RESEARCH PAPER

# Natural Adult Survival in Uncorrected Tricuspid Atresia with Functionally Single Ventricle and Severe Right Ventricular Outflow Tract Obstruction: A Rare Case Report

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**ABSTRACT**

Tricuspid atresia (TA) with a functionally single ventricle is a rare cyanotic congenital heart defect characterized by the absence of the tricuspid valve and complete lack of communication between the right atrium and right ventricle. The natural course is usually fatal in early infancy without surgical intervention. We report an unusual case of a 21-year-old male with uncorrected TA and a functionally single left ventricle who presented with fever and altered sensorium. Echocardiography revealed tricuspid valve atresia, single atrioventricular connection, restrictive subaortic ventricular septal defect (VSD), severe right ventricular outflow tract (RVOT) obstruction, and right-to-left shunt through an atrial septal defect (ASD). Despite the complex anomaly, the patient survived into adulthood with balanced systemic and pulmonary circulations and preserved left ventricular function. This case highlights the importance of clinical suspicion, echocardiographic diagnosis, and the adaptive physiology that may permit rare adult survival in unoperated TA.

**Keywords:** Tricuspid atresia, congenital cyanotic heart disease, single ventricle, right ventricular outflow tract obstruction, adult congenital heart disease, echocardiography, natural survival

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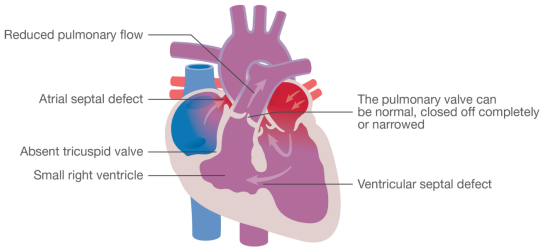
## Introduction

Tricuspid atresia (TA) is a rare form of cyanotic congenital heart disease (CHD), accounting for approximately 1% to 3% of all CHDs [1]. It is defined by the congenital absence of the tricuspid valve, resulting in complete absence of a direct communication between the right atrium (RA) and right ventricle (RV) [2]. Consequently, systemic venous return is shunted across an interatrial communication—usually a patent foramen ovale (PFO) or atrial septal defect (ASD)—into the left atrium (LA), leading to obligatory right-to-left shunting and systemic desaturation.

The left ventricle (LV) becomes the dominant and functionally single pumping chamber supplying both the systemic and pulmonary circulations through associated defects such as a ventricular septal defect (VSD) or patent ductus arteriosus (PDA). The pulmonary circulation is frequently supplied through a VSD or via systemic-to-pulmonary collaterals.




TA is classified anatomically based on the relationship of the great arteries and the presence of pulmonary stenosis or atresia (Edwards and Burchell classification). Type I (normally related great arteries) is most common, followed by Type II (transposed great arteries). The degree of pulmonary stenosis or RVOT obstruction determines the clinical presentation and prognosis [3,4]

The natural history of TA without surgical correction is poor. Approximately 50% of patients die within the first six months of life, and only 10% survive beyond the first decade . Rarely, patients may survive into adolescence or adulthood due to a favourable

hemodynamic balance—specifically, a restrictive VSD that prevents pulmonary overload, adequate pulmonary blood flow, and preserved LV contractility.

We present a case of a 21-year-old male with uncorrected tricuspid atresia, a functionally single left ventricle, and severe RVOT obstruction who survived into adulthood without prior intervention. Such cases provide unique insight into the compensatory mechanisms and adaptive circulatory physiology that can sustain life in the absence of surgical palliation.

**Case Presentation**

A 21-year-old male presented to Sharda Hospital, Greater Noida, with a four-day history of intermittent fever and one-day history of altered sensorium.. The patient had been cyanotic since childhood but had never undergone cardiac evaluation or intervention. His exercise tolerance was modest, and he reported occasional breathlessness but no syncope or chest pain. There was no history of seizures, vomiting, photophobia, or limb weakness

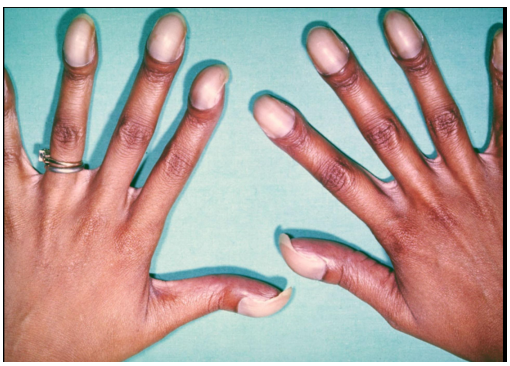
**Past and Personal History**

No history of hypertension, diabetes, tuberculosis, thyroid disease, or asthma. He was a non-smoker, non-alcoholic, and consumed a vegetarian diet. Bowel and bladder habits were normal.

**Physical Examination**

The patient appeared cyanotic but was hemodynamically stable.

