

## BRIEF REPORT

Kazunari Kaneko · Keiichi Kiya  
Kimihiro Nishimura · Toshiaki Shimizu  
Yuichiro Yamashiro

## Nutcracker phenomenon demonstrated by three-dimensional computed tomography

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**Abstract** We report a 15-year-old girl with nutcracker syndrome (NCP) in whom three-dimensional helical computed tomography (3D helical CT) was useful for diagnosis; angiographic imaging using 3D helical CT provided fine outlines of the vascular structures in NCP, i.e., a compression of the left renal vein between the aorta and the superior mesenteric artery and the abnormal acute angle between them. Thus, a 3D helical CT enables precise depiction of the anatomical characteristics of renal vasculature and, therefore, may be a useful alternative imaging technique instead of conventional examinations, such as invasive venography or digital subtraction angiography. Further study in a large number of children with vascular abnormalities would be worthwhile.

**Keywords** Three-dimensional imaging · Helical CT · Spiral CT · Nutcracker phenomenon · Left renal vein entrapment

### Introduction

Nutcracker phenomenon (NCP) refers to the compression of the left renal vein (LRV) between the aorta and the superior mesenteric artery (SMA) which results in renal vein and left gonadal vein varices [1]. This is uncommon, but a well accepted cause of hematuria. The diagnosis of NCP can be established by venographic imaging, which measures the pressure gradient between the

LRV and the inferior vena cava (IVC) or intraarterial digital subtraction angiography (DSA) [2]. Since these examinations are relatively invasive, a less invasive method for diagnosis is desirable.

Recently, it has been reported that magnetic resonance angiography (MRA), which is less invasive compared to conventional angiography, is useful for the diagnosis of NCP [3, 4]. This is not conclusive, however, because of the low resolution, and invasive angiographic study still seems to be necessary.

Development of the three-dimensional (3D) imaging techniques in computed tomography (CT) has enabled surgeons to carry out preoperative planning for brain, maxillofacial, and orthopedic surgery. These areas are particularly amenable to 3D imaging because of the minimal motion artifacts during imaging [5]. In addition, advances in helical CT techniques have improved renal imaging by decreasing volume-averaging artifacts and eliminating respiratory misregistration artifacts [6, 7]. Thus, improvements in helical CT combined with continued advances in computer hardware and software have allowed the production of high quality 3D CT images, even of the renal vasculature in any plane [7].

In this study, we describe the usefulness of 3D helical CT imaging in the case of complex renal vascular abnormality, i.e., NCP.

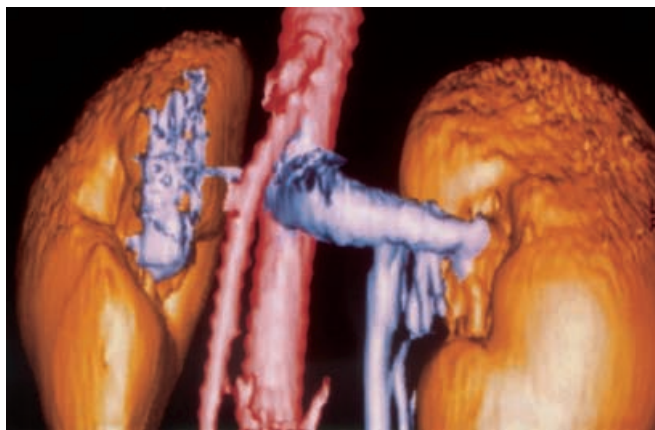
### Case report

A girl, aged 15 years, was admitted to our hospital for the evaluation of a macroscopic hematuria which had started 4 months previously, when she was suffering from bacterial enterocolitis. Past medical history was unremarkable for renal trauma, renal colic, and urinary tract infection. Physical findings were also unremarkable. Initial examinations including complete blood cell count, biochemistry, and blood clotting profiles were all normal. Urinalysis revealed numerous red blood cells per high power field with trace protein. Morphology of urine red blood cells suggested that hematuria was of non-glomerular origin because of the predominance (>90%) of isomorphic urine red blood cells. Repeated urine cultures were negative for pathological bacteria. A renal ultrasound suggested a severe compression of the left renal vein be-

