Eur J Pediatr (2013) 172:135–136
DOI 10.1007/s00431-012-1859-5

CORRESPONDENCE

Renal Doppler ultrasonography in the diagnosis of nutcracker syndrome.

Se Jin Park · Jae Il Shin

Received: 7 August 2012 / Accepted: 2 October 2012 / Published online: 20 November 2012
© Springer-Verlag Berlin Heidelberg 2012

Dear Editor,

We read with great interest the contribution by Waseem et al. [6]. They reported a 17-year-old boy who presented with left flank pain and hematuria and was diagnosed with nutcracker syndrome (NCS) by computed tomography (CT). They also concluded that NCS is a rare but important and potentially treatable clinical condition and its early diagnosis with a high index of suspicion can achieve efficient management. Agreeing to their opinion, however, we would like to add a few comments on their conclusions.

Above all, NCS is really not a rare entity but an underestimated or underrecognized cause of hematuria in children [3]. One of our authors previously reported that NCS was present in 60 (40 %) of the 149 children in whom no other explanation for hematuria was ascertained [5]. When a peak velocity (PV) of the left renal vein (LRV) of at least 4.1 (the calculated cutoff level of the aortomesenteric PV2/hilar PV1 ratios for diagnosing NCS) was defined as abnormal, 72 cases (33.3 %) of 216 children with hematuria and no cases in normal children were diagnosed with NCS [5]. On the basis of these data [5], we speculate that childhood NCS has been underestimated in the past and is more common in children with isolated hematuria than we think.

Although left renal venography with measurement of the pressure gradient between the LRV and the inferior vena cava is the most accurate tool to diagnose NCS, it is an invasive and time-consuming procedure. In the case of Waseem et al. [6], the diagnosis of NCS was established by a CT scan, which has too much radiation. However, renal Doppler ultrasonography with color flow has improved functional diagnostic capabilities by showing the blood flow velocity of the LRV. When the criterion for the PV ratio (>4.7) was applied, the sensitivity and specificity of the PV2/PV1 ratios were 100 and 90 %, respectively, in children with NCS [1]. The detection rate of NCS by renal Doppler ultrasonography was highest among many other diagnostic tests [1, 5].

As for diagnosing NCS by renal Doppler ultrasonography, the upright position has significant effects on the angle of the superior mesenteric artery (SMA) and LRV hemodynamics. In the upright position, the mean PV ratios of the LRV at the aortomesenteric and hilar portions were 13.2± 6.9 and 3.9±2.2 in the NCS patient and control groups, respectively, and the PV ratios were statistically significant between the two groups both in the supine and upright position examinations ($P<0.001$) [2]. However, upright position imaging revealed a significant decrease of SMA angle and more pronounced entrapment findings in patients with NCS [2]. Recently, Romera-Villegas et al. [4] also demonstrated that the PV2 and PV1 in the standing position was 125.96 and 25.54 cm/s, respectively, compared to 115.48 and 25.77 cm/s in the supine position and suggested that the standing PV ratio is the best parameter for detecting NCS.

Therefore, we recommend that NCS should be reconsidered as one of the substantial causes in children with isolated hematuria and renal Doppler ultrasonography should be performed as the initial screening examination in the upright position, if possible, to avoid unnecessary invasive and hazardous procedures in the diagnosis of NCS.

S. J. Park
Department of Pediatrics, Ajou University Hospital,
Ajou University School of Medicine,
Suwon, Republic of Korea

J. I. Shin (✉)
Department of Pediatrics, Severance Children's Hospital,
Yonsei University College of Medicine,
50 Yonsei-ro, Seodaemun-gu, C.P.O. Box 8044, Seoul 120-752,
Republic of Korea
e-mail: shinji@yuhs.ac

References

1. Cheon JE, Kim WS, Kim IO, Kim SH, Yeon KM, Ha IS, Cheong HI, Choi Y (2006) Nutcracker syndrome in children with gross haematuria: Doppler sonographic evaluation of the left renal vein. Pediatr Radiol 36(7):682–686
2. Fitoz S, Ekim M, Ozcakar ZB, Elhan AH, Yalcinkaya F (2007) Nutcracker syndrome in children: the role of upright position examination and superior mesenteric artery angle measurement in the diagnosis. J Ultrasound Med 26(5):573–580
3. Park SJ, Shin JI (2012) Nutcracker syndrome: is it really a rare cause of hematuria? Comment on: Le syndrome casse-noisette (Nutcracker): une cause rare d'hématurie. Arch Pediatr 2011;18:1188–90. Arch Pediatr 19(6):658
4. Romera-Villegas A, Vila-Coll R, Cairols-Castellote M, Poveda-Monge R, Masuet-Aumatell C, Grinyó-Boira M (2009) The importance of a standing position in the diagnosis of Nutcracker phenomenon by duplex sonography. Int Angiol 28(6):461–468
5. Shin JI, Park JM, Lee JS, Kim MJ (2007) Effect of renal Doppler ultrasound on the detection of nutcracker syndrome in children with hematuria. Eur J Pediatr 166(5):399–404
6. Waseem M, Upadhyay R, Prosper G (2012) The nutcracker syndrome: an underrecognized cause of hematuria. Eur J Pediatr 171 (8):1269–1271