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Cloaca reconstruction: a new algorithm which considers the role of urethral length in determining surgical planning

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ABSTRACT

Background: Cloacal malformations represent a uniquely complex challenge for surgeons. The surgical approach to date has been based on the common channel (CC) length with two patient groups considered: less than or greater than 3 cm, which we believe is an oversimplification. We reviewed 19 patients, referred after surgery done elsewhere. Eight had postoperative urinary complications, 3 had constant urinary leakage and had been left after surgery with a urethra <1 cm, .5 with an original 3 to 5 cm common channel, who had undergone total urogenital mobilization (TUM), experienced peri-operative urethral loss needing a vesicostomy, and later, a Mitrofanoff. These patients together with a review of the cloacal and urological literature led us to design a new algorithm where urethral length is a key determinant for care.
Methods: We prospectively collected data on 31 consecutive cloaca patients referred to our team (2014 to 2016) and managed according to this new protocol. The CC length, urethral length, surgical technique employed, and initial outcomes were recorded.
Results: Of 31 primary cases, CC length was 1 to 3 cm in 20, 3 to 5 cm in 9, and greater than 5 cm in 2. In the 1 to 3 cm and the 3 to 5 cm groups, a urethra less than 1.5 cm led us to perform an urogenital separation. We only performed a TUM if the urethra was greater than 1.5 cm. Using this protocol, we performed a urogenital separation in 1 of 20 in the 1 to 3 cm CC group, 6 of 9 in the 3 to 5 cm CC group, and 2 of 2 in the greater than 5 cm CC group. Seven patients underwent separation, who with the previous approach, would have had a TUM. Thus far, no urinary leakage or urethral loss has occurred in any patient, but follow-up is less than 3 years.
Conclusion: Urethral length appears to be a vitally important component in cloacal reconstruction. A short urethra left after repair can lead to urinary leakage. A TUM done under the wrong circumstances can lead to urethral loss. We describe a new technical approach to cloacal repair which considers urethral length but recognize that long term urological outcomes will need to be carefully documented.
Type of study: Clinical cohort study with no comparative group.
Level of evidence: Level 4.

The significant challenge for surgeons repairing cloacal malformations is created by the need to correct a common orifice of the urologic, gynecologic and gastro-intestinal tracts into three reconstructed perineal openings. The goal of the operation is to separate the rectum from the common urogenital tract and place it within the muscle complex to aid fecal continence. In addition, the reconstruction of the urogenital tract should be done in such a way as to promote urinary continence, long term renal health, sexual function and potential fertility. The understanding of both the anatomy and the technical aspects of the repair of cloacal malformations have advanced greatly since the initial attempts at repair, when all patients underwent urogenital separation [1]. The advent of the total urogenital mobilization (TUM) in 1997 [2], was a fundamental step in this development. In addition, the introduction of the cloacagram [3,4], brought further clarity to the anatomy. Based on this literature, the surgical strategy has been guided by the length of the common channel alone [2,5], with patients divided into 2 groups: short (less than 3 cm) and long (greater than 3 cm) common channels [2,3,5,6].

We believe that this approach is an oversimplification of the anatomic problem and that urethral length must also be considered. When examining the large reported series of cloacal repairs, no mention is made of long-term functional bladder outcomes. [5,6] A careful

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[2] critical revision of manuscript.
[3] study conception and design, critical revision of manuscript.

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review of the literature reveals that rates for urinary incontinence are high; 26% in the 1 to 3 cm group (N = 225) and 72% in the greater than 3 cm (N = 175) [5]. In another large series, only 22% (N = 50) voided spontaneously, 12% were kept dry with clean intermittent catheterization (CIC), 46% required urologic reconstruction and 20% remained incontinent [14]. While there are a few other reports of preserved urinary continence after TUM, these are based on very few patients [9,15] and no validated outcomes tools were used.