- BP: 100/70 mmHg
- HR: 74 bpm
- RR: 24/min
- $SpO_2$: 98% on 10 L/min $O_2$ by non-rebreather mask

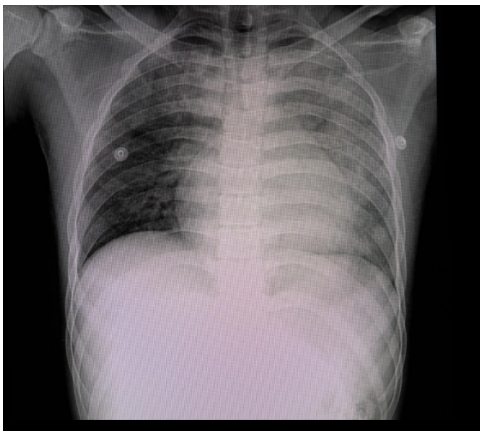


Clinical photograph demonstrating digital clubbing, consistent with long-standing cyanotic congenital heart disease.

- CNS:Confused but Oriented to Time, Place and Person; no focal deficits
- CVS: Loud S2 audible,Ejection systolic murmur present
- Respiratory: Clear bilateral air entry
- Abdomen: Mild hepatomegaly; non-tender

**Investigations:**

- Hemoglobin: 19.2 g/dL
- Hematocrit: 58.2% (secondary erythrocytosis)
- Platelets: 53,000/µL (mild thrombocytopenia)
- LFT: Mildly raised transaminases (SGOT 102 U/L, ALP 383 U/L)
- RFT: Urea 62.5 mg/dL, Creatinine 0.7 mg/dL
- Electrolytes: $Na^+$ 135 mEq/L, $K^+$ 5.9 mEq/L, $Ca^{2+}$ 7.9 mg/dL

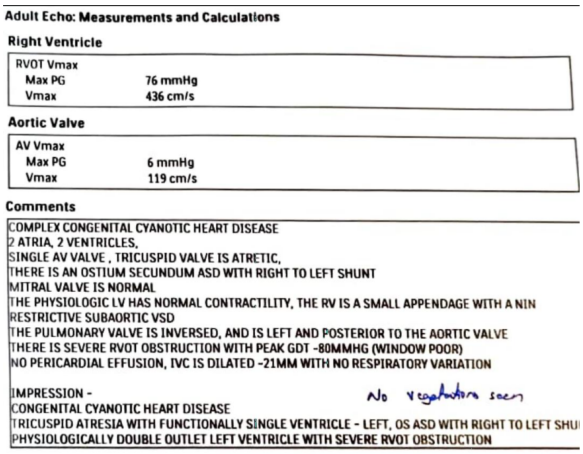


Chest X Ray showing cardiomegaly with relatively reduced pulmonary vascular markings suggestive of decreased pulmonary blood flow.

- Thyroid: Low T3 and T4 with normal TSH (euthyroid sick syndrome)
- CSF: Protein 108 mg/dL, Sugar 83 mg/dL, no organisms — suggestive of aseptic inflammation
- Serology: Malaria, dengue, and typhoid negative
- Ultrasound Abdomen: Mild hepatosplenomegaly, gallbladder wall edema, mild ascites, and right pleural effusion

**Cardiac Evaluation**

- ECG: Left axis deviation, LV hypertrophy pattern, HR 80 bpm, prolonged QTc (440 ms).
- Echocardiography: Tricuspid valve atretic
  - Single atrioventricular connection
  - Functionally single left ventricle
  - Restrictive subaortic VSD
  - Ostium secundum ASD with right-to-left shunt
  - Severe RVOT obstruction (peak gradient 80 mmHg)
  - Dilated IVC (21 mm), normal LV contractility
  - No vegetations seen



**Adult Echo: Measurements and Calculations**

**Right Ventricle**

| RVOT Vmax | |
|---|---|
| Max PG | 76 mmHg |
| Vmax | 436 cm/s |

**Aortic Valve**

| AV Vmax | |
|---|---|
| Max PG | 6 mmHg |
| Vmax | 119 cm/s |

**Comments**

COMPLEX CONGENITAL CYANOTIC HEART DISEASE
2 ATRIA, 2 VENTRICLES,
SINGLE AV VALVE , TRICUSPID VALVE IS ATRETIC,
THERE IS AN OSTIUM SECUNDUM ASD WITH RIGHT TO LEFT SHUNT
MITRAL VALVE IS NORMAL
THE PHYSIOLOGIC LV HAS NORMAL CONTRACTILITY, THE RV IS A SMALL APPENDAGE WITH A NIN
RESTRICTIVE SUBAORTIC VSD
THE PULMONARY VALVE IS INVERSED, AND IS LEFT AND POSTERIOR TO THE AORTIC VALVE
THERE IS SEVERE RVOT OBSTRUCTION WITH PEAK GDT -80MMHG (WINDOW POOR)
NO PERICARDIAL EFFUSION, IVC IS DILATED -21MM WITH NO RESPIRATORY VARIATION

IMPRESSION - No Vegetations seen
CONGENITAL CYANOTIC HEART DISEASE
TRICUSPID ATRESIA WITH FUNCTIONALLY SINGLE VENTRICLE - LEFT, OS ASD WITH RIGHT TO LEFT SHU
PHYSIOLOGICALLY DOUBLE OUTLET LEFT VENTRICLE WITH SEVERE RVOT OBSTRUCTION

**Final Diagnosis:**

Congenital cyanotic heart disease — Tricuspid atresia with functionally single left ventricle and severe RVOT obstruction.

