Outcomes of the rehabilitative procedure for patients with pulmonary atresia, ventricular septal defect and hypoplastic pulmonary arteries beyond the infant period

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Abstract

OBJECTIVES: To determine the effect and safeness of the right ventricle to pulmonary artery connection with occlusion of major aortopulmonary collaterals and pulmonary artery angioplasty to rehabilitate the hypoplastic pulmonary arteries in patients with pulmonary atresia and ventricular septal defect beyond the infant period.

METHODS: From December 2009 to August 2012, 37 consecutive patients (mean age 1.9 ± 1.7 years, range 0.6–7.2 years) diagnosed with pulmonary atresia, ventricular septal defect and pulmonary artery hypoplasia (Nakata index 90.9 ± 42.6 mm²/m²; McGoon ratio 1.0 ± 0.2) were included in this retrospective study. All patients underwent the procedure of right ventricle to pulmonary artery connection, during which most of them received transcatheter occlusion of major aortopulmonary collaterals and/or pulmonary angioplasty. Mean follow-up was 1.6 ± 0.8 years (range 0.6–3.3 years). Continuous variables are expressed as means ± standard deviation.

RESULTS: There were no early deaths, but one patient died of myocarditis 1 year after the rehabilitation. Significant pulmonary artery growth was obtained (Nakata index 215.1 ± 95.1 mm²/m², P < 0.001; McGoon ratio 1.6 ± 0.5, P < 0.001) in all of the 37 patients, and among them, 17 patients (45.9%) whose pulmonary growth was considered adequate obtained a complete repair without perforation of the ventricular septal defect. The preoperative McGoon ratio might be a good predictor for adequate pulmonary growth. There was one early death after anatomical repair. At the last visit, all survivors who underwent anatomical repair were in New York Heart Association class I–II with satisfactory haemodynamics.

CONCLUSIONS: Connection of the right ventricle to the pulmonary artery is safe and effective to promote the growth of the native pulmonary arteries in patients with pulmonary atresia, ventricular septal defect and hypoplastic native pulmonary arteries. Ultimately, this strategy allows complete repair in the majority of patients beyond infancy.

Keywords: Pulmonary atresia • Ventricular septal defect • Right ventricle to pulmonary artery connection • Major aortopulmonary collaterals

INTRODUCTION

Pulmonary atresia with ventricular septal defect (VSD) is a rare and complex congenital heart disease with great morphological variability, such as hypoplastic and even absent native pulmonary arteries (PAs). The most important independent risk factor that influences the possibility for complete repair and the long-term survival rate is the poorly developed pulmonary vascular bed [1–4]. The surgical strategy is therefore focused on the rehabilitation of native hypoplastic PAs through an initial systemic to pulmonary shunt or right ventricle (RV)–PA connection [5, 6]. The latter procedure has more advantages than the former, i.e. uniform enlargement of all segments of the pulmonary artery, a lower incidence rate of obstruction or thrombosis and an easier approach to interventional PA dilatation that helps to prevent PA restenosis [1, 3, 6–10].

In previous reports, the majority of patients were infants (mostly younger than 3 months) and they received unifocalization of the major aortopulmonary collaterals (MAPCAs). Recently, studies have shown that MAPCAs, by their very nature, are bronchial arteries with a high incidence of thrombosis [1]. Furthermore, even when the MAPCAs remain patent, they do not exhibit any growth after unifocalization [1–4]. Nowadays, rehabilitation of the native PA without unifocalization is widely suggested. We used a new hybrid strategy, based on all the above considerations, which consisted of RV–PA connection without unifocalization and PA angioplasty, to promote rehabilitation of the intrinsic PAs. The aim of this study, therefore, was to evaluate the efficacy and safety of this strategy.

