Pulmonary circulation after biventricular repair in patients with major systemic-to-pulmonary collateral arteries

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Abstract

Objective: To determine factors affecting postoperative pulmonary circulation in patients with major systemic-to-pulmonary collateral arteries. Methods: A total of 48 patients underwent biventricular repair subsequent to unifocalization at ages in the range 1–34 years. The preparative procedures consisted of ligation of the collateral arteries in 6, plasty to the pulmonary arteries using no artificial materials in 12 and extensive reconstruction using heterologous pericardial tubes in 30. The number of the pulmonary vascular segments unifocalized was 9–18 (16 ± 3). The amount of flow draining via residual minute systemic-to-pulmonary collaterals measured at the time of repair was 4–58% (24 ± 16%) of the total perfusion by the cardiopulmonary bypass machine. Results: This value was 40 ± 16% in 5 patients dying in the short term after repair. The number of segments was nine or ten after unifocalization in 2 of these. Another 4 patients died in the longer term, 3 of these with CATCH 22 syndrome dying because of pulmonary hypertension. Postoperative catheterization demonstrated mean pulmonary arterial pressures in the range 8–40 (21 ± 9) mmHg and pulmonary resistance in the range 1.7–10 (5.0 ± 2.1) units/m². Pulmonary resistance was correlated statistically to age at repair (r = 0.77), the number of pulmonary vascular segments (r = −0.41) and to percent collateral flow (r = 0.48). The use of a heterologous pericardial tube for unifocalization was also related probably to higher pulmonary resistance. Conclusion: It is essential to accomplish effective unifocalizations followed by earlier definitive repair so as to establish better pulmonary circulation. © 1997 Elsevier Science B.V.

Keywords: Major aortopulmonary collateral arteries (MAPCA); Pulmonary circulation; Pulmonary arteries; Tetralogy of Fallot with pulmonary atresia

1. Introduction

Remarkable strides in cardiac surgery have recently been made in repairing congenital cardiac malformations with severely abnormal pulmonary arterial trees [5–7,11,12,18–20]. An initial matter of surgical concern was how to successful achieve biventricular repair [10,16]. Since surgical results and unifocalization of the blood supply to the lung as well as biventricular repair in patients with major aortopulmonary collateral arteries (MAPCAs) or major systemic-to-pulmonary collateral arteries [1], have obviously improved particularly in the short term [11,20], the next step the surgeon should take is to establish better results in the longer term. From this viewpoint, an efficient pulmonary circulation will play a crucial part in better postoperative outcomes and more preferable functional status. Impaired pulmonary circulation associated with abnormal arterization of the pulmonary arterial tree [3] should be minimized if any alternative surgical strategy will allow it. In the present study, a series of patients with MAPCAs were retrospectively investigated to determine factors affecting postoperative pulmonary circulation in this particular setting.

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Table 1
Previous or concomitant unifocalizations in terms of operative procedures

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Right-sided</th>
<th>Left-sided</th>
<th>Bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ligation of MAPCAs</td>
<td>3</td>
<td>3</td>
<td>—</td>
</tr>
<tr>
<td>Intrapulmonary PA plasty</td>
<td>1</td>
<td>1</td>
<td>10</td>
</tr>
<tr>
<td>PA reconstruction using a pericardial roll tube</td>
<td>2</td>
<td>—</td>
<td>28</td>
</tr>
</tbody>
</table>

MAPCAs, major aortopulmonary collateral arteries; PA, pulmonary arteries.

2. Materials and methods

Between December 1985 and July 1996, 76 patients with MAPCAs had been referred to us for surgical interventions. Of these, 4 had ventricular morphology unsuitable for biventricular repair. Although these patients underwent surgical unifocalization of pulmonary blood supply, no patients have been submitted to the Fontan type procedure because of a high pulmonary resistance > 4 units/m². Of the 72 patients where biventricular repair was considered feasible, to date, 46 patients underwent biventricular repair subsequent to unifocalization, the average interval between unifocalization and biventricular repair being 8.3 ± 6.5 months (mean ± S.D.) and another 2 patients underwent one stage repair. An initial palliative procedure, such as aortopulmonary anastomosis [19] or palliative reconstruction of the right ventricular outflow tract, was intrapericardially needed in 7 patients for severely hypoplastic pulmonary arteries (PA) to grow sufficiently towards surgical unifocalization. Of these, 5 have successfully undergone subsequent unifocalizations via bilateral thoracotomies as well as biventricular repair via median sternotomy.

