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Congenital urethral stenosis in a female infant with a functional solitary kidney: A rare case and review of the literature

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Introduction

- **Congenital urethral stenosis (CUS)** in **female** infants is **extremely rare** and often underdiagnosed.
- May lead **to bladder outlet obstruction, vesicoureteral reflux (VUR)**, and **renal impairment**.
- When associated with a **solitary functioning kidney**, the risk of **irreversible renal damage** is high.
- Diagnosis is frequently delayed due to **nonspecific symptoms** (recurrent UTIs, weak stream, voiding dysfunction).
- This case illustrates **the diagnostic and therapeutic challenges** of CUS in a little girl with a single kidney and multiple comorbidities.

Case presentation

- **Patient:** 33-month-old female, ex-preterm (26 weeks), history of omphalocele repair, ventriculitis, developmental delay, retinopathy of prematurity, and chromosomal anomaly.
- **Past findings:** Left hydronephrosis; right kidney non-functional (solitary left kidney).
- **Presentation:** Recurrent febrile **UTIs** and progressive hydronephrosis of the solitary kidney.
- **Investigations**
  - Ultrasound: Left hydronephrosis, bladder diverticulum (7 mm).
  - DMSA scan: Solitary left kidney with preserved contour but heterogeneous uptake; no scarring.
  - Serum creatinine: 51 µmol/L → eGFR 59.5 mL/min/1.73 m^2.

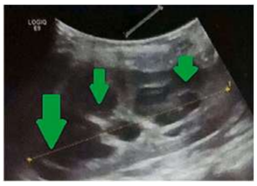

Fig. 1. Renal ultrasound showing urinary dilation of the left kidney (Arrow).

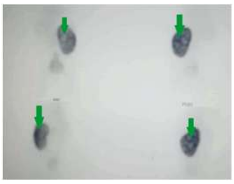

Fig. 2. DMSA scan showing absence of uptake in the right kidney and heterogeneous uptake in the left kidney (Arrow).

- **Catheterization:** Failed repeatedly due to **pinpoint urethral meatus.**

Fig. 3. Physical examination showing a punctiform urethral orifice (Arrow).

- **Diagnostic procedure**
  Examination under anesthesia (EUA):
  - Severely stenotic urethral orifice; endoscopic access impossible.
  - Serial dilations enabled insertion of a 6 Fr catheter.

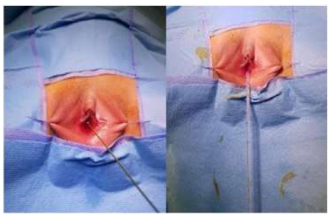

Fig. 4. Gentle dilations of the urethra eventually allowed passage of a 6 French catheter,

Voiding cystourethrogram (VCUG):

Fig. 5. The VCUG showing a thick-walled, trabeculated bladder with multiple diverticula (Star) and severe grade V vesicoureteral reflux (Arrow).

- **Management**
  - Due to persistent infection and poor tolerance of catheterization → **Vesicostomy** performed for continuous bladder drainage.
- **Follow-up (3–12 months):** UTI-free, improved renal function, regression of hydronephrosis.
- Patient remains on **vesicostomy management** due to neurological status.

Discussion

- **CUS in females**: exceptionally rare; results from incomplete canalization of the urogenital sinus or persistent embryologic membranes.
- **Clinical presentation:** recurrent UTIs, voiding difficulty, or hydronephrosis; often misdiagnosed as functional bladder dysfunction.
- **Diagnostic key:**
  - EUA and cystoscopy = most reliable for confirming fixed distal obstruction.
  - VCUG and ultrasound help assess bladder morphology and reflux.
- **Differential diagnoses:** neurogenic bladder, posterior urethral valves (rare in females).
- **Treatment options:**
  - **Urethral dilation** – first-line; may require periodic repetition.
  - **Meatotomy or urethrotomy** – for refractory cases.
  - **Vesicostomy** – effective for children unable to perform clean intermittent catheterization (CIC).
- **Goal:** preserve renal function by ensuring **low-pressure bladder** drainage and preventing infection.
- **In this case:** vesicostomy provided safe, stable drainage and improved renal outcomes.

Conclusion

- **CUS should be suspected** in female infants with recurrent UTIs and signs of bladder outlet obstruction.
- **Examination under anesthesia** is essential for diagnosis when catheterization fails.
- **Vesicostomy** is a valuable interim or long-term option in children with **solitary kidney** or neurological impairment.
- **Early recognition and intervention** are critical to preserve renal function and prevent irreversible damage.

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