

8-14-2014

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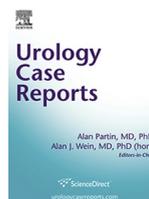
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### Recommended Citation

Peters, C.A., Pathak, N. (2014). Double Trouble: A rare case of bilateral upper pole ureteropelvic junction obstruction. *Urology Case Reports*, 2(5), 156-158.

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## Pediatrics

## Double Trouble: A Rare Case of Bilateral Upper Pole Ureteropelvic Junction Obstruction<sup>☆</sup>



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### ARTICLE INFO

#### Article history:

Received 13 July 2014

Accepted 16 July 2014

#### Keywords:

Ureteropelvic junction obstruction

Hydronephrosis

Robotic surgery

Ureteral duplication

### ABSTRACT

A 16-year-old girl presented with bilateral back pain caused by bilateral upper pole ureteropelvic junction obstructions; an extremely rare phenomenon. Bilateral robotically assisted upper pole pyeloplasties were performed at the same setting with an excellent clinical response. Although rare, upper pole ureteropelvic junction obstruction is a defined entity that urologists should be aware of.

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### Introduction

Upper pole ureteropelvic junction (UPJ) obstruction is an extremely uncommon entity. In duplicated renal systems, it is the lower pole that is typically obstructed at the UPJ. Bilateral UPJ obstruction has been commonly reported, while bilateral upper pole UPJ has not been specifically reported in the literature. A case is presented with a discussion as to the therapeutic options and clinical management.

### Case presentation

A 16-year-old Caucasian girl presented with intermittent bilateral back pain aggravated by activity. She had no clinically significant medical or surgical history. A bone scan demonstrated delayed excretion and retention of radioisotope in the upper poles of both kidneys suggesting renal obstruction (Fig. 1A,B). Ultrasonography revealed bilateral symmetric upper pole hydronephrosis (Fig. 2). Magnetic resonance urogram (Fig. 3) and mercaptoacetyltriglycine diuretic renogram (Fig. 4) revealed bilateral complete duplication and bilateral upper pole ureteropelvic junction (UPJ) obstructions. The lower poles appeared normal.

Surgical repair was recommended, and the patient underwent bilateral robotically assisted upper pole pyeloplasties using a Y-to-V

advancement repair with upper pole double-J ureteral stent placement. Postoperatively, the right ureteral stent became obstructed, requiring replacement on postoperative day 3 because of urinary ascites and pain. She did well and was discharged on postoperative day 8 on prophylactic antibiotics. The stents were removed 6 weeks postoperatively.

The patient showed complete resolution of her symptoms despite vigorous activity. She suffered 2 minor episodes of cystitis, which resolved with treatment. Follow-up imaging showed persisting hydronephrosis, which appeared improved with more parenchyma visible between the calyces (Fig. 5). The family has deferred obtaining subsequent mercaptoacetyltriglycine scan because of her clinical improvement. At the most recent follow-up 3 years postoperatively, she is attending college and is asymptomatic.

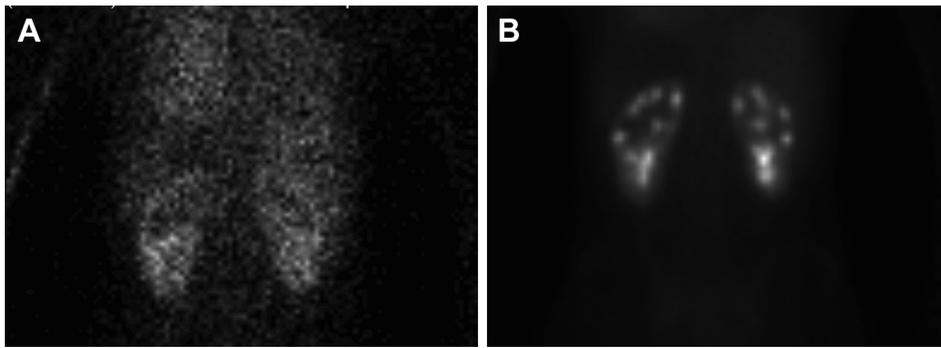
### Discussion

Unilateral upper pole UPJ obstruction is extremely rare<sup>1-7</sup>; bilateral upper pole UPJ obstruction has not been reported to date. Common presentation is with flank pain,<sup>2,8</sup> as well as infection, and through prenatal detection of hydronephrosis.<sup>4</sup> Vascular occlusion is considered a common cause, although the specific details are not well defined in the literature.<sup>1,2</sup> This may have some similarity to the so-called Fraley syndrome of vascular upper infundibular obstruction.<sup>9</sup> This patient's diagnosis was delayed because of confusion with musculoskeletal pain in the absence of lateralizing symptoms. Modern imaging can adequately define the anatomy, but optimal treatment is not well defined. Bilateral upper pole

