CAN ISOLATED VENTRICULAR INVERSION SURVIVE WITHOUT ATRIAL OR VENTRICULAR SEPTAL DEFECT?- CASE REPORT

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July 4, 2022

Abstract

Background: Isolated ventricular inversion is characterized by atrioventricular discordance and ventriculoarterial concordance, clinically akin to transposition of the great arteries. [1] In the absence of a significant atrial or ventricular septal defect, profound cyanosis is expected at birth. **Case Report:** A 5-month-old infant with isolated ventricular inversion presented with mild cyanosis. The left sided tricuspid valve straddled the interventricular septum with a closed interventricular communication, a type of "Double Outlet Left Atrium with three atrioventricular valves", which provided the necessary 'left to right' shunt while severe regurgitation through the straddling segment and a patent ductus arteriosus provided the effective pulmonary blood flow. [2] **Result**: The infant underwent atrial septectomy, closure of the leaky accessory orifice and a Damus-Kaye-Stansel anastomosis and a bidirectional Glen operation. **Conclusion:** A unique natural mechanism permitting survival in a child with transposition physiology is described. The Damus procedure preserved tricuspid valve, right ventricle and pulmonary valve function.

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(Running title: Inverted ventricles, straddling tricuspid)

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Word count:

Abstract: 149 words

Manuscript (excluding title page, abstract, references and legend): 1469 words

The corresponding author takes full responsibility for the veracity of the facts presented.

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Declaration

1. There are no financial or commercial disclosures
2. The authors have not received any financial support and declare no potential conflict of interests with respect to research, authorship, and/or publication of this article. This is an original work and is not under consideration for publication anywhere else. It was submitted as a poster for the Institutional Research Day proceedings of the National Heart Center, Royal Hospital Muscat. Oman held on 18th December 2021.
3. Manuscript has been approved by the institutional review board Ref No.SRC#CR28/2021 and informed consent has been taken from the parent of the patient.

Keywords:

Congenital heart disease; Single ventricle, isolated ventricular inversion, straddling tricuspid valve without ventricular septal defect, double outlet left atrium, Damus Kaye Stansel. Heart ventricles/ abnormalities

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Introduction:

'Isolated ventricular inversion in situs solitus' with atrioventricular discordance and ventriculoarterial concordance is characterized by a morphological left atrium connected to the right ventricle through a tricuspid valve and the right atrium connected to the left ventricle through the mitral valve. The great arteries arise from their respective ventricles; thus, the physiology is akin to transposition of the great arteries, and profound cyanosis is expected after birth in the absence of significant intracardiac shunts.[1]

Straddling of an atrioventricular valve is due to a deviated interventricular septum towards the side of the straddling valve and is accompanied by varying degrees of inflow hypoplasia of the ventricle on the deviated side. In such cases, if there is no atrial or ventricular septal defect, it leads to a form of 'double outlet left atrium' [DOLA] with three atrioventricular valves (ventricular septal malalignment type).[2]

Case Report:

A five-month-old female child [weight 3.7Kg, height 59cm] with mild cyanosis was referred for management of complex congenital heart disease. Following an antenatal diagnosis of isolated ventricular inversion and severe intrauterine growth retardation, she was born by normal vaginal delivery at 36 weeks, with a birth weight of 1.84Kg. Her two older siblings were fully saturated with no cardiac diagnosis and the patient was asymptomatic and gaining weight normally. Her pulse oximeter reading was 90% in room air. She was not dysmorphic and her clinical examination revealed a Grade 3 pansystolic murmur in the left 5^{th} interspace in the anterior axillary line radiating to the axilla. Due to low birth weight and mild cyanosis, she was managed conservatively until her current presentation.

The transthoracic echocardiogram (TTE) revealed situs solitus, levocardia, atrioventricular discordance and ventriculoarterial concordance with normally related great arteries {S, L, D}. The left sided tricuspid valve straddled across a mal-aligned interventricular septum with a closed ventricular septal defect component **(Figure 1A, B)** . There was severe regurgitation from the accessory orifice, with blood flow traveling predominantly from the morphological (right sided) left ventricle into the left atrium **(Figure 1B)** . There was a 2mm patent foramen ovale with left to right shunting **(Figure 1C)** and a 2.5mm patent ductus arteriosus with continuous flow into the pulmonary artery. The right ventricle was relatively hypoplastic and the straddling orifice measured 7.6 mm while the major orifice was 9.5 mm**(Figure 1D).** Presence of Aortomitral continuity and a nonobstructive subpulmonary conus were noted.

