A 50-year-old woman with a history of mild exertional dyspnea for several years was consulted for preoperative cardiovascular evaluation as a result of an abnormal ECG. Physical examination revealed a blood pressure of 154/90 mm Hg and a regular pulse rate of 84 beats per minute. Cardiac auscultation noted a grade 2/6 systolic murmur at the apex and the lower left sternal border. No peripheral cyanosis or clubbing of fingers was noted. Her resting ECG showed a sinus rhythm with a normal PR interval, left axis deviation, Q waves in the inferior leads, and left ventricular hypertrophy with ST-T change (Figure 1). Transthoracic echocardiographic segmental analysis of apical 4-chamber view revealed left atrium entered a morphological right ventricle and right atrium entered a morphological left ventricle (atrioventricular discordance; Movie I in the online-only Data Supplement). There was a moderate-to-severe tricuspid regurgitation from morphological right ventricle into left atrium (Movie II in the online-only Data Supplement). The long axis view showed the morphological left ventricle entering into a pulmonary artery with its left and right branches (Movie III in the online-only Data Supplement), and identified aorta by the presence of coronary artery from sinus of Valsalva which received the severely hypertrophied morphological right ventricle (ventriculoarterial discordance; Movie IV in the online-only Data Supplement). This double discordance resulted in a congenitally corrected transposition of the great arteries (ccTGA). Real-time 3-dimensional echocardiograms identified the morphological characteristic of the 4 chambers and great arteries (Figure 2, Movies V and VI in the online-only Data Supplement).

Cardiac multi-detector computed tomography scan revealed the anatomic left anterior descending and posterior
descending arteries in the interventricular groove. The anatomic left circumflex artery coursed in the right atrioventricular groove (Figure 3A). A single hypertrophied right coronary artery in the left atrioventricular groove perfused the systemic right ventricle (Figure 3B). The coronary sinus coursed along the left atrioventricular groove behind the systemic atrioventricular (tricuspid) valve.

Speckle tracking echocardiographic analysis of the systemic right ventricle free wall recorded a maximal longitudinal strain of −24% and a maximal circumferential strain of −23% (Figure 4). The mean left ventricular apical rotation was 3.0° counterclockwise (Movie VII in the online-only Data Supplement) and mean basal rotation was 1.6° clockwise. This resulted in a net twist of 4.6° (Figure 5). Continuous-wave Doppler interrogation of tricuspid regurgitation of the systemic right ventricle recorded a jet velocity of 5.7 m/sec (estimated right ventricular systolic pressure was 140 mm Hg) and a dP/dt of 1010 mm Hg/sec. A 3-dimensional slice plane view illustrated that the global contraction pattern of systemic right ventricle resembled that of a normal left ventricle (Movie VIII in the online-only Data Supplement).

The systemic right ventricle in ccTGA adapts to systemic load by a shift of contraction pattern from normal right ventricle to normal left ventricle. She was discharged after an uneventful hysterectomy of the uterine myoma and received angiotension receptor blocker antihypertensive treatment. ccTGA is a rare congenital anomaly in which a double discordance results the blood to flow in a normal direction but passes through the wrong ventricles. The morphological right ventricle supports the systemic circulation in a ccTGA. The systemic right ventricle adapts to the systemic load by a predominant increase in circumferential over longitudinal strain (normal left ventricular, −25.7±3.1% versus −16.5±1.7%; normal right ventricular, −15.8±1.3% versus −30.7±3.3%). In addition, the systemic right ventricle in this case displays a net twist, though in a much smaller magnitude than normal left ventricle. This net twist of the systemic right ventricle is not demonstrated in patients with the surgically corrected (Senning-operated) transposition of the great arteries. Cardiac multi-detector computed tomography scan showed the coronary sinus coursed along the left atrioventricular groove behind the systemic atroventricular (tricuspid) valve. This serves as an

**Figure 2.** A real-time 3-dimensional echocardiogram identified the morphological characteristic of the four chambers. Two black arrows identify the insertion of mitral and tricuspid valves to the septum. LA indicates left atrium; LAA, left atrial appendage; MB, moderator band; MLV, morphological left ventricle; MRV, morphological right ventricle; MV, mitral valve; RA, right atrium; and TV, tricuspid valve.

**Figure 3.** A, Cardiac multi-detector computed tomography (MDCT) scan revealed the anatomic left anterior descending and posterior descending arteries in the interventricular groove. The anatomic left circumflex artery coursed in the right atrioventricular groove. B, A single hypertrophied right coronary artery in the left atrioventricular groove perfused the systemic right ventricle. The coronary sinus coursed along the left atrioventricular groove behind the systemic atrioventricular (tricuspid) valve. CS indicates coronary sinus; LAD, left anterior descending artery; LCx, left circumflex artery; MLV, morphological left ventricle; MRV, morphological right ventricle; MV, mitral valve; PDA, posterior descending artery; RCA, right coronary artery; and RV, right ventricular branch.
anatomic landmark feasible for percutaneous annuloplasty for tricuspid regurgitation.

Disclosures
None.

References

Figure 4. Speckle tracking analysis of the systemic right ventricle free wall recorded a maximal longitudinal strain (A) of −24% and a maximal circumferential strain (B) of −23%.

Figure 5. The mean left ventricular apical rotation was 3.0° counterclockwise (A), and mean basal rotation was 1.6° clockwise (B). This resulted in a net twist of 4.6°.
Adaptation of the Systemic Right Ventricle in a Congenitally Corrected Transposition of the Great Arteries
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