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COMPREHENSIVE ECHOCARDIOGRAPHIC EVALUATION OF TRICUSPID ATRESIA AFFLICTING AN ADOLESCENT: A DISTINCTIVE CASE REPORT WITH LITERATURE REVIEW

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ABSTRACT

Tricuspid atresia (TA) is a complex congenital heart disease that presents with cyanosis in the neonatal period. TA is associated with high mortality without early intervention. TA arises from either the agenesis (absence of right atrioventricular connection) or the imperforation of the tricuspid valve. Consequently, there is no direct communication between the right atrium and the right ventricle. Survival in patients with TA has been shown to be hemodynamically dependent upon the presence of adequate pulmonary blood flow (PBF), a lack of elevated pulmonary vascular resistance, and the degree of ventricular shunting. PBF is the most integral component of these factors. We are presenting a remarkable case of a seventeen year old female adolescent afflicted with TA alongwith severe central cyanosis, conspicuous clubbing in fingers of all the four extremities, atrial septal defect (ASD), ventricular septal defect (VSD), normally related great arteries (NRGA), hypoplastic right ventricle (RV) and dilated left ventricle (LV). It is noteworthy, that despite the protracted course of the illness, the patient has not undergone any palliative or surgical interventions till date.

KEYWORDS: Tricuspid atresia, Tricuspid atresia without pulmonary obstruction, Tricuspid atresia with prolonged survival, Tricuspid atresia with dilated pulmonary artery.

INTRODUCTION

Tricuspid atresia may be defined as congenital absence or agenesis of the tricuspid valve (Figures 1,2).[1] It is the third most common cyanotic congenital heart defect; the other 2 frequently observed cyanotic congenital cardiac anomalies are transposition of the great arteries and tetralogy of Fallot. Tricuspid atresia is the most common cause of cyanosis with left ventricular hypertrophy.[2]

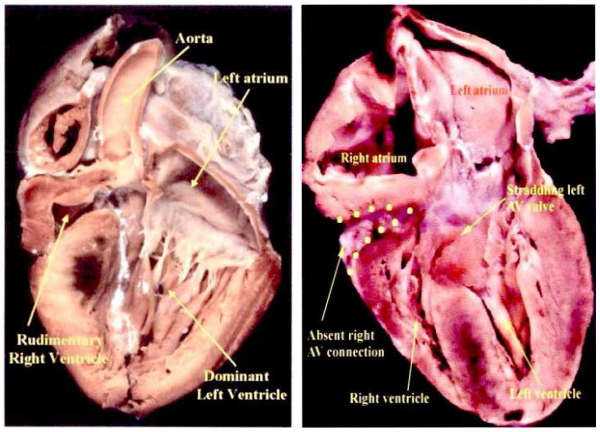

Figure 1: (A) Pathological specimen of Tricuspid Atresia demonstrating a dilated LV, rudimentary RV. LA is connected to the LV. (B) Another Specimen of Tricuspid Atresia identifying the absence of right atrioventricular connection, rudimentary RV and dilated LV.

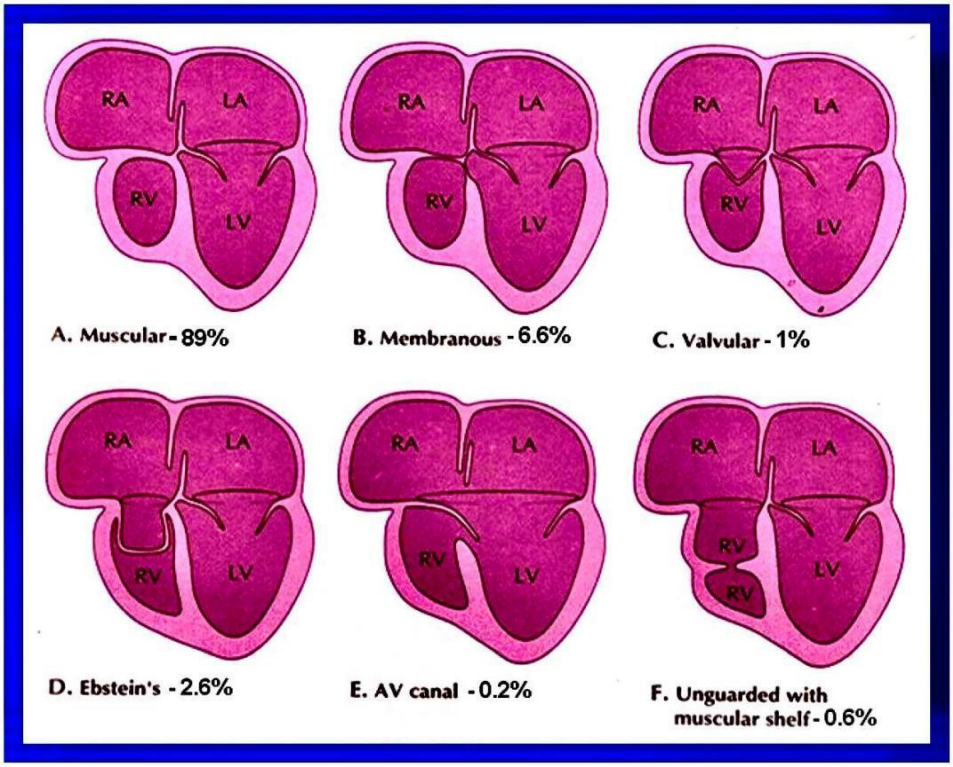

Figure 2: Diagrammatic portrayal of anatomic types of TA based on the morphology of the atretic TV. A: muscular type; B: membranous type; C: valvular type; D: Ebstein's type; E: AV canal type; and F: unguarded valve with muscular shelf. The prevalence of each is shown under each diagram. For the sake of simplicity, great vessels are not shown. Also, note that no VSDs are shown. LA, left atrium; LV, left ventricle; RA, right atrium; RV right ventricle.

