**RESEARCH PAPER**

# A Rare 43-Year Survival of Unoperated Tetralogy of Fallot with Pulmonary Atresia with Huge MAPCAs with High Pulmonary Vascular Resistance Surviving 2 Pregnancies: A Case Report

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## ABSTRACT

**Background:** Tetralogy of Fallot with pulmonary atresia (TOF-PA) is one of the most severe cyanotic congenital heart diseases. Long-term survival without surgical intervention is exceedingly rare.

**Case Summary:** We report a 43-year-old female from Karnataka, India, who presented with dyspnea on exertion, headache, and blurring of vision for 2 months. She had never undergone any cardiac surgery, surviving 2 pregnancies with no postnatal complications. On examination, she had $SpO_2$ 78%, marked clubbing, and a continuous murmur over all lung fields, suggestive of major aortopulmonary collateral arteries (MAPCAs). Hyperviscosity symptoms were also present. Echocardiography confirmed TOF-PA physiology, and her long-term survival is attributed to extensive systemic-pulmonary collateralization.

**Conclusion:** This case exemplifies survival in unoperated TOF-PA due to well-developed MAPCAs, emphasizing the need for lifelong monitoring.

**Keywords:** Tetralogy of Fallot, pulmonary atresia, MAPCAs, cyanotic congenital heart disease, hyperviscosity, adult congenital heart disease, India.

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## INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect (CHD). Without surgical intervention, less than 3% of patients with TOF can survive over 40 years. Pulmonary atresia with ventricular septal defect (PA-VSD) is at the extreme end of the spectrum of TOF. Survival rates in PA-VSD without surgical

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intervention are 50% at age 1 year and 8% at age 10 years.[1]

Reports of 40-year survival without surgical intervention are exceptional. Mechanisms of survival in such cases often relate to the development of major aortopulmonary collateral arteries (MAPCAs) that provide partial pulmonary blood flow.
Here we describe a 43-year-old Indian woman with unoperated TOF-PA who survived 2 pregnancy- a rarity even in global literature.

## CASE PRESENTATION

### Patient Information

A 43-year-old female presented with progressive dyspnea on exertion for 2 years of age which has increased in last 2 months (NYHA CLAS II–III). Patient was born as full term normal vaginal delivery and was alright until 2-3 months of age when her parents noticed bluish discoloration of skin and mucus membrane while crying and feeding which was relieved by rest. She started having cyanotic spells by 1 year of age and was relieved by squatting till 2-3 years of age after which such episodes became infrequent. She also had easy fatiguability while playing with peers during childhood. For these complaints she went to a doctor and was advised symptomatic treatment. There was no history of recurrent lower respiratory tract infection, chest pain , palpitation, fever, skin lesions or any features suggestive of infective endocarditis. She achieved all developmental milestones till age and is vegetarian by diet with normal sleep and bladder & bowel habits and no family history of any congenital heart disease. Patient also gave birth to 2 children, one 9 years old and other one 4 years old, both of them born through caesarean section with no antenatal or postnatal complication and living a healthy life. There is past history of hypothyroidism with no other chronic illness. Now presented to KLE hospital with exertional dyspnea and also reported headache and blurring of vision for last 2 months, consistent with hyperviscosity symptoms and was admitted for further management. On admission basic vitals were notes which were as follows-

**Heart Rate:** 80/min, regular in rhythm, normal in volume and character, no radio-radial or radio femoral delay, normal condition of the vessel wall

**Blood Pressure:** 130/70 mmHg taken in right arm in supine position

**Respiratory rate-** 18 breaths/min

**$SpO_2$ (room air):** 78%

**General:** Height- 155cm, weight- 64kg, BMI- 26.6 $kg/m^2$, Central cyanosis present, marked pandigital clubbing of grade 3.

**Figure 1: Pan Digital Clubbing**

**CVS:** A continuous murmur audible over all lung fields, strongly suggestive of MAPCAs along with another high pitched blowing decrescendo diastolic murmur in left third intercoastal space, best heard with diaphragm in leaning forward position and it increases in intensity with isometric hand grip exercises.

