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REVIEW

Nutcracker Syndrome: An Update on Current Diagnostic Criteria and Management Guidelines

K. Ananthan [a], **S. Onida** [b], **A.H. Davies** [b,*]

[a] North West Thames Foundation School, Watford General Hospital, Watford, UK
[b] Academic Section of Vascular Surgery, Division of Surgery, Department of Surgery & Cancer, Imperial College London, Charing Cross Hospital, London, UK

WHAT THIS PAPER ADDS

This review provides an update and reviews the most recent literature relating to nutcracker syndrome. Specifically, it reviews the latest evidence base for diagnosing nutcracker syndrome using a combination of history and clinical examination, as well as various imaging modalities and outlines key diagnostic criteria. Furthermore, the approach to management of this condition is discussed in detail. Compared with other reviews, this paper also explores the role of minimally invasive approaches, such as endovascular and laparoscopic surgery.

Background: Nutcracker syndrome (NCS) describes left renal vein compression between the superior mesenteric artery and the aorta. Although uncommon, it is an important diagnosis due to the important morbidity associated with it, including the risk of chronic kidney disease from long-term left renal vein (LRV) hypertension and the risk of LRV thrombosis.

Methods: This article reviews the literature on NCS, particularly with respect to the diagnostic accuracy of different imaging modalities and the success rates, complications, and long-term follow-up data associated with various surgical interventions.

Results and discussion: The diagnosis of this condition is based on a stepwise work-up with history and clinical examination, followed by Doppler ultrasonography, computed tomography, magnetic resonance imaging, intravascular ultrasound (IVUS) and phlebography with measurement of the renocaval pressure gradient. Management is determined by symptom severity; often symptom resolution occurs following a conservative approach. However, in some cases, surgical management is required, particularly when conservative management is unsuccessful. When it comes to the surgical management of NCS three main pathways exist: open surgery, laparoscopic surgery and endovascular approaches, with the latter 2 becoming increasingly popular due to their minimal invasiveness. Additionally, cases involving the use of robotic surgery in the management of NCS have been reported.

Conclusion: Despite the rarity of NCS, its recognition and management are important. This article has explored the evidence basis for conservative, medical and surgical options.

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INTRODUCTION

Nutcracker syndrome (NCS), also known as left renal vein (LRV) entrapment syndrome, was first defined anatomically by Grant in 1937 as follows: "… the left renal vein, as it lies between the aorta and superior mesenteric artery, resembles a nut between the jaws of a nutcracker."[1] This condition leads to stenosis of the aorto-mesenteric region of the LRV, with dilatation of the distal portion of the vessel. In 1950, El Sadr and Mina first described the compression of the LRV by the aorta and superior mesenteric artery (SMA).[2] Despite the term "nutcracker" being coined in 1971,[3] it was in 1972 that De Schepper, a Belgian radiologist, named the disorder as nutcracker syndrome.[4] Although the term is used interchangeably with nutcracker phenomenon, it is clear that such anatomy does not always lead to clinical symptoms. Hence the term NCS should be limited to patients who

* Corresponding author. Academic Section of Vascular Surgery, Division of Surgery, Department of Surgery and Cancer, Imperial College London, Floor 4 East, Charing Cross Hospital, Fulham Palace Road, London W6 8RF, UK.

E-mail address: a.h.davies@imperial.ac.uk (A.H. Davies).

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present with the characteristic clinical signs and symptoms,[5] specifically haematuria, proteinuria, flank pain, pelvic congestion in females, and varicocele in male patients,[6] alongside diagnostic imaging of the anatomy associated with the syndrome.[5,7,8] This is important, as asymptomatic dilatation of the LRV found on diagnostic imaging such as Doppler ultrasound (DUS) and computed tomography (CT), is considered to be a normal variant.[9] This review aims to identify the common clinical presentations, key diagnostic criteria, and diagnostic accuracy of various imaging modalities for NCS, as well as reporting follow-up data on different NCS management strategies ranging from conservative to open surgery and endovascular techniques.

