Internal jugular phlebectasia: A systematic review

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INTRODUCTION

Internal jugular phlebectasia (IJP), a nontortuous dilatation of the internal jugular vein (IJV), is typically considered a benign anatomical variant of unknown etiology. Clinically, its most common presentation is as a soft and painless mass in the lateral neck, which transiently appears in periods of increased intrathoracic pressure, such as when performing a Valsalva maneuver. At the present time, IJP treatment is not well defined. Both a conservative approach and surgical removal are described in literature. Surgery is mostly reserved for cosmetic reasons or to prevent complications such as enlargement or thrombosis of the dilatation.

In literature, multiple terms have been applied to describe IJP such as venoma, venous cyst, venous aneurysm, and venous ectasia. It was first described in a case report by Harris on a 5-month-old infant with a right-sided neck mass. The infant died during the surgery to remove the mass, and the only finding considered to explain the symptoms was a blood-filled cyst. It was controversial whether the original lesion was, in fact, IJP.
IJP was published by Zuckschwerdt, in 1929, and the anomaly was further characterized by Gerwig, in 1952.[30] Since then, there have been multiple case reports of IJP.[37,69,82] To the best of our knowledge, however, no systematic review defining the most common presentation of IJP and treatment of this anomaly can be found in literature. The aim of this study was to describe the most common clinical presentation, method of diagnosis, and treatment of IJP in adult and pediatric patients based on published data.

METHODS

Following the preferred reporting items for systematic reviews and meta-analyses (PRISMA) guidelines,[60] literature search for IJP was conducted in COCHRANE, PUBMED, EBSCOHOST, SCOPUS, OVID, and SCIELO databases up to October 11, 2018. As search terms, we used “phlebectasia” or its synonyms, “venous cyst,” “aneurysmal varix,” “venoma,” “venectasia,” “venous aneurysm,” and “venous ectasia” in the title and/or abstract, with the word “jugular” included in all fields, excluding articles that had in the title the words “external” or “anterior.” Only papers written in English or Spanish were considered for this review. The search was not limited by date of publication. This search strategy resulted in 211 unique articles. Titles and abstracts of the articles were screened. Potentially suitable studies for IJP were read in full by three independent reviewers. A total of 114 of the original articles were excluded. Defined variables (patient characteristics, clinical presentation, IJP characteristics, method of diagnosis, treatment, and outcome) were extracted independently by the reviewers and disagreements were solved by consensus. All the statistical analyses were performed using the statistical program MATLAB R2016a (Mathworks Inc.) the Chi-squared test was used for dichotomous variables and the Mann–Whitney U-test for quantitative variables, with a confidence interval of 95%. We considered \( P < 0.05 \) to be statistically significant.

RESULTS

Following PRISMA guidelines, a total of 97 articles were included in the analysis [Figure 1]. These articles covered a total of 247 patients with IJP, including both pediatric and adult patients [Supplemental Table 1]. In the analyzed papers, not all searched variables were described; therefore, the number of cases described for each variable could vary in the description and statistical analyses [Table 1].

We found that IJP predominantly affected the pediatric population, with 206 patients (83.4%) younger than 18 years and only 41 adult cases (\( P < 0.001 \)) reported in literature. The mean age of presentation was 47.8 years in adults (range: 17–74 years) and 6.4 years in pediatric patients (range: 6 weeks–15 years). The sex most affected was female in the adult group (61%) and male in the pediatric group (71%).

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In just 150 of 247 cases, the primary diagnostic study was reported. Ultrasound (US) was employed for 108 patients (72%) as the primary study, a percentage that was higher in the pediatric population compared with that in adult patients. A contrast-enhanced computed tomography (CECT) was performed on 22 patients (14.6%), venography on 8 patients (5.3%), angiography on 4 patients (2.7%), magnetic resonance imaging (MRI) on 3 patients (2%), xenography on 2 patients (1.3%), and cinefluorographic studies, plain X-ray, and scintigraphy 99mTc-fibrinogen on 1 patient each (0.7%). The most common secondary study was CECT in 25 cases, MRI in 13 cases, US in 7 cases, barium swallow in 4, and unenhanced CT in 1 (2%).

