International Journal of Cardiology Sciences
Online ISSN: 2664-9039, Print ISSN: 2664-9020
Received: 05-04-2020; Accepted: 20-04-2020; Published: 06-05-2020
www.cardiologyjournals.net
Volume 2; Issue 1; 2020; Page No. 08-11

Aorto pulmonary window manifesting as Eisenmenger syndrome at initial presentation in an adolescent female: A rare case

Sreekanth Y Bellamkonda[1], Naga Chaitanya Karella[2], Praveen Nagula[3], Srinivas Ravi[4], Krishna Mala Konda Reddy P[5]

[1, 2] Resident in Cardiology, Department of Cardiology, Osmania General Hospital, Hyderabad, Telangana, India

[3] Assistant Professor in Cardiology, Department of Cardiology, Osmania General Hospital, Hyderabad, Telangana, India

[4] Associate Professor in Cardiology, Department of Cardiology, Osmania General Hospital, Hyderabad, Telangana, India

[5] Professor and Head of the Department of Cardiology, Department of Cardiology, Osmania General Hospital, Hyderabad, Telangana, India

Abstract
Aortopulmonary window a rare congenital abnormality of the great vessels, manifests as heart failure in the early age with less chances of survival beyond first decade. It presenting as the Eisenmenger syndrome is rare. A 16 year female with breathlessness and bluish discoloration of 3 months duration on evaluation found to have a large aortopulmonary window with reversal of shunt on echocardiography. She was kept on pulmonary vasodilators and oral anticoagulants. she improved symptomatically as assessed by improved six minute walk test distance.

Keywords: Aortopulmonary window , Pulmonary Hypertension , Eisenmenger Syndrome, Pulmonary vasodilators

Introduction

A 16 year female came with breathlessness on doing less than her usual physical activity with bluish discoloration of her fingernails since 3 months. There was no history of chest pain, palpitations, syncope. There was no history of similar complaints in the past. There was no history of cyanotic spells or recurrent respiratory tract infections during her childhood. She was born out of nonconsaguineous marriage with no significant history perinatally and childhood.

On examination, her saturation was 85% on room air, she was cyanotic and pan digital clubbing was noticed (figure1). She was conscious, coherent and well oriented. The pulse was normal 90/min, regular, with no special character. Her blood pressure recorded 100/70 mm Hg over the arm in sitting position. The cardiac examination revealed precordial bulge, with grade 3/3 parasternal heave and heaving apex. There was dull note on percussion in the pulmonary area with palpable P2. On auscultation, there was loud P2, and pan systolic murmur at the left lower sternal border increasing with inspiration. Other systemic examination was normal.

On biochemical profile, she was found to have polycythemia with hematocrit of 56% and other lab parameters were normal. Her electrocardiogram showed sinus rhythm with right axis of +120 and right ventricular hypertrophy and secondary repolarization changes in v1-v3 (figure 2). The chest x ray showed cardiomegaly with RV apex and pulmonary oligemia (figure3).

The two dimensional echocardiography showed dilated right atrium, right ventricle with right ventricular hypertrophy (figure4). There was defect between the aorta and the main pulmonary artery in the suprasternal view with opacification of the both great arteries on bubble contrast echo(figure 5). There was reversal of shunt on color Doppler. The tricuspid regurgitation velocity was 4.5m/sec with pressure gradient of 78 mm Hg(figure 6). The inferior vena cava was dilated with less than 50%variation with respiration. There was D shaped left ventricle in the short axis view(figure 7). The Pulmonary valve M mode showed wandering W sign suggestive of severe pulmonary hypertension(figure 8). The pulse wave Doppler of the Right ventricular outflow tract showed pulmonary acceleration time of 78 msec(figure 9). The right ventricular function was normal as noticed by TAPSE of 17 mm and TAPSV of 12 cm/sec.. The left ventricular function was normal(figure 10).There was mitral regurgitation secondary to conformational change in the left ventricle. The aortic arch was normal with left side aorta. There was no PDA or any other abnormalities. A diagnosis of large Aortopulmonary window with reversal of shunt and severe pulmonary hypertension and normal biventricular function was made.

For correct delineation of the defect she was subjected to contrast enhanced computed tomography angiography of the great vessels and the heart. There was 2.1 cm type 1 aortopulmonary defect noticed and no other abnormalities notified (figure11).

She was kept on pulmonary vasodilators (SILDENAFIL 20mg BD) with gradual uptitration upto 20mg TID. She was kept on oral anticoagulation warfarin 4 mg after attaining target INR of 2.5.She was initially assessed of her cardiopulmonary efficacy by six minute walk test. She walked 230 meters in six minutes. She is under regular follow up with improvement of the six minute walk distance to 360 meters after treating for four months.

