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Review

Considerations in Timing of Surgical Repair in Tetralogy of Fallot

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ABSTRACT

Certain aspects of the treatment of tetralogy of Fallot (TOF) repair remain controversial. The optimal timing of the elective repair of asymptomatic patients and the ideal strategy for managing symptomatic neonates and infants with TOF are still debated despite years of experience in TOF treatment. In this article, we discuss why a surgical correction at 3-6 months of age is likely the ideal time frame for the elective repair of TOF. We also elaborate on our strategy for managing symptomatic neonates and infants with TOF and why we prefer an early single-stage primary repair.

RÉSUMÉ

Certains aspects du traitement de la tétralogie de Fallot (TF) par correction chirurgicale demeurent controversés. Le moment le plus approprié pour l'intervention non urgente chez les patients qui ne présentent pas de symptômes et la meilleure stratégie de prise en charge des nouveau-nés et des nourrissons atteints de TF symptomatique font encore l'objet de débats, même après de nombreuses années d'expérience dans le traitement de la TF. Dans le présent article, nous expliquons pourquoi la période de 3 à 6 mois est probablement idéale pour réaliser une correction chirurgicale non urgente de la TF. Nous expliquons également notre stratégie de prise en charge des nouveau-nés et des nourrissons qui présentent une TF symptomatique et les raisons pour lesquelles nous préférons réaliser une correction primaire en une seule étape à un âge précoce.

Tetralogy of Fallot (TOF) is a group of congenital cardiac malformations with biventricular atrioventricular connections characterized by the anterosuperior deviation of the conal septum or its fibrous remnant resulting in narrowing or atresia of the pulmonary outflow, a ventricular septal defect of the anterior malalignment type, a biventricular origin of the aorta (over-riding aorta), and most often right ventricular (RV) hypertrophy.[1,2] TOF was first described by Neils Stenson in 1671, later by Etienne-Louis Arthur Fallot in 1888, and subsequently termed "tetralogy of Fallot" by Maude Abbott in 1924.[3] It accounts for approximately 7% of congenital heart disease diagnoses and is encountered in approximately 4 per 10,000 live births.[4,5]

Despite 7 decades of surgical experience with the treatment of TOF, controversies in the management of this malformation remain. In this review article, we discuss the optimal timing of repair in asymptomatic patients and our preferred surgical approach for the repair of symptomatic neonates and infants with TOF with pulmonary stenosis.

Morphology of Tetralogy of Fallot

TOF encompasses a wide spectrum of malformations from TOF with pulmonary stenosis to TOF with pulmonary atresia. This article mainly focuses on the timing of the repair of TOF with pulmonary stenosis. The hallmark clinical sign of TOF is cyanosis, the degree of which is contingent on the degree of the RV outflow obstruction. This produces a wide range of clinical presentations from severe cyanosis at birth with ductal-dependent pulmonary circulation to patients who are asymptomatic and acyanotic at rest with only mild cyanosis during exertion.[6]

Early History of TOF Repair

The goal of TOF repair includes the elimination of intracardiac shunting by closure of the anterior malalignment ventricular septal defect and relief of RV outflow tract obstruction ideally with preserving the pulmonary valve and protection of normal myocardial function.[6] The first surgical treatment of TOF was done using a palliative shunt

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(subclavian to pulmonary artery) by Blalock and Taussig in 1945,[7] whereas the first surgical corrective intracardiac repair of TOF was performed in 1954 by Lillehei et al.[8] using cross-circulation and later in 1955 by Kirklin et al.[9] using a pump-oxygenator. During the earlier years of corrective cardiac surgery, the higher mortality rates encountered with early repair lead to the adoption of a 2-stage repair approach of TOF in which an initial palliative shunt was performed followed by a definitive repair in early childhood. Later experiences led to adopting a routine 1-stage repair,[10] and further advancements permitted repair during infancy[11] and extended the concept of a 1-stage complete repair of TOF to the symptomatic neonate with excellent results.[12]

