

AMSER Case of the Month

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7-day-old male with low urine output and gross hematuria

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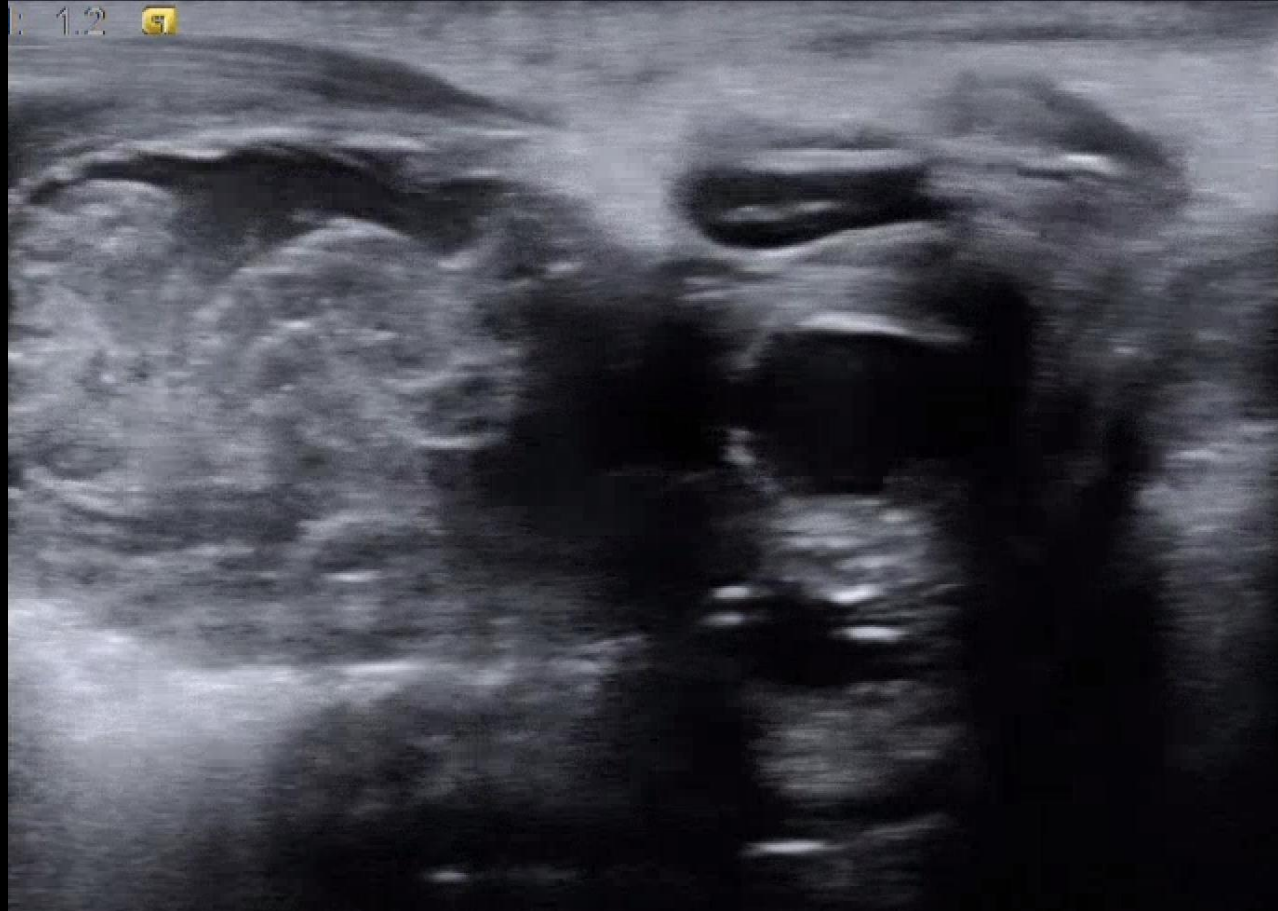
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Patient Presentation