Secondary invasive studies were venography in 22 cases and angiography in 10 cases.

The treatment most frequently reported was a conservative approach in 85 patients (62%); however, the proportion of patients treated conservatively in the pediatric population was greater than that in the adult population. In the adult group for whom the treatment was described, no complications were found during conservative treatment, and complications from surgical resection were reported in 4 (11.4%) patients ($P < 0.001$): massive hemorrhage in 2 cases and paralysis of the left vocal cord and incomplete surgical resection in one case each. In the pediatric population, 102 patients had their treatment described. No complications were found during conservative treatment, and two (2%) patients had surgical complications ($P < 0.001$). The complications were subjective congestion of the left side of the head for 24 h after surgery and a postoperative transient increase in blood pressure. One patient treated conservatively died 14 months later due to Menkes disease complications.

### DISCUSSION

IJP remains an infrequently diagnosed vascular anomaly. Although it is becoming increasingly recognized, partly due to improved diagnostic techniques,[44] medical guidelines have not yet been established. Clinical decisions for the diagnosis and treatment of this condition are based almost exclusively on personal experience. In this study, we analyzed clinical data gathered from literature on 247 patients with IJP. IJP is classically found unilaterally, although a few bilateral cases have been reported. In this review, bilateral IJP was only found in a few pediatric cases. IJP is most commonly found incidentally during a physical examination as a mass in the neck that increases with common efforts such as talking, coughing, or swallowing. Other symptoms include voice alteration, paralysis of the vocal cords, and/or dysphagia, all of which are caused by the proximity of the vagus nerve and other lower cranial nerves to the IJP.[44,63,82]

Multiple pathologies affecting the neck are considered to be differential diagnoses, especially laryngocele, branchial cyst, cystic hygroma, hemangioma, and paraganglioma.[6,7]
In this review, we found some cases with initial inaccurate diagnoses that varied depending on the age group. In the adult population, four patients had other causes of the neck mass including globus pharyngeus, an infectious process, aneurysm of the subclavian artery, and an ovoid-shaped neuroma. In the pediatric patients, the erroneous diagnoses included goiter, laryngoele, adenopathy, adenoidectomy, and aerocoele.

Although the etiology of IJP is not completely understood, some authors have suggested previous direct neck injuries or medical procedures such as central venous catheterization, positive-pressure ventilation, neck surgeries, or tumors.[11,26,63,88] In this review, no definitive association with these variables was found.

Histopathological changes in those cases treated with surgical resection include a thinning of the muscular and elastic layers; however, the great majority of cases showed no significant changes compared with the normal venous wall.[17,92] Several authors have found this abnormality in patients with neurofibromatosis type 1 and in Ehlers–Danlos syndrome.[18,39,49,67] In the present review, there was no significant association with these pathologies.

IJP most commonly presents as a unilateral right-sided anomaly.[91] In this review, the right-to-left ratio was 4:1. Although no clear etiology has been elucidated for IJP, multiple hypotheses have been formulated to explain the right-sided predominance.[71] LaMonte first proposed that, given the right innominate vein is in close contact with the right apical pleura, an increase in intrathoracic pressure would be transmitted to the right IJV and thus predispose an individual to unilateral phlebectasia. The left IJV, anatomically located more medially, does not receive such stress.[54] More recently, Paleri and Gopalakrishnan hypothesized that intrathoracic pressure could be easily transmitted to the right jugular bulb due to several anatomic factors including the following: (1) the fact that the right IJV valves, which are involved in preventing retrograde blood flow, are located more cephalad than their left-sided counterparts; (2) the larger diameter of the right IJV compared with the left side; (3) the direct continuity of the superior vena cava with the right brachiocephalic vein; (4) the higher number of valves in the left brachiocephalic vein in comparison with that of the right side; and (5) the higher number of competent valves in the right subclavian vein compared with that of the left side.[71] However, not all reported cases agree with the theory proposed by Paleri,[61] and more studies are needed to fully understand the side predominance of IJP.

