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(CASE REPORT)

Echocardiographic imaging of neonatal transposition of great arteries: Rare case report and review of literature

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Abstract

Transposition of the great arteries (TGA) is a congenital conotruncal abnormality characterized by discordant connections between the ventricles and great arteries, with the aorta originating from the right ventricle (RV), and the pulmonary artery (PA) originating from the left ventricle (LV). The two main types of TGA are complete transposition or dextro-transposition of the great arteries (D-TGA), commonly referred to as D-loop, and congenitally corrected transposition (CCTGA), commonly referred to as L-loop or L-TGA. In D-TGA, the connections between the ventricles and atria are concordant, whereas in CCTGA they are discordant, with the left atrium connected to the RV, and the right atrium connected to the LV. Imaging plays an important role in the evaluation of TGA, both before and after surgery, for helping define the anatomy, quantify hemodynamics, and evaluate complications. Transthoracic echocardiography is the first-line imaging modality for pre-surgical planning in children with TGA. MRI provides comprehensive morphologic and functional information, particularly in adults after surgery. CT is performed when MRI is contraindicated or expected to generate artifacts.

We are reporting an extremely rare and unique case of 25-day old male neonate, suffering from TGA with a side to side alignment of great arteries, accompanied by a moderate sized restrictive atrial septal defect (ASD) and an ascending aortic aneurysm (AAA).

Keywords: Neonatal TGA; Neonatal ascending aorta aneurysm; Side by side great artery relationship; Transposition of great arteries

1. Introduction

Transposition of the great arteries (TGA) is a rare anomaly with an estimated prevalence of 5%-7% among all congenital heart diseases (CHD) (Figure 1, 2) [1].

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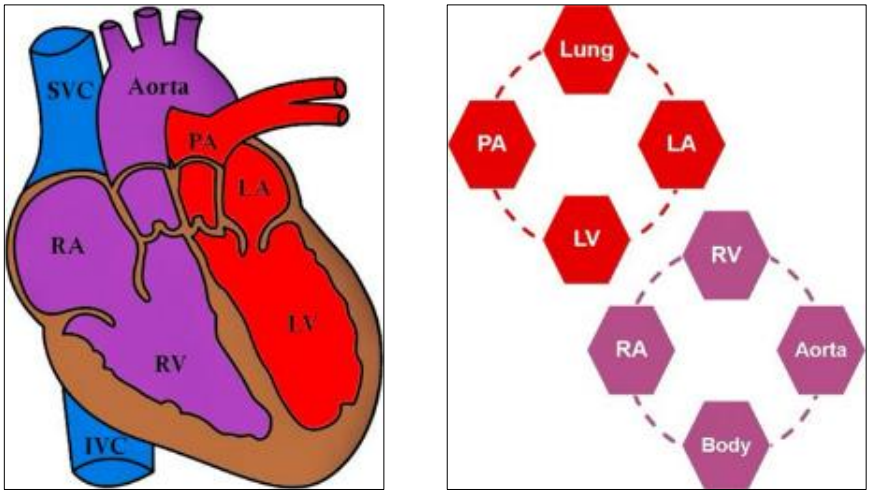

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Figure 1 D-TGA. LA=left atrium. (A) Illustration shows the anatomy of the heart and great arteries in D-TGA. The morphologic RV is connected to the aorta, and the morphologic LV is connected to the PA. There is concordance between the atria and ventricles. IVC=inferior vena cava, SVC=superior vena cava. (B) Schematic diagram shows the hemodynamics in a patient with D-TGA. There are essentially two parallel circulations

Figure 2 Anatomic specimen of complete TGA with intact septum. Pulmonary trunk arises from the left ventricle and aorta from the right ventricle (discordant VA connection). The ventricular septum is intact

The main clinical manifestations of TGA are cyanosis and dyspnea [2]. TGA has a normal atrioventricular connection but a ventriculoarterial discordance, in which the pulmonary artery arises from the morphological left ventricle and the aorta arises from the morphological right ventricle (RV) [3]. TGA is considered to account for a variety of organ damage in infants because of the decreased level of brain oxygen [4, 5]. Thus, a delayed diagnosis and intervention will expose patients to increased risk [6]. It is vital that the choice of operation is based on the knowledge of the spatial relationship of the great arteries and the coronary artery abnormalities. Accurate and comprehensive evaluation of TGA and associated abnormalities is thus critical before surgery.

2. Imaging in neonatal transposition of great arteries

2.1. Goals of Imaging

The goals of imaging in patients with TGA [7] are to provide accurate and reproducible anatomic and hemodynamic information that facilitate medical and surgical planning and to provide surveillance imaging to evaluate potential issues related to the type of surgical operation that has been chosen.

2.2. Imaging Modalities

Multiple options available for imaging in neonatal TGA are outlined (Figures 3-6) [7]:

- Echocardiography
- Cardiovascular Magnetic resonance (CMR)
- Cardiovascular Computed Tomography (CT)
- Cardiac Catherisation and Angiography

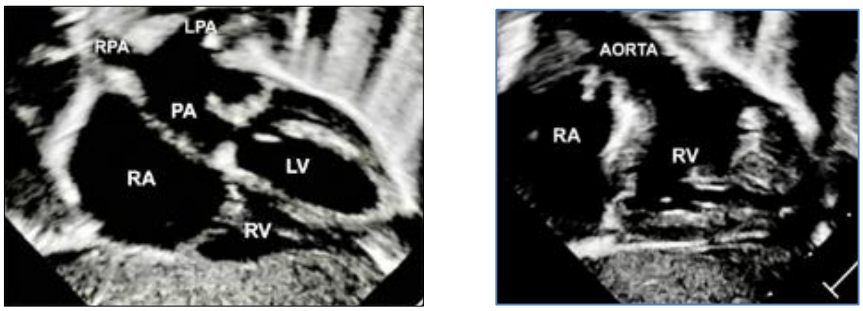

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Figure 3 Echocardiography in D-TGA. (A) Five-chamber echocardiographic image shows the PA originating from the LV. LPA=left PA, RPA=right PA. (B) Right ventricular outflow tract echocardiographic image shows the aorta originating from the RV

