



**Society of Radiologists in Ultrasound
2012 Toshiba Residents Program**

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Clinical History: 29 yo G1P0 referred from an outside clinic for fetal bladder distention.

Figures:

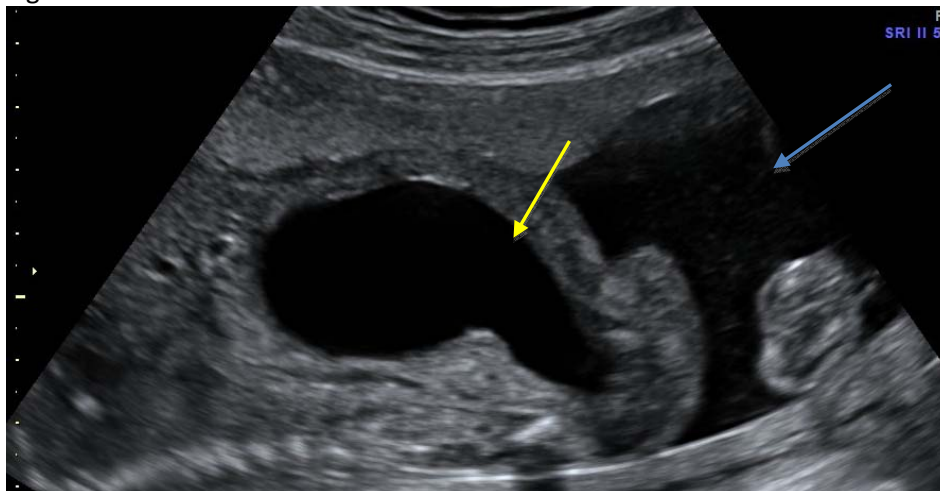


Figure 1. Longitudinal image of the 21 wk 1 day fetus demonstrates a markedly distended fetal urinary bladder (yellow arrow). Normal amniotic fluid is also seen (blue arrow).

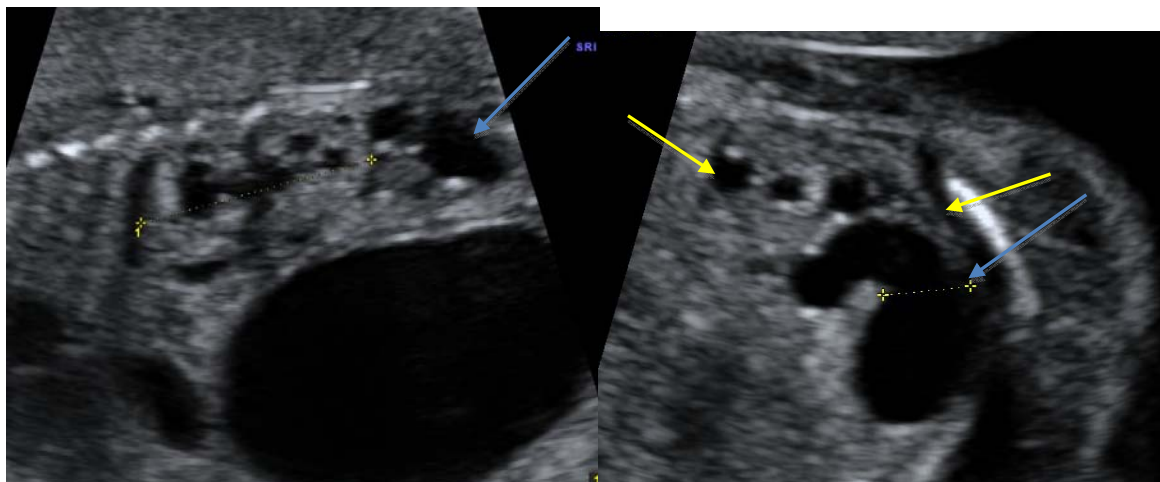


Figure 2A

2B

Both kidneys demonstrate hydronephrosis and increased parenchyma echogenicity. 2A calipers show margins of echogenic right kidney with dilated collecting system. Anechoic perinephric fluid on the right is likely related to a urine leak. 2B yellow arrow points to the left kidney. Bilateral distended ureters are also seen (blue arrows, 2A and 2B).

