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Tetralogy of Fallot with Major Aortopulmonary Collaterals: Early Total Repair

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Abstract. Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collaterals is a complex lesion distinguished by marked heterogeneity of pulmonary blood supply. Over the past two decades, investigators have developed various approaches to the management of this anomaly generally based on the concept of staged unifocalization of pulmonary blood supply. Although such approaches may represent an improvement on the natural history of this lesion, they remain inadequate for a substantial portion of patients born with tetralogy of Fallot and major aortopulmonary collaterals. Since 1992, our approach has been to perform one-stage complete unifocalization through a midline approach in all but a few extremely complicated patients. We aim to repair these patients early in infancy, with an emphasis on native tissue–tissue reconstruction, in order to optimize prospects for survival with a good functional outcome in as many patients as possible. In this review, we present our philosophy and our experience with unifocalization and repair in 72 patients.

Key words: Pulmonary atresia — Pulmonary blood flow — Pulmonary arteries — MAPCAs

Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries (MAPCAs) is unique from most other forms of complex congenital heart disease inasmuch as its complexity is a function of heterogeneous and frequently severe anomalies of pulmonary blood supply. MAPCAs, which are thought to derive from the embryonic splanchnic vascular plexus, contribute to the pulmonary blood supply in 30–65% of patients with tetralogy of Fallot and pulmonary atresia [11, 34]. Highly variable in terms of number, size, origin, course, arborization, and structure, collaterals may constitute the sole source of pulmonary blood flow or they may supply as little as a single lung segment. The extent of distal pulmonary vascular bed supplied by collaterals generally varies inversely to that supplied by the true pulmonary arteries [34], which range from normal in size to completely absent. However, a given lung segment may be supplied by both collaterals and true pulmonary arteries, with connections between the two sources occurring centrally or peripherally and at single or multiple sites. The complexity of tetralogy of Fallot with MAPCAs is such that, until the mid-1970s, most patients were managed either medically, with surgical palliation, or with right ventricular outflow reconstruction and ligation of MAPCAs [6, 21, 23, 24]. The first major advances in the understanding and treatment of this lesion came with the morphologic and physiologic characterization of the extremely variable pulmonary blood supply [4, 8–10, 13, 17, 18, 27, 38]. On the basis of these studies, Haworth and Macartney postulated that pulmonary blood supply in these patients might be normalized by unifocalizing the individual pulmonary arteries and MAPCAs [8, 9].

Since the early reports by Haworth and Macartney [8–10, 18] and others [4, 13, 17, 27, 38], a number of investigators have developed programs for the management of this challenging lesion. Most of these have been based on the concept of staged unifocalization, with the initial phase of management often designed to increase flow to the true pulmonary arteries in an effort to stimulate growth [5, 11, 12, 20, 25, 26, 31–33, 36, 40]. While these various strategies have served to advance the field and have provided good results for a select group of patients, few include a sizable cohort of young infants and most leave a substantial proportion of patients without complete repair. Nine large series of staged unifocalization are summarized in Table 1. Taking into account natural attrition prior to entry into such a program, which is highest in infancy [1], we estimate that only 20–30% of a cohort of newborns with tetralogy of Fallot and MAPCAs will achieve complete repair with acceptable right ventricular hemodynamics if a delayed staged approach is taken. As Bull and colleagues [1] observed only a few of years ago, ‘‘Infancy is the period of greatest attrition for complex pulmonary atresia, without and with attempts of palliation . . . any surgical focus aiming

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Table 1. Prior series of staged unifocalization for tetralogy of Fallot with MAPCAs

Series	Years of study	No. operated patients[a]	Median age 1st operation (years)	No. (%) completely repaired with low RV pressure[b]
Dinarevic et al. [5]	1972–1992	24	0.14	2 (8)
Hofbeck et al. [11]	1976–1988	32	0.86	4 (13)
Iyer and Mee [12]	1979–1989	58	1.8	26 (45)
Permut and Laks [25]	1982–1994	29	—	12 (41)
Puga et al. [26]	1982–1987	38	Mean 4.7	18 (47)
Rome et al. [31]	1978–1988	43	—	17 (40)
Sawatari et al. [32]	1982–1988	34	Mean 6.6	10 (29)
Sullivan et al. [36]	1979–1986	26	2.0	3 (12)
Yagaihara et al. [40]	1985–1993	50	Mean 5.9	26 (52)

—, Median/mean for entire group not specified.

