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Staged operation for pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries

New technique for complete unifocalization

A new staged operation for total correction of pulmonary atresia and ventricular septal defect with major aortopulmonary collateral arteries has been developed. In first-stage repair (complete unifocalization), intrapulmonary arteries were unified at the hilum with equine pericardial conduits (intrapulmonary bridges). In case of absent or severely hypoplastic central pulmonary arteries, new central pulmonary arteries were created. Finally, the unifocalization was completed by modified Blalock-Taussig shunts with the ligation of collateral arteries. In second-stage repair, right ventricular–pulmonary arterial continuity was established with a trileaflet pericardial conduit and closure of the ventricular septal defect. From January 1982 through July 1988, 34 patients, whose ages ranged from 1 month to 24 years (mean 6.6 years), underwent first-stage repair with two resultant late deaths (mortality rate 5.9%). Second-stage repair has been completed in 16 patients. There were two early deaths (mortality rate 12%) from bacterial infection. Postoperative right ventricular/left ventricular systolic pressure ratios ranged from 0.36 to 1.0 (mean 0.72). In four patients in whom the ratio was 1.0, the ventricular septal defect had been closed with a perforated patch. By complete unifocalization with the intrapulmonary bridge technique and the creation of new central pulmonary arteries, the majority of patients with major aortopulmonary collateral arteries can have successful repair.

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The current standard surgical treatment of pulmonary atresia and ventricular septal defect is generally associated with good results.[1-6] However, when the procedure involves major aortopulmonary collateral arteries (MAPCAs), the pulmonary arterial vasculature has complicated anomalies that still provide serious obstacles to successful corrective operations.[1-8] In this anomaly, the primary pulmonary blood source is multifocal[9]; the central pulmonary arteries, which are frequently severely hypoplastic or absent, connect with only a limited number of intrapulmonary arteries because of arborization abnormalities, whereas isolated segments are directly supplied from MAPCAs.

Our current management for these patients is a staged operation: (1) first-stage repair, complete unifocalization; (2) second-stage repair, establishment of right ventricular–pulmonary arterial continuity[10] (Fig. 1). This report describes our surgical experiences and discusses operability and surgical approach in these patients.

Patients

From January 1982 through July 1988, 34 patients underwent the first-stage repair at The Heart Institute of Japan, Tokyo Women's Medical College. The group comprised 16 male and 18 female patients with ages ranging from 1 month to 24

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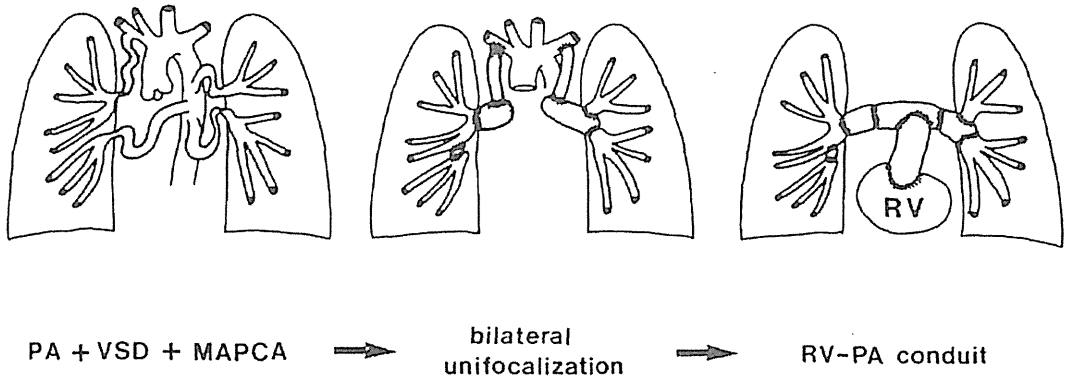

Fig. 1. Schematic representation of the staged operation. *Left,* A typical form of pulmonary atresia and ventricular septal defect with MAPCAs. *Middle,* First-stage repair: Complete unifocalization. *Right,* Second-stage repair: The establishment of right ventricular–pulmonary arterial *(RV-PA)* continuity along with closure of a ventricular septal defect *(VSD).*

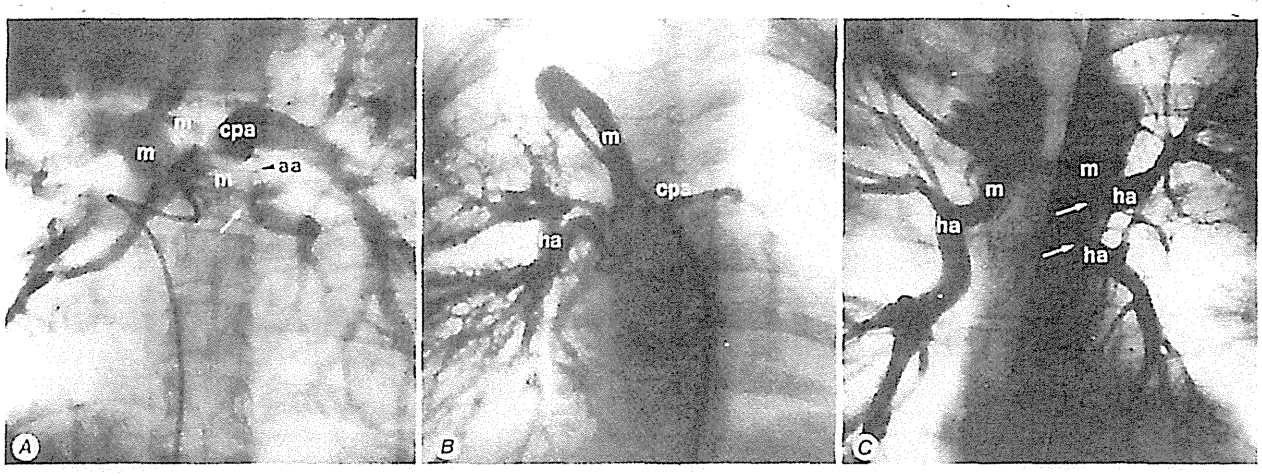

Fig 2. Characteristic types of the central pulmonary arteries in pulmonary atresia and ventricular septal defect with MAPCAs (pulmonary angiograms obtained by selective MAPCA injections). A, Moderately hypoplastic central pulmonary arteries. B, Severely hypoplastic central pulmonary arteries. C, Absent central pulmonary arteries. *aa,* Arborization abnormality; *cpa,* central pulmonary artery; *ha,* Hilar pulmonary artery. *m,* MAPCA; *white arrow,* hilar stenosis of the connection between the MAPCA and the hilar pulmonary artery.

years (mean age 6.6 years); there was no difference in the mean age between male and female patients. Age distribution was as follows: Six patients were younger than 1 year, 12 patients were between 1 and 5 years, and the other 16 patients were older than 5 years. Arterial oxygen saturations with the patients breathing room air ranged from 57% to 90% (mean 76%), and arterial oxygen partial pressures ranged from 27 to 50 mm Hg (mean 42 mm Hg). Sixteen patients (47%) had a right aortic arch, 12 (75%) of them being female. Student's *t* test was used for the statistical analysis.

Pulmonary arterial anatomy and surgical classification. Pulmonary arterial structures are designated extrapulmonary and intrapulmonary. Extrapulmonary arteries are formed by the pulmonary trunk and right and left central pulmonary arteries. Intrapulmonary arteries are formed by secondary and subsequent branches and retain elastic structures from the hilum (hilar pulmonary arteries). MAPCA is defined as a large tortuous collateral artery, which almost always arises from the anterior surface of the descending aorta just beneath the carina or subclavian arteries, loops independent of the course of the bronchus to the lung, and connects with intrapulmonary arteries at the posterior aspect of the hilum.[11] When pulmonary arterial blood flow is diminished in congenital heart diseases, enlarged bronchial arteries, which do not normally communicate directly with the lumina of pulmonary arteries, can affect precapillary anastomosis.[12] MAPCAs should be differentiated from these enlarged bronchial arteries, which run in the wall of the bronchus and connect with pulmonary arteries in the region of capillary arteries.

Preoperatively, the anatomy of pulmonary arterial vasculatures and MAPCAs was carefully delineated by repeated selective injections of every MAPCA. The average number of the

Fig. 3. Lateral view of pulmonary angiogram, obtained by selective MAPCA injection, demonstrating peripheral pulmonary stenosis *(white arrow)*.

MAPCAs at the hilum was 3.4 (range 2 to 6) per patient. Sixty-five percent of the MAPCAs had a stenotic segment localized in the region of the hilum just before the connection with the hilar pulmonary arteries[13-16] (Fig. 2). These hilar stenoses were either long or short segments. The progressive obstruction of hilar stenosis was observed in three patients before the first-stage repair. Peripheral pulmonary stenosis in the hilar region was also found in five patients (Fig. 3). The pulmonary trunk was absent in 31 patients. The central pulmonary arteries showed various degrees of hypoplasia. As a whole, the growth of the intrapulmonary arteries was relatively good compared with that of the extrapulmonary arteries. However, serious complications regarding the growth of intrapulmonary arteries, severe hypoplasia or severe pulmonary hypertension, were present in 10 patients (29%): In five of them, the intrapulmonary arteries were severely hypoplastic. In the other five, severe pulmonary hypertensive obstructive change in most parts of the lungs was suggested by pulmonary angiograms. These 10 patients showed the following characteristics: (1) The mean age at first-stage repair was significantly higher than that of the other 24 patients ($p < 0.05$). (2) A right aortic arch was encountered in nine patients (90%), 80% of them female.

Our surgical classification is given in Table I. All surgical cases are summarized in Tables II and III according to this surgical classification, based on the growth of central pulmonary arteries and the presence or absence of arborization abnormalities (Fig. 2). The size of central pulmonary arteries was evaluated by the pulmonary artery index (PA index)[17], which indicates the pulmonary arterial cross-sectional area divided by the body surface area (normal value 330 ± 30 mm^2/m^2 body surface area). The pulmonary arterial cross-sectional area was calculated by measuring the diameters of the right and left pulmonary arteries just proximal to the first lobar branch on the pulmonary angiogram. The central pulmonary arteries were moderately hypoplastic (type 1, PA index ≧ 60) in 17 patients, severely hypoplastic (type 2, PA index < 60) in 12 patients, and absent (type 3) in five patients (Fig. 2). There were no significant differences in the mean ages at the first-stage repair among these three types. However, the distribution by sex was uneven; 76% of patients with type 1 arteries were male, whereas 92% of those with type 2 and 20% of those with type 3 were female. A total of 33 arborization abnormalities were present in 22 patients (65%). The number of arborization abnormalities per patient ranged from one to three. The size of isolated segments varied greatly from one bronchopulmonary segment to more than one lobe. The degree of arborization abnormality in each patient was represented in six grades, grade 1/6 to grade 6/6, by the ratio of combined isolated area to total pulmonary arterial area, in which the combined isolated area was estimated by considering one sixth of the total pulmonary arterial area as one unit.

Table I. *Surgical classification of 34 patients with pulmonary atresia and ventricular septal defect with MAPCAs*

Surgical classification	*No. of patients*
1. Moderately hypoplastic central pulmonary arteries (PA index ≧ 60)	
a. Arborization abnormalities (−)	7
b. Arborization abnormalities (+)	10
2. Severely hypoplastic central pulmonary arteries (PA index < 60)	
a. Arborization abnormalities (−)	2
b. Arborization abnormalities (+)	10
3. Absent central pulmonary arteries	
a. Arborization abnormalities (−)	3
b. Arborization abnormalities (+)	2

PA index = (RPA area + LPA area)/BSA (normal range 330 ± 30 mm^2/m^2 BSA), where RPA is right pulmonary artery, LPA is left pulmonary artery, and BSA is body surface area.

