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Italian Journal of Medicine

Italian Journal of Medicine 2026; 20:2666
doi:10.4081/itjm.2026.2666

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When congenital anomalies of the kidney and urinary tract are discovered late: incidental unilateral renal agenesis and ipsilateral ureterocele in an elderly patient

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Key words: CAKUT, unilateral renal agenesis, ureterocele, late diagnosis, solitary kidney, chronic kidney disease.

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Dear Editor,

Congenital anomalies of the kidney and urinary tract (CAKUT) are increasingly recognized in adulthood, frequently as incidental findings during imaging performed for unrelated indications.[1] We herein describe an exceptional case of late-discovered CAKUT in an elderly patient and discuss its embryological basis, diagnostic approach, and clinical implications for the practicing internist.

A 73-year-old man with a history of cardiac disease underwent routine abdominal ultrasound during cardiovascular follow-up. He was entirely asymptomatic from a urological and nephrological standpoint, denying lumbar pain, hematuria, recurrent urinary tract infections, lower urinary tract symptoms, and previous urolithiasis. Physical examination was unremarkable, and laboratory tests, including renal function indices, were within normal limits. Ultrasound incidentally revealed non-visualization of the left kidney and a cystic protrusion into the bladder lumen suggestive of an ipsilateral ureterocele (Figure 1). Abdominal computed tomography, performed before and after intravenous iodinated contrast administration, confirmed congenital agenesis of the left kidney with compensatory hypertrophy of the right kidney, and demonstrated an orthotopic intravesical left ureterocele (Figure 2). No proximal or distal left ureteral structures were identifiable, configuring a picture of complete agenesis of the ipsilateral excretory system associated with an isolated ureterocele. No residual renal parenchyma, signs of urinary tract obstruction, hydronephrosis, urolithiasis, or additional congenital anomalies were detected, and the patient had no history of nephrectomy. Transurethral cystoscopy under local anesthesia confirmed a translucent cystic bulge at the left bladder wall with intact mucosa, a thin underlying vascular layer, and no signs of inflammation or papillary lesions. The contralateral right ureteral orifice was single, of regular morphology, and in a normal anatomical position, with spontaneous clear urinary efflux. The finding was entirely incidental in a patient who had reached his eighth decade without prior clinical manifestations attributable to a congenital urinary tract malformation.

This case illustrates the exceedingly rare persistence of a blind-ending distal ureteral remnant despite complete ipsilateral renal agenesis. Ureteroceles are classified as simple intravesical or ectopic extravesical lesions, and may be further categorized according to the Stephens classification into stenotic, sphincteric, sphincterostenotic, and cecoureterocele types. They are thought to arise from failed regression of the Chwalla membrane, a transient embryological structure occluding the ureteral orifice during development. Unilateral renal agenesis, conversely, results from failure of the ureteric bud to induce metanephric mesenchymal differentiation, with involvement of multiple developmental genes; recent exome-based studies have expanded the spectrum of causal genes and strengthened the link between kidney malformations and broader developmental disorders.[2,3] The coexistence of these anomalies suggests an early defect in Wolffian duct formation, with persistence of a distal ureteral remnant. CAKUT encompasses nephropathies and uropathies, representing the most common group of congenital malformations.

Although prenatal screening has markedly reduced late presentations, congenital unilateral renal agenesis remains uncommon but well documented in population-based registries and meta-analyses. In adults, ureteroceles may present with recurrent urinary tract infections, lower tract symptoms, abdominal pain, or hematuria, but are frequently asymptomatic.[4,5] In our patient, the absence of the ipsilateral kidney combined with a ureterocele still poses a theoretical risk to the solitary functioning kidney through reflux, obstruction, or infection.

Accurate characterization requires a multimodal diagnostic approach. Ultrasound is the first-line modality, capable of demonstrating renal absence, assessing compensatory hypertrophy of the contralateral kidney, and identifying the classic cystic bladder lesion; color Doppler may reveal the dilated ureter projecting into the bladder lumen. Computed tomography urography provides detailed anatomic mapping and may exclude an ectopic pelvic kidney not visualized on ultrasound. Magnetic resonance imaging with T2-weighted sequences offers excellent soft-tissue resolution without ionizing radiation and is particularly valuable for repeated follow-up in elderly patients with compromised renal function in whom iodinated contrast is contraindicated. Cystoscopy remains the gold standard for confirming the intravesical cyst and allows simultaneous endoscopic intervention.

