Unroofed Coronary Sinus and Persistent Left Superior Vena Cava as a Cause of Complication after Surgical Correction of a Double Outlet Right Ventricle

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Abstract: Unroofed Coronary Sinus (URCS) is a rare cardiac malformation, which was first described by Raghib *et al.* [1] in 1965, characterized by partial or complete fenestration between the coronary sinus and the left atrium. It is frequently associated with the existence of a left superior vena cava (PLSVC) and the clinical outcome is variable, from behaviour which is similar to an interatrial septal defect to the possibility of paradoxical embolism due to a right-left shunt at the level of the left atrium.

It can also be associated to other heart disorders, such as atrioventricular septal defects, interventricular communication, interatrial communication, cortriatriatum, tetralogy of Fallot or partial anomalous venous drainage and, if not diagnosed prior to the main surgical correction, may lead to post-surgical complications.

We present the case of a young infant with Double Outlet Right Ventricle with subaortic ventricular septal defect and side by side great arteries, persistent left superior vena cava and ostium secundum atrial septal defect who underwent surgery to correct the septal defects. Post-surgical evolution was difficult, however she recovered after diagnosis and treatment for an unroofed coronary sinus.

Keywords: Unroofed Coronary Sinus, Persistent Left Superior Vena Cava, Congenital Heart Disease.

CLINICAL CASE

A 2-month-old female baby, weighing 3,650g, underwent surgery with cardiopulmonary bypass for a Double Outlet Right Ventricle with subaortic ventricular septal defect and side by side great arteries, and ostium secundum atrial septal defect. The post-surgical period was complicated by right ventricular overload and pulmonary pressure increased to 80% of the systemic level. The patient required moderate inotropic treatment (Wernovsky Score of 17), nitric oxide and peritoneal dialysis. She was extubated 46 hours postoperatively; however, she subsequently presented symptoms of cardiac insufficiency with tachycardia, tachypnea and hepatomegalia, concurrent with echocardiographic findings of left ventricular dilation, moderate tricuspid regurgitation, paradoxical motion of the interventricular septum, conserved left ventricular contractility and a small, apparently Gerbode type atrioventricular defect. As a result of low cardiac output she was reintubated and, 5 days after the first operation, underwent a second intervention to close the septal defect. Evolution was difficult with predominantly right sided cardiac insufficiency which required moderate inotropes. Echocardiographic data suggested a persisting shunt which primarily dilated the right cavities and increase pulmonary blood flow and, secondary to this, pulmonary hypertension. Ventilation, either invasive or non-invasive, was required for a further week.

A cardiac catheterization showed no residual ventricular septal defect but revealed a restrictive left ventricle. After the catheters were removed at the end of the study, results of a right atrial blood gases were received and showed a large oximetry data jump for which no explanation could be found. We accept it was a mistake the withdrawal of catheter before to get oximetric values. Medication was intensified with the inclusion of levosimendan. The patient was extubated again 11 days after the 2nd surgery, however non-invasive ventilation was required. The follow-up echocardiographic studies continued to show considerable dilation of the right cavities leaving the left ventricle proportionally small. BNP levels remained higher than 350 pcg/ml, independent of the treatment applied.

Faced with the persistent dilation of the right cavities, and evidence of the persistence of the left

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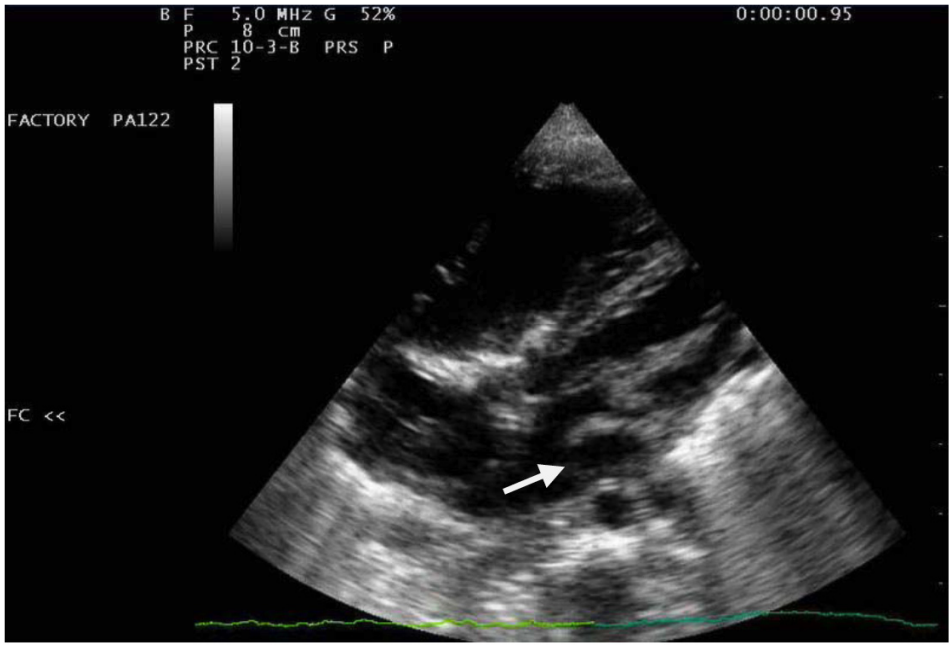

Figure 1: Apical four chambers view. Dilated coronary sinus is open to left atrium (arrow).

cava draining into the coronary sinus, it was suspected that this presented fenestration to the left atrium. An echocardiogram was performed while agitated saline solution was injected into a peripheral vein in the left arm. This revealed that the left atrium was immediately filled with microbubbles (Figures **1** and **2**), thereby demonstrating the existence of communication between the coronary sinus and this cavity, suggesting the existence of a shunt capable of dilating the right, but not the left cavities, and explaining the oximetry data jump in the right atrium detected during the catheterization. Using echocardiography it was then easy to identify the defect in the coronary sinus, which had been previously overlooked.

