Cloacal Anomalies: Role of Vesicostomy

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• **The purpose of this report is to discuss the use of vesicostomy in the treatment of cloacal anomalies. In 4 years, the authors have performed primary reconstruction in four children who had cloacal anomalies. Three children had hydrocolpos, which in two cases failed to decompress with tube vaginostomy or clean intermittent catheterization. In both cases, cutaneous vesicostomy effectively prevented urinary sepsis, which allowed the children to thrive before definitive reconstruction was performed. Vesicostomy is technically simple to perform and is easily reversed without loss of bladder volume. Further, it lends itself well to definitive reconstruction of cloacal anomalies.**

INDEX WORDS: Cloacal anomalies; vesicostomy, cutaneous.

UPPER URINARY tract sepsis and deterioration are a frequent occurrence in children who have cloacal anomalies. Hydronephrosis secondary to urinary malformations, hydrocolpos, bladder dysfunction, and outlet obstruction may occur in more than 65% of patients.[1] In most cases, a high vesical-vaginal fistula occurs in conjunction with an elongated urogenital sinus, leading to massive pooling and stasis of urine within the vagina. The resultant hydrocolpos may compress the bladder neck, causing ureteral-vesical junction obstruction. We have found cutaneous vesicostomy to be superior to other methods of drainage for the prevention of urinary sepsis and upper tract deterioration. Further, vesicostomy lends itself well to definitive reconstruction of cloacal anomalies.

CASE REPORTS

Case 1

A full-term 2,000-g white newborn girl had a blank perineum with no vaginal opening and a small sinus tract at the base of her clitoris. In addition, she was had abdominal distension and a palpable midline pelvic mass. Ultrasound of the abdomen showed a solitary left kidney with hydronephrosis in conjunction with hydrocolpos. She underwent a double-barrel left transverse colostomy and cystoscopy; a long urogenital sinus was observed, as well as communication of the vagina and rectum at the level of the bladder neck.

Postoperatively, multiple episodes of urinary sepsis occurred, and surveillance cultures repeatedly showed colony counts of greater than 100,000 mixed organisms, including *Klebsiella, enterococcus,* and *Escherichiacoli.* A clean intermittent catheterization program was instituted, accompanied by antibiotic prophylaxis. Subsequently, several hospitalizations were required because of intravenous antibiotic treatment of multiply resistant organisms.

At 5 months of age she weighed 3.8 kg; she was anemic and had a hematocrit of 27%. An ultrasound of the abdomen demonstrated a large hydrocolpos with persistent left-sided hydronephrosis. She underwent a Blocksom vesicostomy, and a follow-up sonogram showed reduced hydrocolpos and a lower grade of hydronephrosis. There was dramatic clinical improvement and no subsequent episodes of urinary sepsis. At 9 months of age she weighed 7.8 kg and had a hematocrit of 36%.

At 10 months of age, she underwent posterior sagittal exploration in the prone jackknife position at which time the urogenital sinus was not opened and the colon and vagina could not be visualized adequately. After a total body prep, she was returned to the supine position and underwent a laparotomy in which an extended Pfannenstiel incision was used, which incorporated the vesicostomy. Bilateral ureteral catheters were placed through the open bladder, and the colon, vagina, and bladder were separated by a meticulous dissection. A full-thickness vaginal cuff was created circumferentially down to the common wall between the bladder and vagina whereupon an endovaginal dissection was performed and the vesical-vaginal communication was repaired. The vaginal cuff was mobilized on an ovarian pedicle, the vaginal septum was excised, and the vagina was tapered to an appropriate size. The vesicostomy was closed over a suprapubic cystostomy tube, and the vagina and colon were pulled through to the perineum. Finally, the patient was returned to the jackknife position; the vaginal cuff was marsupialized, and the sphincter complex and rectum were reconstructed in their proper anatomic configurations. Three months later the colostomy was closed without incident.

At 3 years of age, she is infection-free and continent of both urine and stool.

