Surgical management of pulmonary atresia with ventricular septal defect in late adolescence and adulthood

Emre Belli a,*, Loïc Macé b, Mohammed Ly a, Patrice Dervanian a, Emmanuelle Pineau a, Régine Roussin a, Emmanuel Lebret a, Alain Serraf a

a Marie Lannelongue Hospital, Paris-Sud University, Le Plessis-robinson, France
b University Hospital, Nancy, France

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Abstract

Objective: In presence of adequate pulmonary blood flow, patients presenting with unoperated or palliated pulmonary atresia with ventricular septal defect (PA/VSD) can reach adult age. However, they remain symptomatic with a limited life expectancy. Methods: Since 1993, 27 patients underwent surgery for unrepaired PA/VSD. Median age was 20 (range: 15—43) years. Nineteen patients had 33 previous palliative procedures while eight were unoperated survivors. Major aortopulmonary collateral artery (MAPCA) had been observed in all but 2 and were still patent in 23. All bronchopulmonary segments were connected to the native pulmonary arteries (NPA) in 4 (type A), to both NPA and MAPCA in 18 (type B) and only to MAPCA in 5 (type C). The biventricular repair was performed in 17 patients: 3 type A, 12 type B and 2 type C. Ten patients underwent palliative procedure: eight aortopulmonary shunt, with unifocalisation in two and one right ventricle to NPA restrictive conduit. Results: One (4%) hospital death occurred following the failure of a palliative procedure. No clinical improvement was observed in seven patients including one repaired and six palliated survivors. Two late cardiac death occurred 1 and 7 years after repair. At last visit, 15 of 16 repaired survivors were in NYHA class I or II. Only one patient awaits septation, while eight other with subsequent palliation were considered not repairable. Conclusion: The outcome was encouraging in patients who were eligible for completed biventricular repair. Although considered as unique alternative to cardiopulmonary transplantation, the justification for palliative surgery to improve pulmonary blood flow remains to be established.

Keywords: Heart defects; Congenital; Surgery

1. Introduction

The treatment of pulmonary atresia (PA) with ventricular septal defect (VSD) has evolved in time due to progress achieved in surgical techniques as well as investigation methods defining more precisely the pulmonary artery and major aorto-pulmonary collateral artery (MAPCA) anatomy [1]. Although several reports studied the surgical management in infancy, indication criteria for reparative surgery in unoperated or palliated adolescent and adult patients with adequate blood flow that permits survival remain to be established [2—6].

On the other hand, justification for 'definitive' palliation to increase pulmonary blood flow in symptomatic survivor obviously not suitable for septation constitutes an additional controversy.

In light of these issues, we reviewed the records of 27 patients who underwent procedure for PA and VSD.

2. Methods

Since 1993, 27 patients, 15 years old or more, were considered for surgery. Excluded from the study were patients with tetralogy of Fallot (TOF) with evidence of antegrade pulmonary blood flow and PA associated with other congenital malformations. Median age at surgery was 20 (range: 15—42) years. Preoperative median oxygen saturation was 80%. All were dyspneic on exertion with considerable fatigue with exercise. Nineteen patients underwent 33 previous palliative procedures including one failure of repair with reopening of the VSD, while eight were unoperated survivors. Table 1 summarises the previous palliative surgery performed.

MAPCAs had been observed in all but 2 and were still patent in 23. The study cohort was evaluated with regard of
pulmonary blood supply: classification proposed by Barbero-Marcial and Jatene [7] was employed. In four patients, almost all broncho-pulmonary segments were in connection with native pulmonary arteries (type A). In 18 patients, some pulmonary segments were exclusively supplied by non-communicating MAPCAs while others were in connection with native pulmonary arteries (type B). In the remaining (five patients), central pulmonary arteries were absent. Mild aortic valve insufficiency was often present [8]. One patient presented with associated moderate—severe aortic valve insufficiency. Twelve patients presented with moderate ventricular dysfunction.

