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Dietl's Crisis in the Young Adult

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Abstract | **Case Report**

Given its intermittent nature, non-specific symptomatology, and under-recognition, Dietl's crisis remains an important diagnostic consideration in patients—especially children or young adults—with episodic abdominal pain, hydronephrosis on imaging, and no alternate explanation. Documenting cases adds to the understanding of its varied presentations, imaging features and outcomes of intervention.

Keywords: Dietl's crisis, UPJ obstruction, Hydronephrosis, Abdominal pain, Pediatric, Intermittent.

BACKGROUND

Dietl's crisis is a clinical syndrome describing the intermittent and sudden upper abdominal pain that occurs when a patient's ureteropelvic junction (UPJ) is transiently obstructed. The syndrome was first described by Josef Dietl in 1864. It is rare and under-diagnosed. Classically, the presentation involves positional or diuresis-provoked renal pelvic distension. The intermittent nature of the syndrome often complicates evaluation and delays diagnosis.

Incidental data is generally lacking however several retrospective studies suggest that Dietl's crisis accounts for approximately 11-15% of pediatric patients with UPJ obstruction and episodic pain [1].

Pathophysiologically, the obstruction of fluid output from a unilateral kidney is caused by either intrinsic or extrinsic compression. This includes congenital narrowing or a ureter's high insertion in the pelvis due to the crossing of a blood vessel, respectively [2]. The diagnosis is challenging due to its intermittent nature that often mimics other conditions. It is most commonly seen in the pediatric population and episodes can be triggered by high fluid intake.

Symptoms typically include intermittent crampy flank or upper abdominal pain with the absence of any urinary stone disease or infection. In the pediatric population, the symptoms can mimic musculoskeletal or gastrointestinal causes. One study of eight children found that all had experienced pain for more than one year prior to diagnosis and only one expressed urologic complaints [3]. Cases have also been documented in the adult population. One case report presented a 53-year-old woman with two years of recurrent abdominal pain who underwent several GI and surgical evaluations before she was imaged during an acute attack. This revealed clear hydronephrosis due to a vessel compressing her UPJ [4].

Generally, symptoms only occur during the intermittent obstruction phase and spontaneously resolve once the drainage restores, therefore imaging done in between flare ups may appear normal and delay recognition. The risk of this is that prolonged blockages can lead to kidney function loss. Presenting to the Emergency Room when actively in pain for an ultrasound can significantly increase the likelihood of seeing the UPJ obstruction1.

Ultrasound is often the first-line modality to detecting the dilating renal pelvis/hydronephrosis. Advanced imaging is then advised to assess drainage and differential function. To do so, diuretic renography and magnetic resonance urography (MRU) can combine anatomical and functional assessment. In one case of a 7-year-old boy, MRU helped demonstrate delayed transit, time-to-peak enhancement, and worsening hydronephrosis during diuresis, thereby supporting the diagnosis [5]. Interpreting diuretic renography in the setting of intermittent blockage however can be a challenge. One study of 59 patients noted significant variability in renographic patterns6. Thus, the absence of classic renographic findings does not exclude the diagnosis if clinical suspicion is high.

Once recognized and confirmed, management often involves surgical correction of the obstruction commonly via pyeloplasty. This prevents recurrent pain episodes and preserves renal function. In the study of 8 pediatric patients, seven underwent pyeloplasty, one required nephrectomy and all reported resolution of their nausea and vomiting and abdominal pain. Other reports have also described complete symptomatic relief and improvement of renal function post surgery [6,7].

Literature reviews yield less than 20 case reports published in the pediatric population of Dietl's crisis. It is generally an underdiagnosed syndrome. Delays in diagnosis are common and therefore heightened awareness of Dietl's crisis is imperative due to the possibility of progressive renal parenchymal injury.

CASE PRESENTATION

A 19-year-old female with a past medical history of chronic constipation presented with 4 years of unexplained lower abdominal pain. The pain was localized to the right lower quadrant and was intermittent. At the onset, in her mid-teen years, the patient had irregular periods and constipation. Following an abdominal ultrasound and blood investigations, which were all normal, she was diagnosed with irritable bowel syndrome (IBS).

As she got older, the pain worsened and became sharper. Over the course of 9 months the pain migrated to the right upper quadrant and right flank. The patient expressed that there was a palpable lump in the area of her pain however examining providers were unable to appreciate the lump. The patient's symptoms developed whereby she would experience markedly reduced urine frequency to the point of passing urine only once daily. She specifically expressed developing the pain and feeling the mass following water consumption.

During this time, her symptoms were repeatedly attributed to constipation, IBS or biliary colic. Investigation including CBC, LFTs, RFTs and a celiac intolerance screen were all normal. She had a normal H pylori stool test, a normal fecal calprotectin and fecal Immunochemical Tests (FIT).

An updated abdominal ultrasound during this time now showed a grossly hydronephrotic right kidney with a renal pelvis measuring 4cm and no improvement post micturition. She subsequently underwent several renal scans.

A CT urogram confirmed an idiopathic right pelvi-ureteric junction obstruction (PUJO) with no excretion of contrast through the right ureter likely caused by malrotation of the right kidney. There was no evidence of a vessel crossing over the PUJ which is a common cause of obstruction.

