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Symptomatic myocardial bridging: a frequently occurring coronary variation can cause severe myocardial ischaemia in affected children with underlying cardiac conditions

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Abstract

Myocardial bridging is a congenital coronary artery anomaly in which the coronary artery has a partly "tunnelled" intramyocardial course. This tunnelling leads to compression of the affected vessel segment during ventricular systole. It is considered to be a benign variation of the norm in about 25% of the population caused by an aberrancy of embryologic coronary development. The bridging is also thought to cause severe cardiac conditions in a few of those affected. The series of six young patients presented here is the largest series so far to report on symptomatic myocardial bridging in children with different underlying heart diseases. All patients recently presented to our centre with signs of myocardial ischaemia. They subsequently underwent coronary angiography, which revealed myocardial bridging of the ramus interventricularis anterior. In all patients, therapy with β blockers was started to reduce heart rate and myocardial contractility. β Blocker treatment was also given in order to prolong diastole and improve coronary artery blood flow. Two patients underwent surgical exposure of the involved coronary segment: a 2-year-old boy because of recurrent, severe myocardial ischaemia in combination with a reduction of general health, changes in ST-segments, and the presence of a dilative cardiomyopathy; and a 13-year-old girl because of evidence of myocardial ischaemia during exercise testing after surviving sudden cardiac death. Surgery was successful and recovery was complete and uneventful. The presented series shows that myocardial bridging can be symptomatic and may require urgent treatment and even surgical intervention in early childhood in rare cases.

Myocardial bridging is a congenital coronary anomaly in which the coronary artery has a partly "tunnelled" intramyocardial course. It is thought to be a benign variation of the norm caused by an aberrancy of the embryologic coronary development.[1] The tunnelling of the artery leads to compression of the "tunnelled" vessel segment during systole that can persist into diastole.[2,3] Complex and dynamically interacting biomechanical factors may influence coronary blood flow within and distal to the bridged coronary segments.[3] The clinical relevance of this systolic compression is controversial. In most cases, myocardial bridging is thought to be benign, but in a few patients it can cause severe cardiac issues including myocardial ischaemia, ventricular arrhythmia, and sudden cardiac death. Several studies could not find any systematic association between myocardial bridging in hypertrophic cardiomyopathy patients and sudden cardiac death.[4] Despite the lack of association between myocardial bridging and symptoms, the authors speculate that there may be a potential increased risk in individual patients.

Myocardial bridging is found in about one-third of healthy adults. The number of affected people in different studies is highly variable; in autopsy series there is a reported mean of 25%, and in angiographic series it varies between 0.5 and 40% with provocation tests.[1–3,5] Typically, the ramus interventricularis anterior is affected in more than two-thirds of patients.[3]

There are a few reports of myocardial bridging in children. In children with hypertrophic cardiomyopathy the frequency is stated to be about 28%,[2,6] but the condition seems to be rare in children without hypertrophic cardiomyopathy. A number of these children showed severe symptoms with chest pain, syncope, ventricular arrhythmia, palpitations, and cardiac arrest and required surgical treatment with division ("unroofing").[2,7] In young patients without

hypertrophic cardiomyopathy, there are rare single case reports regarding symptomatic myocardial bridging in otherwise healthy children.[8,9] Asymptomatic myocardial bridging has been reported in a child with familial subaortic stenosis[10] and in a study of children with Williams syndrome – 3 out of 38 children.[11] These case reports related to children aged 5–16 years.

Materials and methods

We retrospectively analysed a series of six patients aged 0–16 years, who have been treated in our centre within the past years with symptomatic myocardial bridging and different underlying cardiac conditions. This retrospective analysis was approved by the head of our department. The report was conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

Results

In this retrospective single-centre study, we describe six patients who presented with signs of myocardial ischaemia and subsequently underwent coronary angiography, which revealed myocardial bridging of the ramus interventricularis anterior (Fig 1). All patients underwent echocardiographic function tests including two-dimensional echo and two-dimensional speckle tracking echocardiography, followed by invasive angiography. Our two adolescent patients additionally received magnetic resonance tomography and ergometric testing; one other patient