In addition to this literature review we also retrospectively analyzed the medical records of 19 patients with a cloacal malformation referred to us for evaluation and care after their definitive reconstruction had been done elsewhere. We recognize that this is a unique referral group and that we do not know the denominator of all repaired cloaca patients. Eight of this group had a postoperative urinary complication. We focused our review on the length of the common channel as well as the surgical techniques employed at the time of their repair and length of urethra they were left with after surgery. In 3 of the 8 patients, constant urinary leakage was present, and we found that they had a short urethra (<1.0 cm), after initial TUM, a situation whereby their bladder neck was too close to their perineum. This persistent leakage required bladder neck tightening (n = 1) or permanent closure plus mitrofanoff (n = 2). The other 5 patients had been operated for a common channel of 3 to 5 cm and experienced intra or postoperative urethral loss. In each case a total urogenital mobilization (TUM) was initially performed but was converted to a urogenital separation during the procedure. In all cases, because the TUM did not reach and a urogenital separation was performed after the TUM, the urethra had both an anterior and posterior dissection. This led to urethral loss that we ascribe to a diminished blood supply. All of these patients were converted to a vesicostomy and later a Mitrofanoff for intermittent catheterization.

Based on the literature review and our analysis of referred patients, we were led to consider another approach. A review of the urology literature showed us that urethral length is well described as a fundamental part of decision making and surgical planning in the repair of a urogenital sinus with or without congenital adrenal hyperplasia. [8,9] Not only does this factor into the classification system introduced by Rink et al. [10] but it is thought to be the single most important factor in determining surgical complexity and clinical outcomes for such patients [8]. This concept, we believe, should have similar application to cloaca surgery which has similar urethral challenges.

The length of the urethra is relevant in the following situations: (1) In the group of patients in whom the common channel length is short, less than 3 cm in length, but who in addition, have the rare association of a short urethra (1.5 cm or less). (Fig. 1) (2) In patients with a mid-length (3 to 5 cm) common channel who also have a short urethra, (less than 1.5 cm).

A new algorithm for the care of these patients is shown in Fig. 3. Our hypothesis was that the urological morbidity could be reduced with improved patient selection for total urogenital mobilization (TUM) based on urethral length and not only common channel length.

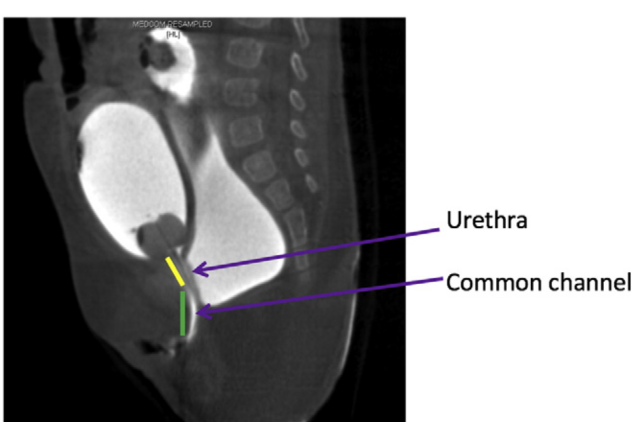

Fig. 1. Short common channel with inadequate urethra.

1. Methods

We prospective collected data on 31 primary cloaca patients referred to our multi-disciplinary team between April 2014 and July 2016 for repair. We recorded the urethral length, common channel length, the operative technique employed and outcomes on all patients.

All patients had an examination under anesthesia in which the child undergoes a careful physical examination, a cystovaginoscopy, examination of the perineum, 3D cloacagram and urodynamics. We followed a standardized data collection during the cystovaginoscopy in which we assessed the gynecologic structures (number of vaginal cavities, presence of a vaginal septum, vaginal length from common channel confluence), the urological structures (bladder capacity, urethral length, bladder neck function, location of ureteral orifices, presence of ectopic ureters, presence or absence of cystitis cystica) and colorectal structures (insertion of the rectum into the common channel). This information is complemented by a 3D cloacagram that is performed during the same anesthetic which gives us an improved spatial understanding of the cloacal defect by distending its hollow structures. Further measurements of urethral length, bladder distention and the existence of vesicoureteral reflux and position of the rectum in relation to the sacrum and coccyx were also collected from the 3D cloacagram. The examination was performed by the same surgical team that performed the operative repair. Having urology, gynecology and colorectal surgery at the assessment gave the whole team first-hand information which aided surgical planning.

The surgical approach was determined by the algorithm we have designed, that can be seen in Fig. 3.

