# Crossed Fused Renal Ectopia and Orthotropic Kidney with Pelviureteric Junction Obstruction: Case Report

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## *Abstract*

Crossed fused renal ectopia (CFRE) is an extremely rare congenital malformation in which both kidneys are fused and located on the same side of the body. It is estimated to occur in 1 per 1000 live births. Most cases of CFRE are asymptomatic and are discovered incidentally, albeit can be rarely symptomatic. CFRE has been associated with Pelviureteric junction obstruction (PUJO) in 32-33% of reported cases. We report on a case of a 16-month-old male who was referred to our centre after an incidental finding of a single left kidney with presumed obstructing parapelvic cyst. Despite extensive evaluation and conservative measures, definitive diagnosis of CFRE and PUJO of the orthotopic kidney was made through surgical exploration followed by an open pyeloplasty of the PUJO. Upon follow up, the patient achieved full recovery with no further sequelae. This case report highlights the challenges we encountered in reaching a definitive diagnosis of CFRE with PUJO of the Orthotopic Kidney due to the limitations of imaging modalities and interpretation, and presence of overlapping clinical mimickers. Furthermore, we aim to raise awareness and review the current literature on this rare entity.

**Keywords:** Crossed Fused Renal Ectopia, Congenital anomalies of kidney, Pelviureteric Junction obstruction, Case report

## Introduction

CFRE is a congenital anomaly characterized by the displacement of a kidney across the midline and fusion with the contralateral kidney. It is the second most common renal fusion anomaly, after horseshoe kidney. While its exact incidence is unknown, it is estimated to occur in 1 per 1000 live births, with a male to female ratio of 3:2.[1] PUJO is defined as the obstruction of urine flow from the renal Pelvis to the proximal ureter. We report on a case of CFRE with PUJO of the orthotropic kidney and underscore the diagnostic challenges faced. We also aim to discuss the relevant literature on this rare condition. To the best of our knowledge, no reports of CFRE have been documented in Oman, especially when associated with PUJO of the orthotropic kidney.

## Case Report

A 16-month-old male toddler, with background of previous bilateral inguinal herniotomy at 9 months, was referred to our centre, after an incidental finding on an Ultrasound (US) abdomen of a non-visualized right kidney and left kidney with a presumed obstructing large parapelvic cyst and

calyceal dilatation. The US abdomen was part of his evaluation of chronic constipation, irritability and recurrent fever. The patient had no perinatal investigation for reference.

On initial assessment, the patient's physical assessment and laboratory work up were unremarkable. A repeat renal ultrasound (US) at our institution confirmed the findings of the initial US abdomen. A MAG-3 diuretic renogram showed delayed uptake and excretion from an upper moiety but normal draining lower moiety from the left kidney, while no radiotracer activity was detected on the right fossa, consistent with absent right kidney. This raised our suspicion for either a duplex left kidney with an obstructed upper moiety or a left kidney with a large parapelvic cyst causing pressure effect.

We proceeded with insertion of a percutaneous nephrostomy (PCN) to drain the presumed parapelvic cyst and decompress the obstructed single left kidney. After PCN insertion, the patient had notable output from both the nephrostomy and urethra. A repeat US abdomen demonstrated that the presumed cyst had collapsed yet the left kidney had persistent calyceal dilatation. A nephrostogram showed the contrast filling the presumed cyst without antegrade flow through the ureter.

To assess for possible upper urinary tract obstruction, we proceeded with a cystourethroscopy and Retrograde pyelography (RGP). During the procedure, no left ureteric orifice could be identified but a single right ureteric orifice was seen and cannulated for (RGP). The RGP demonstrated a crossed ureter- the "presumed left kidney" was a crossed ectopic right kidney (**Figure 1**). A Double-J stent was inserted into the crossed ureter, to encourage antegrade urine flow.

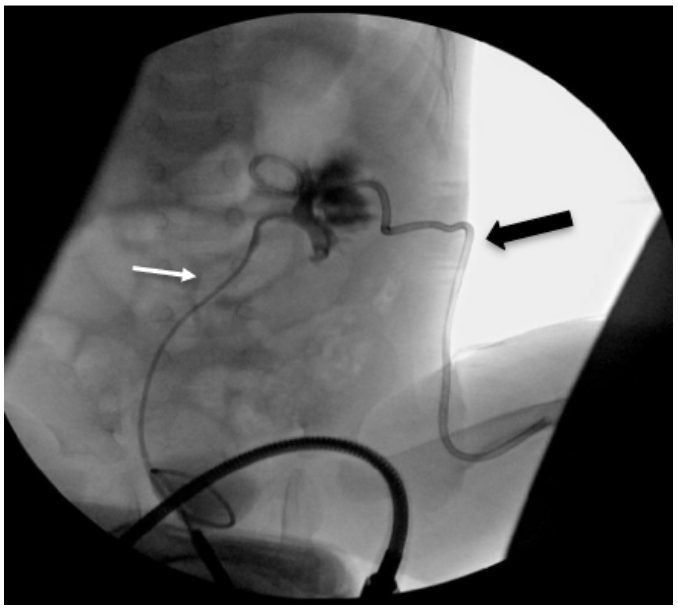

**Figure 1**: Retrograde pyelogram following contrast injection through the right ureteric orifice, demonstrating the right ureter crossing the midline (white small arrow) to join the left kidney. A nephrostomy tube is visible on the left side (black thick arrow).

