Case Report

Dietl crisis: Presentation and imaging findings in a 7-year-old boy [☆],[☆☆]

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ABSTRACT

Intermittent ureteropelvic junction obstruction, or Dietl crisis, is a rare entity with sparse reports in published literature. Establishing the diagnosis is challenging given its intermittent nature. We report a case of Dietl crisis, focusing on ultrasound (US) and magnetic resonance urography (MRU) findings in a 7-year-old boy with recurrent episodes of colicky abdominal pain prompting multiple visits to the emergency department. Severe left hydronephrosis was visualized on US during one episode with complete resolution on follow-up US. MRU demonstrated severe left hydronephrosis with delayed calyceal transit time, time-to-peak enhancement, and excretion. There was no aberrant blood vessel. Surgical pyeloplasty provided complete symptomatic resolution. MRU can be a valuable tool in eliciting and dynamically confirming the diagnosis of Dietl crisis.

Introduction

Dietl crisis is a relatively rare entity, defined as intermittent ureteropelvic junction (UPJ) obstruction causing episodic abdominal pain, most often due to an aberrant accessory renal artery or vein [1]. It was first described in 1864 by Josef Dietl, who suggested initial conservative treatment or abdominal support with a belt or corset, as well as external application of pressure in the area of the affected kidney for prolonged pain [2]. Subsequently, from 1870 until the 1960s, surgical correction in the form of nephropexy was adopted [2]. More recently, laparoscopic pyeloplasty (with or without robotic assistance) has emerged as an adequate surgical intervention that causes complete resolution of symptoms in most cases [3]. While congenital UPJ obstruction is usually identified on routine prenatal ultrasound (US), some children present at an older age with episodes of abdominal or flank pain on the side of the obstruction, that may be associated with nausea and/or nonbilious vomiting, prompting multiple emergency department (ED) visits [4]. In view of complete symptom resolution with corrective surgery, accurate and prompt identification of patients with Dietl crisis with imaging tests becomes crucial. However, because of the vagueness of its presentation

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and the high frequency of nonspecific abdominal pain in school-aged children, routine abdominal US is not often performed and the diagnosis of Dietl's crisis is either delayed or missed [3]. The clinical differential diagnosis includes psychogenic pain, constipation, urinary tract infection, renal calculi, and appendicitis [1]. In the setting of high suspicion of Dietl crisis and when performed while symptomatic, US may identify obstructive hydronephrosis with dilatation of the renal pelvis and/or calyces, prompting further imaging such as computerized tomography, technetium-99m labeled mercaptoacetyltriglycerine (^{99m}Tc-MAG3) diuretic renography, or magnetic resonance urography (MRU) for anatomic and functional assessment of the pediatric urinary tract. We report a case of Dietl crisis in a 7-year-old boy while highlighting associated US and MRU findings.

Case Report

A previously healthy 7-year-old boy (weight = 21.3 kg; body surface area = 0.765 m^2), with a normal antenatal US, presented to the ED with colicky left flank pain that began acutely a few hours prior to presentation. The pain was described as severe, waking him from sleep, and was associated with nausea and vomiting without fever, diarrhea, or urinary symptoms. The patient had previously presented to the ED with similar symptoms on two different occasions. On physical exam, left flank tenderness was noted on palpation with ipsilateral lower abdominal fullness. Complete blood count and basic metabolic panel were normal (Cr = 0.48 mg/dL). US examination of the kidneys was performed and demonstrated severe left hydronephrosis (Fig. 1a) without ureteral dilatation and a normal right kidney. Dietl crisis was suspected, and the patient was admitted to hospital for overnight observation and pain management. A follow-up US was performed 2 weeks later as an outpatient, showing complete resolution of left hydronephrosis (Fig. 1b).

Pre- and postcontrast magnetic resonance imaging was performed based on the MRU protocol described by Dickerson et al. [5]. The patient was prehydrated with intravenous fluids (normal saline 20 cc/kg over 45 minutes). The MRU was performed under general anesthesia with a bladder catheter in place for the duration of the scan. Furosemide (1 mg/kg intravenous, max. 20 mg) was administered 15 minutes prior to gadolinium-based contrast (0.2 mL/kg intravenous, Dotarem, Guerbet). Sequential dynamic, coronal-oblique fat-saturation VIBE images were acquired before, during and after power injection of the intravenous contrast.