No pedal edema, no clinical right heart failure signs

### Investigations

**ECG:** Sinus rhythm with features of right ventricular hypertrophy (FIGURE 2)

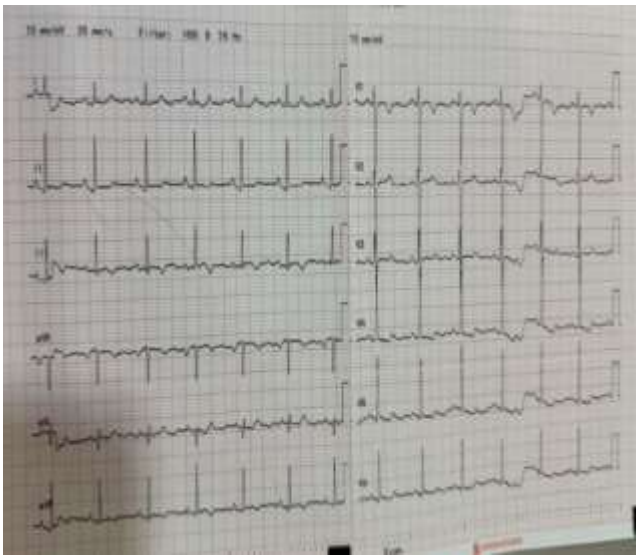

**Figure 2: ECG**

**Chest Radiography-**

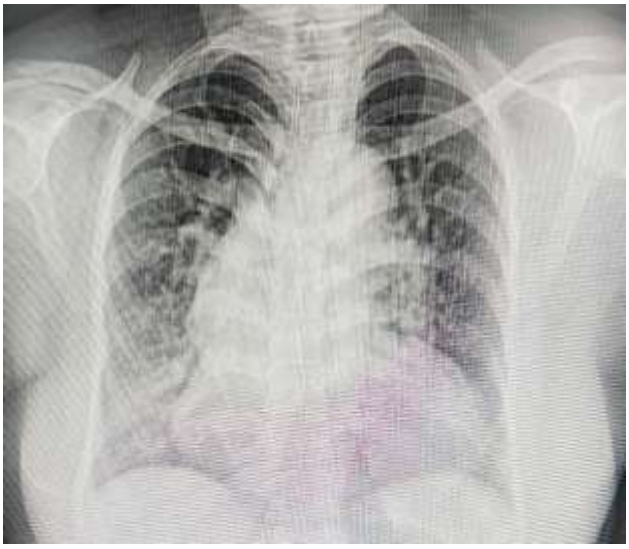

**Figure 3: Chest X- Ray**

Cardiomegaly with reticular opacities involving bilateral mid and lower zones and patchy pulmonary vasculature and dilated arch of aorta (FIGURE-3)

**Echocardiography-**

- Situs solitus d- loop ventricle
- Large malaligned VSD measuring- 1.8-1.9 cm with bidirectional shunt (FIGURE 4)
- Pulmonary atresia
- 50% Overriding of aorta
- Moderate aortic regurgitation
- Right ventricular hypertrophy
- Good biventricular function

