



Case Report: A Rare Case of a 31 year-old Filipino male diagnosed of Double-Inlet Left Ventricle with unrestricted Pulmonary blood flow reaching Early Adulthood



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INTRODUCTION

Tremendous advancement ensued over the past century in the pediatric care, surgery and anesthesia, which led to more than 90% of children diagnosed with congenital heart disease (CHD) reach adulthood.¹ As good as this may seem, CHD is still said to be the leading cause of infant mortality.² Among children aged less than 5 years old, more than 80% die because of CHD-related events.² This is due to congenital defects interfering the normal physiologic processes needed to sustain life. The developing countries, particularly Africa and Asia, have the highest mortality rates among patients with CHD.²

CASE

G.V., 31 year-old male, living in Cainta, was diagnosed with Double-inlet Left Ventricle and transposition of great arteries at birth. He was born to a 20 year-old G1P1 via normal spontaneous delivery. Upon diagnosis of the heart defect, surgery was advised but patient's relatives opted to seek a second opinion. At 4 years of age, cyanosis and clubbing of nails started to appear. He was prescribed with Digoxin 0.25 mg/tablet, half tablet once daily. During adulthood, he had hypertension, and started to develop secondary erythrocytosis, necessitating phlebotomy. He was non-smoker and non-alcoholic. No familial history of congenital heart disease was noted. G.V. had occasional palpitations and orthopnea, with persistent nail clubbing, but was lost to follow-up. The patient sought consult in the emergency department due to recurrent hemoptysis. He was in mild respiratory distress. On physical examination, he had circumoral cyanosis, clubbing of nails (Figure 1), mid-to-base rales on bilateral lung fields, an S3 gallop, and a grade 3/6 systolic murmur which was heard loudest at the apex. ECG showed sinus tachycardia with right axis deviation. Chest X-ray revealed hazy opacities with patchy consolidations in the right upper lobe. There was no cardiomegaly appreciated. Other diagnostics were normal except for erythrocytosis at 166 mg/dL.



Figure 1. Clubbing of nails of G.V.

A segmental TTE showed situs solitus, levocardia and ventriculoarterial discordance. The 4-chamber apical view clearly depicted a single ventricle in connection with both right and left atria. The ventricle had prominent posterior median ridge on the diaphragmatic wall, pointing to a left ventricular morphology (Figure 2). In the parasternal long axis view, two vessels were seen lying parallel with one another, both originating from the single ventricle. 3-D TTE showed the aorta situated anteriorly and to the right of the main pulmonary artery (Figure 3). Other findings were a

