

## Case Report

# IJV Phlebectasia: an approach algorithm

**Karthik N. Rao\*, Shrinivas S. Chavan, Vitthal D. Kale, Amol Hekare, Archana Sylendran, Abhishek Khond**

Department of Otolaryngology and Head Neck Surgery, Grant Medical College and Sir JJ Group of Hospitals, Mumbai, Maharashtra, India

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**\*Correspondence:**

Dr. Karthik N. Rao,

E-mail: [Karthik.nag.rao@gmail.com](mailto:Karthik.nag.rao@gmail.com)

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### ABSTRACT

Internal jugular venous (IJV) Phlebectasia are rare disorders. It is generally diagnosed at an early age, usually unidentified or misdiagnosed or ignored because of the scarcity of the knowledge on the disorder. We encountered a case of 7-year-old child with a right sided intermittent neck swelling which mimicked as an external laryngocele. But, the diagnosis of IJV Phlebectasia was made on “dynamic” ultrasound (USG) doppler study. Cervical adenopathy, mediastinal masses, tuberculosis and certain syndromes of connective tissue disorders were ruled out. The child was managed conservatively with parents and the child being educated about disorder.

**Keywords:** IJV, Phlebectasia

## INTRODUCTION

Internal Jugular vein Phlebectasia is one of the rare clinical entities, more commonly encountered during childhood.<sup>1</sup> The knowledge regarding this unusual clinical disorder is sparse amongst the medical fraternity. There is recent upward trend in the diagnosis of this disorder. The major purpose of this article is to shine the light on the dilemmas faced during the differentials elimination, work up and treatment.

## CASE REPORT

A 7-year-old male child presented with a 4-5 months history of intermittent, non-progressive swelling on the right side of the neck. Which becomes prominent on crying, coughing, laughing, and straining. There were no changes in the voice or any history of trauma.

A 3-cm ill-defined intermittent swelling on the right side of the just lateral to thyroid cartilage, soft and completely

compressible, more pronounced on Valsalva manoeuvre (Figure 1).

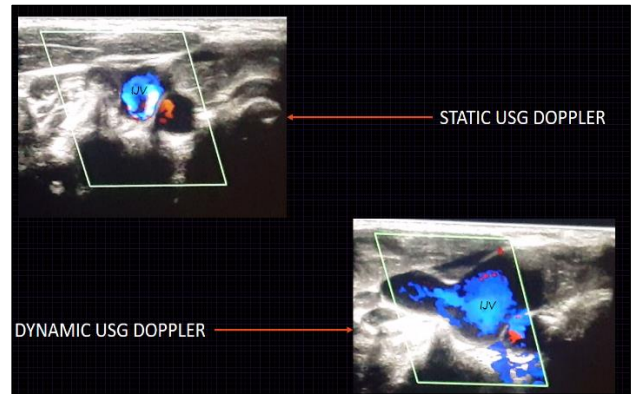


**Figure 1: No swelling on the right side (right) and the swelling prominent upon coughing (left).**

There were no other palpable swellings in the neck. Flexible fiberoptic nasopharyngoscopy was done to look at the endolarynx to rule out any internal or mixed laryngoceles. But was found to be unremarkable. Rest of the physical examination was normal. A differential diagnosis of laryngocele, A-V malformation, branchial cyst, cystic hygroma and IJV ectasia were considered. A decision was made to perform USG neck to rule out the differentials as it is an economical, easy and a non-invasive investigation.

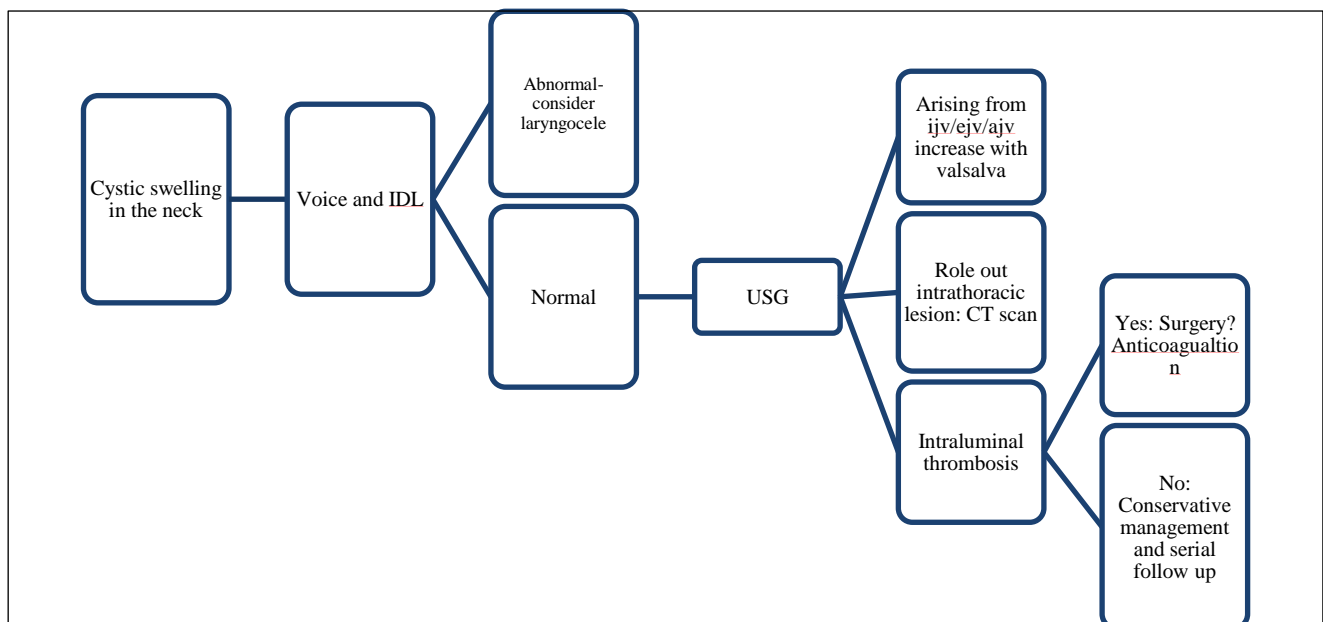
On USG, focal abnormal dilatation of the Right IJV, segment measuring 3 cm, 150% increase in diameter at the level of the dilatation upon Valsalva manoeuvre (dynamic component in USG). There was an evidence of mild turbulence in flow at the focal dilated segment (Figure 2), but no features of endoluminal thrombus was noted. Rest of the right and left IJV is unremarkable. No significantly enlarged cervical lymph nodes were detected on USG.

