Journal of Pediatric Surgery (2011) **46**, 1250–1255

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Pitfalls and challenges of cloaca repair: how to reduce the need for reoperations

Marc A. Levitt*, Andrea Bischoff, Alberto Peña

Division of Pediatric Surgery, Colorectal Center for Children, Cincinnati Children's Hospital Medical Center, Cincinnati, OH 45229, USA

Received 21 March 2011; accepted 26 March 2011

Key words:
Cloaca;
Reoperation;
Vaginal replacement;
Persistent urogenital sinus

Abstract

Purpose: Cloacal malformations represent the most complex of genitourinary/anorectal anomalies. We have encountered a unique group of complications in referred patients after failed attempted repairs elsewhere and chose to review this experience with the hope of identifying pitfalls to avoid during the primary repair.

Methods: In our series of 509 cloacas, 95 were repaired elsewhere but required reoperation. These cases were reviewed for specific indications for reoperation and methods used for reoperative repair. Key findings at reoperation to explain the complication(s) were specifically sought.

Results: Indications for reoperation included the following: persistent urogenital sinus (46), rectal stricture or acquired atresia (45), acquired vaginal atresia or stricture (45), mislocated rectum (36), urethrovaginal fistula (16), rectal prolapse (12), urethral atresia or stricture (7), and rectovaginal fistula (5). Most patients had more than one indication. In cases of persistent urogenital sinus, the surgeons were unaware of the presence of a cloaca, referring instead to the malformation as a "rectovaginal fistula." From our reading of the operative reports of the original operations, we ascertain that rectal stricture, atresia, or fistula that occurred was most likely related to tension or ischemia. Prolapse was associated with poor pelvic musculature. The average length of the common channel of those patients with vaginal and urethral problems was 4.1 cm.

Conclusion: We have observed key complications requiring reoperation in a large series of cloacal malformations that are potentially avoidable. A persistent urogenital sinus can be avoided by properly diagnosing a cloaca and repairing the entire malformation and not just the rectum during the initial repair. Vaginal and urethral complications occurred mainly in patients with a common channel longer than 3 cm. Repair of cloacas with common channels longer than 3 cm requires familiarity with a complex decision-making process, and atresias, strictures, and fistulae can be avoided with adequate mobilization of structures and preservation of blood supply. Rectal prolapse occurrence relates to the quality of the perineal muscles. Reoperations can restore the anatomy, but the functional results are not as good as those achieved after primary repair.

A cloaca is a malformation that affects the rectum and urogenital tract presenting at birth in females as a single perineal orifice. The vagina, urethra, and rectum are fused together inside the pelvis, creating a single common channel that emerges where the urethra normally opens. The length of

* Corresponding author. Division of Pediatric Surgery, Department of Surgery, University of Cincinnati, Colorectal Center for Children, Cincinnati Children's Hospital Medical Center, Cincinnati, OH 45229, USA. Tel.: +1 513 636 3240; fax: +1 513 636 3248.

E-mail address: marc.levitt@cchmc.org (M.A. Levitt).

0022-3468/$ – see front matter © 2011 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.03.064

the common channel varies from 1 to about 10 cm with an average of approximately 3 cm. Surgical reconstruction represents a significant technical challenge with the goals to achieve urinary control, bowel control, menstrual function, sexual function, and obstetric potential.

As we have learned more about the complexity of cloacal malformations, we have become concerned about the reproducibility of some of the techniques used to repair them. We have encountered a unique group of complications in referred patients requiring reoperation and chose to review this experience to identify pitfalls to avoid during the primary repair [1].

1. Methods

Ninety-five patients with cloacal malformations were referred because they required reoperation. These cases were reviewed for specific indications for reoperation. Key findings at reoperation to explain the complication were specifically sought, and these cases were compared with our published series of primarily repaired cloacas [1,2]. We obtained institutional review board approval for this study.

