Aortic Valve Prolapse in Subpulmonic Ventricular Septal Defect

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Abstract

Background : Ventricular septal defect (VSD) is the most common congenital heart disease worldwide. Subpulmonic type VSD is an interesting subtype due to the aortic valve complications rate, which can change the prognosis of the VSD. Higher prevalence rates have been reported with this subtype in Eastern countries but there has been no report from Thailand so far.

Objective : 1. To determine the prevalence of subpulmonic VSD.
2. To determine the prevalence and demographic data of aortic valve prolapse (AVP) and aortic regurgitation (AR) in subpulmonic VSD.

Method : A retrospective study of 1,977 patients with isolated VSD, diagnosed from January 1995 to June 2002 at the Cardiology Unit, Queen Sirikit National Institute of Child Health was reviewed to differentiate types of VSD. Color flow doppler echocardiogram was performed in all cases to confirm the diagnosis and differentiate the types of VSD. Patients with subpulmonic VSD were studied to find out the presence of the aortic valve prolapse and aortic regurgitation. Those who had subpulmonic VSD were called for reevaluation of aortic valve complications, from January 2000 to June 2002.

Main Outcome Measure : Subpulmonic VSD, aortic valve prolapse (AVP) and aortic regurgitation (AR).

Results : Subpulmonic VSD was diagnosed in 312 cases (17.5%). At the mean age of 3.47 yr, AVP was found in 101 cases (32.4%) and AR was found in 54 cases (17.3%) at the first echocardiogram. Subsequent echocardiography follow-up showed that the overall number of AVP cases was 153 (49%) and AR was 84 (26.9%) at the mean age of 5.5 and 6.3 year respectively.

Conclusion : The prevalence of subpulmonic VSD was high among Thai children. Aortic valve complication is common in this group and can occur from early infancy. The present findings support the progressive increase in this complication with age.

Key word : Heart Septal Defects, Ventricular; Heart Defects, Congenital; Aortic Valve Prolapse; Echocardiography

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Isolated ventricular septal defect (VSD) is the most common congenital heart disease worldwide. The prevalence of VSD varies between 24 and 38 per cent of all congenital heart diseases[1-3]. At the authors' institute this figure is about 27 per cent. Perimembranous VSD is the most common subtype reported in all parts of the world but the prevalence of subpulmonic VSD varies in different countries, being higher in the Eastern hemisphere than in the Western (30% *vs* 6-8%)[1-6].

Subpulmonic VSD is an interesting subtype due to the difference in natural history from other types of VSD. Many patients with subpulmonic VSD present with aortic valve prolapse (AVP) and aortic regurgitation (AR) when they become older[1,7,8]. There are reasons to explain how AVP and AR in subpulmonic VSD develop[9]. Firstly, subpulmonic VSD is located beneath both the aortic and pulmonic valve leaflets, thus exposing the right coronary cusp of the aortic valve to the VSD, this lack of support below the right coronary cusp is due to the defect and secondly, the high turbulence flow across the VSD beneath the aortic valve causes distortion of the leaflet especially the right coronary cusp or part of it, resulting in prolapse into the defect. High flow velocity across the VSD usually occurs in small to moderate sized VSDs, hence the size of the VSD becomes an important factor in the development of AVP. If the distortion of the aortic valve is significant enough, AR will occur due to the elongation of the free margin of the prolapsed cusp.

AVP and AR are the natural complications of VSD, which will progressively increase with age and change the prognosis of VSD[1,10,11]. The prevalence of subpulmonic VSD with aortic valve complications has not been reported in Thailand.

Objective

1. To determine the prevalence of subpulmonic VSD.

2. To determine the prevalence and dermographic data of AVP and AR in subpulmonic VSD.

MATERIAL AND METHOD

Retrospective review was done in all patients diagnosed with isolated VSD at Queen Sirikit National Institute of Child Health from January 1995 to June 2002. Diagnosis was confirmed by color flow doppler echocardiogram performed by pediatric cardiologists. Subpulmonic VSD patients and patients without details about the type of VSD were recalled for echocardiographic reevaluation to confirm the type of VSD and to look for aortic valve complications. The study was conducted from January 2000 to June 2002. Demographic data was collected from the medical records and videotape recordings of echocardiograms were reviewed.

