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Neurosurgical implications of the Jugular Vein Nutcracker

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Abstract

In the last ten years, a new variant of Eagle Syndrome is emerging and being described: Styloid Jugular Nutcracker (SJN). In SJN, an elongated or vertically directed styloid process causes jugular vein stenosis by compressing the vein against the arch of C1. The clinical consequences appear to be various and misunderstood, ascribable mainly to venous flow impairment and consequent intracranial hypertension. The aim of this paper is to create an overview of Jugular Vein Nutcracker and to focus on its neurosurgical implications. A PRISMA-based literature search was performed to select the most relevant papers on the topic and to realize a mini-review. Future searches in the neurosurgical field should focus on collecting data about further causes of jugular stenosis compression and the association of SJN with cerebrovascular diseases. It would also be interesting to investigate the potential role of primary and secondary prevention, which is unknown so far.

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Introduction

Intracranial veins, differently from other systemic veins, lack intraluminal valves. The main cerebral venous drain for the intracranial blood in the supine position is represented by Internal Jugular Veins (IJVs).¹ A reduction of jugular vein blood flow could potentially cause intracranial venous hypertension with a subsequent increase of Intracranial Pressure (ICP).²

Skull content is represented by brain parenchyma, blood, and Cerebrospinal Fluid (CSF). Brain parenchyma, blood, and CSF are instead incompressible. The skull (bone and *dura* together) constitutes a non-expandable rigid container, in contrast to neck soft tissues. As a consequence, venous structures are subject to drastically different physiologic laws when flowing in neck soft tissue rather than inside the cranium. The point of transition between intracranial and extracranial veins is represented by the osteo-dural ring.³ This ring is positioned at the end of the bridging veins at the point where these veins enter the dural sinuses. The bridging veins act like a Starling resistor, as they are compressible tubes filled with fluid (blood) that are positioned inside a closed pressure chamber (the intracranial system). Therefore, from a physiologic point of view, the dural sinuses should be considered as extracranial, as they constitute a unique “rigid prolongation” of the jugular veins.⁴

Concerning Styloid Jugular Nutcracker (SJN) and its consequential intracranial hypertension, it is essential to evoke the Monro-Kellie principle included in “Studies in intracranial physiology & surgery; the third circulation, the hypophysis, the gliomas”.⁵ This book was published by Harvey Cushing in 1926 and reports a wrong interpretation of Monro’s doctrine. It includes the equation according to which the sum of the skull contents remains constant: brain volume + CSF + blood vol = K.⁶ Hence, when one of these elements changes in quantity, in order to preserve the constant, the others need to change their quantity, too. Instead, what Monro wrote in his book (1783) wasn’t that. The constant content of the intracranial system was the premise of Monro’s speculations and not the conclusion. He instead wrote “As the substance of the brain, like that of the other solids of the body, is nearly incompressible”, how can intracranial circulation occur? His answer to this question was: “while the heart is performing its systole, the arteries here, as elsewhere, may be dilating, and in the meantime, a

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quantity of blood equal to that which is dilating then, is passing out of the head by veins".⁷ Therefore, Monro stated that the venous outflow of the intracranial system is pulsatile and almost synchronous with arterial pulsations.⁸

The blood enters the skull pulsating into the arteries, the mechanical wave of pulsation is transmitted through parenchyma and CSF over the bridging veins, then the blood leaves the skull pulsating into the extracranial veins (dural sinuses).

Eagle's Syndrome (ES) refers to a rare group of neuropathic and vascular symptoms caused by abnormal elongation or angulation of the styloid process.⁹ The styloid process is a bony outgrowth of the temporal bone with a needle-like shape. It represents the point of attachment of the stylohyoid muscle, whose function is to elevate the hyoid bone. Eagle's Syndrome was first described in 1652 by Italian surgeon Pietro Marchetti.⁹ Watt Eagle in the 1940s outlined the clinical syndrome. He described a typical syndrome involving patients who had undergone tonsillectomy and whose symptoms arose during the post-tonsillectomy convalescence because of the formation of scar tissue stretching the nerve endings.¹⁰ Typical symptoms were: "constant pain or a nagging dull-ache in the pharynx, pain frequently referred to the ear, increased salivation, hesitancy and difficulty in swallowing, gagging, a sensation of a foreign body in the pharynx". Watt also described a Carotid Artery Syndrome characterized by an elongated styloid process producing pain along the carotid artery distribution.¹⁰ In this Eagle variant, the elongated styloid process impinges upon the carotid artery and associated nerve endings, causing pain and facilitating cerebrovascular complications such as cerebral ischemia.¹¹