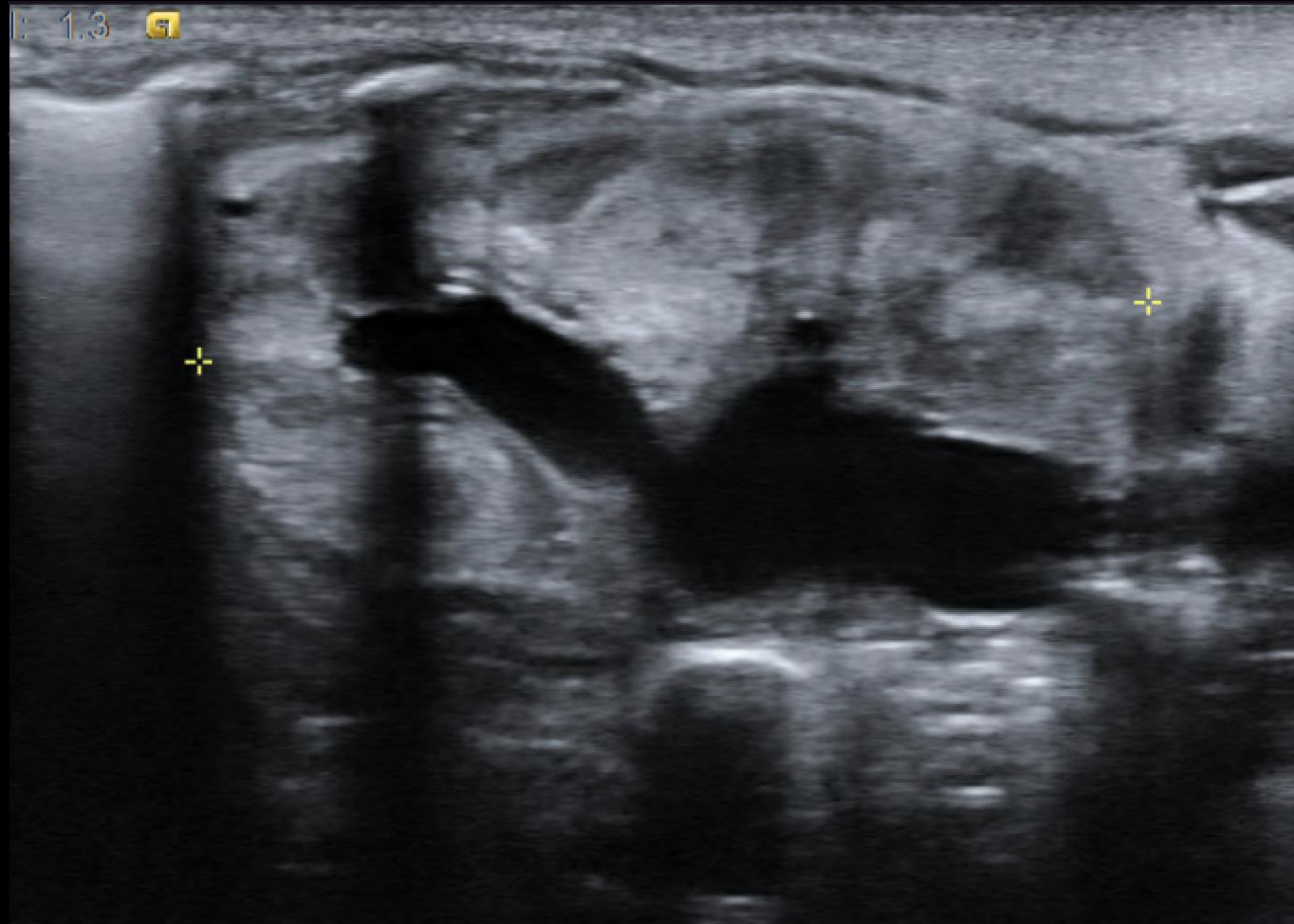
- **HPI:** 7-day-old male born at 31 weeks by C—section complicated by prenatal oligohydramnios presents to neonatal ICU with low urine output on catheterization and gross hematuria.
- **Review of Systems:** Positive for gross hematuria; otherwise, his preterm delivery.
- **Physical Exam (pertinent findings):**
 - Abdominal: Firm, distended bladder just below the umbilicus.
 - Genitourinary: Uncircumcised penis, no blood visible at the tip of the meatus. Bilateral descended testes with no abnormalities felt.
- **Initial laboratory values demonstrate:**
 - Cr 1.13 at time of birth → Cr 3.78 on presentation
- Subsequently, a renal ultrasound was performed

