The Promise, Mystery, and Perils of Stenting for Symptomatic Internal Jugular Vein Stenosis: A Case Series

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BACKGROUND AND OBJECTIVES: Cerebral venous outflow disorders (CVDs) secondary to internal jugular vein (IJV) stenosis are becoming an increasingly recognized cause of significant cognitive and functional impairment in patients. There are little published data on IJV stenting for this condition. This study aims to report on procedural success.

METHODS: A single-center retrospective analysis was performed on patients with CVD that underwent IJV stenting procedures.

RESULTS: From 2019 to 2023, 29 patients with CVD underwent a total of 33 IJV stenting procedures. Most patients (20; 69%) had an underlying connective tissue disorder diagnosis. The mean age of the included patients was 36.3 years (SD 12.4), 24 were female (82.8%), and all were Caucasian except for 2 patients (27; 93.0%). Twenty-eight procedures (85%) involved isolated IJV stenting under conscious sedation, whereas 5 procedures (15%) involved IJV stenting and concomitant transverse sinus stenting under general anesthesia. Thirteen (39%) patients underwent IJV stenting after open IJV decompression and styloidectomy. Three patients had stents placed for stenosis below the C1 tubercle, one of which was for carotid compression. Periprocedural complications occurred in 11 (33%), including intracardiac stent migration in 1 patient, temporary shoulder pain/weakness in 5 (15%), and persistent and severe shoulder pain/weakness in 2 patients (6%). Approximately 75% of patients demonstrated improvement after stenting although only 12 patients (36%) had durable improvement over a mean follow-up of 4.5 months (range 6 weeks-3.5 years).

CONCLUSION: Our experience, along with early published studies, suggests that there is significant promise to IJV revascularization techniques in these patients; however, stenting carries a high complication rate, and symptom recurrence is common. Most neurointerventionalists should *not* be performing IJV stenting unless they have experience with these patients and understand technical nuances (stent sizing, anatomy, patient selection), which can maximize benefit and minimize risk.

KEY WORDS: Cerebral venous outflow disorders, Idiopathic intracranial hypertension, Internal jugular vein, Stenting, Styloidectomy

The recognition of transverse sinus stenosis as a pathophysiological driver of intracranial venous hypertension in patients with idiopathic intracranial hypertension (IIH) has led to a dramatic shift in enthusiasm and attention surrounding IIH and its treatments.[1] The frequent use of catheter venography to evaluate for stenting candidacy has resulted in a marked increase in scientific research being published on cerebral venous anatomy, physiology, and mechanisms of disease.[2] The increasing clinical interest and willingness-to-treat cerebral venous dysfunction and resulting IIH have led to an improved understanding of associated extracranial cerebral venous outflow disorders (CVDs), which may be considered IIH-spectrum conditions. The Society of Neurointerventional Surgery recently formalized this interest by creating the Cerebral Venous and Cerebrospinal Fluid Disorders Section as a means of promoting education, research, and practice guidance on these conditions.[3]

A number of potential causative sites of extracranial venous flow impairment have been identified.[4] In our experience, the most commonly identified site of outflow impairment is in the rostral internal jugular vein (IJV) near the transverse process of C1

ABBREVIATIONS: CVD, cerebral venous outflow disorder; **IIH,** idiopathic intracranial hypertension; **IJV,** internal jugular vein; **MS,** multiple sclerosis.

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and the styloid process, often referred to as styloidogenic jugular stenosis. Patients with symptomatic IJV stenosis present similar to patients with IIH with a predominance of complaints secondary to intracranial venous congestion including headache, skull base pain such as glossopharyngeal neuralgia, swallowing and mastication dysfunction, brain fog, tinnitus, blurred vision, and sleep disorders.[5,6] Because of the recent recognition of IJV stenosis as a pathological driver of neurological symptoms, there are limited data supporting surgical or medical interventions for this condition and limited guidance in determining candidacy for these treatments. In this study, we report our experience with stenting of the jugular vein for symptomatic jugular outflow impairment and report on diagnostic strategies, results, and complications to educate neurointerventionalists regarding the complexity and danger of stenting for this condition.