[a] Does not include patients in these series who did not undergo surgery.

[b] Low right ventricular (RV) pressure is considered <85% systemic pressure.

to make an impact on the overall mortality of this condition must alter the pattern of attrition in infancy. Recent influential reports . . . do not address this most difficult group of patients.'' [p. 496]

Therapeutic Goals

The ultimate goal of surgical therapy in tetralogy of Fallot with MAPCAs is to normalize circulatory physiology. Among patients surviving complete repair, the most important physiologic indicator of outcome is peak right ventricular pressure, which should remain as low as possible [15, 35]. For a right ventricle pumping a single cardiac output, peak pressure is a direct function of pulmonary vascular input impedance, which is generally calculated as resistance. In addition to vascular resistance, which is inversely related to the total cross-sectional area of the distal pulmonary vascular bed, pulmonary arterial input impedance is also influenced by pulse-wave propagation characteristics and the presence or absence of arterial stenoses.

Given the importance of right ventricular pressure in predicting outcome, the most reliable means of ensuring that patients receive the optimal benefit from surgical therapy is to make every effort to minimize right ventricular afterload. This principle forms the basis for our approach to management. In patients with repaired tetralogy of Fallot and pulmonary atresia, the number of lung segments supplied by the pulmonary arterial system has been found to correlate strongly with pulmonary arterial pressure and calculated pulmonary vascular resistance [35]. Thus, in patients with multifocal pulmonary blood supply, it is important to incorporate as many lung segments as possible into the unifocalized pulmonary vascular bed. It is also important to ensure that resistance in a given pulmonary segment is as low as possible, which is largely a function of the health of the microvasculature. Presumably, the microvasculature of the lung in patients with MAPCAs is in its healthiest state at birth. The natural history of MAPCAs often follows a course of progressive stenosis and occlusion, sometimes precluding access to a given segment of lung at the time of unifocalization. Even if a collateral is accessible, a long-standing severe stenosis may lead to distal arterial hypoplasia and underdevelopment of preacinar and acinar vessels and alveoli [27], which may not be able to exhibit full compensatory growth after revascularization [14]. Also, iatrogenic occlusion can occur when MAPCAs are unifocalized in stages using nonviable conduits, sometimes resulting in loss of segments. Just as the health of the pulmonary vascular bed may suffer from stenosis or occlusion of MAPCAs, unrestricted flow through large collaterals without protective stenoses can lead to pulmonary vascular obstructive disease, which also effectively raises resistance. Experimental studies have shown that pulmonary vascular function and structure are altered after as little as 4 weeks of postnatal pulmonary hypertension with high pulmonary blood flow [29]. Likewise, staged unifocalization using systemic–pulmonary arterial shunts may result in pulmonary vascular obstructive disease without necessarily stimulating compensatory flow-related growth [14]. Injuries to the distal pulmonary vasculature due to both hypoperfusion and perfusion at systemic pressure are progressive, time-related processes. Thus, it seems clear that one of the keys to optimizing the health of the pulmonary vasculature, and hence the entire right heart complex, is to normalize the pulmonary circulation as early in life as possible, removing the pulmonary vascular bed from exposure to the inevitable hemodynamic vagaries associated with MAPCAs.

Although the mechanics of the pulmonary vasculature and its coupling with the right ventricle are not understood in their full complexity, especially in the context of the heterogeneous aborization and structural

anomalies characteristic of tetralogy of Fallot with MAPCAs, this may also be an important issue in evaluating the health of the pulmonary circulation [7, 16]. When circumferential conduits are employed in the central and hilar reconstruction of the pulmonary vasculature, the mechanical efficiency of right ventricle–pulmonary arterial coupling is likely to be impaired. Large pericardial manifolds or synthetic conduits, regularly incorporated into the reconstruction in many approaches to staged unifocalization [12, 25, 26, 32, 36, 40], may produce extensive compliance and diameter mismatches in the pulmonary vascular bed, which can decrease the efficiency of right ventricle–pulmonary arterial coupling and predispose to shear stress-related vascular changes at anastomotic junctions [7, 39]. Other potential disadvantages of such methods of repair include lack of growth potential and a tendency to calcify or acquire a pseudointimal peel.

