Clinical Imaging 55 (2019) 144–147

Contents lists available at ScienceDirect

Clinical Imaging

journal homepage: www.elsevier.com/locate/clinimag

Pediatric Radiology

The nutcracker syndrome: The usefulness of different MRI sequences for diagnosis and follow-up

Ali Er[a,*], Nail Uzunlulu[b], Tevfik Guzelbey[b], Sevgi Yavuz[c], Aysel Kiyak[c], Arda Kayhan[b]

[a] Izmir Tepecik Training and Research Hospital, Department of Radiology, Izmir, Turkey
[b] Istanbul Kanuni Sultan Süleyman Training and Research Hospital, Department of Radiology, Istanbul, Turkey
[c] Istanbul Kanuni Sultan Süleyman Training and Research Hospital, Division of Pediatric Nephrology, Istanbul, Turkey

ARTICLE INFO

Keywords:
Nutcracker syndrome
Renal vein entrapment
Magnetic resonance imaging

ABSTRACT

Objective: Nutcracker Syndrome (NCS) is the extrinsic compression of the left renal vein by neighboring arterial, ligamentous, muscular, or osseous structures. Diagnosis is made by Doppler ultrasonography (US), multidetector computerized tomography (MDCT), magnetic resonance imaging (MRI), phlebography. The aim of the current study is to assess the value of MRI and compare the efficiency of different sequences in diagnosis and follow up of children with NCS.
Material and methods: A total of 40 children (female/male ratio 3:1) with NCS were included in this prospective study. A standardized abdominal MRI protocol was used and T2-TRUFI (True Fast Imaging with Steady-State Free Precession), T2-HASTE (Half Fourier Acquisition with Single Shot Turbo Spin Echo), T1-VIBE (Volumetric Interpolated Breath Hold Examination), and out-of-phase (opposed-phase) T1 sequences were obtained. The sequences were compared according to anatomical depiction, measurability, and pulsation artifact.
Results: A four point-scale was used to assess subjective image quality and the results were listed as: 1 = poor, 2 = fair, 3 = good, and 4 = excellent. Both in total and for each individual criterion, the highest scores were obtained with T2-TRUFI (total mean 3.74 ± 0.45, anatomical depiction 3.9 ± 0.3, measurability 3.8 ± 0.4, aortic pulsation artifact 3.52 ± 0.55).
Conclusion: Although Doppler US is the gold standard technique in the diagnosis of NCS, MR imaging may be used as an additional modality, as it is superior to Doppler US in terms of anatomic depiction and a lower rate of imaging artifacts. Non-contrast MR imaging, particularly TRUFI sequence, may have an incremental value in the accurate diagnosis and follow-up of these patients.

1. Introduction

Nutcracker Syndrome (NCS) is the extrinsic venous compression of the left renal vein (LRV) by neighboring arterial, ligamentous, muscular, or osseous structures [1]. Compression of the LRV between the abdominal aorta and superior mesenteric artery (SMA) is known as anterior NCS, while compression of the LRV between the aorta and vertebral column is known as posterior NCS [2,3]. In the case of left-sided inferior vena cava (IVC), the right renal vein (RRV) is compressed and this is referred to as right NCS [4]. NCS may present with symptoms such as flank pain, hematuria, proteinuria, pelvic congestion, and left-sided varicocele [5,6]. NCS may be diagnosed using different radiological methods such as Doppler ultrasonography (US), multidetector computerized tomography (MDCT), magnetic resonance (MR) imaging, and phlebography [7]. The aim of the current study is to assess the value of MR imaging and compare the efficiency of different sequences in the diagnosis and follow up of children with NCS.