**Treatment:**

The patient received intravenous vancomycin, ceftriaxone, acyclovir, dexamethasone, and levetiracetam. Supportive care with oxygen and fluids was provided. Fever and altered sensorium resolved with conservative management. He was discharged after seven days in stable condition with referral to a cardiology unit for long-term follow-up.

**Discussion:**

Tricuspid atresia (TA) is a rare cyanotic congenital heart disease with a typically poor natural history. In the absence of surgical intervention, mortality is extremely high, with only about 33% surviving to 1 year and ~10% to 10 years . Most untreated patients do not reach adulthood, making the present case clinically exceptional.

Adult survival in uncorrected TA has been reported only in isolated case reports and small series:

- A classic case described survival up to 57 years without surgery, representing one of the longest documented survivals .(5)
- Series of adult patients with TA suggest that survival beyond adolescence is rare and often associated with prior palliation or unique hemodynamic adaptations .(6)

More recent analyses of unoperated single-ventricle physiology indicate that selected patients may survive into the fifth or even seventh decade, provided a stable circulatory balance is maintained

**Natural History and Hemodynamic Adaptation**

Most patients with TA succumb early due to inadequate pulmonary blood flow or heart failure. However, survival into adulthood occurs when there is:

i). Balanced pulmonary blood flow — achieved through restrictive VSD or RVOT obstruction preventing pulmonary hypertension.

ii). Adequate systemic flow — through preserved LV function maintaining cardiac output.

In this case, the restrictive VSD and severe RVOT obstruction likely prevented pulmonary overload, while the single LV maintained systemic output, resulting in stable hemodynamics over two decades.

**Hematological and Systemic Findings**

Secondary erythrocytosis (Hb 19.2 g/dL, Hct 58%) develops as a compensatory response to chronic hypoxia, improving oxygen-carrying capacity . Thrombocytopenia and hepatosplenomegaly, as observed in our patient, are known sequelae of long-standing cyanosis and venous congestion. Mild hepatic dysfunction may result from chronic right-sided pressure overload and passive congestion.

**Neurological Manifestations**

Altered sensorium in cyanotic CHD can result from hyperviscosity, hypoxia, or paradoxical emboli.In our patient, absence of infection and normalization with supportive care pointed to metabolic encephalopathy secondary to hypoxia rather than CNS infection.

**Echocardiographic Diagnosis**

Echocardiography remains the cornerstone of diagnosis. It delineates valve morphology, ventricular function, shunt direction, and great artery relationships. The presence of severe RVOT obstruction, paradoxically protective, prolonged survival by limiting pulmonary flow and maintaining balanced circulation . Three-dimensional echocardiography and cardiac MRI are useful adjuncts for preoperative assessment.

**Comparison with Literature**

Few adults with uncorrected tricuspid atresia have been reported in the literature.

- Bhat et al. (2015) described a 25-year-old male surviving without surgery due to restrictive VSD and balanced flow [7].
- Singh et al. (2021) reported a 22-year-old woman with TA and pulmonary stenosis surviving into adulthood due to compensated hemodynamics [8].
- Zhang et al. (2023, Cureus) highlighted that adult survival is possible when pulmonary flow is neither excessive nor critically reduced [9]

Our patient has favorable hemodynamics and preserved LV function, facilitated survival.

**Modern Management Perspective**

Current management strategies emphasize early palliative and definitive surgeries — such as the Blalock–Taussig shunt followed by the Fontan procedure — to separate systemic and pulmonary circulations [10] Adults diagnosed late require comprehensive evaluation for suitability for Fontan-type palliation or heart transplantation. However, in stable adults like our patient, conservative management and periodic cardiology follow-up may be justified to minimize procedural risk.

**Prognosis**

Long-term complications include progressive cyanosis, heart failure, arrhythmias, brain abscess, and thromboembolism . Nonetheless, the longevity in our case underscores the role of favorable anatomy in survival, reinforcing the spectrum of natural adaptation in unoperated congenital heart disease.

**Conclusion**

This case highlights a rare instance of natural adult survival in uncorrected tricuspid atresia with a functionally single left ventricle and severe RVOT obstruction. The patient's survival can be attributed to a unique hemodynamic balance—restrictive shunting, protective RVOT obstruction, and preserved ventricular function. Clinicians should maintain a high index of suspicion for congenital cyanotic heart diseases in adults with unexplained polycythemia, mild cyanosis, or hepatosplenomegaly. Early echocardiographic diagnosis remains essential for appropriate counseling and management

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