Pulmonary Insufficiency

Iatrogenic pulmonary insufficiency after TOF repair was thought to be a benign well-tolerated lesion with no significant effect on the patient outcome, and the use of transannular patch (TAP) reconstruction of the RV outflow tract was prevalent, with some series suggesting an uncommon need for late reoperation due to its use.[13] A growing body of literature has shown that pulmonary insufficiency after TOF repair is indeed not benign, with detrimental long-term effects on RV size and function.[14–16] This has become more evident with the increasing late reoperations to address these late effects of pulmonary insufficiency[17,18] and has also led to the adoption of different techniques of pulmonary valve preservation.[18–20] In particular, greater efforts are being pursued by many surgeons to maintain some pulmonary valve function or avoid TAP reconstruction at the time of initial TOF repair (including intraoperative balloon dilation of the pulmonary valve and augmentation of pulmonary valve leaflets)[19–22] as it has been shown that pulmonary valve annulus–sparing repair has a reduced need for late reintervention in early adulthood.[23] Although the reported long-term outcomes are superior when annulus preservation is performed, this is not always possible, and thus, it is likely that the morphology and degree of pathology may dictate the type of intervention or even the timing of intervention rather than the surgeon's preference.[24] In our opinion, the preservation of dysplastic pulmonary valves with small annuli may lead to a bad combination of significant residual obstruction and regurgitation.

Primary vs 2-Stage Repair

Most patients with TOF are asymptomatic in the neonatal period and will undergo primary repair between 3 and 6 months. Those who are symptomatic may be treated with either primary repair or initial palliation (surgical or transcatheter) followed by delayed repair. Practices vary depending on the centre and surgeon-specific preferences and expertise as both strategies have advantages and disadvantages as demonstrated in Table I.

In a 2010 paper looking at North American patterns of management of TOF with pulmonary stenosis using the Society of Thoracic Surgeons (STS) database, data for 3000 patients operated on between 2002 and 2007 were analysed to reveal that primary repair in the first year of life (most between 3 and 6 months of age) without previous palliation was the most prevalent strategy. The most common type of TOF repair without previous operation was infundibulotomy with a TAP (1329 patients). Only 294 patients had initial palliation before TOF repair mostly in the form of a modified Blalock-Taussig-Thomas shunt (224 patients). Infundibulotomy with a TAP was also the most common type of repair in patients with TOF repair after a previous cardiac operation.[25]

Palliative procedures in the symptomatic neonate with TOF have been abandoned by some groups in favour of early primary repair.[26] Initial palliation of patients with TOF by a modified Blalock-Taussig-Thomas shunt before repair has been associated with morbidity and interval mortality. Morbidities include pulmonary artery stenosis and distortion, smaller pulmonary arteries before repair after neonatal palliation, limited lifespan of the shunt, and increased incidence of late sudden death.[27,28]

It is important to consider the cumulative results of a staged repair approach (initial palliative procedure, interstage interval, and the eventual corrective surgery) when compared with a single-stage primary repair approach. Furthermore, the comparison could be biased by the fact that many centres favour a staged approach for high-risk patients. With that in mind, the reported mortality rates between the 2 strategies appear to be similar.[29,30] An increased early mortality rate in the single-stage primary neonatal repair patient population was noted in some trials, but the overall cumulative mortality rate was similar to the staged repair approach in 5 years' time,[31,32] whereas some reported increased early mortality and worsened long-term survival with a staged repair approach.[33] The risk of reintervention, however, was higher in the staged approach even when not considering the complete repair as an intervention in both surgical and catheter-based palliation.[31,34]

Despite reports of increased pulmonary annulus and branch pulmonary artery sizes after initial palliation before TOF repair,[35,36] the need for employment of a TAP may not be affected with a staged approach.[25,37] The data reported from the 2010 study looking at the patterns of TOF management using the STS database revealed an approximately 54% use of a TAP in TOF repair without a previous operation, compared with a 74% rate of TAP usage in TOF repair after a previous operation.[25] It has been observed that palliated patients tend to have more TAP procedures (83% vs 48%),[38] and it is difficult to discern whether the higher rate of non–valve-sparing TOF repairs is in part related to palliation or the possible inherent bias of palliating patients with more severe forms of TOF with pulmonary stenosis.

