

ORIGINAL ARTICLE

นิพนธ์ต้นฉบับ

Internal Jugular Phlebectasia in Children

*Paisarn Vejchapipat ***Soottiporn Chittmittrapap ***Bidhya Chandrakamol **

Paisarn Vejchapipat, Soottiporn Chittmittrapap, Bidhya Chandrakamol. Internal Jugular Phlebectasia. Thai Journal of Surgery 1996 ; 17 (2) :74-80

- Objective** : To review and to evaluate the patients with internal jugular phlebectasia treated at Chulalongkorn University Hospital.
- Setting** : Division of Pediatric Surgery, Department of Surgery, Faculty of Medicine Chulalongkorn University Hospital, Bangkok, Thailand
- Design** : Retrospective descriptive study.
- Subjects** : Children diagnosed as internal jugular phlebectasia between 1995 and 1996.
- Method** : The data of all patients were reviewed for general information, diagnostic tools, treatment, complications and outcome.
- Results** : Five children were diagnosed as internal jugular phlebectasia. The sex ratio male:female = 4:1 and age ranged from 2 to 12 years. All patients had right-sided lesions. The single presenting symptom was collapsible neck swelling that enlarged with straining. The diagnosis was confirmed by doppler ultrasound in three patients. All children underwent surgical correction. The results of the treatment at 6 month-follow up were satisfactory in all patients.

Index : Phlebectasia, Internal jugular vein, Neck mass

Reprint request : Paisarn Vejchapipat, MD.
Department of Surgery, Faculty of Medicine,
Chulalongkorn University, Patumwan, Bangkok 10330, Thailand

* Division of Pediatric Surgery, Department of Surgery, Chulalongkorn University Hospital

ORIGINAL ARTICLE

นิพนธ์ต้นฉบับ

หลอดเลือดดำ Internal Jugular โป่งพอง

ไพศาล เวชชพิพัฒน์*,

สุทธิพร จิตต์มิตรภาพ*,

พิทยา จันทรภมม*

วัตถุประสงค์	เพื่อรวบรวมและศึกษาผลของการรักษาผู้ป่วยเด็กที่เป็น internal jugular phlebectasia ในคณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย
สถาบัน	หน่วยกุมารศัลยศาสตร์ ภาควิชาศัลยศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย กรุงเทพมหานคร
รูปแบบ	เป็นการศึกษาย้อนหลังแบบพรรณนา
ข้อมูล	ผู้ป่วยเด็กที่ได้รับการวินิจฉัยว่าเป็น internal jugular phlebectasia ระหว่างปี พ.ศ. 2538 และ 2539
การศึกษา	รวบรวมและศึกษาแฟ้มประวัติผู้ป่วยในแง่ของข้อมูลทั่วไป การวินิจฉัย การรักษา ภาวะแทรกซ้อน และผลของการรักษา
ผล	มีผู้ป่วยทั้งหมด 5 ราย เพศชายต่อเพศหญิงเท่ากับ 4 ต่อ 1 อายุอยู่ระหว่าง 2 ปี ถึง 12 ปี ผู้ป่วยทุกคนมาพบแพทย์ด้วยเรื่องก้อนที่คอด้านขวาซึ่งก้อนสามารถขยายขนาดได้เมื่อเบ่ง การยืนยันการวินิจฉัยอาศัยคลื่นความถี่สูงในผู้ป่วย 3 ราย ผู้ป่วยทั้ง 5 รายได้รับการผ่าตัดรักษา จากการตรวจผนังเส้นเลือดทางพยาธิวิทยาไม่พบสิ่งผิดปกติชัดเจน ผลการรักษาในผู้ป่วยทุกคนเป็นที่น่าพอใจ ไม่มีภาวะแทรกซ้อนเกิดขึ้น

* หน่วยกุมารศัลยศาสตร์ ภาควิชาศัลยศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

Internal jugular phlebectasia or venous dilatation of internal jugular vein is a rare condition. Such lesion has been reported in various names, including venous aneurysm, venous pseudoaneurysm, congenital venous cyst, venomes du cou, venous ectasia and internal jugular phlebectasia¹. Most of the patients are children. This condition is believed to be of congenital cause. The lesion may be fusiform or saccular but fusiform dilatations are more common than saccular lesions in both adult and children. Specific histopathologic abnormalities have not been identified in the previous report. Progressive enlargement and possible thrombus formation causing pulmonary embolism indicate surgical correction in most patients².

