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Original Article

Cardiac resynchronisation therapy associated with pulmonary artery banding in an adult with severe right ventricular dysfunction after Mustard repair for complete transposition of the great arteries: results after 2 years of follow-up

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Abstract Late dysfunction of the systemic right ventricle in patients with complete transposition of the great arteries after Mustard or Senning procedures and progressive deterioration of the clinical status has been demonstrated. However, evidence-based data on the effective therapy for systemic right ventricular dysfunction in these patients are yet to be defined. Our patient shows an improvement in the right ventricular systolic function, with a reduction in tricuspid regurgitation and a consequent better exercise tolerance after a hybrid approach consisting of an upgrading of a previous transvenous-implanted dual-chamber Implantable Cardiac Defibrillator to biventricular pacing associated with pulmonary artery banding via an anterior thoracotomy.

Keywords: Complete transposition of the great arteries; cardiac resynchronisation therapy; congestive cardiac failure; echocardiography

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Late dysfunction of the systemic right ventricle in patients with complete transposition of the great arteries after Mustard or Senning procedures and progressive deterioration of the right ventricular function and clinical status has been reported previously.[1–3] However, evidence-based data on the effective therapy for systemic right ventricular dysfunction in these patients are yet to be defined. Beneficial effects of cardiac resynchronisation therapy on systemic right ventricular function in small groups of patients with transposition of the great arteries after atrial switch repair have been reported.[4–6] Most of these patients had previous pacemaker implantation because of sinoatrial or atrioventricular conduction disturbances, which might have caused or worsened an electromechanical dyssynchrony,[7] with a detrimental impact on the systemic right ventricular function. Right ventricular dyssynchrony has been documented using Tissue Doppler Imaging in adults after atrial switch operations for transposition of the great arteries and has been associated with right ventricular systolic dysfunction with impaired exercise tolerance.[8]

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Materials and methods

We report the case of a 40-year-old man who underwent the Mustard operation at the age of 1 year for complete transposition of the great arteries, who developed severe right ventricular dysfunction with impaired exercise tolerance despite optimal medical therapy using digoxin, captopril, and carvedilol in the adult age. Severe right ventricular dilatation and systolic dysfunction with moderate–severe tricuspid regurgitation have been documented using echocardiography over the past 3 years, with a

tricuspid annular plane systolic excursion of 7.5 mm and a dP/dT of 420 mmHg/second measured using a tricuspid regurgitation jet. Cardiovascular magnetic resonance imaging confirmed a severely dilated systemic right ventricle – indicised end-diastolic volume of 220 ml/mq – with severe systolic dysfunction – ejection fraction 27% – including different areas with right ventricular myocardial fibrosis showed using late gadolinium enhancement imaging (Fig 1a and b). The systemic and pulmonary vein pathways were normal without obstruction or residual shunts.

The patient had undergone an Implantable Cardiac Defibrillator implantation for primary prevention of sudden cardiac death and a dual-chamber (Dual Dual Dual) device was chosen because of the associated brady-tachy syndrome. The device was implanted via the left subclavian vein, with the bipolar screw-in transvenous leads, respectively, positioned in the systemic left atrium and on the endocardial surface of the pulmonary left ventricular wall. Nonetheless, 1 year later, his exercise tolerance worsened to New York Heart Association class 3. A cardiopulmonary exercise test obtained using a bicycle ergometer revealed a normal rest/stress oxygen saturation (%); reduced systolic blood pressure increase during exercise, with a normal chronotropic response; reduced work load; and peak indicised oxygen consumption and pulse with increased minute ventilation/rate of CO_2 elimination slope (Table 1). The rest electrocardiogram revealed a sinus rhythm with a low-amplitude P wave and a broad QRS complex (182 ms) with right bundle branch block morphology. A Tissue Doppler Imaging study performed during sinus rhythm revealed a significant intraventricular mechanical delay (102 ms) and a significant interventricular delay (145 ms; Fig 2a and b). Surgery was proposed to the patient in order to upgrade the Dual Dual Dual-Implantable Cardiac Defibrillator system to a biventricular pacing system and to reduce his systemic tricuspid regurgitation by remodelling the right ventricular shape using pulmonary artery banding. Through a redo-midline sternotomy, under trans-oesophageal echocardiography monitoring, a pulmonary artery banding was applied and tightened until a subpulmonary left/right ventricular pressure ratio of 2/3 was achieved. Tricuspid regurgitation decreased immediately from severe to moderate.

