

IJV Decompression in Eagle Syndrome

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Criteria

Internal jugular vein (IJV) decompression surgery is considered medically necessary to treat IJV stenosis from external compression in the setting of **all** of the following criteria:

- **Symptoms:** At least one of the following persistent or recurrent symptoms is present and clinically attributable to impaired cerebral venous outflow:
 - Headache, head pressure, or positional headache
 - Visual disturbance or blurry vision
 - Pulsatile tinnitus or head noise
 - Dizziness or disequilibrium
 - Cognitive slowing or brain fog
- **Objective clinical findings:** At least one of the following objective clinical or examination findings is documented:
 - Papilledema
 - Visual field defect
 - Sixth cranial nerve (abducens) palsy
 - Pain reproduced with palpation of the neck or tonsillar fossa
 - Reproduction of symptoms with head rotation or neck extension
 - Intracranial venous hypertension documented by elevated intracranial pressure (ICP) on lumbar puncture of >25cm H₂O
- **Conservative therapy:** Conservative therapy appropriate to the clinical presentation has been ineffective, contraindicated, or not tolerated. (Conservative therapy may include lifestyle modification, NSAIDs, CSF pressure-lowering medication such as acetazolamide or topiramate, or anticoagulation when clinically indicated).
- **Diagnostic testing:** Diagnostic testing confirms all of the following:
 - Imaging demonstrates an elongated styloid process 3cm or more
 - CT, CTV, or MRV demonstrates IJV compression between the styloid process and the C1 transverse process or lateral tubercle **and** at least **one** of the following:
 - Focal narrowing or stenosis of the IJV at the level of the styloid process
 - Prominent collateral veins or abnormal venous drainage patterns
 - Reproducible pressure gradient across the compressed IJV segment
 - Elevated proximal venous pressures
 - Elevated venous pressure with contralateral head rotation

- **Dynamic assessment with head motion:** Dynamic venography demonstrates worsening stenosis and/or symptom reproduction during head rotation, flexion, or extension, as documented by the radiology report, and there is no alternative intracranial cause (e.g., transverse or sigmoid sinus stenosis) that fully explains the clinical presentation.

Not Medically Necessary

Open or endovascular intervention of the internal jugular vein as a primary therapy or in conjunction with styloidectomy or surgical decompression is considered **not** medically necessary for treatment of stylo-jugular venous compression syndrome, including Eagle syndrome-related internal jugular vein compression, because the evidence is insufficient to establish safety, effectiveness, and long-term benefit.

Background and Supporting Information

Surgical interventions included in IJV decompression

In Eagle syndrome with internal jugular vein compression, IJV decompression is used to treat venous outflow obstruction due to bony IJV compression caused by the elongated styloid and the transverse process of the C1 vertebrae and includes a combination of the following:

- styloidectomy
- IJV fasciotomy
- IJV repositioning
- C1 transverse process resection

Eagle syndrome

Eagle syndrome is a rare condition associated with an elongated styloid process and/or ossification or calcification of the stylohyoid ligament. The styloid process is generally considered elongated when it exceeds approximately 3cm, although the literature emphasizes that length alone is not sufficient to establish clinical significance because many individuals with elongated styloid processes are asymptomatic. Classic Eagle syndrome has been described as a pain syndrome involving the head, neck, pharynx, and ear. Other recent literature describes vascular variants in which the elongated styloid process compresses adjacent vascular structures, including the internal carotid artery or the internal jugular vein (Prasad et al., 2002; Swanson et al., 2023).

Classic Eagle syndrome typically results from irritation or compression of neural and soft tissue structures near the styloid process. Commonly reported symptoms include cervicofacial pain, throat pain, foreign body sensation in the throat, dysphagia,

odynophagia, otalgia, painful head movement, and neck pain. Symptoms may be provoked or worsened by swallowing, yawning, jaw movement, palpation of the tonsillar fossa, or head rotation. Diagnosis is usually based on a combination of compatible symptoms, physical examination findings, and imaging.

