

Congenital Idiopathic Dilatation of the Right Atrium: Antenatal Appearance, Postnatal Management, Long-Term Follow-Up and Possible Pathomechanism

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Key Words

Aneurysm · Dilatation · Congenital · Right atrium ·
Echocardiography

Abstract

Introduction: Idiopathic dilatation of the right atrium (IDRA) is a rare abnormality usually detected by chance at any time between antenatal and adult life. It is defined as isolated enlargement of the right atrium in the absence of other cardiac lesions causing right atrial dilatation. IDRA can be associated with atrial arrhythmia and systemic embolism. The clinical presentation shows high variability ranging from the lack of any symptoms up to cardiac failure. **Methods/Results:** We describe 2 children with antenatally diagnosed IDRA, the intrauterine course in 1 case, the postnatal management and its long-term follow-up. There has been no need for surgical intervention so far because of the lack of arrhythmias and no further progression of right atrial diameters. Thrombus formation in the right atrium, which is a potential risk for pulmonary embolism, led us to initiate anticoagulation in our cases to prevent such complications. Furthermore, we suggest one possible pathomechanism of congenital right atrial dilatation. **Conclusion:** Optimal management of severe IDRA

depends on the individual case. Long-term follow-up of these patients is necessary to monitor a possible further progression of right atrial size and occurrence of arrhythmias. As a possible pathomechanism, a functional partial anomalous pulmonary venous insertion may imitate a structural abnormal pulmonary vein connection in some idiopathic cases of congenital right atrial dilatation.

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Introduction

So far, idiopathic dilatation of the right atrium (IDRA) is a congenital anomaly with unknown pathogenesis. It is defined as isolated enlargement of the right atrium (RA) in the absence of other cardiac lesions known to cause right atrial dilatation [1, 2]. There is no consensus on whether it is congenital or acquired [3]. Some prefer to call it idiopathic dilatation of the RA [1], while others label it as congenital enlargement of the RA [4] or congenital right atrial diverticulum [5]. There are clear characteristics distinguishing an aneurysm, 'as a dilatation of the atrium involving all layers of the atrial wall, from a diverticulum, an entry of fibromuscular strands across

the atrial wall' [6]. However, in the literature this clear distinction is not made continuously, as one can see in the definition by Chockalingam et al. [7]: 'an atrial aneurysm, or diverticulum, is an aneurysmal outpouching from an otherwise normal atrium'. Therefore, because of inconsistent terminology, the first description of IDRA was cited differently by Borrichin in 1676 [8], Semans and Taussig in 1938 (cited by Imren et al., 2006 [4]) and Morrow and Behrendt [9] in 1968. Bailey (cited by Forbes et al., 2007 [1]) was the first to excise a diverticulum of the RA.

The clinical spectrum ranges from the incidental finding of cardiomegaly on chest radiography to antenatal diagnosis because of significant atrial tachyarrhythmias. Most of the IDRA patients are asymptomatic. However, some patients develop arrhythmias or symptoms of congestive heart failure. Since there have been reports of significant symptoms and even sudden death [10], information about the management of this rare disease is essential to correct diagnosis, appropriate patient survey as well as treatment. We describe 2 children with antenatally diagnosed IDRA, providing long-term follow-up on our cases and suggesting a possible pathomechanism for this rare condition.

Methods

Two children with antenatally diagnosed IDRA, the intrauterine course in 1 case, the postnatal management and its long-term follow-up are described. Because of the lack of arrhythmias and no further progression of right atrial diameters so far, there has been no need for surgical intervention. Thrombus formation in the RA with the potential risk for pulmonary embolism has been described by others [9], leading us to initiate anticoagulation in our cases. Guided by an interesting intrauterine finding in case 1, we suggest a possible pathomechanism of congenital right atrial dilatation.

