

Case 112 A gross congenital abnormality



Figure 112.1

Figure 112.1 shows the appearance of the abdomen of a newly delivered baby girl. As well as the gross abnormality seen in the suprapubic area, it was also noted that urine was trickling continuously from below the protruding mass immediately distal of the clamped stump of the umbilical cord. Full examination of the baby revealed no other anomalies.

What is the name of this condition?

Ectopia vesicae – which means literally that the bladder is abnormally placed.

What is the nature of this condition?

The bladder fails to develop normally and the ureters, together with the trigone of the bladder (the triangle between the ureteric orifices and the bladder outlet), open directly onto the anterior abdominal wall. In the male there is associated epispadias of the penis.

This child has a commonly associated abnormality of the pelvic girdle; the visible nodule on each side of the exposed bladder mucosa is the corresponding pubic ramus.

What does this comprise?

Frequently, as in this child, there is a failure of the pubic bones to meet in the midline at the symphysis. This results in a widened pelvis and the child eventually walks with a waddling gait.

Left untreated, what is the natural history of this condition?

The child may die of pyelonephritis due to ascending urinary tract infection. Carcinoma of the bladder rudiment may take place after initial metaplastic change. Note that this tumour is therefore a stratified squamous carcinoma, not a transitional cell tumour, which is the usual carcinoma of the uroepithelium.

What surgical treatment is usually carried out in this condition?

Attempts to reconstruct the bladder have been made, but are usually unsuccessful. Standard treatment usually comprises reimplantation of the ureters into an ileal conduit, which drains into a plastic bag attached around the ileostomy spout stoma. The bladder remnant is excised as a prophylaxis against malignant change. The abdominal wall is closed over the defect; closure may require a pelvic osteotomy to facilitate apposition of the edges.