CASE 5. Twisted heart with congenitally corrected transposition of the great arteries

- Source images: Non-ECG-gated contrast-enhanced MR angiograms.

Summary:
- Situs solitus / Levo cardia / Left aortic arch
- Discordant atrioventricular connection / Discordant ventriculoarterial connection
- Right ventricle, positioned superior and anterior to the Left ventricle
- Right and left ventricular apices pointing the opposite directions
- Non-parallel, twisted atrioventricular connection axes
- Aortic valve located directly anterior to the pulmonary valve
- Well-developed subaortic infundibulum. Very short subpulmonary infundibulum
- Long and narrow muscular subaortic outflow tract.
- Diffusely small aorta with severe tubular hypoplasia of the aortic arch and coarctation of the aorta. A large patent ductus arteriosus
- A large VSD between the inlet of the right ventricle and the outlet of the left ventricle.

Models (5 pieces):

Model 5A. Volume rendered images of the cardiac chambers viewed from anterior (upper left panel), anterior and inferior (upper right panel), left anterior (lower left panel), and left posterior (lower right panel) aspects. Cardiac valve annuli are marked with color.
Model 5B. Volume rendered images showing the interior of the right ventricle viewed from the front at different angles. VSD is marked with green dots.

Model 5C. Volume rendered image (left panel) showing the interior of the right and left ventricles viewed from behind. Corresponding photograph of the model with a plastic bar passed through the left ventricular outflow tract (LVOT). The VSD margin is marked with green dots.

Model 5D. Volume rendered image of the interior of the left ventricle.

Model 5E. Volume rendered image of the base of the ventricles. The VSD margin is marked with green dots.
Findings:

- There is atrial situs solitus. The systemic and pulmonary venous connections are normal.
- There is a superoinferior relationship of the major parts of the atria with the left atrium superior to the right atrium (5A, lower right panel). There is a secundum type defect (5D). The atrial septum is oriented in a slanted coronal plane.
- The right ventricle is superior and anterior to the left ventricle (5A-5D).
- The right atrium connects to the inferiorly located left ventricle. In contrast to the high position of the tricuspid valve within the right atrium in Cases 1 and 2, the mitral valve is located inferiorly and posteriorly to connect the right atrium to the inferiorly located left ventricle (5A, 5C).
- The opening axes of the tricuspid and mitral valves are not parallel. The mitral valve is oriented in a sagittal plane to open directly leftward (5C, 5E). The tricuspid valve is oriented obliquely to open forward and slightly leftward (5B-SE).
- The right ventricular apex points to the right, anterior and inferior, while the left ventricular apex points leftward and anterior (5A, 5B, 5D).
- There is a large VSD between the right ventricular inlet and the left ventricular outlet (5B-SE).
- The right ventricle is moderately hypoplastic.
- The right ventricular side of the ventricular septum accepts the palmar surface of the observer's left hand with the thumb in the inlet, the wrist on the apex and the fingers in the outlet. Therefore, there is a so-called left-hand pattern or chirality of the ventricular topology (5B, 5C).
- The aorta arises from the right ventricle through a long infundibulum. The pulmonary arterial trunk arises from the superior aspect of the left ventricle. The left ventricular outlet consists of a short muscular infundibulum with close proximity of the pulmonary and mitral valve annuli. The right ventricular inlet wraps around the left ventricular outlet (5B, 5C).
- The subaortic outflow tract is narrow and the aortic valve is small.
- The aortic valve is located directly anterior to the pulmonary valve. The aorta courses posteriorly, while the pulmonary trunk courses from the left to the right.
- The aortic arch shows severe tubular hypoplasia. A large patent ductus arteriosus connects the pulmonary arterial trunk to the descending aorta distal to the left subclavian arterial origin. There is focal coarctation due to a posterior shelf above the insertion of the ductus arteriosus to the aorta.

Comparison with Case 4:

Both cases have twisted atrioventricular discordant connection, Case 5 showing a higher degree of twisting than Case 4. Note that the right ventricular apex points leftward in Case 4, while it points rightward in Case 5. In Case 5, the right and left ventricular apices point in opposite directions.