LEVOSIMENDAN IN SINGLE VENTRICLE HEART FAILURE AFTER LONGTERM SURVIVAL OF A MODIFIED BLALOCK-TAUSSIG SHUNT

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Abstract

We report the case of a 44 year-old patient with complex ACHD, admitted with acute decompensated heart failure (ADHF) in hemodynamic profile B. He had a single ventricle with pulmonary atresia, previously submitted to three modified Blalock-Taussig shunts (mBTs) at the age of 2, 12 and 19 years old. Despite conventional treatment with diuretics, β-blockers (BB) and isosorbide dinitrate the patient progressed to profile C and the transthoracic echocardiogram disclosed a reduced systolic function. Likewise, levosimendan was commenced and an appropriate decongestion and a marked reduction in the NT-proBNP were seen. Treatment with angiotensin-converting-enzyme inhibitor, BB, ivabradine and mineralocorticoid receptor was optimized. The patient was discharged home after 26 days in NYHA class III and referred for heart transplant after right heart catheterization. To our knowledge, this is the first report of successful levosimendan's use in ADHF in a mBTs long-term survivor.

INTRODUCTION

A single ventricle anomaly accounts for 1-2% of the CHD and is frequently associated with other cardiac abnormalities.[1] The adequacy of pulmonary blood perfusion will determine the initial clinical presentation (degree of cyanosis) and the need for a surgical intervention. Blalock-Taussig shunt is a surgical procedure first attempted in 1944 by Alfred Blalock and Helen Taussig. Its purpose is to enable blood oxygenation by joining the subclavian and pulmonary arteries. Nowadays, a prosthetic graft tube is used to create the shunt (modified Blalock-Taussig shunt - mBTs). This systemic-to-pulmonary artery shunt optimizes pulmonary blood perfusion until neonates can grow enough to be submitted to a more definitive procedure, a Fontan-like surgery.[2] We report a case of acute decompensated heart failure (ADHD) in a mBTs long-term survivor patient with a single ventricle who was successfully treated with levosimendan.

CASE DESCRIPTION

A 44 year-old man with a single ventricle with pulmonary atresia, previously submitted to three mBTs at the age of 2, 12 (Figure 1) and 19 years old (this last procedure was a Fontan-like surgery attemptation that ended in another mBTs (due to technical problems), presented with dyspnea on exertion, peripheral edema, orthopnea and paroxysmal nocturnal dyspnea shortly after arriving from an intercontinental flight. He recalled having coryza 2 weeks before. On physical examination the patient was tachycardic, hypertensive and tachypneic. He was afebrile, cyanotic and the peripheral oxygen saturation was 80% while breathing with a facial mask at 40% fraction of inspired oxygen.

Pulmonary congestion, lateral and inferior ictus cordis, and a grade 4 systolic murmur with a thrill were noticed. The electrocardiogram showed sinus rhythm, right bundle branch block and marked repolarization abnormalities. The hemoglobin was 21.9 g/dL, the N-terminal pro-B-type natriuretic peptide was 2324 pg/mL, and troponin I was 2.19 ug/L. Serum chemistry was unremarkable. A transthoracic echocardiogram (TTE) performed at admission (Figure 2) documented a severe ventricular systolic dysfunction, not present in previous exams. A thoracic CT angiography confirmed shunt patency (Figure 3) and had no signs of pulmonary emboli.

Conventional ADHF treatment (diuretics, beta-blockers, intravenous dinitrate) was initiated but the patient

