ANORECTAL ANOMALIES IN THE FEMALE

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Malformations of the anus and rectum are among the most difficult lesions treated by pediatric surgeons. The classification and embryologic basis for imperforate anus are controversial; comparison of results reported by various surgeons is difficult because of different evaluation techniques. In the surgical literature, more attention has been focused upon the male with the high lesion than upon female anomalies. This is partly because the most frequent anorectal defects in females are “low lesions” for which one assumes a good result is always obtained. Unfortunately, in a review of the experiences with imperforate anus at the Cook County Children’s Hospital and the Children’s Memorial Hospital in Chicago, I found a number of girls born with an anorectal anomaly who had a poor long term result. In all of these children, there had been an error in diagnosis which had led to a poorly planned initial operation and complications including incontinence, constipation and urinary tract deterioration. The most frequent initial error was in confusing a rectovestibular fistula with an anovestibular defect. A perineal anoplasty for a rectovestibular anomaly often led to retraction of the bowel, stricture and numerous secondary operations. The second most common error was in leaving a largely dilated “megarectum” which led to intractable chronic constipation. Finally, we have observed that when a simple colostomy is done for a cloacal malformation and the urinary tract is not separated from the vagina, the result is urinary tract sepsis with deterioration of the kidneys and ureters. This paper reviews my own experiences with anorectal malformations in the female and is based upon experience over the past 20 years at the Cook County Hospital and The Children’s Memorial Hospital in Chicago. I have identified these three problems and have attempted to circumvent them over the past 10 years. When possible, these patients are classified according to the international classification proposed by Santulli and Kiesewetter (1) (figure 1).

Diagnosis

An infant born with imperforate anus requires immediate, complete examination to determine the presence of associated esophageal, cardiac, vertebral and gastrointestinal anomalies. Initially this consists of the passage of a radiopaque nasogastric tube together with a roentgenogram of the chest and abdomen. The diagnosis of a specific anorectal anomaly can usually be made by examination of the perineum together with abdominal palpation. The perineum is visually inspected with a good light while the infant is held in the lithotomy position. Next the vagina is examined with a small nasal speculum. The urethra is located and catheterized and finally, if an anal opening is observed, it too is catheterized. The rectum is irrigated with warm saline to aid in the initial passage of meconium. If the urethra is identified together with a separate vagina from which one can irrigate meconium, there is a high rectovaginal fistula. A single perineal opening indicates a cloacal malformation; a palpable lower abdominal mass suggests an associated hydrocolpos. The usual invertogram is of no use in establishing the level of the rectal pouch in girls. However, contrast material injected into the vaginal, cloacal, or anal orifice will aid in the arrival at a correct diagnosis. A cystogram and intravenous pyelogram are always obtained in every patient with an anorectal anomaly. Associated genitourinary anomalies such as vesicoureteral reflux or an absent kidney are rarely found in girls with low lesions. All of our

(80)
patients with a high rectovaginal fistula or a cloaca had associated urinary tract anomalies. If a satisfactory visual examination cannot be carried out initially, then the infant must again be examined with a speculum and, if necessary, by endoscopy under anesthesia prior to any surgical therapy.

Treatment and Results
We shall consider treatment and the long term results together in our discussion of each category of patients.

Anterior Perineal Anus
The anterior perineal anus opens upon the perineum between the anal dimple and the vestibule. There is dry perineal skin between the anus anteriorly and the genital tract (figure 1-a). There may be some protrusion of mucosa from the anus because this is not a true anal canal with only a rudimentary external sphincter. If there is no associated stenosis, no treatment is indicated. Otherwise, a minor degree of narrowing is merely dilated. More significant degrees of anterior displacement and those associated with stenosis are treated with V-Y modification with a classical anal "cutback" operation. An inverted V-shaped flap of skin is dissected backwards from the anal opening. The mucosa is then incised in the midline, but any muscular fibers are pushed back and saved. The flap of skin is then sutured into the mucosal incision. The resulting anal canal is skin-lined and requires only minimal post-operative dilatations. This procedure is illustrated in figure 2. Minimal anterior displacement of the anus may result in constipation in older children (2). The V-Y cutback will also alleviate these symptoms.