2. Results

In the 31 consecutive primary cloaca patients who presented for repair to our team during the 2 years (April 2014 and July 2016), there

Table 1
Cloaca series; demographic, anatomic and surgical summary.

Patient	Age in years	Common Channel length	Urethral length	Procedure
1	0.9	1.9	2	TUM
2	0.8	2.4	3	TUM
3	2.2	3.2	2.4	TUM
4	1.1	2.1	3.1	TUM
5	1.3	2.0	1.5	TUM
6	0.9	2.0	1.8	TUM
7	0.7	1.0	1.3	TUM
8	0.9	2.8	2	TUM
9	0.5	1.2	3.	TUM
10	9.2	1.2	2.5	TUM
11	1.3	2.1	2.5	TUM
12	0.7	2.2	1.8	TUM
13	3.7	2.9	2.8	TUM
14	1.5	2.1	2.1	TUM
15	2.6	1.1	2.4	TUM
16	2.6	2.3	2.5	TUM
17	2.1	2.9	1.6	TUM
18	0.9	2.8	1.8	TUM
19	0.9	2.2	1.9	TUM
20	0.8	2.9	1	Separation
21	4.5	3.2	1.5	TUM
22	1.3	3.5	1.5	TUM
23	6.0	4.1	1.8	TUM
24	1.9	3.8	0.5	Separation
25	1.7	4.4	1	Separation
26	7.7	4.3	1.1	Separation
27	9.1	3.8	0.8	Separation
28	6.8	3.6	0.4	Separation
29	8.1	3.5	0.5	Separation
30	3.2	5.5	0.5	Separation
31	1.6	6.1	0.7	Separation

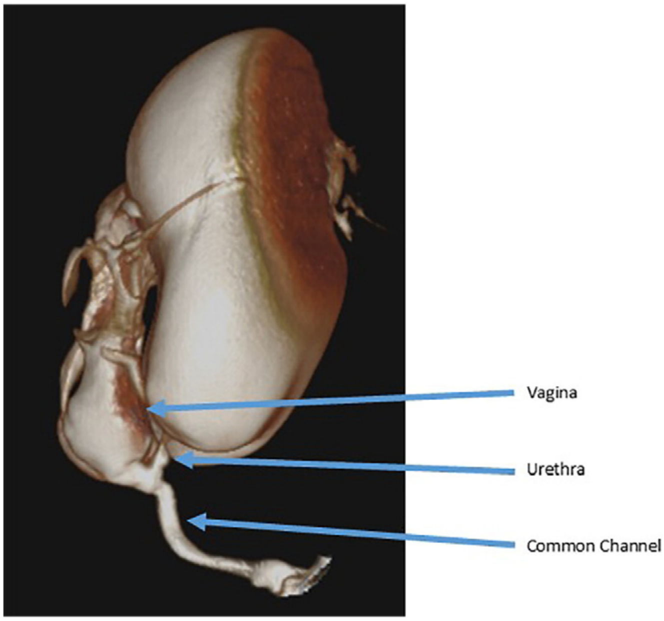

Fig. 2. Long common channel with short urethra.

were 20 patients with a common channel of 1 to 3 cm. Nineteen of these underwent TUM, 1 patient underwent urogenital separation because the urethra was only 1 cm in length. There were 9 patients who had common channels of 3 to 5 cm. In 6 of these, we determined preoperatively that we would perform a urogenital separation due to a short urethra (less than 1.5 cm) leaving the common channel to be the neo-urethra. With preservation of the common channel as the neo-urethra, these patients were left with a urethra of 3.5–5.7 cm. In 3 of these patients a total urogenital mobilization was successfully performed and was chosen because the urethral length, after splitting of the common channel, was able to be left as at least 1.5 cm. There were 2 patients with common channels greater than 5 cm and urethral length less than 0.5 cm. These were both repaired with a urogenital separation, thus preserving the entire common channel length for urethra. Thus 7 patients underwent urogenital separation who using the prior strategy would have undergone a TUM. No patients experienced peri-operative urethral loss and no patients have constant leakage of urine. See Table 1.