The electrocardiogram revealed sinus rhythm with a heart rate of 159 beats/minute with a frontal QRS axis of 97 degrees, a P wave axis of 70 degrees, normal septal activation, clockwise looping (q waves in leads II, III and aVF), left atrial enlargement and features of biventricular hypertrophy **(Figure 2A).**

A cardiac catheterization study revealed increased pulmonary blood flow with a Qp:Qs ratio of 3.9 with the PVRI measuring 0.74 Wood units. The main pulmonary artery pressure was 48/19 mm Hg with a mean of 30mmHg and the aortic pressure was 73/34 with a mean pressure of 50 mmHg. The left atrial pressure was 15mmHg and the right atrial pressure was 7mmHg.

The patient was taken for elective surgery **(Video 1).** Under mildly hypothermic cardiopulmonary bypass with cardioplegic arrest, the right atrium was opened and the atrial septum was widely excised. The AV valve connected to the right atrium was the mitral valve and led to the left ventricle and thence to the aorta **(Figure 2B, C).** The left atrium led to the two orifices, the posterior of the two led to the hypoplastic right ventricle while the anterior 'accessory orifice' led to the morphological left ventricle. There was no interventricular communication. The accessory orifice was closed with pledgetted 5-0 polypropylene sutures and competence of the valve and adequacy of closure of the orifice were confirmed by saline injection into the respective ventricles. The pulmonary artery was transected and the distal end closed with a bovine pericardial patch. The ascending aorta was opened obliquely and an end-to-side anastomosis was created between the pulmonary artery and the aorta (Damus-Kaye-Stansel shunt [DKS]) and the anastomosis was augmented anteriorly with a patch of bovine pericardium. The bidirectional Glenn anastomosis was performed after release of the aortic cross-clamp. The child was weaned off cardiopulmonary bypass uneventfully in sinus rhythm with good hemodynamics and a Glenn pressure of 12mmHg, a common atrial pressure of 7mmHg and an oxygen saturation of 85% on 0.5 FiO2. At 10 months follow-up she weighed 8.3Kgs and was saturating 92% on room air. Echocardiography revealed no leak or flow across the surgically closed accessory orifice of the straddling tricuspid valve, laminar flow across the other two atrioventricular connections with no regurgitation and laminar flows through the Glenn shunt and the DKS with trivial aortic regurgitation **(Figure 3A, B and C)** .

Comment:

The term 'isolated' in 'isolated ventricular inversion' (IVI) was originally used to differentiate it from other forms of ventriculoarterial connections including discordant [double discordance – congenitally corrected transposition of great arteries, CCTGA], double outlet from a morphologically right ventricle and single outlet in case of pulmonary atresia. [1]This case would belong to the 'type A' isolated ventricular inversion {S, L, D} in the classification proposed by Konstantinov et al.[3] Situs solitus, 'normally' related great arteries

and presence of aorto-mitral continuity distinguish it from the other types.

A straddling tricuspid valve without VSD is extremely rare and one with AV discordance, rarer still.[4,5] The mechanism of closure of the inter-ventricular communication has been likened to that in 'atrioventricular septal defect without ventricular septal defect' with a bar of tissue attaching the leaflets to the crest of the interventricular septum.[4,6]

Straddling of the tricuspid valve is the result of malalignment of the ventricular septum and results in varying degrees of hypoplasia of the right ventricular inflow ranging from mild (biventricular) to the extreme – the double inlet left ventricle – end of the spectrum.[5] However, the degree of override is usually between 20 and 45% (which, in the current patient, was about 40%).[5]

The conduction system in isolated ventricular inversion is not completely understood, and is of most relevance while closing a VSD, which is not of concern in this case. The lack of extreme wedging of the pulmonary artery in IVI precludes such conduction anomalies as in CCTGA. Straddling, in fact, may play a more important role: the straddling mitral valve usually straddles the anterior part of the interventricular septum while the tricuspid usually straddles the posterior part (due to which the interventricular septum does not reach the crux of the heart). However, a straddling *left sided* tricuspid valve possibly overrides the *anterior* interventricular septum and the conduction system is formed normally as suggested by the normal conduction axis in this patient.