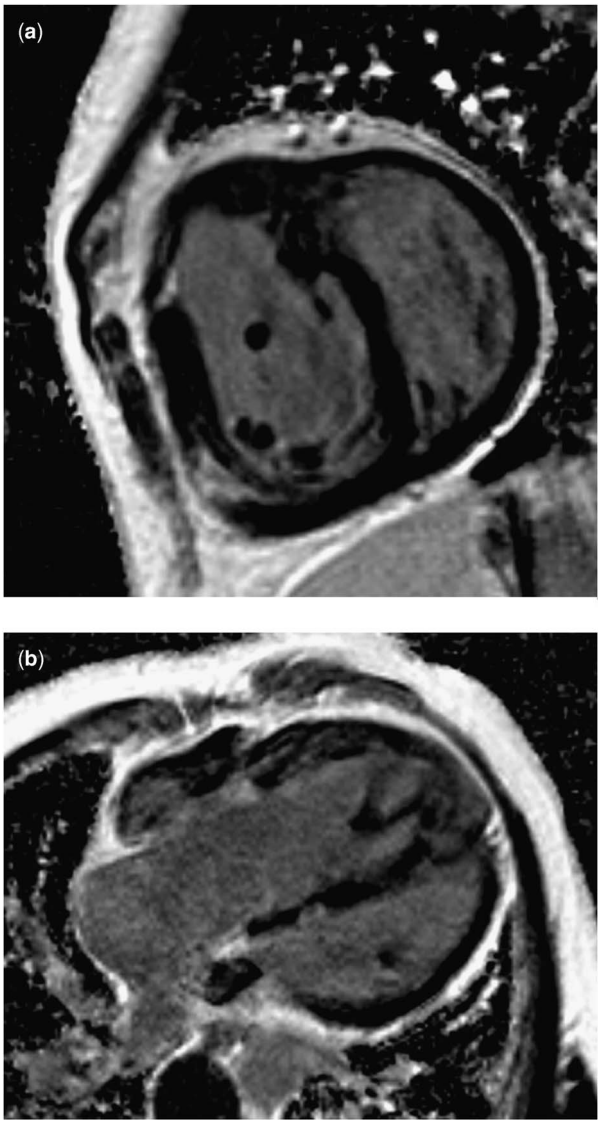

Figure 1.
*Cardiovascular magnetic resonance with late gadolinium enhancement imaging: severely dilated systemic right ventricle with different areas of myocardial fibrosis. (**a**) Short-axis view and (**b**) long-axis view.*

Results

At the 2-year follow-up, his clinical evaluation revealed an improvement in the functional class, from New York Heart Association class 3 to 2, with an appreciable improvement in the cardiopulmonary exercise test. In particular, he showed an improvement in the peak workload, peak indicised oxygen consumption, oxygen pulse, and a reduction in the minute ventilation/rate of CO_2 elimination slope (Table 1). The electrocardiogram revealed sinus rhythm and paced QRS complexes (140 ms). In comparison with the pre-surgical study, the echo Doppler examination revealed a mild tricuspid regurgitation with reduced systolic flattening of the interventricular septum due to the pressure overload imposed on the left ventricle from the pulmonary artery banding, a reduction in the right ventricular end-diastolic dimension, and an improvement in

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Table 1. Parameters obtained using CPET.

	Pre-CRT	Post-CRT
Rest/stress O_2 saturation (%)	96/93	95/93
SBP during exercise (mmHg)	140	135
Peak HR (bpm)	162	149
Watt, (Watt%), METs	80, (40%), 5.4	110, (50%), 6.6
Peak VO_2i (ml/kg/min) (peak VO_2i%)	17 (44%)	22.8 (60%)
O_2 pulse (ml/b) (O_2 pulse%)	6.3 (37%)	10.6 (63%)
VE/VCO_2 slope	37	27.5

CPET = cardiopulmonary exercise testing; CRT = cardiac resynchronisation therapy; HR = heart rate; SBP = systolic blood pressure
Percentage of predicted value is given within parentheses; MET = Metabolic Equivalent of Task