First-stage repair (complete unifocalization). First-stage repair, complete unifocalization, is aimed to remodel the pulmonary arterial structures (Fig. 1). Here the term *unifocalization* means to convert multifocal to unifocal pulmonary blood supply, according to the definition suggested by Macartney and Haworth[11]: For the purpose of unifocalization, "unification" of intrapulmonary arteries is necessary. This first-stage repair is performed separately on the right and left lungs by the standard technique through a posterolateral thoracotomy. Understanding the anatomy of pulmonary arteries and MAPCAs at the hilum is indispensable for complete unifocalization. Because of "wash-in" and "wash-out" phenomena,[18] the preoperative information by MAPCA injections may be erroneous; therefore the connections between the central pulmonary arteries, the hi-

Table II. *Surgical cases and results of type 1 disease (moderately hypoplastic central pulmonary arteries)*

						First-stage repair			Second-stage repair			
Patient	*Sex*	*Aortic arch*	*Surgical classification (right, left)*	*Grade of arborization abnormality*	*Severities of intrapulmonary arteries*	*Age (yr)*	*PA index*	*Intrapulmonary bridge (d)*	*Age (yr)*	*PA index*	*Postop. RVP/LVP*	*Death*
1	M	R	(1a, 1a)	0/6		0.1						
2	M	L	(1a, 1a)	0/6		0.3	136		3.2	240	0.60	
3	M	R	(1a, 1a)	0/6		0.5	102		2.5	171	0.68	
4	M	L	(1b, 1a)	2/6		0.6	108	8 mm	3.1	175	0.65	
5	M	L	(1b, 1a)	2/6		3.0	72	12 mm				
6	M	L	(1b, 1b)	4/6		3.0	70	12 mm				
7	M	R	(1b, 1a)	1/6	Severe hypoplasia	3.0	66					
8	M	L	(1a, 1a)	0/6		3.4	125					
9	F	R	(1a, 1a)	0/6	Severe hypoplasia	4.0	74					
10	M	R	(1a, 1b)	2/6	Severe PH (broad)	5.0	150	12 mm	5.2	196	1.00*	
11	M	L	(1b, 1a)	3/6		6.0	167	13 mm	6.5	296	0.73	
12	M	L	(1a, 1a)	0/6		7.0	100		12.0	160	0.52	
13	F	R	(1b, 1b)	4/6		8.0	230	10 mm	8.6	230	0.75	
14	*M*	*L*	*(1a, 1b)*	*2/6*		*9.0*	*360*	*8 mm*	*9.1*	*360*	*0.72*	
15	M	L	(1a, 1a)	0/6		10.0	193		10.0	190	0.65	
16	F	L	(1b, 1a)	1/6		11.0	208		13.0	207	0.65	Death
17	F	R	(1b, 1b)	4/6	Severe PH (broad)	15.0	145	12 mm				

For aortic arch: R, Right aortic arch; L, left aortic arch. Grade of arborization abnormality: See text. PA index: See text and legend to Table I. PH, Pulmonary hypertension; d, diameter; RVP/LVP, peak systolic pressure ratio of right ventricle to left ventricle.

*Perforated patch closure of ventricular septal defect.

lar pulmonary arteries, and MAPCAs were verified by dissecting the parietal and visceral pleura of posterior hilum toward the lung.

Unification of intrapulmonary arteries (Figs. 4 and 5). When arborization abnormalities were present, the intrapulmonary arteries were unified by restoring the continuity between hilar pulmonary arteries in the hilum or inside the lung. By careful dissection of the visceral pleura toward the lung and separation of interlobar fissures, the interrupted portion of the hilar pulmonary arteries was exposed. Hilar stenoses of the MAPCAs, located at the junction to the hilar arteries, were avoided by not using MAPCAs to unify the intrapulmonary arteries. Instead, the continuity between hilar pulmonary arteries was restored by interposition of a prosthetic conduit (intrapulmonary bridge). Intrapulmonary bridges, 8 to 13 mm in diameter, were tailored of equine pericardium (Xenomedica AG, Luzern, Switzerland) according to each individual situation. In placing intrapulmonary bridges, we were careful to avoid compressing the surrounding pulmonary veins and bronchial tree.

The intrapulmonary bridge technique was used in 15 patients in whom arborization abnormalities were present. It was also used in three patients to relieve peripheral pulmonary stenoses in the region of the hilum. However, when the isolated segment was only a small bronchopulmonary segment, the segmental artery of the isolated segment was directly anastomosed to the appropriate portion of the hilar pulmonary arteries communicating with the central pulmonary artery (Fig. 6). This direct anastomosis between intrapulmonary arteries was used in nine patients.

Remodeling of central pulmonary arteries (Figs. 6 and 7). As a part of complete unifocalization, we remodeled the central pulmonary arteries so that they would be large enough to attach to hilar pulmonary arteries. Besides, central pulmonary arteries should be large enough (PA index $\geqq$ 120) to function as the conduit between the right ventricle and intrapulmonary arteries in the second stage of the repair. We tried to enlarge hypoplastic central pulmonary arteries in the following fashion: When the central pulmonary arteries were moderately hypoplastic (type 1, PA index $\geqq$ 60), we expected the modified Blalock-Taussig shunts to stimulate their growth. If they did not grow sufficiently, an additional extensive patch angioplasty to the hilum was done in the second-stage repair. When the central pulmonary arteries were severely hypoplastic (type 2, PA index $<$ 60) or absent (type 3), new central pulmonary arteries were created. However, this was not done in the first six patients with type 2 arteries.

New central pulmonary arteries were created with Xenomedica conduits, which are tailored to each individual anatomic situation. These conduits were designed to imitate the natural central pulmonary arteries; thus they were slightly curved and tapered. In type 2 the hypoplastic central pulmonary arteries gradually passed into the hilar pulmonary arteries at the hilum (Fig. 6), whereas in type 3 the MAPCAs were directly connected to the hilar arteries at the hilum (Fig. 7). In both cases the transitional part of the hilar pulmonary arteries from the central pulmonary arteries or MAPCAs was relatively narrow. Hence dissecting into the lung and separating the interlobar fissures exposed hilar pulmonary arteries whose orifices were large enough to allow anastomosis with the Xenomedica conduits. In type 2 the native hypoplastic central pulmonary arteries were

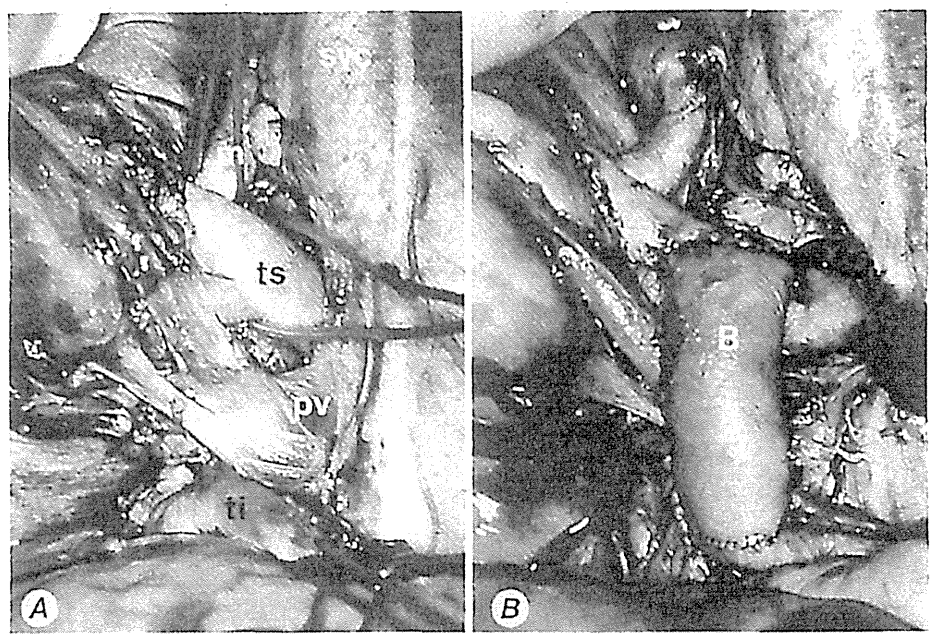

Fig. 4. Intrapulmonary bridge technique: Intraoperative photographs of right complete unifocalization during first-stage repair (patient 11). A, Preoperative view. The inferior trunk *(ti)* was isolated from the superior trunk *(ts)*, which communicated with the central pulmonary artery. *pv*, Right upper pulmonary vein; *svc*, superior vena cava. *m*, MAPCA B, Postoperative view. Intrapulmonary bridge *(B)* was inserted between the superior and inferior trunks in the hilum.

Table III. *Surgical cases and results in type 2 disease (severe hypoplastic central pulmonary arteries) and type 3 disease (absent central pulmonary arteries)*

						First-stage repair				*Second-stage repair*			
Patient	*Sex*	*Aortic arch*	*Surgical classification (Right, Left)*	*Grade of arborization abnormality*	*Severities of intrapulmonary arteries*	*Age (yr)*	*PA index*	*Central creation (d)*	*Intrapulmonary bridge (d)*	*Age (yr)*	*PA index*	*RVP/LVP*	*Death*
18	F	R	(2b, 2b)	4/6	Severe PH (broad)	2.0	54		8.12 mm	2.3	75	1.00*	
19	F	L	(2b, 2b)	2/6		4.0	34				50		
20	F	R	(2b, 2b)	3/6	Severe hypoplasia	4.0	41	16 mm, 16 mm	12 mm				
21	F	L	(2a, 2a)	0/6		5.0	50				70		
22	F	R	(2a, 2a)	0/6		5.0	43	12 mm, 12 mm		8.0	—	0.80	
23	F	R	(2a, 2b)	3/6		5.0	50		12 mm		50		
24	F	R	(2b, 2b)	2/6		8.0	37				57		
25	F	L	(2b, 2a)	1/6		8.0	32	16 mm, 16 mm					
26	F	R	(2a, 2b)	2/6	Severe hypoplasia	8.0	45	18 mm, 20 mm	12 mm				Death
27	M	L	(2a, 2b)	1/6		10.0	30	13 mm, 18 mm		11.5	—	1.00*	
28	F	R	(2b, 2b)	4/6		13.0	45		13 mm		60		
29	F	R	(2a, 2b)	2/6	Severe PH (broad)	14.0	44	12 mm, 12 mm	12 mm				
30	F	L	(3a, 3a)	0/6		1.0	0	14 mm, 14 mm		2.3	—	0.36	
31	F	L	(3a, 3a)	0/6		1.0	0	14 mm, 16 mm		1.1	—	0.39	
32	M	L	(3b, 3b)	2/6		6.0	0	16 mm, 16 mm					
33	F	R	(3a, 3a)	0/6	Severe PH (broad)	15.0	0	20 mm, 21 mm		16.0	—	1.00*	Death
34	F	L	(3b, 3b)	4/6	Severe hypoplasia	24.1	0	20 mm, 21 mm	12 mm				Death

For legend see Table II.

*Perforated patch closure of ventricular septal defect.

