

Rare exstrophy variant with unilateral renal agenesis

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DESCRIPTION

A 3-month-old male child presented with continuous dribbling of urine from the lower abdominal wall. The child had an exposed bladder plate with polypoidal mucosa ([figure 1](#)). The penis was normal and had no chordee. The prepuce was intact. The urethral meatus was located at the glans tip. An 8-French infant feeding tube could be easily passed through the urethra. Bilateral inguinal hernia was present, which was reducible. The position of the anus was normal.

Ultrasonography showed a normal right kidney and absent left kidney. The urinary bladder was not visualised. Dimercaptosuccinic acid scan demonstrated non-visualised left kidney and normal right kidney ([figure 2](#)).

Only two such cases have been reported previously in the English literature.^{1,2} Both the previously described cases did not have any associated anomaly. Our case is unique as it has associated renal agenesis. Kulshrestha *et al* described it as a variant of the superior vesical fissure, which itself is an exstrophy variant.² Exstrophy variants are rare congenital anomalies with an incidence of 1 in 4–5 lakhs.³ The term exstrophy variant is under debate because they are similar to classical exstrophy only in having pubic diastasis and divergence of rectus muscles. Their prognosis in terms of urinary continence is better because of the intact sphincteric mechanism. They are associated with different types of congenital anomalies as opposed to classical exstrophy–epispadias

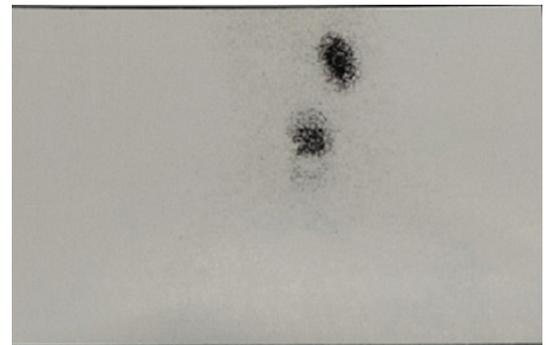


Figure 2 Dimercaptosuccinic acid scan showing left renal agenesis.

complex.⁴ Different types of exstrophy variants have been described, and we also identified a rare exstrophy variant with left renal agenesis.

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Figure 1 Exposed bladder plate with normal phallus.

Perspective of patient's mother

I am worried about my child as he is suffering from a rare disease and cannot hold urine. His one kidney is also absent and there is continuous dribbling of urine. The elderly in the family do not want to play with the child because of urine smell. I am afraid that my child will not adjust well in the society if the disease is not cured.

Learning points

- ▶ Exstrophy variants are of different types and have various associated anomalies.
- ▶ Exstrophy variants must be investigated for associated anomalies.

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