The rates of pulmonary valve–sparing repairs vary in the literature with some institutions reporting pulmonary valve–sparing rates of 42% in their staged repairs (compared with 74% in their single-stage complete elective repair of TOF).[36] Others have reported lower (pulmonary valve–sparing rate of 17% in staged TOF repair)[38] and higher rates (pulmonary valve–sparing rate of 60% in staged TOF repair).[39] Even though initial palliative shunting may have an uncertain effect on the rates of pulmonary valve preservation, some groups still advocate for a staged repair approach in the neonatal period to avoid the higher rates of transannular patching associated with a neonatal repair.[40] However, it is likely that a patient with a more severe form of TOF with pulmonary stenosis that would require transannular patching would present earlier. By that logic, a neonatal repair of TOF (that has been associated with

TABLE I. Comparison of advantages and disadvantages between a primary neonatal repair and staged repair strategy in neonates with tetralogy of Fallot

Primary neonatal repair[12,25,27,49,50,69,70]	Initial palliation with delayed repair[29,36,45]
Advantages	
Reduction of duration of cyanosis with earlier resumption of normal physiology	Reduction of exposure to cardiopulmonary bypass and its complications in the neonatal period
Avoiding palliative procedures	Reduction of early morbidity and mortality associated with neonatal repair
Eliminating interstage morbidity and mortality	Possible reduction of the need of transannular patch
Improved pulmonary artery growth and avoidance of pulmonary artery stenosis and distortions associated with palliative procedures	
Disadvantages	
Greater early morbidity	Shunt-related morbidity (thrombosis and stenosis)
Potentially greater initial mortality for patients with high-risk features	Persistent cyanosis and right ventricular hypertrophy
	Combined mortality of initial palliation, interstage period, and delayed repair

higher rates of transannular patching) would require a non-pulmonary valve–sparing repair by virtue of the disease morphology itself and not by the age at which the repair was performed. The observed greater increase in the pulmonary artery size associated with earlier relief of RV obstruction (as evidenced with a significantly greater increase in the Nakata index in a neonatal repair of TOF)[41] further makes the case for a neonatal single-stage repair approach.

When looking at the interstage growth of the pulmonary arteries (pre-shunt size vs pre-complete repair at 4-12 months of age in millimetres by echocardiographic measurements) following a palliative procedure (mostly a modified Blalock-Taussig-Thomas shunt) in TOF with pulmonary stenosis, although the absolute value in millimetres was found to be significantly increased, most growth occurred in the branch and main pulmonary arteries rather than the pulmonary "annulus" itself, and the *z*-score of the "annulus" remained mostly unchanged after the palliative shunt.[36] Despite other papers demonstrating different results showing growth of the pulmonary valve "annulus," it is difficult to say with certainty if the observed growth is a direct result of the initial palliative procedure or a natural history growth of the patient afflicted with TOF. In one paper that investigated pulmonary "annular" growth with the use of a Blalock-Taussig-Thomas shunt in patients with TOF and double-outlet ventricle, significant *z*-score growth of the pulmonary "annulus" was demonstrated (pulmonary annulus size *z*-score increased from -5.1 ± 2.5 to -2.8 ± 2.1). It is worth noting, however, that even though growth was observed in the pulmonary shunt group, there was no significant difference between pulmonary "annular" growth seen in the shunt group and the "annular" size in the primary repair group before the intracardiac repair.[42] Other important points to consider about the interval growth with a systemic to pulmonary artery shunt are that the observed growth of the pulmonary arterial tree is not uniform as some trials have shown relatively unchanged *z*-score sizes of the pulmonary annulus and main pulmonary artery with a systemic to pulmonary shunt despite the absolute increase in diameters,[43] and despite some trials showing facilitation of significant *z*-score growth with an initial palliative shunt of the pulmonary valve "annulus," preservation of the pulmonary valve may still not be achievable with a staged approach if the initial pulmonary valve annulus is small.[37] It is unknown if the interval pulmonary "annular" increase is a direct result of the palliative shunt, or whether the palliative shunt allows time for growth to occur as the child grows as mentioned previously. Furthermore, no correlation was found between the magnitude of size increase and the length of time the shunt existed.[44]

