

# Comprehensive review with pooled analysis on external and internal jugular vein aneurysm

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## ABSTRACT

**Objective:** The aim of the present comprehensive review was to present an overview of the clinical presentation and treatment options for external (EJVs) and internal jugular vein aneurysms (IJVAs) to help clinicians in evidence-based decision making.

**Methods:** A systematic literature search was conducted in accordance with the PRISMA (preferred reporting items for systematic reviews and meta-analyses) statement and included MEDLINE, Embase, Cochrane Library, Scopus, WHO (World Health Organization) trial register, [ClinicalTrials.gov](http://ClinicalTrials.gov), and the LIVIVO search portal. The inclusion criteria were studies of patients who had presented with IJVAs or EJVs. The exclusion criteria were animal and cadaver studies and reports on interventions using the healthy jugular vein for access only (ie, catheterization). Analysis of the pooled data from all eligible case reports was performed.

**Results:** From 1840 identified reports, 196 studies were eligible. A total of 256 patients with JVAs were reported, with 183 IJVAs and 73 EJVs. IJVAs were reported to occur in 66% on the right side compared with the left side ( $P = .011$ ). The patients with IJVAs were mostly children (median age, 12 years; interquartile range, 5.8-45.2 years). The patients with EJVs were young adults (median age, 30 years; interquartile range, 11.0-46.5 years). EJVs were more frequently reported in women and IJVAs in men ( $P = .008$ ). Most of the patients were asymptomatic. Pulmonary embolization in association with thrombosed EJVs was only reported for one patient. A report of the outcomes after surgery and conservative management was missing for ~50% of the patients. No relevant complications were reported after ligation of the EJVA without reconstruction. Intracranial hypertension after ligation of the right-sided IJVA was reported in three children; in one of them, a pontine infarction was observed.

**Conclusions:** JVAs are a disease of the younger population but can occur at any age. It seems to be safe to observe patients with nonthrombosed JVAs. However, in the presence of thrombus or pulmonary embolization, surgical treatment should be considered. A reconstruction technique of the IJVA with venous patency preservation should be preferred. (*J Vasc Surg Venous Lymphat Disord* 2021;■:1-8.)

**Keywords:** Aneurysm; Dilatation pathologic; Jugular veins; Systematic review

Jugular vein aneurysms (JVA) are rare. Harris<sup>1</sup> was the first to describe a venous aneurysm in 1928 in a short communication. Gillespie et al<sup>2</sup> reported the typical anatomic locations where venous aneurysms are located, including 77% in the lower extremities, 10% in the upper extremities, and 13% in the internal jugular veins (IJVs). In addition to the IJVs, aneurysms have also been reported

in the external jugular veins (EJVs). However, no reliable information is available on the prevalence of these pathologies. The diagnosis has often been suspected after a clinical examination has revealed a prominent neck swelling. The finding should usually be confirmed using duplex ultrasound. Computed tomography or magnetic resonance imaging angiography can be used to confirm the diagnosis and exclude any other neck pathologies such as malignancy.

Venous aneurysms are vascular pathologies defined as a segment of venous dilatation attached to a normal size segment. No clear diameter cutoff exists to define dilatations as aneurysms. JVAs have two types: fusiform and saccular. Fusiform aneurysms are mostly congenital and will usually present in childhood—they are also called phlebectasia.<sup>3</sup> Saccular aneurysms are mostly acquired and typically occur in adults. They can evolve spontaneously, secondarily from tumor, trauma, or inflammation, or be iatrogenic after neck surgery or catheterization.

The clinical presentation of JVAs is usually an asymptomatic and harmless swelling that increases in size with performance of the Valsalva maneuver. The

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complications of JVAs include thrombosis, thrombophlebitis, and pulmonary thromboembolism. Although the natural history of a neck vein aneurysm is considered benign, a thrombosed external JVA (EJVA) causing undetected pulmonary embolism has been previously reported.<sup>3</sup>

The management of JVA is controversial. To date, only three reviews of internal JVAs (IJVAs) have been reported. The first reported literature review of venous aneurysms was performed by Calligaro et al<sup>4</sup> in 1995. In 2018, Teter et al<sup>5</sup> analyzed venous aneurysms by their anatomic location. They could identify only four additional reports compared with the review by Calligaro et al.<sup>4,5</sup> Teter et al<sup>5</sup> reported 35 head and neck venous aneurysms, 42 thoracic venous aneurysms, 152 intra-abdominal venous aneurysms, and 279 venous aneurysms of the extremities. The literature search was conducted in three databases, and only 35 reports of IJVAs were identified. The most recent and most comprehensive systematic review on JVAs by Figueroa-Sanchez et al<sup>6</sup> identified 211 reports, of which only 97 studies were eligible.