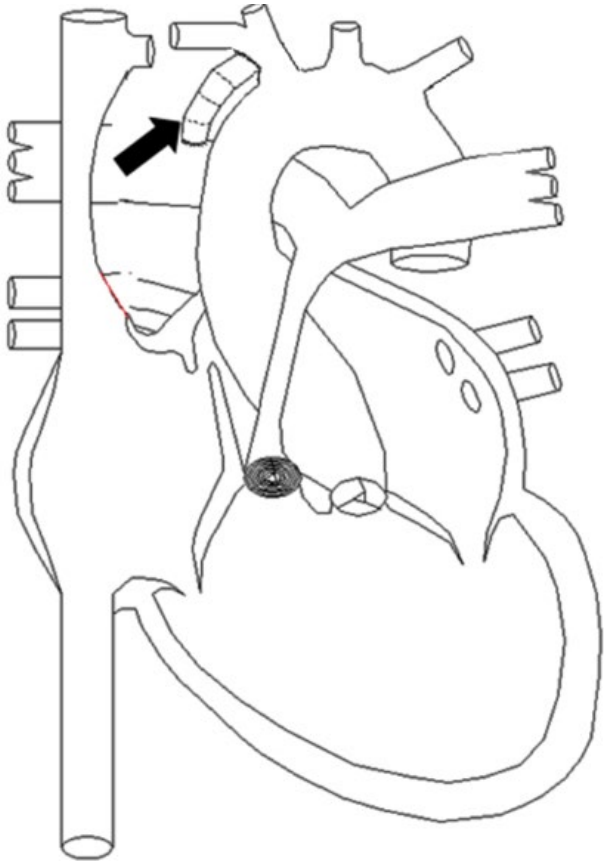

Figure 1 *Patient anatomy. High complexity adult congenital heart disease with univentricular phisiology with two atrioventricular valves opening in a single ventricle, pulmonary artery atresia and anterior and left position Aorta. He had a functional right BTS (arrow).*

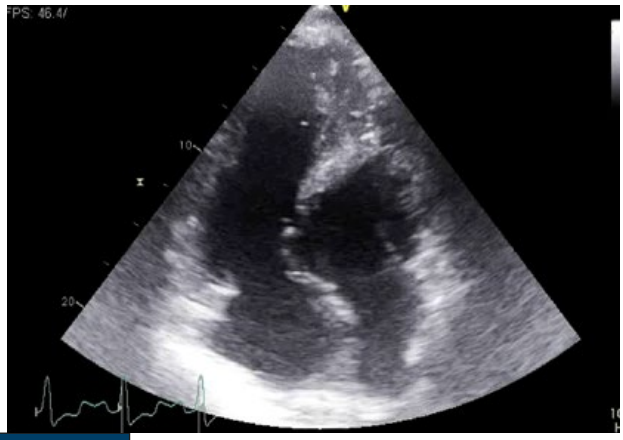

Figure 2 *Admission transthoracic echocardiogram documented a severe ventricular systolic dysfunction, not present in previous examinations.*

progressed to profile C. Levosimendan was added at a perfusion rate of 0.05 mcg/kg/min and maintained for 24 hours. There was a favorable clinical improvement with appropriate decongestion and a marked reduction of the NT-proBNP (Figure 4). A brief episode of atrial fibrillation was observed 24 hour after levosimendan's perfusion

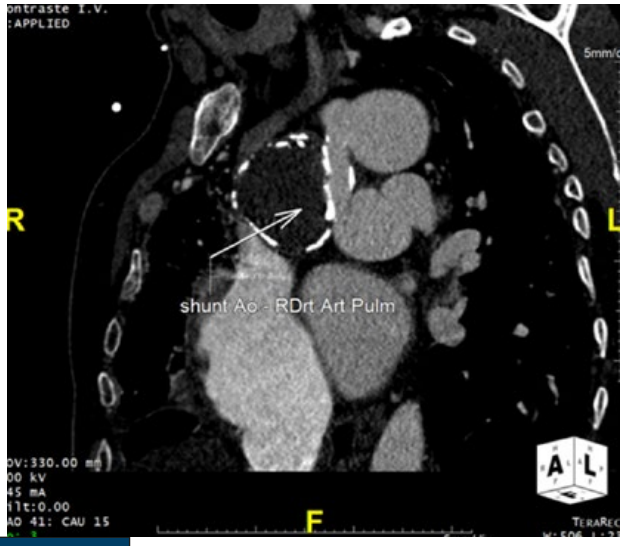

Figure 3 *Left-anterior sight thoracic computed tomography angiography confirmed a patent Blalock-Taussig shunt (arrow).*