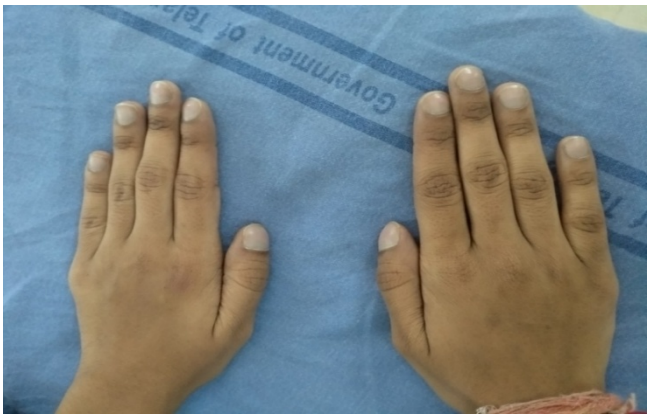

Fig 1: Pandigital clubbing and peripheral cyanosis of both hands

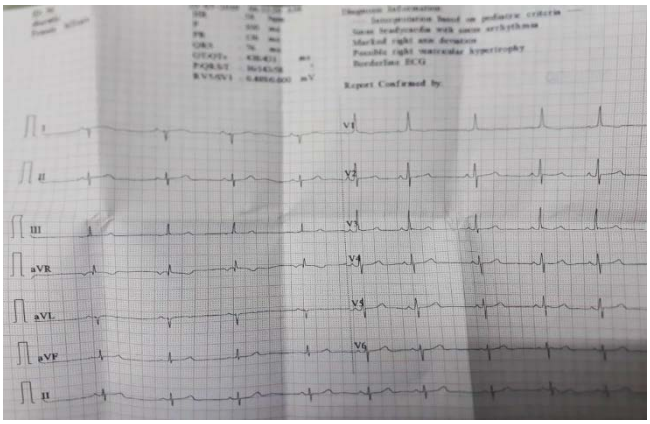

Fig 2: Twelve lead electrocardiogram showing sinus rhythm, right axis deviation of +150 degrees, and right ventricular hypertrophy (monophasic R wave in lead V1 and prominent S wave in the V6).

Fig 3: Chest x ray posteroanterior view showing mild cardiomegaly (right atrial and right ventricular enlargement) with right ventricular apex and pulmonary oligemia. Mild scoliosis of the thoracic spine is also noted.

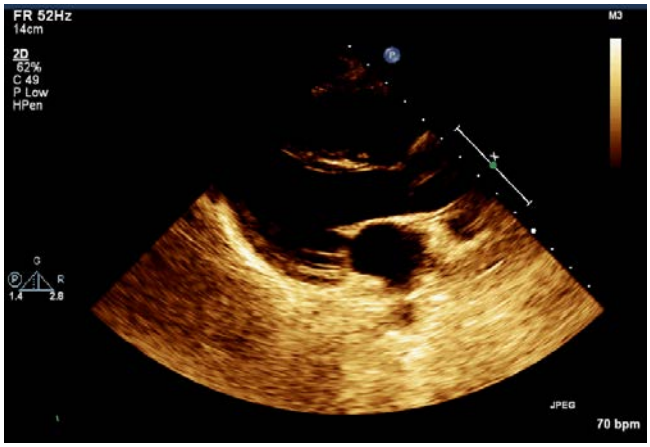

Fig 4: Parasternal Long axis view showing dilated right ventricle with interventricular septum pointing towards the left ventricle in early diastole.

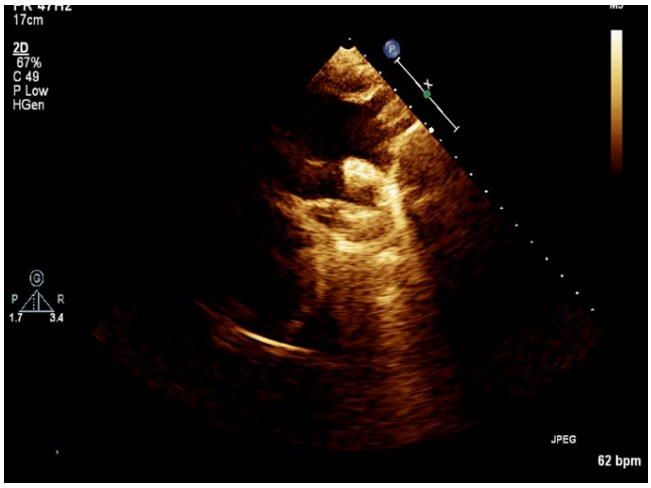

Fig 5: Suprasternal view showing the aorta and the great arteries with defect between the aorta and the pulmonary artery(bold asterix).