In asymptomatic adults with simple intravesical ureteroceles, conservative surveillance may be appropriate; however, in the setting of a solitary kidney, a proactive approach is generally preferred given the infectious risk to the remaining organ.

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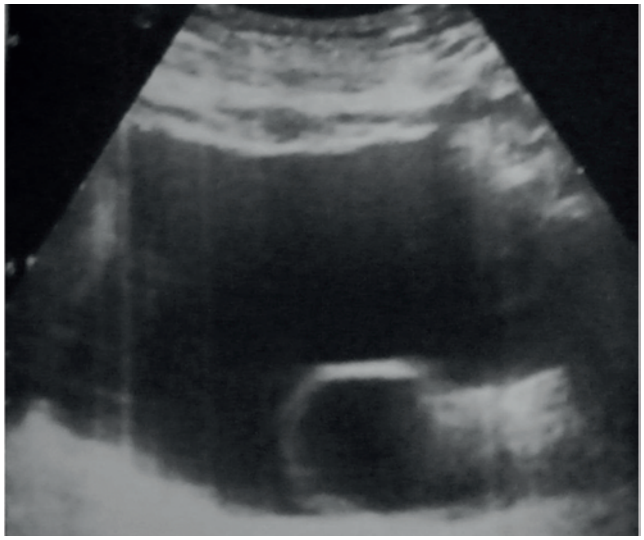


Figure 1. The urinary bladder is adequately distended, with regular wall thickness. At the level of the left ureteral orifice, a well-defined, thin-walled anechoic intravesical cystic lesion is identified, protruding into the bladder lumen, consistent with a left ureterocele.



Figure 2. A well-circumscribed, fluid-attenuation intravesical cystic lesion is seen arising from the left ureterovesical junction and protruding into the urinary bladder lumen, consistent with a left ureterocele.

Endoscopic incision/meatotomy is generally considered first-line in symptomatic or complicated cases, reserving reconstructive surgery for failures or complex anatomy.

The greater clinical challenge lies in the long-term stewardship of the congenital solitary kidney. Contrary to the traditional view of a benign condition, systematic review evidence indicates that unilateral renal agenesis is frequently associated with additional urogenital anomalies and confers clinically relevant lifetime risks of hypertension, albuminuria, and reduced glomerular filtration rate.[1,6] Contemporary consensus recommendations on congenital solitary kidney emphasize structured lifelong surveillance and risk stratification.[7] Patient education regarding the non-benign nature of the congenital solitary kidney is essential to ensure adherence to long-term follow-up.

In conclusion, the incidental discovery of unilateral renal agenesis with an ipsilateral ureterocele in an elderly patient underscores the importance of considering CAKUT even in late adulthood. A structured diagnostic pathway confirms the anatomy, while the internist must assume a proactive role in lifelong nephrological surveillance to mitigate the risks of hypertension, albuminuria, and progressive chronic kidney disease.[1,6,7]

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Received: 30 July 2026; Accepted: 7 October 2026; Early view: 27 October 2026.

Contributions: all authors contributed to the literature search, writing, drafting, and reviewing of the manuscript, doing refinement of sections in alignment with journal guidelines. All authors approved the final version.

Conflict of interest: the authors declare no competing interests. All authors have reviewed the manuscript and confirm the accuracy of the data and the integrity of the work.

Ethics approval and consent to participate: the ethics committee deemed formal approval unnecessary, as no experimental procedures were performed and national and international therapeutic guidelines were followed. The study was conducted in accordance with the Helsinki Declaration. Clinical case concerns a patient admitted to the hospital and directly managed by the authors.

Patient consent for publication: informed consent was obtained from the patient for the publication of the clinical case.

Availability of data and materials: the clinical case data are available.

Declaration of generative artificial intelligence and artificial intelligence-assisted technologies in the writing process: the authors declare that no generative artificial intelligence (AI) tools or AI-assisted technologies were used in the conception, preparation, writing, editing, analysis, or revision of this manuscript.

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