MRU demonstrated left hydronephrosis that gradually worsened in severity over the course of the scan (Fig. 2a and b) as the diuretic took effect. Arterial phase images showed no evidence of an aberrant blood vessel crossing the UPJ. Quantitative analysis was performed using Children's Hospital of Philadelphia Functional MRU analysis software (CHOP-fMRU, https://www.chop-fmru.com/) [6]. Renal parenchymal volume was found to be 77.9 mL on the right and 66.5 mL on the left. Differential renal function (DRF) was as follows: right-to-left ratio of 53.9%-46.1% using volumetric calculation (vDRF), 58.1% to 41.9% using Patlak calculation (pDRF), and 61.9%-38.1% using Volumetric-Patlak calculation (vpDRF).
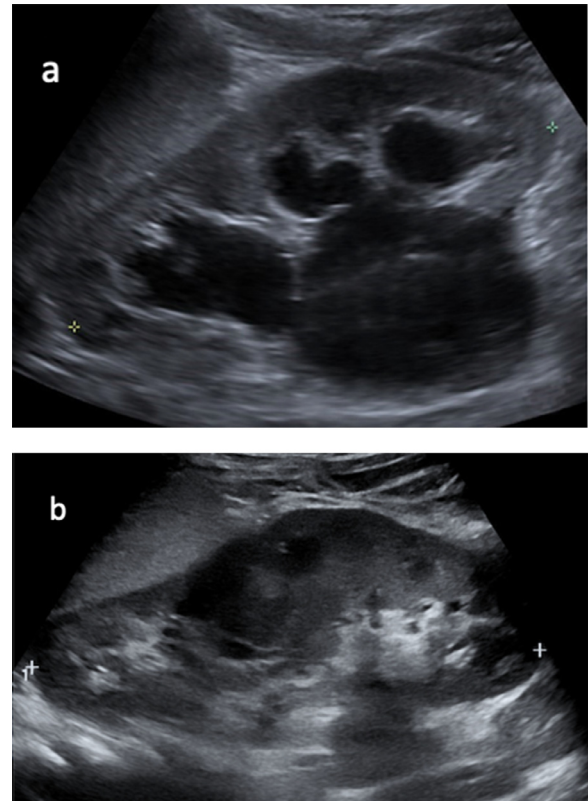

Fig. 1 – Sagittal ultrasound of the left kidney performed at the time of presentation in the emergency department (a). The kidney demonstrated marked dilatation of the renal pelvis and calyces and measured 10.3 cm in length. Repeat ultrasound at outpatient follow-up visit 2 weeks after presentation (b) demonstrated resolution of the previously seen hydronephrosis in the affected kidney which measured 10.0 cm in length.

The difference between vDRF and pDRF (4.2%) was borderline significant (≥4%), suggesting decompensated hydronephrosis. Calyceal transit time (CTT) was delayed on the left (4 minutes 37 seconds) as compared to the right (2 minutes 13 seconds). Time to peak (TTP) of parenchymal enhancement was delayed on the left (5 minutes 33 seconds) as compared to the right (3 minutes 31 seconds). Excreted contrast reached the right proximal ureter (renal transit time, RTT) at 3 minutes 8 seconds. No excreted contrast was visualized in the left ureter over 34 minutes of postcontrast imaging, indicating UPJ obstruction. The delayed time values in the left kidney were further suggestive of decompensated hydronephrosis.

Ultimately, a diagnosis of Dietl crisis was made and the patient underwent robotic-assisted laparoscopic left pyeloplasty with anterograde insertion of a double-J stent. Subsequently, clinical symptoms did not recur, and follow-up US at 3 months post-operative showed complete resolution of left hydronephrosis.