Case 2

A 37-week-gestational-age black girl, weighing 2,360 g, had a blank-appearing perineum with no vaginal opening and a small sinus at the base of her clitoris. During the physical examination, a distended abdomen and palpable mass were noted. A sonogram of the abdomen showed grade 2 bilateral hydronephrosis. She underwent a tube vaginostomy and end-sigmoid colostomy with mucus fistula. Postoperatively, the vaginostomy tube drained very little fluid and was removed. Relapsing fever developed subsequently, and urine cultures showed persistent growth of mixed organisms, with colony counts of more than 100,000 per cubic centimeter. She was treated with clean intermittent catheterization and appropriate antibiotic therapy, which were unsuccessful. A follow-up sonogram showed a markedly distended vagina with grade 3 bilateral hydronephrosis.

At 1 month of age, she underwent vaginoscopy, which showed a markedly dilated bladder and vagina and a vesical-vaginal communication at the level of the bladder neck. A Blocksom vesicostomy was performed, and postoperatively a sonogram showed dramatic reduction in the degree of hydrocolpos and the grade of hydronephrosis. At the same time her clinical condition improved, and there were no more episodes of urinary sepsis.

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At 12 months of age, she underwent definitive reconstruction at which time a posterior sagittal incision was made without opening the urogenital sinus. Because the colon and vagina could not be mobilized adequately from below, she was returned to the supine position and given a total body prep. A laparotomy was performed using a Pfannenstiel incision that encompassed the vesicostomy. Bilateral ureteral stents were placed to facilitate the dissection between the colon, vagina, and bladder. A full-thickness vaginal cuff was created circumferentially down to the level of the common wall between the bladder and vagina, whereupon an endovaginal dissection was performed to the level of the vesical-vaginal fistula at the bladder neck. The fistula was closed and the bladder neck repaired. A vaginal cuff was mobilized on an ovarian pedicle, the vaginal septum was excised, and the vagina was tapered. The vesicostomy was closed around a suprapubic cystostomy tube, and the rectum and vagina were pulled through to the perineum. The patient was returned to the prone jackknife position; the vaginal cuff was marsupialized and the sphincter complex reconstructed with the rectum in correct anatomic position.

At 16 months of age, it was noted that the vaginal orifice was sealed. A limited perineal exploration was performed, and the distal vagina was reconstructed with a split-thickness skin tube. The colostomy was closed subsequently, and at 28 months of age she underwent bilateral neoureterocystostomy for grade 4 vesical ureteral reflux. At 3 years of age she is on a clean intermittent catheterization program; she has been dry and has had no more episodes of urinary sepsis. She is not yet toilet trained.

DISCUSSION

Cutaneous vesicostomy was first introduced by Blocksom[2] in 1957 and adapted to children by Michie[3] in 1962. This tubeless form of diversion provides excellent temporary drainage of the lower urinary tract without loss of bladder volume.[4] Vesicostomy eliminates urinary stasis and high intravesical pressure and may control infection and stabilize renal function. Vesicostomy for cloacal anomalies prevents urinary reflux into the vagina and colon, allowing regression of the hydrocolpos and reversal of hyperchloremic acidosis, respectively. Other specific indications for vesicostomy include posterior ureteral valves in low-birth-weight infants,[5] prune-belly syndrome,[6] neurogenic bladder dysfunction,[7] and vesicoureteral reflux and hydroureteronephrosis in infants,[8] whenever the latter conditions lead to recurrent sepsis or deterioration of the upper urinary tract.

