Cardiology in the Young

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Transcatheter systemic atrioventricular valve-in-valve implantation in a congenitally corrected transposition of the great arteries patient

Miguel Martins de Carvalho[1,3], Ana Paula Machado[2], Rui André Rodrigues[1], João Carlos Silva[1], Cristina Cruz[1] and Filipe Macedo[1,3]

[1]Department of Cardiology, São João Universitary Hospital, Oporto, Portugal; [2]Department of Obstetrics, São João Universitary Hospital, Oporto, Portugal and [3]Cardiovascular R&D Center, Universidade do Porto Faculdade de Medicina, Portugal

Brief Report

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Author for correspondence:
Miguel Martins de Carvalho, Department of Cardiology, Alameda Prof. Hernani Monteiro, 4200-319 Porto, Portugal. Tel: +351967096459. E-mail: miguel.martins.carvalho@chsj.min-saude.pt

Abstract

We present an asymptomatic pregnant patient with congenitally corrected transposition of the great arteries and severe atrioventricular bioprosthesis regurgitation – with increased maternal and fetal risk due to volume overload. She was considered high risk for reintervention and was submitted to an off-label post-partum transcatheter valve-in-valve implantation with a Sapiens 3 valve. The procedure was successful, and she remains asymptomatic 30 months after – and even went through another successful pregnancy.

Congenitally corrected transposition of the great arteries is a CHD characterised by double discordance (atrioventricular and ventriculoarterial) and a systemic right ventricle. The aorta is usually anterior and to the left of the pulmonary artery. The prevalence is less than 1% of all CHD,[1] and it is almost always associated with other abnormalities.[2,3]

Clinical manifestations can range from asymptomatic to fatigue/exercise intolerance, arrhythmias, and cyanosis. Nonetheless, as these patients have a systemic right ventricle, which is not built to support systemic pressures, they will inevitably develop heart failure. Also, the systemic atrioventricular valve (morphologically tricuspid) is prone to develop regurgitation (perpetuating the volume overload cycle).[4] This impairs patient's quality of life and is associated with significant morbi-mortality.[5] Pregnancy is associated with volume overload. In congenitally corrected transposition of the great artery patients, it is linked to an increased risk (mWHO class III), and it can lead to worsening atrioventricular valve regurgitation and heart failure. The gold standard for the treatment of severe valve disease is surgery. However, in patients with a high surgical risk, transcatheter valve implantation may be an option. In the context of CHD, percutaneous interventions are a promising option especially in patients with multiple previous cardiac surgeries. While there is considerable experience with transcatheter pulmonary valve implantation, there are still few examples of valve implantation in the atrioventricular position due to bioprosthesis dysfunction.

Clinical case

A 23-year-old female patient with a previous medical history of congenitally corrected transposition of the great artery presents to the outpatient clinic in December 2019 due an unplanned pregnancy, with 26 weeks gestation. The patient had been diagnosed with congenitally corrected transposition of the great artery by the age of 7 years old, after the finding of a heart murmur on a routine medical observation. The patient also had a history of rheumatic fever (with Sydenham Chorea) as a child and, at 17 years old (in 2013), she was submitted to a systemic atrioventricular valve replacement with a 29-mm Carpentier-Edwards bioprosthesis, due to progressive atrioventricular valve regurgitation. Afterwards, the patient was in NYHA class I and was seen in the outpatient clinic once a year.

As the patient was pregnant, a transthoracic echocardiogram was performed which revealed severe bioprosthesis regurgitation and mild systolic dysfunction of the systemic right ventricle. A transesophageal echocardiogram showed extensive bioprosthesis dysfunction, with two flail leaflets. After a Pregnancy Heart Team discussion, due to her high surgical and pregnancy risk, a transcatheter valve-in-valve implantation was planned, ideally post-partum as the patient was asymptomatic. An elective caesarean section was performed at 34 weeks gestation to minimise the risk of potential obstetric complications. One week after successful delivery, on March 2020, a Sapiens 3 Ultra 26 valve was implanted in the systemic atrioventricular valve position with an antegrade approach. The valve was successfully inserted via trans-septal puncture (preceded by

