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Population study of 332 consecutive newborns with HLHS - a single center experience

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BACKGROUND: Polish Mother's Memorial Hospital, Research Institute is one of the leading centers for hypoplastic left heart syndrome (HLHS) treatment in Poland.
AIM: The aim of the study was the estimation of perinatal and anatomic data of 332 newborns with HLHS operated at our institution.
MATERIAL AND METHODS: Retrospective analysis of perinatal data and anatomical findings of 332 newborns with HLHS (226 boys - 68% and 106 girls - 32%) treated by staged Norwood operation at our institution in 1992-2010.
RESULTS: Prenatal diagnosis was made in 58% of pts (68-75% in last 5 years), mean pregnancy duration was 39.4± 1.8 (31-43) weeks; 77.3% of pregnancies finished by vaginal births, 22.7% by cesarean section, mean mother's age was 26±5 (17-43) years, in 147 cases (44%) child with HLHS came from the first pregnancy. Mean Apgar score was 8.7±1.4 (1-10), mean birth mass was 3236±505.6g (1995-4430g). 10 patients (3%) came from twin pregnancies (in 1 case the second twin with TOF, in remaining 9 - the second twin was healthy). 5 patients (1.5%) had coexistent extracardiac malformations. In 4 cases (1.2%) our patient was the second child with HLHS of the same parents. In 6 families (1.8%) cardiac or extracardiac malformations in HLHS patient's siblings were confirmed (in 2 families TGA in sibling). In one patient Turner syndrome (45,X) was diagnosed.
Anatomic subtypes of HLHS: MA/AA in 123 pts (37%); MS/AA in 113 pts (34%); MS/AS in 93 pts (28%); MA/AS in 3 pts (1%). Myocardial performance index for right ventricle (RV- MPI) 0.521±0.18 (0.2-0.968) vs 0.3±0.078 (0.183-0.445) in control group of 50 healthy newborns. Restrictive atrial communication was confirmed in 33 pts (10%). Mean ascending aorta diameter was 3.8 mm (1-7.5mm). Severe tricuspid regurgitation was diagnosed in 40 patients (12%).
CONCLUSIONS: Patient with HLHS is usually male, good developed, full term delivery newborn of young mother. HLHS rarely coexists with other malformations or genetic disorders. In our material MA/AA, MS/AA, MS/AS subtypes occurred with similar frequency, MA/AS was very rare. RV-MPI for HLHS patients is significantly higher comparing with healthy neonates.