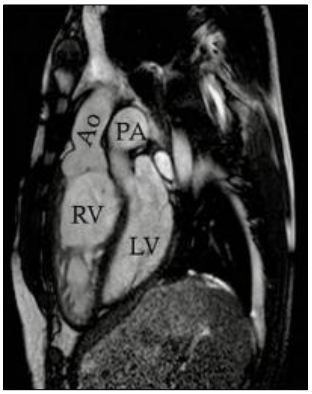


Figure 4 Cardiac CMR in TGA. Sagittal cine SSFP MR image in the same patient shows parallel orientation of the aorta and main PA

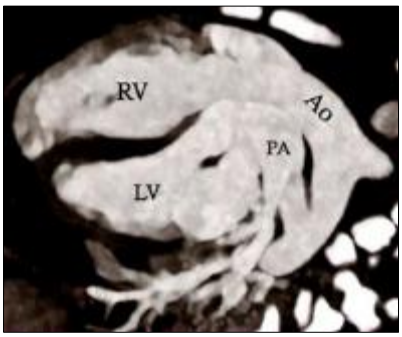
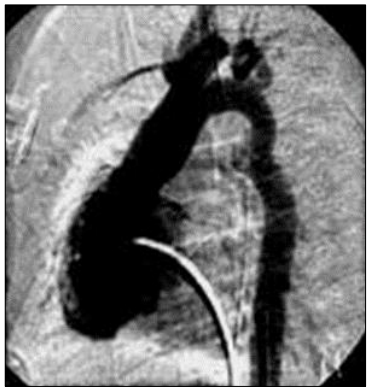


Figure 5 Cardiac CT image of TGA. D-TGA appearance at CT. Axial oblique reformatted maximum intensity projection image from cardiac CT angiography in a patient with D-TGA shows the LV connected to the main PA and the RV connected to the aorta (Ao)



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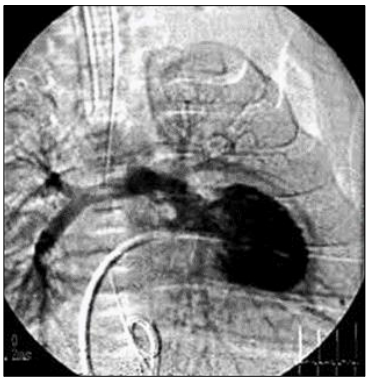

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Figure 6 (A) Right ventricular angiogram. Transposition of the Great Arteries. This left ventricular angiogram shows the pulmonary artery arises directly from the left-sided posterior left ventricle (30° right anterior oblique [RAO]). (B) Left ventricular angiogram. Transposition of the Great Arteries. This right ventricular angiogram shows the aorta arises directly from the right-sided anterior right ventricle (70° left anterior oblique [LAO])

Comparative advantages of echocardiography, CMR and cardiac CT are elucidated in the Table 1 [7].

Table 1 Comparison of non-invasive imaging techniques

Characteristic	**Echocardiography**	**CMR**	**CT angiography**
Availability	++++	++	++
Portability	++++	-	-
Radiation exposure	-	-	+++
Safety with pacers	++++	+	+++
CA anatomy	++	+++	+++
Aortopulmonary collateral vessels	+	++++	++
Supravalvar aortic stenosis (ASO)	++++	++++	++++
Supravalvar pulmonary stenosis (ASO)	++++	++++	++++
Branch PA stenosis (ASO)	++	++++	++++
Neoaortic root dilation (ASO)	++++	++++	++++
Neoaortic regurgitation (severity) (ASO)	++	++++	-
CA stenosis (ASO)	+	+++	++++
Myocardial ischemia (ASO)	+	+++	-
Systemic venous baffle obstruction (AtrSO)	++	+++	+++
Pulmonary venous baffle obstruction (AtrSO)	++	+++	+++
Baffle leak (AtrSO)	++	++	-
RV dysfunction (AtrSO)	++	++++	+++
Residual VSD (Rastelli/Nikaidoh)	++++	+++	+
Subaortic obstruction (Rastelli/Nikaidoh)	++++	++++	+++
Conduit obstruction (Rastelli/Nikaidoh)	+++	+++	+++
Conduit regurgitation (Rastelli/Nikaidoh)	++	++++	+
CA; coronary anatomy, ASO; arterial switch operation, AtrSO; atrial switch operation.			

3. Case Report

A 25 day old male neonate was referred to us for a comprehensive transthoracic echocardiography.

He was a full term normal delivery from a multipara primipara woman of 30 years of age. There was no history of maternal risk factors of CHD (obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure). On clinical examination, the child was very "sick-looking" and was having deep cyanosis, severe respiratory distress, intercostal retractions, tachypnea, facial puffiness (Figure 7A).

He was of average built, highly irritable and persistently crying. His weight was 3.8 kg, respiratory rate was 38/min, pulse rate was 151/mm, blood pressure was 100/70 mmHg and SPO2 was 60% at room air for which he was intubated and CPAP device was installed. Moreover, supplemental oxygen inhalation was given to maintain oxygen saturation of > 96%. The child was cyanosed with bluish coloration of tongue, lips, all the fingers, and toes. There was a conspicuous protruding chest wall deformity (Figure 7B) without any other musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio femoral delay.

