URINARY EXTRAVASATION IN A NEW BORN INFANT ASSOCIATED WITH CONGENITAL STENOSIS OF THE URETHRA

Report of a Case

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Congenital obstructive lesions of the urethra, though uncommon, have been recognized more frequently in recent years due to improved diagnostic methods, notably cystoscopy and intravenous urography. Most of the earlier reports were based upon autopsy findings but today the majority are recognized clinically and the various types have been reported and classified.

The present case is reported not only because it represents an unusual type of congenital urethral obstruction, but also because it was complicated by urinary extravasation.

REPORT OF CASE

A male infant, aged three days, was admitted to the Cleveland Clinic Hospital on May 9, 1938, because he had passed no urine since birth.

The baby was a full term infant, the mother's third pregnancy. Delivery was accomplished by version extraction. The family physician reported that, although it was an unusually large baby, weighing 13 pounds and 13 ounces, no difficulty was encountered in the delivery. Examination of the baby following birth showed it to be normal in every respect. The child emitted a strong cry on being delivered and there were no signs of maldevelopment.

For the first 24 hours, the diapers were apparently somewhat moist and it was assumed that probably some urine was passing, although this is open to question. The child had passed some meconium. However, in the next 48 hours, it became evident that no urine was being passed. Twenty-four hours before admission to the Clinic there appeared for the first time some edema of the scrotum and penis. The family physician, feeling that perhaps a phimosis which was present was causing obstruction, performed a dorsal slit but this did not alter the condition. The edema extended throughout the entire perineum and scrotum and began to appear in the suprapubic region.

Physical examination revealed a rather large baby, well developed but apparently severely ill. He was not crying a great deal and gave no evidence of pain. The skin was very moist and over the entire body there seemed a peculiar redness of the skin with the exception of the hands and feet which had a dusky, almost cyanotic hue. The respirations were rapid but with adequate excursion of the lung field. The

abdomen was not distended. Careful palpation revealed no evidence of retention of urine in the bladder.

The finding of chief interest was the presence of a marked induration which covered the entire perineum and extended up over the symphysis onto the lower part of the abdomen, extending on the left side almost to the costal margin. The scrotum, penis, and prepuce were extremely edematous and swollen. No crepitation was felt but there was marked pitting on pressure. The entire picture was rather characteristic of urinary extravasation and the condition demanded immediate attention without further study.

An attempt was made to pass a urethral catheter but both catheters and bougies met an impassable obstruction in the membranous portion of the urethra. The only alternative was to perform a suprapubic cystotomy which was advised and carried out.

At operation, the subcutaneous tissues were quite edematous and the perivesical space was infiltrated with extravasated urine. It was interesting to note that when the perivesical space was exposed and sponged, the scrotal and penile swelling showed marked regression. The bladder was found to be quite small with a distinctly thickened wall and contained only a small amount of urine. No rupture of the bladder could be demonstrated and as the child's condition did not warrant a prolonged search for the point of extravasation, a small mushroom catheter was fixed in the bladder and the perivesical space thoroughly drained. The wound was loosely closed to permit free drainage.

The condition of the baby immediately following cystotomy was quite satisfactory. On the first postoperative day some improvement was made; the child took nourishment and became somewhat brighter. The temperature during this time remained around 102° F. On the second postoperative day, the temperature came down to 100.4° F. However, on the morning of the third day, the temperature was recorded at 104.4° F. and it was noted that the skin over the entire body had a loose, shriveled appearance. On the back over the right scapular area, there was a separation of a large area of skin about three inches across. This area had the appearance of a large blister. There were numerous large reddish-pink areas on the body and extremities. Overlying these, the skin was easily slipped off and exfoliation of the epidermis soon took place at all points of pressure. The child was seen by a dermatological consultant who made a diagnosis of epidermolysis bullosa. The lips became deeply fissured. The conjunctiva were congested and there was a purulent discharge from both eyes. The lips, fingers, and toes became quite cyanotic. The child's condition grew steadily more grave and death occurred the following day.

Permission for necropsy was granted and in this examination the points of interest were confined almost entirely to the genito-urinary tract (Figs. 1 and 2). Careful examination of all other organs showed no congenital malformations. The kidneys, ureters, bladder, and

