Fellow's Column: Conservative Management of Urinary Stasis in Prune Belly Syndrome

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Case Report:

We are writing in follow up to the interesting case of Prune Belly Syndrome (Eagle-Barret syndrome) presented by Khan et al. 1 published in the June 2019 issue of Neonatology Today. A baby boy was born at 40weeks, 1day gestation to a 35-year-old G3P2012 mother by C-Section. All maternal prenatal laboratory including RPR, HIV, hepatitis B, chlamydia, and gonorrhea were negative. Blood group was B positive. Apgar scores were 8 and 9. Birth weight was 4040 grams, length 51 cm, head circumference 36 cm.

Physical examination showed normocephalic head, flat anterior fontanelle and preauricular pit on the left side. Pupils were equal, round, and reactive to light. Cardiovascular examination was normal. The chest appeared small, but no distress was noted. The abdomen was soft and distended with wrinkled skin (Figure 1-A). Testes were undescended bilaterally. Bilateral clubfeet were noted. The skin was warm and dry.

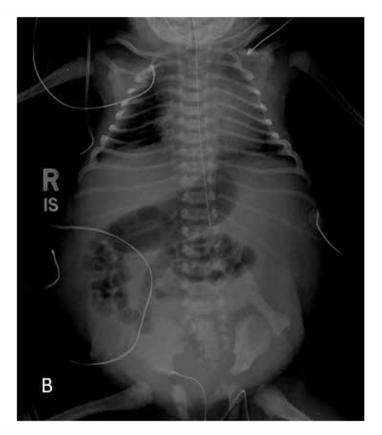
Chest x-ray showed a bell-shaped chest (Figure 1-B). Renal ultrasound showed bilateral hydronephrosis (Figure 2). Pediat-

ric urology and nephrology services were consulted. A voiding cystourethrogram (Figure 3) and Tc-99m MAG3 (mercaptoacetyltriglycine) scan (Figure 4) were ordered. The VCUG showed no reflux and MAG3 showed normal renal perfusion with normal uptake but delayed excretion of tracer by both kidneys. Figure 5 depicts that both ureters and bladder are not seen indicating urinary stasis or obstruction.

"Prenatal history was significant for poor prenatal care and perinatal ultrasound consistent with protruding abdominal wall mass suspecting omphalocele."

Kidney functions were monitored. Serum creatinine (Figure 6) and urine output (Figure 7) remained normal. In view of the preserved renal function, normal serum creatinine, and urine output, the urology team decided against doing a percutaneous nephrostomy tube or vesicostomy. They felt trace retention was most likely due to urinary stasis rather than true obstruction. The infant remained stable and tolerated full enteral feeds of Similac PM 60/40 formula. He was started on amoxicillin 50mg daily for UTI prophylaxis and circumcised at the bedside by urology. He was discharged home with follow up with pediatric urology, pediatric nephrology, and NICU high-risk clinic.





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References:

1. Khan A, Hafeez M, Khan W. Case Report: Prune Belly Syndrome- With An Unusual Presentation. Neonatology Today; June 2019:20-23

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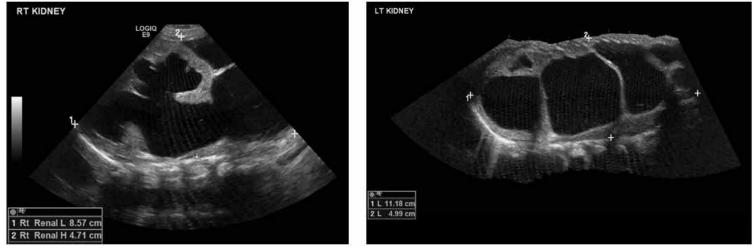
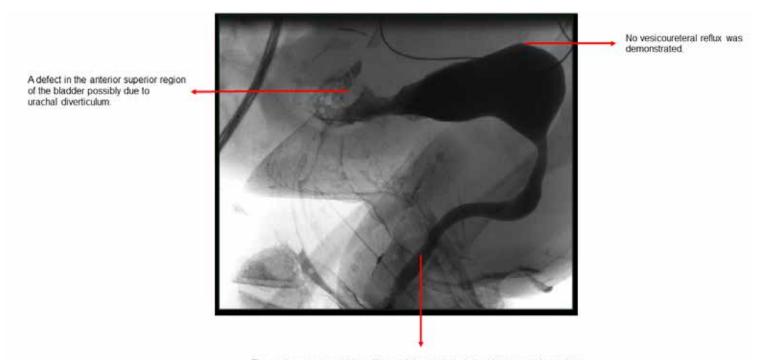


