

Congenital heart defects in twin pregnancies

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Background: The relative risk for congenital heart defects (CHD) is increased in monozygotic twins. The respective contribution of genetics of cardiac development and environmental risk factors such as twinning per se are scarcely known.

Aim: To analyze CHDs in twin pregnancies in which one or two fetuses were diagnosed to have a CHD and to identify the role of chorionicity and amnionicity on CHD type and concordance between twins.

Methods: 226 consecutive twin pregnancies with one or two fetuses with CHD were reviewed.

Chorionicity, amnionicity, twin-twin transfusion syndrome (TTTS), cardiac phenotypes, anatomical/CHD type (7 groups of CHD), concordance between twins and outcomes were analyzed.

Results: Pregnancies were bichorionic-biamniotic (BCBA) in 103 cases and monozygotic in 112 (biamniotic 92-MCBA, monoamniotic 20-MCMA) and the remaining 11 were unknown. Overall the 2 fetuses were affected in 35 cases (15.4%) with the two CHDs belonging to the same group in 23 (65.7%). The two fetuses had a CHD in 35% of MCMA, 14.1% of MCBA and in 10.7% of BCBA. The most frequent defects were conotruncal defects (n=71) with concordance in 13/71 (8/20 MCBA, 3/5 MCMA). For other groups concordance in all pregnancies, in MCBA and in MCMA were respectively : right outflow tract obstructions 5/48, 0/33, 0/0; obstructive left heart diseases 7/42, 2/13, 1/1; laterality defects 2/36, 0/6, 2/12; ventricular septal defects 5/19, 1/9, 0/0; atrioventricular septal defects 3/9, 1/2, 0/0; functionally univentricular heart 1/15, 0/5, 0/1. MCBA pregnancies were more frequent in right outflow tract obstructions (33/48; 69%-p<0.01) and discordance between twins was constant and was associated with proven TTTS in 22/33. MCMA were more frequent in laterality defects (12/36; 33%-p<0.01) and discordance between twin of was observed in 10/12. The outcome of pregnancy was : termination of pregnancies 47 fetuses, intrauterine foetal deaths 20 fetuses, neonatal death 24/387 live births. 116 patients underwent cardiac surgery within the first year of life with 13% mortality.

Conclusion: Concordance for CHD between twins is low even in monozygotic twins. Discordance in monozygotic twins might be related to twinning per se for laterality defects in MCMA and pulmonary stenosis in MCBA-TTTS.