



Girish Gowda SL^{1*}, Seetharama Bhat PS¹, Jayaranganath Mahimarangaiah² and Cholenahally NM²

¹Department of cardiothoracic and vascular surgery, Sri Jayadeva Institute of Cardiovascular Sciences and Research, India

²Department of cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, India

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***Corresponding author:** Girish Gowda SL, Department of cardiothoracic and vascular surgery, Sri Jayadeva institute of cardiovascular sciences and research, Bangalore, Karnataka, India, Tel: 09449046932; E-mail-gowda1212@yahoo.co.in

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Case Report

Double outlet right ventricle, total anomalous venous return, total anomalous hepatic venous drainage and supra mitral ring in a child with Ivemark's syndrome

Abstract

Double outlet right ventricle, total anomalous venous return, total anomalous hepatic venous drainage and supra mitral ring with Ivemark syndrome is an unusual combination of cardiac malformations. These complexities of congenital anomalies can pose problem in preoperative diagnosis and surgical management. We present investigation and surgical management of a three month old child with these mentioned rare combination of anomalies and a brief review of literature.

Introduction

Double outlet right ventricle, total anomalous venous return, total anomalous hepatic venous drainage and supra mitral ring in a child with Ivemark syndrome is an unusual combination of cardiac malformations which has not been reported till now in the world literature. These complex combinations of cardiac malformations can be surgically corrected.

Case Report

A three month old female child was referred to our institution with a history of cyanosis since birth for evaluation and treatment. There was mild cyanosis present with a grade III/IV systolic murmur along the left and right sternal border, heard more over the third left interspace, which radiated well into the second right interspace. All peripheral pulsations were well felt. Chest and abdomen x-ray showed bilateral eparterial bronchi, symmetrical transverse liver and ipsilateral side of liver and stomach (Figure 1). ECHO showed findings of DORV, cardiac type of total anomalous venous return, total anomalous hepatic venous drainage into RA and supra mitral ring with normal drainage of inferior vena cava into RA (Figure 2). With the above mentioned diagnosis patient was taken for surgery. Both atrial appendages were morphologically identical to right atrium and child had LSVC. All three venacavas and anomalous hepatic vein were cannulated for cardiopulmonary bypass (Figure 3) (video). On opening the RA the diagnosis of cardiac total anomalous venous return, DORV and supra mitral ring

were confirmed. Resection of supra mitral ring, rerouting of the pulmonary veins to the left atrium and intra ventricular tunneling of VSD to aorta was performed. Child was extubated on the second postoperative day.

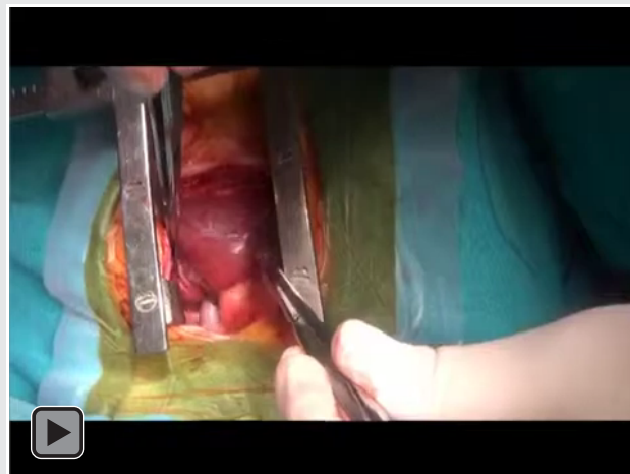
Discussion

Ivemark's syndrome is a rare congenital anomaly which is inherited in an autosomal recessive fashion and is estimated



Figure 1: Chest and abdomen x-ray shows bilateral eparterial bronchi, symmetrical transverse liver and ipsilateral side of liver and stomach showing situs ambiguous features (EB- eparterial bronchi, L- liver, S- stomach).

to occur in 1 in 10,000 to 40,000 live births [1,2]. Ivemark's syndrome consists of situs ambiguus with intrathoracic and abdominal visceral heterotaxy. Intracardiac defects associated with Ivemark syndrome includes common atrium ventricular septal defect with a common atrioventricular valve, common atrium, interruption of the inferior vena cava with azygos venous continuation to the atrium, direct hepatic venous drainage to the atrium, DORV, bilateral superior venacava and various other cardiac anomalies [3]. With extensive searching of world literature through PubMed no case with Ivemark's syndrome associated with supramitral ring were found. Hence this is the first reported case of this combination of anomalies. In some cases of right isomerism, right and left sided atrial origin of the P wave is present at different times of ECG reflecting the activity of bilateral sinus nodes [4]. Our case also had this mentioned ECG finding. Atrioventricular block is very uncommon in right isomerism whereas varying degrees of atrioventricular block are common in left isomerism.



Video : Operative video showing superior venacava, inferior venacava, left superior vena cava, and anomalous hepatic vein draining in to right atrium. Also shows venous cannulation of all three venacavas and anomalous hepatic vein cannulated for cardiopulmonary.

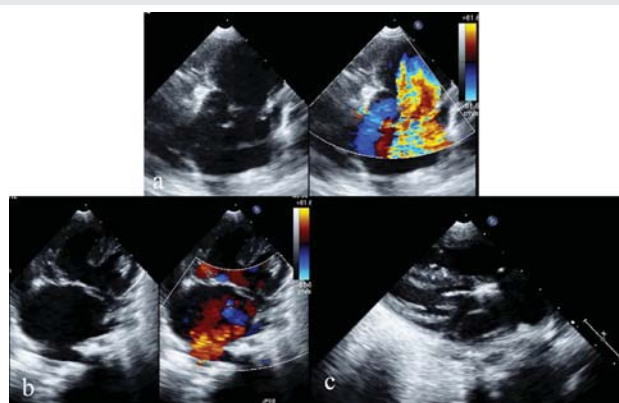


Figure 2: Colour image is available in the online version of the article.

a) Modified apical view showing aorta and pulmonary artery arising from right ventricle. There is turbulent flow across the pulmonary valve
b) Apical four chamber view showing the supramitral ring.
c) Parasternal long axis view showing the supramitral ring.
(RV- Right ventricle, AO- Aorta, PA- Pulmonary artery, SMR- Supramitral ring, AML- Anterior mitral leaflet, PML- Posterior mitral leaflet).



Figure 3: Colour image is available in the online version of the article. All three venacavas and anomalous hepatic vein cannulated for cardiopulmonary bypass. (SVC- superior venacava, IVC- inferior venacava, LSVC- left superior vena cava, TAHVD- total anomalous hepatic venous drainage).

Patients with right isomerism may occasionally present with serious extra cardiac anomalies [5]. Our reported case had no extra cardiac anomalies. The IVC drains into the heart through azygos vein more often in patients with left atrial isomerism whereas in right atrial isomerism the hepatic veins drains into the right atrium with an interruption of the IVC or without interruption of IVC [6,7]. Cardiac anomalies are more frequent in right atrial isomerism and lead to a mortality of 80% to 95% in the first year of life without their prior surgical correction [8]. In the absence of cyanosis or cardiac anomalies, patients with asplenia are at greater risk of dying from sepsis than from congenital heart disease. Our reported case had extremely rare combination of cardiac anomalies which is first in the word literature to be reported.

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