METHODS

A retrospective review of all adult patients who underwent IJV stenting for symptomatic CVD was performed by the primary author from 2019 to 2023 at a single, tertiary hospital. Institutional Review Board approval was obtained for retrospective analysis; patient consent was not obtained for inclusion as per the institutional protocol for retrospective series. Demographic, radiographic, and procedural data were collected. Stenting candidacy was determined by the primary author using an unpublished standard protocol involving diagnostic cerebral arteriography, venography, and spinal puncture with fluid drainage (**Supplemental Digital Content 1**, http://links.lww.com/NEU/E140). IJV provocative testing is performed in all patients. Treatment outcomes were assessed based on patient self-report documentation of qualitative improvement, stability (no change), or worsening at last follow-up. Because most included patients travel long distances for this treatment, most of the follow-up data are from patient self-report through telephone visits.

RESULTS

From 2019 to 2023, 29 patients with CVD underwent a total of 33 IJV stenting procedures by the primary author (Table). Most of the included patients (20; 69%) had either an underlying diagnosis of Ehlers-Danlos syndrome or other connective tissue disorder or were believed to carry a connective tissue disorder diagnosis by referring physicians. The mean age of the included patients was 36.3 years (SD 12.4), 24 were female (82.8%), and all were Caucasian except for 2 patients (27; 93.0%)

Twenty-eight procedures (85%) involved isolated IJV stenting under conscious sedation, whereas 5 procedures (15%) involved IJV stenting and concomitant transverse sinus stenting under general anesthesia. Thirteen (39%) patients underwent IJV stenting after open IJV decompression and styloidectomy. Three patients had stents placed for stenosis below the C1 tubercle, one of which was for carotid compression (Figure 1A-1C). All stents were Cordis Pro Rx Precise carotid stents (Miami Lakes, FL) placed using an 0.070-inch guide catheter positioned in the IJV through 6 F femoral vein access under systemic heparinization and dual antiplatelet therapy. All but one patient had a single stent placed; the one patient had migration of the stent caudally immediately on deployment, necessitating placement of a larger stent to fix the smaller stent in position. No other intraoperative complications were identified.

Nearly all patients noted temporary ipsilateral neck or ear pain and fullness and swallowing discomfort. Periprocedural complications occurred in 11 (33%), including intracardiac stent migration in 1 patient, temporary shoulder pain/weakness in 5 (15%), and persistent and severe shoulder pain/weakness in 2 patients (6%), of which 1 required rescue styloidectomy. Approximately 75% of patients demonstrated improvement after stenting although only 12 patients (36%) had durable improvement over a mean follow-up of 4.5 months (range 6 weeks-3.5 years), and over one-third of patients underwent either contralateral styloidectomy or stenting because of ongoing contralateral symptoms (most commonly pulsatile tinnitus, neck or head pain) but noted improvements in ipsilateral symptoms. Importantly, just over one-third of patients (39%) reported either no symptomatic improvement or worsening after IJV stenting in this series.

Complication Example Case 1

A female patient in her 20s with chronic headaches was found to have severe right transverse sinus stenosis with a 9-mm Hg gradient and bilateral IJV stenosis near C2–3 with a 5-mm Hg gradient on both sides on awake venography (Figure 2A-2D). She underwent right transverse sinus stenting with right IJV stenting under anesthesia without complication. Roughly 8 hours after the procedure, she developed severe chest pain. Computed tomography (CT) of the chest and echocardiogram demonstrated stent migration out of the neck and into the right ventricle. The patient underwent successful stent removal by interventional cardiology. The patient experienced several weeks of shortness of breath and chest pain, which improved over time. CT of the chest demonstrated a small pulmonary embolism and few tiny fragments of stent in the lungs and along the right ventricle wall.

Complication Example Case 2

A nonobese male patient in his 20s with chronic headaches, brain fog, dizziness, and left-sided jaw pain underwent venography, demonstrating stenosis of the dominant left IJV with a 4-mm Hg gradient, collateralization to the contralateral IJV and dilated suboccipital venous collaterals, and positive provocative testing (Figure 3A-3E). No intracranial gradients were present. CT confirmed stenosis at C1 near the styloid process and C1 tubercle but with space to accommodate a stent. The patient underwent stenting, resulting in relief of the gradient and reduction in collateral flow. Roughly 1 week after the procedure, the patient developed left shoulder pain and weakness, which progressed to severe shoulder adduction weakness consistent with severe spinal accessory nerve palsy. The patient was tried on

