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Supportive medical therapy for inter-stage of the Gießen Hybrid procedure.

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Objectives:

To report on our 5-year single-center experience with supportive medical therapy (SMT) for the Gießen hybrid procedure for complex congenital heart disease (CCHD).

Methods:

BLS (Bisoprolol, Lisinopril, Spironolactone) is the cornerstone of the Gießen SMT, which was developed by a pathophysiological driven concept addressing the neuro-humoral and cardiovascular effects of the specific flow pattern of postnatal parallel circulation i.e. I) blocking the inadequately activated neuro-humoral axis and lowering heart rate II) balancing systemic and pulmonary flow III) enhancing anti-fibrotic activity and promote cardiac remodeling. The dosage of B and L was in most patients 0.05 - 0.1mg/kg S 1-2mg/kg; all drugs given once daily. Diuretics were avoided in order not to further activate the neuro-humoral axis. Data was retro- and prospectively analysed (study period 12/2010 to 12/2015).

Results:

52 newborns underwent the "Gießen hybrid" procedure with uneventful surgical bilateral pulmonary artery banding (bPAB) and percutaneous arterial duct stenting for primary treatment of CCHD (hypoplastic left heart syndrome/complex (HLHS/HLHC), n = 30/16; others, n = 6) at a median age of 6 days (range 1 – 50) with a median weight of 3.0 kg (range 1.9 – 4.4; n = 9 ≤2500g) and a median Aristotle score of 17.0 (range 14.5 – 21.5). Prior to bPAB, 16 % of patients received bisoprolol, 4 % lisinopril, 12 % spironolactone, 24 % furosemide, 12 % hydrochlorothiazide. At discharge, 90 % of patients received bisoprolol, 73 % lisinopril, 78 % spironolactone, 18 % hydrochlorothiazide. The mean resting heart rate was reduced from 138/min at admission (range: 112 – 172/min; n=52) to 123/min at discharge (range: 99 – 139/min; n=51) to 110/min before stage II/biventricular repair/heart transplantation (range: 90 – 140/min; n=37). No side effects were observed; the parental compliance for treatment was excellent.

Conclusion:

SMT for the Gießen Hybrid procedure appears to effectively reduce full-body and myocardial oxygen consumption. In single cases reduction of the diastolic left-right flow across the stented duct could be monitored. Further prospective studies are warranted whether the pathophysiological concept sufficiently addresses the specific hemodynamics of the parallel circulation and in which agonistic and antagonistic neuro-endocrine parameters are affected.