Patients and Methods

The records of patients diagnosed as internal jugular phlebectasia admitted to the Pediatric Surgery Division, Department of Surgery, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand between 1995 and 1996 were reviewed. The information registered included age, sex, side involved, physical findings, investigations for diagnosis, indication for surgery, types of surgery, operative findings, histologic findings, peri-operative course, complications and results at 6 month-follow up interval.

Results

General Information

There were five patients diagnosed as internal jugular phlebectasia admitted between 1995 and 1996.

Male: female ratio was 4:1 and the age of the patients ranged from 2 years to 12 years old. All patients had right-sided lesions. (Table 1)

Physical findings and investigations

All patients were identified with a non-pulsatile, non-tender, compressible neck swelling that enlarged during Valsalva maneuver, straining, coughing, shouting and singing. One patient had insidious onset of voice fatigue while singing determined that phlebectasia may compress external branch of superior laryngeal nerve.

The diagnosis of internal jugular phlebectasia were confirmed by doppler ultrasound in first three patients.(Fig.1) Large expansile internal jugular vein was identified in every patient. In the last two patients, the diagnosis was based only on specific physical findings.

Indications for surgery, types of surgery and operative findings

Indications for surgery in most of the patients were cosmetic concern and parents' concern except in one patient, the surgical indication in such case was fatigue of voice. All five patients underwent various type of surgical correction. There were three types of procedures: resection without reconstruction, resection with end to end anastomosis and venous plication as shown in table 2. Patient#1 underwent venous plication. Patient#2, 3 and 4 underwent venous resection and patient#5 underwent venous resection with end to end anastomosis. The size of the phlebectasia ranges from 3-4 cm. to 4-6 cm. and the type of phlebectasia of the patients was fusiform type. (Table 2 and Fig.2)

Table 1 General Informations

Patient	Sex	Age(yrs)	Affected side
1	male	2	right
2	male	5	right
3	male	7	right
4	female	8	right
5	male	12	right

Table 2 Operative findings, procedures and operative time

Patient	Incision	Length(cm.)	Diameter(cm.)	Procedure	Operative time (min)
1	Transverse	4	3	Venous plication	60
2	Transverse	5	3	Resection	75
3	Oblique	5	4	Resection	120
4	Transverse	6	3	Resection	90
5	Transverse	4	3	Resection with EEA	90

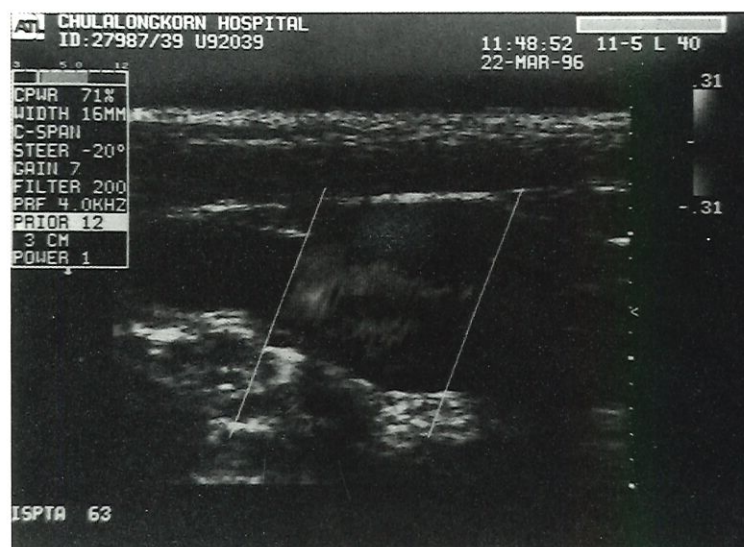


Figure1. Doppler Ultrasound demonstrates an enormously dilated right internal jugular vein during Valsalva maneuver.

Histologic findings

Microscopic examination (H&E stain) of the excised veins (patient # 2, 3, 4, 5) showed no significant abnormalities in the wall of the internal jugular veins.(Fig.3)

Perioperative period, complications and follow-up results

Most of the patient can be discharged from the hospital in the third postoperative day with no any complication. The symptom of voice fatigue in patient #3 disappeared immediately after the operation. The results of all patients on 6-month follow-up were satisfactory and there were no late complications.