Vesicostomy is quick and simple to perform, making it especially attractive for small or critically ill infants. If constructed properly, it is reliable and easily reversed without sacrifice of bladder tissue. Our technique is similar to the Blocksom procedure as modified by Duckett.[8] We use a 2- to 3-cm incision within the skin crease, above the symphysis pubis, which eventually may be used for the abdominal portion of the definitive procedure (Fig 1). The rectus fascia and muscle are opened transversely, and the dome of the bladder is completely mobilized until the urachal remnant or obliterated hypogastric is identified. The dome of the bladder is fixed to the fascia and opened, and the bladder edges are everted onto the skin (Fig 2). Potential complications include stenosis and prolapse, which occur in less than 5% of patients.[4,9]

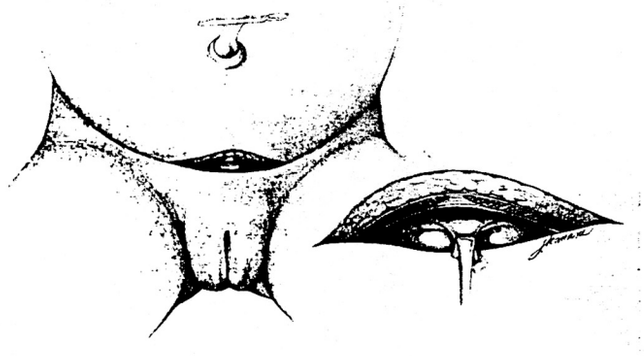

Fig 1. Exposure of the bladder. The dome is mobilized to the urachal remnant.

Vesicostomy for cloacal anomalies was first advocated by Duckett in 1973.[8] Raffensperger's[10] earlier experience showed that suprapubic tube cystostomy and tube vaginostomy often perpetuate urinary sepsis without providing adequate drainage. Various explanations for this include the tube acting as a foreign body, hymenal remnants trapping urine within the vagina, and a septated vagina (in 65% of cases)[11] deterring tube drainage of the hydrocolpos. Also, cystostomy and vaginostomy do not provide dependent drainage and may lead to contraction and fibrosis of the bladder and vagina, respectively.

Clean intermittent catheterization, advocated by Nakayama,[12] may achieve drainage without an indwelling catheter; however, it requires training and strict adherence to a formal catheterization program. Further, it may require several surgical procedures including cutback and dilatation of the urogenital sinus as well as endoscopic incision of the vaginal septum.

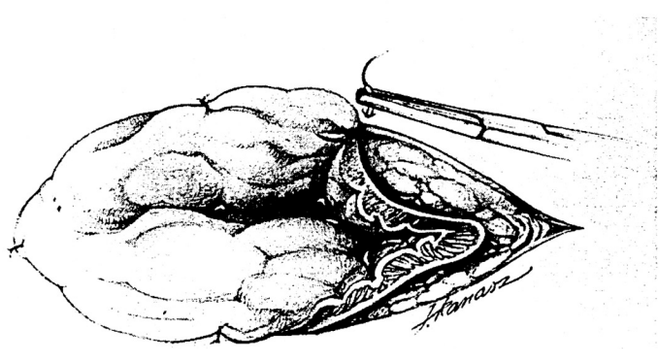

Fig 2. The open bladder is secured to the abdominal wall at the fascial and skin level with absorbable sutures.

Catheterization of the urogenital sinus is usually directed into the vagina rather than the bladder because the contents of the latter are thickened by vaginal mucous. Thus, catheterization does not always provide adequate drainage of the urinary tract.

It is important to emphasize that bladder diversion may not prevent urinary deterioration or sepsis in the presence of upper urinary tract obstruction. Therefore, if bilateral hydronephrosis is demonstrated by sonography in the newborn period, a latex DPTA scan (with a catheter in place) should be obtained to rule out the latter. In the case of ureterovesical junction obstruction, a cutaneous ureterostomy should be performed to preserve renal function.

In conclusion, urinary sepsis is a common complication of cloacal anomalies that frequently lead to failure to thrive and impaired renal function. These complications may be avoided by temporary vesicostomy, which is simple to perform and easily reversed, without loss of bladder volume. Finally, in addition to providing excellent drainage of the urinary tract, vesicostomy allows easy access of the bladder for placement of ureteral catheters that facilitate the abdominal dissection required for reconstruction in the majority of cases.[13] We advocate use of this procedure in infants with cloacal anomalies and hydrocolpos for whom clean intermittent catheterization is either impractical or unsuccessful.

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