Indications were based on cardiac catheterisation studies delineating pulmonary blood supply and haemodynamic data. Intracardiac morphology was determined in echocardiography. In our recent practice, multi-slice scanner investigation contributed to optimise the information on pulmonary artery and MAPCA anatomy.

Indication for biventricular repair was based on the size and accessibility of native pulmonary arteries and MAPCAs, on the blood pressure in the latter and, finally, on the quantity of salvageable pulmonary segments. MAPCA recruitment was considered according to anatomical position, their size (>3 mm) and absence of peripheral stenosis. Decisions were much more subjective and in a ‘case per case’ manner than that applied in paediatric population, the aim being to result in a significantly infra-systemic right ventricular pressure after septation. One patient (No. 14, Table 2) underwent emergency repair because of right ventricular outflow thrombosis (Fig. 1).

Patients were analysed in two groups: Group I included patients for whom the biventricular repair was performed (n = 17) and Group II (n = 10) included patients who underwent palliative procedure in order to increase pulmonary blood flow and for whom subsequent septation was often unforeseen case.

Table 1
Palliative procedures

<p>| | | |</p>
<table>
<thead>
<tr>
<th></th>
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<tr>
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<tr>
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<td></td>
</tr>
<tr>
<td>Central shunt</td>
<td>6</td>
<td></td>
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<tr>
<td>RV/PA conduit</td>
<td>3</td>
<td></td>
</tr>
<tr>
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<td>Waterstone shunt</td>
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<tr>
<td>MAPCA ligation</td>
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</tbody>
</table>

*All type of Blalock systemic-pulmonary shunt; RV, right ventricle; PA, pulmonary artery.

Fig. 1. Preoperative multi-slice tomography of patient No. 14, Table 2; arrow indicates thrombotic occlusion of right ventricular outflow.

Table 2
Group I: patients and results

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Type</th>
<th>Previous procedures</th>
<th>Previous unifocalisation</th>
<th>Nakata index</th>
<th>Associated procedures at repair</th>
<th>Postop. RV/LV</th>
<th>Residual MAPCA</th>
<th>Length of FU</th>
<th>NYHA</th>
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<tr>
<td>1</td>
<td>20</td>
<td>A</td>
<td>2</td>
<td>2</td>
<td>PAs reconstruction</td>
<td>50</td>
<td>0</td>
<td>49</td>
<td>NYHA I-II</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>27</td>
<td>A</td>
<td>2</td>
<td></td>
<td>LPA reconstruction, AVR</td>
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<td>44</td>
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<td>49</td>
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<td></td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>B</td>
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<td></td>
<td>MAPCA ligation</td>
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<td>0</td>
<td>95</td>
<td>NYHA I-II</td>
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<tr>
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<td>B</td>
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<td>1</td>
<td>Alive</td>
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<td></td>
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<tr>
<td>6</td>
<td>17</td>
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<td></td>
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<td>79</td>
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<td>16</td>
<td>B</td>
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<td>0</td>
<td>65</td>
<td>NYHA I-II</td>
<td></td>
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<td>18</td>
<td>B</td>
<td>4</td>
<td>146</td>
<td>Right and left unifocalisation</td>
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<td>0</td>
<td>48</td>
<td>NYHA I-II</td>
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<td>203</td>
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<td>B</td>
<td>2</td>
<td></td>
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<td>47</td>
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<tr>
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<td>2</td>
<td></td>
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<td>34</td>
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<td>43</td>
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<td>27</td>
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<td>29</td>
<td>B</td>
<td>0</td>
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<td>Right unifocalisation</td>
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<td>12</td>
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<td>20</td>
<td>B</td>
<td>3</td>
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<td>60</td>
<td>NYHA I-II</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>23</td>
<td>C</td>
<td>3</td>
<td>Left and right</td>
<td></td>
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<td>0</td>
<td>82</td>
<td>NYHA I-II</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>25</td>
<td>C</td>
<td>1</td>
<td>Left</td>
<td>212</td>
<td>Right unifocalisation</td>
<td>60</td>
<td>0</td>
<td>82</td>
<td>NYHA I-II</td>
</tr>
<tr>
<td></td>
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<td></td>
<td></td>
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</tbody>
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(*) Barbero-Marcial; (**) mm²/m²; (***) PA-pulmonary vein fistula; | and ||, died, see text; FU, follow-up.
from other centres. VSD was closed through right ventriculotomy. Interatrial septum was inspected and defects, if present, were closed. MAPCAs were managed in three ways: concomitant unifocalisation procedure was performed in five patients. PTFE conduits were more likely employed because of difficulties to perform direct connection of MAPCAs and PAs in large patients (Fig. 2). Two patients had associated MAPCA ligation while in six patients at least 1 MAPCA was left patent (Fig. 3). In two patients, percutaneous MAPCA embolisation was performed preoperatively. Six patients who did not have any concomitant unifocalisation procedure required PA branch reconstruction. In 13 patients, the reconstructed pulmonary arteries were connected to the heart by means of a valved conduit: 9 Hancock, 4 pulmonary homograft. In two patients, a bioprosthetic valve was inserted into the reconstructed right ventricle (RV) outflow. The remaining two patients had valveless right ventricle outflow patch reconstruction. Finally, one patient underwent concomitant aortic valve replacement and another patient underwent ascending aorta replacement.