Treatment for classic Eagle syndrome may be conservative or surgical depending on symptom severity. Conservative options described in the literature include analgesics, anti-inflammatory medications, neuropathic pain medications, local anesthetic or steroid injections, and physical therapy. When symptoms are persistent or severe, styloidectomy, performed via transoral or transcervical approaches, is generally described as the definitive treatment Swanson et al., 2023).

Eagle syndrome with internal jugular vein compression

In Eagle syndrome with internal jugular vein compression, also referred to in the literature as styloidogenic jugular venous compression syndrome, the internal jugular vein is externally compressed, most often at the high cervical level, between the styloid process anteriorly and the C1 transverse process or lateral tubercle posteriorly. This variant is more often associated with symptoms suggestive of impaired cerebral venous outflow or intracranial venous hypertension. Reported symptoms include headache, head pressure, positional headache, pulsatile tinnitus or head noise, visual disturbance, papilledema, dizziness, disequilibrium, insomnia, cognitive slowing or brain fog, hearing impairment, neck discomfort, and symptoms worsened by head rotation or neck positioning. Literature states compression may be dynamic, meaning the degree of IJV narrowing can change with head rotation, flexion, or extension, emphasizing that neutral-position imaging may underestimate obstruction in some individuals (Scerrati et al., 2021; Zamboni et al., 2019; Zhao et al., 2019).

Across the literature, recurrent imaging findings demonstrate high-cervical IJV compression between an elongated styloid process and C1, producing focal IJV stenosis, often with collateral venous drainage and dynamic worsening on provocative head positioning. Commonly used imaging modalities include CT venography, CT angiography with venous phase imaging, MR venography, catheter venography and venous manometry. Dynamic imaging and manometry are supportive tools because they can demonstrate positional worsening of stenosis, symptom reproduction, altered flow, or pressure gradients across the compressed segment. However, the literature does not establish one universally accepted stenosis percentage or pressure-gradient cutoff for diagnosis (Bai et al., 2020; Leung et al., 2024; Li et al., 2019; Scerrati et al., 2021).

Treatment for Eagle syndrome with internal jugular vein compression depends on symptom severity such as the presence of intracranial hypertension, collateral drainage, and the anatomy of compression. Approaches include conservative management, anticoagulation when thrombosis is present, intracranial-pressure-

lowering medications, and surgical decompression. Surgical approaches include styloidectomy, C1 transverse process resection or tuberculectomy, IJV fasciotomy, IJV repositioning, division of compressive soft tissue or muscle structures, and combined decompression procedures. Combined approaches are indicated needed when the compression is caused by both the styloid process and C1 anatomy (Scerrati et al., 2021; Zhao et al., 2019).

The evidence base is limited for surgical management. Most published data consist of case reports, small case series, retrospective reviews, and observational cohorts with no robust randomized trials, and no universally accepted diagnostic or treatment guidelines. Reported outcomes suggest that selected patients may improve after decompression, particularly when symptoms, imaging, dynamic venous findings, and anatomy are concordant (Higgins et al., 2017; Pang et al., 2022; Petersingham et al., 2025; Scerrati et al., 2021; Zhao et al., 2019).

Endovascular stenting has also been reported, but the literature raises caution that stenting alone may be ineffective or problematic when the underlying issue is persistent external bony compression. Some reports describe stenting as an adjunct after decompression or in selected residual stenosis cases, but long-term safety, durability, and patient-selection standards remain uncertain. Complications reported across surgical and endovascular literature include wound infection, hematoma, transient or permanent neurologic deficits, facial droop, dysphagia, jaw/neck/ear numbness, accessory nerve palsy, stent restenosis, stent thrombosis, migration, or fracture (Pang et al., 2022; Scerrati et al., 2021).

Evidence Discussion

Surgical interventions

A single-center observational case series conducted from 2023 to 2025 prospectively collected data from 27 adults with symptoms consistent with intracranial hypertension, radiologic evidence of internal jugular vein compression, and persistent impairment despite medical therapy. All patients underwent combined styloidectomy, internal jugular vein fasciotomy, posterior belly of digastric division, and C1 transverse process resection; no comparator group was included. Outcomes were assessed preoperatively and at 3–6 months postoperatively, with a mean follow-up of 132 ± 43 days. Significant improvements were reported in radiologic stenosis ($p < 0.0001$), headache ($p = 0.0002$), head pressure ($p = 0.0032$), brain fog ($p = 0.0008$), visual change ($p = 0.0041$), fatigue ($p = 0.0012$), tinnitus subtypes ($p < 0.05$), and overall quality of life ($p = 0.0060$). Dizziness did not significantly improve ($p = 0.6037$). Interpretation is limited by the uncontrolled single-center design, small sample size, reliance on patient-reported outcomes, incomplete postoperative quality-of-life data, and short follow-up, which limit

conclusions regarding comparative effectiveness and durability of benefit (Abeyasinghe et al., 2026).

