DOI: https://doi.org/10.3329/medtoday.v37i1.79501

CASE REPORT OPEN ACCESS

A Case Report of a 10-Month-Old Child Diagnosed with Tricuspid Valve Atresia

Golam Mahfuz Rabbani*[1]

Abstract

Introduction: *Tricuspid atresia (TA), a cyanotic form of congenital heart disease, is the absence of real or potential joining of the right atrium and right ventricle. This total absence of the right atrioventricular junction is typically caused by the regular position of the tricuspid valve, which forms a dimple close to the right atrium's floor exactly over the right ventricle.* ***Case history:*** *A 10-month-old male patient came to us with complaints of fever and respiratory distress. His oxygen saturation in room air was 76% with a heart rate of 130 and body temperature of 101 F. At first, he was diagnosed with pneumonia by a pediatrician. Then he came to me after not receiving any response from treatment. On examination I founded rhonchi and systolic murmur. So, our initial assessment involved taking CBC, x-ray and echocardiography of the patient's chest. The data from his echocardiography reveals that he has a tricuspid valve atresia.* ***Conclusion:*** *We decided to continue the conservative treatment for TVA with pneumonia (Paracetamol sos, Agoxin, Vitamin B complex for 6 months, and injectable antibiotic for 10 days) that started on him. After 11 months, the patient returned to us with RTI again. We treated his RTI with antibiotics. At the follow-up of the 12th month, his symptoms showed improvement.*

Keywords: *Diagnosis, Management, Tricuspid Valve Atresia.*

Number of Figures: 02; Number of References: 17; Number of Correspondences: 03.

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Introduction:

Tricuspid atresia (TA), a cyanotic form of congenital heart disease, is the absence of real or potential joining of the right atrium and right ventricle. This total absence of the right atrioventricular junction is typically caused by the regular position of the tricuspid valve, which forms a dimple close to the right atrium's floor exactly over the right ventricle[1]. Through the foramen ovale, every venous return from the systemic veins goes to the left side of the heart[1]. Atrial septal defect (ASD) of variable size is typically present[1]. According to the majority of reports, the prevalence of congenital cardiac anomalies, including tricuspid atresia, ranges from 1.5% to 2.5%[1]. The estimated incidence of this uncommon cyanotic congenital heart disease (CHD) is 79 per million live births[2]. According to the modified Edward-Burchells classification, tricuspid atresia can be divided into three categories depending on the degree of pulmonary stenosis and the relationship between the great arteries[3]. If paired with an unroofed coronary sinus, a persistent left superior vena cava (LSVC) may be linked to TA and result in persistent arterial desaturation even following curative surgery[4]. Prenatal diagnostics suggest that these types of treatment are more appropriate for infants who need surgery. Although the prenatal diagnosis of tricuspid atresia has been recorded several times, the prenatal sonographic components of this condition have not received much attention in the literature[5–10]. In our case report, we are presenting the echocardiographic images of a cyanosed ten-month-old child who presented with failure to thrive and fatigue on feeding.

Case History:

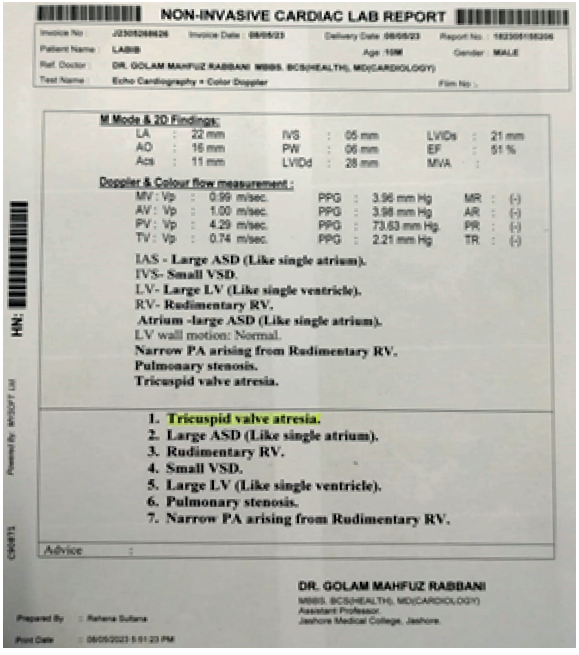
A 10-month-old male patient came to us with complaints of fever and respiratory distress. His oxygen saturation in room air was 76% with a heart rate of 130 and body temperature of 101 F. At first, he was diagnosed with pneumonia by a pediatrician. Then he came to me after not receiving any response from treatment. On examination I founded rhonchi and systolic murmur. Our initial assessment involved taking CBC, x-ray and echocardiography of the patient's chest [Fig.1&2]. The data from his echocardiography reveals that he has a tricuspid valve atresia. Echocardiography showed tricuspid valve atresia, large ASD (Like Single Atrium), rudimentary RV, small VSD, large LV, pulmonary stenosis, and narrow PA arising from rudimentary RV.

