

# EVIDENCE BASED STATEMENT

DOMAIN **05**, Statement **09**

TOPIC: “**Iliac/Renal vein compression management**”

## SEARCH TERMS & SOURCES

(nutcracker syndrome)

### INCLUSION CRITERIA

- English language
- Reviews, Meta-analysis
- Publication < 10 years, only ENG

## SEARCH RESULT BEFORE - AFTER SELECTION

55/20

### PERTINENT LITERATURE NOT IDENTIFIED BY THE LITERATURE SEARCH

1. Miró I, Serrano A, Pérez-Ardavín J, et al. Eighteen years of experience with pediatric nutcracker syndrome: the importance of the conservative approach. *J Pediatr Urol* 2020;16:218.e1-6.
2. Wu Z, Zheng X, He Y, et al. Stent migration after endovascular stenting in patients with nutcracker syndrome. *J Vasc Surg Venous Lymphat Disord* 2016;4:193-9.
3. Hannick JH, Blais AS, Kim JK, et al. Prevalence, Doppler ultrasound findings, and clinical implications of the nutcracker phenomenon in pediatric varicoceles. *Urology* 2019;128:78-83
4. Beinart C, Sniderman KW, Saddekni S, et al. Left renal vein hypertension: a cause of occult hematuria. *Radiology* 1982;145:647-50.

# EVIDENCE BASED STATEMENT

## Domain 5; Statement 9

### IDENTIFIED REFERENCES

1. Barge TF, Uberoi R. Symptomatic pelvic venous insufficiency: a review of the current controversies in pathophysiology, diagnosis, and management. *Clin Radiol*. 2022 Jun;77(6):409-417.
2. Rangel Villalobos E, Busquier Cerdán T, Cortés Sañudo X, et al. Vascular compression syndromes: the value of Doppler ultrasonography. *Radiologia (Engl Ed)*. 2022 Jan-Feb;64(1):17-25.
3. Meyer J, Rother U, Stehr M, Meyer A. Nutcracker syndrome in children: Appearance, diagnostics, and treatment - A systematic review. *J Pediatr Surg*. 2022 Jan 1:S0022-3468(21)00874-5.
4. Kolber MK, Cui Z, Chen CK, et al. Nutcracker syndrome: diagnosis and therapy. *Cardiovasc Diagn Ther*. 2021 Oct;11(5):1140-1149.
5. Sandmann W, Scholbach T, Verginis K. Surgical treatment of abdominal compression syndromes: The significance of hypermobility-related disorders. *Am J Med Genet C Semin Med Genet*. 2021 Dec;187(4):570-578
6. Brahmabhatt A, Macher J, Shetty AN, et al. Sonographic Evaluation of Pelvic Venous Disorders. *Ultrasound Q*. 2021 Sep 1;37(3):219-228.
7. Chait J, Sen I, Kalra M. Nutcracker Syndrome: How to Diagnose It and When/How Should It Be Treated in the Pelvic Venous Disease Population. *Tech Vasc Interv Radiol*. 2021 Mar;24(1):100734.
8. Franco-Mesa C, Gloviczki P, Erben Y. Nutcracker syndrome. *J Cardiovasc Surg (Torino)*. 2021 Oct;62(5):467-471.
9. Farina R, Foti PV, Conti A, et al. The role of ultrasound imaging in vascular compression syndromes. *Ultrasound J*. 2021 Feb 8;13(1):4.
10. Wang RF, Zhou CZ, Fu YQ, Lv WF. Nutcracker syndrome accompanied by hypertension: a case report and literature review. *J Int Med Res*. 2021 Jan;49(1):300060520985733.
11. Granata A, Distefano G, Sturiale A, et al. From Nutcracker Phenomenon to Nutcracker Syndrome: A Pictorial Review. *Diagnostics (Basel)*. 2021 Jan 11;11(1):101.
12. Gozzo C, Giambelluca D, Cannella R, et al. CT imaging findings of abdominopelvic vascular compression syndromes: what the radiologist needs to know. *Insights Imaging*. 2020 Mar 17;11(1):48.
13. de Macedo GL, Dos Santos MA, Sarris AB, Gomes RZ. Diagnosis and treatment of the Nutcracker syndrome: a review of the last 10 years. *J Vasc Bras*. 2018 Jul-Sep;17(3):220-228.
14. Orczyk K, Wysiadecki G, Majos A, et al. What Each Clinical Anatomist Has to Know about Left Renal Vein Entrapment Syndrome (Nutcracker Syndrome): A Review of the Most Important Findings. *Biomed Res Int*. 2017;2017:1746570.
15. Velasquez CA, Saeyeldin A, Zafar MA, et al. A systematic review on management of nutcracker syndrome. *J Vasc Surg Venous Lymphat Disord*. 2018 Mar;6(2):271-278.
16. Park JH, Lee GH, Lee SM, et al. Posterior nutcracker syndrome - a systematic review. *Vasa*. 2018 Jan;47(1):23-29.
17. Siddiqui WJ, Bakar A, Aslam M, et al. Left Renal Vein Compression Syndrome: Cracking the Nut of Clinical Dilemmas - Three Cases and Review of Literature. *Am J Case Rep*. 2017 Jul 6;18:754-759.
18. Ananthan K, Onida S, Davies AH. Nutcracker Syndrome: An Update on Current Diagnostic Criteria and Management Guidelines. *Eur J Vasc Endovasc Surg*. 2017 Jun;53(6):886-894.
19. White JM, Comerota AJ. Venous Compression Syndromes. *Vasc Endovascular Surg*. 2017 Apr;51(3):155-168.
20. Zucker EJ, Ganguli S, Ghoshhajra BB, et al. Imaging of venous compression syndromes. *Cardiovasc Diagn Ther*. 2016 Dec;6(6):519-532.

