

Bladder outlet obstruction in the neonate

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Seven neonates with severe bladder outlet obstruction causing urine retention and presenting with a large, palpable lower abdominal mass are reported. The obstruction was due in two cases to posterior urethral valves, in one case each to prune belly syndrome, prolapsing ureterocele, urethral diverticle, and in two cases to pelvic neuroblastoma. Diagnosis was based on physical examination and roentgenographic studies. Therapy and prognosis of bladder outlet obstruction in neonates are discussed. Of the seven patients, 3 neonates died, the rest are developing well.

Early and proper evaluation of the neonate with a large bladder is essential [5]. Urine retention may be caused by a number of conditions, of these the most important are summarized in Table I. In this paper we shall only deal with mechanical obstruction.

DIAGNOSIS OF BLADDER OUTLET OBSTRUCTION

The diagnosis of urine retention in a neonate is suggested by the presence of a round midline mass above the symphysis, reaching occasionally to the umbilicus. The mass disappears on catheterization of the bladder. Abdominal distension due to urinary ascites is suggestive of subvesical obstruction [4]; dribbling or intermittent passing of small volumes of urine with a slow or weak stream is characteristic. A careful rectal examination is essential to identify a presacral or pelvic tumour.

X-ray of the abdomen before decompressing the bladder may reveal a shadow pointing towards the urine-filled bladder. Displacement of the intestine without intestinal obstruction is a frequent occurrence. The indwelling catheter used for evacuation of the bladder should serve also for cystography but this intervention should be postponed until any infection has been brought under control. A lateral view gives more detail than the anterior-posterior projection. Intravenous pyelography (IVP) gives some indication of the function of the obstructed urinary tract and may determine the site of the obstruction. It should be delayed by 24 to 48 hours after decompression of the bladder to obtain reliable information on the upper tract, and a delay of several hours after injection of the dye is of particular importance for visualizing every detail. Ultrasound studies will usually give a rapid and precise diagnosis of the level of obstruction. In the diagnosis of pre-

TABLE I
Conditions leading to urine retention in the neonate

<i>Mechanical obstruction</i>	
<i>Intrinsic</i>	<ul style="list-style-type: none"> — posterior urethral valve — anterior urethral diverticulum — ectopic ureterocele — urethral polyp — urethral atresia or stenosis (prune belly syndrome) — stricture — meatal ulcer
<i>Extrinsic</i>	<ul style="list-style-type: none"> — pelvic tumours (neuroblastoma, teratoma, rhabdomyosarcoma) — hydrocolpos
<i>Neuropathic bladder</i>	
	<ul style="list-style-type: none"> — neurogenic bladder (myelomeningocele, sacral agenesis, occult neuropathy)

TABLE II
Diagnosis of bladder outlet obstruction

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- Round midline mass, disappearing on catheterization
 - Dribbling of urine, weak stream
 - Inspection of the vulva, penis and abdominal wall
 - Both kidneys may be palpable
 - Careful rectal examination
 - X-ray
 - plain radiograph
 - i.v. pyelography with delayed films
 - cystography
 - Ultrasound
 - Laboratory studies (azotaemia, acidaemia, urinary infection)
-

sacral and pelvic masses displacing the rectum and bladder, a barium enema may be informative if rectal gas is not seen on the plane film.

The clinical picture is characterized by azotaemia and acidaemia. Vomiting, acidaemic hyperpnoea are common complaints and may simulate cardiorespiratory disease. Superimposed urinary infection is common and may cause diarrhoea, jaundice or haemorrhagic diathesis. The vulva should always be inspected as it may reveal an underlying cause of hydrocolpos.

CASE REPORTS

Seven neonates presenting with palpable bladder caused by intrinsic or extrinsic obstruction are briefly reported to illustrate the importance of prompt diagnosis and management. Their data are summarized in Table III.

Patient 1. The first neonate with a palpable bladder and dribbling was referred to our clinic in a very serious condition (pH 7.03, BE not measurable, plasma potassium 2.1 mmol/l, sodium 112 mmol/l, BUN 27.6 mmol/l). A lateral view cystogram showed the typical picture of a posterior urethral valve. The infant died of advanced urinary sepsis 12 hours after bladder drainage. Necropsy clearly demonstrated a well-developed malformation of both kidneys and ureters.

