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Posterior Urethral Valves Leading to Massive Hydronephrosis in A Four-Day Old Baby

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Received Date: 03 November 2023 | **Accepted Date:** 08 December 2023 | **Published Date:** 15 January 2024

Citation: Acharya S., Shukla S., Shinde R., Tapesh D Nagaria., (2024), Posterior Urethral Valves Leading to Massive Hydronephrosis in A Four-Day Old Babys, *Journal of Clinical Surgery and Research*, 5(1); **DOI:10.31579/2768-2757/103**

Abstract:

Posterior urethral valves (P.U.V.s), also referred to as congenital obstructing posterior urethral membranes (C.O.P.U.M.) is a congenital malformation affecting males where the membranous folds of the urogenital membrane obstruct the membranous and prostatic urethra. It is a congenital disorder only seen in males, with an incidence of 1 per 4000 to 1 per 25000 live births in some areas. Type 1 P.U.V. results from the abnormal insertion and absorption of the distal Wolffian duct during the development of the bladder. Type 3 PUV is observed as a membrane in the posterior urethra, originating from the incomplete canalisation between the anterior and the posterior urethra. Type 2 P.U.V. is now considered a normal anatomical variant. Regardless of the type, P.U.V.s present with urinary tract obstruction, bladder distension, abdominal distension, difficulty voiding, poor stream of urine, recurrent Urinary Tract Infection (U.T.I.), diurnal enuresis and a failure to thrive. They are diagnosed Antenatally by Ultrasonography showing distended or thick-walled bladder, bilateral hydroureters, and bilateral hydronephrosis. However, the gold standard for the diagnosis is a Micturating

CystoUrethrogram (MCU), also known as a Voiding Cystourethrogram (V.C.U.) shows dilation/elongation of the posterior urethra during voiding along with signs of vesicoureteral reflux if present. This patient was antenatally diagnosed with posterior urethral valves at seven months of fetal age and came to us for further management. Antenatal diagnosis of Posterior Urethral Valves on Ultrasonogram involves visualisation of 1. Keyhole sign- due to the distension of the urethral proximal to the valves and a distended bladder, 2. marked distention or hypertrophy of the bladder, 3. Hydronephrosis and hydroureter. An Ultrasonogram of the abdomen and pelvis done postnatally revealed bilateral gross hydroureteronephrosis, which was missed on antenatal scans. The baby was born to a G2P1L1 mother and weighed 3.04kg at birth. The baby cried immediately after birth and maintained oxygen saturation at 100% on room air. The patient was managed with Cystoscopy with Posterior Urethral Valve fulguration with circumcision.

keywords: congenital disorder; diagnosis; cystourethrogram

Images-

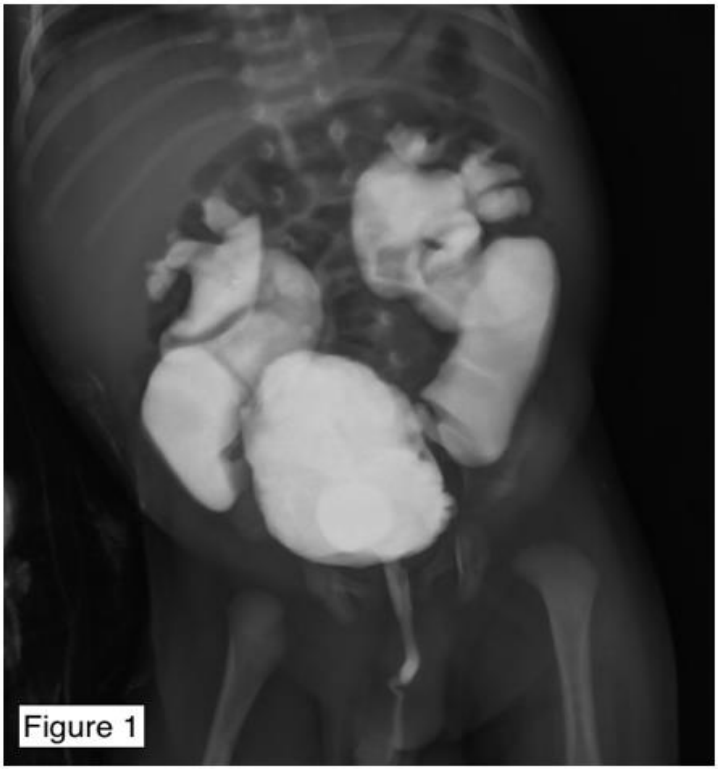

Figure 1: a Micturating Urethrogram showing bilateral grossly dilated ureter and renal pelvis along with posterior urethral valve as a narrowing in the posterior urethra.