Findings (unlabeled)



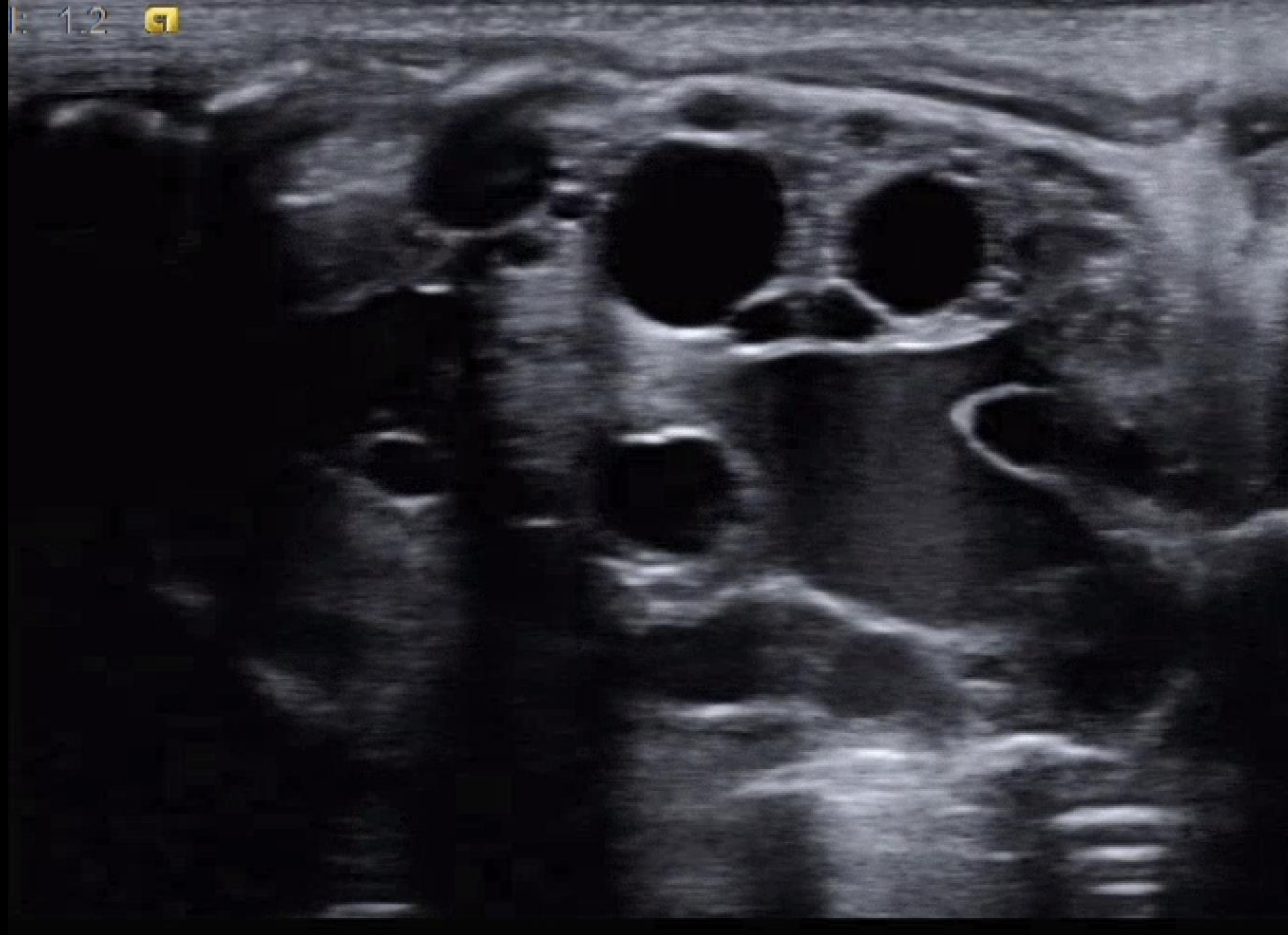
US bladder, sagittal view

Findings (unlabeled)