In the last ten years, a new variant of Eagle syndrome has emerged and is being described in the literature: Styloid Jugular Nutcracker (SJN). SJN, also known as Styloidogenic-cervical spondylotic internal jugular venous compression or Styloid-induced Internal Jugular Vein (IJV) stenosis, is considered a vascular venous variant of Eagle Syndrome (Eagle Jugular Syndrome, EJS).¹² A common element with the other previously described variants consists of an elongated styloid process. In this case, the abnormal styloid process imprints the superior portion of the jugular vein (J3) and compresses it against the C1 arch, causing a jugular stenosis.

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Main clinical symptoms associated to the consequent stenosis of jugular veins are: headache, pulsatile tinnitus, dizziness and impaired cerebral parenchyma drainage.¹² SJN is usually associated with intracranial venous hypertension.

For what concerns SJN radiological diagnosis, the best strategy appears combining different radiological examinations. Computed Tomography (CT) can be used to measure the length of the styloid process, the distance between the styloid process and the C1 tubercle,¹³ and the relative position of the styloid tip along the Chamberlain line (the line which ideally connects the posterior edge of the hard plate with the opisthion).¹⁴ The application of 3-dimensional reconstruction is suggested in order to obtain more precise measurements. Venous phase of cerebral angiography (or venography, 4–8 s from the injections) represents the instrument of choice to show possible jugular stenosis. Cerebral angiography is an invasive procedure, but its employment allows to gain a differential diagnosis between anatomic variants such as sinus atresia/ hypoplasia, normal sinus filling defects caused by arachnoid granulations, or *septa*, asymmetrical drainage.³

Color Doppler Ultrasound (CDU) investigation represents a useful tool in order to assess venous hemodynamics, especially for what concerns flow velocity and flow rate of the internal jugular vein in general and in the upper part of the neck in SJN. In the more severe cases, the internal jugular vein appears empty, with no Doppler detectable flow in the lumen. CDU can also be employed during the follow-up phase of patients treated conservatively, in order to monitor any further flow variation.¹¹ Physical examination turns out to be negative in almost all cases.

With its constellation of symptoms, Eagle syndrome and especially SJN require the involvement of different medical figures for their diagnosis and appropriate treatment. The aim of this mini-review is to create an overview of the Jugular Vein Nutcracker and mainly to focus on its neurosurgical implications.

Materials and Methods

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A literature search was performed in order to select and analyze the main articles concerning Styloid Jugular Nutcracker and in particular its neurosurgical implications in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) statement. The electronic database PubMed (Medline), was searched using the following Medical Subject Headings (MeSH) and keywords: “Styloid Jugular Nutcracker”, “Eagle Jugular Syndrome”, “Neurosurgery Eagle Syndrome” and “Neurosurgery Jugular Nutcracker”. Only English articles were included. No time limits were settled.

Results

Pubmed research with previously illustrated keywords identified 76 studies overall. Before screening, 15 duplicated records were removed. Twenty-nine papers were excluded after title-only selection, 18 after abstract reading and further 5 articles after full article reading, as all of these papers were not pertinent with the aims and scopes of our research. Finally, 9 studies were included in this mini-review (Table 1).

The paper selection process is shown in Figure 1. The main included articles are shown in Table 1.

Discussion

Styloid Jugular Nutcracker (SJN) comprises a heterogeneous ensemble of signs and symptoms. The possible consequences of this syndrome are various, too, and involve different specialists, such as otorhinolaryngologists and brain and vascular surgeons, often demanding a multidisciplinary approach.

The aim of this paper is to focus on those SJN aspects which are of neurosurgical interest.