Volume 98
Number 5, Part 1
November 1989

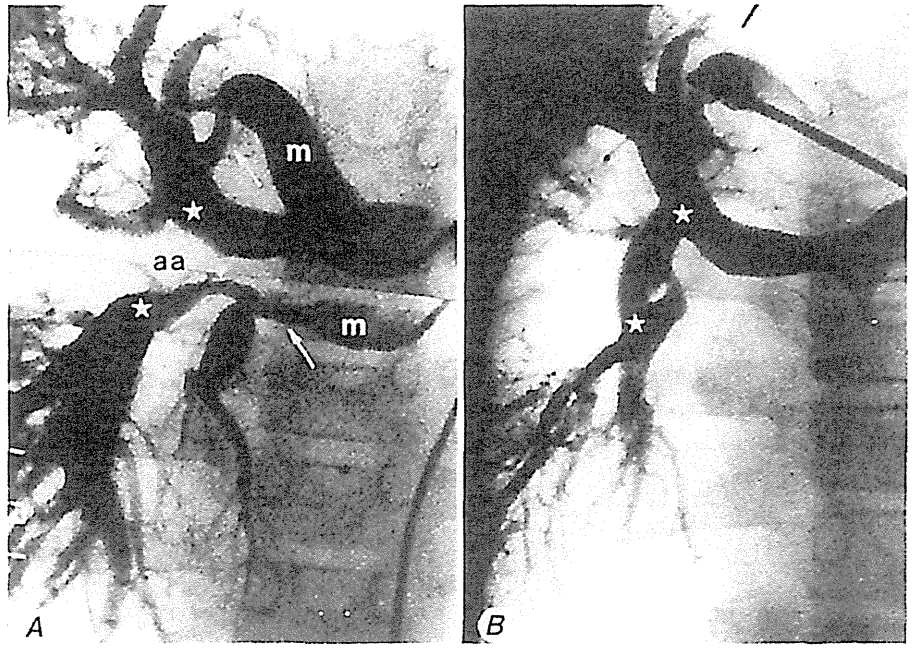

Fig. 5. Pulmonary angiograms of the right lung before and after right complete unifocalization in patient 11. A, Preoperative angiogram composed of two selective MAPCA injections. *aa,* Arborization abnormality; *white stars,* discontinuity between the superior and inferior trunks; *m,* MAPCA; *white arrow,* hilar stenosis of the MAPCA. B, Postoperative angiogram obtained by injection into the modified Blalock-Taussig shunt. Discontinuity *(two white stars)* between the superior and inferior trunks was restored by the intrapulmonary bridge.

left open. New central pulmonary arteries were created in six patients with type 2 and five patients with type 3 arteries.

Modified Blalock-Taussig shunt with MAPCA ligation (Fig. 7). Finally, complete unifocalization was accomplished by a modified Blalock-Taussig shunt with a 5 mm polytetrafluoroethylene (PTFE) tube,* concurrent with ligations of every MAPCA. When a central pulmonary artery was created with a Xenomedica conduit, a modified Blalock-Taussig shunt was placed about 5 mm apart from the proximal end of the conduit, not only to minimize the turbulent flow but also to allow for placement of a vascular clamp in the second-stage repair. Then, the proximal end of the conduit was anchored to the anterior or middle mediastinum, and a PTFE sheet was placed around the modified Blalock-Taussig shunt so that the shunt could be easily ligated through a median sternotomy in the second-stage repair.

Postoperative evaluation. There were no early deaths. The postoperative arterial oxygen saturations with the patients breathing room air ranged from 68% to 89% (mean 80%) and arterial oxygen partial pressures ranged from 36 to 49 mm Hg (mean 44 mm Hg). The late mortality rate was 5.9%. Two patients, who had broad, severe pulmonary hypertensive obstructive disease or severe hypoplasia of the intrapulmonary arteries preoperatively, died of gastrointestinal bleeding and intratracheal bleeding 1 month and 9 months, respectively, after the first-stage repairs. Postoperative pulmonary angiograms were performed in 26 patients to evaluate the result of complete unifocalization. The intrapulmonary bridges were patent and functioning well in all patients (Fig. 5). In one patient in whom new central pulmonary arteries were created, the new left central pulmonary artery was completely obstructed because of an unexpected shunt failure.

In the patients who had not undergone creation of central pulmonary arteries, the growth of the central pulmonary arteries before and after complete unifocalization is indicated as the change in the PA indexes in Fig. 8. When the preoperative PA index was greater than 60, the PA index increased satisfactorily; when the preoperative PA index was less than 60, the PA index did not increase satisfactorily. Among the 32 patients who survived the first-stage repair, 16 patients have undergone the second-stage repair (Tables II and III). The mean interval between first-stage and second-stage repair was 10 months. Unilateral complete unifocalization was performed concurrent with second-stage repair in four patients. Of the remaining 16 patients, 11 are now awaiting second-stage repair as scheduled; the other five patients, in whom broad, severe pulmonary hypertensive obstructive disease or severely hypoplastic intrapulmonary arteries were present preoperatively, are now in the period of postoperative follow-up.

Second-stage repair (Fig. 9). In second-stage repair, right ventricular–pulmonary arterial continuity was established along with closure of a ventricular septal defect through a median sternotomy with moderately hypothermic cardiopulmonary bypass and cold cardioplegia. Before cardiopulmonary bypass was established, the anastomosis between the central pulmonary arteries and a right ventricular–pulmonary arterial prosthetic conduit was performed by placing vascular clamps proximal to the modified Blalock-Taussig shunt on each side. The right ventricular–pulmonary arterial Xenomedica conduit was tailored according to the individual anatomy and equipped with a trileaflet pulmonary valve. When new right and left central pulmonary arteries were created in the first-stage repair, a T-

*Gore-Tex vascular graft, registered trademark of W. L. Gore & Associates, Inc., Elkton, Md.

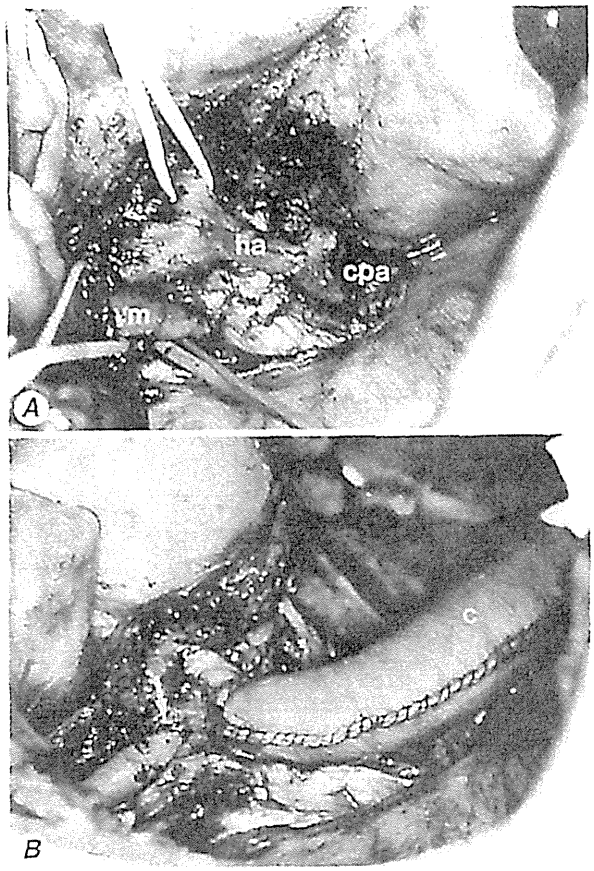

Fig. 6. Creation of a new central pulmonary artery in a case of severely hypoplastic central pulmonary artery (type 2). Intraoperative photographs of left complete unifocalization during first-stage repair. A, Preoperative view. The hypoplastic left central pulmonary artery *(cpa)*, 4 mm in diameter, continued to the hilar pulmonary artery *(ha)*. Note the narrowing feature of the connecting portion of the hilar pulmonary artery. One bronchopulmonary segment was isolated and supplied from a small MAPCA *(m)*. B, Postoperative view. Xenomedica conduit *(c)* was anastomosed to the large hilar pulmonary artery beyond the stenosis. The segmental artery was directly anastomosed to the hilar pulmonary artery *(white arrow)*.

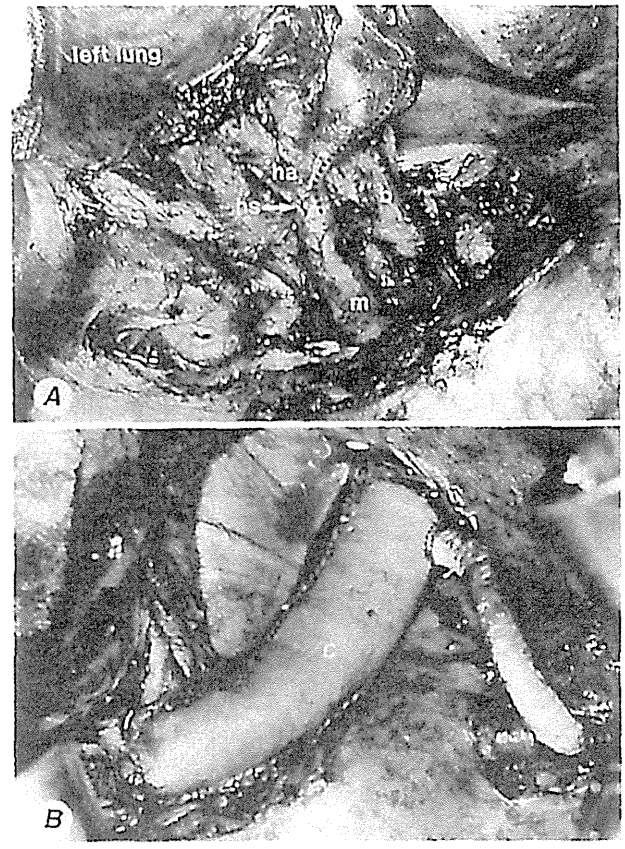

Fig. 7. Creation of a new central pulmonary artery in a case of absence of the central pulmonary arteries (type 3). Intraoperative photographs of left complete unifocalization during first-stage repair. A, The left hilar artery *(ha)* was directly supplied from a single MAPCA *(m)*. Note the stenosis *(hs)* between the MAPCA and the hilar pulmonary artery. *b*, Bronchus; *dashed line*, incision for the anastomosis of the conduit. B, Postoperative view. The conduit *(c)* was anastomosed to the large hilar pulmonary artery with a sufficient orifice, and a modified Blalock-Taussig shunt was placed to the proximal end of the conduit.

shaped Xenomedica conduit was applied for the right ventricular–pulmonary arterial conduit. In this circumstance, the transverse portion of the conduit, which joins the proximal portions of the created right and left central pulmonary arteries, was placed posterior to the ascending aorta or brought up cephalad over the aortic arch to avoid the external compression.

The modified Blalock-Taussig shunts were ligated concurrent with the establishment of cardiopulmonary bypass, and patch closure of the ventricular septal defect was performed. Finally, the proximal anastomosis of the right ventricular-pulmonary arterial conduit was performed. In three patients, the pulmonary trunk was present. Thus patch reconstruction of the right ventricular outflow tract was successfully done.

Results

There were two early deaths (mortality rate 12%) resulting from bacterial endocarditis; one of these cases was also complicated by broad, severe pulmonary hypertensive obstructive disease preoperatively. Sixteen patients survived the second-stage repair and underwent postoperative catheterization about 1 month after the operation. The postoperative peak systolic pressure ratios of the right ventricle to the left ventricle (RVP/LVP) ranged from 0.36 to 1.0 (mean 0.71) and were not correlated with the preoperative PA indexes. The RVP/LVP was 1.0 in four patients in whom the perforated patch closure of the ventricular septal defect was used, for the following reasons: In three patients, severe, broad pulmonary hypertensive

change was suggested in the preoperative evaluation; in the other patient, the created left central pulmonary artery was completely obstructed before the second-stage repair.

Discussion

In the surgical treatment of pulmonary atresia and ventricular septal defect, MAPCAs have presented challenging problems for the cardiac surgeon. In these patients the presence of MAPCAs is associated with the complicated abnormalities of the pulmonary arterial vasculature, which form characteristic features in the morphologic and hemodynamic aspects and prevent successful corrective operations.

Morphologically, the most critical problem for successful corrective operations is arborization abnormalities of the intrapulmonary arteries. When arborization abnormalities are present, only a limited number of the intrapulmonary arteries can be connected to the central pulmonary arteries, whereas the other parts of intrapulmonary arteries are isolated and directly supplied from MAPCAs. The restricted pulmonary blood flow by arborization abnormalities is one of the major causes of postoperative right ventricular hypertension and is the important determinant for early and late outcomes after corrective operations.[2,4,7,8,19] Preliminary unifocalization, therefore, is necessary for successful corrective surgery.

