

**Persistent left superior vena cava as marker of anomalous pulmonary venous connection in patients with isolated atrial septal defect: Descriptive analysis of 441 pediatric cases.**

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**Introduction:**

Atrial septal defects(ASD)are among the most common congenital heart diseases.The persistence of a left-sided superior vena cava(LSVC)is the most common variant of systemic venous drainage(0.5% of the general population, and up to 10% of those with established congenital heart disease),with a typical drainage to the coronary sinus.LSVC has been associated with an increased relation to atrial and ventricular septal defects,and several other malformations,and an increased risk of arrhythmias,most commonly atrial fibrillation in adult patients.We present 441 pediatric cases of isolated ASD and its association with anomaly of pulmonary venous connections and LSVC.

**Methods:**

Retrospective observational study of isolated ASD in pediatric population and a description of the pulmonary venous connections and LSVC,from January 2003 to December 2013.Anatomic and surgical characteristics are also described.

**Results:**

A total of 441 pediatric patients with isolated ASD were identified.Other heart congenital diseases were excluded.Range age was 1 month-18 years(mean 4,95),and data showed predominance in females(58%).Of these isolated ASD,had a surgical correction in 52%(232/441),and the other cases(209/441)had interventional catheterisation with a device or had a spontaneous closure.The most common reasons for the surgical correction were the anomalous pulmonary drainage and the impossibility to closure with a device due to anatomic incompatibility.Of all 441 isolated ASD,69 patients(15%)had a partially or totally pulmonary anomalous drainage.

LSVC was present in 19 cases(4,3%)and no arrhythmia has been described,but a 24h cardiac-Holter was not in the cardiac protocol when a LSVC was detected.Of these 19 ASD combined with LSVC,we found an anomalous pulmonary venous connection in 9 cases(47%,sinus venous ASD),and corresponding to right pulmonary veins draining to right atrium(5),right pulmonary veins draining to right superior vena cava next to the venous sinus(3),and left inferior pulmonary vein draining to inferior vena cava next to the venous sinus(1).In these 19 patients,and the others with a partially or totally pulmonary anomalous drainage,surgical correction of the venous drainage and the ASD with a pericardial patch was required.

**Conclusions:**

When an isolated ASD is related to LSVC,an anomalous pulmonary drainage is most frequently detected,and surgical repair will be required.Because the relation of LSVC to arrhythmias published in several reviews,a cardiac-Holter study seems recommendable in these patients and we added it at our protocol.