

Hypoplastic Left Heart Syndrome with Restrictive Atrial Septum: Echocardiographic Criteria for the Need of In-Utero Catheter Intervention and First Results of Human Fetal Atrial Stenting

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Background. Fetal pulmonary venous (PV) Doppler flow has been correlated with the severity of atrial septal restriction and the need for emergent atrial septostomy in newborns (NEAS) with hypoplastic left heart syndrome (HLHS). Despite NEAS, many infants with severely restrictive atrial septum (RAS) will eventually die from persistent severe pulmonary vascular disease.

Methods. To develop selection criteria for fetal intervention, we reviewed 67 HLHS fetuses with active postnatal care and studied the correlation between PV Doppler, NEAS at day 1, and survival to infancy. PV Doppler assessments included S-, D- and A-wave peak velocities (cm/s), A-wave duration (ms), and forward/reverse PV VTI ratios. Moreover, the outcome of four recent RAS fetuses with in-utero atrial stenting is reported.

Results: Four (6%) of the 67 cases without fetal intervention required NEAS. Two other fetuses with the same PV Doppler findings developed fetal hydrops and died during the last trimester. The table indicates the median (range) pre-delivery PV Doppler indices of HLHS cases with vs. without NEAS:

Parameters	NEAS (n=4)		No NEAS (n=63)		P-values
Fetal age (last echo; wks)	36	(35-38)	35	(28-39)	NS
S-wave (cm/s)	48	(25-56)	43	(15-91)	NS
D-wave (cm/s)	0	(0-11)	19	(8-52)	NS
A-wave (cm/s)	-43	(-19-57)	-20	(-1-72)	0.006
A-wave duration (ms)	107	(90-113)	65	(30-88)	<0.0001
Forward/reverse PV VTI	1.5	(1.3-2.3)	8.1	(1-50)	<0.0001

All NEAS cases had a forward/reverse PV VTI < 2.5 and PV A-wave duration >90 msec. This was the case in 0/63 (0%) cases without NEAS (p=0.0001; sensitivity 100%; specificity: 100%). The need of NEAS was associated with 0% survival during the first 2 months due to severe pulmonary vascular disease, while 83% without NEAS were alive (hazard ratio 7.7; p<0.001). Of 4 recent RAS cases with fetal atrial stenting at 27, 31, 32 and 35 weeks, 2 did not require NEAS, 4 underwent stage 1 surgery, and 3 are alive.

Conclusions: Two PV Doppler parameters (A-wave duration >90 ms; PV VTI <2.5) accurately predicted the need for NEAS, but none survived beyond early infancy. Fetal atrial septal stenting improved the postnatal outcome of our latest cases.