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Complex cyanotic congenital heart disease and its complications- it is never too late to treat

Radu DN.¹; Stoica S.²; Dorobantu DM.¹; Olaru-Lego G.¹; Buta AS.¹; Platon P.¹; Iosifescu A.¹; Coman IM.¹; Ginghina C.¹; Popescu BA.¹; Enache R.¹

¹Carol Davila University of Medicine and Pharmacy, Eurocolab, Prof. Dr. C. C. Iliescu Institute, Bucharest, Romania

²Bristol Heart Institute, Bristol, United Kingdom of Great Britain & Northern Ireland

Noncardiac complications significantly contribute to the morbidity and mortality of adults with congenital heart disease (CHD), mainly because life expectancy and quality of life for those born with CHD have greatly improved in the last decades. Double-outlet right ventricle (DORV) with a subaortic ventricular septal defect (VSD) associated with subpulmonary stenosis is a complex cyanotic congenital heart disease from the spectrum of tetralogy of Fallot.

We present the case of a young male who was diagnosed at the age of 18 with DORV with subaortic VSD (Figure A), subpulmonary stenosis (Figure C) and mitral valve malformation (anterior leaflet cleft (Figure B) and both leaflets prolapse) with secondary moderate mitral regurgitation (MR), associated with major aortopulmonary collaterals, without pulmonary hypertension. There was no surgical correction at that moment. At the age of 26 he presented with recurrent hemoptysis and embolisation of bronchial arteries was performed. In the same year, a cerebral MRI showed signs of multiple infratentorial and supratentorial ischemic strokes. One year later, he was diagnosed with brain abscess and otomastoiditis that were surgically managed; multiple microorganisms were isolated from the two sources (*M. morganii*, *K. pneumoniae*, *P. aeruginosa*) and the patient received prolonged antibiotic therapy. Two months later he presented with clinical, biological and echocardiographic signs of infective endocarditis (IE) and blood cultures confirmed the diagnosis of *Candida albicans* associated mitral valve IE (Figure D, E, F, G). Antifungal therapy was administered for one month, with the complete disappearance of the vegetation, but worsening of the mitral regurgitation and moderate left ventricular systolic dysfunction. Thus, the patient had a clear indication for mitral valve replacement together with complete repair of the cardiac malformation that seemed still feasible as the pulmonary arteries were well developed, there was no pulmonary vascular disease and the systolic function of the RV was normal. Correction of CHD was performed in March 2019 consisting of repair of the DORV (Figure H), mitral valve replacement (33 mm bileaflet mechanical valve) (Figure I), tricuspid valve repair with RVOT remodelling. Postoperative, the patient clinical status improved significantly and transthoracic echocardiography revealed a mild residual subpulmonary stenosis (Figure J) and normally functioning mitral prosthesis.

There are frequent, various complications in the natural history of congenital heart diseases, especially unrepaired. This case illustrates how vicious the circle of complications can get in a case of unrepaired cyanotic cardiac malformation and the difficulty of breaking this circle. The correction of CHD is the best prevention method and should be performed whenever it is feasible, even at an adult age.

Abstract P719 Figure.