In addition to the pulmonary vascular bed and postrepair right ventricular afterload, there are other factors to consider in defining therapeutic goals of management for tetralogy of Fallot with MAPCAs. Long-term right ventricular function may also be affected by prolonged pressure load prior to repair. Two of the major long-term problems that beset patients with repaired tetralogy of Fallot are ventricular dysfunction and ventricular arrhythmias. While in many cases these are related to ventriculotomy [19], hypertrophy due to the pressure load and myocardial ischemia associated with cyanosis both contribute to myocardial fibrosis. As a consequence, right ventricular compliance and systolic function are both decreased. In one study of tetralogy patients without MAPCAs, age at repair was the sole independent predictor of postoperative dysrhythmias, whereas duration of follow-up and era of surgery were not significant [27]. This finding may also be related to ventricular hypertrophy and fibrosis. The systemic circulation can be affected as well because the increased left ventricular volume load incurred with prolonged systemic–pulmonary arterial shunting (through either MAPCAs or palliative shunts) may have a detrimental influence on both left ventricular and aortic valvar function [2, 3]. Other potential benefits of normalizing the circulation as early as possible include avoiding the potential consequences of hypoxemia and polycythemia, which may affect development of the brain and other organs.

In light of the poor natural history of tetralogy of Fallot with MAPCAs and the inadequacy of staged approaches for achieving successful complete repair in the majority of patients, we have developed a program for managing this lesion that is founded on the principle of early total repair with one-stage unifocalization of all lung segments and maintenance of native tissue continuity [28]. The idea of this program is to provide complete repair with optimum results for as many patients as possible. Keeping in mind the therapeutic goals outlined previously, we prefer to complete the repair in early infancy. This allows for early normalization of cardiovascular physiology, with preservation of the pulmonary vascular bed, recruitment of all lung segments, alleviation of cyanosis, and prevention of other cardiac sequelae. By performing the repair in a single stage, the number of operations required can be minimized and the number of patients who can be completely repaired is likely to be enhanced.

Techniques of Unifocalization

Our approach to one-stage unifocalization of pulmonary blood supply in tetralogy of Fallot with MAPCAs follows several basic principles. It is essential to control all collaterals before commencing cardiopulmonary bypass and to perform native tissue–tissue reconstruction whenever possible. There are a number of important techniques that can aid in achieving these goals. The mediastinum is entered through a median sternotomy, which is extended and retracted widely in order to improve exposure. Other investigators have described the use of a bilateral transsternal thoracotomy, which will also provide good exposure [22]. Collaterals are identified and dissected using a variety of approaches. Both pleural spaces are opened widely anterior to the phrenic nerves and the lungs are lifted out of their respective pleural cavities, allowing identification of collaterals at their aortic origins. Additional collaterals from the upper descending aorta are identified and dissected in the subcarinal space (between the tracheobronchial angle and the roof of the left atrium) by an approach between the right superior vena cava and the aorta (Fig. 1). The floor of the pericardial reflection in the transverse sinus is opened and the posterior mediastinal soft tissues are dissected to expose the aortic segment and the collaterals in this region. This is an important measure not only for gaining access to MAPCAs, which often arise from this location, but also for providing a direct pathway for rerouting of MAPCAs in order to facilitate tissue–tissue anastomoses. Collaterals arising from the aortic arch, brachiocephalic vessels, or coronary arteries are also exposed and dissected. Immediately before instituting cardiopulmonary bypass, all collaterals are ligated at their origin in order to achieve controlled perfusion.

As many collaterals as possible are ligated, mobilized, and unifocalized without cardiopulmonary bypass. With the ligation of each collateral, the decrease in arterial oxygen saturation is reflected by the pulse oximeter, so it is possible to proceed with unifocalization off bypass until the desaturation approaches a compromising level. At this point, moderate hypothermic partial cardiopulmonary bypass is instituted with the heart beating, and the remaining collaterals are unifocalized. A calcium supplemented blood prime is used to maintain normal