All the 48 patients undergoing biventricular repair were clinically reviewed. Seventeen patients possessed the so-called CATCH 22 syndrome. In 18 patients, the Nakata’s index was > 120. In 6 of these, some pulmonary vascular segments were dually supplied via the MAPCAs as well as via the native intrapericardial PA and ligation of MAPCAs was employed. In the other 12 patients, plasty to the intrapulmonary PA was carried out by directly connecting the autologous tissues. In almost all these patients, a systemic-to-pulmonary shunt was concomitantly placed. Of the remaining 30 patients, the intrapericardial PA was hypoplastic with the Nakata’s index [9] < 120 in 12, was vestigial with a diameter of < 2 mm in 10 and no intrapericardial PA could be detected in 8. Extensive reconstruction of the pulmonary arterial tree was needed using heterologous pericardial roll tubes [5,20] in these patients (Table 1).

The age at biventricular repair ranged from 1 to 34 years old, with a mean of 7.6 ± 7.7 years old (Fig. 1). Although our preference has been for definitive repair at a relatively younger age, several patients were referred to us at their adolescent or adult stages. This explains why patients at various ages were operated. The number of MAPCAs was initially from one to 6 (3.4 ± 1.2) per patient. Angiography subsequent to unifocalization and prior to biventricular repair demonstrated that the number of the pulmonary vascular segments preserved was between 9 and 18 (16 ± 3) (Fig. 1).

At the time of definitive repair on cardiopulmonary bypass, the amount of venous drainage to the left atrium per min was measured during aortic cross-clamping. Then, the value was divided by the total amount of aortic flow per min perfused by the machine for cardiopulmonary bypass, calculated as percent cardiac return. This proportional value would represent the amount of flow draining via residual minor systemic-to-pulmonary collateral vessels including the bronchial arteries [4,8]. The cardiac return was from 4 to 58%, a mean being 24 ± 16%. There was a correlation seen between percent cardiac return and age at repair (Fig. 2). The procedures employed for unifocalizations were not influential on percent cardiac return. A multiple linear regression demonstrated that age at...
repair and percent cardiac return were correlated factors and that CATCH 22 syndrome, the number of pulmonary vascular segments unified by the preparative procedures and the method for unifocalizations, proved to be independent factors.

3. Results

Five patients could not survive the repair because of either oesophageal or airway bleeding in 4 and intractable thrombosis within the pulmonary arteries in 1. These were related probably to abundant minor aortopulmonary collaterals around the oesophagus and the bronchial tree and to high pulmonary resistance, respectively. The follow-up term after definitive biventricular repair in operative survivors ranged from 12 to 122 months with a mean of 54 ± 30 months. Of the 43 operative survivors, 4 patients died in the longer term; 3, 5, 32 and 36 months after repair, respectively. The cause of death was pulmonary haemorrhage in the patient dying 3 months after repair and high pulmonary resistance as well as pulmonary hypertension in the other 3.

Of 5 early deaths, older age was seen in 2, excessive cardiac return in 2 and an inadequate number of segments in 2 (Fig. 3). Of 4 late deaths, 1 patient dying of pulmonary haemorrhage underwent repair at the age of 9 years with 13 pulmonary vascular segments. In the other 3 having CATCH 22, high pulmonary resistance had progressed in spite of younger ages at repair, smaller amounts of cardiac return and adequate numbers of segments (Fig. 3).

Postoperative catheterizations have so far been carried out in 31 patients 16 ± 13 months (1–40 months) after biventricular repair. Pulmonary arterial pressure was 8–40 mmHg, with a mean of 21 ± 9 mmHg. Pulmonary resistance was calculated to be 1.7–10 units/m², with a mean of 5.1 ± 2.1 units/m². Correlations were statistically seen between these parameters and age at repair (Fig. 4). Patients with CATCH 22 syndrome, including 2 late death cases, demonstrated relatively high pulmonary arterial pressure and high pulmonary resistance for their ages at repair.