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PATIENTS AND METHODS

Study population

This is a retrospective single-centre study, for which the protocol was reviewed and approved by our institutional ethic committee. From December 2009 to August 2012, 37 consecutive patients beyond infancy (older than 6 months) with pulmonary atresia, VSD and hypoplastic PAs were included in our study. They all underwent the RV–PA connection as a palliative procedure to promote the development of the hypoplastic PAs. All of them had confluent PAs without malformation of coronary arteries, such as a RV-dependent coronary circulation. The medical records, angiographic studies, computed tomography (CT) images, echocardiogram records and clinical follow-up reports of all patients were reviewed. The details of the patients’ characteristics are shown in Table 1 and Figure 1.

Pulmonary artery rehabilitation strategy

Connection of the right ventricle to the pulmonary artery.

Patients were operated on via a median sternotomy and cardiopulmonary bypass on a beating heart, exactly as described by Metras et al. [5]. If the main PA existed, it was incised longitudinally, and a small oval autologous patch of pericardium prepared with glutaraldehyde was sutured to the edges of the arteriotomy as augmentation. The incision was then extended to the RV infundibulum if there was no major coronary artery crossing the infundibulum, and the pericardial patch was sewn to the edges of this incision, making sure the opening in the RV was sufficient by part resection of hypertrophied muscle. If no central PA existed or if it was discontinuous from the RV, the RV to PA connection was reconstructed using an autologous pericardial roll, Gore-tex conduit or bovine jugular vein, depending on the surgeon’s choice.

The diameter of the connection between the RV and the PA was 5–12 mm, which was determined by the patient’s weight and the oxygen saturation (SPO2) after the connection was made. For patients without MAPCAs, the diameter was controlled to allow the SPO2 to rise by approximately 10%. For patients whose SPO2 dropped sharply after the occlusion, the diameter was controlled to maintain the SPO2 just equal to its value before the palliative procedure.

The strategy for major aortopulmonary collaterals. Before the operation, a CT scan and/or angiography were used to analyse the features of the MAPCAs, i.e. their origin, size, stenosis and distribution. Additionally, all communications between collaterals and the true PA system needed to be identified clearly, in order to define lung segments that were supplied solely by the collaterals or perfused dually by both the collaterals and the true PAs simultaneously. If MAPCAs were not accompanied by significant stenosis or did not provide the sole supply to the lung, catheterization was performed to occlude the MAPCAs using coils before the connection procedure in our hybrid operation room. If occlusion of the MAPCAs would result in a precarious hypoxia, the MAPCAs would be occluded after the RV–PA connection or ligated during the procedure. If the MAPCAs provided isolated perfusion into a pulmonary segment or were accompanied by severe stenosis that would not induce pulmonary overperfusion after the RV–PA connection was made, they would be ignored and the occlusion would not be done.

Pulmonary artery angioplasty. If the origin of left and/or right PA stenosis existed, it would be surgically enlarged to meet the criteria of normal children of the same age, using the autologous pericardium during the procedure.

The follow-up strategy after the palliative procedure. After the RV–PA connection procedure, all patients were carefully followed up using echocardiography in our outpatient clinic, at 3

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Table 1: Demographic data and morphological features before RV–PA connection

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>n (%)</th>
</tr>
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<tbody>
<tr>
<td>Male/female</td>
<td>16/21</td>
</tr>
<tr>
<td>Weight</td>
<td>9.1 ± 2.5 (6.2–18.5) kg</td>
</tr>
<tr>
<td>Age</td>
<td>1.9 ± 1.7 (0.6–7.2) years</td>
</tr>
<tr>
<td>Without central pulmonary artery (n [%])</td>
<td>11 (29.7%)</td>
</tr>
<tr>
<td>Patients with MAPCAS (n [%])</td>
<td>24 (64.9%)</td>
</tr>
<tr>
<td>MAPCAS with stenosis (n [%])</td>
<td>34 (53.1%)</td>
</tr>
<tr>
<td>Number of MAPCAS</td>
<td>64</td>
</tr>
<tr>
<td>Number per patient</td>
<td>2.9 ± 1.3 (1–5)</td>
</tr>
<tr>
<td>Patent ductus arteriosus (n [%])</td>
<td>20 (54.1%)</td>
</tr>
<tr>
<td>Mean Nakata index*</td>
<td>90.9 ± 42.6 (25.9–164.7) mm²/m²</td>
</tr>
<tr>
<td>Mean McGoOn ratio*</td>
<td>1.0 ± 0.2 (0.6–1.5)</td>
</tr>
<tr>
<td>Mean O₂ saturation</td>
<td>76% ± 10%</td>
</tr>
<tr>
<td>Mean haemoglobin</td>
<td>171.6 ± 32.5 g/l</td>
</tr>
<tr>
<td>Left PA stenosis (n [%])</td>
<td>13 (35.1%)</td>
</tr>
<tr>
<td>Right PA stenosis (n [%])</td>
<td>1 (2.7%)</td>
</tr>
<tr>
<td>Both PA stenosis (n [%])</td>
<td>7 (18.9%)</td>
</tr>
</tbody>
</table>

