CASE 10. So-called topsy-turvy heart with double outlet right ventricle

- Source images: ECG-gated contrast-enhanced CT angiograms.

Summary:
- Situs solitus / Levocardia / Left aortic arch
- Unusual superoinferior relationship of the cardiac chambers with the left atrium and left ventricle above and the major part of the right atrium and right ventricle below.
- Juxtaposition of the atrial appendages on the right side
- Large secundum atrial septal defect
- Concordant atrioventricular connection with parallel atrioventricular connection axes
- Right ventricular outflow tracts arising from the far posterior aspect of the right ventricle.
- Double outlet right ventricle through a muscular infundibulum that is evenly divided into the subaortic and subpulmonary outflow tract
- A large confluent VSD that is closer to the subaortic outflow tract
- Aortic valve located anterior and slightly to the right of the pulmonary valve
- Long ascending aorta and pulmonary arterial trunk taking acutely angulated proximal courses.
- Confluent pulmonary arteries with a tortuous patent ductus arteriosus.

Models (4 pieces):

Model 10A. Volume rendered images of the cardiac chambers with the cardiac valve annuli marked with color.
Findings:

- There is atrial situs solitus and levocardia. The systemic and pulmonary venous connections are normal.
- The right atrium is located to the right, anterior and inferior relative to the left atrium. There is right juxtaposition of the atrial appendages with the left atrial appendage being displaced to the right side and sitting above the right atrial appendage (10A). There is a large secundum type atrial septal defect.
- The four-chambers of the heart are superoinferiorly related with the left atrium and ventricle located on top of the right atrium and ventricle (10A and 10B). The atrial and ventricular septa are horizontally oriented. Note that the left atrial vestibular part is markedly elongated between the pulmonary venous confluence and the mitral valve (10D).
- There is a concordant atrioventricular connection (10A and 10B). Despite a superoinferior relationship between the ventricles, the opening axes of the tricuspid and mitral valves are parallel and the apices of the ventricles are pointing the same leftward direction.
- 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 left-sided apex and the fingers in the outlet. Therefore, there is a so-called left-hand pattern or chirality of the ventricular topology (10B and 10C).
- The right ventricular outlet is a completely muscular infundibulum projecting backward from the inferiorly located right ventricle (10A right upper panel). It is divided equally into the right-sided subaortic and left-sided subpulmonary outflow tract by the outlet septum.
- There is a large confluent VSD (10B-C). The VSD is closer to the aortic valve.
- The ascending aorta is superoinferiorly elongated as it arises from the inferiorly located right ventricle. It takes an acutely angled course in its proximal part (10A). The main pulmonary artery is also long and takes an angulated course.
- A long and tortuous patent ductus arteriosus connects the descending aorta to the top of the pulmonary arterial trunk (10A left lower panel).
Figure. Hypothetical pathogenesis of topsy-turvy ventricular relationship in this case. The usual orientation of cardiac chambers and great arterial trunks is shown on the top. With 90 degrees of organoaxial rotation of the cardiac chambers as seen from the apex of the ventricles, the atria and ventricles show a superoinferior relationship with the right atrium and right ventricle located above the left atrium and left ventricle. Further rotation to 180 degrees results in the left atrium and left ventricle lying in front of the right atrium and right ventricle. With 270 degrees of rotation, the left atrium and left ventricle are placed on top of the right atrium and right ventricle, respectively. As both arterial trunks leave the inferiorly located right ventricle, they are markedly elongated.