Weak adverse correlation was seen between percent cardiac return and pulmonary resistance (Fig. 5). The number of pulmonary vascular segments could have an inverse affect on pulmonary resistance. Calculated pulmonary resistance values < 4 units/m² were mainly seen in patients with no segmental defects (Fig. 5). The use of a heterologous pericardial roll tube for reconstruction of the pulmonary arterial tree possibly resulted in higher pulmonary resistance (Fig. 5).

4. Discussion

Pulmonary circulation in patients with MAPCAs has been documented in the course of natural prognosis without any surgical interventions [3] or in terms of evaluation of status before definitive repair [2]. The circumstances of the circulation subsequent to surgical interventions, biventricular repair and unifocalizations, are also increasingly being described in a series of patients [5,12,14,15]. In the setting of abnormal arborization of the pulmonary arterial tree, the amount and distribution of pulmonary perfusion in one patient varies from that in another patient. In pulmonary vascular segments supplied by large and unobstructed MAPCAs, hypertensive pulmonary vascular change would readily progress, leading to undesirably high pulmonary resistance. In contrast, if a significant stenosis is present within a MAPCA or at the junction between the intrapulmonary PA and a MAPCA, pulmonary vasculature in the pulmonary parenchyma can be rather hypoplastic. Abnormal pulmonary perfusion is, therefore, closely associated with impaired pulmonary circulation with higher pulmonary resistance, duration of abnormal pulmonary perfusion probably being related to progression and severity of the pulmonary vascular change. In this respect, age at repair subsequent to, or concomitant with unifocalizations, is significantly correlated with postoperative pulmonary resistance.

In patients with severe cyanosis, development of minor systemic-to-pulmonary collateral vessels is commonly seen. The correlation between percent cardiac return and age at repair detected in this study is supportive of this clinical impression. As has been previously reported by our colleagues [4,8], the total amount of perfusion via such collaterals influence surgical re-
results after the Fontan type procedure. The present study also indicated that, in patients with MAPCAs, excessive development of such collaterals could be a risk factor for successful definitive repair as well as an unfavourable sign for efficient postoperative pulmonary circulation. Abundant collaterals around the oesophagus and the bronchial tree might be related to intractable haemorrhage shortly after extensive surgery. Furthermore, in patients having cardiac return >30%, postoperative pulmonary resistance was not <4 units/m².

Another important factor to be taken into account for better postoperative pulmonary circulation is undoubtedly the technical aspect of the operative procedures for unifocalizations. Since an adequate number of pulmonary vascular segments is needed for establishing successful surgical results in the short term after biventricular repair, as well as better pulmonary circulation in the longer term, the surgeon should strive to unify MAPCAs as much as possible by sensible operative procedures. Residual significant stenosis in the pulmonary arterial tree would be less attractive. In this respect, our preference has been for achieving unifocalization at the level of the intrapulmonary PA through the pulmonary fissures [5,20], because obstruction was frequently found at the junction between the MAPCA and the intrapulmonary PA in our patients. In patients where such an obstruction is lacking and the pulmonary arteries have sufficient sizes, earlier one stage repair, biventricular repair concomitant with unifocalization [11,13], should be the best option of choice, avoiding unfavourable development of hypertensive pulmonary vascular change. We use a prosthetic material as an artificial main pulmonary artery when the intrapericardial pulmonary arteries are severely hypoplastic or even absent. The operative procedures, as well as surgical strategy, should be considered according to the morphology of the pulmonary arterial tree [5,20].

It remains controversial whether use of a prosthetic material for unifocalization is reasonable. Of course, this is not ideal. Reconstruction of the pulmonary arterial tree using the intrapericardial PA, if present and not vestigial, is undoubtedly more attractive. It was suspected, on the basis of our present study, that use of prosthetic roll tubes was seemingly associated with higher postoperative pulmonary resistance. It should be noted, however, that more extensive operative proce-
Fig. 5. Influence of percent cardiac return, number of pulmonary vascular segments and operative procedures employed for unifocalization on postoperative pulmonary resistance.