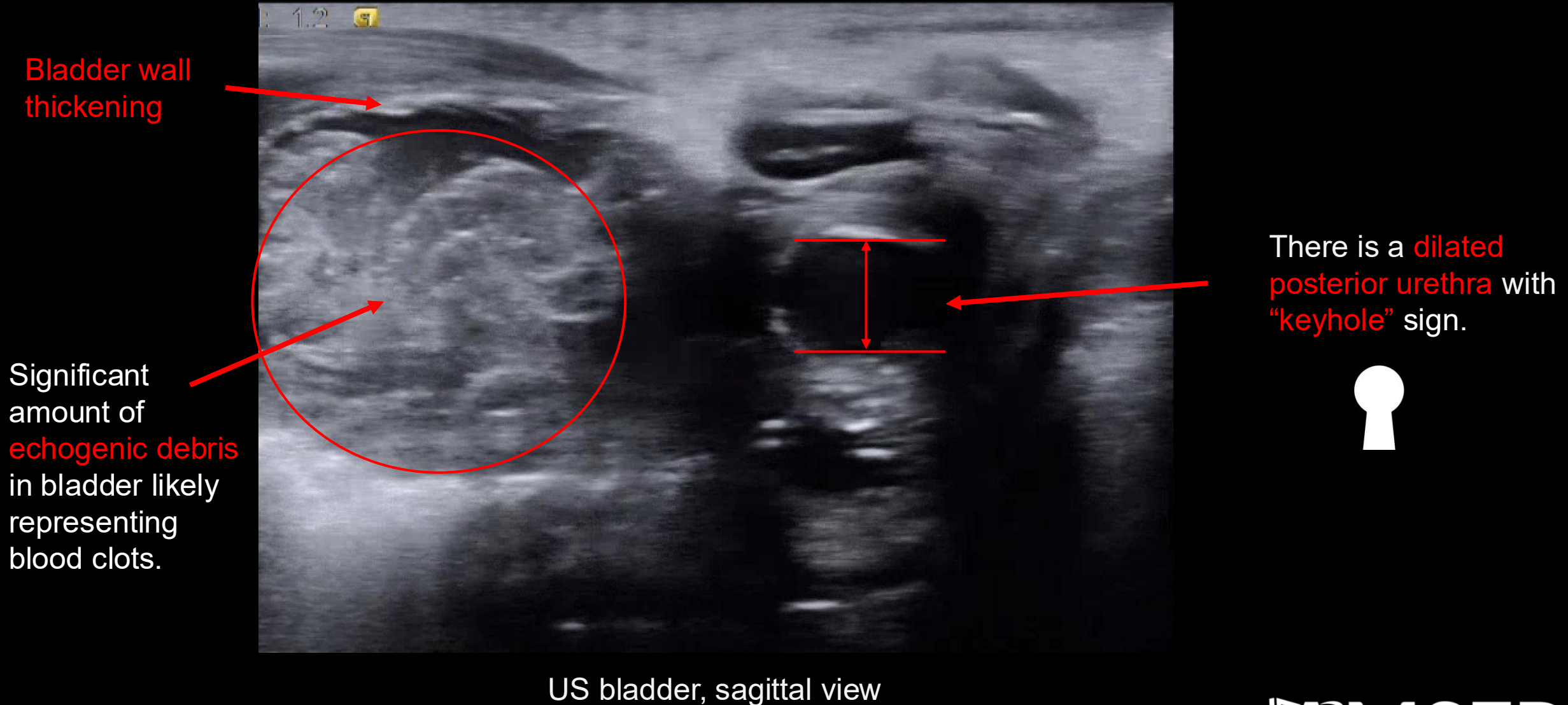
US left kidney, sagittal view

Findings (unlabeled)