A NM renogram of the right kidney demonstrated normal uptake and excretion however there was a hold up in the right PUJ with an upsloping curve. The split function was 48% on the right side, and 52% on the left side.

The patient was diagnosed with Dietl's Crisis and underwent a laparoscopic pyeloplasty and ureteric stent insertion with resolution of symptoms and regular screenings.

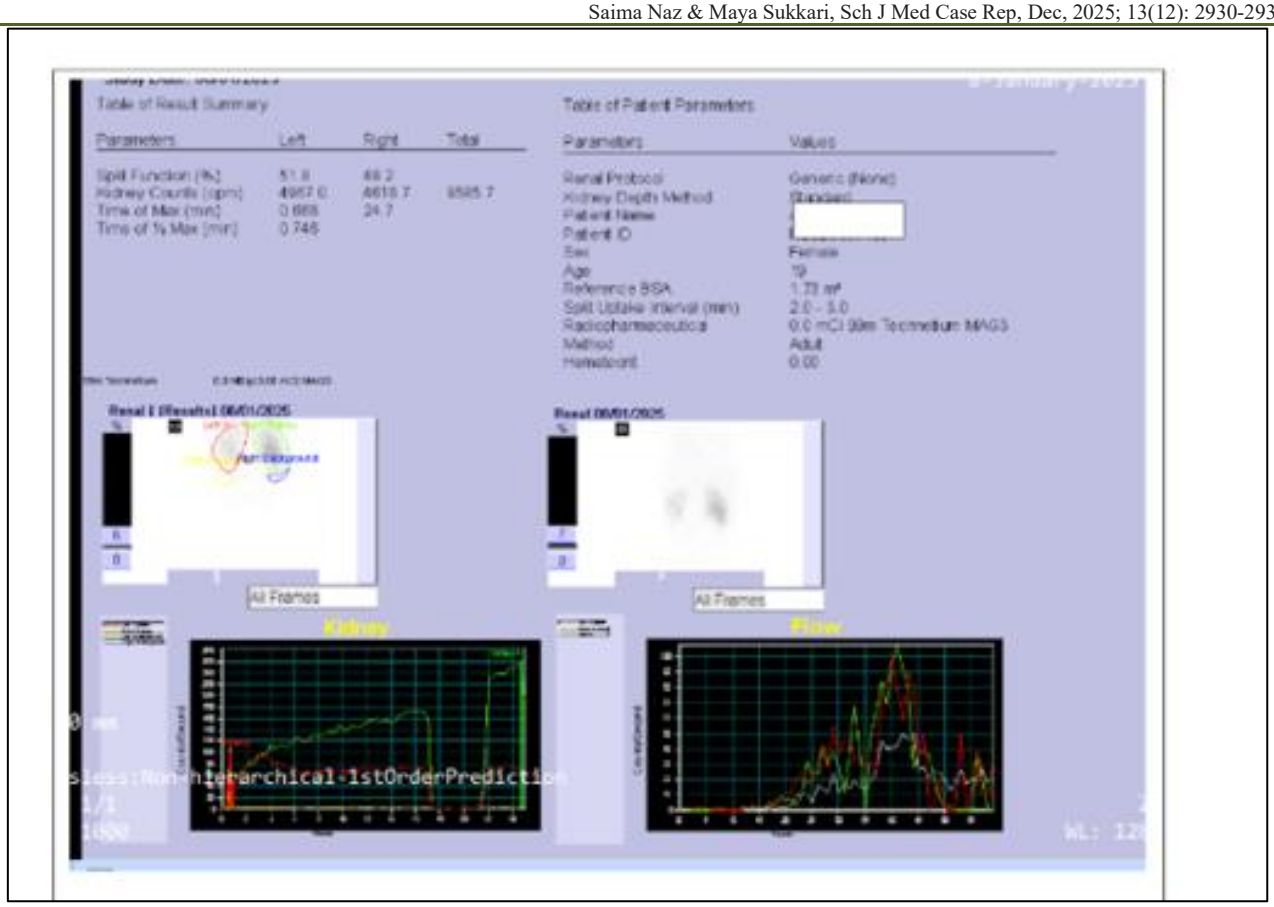
INVESTIGATIONS

A. Ultrasound Abdomen - December 2024

Finding: Previous normal abdominal ultrasound dated 13.12.20 noted. The right kidney is grossly hydronephrotic with the renal pelvis measuring 4cm. This does not improve post micturition. No cortical thinning noted. No hydroureter seen. The urinary bladder appears normal and completely empty post