Hemodynamically, as a consequence of arborization abnormalities, the regional imbalance of pulmonary blood flow arises to cause the regional imbalance of pulmonary arterial growth and of pulmonary vascular resistance. This circumstance presents two problems. First, high-resistance pulmonary hypertension segments coexist with low-resistance hypoplastic segments in the same lung.[20,21] If the regional imbalance of pulmonary vascular resistance is not normalized after the corrective operation, the increased pulmonary blood flow via the right ventricle is forced to concentrate in the low-resistance hypoplastic segments and causes postoperative pulmonary edema. Second, the growth of intrapulmonary arteries varies widely according to the degree of pulmonary blood flow, and the intraacinar arteries, as a whole, are considerably smaller than normal.[20,21] There exist many bronchopulmonary segments in which the blood flow should be increased to stimulate the growth of the hypoplastic intrapulmonary arteries, although stimulation of all the segments is unattainable via a single surgical intervention. Therefore early unifocalization is needed to avoid the regional imbalances of pulmonary vascular resistance and pulmonary arterial growth.

The concept of unifocalization was first advocated by Macartney and Haworth[11,22] and their colleagues to convert multifocal to unifocal pulmonary blood supply. Several unifocalization techniques have since been described as follows: (1) direct anastomosis between a central pulmonary artery and an MAPCA,[23] (2) anastomosis between a central pulmonary artery and an MAPCA by interposition of a prosthesis,[24,25] and (3) anastomosis between an isolated segment of the descending thoracic aorta that gives rise to MAPCAs and the right ventricle.[6,26] In these techniques, however, MAPCAs, including their connections, where progressive stenoses frequently occur, inevitably remain in the pathway from the right ventricle to the intrapulmonary arteries. These hilar stenoses, although they apparently protect the intrapulmonary arteries from pulmonary hypertensive obstructive changes preoperatively (Fig. 10),[13-16] prevent successful unifocalization. Therefore, as suggested by Macartney, Scott, and Deverall,[9] the most logical way to achieve successful unifocalization is to unify intrapulmonary arteries at the hilum even though it is technically difficult.

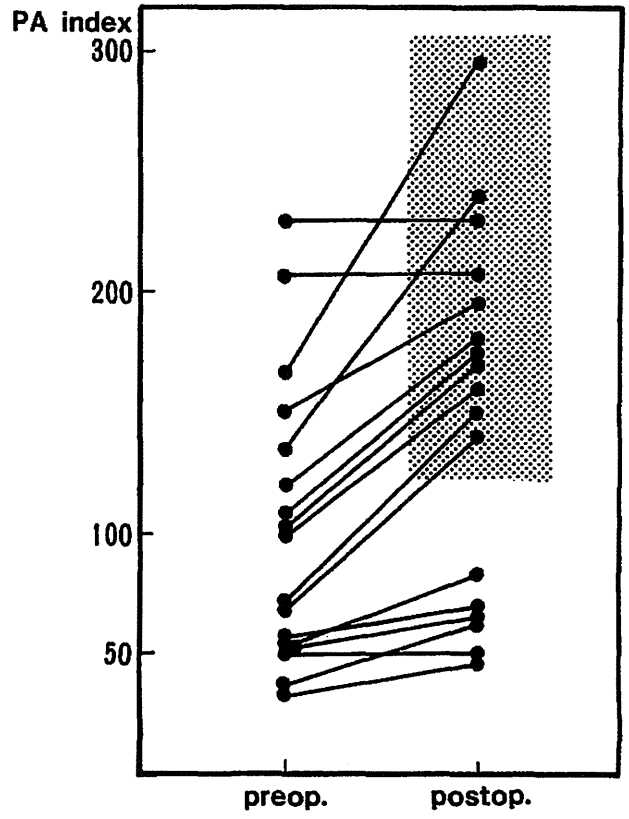

Fig. 8. Changes in PA indexes before and after first-stage repair, complete unifocalization. *Dotted area,* Desirable PA indexes for second-stage, right ventricular–pulmonary arterial conduit repair.

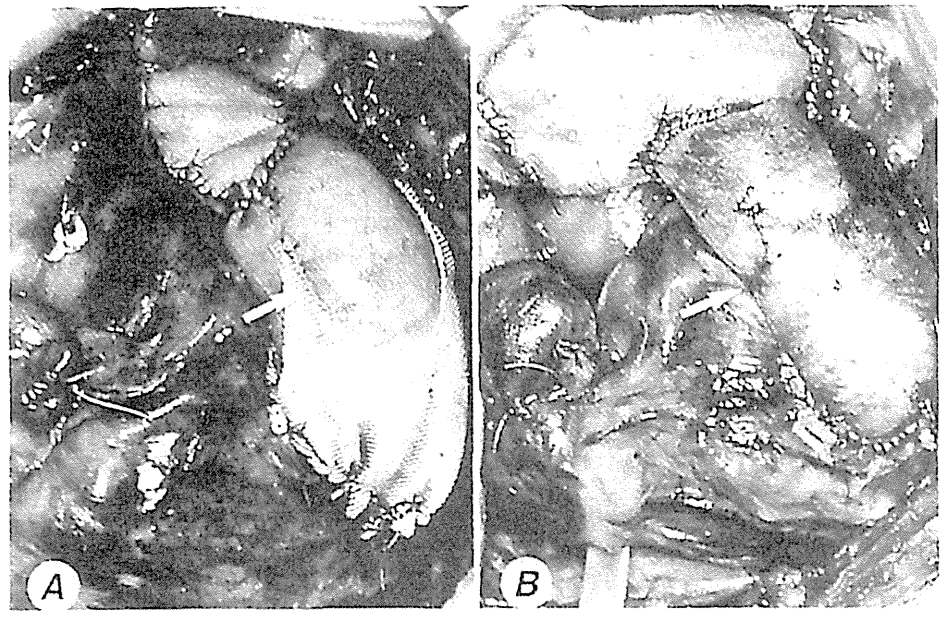

Fig. 9. Second-stage, right ventricular–pulmonary arterial conduit repair. Intraoperative photographs of two patients. Position of tailored, trileaflet pulmonary valve is indicated by the *white arrow*. A, The transverse portion of the conduit was placed posterior to the ascending aorta (patient 30). B, The transverse portion of the conduit was brought up cephalad over the aortic arch (patient 31).

In our complete unifocalization, we have aimed to restore the continuity between intrapulmonary arteries completely at the hilum or even inside the lung.[10] In this disease, hilar pulmonary arteries are relatively large compared with the other parts of the pulmonary arteries, so that we can perform satisfactory anastomoses between them. However, as a method for restoring the continuity between hilar pulmonary arteries, direct anastomosis is not applicable for the following reasons: (1) The interrupted portion of hilar pulmonary arteries is generally long; (2) hilar pulmonary arteries are tightly buried in the lung tissue and surrounded by pulmonary veins and the bronchial tree, so that they are difficult to mobilize; (3) compression of the pulmonary veins or the bronchial tree should be avoided in restoring the continuity of hilar arteries. Therefore we have developed an intrapulmonary bridge technique, which is widely applicable for the various forms of arborization abnormalities and peripheral pulmonary stenoses.

In tetralogy of Fallot, the size of central pulmonary arteries is generally considered to be correlated with the growth of intrapulmonary arteries; thus, it is an important determinant of operability and outcome. In pulmonary atresia and ventricular septal defect with pulmonary blood supply from MAPCAs, however, the size of central pulmonary arteries does not exert such an important role for the following reasons. As an anatomic feature, MAPCAs connect directly with hilar arteries at the hilum, skipping over central pulmonary arteries. They supply pulmonary blood flow mainly to the ipsilateral lung and occasionally contribute negligibly to the blood flow of the contralateral lung. Thus the blood flow of intrapulmonary arteries considerably exceeds that of the central pulmonary arteries. In Fig. 2, *B*, for instance, the blood flow in the right central pulmonary artery was only 50 ml/min, whereas the combined intrapulmonary arterial flow of the right lung was 400 ml/min when measured intraoperatively by electromagnetic flowmeter. Therefore we believe that the size of central pulmonary arteries is not always correlated to the growth of intrapulmonary arteries and, therefore, is not a suitable selection criterion for corrective operations.

On the contrary, the size of hilar pulmonary arteries exactly reflected the size of intrapulmonary arteries in our series. As suggested by Sotomora and Edwards,[27] independent of the presence or absence of central pulmonary arteries, there are always patent—although frequently interrupted—hilar pulmonary arteries, which can receive a conduit from the right ventricle. We think the efficacy of corrective operations should be determined by the growth of hilar pulmonary arteries. However, hypoplastic or absent central pulmonary arteries should be reconstructed to act satisfactorily as a conduit to receive the whole blood flow from the right ventricle during the second-stage repair. We believe that a PA index greater than 120 is necessary for this purpose. When central pulmo-

nary arteries are moderately hypoplastic, sufficient growth of the hypoplastic central pulmonary arteries can be stimulated by effective modified Blalock-Taussig shunts. Even if the shunts do not stimulate sufficient growth, extended patch angioplasty of the central pulmonary arteries can be successfully applied during the second-stage repair. When central pulmonary arteries are severely hypoplastic, however, they remain so hypoplastic even after satisfactory modified Blalock-Taussig shunts that additional extended patch angioplasty to the hilum cannot be applied effectively. Therefore the creation of new central pulmonary arteries is a more reliable method to obtain sufficient central pulmonary arteries for second-stage repair. At present, we think patients with a PA index less than 60 are candidates for this method.

At second-stage repair, a valved conduit is generally placed from the right ventricle to the central pulmonary arteries. We recommend the use of a valve-containing conduit in these patients on the supposition that postoperative right ventricular hypertension may occur. This tailored valve has been proven by echocardiography to function effectively; it is able to sustain high pulmonary arterial pressure and prevent pulmonary regurgitation during the early postoperative period when the right ventricle most likely has to work against pulmonary hypertension. In addition, by fitting itself within the retrosternal space, the pericardial conduit has the advantage of avoiding retrosternal compression, which may be a cause of right ventricular hypertension.

As stated earlier, the growth of central pulmonary arteries and arborization abnormalities are no longer the major determinants of operability and outcome in our staged operation. Instead, the final determinant is the growth of intrapulmonary arteries, which is well indicated by the growth of the hilar arteries; however, the minimum acceptable size of hilar pulmonary arteries for successful corrective surgery remains to be determined. Although complete unifocalization obviously stimulates the growth of hypoplastic hilar arteries in many cases, we cannot expect satisfactory growth of the intrapulmonary arteries when hilar arteries are extremely hypoplastic. Under this circumstance, the establishment of right ventricular–pulmonary arterial continuity in the neonate is possibly the sole way to stimulate the growth of intrapulmonary arteries sufficiently.

Another critical situation with regard to the growth of intrapulmonary arteries concerns pulmonary hypertensive obstructive disease. When most of the pulmonary vascular bed is supplied from a single large MAPCA in older patients, the presence of a broad, severe pulmonary hypertensive obstructive change, which critically interferes with successful surgical interventions, is important.

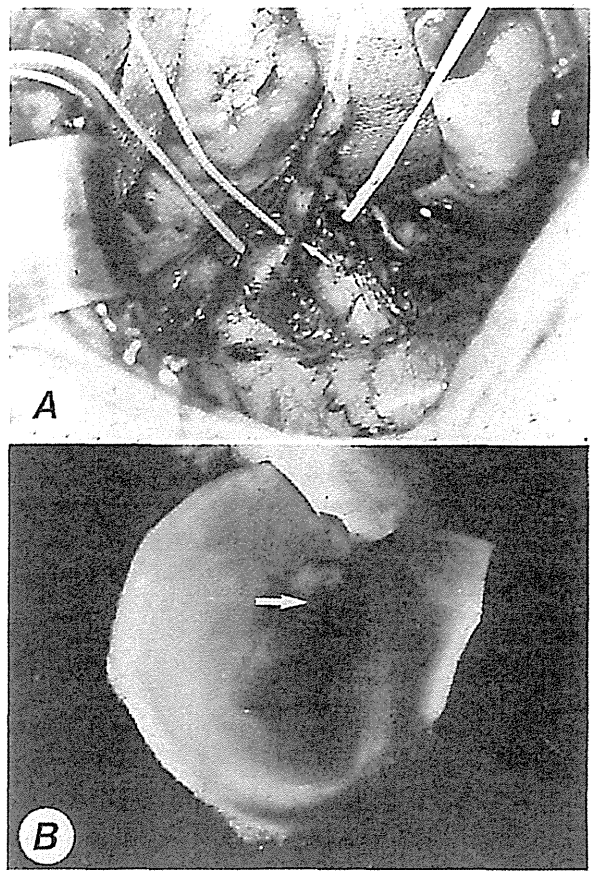

Fig. 10. Hilar stenosis of MAPCA. A, Intraoperative photograph showing hilar stenosis *(white arrow)*, located in the MAPCA just before connecting to the intrapulmonary artery. The intrapulmonary artery (the part distal to the stenosis) has been already exposed by dissection into the interlobar fissure. B, Inside view of the same hilar stenosis *(white arrow)* from the aortic side. The degree of the stenosis is more severe than its external appearance indicated. Note the intimal proliferation around the internal orifice, which probably explains the progressive obstruction of the MAPCA.

