Hypoplastic left heart syndrome with prenatally diagnosed foramen ovale restriction: diagnosis, management and outcome.

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In up to 22% of patients with hypoplastic left heart syndrome (HLHS) restriction of the atrial septum develops at some point of gestation and often progresses until birth.

We carried out this study in order to determine survival predictors as well as possibly find out why restriction occurs in some HLHS patients and not others.

**OBJECTIVES**

- Earlier development and longer presence of foramen ovale restriction in the setting of HLHS was associated with higher short-term mortality regardless of the degree of restriction.
- VTI(f/r) ratio was not correlate with foramen ovale size and Vmax, and had not influence the survival rates
- Infection of the fetus should be considered as a potential risk factor of restriction development in patients with HLHS

**METHODS**

It was a retrospective analysis of prenatal history (with emphasis on parameters describing restriction - FO size and Vmax, pulmonary venous Doppler – VTI(f/r) and postnatal follow-up of 22 patients with HLHS and restriction. Restriction criteria of FO were: FO size of ≤ 4mm, and FO flow of >70cm/s with reverse pulmonary venous flow at the level of left atrium. Additionally, any signs of maternal or fetal infection were analyzed.

**RESULTS**

- There were 11 survivors and 11 non-survivors. Only 2 newborns required an emergent balloon atrioseptostomy immediately after birth. The most significant difference between the two groups pertained to the average time of restriction diagnosis which was 33hbd for survivors and 28hbd for non-survivors (p=0.0416) and the duration of in-utero restriction (9 weeks vs 5 weeks, p=0.0213).
- Additionally, 20 out of 22 patients exhibited cardiac and extracardiac anomalies which were unusual for HLHS itself but could have been a sign of an ongoing infection of the fetus (hydrops testis, tricuspid regurgitation, pericardial effusion, cardiomegaly, placentitis, hyperechogenic bowel/intestines/hepar/amniotic fluid, ultrasonographic features suggesting hepatitis) or there was a record of treated/untreated maternal infection.

**CONCLUSION**

1. Earlier development and longer presence of foramen ovale restriction in the setting of HLHS was associated with higher short-term mortality regardless of the degree of restriction.
2. VTI(f/r) ratio was not correlate with foramen ovale size and Vmax, and had not influence the survival rates
3. Infection of the fetus should be considered as a potential risk factor of restriction development in patients with HLHS