2. Results

In our review of 95 reoperations for cloacal malformations, we determined the following indications: persistent urogenital sinus (the surgeon repaired the rectal component but left the urogenital sinus) (46), rectal stricture or atresia (45), acquired vaginal atresia or stricture (45), mislocated rectum (36), urethrovaginal fistula (16), rectal prolapse (12), urethral atresia or stricture (7), and rectovaginal fistula (5). Most patients had more than one indication.

All of these patients required a reoperation that was performed at our center. Based on our operative findings, reading of the operative reports, and following discussions with the surgeons, we concluded the following: (1) persistent urogenital sinus related to an initial misdiagnosis whereby the surgeon failed to recognize the type of defect and did not identify that the patient had a cloaca; (2) rectal stricture or acquired atresia or fistulas were related to tension or ischemia and inadequate mobilization of structures; and (3) prolapse was associated with poor pelvic musculature. The average length of the common channel in patients presenting with urethrovaginal fistula, vaginal and/or urethral acquired atresias, or stenosis was 4.1 cm. Reoperation for vaginal problems were performed via a posterior sagittal approach (11), with a laparotomy (17), and vaginal replacement was required in 16 patients.

The complications that were encountered were significantly higher than in our previously published primary group [1,2].

3. Discussion

The first challenge in reducing complications in cloaca repair is to determine the complexity of the malformation before embarking on the reconstruction. To do this, we recommend a cystoscopy/vaginoscopy during a separate anesthetic, outside the newborn period, which can help make the distinction between the 2 very different groups of cloacas, those with a common channel less than 3 cm and those with a common channel greater than 3 cm (Table 1). In addition, a cloacagram with 3-dimensional reconstruction is very helpful [1,3].

By doing these evaluations, the surgeon can (1) determine whether they feel comfortable doing the operation, or prefer to refer the patient to a specialized center; (2) determine whether or not it will be necessary to open the abdomen for the reconstruction, which helps predict equipment needs and operating time; (3) determine whether or not the patient needs a total bowel preparation; (4) determine whether some form of vaginal replacement is necessary; and (5) help predict the final functional prognosis with regard to urinary, bowel, menstrual, sexual, and obstetric function.

Another important moment in time is to inspect the anatomy before colostomy closure. Our routine, after cloaca repair, is to close the colostomy at 8 to 12 weeks, provided the perineum is healing well and the anus has reached the desired

Table 1 Two types of cloacas; our primary cloaca experience, excluding redos, with complete follow-up information

	Group A	Group B
Common channel	Short, <3cm	Long, >3cm
Type of operation	Only posterior sagittal	Posterior sagittal and laparotomy
Length of procedure	3 h	6-12 h
Postoperative hospitalization	48 h	Several days
Incidence in our series	58% (n = 217)	42% (n = 155)
Voluntary bowel movements	66%	34%
Voluntary urinary control	73%[a]	26%
Intraoperative decision making	Relatively reproducible operation	Complex and technically demanding[b]

[a] The remaining patients are dry on intermittent catheterization.

[b] Bladder/vagina separation, ureteral catheterization, vesicostomy, bladder neck reconstruction or closure, vaginal switch, and vaginal replacement (with rectum, colon, or small bowel).

size after the protocol of dilatations has been completed. We perform a vaginoscopy and cystoscopy to confirm that the urethra and vagina are patent and healthy before proceeding. In the event of finding problems with these, they are best taken care of before the colostomy closure.

The high frequency of complicated cases we have been referred, which were previously operated on, clearly represents the fact that the surgical maneuvers to repair cloacas are not highly reproducible. Cloacas are not an exception when we affirm that children with congenital malformations "have a single opportunity" to be repaired, meaning that if that first attempt is not successful, the functional prognosis worsens after a secondary procedure [4].