TABLE. Radiographic and Procedural Specifications for the Included Patients

Factor	N (%) or mean (SD)
Sex	
Female	24 (82.8%)
Male	5 (17.2%)
Age (y)	33.2 (12.7)
Opening pressure (cm of water)	22.2 (8.2)
Stented side (N = 29)	
Right IJV	13 (45%)
Left IJV	12 (41%)
Bilateral IJV (all performed at least 6 weeks apart)	4 (14%)
Location of stent (N = 33)	
Across C1 tubercle	30 (91%)
C2–3	2 (6%)
C4–5 (carotid compression)	1 (3%)
Gradient prestent (mm Hg)	2.9 (2.2); range 0–8
Gradient poststent (mm Hg)	0.7 (1.2); range 0–4
Previous open jugular decompression/styloidectomy	13 (39%)
Temporary shoulder pain/weakness	1 (8%)
Severe antiplatelet agent allergy	1 (8%)
No previous open jugular decompression/styloidectomy	20 (61%)
Temporary shoulder pain/weakness	4 (20%)
Persistent shoulder pain/weakness	2 (10%)
Groin hematoma	1 (6%)
Stent migration from undersized stents	2 (13%)
Clinical outcome (N = 33 procedures)	
Durable improvement	12 (36%)
Temporary improvement with recurrence	8 (24%)
No improvement	12 (36%)
Symptom worsening	1 (3%)
Need for contralateral surgical treatment or stenting (N = 29)	10 (34%)

IJV, internal jugular vein.

multiple rounds of oral steroids and physical therapy; however, his pain and weakness did not improve. Four months after the stent procedure, he underwent left styloidectomy and jugular decompression. Intraoperatively, a network of large lymph nodes was discovered surrounding the IJV and was removed along with the styloid process. The patient has ongoing weakness and pain with scapular winging.

DISCUSSION

Key Findings

This is the first large series to evaluate the safety and efficacy of IJV stenting for patients with CVD. While most patients experienced benefit from stenting, complications were frequent and many patients experienced either temporary benefit with recurrence of symptoms or need for subsequent contralateral treatment.

Interpretation

Anatomic Considerations in the Treatment of IJV Stenosis

The IJV descends from the intracranial jugular bulb and is typically positioned anterolaterally to the C1 tubercle. The IJV is draped over the lateral margin of C1 such that it usually takes a slight lateral course over C1 and acquires an ellipsoid shape with the long axis of the ellipse in an oblique position, pointing posterolaterally. The vein is often constrained anterolaterally by the styloid process, which takes an anteromedial course from the skull base toward the hyoid bone.

In some instances, the styloid is immediately adjacent to the C1 tubercle; in others, the bones are widely separated or the styloid is incompletely ossified or absent. In most patients with IJV stenosis near C1, stenosis occurs because of bony constraints from C1 posteromedially and the styloid process and its associated ligaments anterolaterally. The digastric muscle may also constrain the IJV at this site. Below C1, the IJV becomes more circular but may become constrained at C2–C6 from the sternocleidomastoid anterolaterally, a posteriorly displaced or elongated posterior cornu of the hyoid bone, the scalene muscles posteromedially, the omohyoid muscle, or the carotid artery medially.

Aside from bony and muscular relations, the IJV has an intimate relationship with cranial nerves and lymphatics. Cranial nerves IX, X, XI, and XII are closely related to the IJV, and most share space within the bony confines above C2 and within the jugular foramen itself. Furthermore, there are dense lymphatics surrounding the medial, lateral, and anterior sides of the IJV. Anecdotally, swollen and compressive lymph nodes may be found at the time of IJV decompression, which may implicate an inflammatory or infectious process (COVID-19, for example) in disease pathogenesis.

In addition, unlike the anchored venous sinuses, the IJV is subject to mechanical stresses and rotational forces, which are not well understood. There are fairly robust ultrasound data demonstrating that the IJV is at its largest when supine and becomes

