Biatrial Drainage of Right Superior Vena Cava with Anomalous Right Pulmonary Venous Connection

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Summary

A case of right superior vena cava draining to both atria, predominantly to the left atrium, with anomalous right pulmonary venous connection to the lower right superior vena cava is reported. The haemodynamic significance of these anomalies is discussed, and the technique of surgical repair is described. The literature on this rare but interesting clinical entity is briefly reviewed.

Key words

Anomalous right superior vena cava – Anomalous pulmonary venous connections

Zufluß der oberen Hohlvene in den rechten und linken Vorhof in Verbindung mit einer fehlmündenden rechten Lungenvene

Es wird über einen 7jährigen Jungen berichtet, der aufgrund eines linksseitigen perietalen Hirnabszess stationär aufgenommen wurde. Im Rahmen der weiteren Untersuchungen zeigte sich bei einer Farbdoppler-Echokardiographie eine in den linken Vorhof mündende rechtsseitige obere Hohlvene. Die Herzkatheteruntersuchung bestätigte diese Diagnose, zusätzlich zeigte sich eine Verbindung zwischen der oberen Hohlvene und dem rechten Vorhof mit einem Druckgradienten von 6 mmHg zwischen beiden. Bei der Korrekturoperation sahen wir eine in den unteren Teil der oberen Hohlvene fehlmündende rechte Lungenvene, sowie eine in den rechten und linken Vorhof mündende Hohlvene. Die Einmündung in den rechten Vorhof war stenotisch verändert, die gesamte übrige Anatomie war normal. Mit einem autologen, in den unteren Teil der oberen Hohlvene eingenähten Perikardpatch wurde dann die Korrektur derart vorgenommen, daß die rechte Lungenvene in den linken Vorhof und die obere Hohlvene ausschließlich in den rechten Vorhof mündete. Die Stenose der Hohlvene wurde durch Quervernähung einer Longitudinaleröffnung erweitert. Der postoperative Verlauf war völlig komplikationslos, der Patient befindet sich heute in ausgezeichnetem, körperlichen Zustand. Es wird die Literatur dieser seltenen angeborenen Fehlbildung ausgewertet und diskutiert.

Introduction

Different types of anomalous systemic venous connections to the left atrium (LA) have been described. Biatrial connections of an isolated right superior vena cava (SVC) may present as a diagnostic enigma. After excluding haemoglobinopathies and chronic lung conditions, cyanosis out of proportion to the apparent physical and cardiac status of the patient, left-ventricular enlargement, and lack of radiographic findings compatible with the other forms of cyanotic heart disease should make the correct diagnosis possible. Diagnosis of this type of anomaly is very important because of the excellent surgical results obtainable.

We herein report a case of isolated Right SVC connected to both atria. The diagnostic work up and corrective surgical repair are discussed.

Case Report

A 7-year-old boy, weighing 17 kg, presented with fever and convulsions on the right side of the body and from a CT-scan was diagnosed to have a left parietal cortical abscess. Left parasagittal parietal burr hole was made and 70 ml of foul-smelling pus was aspirated. His condition showed marked improvement thereafter with appropriate antibiotics and supportive therapy.

His physical examination revealed mild cyanosis and Grade I clubbing, a pulse of 102 per minute, regular, normal volume, and blood pressure of 120/70 mmHg. The jugular venous pressure and the peripheral arterial pulses were normal. On cardiac examination, the precordium was quiet, apex beat was at normal position. S_1 and S_2 were normal. No murmur was heard. There were no signs of cardiac failure or infective endocarditis. The central nervous system examination revealed mild right-sided residual hemiparesis. All other systems were normal.

His hematocrit was 46 % and the white cell and platelet counts were normal. Serum meth- and sulphhaemoglobinemia were not detected by spectroscopic examination. The haemoglobin electrophoresis was normal. Respiratory function tests were normal. Occult pulmonary embolism, as a cause of patients's systemic oxygen de-

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Table 1 Cardiac catheterization data.