Figure 2: Renal ultrasound showing hydronephrosis



The urethra was normal in caliber and demonstrated no strictures or obstructions

Figure 3: Voiding cystourethrogram (VCUG)- no vesicoureteral reflux, normal urethra

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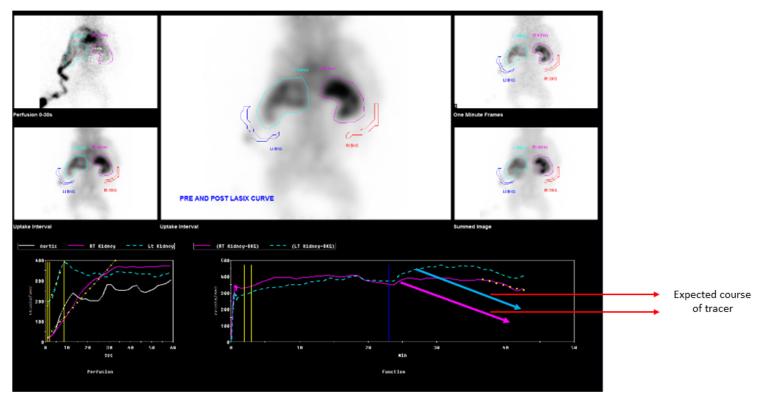


Figure 4: Renal scan: Tc-99m MAG3 (mercaptoacetyltriglycine). Posterior abdominal radionuclide angiogram.

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Figure 5: Pre and post Furosemide scan

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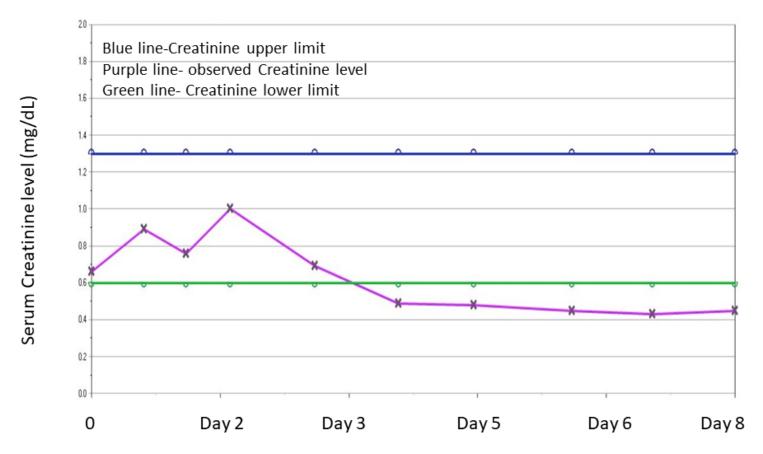


Figure 6: Serial serum creatinine level



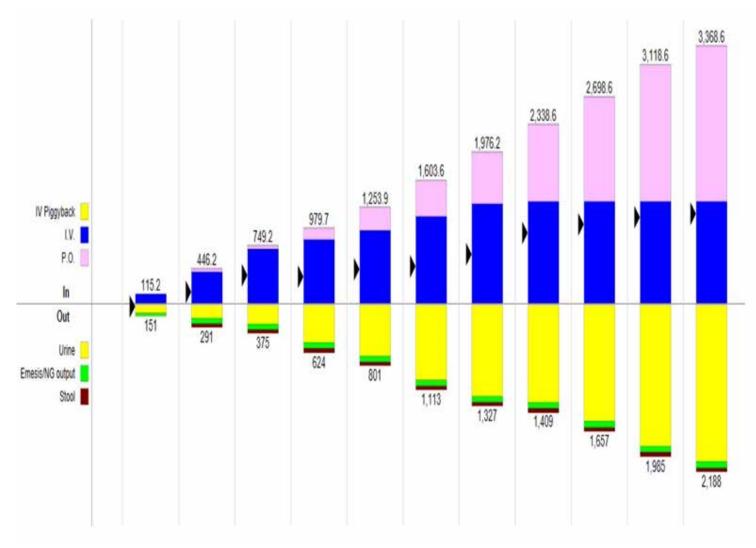


Figure 7: Graphic representation of cumulative intake and output (days)



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