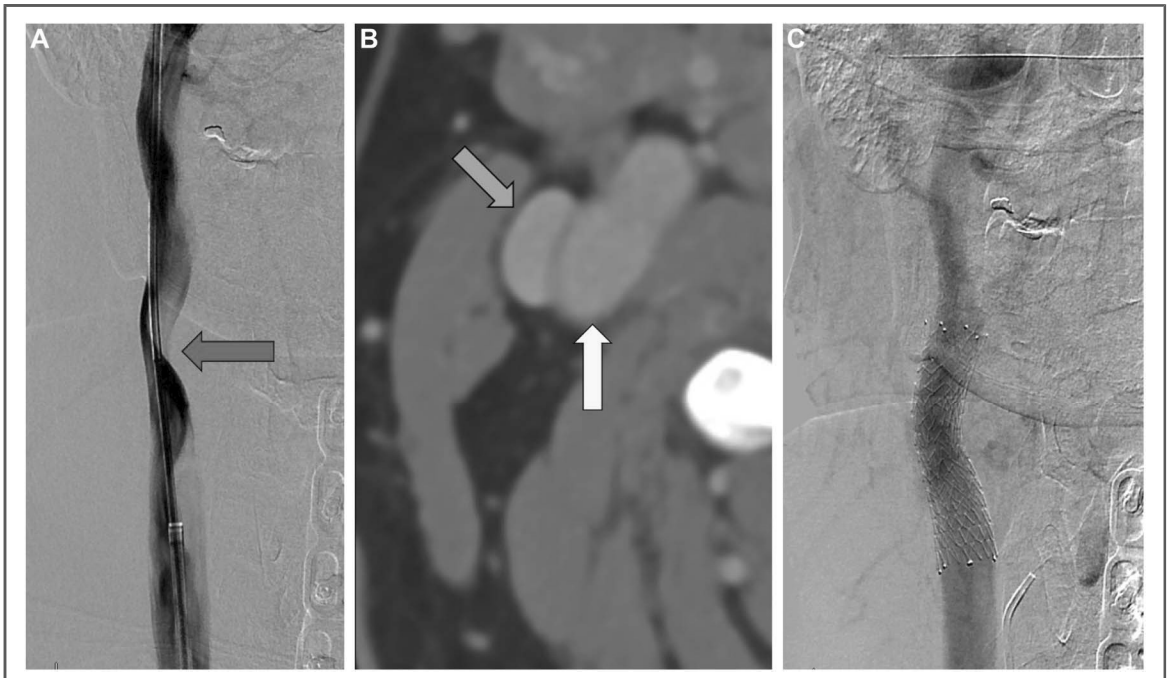

FIGURE 1. *Venography demonstrates severe stenosis of the right IJV at C4–5 with a 4-mm Hg trans-stenosis gradient (*A*, red arrow). Computed tomography confirmed extramural compression of the IJV (*B*, blue arrow) between the sternocleidomastoid muscle and the right carotid artery (yellow arrow).* C*, Stenting was performed with an oversized 12-mm stent without complication, resulting in alleviation of the gradient and significant symptomatic improvement that has persisted. IJV, internal jugular vein.*

progressively more narrowed as the patients sit upright; in the upright position, the IJV is 10% of its supine caliber, with paravertebral veins assuming most of the venous drainage.[7-9] Anecdotally, patients with symptomatic IJV stenosis often report symptom improvement while lying flat (which may mimic and be confused with spontaneous intracranial hypotension), likely because of improvement in jugular outflow in this position. Head rotation often narrows the IJV and produces symptoms. Finally, there are dense venous collaterals at the skull base, which may hypertrophy with IJV stenosis, resulting in a large network of suboccipital venous collaterals that disperse pressure through multiple pathways.

The History of Jugular Vein Stenting

There is a fascinating history regarding chronic cerebral venous insufficiency secondary to presumed jugular stenosis and its relationship to multiple sclerosis (MS), a condition of cerebral venular inflammation and demyelination. MS plaques are perivenular with a central vein, an anatomic configuration that suggests some relationship with venous physiology. Some investigators suggested a mechanistic relationship between jugular outflow impairment, the development of MS, and the frequency of MS relapses. While a strict causal association between MS and IJV stenosis has been debunked,[10] the work testing the MS jugular venous hypothesis led to a number of studies evaluating IJV venoplasty and its benefits in this patient population. Initial reports of venoplasty alone were associated with clinical benefit but were associated with very high restenosis rates.[11] In addition, complications with venoplasty were frequent, with jugular thrombosis being the most frequent severe adverse event in a series of over 400 patients.[12] One series of 38 patients having undergone IJV stenting for MS demonstrated a 12% risk of shoulder weakness, 1 patient with intracardiac stent migration requiring thoracotomy for removal, and 1 death. Interestingly, most patients reported improvement in fatigue, cognition, and heat intolerance.[13] While the relationship between MS, neurocognitive symptoms, and IJV stenosis is out of the scope of this article, the history of IJV stenting for MS does provide some important insights. First, IJV venoplasty alone has high restenosis rates; second, stenting of stenotic IJV seems to result in an improvement in general cognitive symptoms in some patients, potentially through an accidental effect on undiagnosed CVD instead of MS disease burden; and third, complications of jugular interventions can be severe and occur frequently.