Although some authors state that Holmes (1824) or Kuhne (1906) first described tricuspid atresia[3], Rashkind's methodical and thorough historical review indicates that Kreysig (1817) reported the first case in 1817.[4] An 1812 report by the editors of the London Medical Review (1812) appears to fit the description of tricuspid atresia, but they did not use this specific term.[4]

Although the true incidence of tricuspid atresia is not well defined, the prevalence of tricuspid atresia among congenital heart defects was estimated to be 2.9% in an autopsy series and 1.4% in a clinical series after extensive review. According to US data, tricuspid atresia may be estimated to occur in approximately 1 per 10,000 live births.[5] Tricuspid atresia has been classified according to the morphology of the valve[6], the radiographic appearance of pulmonary vascular markings[7,8] and the associated cardiac defects.[9-12]

Although these classifications are generally satisfactory, their exclusion of some variations in great-artery relationships and the lack of consistency in subgroups are problematic. Therefore, the following comprehensive-yet-unified classification was proposed[12] (Table 1).

Table 1: A new classification of tricuspid atresia.[12]

Type I. Normally related great arteries
Type II. D-transposition of the great arteries
Type III. Malpositons of the great arteries after other than D-transposition
Subtype 1. L-transposition of the great arteries
Subtype 2. Double-outlet right ventricle
Subtype 3. Double-outlet left ventricle
Subtype 4. D-malposition of the great arteries (anatomically corrected malposition)
Subtype 5. L-malposition of the great arteries (anatomically corrected malposition)
Type IV. Persistent truncus arteriosus
Each type and subtype are divided
Subgroup a. Pulmonary atresia
Subgroup b. Pulmonary stenosis or hypoplasia
Subgroup c. Normal pulmonary arteries (no pulmonary stenosis)

Tricuspid Atresia: Imaging Techniques

Various imaging modalities are mentioned below:

1. Echocardiography
- Fetal echocardiography
- 2Dimensional color doppler transthoracic echocardiography
- Transesophageal echocardiography
2. Cardiac CT
3. Cardiac MR
4. Cardiac angiography and catheterization

Multiple imaging modalities are utilized for detection and portrayal of TA (Figures 3-7).

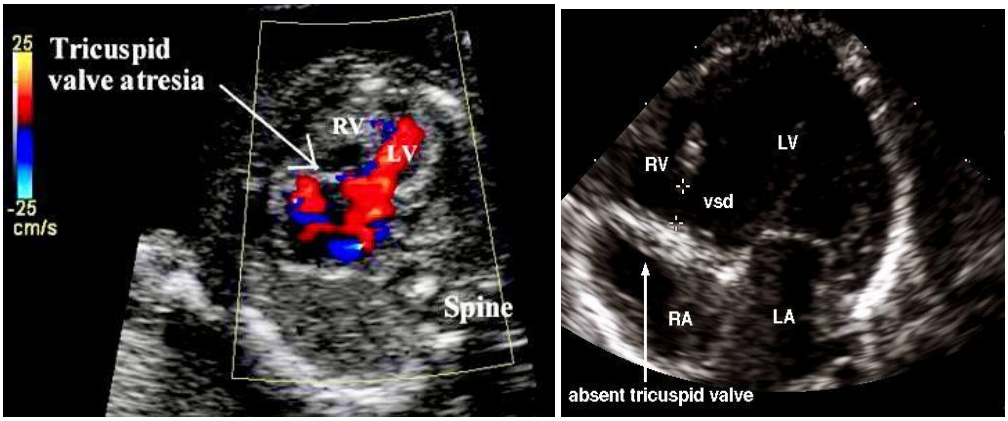

Figure 3: Echocardiographic images of TA. (A) Fetal echocardiogram depicting TA. (B) 2-Dimensional echocardiogram in 4CH view- identifying TA and a restrictive VSD.

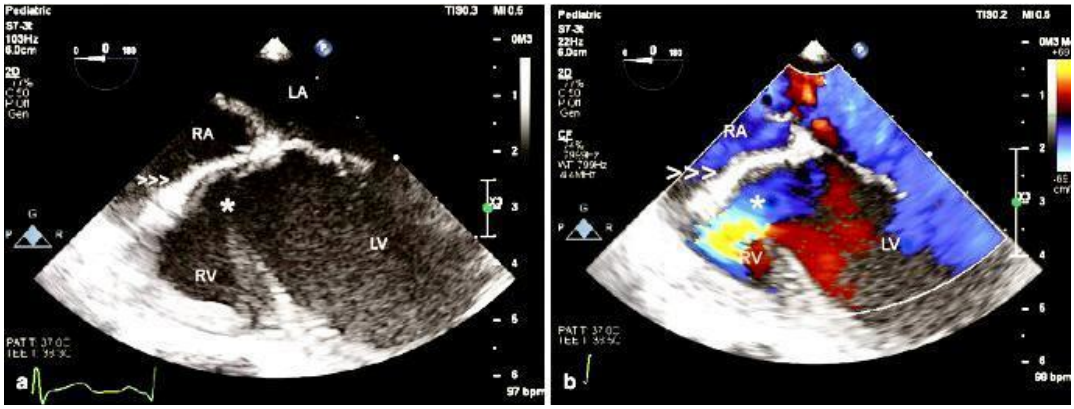

Figure 4: Transesophegeal echocardiogram showing TA (panel a) TA (arrows) alongwith restrictive VSD (asterisk) is identified (panel b) A turbulent mosaic pattern color flow jet is recognized (asterisk) with left to right shunt.

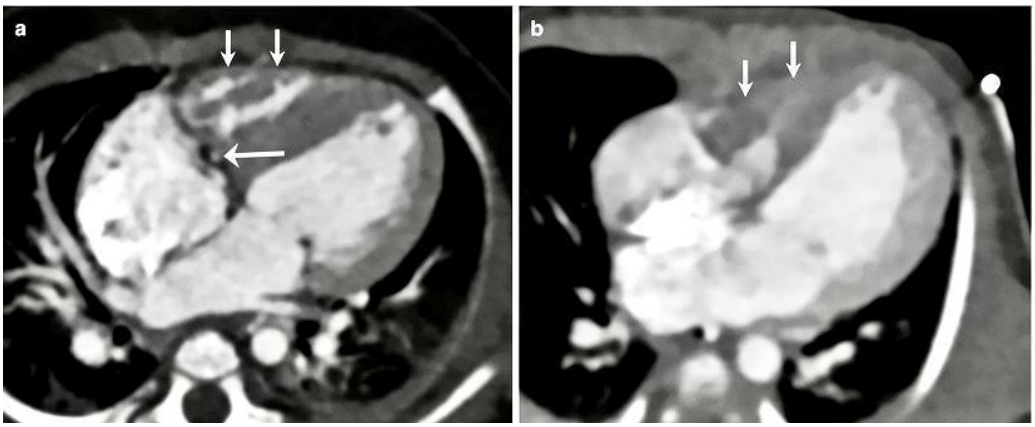

Figure 5: Cardiac CT images of Tricuspid Atresia. Four-chamber CT image shows the absent right atrioventricular connection occupied by areolar sulcus tissue with the deeply invaginated right coronary artery (long arrow) (a). Four- chamber CT image demonstrates the rarer type of tricuspid atresia in which the tricuspid valve is present but atretic (b). In both cases (a, b), the right ventricle (short arrows) appears severely hypoplastic.

