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## Unifocalization in patients with single and two-ventricle physiology

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**Aim:** This retrospective study reviews our results with unifocalization procedure of major aortopulmonary collateral arteries (MAPCAS) in patients with single- and two-ventricle physiology.

**Methods:** From August 2006 to September 2015, 15 patients with pulmonary atresia and MAPCAS have been operated at our institution. Median age was 13 months with interquartile range (IQR) 0.13-109 and median weight 7.8 Kg (IQR 3.2-24), respectively. In 11 patients, unifocalization was the first procedure, while in four patients this was done as a second procedure following modified BT shunt implantation to a pulmonary artery branch, in two patients (in one of them together with correction of a total anomalous pulmonary venous connection of supracardiac type) stent implantation and exploratory sternotomy was done. In three patients, the intracardiac anatomy was not suitable for a two-ventricle correction. In all patients, the unifocalization of the MAPCAS has been performed through a median sternotomy only. The unifocalization was performed concomitant with a modified BT shunt as a pulmonary blood flow source in three patients with a Glenn procedure in two patients and with an intracardiac repair (closure of the ventricular septal defect and right ventricle (RV) to pulmonary artery (PA) valved conduit) in 10 patients. A Glenn procedure and two intracardiac repairs with RV-PA conduit have been performed, thereafter in the three patients with primary unifocalization and shunt procedure. In three patients with unifocalization and intracardiac repair a fenestrated patch has been used for the ventricular septal defect closure.

**Results:** All patients survived the unifocalization procedure at a median follow-up of 57 months (IQR 12-121 months). Two patients with single-ventricle physiology already received the Fontan palliation with a non-fenestrated extracardiac conduit. Four patients with two-ventricle physiology needed catheter interventions for peripheral pulmonary artery stenosis and eventually stent implantation in three cases. In the group of the patients with two-ventricle physiology after unifocalization and intracardiac repair, two patients had an RV pressure estimate of one half systemic pressure, four patients had an RV pressure estimate of more than two thirds systemic pressure while six patients had an RV pressure estimate of less than one third systemic pressure. All three patients with single-ventricle physiology have good hemodynamics after unifocalization and bidirectional Glenn (one patient) as well as after unifocalization and total cavopulmonary connection (two patients).

**Conclusions:** In our experience, very good results can be obtained after unifocalization in patients with single- and two-ventricle physiology. There is a need for conduit replacement due to growth of the patient or conduit degeneration as well as for catheter-based interventions for peripheral pulmonary artery stenosis. Residual high pulmonary artery pressure and right ventricular dysfunction remain of concern for long-term survival in this very difficult group of patients.

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