Total anomalous systemic venous drainage to the left atrium: An entity reviewed and investigated

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ABSTRACT

We present three unusual cases of total anomalous systemic venous drainage (TASVD) where all the systemic veins were abnormally connected to the atrium that drained into the left ventricle (LV). All three patients had features consistent with left atrial isomerism. Based on the available evidence and review of literature we propose that TASVD be included as a part of the syndrome of left atrial isomerism. A classification for TASVD is also proposed with surgical implications.

Keywords: Heterotaxy syndrome, left isomerism, total anomalous systemic venous drainage

INTRODUCTION

Total anomalous systemic venous drainage (TASVD) is an exceptional form of congenital heart disease, wherein all systemic venous flow, including the right superior vena cava (RSVC), persistent left SVC (LSVC), inferior vena cava (IVC) (which may be interrupted), and coronary sinus, drain into the atrium draining into the morphologic left ventricle (LV). We describe three patients of this rare entity. Available literature is reviewed and a classification proposed.

CASE REPORTS

Case 1

A 5-year-old girl was admitted to our hospital for evaluation of mild cyanosis with a room air oxygen saturation of 86%. There was no evidence of congestive heart failure. Her height and weight both ranked in the 90th percentile. Echocardiography revealed left atrial isomerism [Figure 1]. There was ventriculo-arterial concordance. Left atrium and LV were dilated (LV end diastolic Z score +2.8) [Figure 2]. The branch pulmonary arteries were normal in size. All the pulmonary veins and systemic veins including the hepatic veins drained into the left-sided atrium and connected to the LV. A sizable fossa ovalis atrial septal defect was seen. The aortic arch was right sided (the features have been highlighted in [Table 1]. Findings were confirmed by computerized tomography (CT) scan [Figure 3](Table 2,3). The child underwent biventricular repair via an atrial septation and a baffle, which routed venous drainage to appropriate ventricles. At surgery, both atrial appendages were left sided in morphology. At 1-year follow-up, the child was doing well with normal oxygen saturation and unobstructed systemic venous drainage.

Case 2

A 5-year-old child was observed to have cyanosis (oxygen saturation of 68%). There was no evidence of congestive

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DOI: 10.4103/0974-2069.132476

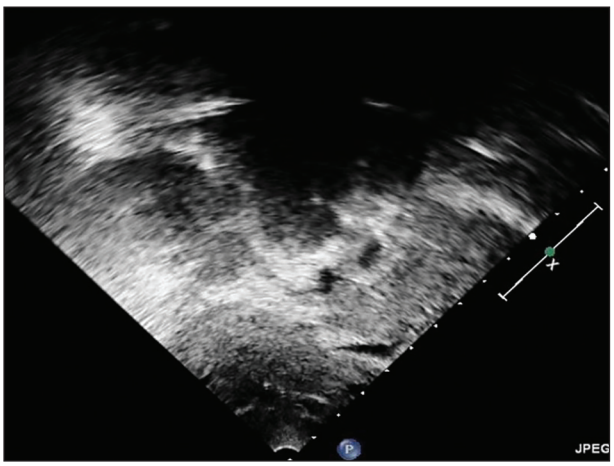

Figure 1: Two-dimensional (2D) echocardiography with substernal coronal view showing the relationship of the aorta and inferior vena cava (IVC) suggestive of left isomerism

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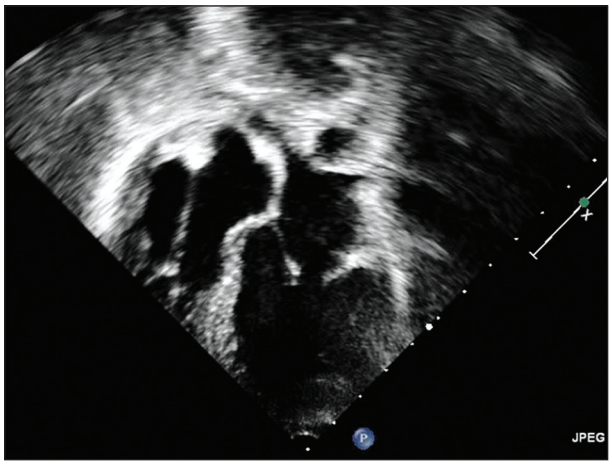

