

Left isomerism with bilateral superior vena cava, interrupted inferior vena cava and tetralogy of Fallot

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ABSTRACT

The association of left isomerism with tetralogy of Fallot (TOF) is rare and reported as scattered case reports. Complex congenital cardiac and noncardiac malformations are known to be associated with left isomerism, but right-sided obstructive lesions are rare. We present our experience with a case of left isomerism with bilateral superior vena cava, interrupted inferior vena cava, and TOF managed by atrial septation and intracardiac repair.

Keywords: Atrial septation, bilateral superior vena cava, interrupted inferior vena cava, left isomerism, tetralogy of Fallot

INTRODUCTION

The association of left isomerism with tetralogy of Fallot (TOF) is rare and reported as scattered case reports.^[1-3] An autopsy study found isomerism in 3.1% of patients with right isomerism in 1.9% and left isomerism in 1.2%.^[4] Complex cardiac and noncardiac malformations are known to be associated with isomerism, but right-sided obstructive lesions are more common with right isomerism.^[1,2,5] We present our experience with one such case of left isomerism with TOF, managed successfully by biventricular (BV) repair.

CASE SUMMARY

A 1-year-old child presented with blue discoloration of skin and delayed milestones. She had a weight of 8.4 kg and room air saturation of 86%. On echocardiography, she had isomerism of left atrial appendages with L-looped ventricles, with right-sided atrium draining into the left ventricle (LV) and left-sided atrium draining into the right ventricle (RV) and ventriculo-arterial concordance. She had bilateral superior vena cava (SVC) with absent

innominate vein and left SVC (LSVC) draining into the right-sided atrium via the coronary sinus (CS). All four pulmonary veins (PVs) were also draining into the right-sided atrium. The atrial septum was absent with only small septal tissue remnant in the superior part. She had TOF morphology with aortic override of 50%, aorto-mitral continuity, large malaligned perimembranous ventricular septal defect (VSD) routable to LV, severe pulmonary stenosis with a gradient of 60 mmHg, and confluent and good-sized pulmonary arteries (PAs). The findings were confirmed by computed tomography (CT) [Figures 1-3]. CT also revealed an interrupted inferior vena cava (IVC) with large azygous vein draining into the right-sided SVC and hepatic veins draining directly into the right-sided atrium. Left bronchial isomerism (bilateral hyperarterial bronchi) with normal abdominal situs (absence of polysplenia) was also noted.

The patient was planned for intracardiac correction with atrial septation. Cardiopulmonary bypass was instituted by aortic, bilateral SVC and hepatic venous cannulation.

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Del Nido cardioplegia was used with core cooling to 28° for myocardial protection. Findings of pectinate muscles not reaching to a crux in either of atria and venous drainage as described previously were confirmed intraoperatively. VSD was closed with Dacron patch via RV incision and infundibular resection was done with transannular patch augmentation of pulmonary annulus with autologous nonfixed pericardial patch. CS was cut back to avoid the obstruction to LSVC flow. After CS cutback, the opening lay laterally to the orifice of PVs so that it lay outside the proposed patch. Superior and inferior PV orifices were widely separated so there was no way of routing all of the systemic venous return to tricuspid valve (TV) without obstructing PV inflow [Figure 4]; therefore, PVs were routed to the mitral valve using nonfixed autologous pericardial patch, making sure that systemic venous return to TV was not obstructed [Figure 5].

The postoperative period was uneventful, with the child being shifted to the intensive care unit with adrenaline support of 0.05 mcg/kg/min and milrinone of 0.7 mcg/kg/min. She was extubated on postoperative day 2 and discharged in asymptomatic condition on postoperative day 8. Echocardiography at 3 months showed no residual VSD, no RV/LV outflow obstruction, no gradient across systemic, or PV return with good BV function.

DISCUSSION

In a recent autopsy analysis,^[6] left isomerism was found to be frequently associated with absent atrial septum, bilateral SVC, interrupted IVC, atrio-VSD, and left ventricular outflow tract obstruction. Right isomerism, on the other hand was found to be commonly associated with absent CS, ventriculo-arterial discordant connection,

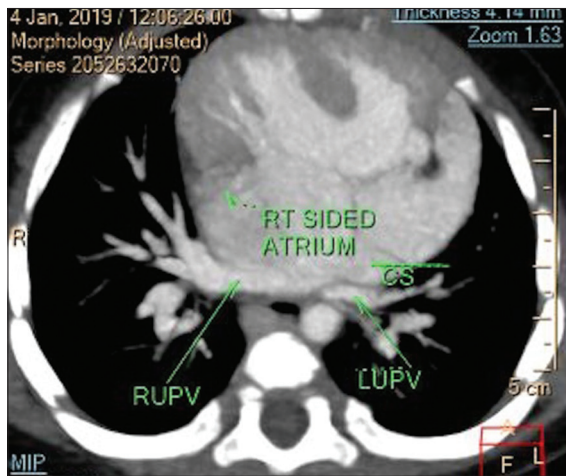


Figure 1: Preoperative computed tomography showing right upper and left upper pulmonary veins and coronary sinus draining into the right-sided atrium. RUPV: Right upper pulmonary vein, LUPV: Left upper pulmonary vein, CS: Coronary sinus

