Median sternotomy single stage complete unifocalization for pulmonary atresia, major aorto-pulmonary collateral arteries and VSD-early experience

Kona S. Murthy*, Shivaprakasha Krishnanaik, Robert Coelho, Anil Punnoose, Sarasa B. Arumugam, Kotturathu M. Cherian

Institute of Cardiovascular Diseases, Madras Medical Mission, 4A, Dr. J. Jayalalitha Nagar, Mogappair, Chennai 600 050, India

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Abstract

Objective: It is a prospective study to assess the results of median sternotomy, single stage complete unifocalization and repair for ventricular septal defect (VSD), pulmonary atresia and major aorto pulmonary collateral arteries (MAPCAs). Methods: From June 97 to August 98, 20 patients were treated with single stage complete unifocalization and repair. Their ages ranged from 6 months to 11 years. Through median sternotomy, all MAPCAs were dissected and looped. On cardiopulmonary bypass, MAPCAs were anastomosed to native pulmonary arteries (PAs) or to MAPCAs. VSD was closed if possible and RV to PA continuity was established with a homograft conduit. If complete repair was not suitable, central shunt was done from ascending aorta to reconstructed PA with a polytetrafluoroethylene graft. The patients were divided into three groups according to the arborization pattern in the lungs. Group 1 had well formed native PAs with MAPCAs, group 2 had hypoplastic PAs with MAPCAs and group 3 had only MAPCAs. Results: Twenty patients had 21 procedures. All MAPCAs were unifocalized with tissue-to-tissue anastomosis for future growth, except one in whom polytetrafluoroethylene tube graft was used to attain the confluence. In group 1, all seven patients had complete unifocalization and repair. In group 2, four patients had RV to PA conduit and two patients had central shunt. In group 3, three patients had complete repair, three patients had RV to PA conduit and one patient had central shunt. There were three deaths, two in group 2 and one in group 3. The first patient died due to a wrong decision to close the VSD, the second patient died due to missed large MAPCA in preoperative angio and the third patient was a 7-year-old boy who died with irreversible pulmonary vascular changes due to unprotected MAPCAs. Conclusions: To conclude, complete repair/RV-PA conduit/central shunt should be done according to the size of the total pulmonary vasculature in patients with group 1, 2 and 3 with protected PAs/MAPCAs and in hypoplastic or absent PAs with unprotected MAPCAs (less than 1 year) and protected MAPCAs. We are yet to determine the surgical procedure to be performed in hypoplastic/absent PAs with unprotected MAPCAs more than 1 year. It is very essential to delineate all the MAPCAs up to the level of the diaphragm preoperatively. © 1999 Elsevier Science B.V. All rights reserved.

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1. Introduction

Ventricular septal defect (VSD) pulmonary atresia and major aorto pulmonary collateral arteries (MAPCAs) is a complex congenital cardiac anomaly characterized by variable sources of pulmonary blood supply and arborization abnormalities. The native pulmonary arteries (PAs) may either be present or absent and if they are present, they are either normal in size or hypoplastic and non-confluent. The MAPCAs are variable in number, origin, size and course. An area of the lung may receive its blood supply from the native PA or MAPCAs alone, or from both. They present with symptoms of decreased or increased or balanced pulmonary blood flow (PBF). The conventional approach for these patients has been multistaged unifocalization followed by final correction [1–3]. Recently, median sternotomy single stage unifocalization and complete repair has been reported by Reddy and Hanley [4]. We report our early experience with single stage unifocalization and repair for this anomaly.
2. Methods

From June 1997 to August 1998, 20 patients were treated by median sternotomy, single stage complete unifocalization and repair. The diagnosis was made or suspected by echocardiography and confirmed by cardiac catheterization and angiography. An effort was made to get the pressure measurements of individual MAPCAs along with selective delineation of them by angiography. Clinical details are given in Table 1.

They were divided into three groups according to the morphological features of native PAs and MAPCAs and their arborization pattern in the lungs. Group 1 had well-formed PAs. Group 2 had hypoplastic PAs and group 3 had absent PAs. All three groups were further subcategorized into (a) with protected MAPCAs (b) with unprotected MAPCAs. The details are given in Table 2.