The following day, a catheter was inserted through the left external jugular vein to the coronary sinus, it was then withdrawn and located in the left vena cava. Successive gasometries were taken from both points and the following values were obtained: pO2 79mmHg and SatO2 97% in the coronary sinus and pO2 29 and Sat.O2 59% in the left vena cava. The interauricular

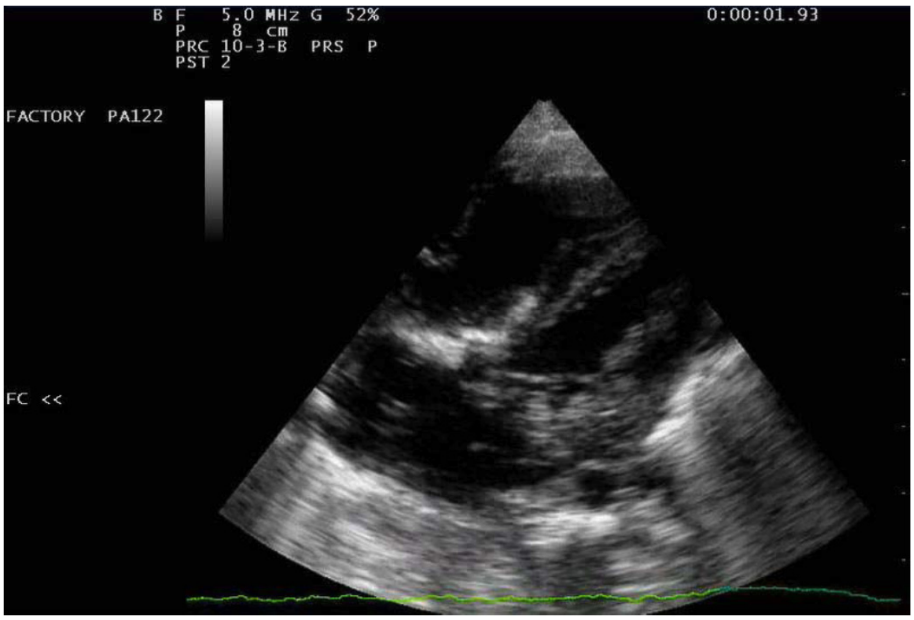

Figure 2: Apical four chambers view. After agitated saline solution was injected into a peripheral vein in the left arm, the left atrium was immediately filled with microbubbles.

shunt was confirmed taking advantage of the defect in the roof of the coronary sinus in its passage to the left atrium.

Two days after the diagnosis, the defect, which was quite large and found to be in the mid-coronary sinus and draining the left superior vena cava into the right appendage, was surgically closed. The patient quickly improved with the size of the left cavities becoming normal and the tricuspid regurgitation and the pulmonary hypertension, disappearing. The patient was discharged from the PICU five days later.

DISCUSSION

URCS is a rare cardiac malformation which was first described by Raghib *et al*. in 1965 [1]. It is frequently associated with venous drainage disorders, such as a persistent left vena cava [2].

Persistent left superior vena cava (PLSVC) is found in 0.2-0.5% of the population, and in 9% of those affected by congenital cardiopathy. In this latter group, approximately 41% are carriers of a left or right heterotaxy syndrome [2]. The cardiac defects most commonly found in the heterotaxy syndrome group are atrioventricular septal defects. However, those most frequently found in the PLSVC group without heterotaxy syndrome are interventricular communications, conotruncal anomalies and left ventricular outflow tract obstructions [2].

In approximately 8% of patients with PLSVC, due to the detachment of the coronary sinus, this vein drains into the left atrium between the appendix and the pulmonary veins. Complete detachment of the coronary sinus is highly related to heterotaxy and, as it is a complete defect, is not associated to dilation of the coronary sinus [3].

The 4 morphological types of URCS were described by Kirklin and Barratt-Boyes [4]: type I, completely unroofed with left superior vena cava (LSVC); type II, completely unroofed without LSVC; type III, partially unroofed midportion, and type IV, partially unroofed terminal portion. These latter types can be with or without PLSVC. Our case belonged to classification type III.

Although this defect can be diagnosed with transthoracic and/or transesophageal echocardiogram imaging, this may not be so easy in the presence of complex cardiopathies. Oktaki *et al*. [5] retrospectively analyzed 11 consecutive cases of patients affected by URCS without heterotaxy syndrome who underwent surgery: in 6 of these patients the diagnosis that concerns us here was made prior to surgery, another 3 cases were confirmed during surgery and, in the remaining 2, a residual shunt, which led to complications during the postsurgical period, led to the diagnosis. These were incomplete defects associated to PLSVC and the diagnoses were made either by angiography or by injecting a contrast material with saline solution into a subsidiary LSVC vein. Oktaki *et al*. [5] proposed that this procedure should be routinely performed in patients with congenital cardiopathy associated to PLSVC.

The symptoms of PLSVC+URCS are varied, from that corresponding to an interatrial defect, to the possibility of a paradoxical cerebral embolism or subcyanosis[5]. These may not be discovered until decades later in life, however, in the context of the postoperative period of a complex congenital heart disease in a young infant, this may impede the patient's recovery. Therefore, like Otaki *et al*. [5], in cases of LSVC drainage into the coronary sinus and other congenital cardiopathies, we propose the injection of agitated saline solution in the territory of the LSVC.

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