Case report

Unusual complication of arterial switch operation: ventricular septal aneurysm treated with surgical ventricular restoration

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Abstract

Despite current low overall perioperative mortality, ischemic complications due to coronary artery translocation remain the most unwanted early complication of the arterial switch operation. We describe the case of a boy who underwent, at five days of age, one-stage arterial switch operation for transposition of the great arteries with repair of ventricular septal defect and coarctation of the aorta. Early postoperative course was complicated by severe ischemic left ventricle (LV) dysfunction and development of a ventricular septal aneurysm. At four years of age he underwent successful surgical ventricular restoration associated with implantation of a biventricular pacing, with complete resolution of heart failure symptoms.

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1. Case report

A five day old boy, with transposition of the great arteries, ventricular septal defect and aortic coarctation underwent one-stage correction by arterial switch operation and repair of the associated defects in another center. Early postoperative course was complicated by low output syndrome with echocardiographic evidence of severe ventricular dyskinesia with an early aneurysm formation on the left ventricle (LV) antero-septal wall. Furthermore, complete atrioventricular block required an epicardial pacemaker implantation on second postoperative day. The patient was discharged home in NYHA class II on full medical therapy for heart failure. Cardiological follow-up showed a progressive worsening of his clinical status. A myocardial radionuclide scan, performed two years later, showed no viability in the mid septal wall and a later cardiac catheterization confirmed the diagnosis of aneurysm of the ventricular septum, bulging into the right ventricle, with severe stenosis of the first septal branch.

At four years of age, this patient was admitted to our department for heart transplant evaluation, with rest dispnea and recent multiple episodes of congestive heart failure despite full medical therapy. A transthoracic echocardiogram showed an ejection fraction of 10% and a large ventricular septal aneurysm. The indexed left ventricular end diastolic volume (LVEDV/m²) and the left ventricular end systolic volume (LVESV/m²) were, respectively, 200 and 185 ml/m², with a more spherical LV shape and LV transverse diameter superior to the LV long axis diameter in the apical four chamber view, mild mitral regurgitation and moderate tricuspid regurgitation (Fig. 1).

Surgery was performed through a median resternotomy using cardiopulmonary bypass, with antegrade normothermic ematic cardiopegic arrest. A longitudinal incision was made through the fibrotic apex of the LV and the septal aneurysm was plicated by means of a 4/0 prolene continuous suture and then covered by a bovine pericardial patch sutured in place with a prolene 4/0 suture (Fig. 2). The ventriculotomy was then closed with a 4/0 prolene running suture. For biventricular pacing, permanent three screw-in type epicardial leads (4965-35 Medtronic, Minneapolis, MN) were placed on the LV, at the base of the left atrial appendage, on right venticle outflow tract and on the right atrium. The new pace maker (Guidant Contak TR, 1231) was placed in the abdominal pocket. Weaning from cardiopulmonary bypass was achieved with infusion of adrenaline and enoximone. The patient underwent an uneventful recovery and on postoperative day 7 a transthoracic
echocardiography showed an improved EF to 40% with ventricular resynchronization. The patient was discharged home two weeks postoperatively on medical therapy with ace-inhibitors, amiodarone and diuretics.

Six months postoperatively a transthoracic echocardiography showed significant improvement of LVEDV/m^2 157 ml/m^2 and of LVESV/m^2 78 ml/m^2 with an ejection fraction of 50%. The patient was asymptomatic in NYHA class I, on medical therapy with amiodarone and ace-inhibitor.

2. Discussion

Perioperative coronary complications after arterial switch operation usually result from a combination of anatomic and surgical factors. Stretching, kinking and compression of a coronary artery may not be infrequent following coronary translocation and myocardial ischemia with ECG modifications are quite common in the immediate postoperative period, ranging from 3.7 to 22% in different reports [1]. Despite this, almost 70% of patients generally fully recover, from myocardial ischemia with normal ECG and echocardiographic patterns, one month after operation. Different pathophysiology is present in late coronary abnormalities following arterial switch operation, generally between 1 month and 1 year of age. This can be divided in occlusion, major or minor stenosis and stretching of the coronary artery.

To the best of our knowledge this is the first case of transposition of the great arteries with type A coronary anatomy which developed a large septal aneurysm following anatomic correction. Reoperation which was required because of medically refractory congestive heart failure, was performed successfully by means of SVR and biventricular pacing.

The shortage of donors, especially of pediatric age, and the complications of heart transplantation as rejection, infection, graft vasculopathy and side effects of long lasting immunosuppressive therapy, forced us to revaluate a conventional surgical option, despite the LV indexed volumes, considering also that SVR and biventricular pacing have been proved by previous study, to be a safe and effective solution for treatment of the post-ischemic LV in adults with low ejection fraction and prolonged QRS duration [2–7].

Despite this, these associated techniques are still anecdotal and, to our knowledge, never experienced before in pediatric patient, since post-ischemic cardiomyopathy is rare in these patients.

In conclusion, combined SVR and biventricular pacing seems useful techniques even for pediatric patients with severe impaired post-ischemic LV dysfunction and LV asynchrony, allowing to delay heart transplantation and assuring a good quality of life at medium term.

Long-term follow-up and longer series is needed to verify the durability of this solution in young patients.

References