*If stenosis existed, the PA was measured at the level of the hilus of the lung; we used this diameter to calculate the Nakata index and McGoOn ratio. Five patients with an approximately satisfactory PA development index but extremely diminutive PA on one side were also included in our procedure (shown in Fig. 1).

MAPCAS: major aortopulmonary collaterals; PA, pulmonary artery.
month intervals. When the PAs achieved satisfactory growth or the shunt and/or pulmonary branch showed obvious obstruction, a CT scan and/or angiography was performed. If there was an obstruction or stenosis that would not permit adequate blood flow to the distal PAs, balloon angioplasty or stent implantation was attempted. The only exception was residual proximal native PA stenosis when the patient’s Nakata index or McGoon ratio was satisfactory to allow complete repair, in which case it would be convenient and wise not to treat such stenosis until the final operation.

**Complete repair**

When the Nakata index and the McGoon ratio were considered adequate (usually, Nakata index >160 mm²/m²; McGoon ratio >1.6), a complete repair would be performed under cardiopulmonary bypass, with cardioplegic cardiac arrest. This consisted of a redo-sternotomy and complete repair via right ventricular outflow reconstruction and VSD closure. For the RV–PA continuity, a bovine jugular vein or a bovine jugular vein patch was used. The patch VSD closure using Dacron material was completed by a transatrial or transventricular approach. After the completion of cardiopulmonary bypass, pressures were recorded, and if the RV/left ventricle (LV) pressure ratio was less than one, the correction was considered satisfactory; otherwise, a fenestration was made in the VSD patch.

**Statistical analysis**

Continuous variables were expressed as the mean ± standard deviation (minimum, maximum). But the outcomes such as the duration of ventilatory support and the length of stay in the intensive care unit often showed skewed distributions and were summarized by the median (first quartile, third quartile). Categorical variables were compared using a χ² test or Fisher’s exact test, and continuous variables before and after the RV–PA connection procedure were compared using Student’s paired t-test. To identify the factors associated with complete repair, we used univariate analysis for the small sample size with an independent t-test for the age, Nakata index and McGoon ratio before the palliative procedure and a χ² test or Fisher’s exact test for the categorical variables, such as with or without MAPCAs, the type of RV–PA connection, etc. The cumulative rate of complete repair was estimated by Kaplan–Meier analysis. The statistical analysis was performed using SPSS version 17.0 for Windows (SPSS Inc., Chicago, IL, USA).

**RESULTS**

Thirty-seven patients were included in our study. The flow of patients and data is shown in Figure 2. All of the patients achieved significant PA growth. Among them, 17 patients achieved a complete repair, but one died. At the time of writing, three patients with satisfactory PA development were waiting for the final operation. One patient died of myocarditis 7 months after RV–PA connection when her PA was considered favourable for complete repair. Sixteen patients whose PA index had not yet met the criterion for complete repair were still in the follow-up process. When development of the PAs met the criterion, the complete repair would be done, and if PA stenosis occurred, which could inhibit the PA from developing, balloon angioplasty or stent implantation was attempted.