We recommended conservative treatment for the patient. He was treated with injectable antibiotics, Paracetamol, Agoxin, and Vitamin B complex. In the second week of his follow-up, he

Received: *15 June 2024* ***Accepted revised version:*** *25 August 2024*

improved. After treatment, his oxygen saturation in room air was 92% with a normal body temperature of 98 F. We continued the conservative treatment with Agoxin, Vitamin B, and Zinc on the patient.

After that, the patient was taken to India for further treatment. They also suggested the same conservative treatment as we did. After receiving the treatment for 11 months, the patient returned to us with RTI again. We treated his RTI with antibiotics. At the follow-up of the 12th month, his symptoms showed improvement.


NON-INVASIVE CARDIAC LAB REPORT

Patient Name : LABIB
Ref. Doctor : DR. GOLAM MAHFUZ RABBANI MBBS, BCS(HEALTH), MD(CARDIOLOGY)
Test Name : Echo Cardiography + Color Doppler

M Mode & 2D Findings:

LA	22 mm	IVS	05 mm	LVIDs	21 mm
AO	16 mm	PW	06 mm	EF	51 %
Acs	11 mm	LVIDd	28 mm	MVA	:

Doppler & Colour flow measurement :

MV : Vp	0.99 m/sec.	PPG	3.96 mm Hg	MR	(-)
AV : Vp	1.00 m/sec.	PPG	3.98 mm Hg	AR	(-)
PV : Vp	4.29 m/sec.	PPG	73.63 mm Hg.	PR	(-)
TV : Vp	0.74 m/sec.	PPG	2.21 mm Hg	TR	(-)

IAS - Large ASD (Like single atrium).
IVS- Small VSD.
LV- Large LV (Like single ventricle).
RV- Rudimentary RV.
Atrium -large ASD (Like single atrium).
LV wall motion: Normal.
Narrow PA arising from Rudimentary RV.
Pulmonary stenosis.
Tricuspid valve atresia.

1. Tricuspid valve atresia.
2. Large ASD (Like single atrium).
3. Rudimentary RV.
4. Small VSD.
5. Large LV (Like single ventricle).
6. Pulmonary stenosis.
7. Narrow PA arising from Rudimentary RV.

Advice :

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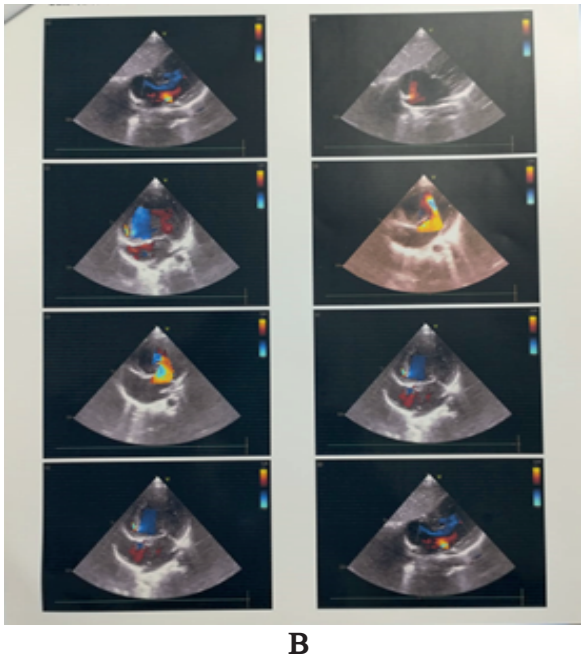

B

Figure 1: A) & B) Echocardiography of the patient

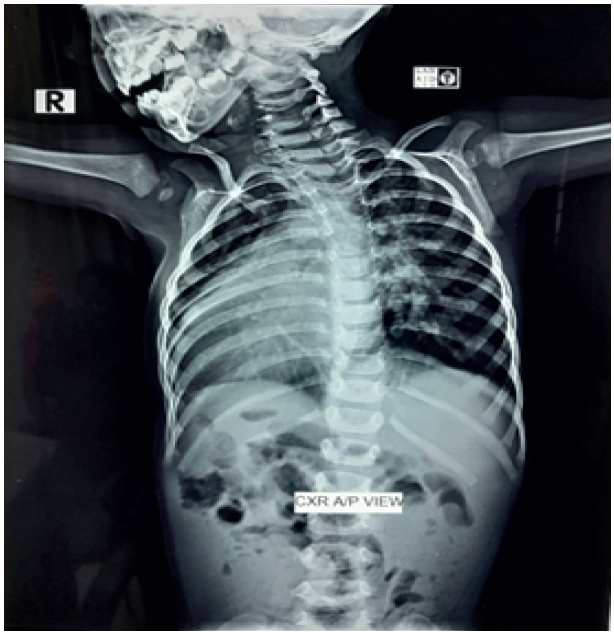

Figure 2: X-ray of the patient

Discussion:

Tricuspid atresia has been categorized using the modified Edwards-Burchells classification system[3]. Three varieties are known to exist: type I features big vessels that are ordinarily connected, type II features great vessels that are transposed, and type III has a persisting truncus arteriosus[3]. Three subtypes can be distinguished from each type: subtype A has pulmonary atresia, subtype B has pulmonary or sub-pulmonary stenosis, and subtype C is not associated with pulmonary or subpulmonary stenosis[3]. One of the rare congenital disorders identified during pregnancy is TA. Still, screening during standard obstetric sonographic scans may be able to recognize it, as it is typically linked to an aberrant 4-chamber image[5]. The present case is of a 10-month-old with tricuspid valve atresia associated with large ASD (Like Single Atrium), rudimentary RV, small VSD, large LV, pulmonary stenosis, and narrow PA arising from rudimentary RV. A persistent LSVC was present in 88 out of 2663 children (3.3%) in the biggest series on the presence of persistent LSVC with congenital cardiac disease[11]. 53 TA patients in the same series included three (5.6%) with an LSVC. A surgical repair may eventually need the identification of an LSVC. For those with TA, the Fontan's operation is the preferred course of care. It was necessary to carry out the Fontan operation in phases. The Glenn's shunt—a connection between the right pulmonary artery and the superior vena cava (SVC)—was made in the first step. After that, a four-step treatment was carried out in stages: the inferior vena cava (IVC) was connected to the PA through the right atrium (RA), a valve was inserted into the IVC, the ASD was closed, and the right ventricle (RV)

to PA link was obliterated[12]. The current recommended method involves modifying this to include an extra-cardiac conduit connected to the RA through a fenestration. The drawback of RA dilation, as shown in the atrio-pulmonary Fontan operation, is eliminated by the extra-cardiac conduit[12]. The fundamental idea behind the Fontan technique is to avoid the hypoplastic right ventricle by rerouting the systemic venous drainage directly to the pulmonary arteries. In a classical series, 90% of TA patients with cyanosis who were not operated on had died within a year[12] . However, survival rates were 90%, 81%, 70%, and 60%, respectively, at one month, one year, ten years, and twenty years following a large series of 225 patients who received the Fontan operation[13]. Most of the time, having an LSVC does not affect the treatment strategy or the likelihood of problems following surgery. Deoxygenated blood from the LSVC enters the left atrium through the unroofed coronary sinus and which can lead to prolonged arterial desaturation even after shunt surgery if the CS is unroofed and there is a persistent LSVC[14]. Contrast echocardiography makes it simple to diagnose CS unroofing. A cardiac computed tomography scan could support the same findings as well. Therefore, it is essential to determine whether a dilated CS and a persistent LSVC exist in children with TA or single ventricle physiology before surgery[15]. The most prevalent variant is a missing right atrioventricular connection with concordant ventriculoarterial connections, as observed in postnatal life. In contrast, discordant connections (transposed great arteries) and other abnormalities of arterial connections occur in around 20% of instances, though they are less common[16]. The neonate's cyanosis will increase if the major arteries are joined normally. The systemic venous return crosses over to the left side of the heart at the atrial level, resulting in a right-to-left shunt. Palliation may not be necessary if the atrial septum is unrestrictive and the VSD is of a reasonable size. However, because the VSD tends to contract with growing, surgical shunt creation might be necessary. In case the major arteries are transposed, the newborn may show signs of unrestricted pulmonary blood flow or a coarctation lesion. If there is not enough VSD and the aorta comes out of the right ventricle, there is a failure to thrive[17]. The presence of a tiny right ventricle and associated VSD, together with the lack of a patent tricuspid valve on the 4-chamber imaging and no flow across the valve on pulsed or color Doppler flow mapping, can be the primary findings for diagnosing various cases.

Conclusion and recommendations:

In our case report, we decided to continue the conservative treatment for TVA with pneumonia (Paracetamol sos, Agoxin, Vitamin B complex for 6 months, and injectable antibiotic for 10 days) that started on him. After 11 months, the patient returned to us with RTI again. We treated his RTI with antibiotics. At the follow-up of the 12th month, his symptoms showed improvement. The patients who came to us with recurrent pneumonia or respiratory tract infection, careful auscultation and echocardiogram needs to be done on them. If systolic murmur is found in auscultation, we recommend to do an echocardiography. It is necessary to obtain additional data from case series and case-control studies to elucidate the late complications and re-intervention rate after a 5-year follow-up.

Conflict of Interest: None.

Ethical approval: The study was approved by the Institutional Ethics Committee.

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