In summary, the reported data on this rare pathology are scarce, and no clear consensus has been reached regarding when and how to treat it. This is reflected by the current treatment recommendations for patients with JVA, which have been determined from experts' opinions only (level C). Furthermore, the current clinical practice guidelines from the Society for Vascular Surgery and the European Society for Vascular Surgery have not covered the subject.<sup>7,8</sup>

We have presented a comprehensive review on the clinical presentation and management of both EJVAs and IJVAs, including all relevant databases, and an analysis of the aggregated data from all identified reports. We aimed to define the relevant characteristics of JVAs that can help in the management of this pathology. Specifically, we sought to elaborate the criteria of when and how JVAs should be treated.

## METHODS

**Search strategy and selection criteria.** For the present comprehensive review, the inclusion criteria were reports of patients who had presented with IJVAs or EJVAs. All types of studies reported in English or German language were included. The exclusion criteria were animal and cadaver studies, reports on anterior and posterior jugular vein or carotid artery, and reports on interventions using the healthy jugular vein as access only (ie, catheterization).

The search strategy was set up by a medical information specialist (H.J.) for each information source using database-specific controlled vocabulary (thesaurus terms; subject headings) and text words. The following information sources were searched on August 14, 2019 (for the entire indexing period) and on November 3, 2020 (search

update for the period from August 14, 2019 to November 3, 2020): MEDLINE via Ovid (including e-pub ahead of print, in-process, and other non-indexed citations), Medline Daily, and Ovid Medline versions (1946 to November 2, 2020); Embase via Ovid (1974 to November 2, 2020); the Cochrane Library (1996 to November 3, 2020); Scopus (1788 to November 3, 2020); and the WHO (World Health Organization) trial register, [ClinicalTrials.gov](https://www.clinicaltrials.gov), and LIVIVO search portal. In addition to the electronic database searches, the reference lists from the relevant reports were manually reviewed for additional studies. The detailed search strategies are presented in the [Appendix](#) (online only). All identified citations were imported into EndNote and duplicates removed by one of us (H.J.). Two of us (M.N. and L.M.) had independently performed the searches and selected the final studies. Conflicts were resolved by discussion between the two authors (M.N., L.M.), and the senior author (V.M.).

**Statistical analysis.** Data extraction was performed by one of us (M.N.). All the identified studies were case reports or case series. None of these studies had conducted a statistical comparison of the different treatments used for JVAs. Therefore, no formal assessment for the risk of bias (eg, due to flawed blinding, random sequence generation, or concealment) and no meta-analysis were conducted. Thus, we performed a descriptive analysis of the pooled case reports. The baseline characteristics of all eligible cases are summarized in [Table I](#). Continuous variables were summarized using the mean  $\pm$  standard deviation if normally distributed or the median and interquartile range. Explorative analyses on the proportions of the side of presentation and EJVA vs IJVA were conducted using the  $\chi^2$  test. The age at presentation between the patients with IJVAs or EJVAs was compared using the Wilcoxon rank sum test. The statistical analysis was conducted by one of us (L.M.) using R Studio, version 3.6.3 (R Foundation for Statistical Computing, Vienna, Austria). All *P* values were two-sided with an  $\alpha$  level of 5%. The present systematic review was conducted and reported in accordance with the PRISMA (preferred reporting items for systematic reviews and meta-analyses) statement and registered at OSF (available at: <https://osf.io/qktmv/>).<sup>10</sup>

## RESULTS

Of 1840 screened reports, 1034 were excluded after a review of the abstracts and 156 after the full-text reading. Thus, 196 reports met the inclusion criteria for the compound analysis. A flow diagram of study inclusion using the PRISMA guidelines is shown in [Fig 1](#). The 196 eligible studies had reported on 256 patients. Two reports had described cases of concomitant EJVAs and IJVAs. Ten studies were case series. EJVAs were described in 73 patients and IJVAs in 183 patients ([Table I](#)).

**Table I.** Baseline characteristics

Characteristic	EJV (n = 73)	IJV (n = 183)	P value
Sex			.008 <sup>a</sup>
Male	28 (39.4)	101 (58.0)	
Missing data	2 (2.7)	9 (4.9)	
Age, years			.017 <sup>b</sup>
Median (IQR)	30.0 (11.0-46.5)	12.0 (5.8-45.2)	
Missing data	2 (2.7)	7 (3.8)	
Laterality			.011 <sup>a</sup>
Right	36 (49.3)	121 (66.1)	
Bilateral	1 (1.4)	6 (3.3)	
Missing data	2 (2.7)	6 (3.3)	
Clinical presentation	73 (100.0)	183 (100.0)	.111 <sup>a</sup>
Asymptomatic	48 (65.8)	115 (62.8)	
Pain	18 (24.7)	23 (12.6)	
Voice disorder	0 (0.0) <sup>c</sup>	8 (4.4)	
Dyspnea	1 (1.4)	3 (1.6)	
Cough	0 (0.0) <sup>c</sup>	3 (1.6)	
Dysphagia	0 (0.0) <sup>c</sup>	4 (2.2)	
Other	1 (1.4)	3 (1.6)	
Missing	5 (6.8)	24 (13.1)	
Maximum diameter			NA
Median (IQR), mm	30 (30-40)	36 (28-45)	
Missing	24 (32.9)	72 (39.3)	
Personal history			NA
None, healthy	25 (34.2)	72 (39.3)	
Neck trauma	3 (4.1)	5 (2.7)	
Hypertension	2 (2.7)	6 (3.3)	
Heart disease	1 (1.4)	6 (3.3)	
Neurofibromatosis type 1	1 (1.4)	6 (3.3)	
Asthma	0 (0.0) <sup>c</sup>	4 (2.2)	
Hernia	1 (1.4)	3 (1.6)	
Menkes disease	0 (0.0) <sup>c</sup>	3 (1.6)	
Missing	34 (46.6)	70 (38.3)	