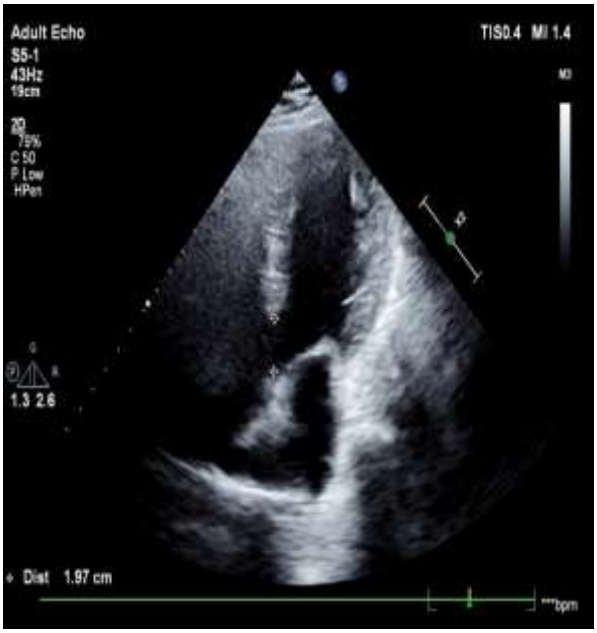


**Figure 4: Large Maligned VSD**

**Coronary Angiography**- Normal epicardial coronaries

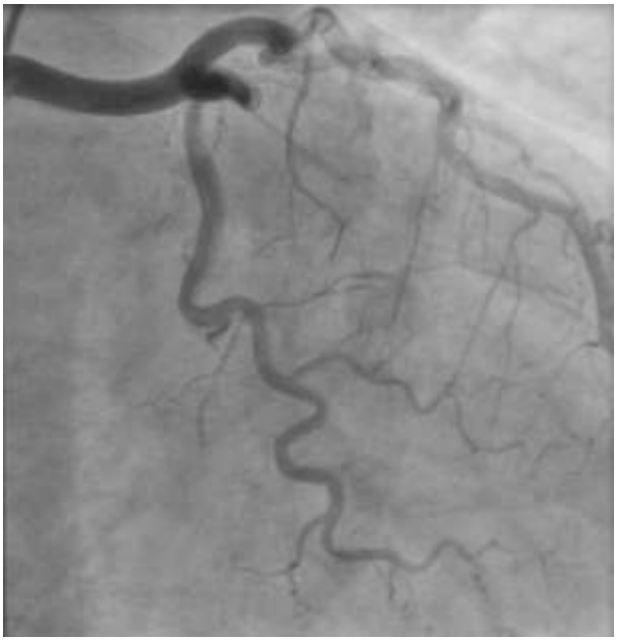

**Figure 4: Normal Left Coronary Arteries**

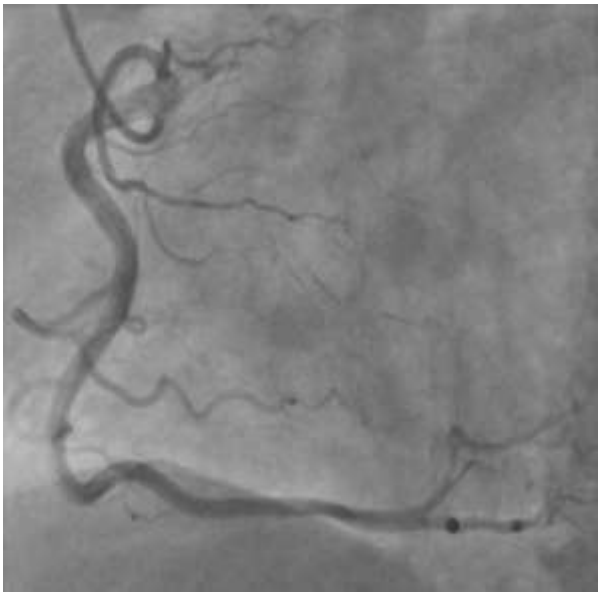

**Figure 5: Normal Right Coronary Artery**

**Cardiac catheterisation study-**

- Large subaortic VSD with bidirectional shunt
- 50% Overriding of aorta

- Aorta is filling through VSD
- Multiple large MAPCAs seen(FIGURE-5,6,7)

