

# Macroscopic Haematuria in a 15-year old Male: A Case of Nutcracker Syndrome Managed by Endovascular Stenting

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## Abstract

Nutcracker Syndrome (NCS) is the compression of the left renal vein between the aorta and superior mesenteric artery. NCS is a rare cause of haematuria in children and its treatment remains controversial. In this paper, we report the case of a 15-year old boy with macroscopic haematuria diagnosed as NCS using renal angiography. After two years of conservative management, endovascular stenting was performed due to persistence of the macroscopic haematuria and progression of left flank pain. This resulted in the patient's gradual resolution of haematuria

## INTRODUCTION

Nutcracker syndrome (NCS) is the impingement of the left renal vein between the aorta and superior mesenteric artery [1]. It is an uncommon cause of haematuria in children [2] with only a few documented cases in medical literature. The typical presentation of NCS are microscopic or macroscopic haematuria with or without proteinuria [3]. Although there are case reports of NCS [4, 5] which resulted in spontaneous remission, massive and persistent macroscopic haematuria may require appropriate intervention [6, 7]. The treatment for NCS is controversial [8]. Case studies in medical literature propose several invasive treatment options such as nephrectomy, nephropexy, renal vein transposition, and autotransplantation [8, 9]. The use of expandable metallic stents has recently been reported to be effective in the treatment of NCS with persistent haematuria [8, 10].

We present the case of 15-year old boy diagnosed with NCS who underwent endovascular stenting of the left renal vein after two years of conservative management which failed to show any resolution of the macroscopic haematuria.

## CASE

A 15 year-old male was initially referred for evaluation of a three-month history of intermittent macroscopic haematuria and left flank pain. His past medical history was unremarkable for any congenital problems, preceding infection, abdominal trauma, or renal colic. There was no family history of hematuria or any hematologic or renal diseases. Physical examination was unremarkable.

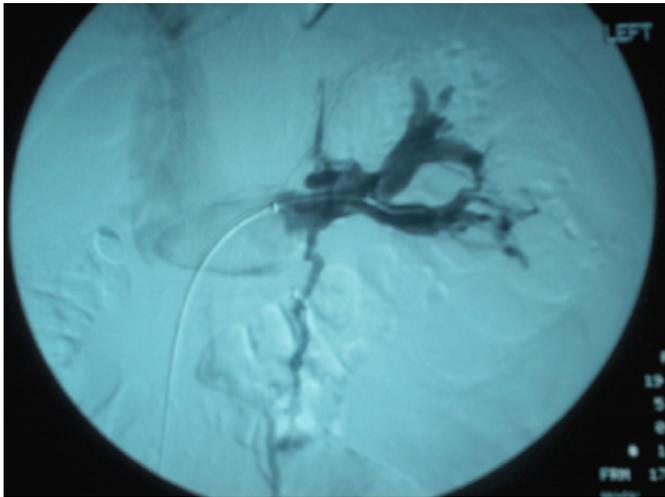
Urinalysis revealed numerous red blood cells (RBC) per high power field with RBC casts. RBC morphology was predominantly (>90%) isomorphic which suggested that the hematuria was of non-glomerular etiology. Other pertinent hematologic and clinical chemistry investigations were all within normal range which ruled out coagulopathy or blood dyscrasias. Screening urinalysis for the patient's family members was also negative.

Based on the patient's medical records, renal ultrasound and abdominal CT scan did not detect any nephrolithiasis, tumors or lesions within the kidneys, ureters or bladder. Interestingly, patient's cystoscopy showed bleeding that solely emanated from the left renal system. In addition, a renal biopsy was done but failed to show any glomerular pathology on light and electron microscopy. Immunofluorescence microscopy was also negative for immune reactants.

In order to rule out any undetected renovascular pathology, we requested for biplanar abdominal aortography with left renal arteriography which showed an abnormal venous phase on the left renal vein with varicosities of collateral veins secondary to the left renal vein compression between the aorta and superior mesenteric artery (See Figure 1). Based on these findings, we made the diagnosis of NCS. After informing the patient and his parents regarding the prognosis of NCS, they decided to pursue conservative management.