Case 1

A 21-year-old gravida 1 para 0 was referred to our outpatient clinic for second-trimester screening at 20 + 5 weeks of gestation. Her medical and obstetrical history was uneventful. Family history did not show any cardiac diseases or chromosomal abnormalities. There have been no extracardiac malformations. Intrauterine fetal growth and well-being parameters appeared normal. Echocardiography of the fetus showed an abnormal 4-chamber view (4CV; fig. 1a) with an increased cardiothoracic circumference ratio of 0.571 and an especially enlarged RA. During the intrauterine course the known RA dilatation persisted (fig. 1a). In addition, we were able to demonstrate an abnormal left-to-right (L-R) shunt across the foramen ovale demonstrated in figure 1b. A female infant was delivered at term (gestational age 39 + 0 weeks) via spontaneous vaginal delivery with a birth weight of 3,020 g (Apgar 8/9/9). Postnatal two-dimensional transthoracic

Table 1. Parameters measured during cardiac catheterization in case 1 at the age of 10 months

	Pressure, mm Hg			SaO ₂ %
	middle	systolic/diastolic -end-diastolic	systolic/diastolic -middle	
RA	2			
RV		20/0-2		
LA	4			
LV		85/0-7		
PA			15/5-9	
SVC				70.5
IVC				81.4
Q _p :Q _s = 1.0				

RA = Right atrium; RV = right ventricle; LA = left atrium; LV = left ventricle; PA = pulmonary artery; SVC = superior vena cava; IVC = inferior vena cava; Q_p:Q_s = ratio of pulmonary blood flow (Q_p) to systemic blood flow (Q_s).

echocardiography demonstrated that the RA was dilated massively with low blood flow velocity and spontaneous contrast within the cavity but without signs of intracardiac thrombus formation (fig. 1c). Thrombosis prophylaxis with salicylic acid was started. Alongside a patent foramen ovale with persistent L-R shunt, the remainder of the intracardiac anatomy, including the tricuspid valve, was normal. Particularly, there was no tricuspid regurgitation noted in ante- or postnatal echocardiograms.

At 4 weeks, the child remained well (body weight 3,970 g) and the echocardiogram was unchanged, showing the RA with an area of 10 cm² (compared to left atrial area of 3.2 cm²) (fig. 1d). Chest radiography disclosed an enlarged cardiac silhouette due to an enlarged RA (not shown). At the age of 4 months the RA area was 18 cm² (fig. 1d). The electrocardiogram (ECG) showed sinus rhythm. At 10 months the RA area increased to 24 cm² and a cardiac catheterization (Heart Center, University of Leipzig, Germany) confirmed the diagnosis of isolated and IDRA, showing no further intracardiac abnormalities (fig. 1e), especially no anatomical partial anomalous pulmonary venous connection. The pulmonary artery pressure measured during cardiac catheterization was 15/5-9 mm Hg (systolic/diastolic-middle). There was no measurable shunt through the small patent foramen ovale (Q_p:Q_s = 1.0) (table 1).

Between ages 2 and 4, the RA area stayed relatively stable in relation to normal body growth, totaling 34 cm² now (fig. 1d). Subsequent Holter monitoring showed sinus rhythm and no atrial arrhythmia. The child's growth, development, and exercise tolerance have all remained normal. The child is now 4 years of age without any clinical symptoms.

Case 2

Diagnosis of RA dilatation was made during fetal life by echocardiography (in a different hospital). Postnatal two-dimensional echocardiogram at the age of 2 weeks confirmed the presence of