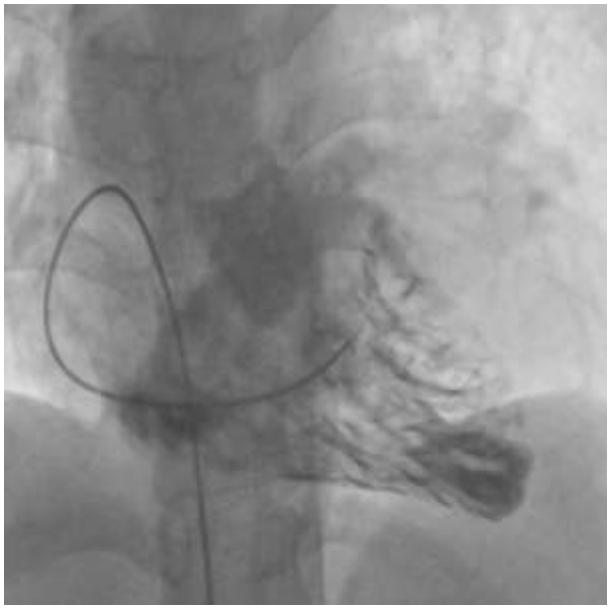

**Figure 6: RV Angio- Coarse Trabeculated Good Contracting RV With Pulmonary Atresia And RV Blood Going Into Aorta.**

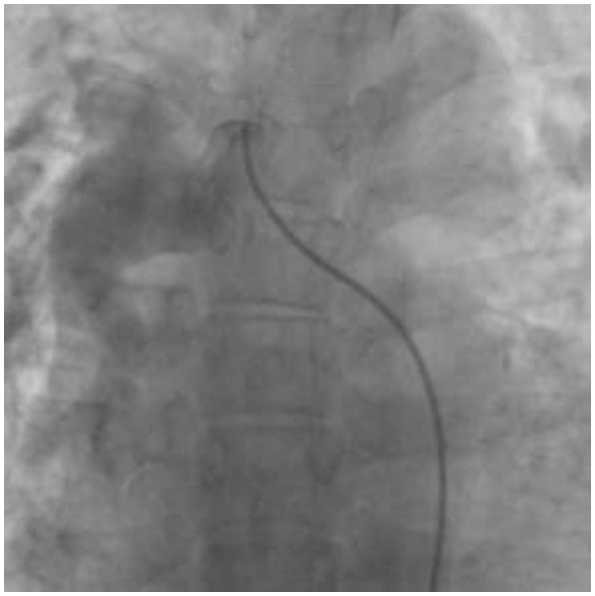

**Figure 7: Large MAPCA Seen Over Right Side**

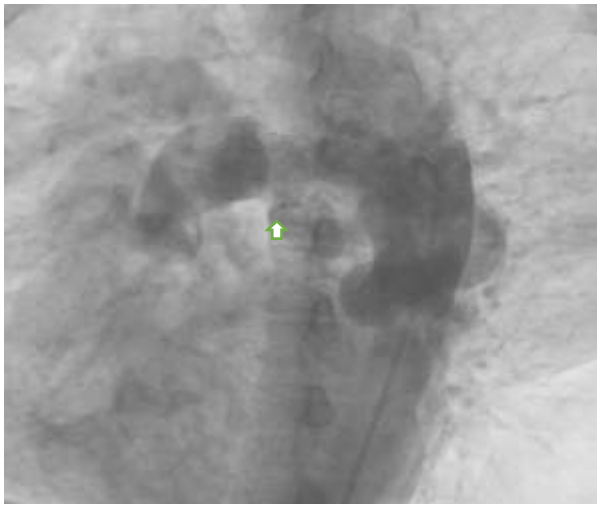

**Figure 8: Large MAPCA From Aorta To Right Side Of Lung**

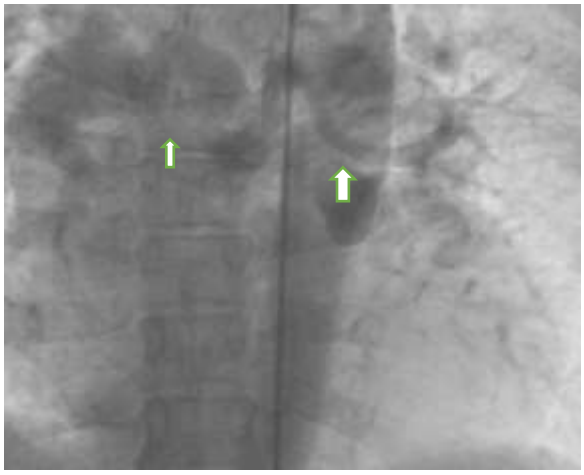

**Figure 9: MAPCAS Filling Both Left And Right Lungs**

CT pulmonary angiography- Suggestive of sub aortic VSD of 9mm (FIGURE 8), overriding of aorta, pulmonary artery atresia (FIGURE 9) with MAPCAs (FIGURE-10,11,12) with pulmonary oligemia