Subpulmonic VSD was diagnosed by echocardiogram in long and short axis views of the right ventricular outflow tract. (Fig. 1, 2) Aortic valve prolapse was commonly seen in the left parasternal long axis as shown in Fig. 3 and aortic regurgitation

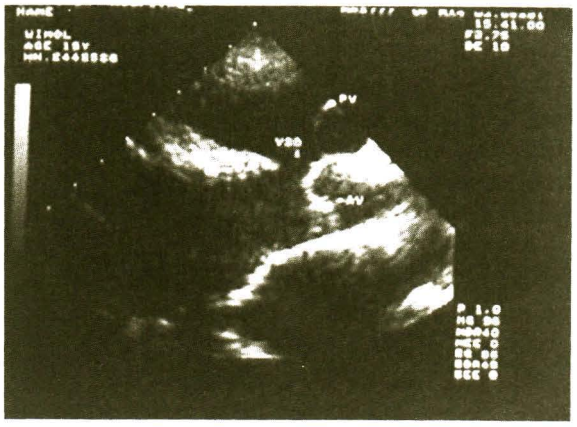

Fig. 1. Long axis view of subpulmonic VSD.

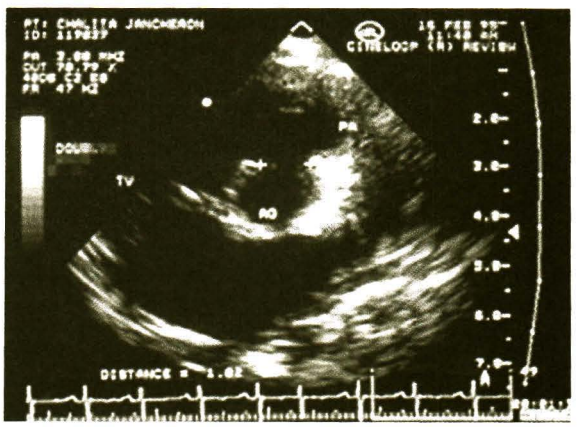

Fig. 2. Short axis view of subpulminc VSD.

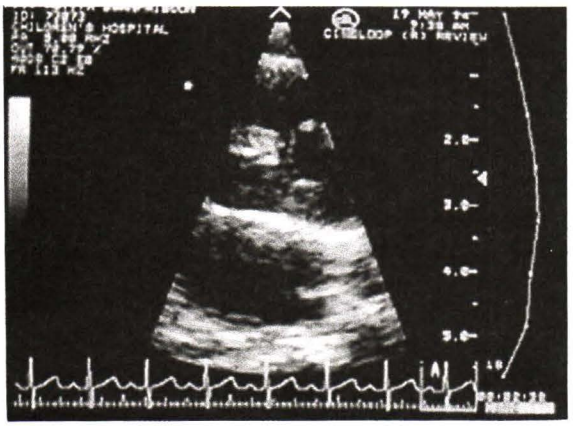

Fig. 3. Aortic valve prolapse in subpulmonic VSD.

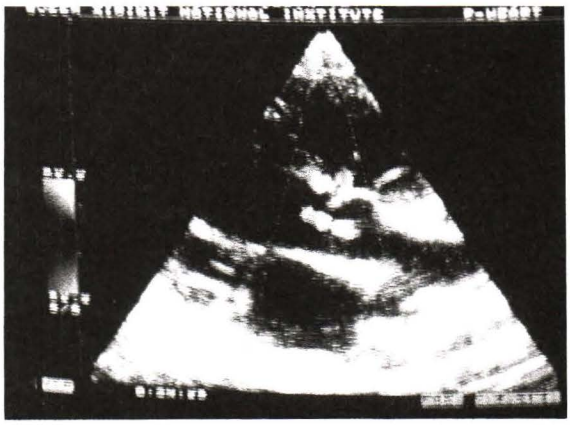

Fig. 4. Aortic regurgitation in subpulmonic VSD.