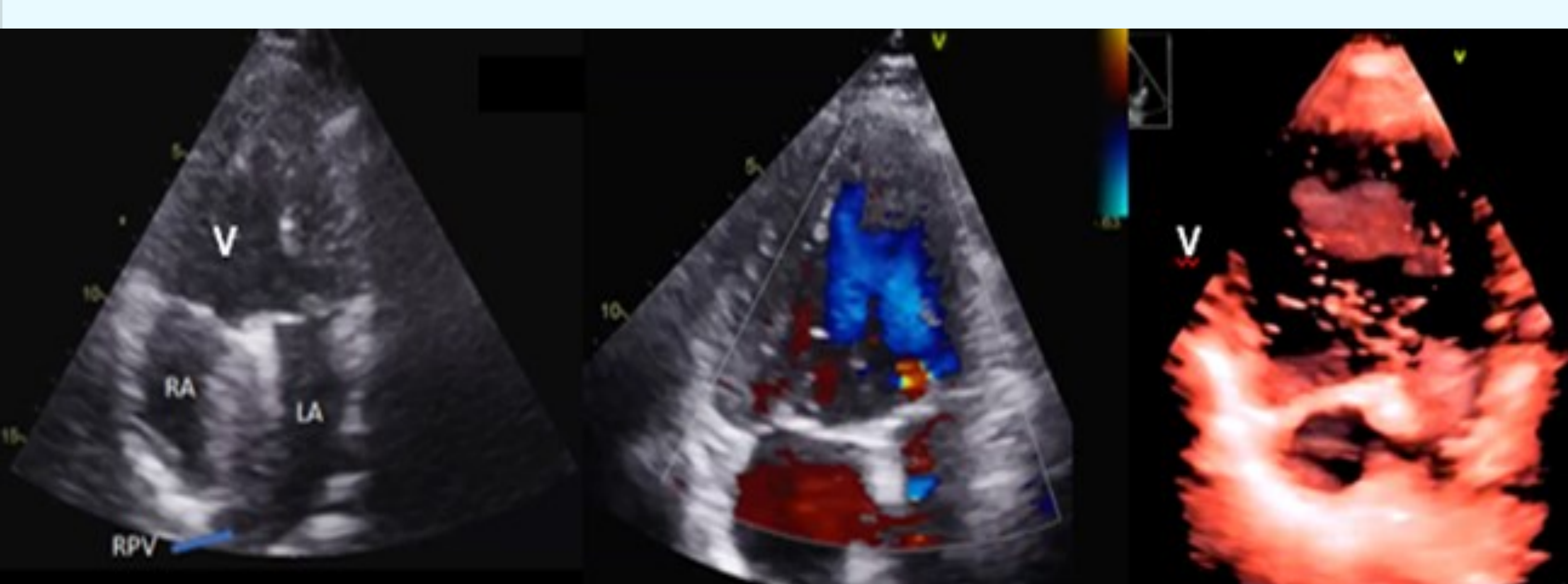


Figure 2. TTE showing a single ventricle connected to both atria. V – univentricle. RPV – right pulmonary vein. LA – left atrium. RA – right atrium.

moderate pulmonary regurgitation, and a mild coarctation of aorta having a peak velocity of 2.32 m/s and peak gradient of 21.47 mmHg (Figure 4). There was no pulmonary stenosis appreciated. In addition to treating the pneumonia, pulmonary hypertension was addressed by giving Bosentan 125 mg/tab, ½ tablet twice daily, and Sildenafil 50 mg/tab, 1 tablet thrice daily. Digoxin was increased to 0.25 mg/tab, 1 tablet daily. Despite medical treatment, patient succumbed to death.

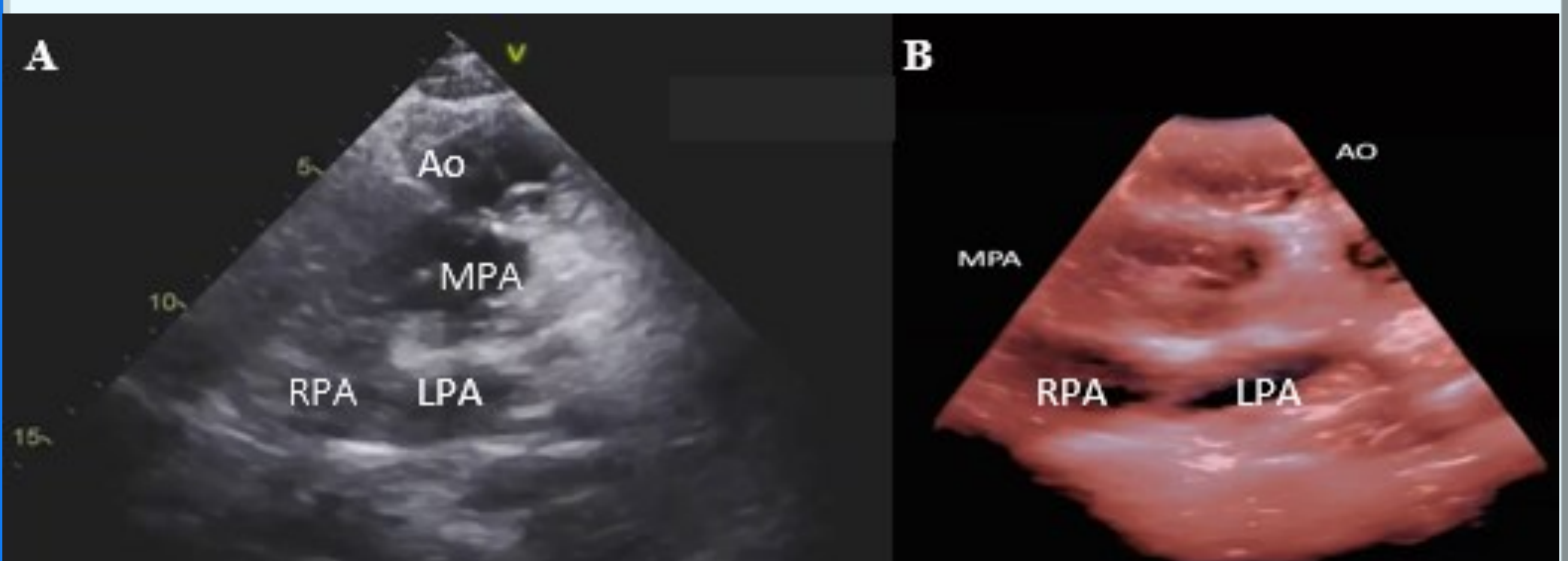


Figure 3. Transposition of the great arteries with the aorta originating on the right side of the main pulmonary artery in 2-D (A) and 3-D (B) images of parasternal short axis view. MPA – main pulmonary artery. RPA – right pulmonary artery. LPA – left pulmonary artery. Ao-aorta.

