Case Report: Prune Belly Syndrome- With An Unusual Presentation

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Prune belly syndrome (PBS) is also known as Eagle – Barret syndrome.

It is classically characterized by

- Deficient development of abdominal muscles that causes the skin of the abdomen to wrinkle like a prune.
- Bilateral cryptorchidism.
- Abnormalities of the urinary tract such as bilateral gross hydronephrosis, megaureter and megacystis. (1)

Our case had an unusual presentation with a hugely distended abdomen with smooth, shiny skin due to the presence of massive urinary ascites.

Case:

A 30 weeks gestational age male preterm infant was born by Cesarean section to a 33 year old Para 3 mother. Antenatal ultrasound scan showed megacystis, and dysplastic kidneys the bladder was filled with hyperechoic shadows with suspected vesicoenteric fistula. Initially, there was anhydramnios but later normal amount of amniotic fluid was noted. Abdomen showed evidence of ascites on the antenatal scan. A large mass compressed the chest with evidence of pulmonary hypoplasia. (fig 1 and 2)

After birth, the baby required ventilator support due to severe respiratory distress and lung hypoplasia. He was extubated at 62 days of life after several attempts of failed extubation.

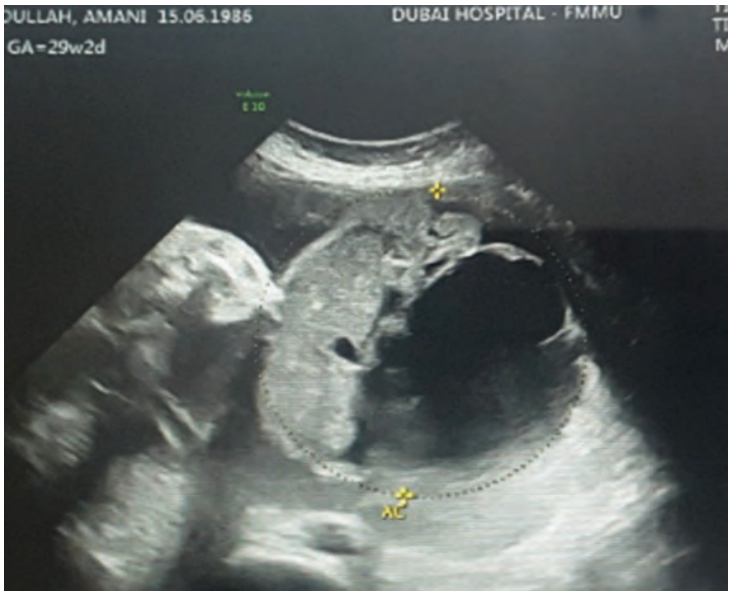


Fig 1 – Figure showing evidence of ascites on antenatal scan

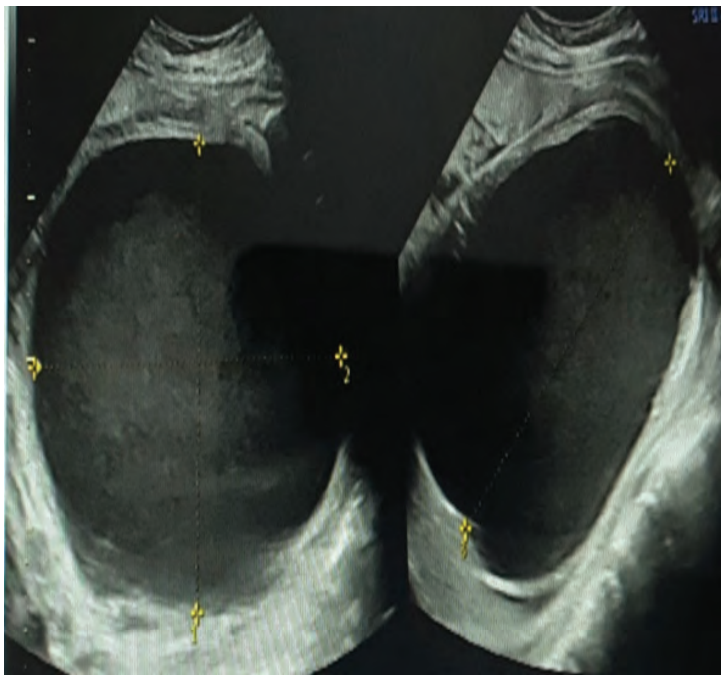


Fig 2- Figure showing antenatal megacystis

His systemic examination at birth showed a narrow thorax, severely distended abdomen with shiny skin (Figure 3 and Figure 4), palpable bladder, male genitalia with bilaterally undescended testes, and imperforate anus. His cardiovascular examination was normal.