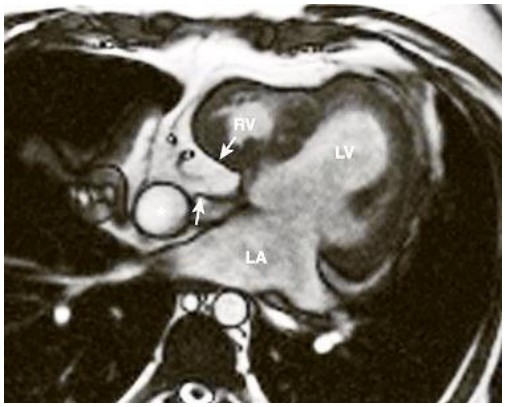

Figure 6: Cardiac MR image of TA. An axial magnetic resonance image of the heart shows a ridge of fatty tissue (arrows) in the floor of the right atrium at the site of the atretic tricuspid valve. A small caliber right ventricle (RV) also is present. LA, Left atrium; LV, left ventricle.

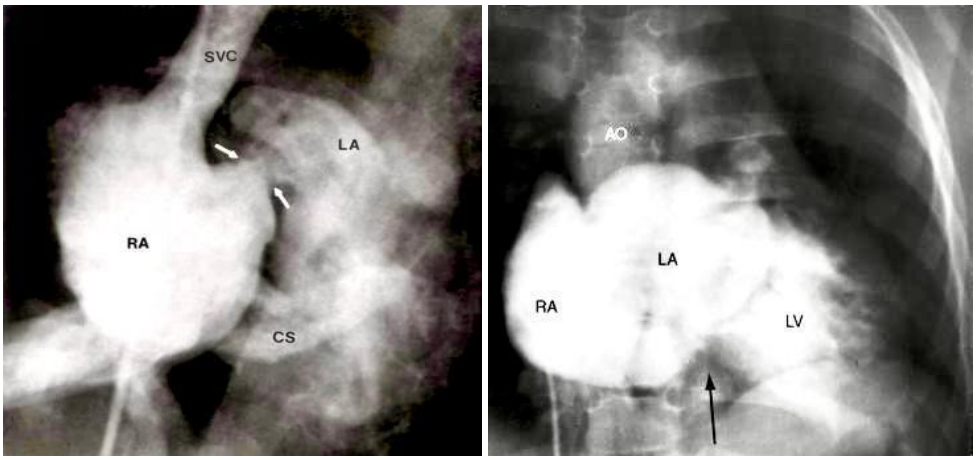

Figure 7: Cardiac angiography in TA. (A) Selective superior vena caval (SVC) injection for a 4- chamber projection (hepatoclavicular) shows tricuspid atresia and filling of the left atrium (LA) through a somewhat restrictive atrial septal defect (arrows). Note the retrograde filling of the coronary sinus (CS). RA = right atrium. (B) Angiocardiogram from a patient with tricuspid atresia, normally related great arteries, and an initially restrictive ventricular septal defect that closed spontaneously. Contrast material injected into the right atrium (RA) sequentially filled the left atrium (LA), left ventricle (LV), and aorta (AO). The right ventricle (unmarked arrow) remains unopacified as a wedge-shaped area of negative contrast.

Transthoracic echocardiography (TTE) is the first-line cardiovascular imaging modality in all types of tricuspid atresia patients. A computed tomography scan can provide the 3D spatial resolution imaging.

Poor prognosis of untreated tricuspid atresia patients is well known; only 10-20% of infants may live through the first year of life.[13] Considerable early mortality occurs and may be related to hypoxemia, cardiac failure, surgical intervention, or their combination. Surgical palliation to normalize pulmonary blood flow by means of systemic-to-pulmonary artery shunts in neonates with pulmonary oligemia and banding of the pulmonary artery in infants with markedly increased pulmonary flow improves survival rates.

CASE REPORT

A seventeen year old healthy looking female adolescent was referred to us for comprehensive cardiac evaluation and transthoracic echocardiography. She was a full-term normal delivery born out of non-consanguineous marriage. There was no history of maternal risk factors of CHD (obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure). The history of neonatal period, infancy and early childhood was narrated by the parents. They informed that the child was mildly cyanotic since birth and moreover the cyanosis became more apparent when the child used to cry. Otherwise, they denied any history of recurrent chest infections, intercostal retraction or failure to thrive.

Now the patient is in adolescent stage and she gives a history of bluish coloration of tongue, finger tips and toes of all the fingers since early childhood. Alongwith this, she also developed shortness of breath and fatigue on moderate effort. There is no history of chest pain, palpitation, loss of consciousness or pedal edema.

On clinical examination, the patient was of normal built, healthy looking, despite the presence of deep central cyanosis (Figure 8). Her weight was 51 kg, respiratory

rate was 15/min, pulse rate was 93/mm, blood pressure was 118/60 mmHg and SPO2 was 40 % at room air. There was no chest wall deformity or any other musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio femoral delay. However, notable clubbing was present at the tips of fingers and toes, of all the four extremities.

