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Surgical Management of Fallot's Tetralogy With Pulmonary Atresia and Major Aortopulmonary Collateral Arteries: Multistage Versus One-Stage Repair

Adriano Carotti, MD[1]

Abstract

A strict and rational approach to Fallot's tetralogy with pulmonary atresia and major aortopulmonary collateral arteries allows to achieve optimal results. Rehabilitative and unifocalization strategies do not constitute separate philosophies; instead the surgical strategy should be tailored to each individual patient. Based on our previous experience, the ability to achieve definitive intracardiac repair is the real determinant of both improved survival and adequate systolic right ventricular performance on mid-term follow-up.

Keywords

Fallot's tetralogy, major aortopulmonary collateral arteries, unifocalization, rehabilitation

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Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries (TOF, PA, MAPCAs) should be considered a cardiopulmonary disease: While intracardiac anatomy is somewhat repetitive, the pulmonary vascularity is variegated, with sometimes extremely complex patterns. Native pulmonary arteries can show varying degrees of hypoplasia or even be absent; on the other hand, MAPCAs perfuse with varying modalities all or part of the lung parenchyma.[1,2] Due to the presence of collaterals, onset of symptoms in neonatal age is rare, and clinical manifestation of the disease can vary widely depending on the anatomy and physiology of pulmonary blood sources.

The surgical treatment aims at obtaining a completely separated two-ventricle circulation with the lowest possible post-repair right ventricle (RV) pressure.[3] Therefore, the extent of pulmonary vascular bed affects surgical options and outcome.

The majority of patients have lungs partially perfused by MAPCAs and also show variable degrees of pulmonary arterial arborization defects (Figure 1). Considering such heterogeneity of the pulmonary vascular supply patterns, what kind of approach should be chosen? And what should be the timing for surgery?

In our center, all patients with TOF, PA, MAPCAs undergo cardiac computed tomography angiography with three-dimensional rendering reconstructions to evaluate the relationship between the MAPCAs and the esophagus and bronchi (Figure 2A and B). Cardiac catheterization, however, with the pulmonary vein wedge-injection remains the gold standard for evaluating the presence of true pulmonary arteries.

Pulmonary arteries rehabilitation aimed at inducing native pulmonary arterial growth was the first surgical treatment described for TOF, PA, MAPCAs.[4,5] The Gates-Laks central shunt is probably the most commonly performed procedure for its effectiveness and for the possibility of off pump realization,[6] although some groups still prefer an RV to pulmonary artery connection.[7]

A strong rehabilitation policy is advocated by the Melbourne group, which supports an early approach by central shunt, followed by revaluation of pulmonary arterial growth, conversion to RV to pulmonary artery conduit and, hopefully, complete repair with or without pulmonary arterial branches augmentation.[8] However, the policy of the Melbourne group relies almost exclusively on the presence of native pulmonary

[1] Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital and Research Institute, Rome, Italy

Corresponding Author:
Adriano Carotti, Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital and Research Institute, P.zza S.Onofrio, 4, 00165 Rome, Italy.
Email: adriano.carotti@opbg.net

Abbreviations and Acronyms

RV	right ventricle
RVSP/SBP	right ventricular systolic pressure/ systolic blood pressure
TOF, PA, MAPCAs	tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries
VSD	ventricular septal defect

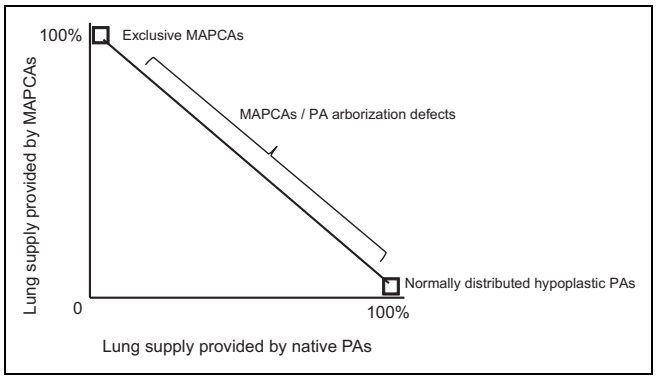

Figure 1. Perfusion provided by the native pulmonary arteries (abscissa axis) and major aortopulmonary collateral arteries (ordinate axis). When excluding the two extremes, the majority of patients is constituted by those with major aortopulmonary collateral arteries and variable degrees of pulmonary arterial arborization defects. MAPCAs indicate major aortopulmonary collateral arteries; PAs, pulmonary arteries.

