Severe congenital heart disease: mortality and impact of prenatal diagnosis

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OBJECTIVES: describe mortality rate of patients diagnosed with severe congenital heart disease (CHD) in the prenatal and postnatal setting, compare prenatal and postnatal overall mortality and evaluate the impact of associated pathology.

METHODS: retrospective database research of 542 patients with severe CHD, diagnosed and/or treated with CHD at the University Hospital of Ghent (pre- and postnatal) or Antwerp (prenatal) between 01/01/2006 and 31/08/2014. CHD was divided in 11 pathologies: coarctation (CoAo, N124), tetralogy of Fallot (TOF, N118), univentricular heart defect (UVHD, N105), transposition of the great arteries (TGV, N74), atrioventricular defect (AVSD, N69), isomerism (N15), truncus arteriosus (N11), pulmonary atresia with intact interventricular septum (N10), double outlet with transposition (N9), Ebstein anomaly (N4) and double discordance (N3).

RESULTS: Overall, CoAo was most frequent (23%), followed by TOF (22%) and UVHD (19%). In 162 of 542 patients (30%), diagnosis was made prenatally. In the prenatally diagnosed group, UVHD accounted for 45%, TOF for 16% and TGA for 10%. Termination of pregnancy (TOP) was carried out in 43% of prenatal diagnoses, 67% being for UVHD. In the UVHD group, 11% of the fetuses had an associated genetic or structural abnormality, in contrast to 100% of fetuses with AVSD (6% of TOP).

Of all patients born alive (N=477), 87 had a prenatal diagnosis. The overall survival was 81% with higher survival rates for patients with TGA (71/74), CoAo (111/119) and AVSD (57/64). The overall mortality of patients born alive was 19%. Compassionate care was given in 25 patients (29% of postnatal deaths), of which 11 had TOF with significant additional pathology. Spontaneous pre-operative death was noted in 10 newborns. 432 patients underwent interventional treatment with an overall survival of 88%.

CONCLUSION: The overall survival after diagnosis of a severe CHD is 70%, with important differences between pathology groups. Despite organised prenatal screening, only 1/3 diagnoses is made prenatally. Prenatal diagnosis is associated with a high mortality due to the high incidence of TOP in our centre. Postnatal mortality is 19% with a high attribution of postnatal compassionate care, spontaneous pre-operative demise and late death related to non-cardiac causes.