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### TEXT FOR INCLUSION IN THE DOCUMENT

DOMAIN 05, Statement 09, TOPIC: “**Iliac/Renal vein compression management**”

Nutcracker Syndrome is a condition arising from an aortomesenteric (AM) angle compressing the left renal vein or, more rarely, from a retroaortic left renal vein compressed against the vertebral column. Circumaortic or duplicated left renal vein have also been reported in simultaneously anterior and posterior compression syndromes. Other causes of left renal vein compression have been identified in association with lymphadenopathy or malignancy, pregnancy, lordosis, pregnancy, weight loss and intestinal malrotation. Its diagnosis includes a variety of heterogeneous signs and symptoms, therefore the exact prevalence remains undetermined. Typical manifestations include hematuria, proteinuria and flank or pelvic pain

**[Orczyk K, Wysiadecki G, Majos A, et al. What Each Clinical Anatomist Has to Know about Left Renal Vein Entrapment Syndrome (Nutcracker Syndrome): A Review of the Most Important Findings. Biomed Res Int. 2017;2017:1746570].**

Ultrasound scanning followed by computed tomography and/or magnetic resonance imaging can provide useful findings, catheter venography and pressure measurement are usually necessary for the diagnosis. Univocal ultrasound criteria are still missing. In the same way, left renal vein caliber and aorto-mesenteric angle values variations have been reported in the literature, yet with a sensitivity and specificity never superior to 91% up to our knowledge. Intravascular ultrasound can be of value in the completion of the diagnosis, as well as a cystoscopy assessing the eventual hematuria.

**[Barge TF, Uberoi R. Symptomatic pelvic venous insufficiency: a review of the current controversies in pathophysiology, diagnosis, and management. Clin Radiol. 2022 Jun;77(6):409-417].**

Conservative approach, eventually by angiotensin converting enzyme inhibitors in case of orthostatic proteinuria, should be first taken into consideration in pediatric nutcracker syndrome in whom the growth is usually associated with a spontaneous resolution.

**[Chait J, Sen I, Kalra M. Nutcracker Syndrome: How to Diagnose It and When/How Should It Be Treated in the Pelvic Venous Disease Population. Tech Vasc Interv Radiol. 2021 Mar;24(1):100734].**

Renal vein transposition remains the goldstandard treatment for symptomatic patients. Laparoscopic or robot-assisted procedures have been reported, yet the percentage of re-stenosis has been reported to be over 65%.

In case of need, auto-transplantation remains a valid option, despite the technically associated more possible complications.

Stent placement has also been proposed in small series. Migration has been already reported in 6.7% cases, confirming the need of relying always only on specialized centers and of producing more data on the topic.

**\*[Kolber MK, Cui Z, Chen CK, et al. Nutcracker syndrome: diagnosis and therapy. Cardiovasc Diagn Ther. 2021 Oct;11(5):1140-1149].**

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### STATEMENT FOR PUBLIC EVIDENCE-BASED AWARENESS

DOMAIN 05, Statement 09

**“Left flank or pelvic pain and blood in the urine can be caused by an anatomical compression of the left renal vein (Nutcracker syndrome): the management of the condition requires a highly specialized center”**

### SELECTED REFERENCES

1. Orczyk K, Wysiadecki G, Majos A, et al. What Each Clinical Anatomist Has to Know about Left Renal Vein Entrapment Syndrome (Nutcracker Syndrome): A Review of the Most Important Findings. *Biomed Res Int.* 2017;2017:1746570
2. Barge TF, Uberoi R. Symptomatic pelvic venous insufficiency: a review of the current controversies in pathophysiology, diagnosis, and management. *Clin Radiol.* 2022 Jun;77(6):409-417
3. Chait J, Sen I, Kalra M. Nutcracker Syndrome: How to Diagnose It and When/How Should It Be Treated in the Pelvic Venous Disease Population. *Tech Vasc Interv Radiol.* 2021 Mar;24(1):100734
4. Kolber MK, Cui Z, Chen CK, et al. Nutcracker syndrome: diagnosis and therapy. *Cardiovasc Diagn Ther.* 2021 Oct;11(5):1140-1149

### identified LITERATURE BIAS

Lack of correlation between clinical and hemodynamic findings with possible heterogeneous study populations

### SUGGESTED NEXT LINES OF RESEARCH

Long term head to head comparison among the different treatment options