

Case Report

Surgical treatment of persistent left superior vena cava draining to the left atrium with partial endocardial cushion defect: a case report

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Abstract: Background: Persistent left superior vena cava is a rare but important congenital vascular anomaly. However, persistent left superior vena cava draining to the left atrium with partial endocardial cushion defect is a very rare congenital anomaly occurring in postnatal life. We report on a rare case of persistent left superior vena cava draining to the left atrium with partial endocardial cushion defect. Case Report: This malformation was diagnosed in a 4-year-old girl during the operation. In operation, persistent left superior vena cava draining to the left atrium with partial endocardial cushion defect was found. In operation, we repaired atrial septal defect, repaired mitral valve, repaired tricuspid valve, left superior vena cava cut-off experiments negative, and ligated the persistent left superior vena cava. Conclusions: Data shows, preoperative ultrasonic cardiogram often cannot find persistent left superior vena cava, due to the permanent left superior vena draining to the left atrium is right to left shunt, so it has to be operated on to flow the bloodstream into the right atrium. The most accurately method still is the persistent left superior vena cava ligation, judging the indication is very important. This patient had a long term history, preoperative heart function was poor, postoperative effect still needed long-term follow-up observation. We think, such patients should be operated on as early as possible, returning to normal hemodynamics as soon as possible, the greatest degree of recovery of cardiac function.

Keywords: Persistent left superior vena cava, endocardial cushion defect, left atrium, ligation

Introduction

Persistent left superior vena cava (PLSVC) is a common internal thoracic vein malformation, during pregnancy, every important venous system of the whole body have different degrees of degradation or development, which is a complex physiological process. Persistent left superior vena incidence is 0.3-0.5% of the general population, and incidence is 3-10% in the congenital heart disease patients. The vast majority of permanent left superior vena draining to the coronary sinus, 92% cases of the patients' bloodstream draining to the right atrium, and do not cause clinical consequences. 10-20% cases of the patients were accompanied with absent right superior vena cava. And other 8% cases of the patients, left superior vena venous draining to the left atrial or the coronary sinus which is abnormal developed, causing right to

left shunt, causing cyanosis, globulism, thrombosis and other clinical consequences [1]. We have a surgical treatment of persistent left superior vena cava draining to the left atrium with partial endocardial cushion defect (PECD). Report as follows now.

Case report

A 4-year-old girl admitted to the clinic of Cardiac murmur was found more than one year. Her mother, pregnant 3 produce 2. She is the younger of the twins. 39+1 gestational weeks caesarean birth, birth weight 2.6 kg, crying sound weak, no birth trauma history and asphyxia history, Apgar score is unknown. This patient is poor growth, repeatedly upper respiratory tract infection, pneumonia, heart failure history, after crying, nose and lips without obvious cyanosis, had no difficulty in feeding. This



Figure 1. Chest X-ray showed: congenital heart disease, pulmonary blood increased.

patient with mental sleep, normal urine, normal bowel. Without hepatitis, tuberculosis and other infectious disease, without food and drug allergy history, without operation and trauma history. Physical examination: Heart sound is strong, left parasternal intercostal audible 2-3 and V/VI level systolic murmur, hyper P_2 . The electrocardiogram showed: right ventricular hypertrophy, P wave tip. Chest X-ray showed: congenital heart disease, pulmonary blood increased (**Figure 1**). Ultrasound cardiogram (UCG) showed: partial endocardial cushion defect, double superior vena cava, recommend intraoperative exploration of how does the left superior vena cava draining to the heart and with the exception of anomalous pulmonary venous malformations, etc. (**Figure 2**).

After the preparation of the operation, we did the partial endocardial cushion defect neoplasty, mitral valve neoplasty and tricuspid valve neoplasty by the cardiopulmonary bypass. In the operation, we saw that, the heart had been enlarged, primary atrial septal defect, the diameter was 30 mm, cuspid anterior valvulae bicuspidalis cracked $II^\circ C$, normal tricuspid valve parietal septum cracked $II^\circ C$, the diameter of the PLSVC was 1 cm, opening near the auricula sinistra, normal coronary sinus. Left superior vena cava cut-off experiments negative, ligated the PLSVC. We repaired cuspid anterior valv-

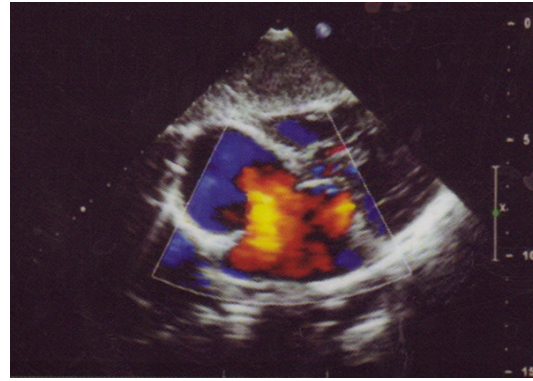


Figure 2. UCG showed: left atrium, right atrium and right ventricular enlargement. Atrial septal echo seen huge interruption maximum diameter of about 37 mm, the lower edge of the defect atrioventricular annulus, only on the edge of a small amount of residual atrial septal tissue, seen left to right shunt blood flow. Mitral valve roots visible cracks up, systolic blood visible regurgitation, pressure gradient (PG): 123 mmHg. Tricuspid valve septal leaflet visible cracks up the roots, systolic blood visible regurgitation, PG: 60 mmHg. No significant septal continuous interruption. Sternum fossa section visible remnant down the left superior vena cava drainage, drainage position display is unclear.