**Figure 10: MAPCA Arising From Descending Thoracic Aorta To Left Upper Lobe- 8.7mm In Diameter**

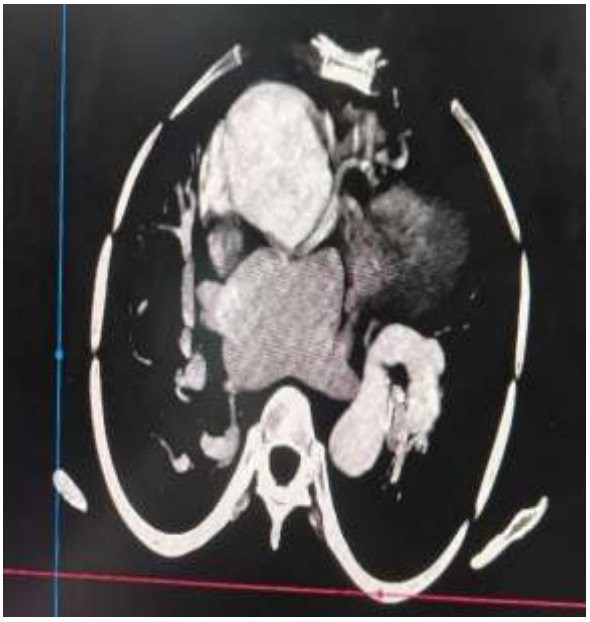

**Figure 11: MAPCA Arising From Descending Thoracic Aorta To Right Upper And Lower Lobes- 1.6cm In Diameter**

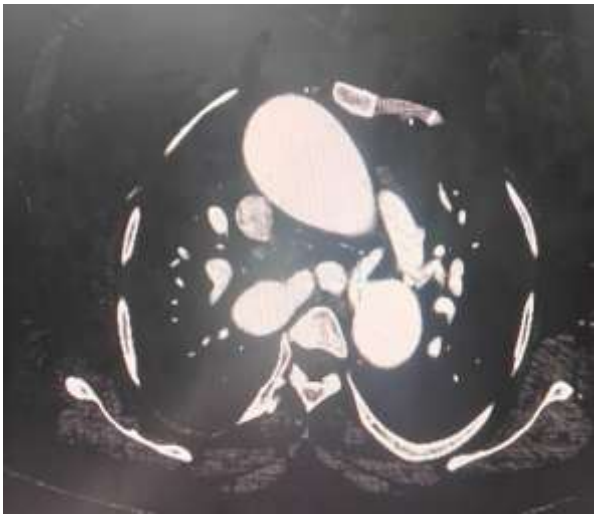

**Figure 12: MAPCA Arising From Descending Thoracic Aortawhich Communicates With Left Pulmonary Artery- 1.6 Cm At Origin**

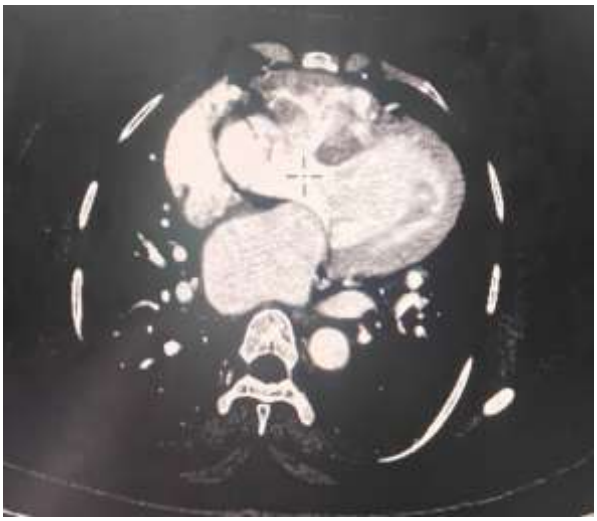

**Figure 8: Sub Aortic VSD**

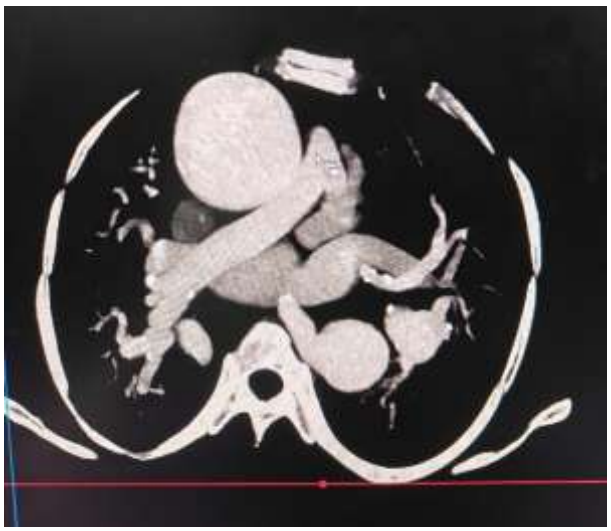

**Figure 9: Main Pulmonary Artery Atretic**

**$SpO_2$ and Hematology**

- $SpO_2$: 78% on room air
- Haemoglobin- 17gm/dl, PCV- 59.5%
- Renal profile- Normal

Hence, the diagnosis of Tetralogy of Fallot with pulmonary atresia with hypertensive MAPCAs with hyperviscosity syndrome was made.