Our aim was to establish a single source of blood supply to the lungs from PAs and MAPCAs and promote their growth in children. Even though there was dual supply and the MAPCAs were big size (>2 mm), they were still unifocalized to maximize the number of pulmonary arterial segments available for eventual correction, on the assumption that the postoperative RV pressure would be lower. All patients had single stage unifocalization and complete repair (VSD closure and RV to PA conduit) or RV to PA conduit alone or central shunt, according to the availability of size and number of pulmonary vascular segments. If the pulmonary arterial size was more than 75% of expected or more than 15 segments of the lung, complete repair was indicated. If the PA size was between 50–75% of normal or the pulmonary segments were between 10–14, RV to PA conduit was done without closing the VSD. If the PA’s size was less than 50% of expected or PA segments less than ten, only central shunt was performed.

2.1. Surgical technique

After median sternotomy, a pericardial patch was removed for subsequent use. The native PAs (if present) were dissected and isolated up to their hilar region. The ascending aorta (AO) and the superior vena cava (SVC) were freed from their surrounding tissues in order to retract them freely to get good access to the deeper plane for dissection. The MAPCAs were approached by dissecting along the aorta and the brachiocephalic arteries, as per their course delineated by the preoperative angiography, to a sufficient length from their origin to bring them to transverse sinus without any tension during anastomosis. Opening of the pleura was not necessary because of limited dissection. The descending aortic MAPCAs were reached by dissection in the posterior mediastinum after opening the posterior pericardium. The right side MAPCAs were reached by approaching between ascending AO, SVC and the roof of the left atrium (LA), usually above or below the carina and right main bronchus. Left side MAPCAs were approached by dissecting between the area of left side of ascending aorta and LA and above or below the left main bronchus. Presence of left SVC and mediastinal lymph nodes make the dissection more difficult. During the dissection, care was taken to avoid hemodynamic compromise by momentarily stopping the dissection and retraction. Precautions were taken not to injure the trachea, bronchi, oesophagus and phrenic, vagus and recurrent laryngeal nerves. All the MAPCAs were controlled before going on to cardio pulmonary bypass (CPB).

Under CPB and beating heart, all the MAPCAs were disconnected from their origin and the proximal end was closed. They were anastomosed end-to-side or side-to-side to native PAs if present; otherwise MAPCAs-to-MAPCAs (Fig. 1). Anastomosis was done using 8/0 polypropylene continuous sutures. Tissue-to-tissue anastomosis was preferred to allow future growth in the children. Under cardioplegic arrest, the VSD was closed with a PTFE patch and RV to PA continuity was established by cryopreserved aortic or pulmonary homograft conduit. If they were not suitable for complete repair, VSD was left alone or central shunt was done from ascending AO to PA with a PTFE interposition graft on a beating heart.
3. Results

All patients had single stage complete unifocalization through median sternotomy. The postoperative results are given in Table 1. Twenty patients had 21 surgical procedures. Ten (50%) had complete repair (VSD closure and RV to PA conduit), seven (35%) patients had RV to PA conduit alone (VSD left open), three (15%) patients had central shunt. Tissue-to-tissue anastomosis was achieved in all patients except one, in whom a 14 mm PTFE tube graft was used to achieve the confluence of the reconstructed PAs (Fig. 2). The mean bypass time was 151 ± 28 min (120–208), the mean AO cross clamp time was 47 ± 20 min (18–95). In the complete repair group, the mean RV, LV pressure (P RV/LV) ratio was 0.66 (0.38–0.87) and the systemic oxygen saturation ranged 95–100%. In situations where the VSD was left open and RV to PA conduits or central shunts were performed, the systemic oxygen saturation ranged from 92–97%. A summary of MAPCAS is given in Table 2.

Table 2
Profile of MAPCAS

<table>
<thead>
<tr>
<th>Condition</th>
<th>Occurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total no. of MAPCAS</td>
<td>54 (range 1–5) median = 3</td>
</tr>
<tr>
<td>Descending Ao</td>
<td>36 (66%)</td>
</tr>
<tr>
<td>Arch</td>
<td>5 (9.2%)</td>
</tr>
<tr>
<td>Ascending Ao</td>
<td>2 (3.7%)</td>
</tr>
<tr>
<td>Subclavian art</td>
<td>11 (20%)</td>
</tr>
</tbody>
</table>

Fig. 1. Reconstruction of MAPCAS to neopulmonary artery. (A) Preoperative diagram. (B) (a–d) Reconstruction of neo PA. (C) Postoperative diagram. (D) Postoperative angio. PAS, pulmonary arteries; MAPCAS, major aorto pulmonary collateral arteries; Preop, preoperative; Postop, postoperative; RPA, right pulmonary artery; LPA, left pulmonary artery; LSA, left subclavian artery; RL, right lung; LL, left lung; Ao, aorta; RV, right ventricle; LV, left ventricle.
85 to 96% (mean 92%) and 74–88% (mean 82%), respectively.