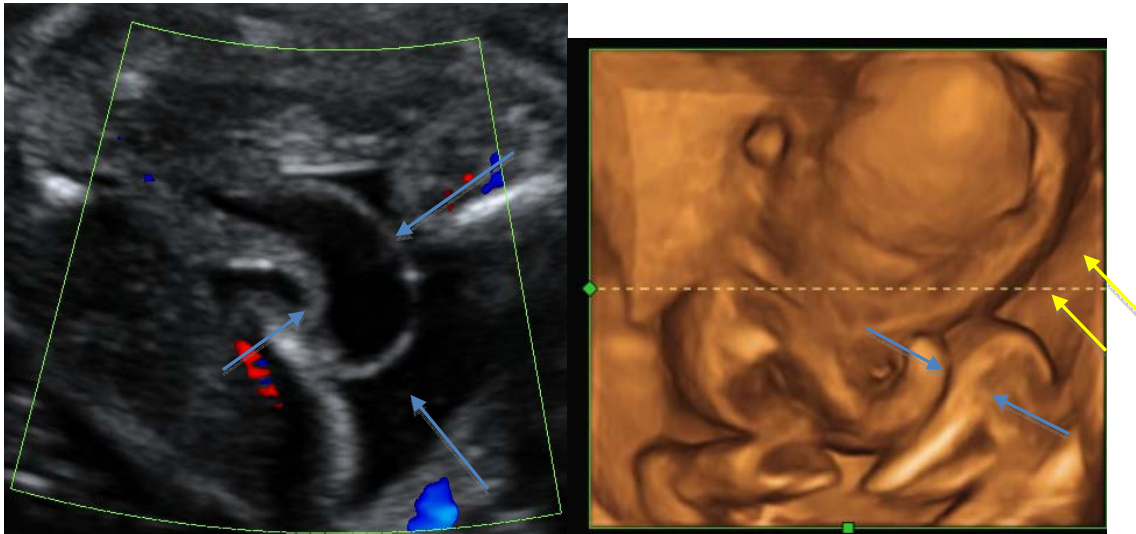


Figure 3A

3B

Fetal penile urethra is distended with fluid (blue arrows, 3A). 3D ultrasound demonstrates the dilated penis (blue arrows, 3B) and protuberant abdomen (yellow arrow, 3B)