Table 1. Types of the VSD.

Type	Number	%
Perimembranous	1,332	74.8
Subpulmonic	312	17.5
Muscular	69	3.9
Inlet	39	2.2
Multiple	28	1.6

jet was seen by color flow doppler echo as shown in Fig. 4.

RESULTS

There were 1,977 cases diagnosed with isolated VSD in the 7.5year study period. Data and video tape recordings were available to differentiate VSD type in 1,780 (90%) cases. Different types of VSD are shown in Table 1.

There were 312 cases of subpulmonic VSD, of which 191 (61.2%) were male and 121 (38.8%) were female. Mean age at diagnosis of subpulmonic VSD was 3.5 years, range from 2 days to 21 years.

The initial echocardiogram of the patients with subpulmonic VSD showed 101 cases associated with AVP. Of these prolapsed cases, 54 cases also had AR. The youngest patient with AVP was one month of age and was found to have developed AR on a subsequent follow-up echocardiogram done at 2 months of age.

By the end of the present study, 735 echocardiograms had been performed in all 312 cases (once in 129 cases, twice in 107 cases, three or more times in 76 cases). 183 cases had echocardiogram more than twice which revealed an additional 52 cases of AVP and 30 cases of AR. The time to development of AVP and AR from the first echo was 3.3 and 3.4 years respectively. The overall prevalence of AVP was 153 cases (49%) and AR was 84 cases (26.9%) at the end of the study. The average age groups of all patients who had AVP and AR were 5.5 and 6.3 years respectively.

Accumulation of the percentage of patients who were found to have AVP and AR at the time of echocardiogram is shown in the graph (Fig. 5).

Outcome of treatment

Of 312 cases in the 7.5 years review, 3 cases had severe pulmonary vascular disease at the initial presentation and 3 cases died due to congestive heart failure and pneumonia prior to surgery. Surgery was performed on 85 cases. Of these, 40 cases with large VSD and associated congestive heart failure had VSD closure during the infancy period with one peri-operative death. All patients who had AR (84 cases) were counseled for surgery but only 45 cases had surgical correction in the present study period; 35 with mild AR had simple VSD closure, 6 had VSD closure with AV repair for moderate to severe AR and 4 cases with severe AR and severe deformity of AV had VSD closure with Ross's operation. All had a favorable outcome after surgery. 72 patients are presently waiting for surgery while 42 cases failed to return for

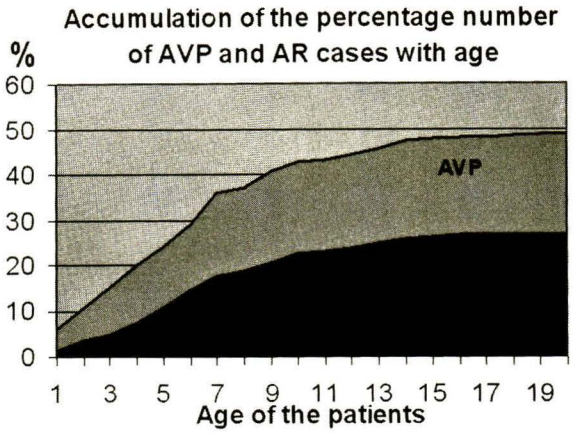

Fig. 5. Accumulation of the percentage of AVP and AR cases with age.

follow-up. There was no case of spontaneous closure during this period.

DISCUSSION

Queen Sirikit Institute is a tertiary care center and most of the patients are referred from all over the country. During the 7.5 year study period there was quite a large number of isolated VSD. The prevalence of subpulmonic VSD from the authors' review suggests that this subtype of VSD is high in the Thai population. The average age of the patients was 3.5 years at the initial diagnosis, which could be due to late referral of the patients. It is possible that before this age, some VSDs may have spontaneously closed especially the muscular and perimembranous type[12] but such a closure rarely occurs in the subpulmonic type. So, the true prevalence of subpulmonic VSD may actually be less than 17.5 per cent due to the referral bias.