In the other patients, we unifocalize every isolated segment irrespective of the degree of pulmonary hypertension because we cannot evaluate the degree of pulmonary hypertension in each individual segment preoperatively. Moreover, after complete unifocalization, pulmonary arterial pressures of each segment are equalized to the value that is determined by the equivalent parallel resistance of all segments under modulated pulmonary flow. The postoperative equalization of their pulmonary arterial pressures leads to the decrease of pulmonary arterial pressure in most segments including those perfused at systemic pressure preoperatively; thus it prevents the further progression of pulmonary hypertensive obstructive change. We think it is useful to unifocalize every segment

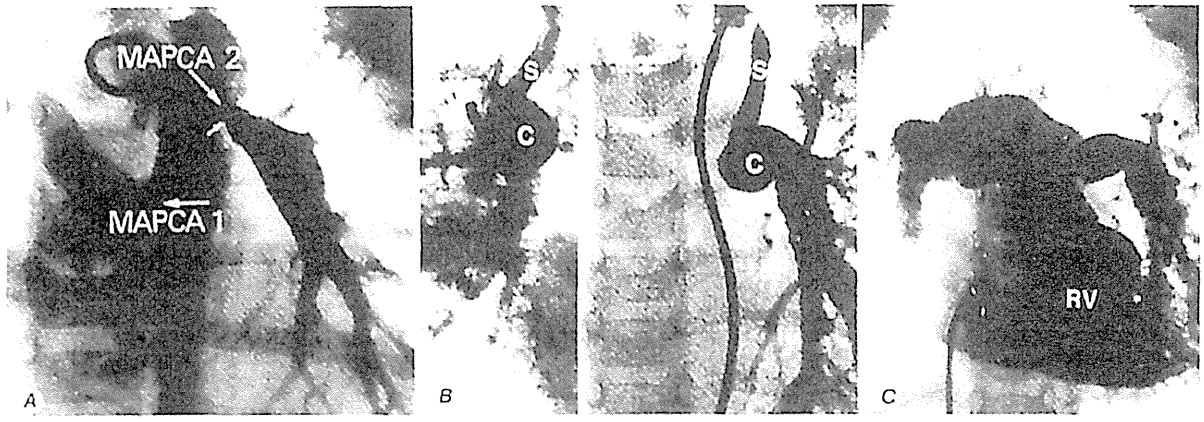

Fig. 11. A case presentation of the staged operation in a 1-year-old patient with absence of the central pulmonary artery (patient 30). A, Preoperative pulmonary angiogram by selective MAPCA injections. *Large arrow*, Origins of MAPCAs; *small arrow*, hilar stenosis of MAPCA. B, Pulmonary angiogram by injections into modified Blalock-Taussig shunts after the first-stage repair. Note the created new central pulmonary arteries. *c*, Xenomedica conduit as the new central pulmonary artery. *s*, The modified Blalock-Taussig shunt. C, Right ventriculogram after second-stage repair. *RV*, Right ventricle.

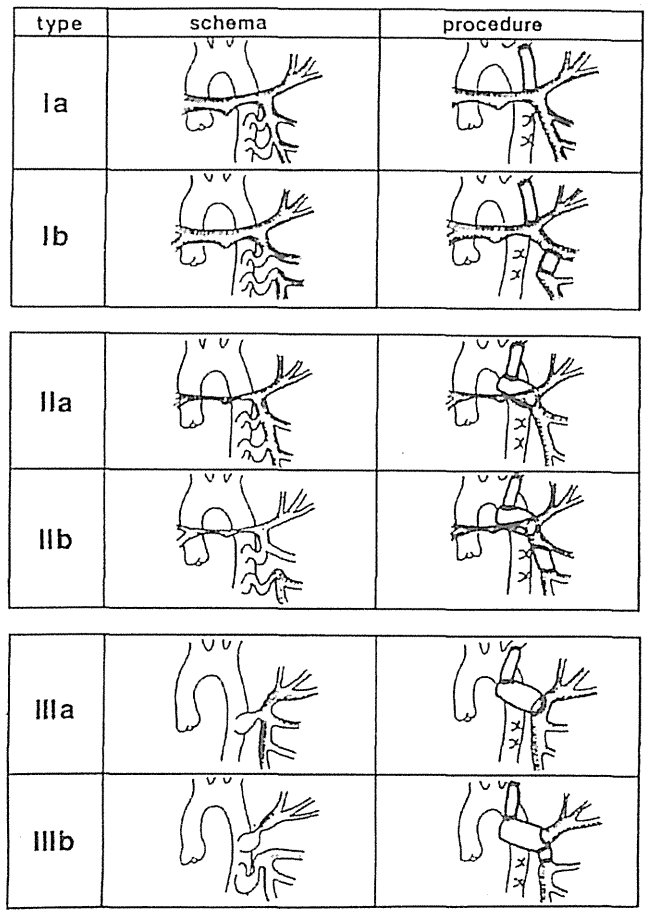

Fig. 12. Schematic representation of the surgical classifications and their surgical procedures is complete unifocalization.

irrespective of the degree of pulmonary hypertension. We should make every effort to use as many bronchopulmonary segments as possible to decrease the degree of postoperative right ventricular hypertension.

The age at first-stage repair is a certain risk factor for the growth of intrapulmonary arteries. The severity of hypoplasia and hypertensive obstructive changes of intrapulmonary arteries certainly increases with age in many patients. As previously mentioned, the development of intrapulmonary arteries extends over a wide spectrum, from severe hypoplasia to severe hypertensive obstructive change, by the patient or even by the lung, depending on the pulmonary blood flow. Although this spectrum is partly inherent in its origin, we believe it mainly extends toward both extremities with age and gradually forms the severe patterns of hypoplasia and hypertension. The progressive obstruction of MAPCA (Fig. 10) plays an important role in this alteration, although it exerts both advantageous and disadvantageous influences on the growth of intrapulmonary arteries according to the relationship between the primary blood flow and the degree of progression. The favorable postoperative RVP/LVP ratios in patient 30 (Fig. 11) and patient 31 support our theory. The schedule for the staged operation should be determined according to the individual anatomy and the complexity of the surgical procedure; however, we consider it beneficial to start a first-stage repair before the child is 1 or 2 years old and complete a second-stage repair before age 3 or 4. If obstructive changes are severe, an appropriate period of follow-up should be established

after the first-stage repair, and delayed closure or perforated patch closure of a ventricular septal defect should also be considered in the second-stage repair.

Because of various and multiple anomalies, no identical pulmonary arterial vasculatures could exist. The first step in the surgical treatment of these patients is to understand the complicated anatomy of the pulmonary arterial vasculature in each patient. The second step is to determine the most desirable surgical approach. The essential abnormalities of the pulmonary blood supply exist in the hilum. Incomplete unifocalization by an extrapulmonic operation does not prepare the patients for a successful corrective operation. In our staged operation, we can easily classify the complicated features of the anomalies, and we can perform the complete unifocalization by an intrapulmonic operation (Fig. 12). By our complete unifocalization technique, with the combination of an intrapulmonary bridge and the creation of new central pulmonary arteries, we can successfully correct the majority of cases of pulmonary atresia and ventricular septal defect with the pulmonary blood supply from MAPCAs. If a regional imbalance of pulmonary blood flow can be prevented or corrected by complete unifocalization in infancy or early in childhood, an avenue to successful corrective surgery can be opened.

REFERENCES

1. Pacifico AD, Kirklin JW, Bargeron LM Jr, Soto B. Surgical treatment of common arterial trunk with pseudotruncus arteriosus. Circulation 1974;49,50(Pt 2):II20-26.
2. Olin CL, Ritter DG, McGoon DC, Wallace RB, Danielson GK. Pulmonary atresia: surgical considerations and results in 103 patients undergoing definitive repair. Circulation 1976;54(Pt 2):III35-40.
3. Gill CC, Moodie DS, McGoon DC. Staged surgical management of pulmonary atresia with diminutive pulmonary arteries. J THORAC CARDIOVASC SURG 1977;73:436-42.
4. Alfieri O, Blackstone EH, Kirklin JW, Pacifico AD, Bargeron LM Jr. Surgical treatment of tetralogy of Fallot with pulmonary atresia. J THORAC CARDIOVASC SURG 1978;76:321-35.
5. Piehler JM, Danielson GK, McGoon DC, Wallace RB, Fulton RE, Mair DD. Management of pulmonary atresia with ventricular septal defect and hypoplastic pulmonary arteries by right ventricular outflow construction. J THORAC CARDIOVASC SURG 1980;80:552-67.
6. Shimazaki Y, Kawashima Y, Hirose H, et al. Operative results in patients with pseudotruncus arteriosus. Ann Thorac Surg 1983;35:294-9.
7. Millikan JS, Puga FJ, Danielson GK, Schaff HV, Julsrud PR, Mair DD. Staged surgical repair of pulmonary atresia, ventricular septal defect, and hypoplastic, confluent pulmonary arteries. J THORAC CARDIOVASC SURG 1986;91:818-25.
8. Kirklin JW, Bargeron LM Jr, Pacifico AD, Soto B, Kirklin JK. Management of the tetralogy of Fallot with large aortopulmonary collateral arteries. In: Godman MJ, ed. Pediatric cardiology. Vol 4. Edinburgh: Churchill LIvingstone, 1981:483-91.
9. Macartney FJ, Scott O, Deverall PB. Haemodynamic and anatomical characteristics of pulmonary blood supply in pulmonary atresia with ventricular septal defect—including a case of persistent fifth aortic arch. Br Heart J 1974;36:1049-60.
10. Sawatari K, Imai Y, Kurosawa H, et al. Surgery for pulmonary atresia and ventricular septal defect with major aorto-pulmonary collateral arteries: remodelling and unifocalization of pulmonary arteries. J Cardiovasc Surg 1987;suppl 10:28.
11. Macartney FJ, Haworth SG. The pulmonary blood supply in pulmonary atresia with ventricular septal defect. In: Godman MJ, Marquis RM, eds. Pediatric cardiology. Vol 2. Edinburgh: Churchill Livingstone, 1979;314-38.
12. Cudkowicz L, Armstrong JB. Injection of the bronchial circulation in a case of transposition. Br Heart J 1952;14:374-8.
13. Jefferson K, Rees S, Somerville J. Systemic arterial supply to the lungs in pulmonary atresia and its relation to pulmonary artery development. Br Heart J 1972;34:418-27.
14. Macartney F, Deverall P, Scott O. Haemodynamic characteristics of systemic arterial blood supply to the lungs. Br Heart J 1973;35:28-37.
15. McGoon MD, Fulton RE, Davis GD, Ritter DG, Neill CA, White RI Jr. Systemic collateral and pulmonary artery stenosis in patients with congenital pulmonary valve atresia and ventricular septal defect. Circulation 1977;56:473-9.
16. Haworth SG, Macartney FJ. Growth and development of pulmonary circulation in pulmonary atresia with ventricular defect and major aortopulmonary collateral arteries. Br Heart J 1980;44:14-24.
17. Nakata S, Imai Y, Takanashi Y, et al. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. J THORAC CARDIOVASC SURG 1984;88:610-9.
18. Fäller K, Haworth SG, Taylor JFN, Macartney FJ. Duplicate sources of pulmonary blood supply in pulmonary atresia with ventricular septal defect. Br Heart J 1981;46:263-8.
19. Marcelletti C, McGoon DC. Pulmonary infarction following ligation of terminally shunted pulmonary artery. J THORAC CARDIOVASC SURG 1976;71:746-9.
20. Thiene G, Frescura C, Bini RM, Valente M, Gallucci V. Histology of pulmonary arterial supply in pulmonary atresia with ventricular septal defect. Circulation 1979;60:1066-74.
21. Rabinovitch M, Herrera-deLeon V, Castaneda AR, Reid L. Growth and development of the pulmonary vascular bed in patients with tetralogy of Fallot with or without pulmonary atresia. Circulation 1981;64:1234-49.

