Progress in Pediatric Cardiology 52 (2019) 57–59

Contents lists available at ScienceDirect

Progress in Pediatric Cardiology

journal homepage: www.elsevier.com/locate/ppedcard

Isolated right ventricular non-compaction in a newborn

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ARTICLE INFO

Keywords:
Isolated non - compaction of the right ventricle
Echocardiogram
Cardiac magnetic resonance imaging
Newborn

ABSTRACT

Isolated right ventricular non - compaction is a rare congenital cardiomyopathy resulting from an arrest in normal endomyocardial embryogenesis. It is characterized by excessive prominent trabeculations and deep inter - trabecular recesses in the ventricular wall. The clinical syndrome includes systolic and diastolic dysfunction, heart failure, ventricular arrhythmias and cardioembolic events. Here we describe a newborn with isolated right ventricular non -compaction who had right heart failure and biventricular systolic dysfunction. Echocardiography and cardiac magnetic resonance imaging demonstrated the morphological abnormalities of the right ventricle.

1. Introduction

World Health Organization/International and Federation of Cardiology Task Force categorized non- compaction of the ventricular myocardium as an “unclassified cardiomyopathy” in 1995. American Heart Association in its scientific statement in 2006 classified it under the “genetic” section of “primary cardiomyopathies (1). Its prevalence is 0.14% in the paediatric population with a male to female ratio of 5.7 to 1.2. The usual site of involvement is the left ventricle. Involvement of both ventricles or isolated non - compaction of the right ventricle has been described rarely (3).

2. Case Report

A baby girl was admitted to the neonatal intensive care unit with cyanosis and respiratory distress at day 3 of life. She was delivered via spontaneous vertex delivery at 37 weeks with good Apgar scores and a birth weight of 3.12 kg. Her foetal echocardiogram revealed dilated right atrium and ventricle with mild tricuspid regurgitation. There was no family history of congenital heart disease. She was not dysmorphic, oxygen saturation was 85%–88% on room air, heart rate of 150 beats per minute and respiratory rate of 65 breaths per minute. Cardiovascular examination revealed regular pulses, equal femoral pulses, apex beat at the 4th intercostal space midclavicular line and normal heart sounds.

Chest radiograph showed cardiomegaly. Electrocardiogram showed ST segment elevation in the inferior leads. Her echocardiogram showed a dilated right ventricle with right ventricular hypertrophy (Fig. 1). Deep trabeculations were seen in the right ventricular wall and apex with blood flow in the deep recesses (Fig. 2). The non-compacted/compacted ratio was 2.90. Tricuspid annular plannar systolic excursion was 1.9 mm. There was mild tricuspid regurgitation with a peak pressure gradient of 24 mmHg. Left ventricular ejection fraction was 48%. The shunt through the foramen ovale was right to left. Cardiac magnetic resonance imaging showed a mildly dilated right atrium and grossly dilated right ventricle. The right ventricle myocardium was slightly thickened. There was thinning at the right ventricle apex and inferior wall with multiple trabeculations and septations within the right ventricle apex (Fig. 3). There was a 2 - layered appearance of myocardium in these regions, the inner layer appeared more hyperintense and communicated with the right ventricle cavity. Inborn error of metabolism screening was normal.

She required continuous positive airway pressure ventilation for 1 day and oxygen therapy via nasal cannula for 15 days. Her oxygen saturation improved to above 95% on non-invasive ventilation and nasal cannula oxygen. She was initially treated for right heart failure with frusemide and captopril. Frusemide was changed to hydrochlorothizade due to increasing urea level. At day ten of life, echocardiogram showed improvement of biventricular systolic function. Tricuspid annular plannar systolic excursion increased to 2.7 mm and left ventricular ejection fraction to 79%. At discharge at day 18 of life, she was comfortable and not in respiratory distress or overt cardiac failure. She was breastfeeding well.

She has gained weight to 4.9 kg at 3 months old at her latest outpatient visit and has remained well at home. Cardiovascular examination was unremarkable. The ST segment elevation in the inferior leads has resolved. Her latest echocardiogram showed normal dimension of the right ventricle, tricuspid annular plannar systolic excursion of

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https://doi.org/10.1016/j.ppedcard.2018.08.008
Received 5 March 2018; Received in revised form 21 August 2018; Accepted 27 August 2018
Available online 24 September 2018
1058-9813/

