# Persistent left superior vena cava: does it have a role in the pathogenesis of hypoplastic left heart syndrome?


## Abstract

The coexistence of a persistent left superior vena cava (PLSVC) and congenital anomalies, both cardiac and noncardiac, is well documented, but whether PLSVC contributes to the development of cardiac malformations is controversial. We conducted a retrospective review of perinatal and pediatric autopsies to determine the association between PLSVC and other congenital anomalies. Of 362 patients, 91 (25%) had congenital heart disease and 19 (5.2%) had PLSVC. Eight cases (47%) were associated with specific syndromes, including heterotaxy syndrome, trisomy 18, trisomy 13, and Jacobsen syndrome. Seventeen cases of PLSVC (89%) were associated with congenital heart disease, most of which were complex. Isolated PLSVC was found in 2 cases (11%). Eight of the 19 PLSVC cases (47%) were associated with hypoplastic left heart syndrome (HLHS), a result that was statistically significant (P = 0.041). Left ventricle inflow/outflow obstruction is believed to be a critical pathogenic factor in the development of HLHS. Whereas 5 of 8 cases of HLHS had additional obstructive cardiac outflow tract lesions, 3 of 8 cases did not. PLSVC is known to be able to compromise left ventricle inflow via a dilated coronary sinus, and we speculate that PLSVC may have played a contributing role in the pathogenesis of HLHS in these three cases. As an isolated lesion, PLSVC would not be sufficient to cause HLHS, but it might contribute in combination with other obstructive lesions, or in the setting of other genetic and/or environmental factors still to be defined for HLHS. A larger series will be needed to confirm this hypothesis.

Taweevisit M, Thorner PS. Persistent left superior vena cava: does it have a role in the pathogenesis of hypoplastic left heart syndrome? Pediatr Dev Pathol. 2011 Mar-Apr;14(2):105-10. doi: 10.2350/10-05-0823-OA.1. Epub 2010 Aug 17. PMID: 20715969.