Fortunately, most cloacas have a common channel shorter than 3 cm [1]. This means that they can reliably be repaired via a posterior sagittal approach, without opening the abdomen and using the maneuver called the "Total Urogenital Mobilization" [5]. For those cases of cloaca with a common channel longer than 3 cm, we believe that the repair is best done at specially dedicated centers experienced in dealing with these malformations. This was the group, as one could have predicted, with the highest complications. The key aspects of the primary repair to mobilize structures without tension that we have used in a long, common channel cloaca are the transabdominal total urogenital mobilization, using a vaginal replacement when appropriate, and in case of a common channel greater than 5 cm, leaving the common channel untouched to be used as the neourethra [1].

When we assessed the operative reports and discussions with the original surgeons for the 95 reoperations, we frequently found descriptions illustrating findings of complex and unpredicted anatomy and structures that were high

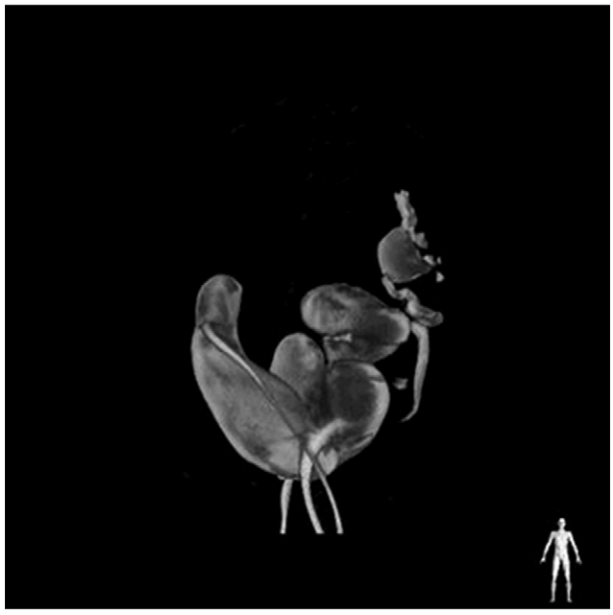

Fig. 1 Three-dimensional fluoroscopic cloacagram performed in interventional radiology. Reprinted from Pediatric Surgery, 7th edition by permission from Elsevier.

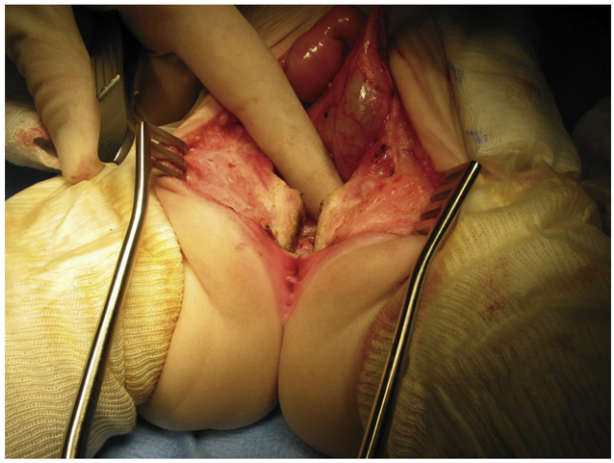

Fig. 2 Transpubic approach.

and difficult to mobilize. It is difficult to determine exactly what happened in each case. After all has healed many months later, we often see cases in which the patient has a blind perineum; in other words; they have no urethra, no vagina, and no rectum, or they may have only 1 or 2 orifices. A study with contrast material injected through the different openings (colostomy, cystostomy, vaginostomy, vesicostomy, or rectum) helps to determine what has become of these structures. This is ideally done in the interventional radiology suite with 3-dimensional fluoroscopy (Fig. 1). We often use this modality in complex primary cloaca cases as well [3]. It is not unusual to find patients with dehiscences, retractions, fistula formations, or abscesses that result in what can be described as "frozen" pelvis, and therefore, these patients are extremely difficult to approach.

