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Case Report

Quantification of end diastolic forward flow in two cases with pulmonary atresia with intact ventricular septum ☆☆☆☆

Yuji Doi*, Kenji Waki

Department of Pediatrics, Kurashiki Central Hospital, Kurashiki-city 710-8602 Okayama, Japan

ARTICLE INFO

Article history:

Received 30 May 2020

Revised 7 December 2020

Accepted 8 December 2020

Keywords:

Pulmonary atresia with intact ventricular septum
 Biventricular repair
 End-diastolic forward flow
 Restrictive physiology

ABSTRACT

Similar to patients with repaired tetralogy of Fallot, patients with repaired pulmonary atresia with intact ventricular septum may need a reintervention at a later stage. Although the role of MRI in the long-term follow-up of patients with repaired tetralogy of Fallot has been established, the same has not been established for patients with repaired pulmonary atresia with intact ventricular septum. To define this role, we quantified the end-diastolic forward flow by fractioning it by the total flow of the main pulmonary artery in two cases during their long-term follow up after biventricular repair. In case 1, a male patient had hepatic congestion and a high end-diastolic forward flow fraction and underwent surgical take down to one and one-half ventricle repair at the age of 18 years. In case 2, a female patient, currently 13 years old, has an increasing end-diastolic forward flow fraction. She is under close observation as a potential candidate for one and one-half ventricle repair in the near future. Both patients had a high end-diastolic forward flow fraction of the total right ventricle output, suggesting that end-diastolic forward flow fraction may become a possible indicator of the adequacy of biventricular repair and the optimal timing for reintervention.

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Introduction

As in patients with repaired Tetralogy of Fallot (TOF), some patients with repaired pulmonary atresia with intact ventricular septum (PAIVS) require reintervention of the right

ventricle outflow tract (RVOT) [1]. The optimal timing of reintervention can be evaluated by performing a magnetic resonance imaging (MRI) in patients with repaired TOF [2], yet the role of MRI in patients with PAIVS has not been well established.

☆ Competing Interests: The authors have no conflict of interests to declare.

☆☆ Funding: The authors have no fundings related to this manuscript.

* Acknowledgment: None.

* Corresponding author.

E-mail address: yd16549@kchnet.or.jp (Y. Doi).

<https://doi.org/10.1016/j.radcr.2020.12.029>

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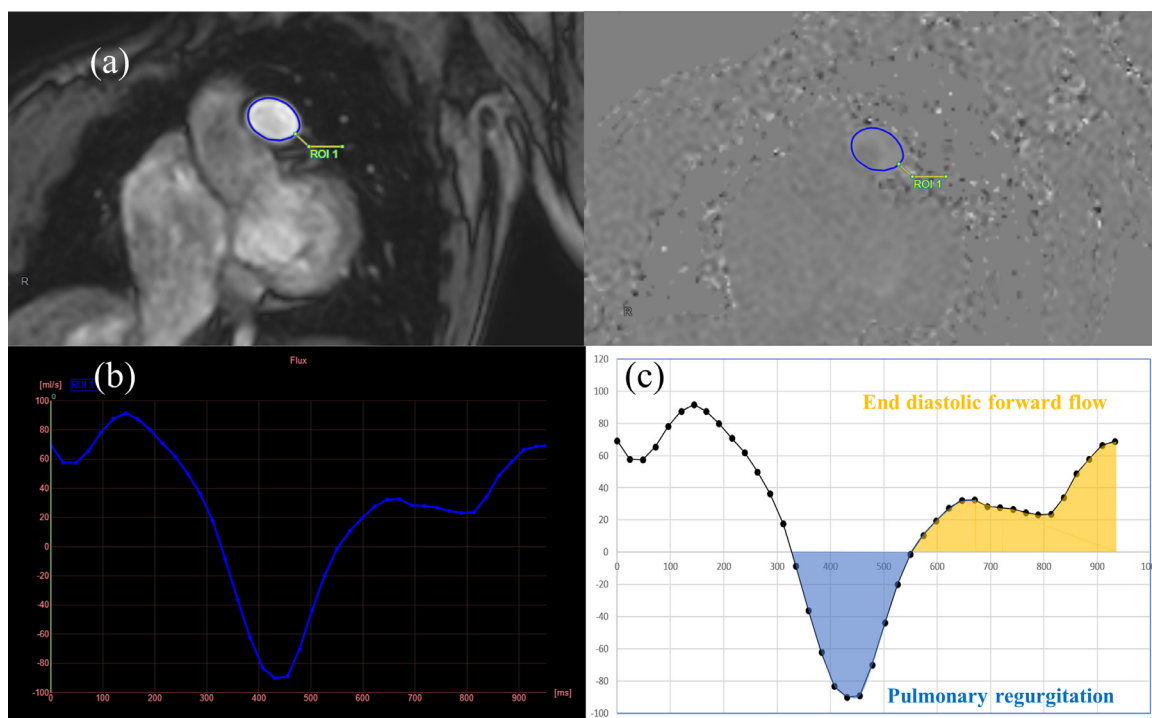