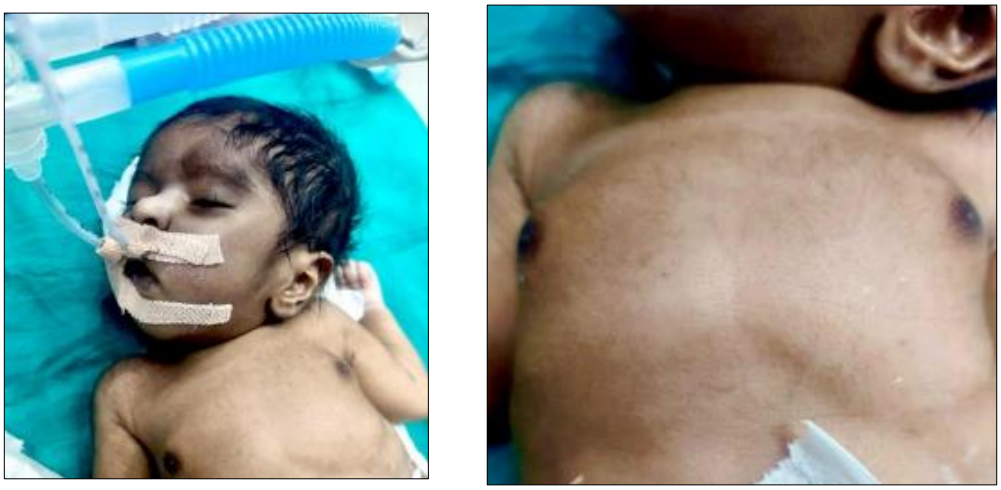

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Figure 7 (A) Sickly looking neonate in altered sensorium and deeply cyanosed. (B) Markedly protruding chest with intercostal retraction and severe respiratory distress

On cardiovascular examination there was presence of Grade 2/6 pansystolic murmur over precordium, best head over lower right sternal border. Rest of the systemic examination was unremarkable.

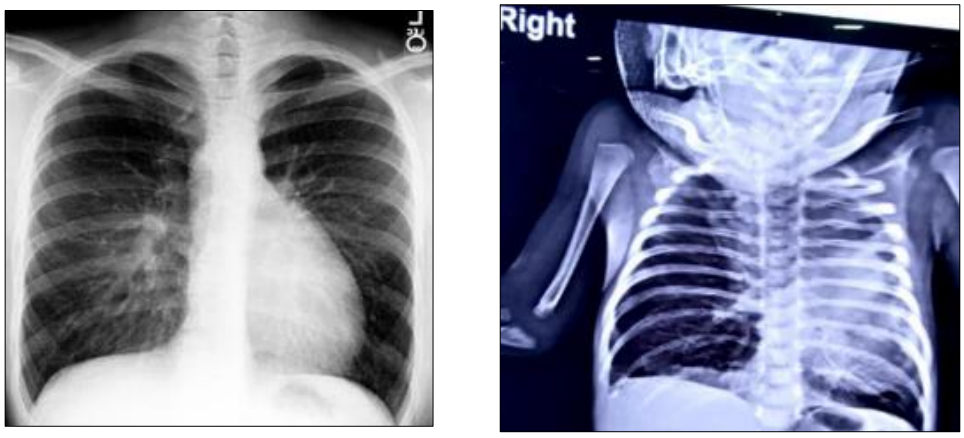

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Figure 8 (A) X-ray chest PA of another patient with TGA. There is a classical "egg on string appearance". (B) Our index patient X-ray chest AP view. Resembling the "egg on string appearance"

Xray chest AP view depicted a resemblance of "egg on string appearance" (Figure 8). Resting ECG revealed (Figure 9) sinus tachycardia with a ventricular rate of 150/min right axis deviation, prominent R waves in right precordial leads and increased S waves in the left precordial leads, consistent with RV hypertrophy.

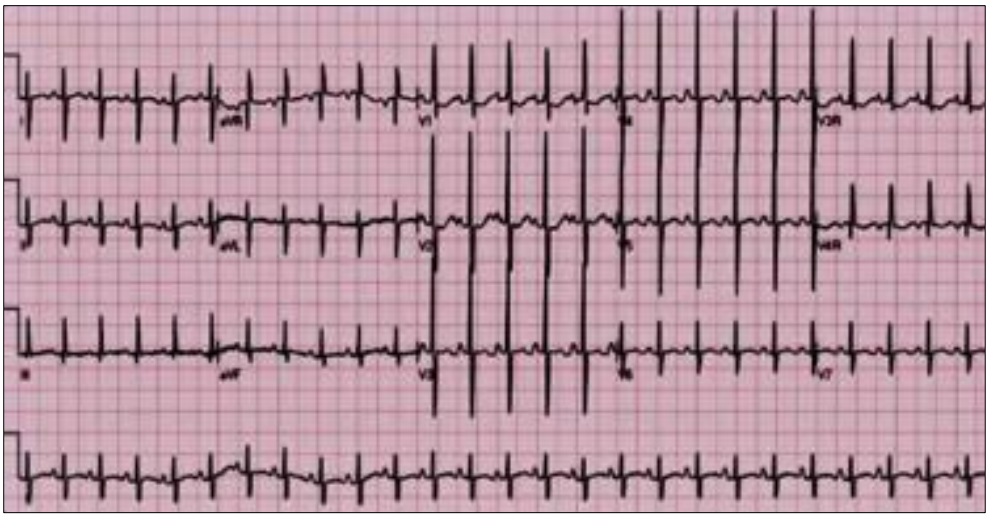


Figure 9 Resting ECG. There is right axis deviation and prominent R waves in right precordial leads and increased S waves in the left precordial leads consistent with RV hypertrophy

3.1. 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 position.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) echocardiography 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 patient was accomplished and the characteristics of the neonate are outlined (Figure 10-15):

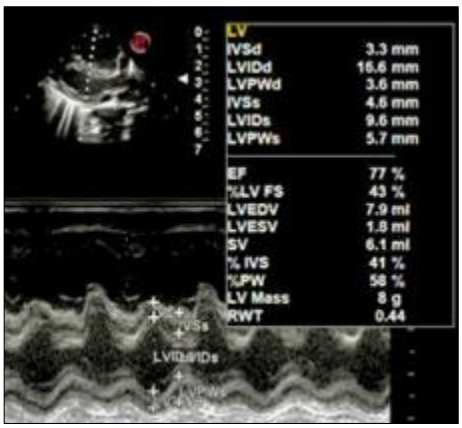
3.2. M-mode Echocardiography

The features of M-mode echocardiography are mentioned:

LV	
IVS d	1.5 mm
LVID d	18.0 mm
LVPW d	2.3 mm
IVS s	3.6 mm
LVID s	13.2 mm
LVPW s	4.1 mm
EF	56 %
%LVFS	27 %
LVEDV	9.7 ml
LVESV	4.3 ml
ESV	5.4 ml

3.3. 2-Dimensional Color Echocardiography

- Levocardia
  - Situs Solitus
  - AV Concordance
  - VA Discordance
  - Transposition of Great Arteries
  - Both great arteries are lying side by side
  - Left Aortic Arch, Confluent Pulmonary Arteries.
- Transposition of great arteries
  - Aorta is arising from morphologic RV.
  - PA is arising from morpholic LV.
  - Both great arteries were lying side by side; PA is lying on the right and the aorta on the left.
- Atrial septal defect (Moderate) –
  - Size : 3.7 mm
  - Ostium secundum type.
  - Peak/mean gradient across ASD = 13.1/5.5 mmHg
  - Lt to Rt shunt.
- Patent ductus arteriosus (Large) –
  - Size : 5.2 mmHg
  - Lt to Rt shunt
- Tricuspid regurgitation (Mild)
  - TV normal
  - TR velocity = 3.86 m/sec (gradient 59 mmHg)
- Both great arteries were dilated
  - Aortic annulus (D) 15.1 mm
  - Ascending aorta (D)17.9 mm
  - PA annulus (D) 12.2 mm
  - Right pulmonary artery (D) 5.9 mm
  - Left pulmonary artery (D) 2.4 mm
- Normal biventricular dilation with normal systolic function.
  - Normal LVEF = 77 %
- No evidence of VSD, COA, AS, PS.



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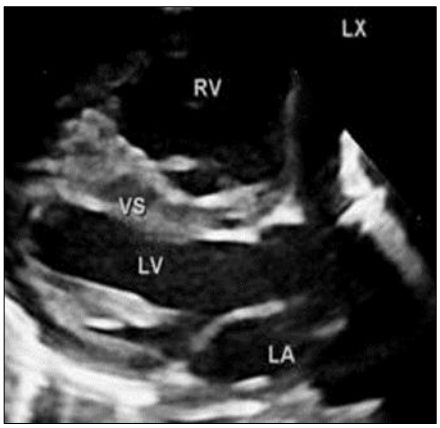

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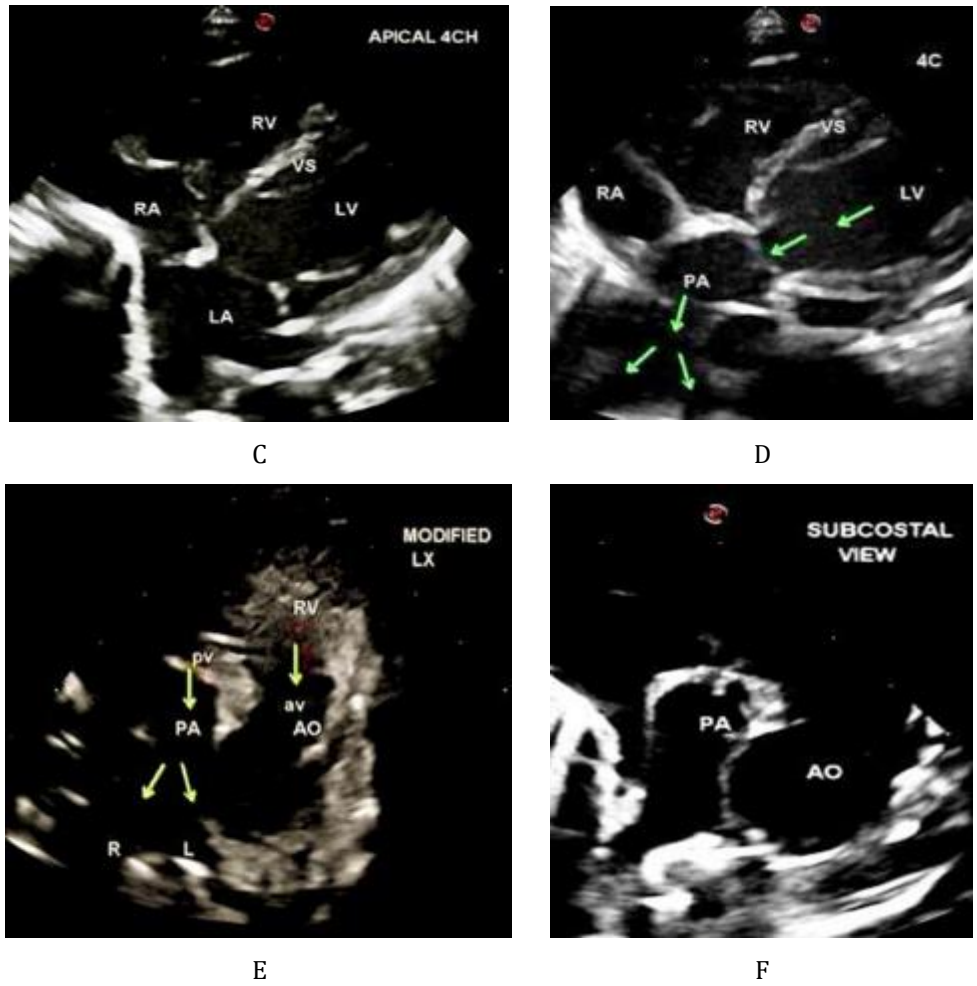

C D

E F

Figure 10 (A) M-mode measurements at the level of LV. (B) LX View. LV is connecting to pulmonary artery. There is RV dilatation. (C) 4CH View. There is presence of atrio-ventricular concordance. (D) 4CH View. LV connects with the pulmonary artery suggesting ventriculo-arterial discordance. (E) Modified LX View. Depiction of ventriculo-arterial concordance consistent with TGA is identified. RV connects to aorta and LV to pulmonary artery. pv; pulmonary valve, PA; pulmonary artery, R; right pulmonary artery, L; left pulmonary artery, RV; right ventricle, av; aortic valve, AO; aorta. (F) Subcostal view. Side to side arrangement of great arteries with PA lying on the right side of aorta