EJV, External jugular vein; IJV, internal jugular vein; IQR, interquartile range; NA, not applicable.  
Data presented as number (%), unless noted otherwise, and were complete if not stated otherwise, with percentages computed from category total.  
<sup>a</sup> $\chi^2$  test.  
<sup>b</sup>Wilcoxon rank sum test.  
<sup>c</sup>Data from Ascher et al.<sup>9</sup>

**Baseline characteristics and clinical presentation.**

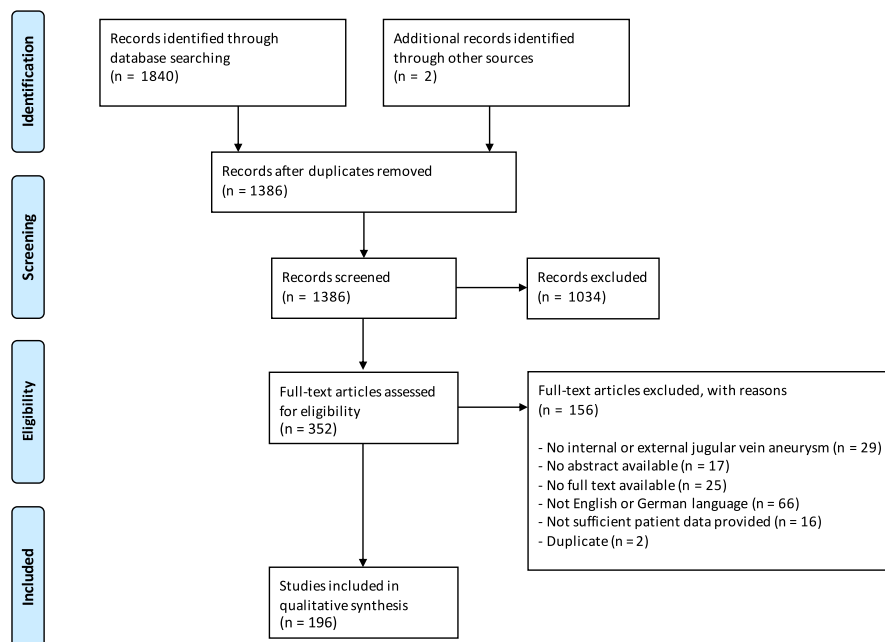
EJVs had occurred significantly more often in women and IJVs more often in men ( $P = .008$ ). JVs have been described in patients of all ages. However, the patients presenting with IJVs were significantly younger than were the patients with EJVs ( $P = .017$ ; [Table I](#), [Fig 2](#)). IJVs seem to predominantly affect children (median age, 12 years; interquartile range, 5.8-45.2 years). More than 60% of the reported cases were asymptomatic, with painless neck swelling observed. One in four

patients with an EJVA was reported to have experienced pain. In contrast, only 12.6% of the patients with an IJVA had reported pain. In contrast, voice disorders (4.4%), cough (1.6%), and dysphagia (2.2%) were only reported for patients with IJVs. EJVs were reported on both sides with the same frequency. In contrast, IJVs were seen significantly more often on the right side (121 vs 50 cases, respectively;  $P = .011$ ; [Table I](#), [Fig 3](#)).

The personal history was reported for 60% of the patients; 34.2% of the patients with EJVs and 39.3% of the patients with IJVs were otherwise healthy. A history of trauma involving the neck was reported for three patients with an EJVA (4.1%) and five patients with an IJVA (2.7%). The reported comorbidities included arterial hypertension, heart disease (ie, atrial fibrillation, coronary artery disease), neurofibromatosis type 1, and hernia repair, with asthma and Menkes disease found exclusively in patients with IJVs ([Table I](#)).

