Case Report

A Rare Case of Adult Aortopulmonary Window With Eisenmengerization Complicated by Severe Aortic Stenosis and Ortner Syndrome

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ABSTRACT

Aortopulmonary window (APW) is a rare congenital cardiac condition resulting in a communication between the aorta and the pulmonary artery. APW can occur as a single lesion or may be associated with other cardiac defects such as ventricular septal defects, atrial septal defects, and tetralogy of Fallot. More than 50% of APW cases are associated with other additional congenital cardiac defects that require surgery. Most patients can develop congestive heart failure at an early age due to left-to-right shunts. The patient survival rate with untreated APW is very low, with a mortality rate of 40% in the first year. Survival into adulthood without treatment is very uncommon. We herein describe a 62-year-old man with Eisenmengerized APW with acquired severe aortic stenosis, mild pulmonary valvular stenosis, and Ortner syndrome without surgery. Ortner syndrome is mainly caused by the compression of the recurrent laryngeal nerve due to the dilatation of the pulmonary artery and the aorta. ***(Iranian Heart Journal 2022; 23(3): 114-119)***

KEYWORDS: Aortopulmonary window, Eisenmengerization, Aortic stenosis, Ortner syndrome, Echocardiography

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Aortopulmonary window (APW) is a rare congenital cardiac condition resulting from the absence of the wall that is present between the aorta and the pulmonary artery. It accounts for 0.2% to 0.6% of all congenital cardiac defects.[1]

More than 50% of APW cases are associated with other additional congenital cardiac defects that require surgery.[2] Most patients could develop congestive heart failure at an early age as a consequence of left-to-right shunts. The survival rate of patients with untreated APW is very low, with a mortality rate of 40% in the first year. Some patients with APW may survive into adulthood. We herein describe a 62-year-old male patient with Eisenmengerized APW presenting with acquired severe aortic stenosis, mild pulmonary valvular stenosis, and Ortner syndrome without surgery. Ortner syndrome was caused mainly because of the compression of the recurrent laryngeal nerve due to the dilatation of the pulmonary artery and the aorta.

Case Report

A 62-year-old male patient was admitted to Kasturba Medical College (KMC), Manipal,

because of exertional dyspnea and fatigue of 12 years' duration, as well as recurrent coughs of 10 years' duration and hoarseness of voice. He had a past history of fever and difficulty in breathing at 5 years of age and was diagnosed with a heart problem, for which he received medications. In 2009, the patient suffered an episode of loss of consciousness when he was standing at a bus stop in Shivamogga. He was transferred to KMC, where he regained consciousness after 4 hours. The patient was told that he had heart problems and was being managed for the condition.

On physical examination, he had a blood pressure of 110/60 mm Hg, a heart rate of 70 beats per minute, cyanosis and clubbing, an elevated jugular venous pressure (8 cm from the sternal angle), palpitation, and a heaving apex at the fifth intercostal space and the mid-axillary line. Body mass index was 16.6 (height=153 cm and weight=39 kg). Auscultation revealed late peaking murmurs at the aortic area with thrills. Additionally, P2 was long with a narrow split, and there were diastolic murmurs.

An electrocardiogram suggested sinus rhythm with a right bundle branch block and biventricular hypertrophy with right ventricular (RV) strain and biatrial enlargement (Fig. 1).

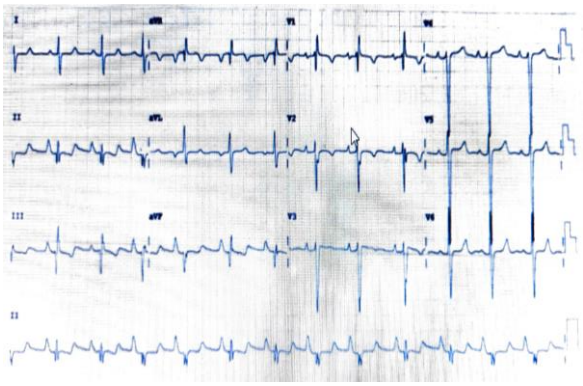

Figure 1: The electrocardiogram shows a right bundle branch block and biventricular hypertrophy with a right ventricular strain pattern and biatrial enlargement.