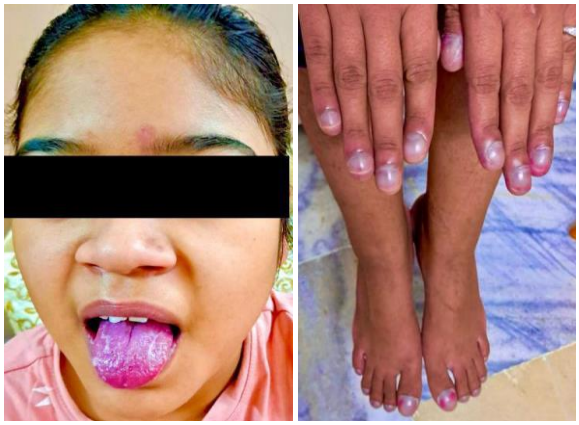

Figure 8: (A) Deep cyanosis of tongue and lips (B) Marked cyanosis and clubbing of tips of all the fingers and toes.

On cardiovascular examination there was presence of Grade III/IV pansystolic murmur heard all over precordium, best heard in IInd Rt & Lt intercostal space at the sternal border. The first heart sound was normal and the second heart sound was loud & closely split. There was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Pathological examination

The highlighting feature of the pathological examination results was elevated haemoglobin levels - 15.9 gm%, increased packed cell volume -52.2%, reduced platelet count- 53,000, raised serum uric acid - 7.8 mg% and a normal prothrombin time - 14.0 sec.

Xray chest PA view was consistent with levocardia, cardiomegaly with evidence of normal pulmonary blood flow (Figure 9).

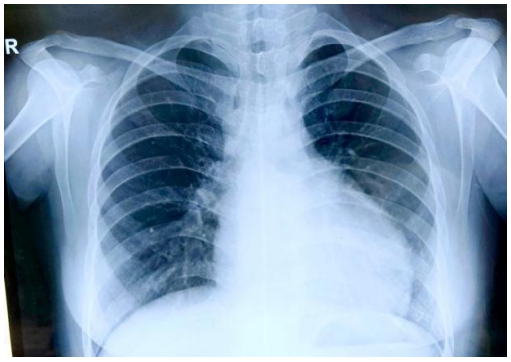

Figure 9: Figure 9: X-ray chest PA: There is conspicuous cardiomegaly with normal pulmonary blood flow.

Resting ECG identified (Figure 10) P pulmonale, bi-atrial enlargement, left axis deviation, left ventricular hypertrophy with strain and normal sinus rhythm.

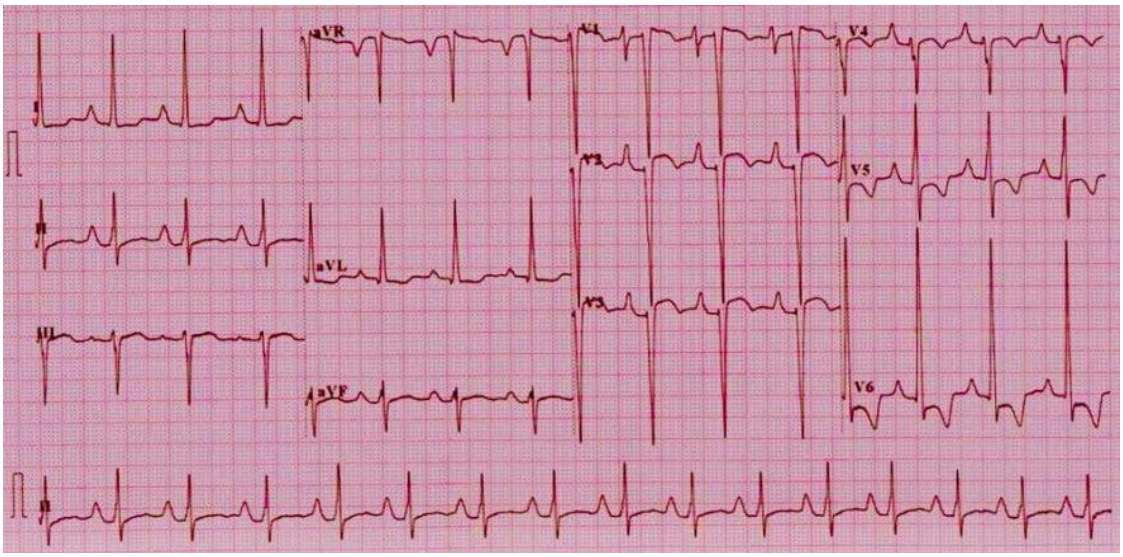

Figure 10: Resting ECG identifies P pulmonale, bi-atrial enlargement, left axis deviation, left ventricular hypertrophy with strain and normal sinus rhythm.

Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using a pediatric probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) and continuous wave doppler (CWD) echocardiography was performed in the classical

subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views. Contemporary sequential segmental approach for echocardiographic analysis of our index patient was accomplished and the characteristic features were outlined (Figures 11-17).

M-mode Echocardiography

Table 1: Calculations of M-mode echocardiography.

Measurements	LV
IVS d	6.7 mm
LVID d	67.6 mm
LVPW d	11.6 mm
IVS s	9.2 mm
LVID s	46.2 mm
LVPW s	18.1 mm
EF	58 %
%LVFS	32 %
LVEDV	236.4 ml
LVESV	98.4 ml
SV	138.0 ml
LV Mass	272 g

Summary of M-mode echocardiography

The LV was dilated (LVEDV = 236.4 ml, LVESV = 98.4 ml) with normal LV systolic function - LVEF was 58 % by M-mode and 55 % by Biplane Simpson's echocardiography. RV was hypoplastic (RV internal dimension in diastole - 13.4) with normal RV systolic function - RVEF was 55%.

2-Dimensional Color Echocardiography

2-Dimensional color echocardiography exhibited multiple features as mentioned below:

1. Levocardia

Situs Solitus
AV concordance
VA concordance
Concordant D-bulboventricular loop
Normally related great arteries (NRGA)
Left aortic arch, confluent pulmonary arteries.
Normal systemic venous drainage
Normal pulmonary venous drainage

2. Atresia of the tricuspid valve

3. Atrial septal defect (large)
Size : 24.8 mm
Ostium secundum type.
Rt to Lt shunt.

4. Ventricular septal defect (small)
Size 2.90 mm.
Perimembranous type
VSD jet peak velocity = 4.96 m/sec (gradient 98.6 mmHg)
Lt. to Rt. Shunt.