SJN, as already explained, can be considered as the jugular variant of Eagle syndrome. It is generally characterized by the compression of jugular vein between the arch of C1 and the styloid process of the temporal bone. The cause of this compression was identified as an

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abnormal length of the styloid bone, which results elongated in most cases. Nevertheless, a recent study¹² considered the spatial orientation of the styloid process, hypothesizing its pathogenic role in SJN. This study concluded that a more vertically directed styloid is even more significant than the absolute distance between the styloid process and the C1 arch in SJN pathogenesis. An elongated or vertical styloid process compresses the jugular vein against the arch of C1, causing jugular stenosis. The most commonly involved jugular segment is J3, and in more than 50% of the patients the stenosis is bilateral.

The more frequent consequence of jugular stenosis is reported in the literature to be intracranial hypertension related to an impaired central venous system outflow.

The most frequent intracranial hypertension correlated symptoms are: headache (46.3%), tinnitus (43.6%), insomnia (39.6%), visual disturbances (28.9%), hearing impairment (24.2%).³ Headache indicative of an increase of the intracranial pressure is generally described as diffuse, and it often progresses in severity over days to weeks.¹⁵ Papilledema is typical of intracranial hypertension because of sixth nerve palsy.¹⁶ In the literature, the association between SJN and intracranial hypertension is proved by some case reports. Xiong *et al.*¹⁷ describe the case of a young man presenting with a constant non-throbbing headache, episodes of blurred vision, and tinnitus. Opening pressure at a lumbar puncture was 260 mmH₂O. A cranial and cervical computed tomography venography documented a bilateral jugular venous occlusion caused by bilaterally elongated styloid processes. The treatment of choice was styloidecotmy and C1 tuberculectomy via transcervical approach, after which the patient recovered completely. Dashti *et al.*¹⁸ reported a similar case, definitively solved with surgical approach by drilling the styloid process. Zhao *et al.*¹⁹ retrospectively recorded data from 10 patients with Styloidogenic Jugular Venous Compression Syndrome (SJVCS) presenting with headache, compared to a control group. Seven of SJVCS patients referred headache aggravation with neck flexion, 5 blurry vision, and just 1 had nausea and vomiting. Venography with manometry in the SJVCS group documented a significantly higher global venous pressure and a higher pressure gradient across the stenosis site than controls (mean [SD] pressure, 2.86 [2.61] vs 0.13 [1.09] cm H₂O; p=0.09). Neck CT with 3-dimensional reconstruction showed

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that the space between the styloid process and the C1 lateral tubercle was much narrower during neck flexion than during neck extension. 9 of the SJVCS patients underwent decompression with a C1 tuberclectomy and a styloidectomy. All of them recovered with postoperative improvement or resolution of symptoms; one patient experienced transient postoperative dysphagia and facial drooping, and another patient referred jaw numbness. Similar cases were described by Mejia-Vergara *et al.*²⁰ and Werheim *et al.*²¹ Both describe examples of styloidectomy successfully employed: the first concerns the resolution of papilledema, while the second presents the case of a patient fully recovered from headache, retro-orbital, ear and neck pain, also suggesting to include SJN when dealing with the differential diagnosis of unexplained neurological symptoms. The increase of ICP and the onset of associated clinical symptoms require an accurate differential diagnosis. SJN is a rare but potential cause of ICP increase because of jugular stenosis and should not be ignored in the diagnostic process. CT venography represents a useful diagnostic tool. The treatment of choice for vascular stenosis is typically endovascular stenting (with or without angioplasty), but this approach is ineffective in this scenario. As the cases reported demonstrate, the surgical approach with styloidectomy and C1 tuberclectomy allows to achieve a long-term improvement or even complete resolution of symptoms.

Sinus thrombosis is reported in literature to be sometimes associated with SJN, too. Dashti *et al.*¹⁸ described the case of a 39-year-old man presenting with a severe headache who was found to have superior sagittal sinus thrombosis in a clinical picture of repeated venous thrombosis. CT venography documented a right internal jugular vein stenosis as the result of extrinsic compression between the right styloid process and the lateral tubercle of C1. After lateral neck dissection for removal of the styloid process and the lateral tubercle of C1, the symptomatology was solved completely. In this case, SJN was considered the anatomic factor that likely predisposed the patient to repeat venous thrombosis. Zhang *et al.*²² described a SJN case associated with sinus thrombosis: a 15-year-old patient with a 2-month history of headache, and papilledema. Lumbar puncture indicated intracranial hypertension (330 mm H₂O), and contrast-magnetic resonance venography diagnosed a left transverse-sigmoid sinus thrombosis, CT venography showed that the left jugular vein was compressed by the styloid process. SJN

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should be taken into account also when dealing with cases of cerebral venous sinus thrombosis of unknown origin.