With this dynamic USG, a working diagnosis of focal IJV ectasia was made. Now, quest was to find the cause for this unusual clinical conundrum. In our clinical setup, the most common cause of any cervical swelling is a lymph node enlargement and the most common reason for the same being Koch's disease. A raise in intrathoracic pressure and compression over the great veins are well known to make IJV prominent on clinical examination and USG.



**Figure 2: Upper image showing the image of IJV (blue) and common carotid (red) in static USG doppler, the lower image showing the significant dilatation of the IJV following Valsalva manoeuvre.**

A HRCT thorax was done and no evidence of any mediastinal adenopathy or space occupying lesion were noted. With the aforementioned investigations, it was confirmed that this asymptomatic focal dilatation of right IJV was an incidental finding without any features of complications. Hence a decision was made to manage the patient conservatively and to keep a regular follow up to rule out any complications.



**Figure 3: Summary of the management.**

## DISCUSSION

Internal jugular Phlebectasia is generally rare and benign condition. A few clinical series data are present in the literature regarding their pathogenesis. About 50 cases

have been reported so far in literature. With a recent increase in trend ascribed to the improvement in the clinical detection from past 5 years. Usually diagnosed during childhood. There have been cases that go unrecognized and have been detected on autopsies. A few

were incidentally on table during neck dissection and spine surgeries.

The most common cause of a mass in the neck that appears only on straining is a laryngocele and the most common neck swelling encountered in India is an enlarged cervical lymph node. Internal jugular Phlebectasia manifests as an asymptomatic, soft, compressible neck swelling that increases in size on Valsalva's manoeuvre.<sup>1</sup> These entities may present as a paroxysmal cough, dysphagia or hoarseness: hinting the compression of surrounding structures.<sup>2,3</sup> Phlebectasia are unnatural and permanent distention of any part of the jugular venous system and can involve the external, internal or anterior jugular veins (in the increasing order of rarity).<sup>4</sup> The term aneurysm is to be avoided as their histological architecture is generally normal. The exact aetiology of venous ectasia is poorly understood. High clinical index of suspicion is required to diagnose the condition.

There is evidence suggesting the lack of clinical knowledge pertaining to IJV ectasia amongst the treating doctors.<sup>5</sup> It is essential to rule out the common differential diagnosis of Laryngocele, A-V malformation, branchial cyst and cystic hygroma. A USG neck is done to repudiate the differential diagnosis and static and dynamic (Valsalva) USG doppler study is done to determine the site, size, flow, endoluminal architecture, presence of thrombus, compression effects over the surrounding structures and effects following the Valsalva manoeuvres.

Once, the diagnosis of IJV Phlebectasia is made, it is imperative to rule out any mediastinal mass such as a Thymoma, aneurysms, cervical rib, lymph node enlargement and tracheobronchial hamartomas. In clinical setting with high incidence of Koch's disease, the same needs to be prioritized in the differentials. Even though there are meagre clinical evidence on Koch's as a cause for the Phlebectasia.

The primary clinical risk associated with venous ectasia is stasis. Venous thromboembolism may develop in up to 71%. Ectasia is not really at risk for rupture per se.<sup>6</sup> Internal jugular vein Phlebectasia is not known to progress. No instances of spontaneous rupture of the swelling. It usually does not require any active intervention.<sup>7</sup> Histologic analysis shows vein wall fragmentation with degeneration of elastin.<sup>8</sup> Most of the authors advice only reassurance to the patients. The contralateral IJV in majority of these reported cases are normal. The main indication for surgical management of these lesions is the fact they may be a nidus for thrombus.<sup>6</sup> Though some authors advise surgical management for Phlebectasia: tangential venorrhaphy, grafting, stenting, sheathing IJV, autovenous banding,

PFTE/ Dacron, focal ligation and complete IJV excision. Regular follow up is to maintain to look for the progression, thrombus formation and flow. A follow up is advisable to include a detailed history discarding the IJV thrombosis, clinical examination and dynamic USG doppler biannually.

## CONCLUSION

IVJ Phlebectasia is a rare disease with a recent increasing trend in the diagnosis attributed to the sophistication, ease of access for advancements medical technologies and upward drift in doctors' knowledge. In spite of IJV Phlebectasia being a benign condition it is crucial to consider it as a potential sign of a more sinister condition. Conservative management is to be opted when there are no evidences of a thrombus formation (summarized in Figure 3). Further detailed studies are required to determine the need for prophylactic anticoagulation and surgery.

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