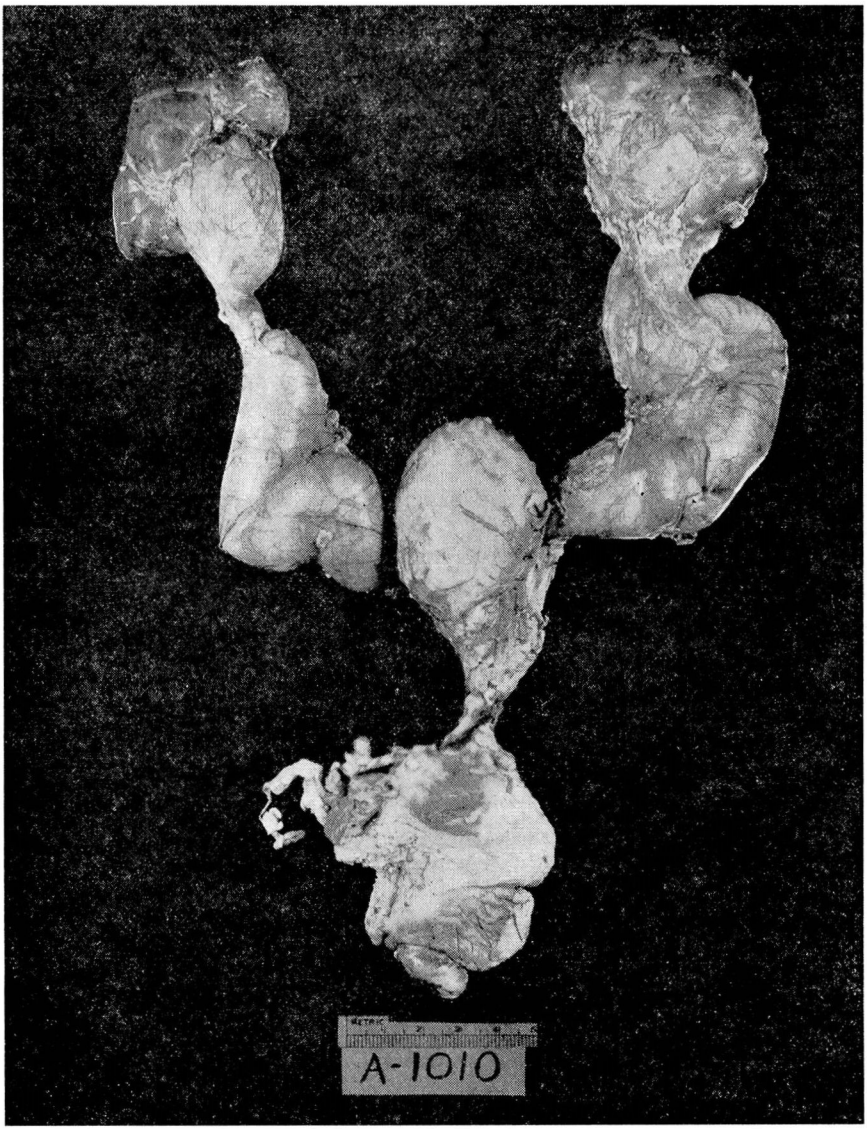

FIGURE 1: Photograph of gross specimen showing marked bilateral dilatation of kidney pelves and ureter.

urethra were examined intact and one was immediately impressed with the marked bilateral hydronephrosis and the dilated, tortuous ureters. The kidneys showed some fetal lobulation; both were about equal in

size and on section were seen to be flattened out into a shell, the kidney tissue varying in thickness from 1 mm. to 5 or 6 mm. There was no indication of remaining pyramids, and only an occasional depression marked the position of a calix. Both ureters were enormous in size, very