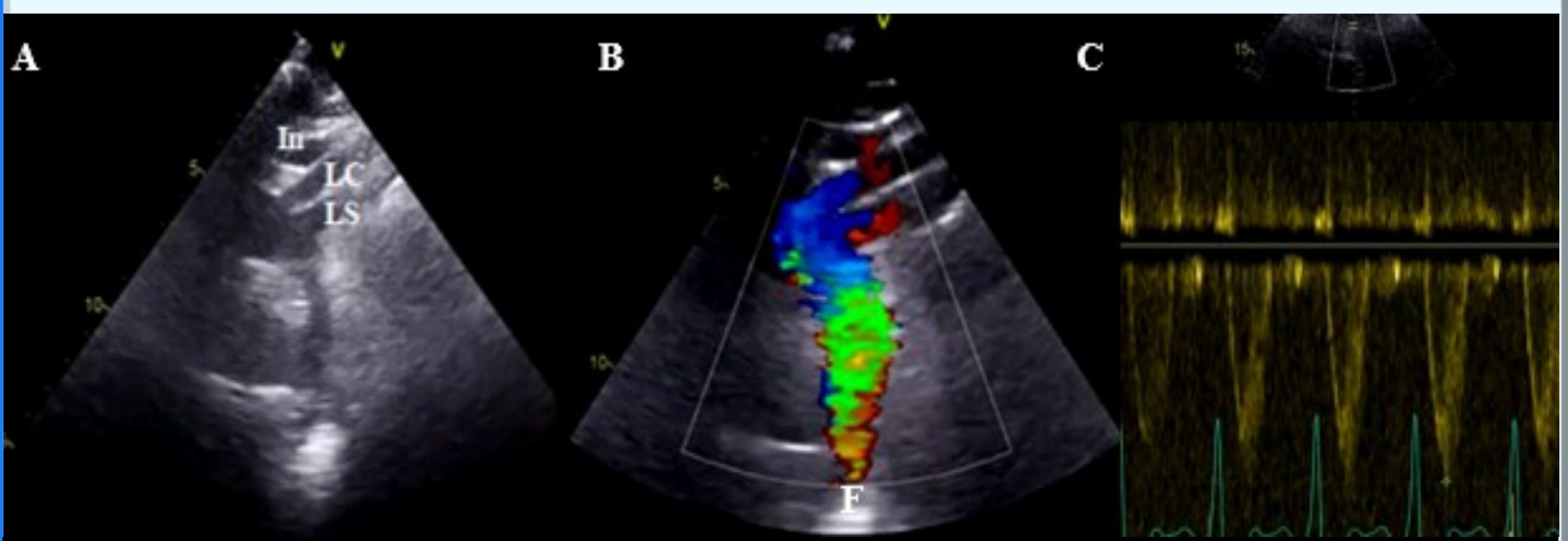


Figure 4. TTE suprasternal view showing the aortic arch and its branches (A), mosaic color flow in the descending aorta with segment narrowing (B), and continuous wave Doppler in descending aorta (C). In – innominate artery. LC – left common carotid artery. LS – left subclavian artery.

DISCUSSION

The development of symptoms and prognosis of these patients are said to be dependent on the presence or absence of an obstruction to pulmonary blood flow and the level of pulmonary vascular resistance.⁴ If there is no anatomic restriction to the pulmonary circulation at all, an intractable heart failure may develop and subsequently result to early death.⁶ Without surgical repair, survival into late adulthood is said to be rare.⁴ And if they survive, these “unoperated” patients develop severe pulmonary vascular disease⁶, increasing the risk for morbidity and mortality. Nugraha *et al* mentioned in their study done in 2017 that hemoptysis is a serious complication of pulmonary arterial hypertension which is rarely seen, and if reported, it signifies pulmonary arterial hypertension at its terminal stage.¹² Our patient consulted due to recurrent hemoptysis which was attributed to pulmonary hypertension as consequence of heart failure.

CONCLUSION

In rare conditions involving anomaly in cardiac development, TTE provides a good anatomic evaluation and assessment of pre-existing hemodynamics. In our patient, there was no obstruction in the pulmonary circulation noted. The transposition of the great arteries and the mild coarctation of the aorta will not cause restriction in the blood flow. Hence, we can only attribute his survival to early adulthood to an extraordinary hemodynamics sustaining him good oxygenation despite continuous mixing of blood in the arterial and pulmonary circulations. Furthermore, the patient's eventual demise was due to complications of heart failure with concomitant pulmonary hypertension.

References