2. Material and methods

2.1. Patients

The study was approved by the institutional ethics committee. Written informed consent was obtained from at least one parent (or legal guardian) of each child. A total of 40 pediatric patients (9 male, 31 female) who previously had clinical and ultrasonographic diagnosis of NCS were included in this prospective study conducted between 2015 and 2017. The age distribution was between 9 and 17 years (mean: 15.6 ± 2.34 years). The patients with symptoms of pain, hematuria (microscopic/macroscopic), or proteinuria were admitted to our

* Corresponding author at: Tepecik Training and Research Hospital, Department of Radiology, 1140/1 Street No: 1, Yenisehir, Konak, Izmir, Turkey.
E-mail address: alier1717@yahoo.com (A. Er).

https://doi.org/10.1016/j.clinimag.2019.02.003
Received 22 May 2018; Received in revised form 31 January 2019; Accepted 5 February 2019

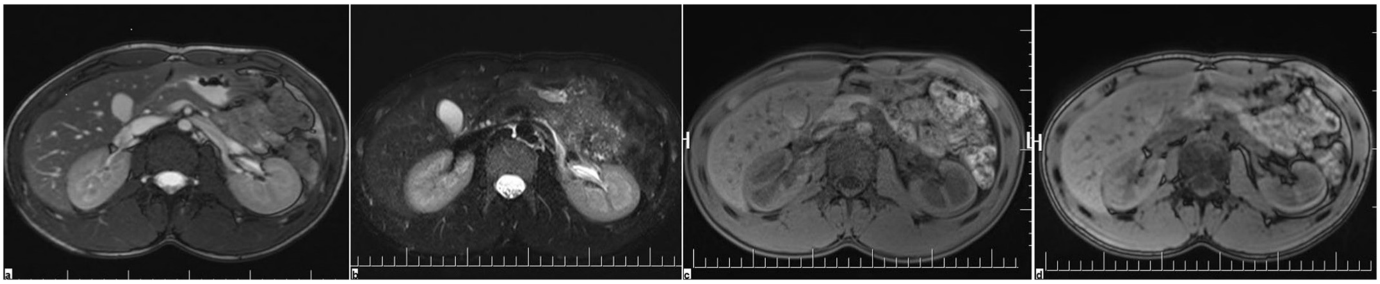

Fig. 1. Axial MRI images (a; T2-TRUFI, b; T2-HASTE, c; T1-VIBE, d; Out-of-phase T1 sequence): compressed left renal vein between aorta and superior mesenteric artery.

pediatric nephrology outpatient clinic for diagnosis of NCS. Children with intolerance and contraindication to MR imaging were excluded from the study. Children under 9 years of age were also excluded to avoid using sedation as an invasive procedure.

2.2. MR imaging protocol

The Siemens Magnetom Aera 1.5 T MR imaging device (Siemens Medical Systems, Erlangen, Germany) was used for MR imaging. The patients were in supine position. A standardized abdominal MR imaging protocol was used for all patients and the images were obtained with T2-TRUFI (True Fast Imaging with Steady-State Free Precession), T2-HASTE (Half-Fourier Acquisition with Single Shot Turbo Spin Echo), T1-VIBE (Volumetric Interpolated Breath Hold Examination), and out-of-phase (opposed-phase) T1 sequences (Fig. 1a–d). The imaging parameters used for each MR imaging sequence are presented in Table 1.

2.3. Image analysis

MR images were evaluated by two experienced radiologists who were blinded to the Doppler US (T.G, N.U). The axial images were transferred to a workstation (Magic-View, Siemens, Germany) for analysis. The scoring criteria included anatomical depiction, measurability, and pulsation artifacts. The total scores for each sequence were compared in terms of diagnostic performance. For anatomical depiction, the scoring was based on the ability of the sequences to assess the anatomic position of left renal vein. Measurability was assessed as the ability of the sequences to discriminate the neighboring structures and measure the diameter at the most narrow and wide segments. For pulsation artifacts, the degree of depiction of the aorta and left renal vein was considered.

A radiologist reviewed all axial images of each patient and measured the maximum AP diameters of the portions of the affected renal vein (Fig. 2a–b). A four-point scale was used to assess subjective image quality and the results were listed as: 1 = poor, 2 = fair, 3 = good, 4 = excellent.

Table 1
Magnetic resonance imaging (MRI) parameter.