Fig. 1 – Magnetic resonance image of the main pulmonary artery during phase contrast in case 2 (a), and the time-flow curve calculated from the image (b). The data are extracted and the flow curve is reconstructed using Microsoft Excel® (Microsoft, Redmond, USA). The total flow and the flow for each segment are calculated. The area below zero represents the flow of pulmonary regurgitation (blue area), and the subsequent antegrade flow represents the end-diastolic forward flow (yellow area) (c).

Case presentation

Case 1

A term male infant diagnosed postnatally with PAIVS and hypoplastic right ventricle (RV) underwent Blalock-Taussig shunt and pulmonary valvuloplasty soon after birth, and then proceeded to undergo RVOT repair with RV overhaul at the age of 9 months. At 2 years of age, he underwent another RVOT repair with RV overhaul due to RVOT obstruction. He was in a stable condition for years after the second RVOT repair; however, his direct bilirubin level increased gradually to 2.9 mg/dL when he was 16 years old. Catheter evaluation showed a central venous pressure (CVP) of 12 mmHg and a right ventricle end-diastolic pressure (RVEDP) of 14 mmHg. An MRI was performed and the end-diastolic forward flow (EDFF) was evaluated by phase contrast of the main pulmonary artery. The region of interest was determined by pediatric cardiologists, and the flow volume curve of the main pulmonary artery was depicted using Ziosation 2 (Ziosoft, Tokyo, Japan) as the analysis software. As the MRI was electrocardiogram gated, EDFF was calculated as the area under curve after the retrograde flow which represents pulmonary regurgitation (PR). The EDFF volume was 16.3 mL, and its fraction was 30.1% of the total RV forward flow. With hepatic congestion and high CVP due to the restrictive RV physiology, he underwent one and one-half ventricle repair at the age of 18 years. Although his hemodynamic status improved, he developed atrial flutter after the

surgical intervention and underwent catheter ablation. He still has repetitive episodes of atrial flutter and is scheduled for another session of catheter ablation.

Case 2

A term female infant with a prenatal diagnosis of PAIVS and hypoplastic RV underwent balloon valvuloplasty soon after birth. Since the saturation remained low after the initial intervention, she underwent Blalock-Taussig shunt at 2 months of age. Subsequently, she underwent an RVOT repair with RV overhaul at 1 year of age, and the atrial septum was left partially open. The atrial septal defect closed spontaneously during the clinical course, yet she was clinically stable thereafter. She underwent catheterization at the age of 10 years, and had a CVP of 9 mmHg and RVEDP of 15 mmHg. An MRI revealed an EDFF of 8.27 mL (28.6% of the total RV forward flow) measured with the same method used in case 1. She is currently 13 years old, and has undergone another catheterization and MRI. The results revealed a CVP of 7 mmHg, RVEDP of 11 mmHg, and an EDFF measured by phase contrast of the main pulmonary artery (Fig. 1) of 14.2 mL (38.9% of the total RV forward flow). PR and tricuspid regurgitation (TR) detected by echocardiography were both mild and did not change in the recent years. She had a lower CVP or RVEDP compared to when the catheterization was performed at 10 years of age. She experienced dyspnea on exertion, and the RV pressure wave form progressed into a dip and plateau pattern (Fig. 2). She has not received any intervention, as there are no signs of hepatic dysfunction or

