นิพนธ์ต้นฉบับ

Aortopulmonary window: Echocardiography, hemodynamics, and operation

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Objective : *To study the clinical manifestations, hemodynamics, echocardiographic findings, operative findings, procedures and results.*

Design : *Retrospective study*

Setting : *Pediatric cardiology unit, Department of Pediatrics, Faculty of Medicine, Chulalongkorn University.*

Subjects : *Nine patients with age ranged from 1 month to 14 years and with diagnosis of aortopulmonary window*

Methods : *Clinical, radiographic, electrocardiographic, echocardiographic, cardiac catheterization, angiocardiographic, and operative finding data were retrospectively studied.*

Results : *Over a period of 10 years there were 9 patients with diagnosis of aortopulmonary window. Clinical features mimicked a large left-to-right shunt lesion but clues to correct diagnosis were signs of congestive heart failure with wide pulse pressure, bounding pulses and significant pulmonary hypertension even early in life. The combined use of two -*

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dimensional and Doppler echocardiography could accurately make a definite diagnosis, and cardiac catheterization data showed pulmonary hypertension and elevated pulmonary vascular resistance in all patients. The catheter manipulation directly from the main pulmonary artery into the ascending aorta was of diagnostic importance. Surgical closures were done in 7 patients, mostly by prosthetic patch closure through a transwindow approach and with good results.

Conclusion : *Aortopulmonary window is a rare congenital heart disease presented with early congestive heart failure and pulmonary hypertension. A definite diagnosis can be made by echocardiography. Early surgical correction before irreversible pulmonary vascular changes developed had good results.*

Key word : *Aortopulmonary window.*

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วัตถุประสงค์ : *ศึกษาลักษณะทางคลีนิค, ลักษณะทาง hemodynamic, ผลของการตรวจคลื่นเสียงสะท้อนหัวใจ, การผ่าตัด รวมทั้งผลของการผ่าตัดในคนไข้ aortopulmonary window*

วัสดุและวิธีการ : *ศึกษาแบบย้อนหลังในผู้ป่วย 9 ราย ที่ได้รับการวินิจฉัยว่าเป็น Aortopulmonary window ในเวลา 10 ปี ตั้งแต่ มกราคม 2529 ถึง พฤศจิกายน 2539 โดยอาศัยลักษณะทางคลีนิค, ภาพรังสีเอกซเรย์, การตรวจคลื่นหัวใจ, การตรวจคลื่นเสียงสะท้อนหัวใจ, การสวนหัวใจและการฉีดสี, รายงานจากการผ่าตัดและผลการผ่าตัด*

ผลการศึกษา : *ลักษณะทางคลีนิคที่ทำให้สงสัยว่า ผู้ป่วยเป็น Aortopulmonary window คือ ส่วนใหญ่จะมีอาการ congestive heart failuse ได้เร็วในระยะเดือนแรกๆ ของชีวิต ตรวจร่างกายพบมี wide pulse pressure, bounding pulses และมีภาวะ pulmonary hypertension ได้เร็ว, การตรวจโดยอาศัย Echocardiography ทั้ง two-dimension และ Doppler สามารถให้การวินิจฉัยได้ถูกต้องแม่นยำในผู้ป่วยทุกราย จากการสวนหัวใจ พบว่าทุกรายมีภาวะ pulmonary hypertension และมีค่า pulmonary vascular resistance สูงกว่าปกติ ตำแหน่งของ catheter จากการสวนหัวใจที่สามารถผ่านจาก main pulmonary artery ไปยัง ascending aorta ได้โดยตรง ช่วยสนับสนุนการวินิจฉัยโรค ผู้ป่วย 7 ราย ได้รับการผ่าตัดปิดโดยใช้ prosthetic patch โดย transwindow approach ซึ่งการผ่าตัดส่วนใหญ่ได้ผลดี*

สรุป : *Aortopulmonary window เป็นโรคหัวใจในเด็กที่พบได้ไม่บ่อย ส่วนใหญ่จะมาด้วยอาการ congestive heart failure และมีภาวะ pulmonary hypertension ได้เร็ว สามารถตรวจโดยอาศัย Echocardiography เพื่อยืนยันการวินิจฉัยได้ถูกต้องแม่นยำ และเป็นโรคที่ควรได้รับการผ่าตัด แก้ไขโดยเร็ว ซึ่งจะทำให้ผลการผ่าตัดดี*

Aortopulmonary window, or aortopulmonary septal defect, is a relatively rare congenital heart disease. In a review of patient records and autopsy specimens, Kutsche and Van Mierop [1] found aortopulmonary window in 0.2-0.6% of patients with congenital heart disease.

In the majority of cases, this defect is located in the proximal portion of the ascending aorta midway between the semilunar valves and the pulmonary bifurcation and has previously been designated as type I. Type II is a more distal defect, the distal border of which is formed by the pulmonary bifurcation and opens into the origin of the right pulmonary artery. Type III is a large defect involving the entire aortopulmonary septum.

