

Diagnostic Criteria of Internal Jugular Phlebectasia in Korean Children

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Internal jugular phlebectasia (IJP) is a fusiform dilatation of the internal jugular vein (IJV), usually presented as a neck mass in children. Accurate diagnosis from carefully directed history, physical examination, and radiological study could result in lifesaving therapy. We performed our study to suggest possible clinical diagnostic criteria for IJP in Korean children. We reviewed three cases of IJP (patients group) and compared the diameter of the internal jugular phlebectasias with diameters of IJVs in ten normal children (control group) using ultrasonography (USG). There were no significant differences in the range of diameters in the resting state between the two groups. The diameters on the right side, compared with those on the left side, showed no statistical significance ($p > 0.05$). The range of expanding diameter and average expanding ratios (resting state to Valsalva maneuver $\times 100\%$) showed a statistical difference between the two groups ($p < 0.05$).

Key Words: Internal jugular vein, phlebectasia, diameter, expanding ratio

INTRODUCTION

Phlebectasia of IJV is a fusiform dilatation, usually presenting itself as an asymptomatic mass in the supraclavicular area along the anterior border of the sternocleidomastoid muscle. The characteristics of the mass are, enlargement of size on straining, crying, bending, sneezing, coughing, and Valsalva maneuver. The mass is soft, non-pulsatile, non-tender, and its size decreases when the

patient is calm. It is usually a childhood disease, thought to be of congenital origin. Diagnosis is confirmed on the basis of ultrasonography combined with Doppler flow imaging or computerized tomography (CT) scanning with contrast. Many clinicians strongly recommend conservative treatments, because the disease has a benign behavior and histopathology, and usually shows similar vascular structures on surgical specimens. Therefore, accurate diagnosis is very important to avoid unnecessary surgical exploration.

However, clinically, its diagnosis may be difficult. We measured IJV diameters, at resting, and the Valsalva state in patients with IJP, and in normal children who had no specific complaint.

MATERIALS AND METHODS

Patients group

Three children affected by IJP visited Chungbuk National University Hospital between 1998 and 2000. Two were six-year-old boys with neck masses on the right side, and one was a two-year-old boy with neck masses on both sides. The maximum size of the neck mass, on Valsalva maneuver, was about 2.5×3 cm in each of the unilateral cases, and about 2.5×3 cm on the right and 1.5×2 cm on the left, in the bilateral case. In all three cases, there were no bruits or venous hums overlying the masses. No swelling in the neck was observed at rest. Examinations of ear, nose and throat showed no specific findings.

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Chest radiography and electrocardiogram revealed no abnormality of lung fields, cardiac silhouette, or mediastinum. They had no history of trauma or head and neck surgery (Fig. 1).

Control group

Eight boys and two girls, as volunteers, were age and sex matched with the patients group. Their parents agreed to their participation in this study. The children had no history of swelling or trauma, and no abnormal findings at rest or during Valsalva maneuver in the neck. Chest X-rays and electrocardiograms revealed no abnormalities.

Measurement of the diameter of IJV

Using ultrasonography (Diasonic Master Series, Diasonic Co., Milpitas, C.A., U.S.A.), we selected

two levels in the course of IJV for measuring its diameter. Level 1 was underneath the carotid bulb, where compression of IJV by surrounding structures was minimal, and level 2 was at the cricoid cartilage, which was near the largest expanding site. Both diameters were measured in the two groups, see Fig. 2.

All diameters were calculated from the average of the widest diameter plus the narrowest diameter multiplied by 0.5 of IJV in the resting state. These were obtained by averaging the maximum and minimum diameters due to pulsation, with the diameters during Valsalva maneuver, measured using the same method.

The expanding ratio was calculated by dividing the diameter, during Valsalva maneuver, by diameter in the resting state as a percentage.

The following statistical analyses were performed; Kruskal-Wallis, and Wilcoxon, tests with significance set at $p < 0.05$.

