Introduction

Left superior vena cava (LSVC) to the left atrium (LA) communication is a congenital malformation of the sinus venous and caval system. It is a rare congenital cardiac anomaly which may appear as an isolated one, or as a part of more complex cardiac anomalies [1]. Drainage of the LSVC to the coronary sinus is well tolerated, but drainage to the LA produces right to left shunt and may be related to brain abscesses and/or embolization secondary to intravenous therapy administered through the left arm.

We present here a very practical method of bedside echocardiography diagnosis of LSVC-LA communication missed both by preoperative echocardiography and during the operation.

Case Report

A one-year-old girl with Down syndrome was admitted to the hospital with congestive heart failure symptoms. The echocardiographic examination revealed perimembranous large ventricular septal defect (VSD), patent ductus arteriosus (PDA) and pulmonary hypertension. The patient was scheduled for elective VSD closure and double ligation of the PDA under cardiopulmonary bypass (CPB). The arterial blood gases, which were totally normal during CPB, deteriorated at the end of the procedure. The surgical team thought that this deterioration could be related to the effect of CPB in a pulmonary hypertensive patient as no additional intracardiac pathology was present to explain this situation. Postoperatively, a low arterial oxygen saturation and 50-60% partial oxygen pressure persisted under mechanical ventilation with 90% inhaled oxygen. The postoperative physical examination was totally normal with no cardiac murmur. However on the chest x-ray we noticed that the tip of the central venous catheter inserted through the left internal jugular vein was directed to the LA (Fig. 1) via a possible LSVC-LA communication.

The postoperative echocardiography was totally normal with complete closure of the VSD. Under bedside echocardiographic examination, injection of 10 ml isotonic saline solution through this left-sided catheter filled the LA and the left ventricle (Fig. 2) via a direct communication between LSVC and LA. The patient was taken back to the operating room. The LSVC was explored distally to the innominate vein and snare-controlled with the pressure monitoring proximal to the snare. As the proximal pressure did not exceed 13 mmHg, the LSVC was doubly ligated.

The arterial blood gases immediately regained normal values and the patient was discharged at the 8th postoperative day following an uneventful postoperative period.

Discussion

Contrast echocardiography (CE) has been used as a clinical method for more than 20 years. Despite of the developments in Doppler methods and transesophageal echocardiography, right heart CE is still needed in some patients with atrial and pulmonary shunts, complex congenital heart disease, noisy Doppler recordings of tricuspid regurgitation (2). Of surgical importance, in suspected patients,
LSVC drainage into the LA or into the coronary sinus can be very easily diagnosed by CE where conventional transthoracic echocardiography can not be predictive.

LSVC is an uncommon congenital malformation. Incidence in general population being 0.3 – 0.5 % and in cases with associated malformation being 1.5 – 10 % (3). The LSVC-LA communication may cause right-to-left shunt and cyanosis, and as in our case, can be ligated if the pressure above the occlusion level is under 15 mmHg (4). Otherwise one of the surgical choices which are; division and reimplantation of the left superior vena cava (LSVC) to the right atrium (5), an intraatrial baffle to divert flow from the LSVC to the right atrium and close the atrial septal defect, or anastomosis of the LSVC to the left pulmonary artery (1) should be performed to restore the continuity of systemic venous return. Alternatively, transcatheter closure of LSVC by occlusion devices is reported in post-surgical patients (6,7).

The LSVC – LA communication which is a rare systemic venous return anomaly, should be considered in cases where intracardiac pathology does not explain the desaturation. Nevertheless, this pathology cannot be diagnosed exactly by two dimensional and color Doppler echocardiography. The exact diagnosis can be made by angiography or by the simple method of the bedside contrast echocardiography as in our case. We, therefore, think that CE is safe and highly informative for the definite diagnosis of LSVC with drainage into the LA or even into the coronary sinus.

References