Diagnosis, Management and Mid Term Outcome of Congenitally Corrected Transposition of the Great Arteries in the Fetus: a single centre experience

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Objectives: to describe the morphologic features and outcome of congenitally corrected transposition of the great arteries (ccTGA) as diagnosed during fetal life.

Background: since now few case series have been published regarding the natural and surgical history of prenatally diagnosed ccTGA

Methods: we included patients with a prenatal diagnosis of ccTGA between January 2005 and December 2015.

Results: 18 fetuses were diagnosed with ccTGA. Mean gestational age at diagnosis and at last fetal echocardiogram were respectively 23 weeks (17-33 weeks) and 35,5 weeks (±1,5). 17 fetuses were referred for suspicion of cardiac malformation, one for dextrocardia. Only 2/18 fetuses presented an isolated form of ccTGA. Associated cardiac defects included an abnormal cardiac position in 6/18, a ventricular septal defect (VSD) in 13/18, pulmonary atresia in 6/18 and pulmonary stenosis in 5/18, tricuspid dysplasia in 5/18, a right aortic arch in 2/18. Two fetuses developed a complete atrioventricular (AV) heart block in the third trimester. None of them presented associated extracardiac abnormalities. Karyotype was tested prenatally in 4 fetuses and resulted normal. Five (28%) pregnancies were interrupted whereas one fetus was lost at follow up. Mean gestational age at birth was 39 weeks, mean birth-weight 3200 g and mean oxygen saturation 95%. 9/12 patients required one or more surgical procedures. Among them, 2 completed the Fontan circulation, 4 received the anatomical repair (2 the Senning and Rastelli procedure and 2 the double switch operation). One received a bidirectional cavopulmonary connection. Both fetuses with complete AV block required a permanent pacemaker implantation at birth. At a mean follow up of 5,1 years (±3,3) 2/12 patients were lost at follow up whereas the others are alive and clinically well.

Conclusions: isolated cases of ccTGA as well as cases of ccTGA with extracardiac or chromosomal anomalies were rare prenatally. The mid term outcome was reasonably good for all liveborn infants even if most of them needed one or more cardiac treatment. Prenatal counselling remains challenging for the fetal cardiologist because fetuses with an apparently favourable condition may then develop major changes particularly regarding to tricuspid valve and cardiac rhythm with significant modification of prognosis.