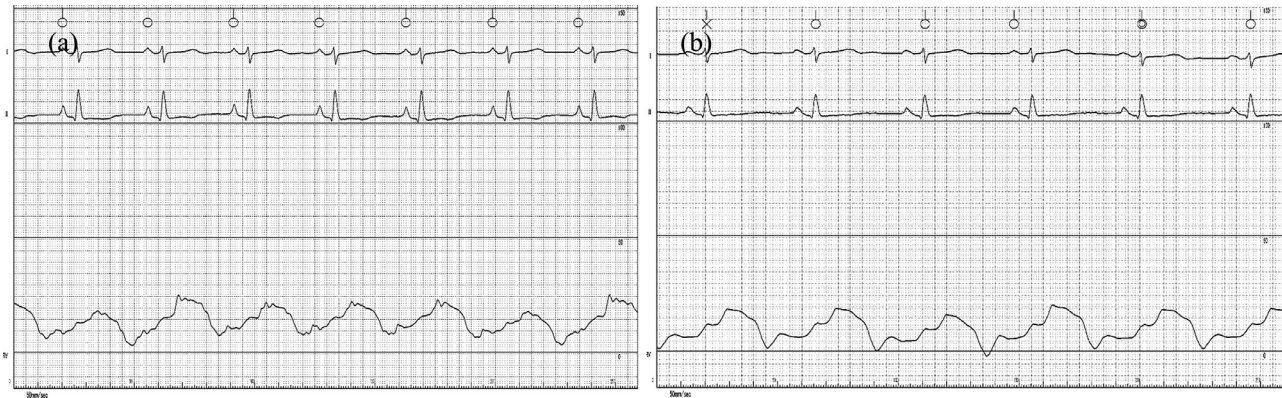


Fig. 2 – Right ventricle pressure wave form in case 2 recorded at 10 years of age (a) and at 13 years of age (b). The wave form has progressed into a dip and plateau pattern.

arrhythmias. She is being observed closely as an outpatient with an intention to perform one and one-half ventricle repair before severe symptoms related to restrictive RV physiology develop.

Discussion

The treatment strategy for PAIVS depends on the development of the right heart and the presence of sinusoidal communication [3]. Some patients undergo biventricular repair using RV overhaul to avoid Fontan repair, but over-enthusiastic biventricular repair may still yield unfavorable outcomes [4]. Thus, it is important to validate its long-term feasibility. John and Warnes reported that after biventricular repair, patients often require reinterventions such as tricuspid valve repair/replacement, or pulmonary valve repair, and some of them experience arrhythmias [5]. Ideally, reintervention should be performed before the occurrence of prominent symptoms such as liver dysfunction due to elevated CVP or arrhythmias because these symptoms may not fully subside after reintervention, as seen in case 1.

Detecting the downtrends of right heart function is often difficult, but in patients with repaired TOF, MRI has an established role to determine the optimal timing for reintervention, mainly by evaluating the RV end-diastolic volume index [2]. We propose that MRI may play the same role in patients with PAIVS to detect the downtrends of right heart function. However, unlike in patients with TOF, the RV end-diastolic volume index does not increase in patients with PAIVS due to the restrictive physiology of the RV, suggesting a need for other indicators. For these cases, EDFF, measured by phase contrast of the main pulmonary artery can assess the overall function of the RV. Although EDFF represents a restrictive physiology of the heart, its presence reflects a different mechanism (primary and secondary) [6]. A primary restrictive physiology represents decreased diastolic function of the myocardium, while a secondary restrictive physiology is a result of RV remodeling due to PR and TR [7]. The RV in patients with both these physiologies will have very limited

capacity for filling (unfillable RV) as seen in patients with PAIVS after biventricular repair. As the presence of EDFF may vary during the cardiac cycle [8], qualitative diagnosis is not helpful and quantification is required. As there is no cut-off value for EDFF, we tried to quantify it by fractionating EDFF by the total RV output. In both the patients, the fractionated EDFF was very high. Although quantification of EDFF itself did not change the clinical decision making in both patients, the fraction of EDFF did seem to represent the progressive restrictive RV physiology well in both cases. In the case 2, especially, the fraction increased as the restrictive physiology seemed to progress clinically despite no deterioration in PR, TR, and elevation of CVP and RVEDP. Output from the RV depends on many factors, such as RV volume, PR, TR, pulmonary stenosis, compliance of the myocardium, and preload of the RV (which is somewhat represented by CVP). Maintaining optimal RV output at permissible preload is probably the key for better prognosis in patients with PAIVS. We consider that EDFF may represent the restrictive RV physiology in these patients reflecting all the other factors, as an unfillable RV results in decreased output during systole and increased output by atrial contraction as a compensation to maintain output which will present as EDFF. A recent study showed that patients with PAIVS showed decreased diastolic function, which was presented as a higher flow velocity of EDFF compared to that in patients with a repaired TOF [9], which supports our hypothesis that quantified EDFF could be used as a possible surrogate marker of restrictive physiology. Although further validation is necessary, quantification of EDFF using MRI may be an ideal method to assess the adequacy of biventricular repair and to detect the optimal timing for re-intervention in patients with PAIVS (especially those with a hypoplastic RV) in the long term. Further research is necessary for validation and for setting a cut-off point for quantification.

Ethics approval

The authors assert that all procedures conducted herein comply with the ethical standards of the relevant national

guidelines on human experimentation and with principles of the Helsinki Declaration of 1975, as revised in 2008. This study has been approved by the institutional committee.

Consent to participate

Written informed consent was obtained from each patient or from their care giver before all the procedures.

Consent for publication

Consent for publication was obtained from each patient.

Availability of data and material

All the data were collected from the procedure performed in our hospital. No statistical analysis was performed.

Code availability

Not applicable.

Authors' contributions

Yuji Doi took care of the patient as a primary physician from the beginning to the end and was the major contributor in writing the manuscript. Kenji Waki took care of the patient and, read and approved the revised manuscript.

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