

MP2-5

Therapeutic Use of Transplacental Nonsteroidal Anti-inflammatory Drugs (NSAIDS) for Fetal Right Heart Failure with Circulatory Shunt

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Background: Circular shunt (CS) describes the disastrous pathophysiology resulting from a widely patent arterial duct (DA), severe pulmonary and tricuspid regurgitation and a failing right ventricle. In this instance, a significant amount of blood recirculates ineffectively between the ventricles, contributing to poor systemic perfusion, hydrops and death.

Methods: In three hydropic fetuses with Ebstein anomaly (EA) and CS, pharmacological DA constriction to birth, followed by immediate surgical pulmonary artery ligation and tricuspid valve closure was attempted. Treatment response to transplacental indomethacin (IND; loading: 100 mg; maintenance: 100-300mg/d) was defined as DA constriction significant enough to resolve fetal hydrops.

Results: The table summarizes the findings at baseline and with indomethacin. Prior to IND, all three fetuses had normal sized DA, continuous retrograde DA flow with low flow velocities, and absent or reversed end-diastolic umbilical artery (UA) and middle cerebral artery (MCA) flows. Case 1 showed no signs of improvement with IND for 9 days and died from postoperative complications. Cases 2-3 responded instantly to IND with DA constriction, normalized UA and MCA flows and resolution of hydrops. Case 2 was electively delivered at 32 weeks when hydrops recurred while on maximal IND. Sustained DA constriction to birth was achieved in case 3. Both survivors required postoperative ECMO and hemofiltration for transient respiratory and renal failure. They are currently well and awaiting second-stage single ventricle surgery at home.

Conclusions: This is the first report of the successful use of IND to manage the circulatory failure and delay the delivery and surgery of two fetuses with EA/CS. The long-term risks of a significant CS on the neurodevelopment and the use of IND on renal function should be further addressed in a prospective longitudinal study.

		Case 1	Case 2	Case 3
Gestational age (weeks)		30	26	30
DA diameter (mm)	Baseline	2.8	3.8	3.1
	IND	3	1.8	1.3
UA end-diastolic flow	Baseline	reversed	absent	reversed
	IND	reversed	anterograde	anterograde
MCA pulsatility index	Baseline	2.44	2.81	3.1
	IND	1.9	1.9	1.85
DA max/min flow (m/s)	Baseline	0.82/0.12	1.1/0.1	1.6/0.35
	IND	1.16/0.14	3.2/0.8	2.8/1.4
Treatment (days)	Total	12	49	42
Outcome		Died	Alive	Alive