5. Mitral regurgitation (mild)
Large AML and PML present
MR jet velocity = 4.19 m/sec
On color flow mapping MR JET area 1.59 sqcm; 10 % of LA area, central jet.

6. Prominent dilated pulmonary artery and its branches were depicted.
PV annulus (D) 26.3 mm
Main pulmonary artery (D) 20.8 mm
Left pulmonary artery (D) 9.2 mm
Right pulmonary artery (D) 12.7 mm
Arotic valve annulus (D) 23.2 mm

7. Dilated LV, hypoplastic RV
Normal biventricular systolic function.
LVEF = 58 % (M-mode), 55 % (Biplane), RVEF = 55 % (M-mode).

8. No evidence of PDA, COA, AS, PS. Infundibular obstruction.

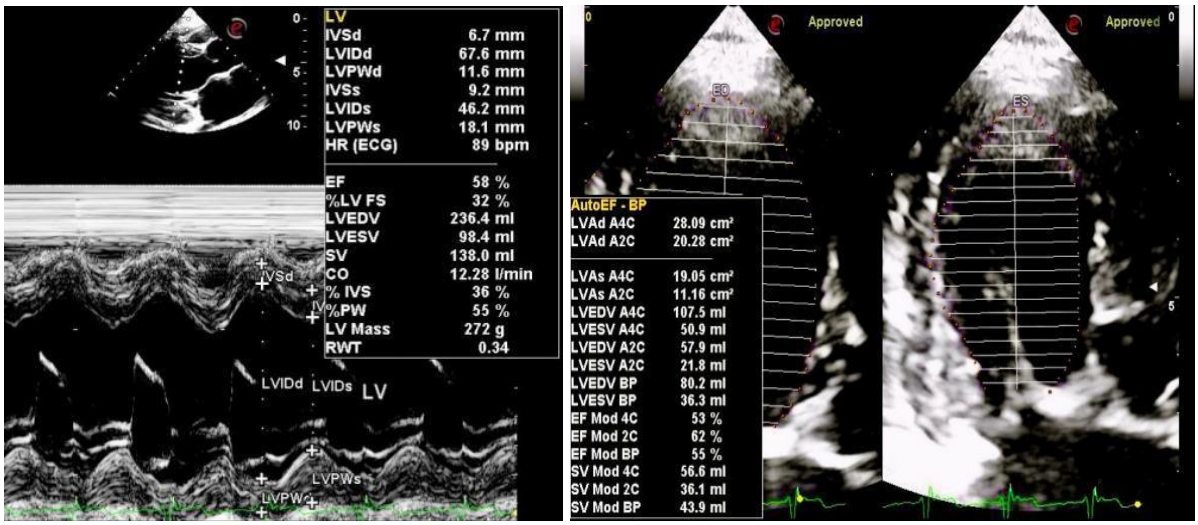

Figure 11: (A) M-mode echocardiography of the left ventricle. (B) Biplane Simpson's method to derive left ventricular systolic functions.

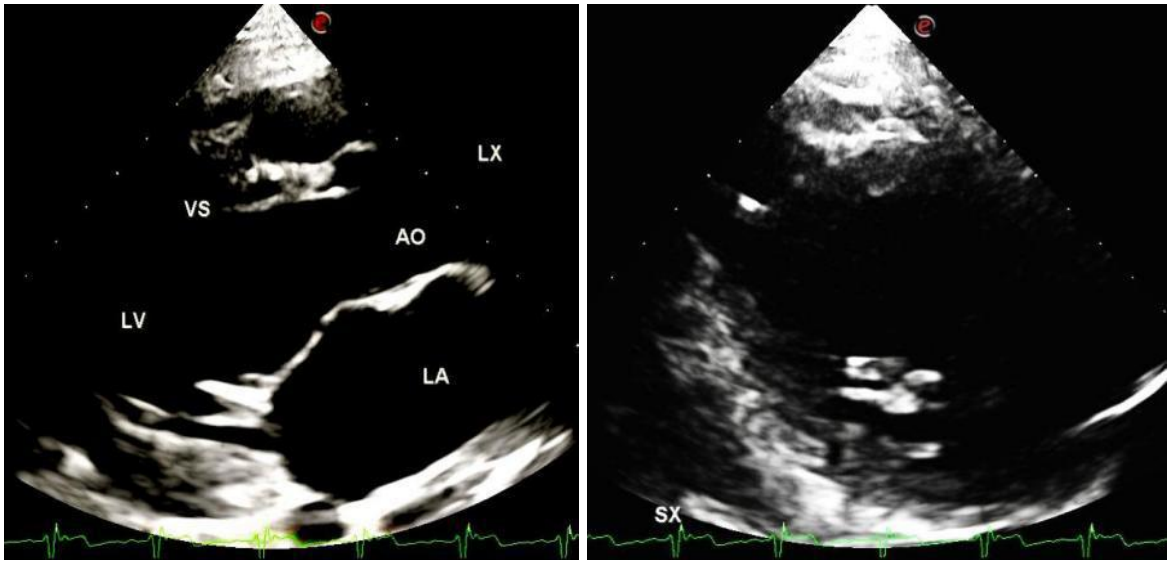

Figure 12: (A) LX view and (B) SX view portraying a large left ventricle.

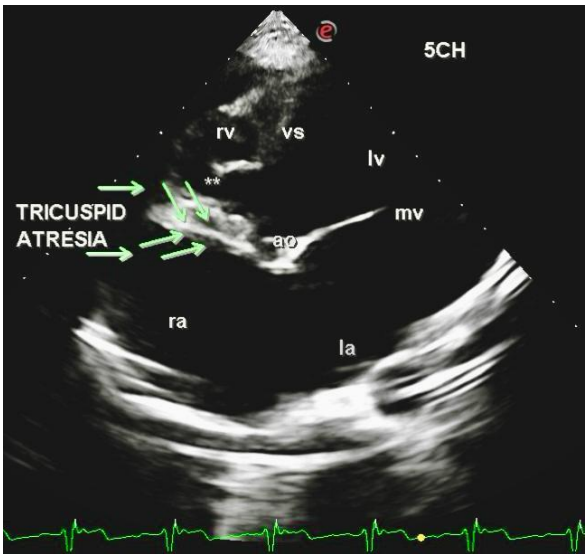

Figure 13: Classical portrayal of tricuspid atresia in the 5CH view. Arrows point towards a fibro-muscular ridge consistent with tricuspid atresia. Asterisk (**) denotes the presence of a small perimembranous VSD. Right ventricle is hypoplastic with conspicuous dilatation of left ventricle. There is also presence of dilated left and right atrium. ao, aorta; rv, right ventricle; vs, ventricular septum.