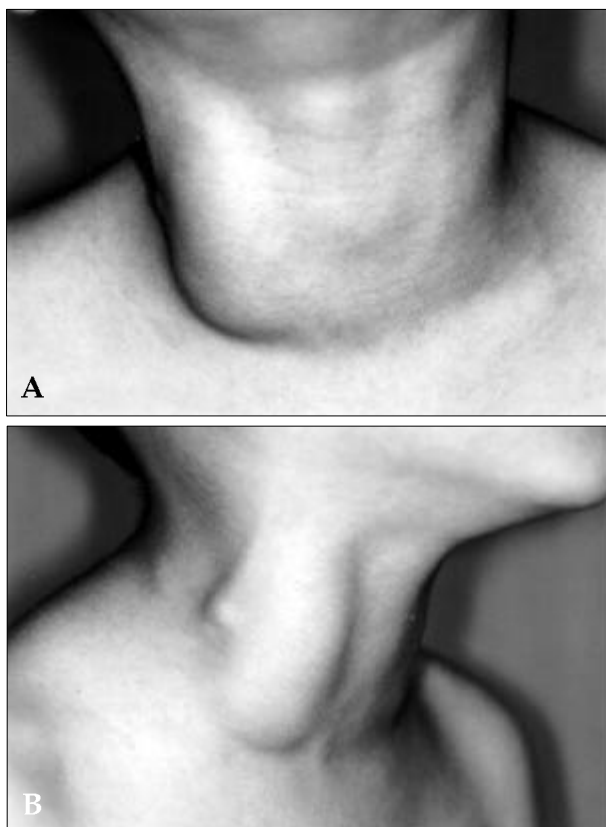


Fig. 1. Patient's neck while performing Valsalva maneuver. Note the swelling in the anterior triangle on the right side. (A) anterior view. (B) right lateral view.

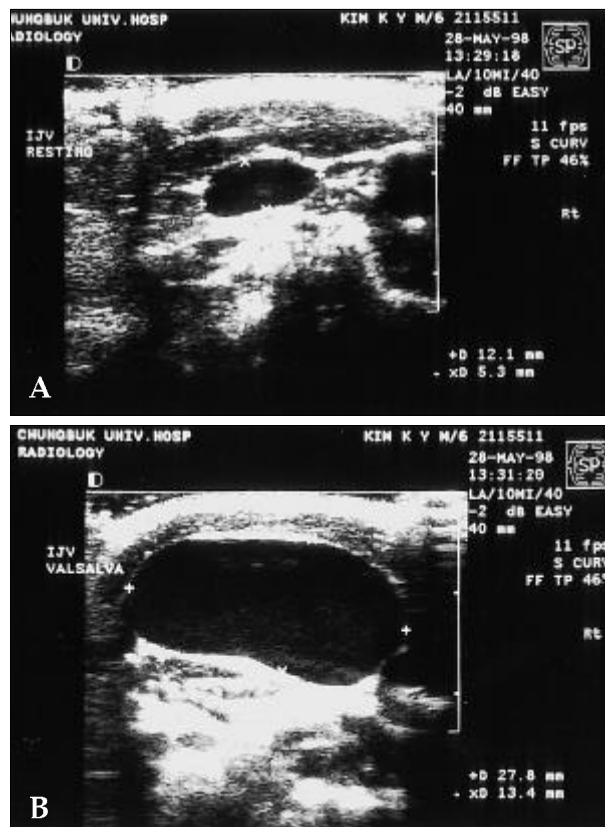


Fig. 2. Ultrasonography of the neck. (A) At rest, a normal sized right IJV is seen. (B) On exertion, an enormously dilated right IJV is seen, giving rise to a swelling in the neck.

RESULTS

Range of resting diameters & expanding diameters

In the patient group, average diameters, for IJVs, were 8.25 ± 0.88 mm (range: 7.25 to 8.90) (level 1), and 9.38 ± 2.54 mm (7.25 to 12.20) (level 2) in the resting state, 11.48 ± 2.55 mm (8.55 to 13.15) (level 1), and 21.92 ± 1.61 mm (20.60 to 23.70) (level 2), on Valsalva maneuver, while those of the left IJVs were 7.10 ± 1.53 mm (6.05 to 6.40) (level 1), and 7.50 ± 2.07 mm (5.95 to 6.70) (level 2), in the resting state, and 9.97 ± 1.40 mm (8.75

to 9.65) (level 1), and 11.01 ± 0.99 mm (10.35 to 12.15) (level 2), on Valsalva maneuver (Table 1).

