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Brief Report

Left and right ventriculo-arterial coupling in a patient with congenitally corrected transposition

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Abstract The single beat method was used to evaluate right and left ventriculo-arterial coupling in an asymptomatic patient with congenitally corrected transposition. The ratio of ventricular end-systolic to arterial elastances was normal for the left ventricle coupled to the pulmonary circulation, and depressed for the right ventricle coupled to the systemic circulation. This result suggests that chronic uncoupling of the right ventricle to the systemic circulation might play a role in the pathophysiology produced by congenitally corrected transposition.

Keywords: Congenital heart disease; heart failure; right ventricle

The clinical course of severe pulmonary hypertension is determined by the ability of the chronically overloaded right ventricle to adapt its output to the peripheral demand for oxygen. Recent studies using the relationship between pressure and volume indicate that the right ventricle, in the setting of pulmonary hypertension, presents with an increased contractility, but an imperfect coupling to increased afterload.[1] Congenitally corrected transposition is a rare congenital cardiac malformation associating discordant atrio-ventricular and ventriculo- arterial connections.[2] These patients offer a natural model of a morphologically right ventricle facing systemic pressures from birth. Usually, they lead a normal life until adulthood, and then start to deteriorate, generally in the third decade of life, due to progressive failure of the systemic ventricle.[2] The reasons why the morphologically right ventricle appears adapted to systemic pressures during decades, and then progressively fails, are not completely understood. We present here measurements of left and right ventriculo-arterial coupling in an asymptomatic patient with congenitally corrected transposition. The results may suggest that the morphologically right ventricle is already uncoupled from the systemic circulation before the clinical onset of systemic ventricular failure.

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Case report

A 17-year-old man with known congenitally corrected transposition was referred for surgical closure of a large atrial septal defect. He gave informed consent to the present study, which was approved by the Institutional Review Board. A transoesophageal echocardiogram had demonstrated an atrial septal defect, which was too large for interventional closure, and normal atrioventricular valves. The indication for closure of the atrial septal defect was a previous transient ischaemic attack, which had been attributed to a paradoxical embolism. The patient was asymptomatic. His clinical examination was unremarkable. Under general anaesthesia using midazolam and sufentanyl, we floated a balloon-tipped thermodilution Swan Ganz catheter (131H-7F; Baxter Edwards, Irvine, CA) into the pulmonary artery, placed 5 french micromanometer-tipped Millar catheters (SPC 350; Millar Instruments, Houston, TX) into the two ventricles, and fitted non-constricting transonic flowmeters (T206; Transonic Systems, Ithaca, NY) around the pulmonary trunk and the aorta. We then measured pulmonary vascular pressures, pulmonary and aortic blood flow, and right and left ventricular pressures before proceeding with

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uneventful closure of the atrial septal defect. End-systolic right ventricular and left ventricular elastances, aortic and pulmonary elastances, and ventriculo-arterial coupling calculated as the ratio of ventricular end-systolic to arterial elastances, were derived using a single beat approach, as previously reported.[3]

The results show a pulmonary vascular resistance of 1.9 Wood units, suggesting absence of pulmonary hypertension, near-equal pulmonary and systemic flows, indicating absence of a left-to-right shunt at the time of the measurements, a normal ratio of left ventricular end-systolic to arterial elastances, and a markedly decreased ratio of right ventricular end-systolic to arterial elastances (Fig. 1, Table 1).

Discussion

The present results suggest that, in this asymptomatic patient with congenitally corrected transposition, the coupling between the morphologically right ventricle and the systemic circulation was altered, while the morphologically left ventricle was adequately coupled to the pulmonary circulation. Evaluation of right ventricular function is notoriously difficult because of its complex geometry and its non-concentric pattern of contraction. Recent progress in techniques for imaging, and development of a single beat approach, has improved the understanding of the adaptation of the morphologically right ventricle to chronic pressure overload in humans.[1] These results indicate that the right ventricle adapts to pulmonary hypertension, essentially by increasing its contractility, and that this can be quantified by determination of end-systolic and arterial elastances. The calculated ratio of ventricular end-systolic to arterial elastances can then be used as a measure of the adequacy of coupling.[1] Previous studies in experimental animals with various types of acute or chronic pulmonary hypertension have validated this approach, which requires the measurement of right ventricular pressures and volumes,[1] or an integration of pulmonary arterial flow.[3–5] Indeed, the measurement of end-systolic elastance as a load-independent index of contractility can be obtained either by recording pressure and volume over multiple cardiac cycles with progressively decreasing venous return, or by a determination during a single cycle of a maximum right ventricular pressure from nonlinear extrapolation of early and late systolic portions of the right ventricular pressure curve, measuring relative changes in volume by integration of the pulmonary arterial flow.[1,3] Single beat determinations of right ventriculo-arterial coupling may also allow for a better understanding of the effects of pharmacological interventions with concomitant inotropic and vascular effects.[3] In all these studies, the normal ratio of ventricular end-systolic to arterial elastances appears to be between 1.5 and 2, in keeping with mathematical modeling, suggesting that this value is compatible with maximal mechanical efficiency.[6]