The prevalence of AVP at 49 per cent and AR at 26.9 per cent in the present series is comparable to reports from Japan, which showed AVP at 43.3 per cent and AR at 24 per cent[10] and those from Taiwan which reported AVP and AR at 28 per cent[6]. In the study period, the authors found 52 additional cases of AVP and 30 cases of AR. This finding further supports the hypothesis that progressive aortic valve disease is quite a common complication in subpulmonic VSD. Nonetheless, from the present study it was also found that 6.1 per cent of AVP and 1.6 per cent of AR cases were infants less than one year of age. The youngest patient who had AVP in the present study was a month old infant. AVP in young VSD patients was also reported by Edagawa T[13], who in his study of 140 cases, found 5 infants less than one year of age with AVP. The finding of AVP in these small infants raises the question of association of congenital abnormality of the aortic valve leaflet itself as one of the factors for the development of AVP and AR in later life.

The average age group of AVP and AR in the present review was similar to that reported by Momma et al[10]. This study was a retrospective one. It was able to show the average age of patients who had AVP and AR but was unable to show the onset of the problem. Diagnosis of AR in VSD patients by using the clinical criteria of murmur alone can be easily missed with a false prevalence as low as 8.9 per cent[14]. Early AR can be diagnosed easily by color flow doppler echocardiogram. Echocardiographic follow-up[15] along with clinical follow-up of patients with VSD is recommended for early detection of AR, which would need a prospective study starting from early infancy.

The accumulation of the percentage of AVP and AR cases from the present series showed increasing numbers of cases with age with the rate of rise being high in the first decade. However, 129 of 312 cases had only one echocardiogram performed. Echocardiographic follow-up is necessary to get a higher prevalence of the AVP and AR cases.

The presence of AVP and AR changes the prognosis of the patient with small to moderate size VSDs. In the past, we thought that all small VSDs were benign lesions and would close spontaneously. Those who have significant aortic valve complications need VSD closure as well as some form of aortic valve surgery. These operations have some mortality and morbidity. At the moment, surgical treatment for subpulmonic VSD is recommended Queen Sirikit Institute in all patients with a moderate to large VSD, or those associated with significant AV deformity or AR. Simple VSD closure will be performed in cases of VSD with trivial to mild AR[16,17]. This suggestion is supported by Hisatomi K et al, who found that after simple VSD closure, AR resolved in 7/13 cases and there was no change in 6/13 with mild AR preoperatively[18]. Komai et al showed that a long period from the onset of AR to the operation may be a factor for persistent AR post-operatively[19]. In cases of moderate AR or aortic valve not severely deformed,

our surgeons prefer to close the VSD and repair the aortic valve[20]. Some failure rate has been seen with this procedure and some cases can have progressive deterioration after aortic valvuloplasty[21,22]. In cases of severe AR from marked deformity of aortic valve, our surgeons have a tendency to perform Ross's operation instead of aortic valve replacement. The type of operations performed will also depend on the experience of the surgeons.

There is debate about when to operate on small subpulmonic VSDs with or without mild AVP. Sim suggested operating on all subpulmonic VSDs with AVP before the development of AR[23,24]. On the other hand, Hung found that AR can progressively be seen even after VSD closure but the prevalence of AR occurring post-operatively was lower than the natural progression of AR in non-operated cases, so he suggested operating on all patients with subpulmonic VSD especially when associated with abnormal aortic valve[1]. Tomiya H. studied the prevalence of AR in the long-term follow-up of patients post simple VSD closure and found a higher number of AR cases (26%, 6 of 23 cases) in the patients without AVP compared to patients with AVP but without AR pre-operatively[25] (13.3%, 2 of 15 cases). Short and long-term results of any treatment regimens are very important for the management of patients with subpulmonic VSD.