In the control group, those on the right side were 7.80 ± 1.78 mm (range: 6.0 to 11.85) (level 1), and 9.75 ± 2.27 mm (5.85 to 12.70) (level 2), in the resting state, 11.14 ± 3.69 mm (8.20 to 20.15) (level 1), and 13.51 ± 3.87 mm (9.15 to 19.10) (level 2), on Valsalva maneuver, while those on the left side were 7.58 ± 1.17 mm (5.90 to 8.65) (level 1), and 9.33 ± 1.7 mm (6.75 to 11.85) (level 2), in the resting state, and 9.71 ± 3.92 mm (7.65 to 15.25) (level 1), and 11.07 ± 2.47 mm (7.25 to 15.35) (level 2), on Valsalva maneuver (Table 2).

Table 1. Diameter and Expanding Ratio of IJV in the Resting and Valsalva States in Patients

Site state	Diameter, mm				Expanding ratio, % range (average \pm SD)
	M/6*	M/6*	M/2*	(average \pm SD)	
Rt. level 1. resting	7.25	8.9	8.6	8.25 ± 0.88	17.93 - 52.91 (38.03 ± 18.07)
Valsalva	8.55	12.75	13.15	11.48 ± 2.55	
level 2. resting	12.2	8.7	7.25	9.38 ± 2.54	75.82 - 226.90 (146.50 ± 76.0)
Valsalva	21.45	20.6	23.7	21.92 ± 1.61	
Lt. level 1. resting	6.4	8.85	6.05	7.10 ± 1.53	29.94 - 59.50 (42.05 ± 15.48)
Valsalva	8.75	11.5	9.65	9.97 ± 1.40	
level 2. resting	6.7	9.85	5.95	7.50 ± 2.07	23.35 - 73.95 (51.58 ± 25.81)
Valsalva	10.55	12.15	10.35	11.01 ± 0.99	

*sex/age, $p < 0.05$.

Table 2. Diameter and Expanding Ratio of IJV in the Control Group

Site state	Diameter, mm range (average \pm SD)	Expanding ratio, % range (average \pm SD)
Rt. level 1 resting	6.0 - 11.85 (7.80 ± 1.78)	26.15 - 70.04 (39.35 ± 12.12)
Valsalva	8.20 - 20.15 (11.14 ± 3.69)	
level 2 resting	5.85 - 12.70 (9.75 ± 2.27)	9.70 - 109.40 (38.98 ± 31.15)
Valsalva	9.15 - 19.10 (13.51 ± 3.87)	
Lt. level 1 resting	5.90 - 8.65 (7.58 ± 1.17)	7.22 - 60.53 (26.83 ± 19.03)
Valsalva	7.65 - 15.25 (9.71 ± 3.92)	
level 2 resting	6.75 - 11.85 (9.33 ± 1.7)	1.40 - 57.47 (19.01 ± 19.35)
Valsalva	7.25 - 15.35 (11.07 ± 2.47)	

$p > 0.05$.

Average expanding ratio in each group

In the patient group, the average expanding ratio, of suspected phlebectasia, (right side) was $38.03 \pm 18.07\%$ (range: 17.93 to 52.91) (level 1) and $146.50 \pm 76.0\%$ (75.82 to 226.90) (level 2). On left side it was $42.05 \pm 15.48\%$ (29.94 to 59.50) (level 1), and $51.58 \pm 25.81\%$ (23.35 to 73.95) (level 2) (see Table 1).

In the control group, the right side was $39.35 \pm 12.12\%$ (26.15 to 70.04) (level 1), and $38.98(31.15\%$ (9.70 to 109.40) (level 2), while that on the left side was $26.83 \pm 19.03\%$ (7.22 to 60.53) (level 1) and $19.01 \pm 19.35\%$ (1.40 to 57.47) (level 2). The expanding ratio of one child inform the control group was 109.40%, while those of the other nine were under 60%, at level 2, on the right side. If this case was excluded, the average expanding ratio was 33.16% (Table 2).

Statistical analysis

There was a significant difference at level 2 for the expanding ratio between the two groups (Fig. 3). The range of diameters between these groups, at level 2, also showed a significant difference (Fig. 4), although, there was no significant difference in the resting state between the two groups at levels 1 and 2 (Fig. 5 and 6).

The diameters on the right side seemed to be larger than those on the left in the control group, but there was no significant difference (Fig. 7). The expanding ratios on the right side seemed to be slightly higher than those on the left in the control group, but there was no statistical significance (Fig. 8).