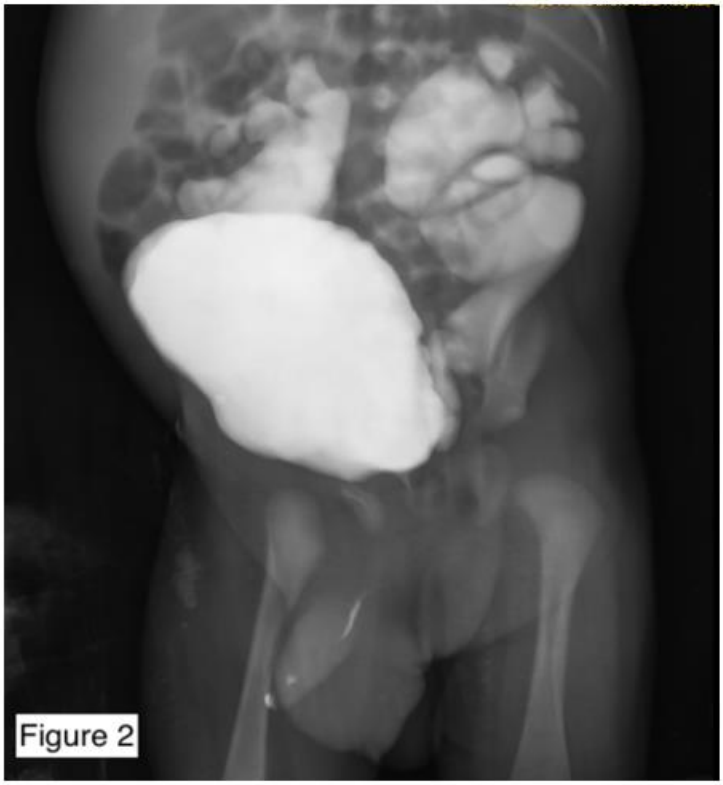

Figure 2: A Micturating Urethrogram - Lateral view - showing bilateral grossly dilated ureter and renal pelvis along with posterior urethral valve as a narrowing in the posterior urethra.

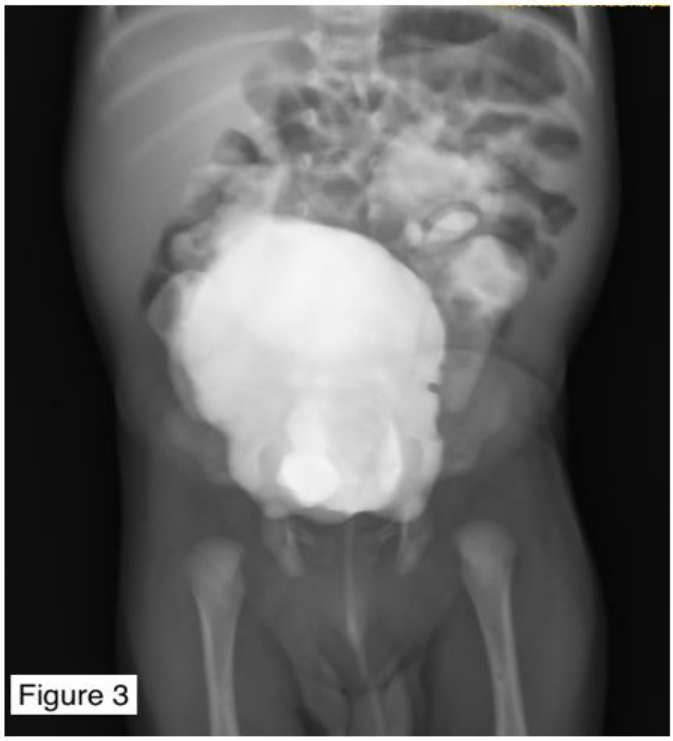

Figure 3: A Micturating Urethrogram - showing filled-up urinary bladder.

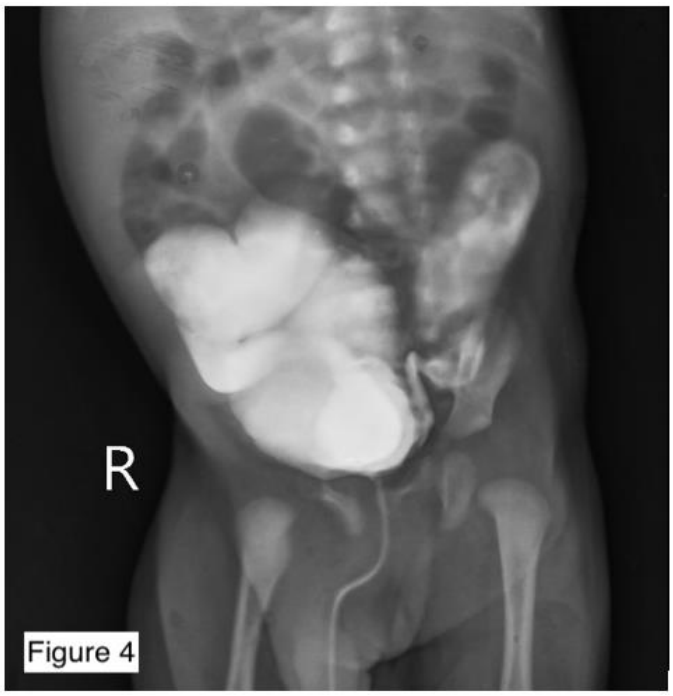

Figure 4: A Micturating Urethrogram - Lateral view - showing bilateral grossly dilated ureter and renal pelvis along with posterior urethral valve as a narrowing in the posterior urethra.

Final Diagnosis-

Bilateral Grade V Hydronephrosis with Posterior Urethral Valves

Differential Diagnosis-

Urethral Stricture

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