arteries. This is mainly because the long-term results of their historical experience with staged unifocalization have shown that MAPCAs are unstable vessels, responsible for underdevelopment of pulmonary vascular bed, and high RV pressure of repairs leading to late RV failure.[9]

On the other hand, the Stanford group favors the one-stage approach.[10] Through a midline sternotomy and after extensive mediastinal dissection, all collaterals are detached from their aortic origin and anastomosed to each other and/or to the native pulmonary arteries to create a new pulmonary vascular tree, appropriately augmented with the use of homograft tissue.

The assumption of unifocalization, supported by clinical data, is that collaterals are not unstable vessels but innocent bystanders, and that their early removal from systemic circulation avoids both MAPCAs degeneration and involution of the native pulmonary arteries. Therefore, early unifocalization recruits vascular supply to all healthy lung segments using both native pulmonary arteries and collaterals as raw material.[11]

The attitude of the world's leading center, Stanford, is well known, basing its approach mainly on unifocalization, reserving very restricted indication to pulmonary arterial rehabilitation[12,13] just for cyanotic patients with small intrapericardial branch pulmonary arteries that are confluent, have normal arborization, and have dual-supply collateral vessels (about 10% of patients with a diagnosis of TOF, PA, MAPCAs). In addition, a subset of patients undergoes complete unifocalization with placement of a shunt to provide pulmonary blood flow rather than simultaneous intracardiac repair. This approach is reserved for patients with multifocal pulmonary blood supply who will not be able to undergo single-stage

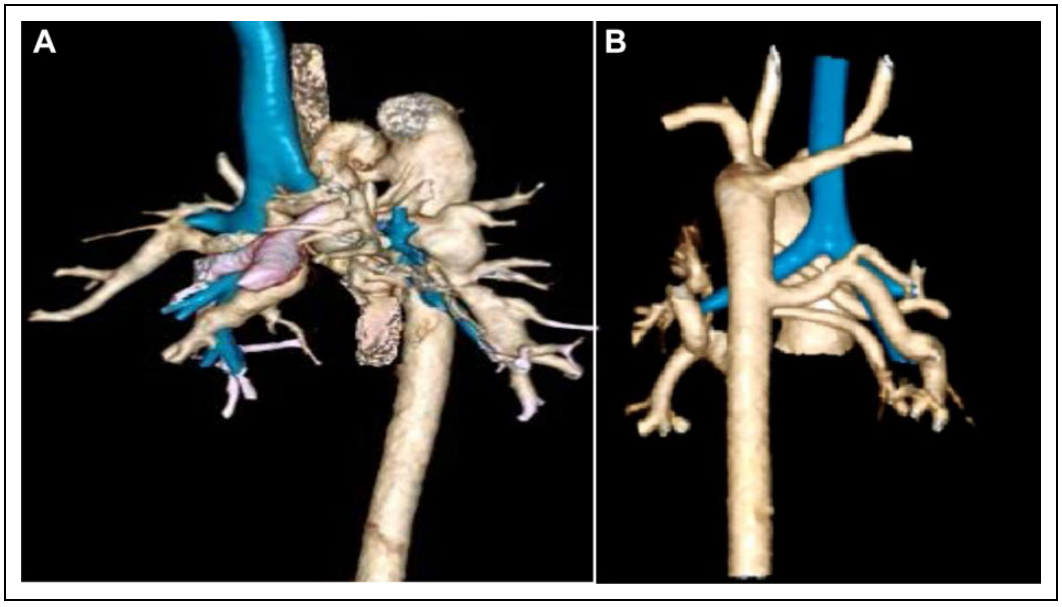

Figure 2. Three-dimensional rendering reconstructions of computed tomography scans highlighting the relationship of major aortopulmonary collateral arteries with both airway (in blue) and esophagus (in dotted ocher). A, A case with true pulmonary arteries (in pink). B, A case with exclusive major aortopulmonary collateral arteries.