K. Kaneko (✉) · T. Shimizu · Y. Yamashiro  
Department of Pediatrics,  
Juntendo University School of Medicine, 2-1-1 Hongo,  
Bunkyo-ku, Tokyo 113-8421, Japan  
e-mail: kkaneko@med.juntendo.ac.jp  
Tel.: +81-3-38133111, ext. 3324  
Fax: +81-3-58000216

K. Kiya  
Department of Pediatrics, Tanashi Dai-ichi Hospital, Tokyo, Japan

K. Nishimura  
Department of Radiology, Tanashi Dai-ichi Hospital, Tokyo, Japan



**Fig. 1** Three-dimensional helical CT in a patient with NCP. Left renal vein (blue) was compressed by the aorta (red) and the SMA (red). The angle between the aorta and SMA was abnormally acute ( $<45^\circ$ , normal  $90\pm 10^\circ$  [1])

tween the abdominal aorta and the SMA. From these findings, a presumptive diagnosis of NCP was made.

She received scanning with spiral volumetric acquisition CT, which obtains images during continuous rotation of the X-ray source while the patient moves at a constant velocity through the gantry. Briefly, spiral CT (Lumage Supreme, GE, USA) was performed first with no intravenous contrast medium and then after an intravenous bolus injection with 90 ml contrast medium (300 mg I/ml at 2 ml/s). Scans were taken at 120 kV and 160 mA s. For the vascular phase scan, a delay of 45 s was used; this scan (CT angiography) was taken with 5 mm collimation and a 7 mm/s table speed, which provided a scan time of 15 s at full inspiration. Overlapping images were reconstructed at 2.5-mm intervals. Images were reconstructed using a volume-rendering workstation and computer software (Advantage Windows Ver 3.1, GE, USA) via a reading workstation.

As shown in Fig. 1, 3D reconstruction of helical CT angiographic imaging provided useful information that gave a precise depiction of both a compression of the LRV between the aorta and the SMA and the acute angle between the aorta and the SMA.

Based on these findings, the diagnosis of NCP, which was responsible for her hematuria, was made. Although treatment of the NCP is controversial, therapy is dictated principally by the clinical situation [1]: she has been followed in conservative fashion, i.e., urinalysis because of the lack of pain. Routine follow-up by urinalysis performed bimonthly and physical examination have revealed the improvement of hematuria assessed by the dipstick test (4+ in April 2000, 2+ in April 2001) and no further episodes of macroscopic hematuria. This suggests the natural regression of nutcracker phenomenon, which is occasionally seen in this condition as previously described [8].

## Discussion

NCP is characterized by left-sided renal bleeding due to compression of the LRV in the fork between the abdominal aorta and the proximal SMA, close to its origin. This results in left renal venous hypertension leading to the development of collateral veins with intrarenal and perirenal varicosities which can cause hematuria if the thin-walled septum separating the veins from the collecting system ruptures [8]. The main presenting symptom is hematuria, with or without left flank pain. Some patients

may present with left flank pain alone and, in a few, varicocele might be the only complaint. Exercise seems to aggravate the symptoms.

Although the pathophysiology of NCP is not fully recognized, recent observations revealed that abnormal branching of the SMA from the aorta is the underlying principle [1]. The diagnosis of NCP can be established by venographic imaging or intraarterial DSA. Since these examinations are relatively invasive, a less-invasive method for diagnosis is desirable and several methods to confirm the presence of compression of LRV have been proposed for this purpose: angiographic CT [1], simple MR imaging [9], and recently MRA [3, 4]. They appear, however, not to be conclusive, because of the low resolution.