**Management and outcomes.** The treatment strategy had been reported for 69 patients (96%) with EJVs and 160 patients (87%) with IJVs. However, a clear indication for treatment had not been reported in most of the studies. More than 70% of the patients with EJVs and 50% of the patients with IJVs had undergone surgery ([Table II](#)). Ligation of the vein next to the aneurysm and excision of the pathology was the most common treatment approach and was performed in 21 patients with EJVs and 38 patients with IJVs. Preservation of the native vein was attempted in only 10 patients. Aju-luchuku et al<sup>11</sup> reported the case of a patient with EJVA who had undergone excision and direct anastomosis. Venorrhaphy was performed in six patients with an IJVA, and venorrhaphy with subsequent coating by a prosthesis was performed in three patients.<sup>12-16</sup> In 2012, Chua et al<sup>17</sup> described a hybrid approach to treat a patient with an IJVA. Two children, aged 6 and 8 years, respectively, were treated for a right-side IJVA with endoscopic transaxillary subfascial access with favorable cosmetic results.<sup>18</sup>

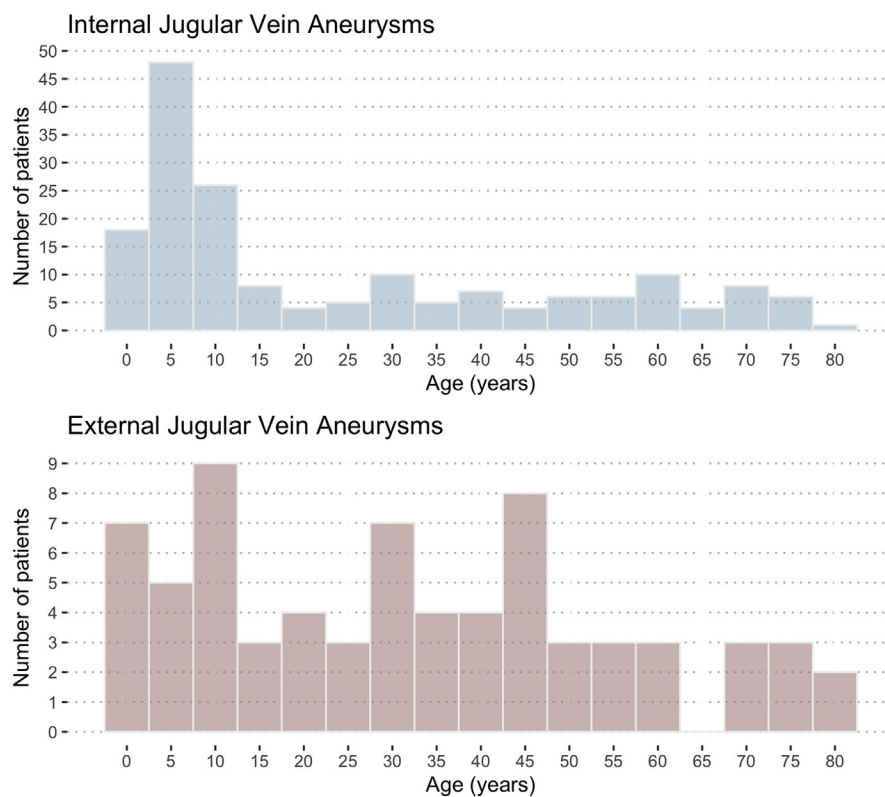
A report of the outcomes was missing for approximately one half of the patients. Therefore, no statistical analysis of the outcomes was performed, and the data were only summarized ([Table II](#)). Complications were reported for five patients. One patient had required redo surgery, and one had developed a wound infection. In addition, three children had developed intracranial hypertension after ligation of a right-sided IJVA.<sup>19</sup> In two of these children, an ipsilateral neck and craniofacial swelling appeared soon after the surgery but had resolved spontaneously within 3 days. In one child, vomiting, headache, and a pontine infarction were observed. Complete recovery was reported at 3 months after ligation of the right-sided IJV in this patient.<sup>19</sup> Only one patient with a previously undetected pulmonary embolism in association with a



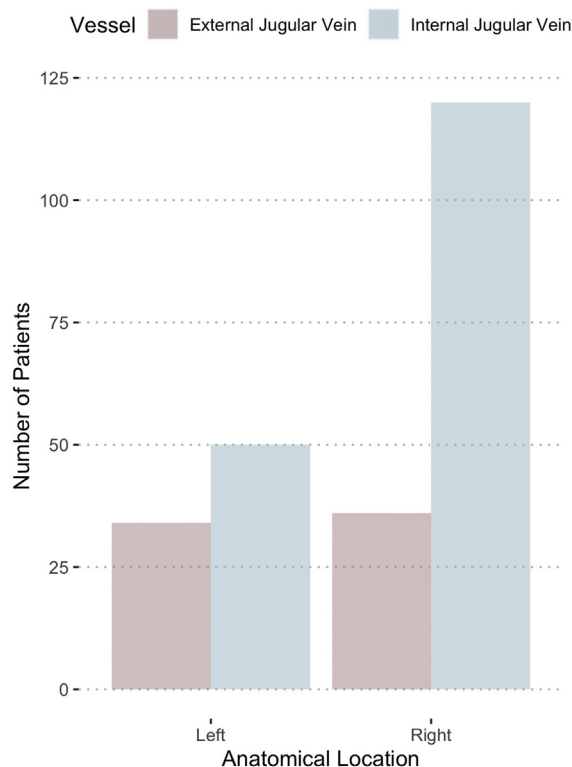
**Fig 1.** PRISMA (preferred reporting items for systematic reviews and meta-analyses) flow diagram showing inclusion and exclusion of studies.

