STAGED REPAIR OF PULMONARY ATRESIA, VENTRICAL SEPTAL DEFECT AND MAJOR AORTO-PULMONARY COLLATERAL ARTERIES WITH SEVERE HYPOPLASIA OR ABSENCE OF PULMONARY ARTERIES

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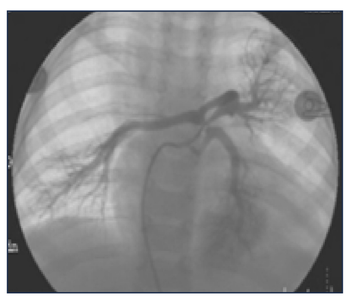
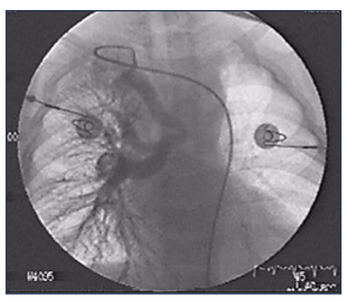
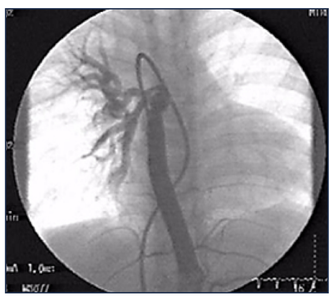
Background

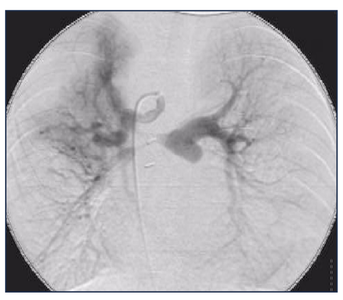
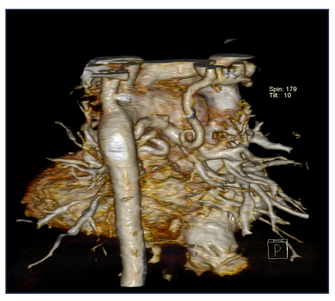
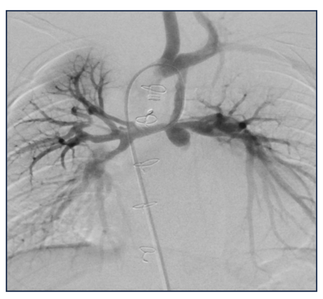
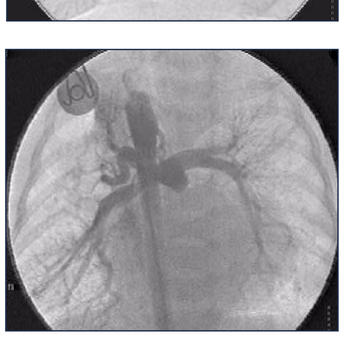
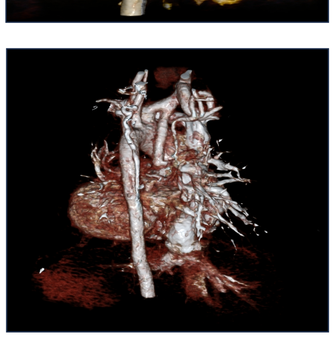
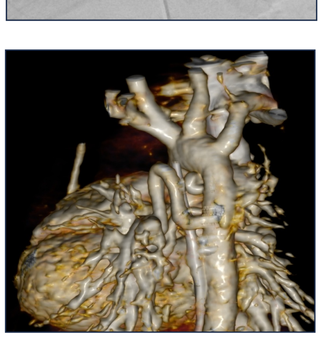
Surgical management of pulmonary atresia, ventrical septal defect and major aorto-pulmonary collateral arteries with severe hypoplastic or absent pulmonary arteries is still a challenge. We evaluated the results of staged surgical treatment in such challenging patients.

Methods

From 2000 to 2015, 32 consecutive patients with pulmonary atresia, ventrical septal defect and major aorto-pulmonary collateral arteries were surgically treated. Primary complete repair was done only in 4 patients, the other ones underwent staged procedures. Treatment of 28 staged patients began with 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. Among all staged patients, 22 had severe hypoplastic pulmonary arteries (median Nakata index 46,7 mm/m^{2,} range 13,1-135,4 mm/m^2), and 6 had inconfluence or absence of the pulmonary arteries.