The image study most frequently used for the diagnosis of IJP is US, with color Doppler as the study of choice to confirm the flow. This study continues to be useful because it is safe, feasible, sensitive, and of low cost. Other studies, such as MRI or CT, are usually helpful in complementing the diagnosis. Invasive studies such as angiography and venography are less commonly used, with only 10 (4%) and 22 (9%) cases, respectively, diagnosed by such methods.

Classically, IJP has been considered a benign entity; however, a serious possible complication resulting from abnormal vascular flow is thrombosis. In this review, the presentation of this complication was more frequent in adults, with 7 (17.1%) cases reported in literature, than in pediatric patients, with only 3 (1.5%) cases. Some authors describe Horner syndrome[46] as another complication. However, no significant association with that complication was found in this study.

Conservative treatment was most frequently reported for IJP in the pediatric population, given it is classically considered a benign vascular abnormality. In adults, surgical resection was selected in almost the same proportion as conservative treatment. Alternative treatments consisted of endovascular angioplasty, surgical wrapping, and endoscopic resection, but these were only performed on the pediatric population. In general, the clinical outcome in both groups was described as good, although the follow-up was not described in a standardized manner. Descriptions of surgical treatment revealed a complication rate of 6.9% in the pediatric population and 11.4% in the adult population. In the cases treated conservatively, no complications were reported in either group. In general, conservative treatment was a safer option than surgical treatment in both groups.

### Table 2: Average IJP dimensions.

| Affected side | Adult | Pediatric |
|---------------|-------|-----------|
|               | Average | Maximum | Minimum | Average | Maximum | Minimum |
| Right sided   | Diameter  | 3.95 cm | 5.1 cm | 1.1 cm | 3.35 cm | 12 cm | 1.1 cm |
|               | Length   | 3.18 cm | 7.0 cm | 1.1 cm | 3.43 cm | 7.0 cm | 1.0 cm |
| Left sided    | Diameter  | 4.73 cm | 10 cm  | 3.0 cm | 3.59 cm | 5.5 cm | 2.0 cm |
|               | Length   | 3.98 cm | 6.9 cm | 1.5 cm | 3.96 cm | 7.0 cm | 2.8 cm |

IJP: Internal jugular phlebectasia
CONCLUSIONS

Following PRISMA guidelines, we analyzed a total of 247 patients with IJP, including pediatric and adult patients. Due to the low incidence of this abnormality, there are not enough original prospective studies to perform a meta-analysis. In the analyzed papers, not all the studied variables were described, and the heterogeneity of the reports prevented the homologation of the variables. However, we conducted a systematic review, in which we compiled all the available literature at the present time including all reported cases in the English and Spanish literature. IJP is considered by most authors to be a benign abnormality, is most frequently found in children, and it affects predominantly the right IJV. The clinical outcome in most cases was benign regardless of the treatment, which was either conservative or surgical. Conservative treatment is preferred for children but not for adults. To the best of our knowledge, this is the largest review of IJP to date. Future prospective multicenter studies that study diagnostic and treatment options are necessary to be able to develop guidelines on approaching this vascular abnormality.

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Conflicts of interest

There are no conflicts of interest.

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### Supplemental Table 1: Articles included in data analysis.