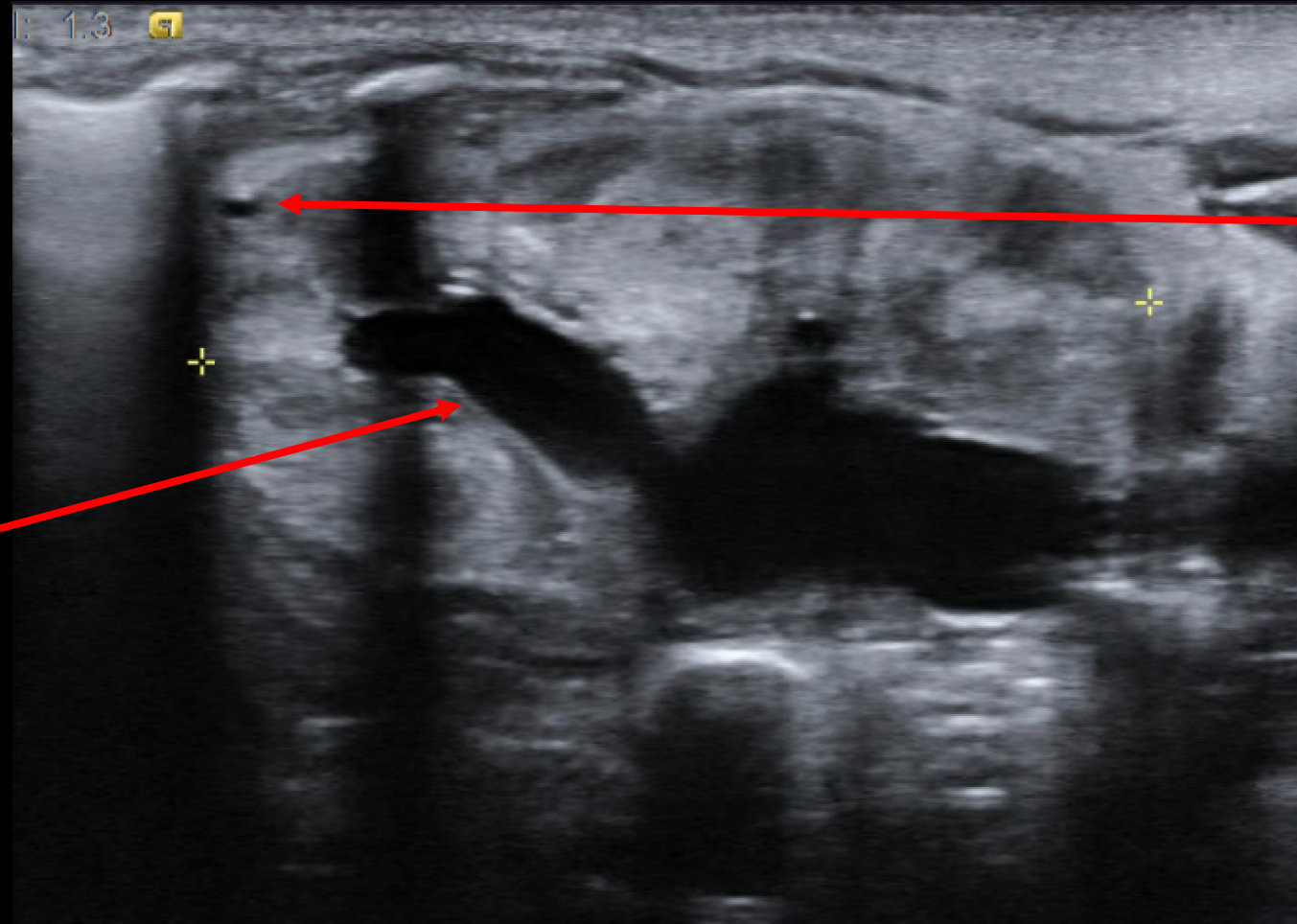


US right kidney, sagittal view

Findings (labeled)