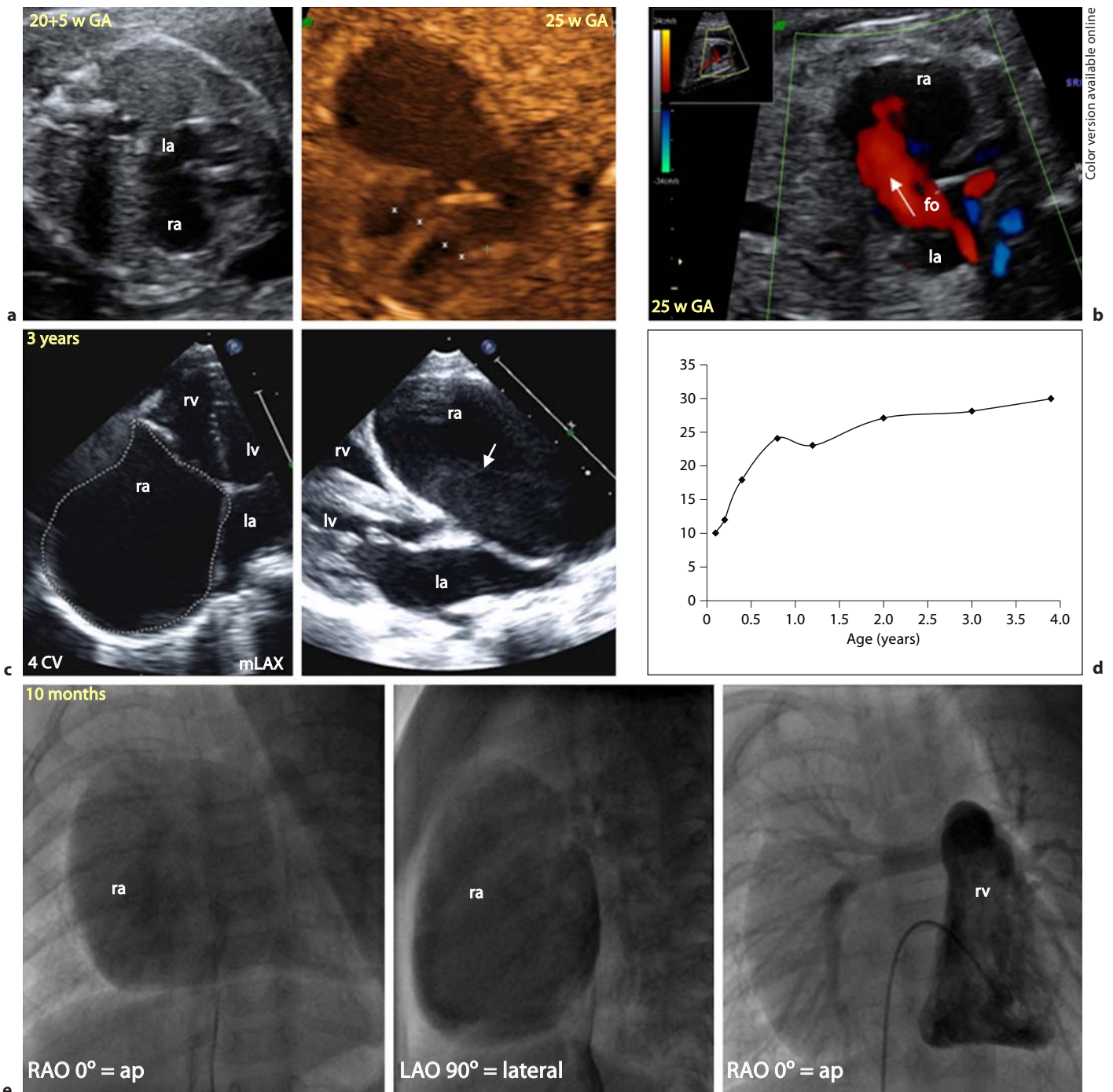


Fig. 1. Case 1. **a** Prenatal echocardiography at 20+5 weeks of gestation (w GA) showing the abnormal 4CV with an increased cardiothoracic circumference ratio of 0.571 due to massive right atrial dilatation and intrauterine course. **b** Abnormal L-R shunt across the foramen ovale (white arrow) shown at 25 weeks of gestation. **c** Postnatal transthoracic echocardiography at the age of 3 years. 4CV and modified long axis (mLAX) of the heart demonstrating a very large RA, normal right ventricle and normal left-sided chambers. Spontaneous echo contrast (white arrow) was present within the dilated RA as a sign of low flow. **d** Diagram

showing the right atrial dimension (cm²) over a 4-year period. There is no evidence of further progressive right atrial enlargement. The y-axis shows the right atrial dimension in cm². **e** X-ray during cardiac catheterization at the age of 10 months excluding further intracardiac anomalies. Contrast image of the RA in anterior-posterior (ap) projection (left panel), lateral projection (middle panel), and contrast image of the right ventricle (rv) in ap projection on the right panel. ra = Right atrium (RA); rv = right ventricle; la = left atrium; lv = left ventricle; fo = foramen ovale.