> ***"Our case had an unusual presentation with a hugely distended abdomen with smooth, shiny skin due to the presence of massive urinary ascites. "***

Postnatal ultrasound showed hydronephrosis of both kidneys with bilateral megaureter. Paracentesis was performed postnatally, and 450 ml of urine was drained. The pediatric surgeon deflated the ureters by inserting bilateral ureteral drains and draining 750 ml of urine. A urinary catheter was also inserted.

The abdominal distension decreased, and skin became wrinkled, showing evidence of absent anterior abdominal wall muscles (Figure 5 and Figure 6).

Later, laparotomy was performed revealing a large thick walled urinary bladder occupying the whole of the lower abdomen with vesicoenteric fistula. The Vesicoenteric fistula was resected, and vesicostomy was

completed with fixation of the bladder catheter. Colostomy bypassed the imperforate anus.

The baby is passing good amount of urine through the vesicostomy and his renal Ultrasound scan done post surgery shows small right kidney and left kidney with mild dilatation of the renal pelvis. Both ureters are not visualized now.

The vesicostomy tube was removed on day 118 of life and kept as free drainage into the pamper, with clean intermittent catheterization

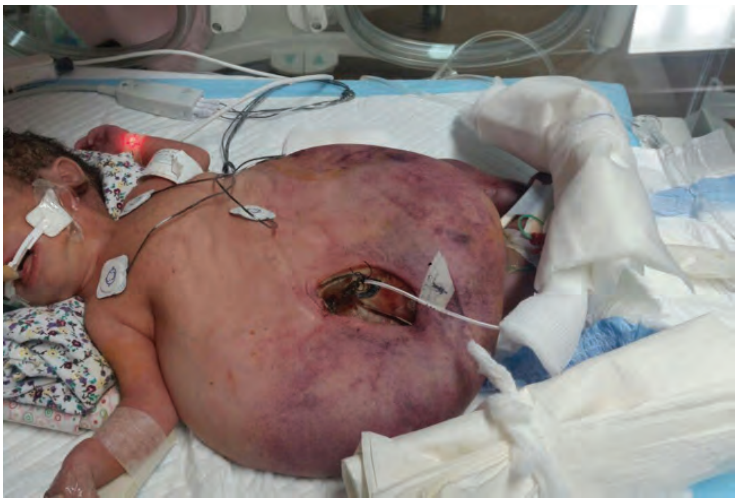
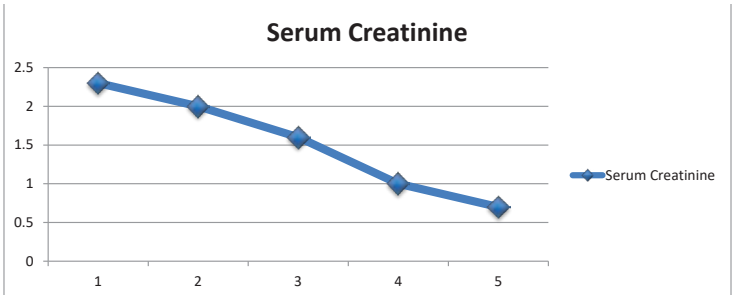


Fig 3 –Severely distended abdomen on admission

through the vesicostomy every 8 hours to empty the residual urine which is continued by the caregiver at home.

His cardiac evaluation, including echocardiography, was normal. He has a normal 46XY karyotype.



Graph 1 - Decreasing serum creatinine

His initial creatinine level was high (maximum of 2.3 then gradually decreased to a normal level, graph 1). He is on prophylactic antibiotics.

After discharge baby had a multidisciplinary follow-up and further investigation in the outpatient clinic:

- DMSA Scan – showed normal left kidney with good and uniform uptake while the right kidney shows no evidence of functioning renal tissue.
- MCUG scan done shows grossly enlarged urinary bladder, dilated ureters with evidence of grade 3 vesicoureteric reflux (VUR) on the right side.
- He had recurrent Urinary tract infection requiring admission and IV antibiotics. His prophylactic antibiotics were continued.

Discussion:

Fig 4- X ray showing severely distended abdomen with narrow thorax.