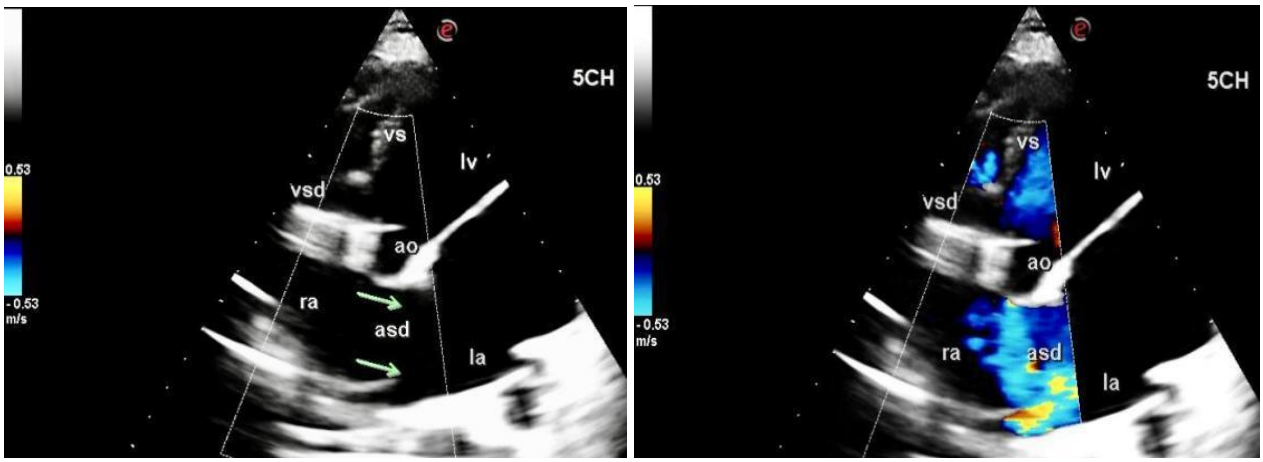

Figure 14: (A) 5CH view identifying a large ostium secundum ASD with (B) Right to left shunt. A small VSD is also visualised.

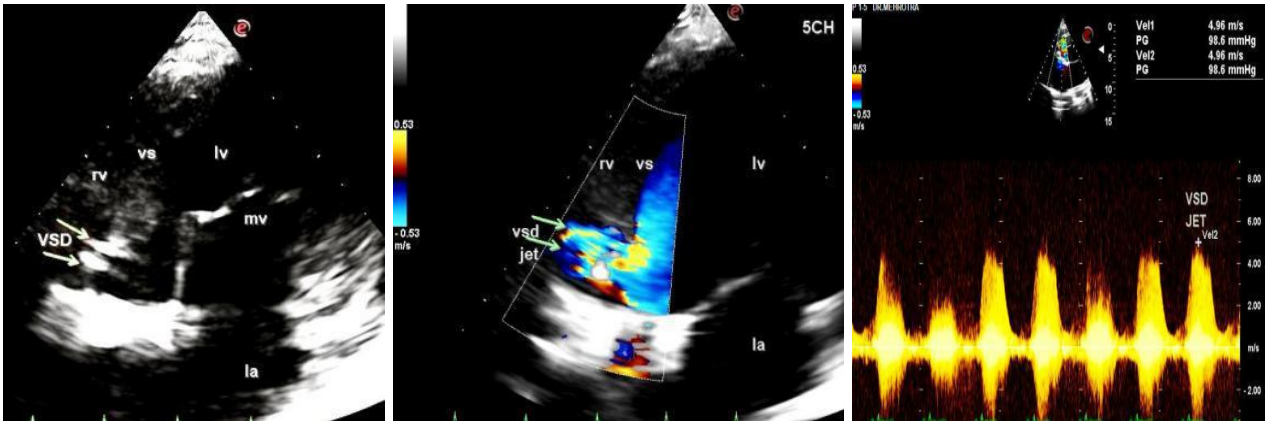

Figure 15: (A) 5CH view illustrates a small perimembranous VSD (arrows) with (B) an obligatory left to right shunt. There is marked hypoplasia of RV with a conspicuously dilated LV. (C) On continuous wave Doppler (CWD) analysis across VSD reveals a high velocity VSD jet of 4.96 m/ sec with a peak gradient of 98.6 mmHg.

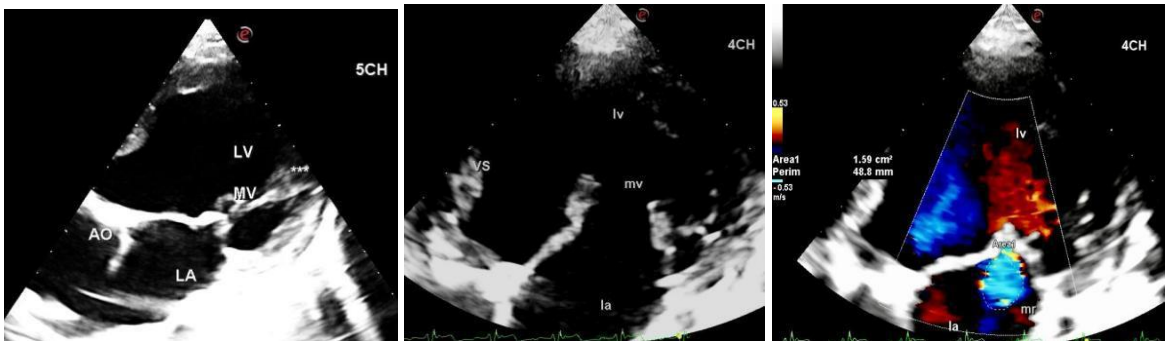

Figure 16: (A) 5CH view and (B) 4CH view demonstrates large anterior and posterior mitral valve leaflets with dilated left atrium and left ventricle (C) A central blue mosaic turbulent jet of mild mitral regurgitation is recognized (area 1.59 sqcm).

