Partial anomalous hepatic venous drainage into left-sided atrium with right isomerism: A case report with review of literature

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Abstract

Right isomerism, also known as Ivemark syndrome, is an unusual degree of symmetry of some of the abdominothoracic viscera reflecting bilateral right-sidedness. We report an exceedingly rare occurrence of anomalous drainage of the left hepatic vein to the left-sided atrium in a patient of right isomerism. With this case report, we further endorse that the isomerism of atrium might extend beyond the appendages, a view that has been long dismissed by the existing literature.

Key words: Anomalous; drainage; hepatic; left atrium; venous

Introduction

Hetrotaxy syndromes (HS) constitute disorders involving abnormal lateralization and arrangement of the abdominal viscera, thoracic organs, and atria of the heart along right-left axis of the body. HS is categorized into two major recognized groups, namely, the right and left atrial isomerism because atrial isomerism forms the major component of these disorders. However, the current literature strongly favors the use of the term atrial appendage isomerism because it has been shown that, based on morphological assessment, the degree of isomerism of atrium has been unequivocally present only in atrial appendages. Various cardiovascular and noncardiovascular complex disorders are often associated with these syndromes, which are in general more severe in right isomerism.

Right isomerism reflects bilateral right-sidedness of abdominothoracic organs and is commonly associated with asplenia. The incidence of this disease is approximately 1/10000–1/40000 births. We report an exceedingly rare occurrence of anomalous drainage of left hepatic vein in a patient of right isomerism.

Case Report

A 3-month-old infant presented to the pediatric clinic of our institute with chief complaints of respiratory distress since birth. There was no history of bluish discoloration of the skin, fever, or cough. Transthoracic echocardiography revealed mild enlargement of the right atrium and right ventricle with double outlet right ventricle (DORV),
atrophic ventricular canal defect, and partial anomalous pulmonary venous return.

Computed tomography angiography (CTA) of the thorax was planned to delineate the vascular anatomy prior to surgery. CTA was performed on a 64-slice multidetector computed tomography (MDCT) scanner (Toshiba Aquilion V3.30ER001 Toshiba America Medical Systems) and was evaluated on Aquarius Intuition workstation (Terarecon version 3.4). CTA showed situs ambiguous in the form of bilateral trilobar bronchi, with D-loop [Figure 1A].

Figure 1(A-E): (A) MDCT coronal reconstructed image showing bilateral trilobed lungs. (B) MDCT coronal maximum intensity projection (MIP) image showing hepatic veins draining into the left atrium (black arrow). In addition, anomalous drainage of the pulmonary veins from the left upper lobe draining into the persistent vertical vein is also seen (white arrow). (C) MDCT volume rendered (VR) image showing the common channel formed by left and right-side pulmonary veins (black arrow). Also seen are the hepatic veins draining into the left atrium (white arrow). (D) MDCT VR image showing the origin of the aorta and pulmonary artery from the right ventricle suggestive of double outlet right ventricle.
Abnormal systemic venous return has been previously described with the so-called “congenital asplenia syndrome.” In a study by Hashmi et al. among 91 patients of right HS, various anomalous venoatrial discordance were identified, predominantly in the form of double SVC, single left SVC, bilateral SVC without a connecting innominate vein, IVC draining into left-sided atrium, total and partial anomalous hepatic venous drainage to the right-sided atrium. However, direct connection of a hepatic vein to the left-sided atrium, as in the index case, is extremely uncommon.

Only few cases of abnormal congenital hepatic venous return to the left atrium has been described in the literature. Even minority of these are, however, in the clinical setting of HS. Tofeig et al. described a case of right isomerism, in which left-sided shunt of hepatic veins developed as a consequence of surgical procedure. Giamberti et al. described two patients with right isomerism, which was corrected with fenestrated Fontan’s procedure, in which right to left shunts developed from the IVC through the liver to a hepatic vein draining to left atrium. Rubino et al., in postmortem study of the preserver hearts, also described drainage of the hepatic veins to the left-sided atrium in the right isomerism. They also observed that the separately draining IVC, and hepatic veins are interconnected via a large venous sinus in many of these. In another retrospective analysis of postmortem specimens, Uemura et al. noted bilateral atrial drainage of the right and left hepatic venous channels in the setting of HS, although he found it to be more common in left isomerism. To the best of our knowledge, the index case is first to report congenital bilateral hepatic venous returns based on imaging of the living patient.

Various other anomalies that have been reported in association of this anomaly include the presence of intrapulmonary shunts, interrupted IVC, ventriculoarterial discordance (Tetralogy of Fallot and DORV), anomalous pulmonary venous drainage, right-sided aortic arch, patent foramen ovale, bicuspid pulmonary valve, and infundibular pulmonic stenosis. Development of the intrapulmonary shunts in these patients is postulated to be caused due to the deprivation of an unknown hepatic factor as a result of exclusion of hepatic venous blood from the lung, as often seen as the complications of Glenn shunt and Fontan procedure. The absence of intrapulmonary shunts in the index case can be explained by the fact that only left-sided veins were anomalous, and right and middle hepatic vein were draining into the right atrium, and thus entering pulmonary circulation. Furthermore, the presence of atrioventricular septal defect allows the blood mixture at the atrial level, which would be sufficient to provide hepatic venous flow to the lungs.

The index case has several striking features:

- Right-to-left shunt due to partial anomalous hepatic venous drainage to the left atrium
- Left-to-right shunt due to partial anomalous pulmonary venous return to right atrium
- A complete atrioventricular canal defect which might be allowing unrestricted interatrial and interventricular communication
- DORV which further hypothetically brings about the averaging of the saturations of pulmonary and systemic flow

Presence of both the left-to-right and right-to-left shunt with a large intercommunicating cardiac AVCD raises a dilemma as to what should be the appropriate treatment and whether or not a surgical intervention is necessary. With bidirectional shunt anomalies, it is possible to have an equal ratio of blood flow to pulmonary and systemic circulation. The unusual occurrence of bidirectional isolated shunts in this case mandates an embryological and etiological explanation. Persistence of left vitelline vein is postulated to provide connection between the liver and left atrium. It is again imperative to note that isolated left-sided anomalous hepatic venous return to the left atrium has never been reported, which might explain and reflect in its unique association with HS.

Our case is also remarkable by the fact that it raises important question contrary to the long held view that atrial isomerism is not a real thing and should be more aptly described as atrial appendage isomerism. Separate hepatic venous drainage of the left and right-sided hepatic veins into the right and left-sided atrium invites explanations that whether the degree of isomerism extends beyond the appendages.

**Conclusion**

To conclude, this case report augments the existing sparse literature regarding this anomaly and describes its new associations. To the best of our knowledge, this is the first reported case of bilateral drainage of hepatic veins in the clinical setting of HS demonstrated by MDCT angiography, which is a very effective investigation in the diagnosis of complex congenital heart disease over direct invasive procedures.

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**Conflicts of interest**
There are no conflicts of interest.

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