**The palliative procedure**

The materials used for RV–PA connection were an autologous pericardial patch (17 patients, 45.9%), an autologous pericardial roll (10 patients, 27%), a bovine jugular vein (four patients, 10.8%) or a Gore-tex conduit (six patients, 16.2%). Major aortopulmonary collaterals were occluded using the interventional procedure before the operation in 10 patients (a total of 22 MAPCAs) and after the operation in four patients (five MAPCAs). Two MAPCAs in two patients were ligated using surgical sutures during the operation. Patent ductus arteriosus was ligated during the operation in all cases except one, because the left PA was more diminutive than the right. For the same reason, in two patients’ right PAs were banded and a modified Blalock-Taussig shunt from the left subclavian artery to the left PA was done. In all cases, the stenosis of the PA was reconstructed during the palliation; left PA angioplasty was performed on 13 patients (35.1%), right PA angioplasty on one (2.7%) and bilateral angioplasty on seven (18.9%).

The mean diameter of the RV to PA shunt was 7.8 ± 1.6 (5–12) mm, and the mean SPO₂ was significantly improved to 87.5 ± 3.6 (82–96) % (P < 0.001) postoperatively. The shunts of two patients had to be banded to avoid a sharp increase in SPO₂. The shunt of another patient, who could not be weaned off mechanical ventilation because of pulmonary oedema, was banded by redo-sternotomy.

After construction of the connection, the median intensive care unit stay was 3 (2, 6) days and the median mechanical ventilation time was 25 (18, 71.5) h. There were no early deaths, but five patients suffered from pneumonia (13.5%) and two patients experienced capillary leak syndromes (5.4%). All these patients recovered before discharge.

**Follow-up after the rehabilitative procedure**

The mean duration of follow-up was 1.6 ± 0.8 (0.6–3.3) years. During the follow-up, no right or left ventricular dysfunction was noted, but one patient died of myocarditis 7 months after RV–PA connection when her PA was considered favourable for complete repair. Left PA stenosis was observed in seven patients and right PA stenosis was detected in five, but only three needed intervention by the PA balloon angioplasty and stent implantation (see Fig. 3), of whom one had right ventricular outflow tract stent implantation.

**Complete surgical repair**

After an average of 1.2 ± 0.8 years, all patients had achieved significant growth of the PAs (Nakata index 215.1 ± 95.1 mm²/m², P < 0.001; McGoon ratio 1.6 ± 0.5, P < 0.001) compared with the level before operation. Seventeen patients (45.9%) whose PAs were considered favourable achieved a complete repair, 1.2 ± 0.4 (0.7–2.0) years after rehabilitation. At the time of writing, another three patients who had obtained sufficient PA growth were waiting for the complete repair. If we were to include them, the rate of complete repair would be up to 54.1%.

The RV–PA connection was replaced with a bovine jugular vein or patch (diameter 14 ± 2 mm, 10–16 mm), and the VSD was closed with a Dacron patch. Additionally, 10 left PA and two bilateral PA angioplasties were done simultaneously.
At the end of the procedure, the RV/LV pressure ratio was $0.5 \pm 0.1$ (0.3–0.7). The median duration of ventilatory support and the median length of stay in the intensive care unit was 23 (18.8, 38) h and 2.5 (1.75, 4.5) days, respectively. One patient died of multiple organ failure, which was induced by pneumonia 2 weeks after the complete repair. One patient, who suffered from low cardiac output syndrome 1 day after the complete repair, was successfully weaned off extracorporeal membrane oxygenation 3 days later. Recovery of all of the other patients was uneventful during the postoperative period. We use the same standard for the complete repair, and we could not find any difference in the details of the clinical characteristics before the complete repair between these two patients and the others who had uneventful postoperative recoveries.

Some MAPCAs were left unoccluded during the palliation, but only four were detected by radiography and/or CT scan before the complete repair. These four MAPCAs, two of which were judged to be providing isolated supplies to the pulmonary segments, were occluded using coils before the complete operation in our hybrid operation room. Our study confirmed that stenosis existed in 53.1% of MAPCAs before the first operation, and most of them (88.2%, 30 of 34) could not be detected by angiography or CT scanning before the complete surgical repair.

During the follow-up period, no deaths occurred, and all survivors who underwent anatomical repair were in New York Heart Association class I–II with satisfactory haemodynamics.