Patient 2. The second patient with a posterior urethral valve was also admitted

TABLE III

Main clinical data of the neonates with bladder outlet obstruction

Pat. No.	Diagnosis	Weight Sex	Age at admission, days	Volume of retained urine, ml	Blood urea mmol/l		Treatment after bladder drainage	Result
					initial	bladder drainage after		
1	Posterior urethral valves	3050 male	28	17	28.5	—	—	died
2	Posterior urethral valves	2200 male	5	105	19.6	8.9	bilateral uretero-cutaneostomy, later reimplantation and refunctionalisation of both ureters	good
3	Prune belly syndrome	2750 male	3	130	21.5	23	—	died
4	Double renal unit, prolapsing ureterocele	3300 female	6	?	10.7	7.5	excision of ureterocele and cutaneous ureterostomy, hemi-ureteronephrectomy, reimplantation of the remaining ureter	good
5	Anterior urethral diverticulum and urethral stenosis, anorectal agenesis	2860 male	0.5	160	12.2	—	suprapubic cystostomy	died
6	Pelvic (presacral and coccygeal) neuroblastoma	3500 female	27	180	7.5	4.3	removal of tumour	excellent
7	Pelvic (presacral) neuroblastoma	3650 female	21	80	3.9	3.2	removal of tumour	excellent

with a palpable round, firm abdominal mass and dribbling urine. Intravenous pyelography showed very poor concentration of the radio-opaque dye but the diagnosis was confirmed by cystography (Fig. 1a). Bladder catheterization resulted in slight improvement. IVP, a few days later suggested obstruction of both ureters at their entries into the bladder. Bilateral cutaneous end ureterostomies then led to rapid improvement. At the age of 8 months the ureters were reimplanted into the bladder. At present the patient is 3 years old and thriving well (Fig. 1b).

Patient 3. A 3-day-old male infant was admitted with a severe prune belly syndrome (Fig. 2a). All laboratory data were indicative of advanced renal insufficiency with sterile urine. The volume of urine retained in the bladder was 130 ml. Excretory urography with delayed films showed no opacification. A cystogram revealed an enlarged bladder and wide, elongated

and tortuous ureters. High temporary urinary diversion was considered, but the parents refused any surgical intervention, and only a small polyethylene catheter was introduced into the bladder. The neonate died of renal failure at the age of one month. A post mortem showed no renal tissue on the right side and a hypoplastic kidney on the left. Both testes were found in the abdominal cavity. The prostatic urethra was dilated and there was an abrupt change in calibre at the junction with the membranous urethra (Fig. 2b).

Patient 4. In this neonate a large prolapsing ureterocele appeared in the vulva and caused urine retention. IVP demonstrated a left double kidney system with poor function. An emergency cutaneous ureterostomy was performed with excision of the ureterocele. A hemiuretero-nephrectomy was subsequently carried out and the remaining ureter was reimplanted into the bladder.

