The Treatment of Imperforate Anus: Experience With 108 Patients

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Background/Purpose: The authors present their experience and results in the treatment of infants with imperforate anus over a 10-year period. Differences between these and previously published western results are noted and discussed.

Methods: One hundred eight patients with imperforate anus were treated from June 1988 to July 1998. Of these patients, 66 were boys and 42 were girls. Associated anomalies include congenital heart disease, anomalies of bone and cartilage, and Down's syndrome. Thirty-five patients with a low lesion received a limited posterior sagittal anorectoplasty. Seventy-one patients had a high lesion and received 3-staged operations including colostomy, posterior sagittal anorectoplasty, and takedown of colostomy. All patients underwent follow-up by the author. Postoperative anorectal function was evaluated based on the following criteria: ability to have voluntary bowel movement, soiling, and constipation. The duration of follow-up ranges from 6 months to 10 years.

Results: One patient died of multiple congenital anomalies after colostomy. One patient died of hyaline membranous disease. All except 2 patients had voluntary bowel movement. Three patients had soiling, and 19 suffered from constipation after operation. The constipation improved with medical treatment and time. Four patients who received the first operation at another hospital (3 underwent posterior sagittal anorectoplasty and 1 had cutback anoplasty) had problems with soiling. In these patients, soiling improved after redo posterior sagittal anorectoplasty.

Conclusions: Utilizing the posterior sagittal operation described by Peña, most patients were continent and able to have voluntary bowel movements. Constipation occurred in a substantial number of patients with high-type lesions, but few of these patients needed medication or enemas. There were significantly fewer sacral and urogenital anomalies than have been reported in most western series. This may explain the excellent results.

INDEX WORDS: Imperforate anus, posterior sagittal anorectoplasty.

IN THE LATE 1960s, Stephens and Smith[1] proposed a radical operation for imperforate anus in which great importance was placed on puborectal muscle function. However, fecal incontinence or sphincter dysfunction developed frequently after this operation.[2,3] Peña and deVries[4] described their posterior sagittal anorectoplasty (PSARP) for the treatment of imperforate anus in 1982. The advantage of this procedure is the extensive exposure allowing the meticulous anatomic repair of all the muscular structures present to achieve optimum continence.[4]

Our hospital opened in June 1988, and we treated our first patient with imperforate anus that month. Since then, we have completed the treatment course of 106 patients with imperforate anus utilizing the PSARP. Postoperatively, these patients underwent follow-up by the author for evaluation of the anorectal functions.

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MATERIALS AND METHODS

Of the 108 patients, 66 were boys and 42 were girls. The types of defects in boys are listed in Table 1. Nineteen boys had a low defect, and 46 had a high-type defect. Nineteen of high-type cases were rectobulbar fistula and only 3 patients had a rectovesical fistula. In 6 boys, no fistula was found on physical examination at birth. Classification of girls is shown in Table 2. Thirteen girls had a low defect. Thirteen patients had a rectovestibular fistula. Two patients had a persistent cloaca with a short common channel. In 2 patients, a colostomy was performed at another hospital. Three patients were admitted for severe constipation, soiling, and excoriation of perianal skin after PSARP at another hospitals. A single surgeon performed all the other operations.

After the diagnosis of imperforate anus is made, a nasogastric tube is inserted to decompress the stomach and rule out the possibility of esophageal atresia and tracheoesophageal fistula. A whole-body x-ray is taken to evaluate the spine, the sacrum, extremities, the chest, and the abdomen. A detailed physical examination is performed, looking for a fistula, the contour of buttocks, and contraction over the anal dimple. The cardiac and renal echography is performed before or after operation, depending on the clinical condition of patient. Ultrasonography of the lumbosacral spine is done on some patients with high malformation with suspicious physical or x-ray findings. In patients in whom the level of the defect cannot be determined, an invertogram of Wangensteen and Rice[5] is performed.

For a low defect, a limited PSARP is done. Two patients with an anal membrane were treated with membrane resection and simple perineal anoplasty. For high-type lesions, a double-barrel colostomy is performed initially, and then an augmented-pressure distal colography[6] is performed to identify any fistula when the infant reaches 5-5.5 kg of body weight. PSARP is done 1 day after distal colography. Three

