STAGED SURGICAL REPAIR OF PULMONARY ATRESIA WITH VENTRICAL SEPTAL DEFECT AND SEVERE PULMONARY ARBORIZATION ABNORMALITIES

Aleksandr Morozov[1], Ruben Movsesian[2], Alexey Shikhranov[2], Evgeny Grekhov[1], Aleksandr Latypov[1], Andrey Tzytko[2], Gennady Chizhikov[2], Nikolay Antsygin[2], Vadim Lubomudrov[3]

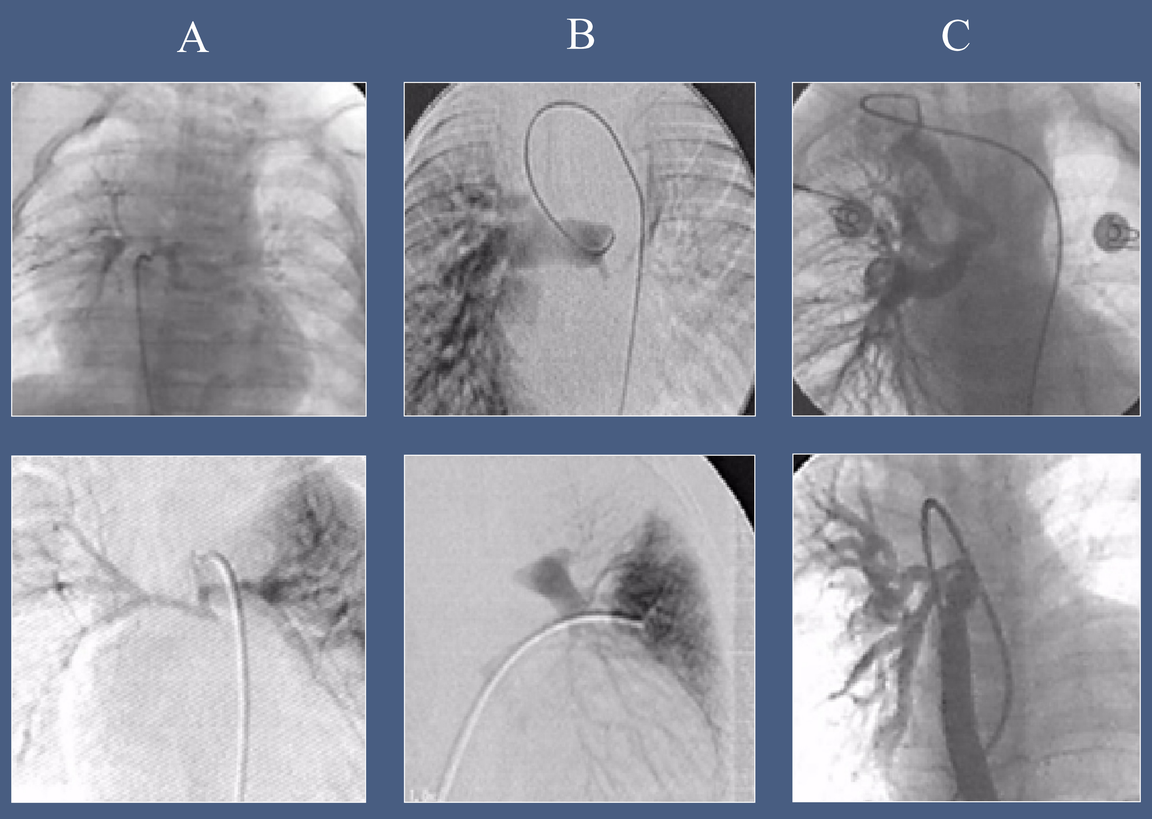
1- Almazov National Medical Research Centre, Department of Pediatric Cardiac Surgery, St.Petersburg-Russia;
2 - Children's City Hospital №1, Department of Pediatric Cardiac Surgery, St.Petersburg-Russia;
3 - Chest Disease Hospital, Department of Pediatric Cardiac Surgery, Al-Kuwait-Kuwait

Background

Surgical management of pulmonary atresia with ventrical septal defect and pulmonary arborization abnormalities is still a surgical challenge. The pulmonary arborization abnormalities in this complex cardiac malformation are characterized by hypoplastic or even absent native pulmonary arteries and multiple sources of pulmonary blood supply. We estimated our results of staged surgical treatment in such patients.