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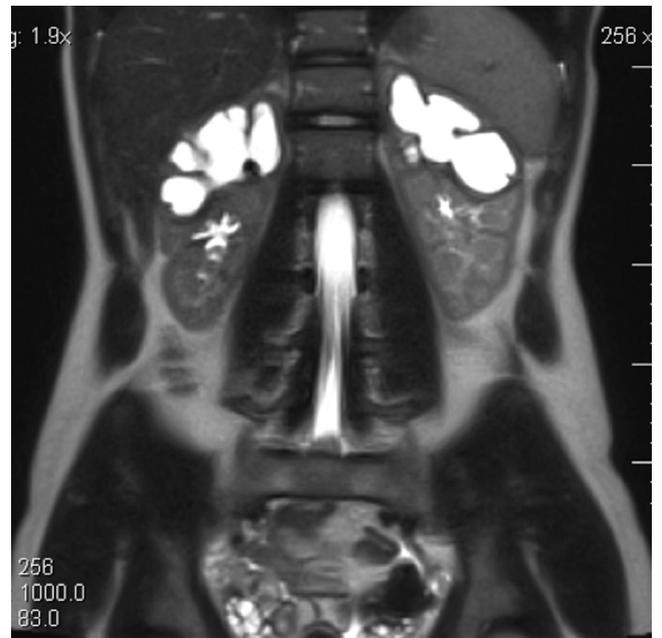
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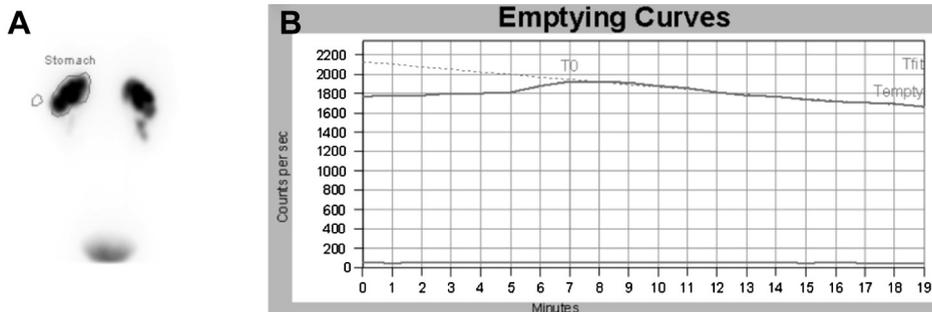
**Figure 1.** Bone scans showing relative photopenia of the upper poles indicative of hydronephrosis (A) and subsequent retention of tracer in the upper pole calyces (B).



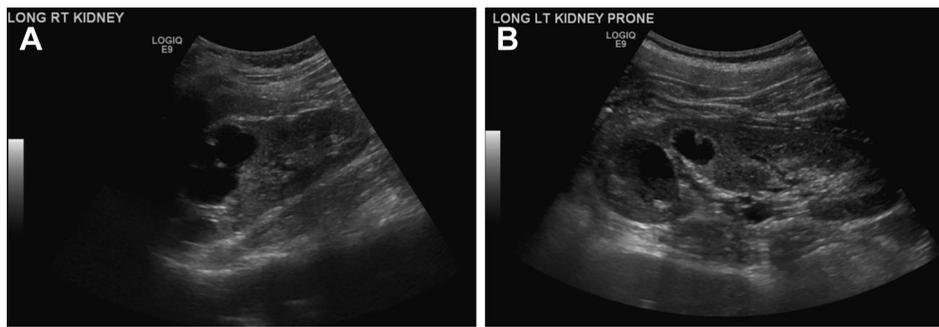
**Figure 2.** Ultrasonogram showing severe upper pole hydronephrosis of the right kidney. The left kidney was of similar appearance.



**Figure 3.** Magnetic resonance urogram of bilateral upper pole ureteropelvic junction obstructions.



**Figure 4.** MAG-3 diuretic renogram demonstrating markedly delayed washout of tracer in both upper pole moieties of the kidneys (A) with an essentially flat-line curve (B).



**Figure 5.** Renal ultrasonogram at 3 years postoperatively, showing reduced hydronephrosis and more pronounced renal parenchyma in the upper poles of the right (A) and left (B) kidneys.

partial nephrectomies could be a viable option. However, renal preservation seemed to be a worthwhile goal. The renal pelvises were not markedly dilated making an upper-to-lower pyelopyelostomy less likely to be feasible. Bilateral robotically assisted pyeloplasties proved to be a successful intervention in this patient. Bilateral renal robotic procedures at the same setting can be accomplished with 4 ports, including the umbilical camera port, a midline subxyphoid port, and 2 midclavicular lower quadrant ports.<sup>10</sup> The use of the Y-to-V flap approach was determined by the intrarenal location of the UPJ segment, which made access challenging. Although her postoperative stay was prolonged because of an obstructed stent, her overall recovery was rapid and permitted a return to full activity with satisfactory long-term follow-up.

### Conclusion

A unique case of bilateral upper pole UPJ obstruction is presented to illustrate the imaging appearance and discuss various management options. Bilateral simultaneous robotically assisted upper pole pyeloplasties using a Y to V advancement technique has been clinically successful.

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