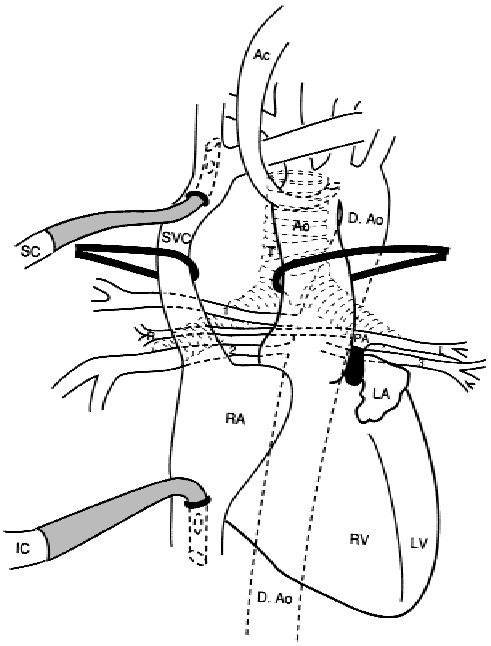

Fig. 1. The transverse sinus is an important avenue for dissecting, rerouting, and unifocalizing collaterals that arise from the descending aorta in the region of the subcarinal space. In this diagram, the MAPCAs (1–3) are depicted as anterior to the tracheobronchial tree. However, collaterals in this region have a variable relation to the extraparenchymal airways and esophagus, and the technique of rerouting and unifocalization must be individualized to optimize the lie of each collateral. *Ac*, aortic cannula; *Ao*, ascending aorta; *D.Ao*, descending aorta; *IC*, inferior caval cannula; *L*, left pulmonary artery; *LA*, left atrium; *LV*, left ventricle; *PA*, atretic main pulmonary artery; *R*, right pulmonary artery; *RA*, right atrium; *RV*, right ventricle; *SC*, superior caval cannula; *SVC*, superior vena cava. Reproduced with permission from Reddy et al. [28].

cardiac function. During the unifocalization process, emphasis is placed on avoiding nonviable conduits in the periphery and achieving unifocalization by native tissue–tissue anastamosis. Important concepts in achieving this type of unifocalization are flexibility regarding reconstruction, aggressive mobilization, maximizing length of the MAPCAs, and creative rerouting. Avenues for collateral rerouting are developed by opening the pleura on both sides posterior to the phrenic nerves in the hilar regions and by opening the subcarinal space through the transverse sinus. In order to meet the objective of complete unifocalization without peripheral conduits and maximize native tissue–tissue anastomosis, the following peripheral and central reconstructive techniques are employed:

- Side–side anastomosis of collaterals to other collaterals or to peripheral true pulmonary arteries
- Oblique end–side anastamosis of collaterals to other collaterals or to peripheral true pulmonary arteries
- Anastamosis of true pulmonary arteries to an aortic button giving off multiple unobstructed collaterals
- Long onlay or side–side anastamosis of collaterals to the central pulmonary arteries
- End–end or end–side anastamosis of collaterals to a central conduit
- Allograph patch augmentation of distal collateral stenoses
- Allograph patch augmentation of the reconstructed central pulmonary arteries
- In rare cases, allograft conduit reconstruction of central pulmonary arteries

These techniques are used as necessary in a given patient and frequently combined, depending on the particular anatomy. Figure 2 depicts an example of complex reconstruction using several of these methods. Direct tissue–tissue anastomoses are achieved by bringing collaterals through the transverse sinus, or below or above the lung hilum, utilizing as much of the collateral length as possible. Collateral length is given the highest priority in order to achieve tissue–tissue anastamosis. For example, if a discrete stenosis is present in the midportion of a collateral, the entire collateral is still used, and the stenosis is relieved by side–side reconstruction with another collateral or true pulmonary artery or by patch augmentation if necessary. All collaterals are incorporated into the reconstruction using these methods, including those which provide dual supply to a lung segment along with a true pulmonary artery, in order to build up the neopulmonary arteries with as much native tissue as possible.

In all but a few patients, blood flow to the unifocalized pulmonary arteries is provided via an aortic allograft valved conduit from the right ventricle to the central pulmonary arteries. Following unifocalization, the distal end of the allograft conduit is anastomosed to the central pulmonary arteries. If necessary, allograft tissue from the conduit can be extended distally to help augment the central branch pulmonary arteries. Prior to completing the proximal anastomosis, intracardiac repair is performed.