The first option is to repair these posterior sagittally, although sometimes via the posterior sagittal approach, it is not possible to mobilize the structures. The next option is an abdominal midline incision. However, even through the abdomen, it is sometimes not possible to find the plane of dissection in the pelvis. At that point, the last alternative that provides the best exposure is the transpubic route, which is ideal for urethral or bladder neck problems for which the posterior approach can be avoided. Of these 42 cases, 1 had an osteomyelitis of the pubis and 1 had a dehiscence requiring resuturing (Fig. 2).

3.1. Persistent urogenital sinus (and associated rectal mislocation, stenosis, stricture, or acquired atresia)

The most common reoperation that we perform in patients with cloacas consists in repairing a persistent urogenital sinus [6] (Fig. 3). Interestingly, from the cloaca cases we have seen with persistent urogenital sinus, very few had come to us with an original diagnosis of a cloaca rather the majority came with the diagnosis of "recto-vaginal fistula." In other words, the surgeons were unaware of the fact that the patient had a cloaca; they tried to repair the malformation, but they only ended up

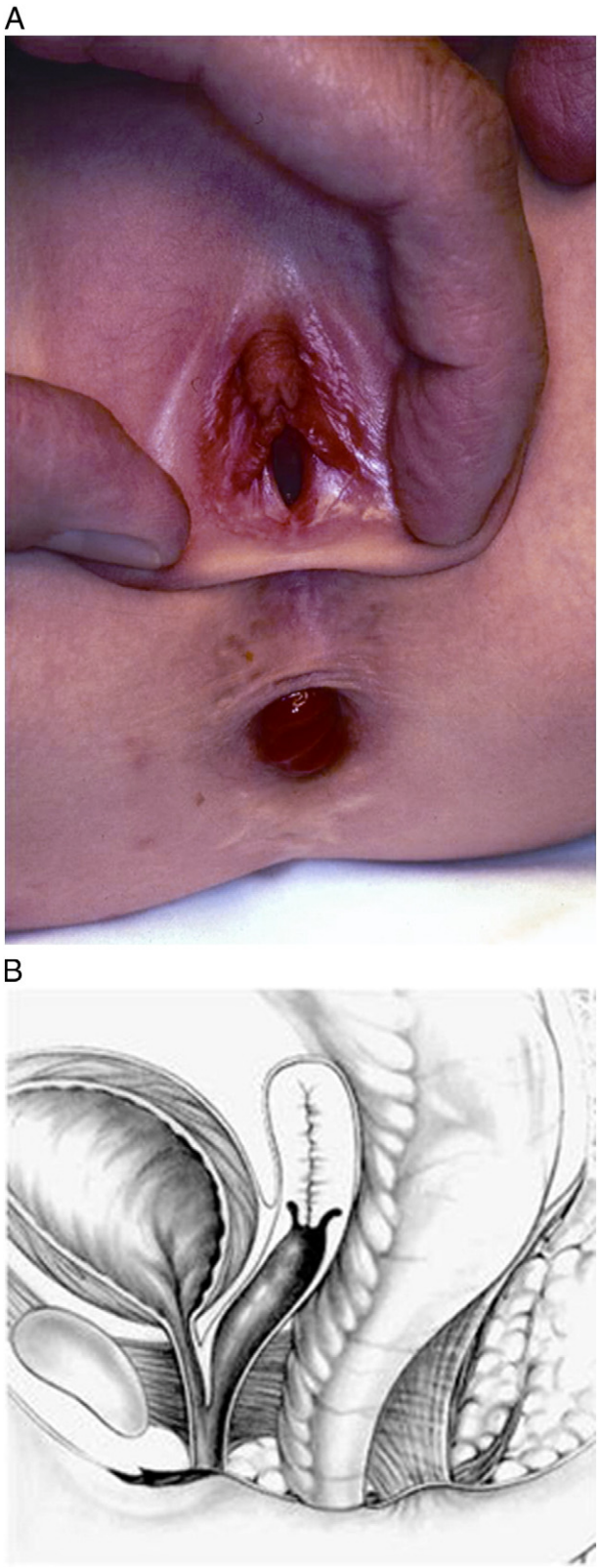

Fig. 3 A and B, Persistent urogenital sinus; only the rectal component of the cloaca was repaired, leaving the urogenital sinus untouched, requiring a complete reoperation. Illustration (B) reprinted from Peña A. Atlas of surgical management of anorectal malformations, Springer Verlag, New York 1989 with kind permission of Springer Science + Business Media.

