Story behind the wrinkle

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A 16-year-old boy born of a consanguineous marriage presented for evaluation of recurrent urinary tract infections (UTI) from the age of 2 years. His urine stream was slow and he had sense of incomplete bladder emptying, increased urinary frequency, and nocturia three to four times per night. Examination revealed a marfanoid body habitus, and absent testes with minimal secondary sexual characteristics. The lax and wrinkled abdomen wall depicted the absence of abdominal muscles [Figure 1]. Ultrasonography confirmed bilateral cryptorchidism and bilateral hydronephrosis. His serum creatinine was 2.3 mg/dl. A plain X-ray abdomen at the age of 2 years [Figure 2] revealed absent abdominal musculature and visceroptosis. Micturating cystourethrogram at the age of 8 years showed bilateral grade IV vesicoureteric reflex, with kidney and ureter shifted to extreme right due to the lax abdominal wall [Figure 3] and no other genitourinary abnormalities. A diagnosis of prune belly syndrome was confirmed.

He presented to us at the age of 16 years. Urodynamic study showed acontractile large capacity (1,200 ml) bladder with normal compliance and safe end-fill pressure during storage phase and abdominal strain pattern during voiding phase together with a large residual urine volume. He was started on clean intermittent self-catheterization (CISC) with low-dose ciprofloxacin (100 mg) for 6 weeks and standard care for chronic kidney disease (CKD). He subsequently underwent laparoscopic bilateral orchidopexy. At a follow-up of 7 years, he is doing well (stable serum creatinine at 2.2 mg/dl) with excellent somatic growth. There were no further hospitalizations for urosepsis despite no prophylaxis.

Prune belly syndrome (also called Eagle–Barrett syndrome/Triad syndrome) is characterized by the triad of deficient abdominal musculature, urinary tract anomalies, and cryptorchidism.[1] It has an incidence of about 1 in 50,000 live births with 95% being male.[2] As depicted in our case, the skin of the anterior abdominal wall is wrinkled, thin, and lax, giving it a prune-like

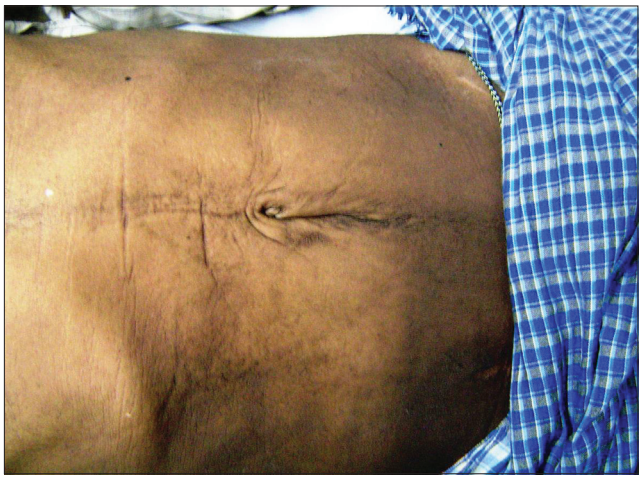

Figure 1: Lax, wrinkled abdominal wall of a patient with prune belly syndrome

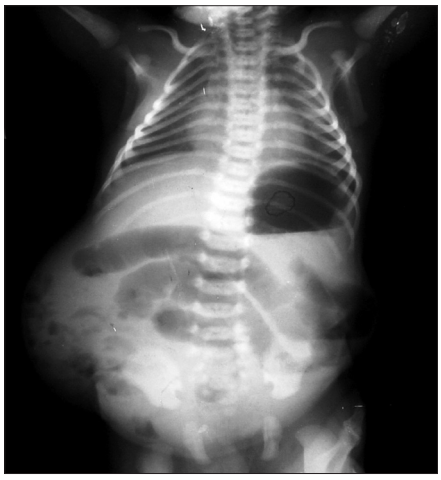

Figure 2: A plain X-ray abdomen done the age of 2 years depicting absent abdominal musculature and the resultant visceroptosis

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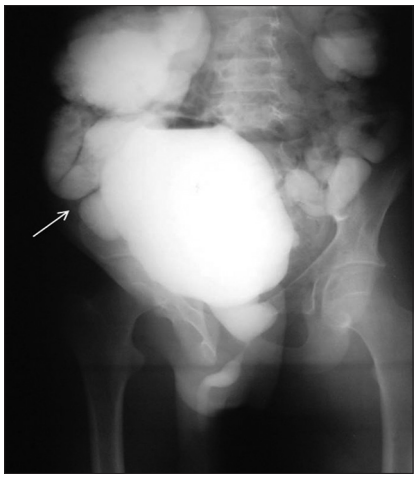

Figure 3: Micturating cystourethrogram done at the age of 8 years showing bilateral vesicouretric reflex (grade IV) with kidney and ureter (dilated) shifted to extreme right due to the lax abdominal wall

appearance. Apart from absent abdominal musculature (complete in 30%, partial in 70%)[3] and cryptorchidism (in about 95%), renal hypoplasia, dysplasia, ureteral dilatation, megacystis, dilated prostatic urethra, and prostatic hypoplasia are commonly seen.

CISC plays a salutary role in dealing with such congenital anomalies in preventing recurrences of UTI and the progression of CKD, as exemplified in our case.

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