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 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.

LVSC has been associated with an increased relation to atrial and ventricular septal defects, and several other malformations, and an increased risk of arrhythmias.

We present 441 pediatric cases of isolated ASD and its association with anomalous pulmonary venous connection and LSVC.

RESULTS
A total of 441 pediatric patients with isolated ASD were identified. Range age was 1 month-18 years (mean 4.95).

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 catheterization 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%) of isolated ASD. No arrhythmias has been described, but a 24h cardiac-Holter was not in the cardiac protocol when a LSVC was detected. Of these 19 combined LSVC and ASD we found an anomalous pulmonary venous connection in 9 cases (47%). All 9 cases were venous sinus ASD and corresponding to right pulmonary veins draining to right atrium (5 cases), right pulmonary veins draining to right superior vena cava next to the venous sinus (3 cases), and left inferior pulmonary vein draining to inferior vena cava next to the venous sinus (1 case). Surgical correction of the venous drainage and the ASD with a pericardial patch was required in all 19 cases and the other with a partially or totally pulmonary anomalous drainage.

CONCLUSIONS
When an isolated ASD is related to LSVD, and anomalous pulmonary drainage is most frequently detected and surgical repair will be required.

Because of the relation of LSVD to arrhythmias published in several reviews, a 24h cardiac-Holter study seems recommendable in these patients and we added it at our protocol.