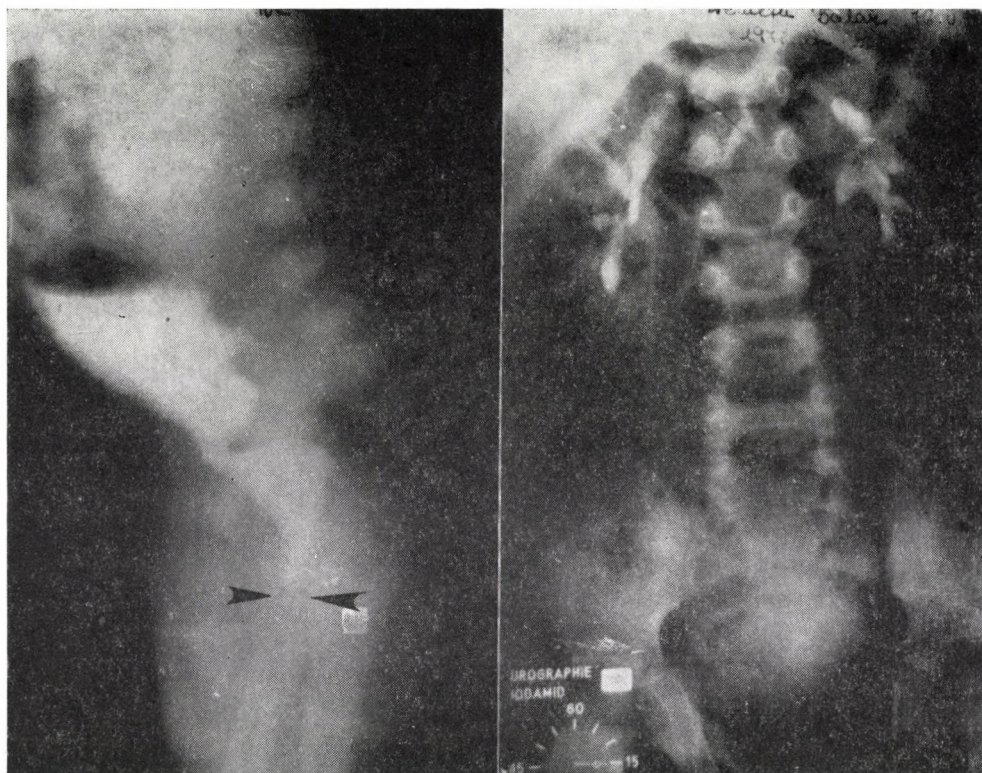


FIG. 1. a) obstructing posterior urethral valve (arrows); b) excretory urogram at 3 years of age

Patient 5. A rare urethral lesion responsible for obstructive uropathy was found in this newborn. A long defect in the spongy corpus of the urethra resulted in a wide diverticulum which was causing almost complete obstruction distally (Fig. 3a). At 16 hours of age suprapubic cystostomy was performed but the infant died on the second day of life. Necropsy showed very advanced urological abnormalities (Fig. 3b).

Patients 6 and 7. These two neonates presented with urine retention due to pelvic and presacral tumours. The masses had dislocated the rectum and obstructed the bladder outlet. Radical removal of the well-capsulated masses was done in both babies. Histologically the tumours proved to be fairly well differentiated neuroblastomas. In one of these neonates extirpation of the mass required an abdomino-retrorectal approach (Fig. 4). Because of their early age and favourable histology neither was

given chemotherapy or radiotherapy. They are now 44 and 40 months old and develop well.

THERAPY AND PROGNOSIS

Bladder outflow obstruction in a neonate should be regarded as an emergency. The programme of medical management, prompt drainage and definitive surgery must be integrated carefully in order to correct dehydration, disturbances of electrolyte balance and acidosis. It is widely accepted that the success of any surgery is greatly promoted if first the

biochemical status is corrected. Close operation between neonatologist, radiologist and paediatric urologist (surgeon) is therefore mandatory.

During the period of resuscitation the bladder should be drained by a small polyethylene catheter passed via the urethra. If the infant's condition does not improve or actually deteriorates on catheterization, temporary upper tract urinary diversion by nephrostomy must be considered

as it provides the best emergency drainage of the upper tract.

Cutaneous ureterostomy may sometimes be necessary. Definitive surgery such as endoscopic ablation of the valve, excision of an ureterocele or urethroplasty can then be performed as an elective procedure.

Duckett [2] does not feel that the dilated urinary tract seen with prune belly syndrome should require the same aggressive therapy as obstruct-