lae bicuspidalis, repaired tricuspid valve parietal septum. We used biological patch to repair the atrial septal defect. After the operation, we loosed transverse sinus forceps, ventricular fibrillation happened, we defibrillated 20 W/S twice, cardioversion returned to sinus rhythm, placed a temporary pacemaker. Blocking the transverse sinus for 80 minutes, bypass time was 137 minutes, the minimum temperature is $27.2^\circ C$. Bedside chest X-ray of the postoperatively of the first day showed: congenital heart disease postoperative changes seen and be seen intubation and pacing lead (**Figure 3**). The patient whose breathe with respirator assisted breathing for 4 days, after that, cardiopulmonary function recovered gradually, stopped using respirator, without arrhythmia, the heart function recovery, no facial edema, temporary pacemaker was removed on the tenth days after surgery. UCG of the two weeks after surgery showed: Changes after the partial endocardial cushion defect surgery (**Figure 4**). The patient leaved hospital after 14 days.

Discussion

Later in embryonic development, after left and right anterior cardinal vein anastomosis, left



Figure 3. Bedside chest X-ray of the postoperatively of the first day showed: congenital heart disease postoperative changes seen and be seen intubation and pacing lead.

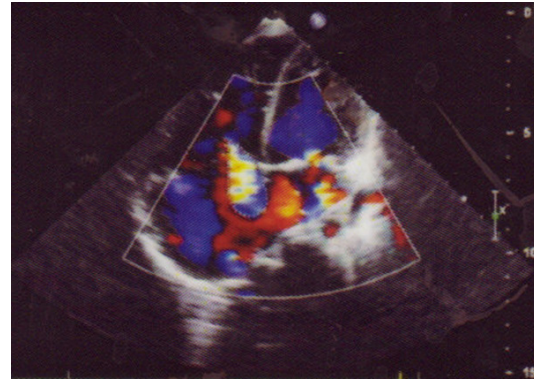


Figure 4. UCG showed: left atrium, right atrium, right ventricular enlargement, septal and left ventricular posterior wall was moved in the same direction. Atrial septal mended reflection enhancement and thickened, the lower edge of the patch be seen diameter of about 10 mm across left to right through the bloodstream. Septal echo is continuous and complete. Ventricular septum blood levels showed no wear. Mitral regurgitation systolic blood be seen, regurgitation beam area of 3.3 cm². Tricuspid regurgitation is seen systolic blood flow, regurgitation beam area of 3.5 cm².

front main vein degenerates into the Marshall ligament. Few people left and right anterior vein hypoplasia, left anterior vein developed into permanent left superior vena. Persistent left superior vena divided into four types [2]: 1) Isolated type, is characterized with extremely dilated coronary sinus, is a very rare type of them; 2) Left superior pulmonary vein returned to the top of left atrial, between auricula sinistra and pulmonary veins, without coronary sinus top, at this point, right superior vena cava often reduced or absent; 3) PLSVC returned to the right atrium alone, or PLSVC and superior vena cava returned to the right atrium together; 4) Rare cases, showing the permanent left superior vena connected to left atrium, and normal coronary sinus.

Persistent left superior vena diagnosis often need the help of chest enhanced Computed Tomography (CT), with or without 3-dimensional (3D) reconstruction, cardiac magnetic resonance imaging, ultrasonic cardiogram and other detection methods [3]. At the same time, the data shows, preoperative UCG often cannot probe PLSVC. This requires more attention to the different clinical signs of PLSVC, such as persistent hypoxemia, left atrial abnormality expansion and cyanosis [4]. Due to the PLSVC turns to the left atrium is right to left shunt, so

it needs operation to turn the bloodstream to the right atrium. Operation methods which are commonly used divided into three types: (1). Posterior left atrial wall sewn into the channel, turns PLSVC's bloodstream into the right atrium. (2). Supplemented with large pericardial or polyester film, separating and forming to right atrial inflow tract. (3). Anastomosed the PLSVC and right atrial, and other methods. These methods are that turning the PLSVC bloodstream to the right atrium, the operation is difficult, and the myocardial ischemic time is prolonged, while the effects pulmonary venous return in the future. So, the more exact method is still the PLSVC ligation, judging the indications of ligation are very important. As the right superior vena cava is well-developed, PLSVC is not too thick, it could be blocked in operation, Observing the left jugular venous pressure and head and neck venous reflux, if the pressure and (or) head and neck venous reflux is not blocked, then the PLSVC can be ligated [5-7].

This patient with multiple cardiac anomalies, rare in clinical, preoperative UCG only prompted detecting left superior vena cava, and failed to clear the drainage position. In operation, we repaired endocardial cushion defect, left superior vena cava cut-off experiments negative, ligated the PLSVC, restoring normal blood flow

dynamics. But it was a long history of this patient, preoperative heart function was poor, the postoperative effect still needs long-term follow-up observation. At present, there is no accurate results of the optimal operation timing which PLSVC turning into the left atrium, this may because of the less cases, and difficult to compare. But through the process of diagnosis and treatment of this case, we think that, PLSVC turning to left atrium with left to right shunt, when the patient can tolerate the operation, the patient should be surgical treated as soon as possible to restore normal hemodynamics, the greatest degree of recovery of cardiac function.

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Disclosure of conflict of interest

None.

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