$ScvO_2$, %	=	66.5	74.6	73.8	
Pre-Oxygenator samples					
pCO_2, mm Hg	=	50.6	51.4	55.2	=
pO_2, mm Hg	=	46.5	48.8	46.5	=
sO_2, %	=	66.5	74.6	74.4	=
Post-Oxygenator samples					
pCO_2, mm Hg	=	31.2	32.6		=
pO_2, mm Hg	=	412.6	424		=
sO_2, %	=	100	100	100	=
BRF, % (PreOxy sO_2-ScO_2)/(PostOxy sO_2-ScO_2)x100	=	5.54	5.07	1.95	=

Patients BSA=1.73 m^2. All reported data are the mean of 3 consecutive measures.
ABE - base excess; BiPAP - bi-levels positive airways pressure; BRF - blood recirculation fraction with calculation formula (17); BSA - body surface area; CI - cardiac index; CPAP - continuous positive airways pressure; CV -controlled volume; CO - cardiac output; ECMO - extracorporeal membrane oxygenation; FiO_2 - fraction inspired oxygen; Hb - Hemoglobin concentration; Hb - Hemoglobin concentration; HCO_3 - bicarbonate concentration; MV - respiratory minute volume; Lac - lactate concentration; PEEP - positive end-expiratory pressure; PIP - peak inspiratory pressure; $paCO_2$ - arterial carbon dioxide tension; paO_2 - arterial oxygen tension; WBC - white blood cells; SO_2 - oxygen saturation; ctO_2 - Oxygen Content; $ScvO_2$ - central-venous oxygen saturation; TV - tidal volume

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Available Online Date: 18.12.2013

doi:10.5152/akd.2013.5139

Spontaneously regressed congenital idiopathic dilatation of the right atrium in the newborn

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Introduction

Idiopathic dilatation of the right atrium (IDRA) is an uncommon cardiac abnormality that consists of a disproportionately enlarged right atrium (RA) in the absence of any other anatomical pathology of the heart (1). Appropriate treatment of IDRA is controversial. Accurate diagnosis is necessary for proper medical and surgical management. Although long-term follow-up literature reports have been limited, the number of authors supporting RA surgical treatment is more than the ones supporting medical follow-up (2-5). Up to date, only few cases have been diagnosed. In this report we want to present an asymptomatic case with giant RA dilatation diagnosed by fetal echocardiography which was regressed spontaneously.

Case Report

Right atrial dilatation was seen in the fetal echocardiography of 18-week-old fetus and followed up till birth without having any rhythm problems. She was born at gestational 40 week weighting 3800 gr with spontaneous vaginal delivery. Her parents have a second degree relationship marriage with no history of known disease in the family. Clinical examination revealed tachypnea and mild respiratory distress but not cyanotic at postnatal first day. Auscultation revealed systolic murmur best heard at the left upper sternal edge. A 12-lead electro-

cardiogram showed sinus rhythm, right axis and an increase in the P wave's amplitude. A chest radiograph showed obvious cardiomegaly with normal pulmonary vascularity (Fig. 1). Echocardiography showed visceroatrial situs solitus, levocardia, excessive RA dilatation and it was seen that interatrial septum curved to left (Video 1. See corresponding video/movie images at www.anakarder.com). The RA minor and major axis in apical four chamber view were 31x33 mm. Moderate tricuspid regurgitation revealed (velocity is 3.8 m/sec) and there was not stenosis. Abnormal pulmonary venous return connection was not seen. Cardiovascular diseases associated with RA dilatation were excluded by multislice cardiac tomography and cardiac catheterization. Right atrial mean pressure was 14 mm Hg and left atrial pressure was 10 mm Hg in cardiac catheterization. Pulmonary artery pressure was 39/16 mm Hg, mean 25 mm Hg and aortic pressure was 74/47 mm Hg mean 65 mm Hg. During the injection of opaque matter to innominate vein, we detected that enlarged right atrium had became larger than right ventricle as a stomach shape (Video 2. See corresponding video/movie images at www.anakarder.com). The RA homogenous dilatation and IDRA diagnosis were verified by multislice cardiac tomography (Fig. 2). Dysrhythmia was not detected in 24 hours Holter monitoring.

After the patient was also assessed by cardiovascular surgeons, we decided that surgery would be more riskier than probable dysrhythmia and thromboembolic complications of current disease. Therefore, antiagregant treatment was given and patient was followed-up clinically. We did not encounter arrhythmia and progression of RA dilatation, so surgical intervention was not required. The patient is now one year old and asymptomatic with right atrial dilatation (23.7x29 mm) and tricuspid regurgitation obviously decreased (Fig. 3A, B).