Diagnosing Symptomatic IJV Stenosis

There are currently limited published data and no guidelines to assist clinicians in the diagnosis of symptomatic IJV stenosis. Adequate diagnosis is further complicated by the high prevalence of incidental IJV stenosis seen on noninvasive imaging. For instance,

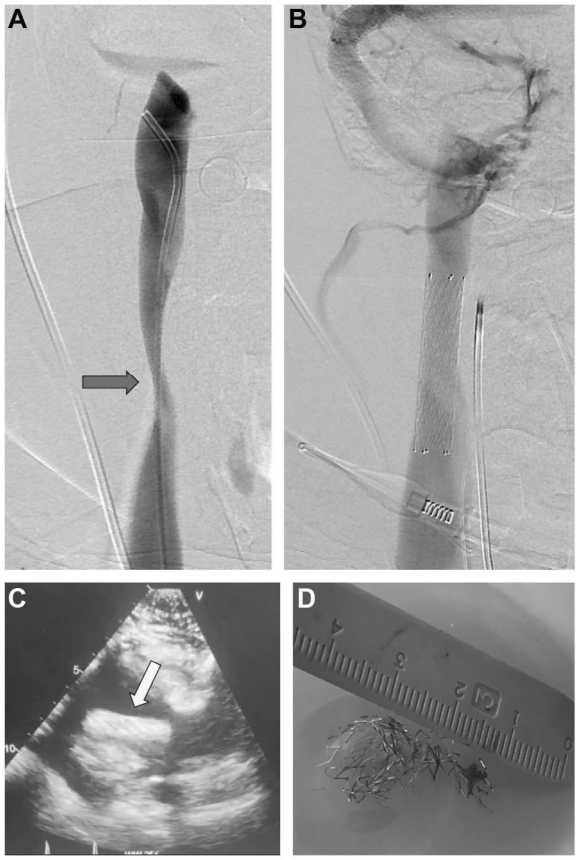

FIGURE 2. *Patient with right transverse sinus stenosis and right IJV stenosis (*A*, red arrow) with a 5-mm Hg pressure gradient in the neutral position at the time of awake venography.* B*, Right transverse sinus stenting was performed along with a single right IJV stent. After implantation, the stent seems undersized compared with the rostral and caudal segments of the vein. Echocardiogram demonstrates the stent in the right ventricle (*C*, yellow arrow).* D*, The stent was successfully snared by interventional cardiology and removed with significant difficulty. IJV, internal jugular vein.*

Jayaraman et al,[14] in a retrospective study of 108 unselected patients who underwent CT angiography of the neck for trauma or atherosclerotic disease, demonstrated that moderate or severe stenosis was present in 57% of right IJV and 44% of left IJV.

Patients presenting with IIH-like symptoms (usually in the absence of papilledema or other imaging findings of high intracranial pressure such as optic hydrops or empty sella) including pressure headache, cognitive dysfunction, memory impairment, pulsatile tinnitus, and barometric pressure sensitivity on noninvasive imaging demonstrating IJV stenosis should be considered for invasive testing. Many of these patients are believed to have a connective tissue disorder contributing to their disease pathogenesis (69% in this series). Cerebral venography and venous manometry, with evaluation from superior sagittal sinus through both transverse-sigmoid sinus and IJV pathways to the right atrium, seem to be most valuable in performing determinations. A Chinese series selected patients based on trans-stenosis IJV gradients of 5.4 cm of water or greater;[15] however, a gradient-based approach seems oversimplified in a Western population, especially as most gradients present tend to be small. Symptomatic intracranial venous stenoses generally show clear pressure gradients.[16,17] Gradients in extracranial veins are generally considerably lower,[18] even when clinical intracranial hypertension is present.