Findings (labeled)



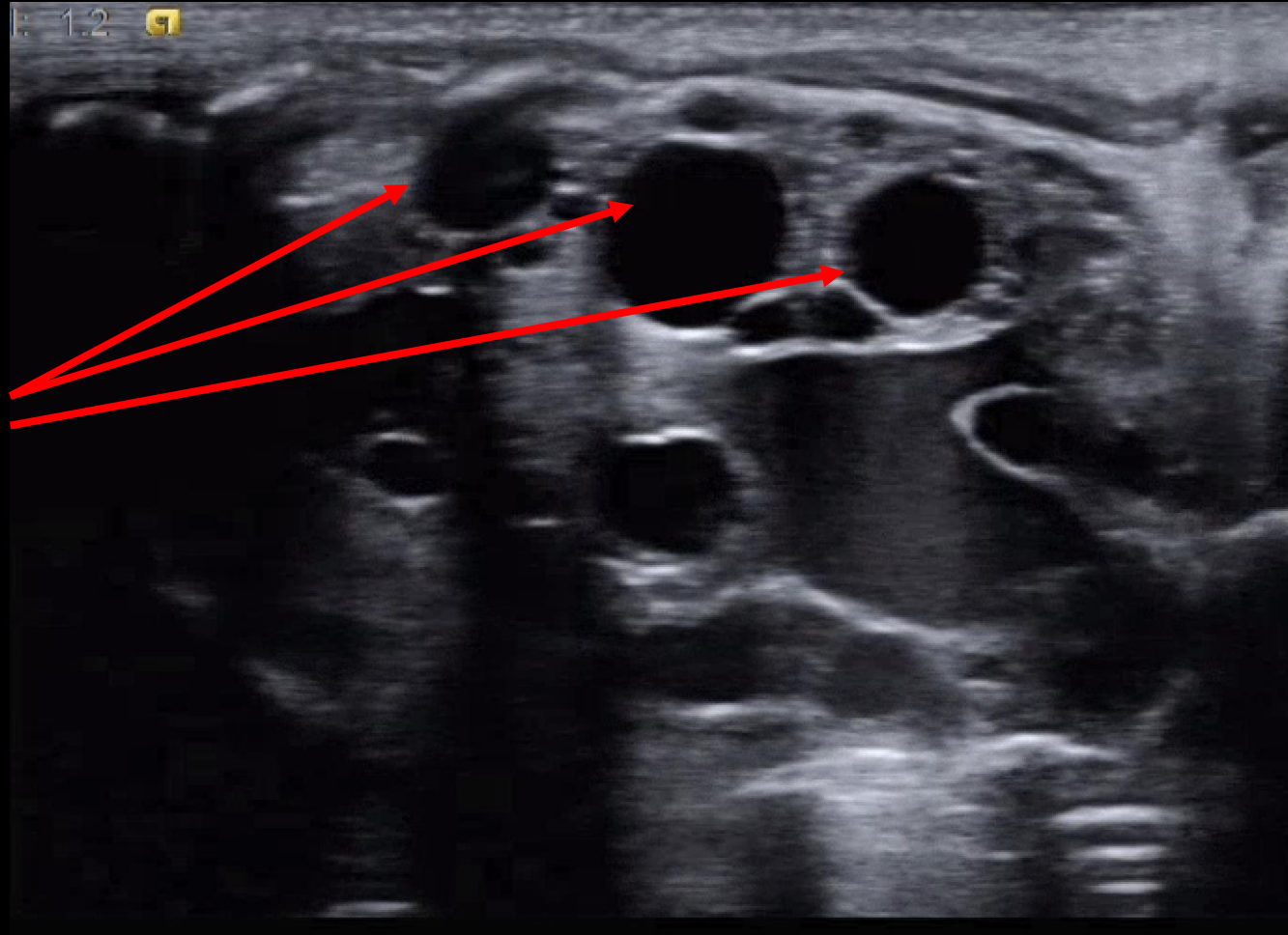
There is moderate
hydronephrosis.