**Hospital Course And Management**

The patient was admitted for evaluation of worsening dyspnea and hyperviscosity symptoms.

**Management included:**

- Phlebotomy was done for hyperviscosity syndrome
- Avoidance of dehydration
- Counseling regarding surgical options — patient was deemed high-risk for any definitive intracardiac repair or unifocalization procedure because of age and chronic hypoxemia.

She improved symptomatically with supportive management and was discharged in NYHA I.

## DISCUSSION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. It has a prevalence of about 0.3–0.6 per 1000 live births. It is characterized by ventricular septal defect, overriding of aorta, right ventricular outflow obstruction and right ventricular hypertrophy. Pulmonary atresia with ventricular septal defect is at the extreme end of the spectrum of TOF. Life expectancy in pulmonary atresia with ventricular septal without surgical intervention is 50% at age 1 year and 8% at age 10 years[1].

The STS-Congenital Heart Surgery Database Committee divides the patients with PA-VSD into three types, according to the presence or absence of native pulmonary arteries and the presence or absence of MAPCA(s).

(1) Type A: native pulmonary arteries are present. Pulmonary blood flow is supplied by the PDA. (2) Type B: both native pulmonary arteries and MAPCAs are present. (3) Type C: there are no native pulmonary arteries. The pulmonary circulation is supplied by MAPCA(s) only. [2]

Patients with adequate but not excessive pulmonary blood flow might survive into adolescence and adulthood. The presence of patent ductus arteriosus or MAPCA is crucial in providing a stable source of pulmonary blood flow. The anatomic variability of the pulmonary vascular bed and its aortopulmonary collateral blood flow in large measure explain the differences in clinical manifestations and in unoperated survival.[3]

A small rate of unoperated patients with PA/VSD can survive until adulthood and the arterial blood supply to the lungs, provided by major aorto-pulmonary collateral arteries (MAPCAs), is one of the main determinants of survival.[4]

So far, there has been 19 adults with unoperated pulmonary atresia with ventricular septal defect have been reported in the world literature. Major aortopulmonary collateral blood supply was frequently multifocal with varied sources in these cases, including the thoracic aorta (12 cases), the coronary artery (4 cases), the internal mammary artery (3 cases), the left brachiocephalic trunk (2 cases), abdominal aorta (2 cases) and subclavian artery (2 cases). [3]

In this case report, we describe a 79-year-old woman with unoperated tetralogy of fallot with pulmonary atresia, who presented with dyspnea on exertion, headache, and blurring of vision for 2 months, along with a history of cyanotic spells in childhood. She had never undergone any cardiac surgery, surviving 2 pregnancies with no postnatal complications. On examination, she had room air $SpO_2$ of 78%, marked digital clubbing, and a continuous murmur in the left scapular region suggestive of major aortopulmonary collateral arteries (MAPCAs). Hyperviscosity symptoms were prominent. Echocardiography and CT pulmonary angiography confirmed TOF-PA physiology, and her long-term survival is attributed to extensive systemic-pulmonary collateralization.

Over decades, she developed robust compensatory mechanisms: Marked right-ventricular hypertrophy, Polycythemia improving systemic oxygen delivery, Gradual adaptation to chronic hypoxemia, Extensive MAPCAs as Lifelines.

By adulthood, TOF-PA patients often become poor surgical candidates due to: Unfavourable anatomy, Fragile MAPCAs and High operative risk. In her case, the risk of destabilizing her chronic compensatory state outweighed the potential benefits for any definitive intracardiac repair or unifocalization procedure.

## CONCLUSION

This 43-year survival without surgery in TOF-PA is extraordinary and underscores the profound adaptability of human physiology when supported by MAPCAs. The case highlights the need for early detection and lifelong structured care for congenital heart disease, particularly in resource-constrained environments. It also emphasizes the importance of recognizing and managing chronic complications such as hyperviscosity syndrome.

## PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

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