Specifications regarding venographic interpretation and patient selection are largely anecdotal and based on physician preference. In our practices, we rely on the presence of specific findings including the degree of IJV stenosis in neutral imaging, the presence of a gradient in neutral head position, the density of venous collaterals present, and findings from provocative testing. Provocative testing seems most important in determining candidacy and most useful when worsened stenosis or larger gradients are discovered in head positions that generate worsened symptoms, retrievable stents or balloon occlusion across the stenosis result in cessation of pulsatile ipsilateral tinnitus, or when strong contrast injection rostral to the stenosis acutely exacerbates and/or replicates daily symptoms.

Treatment Outcomes

There are limited published data on stenting for CVD related to symptomatic IJV stenosis, which mostly consists of small patient series or case reports. One Chinese series of 15 patients who underwent IJV stenting for IJV-induced IIH demonstrated excellent headache improvement (93%), improvements in vision (83%), and tinnitus resolution in 91%. In this series, no adverse events were reported.[15] Higgins and colleagues[19] published a series focused on rescue styloidectomy after IJV stenting for CVD in 11 patients who had stent-related complications including ongoing shoulder weakness, stent constraint from bony structures, or a combination of the two. As in our series, correction of 1 side is often followed by worsening of contralateral symptoms or stenosis, which has been reported previously.[20] Overall, the published literature suggests that there is potential for improvement with IJV stenting; however, contralateral treatment may be necessary; rescue open surgical decompression may be necessary; and the complication rate is quite high and, in fact, considerably higher than that of transverse sinus stenting for IIH.[21]

Open surgical IJV decompression, usually performed using a transcervical approach with styloidectomy and occasionally with additional C1 lateral mass resection, has a wider breadth of evidence supporting its safety given its wide acceptance as a mainstay surgical option in other styloidogenic pain syndromes unrelated to IJV compression. Overall major complications are rare[22,23] although many patients develop temporary swallowing pain or marginal mandibular facial nerve branch injury.[19] There are no large series reporting on the outcomes of styloidectomy for CVD symptoms although the authors of this commentary who perform styloidectomy procedures believe that most patients have symptomatic improvement after styloidectomy, although symptom recurrence is common leading many patients to seek stenting after decompression, and need for bilateral treatment is common (unpublished data).

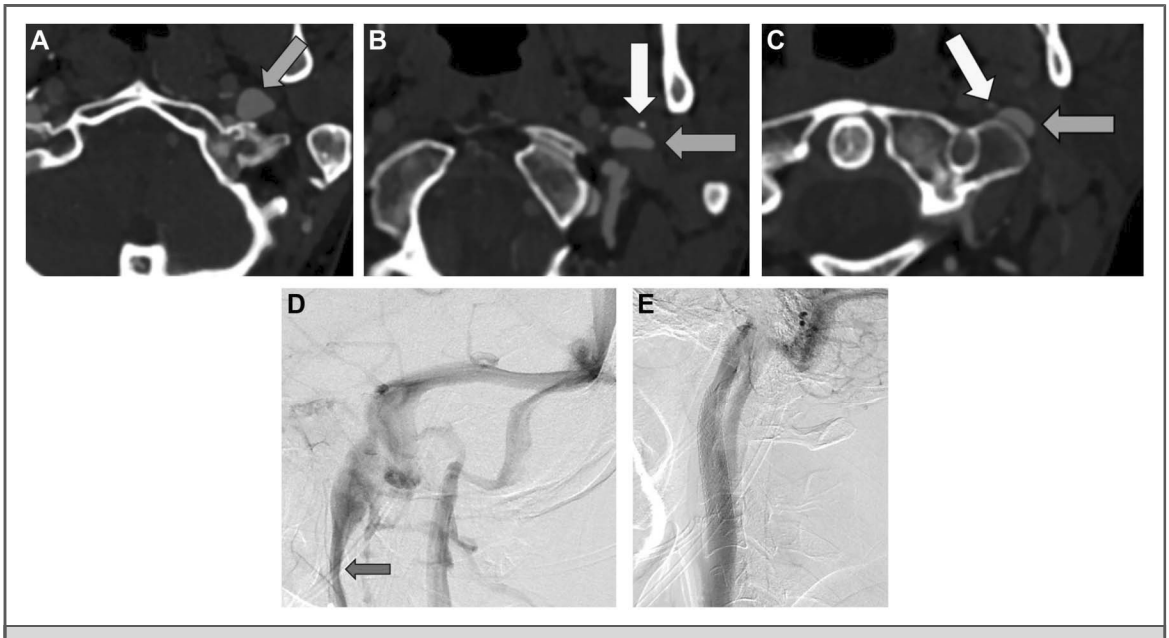

FIGURE 3. A-D, *Patient with dominant left internal jugular vein stenosis (vein shown in blue arrows) and associated 4-mm Hg pressure gradient across C1–2 (red arrow) with potential space to accommodate a stent between the styloid process (yellow arrows) and the C1 tubercle.* E, *After stenting, there is no further stenosis and reduced collateralization is present.*