**The factors associated with complete repair**

As shown in Table 2, there were no significant differences between the completed and uncompleted group, such as in the
Nakata index before the palliative procedure, the presence or absence of MAPCAs/main PA or the type of RV–PA shunt. The McGoon ratio might be a good predictor for complete repair.

DISCUSSION

A low PA bed resistance is an independent factor in complete repair and long-term survival for patients with pulmonary atresia–VSD, so the growth of hypoplastic PAs must be promoted [4, 6–9]. The RV–PA connection as a PA rehabilitative strategy was first described at the Children’s Hospital, Boston [10], where it was also confirmed to be effective among patients who suffered from pulmonary atresia–VSD with PA hypoplasia [3–5, 11–13]. Nevertheless, some surgeons argued that the RV–PA conduit as the source of pulmonary blood flow is contraindicated because the pulmonary flow and pressure are completely uncontrolled, which might induce pulmonary overperfusion and RV dysfunction [9]. To avoid such complications, most authors recommending this strategy have suggested that occlusion of the communicant collaterals should be carried out earlier [14]. We observed a very low incidence of such complications, which we attributed to our control of the diameter of the RV–PA connection according to the patient’s weight and the SPO2 and to the simultaneous occlusion of the MAPCAs, which might have other advantages. In particular, in instances of dual supply, we occluded the collateral artery, in the belief that its competitive flow may inhibit the growth and development of the associated intrapericardial pulmonary arterial tree [15]. Occlusion of the communicant collateral is important, not only to promote growth of the native PA and presumably angiogenesis [14], but also to reduce the occurrence of pulmonary overperfusion.

During establishment of the RV–PA connection, we carried out the PA angioplasty using the native tissue. In this way, we could not prevent stenosis from happening, but we achieved a lower reintervention rate than in other studies [14] and satisfactory PA growth after the rehabilitation, which was similar to the data of Liava’a et al. [16].

Previously, most surgeons suggested unifocalization as a method of obtaining a minimal pulmonary circulation pressure [8, 15, 17–19]. However, at present, more and more surgeons, including the Melbourne group who pioneered the concept, are abandoning unifocalization because the unifocalized MAPCAs show a high incidence of occlusion and no growth [2, 6–8]. Our study confirmed that stenosis existed in about half of the MAPCAs before the first operation, and most of them could not be detected by angiography or CT before the complete surgical repair. We agree with the opinion of d’Udekem et al. that unifocalization provides no long-term benefits [2].

The rate of complete repair was 45.9% (17 of 37 patients). If we were to include the three patients who were waiting for the final operation, this rate would be up to 54.1%. Using the Kaplan–Meier curve, this rate would go up to 70.2% about 2 years after the PA rehabilitation (see Fig. 4). This accumulated rate was similar to that of other reports, which was between 41 and 78%, but our study period was shorter than theirs (3 and 6–18 years.

### Table 2: Factors that influenced completion of the repair

<table>
<thead>
<tr>
<th>Factors</th>
<th>Complete repair (17 patients)</th>
<th>Uncompleted repair (20 patients)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>1.6 ± 1.0</td>
<td>2.2 ± 2.1</td>
<td>0.273</td>
</tr>
<tr>
<td>Nakata index (mm²/m²)</td>
<td>110.4 ± 40.4</td>
<td>79.6 ± 40.6</td>
<td>0.058</td>
</tr>
<tr>
<td>McGoon ratio</td>
<td>1.0 ± 0.1</td>
<td>0.9 ± 0.2</td>
<td>0.013*</td>
</tr>
<tr>
<td>With MAPCAs [n (%)]</td>
<td>11 (64.7%)</td>
<td>13 (65%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Without MAPCAs [n (%)]</td>
<td>6 (35.3%)</td>
<td>7 (35%)</td>
<td>1.000</td>
</tr>
<tr>
<td>With main PA [n (%)]</td>
<td>12 (70.6%)</td>
<td>14 (70%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Without main PA [n (%)]</td>
<td>5 (29.4%)</td>
<td>6 (30%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Type of RV–PA shunt</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>With valve [n (%)]</td>
<td>10 (58.8%)</td>
<td>11 (55%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Autologous pericardial patch [n (%)]</td>
<td>9 (52.9%)</td>
<td>8 (40%)</td>
<td>0.830</td>
</tr>
<tr>
<td>Bovine jugular vein [n (%)]</td>
<td>1 (5.9%)</td>
<td>3 (15%)</td>
<td>0.830</td>
</tr>
<tr>
<td>Without valve [n (%)]</td>
<td>7 (41.2%)</td>
<td>9 (45%)</td>
<td>1.000</td>
</tr>
<tr>
<td>Autologous pericardial roll [n (%)]</td>
<td>4 (23.5%)</td>
<td>6 (30%)</td>
<td>0.830</td>
</tr>
<tr>
<td>Gore-tex conduit [n (%)]</td>
<td>3 (17.6%)</td>
<td>3 (15%)</td>
<td>0.830</td>
</tr>
</tbody>
</table>