CT has evolved continuously since its introduction into medical imaging in the early 1970s. The newest generation of CT, helical CT, provides a technology which allows for continuous data acquisition and improved 3D reconstruction. Although there are increasing numbers of publications evaluating this technology in the adult for vascular, neck, chest and abdominal imaging, there are few reports, to date, which address its role in pediatrics, particularly in terms of vascular imaging [10]. Furthermore, the main indications for 3D CT in children have focused on imaging the craniofacial malformations involving bone [11].

Our case appears to be the first report of 3D CT angiographic imaging used successfully in the detection of NCP. This method could visualize the principal vascular abnormalities of NCP, i.e., abnormal branching of SMA from abdominal aorta associated with LRV. Therefore, it appears that renal 3D CT angiographic imaging can be a very useful, non-invasive alternative to conventional angiography for the assessment of diverse conditions affecting renal vessels, such as renal artery stenosis, screening of potential renal donors, planning repair of ureteropelvic junction stenoses, and identification of relevant renal artery relationships to abdominal aortic lesions as previously described in adults [12].

In conclusion, a 3D reconstructed volumetric CT enables precise depiction of the anatomical characteristics of NCP and, therefore, might be an alternative imaging technique instead of invasive venography or DSA. Further study which focuses on improving the procedure and imaging in a large number of children with abnormal renal vasculature would be worthwhile.

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## LITERATURE ABSTRACTS

M. Levy · J. Feingold

### Estimating prevalence in single-gene kidney diseases progressing to renal failure

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Incidence and prevalence, the measures of “frequency”, are often confused. While in a nonhereditary situation, the useful parameter is the incidence rate, evaluating the impact of an etiologic factor, it is prevalence that is considered useful in a hereditary disease. Prevalence may concern either the whole population or a fraction of this population, that is, males or females or individuals at a given age, for example, at birth. Pathologic phenotype and morbid genotype prevalences have to be clearly differentiated. In this article, we review the epidemiologic surveys allowing an estimation of the distribution of major single-gene kidney diseases progressing to renal failure in different populations. In order to compare their results, the geographic/ethnic composition of the population, the determination of its size, the choice and mode of calculation of the epidemiologic measure, the definition of the disease and modes of diagnosis, the inclusion of cases, the sources of ascertainment and the possible causes of underascertainment, and the period of time during which events were counted should be analyzed accurately. Although their impact in terms of morbidity, hospitalizations, mortality, and cost to society is high, this review shows that information on the prevalence of single-gene kidney diseases is far from complete. To date, the data essentially apply to large populations of European origin. A part of the variation among prevalence data may be due to methodological differences. Not representative are the small populations in which some rare diseases, especially recessive, are found with a high prevalence.

J. Inatomi · K. Yoshioka

### Identification of two novel mutations in the CLCN5 gene in Japanese patients with familial idiopathic low molecular weight proteinuria (Japanese Dent's disease)

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Two Japanese patients, belonging to unrelated families, with idiopathic low-molecular-weight proteinuria (LMWP; Japanese Dent's disease) showed novel mutations of the gene encoding renal-specific chloride channel 5 (CLC-5). Proteinuria was first noticed at the ages of 2 and 3 years in patients 1 and 2, respectively. During follow-up, marked increases in urinary ss(2)-microglobulin levels, hypercalciuria, and high levels of urinary excretion of growth hormone were observed in both patients. Nephrocalcinosis was detected in patient 2. Renal biopsy specimens from both patients showed minimal alterations in glomeruli and tubulointerstitium, except for mild mesangial proliferation in patient 2. DNA sequence analysis of the entire 2238-bp coding region and exon-intron boundaries of the CLCN5 gene showed the presence of two novel mutations in exon 10, consisting of one missense mutation (I524K) in patient 1 and one nonsense mutation (R637X) in patient 2. DNA analysis and measurement of urinary ss(2)-microglobulin levels in family members indicated an X-linked mode of inheritance in patient 1 and sporadic occurrence in patient 2. These results have expanded our understanding of the association between idiopathic LMWP (Japanese Dent's disease) and mutations of the CLCN5 gene.