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Transanorectal Approach for the Treatment of Urogenital Sinus: Preliminary Report

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● **The treatment of the urogenital sinus with normal rectum still represents a challenge. A perineal approach with or without a skin flap seems to be effective for those patients with a low implantation of the vagina. However, in patients with a high vaginal implantation, this treatment frequently fails to provide a good, functional vagina due to a narrow, strictured vaginal opening. Based on previous experience in the treatment of more than 80 patients with a persistent cloaca, a posterior sagittal *transanorectal* approach with a protective colostomy was performed in three patients with urogenital sinus and normal rectum. The pelvis was approached through a midsagittal posterior incision; the coccyx was split and the entire anorectal sphincteric mechanism was divided in the midline. The rectum was bivalved in the midline including both posterior and anterior rectal walls. This provided excellent exposure to the urogenital sinus. The vagina was then fully separated from the urogenital sinus (as described in cases of persistent cloacas), and then mobilized and sutured to the perineum. The rectum and sphincteric mechanism were meticulously reconstructed. A midline incision assures the preservation of anorectal innervation, and provides excellent exposure to the pelvis. Anal dilatations are not necessary to maintain a patent and supple anorectal opening because the rectum has two suture lines, one in front of the other. After the colostomy was closed, all patients had appropriate bowel control for their age; two of them are fully continent for urine and the third one still has a suprapubic cystostomy tube waiting for a repair of an additional urethral malformation.**

INDEX WORDS: Urogenital sinus, transanorectal approach.

THE UROGENITAL sinus in association with a normal rectum is usually seen in patients with adrenal hyperplasia but can also occur in the absence of that condition. The repair of the adrenal hyperplasia type of urogenital sinus involves reduction clitoroplasty, labioscrotal reduction, and a vaginoplasty.[1-16] As with many other defects, the urogenital sinus malformation has a spectrum. In some patients, the vagina communicates with the urethra close to the perineum. In others, the vagina joins the urethra more proximally, resulting in a long common channel (urogenital sinus). In patients with a low vaginal implantation, surgical repair can be performed through the perineum without much difficulty or complication.[1-16] However, patients with high vaginal implantation continue to present a serious challenge in their treatment with the possibility of some complications.[17-19] Attempts to repair a high vaginal implantation in association with a urogenital sinus by the perineal approach can result in severe fibrosis and narrowing of the vaginal opening. For these very high defects, alternative approaches had been attempted including parasacral,[20,21] perineal pull-through vaginoplasties,[1,2,6,20,22] and abdominoperineal operations.[22,23] For this group of patients, a transanorectal approach with a previous protective colostomy seems to be a good alternative method of mobilizing the vagina and achieving satisfactory results, as determined by our experience in managing three patients with a high urogenital sinus. The posterior transanorectal approach includes division of all of the bowel sphincteric structures, both anterior and posterior to the rectum, in order to obtain adequate exposure. This is similar to the management of the persistent cloaca, which involves mobilization of the vagina, reconstruction of the urethra, and then location of the vagina in a more normal site after which the rectum is meticu-

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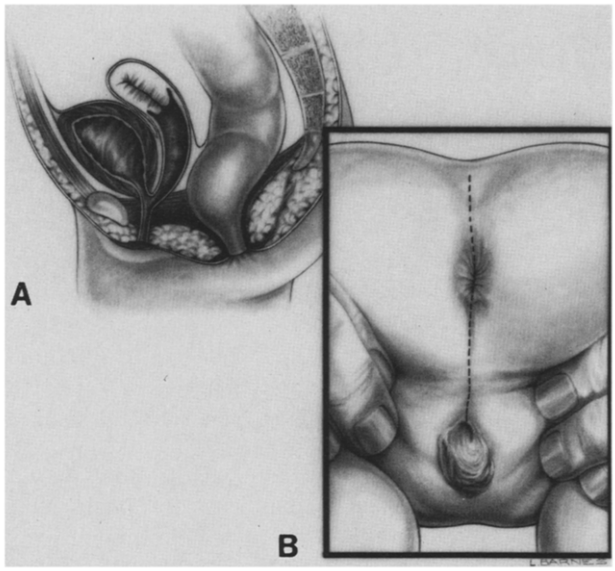

Fig 1. (A) Anatomy of a urogenital sinus with a high implanted vagina. (B) Transanorectal incision.

lously reconstructed. Fecal and urine continence are not affected because the entire operation is performed through the midline.