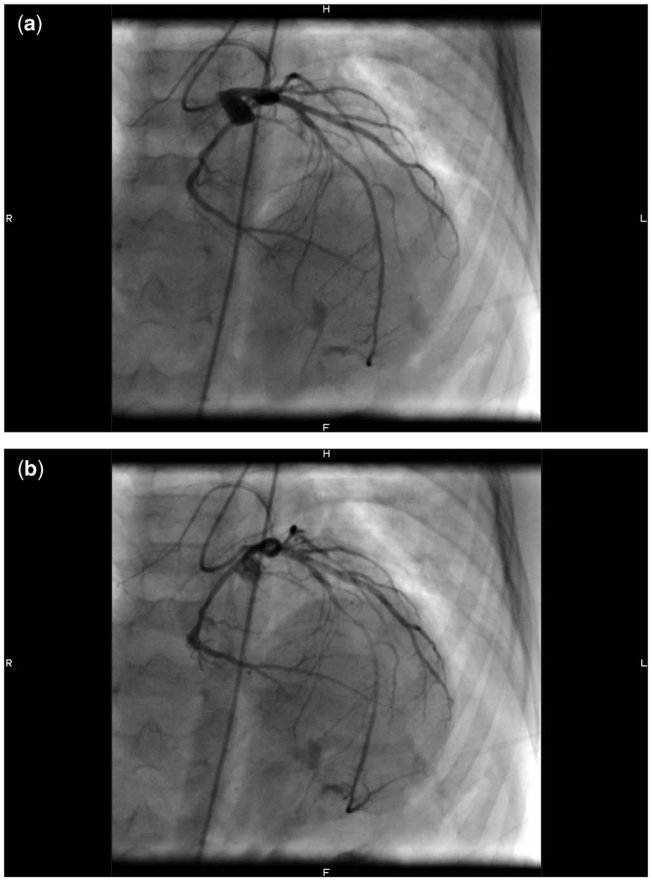

Figure 1. Angiography of the left coronary system in the diastolic (*a*) and systolic (*b*) phase in left anterior oblique view (patient 1). Ramus interventricularis anterior compression in the middle aspect with reduced flow as the ventricle contracts.

Table 1. Clinical characteristics of six children with symptomatic myocardial bridging.

Patients	Sex	Age	Primary diagnosis	Coronary anomaly	Signs and symptoms	Testing	Treatment	Surgical treatment	Follow-up (months)	Adverse events
1	Female	Neonate	Coarctation, ventricular septal defect, "parachute" mitral valve, patent ductus arteriosus, patent foramen ovale	Myocardial bridging in the range of RIVA	Myocardial ischaemia and impaired function of the interventricular septum with elevated myocardial ischaemia markers following corrective heart surgery	2D-E, 2D-STE, Angio	Propranolol, ASS	No	32	None
2	Male	4 months	HCM	Profound, long-segment stenosis of RIVA (suspected history of arteritis) in addition to myocardial bridging	Resuscitation following ventricular fibrillation	2D-E, 2D-STE, Angio	Propranolol, ASS	No	21	None
3	Male	1.5 years	DCM owing to myocardial inflammation	Myocardial bridging in the range of RIVA	6 weeks of cough, intermittent fever, suspected myocarditis, Echo: impaired myocardial function	2D-E, 2D-STE, Angio, EMB	Metoprolol, ASS, Captopril	No	31	None
4	Male	2.5 years	DCM	Myocardial bridging in the range of RIVA	Tachydyspnoea, fatigue, six episodes of acute heart failure	2D-E, 2D-STE, Angio, Cardio-CT, EMB	Metoprolol, ASS, Captopril, Metildigoxin	Yes, surgical exposure of RIVA ("unroofing")	33	None
5	Female	13 years	Cardiomyopathy with LVNC and hypertrophic component	Myocardial bridging in the range of RIVA	Resuscitation following ventricular fibrillation	2D-E, 2D-STE, Angio, Ergo, Stress Echo, Cardio-MRT, EMB	AICD, Metoprolol, ASS, exercise restriction owing to underlying heart disease	Yes, surgical exposure of RIVA ("unroofing")	19	None
6	Male	16 years	HCM, initially suspected myocarditis	Myocardial bridging in the range of RIVA	Pre-syncope, heart murmur	2D-E, 2D-STE, Angio, Ergo, Cardio-MRT, EMB	Metoprolol, ASS, exercise restriction for competitive sports	No	4	None