thrombosed EJVA was reported; however, no case of pulmonary embolism was reported in association with IJVA.<sup>20</sup>

Of the patients followed up with surveillance, 24% had had a stable aneurysm diameter on the follow-up imaging studies, and 6% had had an increase in the aneurysm



**Fig 2.** Histogram of age at presentation. Information was available for 246 patients (data listed in Table I). Patients with internal jugular vein aneurysms (IJVAs) were significantly younger at diagnosis than patients with external jugular vein aneurysms (EJVAs;  $P = .015$ , Wilcoxon rank sum test).



**Fig 3.** Anatomic location. Information was available for 240 patients (missing data indicated in Table I). Internal jugular vein aneurysms (IJVAs) were reported significantly more often on the right side ( $P = .012$ ,  $\chi^2$  test).

diameter. Three patients with an EJVA and six with an IJVA had undergone surgery after an initial surveillance period. These patients had either developed symptoms during the surveillance course or had undergone surgery because of progression of the aneurysm diameter.

**Histologic findings.** The histologic findings were described in 26 reports of EJVAs and 39 reports of IJVAs. The most common finding was a true aneurysm formation, defined as attenuation of the venous wall (thinning of the elastic and muscular layers) and replacement of its structures with fibrous connective tissue, causing wall fragility in 80% of all reported cases. For EJVAs, the second most common finding was a pseudoaneurysm in four cases. In contrast, in patients with IJVAs, the second most common finding was a pathologic vessel wall structure with paucity of the muscle layer in four cases and the absence of elastic fibers in two cases. A partial occlusion by thrombus was present in five patients (19.2%) with EJVAs and seven patients (18%) with IJVA. An occluding thrombosis was only described in two patients with EJVA (Table II).

## DISCUSSION

In the present comprehensive review, we identified >250 reported cases of patients with either IJVAs or EJVAs. The number of reported cases suggests that the

IJV might be affected more often than will the EJV. Furthermore, our review has confirmed the previously reported predominance of aneurysmal disease of the right IJV compared with the left side.<sup>6</sup> However, these findings could have been biased owing to differences in reporting the frequency of these entities. The reports of cases in this field have increased during the past few years, probably owing to improved diagnostic techniques.<sup>6</sup> Nonetheless, robust epidemiologic data on the incidence and prevalence of JVAs are missing, and specific treatment guidelines do not exist.<sup>7,8</sup>

**Timing of JVA Resection.** In contrast to popliteal venous aneurysms with which pulmonary embolization occurs in 6% and rupture in 1.2%, pulmonary embolization was only reported in one patient in association with a thrombosed EJVA and not in patients with IJVAs.<sup>21</sup> No ruptures of JVAs were reported. The risk of pulmonary embolism originating from thrombus in nondilated IJVs has been reported to be <1%.<sup>9</sup> Therefore, we believe it is safe to observe patients with nonthrombosed IJVAs and EJVAs. Surgical resection should only be considered in the presence of pathologic findings such as wall thrombus in a JVA, pulmonary embolization originating from the JVA, or a complex mass with local compression symptoms. The aneurysm diameter was not an important criterion when considering the treatment strategy. The treatment indications for IJVAs in children should be determined by these morphologic findings rather than psychological concerns.<sup>19</sup>

**Optimal surgical method for JVAs.** The short-term results after surgery were not regularly reported but were favorable for most of the patients for whom the information had been provided. It seems safe to resect EJVAs without reconstruction, because we found no complications had been reported. Preservation of the EJV might not be beneficial. Unilateral ligation of the IJV without reconstruction was described in several reports. No relevant postoperative complications were reported after IJV ligation in adults. However, intracranial hypertension after ligation of right-sided IJVAs had been reported in 3 children, 1 of whom had developed a pontine infarction, in a case report series of 32 children.<sup>19</sup> Complications might occur because of insufficient ipsilateral collateral venous drainage owing to the children's age. The intraoperative occlusion test results did not predict for any potential issues that had occurred in the later course. In the case of surgical treatment of IJVAs in children, preservation of IJV patency should, therefore, be recommended. From these limited data, we would recommend that IJVAs should be treated with resection and direct anastomosis, autologous reconstruction, or venorrhaphy, rather than prosthetic reconstruction.

