Double outlet right atrium (DORA) is a rare form of atrioventricular malalignment with dislocation of the left atrium (LA) and the atrial septum to the left and an absent connection of the LA to one of the ventricles.

We report about 2 neonates with DORA and AVSD. Common feature was an LA with regularly connected pulmonary veins, but absent connection to the left ventricle, whereby the LA drained exclusively via an ASD II into the right atrium (RA). Patient 1 showed an unbalanced ventricular morphology with a dominant left ventricle, pulmonary stenosis and left superior vena cava draining into the coronary sinus. Patient 2 presented with two balanced ventricles. Patient 1 was treated by univentricular palliation with BT-Shunt and closure of the pulmonary artery at the age of 8 weeks followed by a bidirectional Glenn-anastomosis at the age of six months. Patient 2 underwent a biventricular repair at the age of 2 months. This was accomplished by complete excision of the interatrial septum, patch repair of the AVSD and separation of the atria by an oblique patch, connecting the left atrium with the left ventricle.

Since DORA is the result of atrioventricular malalignment, patients with this malformation present either with two atrioventricular valves or as in our patients a common atrioventricular valve with a right and left portion. This is in contrast to mitral valve atresia and other forms of a absent left atrioventricular connection. The diagnosis is obtained by echocardiography. Since in patients with DORA the left atrium is able to drain its blood exclusively via an ASD, the issue of left atrial hypertension should be of major concern. Despite an absent left atrioventricular connection a biventricular repair is possible in selected patients with DORA. In our second case, who presented with two adequate size ventricles, corrective surgery was accomplished by oblique septation of the atria connecting the displaced left atrium with the left ventricle.