2D-E = two-dimensional echocardiography; 2D-STE = two-dimensional speckle tracking echocardiography; AICD = automatic implantable cardioverter defibrillator; Angio = angiography; ASS = acetylsalicylic acid; DCM = dilatative cardiomyopathy; EMB = endomyocardial biopsy; Ergo = ergometric testing; HCM = hypertrophic cardiomyopathy; LVNC = left ventricular cardiomyopathy; MRT = magnetic resonance tomography; RIVA = ramus interventricularis anterior

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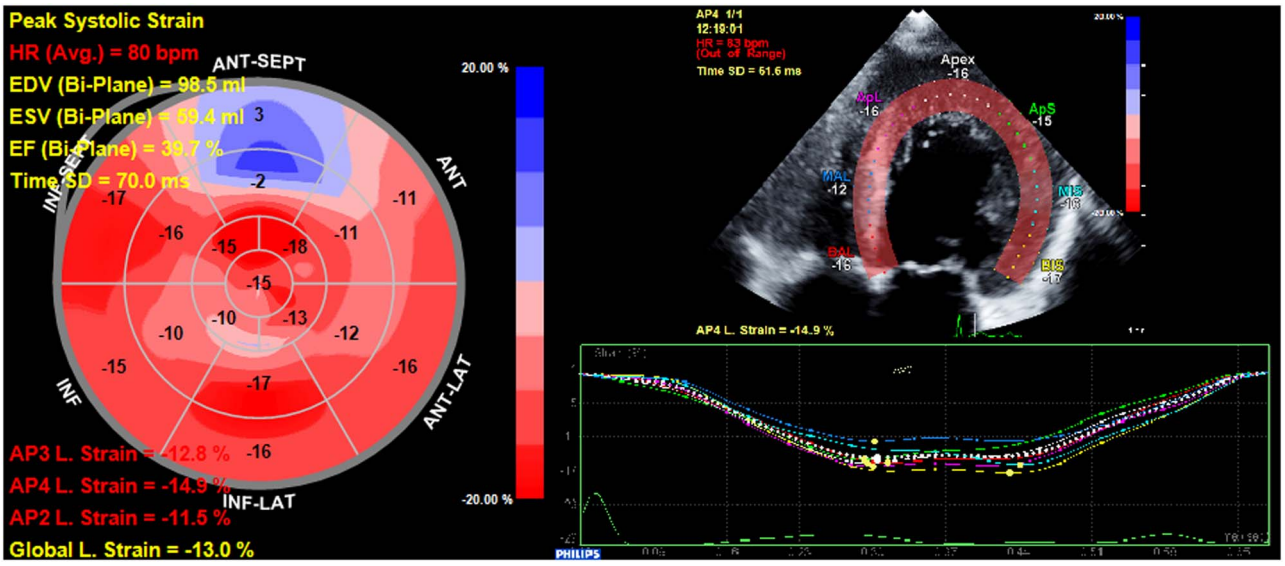

Figure 2. Two-dimensional speckle tracking echocardiography reveals localised myocardial deformation abnormalities in anterior septal left ventricular segments in patient 5 (Epiq 7G, QLab 10.4, aCMQ module; Philips Healthcare North America, Andover, MA, United States of America). ANT = anterior; AP2 = apical two chamber view; AP3 = apical three chamber view; AP4 = apical four chamber view; ApS = apicoseptal; ApL = apicolateral; BAL = basal anterolateral; BIS=basal inferoseptal; EDV = enddiastolic volume; ESV = endsystolic volume; EF = ejection fraction; HR = heart rate; INF = inferior; LAT = lateral; MAL = mid anterolateral; MIS = mid inferoseptal; SEPT = septal.

received an additional CT-scan before being referred to our unit. In four out of six patients, an endomyocardial biopsy was performed (see Table 1). Clinical data for the six study patients are summarised in Table 1. The median age at diagnosis was 5.5 years – ranging from 12 days to 16 years. The median follow-up duration was 26 months – ranging from 4 to 33 months. During the follow-up period, we did not notice any major events with recurrent myocardial ischaemia, cardiac arrest, or death. The clinical manifestation varied from subclinical to severe. The subclinical presentation was an incidental finding of localised myocardial dysfunction demonstrated by two-dimensional speckle tracking echocardiography (Fig 2) and unusually long persistence of elevated myocardial markers of ischaemia following corrective surgery of CHD – closure of ventricular and atrial septal defects and correction of aortic coarctation – in a newborn girl. Two boys aged 1.5 and 2.5 years presented repeatedly with acute heart failure because of acute ischaemic events and dilative cardiomyopathy. Two children were admitted after successful resuscitation because of ventricular fibrillation. One of them suffered from unknown hypertrophic cardiomyopathy and had been admitted to our hospital at the age of 4 months. The other patient, a 13-year-old girl, had a known left ventricular non-compaction cardiomyopathy with a hypertrophic component, but was not on medication and did not attend regular follow-ups. Our oldest patient, a 16-year-old boy, was referred to our centre for further testing with newly diagnosed hypertrophic cardiomyopathy seen in a routine check-up. None of the patients had an obstructive component.