MATERIALS AND METHODS

In order to perform the operation safely, a protective colostomy must be done prior to the operation.

The patient is placed in the prone position with the pelvis elevated. Figure 1A shows the anatomy of a typical urogenital sinus with a high vaginal implantation. A long midline sagittal incision is made from the middle of the sacrum through the anus and its sphincteric mechanism to the perineum. The incision then extends to the single perineal orifice, as shown in Fig 1B. A needle tip cautery is used in order to achieve precise hemostasis. The levator muscle, muscle complex, and external sphincter are divided in the midline; the posterior and anterior rectal walls are then opened, and the incision is then carried into the anterior aspect of the external sphincter. The perineal fat is then divided in the midline, until the urogenital sinus is exposed (Fig 2). The urogenital sinus is then opened on its posterior aspect sagittally, thereby exposing both the urethra and vagina (Fig 3). Multiple 6-0 silk stitches are placed around the cut margin of the vaginal opening immediately above the urethral opening (Fig 4). It is important to note that the vagina and urethra have a common wall; therefore, initially, submucosal dissection of the vaginal wall must be performed for

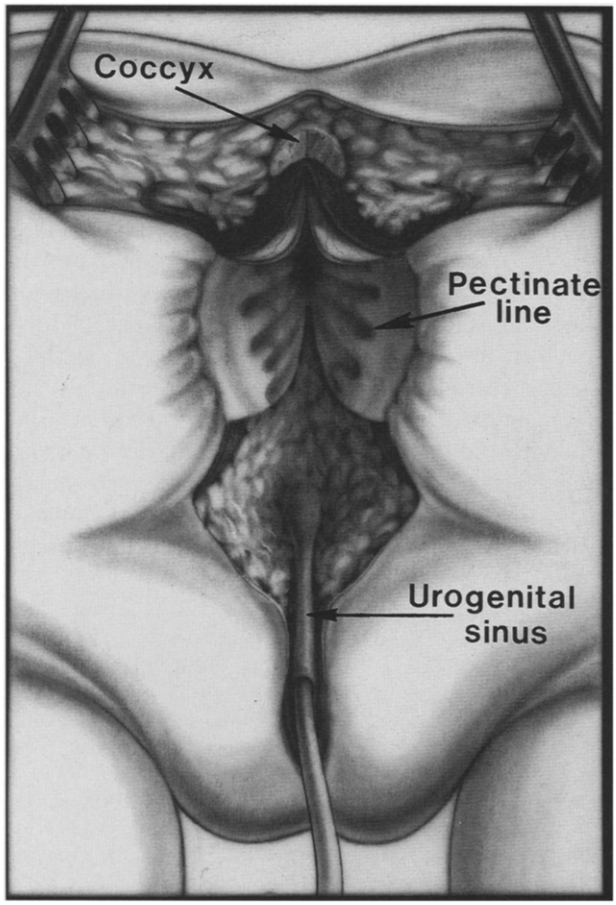

Fig 2. Exposed urogenital sinus showing the bivalved rectum and anus. The pectinate line is indicated.

