bilateral superior vena caval patches at 9 months of age [1]. The Blalock-Taussig shunt was kept open for an additional flow. An angiogram at 20 months of age showed a sufficient development of the PA with a PAI of 181 and no peripheral pulmonary stenosis. The mean PA pressure measured by a catheter was 14 mmHg. Fontan operation with expanded polytetrafluoroethylene extracardiac conduit was completed at 22 months of age. An angiogram at 36 months of age showed excellent development of the PA with a PAI of 228 and smooth venous flow in the pulmonary trees (Fig. 1d). The central venous pressure measured by a catheter was 11 mmHg.

3. Discussion

Functional single ventricle and MAPCAs used to be considered contraindications for the Fontan operation, and surgical treatment is still challenging. For the patients with pulmonary atresia, ventricular septal defect and MAPCAs, however, some authors reported sufficient pulmonary arterial growth after one-stage unifocalization in early infancy [2]. Moreover, a successful case of Fontan operation for a patient with MAPCA has been reported recently [3]. We believe that one-stage unifocalization can also afford sufficient and balanced pulmonary arterial growth for Fontan candidates, even when lacking a central PA, and that it is important for Fontan candidates to solve morphological problems in the PA as earlier as possible without using prosthetic material [4]. In the present case, the PA continued developing with increasing PAI and stable PA pressure throughout the clinical course, even after the Fontan operation. We think that the one-stage unifocalization with autologous pericardium in early infancy and the repeated surgical intervention to the pulmonary artery without prosthetic material resulted in this excellent pulmonary arterial growth. The number of pulmonary segments which are required for the Fontan operation is not clear yet, however, unifocalization of more pulmonary segments without pulmonary hypertension may improve the patient’s prognosis. In the present case, the connections between MAPCAs and all pulmonary segments at the outset enabled the one-stage unifocalization of all pulmonary segments and contributed to the good result.

In conclusion, one-stage unifocalization with autologous material in early infancy, followed by staged Fontan operation, is a good surgical option for selected patients with functional single ventricle and MAPCAs.

References


ICVTS on-line discussion A

Title: Issues regarding unifocalization
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eComment: We read with great interest the article entitled: One-stage unifocalization followed by staged Fontan operation [1].

First described by Choussat et al., the “Ten Commandments” have become the basic criteria for patient selection undergoing the Fontan operation. Choussat listed ten criteria that should ideally be satisfied to minimize morbidity and mortality with the Fontan procedure. These 10 commandments have been adapted over the years and are summarized as: age above 4 years, normal ventricular function, adequate pulmonary artery size, no distortion of pulmonary arteries from prior shunt surgery, low pulmonary artery pressure (below 15 mmHg), low pulmonary vascular resistance, normal systemic venous drainage, no atriocentral valve leak, normal heart rhythm and no right atrial enlargement [2].

The current protocol in patients who fall into the high risk groups is firstly to review all the data in detail. Full consideration is given as to whether there are any surgically (or interventionally) correctable lesions such as AV valve regurgitation amenable to repair or isolated stenoses or hypoplasia within the central pulmonary arteries [3].

The guidelines for the surgical management of this anomaly include that all MAPCAs should be clearly delineated by preoperative aortogram at least up to the level of the diaphragm, so chances of missing would be less. In protected PAs/MAPCAs (proximal stenosis), complete repair or RV to PA conduit or central shunt should be done according to total size of PAs. In hypoplastic or absent PAs with unprotected MAPCAs (<1 year), or protected MAPCAs (proximal stenosis), complete repair/RV to PA conduit/central shunt should be done according to the size of the total pulmonary vasculature. Hypoplastic/absent PAs with unprotected MAPCAs (more than 1 year) are the subsets among these complex anomalies where we have yet to determine the surgical procedure to be performed. In single stage unifocalization, the number of operations, hospitalization and cost are reduced. These patients have early normalization of cardiovascular physiology with good future growth of unifocalized neo pulmonary arteries [4].
The MAPCAs are to be handled in several ways:

1. End-to-side or side-to-side anastomosis to the native pulmonary artery
2. Side-to-side anastomosis to other MAPCAs
3. Beveled end-to-side anastomosis to a polytetrafluoroethylene tube
4. Ligation of adequately communicating MAPCAs to the native pulmonary arteries
5. Ligation of MAPCAs to areas of the lung receiving significant dual blood supply
6. Ligation of small MAPCAs supplying a single bronchopulmonary segment if unifocalization of that segment was too difficult.

Exposure of all of the major aortopulmonary collaterals by opening the posterior pericardium without entering the pleura and completion of the right-sided unifocalization before commencing bypass. To reduce the potential neurologic complications, the right-sided unifocalization usually is accomplished before cardiopulmonary bypass at normothermia. The left-sided collaterals are localized and looped, ready to be snared as soon as bypass is instituted. Transection of the ascending aorta and retracting it to provide easy access to the MAPCAs and then doing the operation is very helpful. It may be easier and much more comfortable to do this kind of procedure with all the MAPCAs on both sides of the descending thoracic aorta. Do not hesitate to transect the ascending aorta if you have to access centrally hypoplastic, stenotic native pulmonary arteries to enlarge them and not to have to work behind the aorta if it is awkward. However, transecting the aorta to expose the MAPCAs implies that bypass is needed, and that increases the ischemia time [5].

References