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CASE REPORT
Adult congenital heart disease

Intraventricular haemodynamic changes caused by increased left ventricular afterload in re-coarctation of aorta: a case report

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Background Long-term re-coarctation of the aorta can cause aortic dilatation, hypertension, and cardiac dysfunction due to increased left ventricular (LV) afterload. It is difficult to detect changes in LV function due to increased afterload if the contractile force of the left ventricle is maintained. Herein, we have reported a case of re-coarctation of the aorta, for which four-dimensional (4D) flow magnetic resonance imaging (MRI) scan was obtained both before and after balloon dilatation for aortic re-coarctation. Ultimately, improvement in aortic helical flow and LV haemodynamics was observed.

Case summary A 29-year-old female was diagnosed with coarctation of the aorta and a bicuspid aortic valve after birth and underwent surgery at 1 month. At 8 years of age, she underwent balloon dilatation for re-coarctation. At the age of 28 years, she was diagnosed with re-coarctation triggered by hypertension. She underwent balloon dilatation as her cardiac catheterization revealed a systolic pressure gradient of 40 mmHg. Pretreatment 4D flow MRI demonstrated helical flow in the ascending aorta and descending thoracic aorta and LV blood flow analysis revealed a decrease in LV kinetic energy during systole; these improved after treatment.

Discussion The use of helical flow evaluation by 4D flow MRI for aortic re-coarctation is well known in clinical practice. However, our report is the first to evaluate intraventricular blood flow before and after the re-coarctation treatment. The MRI evaluation demonstrated that the helical flow and LV blood flow distribution improved after re-coarctation treatment due to the reduction of afterload.

Keywords Re-coarctation of aorta • 4D flow MRI • Left ventricular afterload • Ventricular flow analysis • Kinetic energy • Case report

ESC curriculum 2.3 Cardiac magnetic resonance • 9.1 Aortic disease

Learning points

- Four-dimensional flow magnetic resonance imaging showed that the kinetic energy of left ventricular (LV) blood flow and large-vessel helical flow improved before and after aortic re-coarctation treatment.
- Improvements in the kinetic energy of LV blood flow indicate improvement in LV afterload.
- Combining intraventricular blood flow and helical flow evaluation may indicate re-coarctation treatment non-invasively.

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Introduction

Aortic coarctation accounts for 7% of congenital heart diseases. Most of these lesions are repaired during the neonatal period.[1] In the late postoperative period, repair site re-coarctation develops in 10–15% of cases, accompanied by hypertension, aortic dilatation, and cardiac dysfunction.[2] Cardiac dysfunction is caused by left ventricular (LV) myocardium stiffening due to increased afterload.[3] The quantitative evaluation of the afterload is difficult. Visualization of blood flow in the ventricles and great vessels is enabled by 4D flow magnetic resonance imaging (MRI). Intraventricular blood flow is important as an index for early assessment of diastolic function.[4] However, intraventricular flow analysis has not been reported for re-coarctation cases. We hypothesized that in patients with re-coarctation, improvement of LV afterload would lead to improvement of intraventricular blood flow.

Summary figure

Time	Event
After birth	Diagnosed with aortic coarctation and a bicuspid aortic valve
At 1 month after birth	A left subclavian flap aortoplasty
At 8 years of age	Balloon dilatation for re-coarctation
	Self-discontinued outpatient follow-ups
At 26 years of age	No symptoms
	Diagnosed with hypertension and referred to our hospital
	Contrast-enhanced computerized tomography scan revealed re-coarctation
At 29 years of age	Occasionally noticed fatigue in her legs when walking
	Cardiac catheterization
	• The left ventricular (LV) pressure of 152/e12 mmHg
	• The re-coarctation area of 7.0 × 5.3 mm
	• The systolic pressure gradient of 40 mmHg
	Four-dimensional flow magnetic resonance imaging (MRI)
	• Helical flow was observed in the ascending and descending thoracic aorta
	• Left ventricular blood flow analysis revealed a decrease in LV kinetic energy during systole
At 1 month after catheterization	*Balloon dilatation for re-coarctation*
	• The re-coarctation area expanded to 10.0 mm × 10.5 mm
	• The systolic pressure gradient improved to 6 mmHg
	Four-dimensional flow MRI
	• Helical flow was improved
	• Left ventricular blood flow analysis and kinetic energy were improved