Intracranial hypertension is the most frequent, but not the only, consequence of SJN involving neurological surgeons. As already discussed, also sinus thrombosis could be associated with a diagnosis of SJN. Recent evidence suggests that SJN complications could also include hemorrhagic ones. Scerrati *et al.*²³ investigated the correlation between jugular stenosis and the occurrence of Non-Aneurysmal Subarachnoid Hemorrhage (na-SAH). The rationale of this hypothesis is that jugular stenosis due to SJN causes an impaired venous outflow and it could result in an engorgement of the upstream intracranial veins with transient hypertensive phases facilitating ruptures. A direct or indirect association between systemic venous hypertension and the risk of na-SAH is reported in the literature.^{24,25} This multicenter retrospective study screened 94 patients suffering from na-SAH for eventual stenosis of the internal jugular veins by CT angiography with 3D reconstructions. A significant jugular stenosis was found in 49 (52.1%) of them; the difference with the 100-patients control group resulted in statistically significant ($p < 0.0001$).

As already debated, little is known about Eagle syndrome's variants, in particular, the most recently emerged SJN; their clinical impact on neurosurgical procedures, the best therapeutic approach, and elements which have a role in their pathogenesis should be further investigated. Indeed, evidence about other causes of jugular stenosis causing Eagle syndrome can be found in the literature. For example, De Bonis *et al.*²⁶ described the case of a 63-year-old woman with signs and symptoms of intracranial hypertension (headache and mild bilateral papilledema) and brain Magnetic Resonance Imaging (MRI) evidence of hydrocephalus (Evans index 0.30 vs 0.27 in 2009). A bilateral external compression of the omohyoid muscle on the internal jugular veins was evaluated with intra- and extracranial Venous Doppler ultrasound. Changing posture, turning the head, and/or changing from supine to upright, as well as the activation of the thoracic pump didn't modify the compression. The patient underwent surgical transection of the omohyoid muscle, with the consequent normalization of Intracranial Pressure (ICP) value (6 mmHg) and of ICP waveform. She recovered from all symptoms, and at a 2-year-follow up

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she was still asymptomatic. The compression of the jugular vein between elongated styloid process and omohyoid muscle causing intracranial hypertension and hydrocephalus was denominated JEDI (jugular entrapment, dilated ventricles, intracranial hypertension) syndrome. Nonaka *et al.*²⁷ reported the case of a 68-year-old woman who underwent microvascular decompression for hemifacial spasm of the left side. She developed delayed postoperative subdural hematoma in the left cerebellopontine angle with hydrocephalus and ipsilateral internal jugular vein stenosis between the elongated styloid process and the muscle bundle of the *rectus capitis lateralis* with antero-flexion neck position, which would induce venous congestion in addition to surgical disruption of emissary vein. No surgical treatment was required because the hematoma and the hydrocephalus were solved gradually under management with appropriate neck positioning. Until now, jugular compression between an elongated styloid process and C1 arch have been recognized, but the eventual and emerging role of other elements in jugular vein compression and stenosis, such as muscles, has not been reported in literature yet, except for a few case reports.

While the carotid variant of Eagle syndrome may cause cerebrovascular events, the possibility that SJN does the same is still a questioning. Scerrati *et al.*²³ and Nonaka *et al.*²⁷ explained proves about the plausible association between SJN and hemorrhagic complications, in particular non-aneurysmal subarachnoid hemorrhage and subdural hematoma, respectively. Anyway, larger studies are necessary to confirm these data.

