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 mm²/m²), 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. © 2001 Elsevier Science B.V. All rights reserved.

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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.
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 postoperative 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 mm²/m² [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/m².
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 bypass (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.
The incision was then 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

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.
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) oc- cessional placement of stents in the PA (d) coil occlusion of collaterals if the O2 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 visualization 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 O2 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 O2 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 O2 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, systemicpulmonary 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 angiograms 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 systemic-pulmonary 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, O₂ 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, O₂ 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 collateral, reconstruction of PA bifurcation, VSD closure, embolization of a main collateral.

References


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. Tkáček (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.
of the pulmonary vascular bed. However, we have seen also specific subset of neonates with extremely hypoplastic pulmonary arteries without real MAPCAs. Often, multiple but very small collateral aortopulmonary arteries were present. In these patients we tried to use different approaches to increase the pulmonary blood flow. Unfortunately, however, in these infants central shunt nor right ventricular-to-pulmonary artery patching were not efficient enough to stimulate the growth of the right and the left pulmonary arteries which remained extremely hypoplastic with a diameter in the range between 1 and 3 mm even 2 or 3 years after the first surgery performed during the early infancy. I would like to ask you if you have ever seen such patients in whom the pulmonary arteries would not grow at all.

**Dr Metras:** First of all, I want to say something about one word you said. The shunts on these very small pulmonary arteries, it has been shown very many years ago by Roger Mee that the worst thing for these pulmonary arteries was to branch a shunt on them. So I think it’s better to promote forward flow with opening the RV–PA outflow.

**Excuse me, your question?**

**Dr Tlaskal:** If you have ever seen such patients in whom the pulmonary arteries would not grow at all.

**Dr Metras:** No. In all these patients pulmonary arteries have grown enough to reach correction with the possibility to close the VSD totally. That means probably the RV pressure is satisfactory. So once you have promoted this growth, it works well. The two first patients were sent to Jim Lock, who put stents in the pulmonary arteries. They increase considerably. The video would have shown that nicely.

**Dr W. Brawn (Birmingham, UK):** Your ratio of RV to LV was 60%.

**Dr Metras:** Yes.

**Dr Brawn:** 60%, and some range between 30% and 1. Do you think you’ve done the patient a disservice by closing the VSD, because some of those patients that have a ratio of 1 and probably quite high ratios, what’s going to happen to them in the long-run in terms of RV failure?

**Dr Metras:** The ratio that I mention here is the last ratio in the follow-up. Some of them may be high because there is some obstruction at the level of the RV–PA homograft that may be replaced when we use the 16 mm or 18 mm conduit. So it may not reflect absolutely the situation.

**Dr Brawn:** What sort of pressures would you be happy with for the long term, because otherwise these patients are going to develop RV failure, aren’t they?

**Dr Metras:** Well, if the hemodynamic situation is satisfactory, we will accept immediately the correction, RV/LV ratio up to 1. We don’t have them, but up to 1, it’s all right, we don’t open the VSD if the RV/LV ratio is 1. This has been done the same way. We follow exactly the rules of the Boston group.

**Dr Brawn:** That’s very interesting.

**Dr A. Corno (Lausanne, Switzerland):** I am very pleased that you mentioned a late appearance of collaterals. In the last year we had a series of 5 patients in whom a catheterization study showed the presence of a MAPCA of considerable size, and this was after repair either done by us or in another institution. Instead of asking our cardiologists to coil-embolize this collateral, we decided to go back through a thoracotomy to disconnect the origin of this MAPCA from the aorta and connect it to the pulmonary artery. We don’t have the late results yet because our experience is too recent, but I would like to have your opinion on this.

**Dr Metras:** I think one very interesting thing that we are waiting for in the series of Roberto and, of course, Frank Hanley is how will these collaterals behave in the future in terms of pulmonary vascular disease, since I guess that in your patients the pulmonary artery developed very little if most of the flow goes to all of these big collaterals.