In all patients, therapy with β blocker was started to reduce heart rate and myocardial contractility. Although there is no clear evidence for β blocker treatment, it is still referred to as first-line therapy.[1,3] In theory, β blockers also prolong diastole and improve coronary artery blood flow.[3] Four of our patients were discharged in improved clinical condition under β blocker treatment. A long-term β blocker therapy is planned contingent upon the future clinical course of the patients and the emerging evidence.

In two patients, clinical signs remained with recurrent myocardial ischaemia, severe reduction of general health, and depression in ST-segments, respectively, echocardiography stress testing positive for ischaemia and ST-elevations in ergometric testing (Fig 3). Both were referred for surgery with exposure of the involved coronary segment on cardiopulmonary bypass (Fig 4). Surgical exposure was successful and recovery was complete and uneventful (Supplementary video 1). Post-surgery speckle tracking echocardiography revealed a normalised deformation pattern (Fig 5). The mean follow-up period after surgery has been 22 months with no major events.

Discussion

Clinical relevance of myocardial bridging is controversial because of the high prevalence found in angiography and autopsy compared with the few symptomatic clinical courses. Basso et al analysed the association between hypertrophic cardiomyopathy-related sudden deaths with myocardial bridging and did not find any significant linkage. Nevertheless, the authors included the possibility of increased risk in individual patients.[4] Most data on myocardial bridging in paediatrics are on children with hypertrophic cardiomyopathy.[2] There are few case reports on symptomatic myocardial bridging in otherwise healthy children.[8,9] Three of our patients showed the typical constellation of hypertrophic cardiomyopathy and myocardial bridging. The other three patients suffered from different underlying cardiac conditions. Two patients had known dilative cardiomyopathy of unknown aetiology and presented with acute ischaemic events. The coronary angiography revealed myocardial bridging as a feasible cause of these acute episodes and a possible cause for the presence of dilative cardiomyopathy.

Another useful tool to detect the localised myocardial dysfunction secondary to irregular coronary perfusion is speckle tracking strain echocardiography at rest and on physical stress testing. Its high sensitivity and specificity in detection of

