Lessons from prenatal diagnosis and in utero transfer of fetuses with transposition of the great arteries

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Background: Prenatal diagnosis of transposition of the great arteries (TGA) reduces neonatal mortality and morbidity. The mortality of prenatally diagnosed TGA remains scarcely described.

Objective: To review the experience of prenatal diagnosis and in utero transfer of fetuses with TGA to assess the limits of this program and to describe outcomes of prenatally diagnosed TGA.

Methods: From 1999 to 2017, we reviewed all cases of prenatally diagnosed TGA born at our institution. Files were analyzed for discordances between prenatal and postnatal diagnosis, status at birth, neonatal management, and outcomes.

Results: During the study period, 748 fetuses with prenatal diagnosis of TGA (470 simple, 278 complex) were delivered at our institution and 333 neonates with post-natal diagnosis were referred after having been delivered elsewhere. Discordance between pre- and postnatal diagnosis was noted in 14.8% with two third having consequence on surgical treatment. Median term was 39 WG. The proportion of premature delivery (<37WG) was 5.3%, the majority being late-preterm (>34WG). 100 neonates were small for gestational age (<10th percentile); 10 from twin pregnancies and 10 with very low birth weight (<5th percentile). 14% were intubated in the delivery room and 1.7% required resuscitation maneuvers. 64% had a Rashkind procedure that was done in 15% of cases in the delivery room. 55% received PGE1 infusion and 38% had a Rashkind and PGE1 infusion. There were 9 deaths before surgery (1.2%) with two not related to the TGA (1 foeto-maternal hemorrhage and 1 polymalformation syndrome). A fourth of the cohort experienced neonatal complications including mainly respiratory distress requiring ventilation, infection, necrotizing enterocolitis, pulmonary hypertension and iatrogenic events. Surgical mortality was 1.8% (4 simple TGA and 10 complex). Seven additional deaths occurred during the first year follow-up, 6 being related to late cardiac complications. Overall survival at one year was 96%.

Conclusion: Prenatal diagnosis and in utero transfer of fetuses with TGA does not eliminate the risk of pre-operative mortality. The proportion of immediate distress is high and the need for intensive care in the delivery room is significant. Finally, preoperative mortality accounts for a third of the one-year mortality in this population.