Parameters	T2-TRUFI	T2-HASTE	T1-VIBE	Out–of–phase T1
FOV (abdomen)	40 × 40 cm	40 × 30 cm	40 × 30 cm	40 × 30
Matrix	243 × 256	200 × 320	200 × 320	200 × 320
Slice thickness/ spacing	3 mm	3 mm	3 mm	3 mm
TR	3,71 ms	4000 ms	4,36 ms	180 ms
TE	1,6 ms	110 ms	2,06 ms	2,24 ms
Flip angle (degrees)	55	160	10	70
Total duration	20 s	50 s	1 min 21 s	35 s

FOV = field of view, TR = time to repetition, TE = time to echo.

2.4. Statistical analysis

T2-TRUFI, T2-HASTE, T1-VIBE, and out-of-phase (opposed-phase) T1 sequences were assessed separately in terms of anatomical depiction, measurability, and aortic pulsation artifacts using the Mann-Whitney *U* test, and p-values were evaluated using the Bonferroni correction. Interobserver agreement was evaluated using the intraclass correlation coefficient (ICC). ICC values were classified as: poor reliability (values < 0.5), moderate reliability b (values between 0.5 and 0.75), good reliability (values between 0.75 and 0.9), and excellent reliability (values > 0.90). All statistical analyses were performed using SPSS for Windows, version 23.0 (SPSS Inc., Chicago, IL, USA).

3. Results

Of the total of 40 patients, 4 had retroaortic LRV (posterior NCS) (Fig. 3a–b). Situs inversus with left-sided IVC was detected in one patient and RRV was trapped between the abdominal aorta and SMA, resulting in right NCS (Fig. 4a–b). In terms of anatomical depiction and measurability, interobserver agreement was excellent on all sequences (ICC > 0.75). In terms of aortic pulsation artifacts, interobserver agreement was good (ICC = 0.73) on T1-VIBE sequence and excellent (ICC > 0.75) on all other sequences. The retroaortic course identified on the T2-TRUFI sequence was excellent (Fig. 2a–b). Both in total and for each individual criterion, the T2-TRUFI held the highest scores (total mean 3.74 ± 0.45, anatomical depiction 3.9 ± 0.3, measurability 3.8 ± 0.4, aortic pulsation artifact 3.52 ± 0.55). The T1-VIBE sequence obtained the lowest scores (total mean 2.05 ± 0.79, anatomical depiction 2.02 ± 0.69, measurability 1.5 ± 0.64, aortic pulsation artifact 2.65 ± 0.57). The mean scores for sequences according to the criteria are presented in Table 2.

In the paired comparison of sequences, T2-TRUFI showed significantly higher scores compared to other sequences in terms of anatomical depiction, measurability, and aortic pulsation artifact ($p < 0.05$). T2-HASTE sequence was found to be superior to T1-VIBE in terms of anatomical depiction ($p < 0.05$).

4. Discussion

The accurate prevalence of NCS is unknown, although few studies report an incidence of 0.3–3.7% for posterior NCS; with the development of diagnostic modalities, the actual prevalence of the disease may prove to be higher than estimated. The gender distribution of NCS is unknown, but is predicted to be more prevalent in females [9–11]. In our study, children with NCS were predominantly female with an approximate female/male ratio of 3:1, which is consistent with the literature.

Various imaging modalities such as Doppler US, CT-CT angiography, MR-MR angiography, digital substraction angiography, and phlebography may be used for the diagnosis of NCS [7,12–15]. Although it is possible to measure the reno-caval pressure gradient with phlebography, the invasive nature of the procedure is a drawback [7].