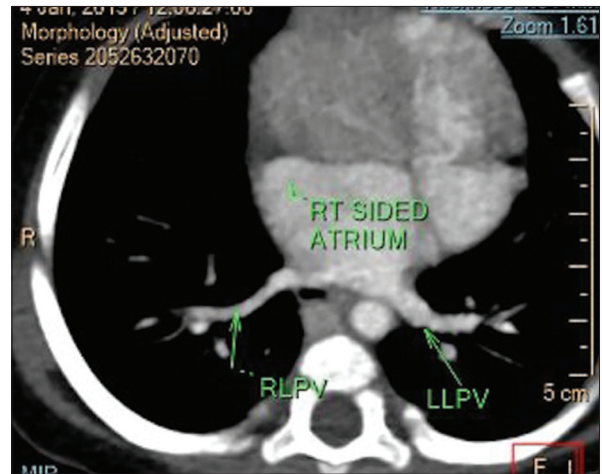


Figure 2: Preoperative computed tomography showing right lower and left lower pulmonary veins draining into right-sided atrium. RLPV: Right lower pulmonary vein, LLPV: Left lower pulmonary vein

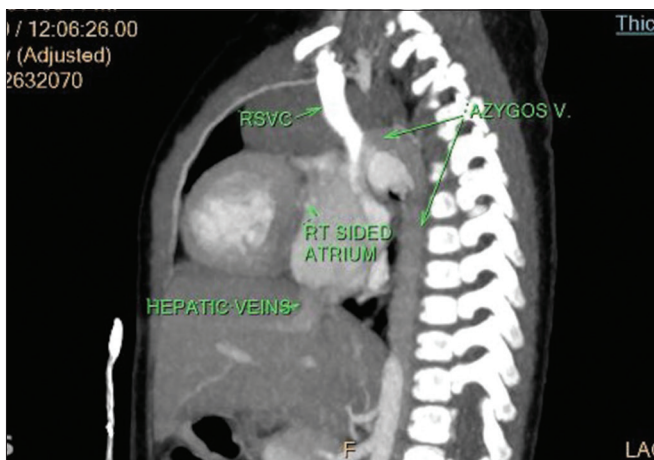


Figure 3: Preoperative computed tomography showing right superior vena cava along with azygos vein and hepatic vein draining into the right-sided atrium. RSVC: Right superior vena cava

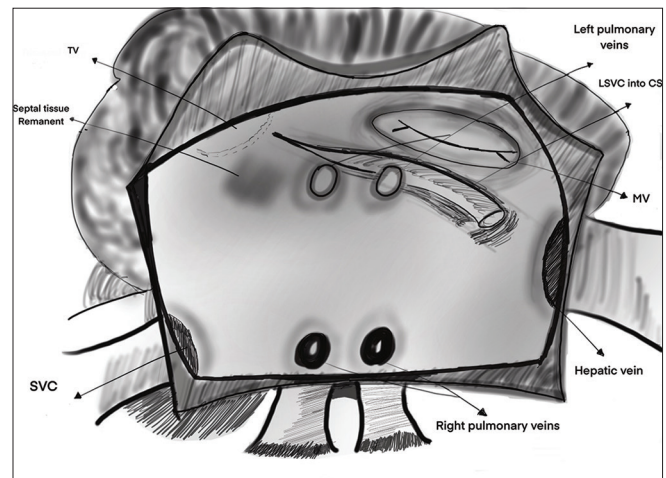


Figure 4: Diagrammatic representation of anatomy as seen intraoperatively after opening right-sided atrium. SVC: Superior vena cava, TV: Tricuspid valve, MV: Mitral valve, LSVC: Left superior vena cava, CS: Coronary sinus

total anomalous pulmonary venous connection (TAPVC), and right ventricular outflow obstruction. Atrial configuration correlates much more with bronchial configuration than asplenia and polysplenia.^[6] Therefore, recognition of isomerism should alert the surgeon to look for associated anomalies so as to plan proper surgical management.