| Author                        | Year | Country of journal | Author origin | ‘of cases |
|-------------------------------|------|--------------------|---------------|-----------|
| Chua et al. [14]              | 2018 | England            | England       | 1         |
| Nasiri et al. [64]            | 2018 | USA                | Saudi Arabia  | 1         |
| Nedumaran et al. [60]         | 2018 | India              | India         | 1         |
| Delvecchio et al. [64]        | 2017 | USA                | USA           | 1         |
| Krstačić et al. [61]          | 2017 | Italy              | Croatia       | 1         |
| Baker et al. [3]              | 2017 | USA                | USA           | 1         |
| Bhattacharya et al. [5]       | 2017 | India              | India         | 1         |
| Phookan et al. [34]           | 2017 | USA                | USA           | 1         |
| Patel et al. [53]             | 2016 | Romania            | England       | 1         |
| Raut et al. [57]              | 2016 | USA                | India         | 1         |
| Sundaram et al. [91]          | 2016 | USA                | India         | 1         |
| Soares-Medina et al. [88]     | 2016 | USA                | Spain         | 1         |
| Yaadhavakrishnan and Navaneethan [98] | 2015 | India              | India         | 2         |
| Malik et al. [55]             | 2015 | India              | India         | 1         |
| Khashram et al. [30]          | 2014 | USA                | Australia     | 1         |
| Daley and Colliver [16]       | 2014 | USA                | USA           | 1         |
| Tanigawa et al. [93]          | 2014 | USA                | Japan         | 1         |
| Hiraki et al. [38]            | 2014 | USA                | Japan         | 1         |
| Huang et al. [42]             | 2013 | England            | China         | 1         |
| Eksioglu et al. [20]          | 2013 | USA                | Turkey        | 21        |
| Nagata et al. [92]            | 2013 | USA                | Japan         | 1         |
| Liu et al. [56]               | 2013 | Germany            | China         | 23        |
| Czyżowski et al. [13]         | 2013 | Czech Republic     | Poland        | 1         |
| Rha et al. [79]               | 2013 | Korea              | Korea         | 1         |
| Bora [7]                      | 2013 | India              | India         | 1         |
| Chakraborty et al. [11]       | 2013 | Egypt              | India         | 1         |
| Bindal et al. [6]             | 2012 | India              | India         | 1         |
| El Fakiri et al. [21]         | 2011 | France             | Morocco       | 1         |
| Aydoğan et al. [2]            | 2011 | USA                | Turkey        | 1         |
| Thulasiraman et al. [94]      | 2010 | India              | India         | 1         |
| Ogbole et al. [90]            | 2010 | England            | Africa        | 1         |
| Chang et al. [12]             | 2010 | USA                | China         | 1         |
| Gundlach et al. [13]          | 2009 | USA                | Netherlands   | 1         |
| Hopsu et al. [40]             | 2009 | USA                | Finland       | 1         |
| Wen et al. [37]               | 2009 | USA                | China         | 4         |
| Haney et al. [37]             | 2008 | Netherlands        | USA           | 1         |
| Momoo et al. [84]             | 2008 | Scotland           | Japan         | 1         |
| Hung et al. [44]              | 2008 | USA                | Canada        | 1         |
| Price et al. [79]             | 2007 | Ireland            | USA           | 3         |
| Fazilah et al. [24]           | 2006 | Malaysia           | Malaysia      | 1         |
| Jianhong et al. [49]          | 2006 | USA                | China         | 39        |
| Grange et al. [14]            | 2005 | USA                | USA           | 1         |
| Hu et al. [41]                | 2005 | USA                | China         | 29        |

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Supplemental Table 1: Continued

