RECONSTRUCTION OF THE HIGH UROGENITAL SINUS: EARLY PERINEAL PRONE APPROACH WITHOUT DIVISION OF THE RECTUM

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ABSTRACT

Purpose: Reconstruction of the vagina and external genitalia in the infant is quite challenging, particularly when a urogenital sinus is associated with high confluence of the vagina and urethra. Many surgeons believe that children with such a malformation should undergo staged or delayed reconstruction, so that vaginoplasty is done when the child is older and larger. Vaginoplasty early in life is thought to be difficult due to patient size and poor visualization. The posterior sagittal approach has been beneficial for acquiring exposure to high urogenital sinus anomalies but it has been thought to require splitting of the rectum and temporary colostomy. We report a modification of this technique.

Materials and Methods: In the last 5 years all patients with urogenital sinus anomalies underwent reconstruction using a single stage approach regardless of the level of confluence. In 8 patients with a high level of confluence reconstruction was performed using a perineal prone approach. Exposure was achieved without division of the rectum. The operative technique is presented in detail.

Results: This midline perineal prone approach has allowed excellent exposure of the high vagina even in infants. In all 8 patients reconstruction was done without difficulty and no patient required incision of the rectum or colostomy. This procedure did not preclude the use of a posteriorly based flap for vaginal reconstruction.

Conclusions: While patients with low confluence can be treated with single posteriorly based flap vaginoplasty, those with higher confluence may benefit from a perineal prone approach to achieve adequate exposure for pull-through vaginoplasty. This prone approach to the high urogenital sinus anomaly can be performed without division of the rectum, provides excellent exposure of the high confluence even in small children and does not preclude the use of posterior flaps for vaginal reconstruction.

Key Words: abnormalities, urethra, vagina, urogenital system

Urogenital sinus abnormalities occur in a wide spectrum (fig. 1). Vaginal reconstruction in these patients can be one of the most complex tasks of the pediatric urologist. This procedure is particularly difficult when the vaginal confluence is high and near the bladder neck. Hendren and Crawford recognized that the variable anatomy of children with urogenital sinus abnormalities necessitates different operative procedures.1 Their classic description in 1969 of the perineal vaginal pull-through procedure for the vagina inserting into the urethra proximal to the external sphincter was a major advance and it remains the basis for reconstruction in these children today. At times such a procedure can be complex. It can also leave an isolated vagina on the perineum, and skin flaps may become stenotic and not produce a functional vagina.

Separation of the vagina from the urogenital sinus is the most challenging portion of the operation, which can be made more difficult by limited exposure. If this separation is not exact, the result may be injury to the urethra or vagina. Resultant urethral complications include stricture, fistula or diverticulum formation from the retained distal vagina. Vaginal injury may potentially lead to stenosis or larger flap requirements. The difficulty of the dissection, which is probably more difficult than any other part of the repair, has led surgeons to recommend delayed vaginal reconstruction until the structures are larger, exposure is better and the sphincter is more easily located.2

Pena et al have made efforts to improve the ease and
results of the procedure in these most challenging patients by using a posterior sagittal approach. However, they performed total division of the rectum and temporary colostomy. We report the use of a prone approach to the high urogenital sinus without division of the rectum or fecal diversion.

MATERIAL AND METHODS

In the last 5 years, all patients with urogenital sinus abnormalities underwent reconstruction at a single stage. In those referred as neonates reconstruction was done in early infancy regardless of the location of the vagina. Eight patients 6 months to 25 years old with urogenital sinus anomalies who presented with high confluence comprise the population of this study (see table). Dumanian and Donahoe have termed high confluence high vaginal atresia. We performed reconstruction in these patients using a posterior prone approach. The older patients were referred to us at that late age. Four patients had pure urogenital sinus abnormalities with nearly normal external genitalia and the remaining 4 had ambiguous genitalia. Three patients had congenital adrenal hyperplasia and 1 had a urogenital sinus anomaly associated with complete genital transposition. Each patient was evaluated promptly by ultrasonography of the abdomen and pelvis, genitography and thorough endoscopic examination. In all patients vaginoplasty and closure of the urethra (urogenital sinus) were done via a posterior midline approach in the prone position. One patient required abdominal transtrigonal vaginal mobilization as well. In those with ambiguity clitoroplasty and labioplasty were performed at the same setting.