Discussion

Idiopathic dilatation of the RA is a rare cardiac anomaly of unknown etiology. Since the initial description of IDRA in 1955, few cases have been reported in the medical literature, predominantly in adults (1-6). Hoffman et al. (7) claimed that a functional partial anomalous pulmonary venous insertion may imitate a structural abnormal pulmonary vein connection in etiopathogenesis of this anomaly.

The IDRA must be confirmed with the enlargement of the RA without having any other abnormalities like Ebstein's Anomaly, RA diverticula, aneurysms, restrictive cardiomyopathy, obstruction of the right outflow-inflow tract and total anomalous pulmonary venous connection to the right heart. It is usually difficult to differentiate IDRA from a right atrial aneurysm. The wall thickness of RA is homogenous in IDRA such as in our case. Conversely, there is fibrous, saccular region and a paper-thin-wall in congenital RA aneurysm (6-9).

Because of these potential risks (right heart failure, malign dysrhythmia, progressive dilatation in the RA and thromboembolic complications), primary surgical treatment is recommended. Therefore, primary surgical treatment was performed in the majority of the cases reported in the literature (2-9). Nonetheless medical follow-up was preferred in a few cases in which the patients were asymptomatic and there was no progression of the RA dilatation in echocardiography (1, 10). We suggest that in asymptomatic patients be followed up closely and conservative, rather than surgical, treatment be provided for probable complications. To our knowledge, this is the first case spontaneously regressed in the literature.

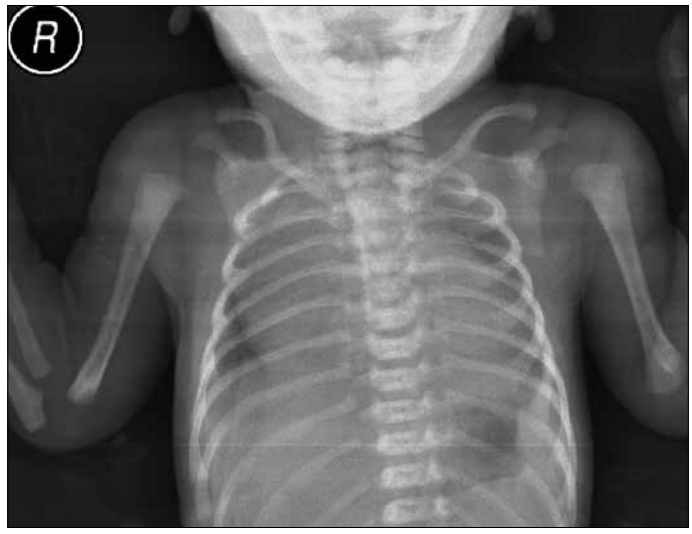

Figure 1. Chest radiography shows prominent cardiomegaly at newborn

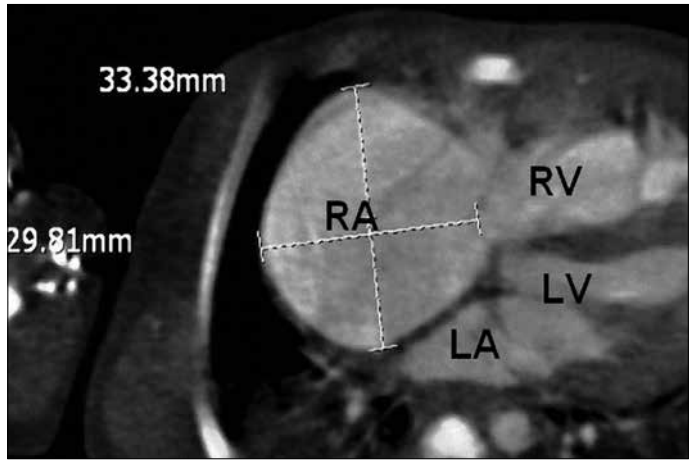

Figure 2. The multislice tomography imaging depicting excessive dilated RA, but normal LA, LV and RV in four chambers, other chambers without abnormalities

IDRA - idiopathic dilatation of the right atrium; LA - left atrium; LV - left ventricle; RA - right atrium; RV - right ventricle