Strategies for Complication Avoidance

First and foremost, IJV stenting carries significant risks and therefore should only be performed by individuals with experience with IIH and CVD after a long discussion with patients about the risks and benefits. We advise performing a cerebral venogram with attention on a complete evaluation of cervicocerebral venous outflow and the use of provocative testing to confirm symptomatic stenosis. Significant attention must be placed on the relationship between the styloid process, C1 lateral mass, and IJV. If the bones are in close proximity, the stent may be constrained, may fail to improve outflow, and may cause local pain (Figure 4A and 4B). If an open jugular decompression with styloidectomy has not been performed, there is a significant risk of shoulder weakness and pain with stenting near C1 even if the styloid seems remote from the C1 lateral mass (30% in the current series; 10% severe); this risk seems largely reduced with stenting after styloidectomy (1 patient with temporary weakness). In our practices, we have adopted a styloidectomy-first approach to mitigate this risk, with IJV stenting offered to those patients with recurrent symptoms or persistent stenosis after decompression. This strategy seems to have the best safety profile and is recommended; however, there is limited evidence supporting this strategy. In our experience, evolving to a styloidectomy-first approach has been important in not only obviating the need for stent placement in a percentage of patients but also lowering the incidence of shoulder complications in those proceeding with stent placement.

For patients where stenting is entertained, accurate sizing and site of implantation seem to be paramount. The rostral portion of the stent only needs to terminate just above the lateral mass of C1; stenting into the jugular bulb is not advised and may be implicated in persistent cranial neuropathy (Figure 4C-4E). Oversizing of the stent should be avoided when stenting in proximity to C1 to reduce the risk of cranial nerve irritation. Usually, a 6- to 8-mm stent is sufficient in this location unless the IJV is very large. By contrast, oversizing of the stent is advisable when stenting below the C1 lateral mass (at least 2 mm greater than the normal widest diameter) to prevent caudal stent migration.

Finally, managing patient expectations is important because symptomatic improvement is usually noticed after local discomfort fades over a period of 2–4 weeks, and worsened contralateral symptoms may occur because of changes in cranial venous outflow.

Limitations and Generalizability

There are a number of important limitations to the present series, including its retrospective nature and single-center, single-operator design. Patient selection is highly nuanced without supportive evidence for candidacy nor interventional technique. Most patients had connective tissue disorders with significant disease severity, which are rare and unique to the primary author's practice and may be only rarely encountered during routine clinical practice. Many patients traveled from long distances, making accurate follow-up and outcomes after stenting challenging. While validated headache and quality-of-life scores were obtained preprocedure, most patients did not complete these