Subtle **cysts** scattered
throughout kidney.

US left kidney, sagittal view

Findings (labeled)

There are **multiple cysts** and poor corticomedullary differentiation representing cystic dysplasia.



US right kidney, sagittal view

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 4. Male. Antenatal diagnosis of hydronephrosis with moderate or severe (SFU grade 3 or 4 or APRPD greater than 15 mm) hydronephrosis on initial neonatal ultrasound, or hydronephrosis associated with parenchymal abnormalities, hydroureter, bladder wall thickening, or posterior urethral dilation.

Procedure	Appropriateness Category	Relative Radiation Level
US kidneys and bladder follow-up in 1-6 months	Usually Appropriate	0
Fluoroscopy voiding cystourethrography	Usually Appropriate	☼☼
MAG3 renal scan	Usually Appropriate	☼☼☼
Voiding urosonography	Usually Not Appropriate	0
MRI abdomen and pelvis with IV contrast	Usually Not Appropriate	0
MRI abdomen and pelvis without and with IV contrast	Usually Not Appropriate	0
MRI abdomen and pelvis without IV contrast	Usually Not Appropriate	0
DTPA renal scan	Usually Not Appropriate	☼☼☼
Nuclear medicine cystography	Usually Not Appropriate	☼☼

This imaging modality was ordered by the physician

Findings (unlabeled)



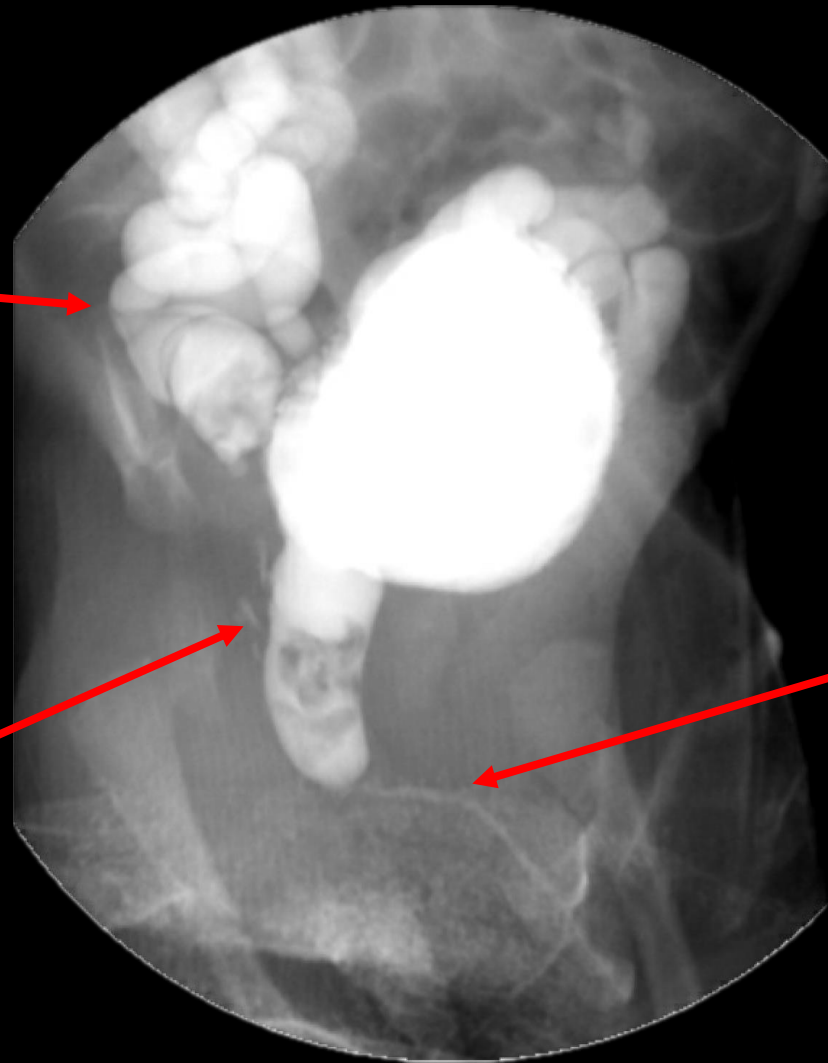
Voiding cystourethrogram (VCUG)

Findings (labeled)

There is bilateral retrograde filling of the **dilated and tortuous ureters**.