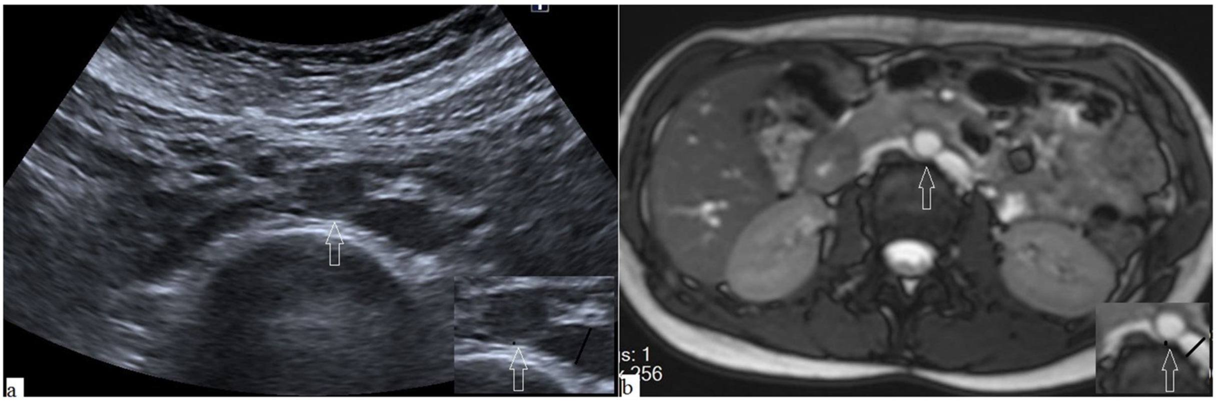

Fig. 2. Right-sided inferior vena cava and compressed left renal vein (arrow) between aorta and columna vertebralis. AP diameter measurements of the proximal hilar and retroaortic portions of LRV are shown in the small figures (short and long black lines) a: Transverse gray scale US b: Axial T2-TRUFI sequence.

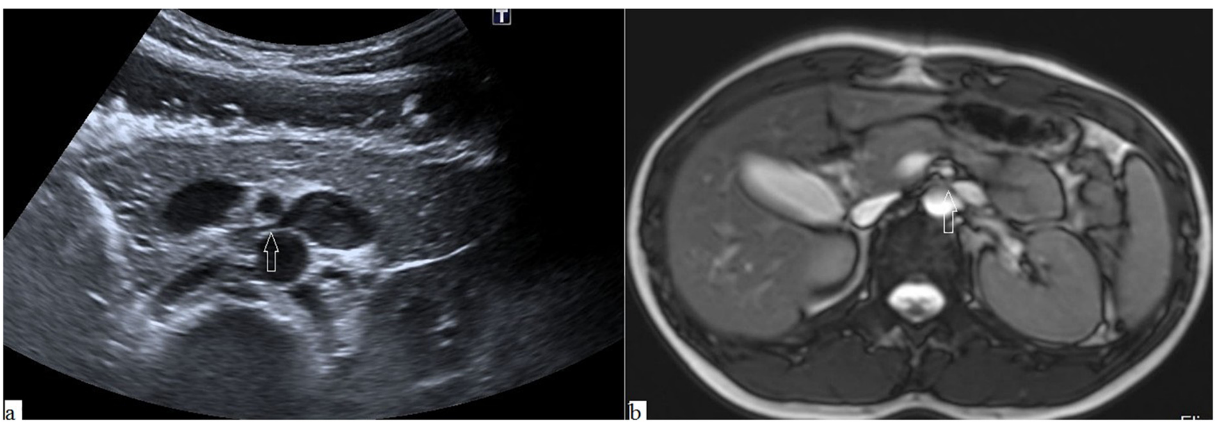

Fig. 3. Right-sided inferior vena cava and compressed left renal vein (arrow) between aorta and superior mesenteric artery a: Transverse gray scale US b: Axial T2-TRUFI sequence.