Anovestibular and Anovestibular Fistula
It is extremely difficult to differentiate the anovulvar from an anovestibular fistula, and both are considered together (figure 1-b). In each, the anal opening is often invisible upon perineal inspection. However, when the vulvae are separated with an anal speculum, the anal orifice is found behind the margin of the vestibule. The urethra and vagina are normal. A catheter or hemostat inserted into the anal opening passes backward along the perineum, indicating that there is a short fistula and that the rectum has penetrated the levator sling to lie close to the perineal dimple. Barium studies demonstrate a rectum of good caliber, although it may be compressed by the levator sling (figure 3). Some surgeons, particularly Stephens, recommend either a simple cutback or a sacroperineal transplantation for an anovestibular fistula (3). Others have been disappointed with the results of a cutback operation because there is no perineal body, but only a thin, moist membrane between the posterior vaginal wall and the anterior border of the anus. Our best results with an anovestibular fistula have followed a formal perineal anoplasty. If the infant is healthy, the procedure can be carried out in the neonatal period. However, if the anal opening is large enough for the passage of stool, the operation may be delayed indefinitely. Currently we perform the operation between three months and one year of age.

Technique for Perineal Anoplasty
The child is given a non-residue elemental diet, rectal irrigations, and 100 mg/kg of Neomycin by mouth 24 hours prior to the operation. After induction of anesthesia, the surgeon must again make certain that it is an anovestibular and not a rectovestibular lesion. The rectum is irrigated with a Betadine solution. The operation is illustrated in figure 4. The initial incision about the fistula includes a bit of skin to hold the 6-0 traction sutures. The dissection between the vagina and anus is commenced before going lateral to the rectum (figure 4-g). The vagina is retracted upward with small skin hooks and the rectum is retracted downward with the traction sutures. Sharp scissors dissection will be aided by a headlight and magnification. Oozing of blood in this operation is controlled by cotton pledgets soaked in 1:100,000 adrenaline and with a needle tip electrocautery. The lateral dissection around the rectal wall is started at least 2-3 cm proximal to the fistula. All dissection must be carried out immediately on the rectal wall and within the levator sling. By starting the lateral dissection from below, it is difficult to avoid the lowermost fibers of the levator sling because they intermingle with those of the muscular coats of the bowel. When one dissects between the vagina and rectum up to the
plane of loose areolar tissue, one may proceed laterally and posteriorly and constantly remain within the levator muscle. A nerve stimulator must be utilized to identify the levator as well as remnants of the external sphincter. The lower posterior fibers of the levator muscle are pushed back just enough to allow transplantation of the anus backward to the anal dimple. The bowel is then brought through the anal sphincter muscle and sutured to the skin flaps. This results in a skinline canal. When this operation is properly performed for an anovestibular fistula, one can expect normal continence and no constipation in all patients. Unfortunately, in the past, my own attempts to perform this operation for a rectovestibular or higher lesion ended in disaster. Frequently a very high dissection was necessary, and the anastomosis performed under tension separated and made it necessary to perform further operations. Some of these patients continue to be incontinent of stool into their teen years.

Rectovestibular and Low Rectovaginal Fistula

A hemostat passed into a rectovestibular fistula (figure 1-c) passes upward almost parallel to the plane of the vagina. It does not reach back to the anal dimple, indicating that the rectum itself has not completely penetrated the levator sling. A perineal anoplasty is contra-indicated in this lesion. Some infants may stool satisfactorily through a rectovestibular fistula, but some with this lesion, and all infants with a higher rectovaginal fistula, should have a colostomy in the neonatal period. I feel that the endorectal pullthrough operations described by Rehbein and Roumaldi are ideal for this lesion (7,8). The dissection is within the muscular layers of the rectum and hugs the back wall of the vagina until the perineum is reached. In our hands, the sacroperineal approach is more difficult to perform and offers greater risk of injury to the vagina and the levator muscles. The endorectal pullthrough has the further advantage of resecting any hugely distended rectum, which leads to intractable constipation if left behind. The technique for an endorectal pullthrough for a rectovestibular fistula is illustrated in figure 5. If a preliminary colostomy has not been performed, the bowel is prepared by feeding the patient an elemental diet and Neomycin. In addition, the bowel is irrigated several days prior to the operation and after the induction of anesthesia. The dissection of mucosa away from the muscular coats of the bowel is commenced at the peritoneal reflection. This is performed with small cotton pledgets while an assistant coagulates all vessels between the muscular levators and the mucosa with a needle tip electrocoagulator. When the mucosa has been removed from the entire length of the rectum and the fistula, a tunnel is made beneath the perineal skin through the external sphincter at the anal dimple. A nerve stimulator is used to identify all perineal muscles. The pulled-through bowel is directed through the muscular tunnel within the levator sling; then, posteriorly to the newly created anus through the fibers of the external sphincter. With this operation, there has been no difficulty with separation of the anastomosis or recurrent fistulae. Dilatations of the sphincter are necessary in the postoperative period, but continence in these children has been achieved by 2–3 years of age and constipation has not been a problem.