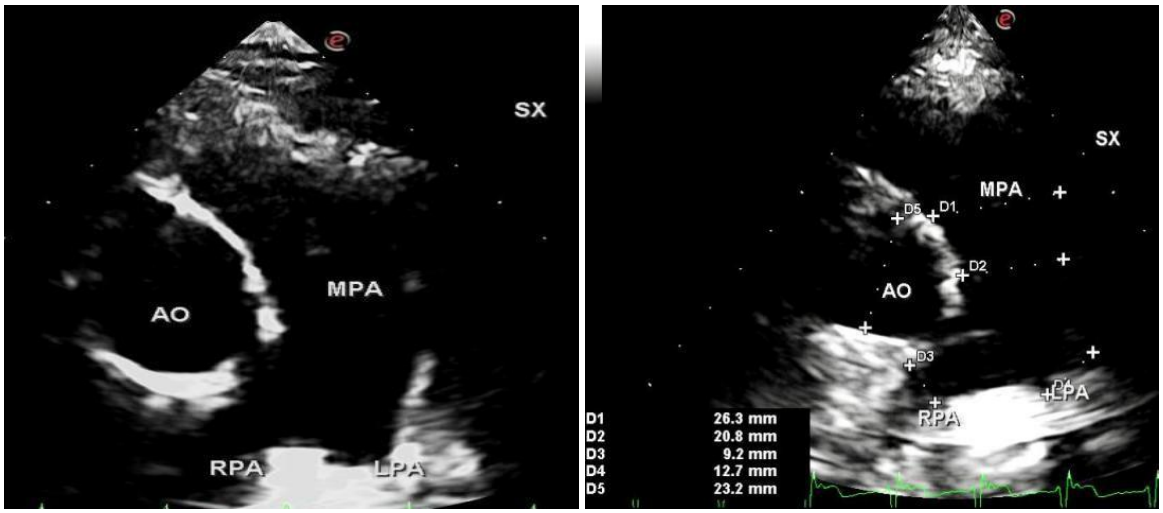

Figure 17: (A) SX view illustrates normally related great arteries with (B) Dilated pulmonary valve annulus, main pulmonary artery, left and right pulmonary arteries.

DISCUSSION & REVIEW OF LITERATURE

Tricuspid atresia (TA) is a cyanotic congenital heart disease (CHD) characterized by total agenesis of the tricuspid valve. It accounts for approximately 1% of all cases of CHD.[14] Its incidence is reported as ~0.1 per 1000 individuals.[15] Risk factors for TA include Down syndrome, poorly controlled diabetes, excess alcohol consumption during pregnancy, and a family history of CHD.[16]

TA is associated with high mortality without early intervention. Innovations in cardiac surgery has altered the dismal prognosis of TA, albeit long term survival is virtually unknown.[17]

Tricuspid atresia - clinical presentation

To better appreciate how these patients present, it is necessary to comprehend the most common route of blood flow through a heart with TA (Figure 18). In TA, the right atrium is separated from the hypoplastic right ventricle. This means that a patent foramen ovale or atrial septal defect (ASD) is required to enable deoxygenated blood from the systemic venous circulation to mix with oxygenated blood returning from the lungs.[18] Usually, this occurs in the left atrium, and the resultant partially oxygenated blood is then pumped out of the aorta by the left ventricle. To reach the right sided outflow tract, this partially oxygenated blood can be shunted from left-to-right across a ventricular septal defect (VSD). It then leaves through the pulmonary trunk, which may or may not be stenosed at its entrance, or entirely hypoplastic. In some cases where a VSD is not present or there is significant pulmonary obstruction, the patient is reliant on a patent ductus arteriosus (PDA) to enable blood flow into the pulmonary circulation. While the cardiac output is mixed in TA, it may still provide sufficient oxygenation to minimize cyanosis if it contains a great enough proportion of oxygenated pulmonary venous return. The degree of pulmonary blood flow (PBF) is, therefore, the main determinant of cyanosis in these infants. This, in itself, is determined by the extent of pulmonary obstruction, the existence and size of a VSD, the relationship of the great arteries, and the presence of a PDA.[19]

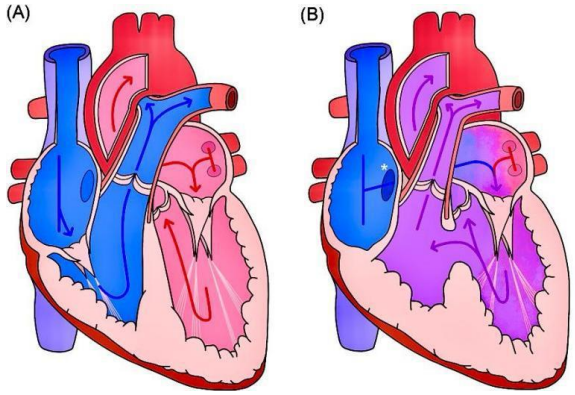

Figure 18: Blood flow in tricuspid atresia. A, Normal. B, TA. *Patent foramen ovale or ASD. ASD, atrial septal defect; TA, tricuspid atresia.

Around 50% of individuals with TA present with symptoms on the first day of life, with 80% being symptomatic by the first month.[20] Generally, two distinct presentations are observed. The first group of patients have high PBF due to left-to-right shunting across a VSD, a lack of pulmonary obstruction, and if transposition of great arteries (TGA) is present; they present with minimal cyanosis initially but go on to develop right-heart strain and cardiac failure in the first few weeks of neonatal life [20]. Patients typically exhibit fatigue, difficulties in feeding, failure to thrive, and recurrent respiratory tract infections.

The second group of patients consists of those with reduced PBF and accounts for the majority of infants with TA.[21] This is usually due to pulmonary obstruction and/or a small or absent VSD.[20] These patients exhibit central cyanosis, tachypnoea and hyperpnoea within the first few days of life.[20] Clinically, their condition may be exacerbated by the closure of the ductus arteriosus, as this is an important means by which these infants get blood flow to their lungs.

