Combination of the aortic interruption in a patient with right atrial isomerism: a previously unreported association

Sağ atriyal izomerizmli bir olguda aortik ark kesintisi: Daha önce bildirilmemiş birliktelik

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A 15-day-old male patient weighing 2.9 kg was referred to our clinic following the observation of dextrocardia on the chest X-ray at a health center consulted for tachypnea. Transthoracic echo-

cardiography evidenced right atrial isomerism, dextrocardia, complete atrioventricular septal defect (unbalanced toward the right), double-outlet right ventricle, supracardiac non-obstructive total anomalous pulmonary venous return via a right-sided vertical vein, left superior vena cava, Type B aortic interruption and pulmonary hypertension (Fig. A-C). A contrast-enhanced multidetector computerized tomography (256 Slices, Somatom Definition; Siemens Medical Solutions, Germany) was performed to evaluate the chest, confirming the echocardiography findings (Fig. D-F). Abdominal ultrasonography was performed and spleen not observed. His didn't have a view of syndromic and genetic examination was normal. Due to these findings, the patient underwent a bilateral pulmonary artery banding proce-



dure. The ongoing prostaglandin treatment was interrupted postoperatively; no need for a ductal stent was observed in the patient, who had no restriction of his ductus arteriosus. Major organ systems are characteristically right-sided in patients with right atrial isomerism. Abnormalities that may be seen in patients with right atrial isomerism include total anomalous pulmonary venous connection, absence of the coronary sinus, common atrioventricular valve, single ventricle, often with a right ventricular conformation and pulmonary atresia or pulmonary stenosis. While many patients with right atrial isomerism exhibit pulmonary atresia or stenosis, aortic atresia is relatively rare in such patients. Published literature does not include a case of right atrial isomerism concomitant with an aortic interruption, to the best of our knowledge. Aortic interruption in this case, in the absence of stenosis or atresia, appears to be an interesting co-occurrence.



Figures- (A-C) Transthoracic echocardiography showed dextrocardia, complete atrioventricular septal defect (unbalanced toward the right), supracardiac non-obstructive total anomalous pulmonary venous return via a right-sided vertical vein and left superior vena cava. (D-F) A contrastenhanced multidetector computerized tomography confirming the echocardiography findings. Ao: Aorta; CA: Common atrium; CAVV: Common atrioventricular valve; DAo: Descendent aorta: IV: Innominate vein: LLLPV: Left lower lob pulmonary vein; LSA: Left subclavian artery; LSVC: Left superior vena cava: LV: Left ventricle; LULPV: Left upper lob pulmonary vein; PA: Pulmonary artery; PDA: Patent ductus arteriosus; RLLPV: Right lower lob pulmonary vein; RV: Right ventricle; RULPV: Right upper lob pulmonary vein: VV: Vertical vein; *Interruption.