Bladder with wall thickening and trabeculations. There is **intermittent filling of the posterior dilated urethra**.

Narrow anterior urethra



Voiding cystourethrogram (VCUG)

Final Dx:

Hydroureteronephrosis & renal dysplasia caused by
congenital posterior urethral valves

Case Discussion

- Definition¹
 - Posterior urethral valves (PUV) is a congenital malformation in males where membranous folds of the urogenital membrane obstruct the membranous and prostatic urethra (posterior urethra).
- Epidemiology¹⁻³
 - Most common cause of urinary tract obstruction in newborn males.
 - Incidence is about 1 per 4,000 live births.
 - Around 25% of patients with posterior urethral valves develop end stage renal failure (ESRF), this being the most common urologic cause of renal failure in childhood.
 - Most cases are identified through antenatal ultrasound screenings.

Case Discussion

- Pathophysiology^{4,5}
 - Bladder outlet obstruction → detrusor/bladder neck hypertrophy, bladder wall thickening, and fibrosis → decreased compliance and increased intravesical pressure.
 - Elevated bladder pressures transmit upstream → vesicoureteral reflux, hydroureteronephrosis, renal tubular injury, and progressive renal dysfunction.
 - Prenatally, oligohydramnios and subsequent pulmonary hypoplasia can be caused in severe settings due to lack of urine output.
 - Clinical symptoms varies with severity:
 - abdominal pain/distension
 - recurrent UTIs
 - urinary retention
 - weak/straining stream
 - hematuria

Case Discussion

- Imaging appearance^{6,7}
 - Ultrasound of bladder and kidney is typically the preferred initial imaging
 - Bilateral hydronephrosis/ hydroureteronephrosis
 - Dilated prostatic urethra (“keyhole” sign)
 - Distended bladder with thickened wall (>3mm)
 - Focal renal parenchymal cysts (demonstrating renal dysplasia)
 - Voiding cystourethrography (VCUG) is “gold standard” for diagnosing PUV
 - Thickened bladder wall with prominent trabeculations
 - Vesicoureteral reflux (unilateral or bilateral)
 - Elongated, widened, or dilated prostatic urethra
 - An abrupt narrowing between the dilated prostatic urethra and the narrow bulbous urethra.

Case Discussion

- Management and outcome⁷⁻⁹
 - In acute retention/inability to pass urine, an attempt to drain the bladder with a catheter is performed.
 - First Line: Cystoscopic ablation
 - Endoscopic procedure involving transurethral incision of the posterior urethral valves in the membranous part of the urethra.
 - Second Line: Vesicostomy (preferred when cystoscopic access to the posterior urethra is challenging)
 - Surgical procedure connecting the bladder and the outside of the lower abdominal wall to allow for drainage of urine.
 - Since PUV are the most common urologic cause of renal failure in boys, renal transplantation is often required later in life.

The patient undergoes an ablation of posterior urethral valves and an eventual right renal transplant.

References:

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2. Thakkar D, Deshpande AV, Kennedy SE. Epidemiology and demography of recently diagnosed cases of posterior urethral valves. *Pediatr Res*. 2014;76(6):560-563.
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4. Sarhan OM. Posterior urethral valves: impact of low birth weight and preterm delivery on the final renal outcome. *Arab J Urol*. 2017;15(2):159-165.
5. Ziylan O, Oktar T, Ander H, Korgali E, Rodoplu H, Kocak T. The impact of late presentation of posterior urethral valves on bladder and renal function. *J Urol*. 2006;175(5):1894-1897.
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