Site	Oxygen saturation %	Pressure (mmHg)
Right SVC		
near atrium ("lower")	78.5	6/0/4
further away	70.2	17/9/10
IVC	71.0	
RA	69.5	9/1/4
RV	69.1	25/4
MPA	68.6	27/9/17
LA	82.0	9/0/5
Aorta	82.0	86/44/60

Abbreviations: SVC – Superior vena cava
IVC – Inferior vena cava
RV – Right ventricle
MPA –Main pulmonary artery
LA – Left atrium

saturation, was excluded by a ventilation-perfusion lung scan.

His electrocardiogram (ECG) showed a rate of 100 per minute, regular sinus rhythm, the QRS axis of +30°, and counter-clockwise loop and left-ventricular hypertrophy (LVH). The chest radiograph revealed normal cardiothoracic ratio. Cross-sectional echocardiography with colour Doppler and contrast studies showed a right SVC draining in the LA. The interatrial and interventricular septae were intact. Left-ventricular ejection fraction was normal.

At the cardiac catheterization the catheter passed readily from the IVC into the right atrium (RA), but there was a mean pressure gradient of 6 mmHg on pull back from the right SVC into the RA. There was an 8 % jump in oxygen saturation in the "lower" (near the atrium) SVC suggestive of a left-to-right shunt at this level. However, there was systemic desaturation (arterial oxygen saturation 82 %). Left innominate vein injection revealed filling predominantly of the LA and also part of the RA (Fig. 1), establishing the diagnosis of a right SVC draining predominantly to the LA and very little to the RA. Pulmonary veins were not entered (Table 1).

The repair was undertaken under cardiopulmonary bypass (CPB) via median sternotomy. There was no left superior vena cava. The right pulmonary veins were seen to drain into the lower SVC. High SVC cannulation was done using a short metal-tipped cannula, besides the usual ascending aorta and IVC cannulation for CPB. The patient was cooled to 28 °C, the ascending aorta was cross-clamped and cold crystalloid cardioplegia was injected into the aortic root. Both cavae were snugged.

A longitudinal paraseptal right atriotomy extending into the SVC revealed that the major portion of the right SVC was opening predominantly into the LA. The part of the right SVC draining to the RA was stenotic. The interatrial septum was intact. This was incised vertically cephalad from the posterior margin of the fossa ovalis. The openings of the IVC and coronary sinus were normal. Left pulmonary veins were draining normally into the LA. The mitral valve was normal.

An autogenous pericardial patch was used to repair the atrial septotomy and divide the lower SVC into two compartments, the posterior portion diverting the pulmonary venous flow to the LA and the anterior compartment leading to the RA. The lower SVC was then completed by suturing the upper part of the longitudinal incision in a transverse manner. This provided unobstructed flow of SVC blood to the RA.

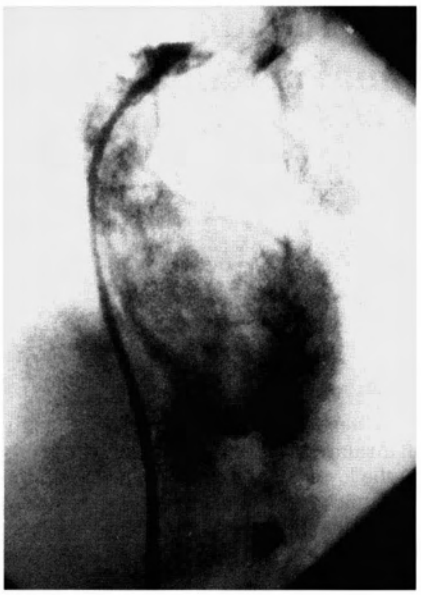

Fig. 1 The angiogram showing injection made into the left innominate vein opacifying the Right Superior Vena Cava draining preferentially into the left atrium, intact interatrial septum, and little contrast entering the right atrium via a narrow channel.