22. Haworth SG, Rees PG, Taylor JFN, Macartney FJ, de Leval M, Stark J. Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: effect of systemic pulmonary anastomosis. Br Heart J 1981;45:133-41.
23. Pacifico AD, Allen RH, Colvin EV. Direct reconstruction of pulmonary artery arborization anomaly and intracardiac repair of pulmonary atresia with ventricular septal defect. Am J Cardiol 1985;55:1647-9.
24. Benson LN, Laks H, Lois J, Dajee H, Child J, Perloff JK. Surgical correction of pulmonary atresia and ventricular septal defect with large systemic-pulmonary collaterals. Ann Thorac Surg 1984;38:522-5.
25. Barbero-Marcial M, Rizzo A, Lopes AAB, Bittencourt D, Junior JOA, Jatene AD. Correction of pulmonary atresia with ventricular septal defect in the absence of the pulmonary trunk and the central pulmonary arteries (so-called truncus type IV). J THORAC CARDIOVASC SURG 1987;94:911-4.
26. Murphy DA, Sridhara KS, Nanton MA, Roy DL, Belcourt CL, Gills A. Surgical correction of pulmonary atresia with multiple large systemic-pulmonary collaterals. Ann Thorac Surg 1979;27:460-4.
27. Sotomora RF, Edwards JE. Anatomic identification of so-called absent pulmonary artery. Circulation 1978;57:624-33.

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Pulmonary atresia with ventricular septal defect, extremely hypoplastic pulmonary arteries, major aorto–pulmonary collaterals ☆

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Abstract

Objective: Among 63 patients with pulmonary atresia and ventricular septal defect (VSD), 10 patients with extreme hypoplasia of the pulmonary arteries (PA) (mean Nakata index 20.6 mm^2/m^2), but with confluent arteries and a diminutive main PA, and major aorto–pulmonary collaterals (MAPCAS), have been submitted to a 'rehabilitation' of the PA with several stages: (i) connection between RV and PAs, (ii) interventional catheterizations, (iii) complete correction with or without unifocalisation. We report here the results of this approach. **Methods**: The RV–PA connection was direct (nine cases) or with an homograft conduit (one case), done under normothermic cardiopulmonary by-pass in patients aged 4.9 months (range 0.1–18 months). Subsequently, six underwent interventional catheterizations (dilations and stents in the PA, MAPCAS occlusion by coils). Complete correction was done in seven patients (mean age 30 months, range 8–49). One patient is awaiting correction. **Results**: One patient died after the first stage. All patients having had the third stage had a satisfactory development of the PA, had a complete closure of the VSD and a satisfactory reconstruction of the PA bifurcation. There was one death of severe pulmonary infection 6 months after repair. All other patients have been followed by catheterization and/or echocardiograms. With a follow-up of 83 ± 65 months, all patients are improved, 50% have no cardiac medications, none has residual shunt, RV/LV pressure ratio is 0.6 (range 0.3–1). **Conclusions**: The strategy of 'rehabilitation' of PA allowing: (i) antegrade flow in the PA, (ii) interventional catheterizations, (iii) growth of the PA with possible angiogenesis, (iv) complete correction, is a logical approach to be undertaken in the young patient and is a valid alternative to strategies relying more on MAPCAS for pulmonary vascular supply. The therapeutic sequences depend upon the individual anatomy.

Keywords: Pulmonary atresia; Rehabilitation; Pulmonary arteries; Correction

1. Introduction

The surgical management of pulmonary atresia with ventricular septal defect (VSD), extreme hypoplasia of the pulmonary arteries (PA), major aorto–pulmonary collaterals (MAPCAS) represents a major challenge. Two main basic concepts have been used in its management. The first is to rely mainly on the MAPCAS, unifocalizing successively the MAPCAS on one side, then the other, including the hypoplastic pulmonary arteries in the reconstruction, and then in a third stage to perform a total correction with closure of the VSD and establishment of a conduit between the right ventricle (RV) and the two unifocalization confluences [1–5]. An alternative of this classical approach in several stages, used in general in older children, has been recently proposed, consisting in a total unifocalization and complete correction in one procedure, done even early in life [6–8].

The second is to rely more on the real pulmonary arteries, and undergo a program of 'rehabilitation' of these hypoplastic pulmonary arteries. This program of rehabilitation is done in several stages. The first stage consists in establishing a direct continuity either between the ascending aorta [9,10] or the RV [11] and the diminutive PA. The following stages are catheterizations studies and interventional catheterizations (with stenosis dilation easier in the RV–PA

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connection, PA stents and coil occlusion of MAPCAS). The final surgical stage is a complete correction with closure of VSD and RV–PA conduit construction. Subsequent post-operative catheterizations for evaluation and/or interventions are then performed.

We report here 10 cases of PA with VSD infancy in extreme hypoplasia of the pulmonary arteries, and MAPCAS, who underwent a program of rehabilitation of the PA, aiming at the complete correction after PA growth and interventional catheterizations on the PA.

2. Material and methods

2.1. Patients

Among a population of 63 pulmonary atresia with VSD (excluding the tetralogy of Fallot with valvular atresia), seen between 1985 and 2000, 10 patients had an extreme hypoplasia of the pulmonary arteries defined by a Nakata index below 90 mm^2/m^2 [11,12], the normal being 330 and an index around 150 allowing the possibility of a correction. However, all had confluence of the diminutive arteries with a diminutive main PA joining the area of the atretic infundibulum of the right ventricle.

They were six boys and four girls with a mean age of 110 days range 3/467 days, 6/10 displaying the 22Q11 deletion, admitted in the hospital for severe hypoxia, two in emergency. Two patients had been previously seen in other centers and considered as unsuitable for any surgical treatment. Excluded from this study have been the patients without pulmonary arteries and having only collaterals, as evidenced by catheterization and angiograms.

All patients were studied pre-operatively by echocardiogram, catheterization and angiogram performed in the descending aorta, and in the pulmonary veins.

In all patients the distribution of blood vessels to the lungs showed:

1. Various MAPCAS originating from the descending aorta or the main branches of the proximal aorta.
2. Extremely diminutive central pulmonary arteries with a confluence and a diminutive trunk of main PA, realizing the 'sea-gull' aspect (Figs. 1 and 2a) were seen by retrograde filling from the collaterals or from pulmonary veins wedge angiogram. The size of the small pulmonary arteries in mm were evaluated by comparison with the catheter diameter and expressed by the Nakata index. They were measured on the right and left pulmonary artery, immediately proximal to their first branch point. In all cases, the 'main' PA branches size, was between 1 and 2.7 mm (mean 1.45 mm) and the Nakata index from 3.5 to 58, mean 20.6 $mm2/m^2$.
3. The distribution of actual pulmonary branches to the lungs, seen in the catheterization studies after RV–PA correction, although difficult to appreciate exactly showed a mean of 16 segments (range 10–20, normal 20), in the patients.

2.2. Surgical technique for RV–PA continuity establishment (stage 1)

All patients were operated upon through median sternotomy with the use of normothermic cardiopulmonary by-pass (CPB) with a beating heart. The analysis of external anatomy showed in all cases a diminutive main PA trunk, connected anatomically to the atretic infundibulum and in no case a major coronary artery crossing the infundibulum that may preclude the direct RV–PA connection. After controlling RPA and LPA with gentle occlusion by rubber loops, the main PA was incised longitudinally, an augmentation small oval patch of autologous pericardium prepared with glutaraldehyde was sutured to the edges of arteriotomy.

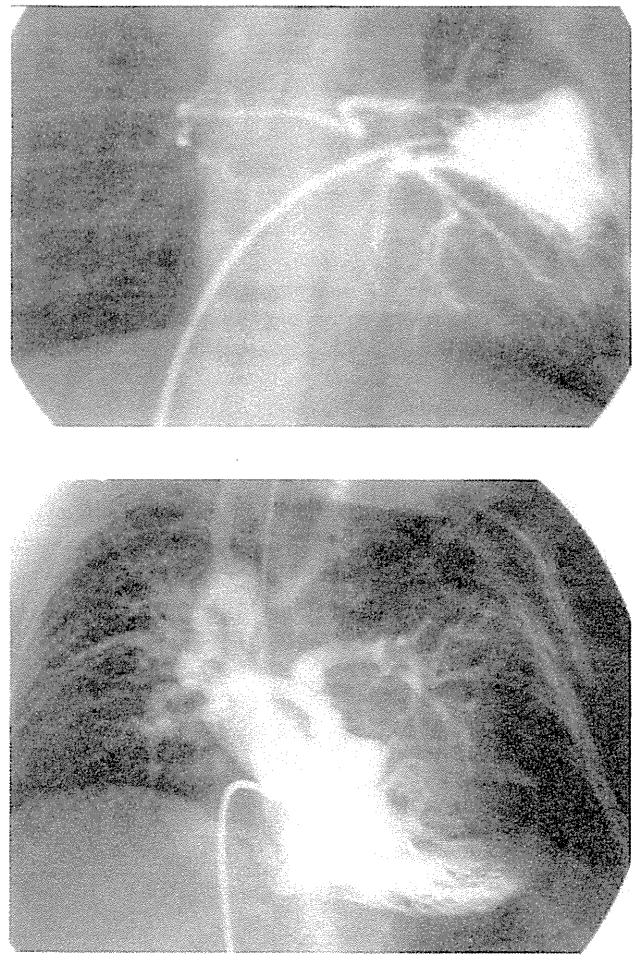

Fig. 1. (Top) extremely diminutive central pulmonary arteries (Nakata: 15) showing the 'sea-gull' aspect, filled by a retrograde angiogram in a pulmonary vein. A MAPCA is also opacified retrogradely. (Bottom) three months after RV–PA connection by patch done at 4 months of age, there is a nice development of the PAs, with normal pressures and satisfactory distribution. The MAPCAS (numerous in this case) are barely seen and they showed a considerable decrease. The patient is scheduled for complete repair.