OPERATIVE PROCEDURE

The patients underwent bowel preparation with Golytely and received preoperative antibiotics. Endoscopy was performed to evaluate the exact anatomy. In some patients a Foley catheter was inserted into the bladder and a Fogarty catheter was placed through the urogenital sinus into the vagina. A marked sponge was placed in the rectum during the procedure. The entire lower half of the patient was prepared with povidone-iodine from the xiphoid process to the toes. The legs were wrapped and the patient was placed through the aperture in the drapes, allowing access to the entire perineum and abdomen, and easy turning from the supine to the prone position. If there was a pure urogenital sinus abnormality with normal external genitalia, the patient was placed in the prone position with the legs spread and pelvis elevated on rolls. If there was significant clitoral hypertrophy, the phallus was first repaired with the patient supine. Vertical incisions were made on the ventral aspect of the clitoris, leaving the urethral plate attached to the glans for improved cosmesis and blood supply (fig. 2, B). The remainder of the preputial skin was maintained for potential use as the anterior flap to the separated vagina. If the urethra was severely masculinized, its distal portion was opened and incorporated into the anterior flap, as described by Passerini. The erectile tissue of the phallus was excised by a lateral approach from the glans back proximal to the corporal bifurcation with care not to disturb the neurovascular bundle. The glans was secured to the pubis. At that time the patient was rotated to the prone position. In either situation a posteriorly based U incision was then made with its corners on either side of the perineal body near the rectum. Flap length was determined by the distance necessary to reach the posterior vagina. It was made generous enough to allow tension-free anastomosis without compromising the perineal body. The flap was retracted posteriorly and dissection was continued in the midline between the rectum and urogenital sinus (fig. 3, A).

As dissection proceeded, the rectum was easily retracted with a Deavor retractor and never required incision or division. The urogenital sinus was opened posteriorly in the midline from the meatus to the vaginal confluence (fig. 3, B). The distal atretic vagina was then opened on its posterior aspect into the more normal proximal vagina. With a retractoren then placed in the vagina and directed upward the critical aspect of the confluence was optimally exposed to facilitate dissection of the anterior wall of the vagina away from the proximal urethra and bladder (fig. 4, C). This position also provided excellent vision for tubularization of the urethra, which was done in 2 or 3 layers with fine absorbable suture over a Foley catheter (fig. 3, D). Generally healthy fatty tissue was brought from either side together in the midline to separate the urethra and vagina. The high vagina usually has an atretic distal portion, which was opened anteriorly and posteriorly in the midline well up into the normal proximal vagina (fig. 4). The posterior perineal flap was sutured to the opened posterior vagina. When a preputial flap was available, it was sutured to the opened anterior vagina (fig. 5, A). If not available, a labial flap was easily constructed to reach the anterior vagina (fig. 5, B). This flap may be developed on either side. In patients with ambiguous genitalia the labial-scrotal tissue was widely mobilized and moved posteriorly as a Y-V plasty to create labia majora lateral to the vagina (fig. 6). In several cases this cosmetic step was performed secondarily if a labial flap had been used to reach the anterior vagina. The Foley catheter remained indwelling and a Penrose drain was placed in the vagina. The rectal sponge was removed and a dressing was placed over the entire perineum.

RESULTS

Followup in this group of patients ranges from 6 months to 5 years. Six of the 8 patients underwent endoscopy 1 to 6
FIG. 3. A, posterior flap is elevated and dissection proceeds in midline, exposing urogenital sinus. B, retractor elevates intact rectum and urogenital sinus is opened in midline posteriorly. C, distal atretic vagina is opened posteriorly and retractor is placed into vagina to expose its anterior wall confluence with urethra. D, with vagina now mobilized opened urogenital sinus is closed in 2 or 3 layers to construct urethra.