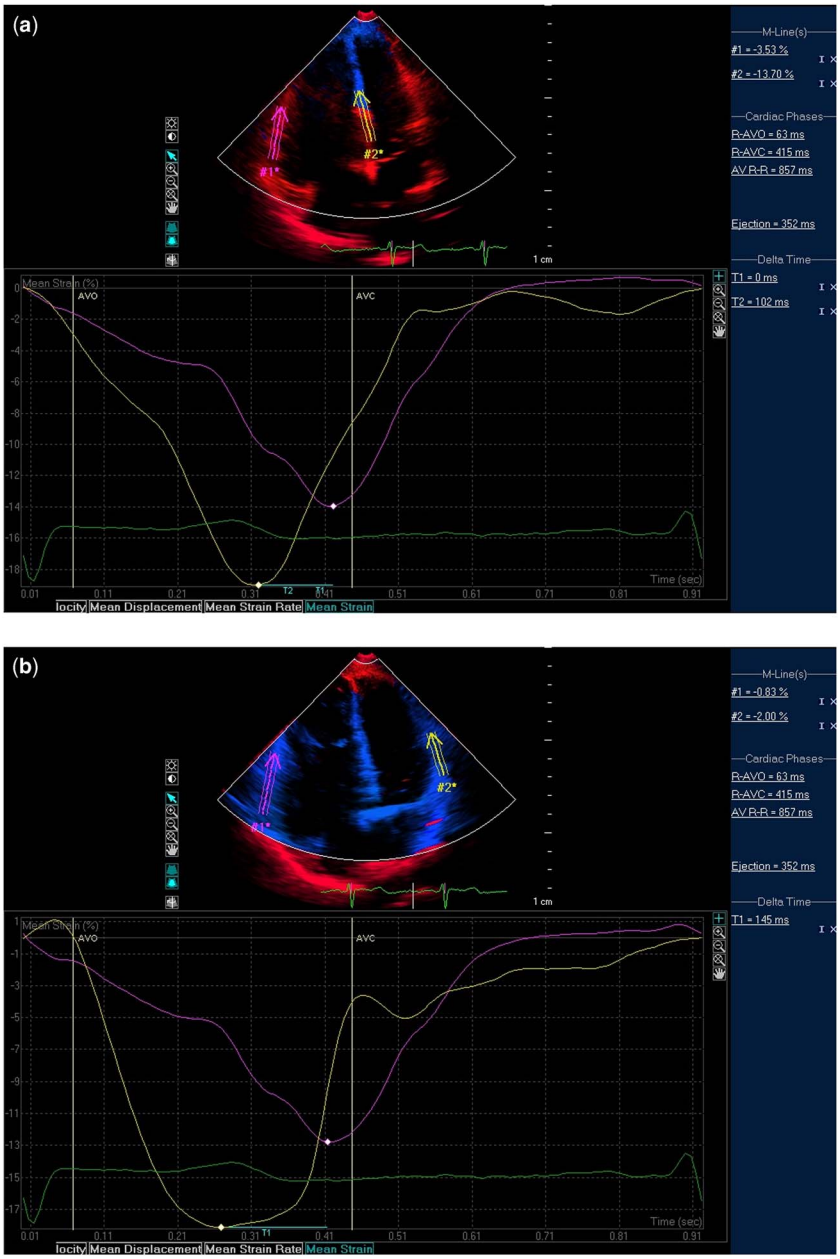

Figure 2.
*Longitudinal strain analysis by Tissue Doppler Imaging (TDI; pre-cardiac resynchronisation therapy): (**a**) intraventricular mechanical delay (102 ms) and (**b**) interventricular mechanical delay (145 ms).*