Intracardiac Repair

The issue of whether to close the ventricular septal defect at the time of unifocalization is critical to successful repair. An incorrect decision may lead to substantial morbidity, if not death. With respect to this issue, one-stage unifocalization differs significantly from staged approaches inasmuch as catheterization and angiographic data are not available following unifocalization but before closure of the ventricular septal defect. Several reports of staged unifocalization have suggested criteria for ventricular septal defect closure based largely on angiographic measurement of the reconstructed pulmonary arteries and measurement of pulmonary vascular resistance [12, 26, 35]. In patients undergoing one-stage unifocal-

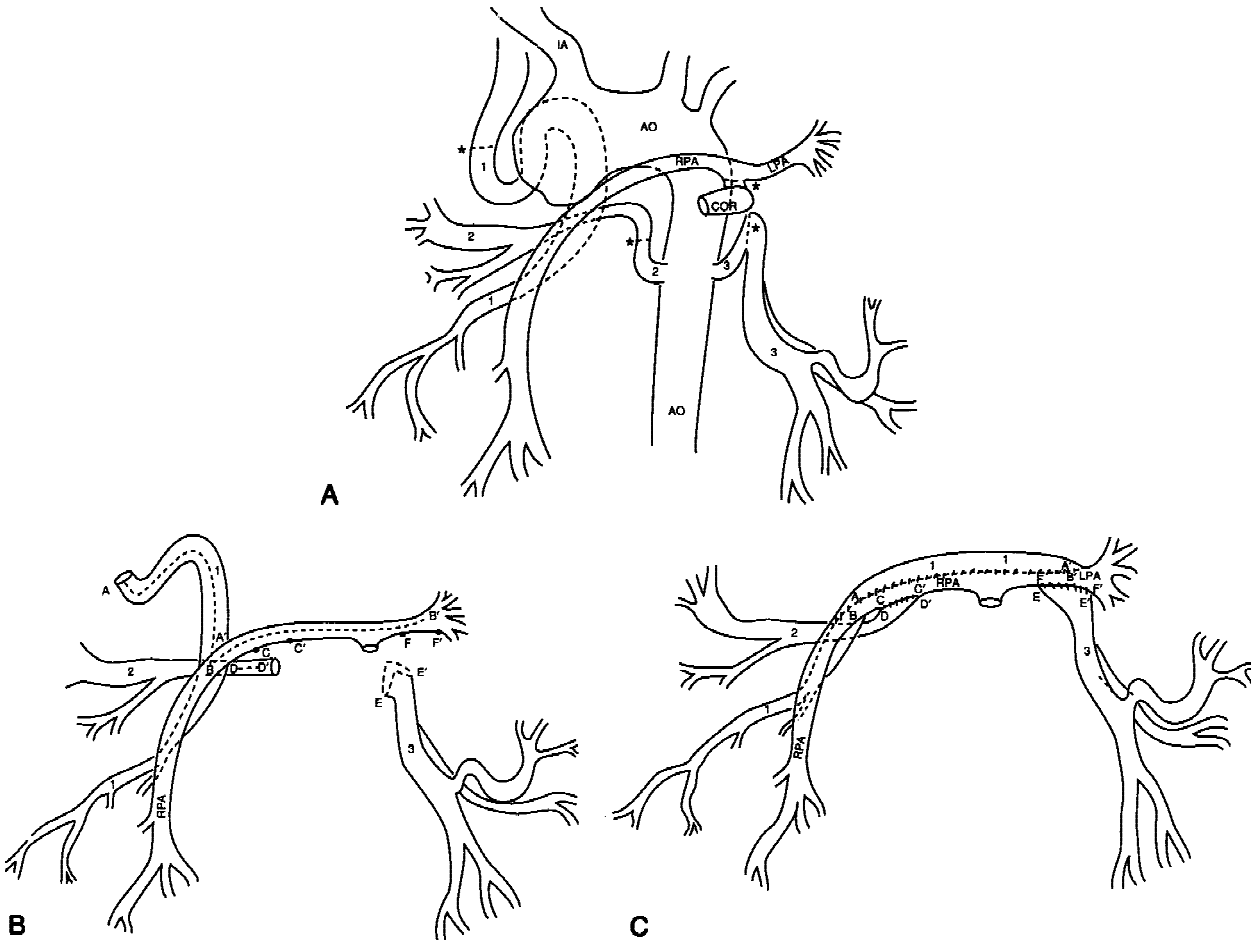

Fig. 2. Anatomy of pulmonary blood supply and unifocalization in a 3.5-month-old child who underwent one-stage complete unifocalization and repair of tetralogy of Fallot with MAPCAs. **(A)** Illustration of pulmonary blood supply, with true left (*LPA*) and right (*RPA*) pulmonary arteries, a coronary arterial collateral (*COR*) connecting to the true pulmonary artery, and three MAPCAs (1–3) from the innominate artery (*IA*) and descending aorta (*AO*). **(B)** MAPCAs are opened along the dashed lines, A–A′, D–D′, and E–E′, whereas the true pulmonary arteries are filleted open from hilum to hilum (B–B′). **(C)** Complete unifocalization and reconstruction of the central pulmonary artery is completed with full native tissue continuity. Collateral 1 A–A′ is anastomosed side–side to the incision B–B′ in the true pulmonary artery as a long onlay. Collateral 2 is anastomosed end–side to the undersurface of the RPA from D–D′ to C–C′. Collateral 3 is anastomosed end–side to the undersurface of the LPA from E–E′ to F–F′. Reproduced with permission from Reddy et al. [28].

ization, such methods are not applicable, and the decision must be made intraoperatively. Angiographic measurements of the collaterals and pulmonary arteries preoperatively can be performed in order to estimate a neopulmonary artery index, which is the sum of the indexed cross-sectional areas of all vessels unifocalized [30]. Although we have found this method to be helpful in defining a neopulmonary artery index above which all patients can undergo closure of the ventricular septal defect (200 mm^2/m^2), it is of little benefit in predicting