Ventricular pressure-volume loops normally present with a square shape for the morphologically left ventricle, and a triangular shape with a rounded left shoulder for the morphologically right ventricle.[7,8] These aspects were mirror-imaged in our patient, which is in keeping with a previous report on left ventricular pressure-volume relationships in patients after the Mustard procedure.[9] This confirms the notion

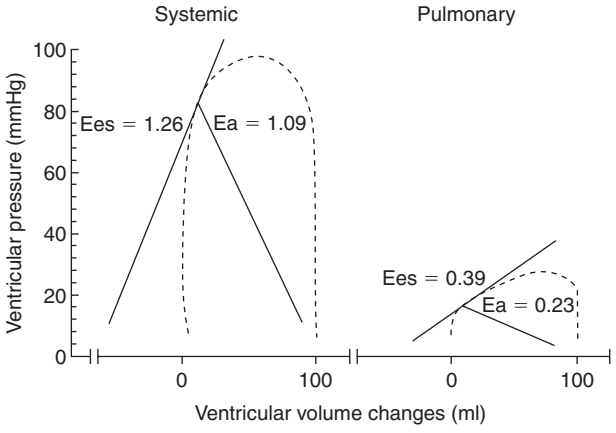

Figure 1.
Pressure-volume relationship of the morphologically right ventricle in systemic position (left panel), and of the morphologically left ventricle in pulmonary position (right panel). Ees: end-systolic ventricular elastance; Ea: effective arterial elastance.

Table 1. Haemodynamic and ventriculoarterial coupling data in an asymptomatic patient with congenitally corrected transposition.

Variables	Systemic right ventricle	Pulmonary left ventricle
Flow (l/min/m^2)	6.9	7.1
Systemic arterial pressure (mmHg)	85	–
Pulmonary artery pressure (mmHg)	23	–
Right atrial pressure (mmHg)	7	
Pulmonary artery occluded pressure (mmHg)	10	–
End-systolic elastance (mmHg/ml)	1.26	0.39
Arterial elastance (mmHg/ml)	1.09	0.23
End-systolic to arterial elastances ratio	1.16	1.67

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that the shape of pressure-volume loops depends on pressure loading conditions, rather than on intrinsic properties of the myocardium.[9]

In our patient, the left ventriculo-pulmonary arterial coupling appeared to be normal, indicating an adapted decrease in left ventricular contractility to the low resistance in the pulmonary arterial circuit. Right ventriculo-systemic arterial coupling, in contrast, was markedly altered, with a value of 1.16, decreased by around half compared to previously reported normal values.[10] This may indicate that the morphologically right ventricle is structurally unable to optimize its systolic function to a systemic arterial hydraulic load.

In the present study, ventriculo-arterial coupling was determined during general anaesthesia. Anaesthetic drugs may have major effects on contractility and vascular tone. There have been no reports until now on such potential effects on ventriculo-arterial coupling. Since in our patient left ventriculo-pulmonary arterial coupling was normal, however, a major uncoupling effect of the given doses of midazolam and sufentanyl is unlikely.

It may appear surprising that the patient had no increase in the ratio of pulmonary-to-systemic flows, in spite of the presence of a large interatrial communication. This may be explained by the fact that the value obtained in the occluded pulmonary arteries had probably slightly overestimated left atrial pressure, so that there was no significant left-to-right atrial pressure gradient to generate a left-to-right flow at the time of the haemodynamic exploration.

The biological mechanisms of systemic right ventricular failure in patients with congenitally corrected transposition are unkown, but could well be related to the mechanical stress of chronic uncoupling of the morphologically right ventricle to an excessive arterial load.

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