*Significant difference. Among five patients with unbalanced development of the PAs, three patients achieved a complete repair.

MAPCAS: major aortopulmonary collaterals; PA: pulmonary artery; RV: right ventricle.
respectively) [3, 8, 20]. Unfortunately, we must mention that the time from the palliation to the complete repair was always extended, for many reasons, in our country, such as the bed availability in our institution or the family’s financial situation.

In contrast to all prior reports, we did not find that age, presence of MAPCAs and the type of RV–PA shunt were factors influencing the complete repair [17–19, 21].

We think that the reason why age had no relationship to the complete repair is that our study excluded patients younger than 6 months, which might be a suitable period for the pulmonary artery development. Even though our results suggested that the older children had a chance of obtaining complete repair by our strategy, which was very important because it meant that the notable proportion of older patients referred to us would be able to obtain a complete repair, we also think that we should rehabilitate the hypoplastic pulmonary arteries as early as possible because the younger patients are most likely to obtain growth of the native pulmonary arteries, as mentioned by Duncan et al. [17].

Most of the MAPCAs of our patients were accompanied by significant stenosis or were occluded during the rehabilitation, which could not contribute to the development of intrinsic pulmonary vascular bed.

Given that the number of patients in different groups according to the type of the RV–PA shunt were very few, we could not determine whether the type of shunt would affect the complete repair, although previous studies have suggested that reconstruction of the RV outflow tract with expandable materials, such as pericardium or bovine jugular vein, would cause aneurysmal dilatation and consequently reduce the energy and weaken the development of the distal PA [19], and that a shunt without a valve might be better for PA promotion [18].

Although our data showed that the patients with a higher McGoon ratio before the palliative procedure might easily achieve a complete repair, we contend that the mean McGoon ratio between the two groups is similar. Therefore, we cannot simply conclude that the patients with the lowest McGoon ratio are not candidates, because the PAs of all patients without complete repair are still developing, and there are three patients with a satisfactory McGoon ratio waiting for operation who are not included in our complete group. Additionally, the children had experienced natural selection; the older children with very poor development in our complete group. Additionally, the children had experienced natural selection; the older children with very poor development of pulmonary arteries may have died had no chance to come to our hospital. Furthermore, other studies have come to the conclusion that patients with severely hypoplastic PAs can achieve a complete repair by staged operation.

In our study, the Nakata index and the McGoon ratio were higher than in other studies. The main reason for this is that if PA stenosis existed, instead of using the diameter before branching to calculate the Nakata index and the McGoon ratio, we used the diameter at the level of pulmonary hilus, which we thought might reflect the true native PA bed, as described by Batra et al. [22]. Moreover, included in our research there were also patients with one well-developed unilateral PA, while the contralateral PA was extremely diminutive.

Limitations in our study are inevitable. First of all, because genetic testing was not carried out routinely in our hospital, the factor of patients with 22q11.2 deletion, which is generally accepted as an independent factor [13, 21, 23, 24], could not be investigated in this study. Secondly, the study has the inherent limitation of being retrospective. Thirdly, the number of patients was too small for us to use multivariate analysis. Finally, as our observation period was only about 4 years, we could not confirm that the PA rehabilitation is contraindicated for patients with a lower McGoon ratio.

