

How does right and left isomerism look like? A portrait of cardiac anomalies and visceral arrangement

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Objectives. To describe principal cardiac anomalies, and the arrangement of organs, in patients with right as opposed to left isomerism.

Methods and Results. We retrospectively reviewed data of patients diagnosed with so-called visceral heterotaxy at our Institutions in the last two years, including echocardiography in all patients and computed tomography angiography in the most complex cases.

We identified right isomerism (RI) in 24 patients (14 males). All had common atrioventricular junctions (AVSD), except for one with tetralogy of Fallot. AVSD was nearly always (85%) unbalanced with prevalent right-dominance. Right outflow obstruction was present in 19/24 cases (79%), and extracardiac anomalous pulmonary venous connections (APVC) in 13/24 (54%). The heart was right-sided in 10 cases (42%), midline in 1. The inferior caval vein (ICV) was left-sided in 9 cases (37%) and interrupted with azygos continuation in 1 case. The spleen was absent in all but 2 patients with a right-sided spleen, and 1 with left-sided multiple spleens, all having right bronchial isomerism. The liver was midline in 18/24 patients (75%).

We identified left isomerism (LI) in 15 patients (4 males). An AVSD with atrial and ventricular shunting was found in 8 cases (53%), and with only atrial shunting in 3 cases (20%). In addition, we observed 2 partial APVC (one associated to subaortic ventricular septal defect), 1 muscular defect and 1 supra-aortic stenosis. Right outflow obstruction was present in only 3/15 cases (20%). The heart was right-sided in 4 cases (27%). A right-sided ICV was noted in 1 case, while ICV was interrupted with azygos continuation in 10/15 cases (67%), and with hemiazygos continuation in 4 cases. Multiple left-sided spleens were found in 11 patients, multiple right-sided spleens in 2, and solitary spleens to the right and left each in 1 patient. All had left bronchial isomerism. The liver was midline in 11 of the patients (73%).

Conclusions. As expected, the majority of patients with RI had AVSDs guarded by a common valve, often with right outflow obstruction, while AVSD was less frequent in LI. Right-sided hearts were found in both subsets. Splenic morphology was not consistently associated with either form. These unexpected arrangements of the organs can be associated with equally unusual cardiac anomalies. The key is to analyse each system in its own rights.