suitability for closure in patients below this level. More useful is an intraoperative pulmonary flow study that we have developed to estimate the resistance of the unifocalized pulmonary arterial bed [30]. Following unifocalization, a pulmonary arterial catheter and perfusion cannula are placed into the valved conduit after the conduit has been attached to the pulmonary arterial system but before it is connected proximally to the right ventricle. The cannula is then snared (Fig. 3). While venting the left atrium vigorously, the lungs are perfused from the bypass pump with gradually increasing flow equivalent to at least one cardiac index (2.5 $L/min/m^2$). In both laboratory and clinical studies, we have found this method to predict reliably mean pulmonary arterial pressure with the ventricular septal defect closed [30]. If the mean pulmonary arterial pressure is $\leqslant$25 mmHg in infants, the ventricular septal defect is usually closed. Otherwise, the defect is typically left open.

After completion of the unifocalization and flow study, the aorta is cross-clamped and cardioplegia is ad-

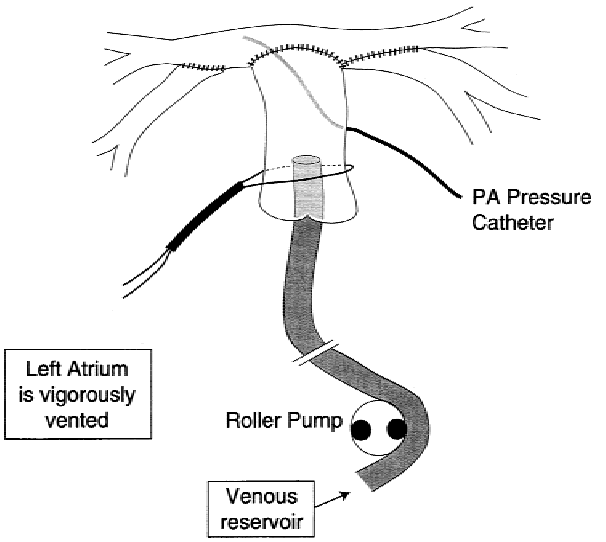

Fig. 3. Drawing of the intraoperative pump flow study. A separate pump head is used to pump blood from the venous reservoir as the cannula is introduced into the allograft conduit. A pulmonary artery (*PA*) pressure catheter is used to determined mean PA pressure while the left atrium is vented vigorously. Reproduced with permission from Reddy et al. [30].

ministered. A longitudinal ventriculotomy is made in the right ventricular infundibulum and the hypertrophied muscle bundles are resected. In patients undergoing closure of the ventricular septal defect, this is performed with an autologous pericardial patch fixed in glutaraldehyde or a Dacron patch using interrupted pledgetted sutures. The right atrium is opened to inspect the atrial septum. If an atrial septal defect or patent foramen ovale is present, it is closed partially to leave a small unidirectional interatrial communication for decompression of the right heart in the event of postoperative right ventricular dysfunction. In some cases with intact atrial septum a small one-way interatrial communication is created. Regardless of whether the ventricular septal defect is closed, right ventricular outflow tract reconstruction is completed by anastomosing the proximal conduit to the infundibulotomy.

