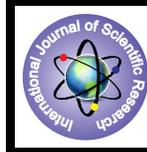


## Association of Crossed Fused Ectopic Kidney with Multiple Congenital Anomalies Presenting as Persistent Ureterohydronephrosis



### Medical Science

**KEYWORDS :** crossed renal ectopia, vesicoureteral reflux, hypospadias, congenital anomalies

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### ABSTRACT

*Crossed fused renal ectopia (CRE) refers to an anomaly where the both kidney situated on the same side of the body. The condition occurs predominantly in males, and inferior ectopia is the most common type. Most cases of CRE are asymptomatic and incidentally diagnosed, except for symptomatic conditions such as urinary tract infections, urolithiasis, and obstruction. The association of CRE with anorectal and urological anomalies has been described separately. Herein, we present a case of inferior-type right fused CRE with imperforate anus, hypospadias with meatal stenosis, bilateral vesicoureteral reflux, and neurogenic bladder with refractory ureterohydronephrosis requiring multiple operations.*

### Introduction

Crossed fused renal ectopia (CRE) was first described by Pan-norlus in 1964 and refers to a condition where one kidney has crossed to the opposite site of the body. Generally, the two kidneys will fuse together; this condition is referred to as CRE (Birmole, Borwankar, Vaidya, & Kulkarni, 1993). There is a male predominance, and the left-to-right crossover occurs more frequently with the inferior ectopia as a common variety (Asghar & Wazir, 2004). The ureter of the ectopic kidney that has crossed the midline is inserted at its normal position into the bladder. Although most cases of CRE are asymptomatic and incidentally diagnosed, early diagnosis may prevent possible urinary tract infection, malignancies, and obstructions. CRE may be associated with other congenital anomalies such as anorectal malformation, anorchia, ureterocele, vesicoureteral reflux (VUR), and ureteropelvic junction obstruction (UPJO) (Arena et al., 2007; Lodh, Kanwar, Singh, & Singh, 2013; Narci et al., 2010; Solanki, Bhatnagar, Gupta, & Kumar, 2013). Imperforate anus constitutes 4% of anomalies associated with CRE (Belekar, Dewoolkar, Desai, Anam, & Parab, 2009; Mülalayim, Demirba? o? lu, & Oral, 2003). The association of hypospadias with CRE was previously described as a rare accompanying congenital anomaly (GUARINO et al., 2004). Imperforate anus, hypospadias, and VUR have each been described previously as accompanying congenital anomalies with CRE. Herein, we present the case of right fused CRE with accompanying imperforate anus, hypospadias, meatal stenosis, and bilateral VUR with refractory ureterohydronephrosis subjected to multiple operations.

### Case report

A 6-year-old boy was referred to the pediatric urology center for persistent ureterohydronephrosis and high creatinine level. At birth, he underwent partial sigmoid resection with double colostomy and anoplasty due to imperforate anus. A month later, he underwent urethromatoplasty for meatal stenosis. A year later, he underwent herniotomy, hydrocelectomy, and hypospadias repair by perineal urethroplasty. Further evaluation with ultrasonography revealed an absent kidney in the left side with inferior-type right CRE. A dimercaptosuccinic acid scan confirmed a zone of radiotracer uptake in the right renal bed with downward extension in the medial portion of the right kidney (Figure 1). A notable finding in this patient was severe bilateral

ureterohydronephrosis confirmed by ultrasonography with the diagnosis of bilateral VUR (Figure 2). First, to resolve the bilateral VUR and lack of possibility of reimplanting both ureters to the small sized bladder, the right ureter was anastomosed to the left one, and then the left ureter was reimplanted to the bladder by the Cohen ureteroneocystostomy approach. Follow-up evaluation revealed persistent bilateral ureterohydronephrosis, particularly in the right side. A diethylenetriamine pentaacetate scan confirmed decreased perfusion in the right kidney and no response to diuretic administration, and therefore, the patient underwent left pyelouretostomy. Despite all of these surgical procedures (which were performed in a district hospital), he was referred to the pediatric urology center with bilateral ureterohydronephrosis and a high creatinine level. Urodynamic study confirmed spastic neurogenic bladder with detrusor sphincter dysynergia (Pdet =40CmHg and high residue). The cystoscopy was performed and a double J stent was placed at the left ureter orifice, which traversed the midline to resolve the obstructive uropathy and improved bilateral ureterohydronephrosis (Figure 3). Ultrasonography confirmed the resolution of bilateral ureterohydronephrosis. In addition, the creatinine value was significantly decreased after the placement of the double J stent. Botox was injected for the improvement of spastic neurogenic bladder. Finally, the patient was discharged with routine clean intermittent self-catheterization.

### Discussion

CRE is a rare congenital anomaly with an incidence of 1:1300–1:7500 generally diagnosed during the third decade (Ahmad, 2007; Nursal & Büyükdere?l, 2005). CRE is the second most common fusion abnormality of the kidney after horseshoe kidney and commonly affects males more than females (Kaur, Saha, Mrigiani, Saini, & Gupta, 2013). CRE may result from the abnormal development of the ureteric bud and metanephric blastema between 4 and 8 weeks of gestation (Solanki et al., 2013). CRE was anatomically classified by McDonald & McClellan into 4 groups: 1) CRE with fusion; 2) CRE without fusion; 3) solitary CRE; and 4) unfused bilateral CRE (Türkvtan, Olçer, & Cumhuri, 2009). The incidence of fused type CRE is more than that of unfused, and is classified as inferior, superior, sigmoid, lump, and disc shape (Sharma & Bargoutra, 2009). Most commonly the upper pole of the inferiorly positioned crossed ectopic kidney is fused to the lower pole of the superior normally placed kidney (Jain,