Case presentation

A 29-year-old woman was diagnosed with aortic coarctation and a bicuspid aortic valve. The patient underwent a left subclavian flap aortoplasty 1 month after birth and balloon dilatation for re-coarctation at the age of 8 years. She subsequently self-discontinued outpatient follow-ups. At the age of 26 years, she was diagnosed with hypertension and referred to our hospital as her contrast-enhanced computerized tomography scan revealed re-coarctation. Her height was 152 cm, weight 53 kg, heart rate 90 b.p.m., percutaneous arterial oxygen saturation 97% (room air), right upper extremity blood pressure (BP) 152/89 mmHg, and right lower extremity BP 120/93 mmHg. She had a Levine III/VI systolic murmur at the second intercostal space left sternal border and left back. The patient occasionally noticed fatigue in her legs when walking. Laboratory findings revealed an N-terminal prohormone of brain natriuretic peptide of 50.6 pg/mL. A chest radiography revealed a cardiothoracic ratio of 48% and no rib notch sign. A 12-lead electrocardiogram revealed sinus rhythm with sV1 + rV5 of 2.6 mV. Echocardiography revealed an LV end-diastolic diameter of 44.6 mm, a posterior wall of 7.9 mm, and an ejection fraction rate of 79%. The LV inflow waveform had a normal pattern with $E/A > 1$. The blood flow velocity at the re-coarctation was 3.2 m/s. The abdominal aortic blood flow waveform was blunted, and the systolic upstroke delayed. Cardiac catheterization revealed an LV pressure of 152/e12 mmHg, a re-coarctation area of 7.0 × 5.3 mm, and a systolic pressure gradient (PG) of 40 mmHg (*Figure 1A*). One month after catheterization, the patient underwent balloon dilatation. Subsequently, the re-coarctation area expanded to 10.0 mm × 10.5 mm, and the systolic PG improved to 6 mmHg (*Figure 1B*). After patient consent, we used 4D flow MRI for research purposes to evaluate the large vessel and LV blood flow before and after treatment. Cardiac MRI was a 3.0 T whole-body imager (MAGNETOM Vida 3 T; Siemens Healthcare, Erlangen, Germany). Scan parameters included: acceleration method, GRAPPA 3; field of view, 340 × 340 mm; flip angle, 8°; temporal resolution, 41 ms; a retrospectively electrocardiogram triggered. The acquired voxel size was $2.5 \times 2.5 \times 2.5\ mm^3$, with velocity encoding 150 cm/s, Segment 2, and 30 frames/cycle. A workstation (cvi42, ver 5.17.0, Circle, Cardiovascular Imaging, Calgary, Canada) was used for the analysis.

Preoperatively, helical flow was observed in the ascending and descending aorta (*Figure 2A and B*). The LV blood flow analysis revealed 39.1% direct flow, 21.7% delayed ejection flow, 23.4% retained inflow, and 15.9% residual volume. Details of the four blood flow components and data on normal subjects are shown (*Figure 3* and Supplementary material online, *Table S1*).[4] No obvious intraventricular blood flow ratio abnormality was observed (*Figure 4A–D*). However, kinetic energy (KE) values of direct flow and delayed ejection flow decreased during systole (*Figure 5A*). Postoperatively, helical flow in the descending aorta was alleviated (*Figure 2C and D*). Left ventricular blood flow analysis revealed 39% direct flow, 19.4% delayed ejection flow, 22% retained inflow, and 19.5% residual volume (*Figure 4E–H*). Although no change was observed in the direct flow ratio, an increase in KE values of direct flow and delayed ejection flow during systole was observed (*Figure 5B*). Left ventricular function and volumes did not change before and after treatment. Two months after treatment, the systolic BP decreased by 20 mmHg.

Discussion

Helical flow in the aorta and systolic KE of the intraventricular blood flow component improved after balloon dilatation. This is the first report to demonstrate improved KE after re-coarctation treatment.