Discussion

Intermittent abdominal pain is a common complaint in the pediatric population, carrying a wide differential, including
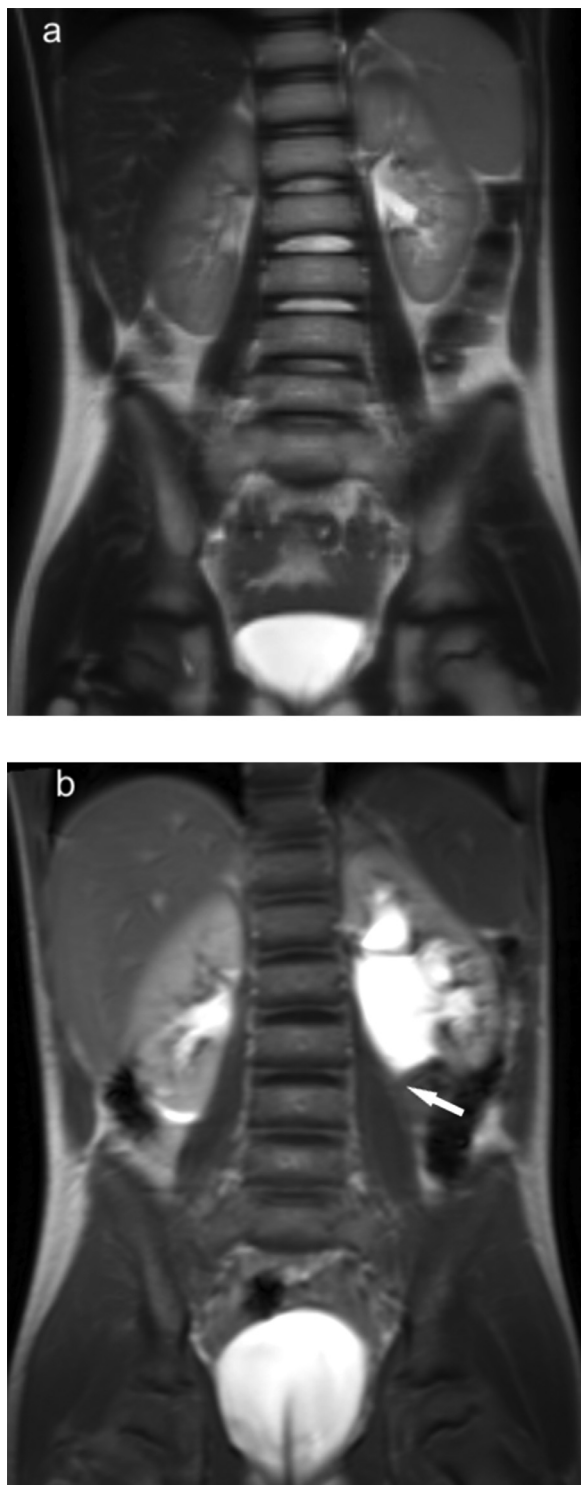
Fig. 2 – MRU performed under general anesthesia and with bladder catheterization. Furosemide was administered intravenously prior to contrast injection. Coronal T2 HASTE (a) and coronal T1 VIBE (b) images acquired 34 minutes after contrast administration demonstrate left hydronephrosis that gradually worsened in severity as the diuretic took effect. There is no dilatation of the ureter distal to the ureteropelvic junction (arrow).

Dietl crisis. US is the first-line modality in the uroradiology work up of pediatric hydronephrosis, allowing real-time assessment of the bladder, perivesicular region, ureteral and pelvicalyceal system, as well as the renal parenchymal and reproductive organs [7]. Doppler US can be used to assess renal perfusion and to detect urine jets at the ureterovesicular junction. However, US is limited in its ability to confirm the presence or absence of UPJ obstruction, and a functional modality is often needed.