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การยื่นของลิ้นหัวใจเอออร์ติคในโรครูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค

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ความเป็นมา : รูรั่วผนังหัวใจห้องล่างเป็นโรคที่พบได้ที่สุดในกลุ่มโรคหัวใจพิการแต่กำเนิดทั้งหมดทุกแห่งในโลก ตำแหน่งของรูรั่วชนิดที่อยู่ใต้ต่อลิ้นหัวใจพัลโมนิคเป็นชนิดที่น่าสนใจเนื่องจากมีรายงานพบได้สูงในแถบประเทศตะวันออกและมีอุบัติการณ์ การเกิดภาวะแทรกซ้อนตามธรรมชาติของลิ้นหัวใจเอออร์ติครั่ว อันเนื่องมาจากการยื่นลงไปอุดรูรั่วที่ผนังหัวใจ อุบัติการณ์นี้ยังไม่มีรายงานในเมืองไทย

วัตถุประสงค์ : 1. เพื่อค้นหาอุบัติการณ์ของรูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค
2. เพื่อค้นหาอุบัติการณ์ของการยื่นและรั่วของลิ้นหัวใจเอออร์ติคในโครูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค

วิธีการศึกษา : โดยการศึกษาย้อนหลังผู้ป่วยเด็กโรครูรั่วที่ผนังหัวใจห้องล่างจำนวน 1,977 รายที่ได้รับการวินิจฉัยตั้งแต่เดือนมกราคม พ.ศ. 2538 ถึง เดือนมิถุนายน พ.ศ. 2545 ที่หน่วยกุมารเวชศาสตร์โรคหัวใจ สถาบันสุขภาพเด็กแห่งชาติมหาราชินี ทำการศึกษาแยกชนิดของรูรั่วที่ผนังหัวใจห้องล่างโดยดูจากผลการตรวจและเทปบันทึกภาพการตรวจคลื่นเสียงสะท้อนหัวใจ และในรายที่เป็นรูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิคจะได้รับการค้นหาอุบัติการณ์ของการยื่นและรั่วของลิ้นหัวใจเอออร์ติคโดยดูจากผลในอดีตและตามมาตรวจซ้ำเพื่อค้นหาภาวะแทรกซ้อนดังกล่าวเพิ่มเติม เริ่มทำการศึกษาตั้งแต่เดือนมกราคม พ.ศ. 2543 ถึง เดือนมิถุนายน พ.ศ. 2545

ตัววัดผลหลัก : รูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค การยื่นและรั่วของลิ้นหัวใจเอออร์ติค

ผลการศึกษา : พบรูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค 312 ราย (17.5%) อายุเฉลี่ยที่ได้รับการวินิจฉัยเท่ากับ 3.47 ปี พบว่ามีการยื่นและรั่วของลิ้นหัวใจเอออร์ติค 101 ราย (32.4%) และ 54 ราย (17.3%) ตามลำดับ จากการติดตามมาตรวจซ้ำพบจำนวนผู้ป่วยเด็กที่มีความผิดปรกติของการยื่นและรั่วของลิ้นหัวใจเอออร์ติค รวมทั้งหมดเป็น 153 ราย (49%) และ 84 ราย (26.9%) ที่อายุเฉลี่ยเท่ากับ 5.5 ปีและ 6.3 ปี ตามลำดับ

สรุป : อุบัติการณ์ของรูรั่วผนังหัวใจห้องล่างชนิดใต้ต่อลิ้นหัวใจพัลโมนิค พบได้สูงในประชากรไทย การยื่นและรั่วของลิ้นหัวใจเอออร์ติค เป็นภาวะแทรกซ้อนที่พบได้บ่อยในผู้ป่วยเด็กกลุ่มนี้โดยพบได้ตั้งแต่ช่วงแรกของวัยทารก การศึกษาครั้งนี้ช่วยสนับสนุนว่าภาวะแทรกซ้อนของลิ้นหัวใจเอออร์ติคจะเพิ่มขึ้นตามอายุ

คำสำคัญ : รูรั่วที่ผนังหัวใจห้องล่าง, โรคหัวใจพิการแต่กำเนิด, ลิ้นหัวใจเอออร์ติกยื่น, คลื่นเสียงสะท้อนหัวใจ

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