DISCUSSION

Phlebectasia was described characterized by Gerwig in 1952 as a fusiform or saccular dilated segment of a vein.¹ IJP is frequently present as an asymptomatic unilateral soft, non-pulsatile and non-tender swelling that enlarges with straining, crying, bending, sneezing coughing, and Valsalva maneuver.^{2,4,5} Hypotheses of etiology have been suggested as idiopathic.^{2,3,6} The histopathologic findings of the excised specimens showed nor-

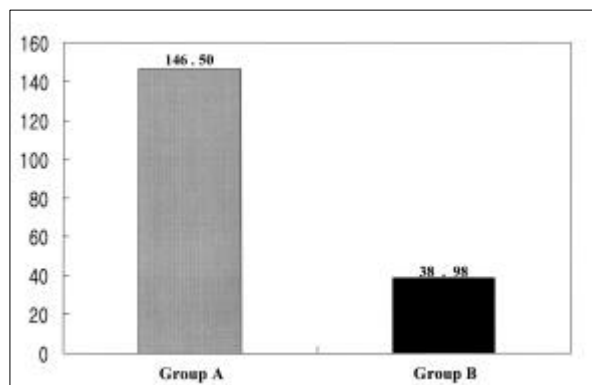


Fig. 3. Expanding ratio between resting and Valsalva states at the right cricoid cartilage level (%). $p < 0.05$.

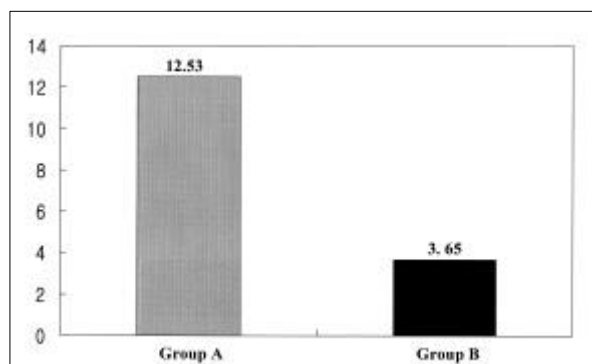


Fig. 4. Differences in the diameter between the resting and Valsalva states in the right cricoid cartilage level (mm). $p < 0.05$.

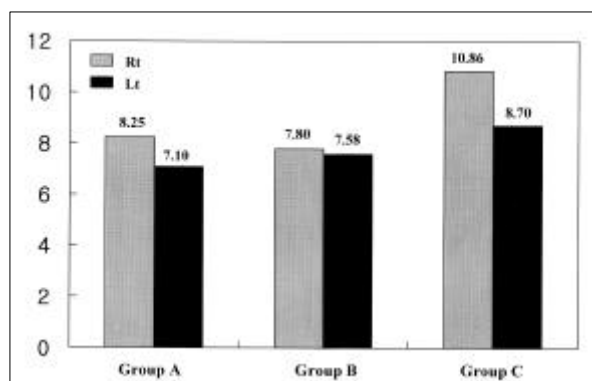


Fig. 5. Diameter of the IJV in the resting state underneath the carotid bulb level (mm). Rt, right side; Lt, left side. $p > 0.05$ between two groups.

mally dilated veins in most cases,⁶ and some specimens showed congenital defects or changes within the muscular layer of the venous wall.^{4,5}

Some pathologically observed changes varied from paucity to absence of the muscular layer,⁴

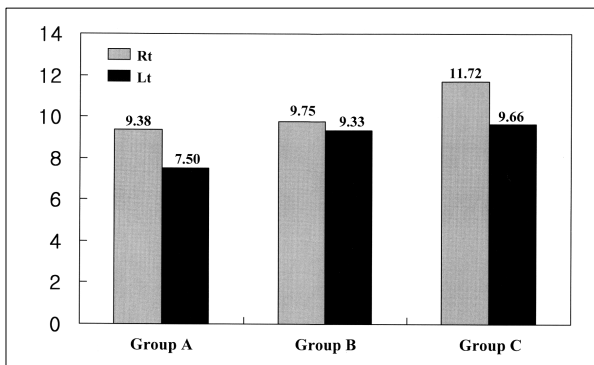


Fig. 6. Diameter of the IJV in the resting state at the cricoid cartilage level (mm). Rt, right side; Lt, left side. $p > 0.05$ between two groups.