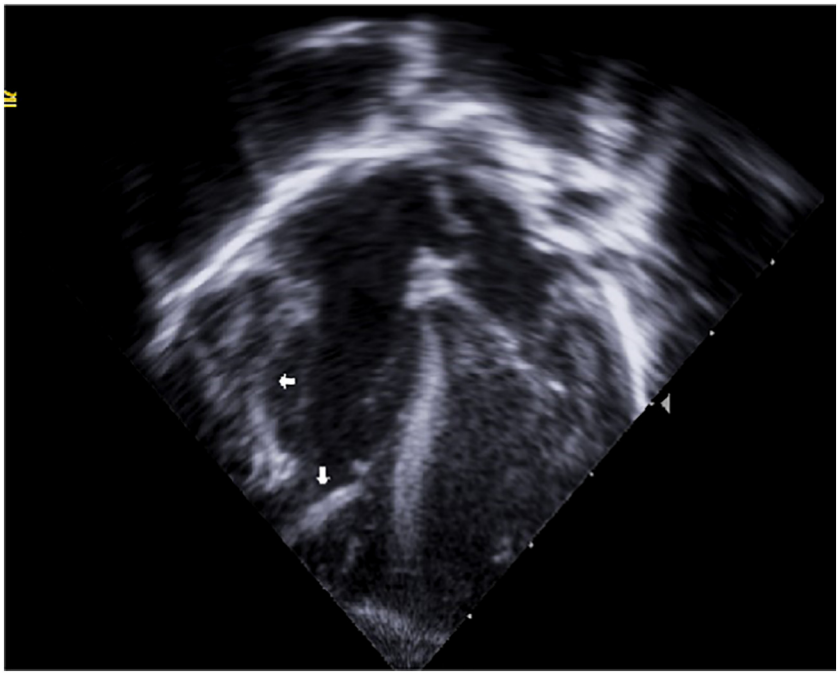

Fig. 1. Echocardiogram showing dilated right ventricle, hypertrophied right ventricle and deep trabeculations in the right ventricle wall and apex.

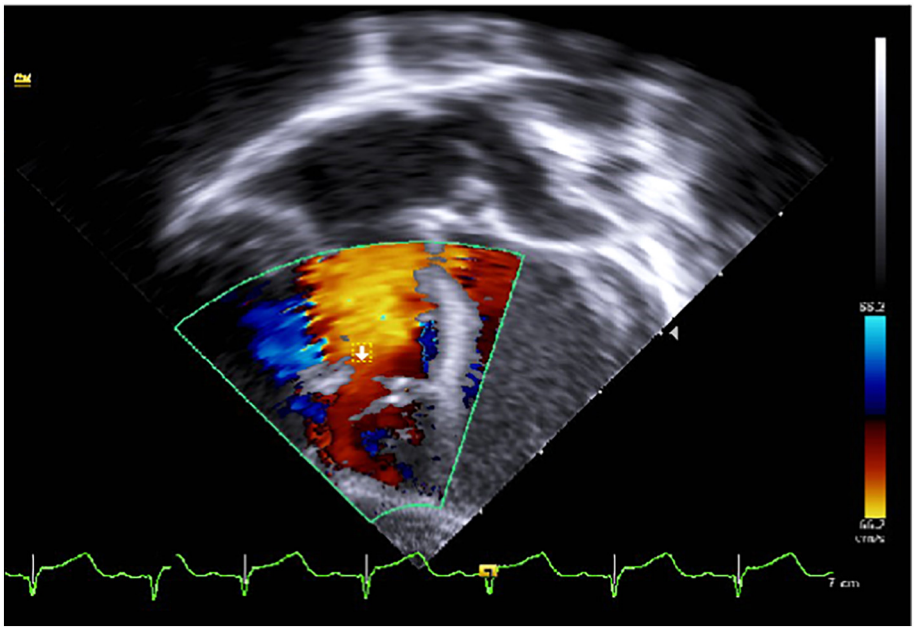

Fig. 2. Blood flow seen in the deep trabeculations in the right ventricle wall and apex.

7.4 mm with mild tricuspid regurgitation. Left ventricular ejection fraction was 65%. Hydrochlorothizade and captopril have been discontinued.

3. Discussion

During normal foetal development, ventricles initially are a meshwork of interwoven fibres and between 5th and 8th week, ventricular compaction occurs from the base towards the apex and from epicardium to endocardium. During compaction, inter-trabecular spaces in ventricular myocardium are obliterated and the recesses in the trabecular network are reduced to capillaries. Failure of this endocardial morphogenesis and regression of ventricular sinusoids results in ventricular non - compaction. The exact pathophysiology of ventricular dysfunction is unclear, but it has been suggested that subendocardial hypoperfusion and coronary microcirculatory dysfunction are a cause of arrhythmogenesis and fibrosis (1).

