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# Persistent left superior vena cava without associated congenital heart defect

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## Objective

The aim of the study is to assess the association of persistent left superior vena cava (PLSVC) without congenital heart defect (CHD) with other extracardiac malformations, chromosomal / genetic abnormalities, perinatal outcome and long term outcome.

## Methods

This is an observational retrospective study performed in two tertiary centers (Hospital Universitari Vall Hebrón and Hospital Universitari Dexeus) in Barcelona between 2008 and 2018. All cases of PLSVC with no associated CHD and complete follow-up were included. The association with extracardiac fetal malformations, chromosomal or genetic anomalies, neonatal outcome and long term outcome was analyzed.

## Results

89 cases of PLSVC with no CHD were included (67 Hospital Universitari Vall Hebrón and 22 Hospital Universitari Dexeus). Median gestational age at diagnosis was 22 weeks (range: 16-39). 55% (49/89) were isolated PLSVC. The remaining 45% of cases had one or more associated anomalies or ultrasound findings: 12% (11/89) had a single umbilical artery, 12% (11/89) had IUGR (intra uterine growth restriction) and 11.2% (10/89) had associated extracardiac malformations. Karyotype and/or microarray analysis studies were performed in 57 cases and found 3.5% (2/57) pathogenic CNV and 3.5% (2/57) VOUS. No chromosomal abnormalities were found in fetuses with isolated PLSVC. 13% (12/89) had an absent right superior vena cava (ARSVC). All 49 cases of isolated PLSVC resulted in a livebirth and had a normal perinatal outcome. 12.5% (5/40) of cases with associated anomalies had an adverse perinatal outcome: 3 cases of termination of pregnancy, 2 neonatal deaths. Postnatal echocardiography found additional heart defects in 3 cases. Long-term outcome: there was 1 case of neurodevelopmental delay in a case with a pathogenic CNV.

## Conclusion

Isolated PLSVC has an excellent outcome. However, the association of other anomalies, including ultrasound markers, worsens the prognosis.