Materials and Methods

From January 1986 through November 1996, nine patients with aortopulmonary window were studied. Six cases were detected, in 1996 likely due to more routine echocardiography. There were 6 boys and 3 girls. Their ages at the time of definite diagnosis ranged from 1 month to 14 years. All clinical, radiographic and electrocardiographic features were recorded. All were diagnosed by echocardiography before sent for cardiac catheterization and angiography for confirming the diagnosis and hemodynamic studies. 7 patients were operated. The operation were undertaken with cardiopulmonary bypass, supplemented by circulatory arrest. Most of them the lesions were closed: with dacron patch via transwindow approach.

Results

Clinical features

All were acyanotic. The clinical features were not specific but were similar to those of patients with a large left-to-right shunt. Signs of congestive heart failure, such as tachypnea, tachycardia, diaphoresis and recurrent respiratory tract infection, usually began in the first few months of life.

Physical examination revealed bounding pulses and wide pulse pressure. On auscultation, the second heart sound was accentuated at the pulmonic valve area in all patients. Also in all patients, an ejection systolic murmur was heard at the upper left sternal border. In 2 patients a grade 3-4/6 holosystolic murmur was heard at apex, correlated with mitral regurgitation that was confirmed by echocardiography. In two patients a grade 2-3/6 early diastolic murmur of pulmonic regurgitation coexisted. None had a continuous murmur heard at the upper left sternal border.

Table 1. Clinical and investigative data.

Patient No.	Sex	Age (mo)	Weight (kg)	Blood pressure (mm Hg)	EKG
1.	F	5	4.4	100/50	CVH
2.	M	7	5.6	120/60	CVH
3.	M	14 yr	37	113/42	LVH
4.	F	8	5	99/51	LVH, $\gamma SR'$ in V_{4R}'V_1
5.	M	9 yr	20	100/56	RAD, CVH
6.	M	14	8	94/41	CVH
7.	M	1	4.4	109/76	CVH
8.	M	3 yr	11	120/55	CVH
9.	F	4	4.2	108/55	CVH

Table 2. Investigative data.

Patients No.	O_2 step up in PA	Pulmonary artery pressure (mm Hg)	Pulmonary/systemic blood flow	PVR/SVR	Associated anomalies
1.	yes	38	1.64	0.77	MVP, MR
2.	yes	55	1.80	0.44	none
3.	yes	78	2.05	0.50	none
4.	yes	60	1.25	0.80	none
5.	yes	79	1.20	0.79	none
6.	yes	57	2.48	0.40	ASD secundum
7.	yes	37	16.40	0.06	none
8.	yes	59	1.45	0.67	Right sided aortic arch
9.	yes	67	3.08	0.28	ASD secondum, VSD

Radiographic features

A moderate to marked enlargement of the heart with prominent pulmonary vascular markings was noted in all patients, indicative of a large left-to-right shunt.

Electrocardiographic features

There were no characteristic findings. The QRS axis was between +45 and +105 in all patients. Almost all patients showed biventricular hypertrophy presented as a tall R wave and/or

deep Q wave in the left precordial leads, deep S wave in left precordial leads or large equiphasic QRS complexes in mid-precordial leads (Katz-Wachtel phenomenon). Isolated left ventricular hypertrophy was present in one patient. In one patient the electrocardiogram showed left ventricular hypertrophy and only mild degree of right ventricular hypertrophy suggested by an γSR' pattern in the right precordial leads.

Echocardiographic features

The two dimentional echocardiography could accurately diagnose aortopulmonary window. The left atrium and ventricle were dilated in all patients due to the large left-to-right shunt. The pulmonary arteries were significantly enlarged. The aortopulmonary windows could be seen by 2D-echocardiography, as dropout areas from high parasternal short axis and suprasternal long-axis cut of the aorta.

Doppler color-flow mappings were helpful in confirming the diagnosis by displaying evidence of flow across the echo-free space into the main pulmonary artery from the aorta. Pulsed-wave Doppler echocardiography was also helpful in confirming the diagnosis by showing retrograde diastolic flow in the aortic arch, or forward diastolic flow in the pulmonary trunk distal to the window.

Cardiac catheterization

All patients had pulmonary hypertension. The right ventricular and pulmonary arterial pressures were at or near systemic levels. Almost all had high pulmonary blood flow ($Q_p/Q_s > 1.5$) and elevated pulmonary resistance ratio ($R_p/R_s > 0.5$). There were O_2 step up in the pulmonary arteries in all patients due to left-to-right shunt at the arterial level. The catheter could be manipulated directly from the main pulmonary artery through the defect into the ascending aorta in all patients. This course of the catheter was of diagnostic importance.

Aortography at the level of the ascending aorta could demonstrate aortopulmonary window by showing opacification of the ascending aorta, the main pulmonary trunk, and the pulmonary arteries simultaneously. Aortography could also demonstrate the type and size of the defects.

Operation

Seven patients were referred to the pediatric cardiothoracic surgeon. The other two were a 9 year old boy and a 14 month boy who were not sent to surgery because the first had a very high pulmonary resistance ratio ($R_p/R_s = 0.79$) and had clinical signs of right-to-left shunt due to pulmonary hypertension presented by intermittent cyanosis, especially during exercise and aortic desaturation. The other had cerebral hypoxia after cardiac catheterization so the surgery was deferred until the neurological status would be more improved. From operative findings 7 of the 9 patient's AP windows were type I, and the other two were type II.