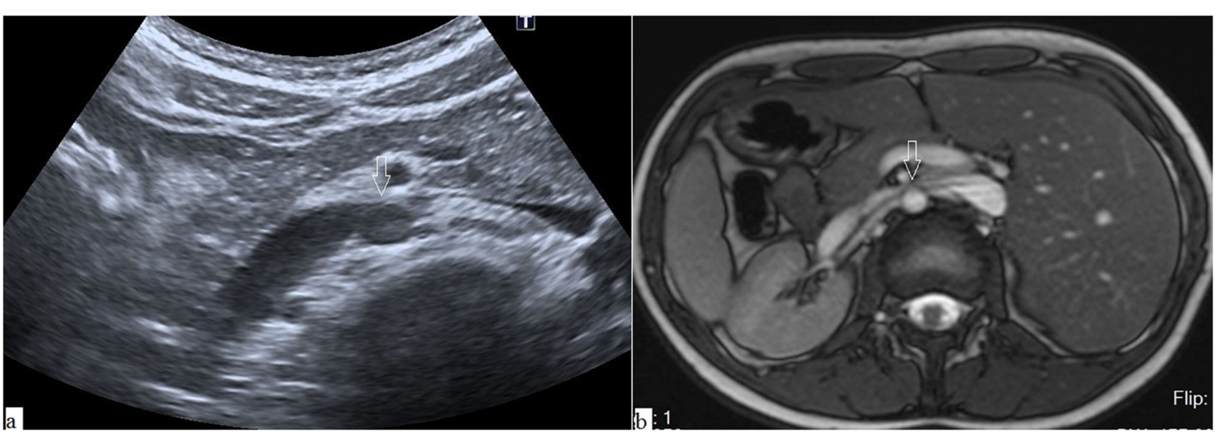

Fig. 4. Left-sided inferior vena cava and compressed right renal vein (arrow) between aorta and superior mesenteric artery a: Transverse gray scale US b: Axial T2-TRUFI sequence.

Table 2
Comparison of the scores according to the sequences.

		T2-TRUFI	T2-HASTE	T1-VIBE	Out–of–phase T1
Anatomical depiction	Mean:	3,9 (+/−, 30)	2,6 (+/−, 81)	2 (+/−, 69)	2,2 (+/−, 77)
Measurability	Mean:	3,8 (+/−, 40)	1,9 (+/−, 57)	1,5 (+/−, 64)	1,7 (+/−, 67)
Pulsation artifact	Mean:	3,5 (+/−, 55)	2,7 (+/−, 53)	2,6 (+/−, 57)	2,7 (+/−, 47)

Doppler US is a noninvasive method for diagnosing NCS and should be the preferred imaging modality. In addition to enabling diameter measurements, it also provides physiological information about blood flow in the RV [16]. In the literature, various significant Doppler US parameters have been defined for the diagnosis of NCS, such as maximal AP diameters, peak flow velocity, and aortomesenteric angle. However, there are potential drawbacks to abdominal Doppler imaging. One of these is insufficient cooperation of patients during the examination which may lead to positional changes and nonstandard measurements. Additionally, it may be difficult to obtain a standardized spectral Doppler signal from the aortomesenteric segment of the LRV due to blood flow [5,8,17]. MDCT angiography is a diagnostic procedure which provides much more substantial and detailed information about collateral vascular circulation and congenital vascular anomalies. It delineates the anatomical structures and enables multi-plan anatomical assessment. The major disadvantages of MDCT angiography are exposure to radiation and intravenous contrast material. MR angiographic findings are similar to MDCT angiography and may be more advantageous than MDCT angiography since no radiation is involved [17,18]. In a case report by Wong HL et al., it was reported that there was high signal intensity LRV in FSE T2WI sequence due to venous hypertension indicating flow stagnation. The investigators suggested that this finding may be used in the diagnosis and follow-up of NCS [17]. For this purpose, we used T2-TRUFI, T2-HASTE, T1-VIBE, and out-of-phase (opposed phase) T1 sequences. We assume that different sequences may be used to yield fast and high resolution anatomical images to show the relation between LRV and neighboring structures. We believe that MR imaging may be used in the follow-up of these patients to standardize incompatible US measurements due to its user-dependent technique. This method is also safer because contrast material is not used.

According to our results, in terms of anatomical depiction, measurability, and aortic pulsation artifact criteria, T2-TRUFI sequence has the most diagnostic value in assessing the LRV. In consideration of these results, it may be advantageous to use T2-TRUFI sequence to measure LRV in the aortomesenteric segment in selective cases with suboptimal US examination due to bowel distension.

There are some limitations of this study. First, non-contrast MR technique does not provide physiological information on LRV blood flow. Doppler US can better delineate information on blood flow dynamics. We believe that non-contrast MR techniques may have an incremental value in the diagnosis of NCS in children with suboptimal US imaging and a more accurate result may be achieved by using these sequences accompanied by US.