Transthoracic echocardiography showed biventricular hypertrophy with severe RV hypertrophy. Further, dilatation was observed in the RV, the right atrium, and the pulmonary artery. RV systolic pressure was approximately 87 mm Hg with a tricuspid regurgitation jet (Fig. 3A), as well as fair RV function with a tricuspid annular plane systolic excursion (TAPSE) value of 15 mm. Moreover, the left ventricular "D Sign" was observed (Fig. 4).

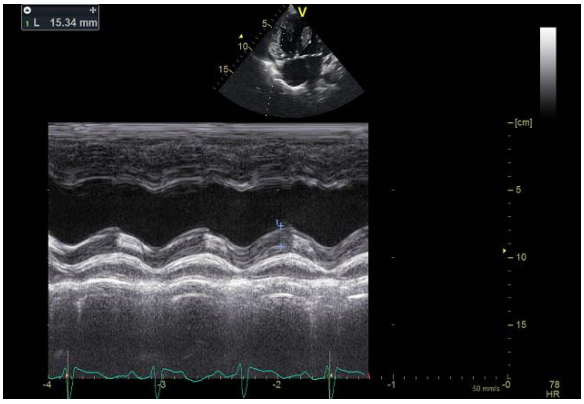

Figure 4: The image shows the M-mode echocardiographic image of the tricuspid annular systolic excursion (15 mm), representing fair right ventricular function.

Left ventricular systolic function was fair with an ejection fraction of 59% and an end-diastolic dimension of 36 mm (Fig. 2A & 2B). A large defect was observed between the left wall of the aorta and the right wall of the pulmonary artery, approximately 15 mm in size. A Doppler study showed a bidirectional shunt across the defect (Fig. 5A & 5B). Additionally, sclerotic aortic valve disease was observed with severe aortic stenosis. The valve area was 0.8 cm^2 by continuity equation (max velocity=4.2 m/s, peak pressure gradient=73 mm Hg, and mean pressure gradient=44 mm Hg). There was also mild aortic regurgitation. Concentric left ventricular hypertrophy was observed due to the presence of aortic stenosis (Fig. 3B & 3C).

Chest radiography (Fig. 7) showed bilateral hilar prominence and pruning with

cardiomegaly. The final diagnosis was APW with severe calcific aortic stenosis. Calcium was deposited over all 4 valves, and mild valvular pulmonary stenosis was noted (max velocity-=2.8 m/s, peak pressure gradient=32 mm Hg, and mean pressure gradient=20 mm Hg). In addition, there was mild pulmonary regurgitation and calcium extending into the RV outflow tract region. The patient was diagnosed with Eisenmenger syndrome, which precluded surgery. He was recommended to receive medicine to reduce pulmonary artery pressure.

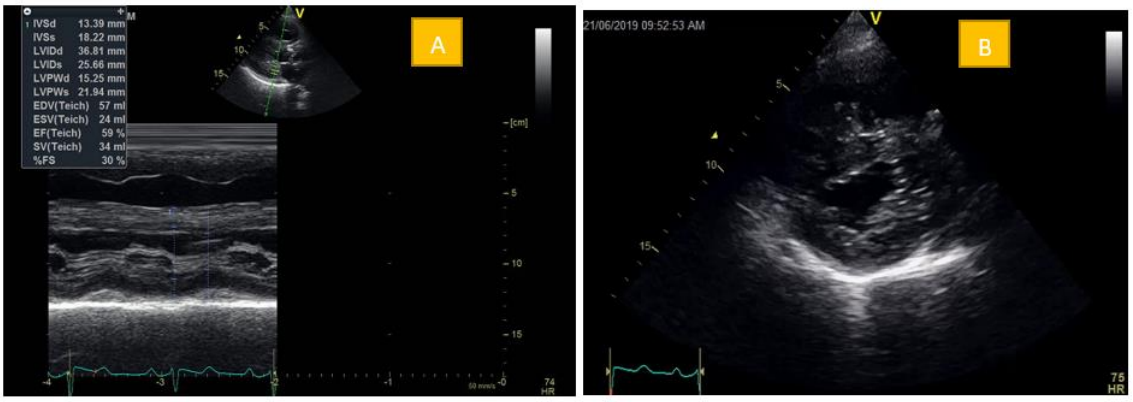

Figure 2A & 2B: The M-mode study shows left ventricular (LV) function, LV end-diastolic dimension, and LV concentric hypertrophy.

