Clinical pictures of newborn infants who have been prenatally diagnosed with congenital heart disease but resulted in hospital mortality.

National Cerebral and Cardiovascular Center, Osaka, Japan

Introduction: It is expected that the prospective clinical care based on prenatal diagnosis using ultrasonography enables us to set up well-planned management and contributes to improvement of prognosis. While the prevalence of prenatally diagnosed congenital heart disease (CHD) has risen over the past decade, we experienced many severe cases die in early postnatal period. Purpose: We reviewed clinical pictures of newborn infants who have been prenatally diagnosed with CHD (includes fetal arrhythmia) but resulted in hospital mortality. Methods: Study population consisted of 89 consecutive newborn infants who were diagnosed with CHD (includes fetal arrhythmia) prenatally in our hospital between January 2011 and November 2012. We retrospectively analyzed the anatomical diagnoses, clinical course and cause of death in the patients who resulted in hospital mortality. Results: Most common fetal diagnoses were fetal arrhythmias (19%), variant hypoplastic left heart syndrome (HLHS) (11%), tetralogy of Fallot (9%), coarctation of aorta or interruption of the aortic arch (9%), single-ventricular lesions (8%) and heterotaxy syndrome (4.5%). Thirteen infants (14.6%) resulted in hospital mortality. Of those, the cause of death was perioperative mortality in 9 (69.2%), chromosomal abnormality (18 trisomy or 21 trisomy) in 3, and the other one died of metabolic disorder. Of the 9 cases who resulted in hospital mortality, the underlying anatomical diagnosis was HLHS in 4, Ebstein’s anomaly with circular shunt in 2, critical aortic stenosis (cAS) with endocardial fibroelastosis (EFE) in 1, pulmonary atresia with intact ventricular septum and cAS in 1 and remain was left atrial isomerism with congenital complete atrio-ventricular block (cAVB). Two cases with HLHS died of fatal capillary leakage syndrome. Conclusions: All the 13 (14.6%) infants resulted in hospital mortality had severe heart defect or systemic disorder. Especially in Ebstein’s anomaly with circular shunt, cAS with EFE and left atrial isomerism with congenital cAVB, the further development of new therapeutic methods or strategies are strongly desired in future. In severe heart defects, it is still difficult to save them even if they have been diagnosed prenatally.