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Hypoplastic left heart syndrome: Intermediate results of a single center

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Introduction: Hypoplastic left heart syndrome (HLHS) is presently a most challenging congenital heart defect. Prenatal diagnosis and improved surgical technique have contributed to improve postnatal outcome, but mortality remains high. The aim of this study was to examine survival in short- and mid-term follow up in a cohort of patients with prenatal diagnosis of HLHS.

Methods: Retrospective descriptive study. Clinical records and echocardiographic clips of patients born in a 4-year period with HLHS were reviewed. Patients with prenatal diagnosis of HLHS and postnatal follow up were included, and those lost to follow up; with termination of pregnancy or who underwent fetal aortic valvuloplasty were excluded. Survival was evaluated in relation to cardiac surgery stages.

Results: Between 2011 and 2015; 18 patients with prenatal diagnosis of HLHS were assessed in our institution. Five were lost to follow up, and presumably underwent termination of pregnancy. One case was selected for fetal aortic valvuloplasty, and thus was referred to another hospital. Twelve babies were born in Fundación Hospitalaria, with no cases of preterm birth nor of low birth weight (<2500 gr). One patient with severe right lung hypoplasia was offered comfort care and died soon after birth. Another case had severe restrictive foramen ovale and died after Norwood surgery (NS). Ten of 11 cases (91%) survived the NS. Two patients (18%) died in the first interstage period; one of them had sudden cardiac death; and the other with right ventricle dysfunction and severe tricuspid regurgitation died after surgical repair of the tricuspid valve. One patient with a borderline left ventricle died after takedown and reconversion to biventricular circulation and 1 patient died after Fontan surgery. There are currently 7/11 (64%) patients alive, 6 in the second interstage period and 1 after Fontan surgery.

Conclusions: A cohort of patients with prenatal diagnosis of HLHS presented high survival rate after NS and significant mortality in the first interstage period. Continuing research and new strategies to improve systemic right ventricle and tricuspid function could further contribute to reduce patient mortality.

Biography

Sofia Grinenco, Medicine Doctor (MD), is a Pediatric Cardiologist member of the Fetal Medicine Unit at Hospital Italiano Buenos Aires, Professor of Fetal Cardiology at Fundación Hospitalaria. She is a Council Member of the Cardiology Committee of the Argentine Society of Pediatrics (SAP), member of the Argentinian Society of Prenatal Diagnosis and Therapy (SADIPT), member of the Association for European Pediatric and Congenital Cardiology (AEPC), with several post-graduate courses on Epidemiology and Statistics and on Clinical Bioethics. Currently her researches focus on optimizing prenatal diagnosis of congenital heart defects, and on these diseases' physiopathology and intrauterine treatment.

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