Historically, left isomerism has been shown to carry a dismal prognosis with more than two-third of patients dying within the 1st year.^[7] However, recent studies have shown long-term survival reaching 87.8% at 40 years, although it has been documented to be an independent predictor for pacemaker implantation.^[8] The patients who underwent BV repair had been shown to have a survival of 80% at 1 year, 71% at 5 years, 66% at 10 years, and 63% after 15 years. Those who underwent single ventricle repair had survival of 73% at 1 year, 61% at 4 years, 53% at 10 years, and 48% after 15 years.^[9] Another study reported survival of 88.1% and 83.3% after 1 and 5 years of BV repair, respectively.^[10]

Left isomerism with TOF is a rare combination, and surgical treatment of such anomalies has been reported sporadically. In various series of left isomerism,^[1-3,11,12] TOF has been reported as an associated anomaly only

in four cases [Table 1]. Management depends on their anatomy and associated anomalies.^[11]

1. Palliative procedure: It may be required in the form of systemic-PA shunt in severe pulmonary obstruction, PA banding in univentricular connection with unobstructed pulmonary blood flow and Norwood in LV hypoplasia or severe subaortic stenosis
2. BV repair: This can be done either as a primary procedure or as a staged procedure depending on anatomy. However, because of frequent association of several malformations, it is usually not possible [Table 1]
3. Orthoterminal repair: The modified Fontan procedure includes right atrium-PA anastomosis after patch closure of atrial septal defect/atrial partition or both and ventricular exclusion.^[13] The presence of associated anomalies makes this a very difficult option and has been accompanied by high surgical mortality.^[11]

Atrial septation in patients with common atrium and anomalies of the venous connections is difficult and should ideally result in correct rerouting and two suitable atrial chambers. For good atrial contractile function, the ratio of the muscular wall to the patch surface area should be maintained.^[11] Methods of atrial septation include using a straight patch when systemic and PV drain into either side of the atrium independently; tailored baffle; and a mustard-type atrial switch when PV drained into the functional right atrium or more than two systemic veins drained into the functional left atrium.

As there are no right atrial appendages in left isomerism, atrioventricular (AV) node is not in normal position and when present, it is usually hypoplastic and location depends on ventricular topology. In right handed, the AV node is present in a regular position, with a posteroinferior penetrating bundle and in left handed, AV node is anterior and the conduction system is found as in congenitally corrected transposition.^[5]

There are no clear criteria to determine suitability for BV repair given the extent of the anatomic variability. BV repair should be considered in the presence of septable venoatrial connections along with two ventricles of

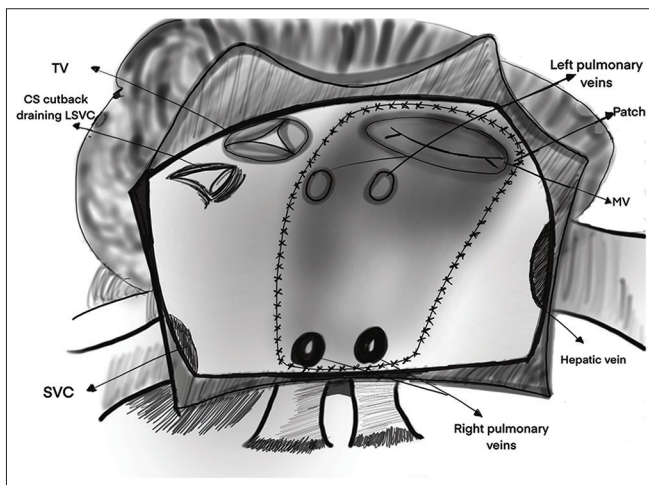


Figure 5: Diagrammatic representation of anatomy after atrial septation. SVC: Superior vena cava, CS: Coronary sinus, LSVC: Left superior vena cava, MV: Mitral valve, TV: Tricuspid valve

Table 1: Incidence of left isomerism, associated lesions, and their management as found in literature

Author	Left isomerism/total isomerism	Bilateral SVC	IVC obstruction	AVSD	TAPVC/PAPVC	TOF	Initial palliative repair	BV repair	UV repair
Hirooka et al. ^[1]	32/93	17	21	18	3	1	8	14	18
Gojo et al. ^[2]	1	1	1	-	-	1	-	1	-
Lim et al. ^[3]	66/91*	39	60	36	6	3	43	66	280/371
Marcelletti et al. ^[11]	5/12	0	3	4	3	0	3	1	1
Sinzobahamvya et al. ^[12]	23/41	3	13	11	6	0	4	17	5

*Data available for 91 patients who underwent biventricular repair out of total 371 patients who had isomerism (right and left). SVC: Superior vena cava, IVC: Inferior vena cava, AVSD: Atrioventricular septal defect, TAPVC: Total anomalous pulmonary venous connection, PAPVC: Partial anomalous pulmonary venous connection, TOF: Tetralogy of fallot, BV: Biventricular, UV: Univentricular

adequate volume and function. Preoperative delineation of exact cardiac morphology and associated anomalies is important to decide the cardiopulmonary bypass strategy and repair technique and improve the chance of BV repair.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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