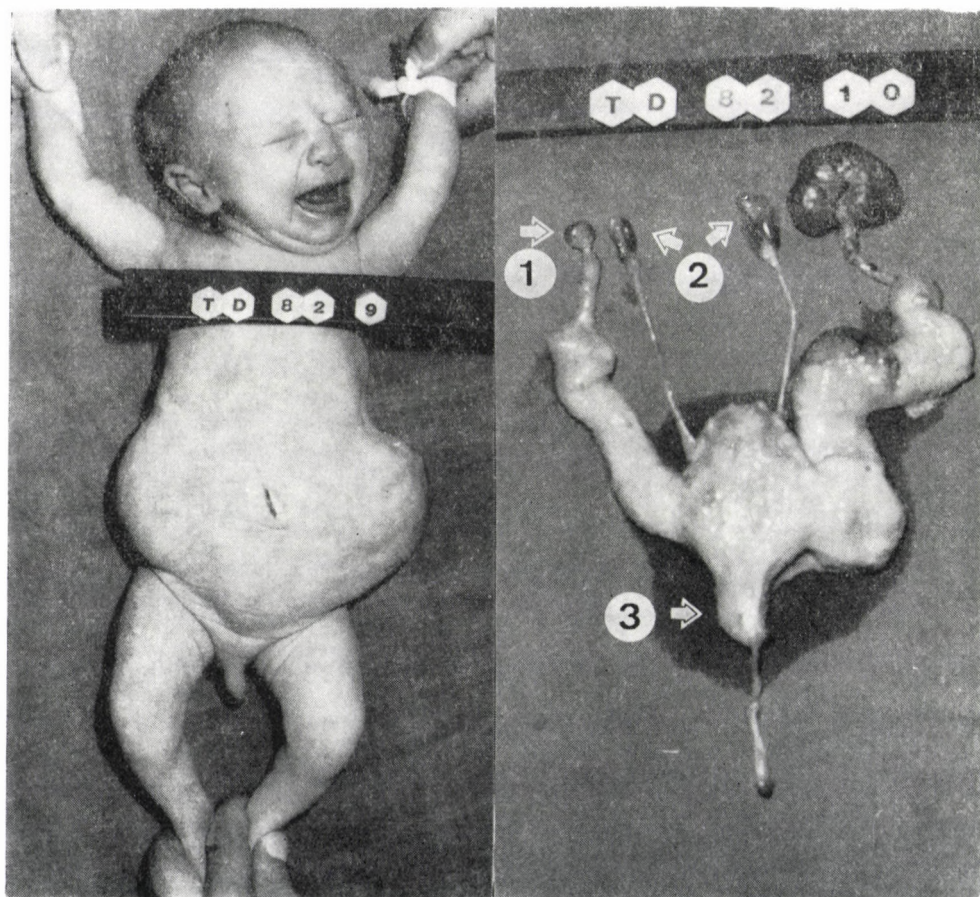


FIG. 2. a) severe abdominal muscular deficiency; b) post mortem: severe hypoplastic right kidney, undescended testes, dilated prostatic urethra

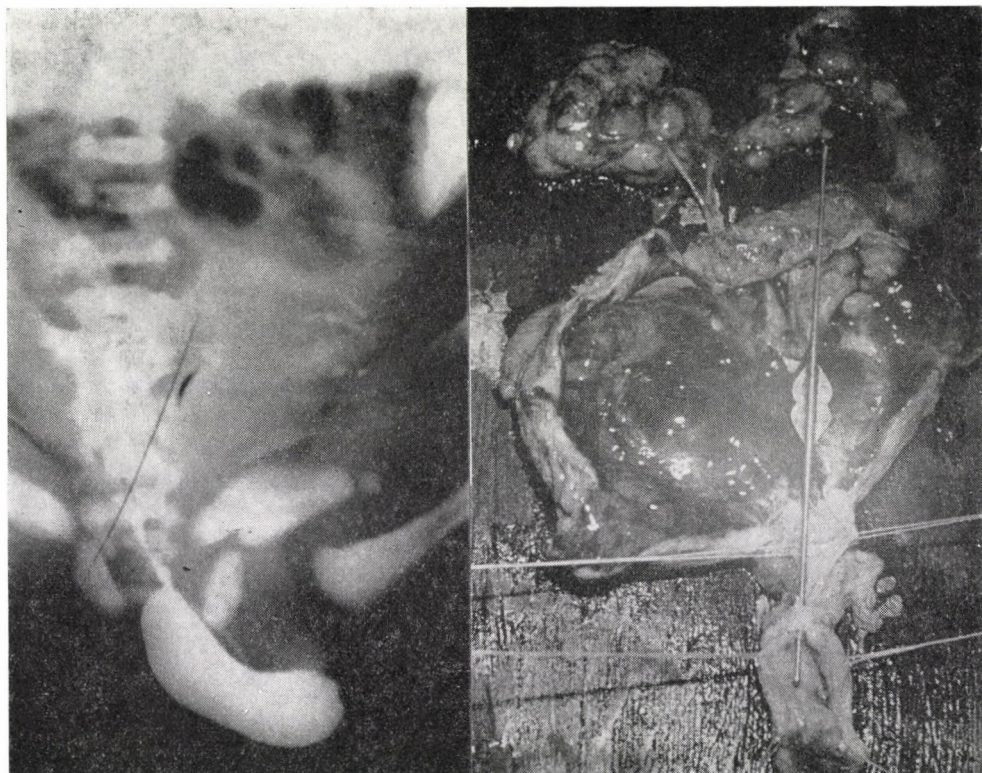


FIG. 3. a) wide anterior diverticulum; b) autopsy specimen: anterior diverticulum and obstruction (arrows)

ed uropathy a baby. Still, there has been a continued enthusiasm for early surgical intervention [7].