A PRISMA-based comprehensive literature review was conducted on 13 studies (12 retrospective studies/case reports/series and 1 prospective study) on a total of 149 total patients with extracranial internal jugular venous (IJV) stenosis. There were no randomized treatment arms or formal comparator group. All patients received medical therapy. Interventions included endovascular treatment, styloidectomy, C1 tuberclectomy, or both, and combined styloidectomy plus stenting. The most frequent symptoms were headache (46.3%), tinnitus (43.6%), insomnia (39.6%) and visual disturbances (28.9%). Increased intracranial pressure (ICP) was reported in 36.2% of patients. General improvement was reported in 58 out of 80 patients (72.5%). In detail, tinnitus resolved or improved in 13 out of 15 patients (86.7%), papilledema in 17 out of 22 (77.2%) patients and visual disturbances improved in 10 out of 23 patients (43.5%). Headache and dizziness were not significantly improved overall. Post procedure imaging revealed improved stenosis in 45 out of 63 surgically treated patients (71.4%), whereas post endovascular procedure stenosis improved in 16 of 63 (25.4%). No statistical testing or p values were reported due to data heterogeneity; therefore, no comparative outcome can be interpreted as statistically significant at $p < 0.05$. Key limitations include low-quality evidence from case reports and case series, heterogeneous definitions and treatments, and incomplete individual patient data (Scerrati et al., 2021).

A 2019 retrospective case series evaluated 10 patients with styloidogenic jugular venous compression syndrome, with comparison against 11 idiopathic intracranial hypertension controls in some analyses. Inclusion included clinical symptoms, imaging-confirmed IJV compression by the styloid/C1 region, and venous pressure gradient assessment. Prior treatment included medical management and endovascular approaches. Interventions included transcervical microsurgical decompression with styloidectomy and/or C1 tuberclectomy. Surgical outcomes included 1 patient with symptoms unchanged, 9 patients reported symptom improvement of which 3 reported resolution of all symptoms and one reported return of headache after 6 months. Clinical features of SJVS patients compared with controls, SJVCS patients had lower obesity prevalence (10% vs 73%, $p < 0.01$), higher positional headache frequency (70% vs 0%, $p < 0.01$), longer styloid length (31.0 vs 19.0 mm, $p < 0.01$), shorter C1-styloid distance (2.9 vs 9.9 mm, $p < 0.01$), and higher venous pressure gradients (2.86 vs 0.13 cm H₂O, $p = 0.03$). Limitations of this review included small cohort, retrospective design, heterogeneous interventions, and limited long-term outcome data (Zhao et al., 2019).

A 2025 literature review evaluated invasive surgical management of cervical internal jugular venous compression, summarizing 14 studies and 127 surgical cases. Inclusion focused on studies reporting surgical intervention for cervical IJV compression; studies limited to medical or endovascular therapy alone were excluded. Interventions included

styloidectomy, C1 transverse process resection, digastric division, fasciotomy, stenting, and combined approaches. Outcomes included complete and partial symptom relief, radiologic decompression, and some complications. Follow-up was variable and often short-term. No formal p-value analysis was reported. Limitations included small sample sizes, case-report/case-series data, heterogeneous diagnostic and surgical criteria, limited long-term follow-up, and inconsistent complication reporting (Petersingham et al., 2025).