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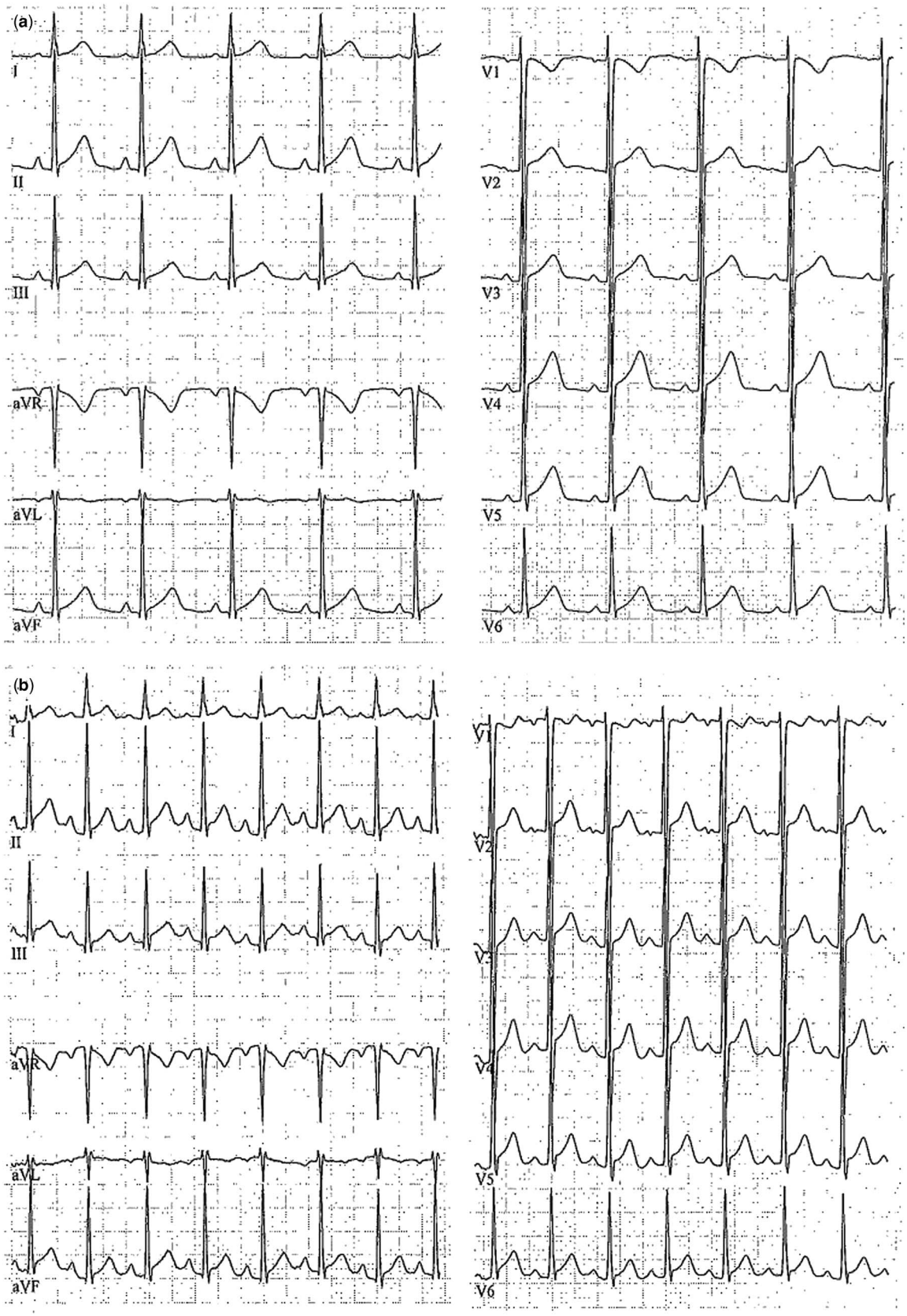

Figure 3. Ergometric testing of patient 5 with signs of myocardial ischaemia with ST-segment elevations in II, aVF, V2-6 at rest (*a*), and under physical stress (*b*). aVF = augmented voltage foot; aVL = augmented voltage left; aVR = augmented voltage right. Limb leads given by I–III and precordial leads given by V1–V6.

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myocardial injury has been extensively reviewed.[12] Although speckle tracking echocardiography at rest was performed in all patients, stress echo testing with strain analysis was only performed in patient number 5, a 13-year-old girl, because there is no appropriate non-invasive test protocol available for young children in our setting. Patient number 6, a 16-year-old boy, was also referred for stress echo testing. However, this patient had initially been suspected to have myocarditis and thus stress testing was thought to be contraindicated.

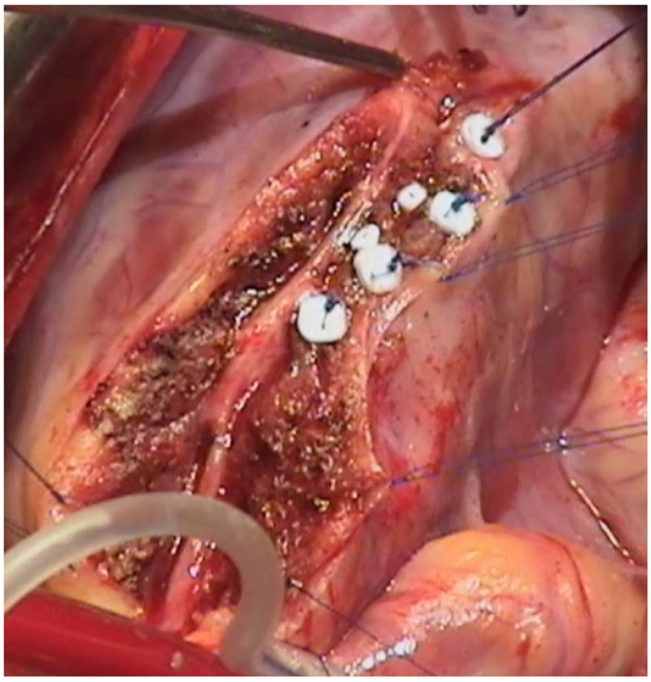

Figure 4. Intraoperative situs of patient 4 after exposure ("unroofing") of the Ramus interventricularis anterior on cardiopulmonary bypass.