Table 1. Classification of Male Cases

Low defect	22
Cutaneous fistula	19
Anal stenosis	3
Rectourethral bulbar fistula	19
Rectourethral prostatic fistula	12
Rectovesical fistula	3
Imperforate anus without fistula	6
Rectal atresia and stenosis	4
Total	66

patients with rectovesical fistula needed laparotomy along with PSARP to divide the fistula and complete the pull-through procedure. Posterior sagittal anorectovaginourethroplasty was performed for 2 patients with persistent cloaca. After PSARP, patients undergo follow-up regularly at my outpatient clinic. Stitches were removed in the operating room on the 12th postoperative day. Strength and symmetry of the contraction of the muscle complex and the parasagittal muscles is assessed with a muscle stimulator. Anal dilatation is begun 2 weeks after PSARP, and the family is taught the technique of home dilation. Takedown of colostomy is performed 6 to 8 weeks after PSARP. The muscle complex is evaluated again with the muscle stimulator. Long-term follow-up by the author included (1) presence or absence of soiling, its frequency and amount; (2) the quality of defecation such as consistency, frequency, amount, and regularity; (3) presence or absence of diarrhea and its frequency and consistency; (4) awareness of the presence of feces in the rectum; (5) presence of perianal skin breakdown and its severity; and (6) urinary continence. Most data were obtained by report from the parents because of the young age of the patients. The average duration of follow-up is 3 years. Ultrasonography of lumbosacral spine was performed in some patients with high lesion that had slower progress in anorectal function. No patient received magnetic resonance imaging (MRI) of lumbosacral spine because no patient was found abnormal by ultrasonographic examination. In patients with longer follow-up, growth and development was assessed, and the social adaptation was evaluated.

RESULTS

Associated Anomalies

The frequently associated anomalies are listed in Table 3. Cardiovascular anomalies were present in 20 cases (18.5%). The most common lesions were ventricular septal defects and atrial septal defects. Endocardial cushing defect, tetralogy of Fallot, pulmonary atresia, and double-outlet of right ventricle also were found. Bone or cartilage malformations occurred in 13 cases. The affected bones varied including polydactyly, microtia, absence of bilateral thumbs, absence of 1 radius, phacomelia of upper and lower extremities, hemivertebrae, and severe kyphoscoliosis. No patient was found to have spinal dysraphism, and only 1 had a sacral anomaly. Seven cases were associated with esophageal atresia and tracheoesophageal fistula (TEF). Ten patients had Down's syndrome. Five patients had hypospadias. Four patients had cryptorchidism, and 4 had inguinal hernia. Other less frequent anomalies included duodenal atresia, malrotation, persistent cloaca, transverse septum of the vagina, cleft lip, cleft palate, and right kidney agenesis (Table 4).

Table 2. Classification of Female Cases

Low defect	13
Cutaneous fistula	9
Anal membrane	2
Anal stenosis	2
Vestibular fistula	13
Vaginal fistula	7
Rectal atresia and stenosis	2
Imperforate anus without fistula	5
Persistent cloaca	2
Total	42

Table 3. Frequently Associated Anomalies

Congenital heart disease	20
Bone or cartilage malformations	13
Esophageal atresia and tracheoesophageal fistula	7
Down's syndrome	10
Hypospadias	5
Cryptorchidism	4
Inguinal hernia	4

Operative Results

The operative results are shown in Table 5. One premature patient had multiple congenital anomalies of imperforate anus, esophageal atresia and tracheoesophageal fistula, duodenal atresia, right renal agenesis, nonrotation of bowel, and hypospadias. A cervical esophagostomy, gastrostomy, duodenoduodeostomy, and colostomy were performed, but the patient died of multiple-organ failure on the third postoperative day. One patient died of respiratory failure caused by hyaline membrane disease. All patients who underwent surgery had satisfactory voluntary bowel movement. However, 3 with high-type lesions suffered from soiling. One had a prostatic fistula, the second had a rectovesical fistula, and the last had a persistent cloaca. Constipation occurred in 19 (17.6%) patients. The incidence of constipation was 11% in low-defect group and 26% in high-lesion group. There was no constipation in patients with rectal stenosis.

Two patients received a colostomy at other hospitals, and 1 of them had surgery 14 years before being admitted to our hospital. We performed a PSARP and takedown of colostomy for these 2 patients. The operative result was good in the aspect of voluntary bowel movement, soiling, and constipation. In 2 patients, a PSARP was performed initially at another hospital. Both of these children suffered from very severe constipation, frequent soiling

Table 4. Infrequent Associated Anomalies

Duodenal atresia	2
Abnormalities of intestinal rotation	2
Cloaca	2
Transverse septum of the vagina	1
Cleft lip	3
Cleft palate	3
Microtia	2
Agenesis of right kidney	1
Gastroesophageal reflux	1

Table 5. Operative Results

Anomaly	No.	Voluntary Bowel Movement	Soiling	Constipation
Low malformation	35	35/35	0/35	4/35
Vestibular	13	13/13	0/13	2/13
Vaginal*	7	6/6	0/6	2/6
Rectal Stenosis	6	6/6	0/6	0/6
No fistula†	11	10/10	0/10	2/10
Bulbar fistula‡	19	19/19	0/19	4/19
Prostatic fistula§	12	12/12	1/12	2/12
Rectovesical fistula‖	3	3/3	1/3	2/3
Cloaca	2	2/2	1/2	1/2
Total	108	108	3	19

NOTE. Duration of follow-up, 6 months to 10 years.