Re-coarctation is a long-term complication after initial repair, caused by contraction of the remaining tissue of the ductus arteriosus, replacement with fibrous tissue, and scar formation.[5–7]

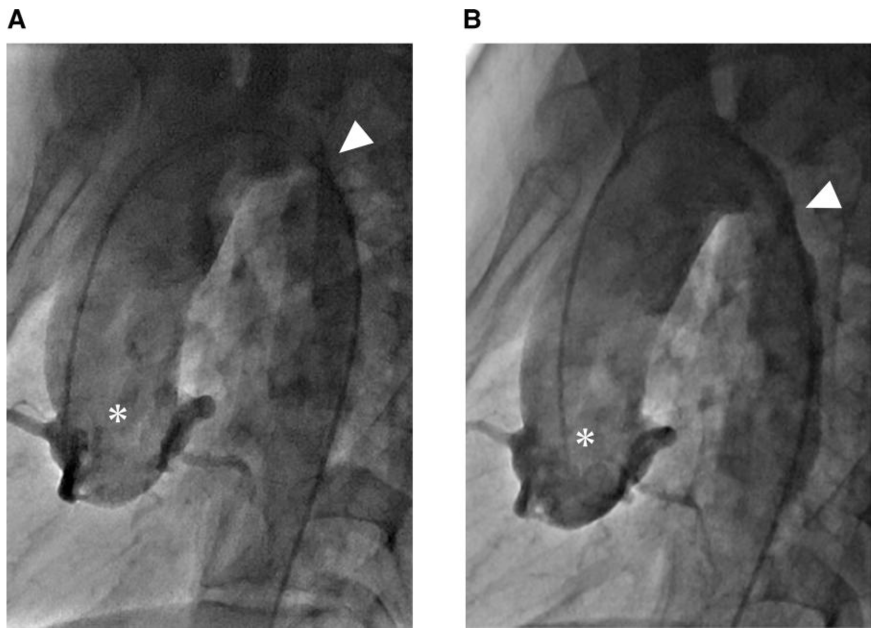

Figure 1 Angiography before (*A*) and after (*B*) balloon angioplasty. The arrow head shows the re-coarctation site. The size of the aortic root (*) before and after treatment was 26.4 and 25.4 mm, respectively.

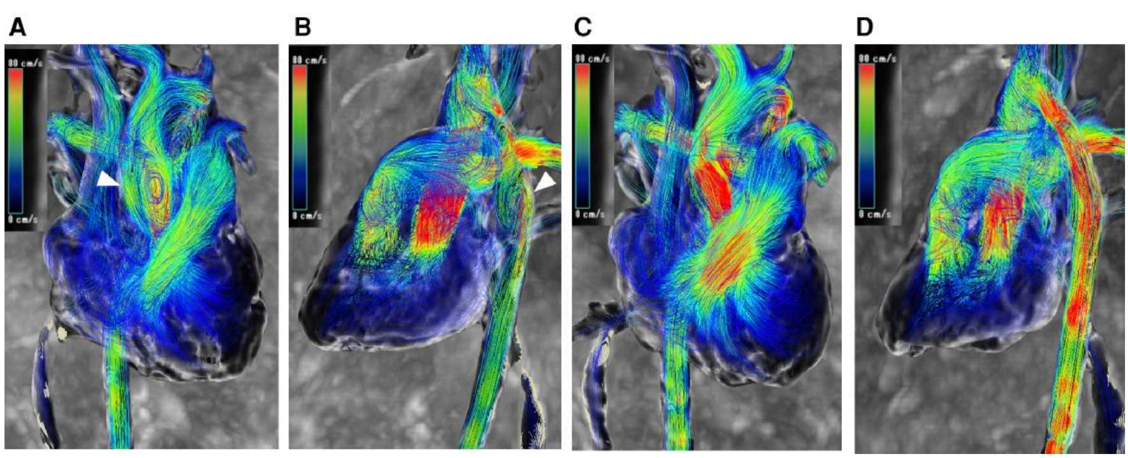

Figure 2 Streamline of four-dimensional flow magnetic resonance imaging. A helical flow (arrow head) is visible in the ascending aorta in the frontal view (*A*) and in the descending aorta in the lateral view (*B*) before balloon angioplasty. After treatment, both flows improved (*C* and *D*).