...dures would be needed in patients with more severe malformation of the pulmonary arterial tree. In other words, more complicated pulmonary circulation could require more extensive surgery. It can be considered, therefore, that vestigial intrapericardial PA or absence of the central PA is frequently related to postoperative higher pulmonary resistance. In addition, as has been previously reported [5], patency of the unified pulmonary vascular segments was not related to operative procedures for reconstruction, but to the various pulmonary resistances of the pulmonary vascular segments initially supplied by MAPCAs independently.

Taking into consideration that postoperative pulmonary resistance would be high in patients with MAPCAs, we have used an external conduit bearing a handmade valve for reconstruction of the right ventricular outflow tract, at the time of definitive repair. As a matter of fact, the use of such an external conduit leads to postoperative morbidity and possible reoperation. If postoperative pulmonary resistance can be anticipated to be <4 units/m², however, use of an external conduit is not mandatory. Repair in the early stage of life is reasonable if the external conduit is not used and earlier repair can provide lower pulmonary resistance, in turn [17]. On the basis of our present study, <3 years of age at repair, <30% of cardiac return and no defect in pulmonary segments are the current criteria for reconstruction of the right ventricular outflow tract using no external conduit at the time of biventricular repair. Similarly, if efficient unifocalizations are established, the Fontan type procedure could be reasonably indicated in patients with ventricular morphology unsuitable for biventricular repair.

Finally, CATCH 22 syndrome has the potential to impair pulmonary circulation. This resembles a similar feature seen in patients with Down syndrome. In this respect, surgical repair should be planned particularly earlier if feasible. Disappointingly, however, an exceptionally high pulmonary resistance at a relatively young age at repair was seen in our patients with CATCH 22 syndrome, resulting in late death. It might be considered that we should have proceeded to surgical treatments much earlier in these patients. As a prospective policy, we will proceed to earlier repair subsequent to or concomitant with unifocalizations, not only in patients with CATCH 22 syndrome, but also in those without the features.

In summary, it is essential to accomplish effective surgical unifocalizations providing sufficient pulmonary perfusion and avoiding deleteriously deficient pulmonary vascular segments. In terms of postoperative pulmonary circulation, it is preferable to proceed to earlier definitive repair, so as to achieve better overall results.

References


Appendix A. Conference discussion

Dr Monro (Southampton, UK): Thank you Dr Uemura. Very nice presentation. Could you perhaps just explain for some people who may not have heard of this before your CATCH-22 syndrome. What do you mean by that?

Dr Uemura: Probably everybody knows about the CATCH-22. The clinical features have been described by several groups of investigators such as the group from Newcastle. The features should be known. The CATCH, C-A-T-C-H, were the initials for the clinical signs. And the basic deficiency is the minor deletion of the 22nd chromosome.

Dr Hagl (Heidelberg, Germany): Dr Uemura, do the data and correlations you have shown us reflect the whole cohort of patients or do they characterize the subgroup of 20 patients (so-called group 3) in whom pulmonary perfusion depends only on MAPCAs. In our experience patients with a small native pulmonary system and MAPCAs may have another course than patients who only depend on multifocal lung-perfusion by major aorto-pulmonary collaterals.

If not yet done, I would suggest to separate both groups to get more information about the pathophysiology of multifoca lung-perfusion.

Dr Uemura: I understand what you mean. As I showed on the slides, the majority of patients had the severe malformation of the pulmonary arterial tree. Only 6 patients had a relatively big size of the intrapericardial pulmonary artery. The majority of the patients, therefore, had MAPCAs with the intrapulmonary PA supplied by the MAPCAs mainly. I agree with you that if the pulmonary arterial anatomy has less severe malformation, then the prognosis can be better. As I showed on one of the latter slides, the technique used for the unifocalization was related to the pulmonary resistance. As I said, it probably reflects the morphology of the pulmonary arteries. All these patients had MAPCAs and of these 42 patients had severe pulmonary arterial arborization abnormality.