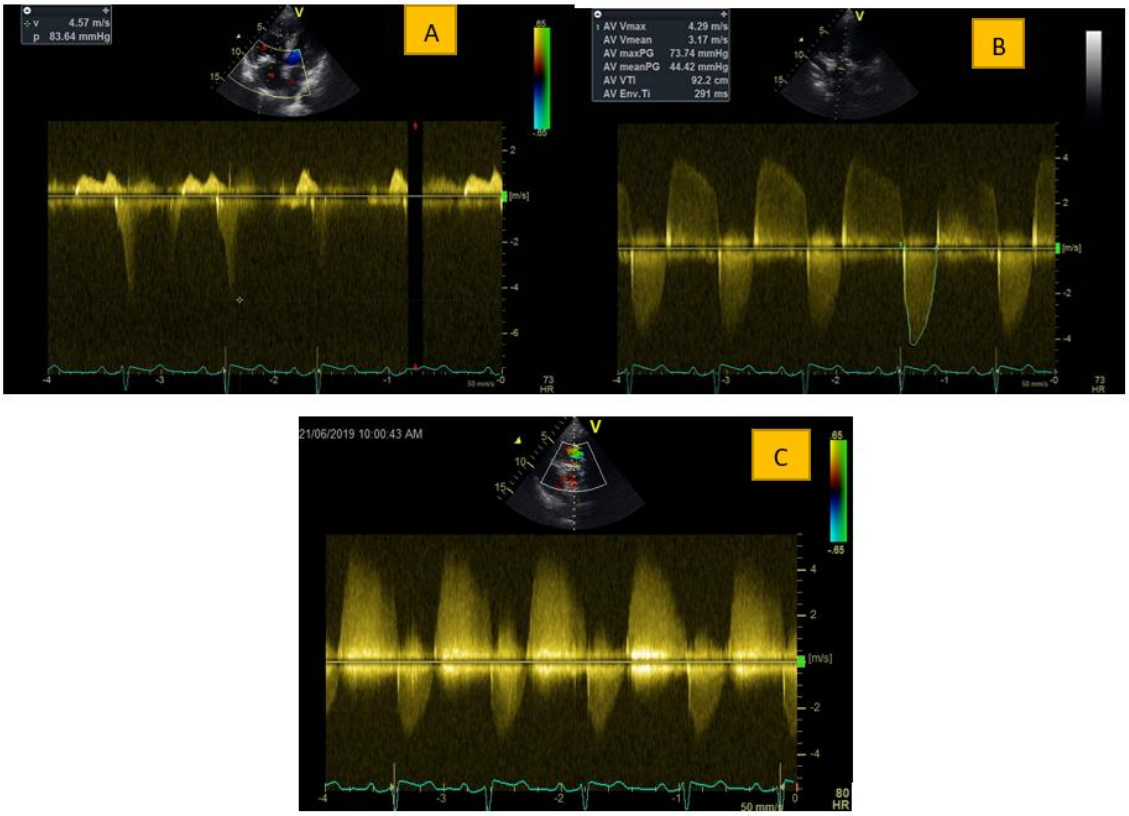

Figure 3A, 3B, & 3C: Tricuspid regurgitation (TR) Doppler spectrum reveals a TR velocity of 4.5 m/s with a pressure gradient of 83 mm Hg. Doppler across the aortic valve in the apical 5-chamber view shows the gradient of severe aortic stenosis, and Doppler across the pulmonary valve shows a mild gradient.

