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Modified Rastelli Procedure at Atresia of Trunk of Pulmonary Artery

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Abstract

18 – years-old patient with tetralogy of Fallot associated with atresia of pulmonary artery type II by Sommerville, and patent ductus arteriosus underwent modified Rastelli procedure with the vascular bifurcation prosthesis with formation of the trunk and branches of pulmonary artery. This method allowed us to achieve total anatomical surgical repair with good functional outcome. Postoperative course was favorable.

Keywords: Atresia of pulmonary artery, Modifications, Rastelli procedure, Tetralogy of Fallot

Introduction

Tetralogy of Fallot is one of common and complex congenital malformations of the cardiovascular system, one of the structural components of which is a wide spectrum of pulmonary artery anatomical variation. The prevalence of PA trunk atresia ranges from 1 to 3% [1,2]. Despite on the achievements in development of TOF surgical repair, atresia of PA still actively developing field of CHD surgery and remains relevant. At present day all existing methods of surgical repair of pulmonary trunk atresia includes creation continuity between RV and LA by conduit. At the same time, the experience of recent decades, represents that valve element inside the conduit necessary to avoid RV failure in the short and long postoperative terms. However primary surgical repair of PA atresia with extended hypoplasia of PA branches remains challenging case in surgical practice.

Our department has experience in performing Rastelli procedure and we would like to represent this brief message.

Case Report

An 18 – years-old patient tetralogy of Fallot associated with atresia of pulmonary artery. At admission patient had follows complaints: fatigue, weakness, decreased tolerance to physical exercise, severe cyanosis.

CHD had been diagnosed at birth. Epidemiologic, allergic and hereditary history is not burdened. Patient had visible cyanosis. Weight 47kg, Height-164cm. BSA - 1,46 m2. Symptom of chronic hypoxia moderately expressed. Chest auscultation: vesicular breathing on both sides, no wheezing. Auscultative tones are clear, rhythmic, systolic murmur in 2-4 intercostal space on left side from sternum. II tone of the pulmonary artery is not accentuated.

Clinical examination of blood revealed severe polycythemia Hb - 242 g/l, WBC - 7,2.; Chest X-ray: Lung fields clear, roots of lungs are sealed, extended. Heart enlarged to the left, apex of heart is raised. ECG: sinus bradycardia with HR-50 beats/min. Hypertrophy of the right ventricle. Echocardiography findings revealed Tetralogy of Fallot. Dextroposition of aorta. VSD -1, 7cm. Hypoplasia of the left ventricle and the trunk of PA. EDV LV - 81ml, EF - 63%. index of EDV LV -55,48ml/m2. Proceeding from data of ECHO patient underwent angiography for specify some components of CHD: Right ventriculography: trunk of PA is absent, RPA proximal part 5mm., LPA proximal part 4mm, distal parts of RPA 11 mm, LPA-18mm, descending Ao 18mm. Left ventriculography: ED LV ~ 141ml, contractility of all segments are good. Aortography: patent ductus arteriosus 3mm, MAPCAs doesn't revealed. Conclusion: Tetralogy of Fallot Pulmonary artery atresia type II according to Sommerville [3,4]. Patent ductus arteriosus Figure 1 A-B.

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17/12/2017 patient underwent surgery: Modified Rastelli operation (simultaneous replacement of trunk and branches of PA with the modeling of monocusp, patch closure of VSD, and suture closure of foramen ovale) under CPB (180min) and cardioplegia (142min), body temperature-29,0 C. Intraoperative revision showed: atresia of PA trunk, but proximal part of PA branches has severe hypoplasia about 0.3-0.4 cm, distal diameters RPA 2,0cm; LPA 2,2cm. Also, patent ductus arteriosus has been revealed, which was exposed and ligated. Intracardiac revision showed presence of juxtaortic VSD 2,0x2,0cm and infundibular obstruction of RVOT. Excision of fibrous obstruction had been performed Figure 2-A. Trunk and proximal parts of PA branches had been replaced by synthetic bifurcatinal conduits number 20, branch number 12 - by continuous suture prolen 4/0 Figure 2-B. Through an incision on RVOT had been performed additional resection of obstructive mass and further creation of proximal anastomosis using auto pericardial patch supplemented by monocusp Figure 2-C. VSD closure was performed with synthetic patch by running suture. Common view of completed reconstruction depicted below Figure 2-D.

