

Pathologic anatomy of the coronary sinus and cardiac veins in double discordance (congenitally corrected transposition of the great arteries).

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Double discordance (DD) is a rare congenital heart defect (CHD) associating discordant atrioventricular and ventriculoarterial connections. Late prognosis in physiologically corrected or unoperated DD depends on the progressive failure of the systemic right ventricle (RV). A possible cause for systemic RV dysfunction could be ventricular dyssynchrony. Cardiac resynchronization therapy (CRT) may thus be indicated in some cases. However, the uncertainties about anatomy of the coronary sinus (CS) and cardiac veins often prevent the cardiologists from implanting a 3-lead CRT transvenous system in these patients.

Aim of the study: To evaluate the anatomy of the CS and cardiac veins in specimens with DD, in order to assess the feasibility of transvenous CRT.

Material and methods: Among the 1337 heart specimens available in the anatomic collection of the French Center of Reference for complex CHD, 22 had DD with 2 ventricles. Hearts were reviewed with special attention paid to the course and drainage of the CS and cardiac veins. Segmental anatomy, location of the VSD, status of the pulmonary outflow tract and anomalies of the atrioventricular valves were reviewed.

Results: Segmental anatomy was S,L,L in 20/22 hearts, S,L,D and I,D,D in 2. There was a VSD in 21/22, pulmonary atresia in 9, subpulmonary stenosis in 3, abnormal tricuspid valve in 17/22 including Ebstein anomaly in 6, straddling in 7. The CS was always located behind the morphologically left atrium (LA). However, its anatomy was normal, with normal drainage into the morphologically right atrium, in only 13/22 (59% of cases). The CS was of reduced length with normal orifice in 6. Orifice was atretic in 3 (1 completely absent CS with direct drainage of coronary veins into the LA, 2 with normal size CS). At least 1 available vein was found in all cases with patent CS orifice.

Conclusion: CS in DD is always located behind the morphologically LA. However, its anatomy is abnormal in 41% of cases. The most frequent anomalies are reduced length (27%) and atretic orifice (14%). This advocates the use of imaging techniques (multislice CT imaging or CS venography) before considering transvenous CRT in these patients.