| Author                     | Year | Country of journal | Author origin | # of cases |
|----------------------------|------|--------------------|---------------|------------|
| Rajendran et al.⁷⁶         | 2004 | India              | India         | 1          |
| Gerek et al.²⁹             | 2003 | USA                | Turkey        | 1          |
| Erdem et al.²²             | 2002 | England            | Turkey        | 3          |
| Jeon et al.⁴⁰             | 2002 | Korea              | Korea         | 3          |
| Fernando et al.²³         | 2002 | Sri Lanka          | Sri Lanka     | 1          |
| Sakallioglu et al.²⁹      | 2002 | USA                | Turkey        | 1          |
| Reed and Grewal⁷⁹          | 2001 | USA                | USA           | 1          |
| Fitoz et al.²⁶            | 2001 | USA                | Turkey        | 2          |
| Yoon and Messner¹⁰¹        | 2001 | Canada             | Canada        | 1          |
| Sommer and Forlê⁶⁷        | 2001 | Canada             | Canada        | 1          |
| Paleri et al.⁷¹           | 2001 | Ireland            | India         | 2          |
| Rossi and Tortori-Donati⁴⁰| 2001 | Germany            | Italy         | 1          |
| Singh et al.⁸⁴           | 2001 | India              | India         | 1          |
| Ng et al.⁶⁶              | 2000 | China              | China         | 1          |
| Kwok et al.⁸¹             | 2000 | Australia          | China         | 3          |
| Fan et al.²³              | 2000 | USA                | China         | 1          |
| Sander et al.⁸²           | 1999 | USA                | Turkey        | 8          |
| Lubiana-Neto et al.³⁶     | 1999 | USA                | Brazil        | 2          |
| Gürpinar et al.³⁶         | 1999 | USA                | Turkey        | 1          |
| Chao et al.¹³             | 1999 | England            | China         | 8          |
| Indudharan et al.⁴⁷       | 1998 | England            | Malaysia      | 1          |
| Sugiyama et al.⁹⁰        | 1998 | Japan              | Japan         | 1          |
| Al-Dousary et al.¹³       | 1997 | Ireland            | Saudi Arabia  | 1          |
| Nopajaroonrri et al.⁸⁷    | 1996 | USA                | USA           | 1          |
| Bosshardt et al.⁹         | 1996 | USA                | USA           | 1          |
| Hussein et al.⁸⁵         | 1996 | Germany            | Germany       | 1          |
| Inci et al.⁸⁶            | 1995 | Germany            | Turkey        | 1          |
| Mickelson et al.¹⁴⁶       | 1995 | USA                | USA           | 1          |
| Calligaro et al.¹⁰⁰       | 1995 | USA                | USA           | 3          |
| Balik et al.¹⁴            | 1995 | Germany            | Turkey        | 1          |
| Walsh et al.⁹⁵           | 1993 | Ireland            | England       | 1          |
| Gorenstein et al.³³³⁵     | 1992 | USA                | Israel        | 1          |
| Kuo et al.³²              | 1992 | China              | China         | 2          |
| Walsh et al.⁹⁶           | 1992 | England            | England       | 2          |
| Shimizu et al.⁸³         | 1992 | Japan              | Japan         | 1          |
| Spiro et al.⁸⁸           | 1991 | USA                | USA           | 1          |
| Dhillon et al.¹⁹⁴         | 1991 | Singapore          | Malaysia      | 1          |
| Yokomori et al.¹⁸⁰        | 1990 | USA                | Japan         | 2          |
| Nwako et al.⁶⁸           | 1989 | USA                | Nigeria       | 1          |
| Zohar et al.¹⁰²           | 1988 | Scotland           | Israel        | 2          |
| Hughes et al.⁹³          | 1988 | England            | England       | 1          |
| Bowdler and Singh⁹¹      | 1986 | Ireland            | England       | 1          |
| Matsuba et al.³⁴⁴         | 1985 | USA                | USA           | 1          |
| Som et al.⁹⁰             | 1985 | USA                | USA           | 1          |
| Yashiro and Iio⁹⁰        | 1984 | USA                | Brazil        | 2          |
| Furukawa et al.²⁷⁷        | 1984 | Germany            | Japan         | 1          |

(Contd..)
Supplemental Table 1: Continued

| Author                | Year | Country of journal | Author origin | # of cases |
|-----------------------|------|--------------------|---------------|------------|
| Stevens et al.         | 1982 | USA                | USA           | 1          |
| Passariello et al.     | 1979 | Germany            | Italy         | 4          |
| LaMonte et al.         | 1976 | USA                | USA           | 2          |
| Gordon et al.          | 1876 | USA                | USA           | 2          |
| Gilbert et al.         | 1972 | USA                | USA           | 2          |
| Okay et al.            | 1970 | USA                | USA           | 1          |
| Garrow et al.          | 1964 | USA                | USA           | 1          |
| Gerwig Jr.             | 1952 | USA                | USA           | 1          |