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Transcatheter Structural Heart Disease Intervention –From ready-made to custom-made therapy

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Short Running Title: TMVR in Congenital Heart

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COI: Dr. Miura is a consultant for Japan Lifeline. Dr. Taramasso is a consultant for Abbott Vascular, Boston Scientific, 4tech, and CoreMedic; and has received speaker honoraria from Edwards Lifesciences. Dr. Zuber is a consultant for Abbott and Edwards Lifesciences. Dr. Maisano is a consultant for Abbott Vascular, Medtronic, Edwards Lifesciences, Perifect, Xeltis, Transseptal solutions, Cardiovalve, Magenta; receives Grant Support from Abbott Vascular, Medtronic, Edwards Lifesciences, Biotronik, Boston Scientific Corporation, and receives royalties from Edwards Lifesciences, 4 Tech, Transseptal solutions, Perifect, and Cardiovalve.
Transcatheter mitral valve repair (TMVR) using the MitraClip system (Abbott Vascular, Santa Clara, CA) has emerged as an alternative treatment for patients with severe symptomatic mitral regurgitation (MR) who are considered inoperable or at high surgical risk. TMVR is rapidly extending to more complex anatomies. Herein, we present the case of a patient with severe symptomatic MR who was also suffering from congenitally corrected transposition of the great arteries (ccTGA).

A 71-yr-old male patient presented with dyspnea NYHA IV and liver cirrhosis. He had ccTGA with heterotaxy (Figure A), and he underwent surgical resection of left ventricular outflow obstruction, and closure of atrial and ventricular septal defect thirteen years ago. Transesophageal echocardiography revealed severe regurgitation with anatomically mitral valve with systolic anterior movement (Figure B, Video S1, S2). After heart team evaluation, the patient was considered eligible for TMVR using MitraClip. A MitraClip XTR was deployed in the middle of mitral leaflets (Figure C), resulting in reduction of MR from severe to trivial (Figure D, Video S3). Unfortunately, he died due to progression of renal failure a few weeks after the procedure.

In technical aspects, steerable guide catheter should be upside-down for adapting the heterotaxy. Moreover, it is difficult to get clear images due to complex anatomy and post-heart surgery.

In the current era of transcatheter intervention, TMVR has been growing mature. The challenges for the anatomy is not only mitral valve but also the complexity of the heart itself. The indication of transcatheter structural heart disease intervention will expand from ready-made to custom-made therapy.
Figure legend

A. Computed tomography revealed heterotaxy.

B. Preprocedural transesophageal echocardiography revealed severe regurgitation with anatomically mitral valve.

C. MitraClip XTR(Abbott Vascular, Santa Clara, CA) was deployed in the middle of mitral leaflets.

D. Postprocedural transesophageal echocardiography revealed improvement of regurgitation from severe to trivial.
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About congenitally corrected transposition of the great arteries

Congenitally corrected transposition of the great arteries (ccTGA) has discordance at both the atrioventricular and the ventriculoarterial level and its incidence has been reported to be around 1/33,000 live births. 4% of the patients have heterotaxy. The anatomy of ccTGA involves systemic venous blood flow to morphologic right atrium, which is connected via the mitral valve to the morphologic left ventricle. And its outflow connect the pulmonary arterial system. The tricuspid valve is located at the entrance to the morphologic right ventricle on the left, and usually it has a dysplastic leaflet with abnormal thickened chordal attachments of the septal and posterior leaflets. Thus patients with ccTGA often suffer from tricuspid regurgitation, and it is rare to suffer from mitral regurgitation.

Video Legend

**Video 1**: Transoesophageal echocardiography showing severe regurgitation with anatomically mitral valve with systolic anterior movement.

**Video 2**: Transoesophageal echocardiography showing severe regurgitation with anatomically mitral valve with systolic anterior movement.

**Video 3**: Transoesophageal echocardiography showing in reduction of MR from severe to trivial after MitraClip.