Figure 2: 2D echocardiography with four-chamber apical view showing dilated left atrium and left ventricle. This image is suggestive of left ventricle volume overload (in case 1)

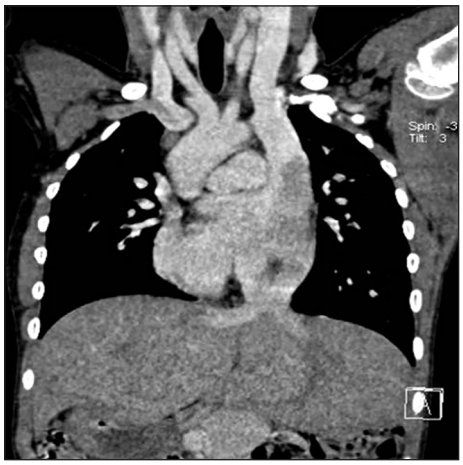

Figure 3: Computerized tomography (CT) of case 1 with coronal view showing the drainage of left superior vena cava (LSVC) and the hepatic veins into the left atrium

Table 1: Echocardiographic features of the three patients with Total Anomalous Systemic Venous Drainage (*n* = 3)

Stucture	Case 1	Case 2	Case 3
Situs	Ambiguous (left isomerism)	Ambiguous (left isomerism)	Ambiguous (left isomerism)
LSVC	To LA	To LA	To LA
RSVC	No	To LA (RSVC > LSVC)	To LA (RSVC > LSVC)
IVC	Interrupted	LA	Interruption
Pulmonary veins	LA	LA	LA
ASD	Yes	Yes	Yes
AV valves	2 AV valves	2 AV valves	2 AV valves
LV	Dilated (+3.1)	Dilated (RV hypoplastic)	Dilated
IVS	Intact	Small apical VSD	Non restricted VSD (routable)
VA	Concordance	Concordance	DORV normally related
PA	No PS	No PS	Severe PS
Arch	Right arch	Normal	Normal

LSVC: Left superior vena cava, RSVC: Right superior vena cava, IVC: Inferior vena cava, AV: Atrioventricular, LV: Left ventricle, LA: Left atrium, PA: Pulmonary artery, PS: Pulmonary stenosis, VSD: Ventricular septal defect, DORV: Double-outlet right ventricle, CS: Coronary sinus, b/l: Bilateral

Table 2: Ultrasonological and computerized tomography (CT) scan profile of our patients (*N* = 3)

Investigation	Case 1	Case 2	Case 3
CT	Bilobed lungs Left isomeric pattern	Bilobed Left isomeric pattern	Bilobed Left atrial isomerism
USG	Polysplenia	Polysplenia	Not available

USG: Ultrasonography

heart failure. Echocardiography showed left isomerism with levocardia. There was a large atrial septal defect that shunted from left to right. The IVC was interrupted and left and RSVC drained into the left-sided atrium. All the four pulmonary veins formed a confluence and also drained to the left sided atrium. There was double outlet of the right ventricle (RV) with side-by-side great vessels. Nonrestrictive perimembranous ventricular septal defect was routable to aorta, with bidirectional shunting. Aortic arch was right sided. There was severe pulmonary stenosis with confluent and normal branch pulmonary artery sizes. CT scan confirmed the echocardiographic findings [Figure 4a and b] [Table 2,3]. The child underwent successful LV to aorta tunneling with an RV to pulmonary artery conduit along with atrial septation. Morphology of both the atrial appendages at surgery was left sided.

Case 3

A 2-year-old boy was incidentally found to be cyanotic with oxygen saturation of 75%. Echocardiographic examination showed left atrial isomerism with pulmonary veins draining to the left-sided atrium. Hepatic vein and bilateral SVC also drained to the left-sided atrium. The IVC ascended on the right of the spine, but crossed the midline and drained to the left sided atrium. There was a small fossa ovalis atrial septal defect with left to right shunt, ventriculo-arterial concordance, D-loop ventricles, and confluent and adequate sized branch pulmonary arteries. The RV was hypoplastic and bipartite. Ventricular function was normal (elaborated under Table 1). CT scan showed a midline liver with polysplenia, with associated agenesis of the tail of the pancreas (Table 2). Rest of the cardiac findings was same as that described in echocardiographic evaluation [Figure 5a and b]. The child underwent bidirectional Glenn shunt and atrial septectomy with banding of the main pulmonary artery. He has been doing well at 4 years follow-up and is now planned for Fontan conversion.