The lower urinary tract and pelvic tumours usually require radical but non-mutilating surgery. Recently, it has frequently been reported that tumours manifest at birth or shortly thereafter display a surprisingly benign behaviour, sometimes despite extremely malignant cellular features. Bolande [1] postulated an "oncogenic period of grace" which begins in utero and extends through the first months of postnatal life. During this period neoplasms tend toward benignity by

arrested growth, regression or cyto-differentiation. Owing to this behaviour the hazards of adjuvant chemotherapy and radiotherapy usually outweigh their possible benefits.

The prognosis of urinary retention in the neonate varies greatly, depending on the severity of the underlying condition. A number of such neonates die of kidney failure and associated infection. In the series of Williams et al [6] 22 of 54 boys with urethral valves presenting under the age of 3 months died. Apart from urethrocele, subvesical outflow obstruction can seriously impair the function of both

kidneys. In recent years improved medical management has made it possible to resuscitate or prolong the survival of many neonates with bladder outlet obstruction who earlier would have succumbed.

A kidney that remains functionless despite the relief of obstruction is best removed, but whenever possible this should be confirmed by radio-nuclide studies rather than by IVP. Following removal of the obstruction, the ureters often recover dramatically both anatomically and functionally. The bladder itself has also some relevance to prognosis, as elimination in time of a subvesical obstruction will lessen sacculatation, trabeculation and

thickening of the bladder wall, and often results in disappearance of the vesico-ureteric reflux. Urinary incontinence following treatment is, however, frequently observed although it usually resolves within a few years or with the onset of puberty when the tone in the vesical neck improves with growth of the prostate [3].

In cases of extrinsic obstruction (pelvic teratoma, neuroblastoma, rhabdomyosarcoma) following removal of the cause of urinary retention, improvement of the ureters and kidneys is more rapid and complete than in patients with an intrinsic obstruction. Even good medical and surgical care may leave a number of such

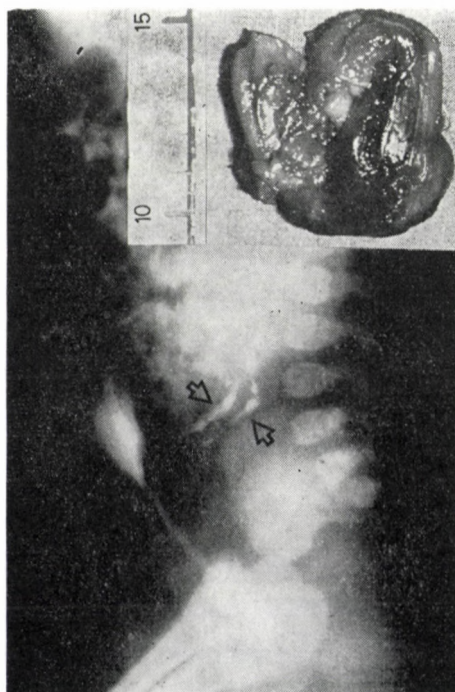


FIG. 4. Pelvic neuroblastoma: bladder is forwarded and displaced upwards, ureters show abnormal position (arrows) upper right: removed tumour

children with severely impaired renal function. They will almost certainly need permanent urinary diversion or a renal substitution programme in later childhood or adolescence.

The overall outlook of our patients was variable. Two of the seven had been admitted in extremely serious condition and died within 48 hours. A third patient succumbed at the age of 1 month; although his bladder was promptly decompressed immediately after admission and a rapid resuscitation was instituted, the severity of the urological and associated malformations was incompatible with life. In the remaining four patients, bladder drainage combined with effective medical treatment brought about considerable improvement of the condition and made later correc-

tive surgery possible. All four of these neonates are thriving well.

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