*Includes 1 case of redo PSARP with anteriorly misplaced anus, and 1 case died of multiple severe congenital anomalies.

†One case died of hyaline membranous disease.

‡Includes 1 case of redo PSARP with anteriorly misplaced anus.

§Includes 1 case of redo PSARP with anteriorly misplaced anus.

‖Includes 1 case of redo PSARP with left misplaced anus.

of underwear, and excoriation of perianal skin. Physical examination showed an anteriorly misplaced anus and anal dimpling posterior to the anus. In both of these patients we performed the redo PSARP pull-through and found the common wall of posterior urethra and anterior rectum had been inadequately separated at the first procedure. After redo procedure, both patients recovered without soiling and without constipation. The other patient who had a cut-back anoplasty performed 25 years ago suffered from severe constipation. A posterior sagittal anorectoplasty had been done at another hospital 5 years ago, but the patient still had constipation and soiled his underwear. He also had an anteriorly misplaced anus. After redo-PSARP at our hospital his symptoms improved.

Three cases of low defects and cutaneous fistulae were not diagnosed at birth. They were ultimately diagnosed during treatment for severe constipation and progressive abdominal enlargement when the patients were 1.5 to 3 years of age. The operative results were still good in these children with a delay in diagnosis. One boy with a prostatic fistula associated with esophageal atresia and tracheoesophageal fistula suffered from severe constipation after PSARP at our hospital. The symptom persisted after medical treatment. Examination with an electrical muscle stimulator in the operating room at the age of 3 years showed that the anus was right misplaced and outside the muscle complex. We performed a redo operation to relocate the rectum exactly at the midline. The symptoms improved after operation.

Complications

The complications encounted are listed in Table 6. The most frequent complication was constipation, which occurred in 19 patients. In most patients, the constipation was temporary, and the symptom improved within several months. Some patients required dietary adjustment, laxatives, or enemas. Only 2 patients failed to respond to regular enemas. In one patient, the symptoms subsided after a redo operation as described previously. One patient required manual disimpaction weekly after 1 year of age. This was 1 of our early patients. However, his symptoms improved dramatically after 3 years of age. By the time he was 5 years old, voluntary bowel control had developed, and he was free of soiling and constipation. His growth and development were normal, and his social interaction was active and cooperative.

Three patients suffered from soiling. All these patients had high malformation requiring extensive dissection at PSARP. The symptom improved somewhat with time but did not disappear. Three patients had anteriorly misplaced anus, and one patient had right misplaced anus. Their clinical course has been described. Rectal mucosal prolapse was found in 4 patients. Three of them were minimal; only 1 needed a revision. Perianal wound dehiscence occurred in 3 patients. In one patient, the whole circumference of anus disrupted from the perianal skin and retracted 1 cm upward. Emergency operation was performed to repair this dehiscence. Colostoma prolapse happened in 3 patients, 1 with proximal colostomy, and 2 with distal colostomy. In 2 patients, the colostomy prolapse required surgical repair.

DISCUSSION

The most common abnormality of the genitourinary system in patients with imperforate anus is the presence of a fistulous tract between the rectum and the urinary tract in boys and with the vagina in girls.[7] The incidence and severity of the associated genitourinary anomaly is directly associated with the severity of the anorectal malformation as defined by the level of the rectal fistula.[8,9] The embryological defect responsible for anorectal malformations is closely associated with the development of the gastrointestinal and genitourinary systems. This would predict the frequent coexistence of development abnormalities in both systems. Renal agenesis-dysplasia or vesicoureteral reflux were the 2 most commonly encountered major abnormalities in the series of

Table 6. Complications

Constipation	19
Soiling	3
Anteriorly misplaced anus	3
Left misplaced anus	1
Mucosal prolapse	4
Minimal	3
Requiring resection	1
Wound dehiscence	3
Colostoma prolapse	3
Proximal	1
Distal	2

Rich et al.[8] However, we have only 1 patient with right renal agenesis, which may originate from abnormal development of ureteral bud from the mesonephric duct. This patient died of multiple congenital anomalies. We also have 5 patients with hypospadias. These show that the genitourinary anomaly is much less severe in our patients than in those of Rich et al.[8]

It is well known that children with anorectal malformations have a high incidence of associated anomalies of the lumbosacral spine.[10-14] Peña[10] found that the functional outcome concerning anorectal function clearly depended on whether the patients had a normal sacrum. The absence of 3 or more than 3 vertebrae is associated with a severe neurological deficit, including neurogenic bladder and lack of bowel control.[15] In our series, no patients were found to have sacral or lumbosacral anomalies. Two patients had severe kyphoscoliosis. One patient had hemivertebrae of upper thoracic spine. Ultrasonography of lumbosacral spine was performed in these patients, but no sacrococcygeal malformations were found. Currently, in these patients there is no evidence of urologic anomaly and neurogenic bladder. The low incidence of anomalies of the upper urologic tract and sacrum in our patients might partially explain our excellent operative results.