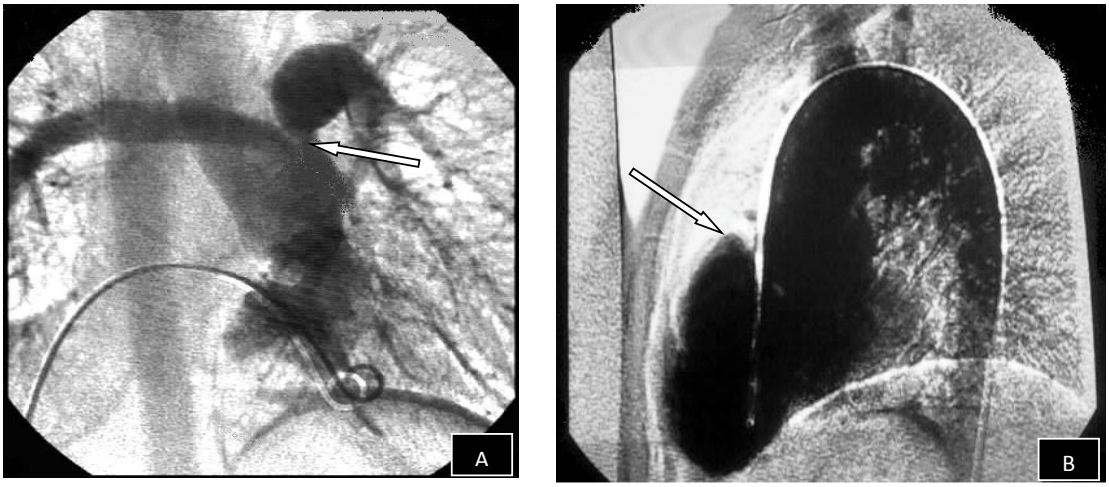

Figure 1A-B: (A) Angiography in A-P projection shows absence of pulmonary trunk and severe hypoplasia of proximal parts of branches of PA (arrow). **(B):** Angiography in sagital projection shows blind ending outflow tract of right ventricle (arrow).