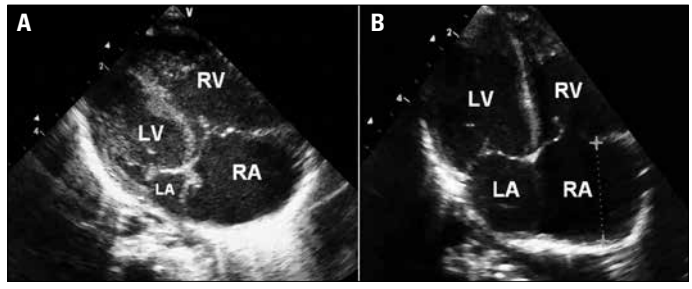

Figure 3. (A) Transthoracic echocardiogram (apical four-chamber view) revealed the right atrial dilatation (31x33 mm) caused by compression of the interatrial septum and tricuspid regurgitation and otherwise normal anatomy of the heart at newborn. Also, the presence of Ebstein's anomaly was excluded with normal tricuspid valve leaflets. (B) Transthoracic echocardiogram (apical four-chamber view) showed spontaneously improved of extreme right atrial dilatation and tricuspid regurgitation at 8 months old

Conclusion

Clinical presentation of this anomaly shows great variability and it is difficult to estimate the anatomical progression. Therefore, the treatment of the disease should be personalized and risk score should be formulated for the objective treatment decision but further studies are needed for this.

Video 1. Echocardiography showed that excessive right atrial dilatation without any tricuspid valve and cardiac anomalies
Video 2. During the injection of opaque matter to innominate vein, we detected that enlarged right atrium had became larger than right ventricle as a stomach shape

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This case was presented as a poster in the 11th National Pediatric Cardiology and Pediatric Cardiovascular Surgery Congres, May 2012, Izmir-Türkiye

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Available Online Date: 18.12.2013

doi:10.5152/akd.2013.5086

A child with L-2 hydroxyglutaric aciduria presenting with dilated cardiomyopathy: Coincidence or a new syndrome?

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Introduction

The etiology of dilated cardiomyopathy (DCM) is generally undetectable; its main feature is dilated ventricles of the heart. While metabolic disorders are among the etiologic factors (1), no patient with L-2 hydroxyglutaric aciduria (L2HGA) and DCM has been reported. We present a 16-year-old male under follow-up with DCM, who was subsequently diagnosed as L2HGA.

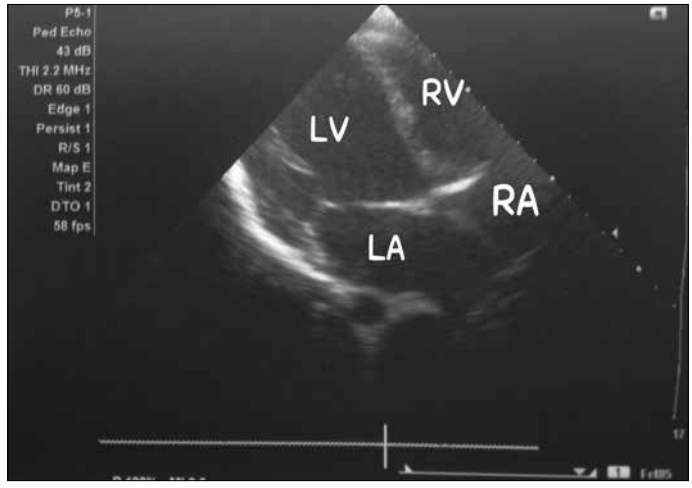

Figure 1. Two-dimensional echocardiogram showing a four-chamber view of the heart in a patient with systolic dysfunction. Note: dilated LV
LA - left atrium; LV - left ventricle; RA - right atrium; RV - right ventricle

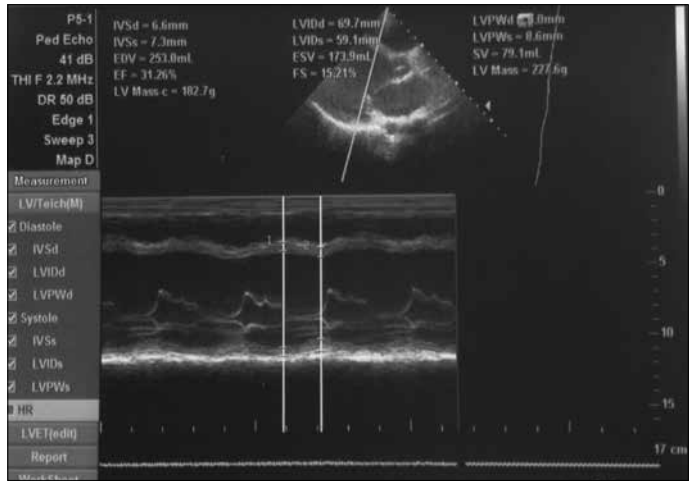

Figure 2. M-mode echocardiogram showing dilated left ventricle and decreased left ventricular contractility.