mobilizing the rectum and left the urogenital sinus untouched [7]. Amazingly, these patients recovered from those operations, continued their life and become aware of their persistent urogenital sinus later, sometimes at the onset of menses or at the age of sexual function.

Looking at the literature, one can find a very obvious shift from the frequency of rectovaginal fistulas and cloacas. Before 1982, the literature had many reports of rectovaginal fistulas and almost no mention of cloacas [7-11]. In retrospect, we know that most of those rectovaginal fistulas operated on and reported in the literature were actually cloacas, or rectovestibular fistulas [7].

The diagnosis of a cloaca is a clinical one and is done by inspection [12]. Vaginoscopy and cystoscopy, as well as a fluoroscopic cloacagram, allow for the determination of the length of the common channel and the characteristics of the vagina(s), which help plan for the magnitude of the operation and determine the likely need for a vaginal replacement.

In our series, persistent urogenital sinus required a full reoperation, including a total urogenital mobilization with or without a laparotomy [4]. In certain cases when the urethra is only 1 cm deep, it can be left alone and somewhat hypospadic, with an introitoplasty only being performed. In such cases, the patient likely will empty her bladder efficiently and will not need intermittent catheterization.

3.2. Acquired vaginal atresia or stenosis and various fistulae

Another common reason why patients are referred to us after an attempted failed repair of a cloaca is that they experience an acquired vaginal atresia or stenosis. In these cases, the surgeon usually knew that he or she was dealing with a cloaca and tried to repair the vaginal component of the malformation. Most of these complications were seen in cloacas with a long common channel. Unfortunately, the vagina was not mobilized properly and had ischemia and acquired atresia. As a consequence, the patient comes to us with a patent urethra and rectum but without a vaginal orifice or one that is significantly narrowed. The vagina may also have a fistulous communication with the urethra. The distal aspect that the surgeon originally attached and sutured to the perineum may have disappeared [13]. The distance between the blind end of the vagina and the perineum varies from patient to patient and, depending on that length, the magnitude and type of operation changes. In such a case, at adolescence, the patient's first manifestation may be menstruation "through the urethra," or she may accumulate menstrual blood (hematometrocolpos) and experience severe abdominal pain with monthly exacerbations.

3.3. Prolapse

Patients with poor pelvic muscles may develop a significant rectal prolapse. The repair uses a technique we have previously described [14].

Vaginal prolapse, if it occurs, can be managed with a similar technique. When the vagina has been replaced with the colon, rectum, or small bowel, some degree of bowel mucosa prolapse may occur. The excess vaginal tissue is mobilized and trimmed. Ideally, this should be treated before the colostomy is closed because it can be done in a single-day admission.

3.4. Acquired urethral atresia, stricture, or stenosis

One may find a patient who comes after a failed attempted repair of a cloaca with an acquired atresia or severe stricture of the urethra. If the urethral stricture is located low enough, the repair can be done posterior sagittally, with mobilization of the rectum, then the vagina and eventually the urethra. Sometimes, the acquired atresia of the urethra is so severe and the length of separation of both healthy portions of the urethra is so great that instead of repairing the urethra, a Mitrofanoff (a conduit for the patient to empty her bladder with intermittent catheterization through an orifice created in the abdomen) is needed. In the infant, a vesicostomy is performed with a urologic reconstruction deferred for a later age. Another option to perform the urethral repair is to use the transpubic approach (Fig. 2).