**Patient characteristics.** Most of the patients presenting with JVAs were reported to be otherwise healthy. The

**Table II.** Management and follow-up

Variable	EJV (n = 73)	IJV (n = 183)
Technical approach	73 (100)	183 (100)
Conservative	14 (19.2)	82 (44.8)
Open surgical repair		
Surgery (not specified)	27 (37.0)	23 (12.6)
Ligation	1 (1.4)	4 (2.2)
Ligation with excision	21 (28.8)	38 (20.8)
Excision with reconstruction	1 (1.4)	0 (0.0)
Venorrhaphy	0 (0.0)	6 (3.3)
Venorrhaphy and coating	0 (0.0)	3 (1.6)
Endovascular repair		
Endovascular (not specified)	1 (1.4)	0 (0.0)
Coiling	1 (1.4)	0 (0.0)
Hybrid (endovascular assisted)	0 (0.0)	1 (0.5)
Endoscopic resection	0 (0.0)	2 (1.1)
Missing	7 (9.6)	24 (13.1)
Follow-up time		
Median (IQR), months	8 (2.5-24)	12 (6-24)
Missing	46 (63.0)	96 (52.5)
Outcome		
Surgical repair	52 (100)	77 (100)
Improvement of symptoms	1 (1.9)	0 (0.0)
Success, no complaints	17 (32.7)	55 (71.4)
Intracranial hypertension	0 (0.0)	3 (3.9)
Repeat intervention needed	1 (1.9)	0 (0.0)
Wound infection	0 (0.0)	1 (1.3)
Not reported	31 (59.6)	18 (23.4)
Conservative	17 (100)	82 (100)
Stable	5 (29.4)	19 (23.2)
Shrinkage	1 (5.9)	5 (6.1)
Shrinkage, completely thrombosed	0 (0.0)	1 (1.2)
Increased	0 (0.0)	2 (2.4)
Surgical repair after surveillance	3 (17.6)	6 (7.3)
Not reported	8 (47.1)	49 (59.8)
Pathologic findings	26 (100)	39 (100)
Vessel wall histologic features		
Normal vein	0 (0.0)	3 (7.7)
True aneurysm	21 (80.8)	29 (74.4)
Pseudoaneurysm	4 (15.4)	0 (0.0)
Hemangioendothelioma	1 (3.9)	0 (0.0)
Neurofibroma	0 (0.0)	1 (2.6)
Paucity of muscle layer	0 (0.0)	4 (10.3)
Absence of elastic fibers	0 (0.0)	2 (5.1)

(Continued)

**Table II.** Continued.

Variable	EJV (n = 73)	IJV (n = 183)
Thrombus/embolus		
Occluded by thrombus	2 (7.7)	0 (0.0)
Partially thrombosed	5 (19.2)	7 (18)
Pulmonary embolism	1 (3.9)	0 (0.0)
No thrombus	1 (3.9)	9 (23.1)
<i>EJV</i> , External jugular vein; <i>IJV</i> , internal jugular vein; <i>IQR</i> , interquartile range. Data presented as number (%), unless noted otherwise, and were complete if not stated otherwise, with percentages computed from category total.		

coincidence of common diseases such as arterial hypertension, atrial fibrillation, and coronary artery disease was reported in several patients. A history of neck trauma was reported in three patients with EJVs and five patients with IJVs.

Seven patients had been reported to have coexisting neurofibromatosis type 1, a rare autosomal dominant disorder that affects ~1 in 3500 individuals and is associated with a predisposition to the development of both benign and malignant tumors.<sup>22</sup> This inherited disease might also affect the nervous system and cause vasculopathies. Common manifestations include renal and cerebral artery stenosis, aortic coarctation and arteriovenous malformations.<sup>22</sup> Vasculopathies are caused by increased proliferation and growth of the endothelium.<sup>23</sup> In addition to these manifestations, numerous other vasculopathies have been described. Kaas et al<sup>24</sup> reported vascular imaging data from 80 children with neurofibromatosis type 1. Moyamoya syndrome, a severe occlusive disease of the cerebral arteries that leads to the development of a collateral circulation with frail vessels prone to bleeding and thrombosis, was the most common vascular pathology, affecting 15% of the examined patients.<sup>24</sup> Involvement of the peripheral vessels was less often examined or reported. Only two children were found to have peripheral vascular disease; both children had had midaortic syndrome requiring surgical therapy.<sup>24</sup> We identified six patients with neurofibromatosis type 1 and a concomitant IJV and one patient with neurofibromatosis type 1 and a concomitant EJV. All patients were middle age (range, 47-63 years) at diagnosis. One patient with neurofibromatosis type 1 was reported to have a huge (>2000 mL in volume) aneurysm of the left IJV that was treated with proximal ligation of the IJV and central ligation of the brachiocephalic vein owing to extension of the aneurysm.<sup>25</sup> The patient developed phlebostatic edema of the left shoulder and necrosis of a neurofibroma of the left arm. Similarly, Menkes disease was diagnosed in three patients with IJVs. Menkes disease is a very rare, inherited X-linked recessive disorder

concerning copper transport throughout the body. Missing copper can affect the function of copper-dependent enzymes, which control the development of the hair, brain, bones, liver, and arteries. However, it remains unclear how this condition can lead to formation of venous aneurysms, especially aneurysms of the IJV.

