

Congenitally corrected transposition of the great arteries: is it really a transposition?

*Arribard N. (1,2), Mostefa-Kara M. (1), Bonnet D. (3), Hascoët S. (1,2), Houyel L. (1,3)
Laboratory of Anatomy of Congenital Heart Disease - M3C, Marie Lannelongue Hospital, Le Plessis-Robinson, France (1); Laboratory of Surgical Research, INSERM U_999, Marie Lannelongue Hospital, Le Plessis Robinson, France (2); Medico-surgical Unit, Congenital and Paediatric Cardiology, Centre de Référence Malformations Cardiaques Congénitales Complexes - M3C, Necker Hospital for Sick Children, Paris, France (3)*

Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital malformation which associates atrioventricular discordance and ventriculo-arterial discordance. Although a ventricular septal defect (VSD) is frequently associated, its anatomy remains controversial. This could be due to the apparently different anatomy of the left-sided right ventricle (RV) compared to a right-sided RV. We wanted to compare the RV septal anatomy between ccTGA, transposition of the great arteries (TGA) and normal heart (NH) and to determine the anatomy of the VSD in ccTGA.

Methods: We analyzed 102 human heart specimens: 31 ccTGA, 36 TGA, 35 NH. According to the last classification of VSD (ICD 11), VSD were classified as outlet if located above the septal insertions of the tricuspid valve, inlet if underneath. We measured the lengths of the anterior (AL) and posterior (PL) limbs of the septal band and the angle between the two limbs. In order to assess the orientation of the septal band, we also measured the angle between AL and the arterial valve above (AL-AV).

Results: VSD was present in 26 ccTGA (83.9%) and was an outlet VSD in 17 cases (65.4%). Mean AL-PL angle was 76.4° for ccTGA compared to 90.6° for TGA ($p=0.011$) and 76.1° for NH ($p=ns$). Mean AL-AV was 70.6° for ccTGA compared to 90.6° for TGA ($p=0.0004$) and 69.1° for NH ($p=ns$). PL was significantly shorter in ccTGA ($p<0.0003$): AL/PL length ratio was 21.4 for ccTGA, 2.2 for TGA and 1.5 for NH.

Conclusion: The typical VSD in ccTGA is an outlet VSD. Its frequent misdiagnosis as an inlet VSD is due to the short PL, which creates the illusion of a posterior VSD. Surprisingly, the orientation of the septal band is similar in ccTGA and NH, despite the atrioventricular discordance, and different in ccTGA and TGA, despite the ventriculo-arterial discordance. Even if a transposition is present, ccTGA should not be considered a sub-group of TGA and the term "double discordance" might be more appropriate. These anatomic characteristics may also play a role in the dysfunction of the systemic RV, which occurs earlier in TGA post-atrial switch than in ccTGA.