3.5. Conclusion

The surgeon must be able to successfully identify a cloaca by clinical inspection and avoid repairing only the rectal component. Endoscopy is helpful in determining the length of the common channel and complexity of the repair and with making the decision to refer difficult cases to specialized centers. Long, common channel cloacas have the greatest challenge in mobilizing the urethra, vagina, and rectum and avoiding ischemia and tension. The primary repair is the best opportunity to achieved the patient's functional potential—reoperation may restore this somewhat but never to the degree achieved with an excellent primary repair [12,15,16].

References

[1] Levitt MA, Peña A. Cloacal malformations: lessons learned from 490 cases. Semin Pediatr Surg 2010;19:128-38.
[2] Levitt MA, Peña A. Imperforate anus and cloacal malformations. In: Holcomb GW, Murphy JP, editors. Ashcraft's pediatric surgery. 5th ed. Philadelphia (Pa): Saunders Elsevier; 2010. p. 468-90.
[3] Patel M, Racadio J, Peña A, et al. Complex cloacal malformations: use of rotational fluoroscopy and 3-D reconstruction in diagnosis and surgical planning. Presented at the Annual meeting of the Radiological Society of North America, Chicago, IL, November 29; 2006.
[4] Levitt MA, Peña A. Reoperations in anorectal malformations. In: Teich S, Caniano D, editors. Reoperative pediatric surgery. Totowa, NJ: Humana Press, Inc.; 2008. p. 311-26.
[5] Peña A. Total urogenital mobilization—an easier way to repair cloacas. J Pediatr Surg 1997;32(2):263-8.
[6] Levitt MA, Peña A. Pitfalls in the management of newborn cloacas. Pediatr Surg Int 2005;21:264-9.
[7] Rosen NG, Hong AR, Soffer SZ, et al. Rectovaginal fistula: a common diagnostic error with significant consequences in girls with anorectal malformations. J Pediatr Surg 2002;37:961-5.
[8] Wangensteen O, Rice C. Imperforate anus: a method of determining the surgical approach. Ann Surg 1930;92:77-81.
[9] Santulli TV. Imperforate anus. In: Benson CD, Mustard WT, Ravitch MM, et al, editors. Pediatric surgery. Chicago (Ill): Year Book Medical Publishers; 1962. p. 821-36.
[10] Stephens FD. Congenital malformation of the rectum and anus in female children. In: Stephens FD, editor. Congenital malformations of the rectum, anus and genito-urinary tracts. Edinburgh: E. and S. Livingstone; 1963. p. 42-61.
[11] Bill AH, Hall DG, Johnson RJ. Position of rectal fistula in relation to the hymen in 46 girls with imperforate anus. J Pediatr Surg 1975;10: 361-5.
[12] Shaul DB, Harrison EA. Classification of anorectal malformations—initial approach, diagnostic tests, and colostomy. Semin Pediatr Surg 1997;6:187-95.
[13] Taghizadeh AK, Wilcox DT. A posterior sagittal approach for revision vaginoplasty. BJU Int 2005;96:1115-7.
[14] Belizon A, Levitt MA, Shoshany G, et al. Rectal prolapse following posterior sagittal anorectoplasty for anorectal malformations. J Pediatr Surg 2005;40:192-6.
[15] Peña A, Grasshoff S, Levitt MA. Reoperations in anorectal malformations. J Pediatr Surg 2007;42:318-25.
[16] Peña A, Hong AR, Midulla P, et al. Reoperative surgery for anorectal anomalies. Semin Pediatr Surg 2003;12:118-23.

Discussion

Anonymous: Question. How do you know when you need a vaginostomy or a vesicostomy? And how do you go about making that decision?