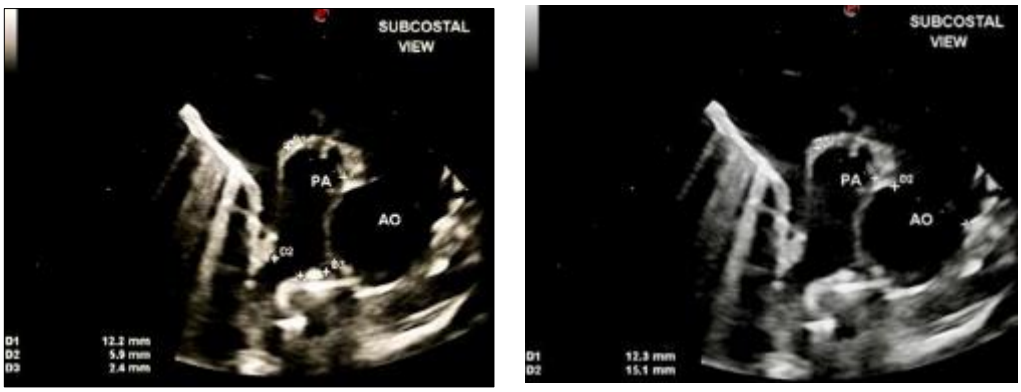

Figure 11 Subcostal view showing side to side spatial relationship of great arteries. Both PA and AO are dilated. Main pulmonary artery (D=12.2 mm), right pulmonary artery (D= 5.9 mm), left pulmonary artery (D= 2.4 mm), AO (D= 15.1 mm)

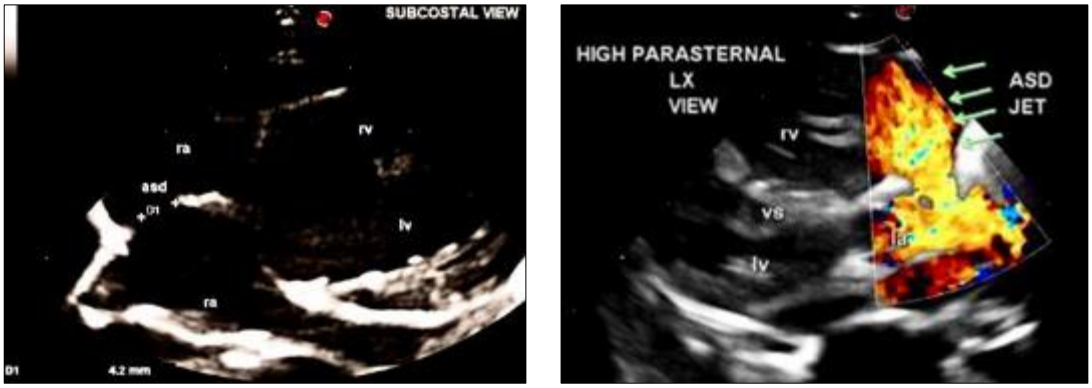

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Figure 12 (A) Subcostal view. Moderate sized ostium secundum ASD (size= 4.2 mm) is present. (B) High parasternal LX view. A conspicuous turbulent jet across ASD is delineated

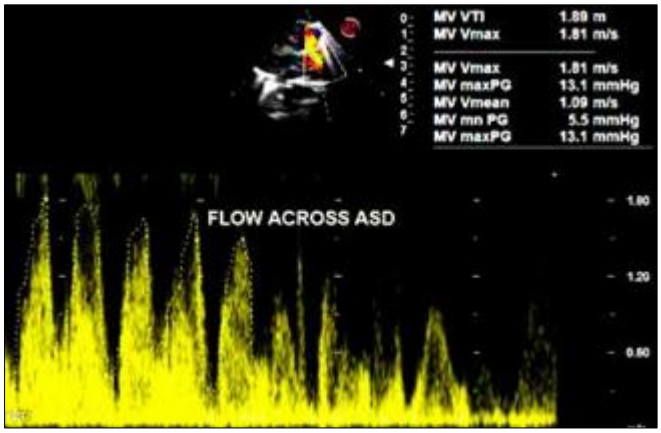

Figure 13 Pulse wave doppler (PWD) analysis across ASD reveals a peak/ mean gradient of 13.1/5.5 mmhg, suggesting a restrictive ASD flow

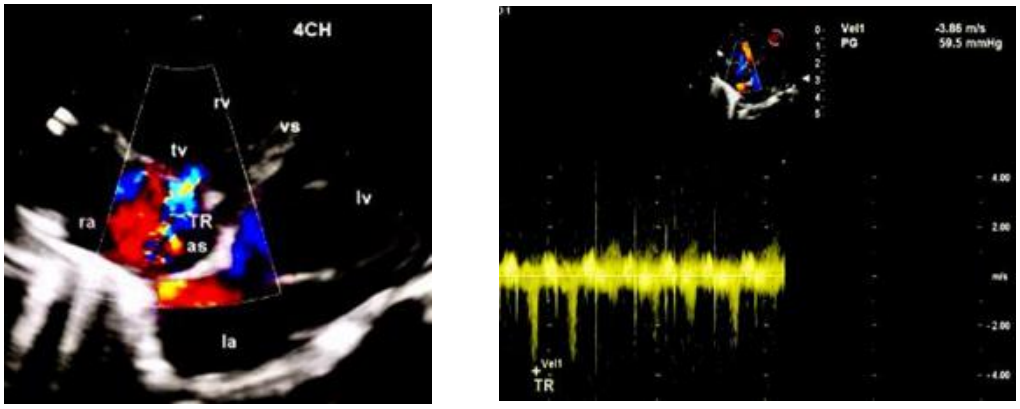

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Figure 14 (A) 4 CH View. Mild tricuspid regurgitation (TR) is observed. tv; tricuspid valve, vs; ventricular septum, as ; atrial septum. (B) On continuous wave doppler (CWD) analysis across the tricuspid valve a TR velocity of 3.85 m/sec (gradient 59.5 mmhg) is delineated

