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Primary Closure and Iliac Osteotomy in the Treatment of Exstrophy of the Bladder

A Case Report

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Exstrophy of the bladder is a rare but serious congenital affliction. Untreated, exstrophy is associated with unacceptable morbidity and mortality. A case report is presented in which primary closure of the bladder and bilateral iliac osteotomy was successfully employed to gain anatomic correction of the bladder defect. However, the value of this surgical technique in restoring normal function of the lower urinary tract system is questionable.

The incidence of exstrophy of the bladder is fortunately low: about one in 40 to 50 thousand births,¹ but the condition is one of the worst congenital afflictions. The untreated patient is urine soaked, foul-smelling, and socially undesirable. He suffers great discomfort from the friction of soaked clothing against the tender, highly sensitive extruding vesical mucosa and adjacent macerated skin edges. Untreated, most patients die or suffer from pyelonephritis before their twentyfirst year. Adenocarcimona of the bladder is a significant late complication.

The condition is manifested by absence of the lower abdominal and anterior vesical walls, with eversion of the

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posterior bladder wall. The uretheral orifices freely discharge urine externally, with complete exstrophy as the usual finding. The posterior wall protrudes in toto. There is epispadias and separation of the pubis. Because of the wide separation, the child commonly waddles or shuffles. This movement, due chiefly to the outward rotation of the femurs, is less noticeable in adults. Malformation of the rectus muscles and fascia are reflected in large inguinal or femoral hernias. Accompanying genital anomalies involve the penis, vulva, clitoris, urethra, ureters and testes. Often, there are nonurologic abnormalities.

Surgical correction can be performed by the time a baby is a year old. At this age the kidneys have not yet been subjected to recurring attacks of infection. The ureters have not dilated,

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so technically it is easier to implant them. The most successful procedure has been ureterosigmoidostomy, cystectomy and revision of the genitalia in staged procedures. The abdominal hiatus is closed with local fascial flaps and undermining of adjacent soft tissue. At Henry Ford Hospital, the procedure of choice has been a ureteral-ileo-sigmoidostomy. However, in recent years, advocates of primary functional closure of the bladder have presented some successful cases. By this technique they attempt to replace the bladder in the pelvis, construct a functional exit passage for urine, and close the abdominal wall defect. Lattimer² states that the advantages are:

- Permanent diversion of the urinary or fecal stream is not required.
- 2. Structures involved are not damaged so that they can be modified at a later date.
- 3. Good cosmetic results can be obtained.
- The procedure is safer than ureterosigmoidostomy as there are no intra-abdominal suture leaks.

It is Lattimer's feeling that one serious attempt at functional closure should be attempted whenever possible.

In 1906, Trendelenberg reported success in the plastic repair of incomplete exstrophy by dividing the sacroiliac ligaments at the posterior synchondrosis, and allowing the pelvis to hinge posteriorly and close anteriorly. In 1958, Shultz⁴ reported the first case in which primary closure was accomplished with the aid of bilateral posterior iliac osteotomies and a second stage plastic repair. The following year, Lloyd-Roberts⁵ reported six cases utilizing pelvic osteotomies and a colleague's experience with manual osteoclasis in newborn patients. By 1960, Lattimer³ was able to report 28 cases of primary closures, in which four had pelvic osteotomies. He found that the osteotomy allowed the soft tissue to be brought together in front of the urethra to a greater degree, and hopefully, permit better urinary control. The osteotomy also obviated or reduced the size of rectus flaps. Operation time was shortened because plastic repair was less extensive. As an added benefit of the osteotomy, gait was improved because the femurs were aligned in a more normal position.

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Procedure

P. G. was first seen at Henry Ford Hospital, in March, 1967 as a sixmonth-old infant with complete exstrophy of the bladder (Fig 1). Aside from genital anomalies, no other disturbances of development were noted. Urinalysis revealed 3+ RBC, 2+ albumin, and a few leukocytes. The BUN

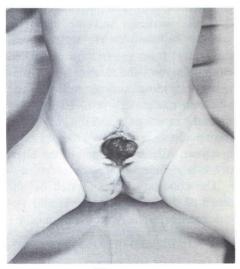


Figure 1 Pre-operative appearance.

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was 13 mg% and hemoglobin was 11.9 gms. An IVP revealed a normal upper urinary tract. When she was admitted for surgical correction in September, 1967, urine culture revealed a growth of proteus, coliform, and enterococci. An IVP again showed no changes of the upper tract.

On September 11, 1967, bilateral iliac osteotomies were performed with the patient intubated and in a prone position (Figs 2, 3). Curved incisions were made along the posterior iliac crest and then extended downward over the posterior iliac spine. The cartilaginous apophysis on the crest was split and by subperiosteal dissection, the gluteal muscles were reflected laterally off the iliac crest and the sacro-spinalis reflected medially. An osteotomy was then made through both inner and outer tables from the crest to the sciatic



Figure 2 Pre-operative x-ray.