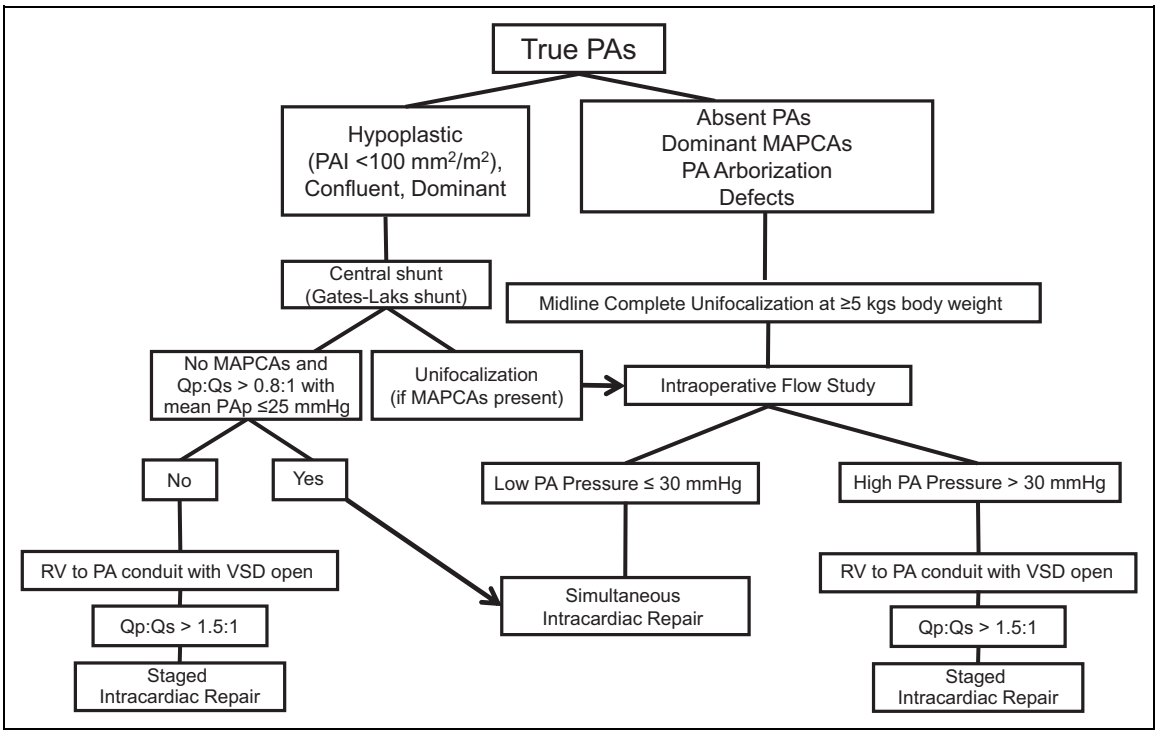

Figure 3. Decision-making tree for the treatment of patients with tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries. MAPCAs, major aortopulmonary collateral arteries; PA/PAs, pulmonary artery/pulmonary arteries; PAI, pulmonary artery index; PAp, pulmonary artery pressure.

repair with acceptably low RV pressure, usually because of small PAs and small MAPCAs with multiple stenoses.[14]

We believe that being able to trace the line of demarcation between the territory of unifocalization and that of rehabilitation, in the context of an integrated approach to such a complex pathology, can lead to optimal results. Our approach to TOF, PA, MAPCAs is similar to that of the Stanford's group, with less stringent indication for pulmonary artery rehabilitation and early timing of unifocalization. Pulmonary arterial rehabilitation is performed in all patients with confluent hypoplastic (ie, pulmonary arterial index less than 100 mm^2/m^2) but dominant pulmonary arteries distributed to the most of the lung parenchyma with eventual MAPCAs supplying the areas nonperfused by the true pulmonary arteries.

Our current technique of choice for rehabilitation is the central shunt. All patients if clinically stable undergo a cardiac catheterization 6 to 12 months after the procedure, and according to the results we might proceed with unifocalization alone, in case of associated terminal MAPCAs, or repair. The decision whether to proceed to concomitant repair is based on the intraoperative pulmonary flow study (Figure 3).