In conclusion, RV–PA connection is an effective and safe strategy to promote the PA growth for patients beyond infancy who have pulmonary atresia, VSD and hypoplastic PAs. Pulmonary artery angioplasty may substantially reduce the reintervention rate after the palliation procedure. Of course, we should maintain a certain level of SPO2, by controlling the diameter of the RV–PA shunt and simultaneously occluding the MAPCAs, to avoid pulmonary overperfusion. The McGoon ratio might be a good predictor for complete repair, but a patient with an unfavourably low ratio is not necessarily a poor candidate.

**Funding**

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**Conflict of interest:** none declared.

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APPENDIX. CONFERENCE DISCUSSION

Dr C. Brizard (Melbourne, Australia): You report a nice sized series of rehabilitation of the pulmonary arteries in patients with pulmonary atresia, VSD and MAPCAs, using a strategy not very different from ours in Melbourne. I have no doubt that with time you will achieve more than 80% of your patients in this series with complete septation and a favourable RV-to-LV pressure ratio.

I have three questions. Your series has relatively favourable confluent pulmonary arteries with a mean Nakata index of 91 mm²/m² and a McGoon ratio of 1. Moreover, 54% of your patients had a patent ductus arteriosus patent at the time of involvement. The first question: Do you think that some of your patients could have had a repair without staging, and of course without unifocalization?

Dr Li: In fact, for most of our patients the Nakata index was less than 90 mm²/m² and the McGoon ratio was less than 0.8. We believe that our patients were not suitable for one-stage repair. In our study, the main reason for the Nakata index and the McGoon ratio to be higher than the other studies is that the five patients who developed the unilateral PA and the other with the diminutive small PA were included. So to help these five patients to recover safely, we performed the hybrid procedure for the native pulmonary arteries.

Dr Brizard: Secondly, your series has no young infants and no very diminutive pulmonary arteries. Why did you choose that? Is this the result of a previous experience, or is it ultimately your goal?

Dr Li: I must say that in China most of the children come to us very late. The main purpose of this study is to evaluate if our strategy is satisfactory for the older children. There are patients with diminutive pulmonary arteries in our study whose Nakata index was 26 mm²/m². Another reason why the pulmonary arteries that developed in our patients look better than others was that the children are a product of natural selection. The older children with very poorly developed pulmonary arteries probably passed away and had no chance to come to our hospital. So, in fact, we did a modified BT shunt and RV–PA connection, and we have these two procedures for patients <6 months of age.

Dr Brizard: And the last question: We are very honoured that you quote the Melbourne approach as being partly your motivation to embark into the rehabilitation without unifocalization. You differ, however, in your approach in the way that you have chosen to embolize the collaterals at the time of the initial palliation, whereas we leave them to regress and to involute. Why did you choose differently?

Dr Li: Simultaneous occlusion of the MAPCAs is helpful for the postoperative course in reducing the harmful effects of overperfusion and right ventricle dysfunction. I noticed in your group, your study also suggests that occlusion of the collaterals should be done before surgery when the pulmonary flow has been increased.

Dr Brizard: For those who remain.

Dr Li: The simultaneous occlusion of the MAPCAs may have other advantages. I think personally that in surviving patients, non-occluded collateral arteries (i.e. competitive flow) may inhibit the growth and development of a natural pulmonary arterial tree. Occluded MAPCAs, I think, may not only still yield the angiogenesis and promote the native PA, but also reduce the magnitude of the pulmonary atresia.

Dr S. Daebritz (Dusseldorf, Germany): One short question. What is, in your opinion, the advantage of not anastomosing or unifocalizing in the first surgical procedure at least those MAPCAs which are big and stenosed; you could probably recruit some healthy, i.e. protected, pulmonary vascular bed without even necessitating cross-clamping or anything else to increase operative risk. Why didn’t you do that?

Dr Li: We don’t do that. I think the collateral is not close and the stenosis of the collateral maybe can occlude spontaneously.