For evaluating the operative results for patients with imperforate anus, the type of malformation and the degree of associated sacral anomaly should be specified. We did not routinely use any radiographic studies or standardized scoring systems to assess the bowel function in our patients. Methods of measuring bowel function such as defecography,[16] rectal manometry,[2] electromyography,[17] MRI,[18] and anal endosonography[19] only evaluate isolated aspects of the very complex physiological mechanism, and; therefore, the results of these objective evaluations often poorly correlate to the quality of life.[10] Clinical scoring methods have been devised by Kelly,[20] Templeton et al,[3] Kiesewetter and Chang[21] and Stephens and Smith[22] according to the degree of continence and the quality of life after management. Kelly's method[20] added strength of puborectalis action on digital examination. However, these methods were developed before the widespread use of the PSARP,[7] and did not use symptoms other than soiling or skin rash for evaluation. Yet, although only 3 of our patients experienced soiling, 19 patients suffered from various degrees of constipation. In this series, constipation was a much more troublesome problem than soiling. In Peña and deVries series, constipation is also a major problem in patients without sacral anomalies.[4] Therefore, our protocol included the postoperative clinical situations and objective observation of strength and symmetry of contraction of muscle complex and parasagittal muscles. The results of this evaluation method correlate much better with the quality of life than other scoring systems and objective methods do. The patients received good marks in our evaluation method and all led a good quality of life.

As previously reported in other series,[23] most of our patients with constipation improved with time after dietary manipulation and other types of medical treatment. However, the symptom in one case improved much later. Some patients improved only after redo operation. If the patient has a normal sacrum, the maturation of the neurological function may be responsible for improvement of constipation. If the rectum is not accurately located in the middle of striated muscle complex in PSARP, the symptom will not subside with time, and redo PSARP is indicated.

Our greatest challenge currently is how to decrease the incidence and severity of constipation after PSARP. It is decreasing over the years with the improvement of surgical techniques. To decrease the incidence of constipation, we adhere to the following principles. (1) Make an augmented-pressure distal colography before PSARP to identify the location of fistula, if present, and the rectal pouch. (2) Do not dissect more extensively than needed. Peña et al[24] proved in the dog that extensive perirectal dissection could impair the rectal function. In the course of PSARP, the distal rectal pouch was distended with normal saline or air by an assistant through a Foley catheter inserted in the distal colostoma similar to the maneuver in the augmented-pressure distal colography. This will promote the visualization of the distal rectal pouch just like the nasogastric tube in the proximal esophageal pouch at the operation for esophageal atresia. Unnecessary dissection around the rectal pouch will be avoided. Use traction sutures instead of forceps to aid dissection and decrease possible damage to the bowel. (3) Do not save the most distal rectal muscle in PSARP. Although Hedlund and Peña[25] proved the most distal rectal tissue has the structure of the internal anal sphincter (IAS) in vitro, others proved that saving of the IAS was not important for continence clinically.[23] Constipation was the major complication of IAS saving PSARP in some series.[26] (4) Keep the plane of dissection exactly at the midline. When a right-handed operator stands to the right side of a prone patient and faces toward the patient's left aspect, he tends to make the line of vertical dissection to the right of the intentional line. We positioned the patients so that the line between buttocks stood vertical to the floor of operating room. Then the dissection was done with the eyesight running in the craniocaudal direction, and the bovi tip was always kept vertical to the floor. With this maneuver, we can keep the dissection strictly at the midline and need only few times of changing direction of dissection by the indication of a muscle stimulator. Our operative results are favorable. The PSARP procedure provides satisfactory results for the majority of our patients with imperforate anus.

We treated 3 patients after a significant delay in diagnosis. They suffered from progressive abdominal distension and severe constipation refractory to medical management. They were diagnosed at my clinic and symptoms relieved after limited PSARP. These patients all had a 3-mm-diameter cutaneous fistula near the normal anal dimpling. They all passed meconium at birth, but developed progressive constipation with time. Detailed physical examination at birth is necessary to avoid this clinical error. Pediatricians should remain alert to the possibility of imperforate anus with perineal fistula in patients with abdominal distension and severe constipation of unknown origin.

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