M. Levitt: That's a very good question. In the newborn, you need to screen the patient for the presence of hydrocolpos and that's ideally done with an ultrasound. You're going to do an ultrasound anyway to make sure you assess whether there are 2 kidneys or a hydronephrosis and you want to see if there is a hydrocolpos. You then take the baby to the operating room with the plan to do a colostomy, and in the operating room I like to palpate the abdomen and see how big that hydrocolpos is. If I know it's there from ultrasound and it's not enormous and not above the umbilicus then I will plan a straightforward left lower quadrant incision for my colostomy, palpate the hydrocolpos and drain it. You have to be careful to open the wall of the dome of the vagina to ensure that you're not dealing with 2 hemivaginas, which you often are and you remove some of the septum so that your vaginostomy tube actually can drain both. If the hydrocolpos is higher and palpable above the umbilicus then I actually do a lower midline incision to get above the hydrocolpos and do the same drainage.

In 95% of the cases, drainage of the hydrocolpos alone is enough to cure the problem of hydronephrosis because the issue is the drainage of the urethras at the trigone, which are being pressed on by the hydrocolpos. If we had

100 urologists in the room, they would say that 95% of the time you need to do a vesicostomy and I think that is incorrect. Most of the time drainage of the hydrocolpos is the way to go because that's the way to resolve the hydronephrosis. On rare occasion when the common channel is very narrow, and the bladder does not drain well, you need to also do a vesicostomy but our routine is to do the colostomy and vaginostomy first, watch the baby over the next week or so, and if the hydronephrosis fails to resolve, if it worsens, or if the bladder fails to empty, then we drain the bladder at that time but it is rarely necessary. Much worse is to drain the bladder alone and ignore the hydrocolpos, and that hydrocolpos then remains filled with fluid and can cause a lot of trouble including urinary tract infections and acidosis.

Tarun Kumar (St Louis, MO): For transabdominal urogenital mobilization, what's your experience with urinary incontinence?

M. Levitt: With urinary incontinence?

T. Kumar: Yes, when you mobilize transabdominally?

M. Levitt: So if you do a total urogenital mobilization, and it does not reach and you then deliver that up into the abdomen for further dissection, we've only really been doing that for the last three or four years. I don't really have a good answer as far as long-term urinary incontinence goes. I can tell you that the vast majority of those patients will need intermittent catheterization. The key is to give them access to a urethra, but whether they have urinary control without the need for intermittent catheterization we don't know. We do know that for those patients with common channels and this is the group that we're talking about, for those patients with common channels greater than 3 cm, 80% of them need intermittent catheterization and don't empty their bladder efficiently on their own. So I would expect that they're going to still remain in that intermittent catheter group.

T. Kumar: Thank you.

Nelson Rosen (New Hyde Park, NY): Would venture to say that in a significant number of those patients you can achieve complete decompression of the hydrocolpos from below and sometimes even with a temporary tube and avoid the tethering effect of a transabdominal vaginostomy, except in the most severe of cases where you describe very narrow long common channels and profound hydro, and by avoiding that tethering effect of the transabdominal tube vaginostomy, you save yourself the detachment of that from the abdominal wall when you go and do the repair. So I've been really trying as a philosophy to be aggressive about decompressing those from below when I can, and it's really helped me in the repairs.

M. Levitt: I think it's a good point. I just would caution that you have to be absolutely certain that you're draining what you want to be draining. And what we would do is teach the family how to do the intermittent catheterization of the common channel and actually teach them in ultrasound to prove that the tube is actually going into the hydrocolpos. Remember the tube could go into the right side, the left side, the bladder or the rectum, and if you're not reliably decompressing it, you're going to get into trouble. Actually, I have not found too much trouble with the tethering as long as you make the tube of vaginostomy relatively low in the lower abdomen. I think the most important thing is to get a complete drainage of the hydrocolpos to avoid trouble.

I would also say that as far as drainage is concerned, we have found that using a curled tube is much better than a Pessar or a Malecot because the hydrocolpos is an inflamed structure, and over time it sort of recedes away from the abdominal wall and then the tube pops out whereas, if you put in a curled, pigtail, tube you'll never have that problem, and we learned that the hard way so I'm telling you to use the curl tube, it's much better.