We describe the case of a 1-month-old infant with complete atrioventricular septal defect with right dominance, situs solitus, and drainage from the persistent left superior vena cava to the coronary sinus. Corrective surgery was carried out without previous cardiac catheterization. During the operation, the right superior vena cava was found to be absent. Cyanosis and head-and-neck edema were observed in the immediate postoperative period. Transthoracic echocardiography carried out after injection of a small volume of stirred saline into an epicraneous vein demonstrated the presence of microbubbles in the left cardiac cavities. A second operation was performed to prevent drainage from the left superior vena cava to the left atrium (via the unroofed coronary sinus) and to insert a PTFE conduit between the innominate vein and the right atrial appendage. The outcome was excellent. In this report, the embryological, clinical, diagnostic and therapeutic characteristics of this entity are discussed.

**Key words:** Right superior vena cava. Left superior vena cava. Coronary sinus.

INTRODUCTION

Persistent left superior vein cava (LSVC) is the most frequent abnormality of the systemic venous system (0.1%-0.5% of the general population) and is not usually associated with any other cardiac defect. Incidence in congenital heart disease varies (2%-5%) and it is more frequent in stenosis or pulmonary atresia, D-transposition, complete atrioventricular septal defects (CASD), and anomalous pulmonary vein drainage. The right superior vein cava (RSVC) is absent in 1% of patients with persistent LSVC and frequently associated with alterations of cardiac situs.

Persistent LSVC usually drains into the right atrium thru the coronary sinus (CS) but in 8% of patients it drains directly into the left atrium (LA) as a consequence of a defect in the wall that separates them (unroofed CS). When this occurs incidence of persistent LSVC is 75%. This combination is usually associated.
with right atrial isomerism and complete or partial atrioventricular septal defect, especially when substan-
tial interatrial communication (IAC) is present.2,3

We report the case of a 1-month-old infant with si-
tus solitus and CASD, absent RSVC and persistent
LSVC draining into the left atrium thru an unroofed
CS.

CLINICAL CASE

A 1-month-old boy was referred from another hos-
pital diagnosed with CASD with pulmonary hyperten-
sion. Pregnancy and birth had been normal.
The boy presented dyspnea with feeding and mini-
mal weight gain. He was being treated with digitalis
and furosemide.

On admission, the boy weighed 3300 g, appeared
undernourished, with normal phenotype, tachypnea,
2/4 pansystolic murmur, loud second sound, without
hepatomegaly. Chest x-rays revealed cardiomegaly
and plethora. The electrocardiogram (ECG) was in
sinus rhythm, with a superior QRS axis and biven-
tricular growth. Echocardiography confirmed CASD
in situs solitus, a large ostium primum-type CIA, in-
terventricular communication (IVC) partially hidden
by common atrioventricular valve chords, cleft mi-
tral valve, right ventricular dominance with viable
left ventricle, and LSVC draining into the CS (Fi-
gure 1).

We performed complete corrective surgery. We
found absent RSVC, using the LSVC for cannulation.

We used the Carpentier (double patch) technique and
acceptable mitral valve competence was achieved.

When disconnected from the pump the patient was in
nodal rhythm with the sternum open. A few hours la-
ter, significant desaturation and mantle or shoulder
girdle edema occurred. Echocardiography revealed ab-
sence of residual shunt, balanced ventricles and mild
tricuspid and mitral valve regurgitation. We did not see
the RSVC. Rapid injection of stirred saline solution in
an epicranial vein produced microbubbles in the left
cardiac cavities. The patient was reoperated immedia-
tely without extracorporeal circulation to introduce an
8 mm polytetrafluoroethylene (PTF) conduit between
the innominate vein and right atrial appendage and
drainage of the LSVC in the left atrium was prevented
thru a clip. The patient came out of the operation with
normal saturation. The postoperative period was posi-
tive and the patient was discharged at 13 days with a
regimen of captopril, furosemide and oral anticoagu-
lants.

DISCUSSION

Few systems are as susceptible to developmental
variations and anomalies as those of the principal sys-
temic veins. Although of scarce functional importance,
they sometime cause problems in the face of invasive
medical-surgical procedures.

In a 4 mm (week 4) embryo, the principal vein for-
mation that can be distinguished is the sinus vein,
where 3 vein groups drain (Figure 2):
– Vitelline vein system. Transports blood from the
vitelline sac.
– Umbilical vein system. Brings blood from the pla-
centa.
– Cardinal vein system, which is completely in-
traembryonic. The anterior and posterior cardinal
veins drain to the right and left of the venous sinus.
The common cardinal veins begin at the point where
they join.

At 15-17 mm, the right umbilical vein disappears
and the left umbilical vein connects distal to the hepa-
tic plexus (venous conduit). The left vitelline vein
atrophies and the right vitelline vein contribute to form
the inferior vena cava. An anastomosis appears (in-
nominate vein) between the anterior cardinal veins.
The common right cardinal will ultimately become the
RSVC and the common left cardinal atrophies leaving

ABBREVIATIONS

LSVC: left superior vein cava.
RSVC: right superior vein cava.
CS: coronary sinus.
LA: left atrium.
CASD: complete atrioventricular septal defect.
Anomalies of superior vena cavae are rare in situs solitus: only occasionally does LSVC draining in the CS appear and it has no hemodynamic consequence. Exceptionally, cyanosis appears in patients with unroofed CS and heterotaxia is then frequent.

Absent RSVC in situs solitus is exceptional (0.1% of patients with cardiomyopathy) and 130 cases had been described prior to 1997. It is associated with LSVC draining in the CS. The RSVC becomes little more than a fibrous chord and in half of these patients some type of cardiac abnormality is found. More frequent abnormalities are those that affect the CS—above all partial or total unroofed CS—which always drains a LSVC causing cyanosis. These cases are associated with CASD, especially in patients with large CIA.

Diagnosis of these anomalies is by clinical symptoms and echocardiography. In our patient, the severe desaturation in the postoperative period was fundamental. In echocardiography, a dilated CS always leads to suspicion of the presence of a LSVC draining in the CS but the diagnosis of defects in the roof makes dilation of the CS less likely. Saline contrast echocardiography provided confirmation of LSVC draining in the left atrium.

Echocardiography of RSVC is not systematic but its absence should have been discounted in this patient (CASD with large CIA). Diagnosis prior to surgery or other invasive procedures is important to avoid difficulties in implantation of pacemakers and catheters, cannulation for surgery, cavopulmonary derivation or transplantation. Transesophageal echocardiography and magnetic resonance imaging can be of great help in the diagnostic process.

We should point out the excellent results when correcting complete atrioventricular septal defect with right dominance. In the reoperation, intraatrial correction was not contemplated for 2 reasons: a small left atrium in a 1-month-old infant with unbalanced CASD, and to avoid extracorporeal circulation. Nor was direct reconnection of the LSVC with the right atrial appendage viable due to the distance between the structures and the aortic interposition. We also rejected using the bidirectional Glenn procedure due to pulmonary pressure in CASD. Implantation of a Gore-Tex conduit between the innominate vein and the right atrium has been described in an older child. In our patient, we foresee the need for replacement of the conduit for another of a larger caliber.

**CONCLUSIONS**

Knowledge of absent RSVC is of great interest for correct medical-surgical treatment of children with cardiomyopathy. In patients with CASD, absent RSVC and persistent LSVC oblige us to discount CS roof defects that can complicate the immediate postoperative period.
REFERENCES