Nine of 10 palliative procedures consisted in the creation of a systemic to pulmonary shunt. Table 3 summarises data concerning group II patients. All presented with MAPCAs. In five, unilateral pulmonary hypertension was documented and that was associated with non-confluent pulmonary arteries in three. Six patients had posterolateral thoracotomy approach and three sternotomy. In two patients, that was associated with unilateral unifocalisation procedure. One patient was re-palliated by the insertion of a restrictive (14 mm) RV to PA valved conduit.

Table 3

<table>
<thead>
<tr>
<th>No.</th>
<th>Age (year)</th>
<th>Barbero type*</th>
<th>Previous procedures (n)</th>
<th>Preop. O₂%</th>
<th>Surgical technique</th>
<th>CPB</th>
<th>Postop. O₂%</th>
<th>Last O₂%</th>
<th>Length of FU (months)</th>
<th>Comments</th>
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<tbody>
<tr>
<td>1</td>
<td>38</td>
<td>A</td>
<td>1</td>
<td>70</td>
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<td>80</td>
<td>78</td>
<td>93</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>29</td>
<td>A</td>
<td>2</td>
<td>75</td>
<td>Left BTS 6 mm</td>
<td>0</td>
<td>78</td>
<td>78</td>
<td>66</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>20</td>
<td>B</td>
<td>2</td>
<td>65</td>
<td>Stent removal/ RV-PA valved conduit 14 mm</td>
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<td>88</td>
<td>85</td>
<td>15</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>15</td>
<td>B</td>
<td>0</td>
<td>75</td>
<td>Central shunt 8 mm</td>
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<td>80</td>
<td>80</td>
<td>59</td>
<td>Awaits septation</td>
</tr>
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<td>5</td>
<td>17</td>
<td>B</td>
<td>1</td>
<td>75</td>
<td>Left BTS 8 mm</td>
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<td>75</td>
<td>4</td>
<td>Died postoperatively</td>
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<td>6</td>
<td>20</td>
<td>B</td>
<td>0</td>
<td>70</td>
<td>Right unifocalisation 8 mm</td>
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<td>70</td>
<td>33</td>
<td></td>
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<tr>
<td>7</td>
<td>17</td>
<td>B</td>
<td>1</td>
<td>75</td>
<td>Left unifocalisation 8 mm</td>
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<td>80</td>
<td>78</td>
<td>9</td>
<td>Unifocalisation thrombosed postoperatively</td>
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<td>8</td>
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<td>B</td>
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<td>80</td>
<td>Central shunt 8 mm</td>
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<td>80</td>
<td>74</td>
<td>120</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>18</td>
<td>C</td>
<td>0</td>
<td>68</td>
<td>Right BTS 6 mm</td>
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<td>78</td>
<td>85</td>
<td>91</td>
<td>MAPCA stenting performed</td>
</tr>
<tr>
<td>10</td>
<td>19</td>
<td>C</td>
<td>0</td>
<td>75</td>
<td>Left double BTS 4 mm</td>
<td>0</td>
<td>80</td>
<td>78</td>
<td>6</td>
<td></td>
</tr>
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</table>