Discussion

Phlebectasia (Phleb = vessel, ectasia = dilatation) is not limited to the internal jugular vein but can also arise from the external jugular vein, anterior jugular vein, facial vein, cervical vein and intercommunicating vein^{3,4}. Internal jugular phlebectasia is a rare entity in which the internal jugular vein, usually on the right side, becomes dilated on straining such as coughing, crying and singing. After searching through the literature, there were approximately 45 cases reported in the world⁵. All of those presented with a swelling in either side of the neck on straining, and none was diagnosed incidentally during other surgery. Most of those cases have been children, or have had an onset of illness

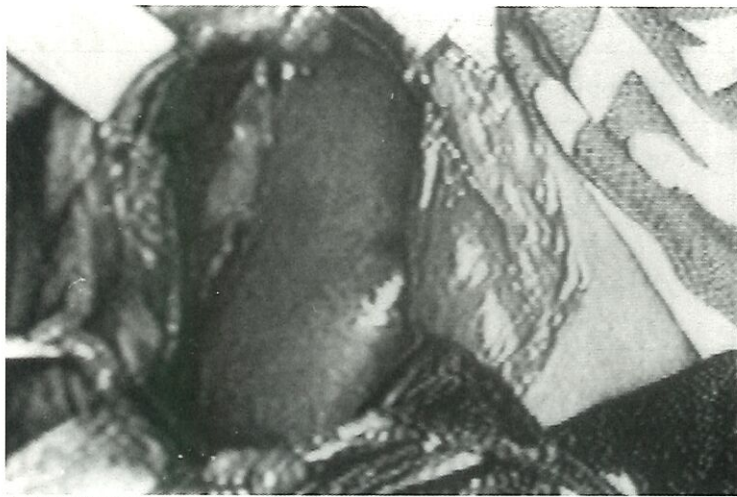


Figure 2. The operative finding shows the dilated internal jugular vein.

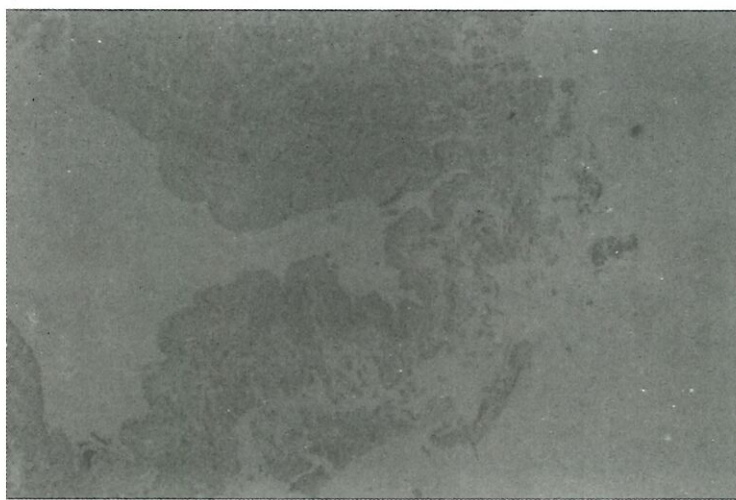


Figure 3. Histologic examination shows normal structure of the venous wall.

that dated back to childhood. Boys are more than twice as often affected as girls. The overwhelming preponderance of right side involvement over the left is obvious^{5,6}. In this report, boys are more common than girls and all patients have right-sided internal jugular phlebectasia.

Every patient with a swelling in the neck that increases in size after the Valsalva maneuver should be suspected of having internal jugular phlebectasia. Diagnostic studies adopted in the literature have included venography⁷, arteriography⁸, doppler ultrasound⁹ and CT scan. In fact, the correct diagnosis of internal jugular phlebectasia was easily made by

physical examination with doppler ultrasound which clearly demonstrated an enormously dilated right internal jugular vein and a pulsating common carotid artery just medial to the dilated vein during exertion. Because of its rarity, the exact pathogenesis remains unclear. Garrow et al¹⁰ reported an elevated venous pressure in the distended left internal jugular vein and a normal pressure in the contralateral vein during spontaneous respiration. However, no mechanical obstruction was identified during surgical removal of the dilated vein. The more lateral placement of the right brachiocephalic vein and the relationship of the other veins (the right subclavian and internal jugular) to the sternoclavicular joint could possibly explain the peculiar presence of internal jugular and other phlebectasia on the right side. In addition, on the right, the brachiocephalic vein comes into direct contact with the right pleura. Therefore, in some individuals, inflation of the cupola of the right lung might produce sufficient pressure on the right brachiocephalic vein (against the head of the clavicle) to produce functional obstruction and temporary distention of the vein. These facts may be indicative of a congenital cause. In addition, most of the patients reported in the literature have been children (or provide a history dating to childhood), suggesting that the underlying anatomic variation may be congenital³.