DISCUSSION

TASVD defies the guiding rule that the drainage of IVC serves as a reliable indicator for the chamber to be classified as right atrium. Although one exceptional case

was reported where all systemic venous blood returned to the coronary sinus,[1] the openings of total anomalous systemic veins are much more commonly connected to the left atrium.[2-10]

All of our cases had features consistent with left isomerism. Literature review provides 14 cases of TASVD [Table 4]. Five of the seven cases reported in the recent literature (after 1980, when the concept of isomerism was described) had features of left isomerism. Thus, it would be reasonable to suggest that the diagnosis of TASVD in any patient should be pointer to the presence of left isomerism. The lack of commitment regarding features of isomerism prior to 1980 is probably on account of the lack of awareness of the concept of isomerism prior to the period[12] (the same has been highlighted in Table 4).

Most of the cases were elegantly compiled by Zhang *et al.*, and the present review owes much to their contribution.[13] We propose a modification of Zhang's classification of TASVD. This has implication in planning cardiopulmonary bypass for the surgical management of the lesion. There are three subsets of TASVD, based on the site of drainage of IVC. These may be further subdivided into type A and B based on the site of drainage of hepatic veins. Extending the proposed classification by Zhang *et al.*, who classified TASVD into

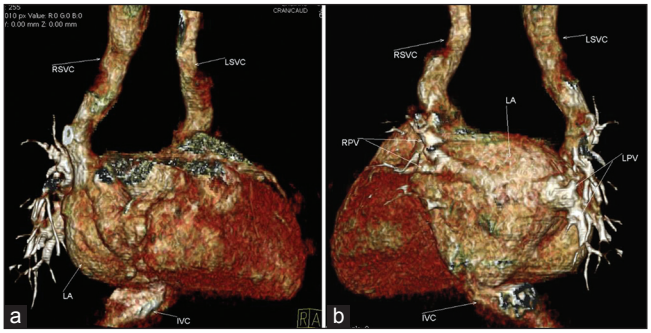

Figure 4: (a) 3D reconstructive CT images of case 2 showing the drainage of bilateral SVC and IVC in the left-sided atrium. (b) The same image as observed from the posterior aspect shows drainage of pulmonary veins in the same chamber (left sided atrium) as that of the systemic veins

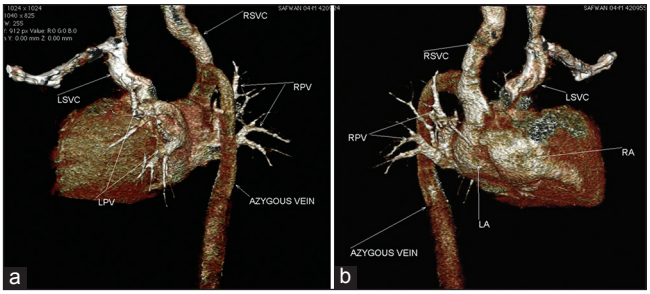

Figure 5: (a) 3D reconstructive images of the patient (case 3) showing IVC interruption and its continuation as azygous and drainage into the right SVC (RSVC). Note also the drainage of the LSVC into the same chamber. (b) Shows the same image from the anterior aspect showing drainage of pulmonary veins also in the left-sided atrial chamber as that of the systemic veins

Table 3: Profile of our reported cases of TASVD (*N* = 3)

Reported cases	Number	RSVC	Drainage of persistent LSVC	Drainage of IVC type	Isomerism	Classification	Surgery
Case 1	1	LA	LA	Interrupted	Left	II	Intracardiac baffling
Case 2	1	LA	LA	LA	Left (CT: b/l left sidedness) polysplenia	I	Glenn
Case 3	1	No	LA (unroofed CS)	Interrupted	Left	II	Rastelli

TASVD: Total anomalous systemic venous drainage, LSVC: Left superior vena cava, RSVC: Right superior vena cava, IVC: Inferior vena cava, LA: Left atrium, CS: Coronary sinus, b/l: Bilateral