**Figure 1**

Figure 1: Biplanar aortogram with venous phase showing dilatation of the left renal vein and presence of varicosities

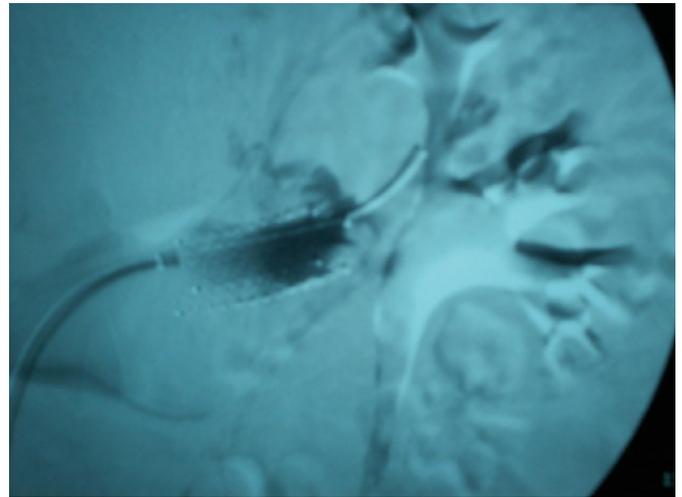


After two years of observation, the patient and his parents decided to seek intervention due to the persistence of haematuria and progressive left flank pain. In the interval time, the patient's haematocrit has begun to decline slightly below normal. Nevertheless, he remained normotensive without any signs of hemodynamic compromise. Renal function was also stable.

The patient underwent endovascular stenting of the left renal vein. Under fluoroscopic guidance, a 14mm X 6cm Zilver® stent was inserted into the left renal vein. A final venogram was performed to check proper placement and adequate vein dilatation. (See Figure 2) Due to the haematuria, the patient was not placed on any form of anticoagulation. The patient was discharged the next day without any complications. The macroscopic haematuria gradually resolved in the succeeding weeks. He subsequently returned to his normal daily activities without any significant discomfort.

**Figure 2**

Figure 2: Placement of intravascular stent on the left renal vein



## DISCUSSION

Nutcracker syndrome (NCS) was first described in the medical literature in 1950 [11]. The compression of the left renal vein between the aorta and superior mesenteric artery was described to resemble a nut trapped between the nutcracker by the anatomist, John C. B. Grant [11]. Its development is usually ascribed to the abnormal branching of the superior mesenteric artery from the aorta [7]. The most common presentation of NCS is haematuria in the absence of any other renal pathology. The haematuria is brought about by the rupture of congested renal veins into the collecting system [12].

After excluding the more common causes of hematuria such as nephrolithiasis, glomerular diseases or tumors, the diagnosis of NCS is usually established by angiography with venographic imaging [13]. Recently, abdominal CT scan [2], three dimensional helical CT [13], and magnetic resonance angiography [14] have been used. Unfortunately, the results have been inconsistent due to several factors. Nevertheless, we recommend that macroscopic haematuria that solely emanates from the left renal system should necessitate further evaluation for the presence of NCS especially in the absence of nephrolithiasis or any renal parenchymal disease that may not fully explain the severity of the haematuria.

The prognosis of NCS in children is also poorly understood. Conservative management is suggested for mild haematuria. Surgical intervention has been advised for persistent macroscopic haematuria [6], severe abdominal or flank pain [2] or autonomic dysfunction [15]. Wendel et al. [16] proposed

medial nephropexy while Stewart et al. [17] used transposition of the left renal vein. Other investigators suggest autotransplantation [18]. Our case provides an example of the benefit of endovascular stenting which is a less invasive procedure. This procedure protects the left renal vein from persistent compression without undergoing major surgery. This allows children to immediately resume their normal daily life with minimal post-operative discomfort. However, more studies are needed to further assess the benefit of endovascular stenting on long term basis.

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