Aortopulmonary Window : A Case Report

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Abstract

Aortopulmonary window (A-P window) is a rare congenital heart disease. The majority of patients come to the hospital with congestive heart failure. If they are left untreated, irreversible pulmonary vascular hypertension inevitably occurs. Although the hemodynamic of this disease resembles large persistent ductus arteriosus, the treatment is quite different. We report one patient of A-P window, a 4 month-old girl, who presented with fever and dyspnea. On investigations, she had A-P window. We successfully treated her by closure the defect under cardiopulmonary bypass. We describe the technique we used.

Key word : Aortopulmonary Window, Case Report

Aorto-pulmonary window is a rare congenital heart disease. The first bedside diagnosis as AP window was described by Elliotson in 1830(1).

Symptoms and signs of congestive heart failure will develop in the first year of life. Decrease in pulmonary vascular resistance related to CHF in infants, pathophysiology of A-P window is similar to persistance ductus arteriosus. But there are many differences in the management among PDA and AP window. Operative mortality is 19 per cent in AP window,(5) but is nearly 0 in PDA. The objective of this report was to emphasize the important points in surgery, especially pitfalls which are of concern during perioperative and postoperative management.

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PATIENT AND METHOD

Patient

The patient was a 4 month-old Thai girl. Her mother brought her to see the doctor when she was 2 months old with the chief complaint of fever and dyspnea. She was found to have congenital heart disease. On physical examination, there was no cyanosis. Normal S_1 and increase S_2 sounds were noted. Bounding pulse was present. There was systolic ejection murmur grade IV/VI at left upper parasternal border and systolic ejection murmur grade II/VI at left lower parasternal border. The liver was one finger breadth below the right costal margin. The patient was referred to a pediatric cardiologist for evaluation and proper management. Chest radiography revealed cardiomegaly and increased pulmonary vasculature. An electrocardiogram showed normal sinus rhythm, normal axis and biventricular hypertrophy. On echocardiogra, there were dilated left atrium, intact interatrial septum, dilated right and left ventricle with intact interventricular septum. There was aortopulmonary window defect of 1 cm in diameter and left to right shunt. Cardiac catheterization was performed and there was significant stepping-up in oxygen saturation at the left pulmonary artery level.

Method

After the diagnosis was made, the patient was prepared for surgery. The operation was performed under general anaesthesia. She was in supine position. Median sternotomy incision was done and pericardial cavity was entered. Operative findings were aortopulmonary window defect connection ascending the aorta to the main pulmonary artery just proximal to the bifurcation and biventricular hypertrophy. The ascending aorta distal to aortopulmonary window was dissected and encircled with black silk no. 2. The superior and inferior vena cava (SVC, IVC) were dissected, encircled and prepared to be snugged with black silk no. 2. Right and left pulmonary artery (RPA, LPA) were encircled like that done on the SVC and IVC. The cardiopulmonary bypass (CPB) was conducted with full flow perfusion *via* aortic cannulation at the ascending aorta and venous return from bicaval cannulation. Both Lt. and Rt. pulmonary arteries, already encircled, were snugged before starting CPB (Fig. 1). The patient was cooled down to 25°C. The venting catheter was inserted to decompress LV *via* right superior pulmonary vein. When the required temperature was reached, the aorta was cross clamped at the level just above the aortopulmonary window. Antegrade cold crystalloid cardioplegia was instilled *via* the ascending aorta every 20 minutes. Arterial blood gas, electrolyte and activated clotting time were monitored during the CPB. Local hypothermia with ice was used. Anterior wall of aortopulmonary window was opened in a longitudinal direction at the junction between the aorta and pulmonary artery (Fig. 2). The defect was inspected and examined, no coronary abnormality was found. The defect was closed by Gore-tex patch with running sutures, starting from the posterior ridge of the defect using prolene 5-0 (Fig. 3).

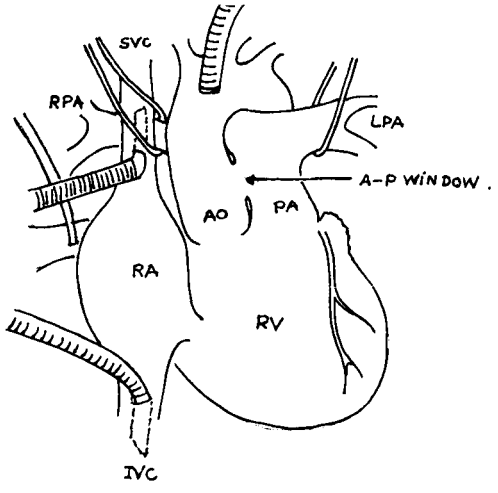

Fig. 1. Show A-P window and cannulation technique for cardio-pulmonary bypass.

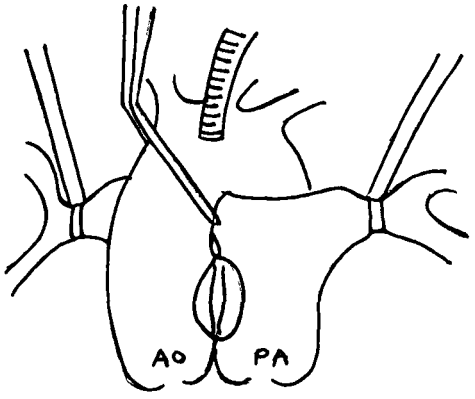

Fig. 2. Show incision at anterior part of A-P window.