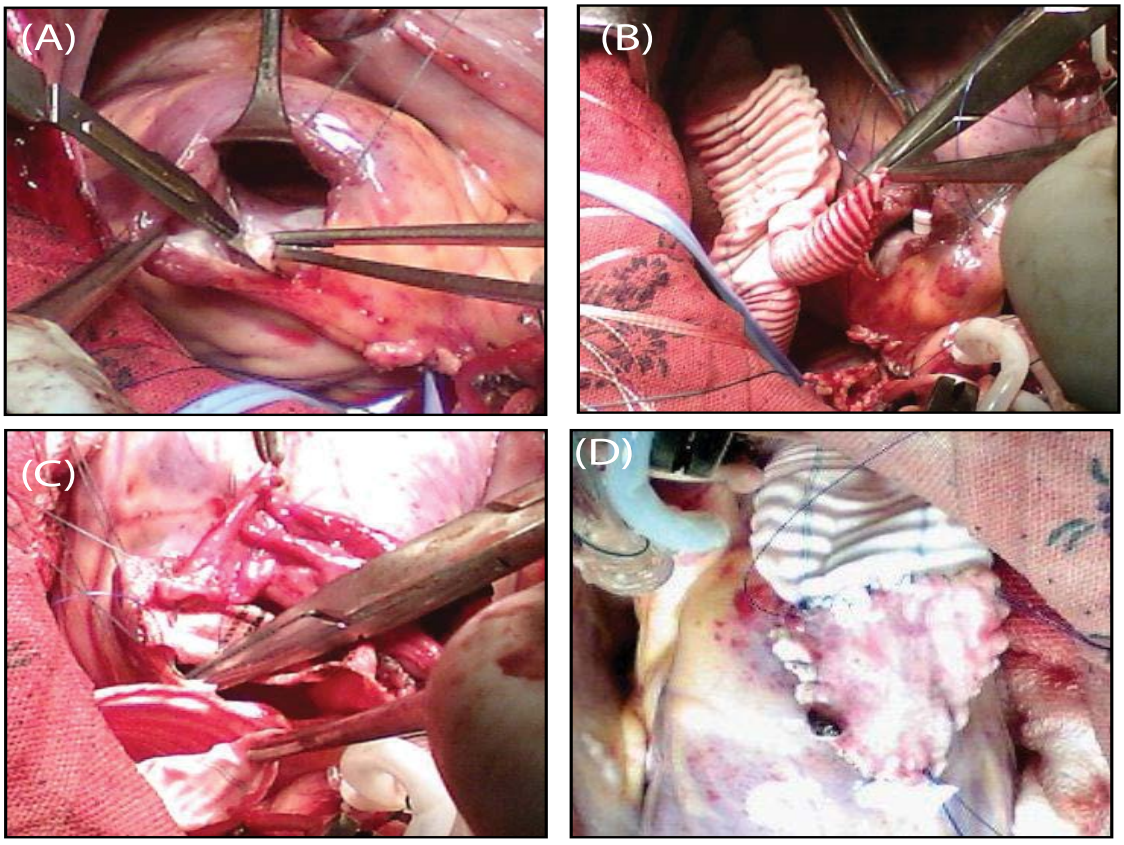

Figure :2 (A): Incision on right ventricle outflow tract **(B):** Distal anastomosis between branches of conduit and pulmonary artery **(C):** Proximal anastomosis between conduit and RVOT **(D):** Common view of performed reconstruction on RVOT

Discussion

In our clinical practice we are using couple of predictive indicators for the planning of reconstructive repair [5-7]: According to data of ECHO and angiography predictive indices was calculated based on distal pulmonary arterial tree. According to calculations we got the following values represented below Table 1.

Analysis of distal part of PA showed a satisfactory development pulmonary arterial tree and sufficient capacity for performing the total correction [8]. Take into consideration patient's age and type of atresia which characterized by the absence of the pulmonary artery trunk accompanying with hypoplasia of the proximal ostia of both pulmonary arteries, as well as absence of conditions for implementation classic Rastelli procedure - we modified classic surgical technique by replacement proximal parts of both pulmonary arteries with branches of bifurcational aortic synthetic prosthesis. Postoperative multidetector CT scans in 3D mode are shown below for complete imagination of reconstruction. Figure 3 A-B.

Postoperatively had been occurred transient right ventricular failure which was eliminated by temporary dobutamine drug therapy. At discharge, the general condition of the patient is satisfactory, FC I by NYHA without surgical wound complication. The control echocardiography showed: IVS without any residual shunt. Systolic peak pressure gradient on PA 5 mm.Hg. EDV LV-60ml, EF LV-57%. In long term follow up for 12 months patient without any cardiac incidents. Control echocardiography revealed: Systolic peak pressure gradient on PA 8 mm.Hg. EDV LV -80ml, EF LV-60%.

Conclusion

We suppose that represented modified method can be recommended for clinical use as alternative way for surgical repair in cases when performing of classic Rastelli procedure impossible for some of objective reasons. However, should be taken into consideration patient selection by age for avoidance of overgrow the prosthesis and increased complexity of surgery repair due to three anastomoses instead two at classic Rastelli procedure.