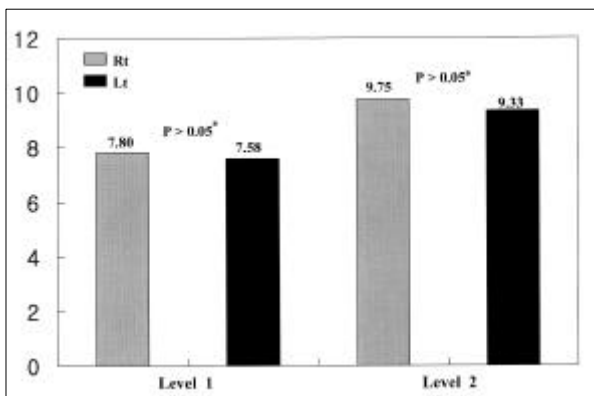


Fig. 7. Diameter of the IJV in the resting state in the control group (mm). Rt, right side; Lt, left side. $*p > 0.05$ between right and left sides at level 1 and 2.

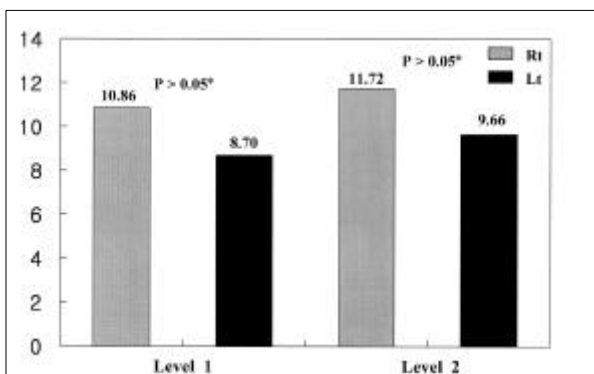


Fig. 8. Expanding ratio between the resting and Valsalva states in the control group (%). Rt, right side; Lt, left side. $*p > 0.05$ between right and left side at levels 1 and 2.

minimal fibrosis, or the amount of connective tissue.⁵

The diagnosis of the IJP was tentatively made

on a clinical basis, and confirmed, either by invasive contrasted radiography, or surgical exploration.^{2,7} Recently, advanced non-invasive or minor invasive technology could substitute for surgical exploration or invasive methods.

Ultrasound with color Doppler flow imaging could be an option for non-invasive, real time diagnostic method^{3,4,7} and can confirm the presence, or absence, of blood flow as well as its direction.⁷

There are no reported studies on the size and anatomical correlation with surrounding structures of the internal jugular vein. We examined IJVs in normal children with USG from the clavicle to the angle of mandible. The image of IJVs varied according to the level. Therefore, we selected two levels for the measurement of IJV diameter. Level 1 was underneath the carotid bulb, and level 2 was at the cricoid cartilage, close to the maximum expanding site. At these levels, the IJV was a round shape on USG, and relatively uncompressed by surrounding structures.

Average expanding ratios at level 2, on both sides of the neck, were higher than those at level 1 in the patient group, which was contrary to that in the control group. It might be that there were slight fusiform or saccular changes at level 2 in the patient group.

In the normal group, the expanding ratio in nine of the ten children was under 60% at the cricoid cartilage level of the IJV, which was relatively free from surrounding structures. However, in one child, at level 2, the right side showed 109.40%. The average expanding ratio of the control group was down to 31.16% from 38.98% when the latter child's result was excluded (Table 2). This finding show there may exist a certain proportion with abnormally expanding IJVs among non-complaining children, and USG can be used as a screening test.

The treatment of IJP in the literature varies due to the rarity of the disease. Most authors strongly recommended conservative treatment, especially if the phlebectasia is asymptomatic, or the patient has minor symptoms because of benign behavior. Surgical intervention should be recommended in cases of phlebitis, thrombus formation, rupture of the lesions, or some cosmetic deformity.⁵ Inflamed lymphatic malformation could be combined with

IJP, and surgical examination may be required.⁸ Potency of the contralateral IJV should be confirmed before elective excision in cases of a unilateral lesion. Bilateral lesions should not be excised because of the possibility of massive edema on the face and head, with associated cerebral edema.

IJP could be diagnosis by careful history taking and physical examination. USG is the best choice of diagnostic and screening method. It could avoid unnecessary surgical exploration. The expanding diameter of the IJV, and the expanding ratio of the IJV diameter, between the resting and Valsalva states, showed good diagnostic criteria.

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