

Total Anomalous Systemic Venous Drainage. A Case Report. Surgical Considerations

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SUMMARY

Total anomalous systemic venous drainage (TASVD) with no concomitant heart defects is an unusual condition. A 24-hour old female new born was referred to our center due to cyanosis and dyspnea. Color-Doppler echocardiography revealed a partial anomalous systemic venous drainage of the superior vena cava to the left atrium. Cardiac catheterization showed a total anomalous systemic venous drainage into the left atrium. The patient underwent corrective surgery at the age of sixteen months with excellent outcomes.

Key words >

Total Anomalous Systemic Venous Drainage - Cyanosis - Cardiac Surgery

Abbreviations >

RA	Right atrium	ECG	Electrocardiogram
LA	Left atrium	CS	Coronary sinus
PA	Pulmonary artery	IVC	Inferior vena cava
TASVD	Total anomalous systemic venous drainage	RSVC	Right superior vena cava
PDA	Patent ductus arteriosus	RV	Right ventricle
ASD	Atrial septal defect	LV	Left ventricle
VSD	Ventricular septal defect	TV	Tricuspid valve

BACKGROUND

Complex congenital heart defects, especially asplenia and polysplenia syndromes, are frequently associated with different types of anomalous systemic venous drainages. (1) Total anomalous systemic venous drainage (TASVD) - in which right superior vena cava (RSVC), inferior vena cava (IVC) and coronary sinus (CS) are connected to the left atrium (LA) - is unusual in the absence of concomitant congenital heart defects. (2-4) This condition requires the presence of a left-to-right shunt [atrial septal defect (ASD), patent ductus arteriosus (PDA) or ventricular septal defect (VSD)] to allow the systemic venous return to reach the pulmonary circulation. We describe the findings of this rare congenital defect and discuss the surgical options available.

CASE REPORT

A 24-hour old female new born (weight: 2.5 kg) was referred to our center with mild dyspnea and cyanosis. Pulse oximetry: 84%. There were no other relevant signs in the physical examination. Chest X-ray: lung fields were normal and cardiothoracic index was 0.55. ECG: left ventricular dominance. Echocardiography: left chambers were moderate dilated and a small PAD was present. Infusion of saline solution through a right

antecubital intravenous access showed the passage of microbubbles into the LA, suggesting the presence of a partial anomalous systemic venous drainage into the LA. Color-Doppler echocardiography revealed systemic venous drainage from the RSVC into the LA. We did not find any other anomalies. As the infant was clinically stable, she was discharged with an indication of surgery once she had gained weight. At the age of 15 months the infant weighted 10 kg; a cardiac catheterization was performed and the following pressures were obtained: left ventricle (LV) 68/0-8; right ventricle (RV): 23/0-6; LA: 12; and right atrium (RA): 12. Angiography: TASVD into the LA, normal pulmonary venous drainage, wide ASD, left chambers dilation. Cyanosis due to complete blood mixing in the atrium. Left chambers hypertrophy. Low pulmonary blood flow. The RV was heavily trabeculated with mild hypoplasia; main pulmonary artery and its branches were also hypoplastic.

The patient underwent corrective surgery at the age of 16 months. After median sternotomy the heart was visualized; its external appearance was normal (*situs solitus*), except for the left anterior descending coronary artery that was remarkably displaced rightwards. Aortic cannulation. Both vena cavae were cannulated 2 cm before reaching the LA, and the patient was placed under pulmonary bypass and hypothermia

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(25°C). A 5-mm PAD was ligated. Hyperkalemic crystalloid cardioplegic solution was used at 4°C. A right atriotomy was made; the RA was small and no veins drained into it. The diameter of the tricuspid valve (TV) was smaller than usual. An ASD with a diameter of 5 mm was present. The smooth portion of the interatrial septum and the crista terminalis were absent. The right atrial appendage was normal. The RV was small and densely trabeculated.

The interatrial septum was resected and both vena cavae and the CS were visualized draining into the LA. Eustachian and Thebesian valves were absent. Pulmonary venous drainage was normal. The morphology of both AV valves was normal, except for the small diameter of the TV. The interatrial septum was closed with a bovine pericardial patch and the systemic venous return was left draining into the RA. Right ventricular trabecules were resected to enlarge the chamber. A 4-mm fenestration was created on the new interatrial septum to maintain patency in case of RV failure or TV or PA obstruction. Finally, the RA was closed with a bovine pericardial patch to enlarge the chamber.

The patient had an excellent recovery with no evidence of heart failure. She did not require inotropic agents and was discharged 5 days after surgery. The girl is currently 12 years old and is in functional class I.