Long term survival in tricuspid atresia

The remarkably small number of adult cases with TA in world literature is indicative of the severe adverse prognosis usually associated with this disease.[22] The key factor to survival in TA is the adequacy of the pulmonary blood flow. Theoretically, the most favourable anatomic situation requires the presence of a large VSD with or without transposition of great vessels but with a sufficient degree of pulmonary outflow track obstruction to prevent excessive pulmonary blood flow. This anatomic type is present in Keith, Rowe and Vlad type I-C and II-B classifications and in few patients in type I-B category with a large VSD.[10] The association of transposition of great vessels with TA has often been said to favour long form survival, particularly if there is some pulmonic or subpulmonic stenosis.[9,23,24]

As mentioned earlier, survival in patients with TA has been shown to be hemodynamically dependent upon the presence of adequate PBF, with a lack of elevated pulmonary vascular resistance, and the degree of ventricular shunting. PBF is most commonly noted as the integral component of these factors.[8,22,25,26,27] Improvement of these factors, and thus of the patient's prospects at survival, are determinants of surgical correction at an early age.[8,28,29] In the recently reported case of Mckinney et al[30], elevated PBF and large VSD are consistent with chronic survival in type IC TA patients. Previously reported cases of uncorrected type I TA hearts with survival to adulthood have been noted to comprise cases where anatomic or functional obstruction to pulmonary outflow protected the pulmonary vasculature from suffering high systemic pressures,[22, 27, 31] This feature is also evident in the case report of Mckinney et al [30], where the pulmonary arteries demonstrate a normal size beyond the dilatation of the pulmonary trunk. This supports the interpretation that the increased volume and turbulent flow within the dilated portion of the pulmonary trunk may have created a functional obstruction balancing the pressure of the PBF with the PVR.

It has thus been speculated that survival is maximized in TA hearts with a large VSD and limited pulmonary outflow tract obstruction, such as in two patients of Jordan and Sanders.[8] However, surgical correction in TA patients with adequate PBF in earlier decades was presumed to be unlikely to improve outcomes until PBF began to decrease. Despite the potential for optimal anatomy in the context of TA, generally survival to

adulthood is very rare. The literature is sparse with only a few cases of type IC hearts with similar survival, and is increasingly rare as these cases are more commonly identified in infancy and treated at that time.[8,17,22,31 32] Most commonly, progression of pulmonary vascular obstruction and chronic heart failure are cited as the determinants of outcome in TA patients with prolonged survival.[8,25, 29, 31,33] From these reports it can be inferred that there is a fine balance between the PVR being low enough to maintain adequate pulmonary blood flow, and being high enough to prevent the shunted systemic pressures from overwhelming the pulmonary vasculature.

Likewise, our index patient of TA falls in Type IC class, according to the new classification.[12] Currently, she is sustaining an adequate pulmonary flow and hence, does not require any surgical palliation. She has been advised medical management until marked cyanosis, hypoxic spells and signs of early congestive heart failure appear.

Associated cardiac defects in tricuspid atresia.[34]
Defects that form the basis for classification are as follow:

- D-Transposition of the great arteries
- L-Transposition of the great arteries
- Double outlet right ventricle
- Double outlet left ventricle
- Other malpositions of the great arteries
- Truncus arteriosus [35]
- Hypoplastic ascending aorta and/or aortic atresia
- Ostium primum ASD
- Parchment right ventricle
- Patent ductus arteriosus
- Persistent left superior vena cava
- Right aortic arch
- Subaortic stenosis
- Total anomalous pulmonary venous connection
- Tubular hypoplasia of the aortic arch
- Valvar aortic stenosis

Other associated defects are as follows

- Juxtaposition of the atrial appendages
- Anomalous entry of coronary sinus into the left atrium
- Common atrium
- Cor triatriatum dexter
- Coronary sinus atrial septal defect
- Double aortic arch
- Double-outlet left atrium
- Hemitruncus

Detects that may need attention before or during palliative or total surgical correction are as follows:

- Absent pulmonary valve
- Aneurysm of the atrial septum
- Anomalous origin of the coronary arteries from the pulmonary artery
- Anomalous origin of the left subclavian artery
- Anomalous origin of the right subclavian artery
- Aortopulmonary fistula
- Coarctation of the aorta

Prognosis

Mortality/Morbidity

Poor prognosis of untreated tricuspid atresia patients is well known; only 10-20% of infants may live through the first year of life.

Considerable early mortality occurs and may be related to hypoxemia, cardiac failure, surgical intervention, or their combination.[36]

Surgical palliation to normalize pulmonary blood flow by means of systemic-to-pulmonary artery shunts in neonates with pulmonary oligemia and banding of the pulmonary artery in infants with markedly increased pulmonary flow improves survival rates.

The availability of PGE1 to keep the ductus open and advances in neonatal care (eg, early identification, safe transport to a tertiary care institution, noninvasive diagnosis by means of echocardiography), anesthesia, and surgical techniques have further decreased the initial mortality rate.[37]

The potential for improved prognosis suggests that every patient with tricuspid atresia should undergo aggressive medical and surgical treatment.

Adult patients who had classic Fontan operation have high initial mortality (28%) and high morbidity rates [38]. The latter is related to reoperation (58%) to revise Fontan connections, arrhythmia (56%) and thromboembolic events (25%). Patients with a total cavopulmonary connection appear to have improved survival and decreased morbidity rates, although follow-up of these patients has been relatively short.

CONCLUSIONS

The current case report demonstrates a unique cardiac and pulmonary physiology after chronic compensation of pulmonary vasculature to systemic blood flow. It is hypothesized that the dilatation of the pulmonary trunk primarily prevented pulmonary hypertension in this patient. It is likely that the patient's unique physiology, including the location of the VSD directly at the base of the aorta and the pulmonary trunk, allowed distention of the pulmonary trunk such that distal pulmonary blood flow was sufficient but at a non-pathologic pressure. The patient's protracted survival can be attributed to the increased PBF, as opposed to decreased PBF more commonly noted in TA.

Patients with TA and prolonged survival reported in the world literature maintain adequate but usually not

excessive pulmonary blood flow. The few patients who survive naturally to adult life probably have adequate pulmonary perfusion and will not benefit from palliative surgery unless there is evidence of a progressive reduction in PBF.

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