FIG. 4. A, posterior vaginal wall is opened. B, anterior vaginal wall is opened.

months after repair. In 1 teenage patient a tiny urethrovaginal fistula noted on cystoscopy has not been clinically significant, since there has been no wetting or infection and she does not desire repair. In 4 patients daily vaginal dilation was started in the postoperative period until endoscopy and in 3 it was stopped. An older patient with mild vaginal stenosis continues routine dilation and has successful intercourse. Cosmesis has been excellent. One patient died 1 year postoperatively of unrelated causes.

FIG. 5. Coverage of neourethra and creation of anterior vaginal wall. A, in pure urogenital sinus abnormalities lateral flap is developed based anteriorly. Occasionally posteriorly based flap is necessary to open vagina. B, in ambiguous genitalia phallic skin is used to construct anterior flap.

DISCUSSION

Urogenital sinus abnormalities occur in a wide spectrum. Powell et al classified the types as I—labial fusion, II—distal confluence, III—proximal or high confluence and IV—absent vagina. The proper surgical approach should be chosen to fit the needs of each case. The key early component in successful
repair is defining the anatomy well. In all patients ultrasound of the abdomen and pelvis is done to identify the uterus, vagina and gonads, and rule out significant upper urinary tract pathology. Genitography is performed by injecting contrast medium through the urogenital sinus meatus. It is imperative that the radiologist understand the need to define the level of the confluence correctly. Endoscopy is then performed and at times it may be a lengthy procedure. In the patient with extreme masculinization the vaginal entrance may be obscure and evident only by a few small openings in the proximal urethra. If the vagina was not seen on genitography, a catheter may be left indwelling at endoscopy and contrast material may be injected later. If the external sphincter is visualized, its relationship to the vaginal take-off is noted.

Vaginoplasty for urogenital sinus abnormalities generally has 3 forms. Cutback vaginoplasty should be done in only the mildest forms of urogenital sinus anomalies, where there is simple fusion of the posterior vaginal introitus. Most surgeons perform posterior flap vaginoplasty for all other urogenital sinus abnormalities (low confluence), except those proximal to the external sphincter (high confluence). High confluence has been better treated with a vaginal pull-through procedure. The choice between these latter 2 procedures is important. The use of simpler flap vaginoplasty should not be overextended to the high proximal vagina, because in reality flap vaginoplasty leaves the confluence of the urethra and vagina in the same position and simply increases the diameter of the distal sinus. It is an appropriate choice only when the confluence is adequately near the perineum. Its application in a case of high confluence leaves the potential for vaginal pooling of urine and urinary tract infection. Careful endoscopy is critical in making a proper choice between flap vaginoplasty and true vaginal mobilization. However, the surgeon should not hesitate to change from a planned flap vaginoplasty to the more aggressive pull-through procedure if surgical exploration defines a higher confluence than expected. With that in mind we now prepare and drape the whole lower body of all patients with urogenital sinus abnormalities to allow rotation to the prone position as indicated.