B. CT Urogram

Finding: No radiopaque urinary tract calculus. Both kidneys have relatively good size and good cortical thickness. On the right, pelvicalyceal dilatation and a prominent extrarenal pelvis with mild malrotation and no excretion of the contrast through the right ureter on the acquired images. On the left is prompt excretion of the contrast through a slightly tortuous ureter but otherwise of normal caliber reaching the urinary bladder. There is no convincing evidence of a vessel crossing the PUJ; the PUJ type of obstruction is most likely idiopathic and possibly related to the minor malrotation of the renal pelvis. No abdominal, retroperitoneal or pelvic lymphadenopathy. Normal appearance of the liver, spleen, pancreas, gallbladder and adrenal glands. Normal caliber small and large bowel. No pelvic mass. Normal height imaged vertebral bodies.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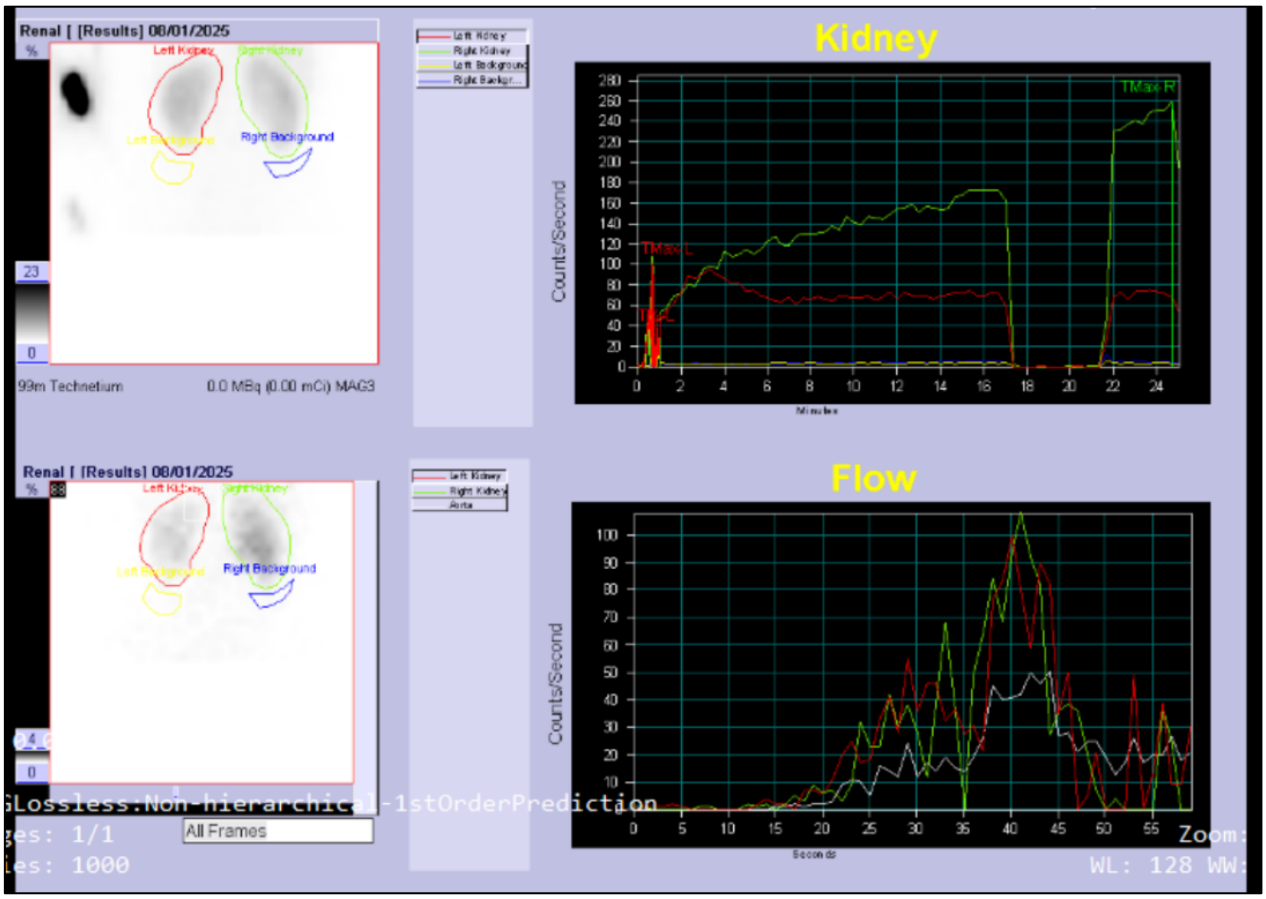
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CONCLUSION/RECOMMENDATIONS

Study confirms the presence of a dilated right pelvicalyceal system with a prominent extrarenal pelvis likely mild dated suggestive of idiopathic PUJ type of obstruction with no convincing evidence of a vessel crossing at the level of the PUJ.

C. NM Renogram - January 2025 (images below)
Findings:

The right kidney demonstrates normal uptake and excretion but hold up in the right PUJ with an upsloping curve. The left kidney demonstrates normal uptake and excretion but very slow drainage. Split function: Left: 52% Right:48%.

CONCLUSION

Right PUJO with good renal function. The left kidney also drains slowly but no complete obstruction.

Outcome and Follow-Up

Following the patient's surgical intervention, symptoms were relieved. Renal function remained stable and regular follow up scans recommended.

Learning Points/Take Home Messages

- Dietl's crisis is a diagnosis of any age, not only pediatric patients
- Pattern of the pain in the history
- The diagnosis must be strongly considered in this case of abdominal pain and cyclical vomiting syndrome.

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