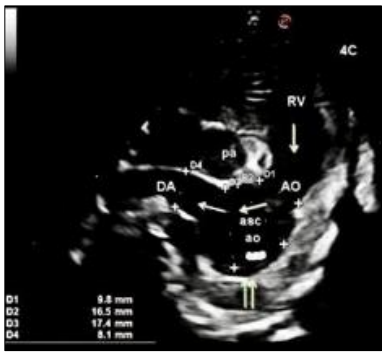

Figure 15 4CH View. Conspicuous dilatation of aorta connected to the RV is discerned

4. Discussion

Infant with d-TGA are usually born at term, while cyanosis appears within a few hours after birth. The amount of mixing of blood of the systemic and pulmonary circulation determines the survival of the TGA patients, and the clinical manifestation and the course of disease are largely dependent on this [8]. Our index case had complete TGA Type 1, with intact interventricular septum (IVS) although it presented late due to high level of intracardiac mixing occurring thought the moderate sized ASD and a large PDA. Similar presentations have been earlier mentioned by many other authors [8-11].

Early administration of PGE1 (alprostadil) within the first 48 hours after birth has been found to reduce early mortality in newborns with TGA [12]. The patient needs to be commenced on PGE1 infusion at the rate of 0.05-0.1 µg/kg/min as soon as the diagnosis of d-TGA is confirmed. This will help to maintain ductal patency and promote left-to-right intracardiac mixing, and thereby increase the level of oxygenated blood reaching the left atrium [10]. The volume-overloaded left atrium is likely to shunt part of its content into the right atrium and improve the oxygen saturation of the aortic blood.

The outcome of the patients would be favourable if oral or parenteral prostaglandin, Rashkind's septostomy and corrective cardiac surgery are available. Cardiac catheterisation and Rashkind balloon atrial septostomy is indicated for a patient who responds poorly to PGE1. The procedure is used to improve atrial level shunt and oxygenation. Improving atrial level shunting will allow the infant to gain more time for the definitive surgery-arterial switch operation (ASO) [10, 13].

The ideal surgical correction for d-TGA Type 1 is ASO or Jantene procedure, which aims to re-establish ventriculo-arterial concordance by detaching both great arteries and re-attaching them to the anatomically correct ventricles. The coronary arteries are also excised and re-implanted to the new aorta. This procedure is best performed within the first 4 weeks of life, when the pressure in the pulmonary vasculature is still high to prevent left ventricular involution. Once the pull pressures drop, the left ventricle becomes used to low pressure performance and involutes (i.e. loses the capacity to pump against high systemic pressures postoperatively [13].

ASO (Senning's or Mustard) is recommended for infants with a late presentation a two-way intra-arterial baffle is created in the top part of the heart using the patient's tissue or synthetic material to direct blood flow into the appropriate ventricles. Complications that may arise are related to baffle obstruction (superior and inferior vena cava syndrome) and heart failure [12]. However, a review of 22 patients with TGA who had an intact ventricular [14] septum reported that ASO is still a feasible and safe option for patients presenting late in developing countries if surgical resources are available. The outcome of those presenting as late as 9 weeks postnatally are similar to those presenting during the neonatal period [14]. There is also evidence of successful late TGA repairs in some developing countries in children older than 1 year[15]. The survival rate following surgery is as high as 85% in developing countries with the necessary resources, while a survival rate of 97% is reported in developed countries [15]. The lower survival rate in developing countries has been associated with pre-operative comorbid conditions, infections and patients presenting late [15].

Our neonatal patient presented to us on 25th day of life and after a thorough clinical, ECG, Xray chest and color transthoracic echocardiographic evaluation a working diagnosis made was; complete transposition of great arteries Type I, intact ventricular septum with associated moderate sized ostium secundum ASD and large PDA with left to right

shunt, biventricular dilation and normal biventricular systolic function. Despite the presence of ASD and PDA the neonate was extremely critical and on continuous ventilation and oxygen inhalation for maintaining oxygen saturation at > 96%. Because of the above mentioned circumstances, the child was referred to a tertiary care pediatric cardiovascular institute for suitable palliative/corrective surgical procedure.

5. Review of Literature

5.1. Epidemiology

The hearts with atrioventricular concordance and ventriculoarterial discordance represent 5-7% of all congenital heart diseases [16], corresponding to an incidence of 20 to 30 per 100,000 live births. There is a male predominance with a male/female sex ratio that varies, in the literature, from 1.5:1 to 3.2:1 [17-19]. There is a definite association with maternal diabetes mellitus.

5.2. Etiology

The exact aetiology of this disease is still unknown. However, some associated risk factors, namely gestational diabetes mellitus [20, 21], maternal exposure to rodenticides and herbicides [22], and maternal use of antiepileptic drugs [23] have been postulated. Significant advances in the understanding of the underlying genetic mechanisms have been achieved over the last decade. Several mutations have been implicated as the cause of discordant ventriculoarterial connections. The genes involved so far are the growth differentiation factor-1 gene [24], the thyroid hormone receptor-associated protein-2 gene [25], and the gene encoding the cryptic protein [26]. They are localised in different chromosomes and their mutations only explain a small minority of the clinical cases.

5.3. Embryology

Currently, there are two main theories regarding the embryological mechanisms of TGA development:

- De la Cruz proposed the theory that the aortopulmonary septum fails to spiral at the level of the infundibulum thus causing a linear development of the septum and TGA [27].
- The second theory, proposed by Goor and Edwards, suggests that TGA is caused by abnormal resorption or underdevelopment of the subpulmonary conus, with persistence of the subaortic conus [28].

5.4. Classification of transposition of great arteries

Complete TGA consists of 4 types [8]: type 1, with intact ventricular septum; type 2, pulmonary stenosis with intact ventricular septum; type 3, ventricular septal defect, and type 4, ventricular septal defect with pulmonary stenosis. Type 1 is the most common one, followed by type 3.