One patient in the 3 to 5 cm group developed a urethral-vaginal fistula. This fistula was below the urinary sphincter and thus far has been of no clinical consequence. To respond to this complication we have altered our technique to include both SIS (Surgisis Cook SIS – Porcine small intestinal submucosa – biological scaffold) sewn directed onto the posterior urethral repair and an ischiorectal fat pad [7] placed directly over the SIS to further augment separation of the urethral and vaginal repairs. We have not had any fistulae since.

3. Discussion

Patients with a common channel less than 1 cm in length (Type 1 cloaca) require no urethral repair. As previously described the urethra should be left untouched and an introitoplasty and PSARP should be performed (posterior sagittal anorectal vaginoplasty) [5]. We agree with prior reports that for those patients with a short common channel cloaca (1 to 3 cm) Fig. 4 a TUM almost always works well [5], however there are rare cases of a short urethra in this group and that is why we believe that an assessment of urethral length is important in all cloaca patients. For long common channels (greater than 3 cm), reconstruction strategies are needed involving complex decision making related to the urethra and vagina [5,6]. The group of patients with common channels between 3 and 5 cm, in our view, present the most significant reconstructive challenges. This specific anatomic type has not been well described in the literature and it is for these patients, that we believe, the urethral length is so vital. Surprisingly, the length of the urethra is not mentioned at all in several recent reviews [3,5,6], with the discussion focused only on the length of the common channel as the key determinant for planning and surgical reconstruction. No mention is made of the length of the urethra from the common channel to the bladder neck and therefore what the length of urethra will be at the end of the repair, when the common channel is split as part of a TUM.

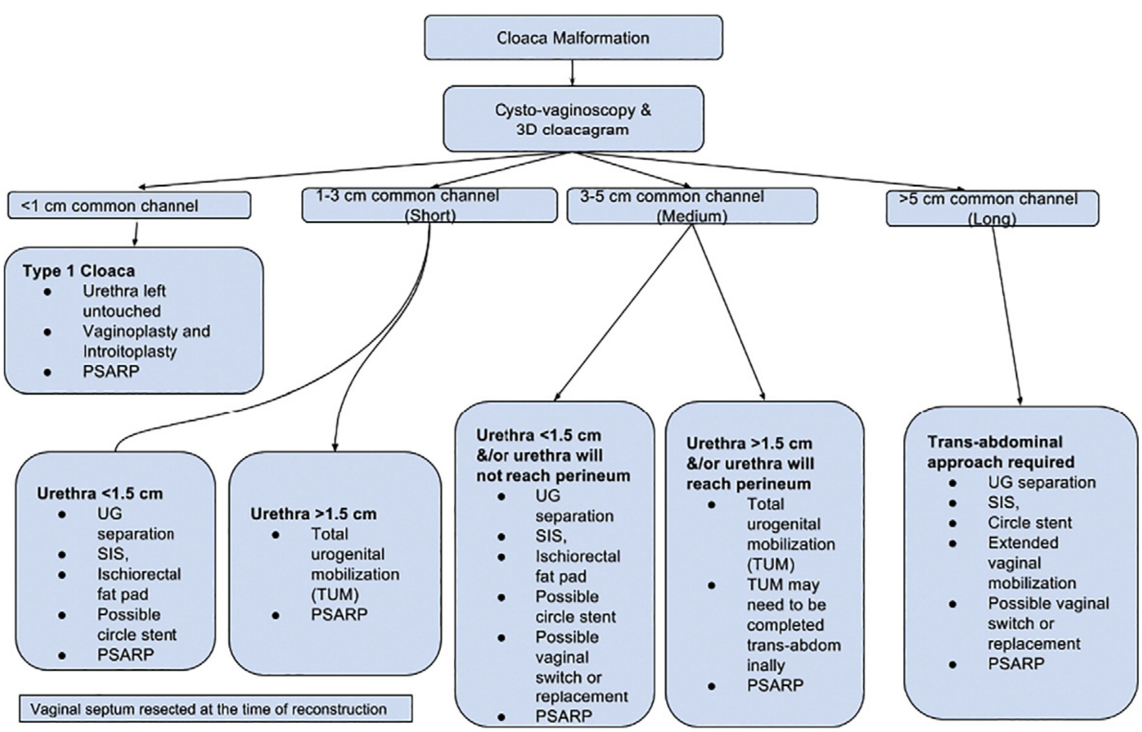

Fig. 3. Algorithm for cloaca management.