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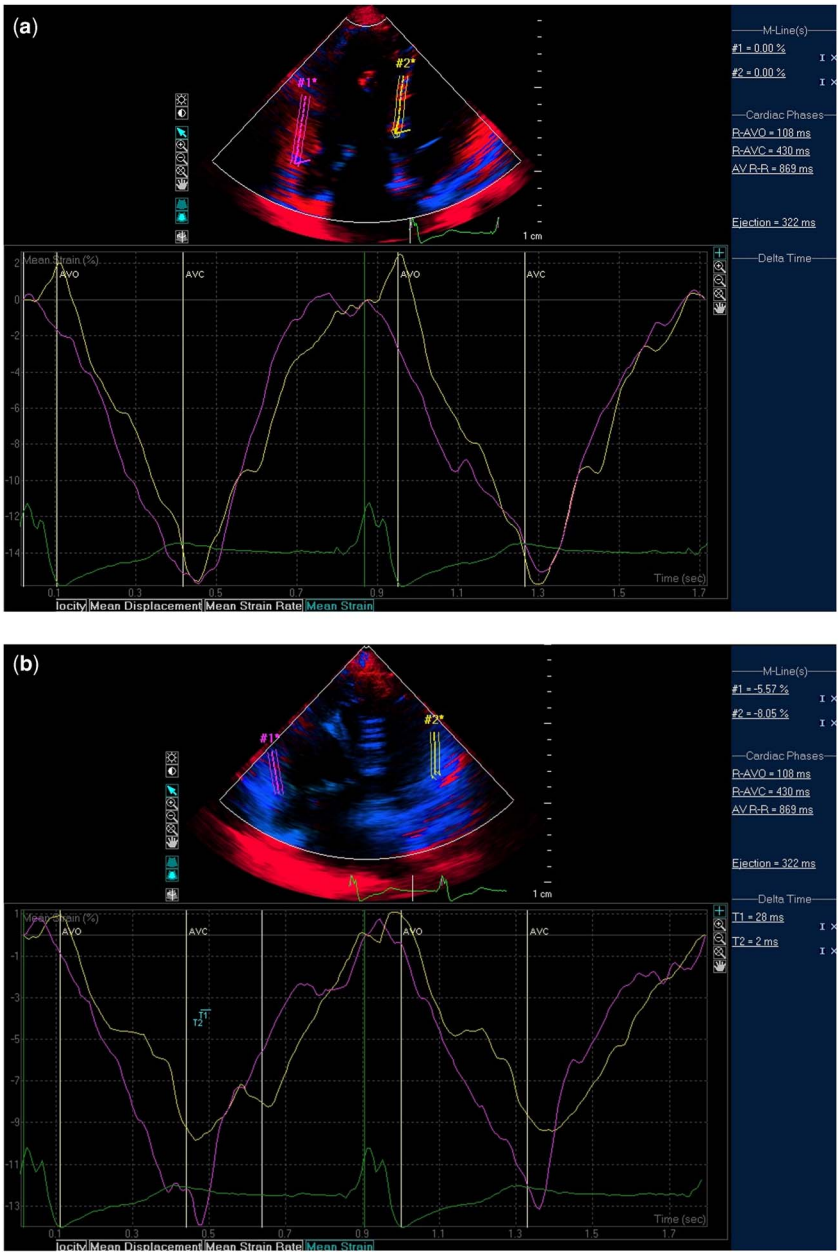

Figure 3.
*Longitudinal strain analysis by Tissue Doppler Imaging (TDI; post-cardiac resynchronisation therapy): (**a**) intraventricular mechanical delay and (**b**) interventricular delay.*

the global systolic function, with an increase in the tricuspid annular plane systolic excursion (12 mm) and dP/dT (770 mmHg/second). The Tissue Doppler Imaging examination of the right and left ventricle performed during biventricular pacing revealed a synchronised mechanical activity of the ventricular chambers, with no significant time delays between the peak longitudinal systolic strains of the right ventricular lateral wall, interventricular septum, and left ventricular lateral wall (Fig 3a and b).

Discussion

Systemic right ventricular dysfunction is well documented in the long-term follow-up after the atrial switch operation for transposition of the great arteries, which was the standard surgical treatment for this congenital heart disease from the early 1960s to the early 1980s. Right ventricular myocardial perfusion defects[9] and reduced myocardial flow reserve[10] have been documented as the causes for ischaemic right ventricular myocardial