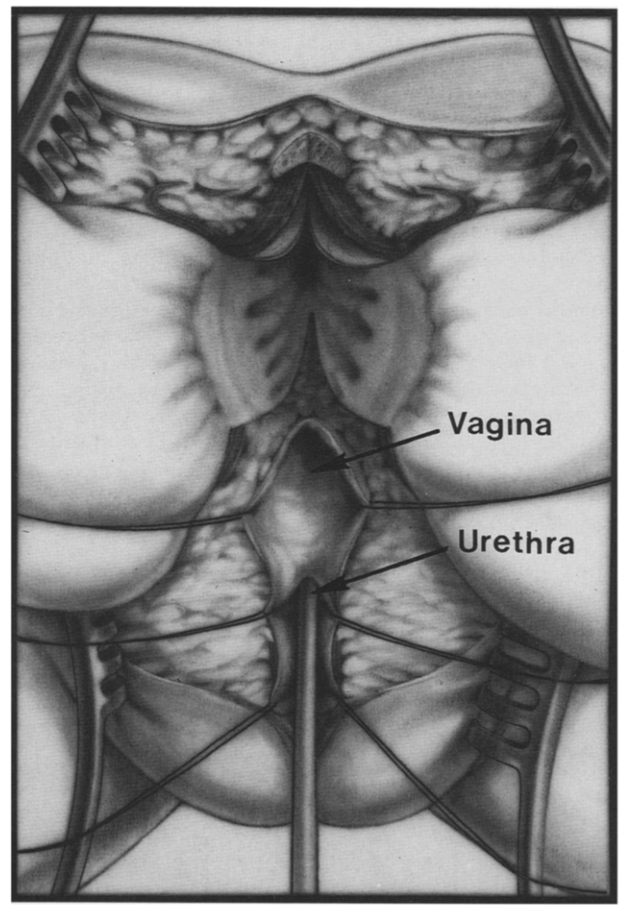

Fig 3. Opened urogenital sinus. The vagina can be seen as well as the urethra.

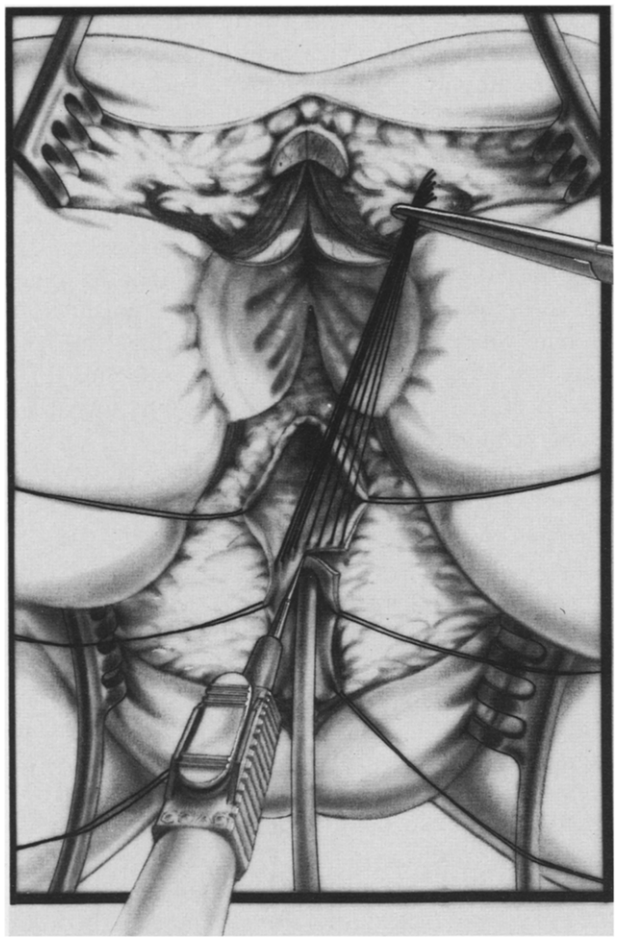

Fig 4. Vaginal dissection.