Kaza, Gupta, & Kumar, 2007). In the present case of inferior-type CRE, the left kidney had crossed and fused to the right kidney. The most frequent abnormalities associated with CRE are skeletal abnormalities, imperforate anus, septal cardiovascular defect, VUR, and UPJO. There are also less commonly associated abnormalities, such as hypospadias, cryptorchidism, posterior urethral valves, and multicystic dysplasia (Kramer & Kelalis, 1983; Sharma & Bargetra, 2009). The present case had accompanying anomalies such as imperforate anus, bilateral VUR, and hypospadias with meatal stenosis. The coincidence of bilateral VUR and neurogenic bladder in this case maybe attributable to the presence of lower tract obstruction due to hypospadias and meatal stenosis, rather than primary VUR. Most patients with CRE are usually asymptomatic but are more susceptible to developing complications like urolithiasis, urinary tract infection, obstructive uropathy, and abdominal mass (Kaur et al., 2013). The treatment is guided toward the associated problems, leading to either symptoms or the deterioration of the upper tract, such as UPJO or VUR, which requires pyeloplasty or the injection of a bulking agent (Solanki et al., 2013). In this case, the presence of bilateral VUR resulted in symptomatic obstructive uropathy with bilateral ureterohydronephrosis. Bilateral ureteroneocystostomy was not possible due to a small sized bladder. Therefore right to left ureteroureterostomy with left ureteroneocystostomy was performed. The association of imperforate anus, hypospadias, and VUR with CRE was previously described as a distinct moiety, but the persistence of these abnormalities together made a complex case, leading to multiple operations. The treatment modality for imperforate anus as the commonly associated anomaly with CRE (4%) was performed at the time of the birth with complete resolution and normal sphincter function. Association of CRE with hypospadias was previously described as our case underwent perineal urethroplasty and urethromeatoplasty for hypospadias repair and meatal stenosis, respectively. Secondary bilateral VUR with neurogenic bladder is a challenging subject in such a patient. The primary surgical reimplantation of one ureter with subsequent right to left ureteroureterostomy had not resolved the ureterohydronephrosis, possibly due to postoperative secondary stenosis. At referral in our center the patient had high creatinine level and bilateral ureterohydronephrosis. The site of the stenosis in this patient was a challenging subject which required a further imaging study with magnetic resonance urography to exactly define the site of obstruction (Figure 4 A and B). The first procedure with a double J stent, was performed for the resolution of obstructive uropathy and creatinine level, was successful, and the postoperative ultrasonography revealed the absence of upper tract dilation and a notable decrease in creatinine level. After mechanical resolution of the obstructive uropathy with a double J stent, the factors which had contributed to worsening the symptoms would be eliminated. Therefore, Botox was injected to decrease the detrusor pressure and improve the detrusor sphincter dyssynergia. The presence of CRE with persistent ureterohydronephrosis due to bilateral VUR and spastic neurogenic bladder made this a complex case which was challenging to manage.

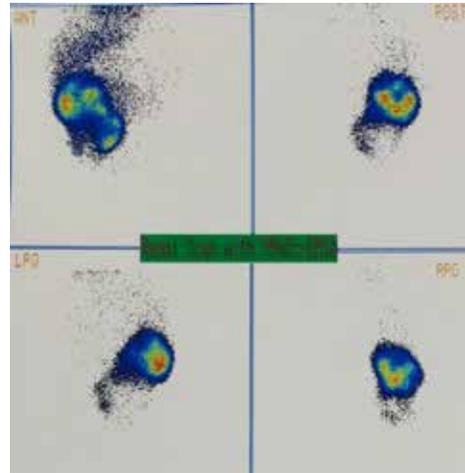
### Conclusions

We present a case of inferior CRE with associated imperforate anus, hypospadias with meatal stenosis, bilateral VUR, and neurogenic bladder leading to a challenging case with refractory ureterohydronephrosis.

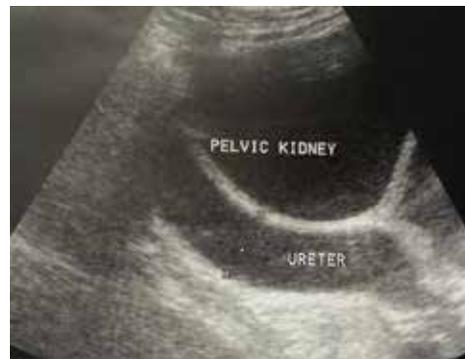
### Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this article.

### Figures and Legends to the figures



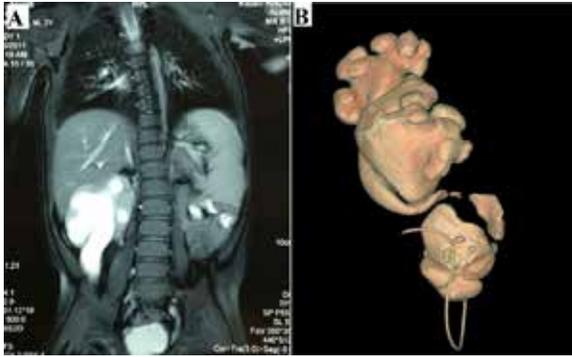
**Figure 1: DMSA scan showed the radiotracer uptake in the right kidney with downward extension and absence of uptake in the left side that is compatible with inferior type CRE.**



**Figure 2: The presence of massive ureterohydronephrosis was notable with ultrasonography.**



**Figure 3: Postoperative urethrography showed the passage of double J stent through the left orifice crossing the mid-line to reach the left kidney overlying in the right side.**



**Figure 4:** Magnetic resonance urethrography (MRU) showed left to right crossed ectopia kidney with inferior type. The presence of bilateral ureterohydronephrosis with the right ureter attached to the left side was detected (A, B).

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