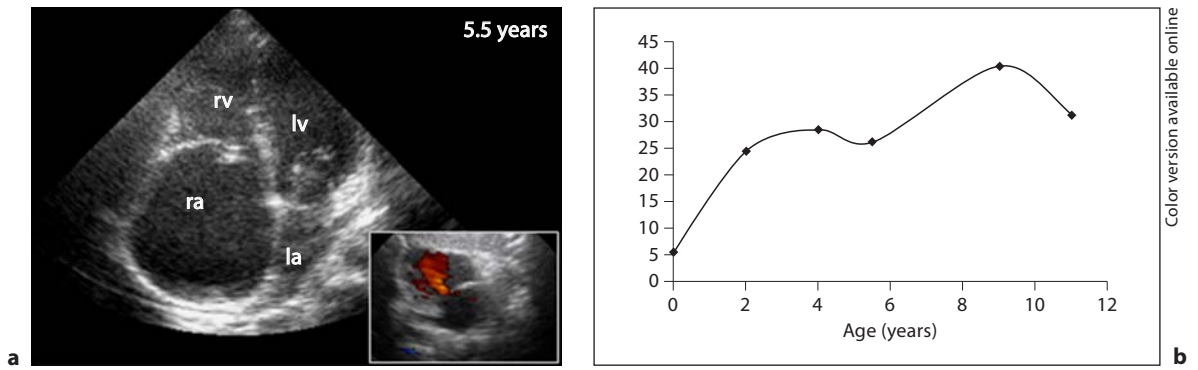


Fig. 2. Case 2. **a** Transthoracic echocardiography at the age of 5.5 years (4CV of the heart) showing cardiomegaly due to right atrial dilatation. **b** Diagram showing the right atrial dimension (cm²) over a 7-year period. The size has been measured two-dimensionally in the apical 4CV and by planimetry. The y-axis shows the planimetered right atrial dimension in cm². ra = Right atrium (RA); rv = right ventricle; la = left atrium; lv = left ventricle.

Table 2. Measurements of right atrial size including major-axis and minor-axis lengths and planimetered areas in 4CV (at end-systole) for both patients

	Age years	Right atrial dimension			BW kg	BL cm	BSA m ² (Haycock)
		major-axis cm	minor-axis cm	planimetered area (4CV), cm ²			
Patient 1	0.1	3.5	2.85	10	3.97	50	0.24
	0.2	3.8	3.7	12	5.4	56	0.3
	0.4	4.5	3.8	18	7.5	66	0.38
	0.8	5.8	4.2	24	9.49	72.8	0.45
	1.20	5.7	4.1	23	10.10	77.5	0.47
	2	5.8	4.6	27	12	86	0.54
	3	5.9	4.85	28	14.5	97	0.63
	3.5	6.2	5.4	34	15.2	100	0.65
4	6.2	5.4	34	17.4	104	0.71	
Patient 2	0	2.6	2.2	5.72	3.8	52	0.24
	2	5.3	4.6	24.38	15	97.5	0.64
	4	5.5	5.2	28.6	18	107	0.73
	5.50	5.7	4.6	26.22	21.6	117	0.84
	9	6.6	6.1	40.26	34	139	1.14
	11	5.8	5.4	31.32	42	152	1.33

4CV = Four-chamber view; BSA = body surface area; BW = body weight; BL = body length.

a large RA (26 × 22 mm). The female child was referred to our hospital at the age of 4. Transthoracic echocardiography at this age showed the known massively dilated RA measuring 55 × 52 mm in the anterior-posterior plane (fig. 2b; table 2). There was only mild regurgitation of a normal tricuspid valve. There was a small fenestrated atrial septal defect from the secundum type with small L-R shunt, which persisted over the years (fig. 2a). Ventricular size and function were normal, as was the remainder of the cardiac anatomy. At the age of 5.5 years the RA dimension was

almost unchanged (57 × 46 mm; fig. 2a, b; table 2). Holter monitoring showed normal ECG, and ergometry was normal. At the age of 9, the RA dimension was 66 × 61 mm (left atrium 46 × 27 mm; fig. 2b; table 2). Two years later, at the age of 11, the RA dimension showed slight regression (54 × 58 mm) in the 4CV (fig. 2b; table 2). There was no sign of cardiac failure. The physical strain remained normal and subsequent ECGs did not show any abnormalities.

