# *Robotic treatment of ureteropelvic junction obstruction in Eagle-Barrett Syndrome*

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FABER LS, RILEY JM. Robotic treatment of ureteropelvic junction obstruction in Eagle-Barrett Syndrome. *Can J Urol* 2022;29(1):11116-11118.

*Eagle-Barrett Syndrome (EBS) is a rare congenital condition characterized by the triad of absent or defective abdominal wall muscles, urinary tract abnormalities, and bilateral cryptorchidism. Ureteropelvic junction obstruction (UPJO) is seldom reported in these patients, despite it being a common cause of childhood obstructive uropathy. We present the case of a patient with EBS who was subsequently identified as having symptomatic UPJO that was successfully treated with robotic pyeloplasty.*

**Key Words:** ureteropelvic junction obstruction, Eagle-Barrett Syndrome

## Introduction

Eagle-Barrett Syndrome (EBS) is a rare congenital condition, seen more commonly in males, characterized by the triad of absent or defective abdominal wall muscles, urinary tract abnormalities, and bilateral cryptorchidism.[1] Commonly reported urinary tract abnormalities include varying degrees of renal dysplasia, urethral dilation, megacystis, hydroureter, and vesicoureteral reflux.[2-4] Ureteropelvic junction obstruction (UPJO), a common cause of obstructive nephropathy in children, is seldom reported.[3-5] We present a patient with EBS presenting with symptomatic UPJO that was successfully repaired with robotic pyeloplasty. To the best of our knowledge, this is the first reported case of symptomatic UPJO in a patient with EBS.

Accepted for publication February 2022

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## Case description

A 21-year-old male with a history of EBS, bilateral cryptorchidism status post orchiopexy, bilateral hydronephrosis, recurrent urinary tract infections, and neurogenic bladder status post vesicostomy with subsequent closure now requiring self-catheterizations presented to the emergency department with acute onset severe, sharp left flank pain accompanied by subjective fever, chills, nausea, and vomiting. In the emergency department he was afebrile at 37.4 degree Celsius, mildly tachycardic, and hypertensive. Physical exam was notable for left costovertebral angle tenderness. Complete blood count showed mild leukocytosis (12,000 WBC) and urinalysis was positive for leukocyte esterase and white blood cells. Urine culture had no bacterial growth. Creatinine was 1.43 mg/dL, elevated from the patient's usual baseline of 1.0 mg/dL. CT scan revealed moderate perinephric edema of the left kidney as well as severe left hydronephrosis and dilation of the renal pelvis consistent with ureteropelvic junction obstruction

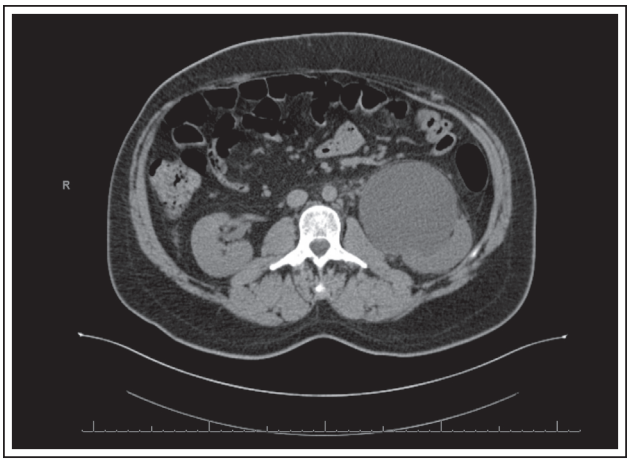


**Figure 1.** CT scan performed in the emergency department showing severe left hydronephrosis.

(UPJO), Figure 1. A nephrostomy tube was placed and drained appropriately. Outpatient follow up was arranged.

Renogram 2 weeks later showed a split function of 46.5% left and 53.5% right with a significant UPJO, Figure 2. Definitive management by robotic pyeloplasty was arranged. Intraoperative retrograde pyelogram confirmed a low insertion site of the left ureter on a dilated pelvis with a narrowed, adynamic appearing segment consistent with UPJO. No evidence of a crossing vessel was found on dissection. The adynamic segment of the ureter was removed and the remaining healthy appearing ureter was re-anastomosed. The patient recovered well from surgery. On follow up greater than a year later, the patient had less frequent UTIs, was no longer on prophylactic antibiotics, and did not have recurrence of UPJO, Figure 3.