Table 3. Surgical findings and various procedures.

Patients No.	Type of AP window	Size of AP window (cm)	Technique (Approach)	Result
1.	I	1.5	division and suture	good
2.	II	1.2	dacron patch (transaortic and transpulmonic)	good
3.	II	1.7	dacron patch with one way valve flap (transaortic)	death
4.	I	1.5	dacron patch (transwindow)	good
5.	I	1.4	not surgery	
6.	I	1.0	surgery not yet	
7.	I	1.5	dacron patch (transwindow)	good
8.	I	2.5	dacron patch (transwindow)	good
9.	I	1.6	dacron patch (transwindow)	good

Operations were undertaken with cardiopulmonary bypass supplemented by circulatory arrest. Median stenotomy were done. Six patient's AP windows were closed by sauvage dacron patch. Four of these six patient's incisions were through the anterior part of the AP window. One patient's closure was done via a tranaortic approach, and the last via both a transaortic and a transpulmonic approach because of the large size of defect and for clear operative views. Only one patient's AP window closure was done by division and suture. In one patient (a 14 yr old boy) a one-way valve flap was inserted, to give the way to release pressure if the pulmonary artery pressure was far more than aortic pressure. One patient died perioperative period but others had good outcome. All had residual small AP windows detectable only by echocardiography.

Discussion

Aortopulmonary window is an extremely rare congenital heart disease. Most clinical features are presented with signs of congestive heart failure which usually begin in the first weeks of life due to large defects and high pulmonary blood flow. Clinically, this lesion often mimics either a VSD, PDA, or both. Distinguishing AP window from large PDA is extremely difficult by physical examination alone. Significant clinical symptoms in the first weeks of life are very unusual for PDA but not for AP window. The murmur of VSD usually is heard at the lower left sternal border, and the pulses are not bounding.

Nearly half of the patients have associated lesions. The most common associations are aortic origin of the right pulmonary artery,[5] interrupted aortic arch type A,[1,8] tetralogy of fallot,[6] anoma-

lous origin of the right coronary artery from the pulmonary artery and right sided aortic arch.[1,12]

But in our study there were only a few associated lesions, such as ventricular septal defect, atrial septal defect, right sided aortic arch and mitral valve prolapse.

Unlike truncus arteriosus, aortopulmonary window has never been reported in association with Digeorge syndrome. Interrupted aortic arch type B is associated with truncus arteriosus, whereas interrupted aortic arch type A is seen more frequently with aortopulmonary window. Thus AP window appears to be unrelated embryologically due to neural crest abnormality such as truncus arteriosus, transposition of the great arteries, and aortic interruption.[1]

At the time of definite diagnosis, all of our patients had pulmonary hypertension, high pulmonary blood flow and highly elevated pulmonary resistance ratios. Therefore, surgical correction should be performed early in life before irreversible pulmonary vascular changes develop. Clinically this lesion often mimics other large left-to-right shunt lesions such as VSD and PDA. Clinical clues to suspect aortopulmonary window and distinguish from other lesions are early CHF, wide pulse pressure, bounding pulses, accentuation of the second heart sound and loud systolic ejection murmurs at the left upper sternal border suggesting pulmonary hypertension even in early life. Echocardiography can accurately diagnose aortopulmonary window. All of the patients in our study were already diagnosed before being sent for cardiac catheterization by two dimensional, Doppler color flow and pulsed Doppler echocardiography. False dropout on two dimensional echocardiography is often seen in the aortopulmonary septal area of normal patients due to thin contiguous proximal aortic and pulmonary arterial walls.[3,11] Balaji et al described a " T " artifact at the edges of the defect to help distinguish it from normal dropout.[2] In addition, Doppler color flow can verify the defect by displaying flow across the echo-free space from the aorta into the main pulmonary artery, continuously.[3,11] Pulsed Doppler echocardiography can verify the defect by displaying retrograde diastolic flow in the aortic arch, or forward diastolic flow in the pulmonary trunk distal to the window.[2,3,11] The combined use of imaging and Doppler echocar-diography can accurately diagnose aortopulmonary window.

Since the first reported correction by Gross in 1952 by simple ligation, several types of surgical corrections have been attempted. Most authors recommended a tranaortic patch closure using a median stenotomy and cardiopulmonary bypass.[5,12] For four of our patients, the approach to the aortopulmonary window was through the anterior wall of the communication, the same as Johnasson L. et al reported in 1978.[7] This makes the operation performed more rapid due to good exposure. There is also a newly described technique to repair aortopulmonary window using an inverted pulmonary flap instead of a dacron patch, thus eliminating foreign material.[9,10]

Conclusion

Aortopulmonary window is an extremely rare congenital heart disease. All patients presented with clinical of congestive heart fallure and early pulmonary hypertension. Echocardiography both two-dimensional and Doppler could accurately diagnose these lesions. Surgical correction were undertaken with cardiopulmonary bypass, supplemented by circulatory arrest. Most of them were closed by prosthetic patch via transwindow approach with good results.

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