The primary anatomic subtypes according to Ossa Galvis et al. [29] are : (1) transposition of the great arteries with intact ventricular septum, (2) transposition of the great arteries with ventricular septal defect, (3) transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction, and (4) transposition of the great arteries with ventricular septal defect and pulmonary vascular obstructive disease.

5.5. Great artery relationship in transposition of great arteries

The great artery connections in TGA are also known as "malposition of great arteries". Four main types of the great arteries can be found [30]: (1) "Dextro (D)-malposition" in which the aortic valve is to the right of the pulmonary valve; (2) "Levo (L)-malposition" in which the aortic valve is to the left of the pulmonary valve; (3) "Anterior (A)-malposition" in which the aortic valve is anterior to the pulmonary valve, and (4) "Posterior (P)-malposition" in which the aortic valve is posterior to the pulmonary valve.

The classification used to define the spatial relationships of the great arteries illustrated in the postmortem study by Massoudy et al. [31] included five different variants. These are, in decreasing order of frequency: (1) aorta anterior to and to the right of the pulmonary trunk; (2) aorta directly anterior to the pulmonary artery; (3) side by side, with the aorta on the right and the pulmonary trunk on the left; (4) aorta anterior to and to the left of the pulmonary artery; (5) aorta posterior to and to the right of the pulmonary artery.

5.6. Coronary artery anomalies in transposition of great arteries

In approximately one third of patients with transposition of the great arteries, the coronary artery anatomy is abnormal [29] with a left circumflex coronary arising from the right coronary artery (22%), a single right coronary artery (9.5%), a single left coronary artery (3%), or inverted origin of the coronary arteries (3%) representing the most common variants. Moll et al [32] in their series of 715 patients of isolated and complex TGA in children found a configuration of coronary artery anomalies as depicted in Figure 16.

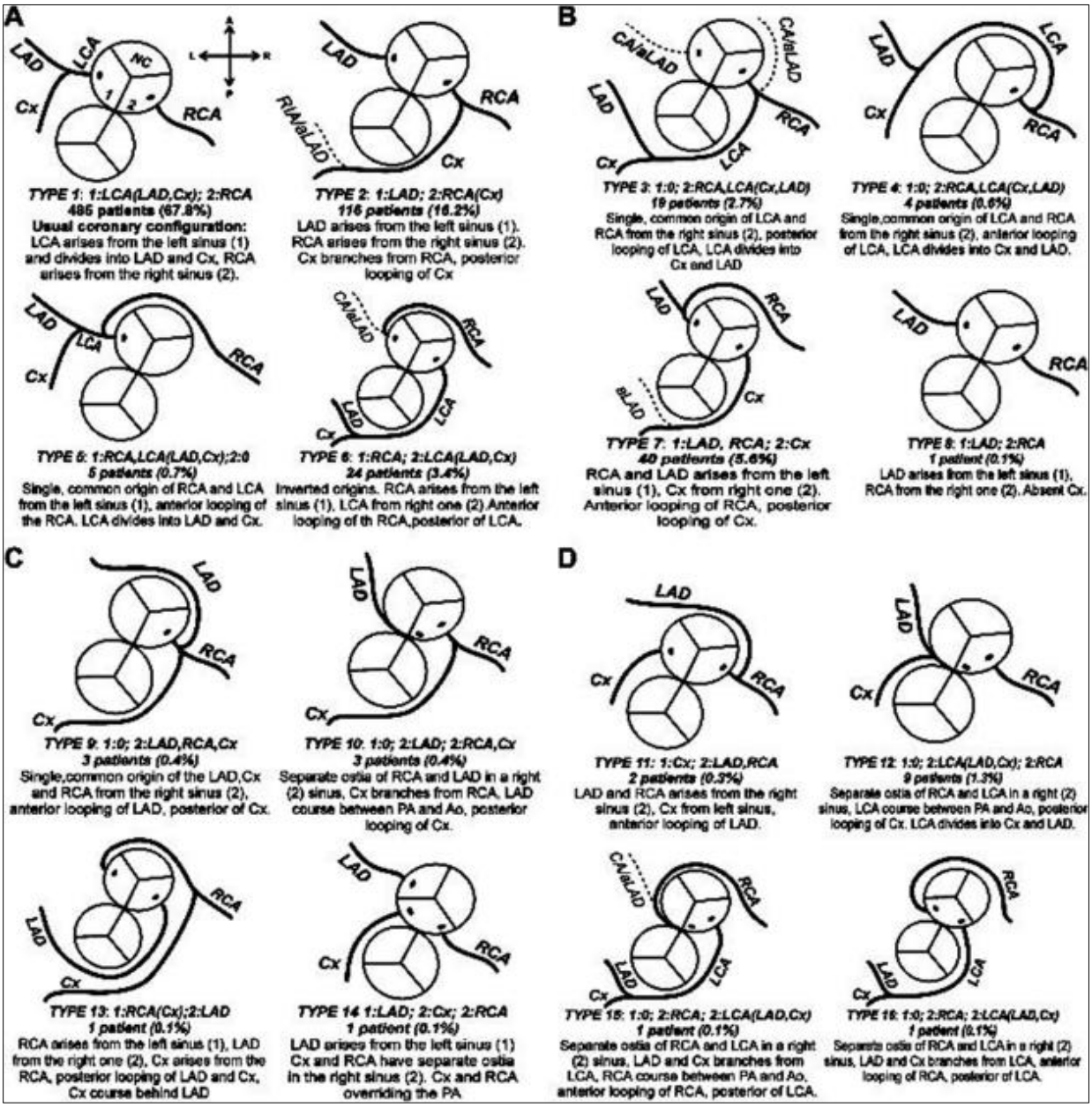

Figure 16 Coronary artery anomalies in TGA. (A-D) Coronary artery configurations in children with transposition of the great arteries. The dotted lines indicate possible coexisting coronary arteries requiring translocation during arterial switch operation. (aLAD=accessory left anterior descending artery; Ao=aorta; CA=coronary artery; Cx=circumflex artery; LAD=left anterior descending artery; LCA=left coronary artery; NC=noncoronary sinus; PA = pulmonary artery; RCA=right coronary artery; RIA=right innominate artery)

Cardiac anomalies associated with transposition of great arteries.