A 2022 narrative review including 17 studies and 300 patients evaluated surgical and endovascular interventions for intracranial hypertension associated with internal jugular vein stenosis (IJVS). 100 patients underwent surgical intervention which included styloidectomy, IJV angioplasty/stenting, C1 transverse process or lateral mass resection, mass/osteophyte resection, and myomectomy. Comparators were not controlled treatment arms. Included studies were original reports of surgical or endovascular treatment for intracranial hypertension/IJVS. Outcomes were primarily reported as symptom improvement/resolution, with some studies reporting venous pressure gradients, and imaging patency or stenosis. Follow-up was heterogeneous not standardized, ranging from early postoperative assessments to reported 1-year symptom self-evaluation and isolated longer follow-up examples. Styloidectomy was associated with symptom improvement/resolution in 49/62 patients (79%) compared with 33/51 patients (66%) undergoing angioplasty/stenting; however, no statistical testing or p-values were reported, so comparative statistical significance cannot be determined. The authors noted major limitations including heterogeneous study designs and outcome definitions, small surgical cohorts, inconsistent selection criteria, limited demographic and follow-up data, lack of randomized controlled trials, and potential publication bias, which substantially limit certainty and preclude conclusions regarding comparative effectiveness (Pang et al., 2022).

A 2020 prospective observational cohort study enrolled 46 patients with cervical spondylotic internal jugular vein stenosis (IJVS) from an initial consecutive IJVS population of 118 patients. Patients were included if they had $\geq 50\%$ IJV narrowing on MRV/CTV/CV, abnormal collateral venous drainage, CTV-confirmed compression from the C1 lateral mass with or without the styloid process, and unexplained nonfocal neurologic or related symptoms. All patients received standard medical therapy, and only 1 patient underwent decompressive bone resection with unilateral balloon dilation, without a controlled comparator. Outcomes included clinical features of head noise (33/46, 71.7%), tinnitus (32/46, 69.6%), insomnia (31/46, 67.4%), dizziness (25/46, 54.3%), hearing impairment (22/46, 47.8%), and headache (18/46, 39.1%). Imaging findings of vascular stenosis was present in 69 out of the 92 IJVs, in which, 50.7% (35/69) was caused by the compression from the transverse process of C1, 44.9% (31/69) by the compression from the transverse process of C1 combined with the styloid process, and 2.9% (2/69) by the styloid process. Only 1 (1.4%) stenotic vessel was not due to extrinsic compression. Meanwhile, 5 (7.2%) stenotic vessels

were compressed by both the surrounding bony structures and carotid arteries. In the single surgically treated patient, headache and dizziness improved and head noise resolved by 6 months, but no statistical inference can be made. Limitations include small sample size, single-center design, primarily descriptive outcomes, lack of symptom severity quantification, no control group, and only one surgically treated patient with incomplete long-term follow-up, precluding conclusions regarding efficacy or safety of IJV decompression surgery (Ding et al., 2020).

IJV stenting

The available literature consists primarily of case reports and small retrospective case series and does not provide adequate comparative evidence to establish durable clinical benefit, long-term safety, or appropriate patient selection.

A 2017 retrospective case series evaluated 29 patients who underwent 34 styloidectomies as an adjunct or alternative to jugular venous stenting for idiopathic intracranial hypertension (IIH) or other disturbances of cranial venous outflow suggests that styloidectomy may improve symptoms or facilitate jugular stenting in selected patients with suspected extracranial venous outflow obstruction. Some patients improved after styloidectomy alone, while others improved only after subsequent jugular stenting, making it difficult to determine the independent treatment effect of styloidectomy. No comparative treatment arm, statistical testing, or p-values were reported; therefore, comparative effectiveness and statistical significance cannot be established. Outcomes included symptom resolution or improvement, stent configuration, accessory nerve symptoms, complications, and need for subsequent stenting; mean follow-up was 14 months, with a range of 1 to 39 months. Reported styloidectomy complications included no procedure-related mortality. Limitations include retrospective design, small sample size, heterogeneous diagnoses and prior procedures, no control group, subjective outcome assessment, individualized treatment selection, absence of normative pressure-gradient data, and inability to isolate the effect of styloidectomy from subsequent jugular or intracranial stenting (Higgins et al., 2017).

Coding

IJV Decompression

Code	Description
21499	Unlisted musculoskeletal procedure, head

Not Medically Necessary

Code	Description
37799	Unlisted procedure, vascular surgery

References

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