the first few millimeters until the walls of the vagina and urethra have been fully separated. At that point, a full-thickness dissection of the vagina can be performed. This dissection is quite similar to that needed for patients with a persistent cloaca. Once the vagina has been completely separated from the urethra and bladder neck area, a circumferential dissection is carried out dividing all of the bands and vessels which hold up the vagina within the pelvic cavity. The previously inserted silk sutures around the vaginal opening are clamped together in order to provide homogeneous traction during the circumferential dissection of the vagina and its mobilization (Fig 5). The neourethra (previous urogenital sinus) is reconstructed in two layers with interrupted 6-0 long-term absorbable sutures. The vaginal orifice is then sutured near the urethral orifice as it would be in the normal child (Fig 6). In the event of damage to the anterior vaginal wall, which can occur during mobilization of the vagina, it is convenient to rotate the vagina by 90° so that there is normal, healthy, well-vascularized vaginal wall posterior to the urethral suture line. The perineal body is then reconstructed with interrupted 5-0 long-term absorbable sutures. The rectum must be reconstructed in a meticulous manner bringing together the corresponding opposing anatomical landmarks, including the anterior and posterior limits of the anus. The posterior rectal wall, the external sphincter, muscle complex, and levator mechanism are meticulously reconstructed and evaluated by using the electrical stimulator (Figs 6A and 6B). Figure 6C shows the completed reconstructed malformation. The colostomy is closed in a separate operation 1 month after the main repair.

RESULTS

This technique was used for three consecutive patients. All of them had a good recovery from their operation. Two of them are fully continent of both urine and stool after their colostomy was closed. The third patient shows a normal bowel movement pattern for her age, but still has a suprapubic cystostomy tube because she suffers from a severe congenital urethral malformation soon to be repaired. Postoperative anal dilation was done once to confirm the patency of the anus. Thereafter, it was not necessary to do any additional anal dilation because stricturing of the anus and rectum does not occur in the

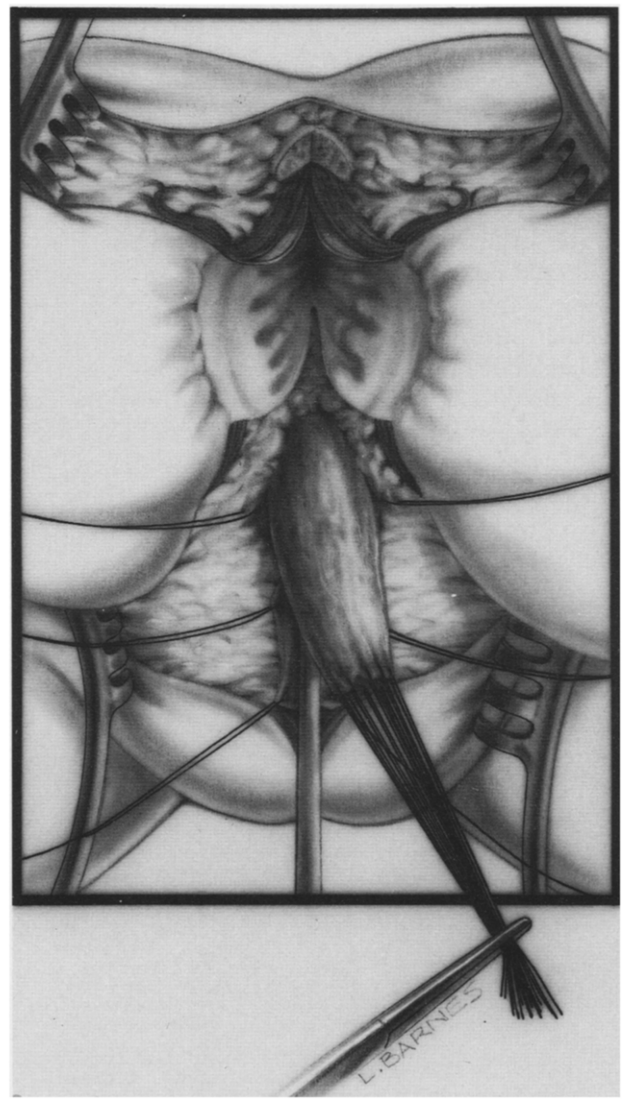

Fig 5. Fully mobilized vagina.

