



**FIG 26-5** Exstrophy of bladder. (Courtesy of H. Gil Rushton, MD, Children's National Medical Center, Washington, DC.)

## Pathophysiology

Exstrophy results from failure of the abdominal wall and underlying structures, including the ventral wall of the bladder, to fuse in utero. As a result, the lower urinary tract is exposed, and the everted bladder appears bright red through the abdominal opening. This is accompanied by a constant seepage of urine from the exposed ureteral orifices, making the area malodorous and susceptible to infection. The constant accumulation of urine on the surrounding skin produces tissue ulceration and further infection. Progressive renal damage from infection and obstruction may cause renal failure if left untreated.

In males with bladder exstrophy, the defect of the genitalia includes epispadias and upward curvature of a shortened penis and may include other problems, such as undescended testes and inguinal hernias. In females, there is epispadias, a bifid clitoris, and small labia minora. The vagina is shortened compared with normal and vaginal dilation may be needed to allow for sexual intercourse. In cloacal exstrophy patients, there are often more severe anomalies, such as bifid or duplicated uterus, split clitoris, completely separated labia, and a duplicate or absent vagina in females. Males may have a split penis and scrotum or a short, flat penis with hypospadias. In either sex, separation of the pubic bones is generally corrected by pelvic osteotomy, particularly if there is