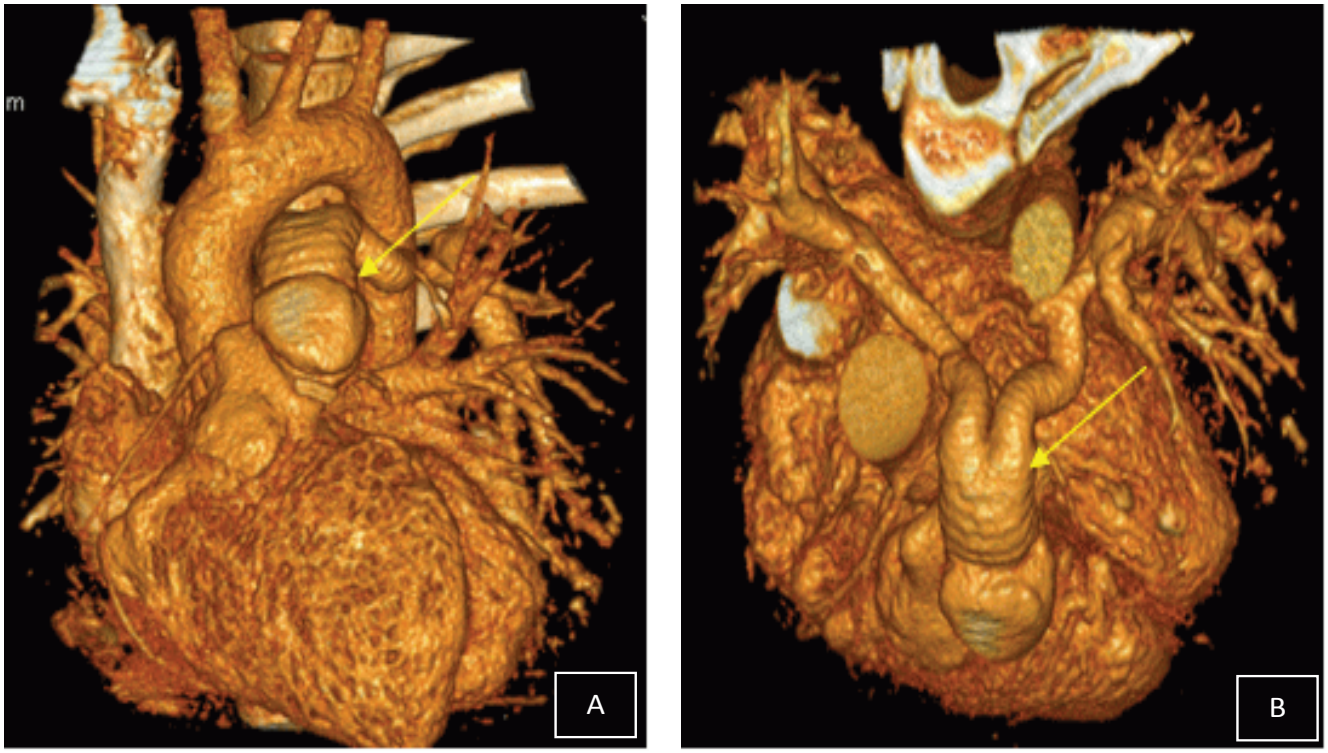

Figure 3-A: Clearly visible recreated trunk and left branch of the pulmonary artery by bifurcational aortic synthetic prosthesis from A-P position. **(B):** Clearly visible recreated trunk and branches bifurcation of the pulmonary artery by bifurcational aortic synthetic prosthesis from cranial position.

Table1: The calculated predictive indicators based on data of angiomorphometry.

		Nakata index mm2/m2	**238,7**
Ø FAPA (assumed)	17,2	McGoone index mm2/mm2	1,61
Z - factor of PA	-1,9	Z – factor of sum RPA&LPA	1451,7
EDV LV ml	141	EDV LV index ml/m2	96,4
Ø FAPA mm (actual)	15	Orifice area of FAPA mm2	176,6
Ø RPA mm (actual)	11	Orifice area of RPA mm2	95,0
Ø LPA mm (actual)	18	Orifice area of LPA mm2	254,3
Ø Ao (phrenicus)	18	Orifice area of Ao mm2	254,3

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