Functional assessment for urinary obstruction is typically performed with ^{99m}Tc-MAG3 diuretic renography, which similar to MRU, includes intravenous pre-hydration and furosemide. The relatively low spatial resolution and exclusive use of planar (ie, nontomographic) images limit the utility of ^{99m}Tc-MAG3 diuretic renography in suspected intermittent UPJ obstruction. For example, gradual increase in the severity of hydronephrosis may not be apparent. Time-activity curves are used to quantitate differential renal function, time to peak activity, and urinary drainage. In UPJ obstruction, the affected kidney can demonstrate diminished differential renal function, and prolongation of both time-to-peak activity and urinary drainage half-time. In Dietl's crisis, however, the intermittent nature of symptoms can result in inconclusive or even normal diuretic renography results, commonly necessitating repeat diuretic renography. Renal parenchymal volume and function may be underestimated by planar scintigraphy due to attenuation of radiation by the dilated collecting system. Furthermore, the low spatial resolution of scintigraphy may not be adequate to detect radiotracer in a nondilated ureter.

MRU has emerged as an important advanced imaging modality, allowing both morphological and functional evaluation of the pediatric urinary tract with high spatial resolution and without exposure to ionizing radiation [5]. It is a valuable tool in assessing possible UPJ obstruction, such as in Dietl crisis. Precontrast T2-weighted sequences of urine (MR hydrography) provide detailed visualization of a dilated or nondilated urinary collecting system. Dynamic contrast-enhanced T1-weighted images provide additional structural evaluation of the urinary collecting system and allow for functional analysis including DRF [5]. The most common cause of UPJ obstruction is primary congenital narrowing, but extrinsic compression from an aberrant blood vessel is also common, appearing as flow voids on T2-weighted images [5]. Dynamic contrast-enhanced imaging can distinguish between arterial versus venous structures. The presence of an aberrant blood vessel may or may not alter the surgical approach of the pediatric urologist. In the case presented here, no extrinsic abnormality was visualized, and obstruction was most likely caused by an intrinsic anatomic abnormality at the UPJ that became accentuated with a full renal pelvis, for example stenosis, kinking, or a combination of the two.

Moreover, functional analysis of MRU can be performed using the freely available CHOP-fMRU software. It allows the generation of postcontrast time-intensity curves for the aorta (arterial input function) and for each kidney. In cases of decompensated hydronephrosis, the affected kidney shows delayed TTP, as well as slow washout of parenchymal signal intensity. In this case, TTP was delayed on the left and normal on the right (Fig. 3). Furthermore, the CHOP-fMRU software allows the generation of excretion curves showing the change
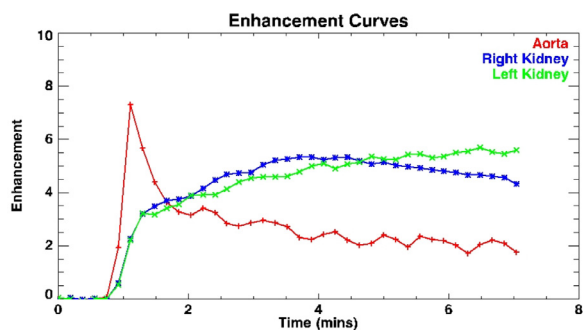
Fig. 3 – Plot demonstrating enhancement over time of the aorta (red), right kidney (blue), and left kidney (green). Time-to-peak (TTP) enhancement was delayed in the left kidney and normal in the right.
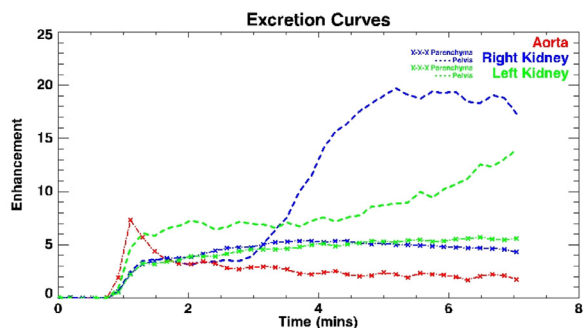

Fig. 4 – Enhancement over time in the right renal pelvis (blue dashed line), and left renal pelvis (green dashed line), superimposed on the plot shown in Fig. 3. There is delayed excretion into the left renal pelvis, while excretion on the right is normal.
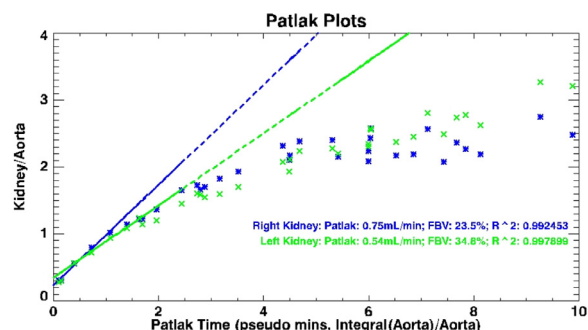