Re-coarctation causes helical flow, leading to increased energy loss and wall shear stress.[8] The afterload of the LV increases with re-coarctation. Increased afterload causes myocardial hypertrophy and LV stiffening, resulting in decreased cardiac output and diastolic dysfunction.[3] Afterload changes can be indicated by the relationship between ventricular pressure and volume. However, it is difficult to quantitatively evaluate the afterload. Therefore, we hypothesized that the intraventricular blood flow component might change with improved afterload. Left ventricular blood flow components include direct flow, delayed ejection flow, retained inflow, and residual volume.[9] The direct flow ratio is high in healthy subjects. The residual volume increases in patients with significant diastolic dysfunction such as dilated cardiomyopathy.[4] However, with unremarkable diastolic dysfunction, only a mild residual volume increase is observed, and the effect of afterload on the LV cannot be evaluated only by the intraventricular blood flow component ratio. Therefore, we obtained the blood flow component ratio and KE change for each cardiac cycle and examined the overall LV KE change. In a normal LV, KE peaks are formed during systole and diastole are represented by the KE curve.[10] Although the LV contractility was maintained, the KE produced by the LV during systole decreased due to increased afterload. As the afterload was reduced, the ejection of blood from the LV

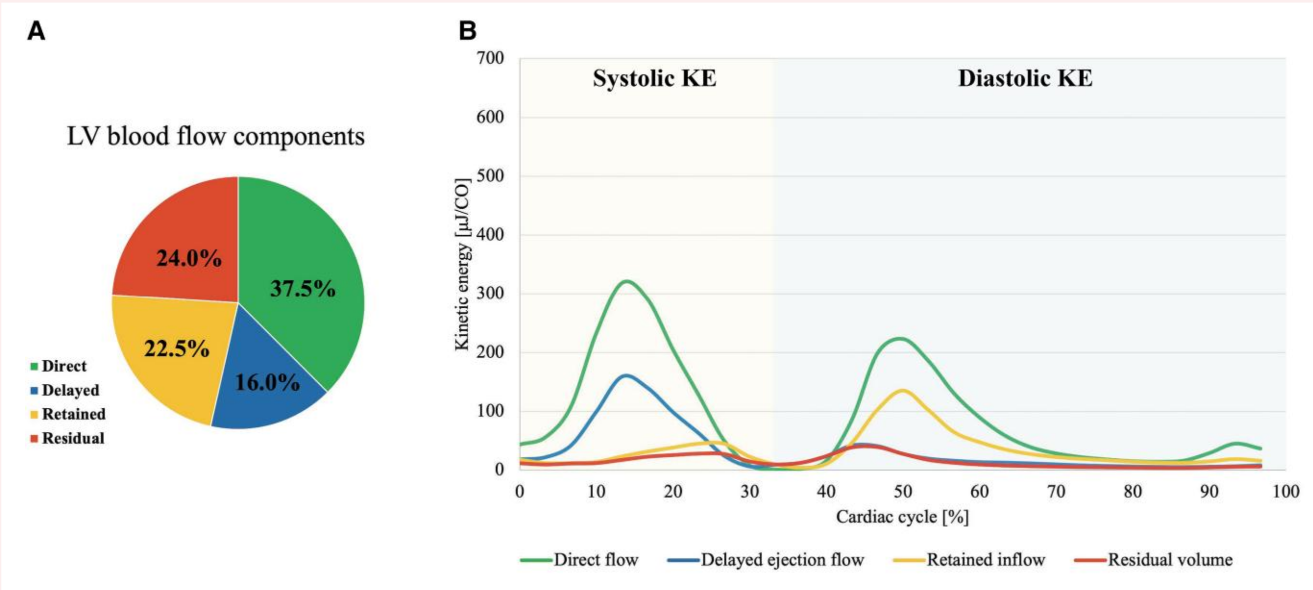

Figure 3 Left ventricular blood flow components (*A*) and kinetic energy (*B*) in healthy subjects in our hospital are shown. The left ventricular kinetic energy of healthy subjects forms bimodal peaks during systole and diastole. Left ventricular blood flow has the largest proportion of direct flow among the four components, followed by residual volume.