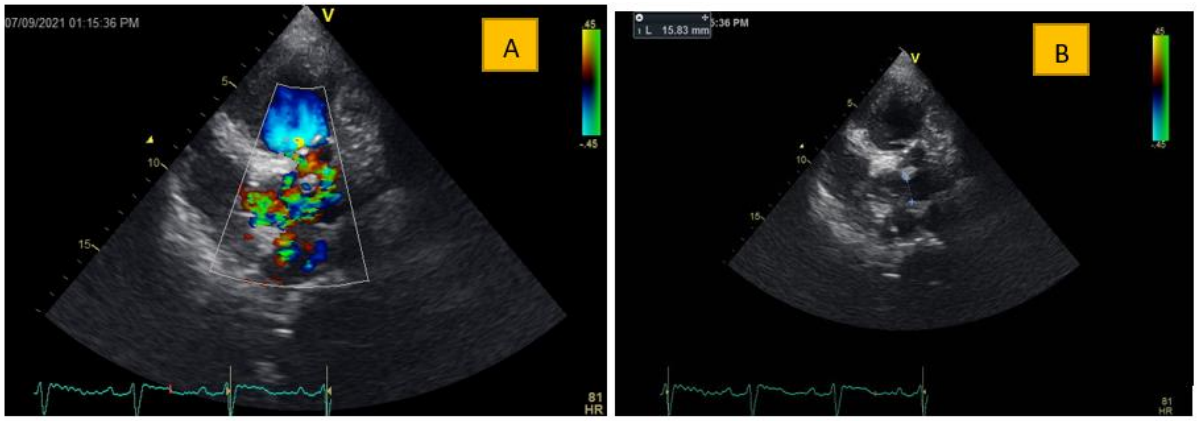

Figure 5A & 5B: The Color Doppler study shows a shunt across the defect, and the 2D image shows the size of the defect.

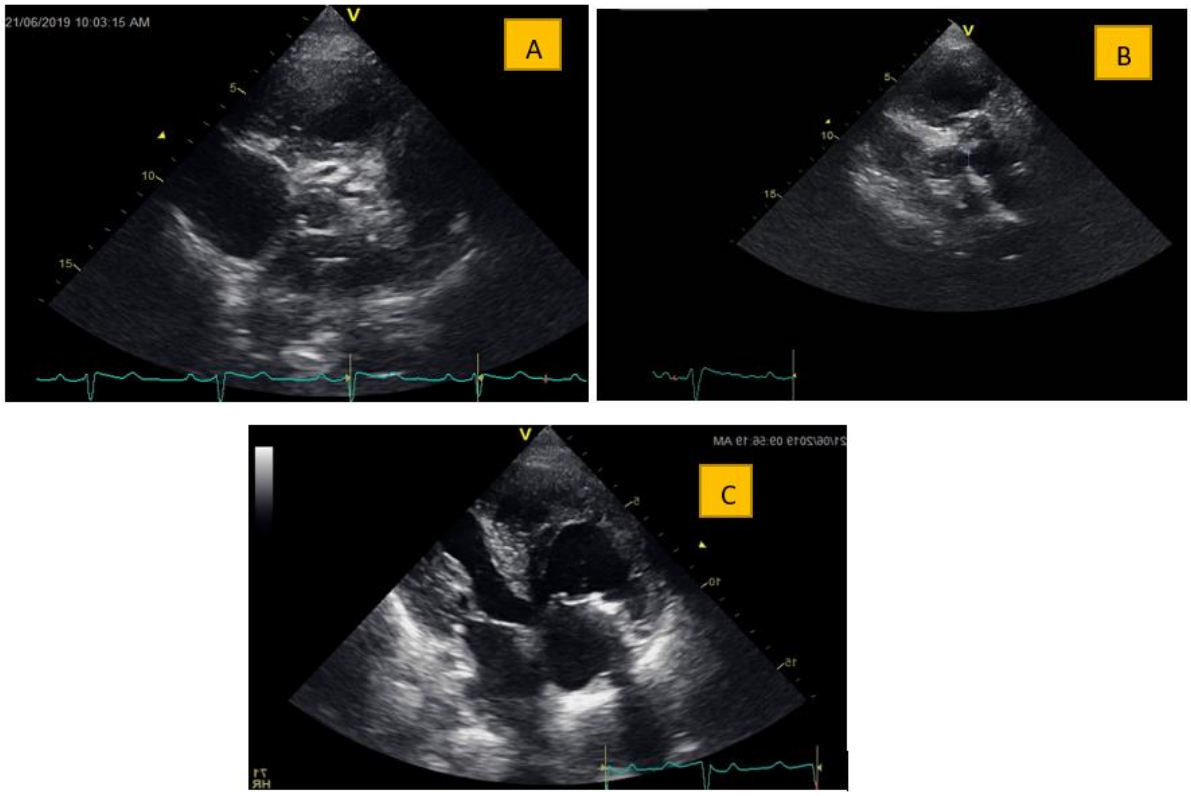

Figure 6A, 6B, & 6C: The image shows calcium deposition in the aortic, pulmonary, and atrioventricular valves, respectively.