Considering pulmonary rehabilitation a straightforward procedure, we usually perform it off-pump, usually avoiding the neonatal period. On the other hand, for complete unifocalization and repair we try to perform surgery in patients of at least 5 kg of body weight, irrespective of patient's age. This is because, from the analysis of the results of the first 15 years of our experience, neonatal age and low body weight at

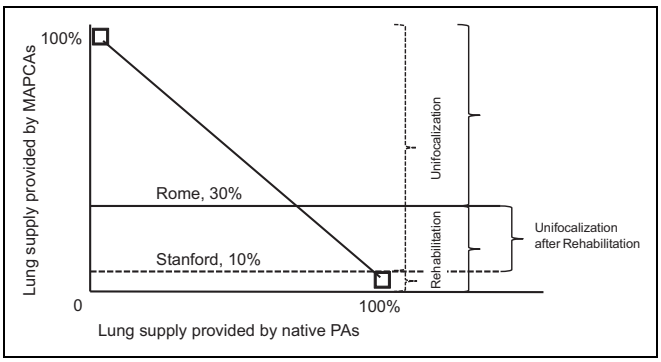

Figure 4. Rehabilitation and unifocalization policy in Stanford (10% vs 90%) and Rome (30% vs 70%). In our center, rehabilitation is utilized also in the 20% of patients with no exclusive distribution of native pulmonary arteries to the lungs which may undergo a second-stage unifocalization procedure. MAPCAs indicate major aortopulmonary collateral arteries; PAs, pulmonary arteries.

unifocalization proved to be determinant of unsatisfactory outcomes.[15] In our experience, we do believe that pulmonary artery rehabilitation should be considered also for patients with nonexclusive distribution of native pulmonary arteries to the lungs which may receive in a second-stage unifocalization after rehabilitation (Figure 4).

Based on the aforementioned considerations, since 1994 to date, we treated 152 patients with TOF, PA, MAPCAs (Figure 5) with a median age of 12 months (range, 10 days-35 years). This retrospective study was approved by the Institutional Review

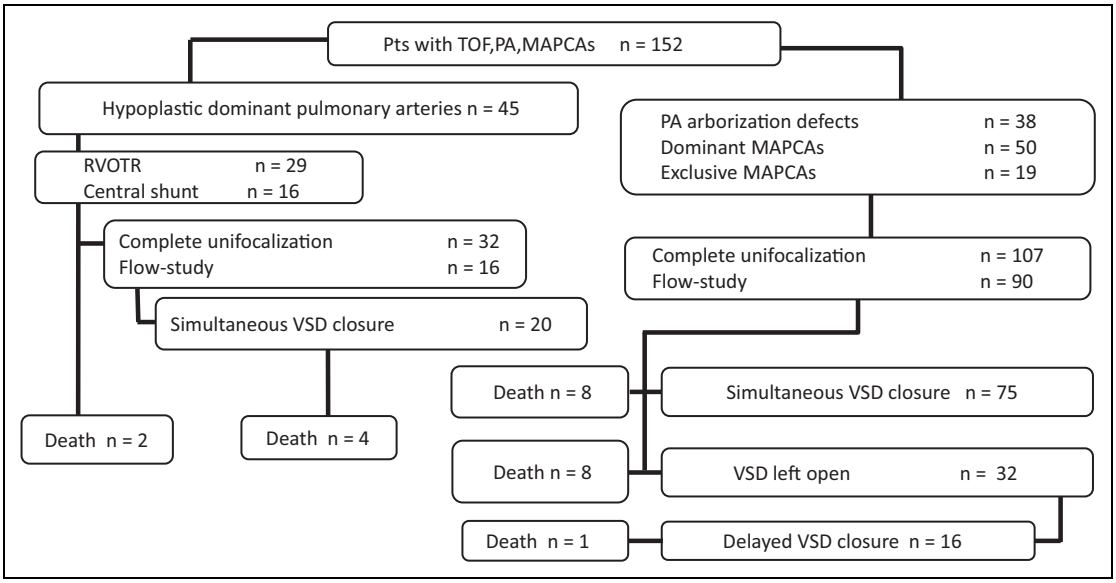

Figure 5. Overall series of Bambino Gesù Children's Hospital and Research Institute (152 cases operated between 1994 and 2019). TOF, PA, MAPCAs indicate tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries; RVOTR, right ventricular outflow tract reconstruction; VSD, ventricular septal defect.