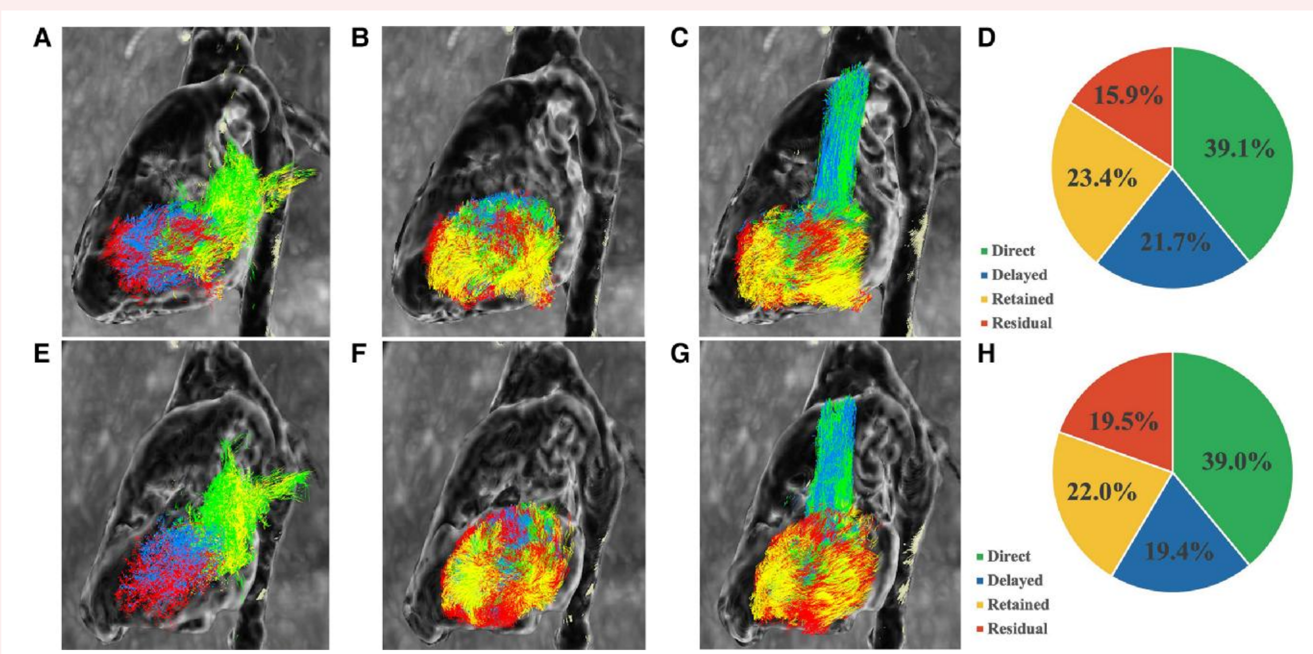

Figure 4 Ventricular flow analysis of four-dimensional flow magnetic resonance imaging. Before balloon angioplasty: (*A*) early diastolic phase, (*B*) end-diastolic phase, (*C*) peak systolic phase, and (*D*) percentage of the components of left ventricular flow. After balloon angioplasty: (*E*) early diastolic phase, (*F*) end-diastolic phase, (*G*) peak systolic phase, and (*H*) percentage of the components of left ventricular flow.

became smoother, and the KE during systole increased. Hence, an increase in KE was considered to indicate the improvement of afterload, and we could show the effects of LV afterload by evaluating intraventricular blood flow.

The ESC guidelines state that the criteria for treatment of re-coarctation are hypertension and elevated PG.[11] Further, it is known that helical flow occurs due to re-coarctation and affects aortic dilatation.[12] However, it has been reported that helical