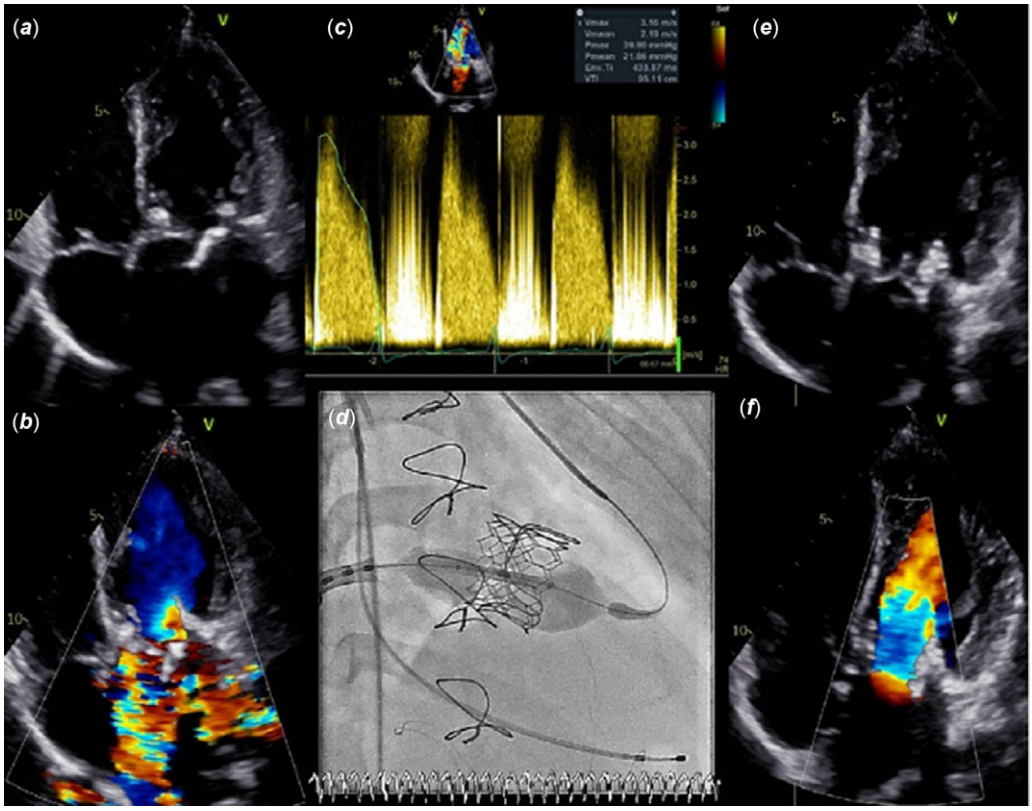

Figure 1. (***a and b***) Apical four chambers – exuberant prosthetic regurgitation with flail leaflet; (***c***) transprosthetic obstructive gradients; (***d***) Transcatheter systemic valve in valve implantation (balloon dilation during rapid ventricular pacing); (***e and f***) Apical four chambers – normofunctional prothesis.

balloon dilatation), with transoesophageal echocardiogram and fluoroscopy guidance. An arteriovenous wire loop technic was applied: a guidewire was inserted in the right femoral vein access; using a snare inserted from the left femoral artery access, the guidewire was snared and pulled until externalisation. During valve deployment, rapid ventricular pacing (temporary pacemaker inserted via internal jugular vein access in the subpulmonary ventricle) was applied. These two manoeuvres prevented valve malposition or embolisation. There were no signs of interference with the adjacent cardiac structures. The mean final atrioventricular gradient was 7 mmHg. There were no periprocedural complications. The patient was discharged 3 days after this intervention. The patient was on anticoagulation with warfarin and remained asymptomatic. In October 2020, she presented with a second unplanned pregnancy. She remained asymptomatic throughout pregnancy and with a successful delivery, with a transthoracic echocardiogram showing preserved biventricular systolic function and a normofunctional valve-in-valve. During a 30-month follow-up, the patient was stable in NYHA I functional class, without any hospitalisations. The transthoracic echocardiogram showed a mean valve-in-valve bioprosthesis gradient of 8 mmHg.

Conclusions

There is a growing experience in transcatheter interventions for CHD. The Sapiens 3 balloon expandable valve can be used for valve-in-valve procedures in degenerated bioprothesis.[6,7] Nowadays, it is an off-label use in patients who have a high risk for surgery. To our knowledge, this was the first case of transcatheter systemic atrioventricular valve-in-valve implantation in a post-partum patient.[8] There is also another published report of a similar case[9] in literature, also with a good result at 1 year of follow-up. Despite the same pathology, the valve implantation technic differed – an arteriovenous loop technic was used in our case. These percutaneous interventions may be a feasible option in selected CHD patients with high surgical risk, after careful Heart Team discussion.

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Conflicts of Interest. None.

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