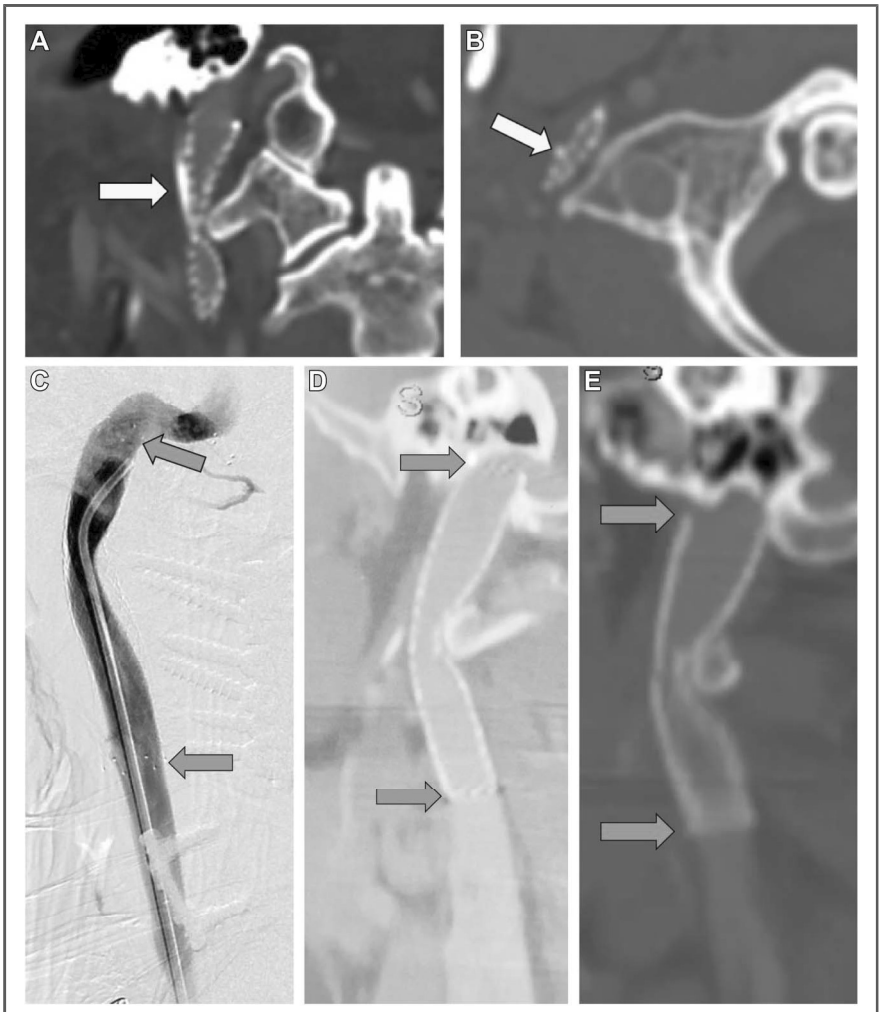

FIGURE 4. *Example patients referred to the authors after IJV stenting elsewhere with complications.* **A** *and* **B**, *A man in his 40s underwent right IJV stenting without consideration of the bony anatomy and has a severely constrained stent between C1 and the styloid process (yellow arrows) with ongoing symptoms and pressure gradient but new right neck pain after stenting. A woman in her 20s underwent left IJV stenting 6 years ago (ends of stent marked with green arrows) and has had a shoulder drop with muscular atrophy since that time with inability to lift her left arm above her head.* **C**, *She has undergone physical therapy without improvement and extensive workup for cervical radiculopathy which was negative. Two separate patients who underwent right IJV stenting with the rostral stents placed high in the jugular bulb (green arrows) with* **D**, *severe right skull base and ear pain since implantation and* **E**, *persistent spinal accessory palsy that has worsened even after rescue styloidectomy. IJV, internal jugular vein.*

postprocedure because most did not return for in-person follow-up, and therefore, these data were not available for comparison with preoperative values. There is no concise validated scale that exists that encompasses CVD symptoms of headache, cognitive dysfunction, tinnitus, dizziness, and visual symptoms to accurately study disease severity and treatment effect. As such, this study is limited by its inability to assess the degree of outcome improvement or provide commentary about individual symptoms that may be more or less amenable to stent treatment.

CONCLUSION

CVD secondary to IJV stenosis is becoming an increasingly recognized cause of significant cognitive and functional impairment in patients. Our experience, along with early published studies, suggests that there is promise to IJV revascularization techniques in these patients. IJV stenting for symptomatic stenosis may be of benefit in select patients but carries a high complication rate, which can include severe complications including persistent cranial neuropathy and intracardiac stent migration. In addition, while

most patients experience symptomatic improvement after the procedure, delayed symptom recurrence is common. Overall, these findings suggest that most neurointerventionalists should *not* be performing IJV stenting unless they have extensive experience with these patients and understand critical technical nuances. Procedural complications can be reduced through accurate patient selection, attention to patient bony and nonosseous adjacent anatomy, performing stenting only if symptoms persist after open decompression, and accurate stent sizing.

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Supplemental Digital Content 1. Supplementary Materials: Rationale and Methods for the Diagnosis and Selection of Patients with Jugular Venous Outflow Impairment for Stenting: **A**, most common symptoms of jugular outflow disorders; **B**, jugular narrowing and connective tissue disorders; **C**, making the diagnosis; **D**, comprehensive testing specifications; **E**, Patients can be segregated into 4 categories based on comprehensive testing; **F**, Treatments for patients with symptomatic internal jugular vein stenosis; **G**, Patient candidacy for jugular vein stenting.