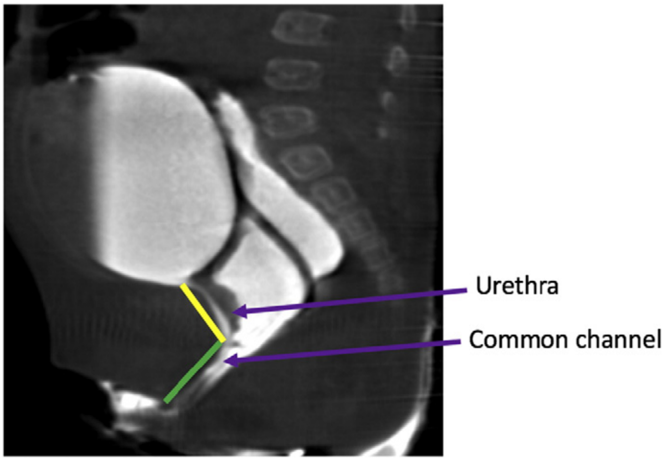

Fig. 4. Short common channel cloaca with adequate urethra.

We recommend that the preoperative assessment, in addition to the common channel length, include the length of the urethra as well as the likelihood of the urethra reaching the perineum if an extended or transabdominal TUM were to be done. This is best performed by a combination of cystoscopy and vaginoscopy and 3D cloacagram [13]. If the urethra is less than 1.5 cm, or the urethra may not reach using a TUM, then a preoperative decision should be made to separate the urogenital tract and thus leave the common channel as neo-urethra. Making this decision preoperatively is very important because if a TUM is performed, in the presence of a short urethra, urinary leakage may result and then, further surgery to deal with a leaking bladder neck will be necessary. In addition, if a TUM is attempted and the urethra does not reach, then a urogenital separation becomes necessary with mobilization of the circumferentially dissected urethra. This should be avoided as it can lead to urethral loss. By contrast, the preoperative decision to separate means that both the anterior urethra and anterior bladder neck remain untouched. We believe that the morbidity associated with urethral loss and incontinence can be avoided with this approach.

The importance of collaboration with urology on the management of cloacal malformations cannot be over emphasized, and this urethral issue is a great example. It is our contention that urethral length needs to be a fundamental component of the cloaca assessment, which if overlooked may lead to incorrect choices during surgical repair and can lead to long-term morbidity, with implications on renal health and urinary continence. While the similarities in the anatomy of urogenital sinus and cloaca led us to this point, it is important to remember the fundamental differences in bladder function between these two groups. Urogenital sinus patients, almost always, have normal bladder function, but many cloaca patients have abnormal bladder function. We settled on the urethral length of greater than 1.5 cm consistent with the recommendations for the UG sinus patients.

In our literature review on cloaca malformations we found only mention of one strategy to deal with the 3 to 5 cm length common channel malformation; an extended total urogenital mobilization [5] in which the entire urogenital complex is delivered up into the abdomen, length gained, and then pulled through. This is called a “transabdominal total urogenital mobilization” [5]. It is specifically this strategy that we caution against, because if this does not work to allow the urethra and vagina to reach the perineum, the only choice at that point is to separate the vagina or vaginas from the urinary tract. This leads to a situation whereby the urethra has been circumferentially dissected and leaves the urethra subject to loss or severe dysfunction due to ischemia. The patient may then require bladder neck reconstruction or closure and a Mitrofanoff.

We believe that the classification for cloacal malformations needs to be expanded (Fig. 3) to include, a description of the urethral length in addition to the common channel length in cases. We propose that if the ultimate urethra would be greater than 1.5 cm (Fig. 4), a TUM should be employed. If the urethra will end up being less than 1.5 cm (Fig. 2), a urogenital separation should be used. In mid-length cloaca malformations (3–5 cm), the default strategy should not be a transabdominal total urogenital mobilization (TUM) as has been previously described [5]. Instead an assessment of the urethral length and the likelihood of the urethra reaching the perineum needs to be done prior to surgery. In cases where the urethra is short (<1.5 cm) or there is doubt that the urethra will reach, a separation of the vagina from the common channel (Figs. 5,6,7) as is recommended for 5 cm or greater common channel cases should be performed [5,6]. This separation of the vagina from the common channel is reminiscent of the original cloacal repair in which the vagina was always separated [1,11,12]. In key cases, when there is concern for urethral length (Fig. 4), this idea remains a valid strategy.