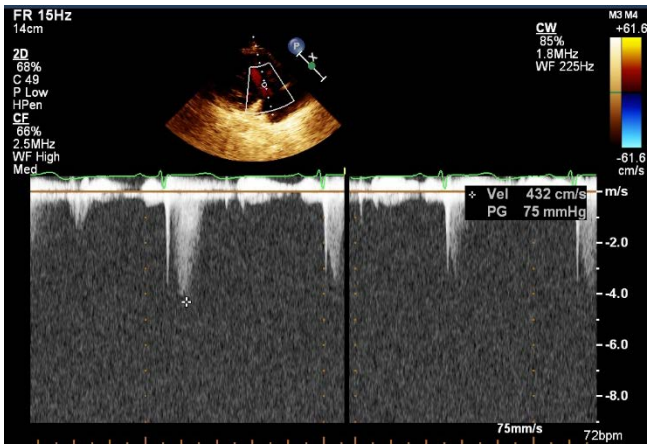

Fig 6: Modified parasternal long axis view of the right ventricular inlet view, showing the continuous wave Doppler of the tricuspid regurgitation with a pressure gradient of 75 mmhg.

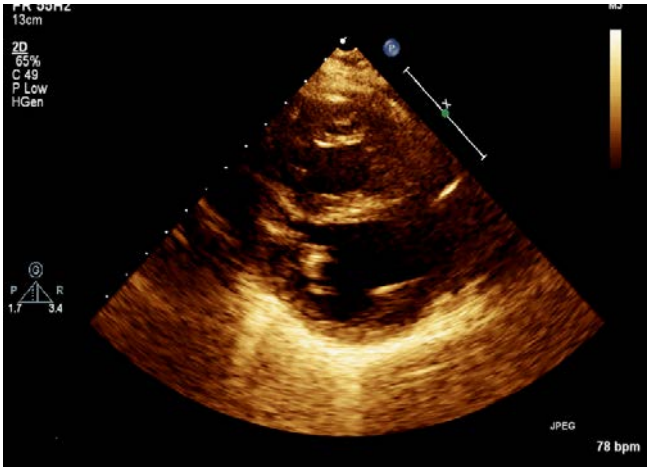

Fig 7: Parasternal shortaxis of the right and left ventricle showing flattened interventricular septum suggestive of right ventricular pressure overload.

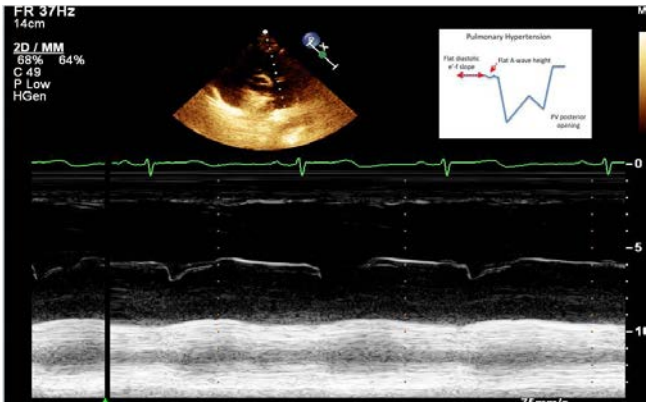

Fig 8: M mode of the pulmonary valve showing the wandering W wave sign suggestive of severe pulmonary arterial hypertension, with small a wave. The inlet showing the graphical representation of the wave pattern.

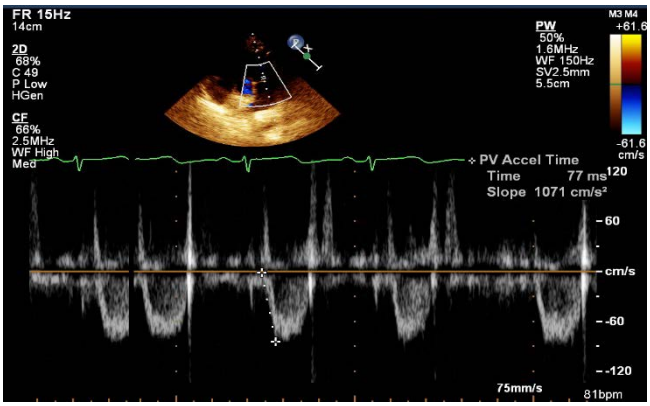

Fig 9: Pulse wave Doppler interrogation across the pulmonary valve showing the pulmonary acceleration time of 77 msec suggestive of severe pulmonary hypertension.