(*) See text; † , died CPB, cardiopulmonary bypass; BTS, modified Blalock Taussig Shunt; MAPCA, major aorto-pulmonary collateral arteries, FU, follow-up.
3. Results

3.1. Group I (Table 2)

No hospital mortality occurred following biventricular repair. Mean RV/aorta pressure ratio at end of procedure was 0.5 ± 0.2. One patient required VSD patch fenestration because of supra-systemic RV pressure. One patient developed reversible neurological stroke and another patient had phrenic nerve paralysis.

Two patients required reoperation. One early reoperation (seventh postoperative day) was destined to eliminate surgically created pulmonary arterio-venous fistula (No. 13, Table 2). The second reoperation was performed 3 months after repair for residual VSD closure. Subsequent percutaneous residual MAPCA embolisation was necessary in two patients. The patient who underwent reoperation for residual VSD closure required pulmonary bifurcation stenosis release by percutaneous angioplasty and stent implantation 6 years after repair, and suddenly died 1 year later (No. 3, Table 2). In one patient, cardiac catheterisation confirmed the partial occlusion of unifocalised MAPCA without significant increase in RV pressure. Finally, one patient (No. 17, Table 2) died 1 year after repair because of endocarditis and multiple organ failure. One overseas patient was lost at follow-up. At last visit, 14 of the 15 survivors were in NYHA class I—II without specific medical treatment. Control echocardiography confirmed the stability of initial functional result associated with minimal increase of right ventricular outflow gradient. One patient developed pulmonary vascular disease obviously because of a residual patent MAPCA and requires medical treatment.

3.2. Group II (Table 3)

One patient died postoperatively because of hypoxemia due to unilateral pulmonary reperfusion oedema. Significant improvement of arterial O2 saturation was observed only in three patients: one of them awaits septation while the other remains unsuitable for repair. All palliated patients remain symptomatic with various degree of exercise intolerance. Fig. 4 is the postoperative multi-slice CT scan of the patient who underwent palliation with two left-sided 4 mm modified BT shunt on the lobar arteries.

4. Discussion

A small percentage of patients presenting with PA/VSD can survive adulthood without subsequent repair [2, 9]. In this series, the oldest patient was 43 years old with a central shunt performed at 26 years of age while the oldest ‘natural’ survivor was 29 years old. All group I patients were deemed ‘unrepairable’ in the past: progress achieved in surgical concepts and techniques permitted to reconsider them for repair and the latter to ameliorate their functional capacity as well as life expectancy, although left and right ventricular end-diastolic pressures were often elevated when measured.

Indication criteria for biventricular repair included usual parameters: ventricular function, size and blood pressure of available PAs as well as MAPCAs considerable for recruitment, and, in some cases, number of pulmonary segment involved as well as Nakata index (one of seven surgeons who performed procedures dealt with Nakata index and segment number) (Table 2) [10]. Evaluation of indication criteria was difficult because of the extreme variability of those parameters and the limited number of patients. The principal difference with the paediatric patient population was to deal with the patients’ definitive pulmonary vascular morphology. Acceptable (>8 mm) distal PA branch size was often associated with low (<50%) postoperative right ventricular pressure (Fig. 5 a and b). The reduced number of pulmonary segment involved and the high preoperative PA pressure appeared to be unfavourable. Although repair was planned with the aim to reach significantly infra-systemic right ventricular pressure, in patients with higher right ventricular pressure after septation, postoperative disappearance of cyanosis was associated with improvement in functional capacity.