The second limitation of the current study is the number of patients. Since NCS is a rare condition and requires specific clinical features or criteria, its diagnosis may be challenging. Therefore, the number of children with NCS was limited in our study. We believe that more statistically accurate data may be obtained from long-term multicenter comprehensive studies.

5. Conclusion

Although Doppler US is the gold standard technique in the diagnosis of NCS, MR imaging may be used as an additional modality, and is superior to Doppler US in terms of anatomical depiction and its lack of artifacts due to patient cooperation. Non-contrast MR imaging, especially using TRUFI sequence, may hold an incremental value in the accurate diagnosis and follow-up of these patients.

Acknowledgments

We thank doctor Elif Cigdem Altunok, from the Department of Biostatistics and Medical Informatics, Faculty of Medicine, Yeditepe University, for their contribution in statistical analysis.

Declarations of interest

None.

References

[1] Lamba R, Tanner DT, Sekhon S, et al. Multidetector CT of vascular compression syndromes in the abdomen and pelvis. Radiographics 2014;34:93–115.
[2] de Schepper A. "Nutcracker" phenomenon of the renal vein and venous pathology of the left kidney. J Belge Radiol 1972;55:507.
[3] Lau JL, Lo R, Chan FL, et al. The posterior nutcracker. Haematuria secondary to retroaortic left renal vein. Urology 1986;28:437.
[4] Yildiz AE, Cayci FS, Genc S, et al. Right nutcracker syndrome associated with left-sided inferior vena cava, hemiazygos continuation and persistant left superior vena cava: a rare combination. Clin Imaging 2014;38:340–5.
[5] Kurlinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. Mayo Clin Proc 2010;85:552–9.
[6] He Y, Wu Z, Chen S, et al. Nutracker syndrome–how well do we know it? Urology 2014;83:12–7.
[7] Menard MT. Nutcracker syndrome: when should it be treated and how? Perspect Vasc Surg Endovasc Ther 2009;21:117–24.
[8] Fitoz S, Ekim M, Ozcakar ZB, et al. Nutcracker syndrome in children the role of upright position examination and superior mesenteric artery angle measurement in the diagnosis. J Ultrasound Med 2007;26:573–80.
[9] Preza Fernandes J, Amorim R, Gomes MJ, et al. Posterior nutcracker syndrome with left renal vein duplication: a rare cause of haematuria in a 12-year-old boy. Case Rep Urol 2012;2012.
[10] Singla RK, Sharma T, Gupta R. Retro-aortic left renal vein with left suprarenal vein draining into inferior vena cava. IJAV 2010;3:134–7.
[11] Kurklinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. Mayo Clin Proc 2010;85:552–9.
[12] Hohenfellner M, Steinbach F, Schultz-Lampel D, et al. The nutcracker syndrome: new aspects of pathophysiology, diagnosis and treatment. J Urol 1991;146:685–8.
[13] Kaneko K, Ohtomo Y, Yamashiro Y, et al. Magnetic resonance angiography in nutcracker phenomenon. Clin Nephrol 1999;51:259–60.
[14] Chen YM, Wang IK, Ng KK, et al. Nutcracker syndrome: an over- looked cause of hematuria. Chang Gung Med J 2002;25:700–5.
[15] Takemura T, Iwasa H, Yamamoto S, et al. Clinical and radiologi- cal features in four adolescents with nutcracker syndrome. Pediatr Nephrol 2000;14:1002–5.
[16] Takahashi Y, Sano A, Matsuo M. An ultrasonographic classification for diverse clinical symptoms of pediatric nutcracker phenomenon. Clin Nephrol 2005;64:47–54.
[17] Wong HL, Chen MCY, Wu CS, et al. The usefulness of fast-spin-echo T2-weighted MR imaging in nutcracker syndrome: a case report. Korean J Radiol 2010;11:373–7.
[18] Gulleroglu K, Gulleroglu B, Baskin E. Nutcracker syndrome. World J Nephrol November 6 2014;3:277–81.