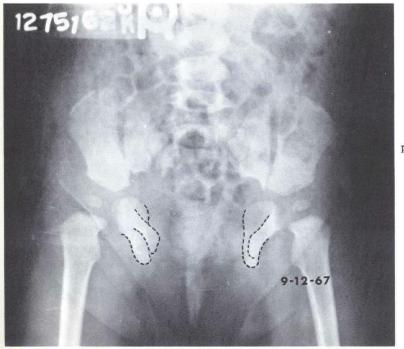


Figure 3 After first procedure. Note iliac osteotomies. notch. The sacrospinous and sacrotuberous ligaments were cut to aid in allowing the pelvis to be brought together anteriorly. A simple light dressing was applied and the patient was returned to bed with no restraints. Postoperative recovery was good, except for temperature elevation, due to urinary tract infection, which responded to appropriate antibiotics.

Nine days after the first procedure, the second stage was performed under general anesthesia. The bladder was dissected free and placed in the pelvis, drained by suprapubic catheter. The urethra was reapproximated over a #10 catheter that was brought out through an abdominal stab wound. The intersymphyseal ligament was isolated and used to form an artificial external sphincter. The anterior pelvis was approximated by pressing on the lateral wings of the ilium and placing a wire

suture in a "figure-8" arrangement across the pubic bones (Fig 4). Impingement of the urethra by the wire suture was avoided by staving anteriorly. The meatus of the urethra was then sutured to the inner margin of the vulva. The defect of the abdominal wall was closed in layers. A double hip spica cast was applied, with the hips in internal rotation and lateral compression across the iliac wing. After being changed under general anesthesia on November 9, the cast was removed on November 29. Endoscopy revealed a narrow but unobstructed urethra. This was then dilated with a temporarily indwelling catheter.

When last seen on December 12, 1968, 15 months after surgery, the child appeared healthy and had a good walking gait. She continued to leak urine daytime and nighttime, but could void up to two ounces at one time.

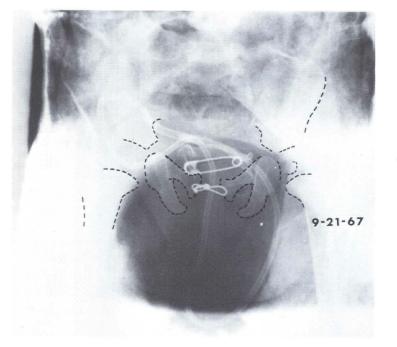


Figure 4 After second procedure.

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The abdominal closure had healed uneventfully. As seen in x-rays (Fig 5) a wire had cut out of the soft bone of the pubis, but by that time the primary closure of the soft tissue had healed. The transpubic distance has remained 3.0 cm as compared with the 4.9 cm preoperatively. An IVP showed a normal appearing bladder without vesical reflux, but with considerable increase in bladder capacity. The child demonstrated interest in toilet training.

Discussion

In their cases of pelvic osteotomies, Ryder and Lattimer,⁶ noted the complication of the wire suture cutting across the lumen of the urethra and advised against the use of metal or nonabsorbable sutures. They recommended applying a spica cast after the osteotomy, and then removing an anterior wedge immediately after operation, and also three days postoperatively, each time allowing the cast to be approximated anteriorly. Seven days postoperatively they reconstruct the bladder through an anterior window. It would seem advantageous to have the pelvis already closed at the time of reconstruction, as this would facilitate approximation of deeper structures, especially the external sphincter. Urinary tract infection imposed a delay between the two stages in our case, necessitating additional fixation at the time of closure of the pelvis. It should be emphasized that the amount of correction of pelvic closure obtained initially is always partially lost whether or not the wires are used. However, (as in our case) the soft tissues have healed by this time, and that is our primary concern. It must be acknowledged that any degree of separation of the pubic bones may vitiate reconstruction of a functional sphincter.

O'Phelan,7 reporting 25 cases in

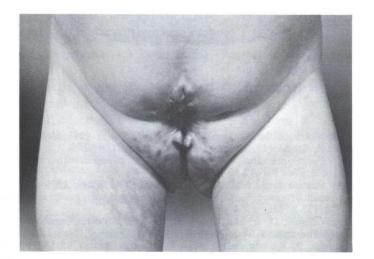


Figure 5 Appearance one year after surgery.

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1963, recommended a two-stage procedure with wiring of the symphysis after osteotomy. To avoid erosion of the urethra by the circumferential wires, he removed the sutures after the child started to walk. Lately, he has modified his wiring techniques by bringing the wires from each side of the superior ramus and tying them anteriorly. In our case, we used a figure-8 wire suture, which was placed anteriorly to avoid impingement of the urethra.

The long term results of this operation have not been defined. The osteotomy appears to have contributed greatly to success in obtaining primary functional closure of the bladder in selected cases. Although this operation has been done as a combined procedure,8 we believe that it would make too formidable an operation unless one has considerable experience. Children on whom this procedure has been done are gaining increasing continence of urine through the years, but it remains to be seen how many will need revision of the urinary tract system at a later date.



Figure 6 Pelvis, four months after surgery.

Summary

A case report of primary closure combined with bilateral iliac osteotomies for repair of exstrophy of the bladder is presented. The indications, technique and complications of this procedure are discussed. Only longterm studies will determine if this reconstructive procedure is of real value.

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