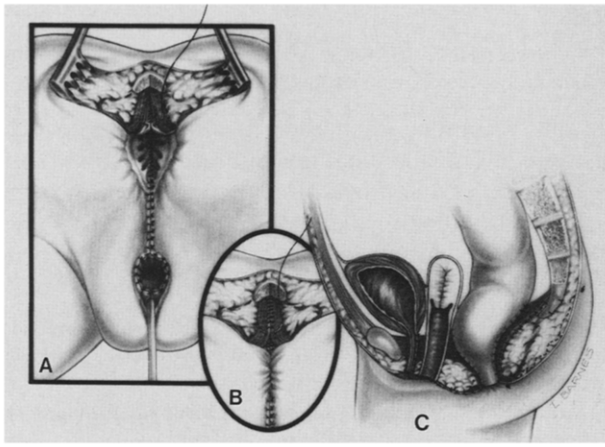

Fig 6. (A) Reconstructed vagina, perineum, and anterior rectal wall. The posterior rectal wall is being sutured. (B) Reconstruction of the posterior sphincteric mechanism. (C) Fully repaired malformation.

presence of two parallel anterior and posterior suture lines.

Table 1 shows the characteristics of these patients.

DISCUSSION

We are aware that operating on the urogenital tract by bivalving the normal rectum and anus has a potential of compromising bowel control and represents a rather bold maneuver. However, this approach provides excellent exposure and changes a rather difficult operation into a relatively easy one. The mobilization of the vagina in these patients seems to be easier than that encountered in patients with a persistent cloaca. We do not recommend this technique in patients with a low vaginal opening into the urogenital sinus because other accepted techniques have proven to be very effective.[1-16,20-23]

It is tempting to perform this operation without a protective colostomy. However, an infection of the reconstruction carries with it the result of compromising bowel control, which would be unacceptable. Therefore, we prefer to perform a protective temporary colostomy prior to the main repair and close the colostomy about a month after the reconstruction.

During the reconstruction, it is very important to do the operation exactly in the midline in order to preserve rectal and urinary tract innervation. A pararectal approach that would involve pushing the rectum laterally could dennervate the rectum with resultant potential complications of bowel control.

Although our experience with this surgical approach is recent and the number of patients few, to date, this approach has proven to be excellent for the treatment of the high urogenital sinus. Based on our experience with three patients, we feel that the use of the technique described should be limited to those patients with a urogenital sinus longer than 2 to 3 cm, as measured cystoscopically. Also, the operation may be a useful alternative for those patients for whom other techniques have failed.

Fortunately, patients with a high vaginal opening are unusual.[1,6] Most authors recommend a vaginal perineal pull-through with the use of skin flaps as recommended by Hendren and Donahoe as the optimal way of repairing this condition.[1,2,6,20,22] Stenosis of the vaginal opening has been mentioned as the most frequent complication of these techniques. Therefore, vaginal dilation has been mandatory for these patients. Our approach allowed for significant vaginal mobilization in all three patients but repeated vaginal dilation have not been necessary. The vaginal opening has been quite satisfactory 6 months to 2 years after the operation.

Most publications emphasize the need to preserve the integrity of the external urinary sphincter in order to avoid urinary incontinence.[1,2,6,20,22] Furthermore, the external urinary sphincter is depicted in those publications as a sling surrounding the urethra distal to the vaginal opening in patients with high vaginal implantation into the urogenital sinus. Based on our experience with this condition, together with our previous experience in treating patients with a cloacal canal anomaly,[24] we feel that the voluntary urinary sphincter is better represented as a muscular structure homogeneously distributed along the length of the urogenital sinus (neourethra). The voluntary urinary sphincter can be divided along its midline posteriorly (in a sagittal manner) and subsequently resutured meticulously without interfering with urinary continence.

Table 1. Patient Characteristics

Case No.	Age at Operation (mo)	Malformation	Common Channel	Results
1	24	UG sinus, adrenal hyperplasia	2.5 cm	Urinary and fecal continence
2	17	UG sinus, no adrenal hyperplasia	4 cm	Urinary and fecal continence
3	16	UG sinus, no adrenal hyperplasia, esophageal atresia, tracheoesophageal fistula, hydrocolpos	Approximately 1.5 cm	Bowel movement pattern appropriate for her age; voiding pattern to be evaluated after urethral repair

Abbreviation: UG, urogenital.

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