The basic principle of surgical reconstruction of the high urogenital sinus was detailed in the landmark report by Hendren and Crawford. Most significantly, they noted that cutback vaginoplasty was ill advised for the high urogenital sinus, since it may result in incontinence, anastomosis under tension or a short urethra. Separation of the vagina from the urogenital sinus and a vaginal pull-through procedure avoid those complications. While this method was a tremendous advance, concerns regarding the procedure became apparent with time. At times this operation resulted in an isolated vaginal opening that was not part of the introitus. Furthermore, the procedure was technically demanding and vision was often limited in the most critical areas. The former concern has been addressed by several surgeons. Gonzalez and Fernandes used preputial skin as an anteriorly based flap to create a more normal appearing vestibule and anterior vaginal wall. Passerini described use of the distal urethra (urogenital sinus) combined with preputial skin to create the whole distal vagina in patients with severe masculinization. Those tissues are more like the normal anterior introitus in appearance and they provide excellent coverage of the urethral closure. Even with these advancements repair remains complex and often quite difficult. To improve the exposure and ease of the procedure Hendren and Atala reported attempted retraction of the rectum laterally but it was too difficult. We have not tried lateral mobilization due to these warnings. They also mentioned a similar posterior prone approach mobilizing the anterior wall of the rectum. Pena et al applied the posterior sagittal approach for cloacal anomalies to the high urogenital sinus, which provided excellent exposure but required division of the rectum and temporary colostomy. We have also found that the posterior midline approach allows optimal exposure but we have been able merely to retract the rectum posteriorly without dividing the rectum. Opening the entire length of the urogenital sinus adds significantly to exposure of the confluence. At times it may seem tempting to leave the distal sinus intact to decrease the length over which a fistula may develop. While that concern is worthwhile, the critical area of repair is at the confluence and that area has the greatest risk for fistulization or stricture. It is our belief that anything done to improve exposure to that area and the quality of the closure at that level is justified and, therefore, we have opened the whole sinus posteriorly. We have used this approach in pure urogenital sinus abnormalities and in the high urogenital sinus associated with congenital adrenal hyperplasia, although only about 5% of patients with congenital adrenal hyperplasia have high vaginal atresia. In patients with ambiguous genitalia we think that vaginal reconstruction is best combined with clitoral recession and reconstruction of the labia at a single stage. At that time the redundant phallic skin may be used for vaginal reconstruction, providing flexibility for the creative surgeon. Options are more limited if the labial and phallic skin has been previously mobilized in a staged repair. We have generally preserved the ventral phallic skin with the glans and have found that a modified Gonzalez preputial flap for the anterior vaginal area results in a much more normal appearing introitus. For the pure urogenital sinus abnormality the anterior vaginal flap is easily constructed from the labia and perineum. The flap can be based anteriorly or posteriorly. At times we have used 1 flap from each side to provide coverage of the urethra and create a distal vagina of good caliber. We have found that endoscopy in the early postoperative period is quite helpful in these highly complex cases, allowing assessment of the healing of urethral and vaginal reconstruction. If there is concern regarding urethral healing, the catheter remains indwelling for a longer period. Vaginal calibration can also be done. In the past with other repairs vaginal dilation was somewhat routine. Early in this series we performed vaginal dilation but we have not found vaginal stenosis to be significant except in 1 case. We now think that dilation is not in the best interest of the child and we recommend it only when significant stenosis has developed. Nearly all others have recommended delayed vaginal reconstruction when there is high confluence until the child and urogenital sinus structures are larger. Delay usually necessitates a second operation and may prevent use of the
phallic skin for the anterior flap. We have performed total reconstruction in patients with high confluence at age 6 months and found that this posterior approach allows the procedure to be done easily. Vision is excellent. Donahoe and Gustafson recently reported their experience reconstructing high vaginal atresia in 3 patients 8 to 12 months old. Cos- 

metic and functional results were favorable, and the tissues were more mobile in infancy. DeJong and Boemers recently reported 1-stage clitorovaginoplasty done before age 3 weeks. They noted that maternal estrogens produced vaginal enlargement and cleavage planes were more easily identified. We have also found early reconstruction to be easily performed, since good exposure was obtained via this posterior prone approach. It has been no more difficult than in older patients and we agree that tissues are more mobile. Clearly there is more flexibility for the surgeon when all tissues are available at single stage repair.

CONCLUSIONS

Reconstruction of the high urogenital sinus by a vaginal pull-through procedure has usually been delayed due to complexity of the repair. The posterior prone approach allows better exposure for dissection of the vagina from the urethra and urethral closure, and it does not require division of the rectum. Repair in infancy is no more difficult than a delayed procedure. Therefore, the entire genital reconstruction can be done at a single stage. Although ultimate success will take many years to determine, our early results with this approach have been good.

REFERENCES