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dysfunction. Right ventricular myocardial fibrosis has further been shown in these patients using cardiac magnetic resonance with late gadolinium enhancement imaging.[11,12] Right ventricular mechanical dyssynchrony has also been documented in patients after the atrial switch operation for transposition of the great arteries associated with right ventricular systolic dysfunction and consequent impaired exercise tolerance.[8] The QRS duration has been shown to correlate with right ventricular end-diastolic dimension, suggesting a possible mechanoelectrical relation.[13] Although only 11% of their patients had QRS duration >120 ms and 32% were found to have right intraventricular dyssynchrony, Chow et al[8] failed to demonstrate a significant correlation between right intraventricular mechanical delay and QRS duration. Regional differences in the timing of the right ventricular contraction might cause ineffective right ventricular pressure build-up and reduced pumping function. The improvement with cardiac resynchronisation therapy of the right ventricular systolic function in post-Mustard patients[4–6,14] suggests that mechanical right ventricular dyssynchrony might play a role in the progression of right ventricular dysfunction in this patient group. Right ventricular mechanical dyssynchrony could also explain the high prevalence of significant tricuspid regurgitation in post-Mustard patients with right ventricular systolic dysfunction. However, the extent to which intraventricular and interventricular dyssynchrony contributes to the severity of right ventricular systolic dysfunction remains undefined. In contrast to severe left ventricular systolic dysfunction with mechanical dyssynchrony, there are no specific guidelines for cardiac resynchronisation therapy in systemic right ventricular failure in adults with congenital cardiac disease. An analysis of feasibility and early experience in this cohort of patients demonstrated that 4–9% of patients with systemic right ventricle appear to be potential candidates for cardiac resynchronisation therapy by using the inclusion criteria used in landmark cardiac resynchronisation therapy trials[15,16] such as: New York Heart Association class >3, at least moderate ventricular dysfunction, at least moderate ventricular dilatation, and a QRS duration >120 ms. In the last European Society of Cardiology guidelines for Grown-Up Congenital Heart Disease Patients,[17] cardiac resynchronisation therapy for systemic right ventricular failure after atrial switch operation is still considered experimental.

In our patient, we found an improvement in the right ventricular systolic function, with a reduction in tricuspid regurgitation and a consequent better exercise tolerance at 2-year follow-up after cardiac resynchronisation therapy and pulmonary artery banding. These results could be attributed to the right ventricular mechanical resynchronisation associated with pulmonary artery banding with consequent septal shift, which might also have contributed to the reduction in the tricuspid regurgitation, as previously described.[18] Owing to the fact that the transposition of the great arteries after atrial switch repair with severe right ventricular systolic dysfunction belongs to the category of congenital defects with the greatest known risk of late sudden cardiac death, we opted to implant an Implantable Cardiac Defibrillator during primary prevention.[19,20] Medical therapy with digoxin and diuretics remains the cornerstone for systemic right ventricular failure, whereas the role of angiotensin-converting enzyme inhibitors and β-blockers is still controversial.[17] Late arterial switch after left ventricular retraining has been described to be a surgical option to cardiac transplantation in children or young adults with severe right ventricular dysfunction after atrial switch repair.[18]

There are some technical aspects to consider. First, there is not yet an accepted method for the assessment of mechanical right ventricular dyssynchrony. There is a potential advantage in using strain analysis by Tissue Doppler Imaging or speckle tracking, because these allow direct assessment of the degree and timing of myocardial deformation during systole and better differentiation between active contraction and passive displacement compared with tissue velocity mapping. This is of particular relevance in patients with segmental myocardial fibrosis,[21] as shown in those who underwent atrial switch operations.[11] Second, the technical approach adopted in our patient consisted of a hybrid implantation with endocardial leads previously implanted in the left atrium and pulmonary left ventricle, whereas after 1 year an epicardial lead was placed on the systemic right ventricle associated with pulmonary artery banding via an anterior thoracotomy. The coronary sinus is ontogenetically part of the morphologic left atrium and is positioned adjacent to the systemic right ventricle in congenitally corrected transposition of the great arteries, as opposed to the pulmonary left ventricle in transposition of the great arteries after atrial switch repair.[22,23] Therefore, in patients who have undergone Mustard/Senning procedures, the coronary sinus and the tributary veins drain blood from the subpulmonary left ventricle. As a consequence, the patients require surgical electrode placement on the systemic right ventricle for cardiac resynchronisation therapy after switch repair for transposition of the great arteries.

We obtained beneficial clinical and functional results after 2 years of cardiac resynchronisation therapy in a 40-year-old man with severe systemic right ventricular dilatation and systolic dysfunction, with right ventricular mechanical dyssynchrony evaluated using Tissue Doppler Imaging. The hybrid approach adopted consisted of an upgrading to biventricular pacing of a previously implanted Dual Dual Dual system by placing an epicardial lead on the systemic right ventricle associated with pulmonary artery banding via an anterior thoracotomy. Other surgical options such as orthotopic cardiac transplantation could therefore be postponed. Large studies are required to establish indications and timing for cardiac resynchronisation therapy, as well as long-term effects on ventricular function and outcome in a variety of adult patients with congenital heart disease.

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