There were three hospital deaths (15%). There were no deaths in group 1, two deaths in group 2 and one death in group 3. The first patient belonged to group 2, who had complete repair and could not come off bypass due to suprasystemic RV pressure. He went back on bypass and VSD patch was removed. He had prolonged bypass time (208 min). Postoperatively, he developed low cardiac output due to RV dysfunction and died on the 7th postoperative day. The second patient was a 1-year-old child from group 2, who had complete unifocalization and RV to PA conduit without closure of VSD. Postoperatively, he developed low cardiac output due to RV dysfunction and died on the 7th postoperative day. The third patient was a 7-year-old boy with unprotected MAPCAs, who died on the 4th postoperative day due to disaturation and ventricular failure. Autopsy of the lung specimen showed grade III pulmonary vascular changes according to the Heath Edwards classification [5]. Three patients were taken up for re-exploration for excessive mediastinal bleeding. One patient had unilateral phrenic palsy and another patient had bilateral phrenic palsy. Both required tracheostomy and prolonged ventilation.

The follow-up ranged from 1 month to 1.3 years. In group 3, one patient who had complete unifocalization and RV to PA conduit presented with congestive heart failure within 2 months of discharge. Investigations showed left to right shunt across the VSD, which was closed to achieve complete repair. In postoperative follow-up, all the survivors were in NYHA class I. The echocardiograms during the follow-up period have shown a mean PRV/LV of 0.72 (0.5–0.9). One patient in whom PTFE tube graft was used to
achieve the confluence developed anastomotic stenosis and he is awaiting balloon dilatation. There were no late deaths.

4. Discussion

Pulmonary atresia, MAPCAs and VSD, remains one of the most challenging groups to manage surgically. In the earlier period, these patients were treated with conventional multistage procedure. Previously published data showed that multistage procedure required a median of three procedures (range 2–6) before complete repair. This culminates in complete repair in 11.5–60.5% of patients. Overall mortality to achieve complete repair has ranged from 10.2–19.2%. From 1993 to May 1997, these patients were treated with multistage unifocalization in our institute. Fourteen patients had 21 procedures (1.5 per patient) in which only three of them had complete repair. There were two deaths (14%), one was due to a blocked shunt in whom pericardial tube was used for unifocalization. Another patient who developed aneurysmal dilatation of homograft tube which was used for unifocalization, died after final correction due to low cardiac output. The two patients who survived complete repair had echocardiogram; cardiac cath and angio cardiogram, RV/LV pressure ratio was 0.5 and 0.92, respectively [6].

From June 1997 onwards, we started doing single stage unifocalization through median sternotomy. Till August 1998, 20 patients were treated with this technique. During this period, all patients were treated with single stage unifocalization and no patient was refused for surgery or used multistage thoracotomy approach. Ten patients (50%) had complete repair, seven patients had RV to PA conduit and three patients had central shunt. Reddy and Hanley reported that 25 infants had complete unifocalization through midline, out of which 17 patients (68%) had complete repair and in eight (32%) patients VSD was left open. There were two deaths (8%) [7].

The surgical management of this complex anomaly should be individualized according to arborization of pulmonary vasculature and amount of pulmonary blood flow. We hence divided the patients into three groups depending upon the pulmonary arterial tree as mentioned earlier. The results we obtained in each group clearly reveals that the group 1 patients are the better spectrum of this anomaly. The hospital deaths in group 2 certainly highlight difficulty in choosing the type of surgery to be undertaken. Evolution of decision-making in these groups and aggressive search for all the MAPCAs during and before surgery would obviate this problem. Minimising the CPB time need not be emphasized. Benefit of doubt of the arborization should be given for not closing the VSD rather than for closing it. Group III patients clearly have all the uncomfortable components of the anatomy, absent PAs, unrestric-

tive MAPCAs and older age. They develop irreversible pulmonary vascular disease if we do not treat them at a younger age, as shown in one of our patients who died with grade III–IV pulmonary vascular changes.

To conclude, based on the experience, we would like to draw up a few guidelines for the surgical management of this anomaly. 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 (less than 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 are 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 cardio vascular physiology with good future growth of unifocalized neo pulmonary arteries.

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References