Table 4: Comparison and evaluation of the reported cases of total anomalous systemic venous connection

Reported cases	Number	Year	RSVC	Drainage of persistent LSVC	Drainage of IVC type	Isomerism	Classification
Gueron *et al.*[6]	1	1969		LA	LA		I
Miller *et al.*[7]	1	1965		LA	LA	Not committed	I
Viart *et al.*[8]	1	1977	No	LA	LA	Non committed	I
Krayenbuhl *et al.*[2]	1	1977		LA	Interrupted	NA	I
Danielson *et al.*[3]	1	1973		LA	Interrupted	Yes (left)	II
Pearl *et al.*[14]	1	1980	NA	NA	NA	NA	I
Johnson *et al.*[4]	1	2002		LA	Interrupted	Not commented	II
Roberts *et al.*[5]	1	1972	LA	LA	Interrupted	No details	II
Zhang *et al.*[13]	1	2009	LA	LA	LA	No	II
Turkoz *et al.*[9]	1	2010	No	LA	Interrupted	Left isomerism	II
Barrea *et al.*[10]	1	2010				Left hetrotaxy	
Ravindranath *et al.*[11]	1	2012	LA	No LSVC	Interrupted	Left hetrotaxy	II
Kadletz *et al.*[1]	1	1997	No	Coronary sinus	Coronary sinus	No	III
Lazzarin *et al.*[15]	1	2007	LA	No	LA		II

TASVD: Total anomalous systemic venous drainage, LSVC: Left superior vena cava, RSVC: Right superior vena cava, IVC: Inferior vena cava, LA: Left atrium

two types, according to the type of vena cava cannulation we would like to add a third type. In type I, the IVC is not interrupted and conventional cardiopulmonary bypass can be instituted. In type II, the IVC is interrupted, and single cannulation of the SVC would suffice for venous drainage. In type 3: With IVC drainage to an accessory chamber like coronary sinus. The accessory chamber may be cannulated or total circulatory arrest instituted for adequate visualization. These three types are further subdivided into type A where the hepatic veins too drained separately to the left-sided atrium and type B where the hepatic veins would drain to an alternative site and need to be specifically looked for.

Proposed classification (modified from Zhang *et al.*):

- IVC to left-sided atrium: Type 1
- IVC interruption: Type 2
- IVC to accessory chamber (like coronary sinus): Type 3
- Type A: Hepatic veins to left-sided atrium
- Type B: Hepatic veins to other site

The embryologic basis is hypothesized as follows. TASVD may occur because of the failure of regression of the right valve of systemic venous sinus (sinus venosus) or because of the systemic venous sinus being incorporated into the left atrium. As for the clinical presentation, these systemic venous anomalies are a rare cause of cyanotic congenital heart disease, with unremarkable precordial examination. Associated cardiac lesions predominantly dictate the clinical features.

Hemodynamically, the left atrium forms the mixing chamber in the circuit, and hence, similar to the case of a total anomalous pulmonary venous drainage, oxygen saturation is equal in all the chambers. Although echocardiography is able to delineate most of the observations, a CT scan helps confirm drainage of all veins concerned to facilitate surgical intervention.

Surgical intervention is guided by associated cardiac morphology. In the presence of isolated TASVD, atrial septation with appropriate routing of veins suffices. In the presence of associated lesions a more complex intracardiac repair with rerouting of the veins may be done. In the presence of associated extreme ventricular imbalance, a univentricular pathway may need to be resorted to.

All three of our cases had bilateral left atrial appendages. Available literature does not always comment on atrial morphology. Hence in such cases, it was impossible to decide laterality, or the lack of it. However, most described cases of TASVD do gravitate into bilateral left sidedness. On the basis of this experience and review of literature, we suggest that that presence of TASVD indicates a strong likelihood of left isomerism.

ACKNOWLEDGMENT

Mr. Vinay Kumar (CT technician) for 3D reconstructive images.

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How to cite this article: Awasthy N, Radhakrishnan S, Kaushal S, Sharma R. Total anomalous systemic venous drainage to the left atrium: An entity reviewed and investigated. Ann Pediatr Card 2014;7:98-102.

Source of Support: Nil, **Conflict of Interest:** None declared