Both familial and sporadic forms of isolated ventricular non - compaction have been described. The familial form was observed in 18% of the adult population with isolated ventricular non - compaction (2). Gene localization has not been determined in isolated right ventricular non - compaction. Our patients first degree relatives echocardiograms were normal. The following diagnostic criteria for isolated left ventricular non - compaction have been used in literature: (i) The absence of coexisting cardiac anomalies; (ii) a two layered structure of left ventricular wall, with the end systolic ratio of the non - compacted

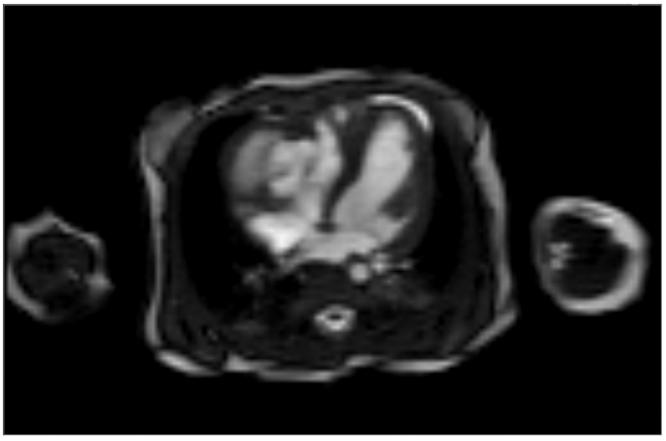

Fig. 3. Cardiac magnetic resonance imaging showed a mildly dilated right atrium and grossly dilated right ventricle. The right ventricle myocardium was slightly thickened. There was thinning at the right ventricle apex and inferior wall with multiple trabeculations and septations within the right ventricle apex.

to compacted myocardial layer > 2; (iii) finding this structure predominantly in the apical and midventricular areas; and (iv) blood flow directly from the ventricular cavity into deep intertrabecular recesses as assessed by doppler echocardiography (1). In the absence of clear cut criterion, the above mentioned criteria had been used earlier by authors and similarly by us for this case. Kohli et al., has classified non - compaction into three morphological categories namely; spongy, meshwork, and prominent trabeculations only. Our patient fulfilled all four criteria and had prominent trabeculations.

Electrocardiogram changes including atrial fibrillation with complete right bundle branch block and ventricular arrhythmia have been reported (1). Ventricular arrhythmias may be due to myocardial ischemia and scar tissue induced by microcirculatory dysfunction. Moreover, the prominent trabecular meshwork on the luminal surface of the ventricle was presumed to increase the probability of an ectopic pacemaker and re-entrant pathways (6).

Cardiac magnetic resonance imaging has been shown to provide good correlation with echocardiogram for localization and extent of non - compaction and has been found to be useful in cases with poor echocardiography quality. Cardiac magnetic resonance imaging has less operator dependence, superior spatial resolution, higher contrast between blood and myocardium, which can provide better delineation of the abnormal trabeculations in non – compaction of ventricular myocardium patients. It is particularly useful when the lesions are confined to the ventricular apex, which is difficult to be detected by echocardiography (7). Characteristic changes included prominent trabeculations, deep intertrabecular recesses and an increase in the diastolic non – compacted to compacted myocardium (NC/C ratio) (7). The diastolic ratio of (NC/C ratio) > 2.3 has been reported as a diagnostic criterion in left ventricular non – compaction (5). The most frequently involved segments were the anterior, lateral and inférolateral segments (7). However, no specific criterion has been proposed for the diagnosis of non – compaction of right ventricle (5).

No definitive consensus on treatment of the condition has been formed. Diuretics, digoxin, angiotensin converting enzyme inhibitors, carvedilol and calcium channel blockers help to control isolated right ventricular non - compaction symptoms and the disease course (4). There are no follow up studies to date of patients with isolated right ventricular non - compaction. Therefore, there are no prognostic criteria proposed for patients with isolated right ventricular non - compaction (1). However, the incidence of heart failure, heart block and coexisting cardiovascular malformations contributed to foetal and neonatal deaths (9). Genotype-phenotype correlations of gene mutations in all cases reported in the literature upto date, revealed no correlation between location or type of mutation and cardiac phenotype or disease severity in isolated ventricular non- compaction (10).

Important differential diagnoses are (8):

- Arrhythmogenic right ventricular cardiomyopathy/dysplasia shows regional right ventricle akinesia, dyskinesia, or aneurysm, epsilon wave in the right precordial leads (V1 to V3) and fatty infiltration on cardiac magnetic resonance imaging.
- Endomyocardial fibrosis is a progressive type of restrictive cardiomyopathy. Echocardiogram shows grossly dilated right atrium, mildly dilated right ventricle and tricuspid r egurgitation.
- Endocardial fibroelastosis is a cause of restrictive cardiomyopathy in children usually presenting within two years of life. Bright endocardial echoes with poorly contracting left ventricle is typical.
- Uhl's anomaly is a very rare disease with partial or total loss of the myocardial muscle in the right ventricle leading to markedly enlarged right ventricle without apical trabeculation and thin hypokinetic parchment like ventricular wall.

4. Conclusion

Knowledge and understanding about aetiology, embryogenesis of the myocardium, genetic background, diagnosis and outcome of left ventricular non - compaction have steadily improved. However, data for right ventricle non – compaction are lacking due to few case reports and under diagnosis of the disease. It is important to consider this rare cardiomyopathy as a differential in a neonate presenting with right heart failure.

Declarations of Interest

Authors declare there are no conflicts.

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