Discussion

Congenital IDRA is a rare condition, reported in fetuses [11, 12], children [1, 13], infants [1] and in adults [14]. There are only 31 reports in the literature about IDRA or congenital RA aneurysm. Only 12 out of these 31 reports describe cases which have been detected antenatally, in newborns or at the stage of early infancy [1, 4, 11, 13, 15–24].

It is difficult to estimate the true incidence of the disease, since most children are asymptomatic [13] and diagnosis is often made incidentally due to cardiomegaly on a chest X-ray. When symptoms are present, they may include palpitations, arrhythmia, non-specific chest pain, dyspnea, fatigue, and syncope.

In some reports, IDRA is claimed to be a benign condition [13] without reduction of expectation of life, but others suggest inevitable progression to right heart failure. The resulting atrial arrhythmias evolve in a recurring way, increasing the potential risk of systemic or pulmonary embolism. Blondheim et al. [8] hypothesize ‘that there may be two types of IDRA: one secondary to a degenerative process of unknown etiology affecting the atrial myocardium’ (apoptotic process), the other ‘a congenital absence of atrial myocardium and perhaps conduction tissue’. The first condition is claimed to have a more benign course, while the second may have associated conduction defects and poor long-term prognosis, including sudden death [25].

Interestingly, in our first described case we were able to show antenatally an abnormal L-R shunt over the foramen ovale (fig. 1b), guiding us to the working diagnosis ‘functional’ partial abnormal pulmonary venous connection. This means that the pulmonary veins are anatomically normally connected to the left atrium. However, there might be an uncommon angle of pulmonary vein insertion leading to a blood flow through the fossa ovalis. This concept of abnormal insertion angle of veins into the heart has already been proposed for fetuses with left-sided liver-up congenital diaphragmatic hernia [26]. Postnatally, there was also a L-R shunt through the foramen ovale, which is very often seen in completely normal newborns. However, in combination with the antenatal images, it might be a possible explanation of the right atrial dilatation. Postnatally, after the shunt through the interatrial septum got smaller, the RA size stayed stable in relation to body growth (fig. 1d). In the second case, similar findings are visible postnatally (fig. 2). However, assumptions are difficult to make due to the missing antenatal data.

Because of its rare occurrence, IDRA may easily be confused with other anomalies that lead to RA enlargement, such as Ebstein’s anomaly [3, 27]. Transthoracic echocardiography yields a definite diagnosis in most cases, as it did in our cases. Nevertheless, transesophageal echocardiography, cardiac catheterization, computed tomography, and cardiac magnetic resonance imaging are other imaging modalities that might be helpful. Transesophageal echocardiography is particularly useful for detecting thrombus in the RA. However, there is a small risk of aneurysmal rupture using this invasive technique [4].

Surgical resection, anticoagulation, catheter-based ablation, and watchful waiting without medical intervention, are therapeutic options for RA enlargement and associated tachyarrhythmia. Patients with arrhythmias have been treated successfully with right reduction atrio-plasty, but the arrhythmias may recur [3]. It still remains controversial whether to excise the aneurysm or not [3, 28]. Some patients manage quite well surgically and others non-surgically [3]. Thrombus formation in the RA, which is a potential risk for pulmonary embolism, has been reported [9]. Therefore, anticoagulant therapy should be considered upon diagnosis of IDRA in order to prevent such complications.

Only in case 1 was an additional cardiac catheterization performed to exclude any further cardiac anomalies, such as a partial abnormal pulmonary venous return to the right heart. Lacking arrhythmias and progression in RA diameters, there has been no need for surgical intervention. Nevertheless, long-term follow-up of these patients with repeated Holter ECG monitoring is necessary to monitor a possible further progressive RA dilatation and occurrence of arrhythmias.

Conclusion

In conclusion, IDRA is a rare congenital heart disease. The clinical presentation shows high variability, ranging from the lack of any symptoms to cardiac failure. Complications are edema, arrhythmia or congestive heart failure, as well as systemic embolism. We described 1 case which might explain the pathophysiology in some idiopathic cases of congenital RA dilatation. ‘Functional’ partial abnormal pulmonary venous connection, described as an uncommon angle of pulmonary vein insertion into the left atrium, may imitate an anatomically incorrect pulmonary venous connection into right heart structures.

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