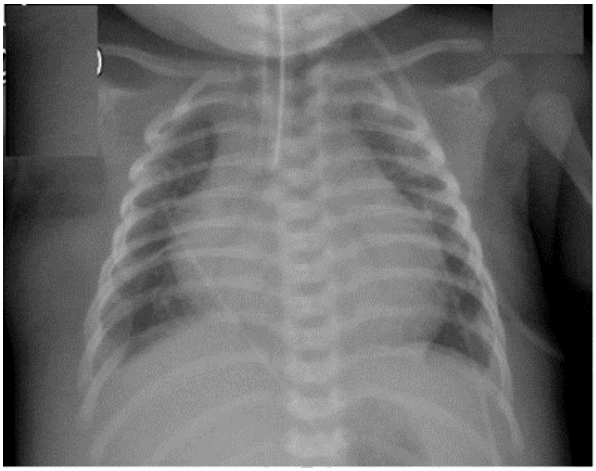


Figure 5 Chest X-ray showing cardiomegaly, suggesting a cardiac malformation confirmed by ultrasound

Additionally, renal ultrasound was abnormal in 3 cases (predominantly uretero-pyelocaliceal dilation), and MRI was performed on 3 patients to plan the surgical procedure, revealing anomalies such as hydrocolpos, bicornuate uterus, bicornuate cervix, didelphys uterus with double vaginal cavities, rectovesical fistula, and duplicated left renal system. Four opacifications were performed after colostomy, revealing 2 rectovesical fistulas and 2 bilateral VUR (vesicoureteral reflux).

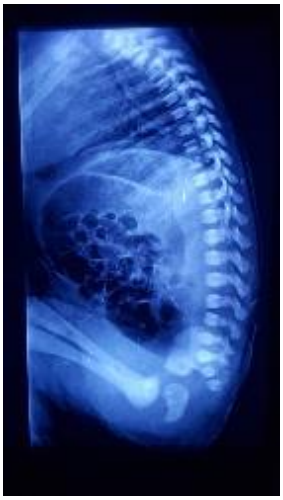


Figure 6 X-ray showing coccygeal agenesis

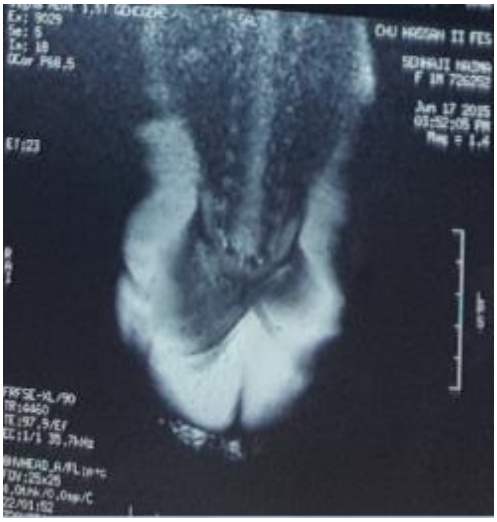


Figure 7 Double left urinary system observed on MRI

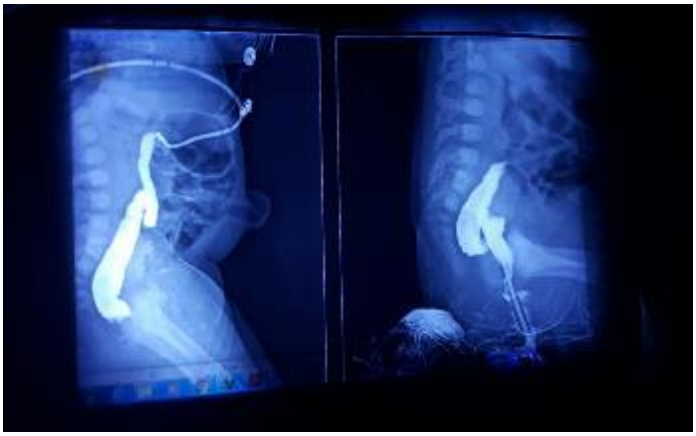
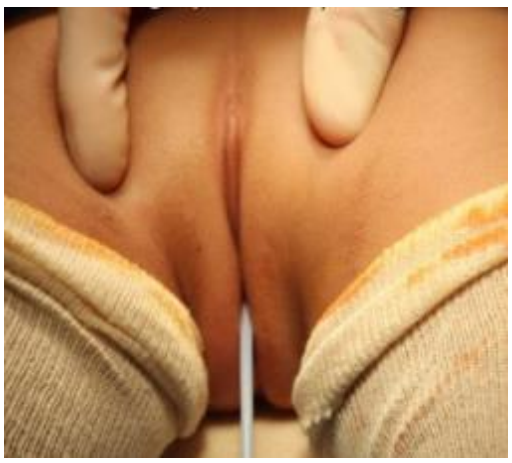


Figure 8 Rectovesical fistula after opacification

The radical repair is performed with an average delay of 9 months using the Peña technique, which involves a posterior sagittal anorecto-vagino-urethroplasty (PSARVUP). Only one case required an abdominal approach because the common channel was beyond 3 cm. A urinary catheter is placed for 2 to 3 weeks, and dilations are initiated on the tenth day.