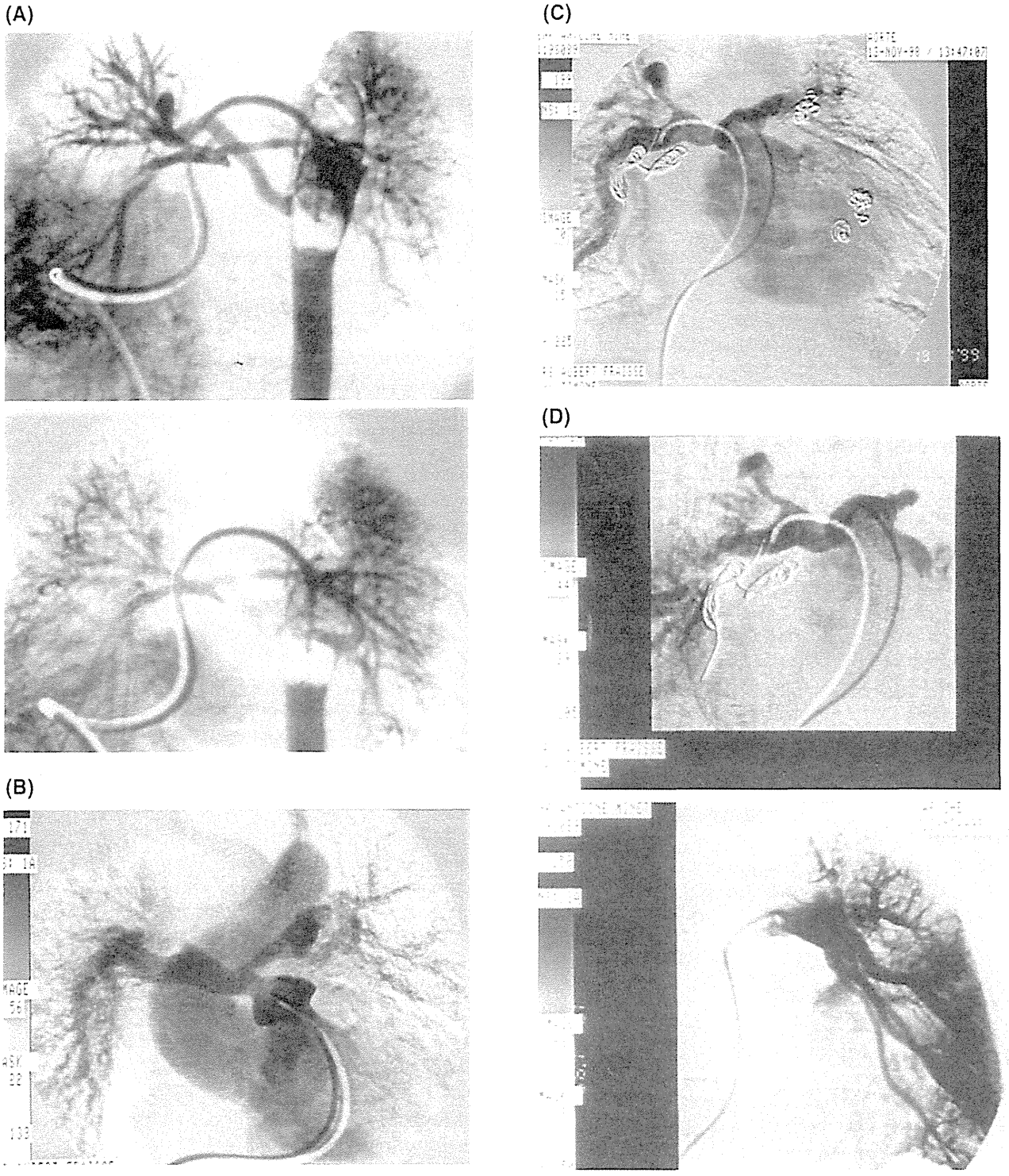

Fig. 2. (A) Aortic angiogram blocked by balloon showing (top) one large MAPCA filling retrogradely a diminutive central pulmonary artery (Nakata: 29), with satisfactory branching (bottom). (B) Angiogram done 8 months after RV–PA patch connection (done at 7 months of age) showing nice development of the PAs. (C) Angiogram done after coil occlusion of several MAPCAS, during the same catheterization showing adequate development of pulmonary arteries on the right side. There is however a stenosis of the right mediastinal artery. (D) Same angiogram. One can appreciate the distribution of PA branching on the left side.

The incision was than extended to the RV epicardium, the patch was sewn to the edges of this incision and finally the RV infundibulum was opened, and the patch insertion finished, making sure the opening in the RV was sufficient by resection of hypertrophied muscle and spreading this opening with scissors or a right angle. In order to avoid

air embolism, only superficial succion was applied in the RV, and a vent was used in the ascending aorta with partial aortic cross-clamping, thus making a bubble-trap.

In 9/10 cases this patch technique was used. In one patient (the first in the series), a small homograft (12 mm diameter) was used, since the patient had previously undergone an unsuccessful anastomosis between the ascending aorta and the small main PA through a 4 mm PTFE small conduit.

The mean age of the patients at this first step was 4.9 months (range 0.1–18 months).

2.3. Subsequent catheterization (stage 2)

After a minimum delay of 3 months, catheterization studies allowed by the RV–PA continuity were done with several goals: (a) as indicated previously, identification of pulmonary artery-branching distribution, (b) identification and dilation of discrete or long segment stenosis, (c) occasional placement of stents in the PA (d) coil occlusion of collaterals if the O_2 saturation allowed it.

2.4. Total correction (stage 3)

It consisted in redo-sternotomy, establishment of hypothermic CPB and complete repair-with:

1. Reconstruction of the pulmonary bifurcation: in all cases, it was done after aortic transection, and enlargement with pulmonary homograft tissue from on hilum to the other, the distal anastomosis being done if necessary for better vizualisation under a short period of hypothermic circulatory arrest.
2. Patch closure of VSD and direct closure of ASD.
3. Establishment of RV–PA continuity with a pulmonary cryopreserved homograft conduit.
4. At the completion of CPB, pressures were recorded, and if the RV–LV pressure ratio was below 1, the correction was considered satisfactory, and the VSD left closed.

3. Results

3.1. RV–PA connection

Nine patients survived after this procedure. The only death was a neonate operated upon at the age of 3 days, admitted in emergency. The angiograms, in addition to hypoplastic pulmonary arteries, showed poor collaterals. The pre-operative O_2 saturation was 71%. Despite an uneventful procedure, an initially hemodynamics satisfactory situation, the flow appearing satisfactory through the patch, he remained severely hypoxic and died on the table.

One other patient exhibited an increased blood flow through the lungs in the post-operative period. He had a long common trunk of collaterals for both lungs that underwent reoperation, was detached from the aorta, and finally simply disconnected and closed, the saturation appearing to be satisfactory with the only antegrade flow from the RV–PA connection. This procedure led to a long post-operative period with 43 days of ICU.

One other patient, also operated in emergency with a diagnosis made at 3 months, probably because acute obstruction of a large MAPCA, with extremely low O_2 saturation (21%), asymptomatic before this event, had a difficult post-operative course, with delayed sternal closure, and spent 27 days in the ICU.

Another patient with associated sub-glottic stenosis, aged 1 month, underwent a Cotton procedure (cervical tracheal plasty with cartilage) 13 days before the RV–PA connection and has a prolonged ICU stay of 19 days.

All the six other patients had a simple post-operative course, with a mean ICU stay of 3.5 days. Mean O_2 saturation rose from 73% pre-operative to 89% post-operative.

3.2. Post-operative catheterizations

After a minimal delay of 3 months, eight patients underwent catheterization studies and had a total of 27 procedures, one to six per-patient.

Fourteen of these procedures were interventionnal catheterizations. During these procedures, 23 pulmonary arteries were dilated, 82% of these were successful, defined by Lock [11] (diameter increase over 75%, gradient decrease over 50%, increase in distal flow by scintigraphy).

Six stents were inserted in the PA [5] or in the RV–PA homograft [1], one year before conduit replacement.

Eight MAPCAS were coil occluded.

The patients were considered suitable for corrective surgery after a mean delay of 19 months (range 4–48 months).

In five patients, the distribution of pulmonary arteries was complete. In two patients unifocalization with MAPCAS was considered necessary at the time of correction due to pulmonary distribution to only 10 segments, two other lobes being vascularized only by MAPCAS.

One patient, having undergone the RV–PA connection, catheterized 4 months post-operatively, exhibits a nice growth of the PA with low distal pressures, and remarkable involution of the large collaterals, is waiting total correction within the next month (Fig. 1). One more patient operated upon 5 months ago is waiting for the first catheterization study. Both have normal pulmonary artery arborization.

3.3. Complete correction

It was done in seven patients. Their mean age at surgery was 28.9 months (range 6–51 months). The complete correction was done after a mean delay between stage 1 and 3 of 19 months (range 4–48 months).

The decision for complete correction depended upon several factors:

1. The size of the pulmonary arteries at the last catheterization evaluation: if the Nakata index was over 150, the

patient was considered operable. If the branching was considered satisfactory but there was stenosis of the mains PAs, the perspective of patching widely the bifurcation made also this decision positive. In fact, it was not always a 'mathematical' decision but a general consensus based upon previous experience.

2. Once the decision was taken, it was tried not to delay the correction, since leaving the pulmonary circulation under systemic pressure might have had a deleterious effect on the pulmonary vascular resistances.

Clinically, all patients were under antifailure treatment with mean cutaneous O_2 saturation of 89%.

Mean CPB duration was 238 mn (range 192–316 mn), aortic-cross-clamping duration was 110 mn (range 71–130 mn). In all cases the VSD could be left entirely closed at the completion of CPB, due to the acceptable RV–LV pressure ratio (mean 0.5, range 0.3–0.8). The reconstructed RV–PA continuity was established by a cryopreserved pulmonary homograft from 17 to 20 mm of diameter. One of the two patients with only 10 segments vascularized by two pulmonary arteries underwent a unifocalization procedure, a collateral artery vascularizing the right upper lobe was disconnected from the aorta through the midline and implanted on a lateral incision of the right pulmonary artery. The other patient underwent correction without unifocalization.

All patients survived the procedure. In the post-operative period, three patients had a delayed sternal closure, and the mean hospital duration was 19 days (range 11–32 days).

The mean follow-up is 45.4 month (range 10–84 months). There was one late death at 6 months of pulmonary infection in an otherwise N.Y.I-LA. class II patient, still under antifailure therapy. All other patients are in class I or II, two patients taking cardiac medication. At the regular echocardiographic controls, there is no residual VSD. One patient underwent a reoperation 6 years after correction: the RV–PA homograft conduit was changed to a 23 mm pulmonary cryopreserved homograft. Surgery was uneventful and the post-operative RV–LV pressure ratio was 0.3.

Four patients have undergone a catheterization from one to 18 months post-operatively after the complete correction. The RV/LV pressure ratio is from 0.32 to 1 the latter being before replacement of the homograft conduit, mean 0.60. One of them has undergone a left branch stenosis dilatation. The mean distal pressure in the pulmonary arteries is from 10 to 27 mmHg (mean 18 mmHg). None has a residual shunt. In most patients adequate size of the pulmonary arteries has been reached (Fig. 2).

4. Discussion

Pulmonary atresia with VSD, MAPCAS and extreme hypoplasia of the pulmonary arteries, filling retrogradely through the MAPCAS blood flow is a therapeutic challenge, with an extremely poor prognosis without surgery [13]. It differs completely from the more frequent setting of pulmonary atresia with sizeable PA, in which the strategy was totally different in the other 53 patients of our experience: modified Blalock–Taussig shunts in the new-born or infant period when necessary and subsequent complete correction with a valve conduit or with direct RV–PA patch connection. The strategy and timing depending upon pulmonary pressures and resistances, importance and distribution of the collaterals when present, confluence or not of the pulmonary arteries, the decision making being based on individual situation analysis.

Several attitudes have been reported:

1. A conservative, symptomatic and medical attitude, historically the oldest method tending to delay maximally surgery. If the patient is symptomatic, with hypoxemia and cyanosis, systemopulmonary shunts have been advocated [14,15]. On the opposite, if blood flow is excessive through the MAPCAS, anti-failure therapy is given, and when pulmonary vasculary disease is present, cardio-pulmonary transplantation is considered.
2. A more aggressive attitude in patients with large collaterals has been advocated. Successive bilateral unifocalisation of MAPCAS with conduits and finally complete correction in a third step with VSD closing and RV to unifocalisation conduits connection has been successfully done, in older children [1–3] This strategy is logical in the complete absence of pulmonary arteries but practically abandons the true pulmonary arteries, in which no attempt at specific enlargement is made. In addition, no consensus on several issues has been reached in the various multi-stage strategies of unifocalization: treatment of the real hypoplastic PA, strategies of unifocalization, criteria for repair. Late functional results and future of pulmonary vascular resistances are lacking with this approach.
3. The overall results of staged approach with unifocalization have been variable [5,14,16] In all series, complete repairs were accomplished in 12 to 60% of patients. When a delayed staged approach is undertaken it has been estimated that only 20–30% of infants with this anomaly will end up with acceptable hemodynamics [5,11,14].
4. More recently, was reported by Hanley's group [6], followed by others [7,8], an early complete correction by median sternotomy or bilateral 'clam-shell' thoracotomy [17] branching together all MAPCAS and hypoplastic pulmonary arteries. In most cases, complete closure of the VSD was achieved and early results were satisfactory. However the pulmonary blood flow is mainly a flow through the MAPCAS. This procedure has been done with all kinds of true pulmonary arteries and there is some uncertainty about the future development of pulmonary vascular obstructive disease [11,18] It has been suggested that the earlier in infancy the operation

is done the better the long term may be, the shear stress in MAPCAS and pulmonary vascular obstructive disease being possibly avoided. However, in a recent publication of this approach [19], the angiogams obtained post-operatively revealed enormous and tortuous pulmonary vessels, some parts looking even aneurysmal so that the future of this vascular supply appears to be very uncertain.