The results from our systematic review have demonstrated the importance of comprehensive and standardized reporting. The cases of 256 patients with JVA had been reported. However, the lack of follow-up information complicated the interpretation of the aggregated data regarding the outcomes of different treatment approaches or conservative management. We have proposed the reporting of (1) the clinical presentation; (2) the diagnostic steps, including the imaging modality, aneurysm diameter, and presence of thrombotic material; (3) the indication for treatment; (4) the chosen treatment approach with sufficient details reported on peri- and postinterventional complications; and (5) patients' follow-up data.

In addition to the unavailability of follow-up information for a substantial proportion of the 256 patients, especially for the conservatively treated patients, our systematic review was limited by other factors. First, to the best of our knowledge, no large case series of patients with prospective follow-up data available have been reported. Second, we did not include studies in languages other than English or German. Finally, the risk of a publication bias was substantial. Investigators are more likely to submit case reports with positive outcomes, and journals are more likely to accept such reports. The vast majority of reported cases in the literature have described positive outcomes.<sup>26</sup> The proportion of patients with comparable conditions but unsuccessful treatment results remains unknown.

## CONCLUSIONS

JVAs are rare pathologies of the younger population but can occur at any age. To the best of our knowledge, information is only available from case reports. Nonoperative management seems to be safe for patients with nonthrombosed IVJAs or EJVAs. However, in the presence of thrombus, pulmonary embolization, or complex masses with local compression, surgery should be considered. A reconstruction technique of the IJVA with venous patency preservation should be preferred. In contrast, EJVAs can be simply ligated.

## AUTHOR CONTRIBUTIONS

Conception and design: MN, LM, VM

Analysis and interpretation: MN, LM, HJ, MS, JS, VM

Data collection: MN, LM

Writing the article: MN, LM

Critical revision of the article: HJ, MS, JS, VM

Final approval of the article: MN, LM, HJ, MS, JS, VM

Statistical analysis: LM

Obtained funding: Not applicable

Overall responsibility: VM

MN and LM contributed equally to this article and share co-first authorship.

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*Additional material for this article may be found online at [www.jvsvenous.org](http://www.jvsvenous.org).*



**APPENDIX (online only).****Summary of database search strategies**

Search strategies were performed on August 14, 2019 and November 3, 2020 (update).

Medline:

- 1.) exp Aneurysm
- 2.) exp Dilatation, Pathologic
- 3.) 1 or 2
- 4.) exp Jugular veins
- 5.) 3 and 4
- 6.) limit 5 to animals
- 7.) limit 6 to humans
- 8.) 6 not 7
- 9.) 5 not 8
- 10.) ((Aneurysm\* or dilatation\*) adj4 (jugular vein\* or vena jugularis\*)).tw.
- 11.) "jugular vein aneurysm\*".tw.
- 12.) "jugular venous aneurysm\*".tw.
- 13.) (Venous aneurysm\* and (jugular vein\* or vena jugularis\*)).tw.
- 14.) (Venous aneurysm\* and neck).tw.
- 15.) (Aneurysmal dilatation\* and (jugular vein\* or vena jugularis\*)).tw.
- 16.) Jugular phlebectasia.tw. (1)
- 17.) ((Neck adj3 (mass\* or lump? or swelling?)) and (jugular vein\* or vena jugularis\*)).tw.
- 18.) "EJV aneurysm\*".tw. (4)
- 19.) (EJV pseudoaneurysm\* or EJV pseudoaneurysm\*).tw.
- 20.) (Phlebectasia adj4 (jugular vein\* or vena jugularis\*)).tw.
- 21.) ((pseudoaneurysm\* or pseudo-aneurysm\*) adj3 (jugular vein\* or vena jugularis\*)).tw.
- 22.) (Neck vein\* adj3 (dilatation\* or aneurysm\*)).tw.
- 23.) (True and false aneurysm\* and (jugular vein\* or vena jugularis\*)).tw.
- 24.) (Vascular mass\* and (jugular vein\* or vena jugularis\*)).tw.
- 25.) or/10-24 26.) 9 or 25 27.) limit
- 26.) to dt="20190814-20201103"

Embase:

- 1.) exp Aneurysm/
- 2.) exp "lesions and defects"/
- 3.) 1 or 2
- 4.) exp Jugular vein/
- 5.) 3 and 4
- 6.) limit 5 to animals
- 7.) limit 6 to humans
- 8.) 6 not 7
- 9.) 5 not 8
- 10.) ((Aneurysm\* or dilatation\*) adj4 (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 11.) "jugular vein aneurysm\*".ab,kw,ti.
- 12.) "jugular venous aneurysm\*".ab,kw,ti.
- 13.) (Venous aneurysm\* and (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 14.) (Venous aneurysm\* and neck).ab,kw,ti.