Adult interventional cardiologists perform intravascular ultrasound and measurement of functional flow reserve on a more regular base.[3,13] Agrawal et al[13] suggested the use of these techniques for risk stratification in children with anomalous coronary arteries and myocardial bridges. These novel techniques could have completed our diagnostic procedures in our teenage patients. A widespread use of these techniques in the future seems possible.

Conclusion

To the authors' knowledge, the presented series is the largest so far to illustrate varying symptomatic courses of myocardial bridging even in early childhood in children with different underlying cardiac conditions. Myocardial bridging seems to be a rare, but a relevant differential diagnosis of acute ischaemic events in early childhood, especially if combined not only with hypertrophic cardiomyopathy but also with other cardiac diseases. This rare entity should be considered even in young children because it may require urgent treatment and in rare cases surgical intervention.

Supplemental material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951118000409

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Conflicts of Interest. None.

Ethical Standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the local head of the department.

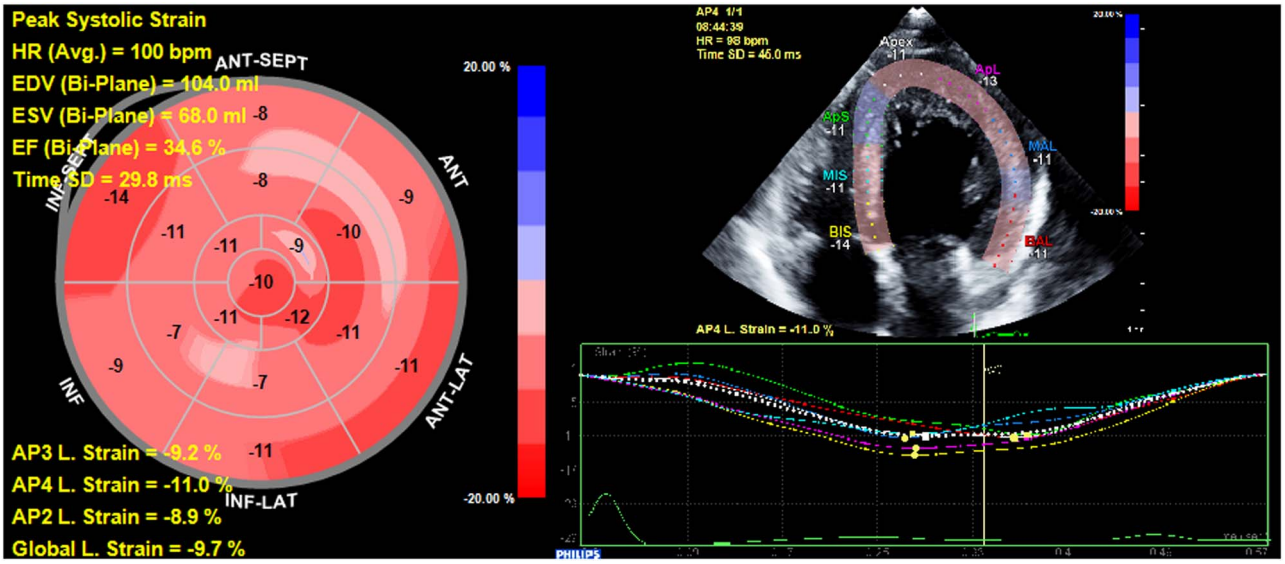

Figure 5. Two-dimensional speckle tracking echocardiography reveals harmonised myocardial deformation after surgery in patient 5 (Epiq 7G, QLab 10.4, aCMQ module). ANT = anterior; AP2 = apical two chamber view; AP3 = apical three chamber view; AP4 = apical four chamber view; ApS = apicoseptal; ApL = apicolateral; BAL = basal anterolateral; BIS = basal inferoseptal; EDV = enddiastolic volume; ESV = endsystolic volume; EF = ejection fraction; HR = heart rate; INF = inferior; LAT = lateral; MAL = mid anterolateral; MIS = mid inferoseptal; SEPT = septal.

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