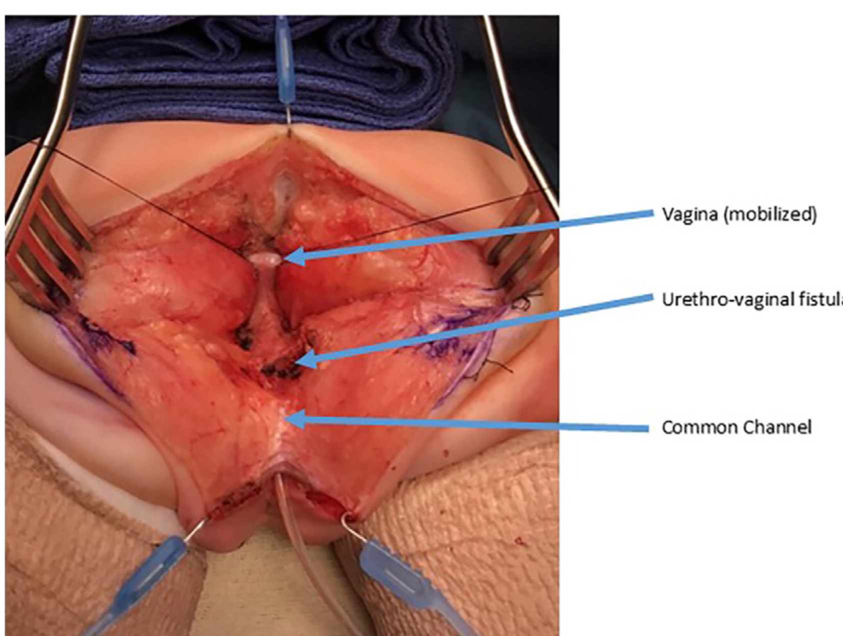

Fig. 5. Operative view showing urogenital separation, leaving common channel as neourethra.

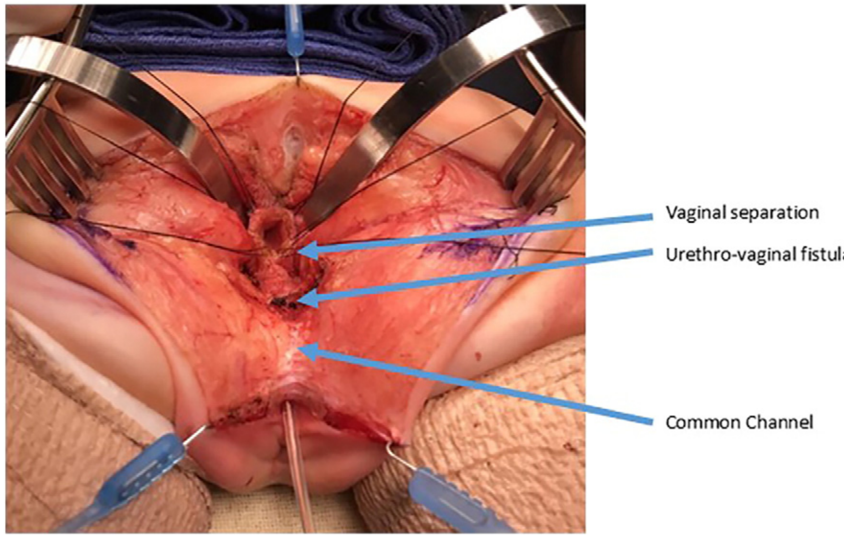

Fig. 6. Operative view showing vagina entering the common channel. The vagina is then separated from the posterior aspect of the common channel.

Long-term outcomes of this new strategy are still not available but in the short term we have not seen any significant postoperative complications and thus far, no patients leak urine and no patients have experienced perioperative urethral loss. We will be able to report our long term results in 2–3 years, once enough time has passed to be able to assess the patient's functional outcomes.