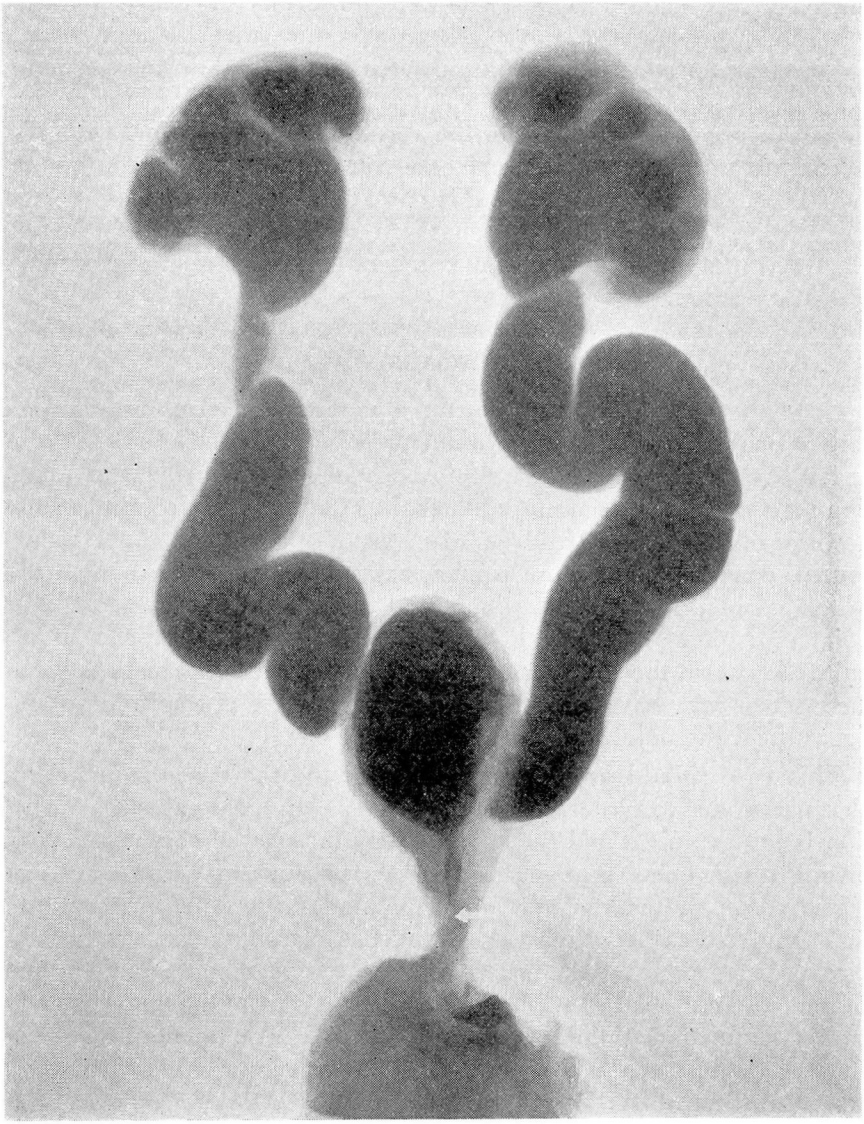

FIGURE 2: Roentgenographic outline of urinary tract after injection with contrast medium —arrow indicates point of obstruction.

tortuous, and irregular throughout their entire course. The ureters passed through the bladder wall in the usual position and manner, but at this point the lumen of each ureter was quite small compared to the

lumen elsewhere. However, a probe could be passed easily into the bladder and no obstructive lesion could be demonstrated in the lower ureter.

The bladder appeared to be about normal size for an infant but on section it was noted that the walls were uniformly thickened and there was rather pronounced trabeculation over the entire bladder mucosa. The prostatic urethra was slightly dilated but careful search showed no evidence of congenital valves or hypertrophy of the verumontanum. However, about 1 cm. distal to the verumontanum there was complete stenosis of the urethra which extended for a distance of about one and one-half centimeters. Distal to this point the urethra was again normal. Just proximal to the point of stenosis there was found a small necrotic opening in the urethra which appeared rather definitely to be the point of extravasation.

Discussion

Congenital urethral stenosis of the type encountered in this case is of rare occurrence. It is an embryological accident and occurs as the result of faulty development of the urethra. The associated urinary extravasation makes the case even more unusual as this is a rare complication of urethral obstruction in children. In speculating as to its cause, it is, of course, conceivable that the version extraction of a large baby with a distended bladder may have increased the intravesical pressure to the point of rupture. Although this may have been a factor, it is also quite likely that the same faulty development which was responsible for the stenosis may have created a point of weakness in the urethra which determined the point of extravasation.

This case should not be confused with the more common urethral obstruction due to posterior urethral valves. In these cases the obstruction is not complete and they are usually discovered some months following birth, being brought to attention because of enuresis, urinary incontinence, palpable distention of the bladder, or when urinary infection complicates the disease. Young has classified these valves into three types according to their relation to the verumontanum, the most common being type I in which the valve passes from the anterior end of the verumontanum to the lateral urethral wall. Treatment of these cases consists in destruction or excision of the valve which can usually be done transurethrally.

The pathology in all cases of congenital urethral obstruction is quite similar. The urinary back pressure first produces dilatation of the bladder with hypertrophy of its musculature, but eventually the bladder becomes decompensated and the back pressure effect is transmitted to the ureters and kidney pelves which become enormously distended. Marked im-

pairment of kidney function results. That this change must go on in utero is shown by our case and others reported in the literature where marked hydronephrosis existed at birth. It is believed that urinary secretion begins in the embryo at about the fifth month. Undoubtedly, serious renal impairment is present in a great many of these cases at birth; few, however, are recognizable at this time and the children live on for several years. The occurrence of disturbance of the urinary tract, most commonly infection, induces complications which require investigation. In this particular case, urinary extravasation called attention to a congenital deformity in a very young infant which necessitated immediate operative intervention.

Several methods of treatment are available. If a normal meatus is present, the passage of a bougie or filiform should first be attempted. The obstruction may be a very thin membrane and give way to gentle sounding. In so doing, however, one must exercise the greatest care in order to avoid undue urethral trauma. The second method is that carried out in this case, suprapubic cystotomy. The high position of the bladder in the infant makes cystotomy a relatively easy procedure, and where the bladder is well distended even a suprapubic puncture may be carried out. Further reconstructive procedures may then be deferred until the immediate emergency has been overcome.