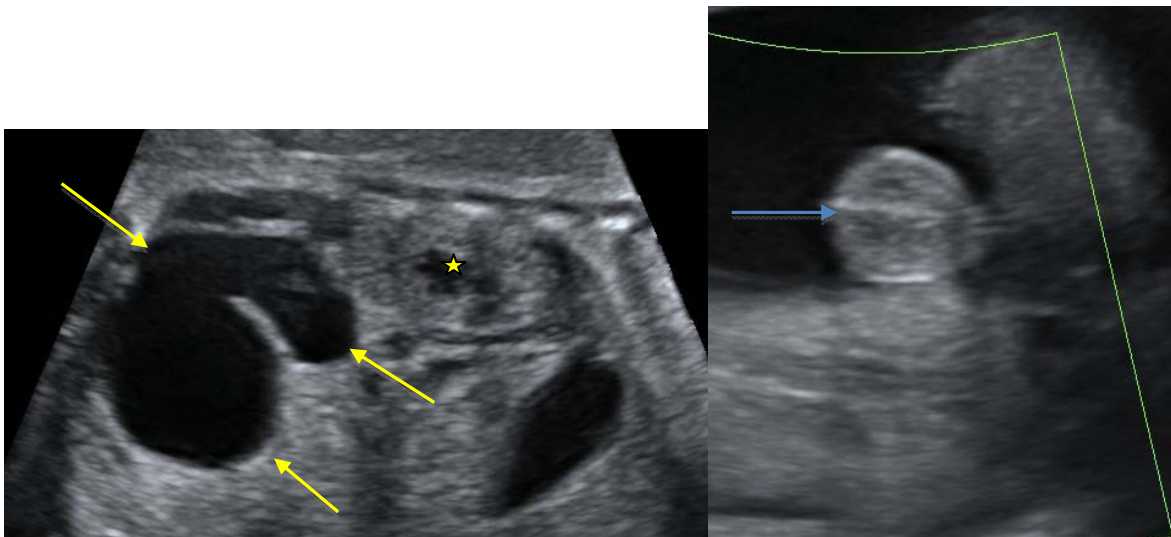


Figure 4A

4B

Follow up ultrasound at 30 wks shows continued massive distention of the ureters, left side shown here (4A, yellow arrows) below the left echogenic kidney (4A, yellow star) and lack of testicles in the scrotum (4B, blue arrow).



Figure 5
Post natal photograph of the newborn shows lax abdominal wall, enlarged penis from megalourethra, and non-descended testicles.



Figure 6A
Postnatal ultrasound of the right kidney demonstrates grade 4 hydronephrosis with a markedly dilated collecting system and thin renal parenchyma (6A). The right ureter is markedly distended (calipers, 6B)

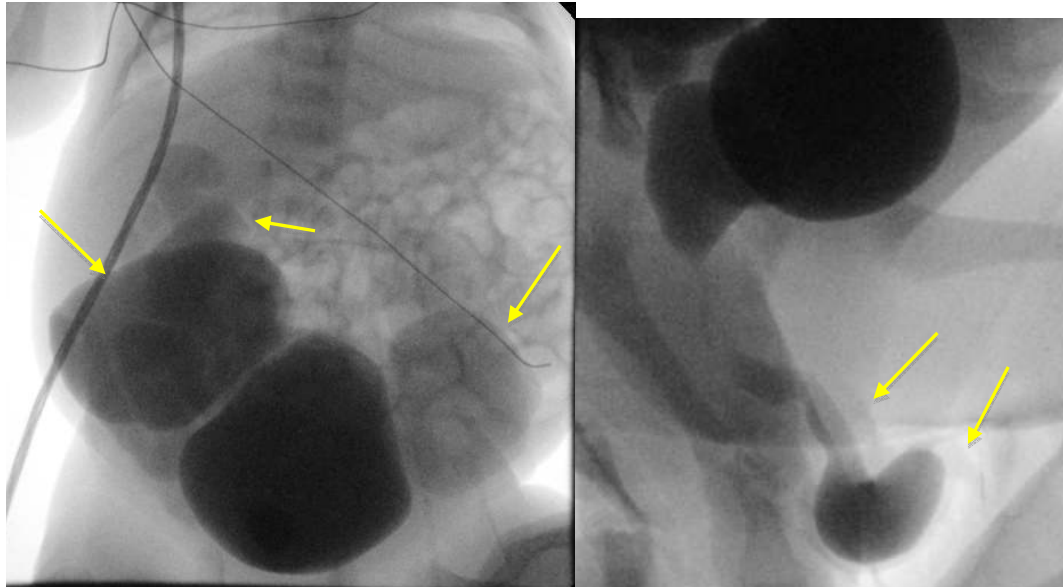


Figure 7A
7B
Retrograde urethrogram shows dilated ureters (yellow arrows, 7A) and urethra (yellow arrows, 7B)

Diagnosis: Prune Belly with Megalourethra

Discussion:

Prune belly syndrome is seen in approximately 1 in 40,000 males [1]. There is no known cause, but many associated abnormalities exist (including skeletal, gastrointestinal, cardiac, and pulmonary anomalies). Prune belly syndrome can be identified in utero and distinction can often be made from posterior urethral valves and other less frequent causes of bladder dilation. This case demonstrates classic findings of prune belly syndrome with megalourethra, with pre- and post-natal imaging.