Methods

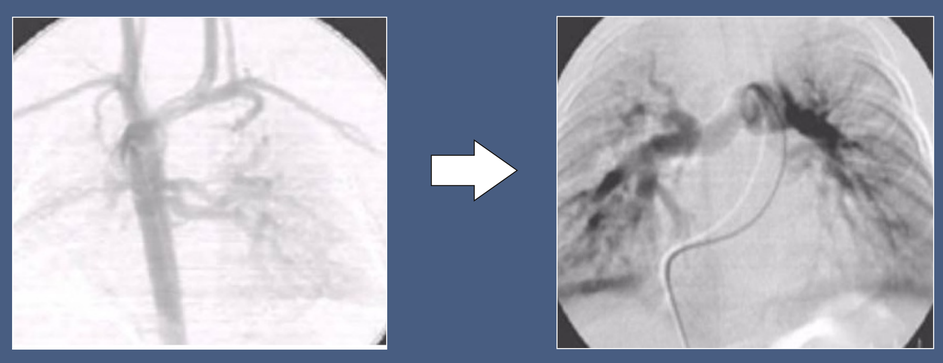
From 2000 to 2015, staged surgical repair was done in 28 patients with pulmonary atresia, ventrical septal defect and pulmonary arborization abnormalities. Severe hypoplastic pulmonary arteries were presented in 22 patients (median Nakata index 46,7 mm/m^2, range 13,1-135,4 mm/m^2), inconfluence or absence of native pulmonary artery was revealed in 6 patients. The pulmonary blood flow was provided by major aorto-pulmonary collateral artery (median 3, range 2-5). Initial surgical procedures were consisted of systemic-to-pulmonary shunt only, unifocalization and systemic-to-pulmonary shunt or palliative right ventricular outflow tract reconstruction, depending on the distribution of pulmonary blood flow.



Column A: diminutive native pulmonary artery; column B: inconfluence of right and left pulmonary arteries; column C: severe pulmonary arborization abnormalities

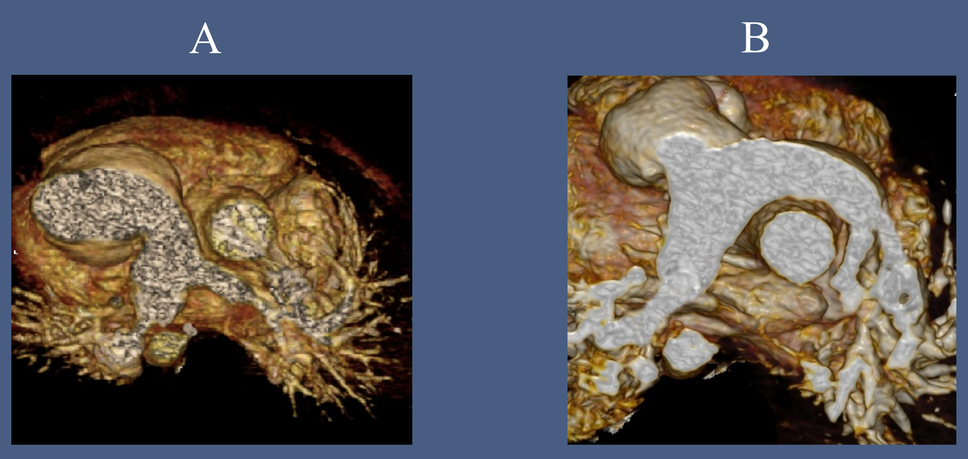
Results

Complete repair with ventricular septal defect closure was achieved in 10 (36%) patients (median 2,5 surgical procedures, range 2-4). Unifocalization of the pulmonary blood flow was finished in 4 patients before the final surgical stage.



Complete 1-stage unifocalization with RVOT reconstruction before VSD closure

Another 6 patients underwent complete repair and final unifocalization of the pulmonary blood flow at the same procedure. The pulmonary artery to the systemic blood pressure ratio after complete repair was 0.66 (range 0.44-1.00). Overall hospital mortality in staged group was 14,3% (3 patients died after staged procedures and 1 – after complete repair). Subsequently, dilation or/and stenting of pulmonary arteries was done in 3 patients, open redo surgery - in 1 patients.



Reconstruction of the central PA's by homograft and main PA by pulmonary valve xenograft (Contegra) using a "Lecompte maneuver" in a patient with bilateral pulmonary branch stenoses after complete repair. Pre-op (A) and post-op (B) CT-scan

Conclusion

Staged surgical approach can be applied in pulmonary atresia with ventricular septal defect and severe pulmonary arborization abnormalities as a way to reconstruction of pulmonary artery to prepare for complete correction with acceptable results.