EPIDEMIOLOGY

The exact prevalence of NCS is unknown, partly because of an absence of definitive diagnostic criteria and partly because of the variability in symptomatic presentation. However, unexplained haematuria is a common feature and NCS has been diagnosed by DUS in 40% of patients with this clinical presentation.[9] Patients can present at any age from childhood to the seventh decade,[5,6,8,10–15] with prevalence peaking in young (second or third decade)[16] and middle aged adults.[11,12] The rapid increase in height and development of the vertebral bodies in puberty results in narrowing of the angle between the aorta and superior mesenteric artery.[16] The prevalence of this condition has been reported as higher in females;[6,17,18] however, further studies have shown that it is equally prevalent among both genders.[11]

AETIOLOGY

There are two main forms of NCS. The most common, anterior NCS, refers to compression of the LRV between the abdominal aorta and SMA (Fig. 1). According to Zhang et al., to diagnose NCS, the angle between the SMA and the abdominal aorta needs to be less than 45° when measured in the sagittal plane;[19] specifically, an angle less than 35° is sufficient for a definitive diagnosis.[20] This is because it is the initial acute angle of descent of the SMA branching from the aorta that causes LRV compression.[13] Supporting this, Kim et al. demonstrated that an angle <39° in the sagittal plane on CT had a sensitivity of 92% and specificity of 89% for detecting symptomatic NCS[21] compared with left renal venography and measurement of the pressure gradients between the LRV and inferior vena cava (IVC). However, as displayed in Table 2, the reliability of these results is limited because of the small sample size. Posterior NCS, although less common with only 19 cases reported in the literature,[22–24] occurs when the retro-aortic or circum-aortic renal vein (incidence 0.8–7.1%)[25] is compressed in the narrow gap between the aorta and vertebral body.[26] The existence of combined (anterior and posterior) NCS has been reported resulting from duplication of the LRV such that it has both a pre- and retro-aortic course (0.3–5.7%).[10] In this case, the anterior tributary of the LRV is compressed between the aorta and SMA, while the posterior tributary is compressed between the aorta and the vertebral column.[27] Less common pathologies and conditions leading to NCS result from compression of the LRV and are demonstrated in Table 1.

CLINICAL FEATURES

The most common clinical presentations of NCS include pelvic pain, flank pain, haematuria, and gonadal varices (varicocele or ovarian vein syndrome, Fig. 2),[6–8,10,11,13–16,19,27–30,32–37] reflecting pelvic and renal congestion. It is hypothesized that elevated pressure in the stenosed LRV results in the formation of venous reflux, venous hypertension, and therefore variceal formation between the renal pelvis and ureter, manifesting with microhaematuria or macrohaematuria.[38] This is secondary to the connection between the dilated

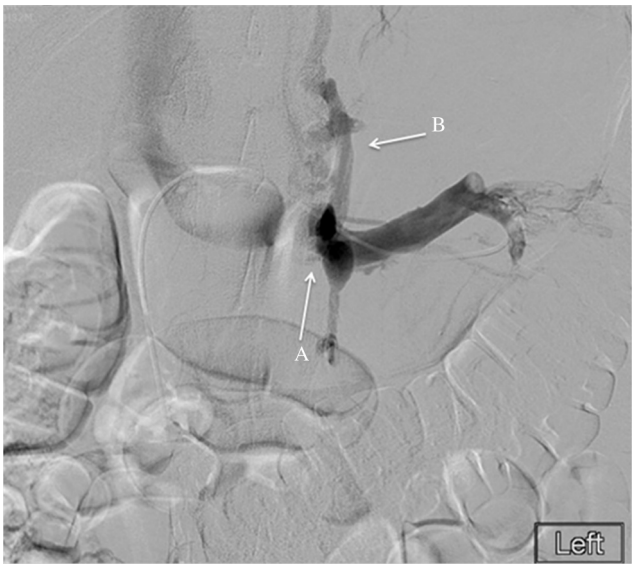

Figure 1. Left renal venogram demonstrating narrowing of the left renal vein prior to entry in the IVC (A) and collateral para-vertebral flow (B).

Table 1. Less common pathologies and conditions leading to NCS as a result of compression of the left renal vein.

Pathologies leading to NCS
Pancreatic neoplasms
Para-aortic lymphadenopathy
Retroperitoneal tumour
Abdominal aortic aneurysm
Overarching testicular artery
Strangulating fibrolymphatic tissue between the aorta and SMA[a, b, c]
High disposition of the LRV[d]
Left renal ptosis resulting in stretching of the left renal vein over the abdominal aorta[e]
Lordosis[e]
Reduced retroperitoneal and mesenteric fat[f, g]
Pregnancy with a gravid uterus compressing the renal vasculature[h]

Less common pathologies leading to NCS based on studies by Menard et al.[a,13], Pastershank et al.[b,28], Neste et al.[c,29], Kurklinsky et al.[d,12], Shokier et al.[e,59], Wendel et al.[f,30], Venkatachalam et al.[g,8], Zhang et al.[h,19], and Itoh et al.[i,31]

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