Prune belly syndrome, also known as Eagle-Barrett syndrome in honor of early describers [2], is defined by a triad of (1) partial or complete lack of abdominal muscles, (2) cryptorchidism, and (3) abnormally dilated urinary tract. Up to 68% have associated urethral malformations, such as megalourethra. While primary urinary tract obstruction is thought to be the etiologic abnormality, the obstructive lesion is not always found. It is hypothesized that the obstructed or dilated bladder may block normal descent of testes and cause reflux of urine into the ureters, resulting in bilateral hydroureter, bilateral hydronephrosis and eventual renal insufficiency. The distended bladder may even exert pressure necrosis on the developing abdominal musculature, although this is theoretical. At times, severe prune belly syndrome in utero can result in oligohydramnios and pulmonary hypoplasia.

Diagnosis can be made in utero with ultrasound and fetal MRI, and in children with ultrasound and vesicoureterogram. On prenatal ultrasound, a fetus with prune belly syndrome will have gross dilation of the urinary system, including a dilated and thin-walled bladder, bilateral hydroureter and bilateral hydronephrosis. If severe megalourethra is present, then a distended fluid filled penis may be seen, as in our case. Oligohydramnios may be present but is often not as severe as cases with complete obstruction from posterior urethral valves. Prenatal MRI will have similar findings.

Postnatal imaging includes transabdominal ultrasound, again demonstrating bilateral hydroureteronephrosis and renal dysplasia. Vesicoureterogram will demonstrate an enlarged, elongated bladder lacking trabeculation, vesicourteral reflux in 85%, and frequently urachal diverticulum [1]. Some degree of megalourethra may also be seen in up to 68% of cases [3].

Prenatal findings suggestive of prune belly syndrome can be seen in the late first trimester. Bladder distention can be visible as early as 12 weeks [4]. However, in patients with early sonographic findings, the prognosis is worse due to more severe obstruction and renal and pulmonary dysfunction. Intrauterine therapy is reserved for the second or third trimester fetus with moderate disease, manifesting as urinary tract dilatation and oligohydramnios, but without pulmonary hypoplasia. Therapeutic options to relieve obstruction and halt ongoing renal injury include intrauterine vesicoamniotic shunting and postnatal vesicotomy. Additional postnatal therapies include abdominal wall reconstruction and orchiopexy.

The differential diagnosis of a grossly dilated bladder in a fetus includes posterior urethral valves and megacystitis microcolon intestinal hypoperistalsis syndrome. The latter is seen primarily in females (4:1) and presents with polyhydramnios, not oligohydramnios. Posterior urethral valves, though, cause proximal urethral obstruction, with resultant bladder dilation and hydroureteronephrosis and oligohydramnios, which can be difficult to distinguish from prune belly syndrome. Posterior urethral valves also presents typically in males. The imaging differences are subtle, including normally descended testes and thickened trabeculated bladder wall with posterior urethral valves. Treatment, associated abnormalities, and prognosis for prune belly syndrome and posterior urethral valves are different. [4]

Prune belly syndrome is a rare disease that can have dire outcomes. Its imaging features on prenatal ultrasound can suggest and even confirm the diagnosis, allowing appropriate prenatal counseling, prenatal interventions, and timely postnatal treatment.

References:

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2. Eagle JF, Barrett, GS. Congenital deficiency of abdominal musculature with associated genitourinary abnormalities: A syndrome. Report of 9 cases. *Pediatrics* 1950;6(5):721-36.
3. Kroovand, R.L., Al-Ansari, R.M., Perlmutter, A.D. Urethral and genital malformations in prune belly syndrome. *J. Urol.* 1982;127:94-95.
4. Papantoniou N, et al. Prenatal diagnosis of prune-belly syndrome at 13 weeks of gestation: case report and review of literature. *The Journal of Maternal-Fetal and Neonatal Medicine.* 2010; 23(10): 1263–1267.
5. Woodward P, Kennedy A, Sohaey R et al. *Diagnostic Imaging: Obstetrics, ed.* Amirsys 2011: 8:68-80.

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