The main concern with urogenital separation has been the development of postoperative urethra-vaginal fistulae. This was one of the main reasons that the TUM was developed [2,3]. The incidence of this complication has been previously reported as 7 to 11% [2]. We had one such case and now employ several surgical strategies to help prevent it. We use (1) a biological scaffold (SIS) placed on the posterior aspect of the repair of the urethra at the (common channel), (2) an ischiorectal fatpad, (previously described in the treatment of recto-vaginal fistulae [7]) mobilized and placed over the repair and the SIS patch and (3) in cases in which the separation has involved a laparotomy, we pass a circle silicone stent through the repaired channel (neo-urethra), into the bladder and out the right or left lower quadrant of the abdomen. Finally, (4) we place a supra-pubic catheter or vesicostomy (if one is not already present) to allow for adequate drainage of urine. The circle stent (with no Foley balloon at the bladder neck), reduces bladder spasms. In addition, a wire can be passed through the stent, and a postoperative cystoscopy can be performed over the wire prior to removal of urethral access. If the urethra is found to be incompletely healed, the stent can be replaced without losing access to the urethra which can allow for further healing. These techniques, which have been described in other situations, we believe will help prevent both fistula development and urethral stenosis and loss.

We believe that functional urologic outcomes have a significant impact on the patient's long-term quality of life and for this reason we perform urodynamics, both pre- and postoperatively, on all patients with cloacal repairs. In addition we record Vancouver scores [16] in every case to assess functional urinary outcomes.

4. Conclusion

We herein report a new strategy and its technical details for cloacal repair. In the future we will report on both the anatomic and functional outcomes of these patients as we longitudinally follow patient reported and urodynamically documented outcomes.

The repair of cloacal malformations remains a significant technical challenge. A pre-operative assessment of both the length of the urethra, as well as the likelihood of it reaching the perineum, is very important (in addition to common channel length) and should be added to the

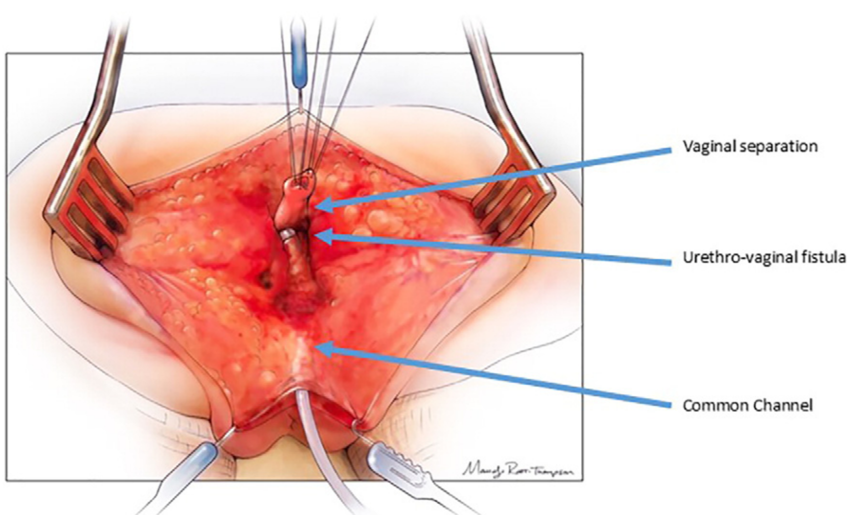

Fig. 7. Artistic drawing of the urogenital separation. Keys: UG: Urogenital Sinus. SIS: Porcine Small Intestinal Submucosa. TUM: Total urogenital mobilization. PSARP: Posterior Sagittal Anorectoplasty.

algorithm (Fig. 3) when one plans a repair. In addition, the use of the total urogenital mobilization technique in patients with a urethra that is less than 1.5 cm long is problematic. A decision to embark on a "transabdominal total urogenital mobilization" should only be done where there is adequate urethra and the surgeon is confident that the urethra will reach the perineum. Separation of urethra and vagina making the common channel become the neo-urethra is a useful strategy that has a key role in certain types of cloaca malformations. Surgical planning which neglects consideration of urethral length can lead to significant urological morbidity.

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