Supraventricular Tachycardia in a Neonate with the Prenatal Diagnosis of a Single Left Superior Vena Cava


Persistence of the left superior vena cava (LSVC) in the absence of the right superior vena cava (RSVC) in a fetus with otherwise normal cardiac anatomy is usually not predictive of postnatal complications. A 37-year-old healthy primigravida was referred to our fetal echocardiography unit because of an abnormal three-vessel view (Figure 1) at the 20-week anomaly scan. Echocardiography revealed visceroatrial situs solitus with normal intracardiac anatomy. The innominate vein connected to a single LSVC, entering the dilated coronary sinus (CS) (Figure 2); the RSVC was not visible. The inferior caval vein drained into the right atrium. The ventricles, aortic valve and arch were of adequate size. No additional malformations were found. Amniocentesis was declined by the parents. Follow-up scans confirmed the suspected diagnosis and showed a normal rhythm.

A healthy girl was born at 39 + 1 weeks by normal vaginal delivery. At the age of 3 weeks the neonate presented at the Paediatric Cardiology Department. The prenatal diagnosis was confirmed by echocardiography. Unexpectedly, an atrioventricular re-entry tachycardia of 250 beats per minute was found, with a good peripheral perfusion. The tachycardia could be converted to sinus rhythm with the administration of intravenous adenosine. Electrocardiography demonstrated a normal sinus rhythm, without a pre-excitation pattern. Sotalol was administered for 2 months. No tachycardia occurred after discontinuation.

Persistence of the LSVC in the absence of the RSVC is very rare. An incidence of 0.09% was reported among a postmortem series of children with congenital heart disease.\(^1\) The expectation is that the incidence during fetal life is slightly higher if isolated cases, spontaneous fetal demise and fetuses with multiple malformations are taken into account.

A persistent LSVC can be associated with cardiac and extracardiac anomalies\(^2\)\(^-\)\(^4\), but in most cases it is an isolated anomaly.\(^5\)\(^-\)\(^6\) On prenatal ultrasound an LSVC presents with an abnormal three-vessel view, which is a transverse plane through the fetal thorax at the level of the pulmonary artery leaving the right ventricle.\(^3\)\(^-\)\(^5\) Usually the RSVC can be visualized in this plane at the right side of the ascending aorta. An LSVC is present at the left side of the pulmonary artery and generally drains into the CS, causing dilatation. A dilated coronary sinus is visible in the four-chamber view or in an oblique parasagittal plane as a ‘tobacco pipe’.\(^7\) The prognosis for an isolated single LSVC is good. It is an asymptomatic condition without hemodynamic significance.\(^5\)\(^-\)\(^6\) Associated arrhythmias have been reported in adulthood, whereas only one study reports on arrhythmias in childhood.\(^8\) The proposed mechanisms for the arrhythmia are stretching of the atrioventricular nodal tissue by the dilated CS, which can cause a re-entry tachycardia\(^9\), or an abnormal development of the sinus node in association with the absence of
the RSVC. A prospective study to determine the prevalence of arrhythmias in these children has not been performed and would be unlikely to be feasible considering the low incidence. This case illustrates that it is vital to remain attentive to arrhythmias when associated with single LSVC and that cardiac anomalies that were assumed to be fairly innocuous can have an unexpected outcome.

**Figure 1** Three-vessel view at 20 weeks of gestation demonstrating the superior vena cava on the left side of the pulmonary artery. AO, aorta; PA, pulmonary artery; SVC, superior vena cava.

**Figure 2** Four-chamber view at 20 weeks of gestation demonstrating a cross-section through the left superior vena cava (arrow) in its course towards the coronary sinus behind the left atrium.
REFERENCES