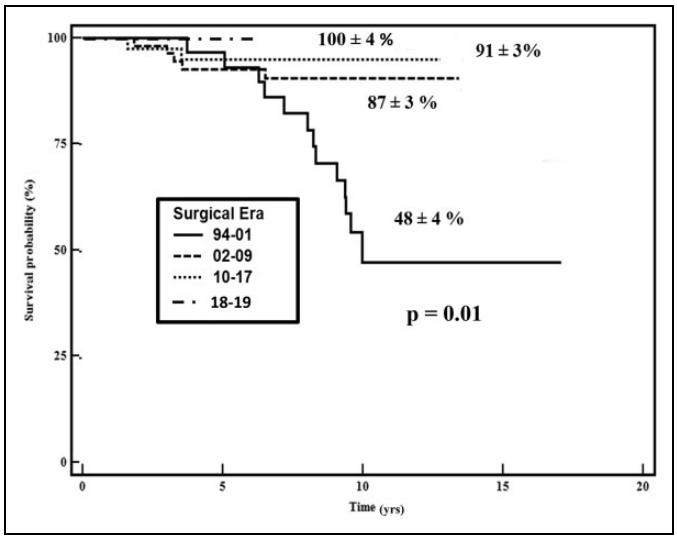

Figure 6. Survival rate by era on an overall series of 152 operated between 1994 and 2019.

Board of Bambino Gesu Children's Hospital and Research Institute, with waiver of individual consent.

Survival significantly improved from the beginning of our experience (thus reflecting both the learning curve and the improvement in postoperative medical care), being above 90% in the recent experience, up to 100% in the last couple of years (Figure 6). Based on a recent analysis,[16] the overall rate of complete intracardiac repair in our series was 83.3% for all patients undergoing unifocalization, either primary or after rehabilitation procedures (16.6%). Postrepair right ventricular systolic pressure/systolic blood pressure (RVSP/SBP) ratio was 0.49 $\pm$ 0.15 (range: 0.2-0.75) in patients who underwent successful concomitant ventricular septal defect (VSD) closure, with no influence by previous staged palliation (RVSP/SBP ratio of 0.47 $\pm$ 0.14 vs 0.50 $\pm$ 0.15, $P = .53$). Furthermore, patients who failed one-stage repair and underwent delayed VSD closure (at a median of 2.3 years after the initial operation) showed a postrepair RVSP/SBP ratio not statistically different compared with patients who underwent concomitant VSD closure (mean: 0.50 $\pm$ 0.17, $P = .86$).

Finally, hemodynamic result showed stability over time, as evidenced by a mean RVSP/SBP ratio of 0.53 $\pm$ 0.18 recorded in a subgroup of patients who underwent cardiac catheterization at a median interval from repair of 95 months.[15] The intraoperative pulmonary vascular compliance test plays a key role in selecting patients suitable for concomitant intracardiac repair carried out after unifocalization and utilizing a mean pulmonary arterial pressure cutoff value of 30 mm Hg, as reported in our original paper in 1998.[17]

In a recent analysis of our overall experience with flow-study aided TOF, PA, MAPCAs repair, flow study accurately predicted successful VSD closure, showing a significant positive correlation between pressures recorded at the time of the flow study and the postoperative RV to systemic blood pressure ratio, in patients who successfully underwent VSD closure.[18] However, flow study pressure values recorded intraoperatively did not affect long-term outcomes, whereas the ability to achieve definitive intracardiac repair (in either one or more stages) is the real determinant for improved survival.

This was also confirmed by the most recent study of our group,[16] which demonstrated that high pressures recorded during the flow study did not preclude a deferred successful VSD closure in about 50% of cases, with good hemodynamics and life expectancy overlapping that of one-stage repairs. In this study, for patients who underwent either concomitant or delayed intracardiac repair, echocardiographic RV systolic function was

retrospectively calculated to assess (1) RV fractional area change and (2) two-dimensional RV longitudinal strain of the free wall of the RV. No differences in terms of systolic RV performance were observed between the two groups.

In conclusion

- Staged and one-stage approach do not constitute separate philosophies, but must be used in the context of an integrated approach.
- Patient selection is critical for both rehabilitation and unifocalization procedures.
- The ability to achieve definitive intracardiac repair is the real determinant for both improved survival and adequate systolic RV performance on mid-term follow-up.

Declaration of Conflicting Interests

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ORCID iD

Adriano Carotti, MD https://orcid.org/0000-0003-3908-5091

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