Cloaca

There are a wide variety of cloacal malformations in which the rectum, vagina and bladder all join in a common channel which opens on the perineum (figure 6). In some, the cloaca appears to enter the rectum while the vagina joins through a narrow opening. In this latter situation, the vagina is distended with secretions and urine, and is often palpable as a separate suprapubic mass. Septate vaginae and double uteri represent a lack of fusion of the mullerian ducts. In our own original series of children with a cloaca, 13/15 cases had severe urinary tract anomalies (9). The diagnosis of a cloaca is made by the observation of a single opening on the perineum. The labia and clitoris are abnormal and the perineum is shortened. There is usually no anal dimple (figure 7). Plain abdominal roentgenograms demonstrate colonic distention and a pelvic mass. If there is a wide communication between the rectum and the obstructed vagina, it will contain an air fluid level representing meconium and intestinal gas. Contrast material injected in the perineal orifice may demonstrate the anatomy on lateral films. Figure 8 illustrates a dilated vagina and the bladder, but no contrast entered the rectal orifice which was severely stenotic. In our hands, roentgen studies and endoscopy have failed to demonstrate the exact anatomy in many infants with cloacal malformations. For this reason, the exact anatomy must
be worked out at surgery by catheterizing each orifice and opening the bladder, vagina and rectum to find their site of junction. Hendren has confirmed our own dismal prognosis in these children when an inadequate operation is performed in the neonatal period (10). A simple colostomy leaves the undrained vagina, which fills with urine and becomes infected. Unless the bladder, vagina, and rectum are separated and each organ given unobstructed drainage, there will be chronic infection, deterioration of urinary function, and eventual infection and loss of the uterus and fallopian tubes. If the infant is too ill for complete correction of this anomaly a colostomy, vesicostomy, and vaginostomy should be performed to provide complete drainage. In some infants, however, there is a partial separation of the vagina from the bladder and adequate drainage. If the vagina is not distended and if the urethral orifice is low on the anterior wall of the cloaca, a colostomy will be sufficient in the newborn period. Figure 9 illustrates the operative repair of an infant with a high rectal atresia, hydrocolpos, septate vagina, and cloaca. The bladder is opened and catheterized from the perineal opening, which is left as the urethra after closure of the vagina fistula and pullthrough of the vagina and rectum to the perineum. It is necessary to dissect very carefully directly along the back wall of the cloaca in order to stay within the levator sling. The child with a short cloaca and an undistended vagina requires only a rectal pullthrough operation. We have now followed seven patients who were operated upon for a cloaca more than five years ago. Four are continent of both stool and urine and are free from urinary tract infections. One girl is continent of urine but requires daily enemas to prevent fecal soiling. The other two are incontinent of urine and are on self-catheterization programs. They also require daily enemas to prevent fecal soiling. All of these children have normal ureters and kidneys; only those on selfcatheterization programs require long term antibiotics for chronic urinary tract infections. These results are not perfect, but are an improvement over those obtained by delayed or incomplete surgical treatment.

In our long term followup of children with anorectal anomalies, we have now identified five children who had perineal repairs for a low imperforate anus but who have severe intractable constipation. All of these children have massive megarectums (figure 10). Diet, laxatives, and chronic enemas have failed to help these children. It has been necessary to perform an endorectal pullthrough with a resection of the massively distended bowel. A preliminary colostomy is necessary if the bowel cannot be emptied by giving the child an elemental diet and irrigations at home or in the hospital. It is my feeling that this complication may be avoided in the newborn period by resection of the distended terminal rectum.

The care of a child with an anorectal anomaly continues for many years. During the first months following the operation the surgeon must dilate the rectum at frequent intervals. It is often helpful to examine and dilate the rectum under anesthesia at least once. At that time redundant tags of mucosa may be excised or, if there is a tight stricture, it may be opened with a minor anoplasty. An intelligent child with well motivated parents who has no neurologic impairment can often be toilet trained by 2½–3 years of age, even when a very high lesion was treated. Every effort should be made to have the child continent before she attends school. When there is neurologic impairment or a serious deficiency in the levator muscle, a small castile soap enema administered in the morning will often induce a bowel movement and keep the child clean during the day. An older incontinent patient can be taught perineal exercises to strengthen any fragments of levator muscle which may be present. The morning enema becomes a part of this exercise as the child is urged to tighten her perineum to hold in the enema fluid. Every effort is made with diet and with a drug such as loperimide to induce a firm stool, since this helps to achieve continence.