As a later hint for future researches, Nastro Siniscalchi *et al.*²⁸ wrote a letter discussing the case report by Zhang *et al.*²² conjecturing a role of SJN in pulmonary embolism: “it is well known that prolonged venous compression can lead to the development of Deep Venous Thrombosis (DVT)”. Thrombosis of the subclavian vein at the costoclavicular junctions, also known as Paget-Schroetter Syndrome, is caused by intermittent venous compression due to muscular stretch in the thoracic outlet and may potentially result in pulmonary embolism initially classified as of unknown origin.²⁹ Similarly, it has already been demonstrated that compression of the jugular vein caused by an elongated styloid process may result in jugular thrombosis. In

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light of such evidence, it is reasonable to hypothesize that jugular vein thrombosis may also lead to pulmonary embolism”.

Conclusions

To conclude, SJN is a rare pathologic entity but is accompanied by annoying symptoms and potentially life-threatening complications. Scientific literature about this topic is heterogeneous and poor and lacks sufficient data. Gaining a correct diagnosis is tough, requires time and often a quagmire of radiological exams. Nevertheless, available scientific literature demonstrates that a correct diagnosis, together with an appropriate treatment, leads to symptom relief, improving life's quality. Future searches in the neurosurgical field should focus on collecting data about further causes of jugular stenosis compression and the association of SJN with cerebrovascular diseases such as non-aneurysmal subarachnoid and subdural hemorrhage or hydrocephalus. It would also be interesting to investigate the potential role of primary and secondary prevention, which is unknown so far.

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First author	Year	Type of paper
Dashti S. R.	2012	Case report
Zhao X.	2019	Case series
Zhang F. L.	2019	Case report
De Bonis P.	2019	Case report
Nonaka T.	2021	Case report
Mejia-Vergara A. J.	2022	Case report and review
Scerrati A.	2022	Retrospective observational
Werheim E.	2023	Case report
Xiong L.	2023	Case report

Table 1. Main selected papers.

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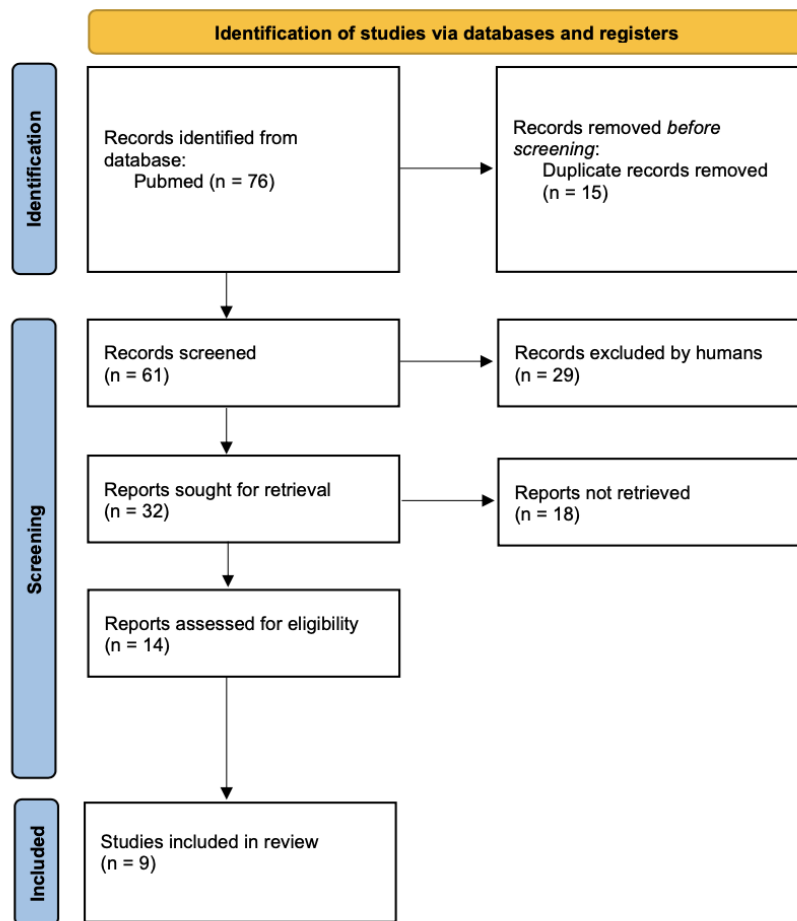


Figure 1. PRISMA Flow Chart.

From: Page MJ, McKenzie JE, Bossuyt PM, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. *BMJ* 2021;372:n71.

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