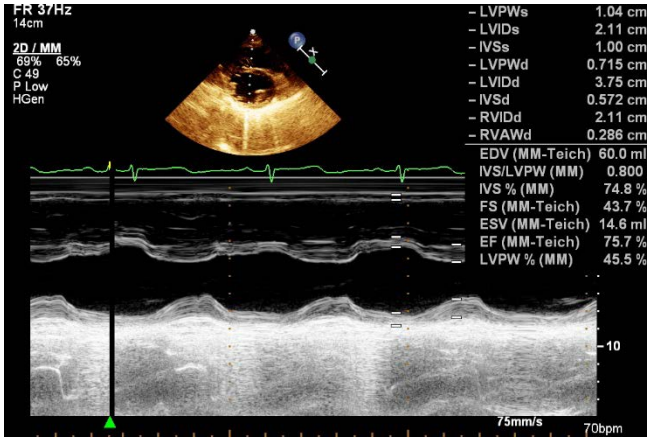

Fig 10: Parasternal short axis view at the level of the papillary muscles of the left ventricle showing the left ventricular ejection fraction of 75%.

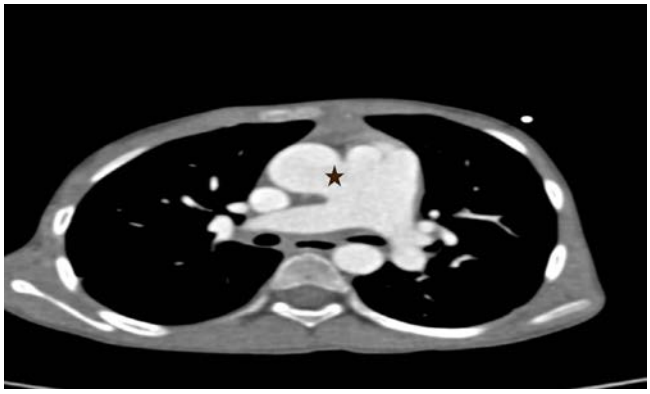

Fig 11: The contrast enhanced computed tomography of the great arteries showing the aortopulmonary window between the ascending aorta and the pulmonary artery before its bifurcation. The main pulmonary artery is also dilated.

Discussion

Aortopulmonary window is a rare congenital abnormality of the great vessels accounting for 0.2% -0.6% of all the congenital heart defects [1]. It is variously named as aorticopulmonary septal defect, truncus arteriosus communis, aorticopulmonary communication [2]. The patients usually present with severe heart failure and pulmonary hypertension. It is usually associated with other defects in half of the cases [3].

Unrepaired defects survival after first decade is rare though anecdotal reports of survival upto seventh decade are present [4]. The defect being asymptomatic in the first decade when present as an isolated abnormality is rare.

First case was reported by Elliotson [5]. Classification is by Mori etal. Most common is type 1 aortopulmonary window 65%, type 2 32%, type iii 05% [6]. The survival rate is 50% in first decade.

Patients presenting with eisenmenger syndrome have poor prognosis as they cannot be subjected for the closure of defect [7]. Medical management of the pulmonary hypertension can be done with varying response to the vasodilators. Six minute walk test is simple reliable estimation of the cardiopulmonary efficacy in patients with significant pulmonary hypertension. The distance walked during the six minutes acts as a prognostic marker of the disorder [8].

Heart-lung transplantation is the treatment of choice in patients with eisenmenger syndrome but the chances of survival is less and there will be increased complications post

operatively [8].

References

1. Kutche LM, Van Mierop LHS. Anatomy and pathogenesis of aortopulmonary septal defect. Am J Cardiol. 1987; 59:443-7.
2. Ganju NK, Kandoria A. Aortopulmonary Window: A Rare Cardiac Anomaly Presenting in Adolescent Age Group. Heart India. 2015; (3):87-9.
3. Moruno Tirado A, Santos De Soto J, Grueso Montero J, Gavilán, Camacho JL, Alvarez Madrid A *et al.* Aortopulmonary window: Clinical assessment and surgical results. Rev Esp Cardiol. 2002; (55):266-70.
4. Su-Mei AK, Ju-Le T. Large un-repaired aortopulmonary window—survival into the seventh decade," Echocardiography. 2007; 24(1):71-73.
5. Elliotson J. Case of malformation of the pulmonary artery and aorta. Lancet. 1830 ; (1):247.
6. Mori K, Ando M, Takao A, Ishikawa S, Imai Y. Distal type of aortopulmonary window. Report of 4 cases. Br Heart J. 1978; (40):681-9.
7. Soares AM, Atik E, Cortˆez et al. "Aortopulmonary window. Clinical and surgical assessment of 18 cases," *Arquivos Brasileiros de Cardiologia*, 1999; 73(1):59–74.
8. Stout K, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS et al (2019). 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. (139):e698–e800. doi: 10.1161/CIR.0000000000000603