Case Report

ECHOCARDIOGRAPHIC DIAGNOSIS OF AORTOPULMONARY WINDOW IN A 4-DAY-OLD BABY

(Received 18 February, 1999)

Figen Akalin, M.D.* / Funda Öztunc, M.D.** / Serap Turan, M.D.***

* Sub-department of Pediatric Cardiology, Department of Pediatrics, School of Medicine, Marmara University, Istanbul Turkey.
** Institute of Cardiology, Istanbul University, Istanbul, Turkey.
*** Department of Pediatrics, School of Medicine, Marmara University, Istanbul, Turkey.

ABSTRACT

Aortopulmonary window is a rare cardiac abnormality leading to pulmonary vascular disease unless diagnosed in early infancy. Echocardiographic diagnosis may be difficult if it is not searched for. Aortopulmonary window must be considered in the differential diagnosis of patent ductus arteriosus and truncus arteriosus because of their similar clinical findings and natural course. Early diagnosis is essential for prevention of complications, especially the Eisenmenger syndrome. We herein present an aortopulmonary window type I case diagnosed by echocardiography and treated surgically in a four-day-old baby.

Key Words: Aortopulmonary window, Echocardiography, Congenital heart disease

INTRODUCTION

Aortopulmonary window (APW) is a rare cardiac abnormality occurring in about 0.2% of the patients with congenital heart disease (1,2). APW was first described by Elliotson in 1830 in an autopsy study (3). The septum between the main pulmonary artery and ascending aorta is incomplete in this lesion (1,2). Several types of APW have been described. Type I defect is located at midway between the semilunar valves and the bifurcation of pulmonary artery. Type II defect is more distally located and the right pulmonary artery usually originates from aorta in this type. Type III defects are large lesions combining the first two types (1,2). Early diagnosis is essential for prevention of pulmonary vascular disease. Clinical findings and natural course of the patients with this abnormality may be similar to truncus arteriosus or large patent ductus arteriosus and APW must be considered in differential diagnosis of these pathologies (1-3).

Complete diagnosis of APW with echocardiography without angiography is reported in recent years (4). We herein present a case of APW diagnosed by echocardiography in a four-day-old baby and review the literature about this rare pathology.

CASE REPORT

A 4-day-old girl has been referred to our hospital because of a murmur noticed during routine examination. She was the first child of non-consanguineous healthy parents. She was born following an uneventful pregnancy.

On physical examination she was an acyanotic, active baby. She was mildly tachypneic and a continuous murmur was audible at the third intercostal region on the left side of the sternum. Second heart sound was normal. The rest of the physical examination was within normal limits.

Chest x-ray showed mild enlargement of the cardiac shadow with increased pulmonary vascular markings and the electrocardiogram showed right axis deviation and right ventricular hypertrophy which was normal according to the age of the baby.

Echocardiographic examination was performed by using an ATL Ultramark 9 echocardiography machine and a 5MHz probe. The left atrium and the left ventricle was found to be enlarged comparing with the right side of the heart. Ventricular contractions were normal, mitral valve was mildly regurgitant. Patency of foramen ovale was detected. Precordial and subcostal short axis views demonstrated an aortopulmonary window 5
mm in diameter, which was about 1.5 cm above the pulmonary valve (Figs. 1 and 2). The shunt through the defect was bi-directional. Pulmonary artery was not enlarged.

The patient was operated under cardiopulmonary bypass surgery and the defect between the ascending aorta and main pulmonary artery was closed successfully. The baby is now doing well following an uneventful postoperative period.

DISCUSSION

Aortopulmonary window is a rare cardiac abnormality and it occurs in about 0.1-0.2% of the patients with congenital heart disease. APW is reported to be present in 0.4% of the patients undergoing surgery for congenital heart disease (1,4). The literature includes small series and case reports of APW and about 350 patients have been reported up to date (1).
The septum between the ascending aorta and pulmonary artery is defective in this abnormality. Our patient had a proximal defect located between the semilunar valves and bifurcation of the pulmonary artery and it was decided to be a type I defect (1,2).

Most of the patients with APW has associated abnormalities (50-80% of the patients). Patency of ductus arteriosus (72%), interrupted aortic arch type A or coarctation of aorta (13-20%), interrupted aortic arch type B (6%), tetralogy of Fallot (6%), right aortic arch (5-20%), agenesis of ductus arteriosus (20%), ventricular septal defect (20%), right pulmonary artery originating from aorta (32%), anomalous coronary artery originating from pulmonary artery (23%), transposition of great arteries (10%) are the common associated abnormalities (3,4). Isolated defects are less common and our patient had an isolated APW. Although association of secondum type atrial septal defects are reported as case reports; the atrial septal defect found in our patient was a small one and it may be considered as a patent foramen ovale in neonatal period. The mild mitral regurgitation detected by color flow Doppler was thought to be caused by the left ventricular overload due to APW.

Clinical findings such as dyspnea, tachycardia, tachypnea, diaphoresis, congestive heart failure and failure to thrive may be present in patients with APW at presentation (1,2). Our patient only had a precordial murmur and mild tachypnea during initial examination probably because of the early diagnosis and continuing physiologic pulmonary hypertension.

Clinical course and prognosis of APW is similar to other cardiac abnormalities causing large left to right shunt, and Eisenmenger syndrome may develop unless it is treated early. Pinto et al (5) have reported that about one third of the patients with APW between 2 to 38 years of age had Eisenmenger syndrome.

Two dimensional echocardiography is usually the method used for diagnosis of APW and the defect may be visualized by using parasternal and subcostal views. Presence of “T” artifact at the margins of the defect is helpful for differentiation from “drop out” that may be seen at the same region. Echocardiographic diagnosis may be difficult in some patients especially in the presence of associated abnormalities (6,7). Color flow Doppler is beneficial in these patients. Occasionally catheterisation and angiography is necessary before surgery. However; in the series of Bertolini et al (3) 8 of 10 cases had undergone cardiac catheterisation. Magnetic resonance imaging is also useful for demonstration of detailed anatomical features (8). In our patient two dimensional echocardiography was sufficient for diagnosis and color Doppler examination showed the turbulent flow in the pulmonary artery.

Surgery is accepted to be the choice of treatment because of the potential complications of APW. Mortality related with surgery is low and long term prognosis after surgery is excellent (9).

In conclusion; APW is a rare cardiac abnormality that may lead life threatening complications. It must be considered in infants with signs and symptoms of left to right shunt and an echocardiographic examination must be performed.

REFERENCES