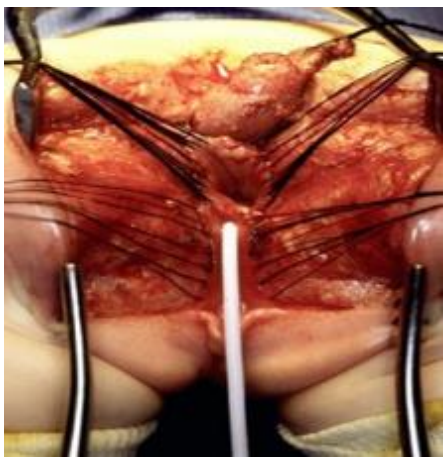


Figure 9, 10 Perioperative images showing the technique of PSARVUP

The colostomy closure is performed after achieving adequate anal and vaginal dilation, combined with vesicostomy closure in one case. Two patients underwent reimplantation using the Cohen technique for vesicoureteral reflux as part of the management of associated malformations.

Immediate postoperative course was uncomplicated, with no dehiscence, stenosis, or obstruction noted. One patient died due to cardiac decompensation at 3 months of age. Long-term outcomes were characterized by the presence of anal incontinence in 3 girls, managed through self-rehabilitation after parental education or biofeedback for girls older than 6 years, with minimal improvement in encopresis.

Vaginoscopy was performed in 2 patients, revealing vaginal stenosis in the middle third of the vaginal canal in one case, and the other presented with a vesicouterine fistula.

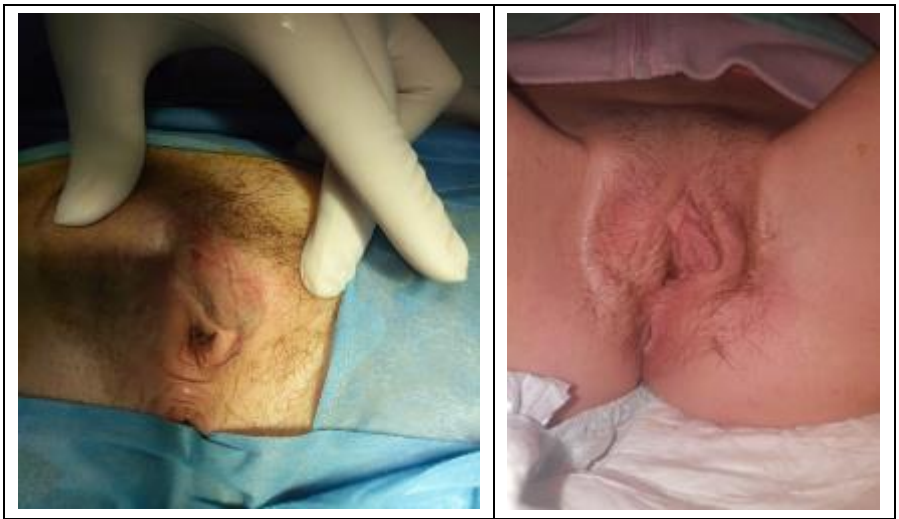


Figure 11, 12 Esthetic aspects after final repair

4. Discussion

Cloacal malformations are rare congenital anomalies characterized by a confluence of the gastrointestinal, genital, and urologic systems which exits the perineum through a single orifice [1]. The etiology of these conditions remains unknown. The definitive diagnosis of cloacal malformation is made in the newborn period by physical examination with the identification of a single perineal orifice, usually posterior to the clitoris. Abdominal examination may divulge the presence of a mass in the lower abdomen which likely represents a hydrocolpos which can be present in as many of 40% of patients with a cloaca [2].

Initial evaluation of a patient with a cloacal malformation involves the identification and treatment of any associated life-threatening anomalies. Like other anorectal malformations, cloacas often correlate with various anomalies in the vertebral, anorectal, cardiac, tracheoesophageal fistula, renal, and limb (VACTERL) association [3, 4].

A thorough urologic assessment is mandatory in the newborn period [5]. Structural and functional urologic anomalies, including hydronephrosis, renal agenesis, renal dysplasia, horseshoe kidney, are common in patients with cloaca and are found in up to 90% of cases [1,6,7].

Identification of spinal anomalies is necessary as there is a high association of cloaca and tethered cord [8].

An assessment of the sacrum can be performed with anteroposterior (AP) and lateral sacral x-rays to assess for sacral anomalies and determine the sacral ratio [9]. The sacral ratio is the length of the sacrum from the sacroiliac joint to the tip of the coccyx over the distance of the iliac crest to the sacroiliac joint.