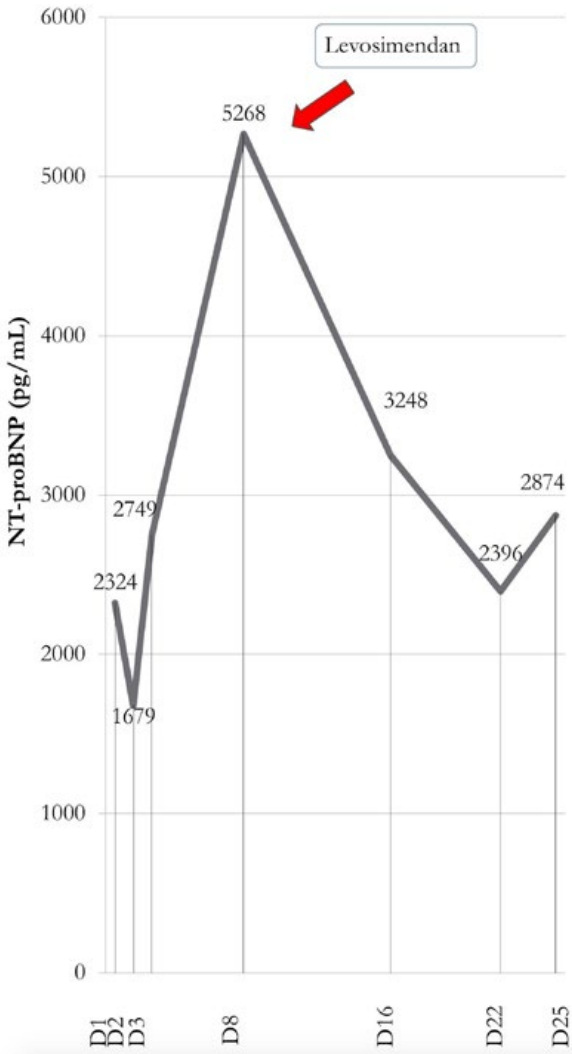

Figure 4 *NT-pro-BNP values evolution, in days after admission (D). There is a coincidence between a drop of Pro-BNP values and the introduction of levosimendan (arrow).*

and successfully managed with amiodarone and correction of the hypomagnesaemia.

Optimization of treatment with carvedilol, ramipril, eplerenone and ivabradine was well tolerated. A repeated TTE 5 days after levosimendan showed qualitative improvement in ventricular systolic function.

A moderate increase in pulmonary artery pressure (PAP) (right PAP 44/31 (38) mmHg; left PAP 48/30 (42) mmHg), and moderately elevated pulmonary vascular resistance (PVR) 3,4 Wood units (mm Hg.min/L) were documented. The patient was discharged home on the 26th day, in functional class III of the New York Heart Association (NYHA) and referred for heart transplant.

At 3 months' follow-up the patient was in NYHA class II. One year later the patient was submitted to heart tranplant in another hospital, but unfortunately died during the procedure due to early right ventricular graft failure.

DISCUSSION

We report a case of ADHF in a mBTs long-term survivor patient with a single ventricle who was successfully treated with levosimendan. This patient survived a total of 44 years (25 years after the last mBTs) without great limitation of his daily routine. To our knowledge, there are only three case reports of such a long-term survival in patients with similar clinical background.[3,4]

We used conventional adult heart failure treatment but, as the patient progressed with hypoperfusion and congestion, inotropic therapy was commenced. Levosimendan was our first choice for inotropic support as the patient was on beta-blockers and systolic blood pressure was above 90mmHg. We decided not to use loading dose. A marked reduction of the brain natriuretic peptide coupled with appropriate decongestion ensued.

Rafik et al[5] reported two cases of complex congenital heart disease [atrial and ventricular septal defect (VSD) and pulmonary atresia with a large VSD] with chronic end stage heart failure treated with pulsed levosimendan. To our knowledge, this is the first report of successful use of levosimendan in acute heart failure of a mBTs long-term survivor. This case report is an example of the unique challenge of adult congenital heart disease management.

Acknowledgments

We would like to thank Dr. João Sá and Dr. Alexandra Bayão Horta for manuscript correction and Dr. Rui Anjos, Dr. Natália Marto and Dr. Daniel Ferreira for patient medical co-management.

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