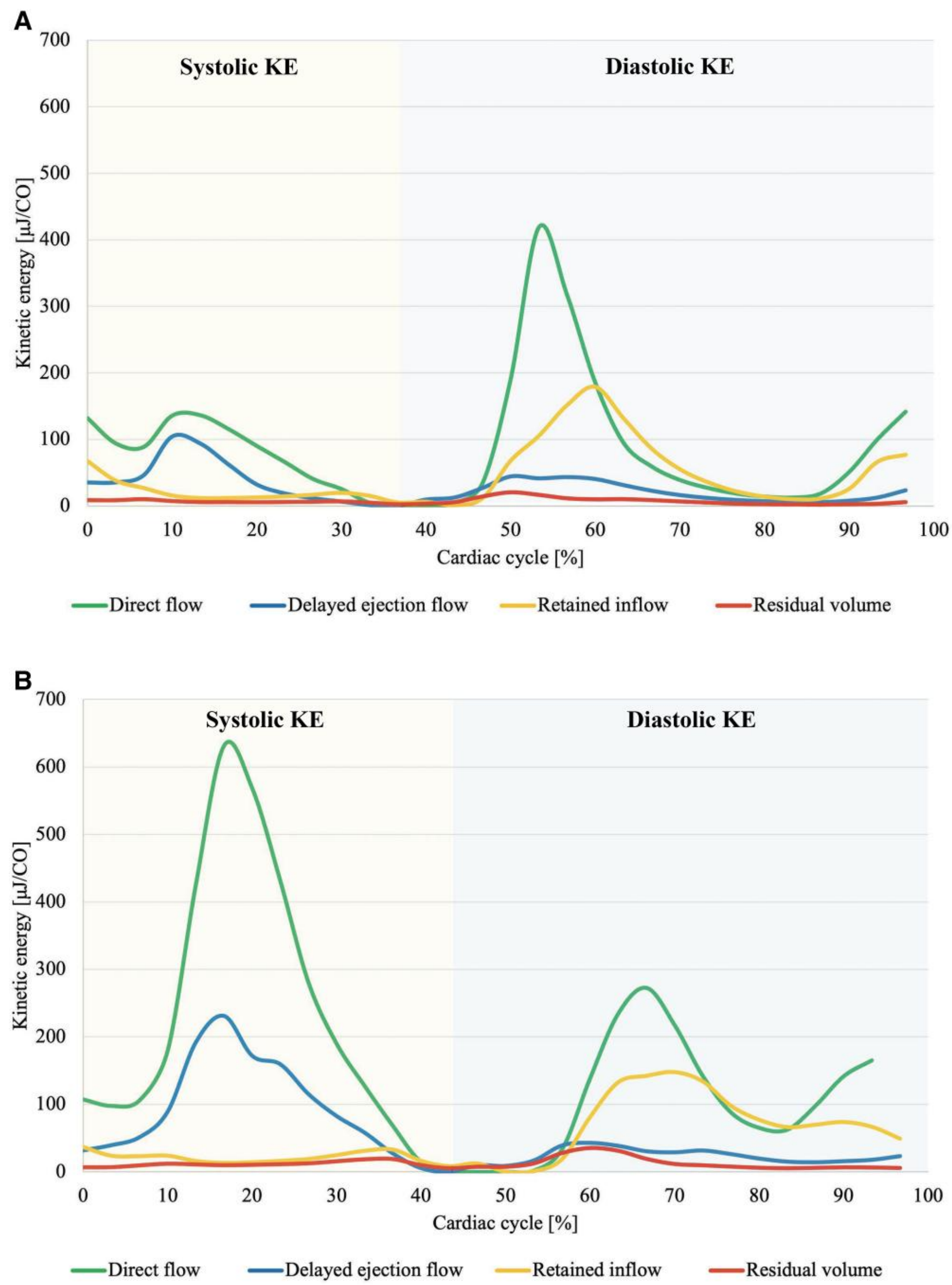

Figure 5 Left ventricular blood flow components kinetic energy analysis whole cardiac cycle; green, direct flow; blue, delayed ejection flow; yellow, retained inflow; red, residual volume. (*A*) The kinetic energy curve before treatment and (*B*) after treatment.

flow occurs even in re-coarctation without a PG.[13] This suggests that helical flow alone is not sufficient to determine the indication for treatment. In this case, LV KE may represent afterload with preserved ejection fraction. Since a decrease in systolic LV KE occurs when PG causes an increase in afterload, the haemodynamic changes caused by re-coarctation should start with the onset of helical flow and progress to a decrease in LV KE. In other words, if a decrease in systolic LV KE occurs in addition to the onset of helical flow, there may be PG to treat. Four-dimensional flow MRI may be used to non-invasively determine the indication for treatment. Further studies are needed to determine the extent to which a decrease in LV KE results in PG as an indication for treatment. In the future, if it will be possible to infer helical flow and intraventricular blood from echocardiography as in 4D flow MRI, the process would be even less invasive.

The effect of LV afterload can be evaluated by 4D flow MRI for re-coarctation through intraventricular blood flow analysis and helical flow evaluation. Combining intraventricular blood flow and helical flow evaluation may indicate re-coarctation treatment non-invasively.

Lead author biography

Yuki Shibagaki graduated from Asahikawa Medical University in Japan in 2018. She currently belongs to Department of Pediatrics, Asahikawa Medical University, and is trained as a pediatric cardiologist.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Data availability

The data underlying this article are available in the article and in its online Supplementary table.

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