Microscopic examination of surgical specimens in the literature demonstrated a wide spectrum of findings^{2,6,11} showing either normal or thinned venous walls and a normal or decreased amount of elastic fibers. The amount of connective tissue may be either increased or decreased. In this study, histologic examinations of the dilated veins showed no significant abnormalities. Because the

internal jugular veins has high flow rates and is not in a dependent position, it is unlikely to develop venous stasis or thrombosis¹¹. Internal jugular phlebectasia is a benign condition. However, the natural history of internal jugular phlebectasia has not been documented so there are somewhat controversy about its treatment. Although most of the reported phlebectasia lesions have been removed, some authors have recommended surgical treatment only if the lesion is tender, increases in size, causes nerve compression or causes cosmetic deformity. Some surgeons prefer conservative treatment because they believe that complications of internal jugular phlebectasia itself is rare and resection of phlebectasia is not without potential morbidity¹¹. While others prefer surgical treatment as a routine, they claim that the lesion would be progressively enlarged and surgical treatment could be accomplished without complication². In our series, surgical correction by resection, resection with end to end anastomosis or venous plication has no morbidity. However, in elderly or in patient with bilateral lesions the surgical treatment for internal jugular phlebectasia may be hazardous⁶. We recommend an individualized approach to these lesions instead of surgical treatment as a routine.

Conclusion

Internal jugular phlebectasia is an unusual neck swelling. The cause of the lesion in children is not known. Finally, internal jugular phlebectasia is a benign condition. Treatment could be limited to reassurance in selected cases that the lesion hardly produces no morbidity or disability. However, surgical treatment for cosmetic purposes alone consists of resection or plication of phlebectasia; this produces no gross side effects.

References

1. Gilbert MG, Greenberg LA, Brown WT, Puranik S. Fusiform venous aneurysm of the neck in children: a report of four cases. *J Pediatr Surg* 1972;7:106-11
2. Danis RK. Isolated Aneurysm of the Internal Jugular Vein: A Report of Three Cases. *J Pediatr Surg* 1982;17:130-1
3. Nwako FA, Agugua NEN, Udeh CA, Osuorji RI. Jugular phlebectasia. *J Pediatr Surg* 1989; 24: 303-5
4. Natarajan B, Johnstone A, Sheikh S, Palmer O, Makhavan KNH. Unilateral anterior jugular phlebectasia. *J Laryngol Otol* 1994; 108: 352-3
5. Yokomori K, Kubo K, Kanamori Y, Takemura T, Yamamoto T. Internal Jugular Phlebectasia in Two Siblings: Manometric and Histopathologic Studies of the Pathogenesis. *J Pediatr Surg* 1990; 25: 762-5
6. Murty GE, Bradley PJ. Bilateral internal jugular phlebectasia. *J Laryngol Otol* 1992; 106: 753-4
7. Gordon DH, Rose JS, Kottheier P, et al. Jugular venous ectasia in children. *Radiology* 1976; 118: 147-9
8. Passariello R, Cozzi F, Casalena G, Colarossi G, Rossi P, Simonetti G. Angiographic diagnosis of jugular venous system dilatation in children. A report of five cases. *Pediatr Radiol* 1979;8: 247-50
9. Leung A, Hampson HB, Singh MP, et al. Ultrasonic diagnosis of bilateral congenital internal jugular venous aneurysms. *Br J Radiol* 1983; 56: 588-91
10. Garrow E, Kirschten H, Soh HI. Internal jugular phlebectasia. *Am J Surg* 1964; 108: 380-3
11. Mickelson SA, Speckler E, Roberts K. Management of internal jugular vein phlebectasia. *Otolaryngol Head Neck Surg* 1995;112:473-5