The heart was deaired and the patient weaned off CPB after rewarming to normothermia. He had excellent postoperative haemodynamics. Analysis of blood from RA and LA after weaning off from CPB showed no shunt and no left-sided desaturation. There was no gradient between right SVC and RA. He was discharged on the fifth postoperative day after an uneventful postoperative course. His postoperative contrast echocardiographic studies showed right SVC draining exclusively to the RA.

Discussion

The most frequently encountered anomaly of the great veins is a persistent left SVC draining to coronary sinus (2, 6). In contrast, total anomalous drainage of the right SVC into the LA is extremely rare. First described by *Wood* (18) in 1956, 12 cases – 6 children and 6 adults have been reported to date.

One of the causes of cyanosis after excluding primary lung diseases, pulmonary arterio-venous fistulae, methaemoglobinaemia and other haemoglobinopathies may be anomalous systemic venous connections (12). The right or left SVC, the IVC or the hepatic veins may drain into the LA (13). It usually occurs in association with additional congenital malformations of the heart, such as atrial septal defect, ventricular septal defect, tetralogy of Fallot, pulmonary atresia, transposition of the great arteries, and patent ductus arteriosus (4, 13). Drainage of persistent left SVC into the LA as an isolated finding has been described, as has drainage of the IVC into the LA (3).

In the present case, the right superior vena cava was draining to both atria. We found six similar cases to be reported in medical literature – 3 presented in childhood;

one died with severe cyanosis and congestive heart failure after unsuccessful surgery. The other three patients, presenting in adult life, were managed medically (13).

The haemodynamic consequences of a vena cava connected to both atria depend on several factors such as place of entry of anomalous pulmonary veins, if any, the relative size of channels to the right und left atria, and the pressures in the atria. The basic haemodynamic disturbance of isolated right SVC draining into the LA consists of an increased blood flow through the left-side chambers and decreased flow to the right-side chambers, a partial bypass of the lungs, and reduction of the systemic oxygen saturation, producing cyanosis.

The right-to-left shunt is generally well tolerated. In the cases reported by *Tuchman* et al. (16) and by *Park* and coworkers (10), the patients were asymptomatic until early adolescence. In the present case, the patient had no cardiac symptoms except for mild cyanosis and Grade I clubbing. Congestive heart failure is rare in these patients, as reported by *Vasques-Péréz* et al. (17). Normal growth and development have been reported in several cases. Three cases of brain abscess in association with this condition have been described (5, 6, 10). Our patient also presented with a brain abscess. Surgical repair is mandatory to prevent further events of paradoxical emboli (1, 7, 11). In their review, *Shapiro* et al. (13) observed that development was normal except in one and, while examination of the heart was normal, two had soft systolic murmur. In only one, who also had a dilated SVC, was the jugular venous pressure elevated and another had evidence of old inferior wall myocardial infarction. All had systemic arterial desaturation and polycythemia (13). None of these patients had associated intracardiac anomalies. The pulmonary vascularity was always normal. The ECG suggested left-ventricular hypertrophy or was normal (13). In the present case, however, counter-clockwise loop and left-ventricular hypertrophy were noticed.

The use of contrast echocardiography for diagnosis has been demonstrated and it provides a rapid and safe detection method (11, 14, 15). Cardiac catherization from the leg, a very common practice did not reveal the true diagnosis in the case reported by *Braudo* et al. (1). With more sophisticated non-invasive methods such as nuclear tracer studies, the preoperative diagnosis may be more feasible (10).

The drainage of the right SVC to the LA is most probably caused by leftward and cephalic malposition of the right horn of the sinus venosus in the presence of normally developed interatrial septum (7). Anomalous right pulmonary veins opening into the lower SVC, as seen in our case, were also noted by *Shapiro* et al. (13). They suggested that abnormal right pulmonary venous drainage into the lower SVC may cause the major portion of the SVC along with its systemic venous blood to shunt into the LA while the portion of the SVC draining to the RA may then become hypoplastic.

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