Fig. 5 – Patlak plot showing kidney/aortic intensities as a function of Patlak time, with the initial slope of each curve representing the GFR index or Patlak number (right kidney in blue; left kidney in green). The Patlak number is expressed in units of (mL/min)/mL.

in relative signal intensity in the renal pelvis over time [6]. In this case, there is delayed excretion into the left renal pelvis, while excretion on the right is normal (Fig. 4). These delayed values, in combination with the delayed CTT and RTT are suggestive of a *decompensated* hydronephrosis in which the renal parenchymal function is compromised.

Functional analysis allows the calculation of DRF using three different methods, based on: (1) volume of enhancing renal parenchyma (vDRF), (2) glomerular filtration rate (GFR) based on Patlak calculation (pDRF), and (3) combination of vDRF and pDRF (vpDRF) [7]. The pDRF method is based on a Patlak plot showing kidney/aortic intensities as a function of Patlak time (obtained through mathematical transformation of aortic signal intensity), with the slope of each curve representing the GFR index or Patlak number (Fig. 5) [6]. The estimated GFR (eGFR in mL/min) for a unilateral kidney is calculated by multiplying its Patlak number ([mL/min]/mL) by its renal parenchymal volume (mL). In *compensated* hydronephrosis, renal parenchymal function is typically preserved and pDRF is symmetric; as such, GFR and serum creatinine usually remain within normal levels. The difference between the vDRF and pDRF for each kidney is normally less than 4%. A

difference ≥4% in combination with other abnormal MRU parameters suggests *decompensated* hydronephrosis.

In our patient, MRU functional analysis demonstrated decompensated left hydronephrosis. The difference between the vDRF and pDRF (≥4%) as well as the delayed CTT, RTT, TTP, and excretion curve are suggestive of *decompensated* hydronephrosis. Moreover, serum creatinine was found to be normal (Cr = 0.48 mg/dL) while eGFR was 73.11 mL/min when calculated using the above formula, which borders on the normal cut-off of 75 mL/min suggested by Pottel et al. [8]. We hypothesize that MRU can potentially detect unilaterally decreased renal function that would otherwise not be suggested by measured serum Cr level and eGFR. As such, MRU can detect irreversible renal damage, namely tubulointerstitial fibrosis, caused by an intermittent pathologic state such as the episodic UPJ obstruction that occurs in Dietl's crisis.

For long-term follow-up, US is used postoperatively to reassess the urinary collecting system. MRU may be repeated when it is necessary to evaluate postsurgical anatomy or in the case of recurrent symptoms.

Conclusion

Establishing a diagnosis of Dietl crisis in the pediatric population can be challenging, as it mimics a wide differential of pathologies. US is typically the first-line imaging modality in the acute setting and depending on the timing of the scan, may or may not reveal hydronephrosis in the affected kidney. MRU is a powerful imaging tool that provides both detailed structural information and quantitative functional assessment of a dilated urinary collecting system, leading to greater confidence when considering the diagnosis of Dietl crisis. Confirming diagnosis is crucial as surgical pyeloplasty typically provides complete symptomatic resolution. In our patient, the real-time observation of worsening diuresis-induced hydronephrosis in combination with decompensated MRU parameters are unusual findings that may be unique to Dietl crisis and warrant future research.

Availability of data and material

The reported material is available.

Code availability

Not applicable.

Authors' contributions

Rita Maria Lahoud: manuscript writing and editing. William Esker: data analysis, figure preparation and manuscript editing. Shirley A. Thurston: data analysis, figure preparation and manuscript editing. Jack Elder: manuscript writing and editing. Ruth Lim: manuscript writing and editing, figure preparation.

Ethics approval

Not applicable (case report).

Consent to participate

Not applicable.

Consent for publication

Obtained.

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