Postoperatively, the patient continued to have significant output from the nephrostomy. Furthermore, attempts to encourage antegrade urine flow by clamping the nephrostomy resulted in recurrent urosepsis. Other laboratory parameter including renal profile were within the normal range. Serial renal US scans demonstrated re-accumulation of fluid in the parapelvic cyst, and persistent renal calyceal dilatation.

Due to the diagnostic uncertainty, a Magnetic resonance Imaging (MRI) abdomen with IV contrast was employed. It demonstrated an inferiorly crossed right kidney "presumed" to be fused with a normal left kidney forming a single mass (**Figure 2**). Additionally, delayed contrast excretion was noted from the right kidney, suggesting a possible PUJO.

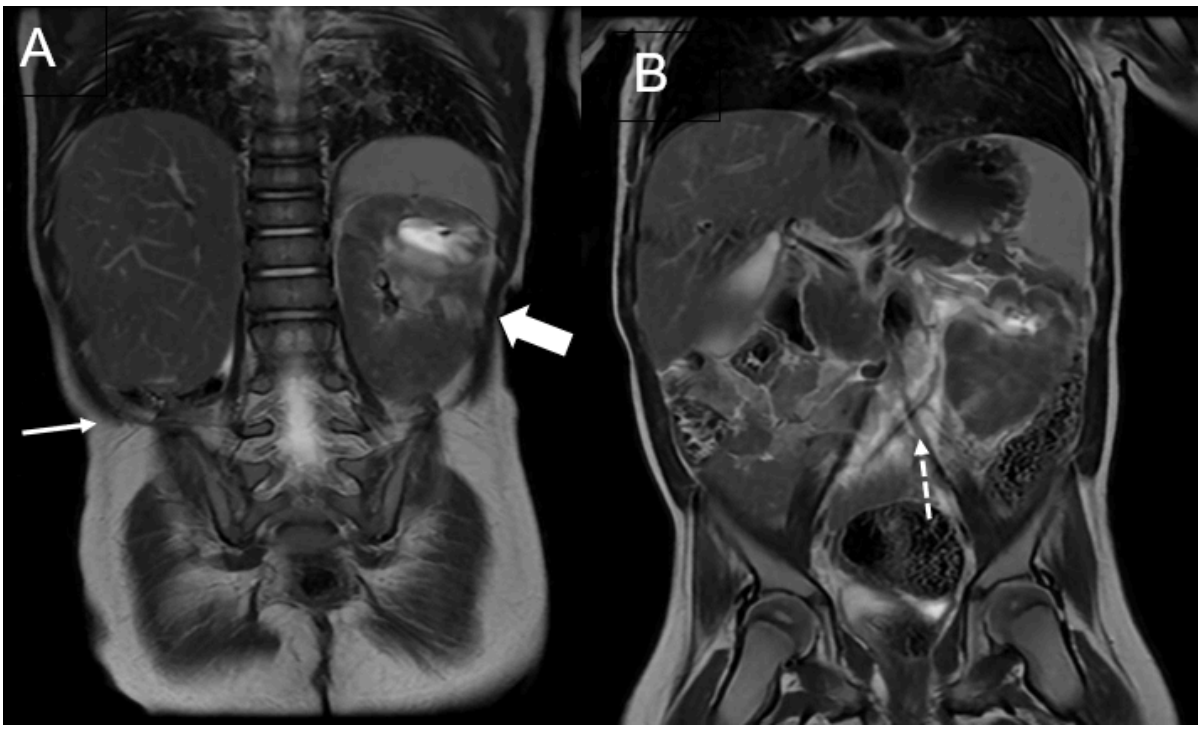


**Figure 2:** Coronal T2-weighted MRI image. **(A)** showing an empty right renal fossa (small white arrow). The left kidney is fused with the hydronephrotic ectopic right kidney, forming a single fused kidney (Thick white arrow). **(B)** shows the right ureter crossing the midline anterior (small dotted white arrow). to the left external iliac artery.

The decision was made to take the patient for exploration of the left renal fossa. Intraoperatively, we found a horizontally lying orthotopic left kidney, displaced superiorly and fused to the parenchyma of crossed ectopic right kidney. A PUJO was identified, it was a 2 cm narrow and fibrous segment; an open pyeloplasty was undertaken over a double-J stent to repair the defect.

