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 epicranial 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.1,2

The right superior vein cava (RSVC) is absent in 1% of patients with persistent LSVC and frequently associated with alterations of cardiac situs.1

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

BRIEF REPORTS

Absent Right Superior Vena Cava With Left Superior Vena Cava Draining to an Unroofed Coronary Sinus

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°Se describe el caso de un lactante de 1 mes de vida con un defecto completo del septo auriculoventricular de predominio derecho, situs solitus y vena cava superior izquierda persistente con drenaje en el seno coronario. Sin cateterismo previo, se realizó una cirugía correctora, durante la que se descubrió la ausencia de la vena cava superior derecha. El postoperatorio inmediato cursó con cianosis y edema en la exclavina. La ecocardiografía transtorácica con inyección de suero fisiológico agitado en una vena epicranial mostró microburbujas en las cavidades izquierdas. El paciente fue reintervenido para cerrar el drenaje de la vena cava superior izquierda en la aurícula izquierda e interponer un conducto entre la vena inominada y la orejuela de la aurícula derecha. La evolución fue excelente. Se exponen y discuten los aspectos embriológicos, clínicos, diagnósticos y terapéuticos de esta asociación.

Palabras clave: Vena cava superior derecha. Vena cava superior izquierda. Seno coronario.
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with right atrial isomerism and complete or partial atrioventricular septal defect, especially when substantial interatrial communication (IAC) is present.2,3

We report the case of a 1-month-old infant with situs 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 hospital diagnosed with CASD with pulmonary hypertension. Pregnancy and birth had been normal.

The boy presented dyspnea with feeding and minimal 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 biventricular growth. Echocardiography confirmed CASD in situs solitus, a large ostium primum-type CIA, interventricular communication (IVC) partially hidden by common atrioventricular valve chords, cleft mitral valve, right ventricular dominance with viable left ventricle, and LSVC draining into the CS (Figure 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 later, significant desaturation and mantle or shoulder girdle edema occurred. Echocardiography revealed absence 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 immediately 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 positive and the patient was discharged at 13 days with a regimen of captopril, furosemide and oral anticoagulants.

DISCUSSION

Few systems are as susceptible to developmental variations and anomalies as those of the principal systemic 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 formation 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 placenta.
– Cardinal vein system, which is completely intraembryonic. 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 hepatic plexus (venous conduit). The left vitelline vein atrophies and the right vitelline vein contribute to form the inferior vena cava. An anastomosis appears (innominate 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.

Figure 1. Echocardiographic image of persistent left superior vena cava (LSVC).
only a small channel (CS). If this does not atrophy, we
call this persistent LSVC draining in the CS.4

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 un-
roofed CS and heterotaxia is then frequent.2,3

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 chord2,5 and in half of these pa-
tients 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 asso-
ciated with CASD, especially in patients with large
CIA.6

Diagnosis of these anomalies is by clinical symp-
toms and echocardiography. In our patient, the severe
desaturation in the postoperatory period was funda-
mental. 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.3 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 diffi-
culties in implantation of pacemakers and catheters,
cannulation for surgery, cavopulmonary derivation or
transplantation.8,9 Transesophageal echography and
magnetic resonance imaging can be of great help in the
diagnostic process.10

We should point out the excellent results when cor-
recting complete atrioventricular septal defect with
right dominance. In the reoperation, intraatrial correc-
tion11 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 viable12 due to the distance between
the structures and the aortic interposition. We also re-
jected using the bidirectional Glenn procedure13 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.14 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 de-
fects that can complicate the immediate postoperatory
period.

Figure 2. Posterior view of the
heart of an embryo at 4 weeks (A),
8 weeks (B), and 10 weeks (C)
geration. We see the atrophying of
the umbilical veins and of the left
vitelline vein (B). The asterisk (C)
represents the atrophying of the
anterior left cardinal vein (persis-
tent LSVC).
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