MAPCA management in this particular group of patients varies from the paediatric patients. In larger patients, extensive MAPCA recruitment is technically more challenging. On the other hand, natural evolution of MAPCA resulted in the development of stenosis or vascular disease of corresponding pulmonary segment. Therefore, only previously unifocalised MAPCAs and MAPCAs presenting with adequate size and pressure level were considered for recruitment. Decision between staged or one stage repair was also a ‘patient specific’ criterion, one stage approach being privileged when complete MAPCA recruitment from midline sternotomy approach appeared feasible. Preoperative percutaneous MAPCA occlusion permitted more optimal operative conditions in two cases. At repair, residual smaller MAPCAs were ignored or ligated (particularly when communicating) according to patients’ specific morphology. Also, postoperative percutaneous residual MAPCA occlusion was considered when poorly tolerated.
Group II patients were considered for surgery because of increasing cyanosis but were not suitable for repair: the aim of surgical procedure was to improve pulmonary perfusion, thus, \( O_2 \) saturation and functional capacity and, in 3/10 cases, to prepare for subsequent repair. At present, only one of them awaits septation. The two others, although with increasing \( O_2 \) saturation, still present restrictive pulmonary vascular morphology. The results of "definitive" palliative procedures were concerning: one patient died postoperatively and six other did not present significant improvement. Septation appeared to be feasible.

In conclusion, PA with VSD can be repaired in properly selected young adult with excellent results. Improvement in clinical status was observed even in patients with restrictive pulmonary arteries resulting in high right ventricular pressure. Patients unsuitable for subsequent repair who were considered for "definitive" palliation did not improve their functional capacity.

References


Appendix A. Conference discussion

Dr Y. d’Udekem (Melbourne, Australia): While reviewing our experience with the multistage approach in Melbourne, we also reviewed our experience with these patients coming late for repair. We’ve looked at the one older than 5 years. And I have to disagree slightly on one point. The impression I get from this excellent presentation is that you should aim for anatomical repair, which effectively gives excellent results. Our finding was that whatever you do to the patients, the outcome is actually much better for natural survivors of the first few years of life than for all-comers with pulmonary atresia, VSD, MAPCs, starting as newborns. So they do have better survival than the younger ones when they’re natural survivors. And actually palliation in patients who are looking extremely desperate is also giving excellent results. We had almost no mortality in any of these patients. And the ones who were palliated can remain well on the long term for as long as 15–20 years with very minimal symptoms. And pulmonary arteries, even diminutive pulmonary arteries, still grow when you shunt them when they’re 14, 15 years of age.

Dr G. Luciani (Verona, Italy): Obviously we are dealing with a spectrum of lesions. My question is how many of these patients represent selected individuals? That is, how many did you discard from surgical option and how many did you transplant, heart-lung transplantation, during the same period?
Dr Belli: In a practical aspect, even if they are in the transplant waiting list, patients with pulmonary atresia, MAPCA and severe terminal stage are never transplanted in our department.

Dr Luciani: So to how many of these do you deny surgery, at all?

Dr Belli: Deny surgery? First of all, all those patients were denied for surgery in the past. Thus, the patients that we refused for surgery are the patients who were obviously not eligible for biventricular repair. But I'm not able to give an exact number of cases in which we didn't perform palliation or repair, because they are often referred from referral centers to us for surgery, and it seems very difficult to give a true number of those patients which are under follow-up without operation or with previous palliation.

Dr J. Vazquez-Jimenez (Aachen, Germany): In our experience, older patients who came to you for operation have very periphery pulmonary stenosis. Have you seen that?

Dr Belli: Of course. For that reason the indication criteria is very difficult to be classified in a guideline. However, overall, even in very limited pulmonary artery size and pulmonary territory, a biventricular repair can be performed. We often had the good surprise to find significantly infra-systemic right ventricle pressure and the interventional catheterization before and after surgery was very helpful in those with surgically inaccessible branch stenosis.