After separation from cardiopulmonary bypass, aortic, pulmonary arterial, and atrial pressures are measured continuously. Transesophageal echocardiography is performed to ensure that there are no significant residual defects. Bilateral pleural and mediastinal tube drains are placed and the sternum is closed. If bleeding or ventilation is an issue, the sternum is left open electively and the skin incision closed with a Silastic patch. The sternum is then closed on the second or third postoperative day.

Follow-Up and Reintervention

Completely repaired patients are followed clinically and scheduled for cardiac catheterization approximately 1 year after surgery or earlier if symptoms develop. Patients who do not undergo closure of the ventricular septal defect at the time of unifocalization are catheterized electively 3 months postoperatively to assess the feasibility of complete repair. At the time of this catheterization, our colleagues in interventional cardiology will often dilate proximal and/or distal stenoses in the unifocalized neopulmonary arteries. If the pulmonary:systemic blood flow ratio is >2:1, the ventricular septal defect is closed, sometimes with additional augmentation of the pulmonary arteries. If the ratio is <2:1, the patient will typically undergo either unilateral or bilateral patch augmentation of the pulmonary arteries.

UCSF Experience

Between July 1992 and September 1997, 72 patients with tetralogy of Fallot and MAPCAs were referred for surgery. Thirteen patients had undergone prior palliative or staging procedures elsewhere, and 5 were deemed inoperable at other centers with programs for managing this lesion. None of the patients referred for surgery were refused. Median age at surgery was 7.3 months (range 2 weeks to 37 years), and 65% of patients were <1 year of age. The number of MAPCAs ranged from 1 to 7 (median 4), and 20 patients had absent or rudimentary (<2 mm) central pulmonary arteries noted at the time of surgery.

Complete unifocalization of the pulmonary blood supply was achieved in one stage through a midline approach in 67 patients (93%), whereas 5 patients with severe distal segmental stenoses or significant comorbid conditions (such as sepsis, biliary atresia, or profound cyanosis and recent treatment with a continuous neosynephrine infusion) were managed with staged unifocalization through bilateral thoracotomies during the same hospitalization. Among the 67 patients who underwent one-stage unifocalization, the ventricular septal defect was closed during the same operation in 46. Therefore, 64% of patients referred for surgery underwent complete unifocalization and repair in a single stage. Among infants, patterns of unifocalization and repair were almost identical. There were 8 early deaths (11%), including 5 children. At follow-up, ranging from 1 to 61 months (median 18 months), there were 6 late deaths and 14 additional patients underwent completion of the repair. Nine patients, including 3 of the 5 who underwent staged unifocalization, are awaiting closure of the ventricular septal defect at a median of 7 months after the initial unifocalization procedure. Actuarial survival is 83% at 1 year and 79% at 2 years and beyond. Among early survivors, actuarial percentage with complete repair is 87% at 1 year and 95% at 2 years.

Conclusion

Despite tremendous conceptual and technical advances in the management of tetralogy of Fallot with MAPCAs over the past two decades, this remains a challenging lesion. Our approach to management is based on the principle of one-stage complex unifocalization early in life, preferably by 6 months of age, with maximal use of native tissue in the reconstruction. We have found this approach to be applicable in almost all patients, even those with absent true pulmonary arteries, and to yield excellent functional results. Nevertheless, there remains room for improvement because actuarial survival 2 years postoperatively is 79%. These results must be appreciated within the context of the natural history of this lesion, according to which an estimated 65% of patients survive to 1 year of age and slightly over 50% survive to 2 years regardless of surgical invention [1]. As other centers begin adopting similar aggressive approaches to early complete repair [22, 37], we hope to see the long-term prospects improve for patients with this difficult lesion.

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