5. A totally different concept has been advocated, using the pulmonary arteries. The concept is to try to promote flow in the true hypoplastic pulmonary arteries the obtain growth of these pulmonary arteries. It had been reported long ago by the Mayo Clinic group but in larger arteries and older patients [1]. The promotion of flow to the pulmonary arteries with a systemic to pulmonary artery shunt (classical or modified Blalock–Taussig shunt) has been particularly disappointing, leading to uneven growth, severe stenosis and severely compromised hypoplastic PA [10,15]. In addition, with PA diameter as in our group of patients, the systemopulmonary shunt appears impossible.

This is why Mee et al. [9,10] have promoted the direct anastomosis of the small main PA in the ascending aorta, or through a small prosthetic conduit. It has led, reportedly, in 28 patients (among 54 patients undergoing other strategies. 30 of them being older than 2 years) to complete repair. During the aorto–pulmonary anastomosis, MAPCAS were ligated or transplanted through additional lateral thoracotomies.

However in the study presented the true size of the hypoplastic PA is not determined and the result of correction is the sub-group of direct aorto–pulmonary anastomosis is not precise.

The approach we report here was reported by the Boston group in 1993 [11] and coined 'rehabilitation' of pulmonary arteries. The basic concept is that with the RV–PA connection, the flow is increased to the native pulmonary arteries, exactly as reported earlier by the Mayo Clinic group and others but in older children.

In our group of patients the age at this operation was even lower than in the Boston series (3.5 months versus 8.7 months).

Despite a very small size of main PA trunk, always present in the sea-gull aspect of PA, this was possible and we have used a beating heart procedure with normothermic cardiopulmonary by pass, rendering easy this RV–PA connection.

Subsequently, diagnostic and interventional catheterizations have been performed showing decrease in the size of MAPCAS, so that some did not need subsequent coil occlusion. Coil occlusion was performed when the MAPCAS were vascularizing the same territories as the true pulmonary arteries (communicating MAPCAS).

The indication of complete correction is taken on the analysis of the pulmonary artery size during echocardiography and catheterization when they are considered large enough.

The result of complete correction in this rehabilitation strategy has been evaluated by the Boston group [11] as favourable when three criteria were achieved (i) mean pulmonary artery pressure below 25 mmHg, (ii) RV/LV pressure ratio below 0.8, (iii) absence of significant residual shunt. These criteria were achieved in our patients that underwent the complete connection.

This rehabilitation approach relies on three findings [1,9–11]. (i) The increase of flow in the PA favours their growth. The angiogenesis of distal vessels is however still hypothetic; (ii) These favourable phenomena would be more important in the first months of life favouring earlier surgery; (iii) the MAPCAS are not a reliable source of pulmonary flow, being frequently tortuous and stenosed or on the opposite evolving towards obstructive vascular disease [18].

Despite theoretical advantages of this staged approach, several issues remain uncertain [11], as the precise quantification of pulmonary arteries, the optimum timing for outflow patch creation, pulmonary artery catheterization dilations, MAPCAS embolizations, the role of unifocalizations.

It remains that a strategy aiming at the development of these pulmonary arteries and a biventricular repair is feasible in most cases of pulmonary atresia, VSD, MAPCAS and severely hypoplastic pulmonary arteries and that this strategy should be undergone early in life, since long term future of true pulmonary arteries may possibly be better than flow through the collaterals.

5. Addendum

Since this paper was written, the two patients waiting for correction were operated after a catheterization study without the need for intervention except a major collateral coil embolization.

5.1. Patient 9

RV–PA connection was done 4 months of age with a very easy post-op case course and a hospital stay of 11 days. Post-operative cardiac catheterization was done at age 8 months. No intervention was done during the catheterization, O_2 saturation was 98%. The collaterals had spontaneously decreased (see Fig. 1).

Complete correction was done at age 9 months, VSD closure, reconstruction of the RV–PA connection with a mono cusp homograft patch.

Relatively easy post-op course, with delayed sternal closure (day 2) and a total hospital stay of 12 days.

Post-operative evaluation: Sao2 100 %, RV/LV pressure by echo 40 %, excellent clinical condition.

5.2. Patient 10

RV–PA connection at age 1 month. Very easy post-op course, hospital stay of 10 days. Post-operative cardiac catheterization at 5 months of age, O_2 saturation 84%, coil embolization of a main collateral.

Complete correction at age 8 months, with ligation of a collateral, reconstruction of PA bifurcation, VSD closure, RV–PA connection with a 20 mm diameter pulmonary homograft. Delayed sternal closure at day 2, easy post-op course, hospital stay 15 days. At follow up, O_2 saturation 100%, RV–LV pressures by echo 60% and excellent clinical condition.

References

[1] Millikian JS, Puga FJ, Danielson GK. Staged surgical repair of pulmonary atresia, ventricular septal defect, and hypoplastic, confluent pulmonary arteries. J Thorac Cardiovasc Surg 1986;91:818–825.

[2] Puga FJ, Leoni FE, Julsrud PR, Mair DD. Complete repair of pulmonary atresia, ventricular septal defect and severe peripheral arborization abnormalities of the central pulmonary arteries. Experience with preliminary unifocalization procedures in 38 patients. J Thorac Cardiovasc Surg 1989;98:1018–1029.

[3] Marelli AJ, Perloff JK, Child JS, Laks H. Pulmonary atresia with ventricular septal defect in adult. Circulation 1994;89:243–251.

[4] Sawatari K, Imai Y, Kurosawa H, Isomatsu Y, Momma K. Staged operation for pulmonary atresia and ventricular septal defect with major aorto-pulmonary collateral arteries. J Thorac Cardiovasc Surg 1989;98:738–750.

[5] Kirklin JW, Barratt-Boyes BG. Tetralogy of Fallot with pulmonary atresia. 2nd edition. Cardiac Surgery, Vol. 2. New-York: Churchill-Livingston, 1993. pp. 942–973.

[6] Reddy VM, Liddicoat JR, Hanley FL. Midline one-stage complete unifocalization and repair of pulmonary atresia with ventricular septal defect and major aorto-pulmonary collaterals. J Thorac Cardiovasc Surg 1995;109:832–845.

[7] Tchervenkov C, Salasidis G, Cecere R, Beland MJ, Jutras L, Paquet M, Dobell AR. One stage midline unifocalization and complete repair in infancy versus multiple stage unifocalization followed by repair for complex heart disease with major aorto-pulmonary collaterals. J Thorac Cardiovasc Surg 1997;114:727–737.

[8] Carotti A, Di Donato R, Squitieri C, Guccione P, Catena G. Total repair of pulmonary atresia with ventricular septal defect and major aorto–pulmonary collaterals: an integrated approach. J Thorac Cardiovasc Surg 1998;116:914–923.

[9] Iyer KS, Mee RBB. Staged repair of pulmonary atresia with ventricular septal defect and major systemic to pulmonary artery collaterals. Ann Thorac Surg 1991;51:65–72.

[10] Watterson KG, Wilkinson JL, Karl TR, Mee RBB. Very small pulmonary arteries: central end-to-side shunt. Ann Thorac Surg 1991;52:1132–1137.

[11] Rome JR, Mayer JE, Castaneda AR, Lock JE. Tetralogy of Fallot with pulmonary atresia. Rehabilitation of diminutive pulmonary arteries. Circulation 1993;88:1691–1698.

[12] Nakata S, Imai Y, Takanashi Y. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. J Thorac Cardiovasc Surg 1984;88:610–619.

[13] Bull K, Somerville J, Ty E, Spiegeihaker D. Presentation and attrition in complex pulmonary atresia. J Am Coil Cardiol 1995;25:491–499.

[14] Kirklin JW, Blackstone EH, Shimazaki Y, Maehara T, Pacifico AD, Kirklin JK, Bargeron LM. Survival, functional status, and reoperations after repair of tetralogy of Fallo with pulmonary atresia. J Thorac Cardiovasc Surg 1988;96:102–116.

[15] Haworth SG, Rees PG, Taylor JFN, Macartney FJ, De leval M, Stark J. Pulmonary atresia with ventricular sepia! defect and major aorto-pulmonary collateral arteries: effect of systemic pulmonary anastomosis. Br Heart J 1981;45:133–141.

[16] Sullivan ID, Wren C, Stark J, De leval MR, Macartney FJ, Deanfield JE. Surgical unifocalization in pulmonary atresia and ventricular septal defect. A realistic goal? Circulation 1988;78:1113–1115.

[17] Moritz A, Manfred M, Wollenek G, Domanig E, Wolner E. Complete repair of PA-VSD with diminutive or discontinuous pulmonary arteries by transverse thoraco-sternotomy. Ann Thorac Surg 1996;61:646–650.

[18] Haworth SG. Collateral arteries in pulmonary atresia with ventricular septal defect: a precarious blood supply. Br Heart J 1980;44:5–13.

[19] Lofland GK. The management of pulmonary atresia, ventricular septal defect, and multiple aorta pulmonary collateral arteries by definitive single stage repair in early infancy. Eur J Cardio-thorac Surg 2000;18:480–486.

Appendix A. Conference discussion

Dr R. DiDonato (Rome, Italy): This is a beautiful series with excellent results.

We in Rome have a series of over 30 patients and we basically follow Dr Hanley's approach of single-stage unifocalization and repair. However, our approach differs a little bit in those cases in whom we have severely hypoplastic pulmonary arteries, although confluent, and small MAPCAS. These patients we treat exactly the way you do. We do a staged approach with an outflow patch. We obtain beautiful growth of the pulmonary arteries to the point that when we do the repair we may not even need to unifocalize all the MAPCAS.

I have three questions. Do you use this approach in all the patients with this disease, even those with good MAPCAS? Second, what do you mean by Nakata index? Is this just applied as it traditionally is to the pulmonary artery size or to the measurement of pulmonary arteries plus MAPCAS? That is what we call neopulmonary artery index, because it's not exactly the Nakata index. And the third question is, how do you decide to close the VSD during the operation? Do you do an intraoperative flow study the way Dr Hanley suggested and that we also use as an intraoperative test?

Dr Metras: Of course I'm perfectly aware of your beautiful work on the Frank Hanley approach. we have tried to go opposite and promote the pulmonary artery bed growth and hope to avoid late development of pulmonary artery vascular disease.

Now, to answer your questions, the last question, if I remember well, was the VSD closure. Well, the VSD closure, once we go ahead to do the total correction is when we think that the pulmonary artery is developed enough, and it's just intraoperatively that we see at the end of bypass if it is tolerated well.

Concerning the Nakata index, the Nakata index, of course, is calculated on the true pulmonary arteries. It's not the corrected Nakata index since we don't use the collaterals. So there is no use of calculating this Nakata index adding pulmonary artery and collaterals.

And your first question?

Dr DiDonato: Do you use this approach in all the patients?

Dr Metras: No. We use the approach only in extremely hypoplastic pulmonary arteries with collaterals.

Dr DiDonato: You end up occluding these MAPCAS?

Dr Metras: Absolutely. When we do this correction, if the pulmonary bed has developed enough, the MAPCAS either have closed spontaneously or have been coil-occluded.

Dr T. Tlaskal (Prague, Czech Republic): In Kardiocentrum of the University Hospital Motol in Prague we have got experience with the surgical treatment of more than 40 patients with pulmonary atresia and MAPCAS. Several different approaches were used for unifocalisation and rehabilitation