Straddling without significant inter-atrial or inter-ventricular communication, in this case, resulted in 'double outlet left atrium with three AV valves due to ventricular septal malalignment'.[2,6]

AV discordance and VA concordance entails transposition physiology and the parallel systemic and pulmonary circulations require bidirectional shunting for survival. The unique physiology in this patient can be summarized as follows:

First, the interatrial communication was tiny. Second, the connection of a volume overloaded left atrium to the right sided left ventricle, through the accessory orifice, ensured systemic oxygenation. Third, the regurgitant jet from the left ventricle to left atrium through the accessory orifice, (which has been also alluded to as a third variant of Gerbode defect, albeit in ventriculoarterial discordance) along-with the fourth, the large patent ductus arteriosus, ensured adequate effective pulmonary blood flow (Q_{ep}).[6]The latter two resulted in left atrial volume overload as well. The net result of all these factors was an Oxygen saturation of 90% on room air and an elevated pulmonary artery pressure of 48/19 (mean 30) mmHg at a PVRI of 0.74 Wood units/sqm.

Surgical management was guided by the right ventricle which was deemed relatively hypoplastic and a bidirectional Glenn was decided by the team. Although the right ventricular inflow was hypoplastic because of the straddling, the rest of the right ventricle was felt to be reasonably well formed. Also considered was the fact that there was no VSD and the pulmonary valve was well formed. In order to avoid closure of the pulmonary valve altogether and to retain right ventricular and tricuspid valve function, a Damus anastomosis was performed in addition to the bidirectional Glen (along with atrial septectomy and closure of the accessory orifice. Retaining the tricuspid valve had additional benefits of an additional drainage for the left atrium should the atrial communication become restrictive, and the possibility of an atrial switch procedure (a hemi-Mustard) or at least a 'biventricular' Fontan in future.

Conclusion:

An infant with isolated ventricular inversion without significant atrial or ventricular septal defects survived without intervention due to a unique natural mechanism – a straddling left sided tricuspid valve with a leaky accessory orifice.

Author contribution:

1. Pranav Subbaraya Kandachar: Concept and design, preparation of manuscript.
2. Eapen Thomas: Concept, preparation of manuscript, Echocardiography images and interpretation.

3. Areej Al Maskary: Echocardiography – image processing, Pre and postoperative data collection.
4. Andrew Campbell: Operating surgeon, Concept and preparation of manuscript.

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6. Lopes BS, Coates L, Anderson RH: Commentary: Shunting between the left ventricle and right atrium can be produced by straddling tricuspid valve with dual orifices. Semin Thorac Cardiovasc Surg. 2020;32(1):143-144.**Figure legend**Figure 1. Pre-operative transthoracic and trans-esophageal echocardiography: transthoracic apical four-chamber view, 2D and color Doppler, depicting the atrioventricular discordance with straddling left sided tricuspid valve (arrows pointing to the three orifices, the mitral valve, the accessory orifice connecting the left atrium and left ventricle and the main orifice of the tricuspid valve) and hypoplastic morphologic right ventricle in diastole **(A)** and systole **(B)** demonstrating the antegrade flow and the regurgitation through the accessory orifice respectively. A distended left atrium and the 2mm patent foramen ovale **(C)** and the two orifices of the straddling tricuspid valve as seen on subcostal 4-chamber view **(D).** *LA= left atrium, LV= left ventricle, PFO= patent foramen ovale, RA=right atrium, RV= right ventricle.* Figure 2. Electrocardiogram **(A)** showing a normal conduction axis and intraoperative photograph showing the connection (arrow) from the right atrium through a right sided (left) ventricle to the aorta (**B**), and the anterior leaflet of the mitral valve (**C**). *LV= left ventricle, MV=mitral valve.* Figure 3. Postoperative transthoracic echocardiogram showing absence of flow through the surgically obliterated accessory orifice (**A**) and laminar flow through the Damus (**B**) and the bidirectional Glen (**C**). *Ao = aorta, LA= left atrium, LV= left ventricle, PA= pulmonary artery, RA=right atrium, RPA= right pulmonary artery, RV= right ventricle. SVC= superior vena cava.***Supplementary material**

Videoclip legend

Videoclip-1. Surgical video of closure of the leaky accessory orifice followed by the Damus Kaye Stansel anastomosis and bidirectional Glen procedure.