- 15.) (Aneurysmal dilatation\* and (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 16.) Jugular phlebectasia.ab,kw,ti.
- 17.) ((Neck adj3 (mass\* or lump? Or swelling?)) and (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 18.) "EJV aneurysm\*".ab,kw,ti.
- 19.) ("EJV pseudoaneurysm\*" or "EJV pseudoaneurysm\*").ab,kw,ti.
- 20.) (Phlebectasia adj4 (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 21.) ((pseudoaneurysm\* or pseudo-aneurysm\*) adj3 (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 22.) (Neck vein\* adj3 (dilatation\* or aneurysm\*)).ab,kw,ti.
- 23.) (True and false aneurysm\* and (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 24.) (Vascular mass\* and (jugular vein\* or vena jugularis\*)).ab,kw,ti.
- 25.) or/10-24 26.) 9 or 25 (875) 27.) limit
- 26.) to dd="20190814-20201103"

Cochrane:

- 1.) MeSH descriptor: [Aneurysm] explode all trees
- 2.) MeSH descriptor: [Dilatation, Pathologic] explode all trees
- 3.) 1 OR 2
- 4.) MeSH descriptor: [Jugular Veins] explode all trees
- 5.) 3 AND 4
- 6.) ((Aneurysm\* OR dilatation\*) AND (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 7.) (jugular vein aneurysm\*):ti,ab,kw
- 8.) (jugular venous aneurysm\*):ti,ab,kw
- 9.) (Venous aneurysm\* AND (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 10.) (Venous aneurysm\* NEAR neck):ti,ab,kw
- 11.) (Aneurysmal dilatation\* NEAR (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 12.) (Jugular phlebectasia\*):ti,ab,kw
- 13.) ((mass\* OR lump\* OR swelling\*) AND (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 14.) (EJV aneurysm\*):ti,ab,kw
- 15.) (EJV pseudoaneurysm\* OR EJV pseudoaneurysm\*):ti,ab,kw
- 16.) (Phlebectasia NEAR (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 17.) (pseudoaneurysm\* AND jugular vein\*):ti,ab,kw
- 18.) (Neck vein\* NEAR (dilatation\* OR aneurysm\*)):ti,ab,kw
- 19.) (True and false aneurysm\* NEAR (jugular vein\* OR vena jugularis\*)):ti,ab,kw
- 20.) (Vascular mass\* AND (jugular vein\* OR jugular vein\*)):ti,ab,kw
- 21.) 6 OR 7 OR 8 OR 9 OR 10 OR 11 OR 12 OR 13 OR 14 OR 15 OR 16 OR 17 OR 18 OR 19 OR 20
- 22.) 5 OR 21
- 23.) 22 (with Cochrane Library publication date from Aug 2019 to Nov 2020)

Scopus:

((TITLE-ABS-KEY (aneurysm\* OR dilatation\*)) W/3 ((TITLE-ABS-KEY ("jugular vein\*" OR "Vena jugularis\*")))) OR (TITLE-ABS-KEY ("jugular venous

aneurysm\*") OR (TITLE-ABS-KEY ("Venous aneurysm\*" AND "jugular vein\*")) OR (TITLE-ABS-KEY ("Venous aneurysm\*" AND neck)) OR (TITLE-ABS-KEY ("Aneurysmal dilatation\*" AND "jugular vein\*")) OR (TITLE-ABS-KEY ("Jugular plebectasia\*")) OR (TITLE-ABS-KEY (neck W/3 (mass\* OR lump? OR swelling?) AND "jugular vein\*")) OR (TITLE-ABS-KEY ("EJV aneurysm\*")) OR (TITLE-ABS-KEY (phlebectasia W/3 "jugular vein\*")) OR (TITLE-ABS-KEY ("True and false aneurysm\*" AND "jugular vein\*")) OR (TITLE-ABS-KEY ("Vascular mass\*" AND jugular)) OR (TITLE-ABS-KEY ("EJV pseudoaneurysm\*" OR "EJV pseudo-aneurysm\*")) OR (TITLE-ABS-KEY (pseudoaneurysm\* OR pseudo-aneurysm\* W/3 "jugular vein\*")) OR (TITLE-ABS-KEY ("Neck vein\*" W/3 (dilatation\* OR aneurysm\*)))

AND (LIMIT-TO (PUBYEAR,2020) OR LIMIT-TO (PUBYEAR,2019))

## LIVIVO:

Jugularvenenaneurysma OR Jugularvenendilatation OR (Jugularvene\* AND Aneurysm\*) OR (Jugularvene\* AND Dilatation\*) OR ("Vena jugularis\*" AND Aneurysm\*) OR "Jugular vein aneurysm\*" OR "jugular venous aneurysm\*" OR ("jugular vein\*" AND aneurysm\*) OR ("jugular vein\*" AND dilatation\*)

## ClinicalTrials.com:

No studies were found for (jugular vein OR vena jugularis) AND (aneurysm OR dilatation)

## ICTRP database (World Health Organization):

(jugular vein OR vena jugularis) AND (aneurysm OR dilatation)