Bladder Agenesis Associated with Crossed Fused Renal Ectopia and Vertebral Anomalies: A Rare Entity

Stephen Lalfakzuala Sailo, 1* Laltanpuii Sailo²

A 9-month-old girl presented with dribbling urine and no normal voiding since birth. There was no history of urinary tract infection. On examination, the child appeared healthy. An abdominal examination was normal. On separation of the labia, a single opening with leaking urine was seen; no separate urethral opening was identified (**Figure 1**). Abdominal ultrasonography revealed a crossed left renal ectopic kidney. Computed tomography also showed a normally excreting crossed fused ectopic kidney/lump kidney (**Figure 2A**), ureters draining into a urogenital sinus, and butterfly and block lumbar vertebrae (**Figure 2B**). The serum creatinine level was normal (0.3 mg/dL). Endoscopy using a pediatric cystoscope inserted through the genital opening revealed a urogenital sinus in which both ureteric openings were located (**Figure 3**); both ureters refluxed when the sinus was filled with contrast (**Figure 4**). We plan to divert the ureters to a cutaneous stoma. Agenesis of the urinary bladder is extremely rare; only 23 living cases have been reported. Agenesis is attributable to injury to the urogenital sinus at weeks 5~7 of embryogenesis. The condition is associated with renal and skeletal anomalies. Young age is not a contra-indication for continent urinary diversion using a self-catheterizable pouch. On the property of the urinary diversion using a self-catheterizable pouch.



Figure 1. Genital opening filled with urine (arrow).



Figure 2. Computed tomographic images of A) the crossed fused ectopic kidneys/lump kidney (arrow) and, B) the crossed fused ectopic kidneys, ureters draining into the urogenital sinus (triangle), block vertebra (arrow), and butterfly vertebra (star).

Department of Urology, and Anaesthesia, Morth Eastern Indira Gandhi Regional Institute of Health and Medical Sciences, Shillong, Meghalaya, India.

*Correspondence: Department of Urology, North Eastern Indira Gandhi Regional Institute of Health and Medical Sciences, Shillong, Meghalaya, India.

Tel: +91 364 2538006. Fax: +91 364 2538010. E-mail: stephensailo@gmail.com.

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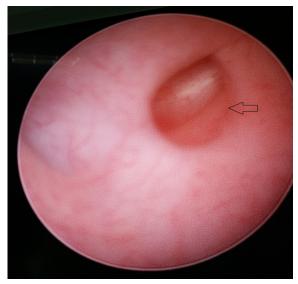


Figure 3. Endoscopy using a pediatric cystoscope shows the urogenital sinus and left ureteric orifice (arrow).



Figure 4. Both ureters and the pelvi-calyceal systems became opacified when the urogenital sinus was filled with contrast.

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