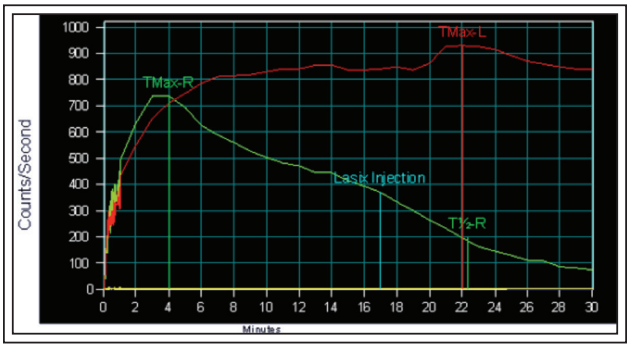


**Figure 2.** Renogram performed during preoperative evaluation showing evidence of a UPJO.

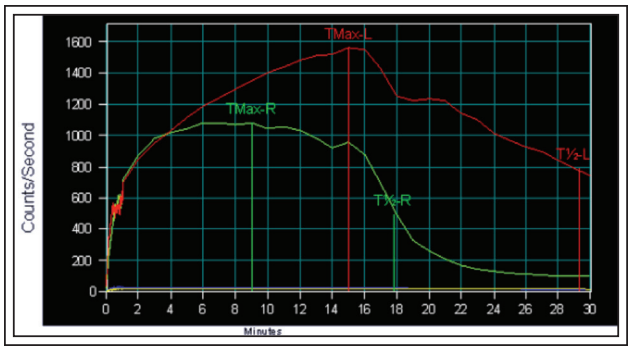


**Figure 3.** Renogram performed during postoperative evaluation showing improvement in drainage.

## Discussion

UPJO is the most common cause of congenital obstructive nephropathy, with an estimated incidence of 1 in 1,500 infants.[6,7] Congenital causes of UPJO include an aperistaltic segment, ureteral stricture, ureteric valve scarring, high insertion, kinking, or a crossing vessel.[6] UPJO can also be an acquired pathology, most commonly caused by instrumentation from prior surgeries, renal calculi, or mass effect from a neoplasm.[7]

Although most cases of UPJO are diagnosed perinatally with ultrasonography, congenital UPJO obstruction can present symptomatically at later ages.[8] Classically, UPJO presents with Dietl's Crisis, characterized by upper abdominal crampy pain or renal colic, nausea, and vomiting.[9] These crises are often induced by increased fluid intake. Patients may also have complaints of chronic back pain, recurrent infection including pyelonephritis, kidney stones due to urinary stasis, or hematuria.[10] If left untreated, the outflow obstruction causes urine to back up into the renal pelvis and calyces, causing hydronephrosis. This permanently damages nephrons and can result in chronic kidney disease, which can even eventually progress to renal failure if left untreated.[7]

Although hydroureter is common in patients with EBS, it is commonly attributed to more distal dysfunction, such as at the level of the bladder, vesicoureteral valves, or urethra.[2-5] UPJO is rarely reported due to its rarity in comparison with other urologic abnormalities seen in EBS, and when reported it is mentioned briefly with attention focused on the more common pathologies such as vesicoureteral reflux or urethral dilation.[3-5] Berdon et al examined 38 patients who were known to have EBS and found no radiographic evidence of upper tract obstruction.[4] Of

their patients who underwent intravenous pyelography, none showed evidence of UPJO.[4] A few years later, Moerman et al reported all of seven patients with EBS had widely patent ureters without obstruction.[5] Recently, Kirsch et al retrospectively analyzed 13 magnetic resonance urograms in patients with EBS, and identified UPJO in one patient with a solitary kidney and no hydroureter.[11] Our patient presented with symptoms of flank pain, nausea, and vomiting with CT evidence of worsening hydronephrosis likely secondary to UPJO. To our knowledge, our patient had no previous instrumentation of the ureter in the past that could have acted as a risk factor for development of UPJO. In addition, no crossing vessel was identified on dissection. Rather, an adynamic segment of ureter consistent with congenital UPJO was found and excised. On postoperative evaluation, he was asymptomatic and recovering well.

## Conclusion

While the true incidence of UPJO in patients with EBS remains unknown, it has been seen in autopsy findings of patients with EBS. Although it is likely very rare, it should be recognized as a possible urologic variant in patients with EBS. Furthermore, robotic interventions may still be a viable approach to treatment, even in the presence of defective abdominal musculature. □

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