Postoperatively, the patient recovered with no additional complications. Laboratory work up remained unremarkable. The PCN was removed on post operative day 7. The patient was readmitted after 4 months for removal of bilateral double-J stents (**Figure 3**). Histopathology of the excised ureteric segment was consistent with obstructive aetiology with features of eosinophilic ureteritis.

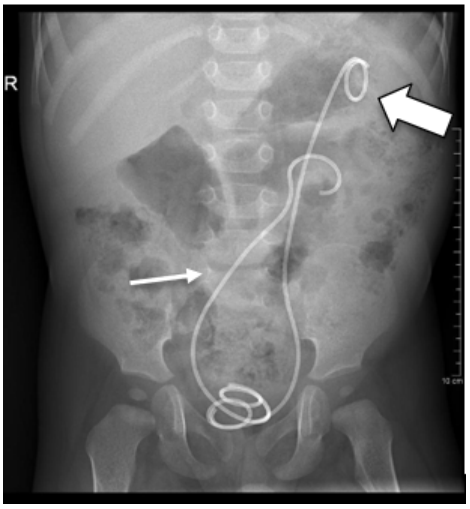

**Figure 3:** An abdominal X-ray demonstrating the right double J stent traversing the midline and extending toward the anatomical location of the left kidney (white small arrow). The left Double J stent can be seen extending to the location of the superiorly displaced orthotropic left kidney (white thick arrow).

## Discussion

As per the Mc Donald and Mc Clellan classification six iterations of CFRE exist (Table 1).[2] Furthermore, left-to-right ectopia is more common than right-to-left ectopia at a ratio of 3:1.[1] In our case, the patient had a variation of Type A ectopia, as the ectopic kidney was fused below a horizontally "displaced" orthotropic kidney, with a right-to-left migration.

**Table 1:** Mc Donald and Mc Clellan classification of CFRE.

| Category | Type of CFRE | Description |
|---|---|---|
| Type A | Inferior crossed fused ectopia | The ectopic kidney is fused below the orthotropic kidney |
| Type B | Sigmoid or S shaped Kidney | The ectopic kidney fuses unilaterally with the orthotropic kidney (hilum faces medially), with its hilum facing laterally forming an S-Shape. |
| Type C | Lump or cake shaped Kidney | Fusion of the two kidneys over a broad margin with the ureter of the ectopic kidney crossing the midline |
| Type D | Tandem or L-Shaped Kidney | The ectopic kidney is fused horizontally to the inferior pole of the orthotropic kidney forming an L-shape |
| Type E | Unilateral Disc Shaped Kidney | Extensive fusion of two kidneys forming a disc-shaped mass. |
| Type F | Superior crossed fussed ectopia | The ectopic kidney is fused above the orthotropic kidney |

CFRE has been associated with PUJO in about 33-52% of cases, in the crossed kidney.[3] However, PUJO in the orthotropic kidney, such as in our case, is extremely rare with only a few documented cases.[3-5] Complications that can be associated with CFRE include ectopic ureteral orifice, hydronephrosis, Vesicoureteral reflux, Cystitis, pyelonephritis, Urolithiasis, tumours and renal dysplasia.[4,6,7]

Most cases of CFRE are asymptomatic and are detected incidentally. Few cases of symptomatic CFRE may present non-specifically with pain, dysuria, frequency, haematuria or palpable mass.[4] In our case, our patient was being investigated for chronic constipation, when the CFRE (presumed as left kidney with cyst at the time) was incidentally detected. In our case, various imaging modalities had been used, before a diagnosis was reached. Our initial presumption was that the patient had a left renal parapelvic cyst and an absent right kidney based on US findings. An RGP confirmed crossed renal ectopia (**Figure 1**), however, it was an MRI that showed features of a Crossed "fused" renal ectopia (**Figure 2**). Furthermore, an US abdomen was useful in demonstrating dilated calyces in the left Kidney, which raised our suspicion for obstruction to urine flow. Similarly, both the MRI and MAG scan demonstrated delayed excretion of contrast consistent with obstruction, which was suspected to be due to PUJO (and later confirmed in surgery).

Primary management of CFRE is not indicated, instead management is directed towards the patients' associated complications.[4-6] In our case, surgical management was directed towards the patients PUJO via open pyeloplasty, after confirming the diagnosis intraoperatively. Laparoscopic pyeloplasty for repair of PUJO in patients with CFRE has been documented.[7]

## Conclusion

CFRE is a rare renal anomaly, especially when associated with PUJO in the orthotropic kidney. Most patients are asymptomatic, with cases being found incidentally or when presenting with complications associated with CFRE. Management is directed towards management of the sequelae or associated anomalies of CFRE.

## Consent

The patient's guardian was contacted via telephone to obtain verbal consent.


## Acknowledgement

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