Our case had an unusual presentation with a hugely distended abdomen with shiny skin due to the presence of massive urinary ascites. Few cases with similar presentation have been reported in the past. (5) He had megaureters and megacystis. He required paracentesis to relieve the abdominal distension on admission. The megaureters were deflated by ureteral drains and vesicostomy performed to drain the urine. Once the abdominal distension was relieved, wrinkling of the skin, a feature of Prune Belly syndrome was evident. Ultrasound abdomen also showed resolution of the megaureters and megacystis. This indicates the probable presence of an obstruction to the urine outflow antenatally resulting in megaureters and a huge bladder leading to a leak of urine into the peritoneal cavity, causing the massive ascites. The huge distension of the bladder has most probably led to vesicoenteric fistula formation.

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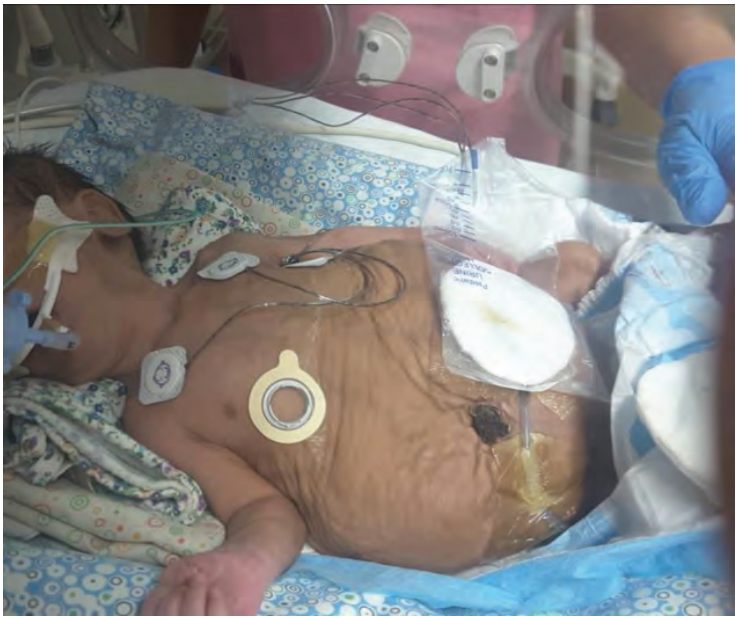

Fig 5 – Baby with vesicostomy drain and showing decreased abdominal distension showing the wrinkles

In unstable patients who cannot tolerate an operation, Cystostomy and pyelostomy are undertaken to shunt the urine temporarily. There is evidence that prenatal vesicoamniotic shunting results in a good outcome with preservation of renal and pulmonary function in a patient with severe lower urinary tract obstruction. (2)

55% of patients with PBS show significant clinical pulmonary manifestations. Hypoplasia of the lungs and cystic adenomatoid malformation are some of the common respiratory manifestations of PBS. (2). Pulmonary hypoplasia is the main cause of mortality in the neonatal period. (2)

Our baby had respiratory distress and lung hypoplasia, requiring prolonged ventilation. In addition, he had an imperforate anus, which was addressed surgically, and a colostomy was performed. 30% of patients with PBS have evidence of gastrointestinal manifestation which includes midgut malrotation and hindgut abnormality leading to anorectal malformation (e.g., the persistence of embryonic cloaca, agenesis of rectum and anus). (3)

Also, the baby had bilateral talipes equinovarus deformity, which is being managed by physiotherapy. 45% of babies with PBS can have skeletal abnormality Skeletal abnormalities. (e.g., clubfeet, limb deficiencies, teratologic hip dysplasia) (4) and 10% of patients can have cardiac anomalies including atrial septal defect, ventricular septal defect, patent ductus arteriosus, and Tetralogy of Fallot. (2)

Long term outcome in a patient with PBS is determined by the severity of urinary tract abnormality and renal function. (2)

Our patient is being managed with a multidisciplinary team approach including neonatologist, pediatric surgeon, pediatric nephrologist, orthopedics and physiotherapist. In the outpatient follow up baby was having recurrent UTI, which was treated by IV antibiotics by the pediatric nephrologist. Baby is on prophylactic antibiotics as MCUG shows grade 3 VUR. DMSA scan showed normal functioning left kidney while the right kidney is non-functioning.

Parents have been taught intermittent catheterization through the vesicostomy. The plan is to refer the patient to plastic surgery for reconstruction of the abdominal wall.

Fig 6 – X ray abdomen showing vesicostomy tube and less distended abdomen.

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Disclosures:
No conflicts of interest or financial disclosures relevant to this article are reported.

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