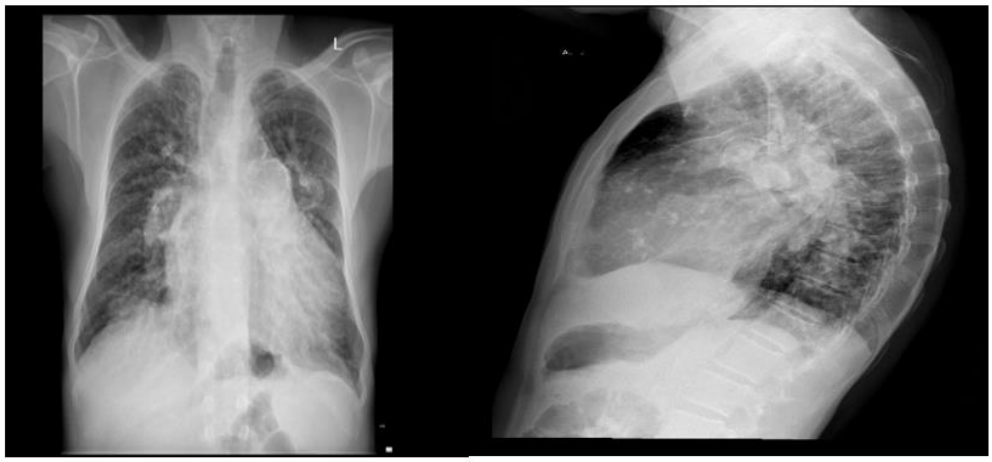

Figure 7: The chest X-Ray shows cardiomegaly and increased pulmonary marking (suggestive of pulmonary venous hypertension) with hilar prominence. Peripheral pruning is also present, and calcification is noted in the pulmonary bay region.

DISCUSSION

APW is a very rare congenital cardiac malformation. It can be defined as a communication between the main pulmonary artery and the ascending aorta resulting in the incomplete separation of these 2 arteries during embryogenesis. APW can occur as a single lesion or may be associated with other cardiac defects such as ventricular septal defects, atrial septal defects, and tetralogy of Fallot. Our patient was diagnosed with type I APW, as well as severe aortic stenosis and mild pulmonary stenosis. APW can be divided into 3 types. In type I, the defect is located in the proximal part of the ascending aorta and the main pulmonary artery. Type II defects are located distally near the bifurcation of the main pulmonary artery and the ascending aorta. Type III defects comprise a combination of types I and II, and these defects can also extend into the pulmonary bifurcation region.

APW was first pathologically described by Ellioton in 1830. A high mortality rate is associated with uncorrected APW, along with young age at presentation, a poor functional class, RV hypertrophy, and arrhythmias. A case report by Angela Koh et al in 2007 described a case of APW who survived until the sixth decade of life and died at the age of 60. Niles et al described a similar case of a female patient who survived until the age of 46. These patients had no associated complicated abnormality, and their condition indicates that biological factors play a very important role in the prognosis.

The present case report is unique because the existing literature contains no case of APW along with acquired severe aortic stenosis. Further, our patient had Ortner syndrome due to the compression of the recurrent laryngeal nerve.

CONCLUSIONS

APW is a very rare cardiac congenital anomaly, and survival in adults without treatment is very uncommon. Our patient, 62 years of age, is being managed medically for irreversible pulmonary hypertension. This case highlights the importance of acquired severe aortic stenosis with Eisenmengerized APW. Diffuse calcification can be seen because of degenerative disease or secondary to calcium disorder (though calcium and parathyroid hormone levels

were within normal limits). Treatment for aortic stenosis remains a higher risk situation and options like transcatheter aortic valve replacement can be considered.

Declarations
We obtained the patient's consent to anonymously publish this case.

Availability of Data and Material
Not applicable

Conflict of Interest
None declared

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None

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