Finally, a gracilis muscle transplantation has been useful in patients who are well motivated and intelligent. I have withheld this operation until the child is 10–12 years old, and after all other measures have failed. It is helpful in slightly more than half of the patients operated upon. Continued diligent, patient supervision of the post-operative care is perhaps as important as operative technique in attaining a satisfactory long term result.

This review of anorectal anomalies is far from complete, but I have reviewed by own conclusions which have been drawn from observations of my patients, as well as those operated upon by colleagues at The Children’s Memorial Hospital.
Figure 1  Female anomalies.

A. Anoperineal. The rectum has penetrated the levator musculature. There is an ectopic opening on the perineum between the posterior vagina and the external sphincter.

B. Anovestibular. The rectum has penetrated the levator musculature. There is a short fistula to the back wall of the vestibule.

C. Rectovestibular. The rectum has not penetrated the puborectalis sling. There is a fistula to the lower portion of the vagina which is long and narrow.

D. High rectovaginal. The entire rectum is proximal to the levator ani. A fistula enters high into the vagina. Note that the puborectalis muscle encircles the vagina.
A. Anterior perineal anus.

B. The inverted V-shaped skin incision is outlined on the skin.

C. Completed V-Y anoplasty.

Figure 2

Figure 3 Anovestibular fistula. Barium enema demonstrating that the rectum has completely penetrated the levator sling, and is compressed. Note the dilated rectum above the levators.
Figure 4  Perineal anoplasty.
A-C. Technique of perineal anoplasty. The cruciate incision over the external sphincter is used in all operations for imperforate anus. The skin flaps are undermined and the external sphincter is identified with a nerve stimulator.
D-F. The anovestibular fistula is circumscribed with sharp dissection and multiple 5-0 traction sutures are inserted through the end of the bowel.
G. The bowel is dissected from the vaginal mucosa and then dissected laterally and posteriorly.
H. When several centimeters of the bowel have been mobilized, the external sphincter is bluntly dissected while the muscle fibers are carefully indentified with a nerve stimulator. The tunnel from the external sphincter to the site of the anovestibular fistula is subcutaneous, so as to avoid injury to the levator ani.

I–J. The bowel is then pulled through and sutured to the sphincter.

K–L. The original cruciate skin flaps are then sutured up in the anal canal to provide a skin-lined anus.
Endorectal pullthrough for a rectovestibular or rectovaginal fistula.

A. The operations commence with a low transverse abdominal incision.
B. The seromuscular layer is opened circumferentially to the mucosa.
C. The mucosa is removed distally, leaving the muscular layers of the bowel intact. When all rectal mucosa has been removed, a tunnel is made beneath the perineal skin to the anal dimple, distal to the lowest fibers of the levator sling.
D. The proximal bowel is pulled through the seromuscular cuff and the levator sling. Finally, the skin is sutured to the bowel.
This is only one type of cloacal malformation. The rectum, vagina and bladder all join together in a common channel. There are several variations of this basic pathology, however.

Figure 6
A cloaca. There is only one perineal opening for urine, stool and vaginal secretions.

Figure 7

Figure 8  Cloacal anomaly.
A. These roentgenograms demonstrate the bladder and a dilated vagina. The rectum which joined the cloaca was stenotic and didn’t fill with barium.
B. The intravenous pyelogram demonstrated mild hydroureter and filling of both the bladder and vagina. This girl had only a colostomy in the neonatal period. She had continuing urinary sepsis and now, at 17 years of age, has an ileal conduit and severely reduced renal function.
Figure 9  Correction of a cloaca.

A–H. The abdomen is opened through a transverse lower abdominal incision and the much distended vagina is brought into the operative field. If there is a vaginal septum, it is excised. A catheter is passed through the urogenital sinus and the fistula between the vagina and the sinus is circumscribed and closed with three layers of polyglycolic sutures. The rectum is separated from the back wall of the vagina, and the vagina and rectum are pulled through the puborectalis tunnel to the perineum.
Closure of the rectal-urogenital sinus opening

Reconstructed anal opening
Figure 10

Megarectum in a 4 year old child who had a perineal anoplasty as a newborn. Enemas and diet partly relieved the severe constipation, but an endorectal pullthrough was required. When a "megarectum" is identified in the neonatal period, a proximal colostomy will not reduce its size. It must be resected at the time of definitive repair.