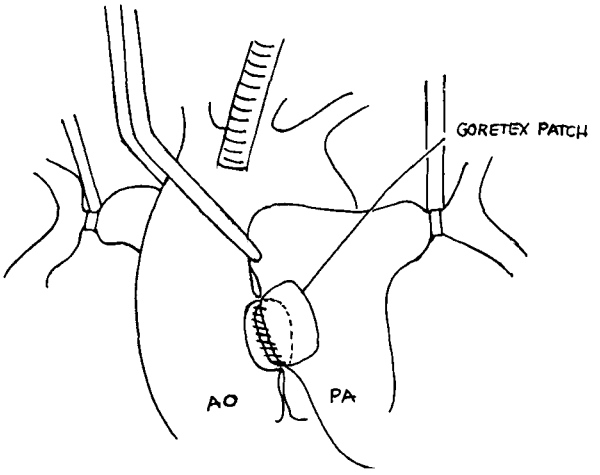

Fig. 3. Closure of A-P window by using goretex and prolene 5,0 continuous suture at the posterior edge of A-P window.

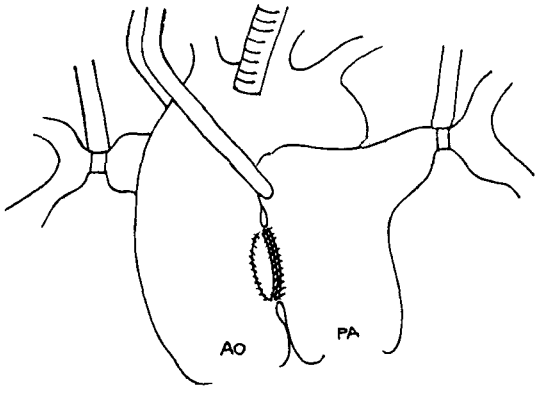

Fig. 4. Anterior part of A-P window was closed double layers by continuous suture with prolene 5,0.

When the sutures reached the anterior part, both sides of the anterior part (aortic and pulmonary rim) were sutured together with Gore-tex patch in between (Fig. 4). After completion of the closure, the patient was rewarmed. The aorta was declamped and air was allowed to come out of the aorta. The heart resumed sinus rhythm spontaneously. When the oral and rectal temperature was 37°C. and 35°C. respectively, the CPB was weaned off. Hemodynamic status was stable. A small catheter was inserted through the right superior pulmonary vein for LAP monitoring. Inotropic drug was started (dopamine 2 μg./kg./min.). Hemostasis was secured. All cannulations were removed. Two intercostal drainages were placed in both pleural cavities. The pericardium was left open. Sternal defect was left open for delayed closure because there was marked cardiomegaly and the pulmonary artery pressure was so high near systemic blood pressure. The defect was covered with silastic sheath sutured to the skin. Aortic cross clamp time was 1 hour 10 min and CPB time was 1 hour 28 min.

In the CCU arterial pressure, left atrial pressure and arterial blood gas were monitored, postoperatively. Postoperative hemodynamic status was stable. The patient gained consciousness at about 6 hours postoperatively. The sternal defect was closed on the 3rd day uneventfully. The patient was extubated on the next day. Inotropic drug dosage was reduced and taken off.

DISCUSSION

Symptoms and signs of congestive heart failure develop early in neonates or infants, which may be associated with a decrease in pulmonary vascular resistance. So decision making to perform surgery is necessary for these patients[2]. Open heart surgery for closure of AP window is highly recommended even in neonates or infants with low body weight[3] (< 4 kg). Difficulties in closure of AP window by open heart surgery are, how to protect the lungs during cardio-pulmonary bypass from aortic run off to pulmonary artery *via* AP window, how to perfuse the cardioplegia to preserve the heart, and how to get good exposure during closure of AP window. We used two venous cannulae at SVC and IVC and cannulated aorta as high as possible. Then we encircled both the right and left pulmonary arteries to control blood from running off to the lungs[4]. This enabled effective perfused cardioplegia to the heart later. Also, the incision to expose the AP window is important because of limitation from aortic cannular. Exposure by opening directly at the anterior wall of the AP window may be adequate for good visualization which makes profound hypothermia and circulatory arrest unnecessary.

Pulmonary hypertensive crisis may develop postoperatively, so delayed sternal closure is recommended in neonates or small infants with

marked cardiomegaly with small collections of clotted blood in the pericardium or pulmonary hypertension crisis may deteriously effect hemodynamic status and delayed sternal closure may be beneficial.

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เอออร์โต พัลโมนารีย์ วินโดว์ : รายงานผู้ป่วย

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Aortopulmonary window (A-P window) เป็นโรคหัวใจพิการแต่กำเนิดที่พบได้ไม่บ่อยนัก ผู้ป่วยส่วนใหญ่จะมาพบแพทย์ด้วยอาการของหัวใจล้มเหลว ถ้าไม่ได้รับการรักษา ผู้ป่วยก็จะเกิดภาวะความดันเลือดในปอดสูง ถึงแม้ว่าการไหลเวียนของเลือดจะคล้ายกับผู้ป่วยที่มี persistent ductus arteriosus (PDA) ขนาดใหญ่ก็ตาม แต่การผ่าตัดรักษานั้นไม่เหมือนกันเลย ผู้รายงานได้รายงานผู้ป่วย 1 รายเป็นเด็กหญิง อายุ 4 เดือน มาพบแพทย์ด้วยเรื่องไข้และหายใจหอบ จากการตรวจพบว่า ผู้ป่วยเป็น A-P window จึงได้ทำการผ่าตัดแก้ไขเป็นผลสำเร็จ โดยใช้เครื่องปอดและหัวใจเทียม และได้รายงานวิธีการที่ใช้ทำผ่าตัดด้วย

คำสำคัญ : เอออร์โต พัลโมนารีย์ วินโดว์, รายงานผู้ป่วย

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