Table 2 Cardiac anomalies associated with TGA [33]

Associated anomalies	
Dextrocardia Persistent left SVC draining in coronary sinus Partial anomalous pulmonary venous drainage in IVC ASD/PFO Appendages juxtaposition	
Tricuspid valve	Hypoplastic Dysplastic Straddling
Mitral valve	Cleft Parachute Arcade Double orifice Straddling
Infundibulum	Subaortic Bilateral
Right outflow obstruction Left outflow obstruction	Deviation of outlet septum Septal hypertrophy Hypertrophy of septoparietal bands Hypertrophy of TSM Insertion of AV valve Deviation of outlet septum
	Septal hypertrophy Septal hypertrophy + fibrous shelf Fibrous diaphragm Anterolateral muscle band Insertion of AV valve Accessory AV tissue
Aortic valve	Bicuspid
Aortic arch	Right arch Coarctation Interruption Retroesophageal subclavian artery
Pulmonary valve	Dysplastic Unicuspid Bicuspid
PDA left/right Bilateral PDA	

ASD, atrial septal defect; AV, atrioventricular; IVC, inferior vena cava; IVS, intact ventricular septum; PDA, patent ductus arteriosus; PFO, patent foramen ovale; SVC, superior vena cava; TGA, transposition of the great arteries; TSM, trabecula septomarginalis; VSD, ventricular septal defect.

5.7. Prognosis

Until mid twentieth century, the treatment of transposition was restricted to few palliative measures and the natural history of the disease with its poor prognosis was an undeniable reality. By that time, the average life expectancy for a newborn with transposition was 0.65 years and the mortality rate at one year was 89.3% [34]. With the advent of newer and improved surgical techniques as well as post operative intensive care, the scenario has completely changed, and very encouraging long-term survival rates almost achieving 90% at 15 years of age have been reported. The potentialities of the current corrective surgery modalities are also underlined by a low 10-year re-intervention rate of (6%) and a corresponding event-free survival of 88% [35].

Nevertheless, recent studies have pointed out to a reduced exercise performance, a compromise in cognitive functioning, and an unfavourable health-related quality of life [35, 36]. Further improvements are therefore necessary and they may be achieved in the future by reinforcing prenatal diagnosis and by establishing strategies to minimise surgical complications.

5.8. Morbidity/mortality

The mortality rate in untreated patients is approximately 30% in the first week, 50% in the first month, and 90% by the end of the first year. Long-term complications are secondary to prolonged cyanosis and include polycythemia and hyperviscosity syndrome. These patients may develop headache, decreased exercise tolerance, and stroke. Thrombocytopenia is common in patients with cyanotic congenital heart disease leading to bleeding complications. With improved diagnostic, medical, and surgical techniques, the overall short-term and midterm survival rate exceeds 90% [37]. Patients with a large ventricular septal defect, a patent ductus arteriosus, or both may have an early predilection for congestive heart failure, as pulmonary vascular resistance falls with increasing age. Heart failure may be mitigated in those patients with left ventricular outflow tract (pulmonary) stenosis.

Arterioplasty in patients with supravalvar pulmonary or pulmonary artery branch stenosis following arterial switch surgery may be an effective and durable management option in the immediate term [37]. In a retrospective study (2004-2013) comprising 223 patients who underwent arterial switch for transposition of the great arteries, 38 patients (16%) developed supravalvar pulmonary stenosis within 12.5 months. The surgical morbidity (eg, main pulmonary artery plasty) was 13%, without hospital or late mortality. At the 41.2 months postsurgical follow-up, all the patients had New York Heart Association (NYHA) functional grade 0 or 1 symptoms [37]. Cardiac catheterization and endovascular stenting of the branch pulmonary arteries is an alternative in older patients versus cardiac surgery.

A retrospective study (1995-2016) that evaluated midterm outcomes in 97 patients with congenitally corrected transposition of the great arteries who underwent different management strategies reported similar transplant-free survival in those who underwent a systemic right ventricle (93%), anatomic repair (86%), and Fontan procedure (100%) (there was a 79% transplant-free survival for pulmonary artery band or shunt) (P= 0.33) [38]. Multivariate analysis demonstrated systemic right ventricular dysfunction as a risk factor for death or transplantation.

A small percentage (approximately 5%) of patients with transposition of the great arteries (and often a ventricular septal defect) develop accelerated pulmonary vascular obstructive disease and progressive cyanosis despite surgical repair or palliation. Long-term survival in this subgroup is particularly poor.

6. Conclusions

TGA is a medical emergency and poses an immediate threat to the newborn in the immediate postnatal life. Early diagnosis and intervention in the immediate postnatal life are paramount to preventing prolonged hypoxemia, acidosis, and death before the definitive surgical correction. Regardless of the missed and unrecognized prenatal diagnosis of TGA and other CHDs, clinicians should have a high index of suspicion when newborns present with clinical manifestations of critical CHD and manage it accordingly to avoid drastic complications.

Mortality from cyanotic congenital heart disease (CCHD) like TGA is still increasing in some developing countries where healthcare facilities are ill-equipped to readily offer either palliative or definitive surgery for this disease. The use of simple, cost-effective and non-invasive methods like routine pulse oximetry screening for all newborns will facilitate early diagnosis and referral. There is a need for continuous advocacy and engagement with governments to improve investment in healthcare delivery through policy changes, universal health coverage and facilitating skills transfer, which will increase access to cardiac surgery and improve outcomes in children with CCHD. Teritary care academic institutions should facilitates teaching and training in complex surgical procedures. Good-quality paediatric cardiac palliative care should be the focus of treatment in resource-limited settings

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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