DISCUSSION

Total and partial anomalous systemic venous drainages are extremely infrequent conditions that occur in hearts with complex congenital defects. (5, 6) Total anomalous systemic venous drainage requires the presence of a left-to-right shunt (PDA, ASD or VSD) to allow the systemic venous return to reach the pulmonary circulation. Functional drainage of systemic venous blood into the left atrium across the atrial septal defect has been described. (7) The aim of this report is to discuss congenital defects with anatomic diversion of systemic venous return into the LA. The abnormal connection between the SVC and the LA may be due to persistence of the right venous valve of the sinus venosus, producing a left-to-right shunt through a superior-type sinus venosus ASD. Malposition of the right horn of the sinus venosus in a left and cephalic direction has also been postulated. (5, 8, 9) The IVC and the CS may drain into the LA when the orifices of the venous valves direct their blood flow towards an ostium secundum ASD. (10, 11) There are other mechanisms which still remain unclear. (6)

Total anomalous systemic venous drainage may occur when the right valve of the systemic venous sinus fails to regress, and partitions the RA into two compartments. (10-12) The upstream chamber receives the venous flow which passes through a patent foramen ovale into the LA. The septation has fenestrations through which the blood reaches the pulmonary circulation. We excised this membrane and found the orifices of the IVC and the CS without their corresponding valves as

this membrane represents persistence of the entire sinus venosus valves. Another theory is that TASVD probably results from the sinus venosus being incorporated into the LA; (13) then, when the interatrial septum develops, systemic and pulmonary veins drain into the LA. Gueron (4) reported the presence of an ostium secundum ASD in a similar case and mentioned the theory of atrial partition when sinus venosus valves fail to regress. If a complete membrane divides the RA into two chambers, the ASD will not be visualized when the RA is excised. The membrane may be perforated and direct the blood flow of the IVC towards the ostium secundum. In this case, the ASD will not be visualized until the membrane is excised. When we excised the RA in our patient, we did not find an ostium secundum ASD; the communication between both atria was higher, probably due to incomplete partition between the right sinus venosus valve and the inferior border of a superior-type sinus venosus ASD.

The presence of a PDA with a left-to-right-shunt might have contributed to the underdevelopment of the right chambers. In the absence of a PDA, the blood flow through the ASD would have entered the RA, RV and PA and these cavities would have developed better.

We conducted a physiological surgical approach; other techniques form a tunnel, rerouting the blood from the pulmonary veins to the mitral valve. (3) This technique creates a small, non-extending chamber that might pose an obstacle to increase cardiac output if required. The one and a half ventricle repair technique might have been attempted if the characteristics of the RV had been thought to be insufficient to carry a complete cardiac output. The technique consists of bidirectional Glenn procedure (SVC to right PA) leaving the IVC connected with the RA. Fortunately we have chosen the correct method as the patient did not present signs of congestive heart failure despite the septal fenestration.

BIBLIOGRAPHY

1. Van Praagh S, Santini F, Sanders S. Malposiciones cardíacas con especial atención a la heterotaxia visceral (Síndromes de asplenia y polisplenia). En: Nadas A, Fyler D, editores. *Cardiología Pediátrica*; 1994. p. 593-612.
2. O'Connor L, Núñez L, Gil M y col. Drenaje venoso sistémico anómalo total a aurícula izquierda: corrección quirúrgica. *Rev Esp Cardiol* 1988;41:566-70.
3. Viart P, Le Clerc J, Primo G, et al. Total anomalous systemic venous drainage. *Am J Dis Child* 1977;31:195-8.
4. Gueron M, Hirsh M, Borman J. Total anomalous systemic venous drainage into the left atrium. Report of a case of successful surgical correction. *J Thorac Cardiovasc Surg* 1969;58:570-74.
5. Alday L, Maisuls H, De Rossi R. Right superior caval vein draining into the left atrium. Diagnosis by color flow mapping. *Cardiol Young* 1995;5:345-9.
6. Meadows W, Bergstrand I, Sharp J. Isolated anomalous connection of a great vein to the left atrium. The syndrome of cyanosis and clubbing, "normal" heart, and left ventricular hypertrophy on electrocardiogram. *Circulation* 1961;24:669-76.

7. Maillis M, Cheng T, Meyer J, et al. Cyanosis in patients with atrial septal defect due to systemic venous drainage into the left atrium. *Am J Cardiol* 1974;33:674-8.
8. Braudo M, Beanlands D, Trusler G. Anomalous drainage of the superior vena cava into the left atrium. *Canad Med Assoc J* 1968;99:715-9.
9. Gorenflo M, Sebening C, Ulmer H. Anomalous connection of the right superior caval vein to the morphologically left atrium. *Cardiol Young* 2006;16:184-6.
10. Lucas R, Krabill K. Anomalous venous connections, pulmonary and systemic. En: Adams F, Emmanoulides G, Riemenschneider T, editors. *Moss' Heart disease in infants, children and adolescents*. 4th ed. Baltimore, USA: Williams & Wilkins; 1989. p. 580-617.
11. Sierig G, Vondrys D, Dachnert I. Anomalous drainage of the inferior caval vein to the left atrium. *Cardiol Young* 2005;15:85-7.
12. Ledden-Klok M, de Mol A, Backx A. Symptomatic divided right atrium in a newborn. *Cardiol Young* 2007;17:110.
13. Roberst K, Edwards J, Astley R. Surgical correction of total anomalous systemic venous drainage. *J Thorac Cardiovasc Surg* 1972;64:803-10.