After stabilizing the child, early surgical management aims to divert the fecal stream with a descending colostomy with mucous fistula creation. Urinary diversion can be performed at the time of colostomy creation with a vesicostomy or placement of a suprapubic cystostomy tube [5].

Definitive surgical repair requires the identification and measurement of several pelvic structures, including:

Length of the common channel, length of the urethra, the presence and length of a vagina, the presence of a hemivagina or longitudinal vaginal septum, the presence of a cervix (or multiple cervices),visualization of the height of the rectum and location of the rectal fistula relative to the pubococcygeal (PC) line [10].

The length of the common channel and urethra are critical for surgical planning (e.g., to decide whether to perform a urogenital separation or total urogenital mobilization). The 3 cm common channel is traditionally taught as the cutoff between simple and complex cloacas [10]. The presence and length of the patient's native vagina are necessary to determine the likelihood of needing to perform a vaginal replacement at the time of PSARVUP (post-sagittal anorecto-vagino-urethroplasty).

No standardized imaging protocols exist at present, but endoscopy and 3D cloacagram are two useful modalities to

assess the patient's anatomy [11]. Some authors have suggested that the role of MRI will probably become more important in the future [12].

The definitive management of cloaca is surgical, with the goal of separating the gastrointestinal, gynecologic, and urologic structures and creating a perineal opening for each of the three structures for optimization of the function of each system, and to create a catheterizable urethra [1,13,14].

The surgical management of this condition has evolved significantly over the last several decades. In 1982, Peña first described the posterior sagittal approach for the repair of a cloacal malformation; an operation termed the posterior sagittal anorecto-vagino-urethroplasty (PSARVUP) [15]. The technique described separating the vagina from the common channel using a technique called a urogenital separation. The fine dissection in the plane between the vagina and urethra often resulted in devascularization of the tissue resulting in urethrovaginal fistula, which can be a very debilitating complication of urogenital separation.

In 1997, the total urogenital mobilization (TUM) was described as a technique to mobilize the vagina and urethra as a single unit, thereby obviating the need to mobilize the tissue between these two structures [16]. This procedure, however, risks leaving the patient with a urethra which is too short and a bladder neck which is below the urogenital diaphragm, resulting in urinary incontinence [17]. Wood, therefore, proposed an algorithm in 2016 which considers both the length of the common channel and native urethra to leave the patient with a urethra of adequate length to maximize urinary continence [10]. Definitive repair is typically performed between 6 and 12 months of age.

Urinary and bowel continence in patients with cloacal malformations are related primarily to the length of the common channel and the presence of other anomalies, in particular, the degree of sacral development and the presence of tethered spinal cord. Patients with a common channel of greater than 3 cm, tethered cord, and a sacral ratio of less than 0.4 tend to have worse functional outcomes [18]. Some patients with poor colorectal and urologic function may require bowel and bladder reconstruction later in childhood which can be performed simultaneously [19].

A Foley catheter placed intraoperatively remains in place for 2 to 4 weeks. Patients should undergo an examination under anesthesia (EUA) approximately 4 weeks after PSARVUP. The Foley catheter can be removed at the time of cystoscopy to ensure that the urethra can be accessed for intermittent catheterization as needed [7].

The anus should be examined to ensure adequate healing of the perineal body and the anoplasty. If both structures well healed, the anus can undergo sizing with a Hegar dilator, and a twice-daily dilation program started with the goal to increase the size of the dilator every week to a final measurement determined by the patient's age. Upon reaching the desired size, the colostomy can be closed [14].

A bowel regimen should be started at the time of colostomy closure to assist colonic emptying regularly and to avoid dilation of the rectosigmoid colon which will impair normal colonic function and make potty training difficult. If the patient is unable to potty train, a formal bowel management program may be needed to determine a laxative or enema regimen that can get the child clean and in normal underwear.

Our results are consistent with what is described in the literature regarding the high frequency of anal incontinence with or without soiling. Studies conducted by the authors have revealed a high frequency of incontinence, especially among patients with sacral or coccygeal agenesis.

Gynecologic counseling should take place before puberty with consideration of the patient's unique gynecologic anatomy. An adequate vagina that allows menstrual egress should be ensured prior to the start of menses. Preliminary to the onset of sexual activity, the patient should be examined to ensure the vagina is adequate for penetrative vaginal intercourse. Pregnancy is possible for patients with the cloaca, although Cesarean sections are the preferred method of delivery [20].

5. Conclusion

Cloacal malformations are rare, multi-system anomalies which require management by a team of an interprofessional specialists including nurses [21]. A quick transfer should be made as soon as the diagnosis is established. Managing anorectal malformations (ARM) requires long-term monitoring of patients, often referred to as "patients for life," and the necessity of raising awareness among parents for better collaboration.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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