Column A | Column B | Column C

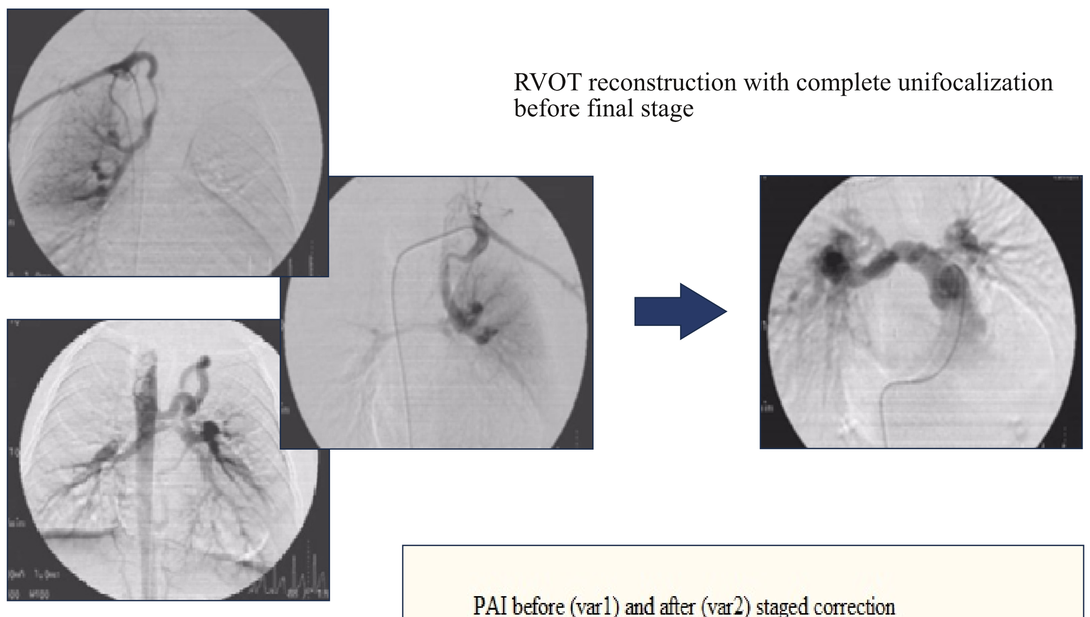







Column A: right side unifocalization; B: BT-shunt to diminutive PA; C: BT-shunt before MAPCA coil embolization

Results

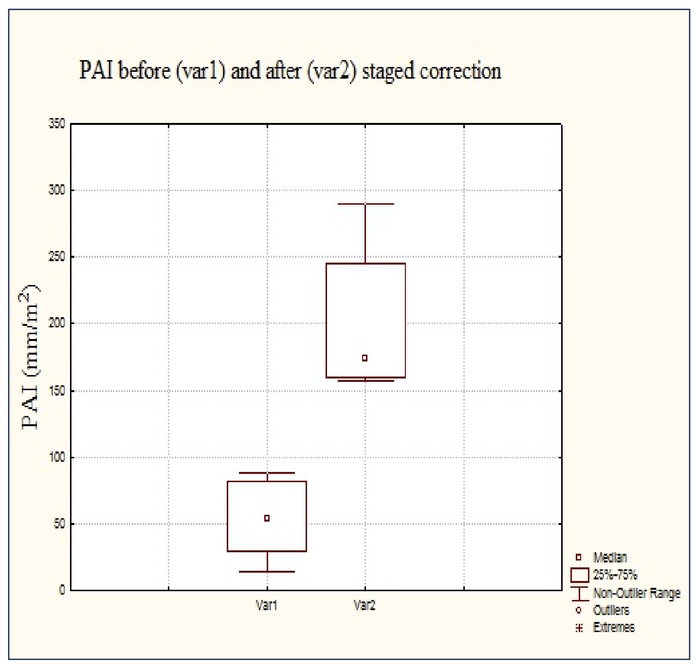
10 staged patients (36%) subsequently underwent complete repair with ventricular septal defect closure (median 2,5 surgical procedures, range 2-4). The median of the pulmonary artery pressure to the systemic blood pressure ratio at the final stage of repair was 0.66 (range 0.44-1.00). Duration of the full surgical course ranged from 6,6 to 69,2 months (median 15,7 months).

Unifocalization of the pulmonary blood flow was finished in 4 patients before the final surgical stage. Another 6 patients underwent complete repair and final unifocalization of the pulmonary blood flow at the same procedure.



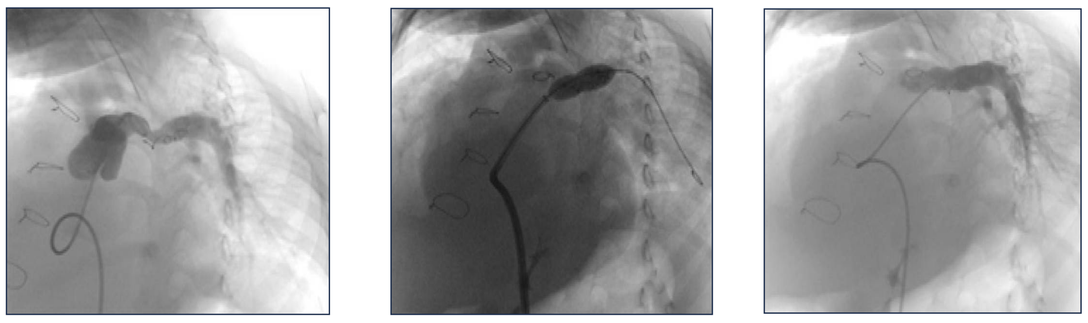
RVOT reconstruction with complete unifocalization before final stage

The mean Nakata index was increased from 53,3 mm/m^2 to 174 mm/m^2 by staged correction of the pulmonary hypoplasia and arborization abnormalities.



Overall hospital mortality in staged group was 14,3% (3 patients died after staged procedures and 1 – after complete repair). Subsequently, 3 patients underwent interventional catheterizations (dilation or/and stenting of pulmonary arteries), and right ventricle – pulmonary artery conduit was replaced in 1 case.

A | B | C



A: LPA stenosis; B: balloon dilation of LPA; C: control angiography

Conclusion

Staged surgical approach can be used in patients with pulmonary atresia, ventricular septal defect, severe hypoplastic or absent pulmonary artery and major aorto-pulmonary collateral arteries with acceptable results.