Prune Belly Syndrome

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A 2-day-old full-term male baby, born out of nonconsanguineous marriage, was admitted with presentation of respiratory distress, oliguria, and flabby abdominal skin since birth. On cutaneous examination, neonate had thin wrinkled skin over abdomen with the lax abdominal wall ballooning out into the flanks [Figure 1]. Urinary bladder was enlarged appearing as a bulge below the umbilical area [Figure 2]. The entire abdominal muscle wall was absent allowing easy palpation of underlying organs. Examination of the testis showed cryptorchidism. Investigations revealed dextrocardia and scoliosis on chest X-ray, [Figure 3] acute renal failure (ARF) (creatinine: 1.96) on routine blood investigations, bilateral echogenic kidneys with multiple cortical cysts, and bilateral hydroureteronephrosis with dilated ureters on abdominal ultrasound. A clinical diagnosis of prune belly syndrome (PBS) with dextrocardia, respiratory distress syndrome (RDS), and oliguric ARF was made. The RDS was managed with moist oxygen inhalation, broad-spectrum antibiotics, and intravenous fluids. In view of inability to catheterize the patient (urethral malformation), enlarged bladder, and investigations suggestive of hydroureteronephrosis along with oliguric ARF, the patient underwent a vesicostomy under the department of pediatric surgery. The neonate was discharged after improvement in RDS and ARF, with regular follow-up visits advised.

PBS, also known as Eagle–Berrett syndrome or triad syndrome, is an extremely rare congenital disorder (3.6–3.8 per 100,000 live male births) characterized by clinical triad of abdominal muscle deficiency, severe urinary tract abnormalities, and bilateral

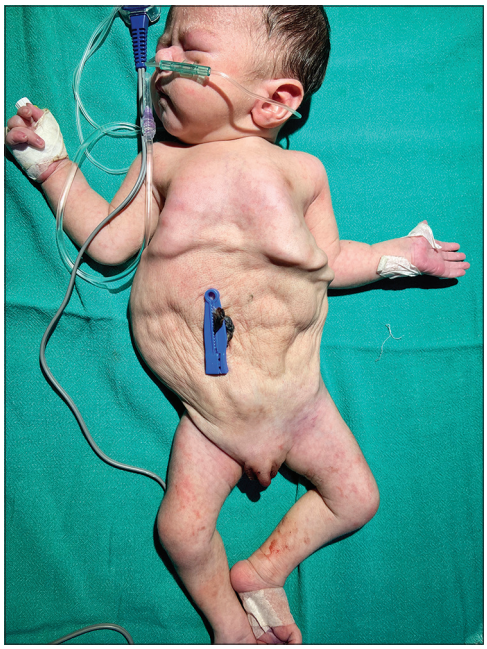

Figure 1: Neonate with thin wrinkled skin over abdomen with lax abdominal wall ballooning out into the flanks

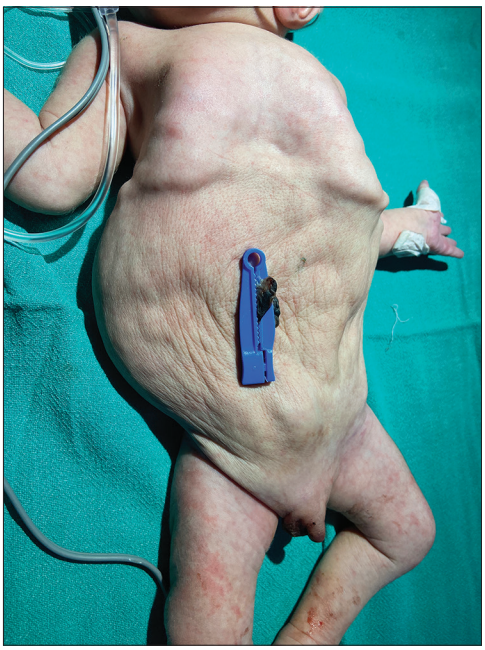

Figure 2: Close-up view illustrating the lax abdominal skin and urinary bladder bulge below the umbilicus

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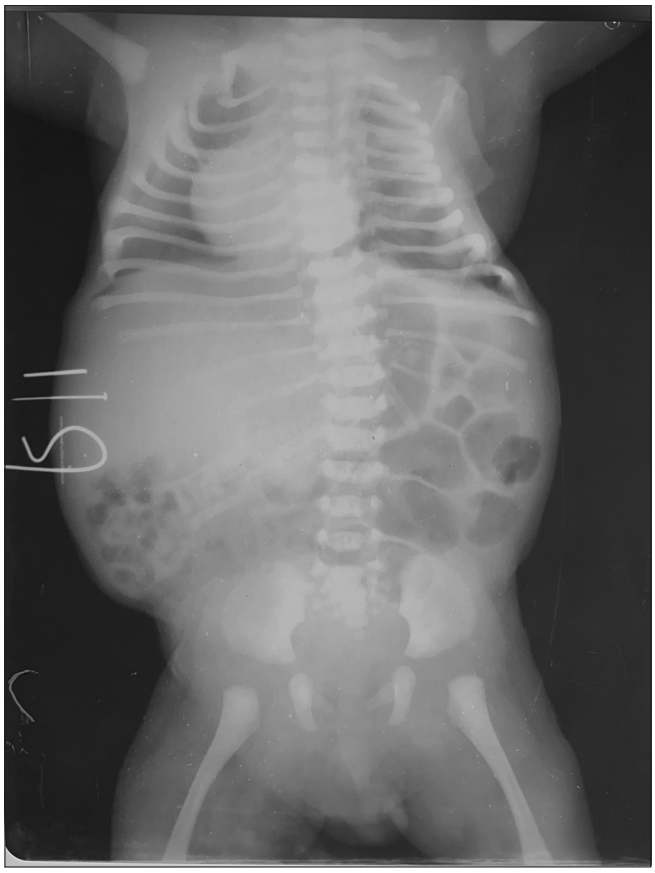

Figure 3: Dextrocardia and scoliosis on chest x ray

cryptorchidism.[1] The protruding hypoplastic abdominal wall looks like a "dried prune," hence the name. The incidence has gone down with advent of prenatal ultrasound allowing early detection and subsequent termination of such pregnancies. Long-term prognosis is poor with renal failure being most common cause of mortality.[2]

Most likely, a dermatologist will come across such a rare presentation from a referral consultation, so he/she must be able to recognize the classical clinical findings and be aware of appropriate investigations and potential complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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