Primary corrective surgery for TOF with pulmonary stenosis is considered the standard of care in many institutions with excellent outcomes, whereas staging with initial palliation is reserved only for infants with high-risk features (low birth weight, prematurity, poor anatomy of the pulmonary arteries, and noncardiac comorbidities) to bridge to complete repair.[45] The primary repair of symptomatic patients with TOF presenting at an age younger than 3 months has been shown to be safe with good outcomes and acceptable morbidity and is the preferred approach by some groups.[46] There are however reports of achieving good outcomes with a 2-stage strategy for a non-neonatal repair for symptomatic patients with TOF,[47] and in a recently published consensus document,[48] both strategies (initial palliative procedure or complete surgical correction) were considered reasonable approaches for the symptomatic neonate with TOF who has a standard risk, and palliation in the case of high-risk patients with a preference for consideration of a transcatheter palliative intervention in lieu of a palliative surgical procedure in the high-risk group (high-risk features being low-birth-weight patients/prematurity, small or discontinuous pulmonary arteries, and other major genetic or extracardiac congenital anomalies or other comorbidities such as intracranial haemorrhage, sepsis, or other end-organ compromise). Many studies combine asymptomatic and symptomatic patients with TOF, but in asymptomatic patients, there is a higher risk of morbidity when the repair is performed in less than 3 months of age. Patients with TOF at the severe end of the spectrum with severe RV outflow tract obstruction are more likely to undergo earlier repair and are more likely to require a TAP. They may have a more complicated postoperative course, thus demonstrating an association rather than a causality between the age of repair and the observed outcomes (pathology dictating outcome rather than patient age at repair). This raises the notion that we may not be able to resolve whether one treatment strategy is better than the other (primary vs staged repaired) until a comprehensive TOF anatomic and physiological preoperative scoring system is developed to be able to reliably compare and analyse the outcomes of surgical interventions in these patients.

In regards to managing symptomatic neonatal patients with TOF, there are concerns of potentially increased operative mortality when comparing primary repair and palliation in the neonatal period. In the 2010 STS database paper, a higher mortality was reported for neonatal TOF repair (7.8% mortality for neonatal repair compared with 1.6% mortality

for repair performed at 1-3 months of age).[25] This neonatal repair group could have included patients with high-risk features that we advocate a staged repair strategy for. But the pertinent comparison should not be between neonatal repair vs repair performed at a later age, as these 2 groups may not be comparable. One needs to compare primary neonatal repair vs neonatal palliation and delayed repair. It is important to take into consideration that the reported palliation mortality at a neonatal age is also increased at 6.2% and that it does not include the cumulative additional mortality of the interstage period and the second-stage corrective procedure.

Neonatal TOF repair was reported to be associated with higher rates of mortality, more postoperative complications, longer hospital stays, and higher hospitalization cost in a study using data from a large national database (over 6800 patients, 8% of them undergoing neonatal repair).[49] An important caveat of this paper is that there is no distinction of the TOF morphology because the data are collected mainly for medical coding and billing. Potentially, the higher neonatal mortality could be in part related to more RV outflow reconstruction in comparison with postnatal repair. The neonatal patient group in comparison with the postnatal group did have significantly more prematurity (14% vs 1.1%), more extracardiac anomalies (43% vs 22%), and more genetic or chromosomal disorders (21% vs 17%).

In another observational study involving over 4500 patients with TOF (approximately 200 of whom underwent neonatal repair), neonatal TOF repair was associated with significantly higher postoperative in-hospital morbidity and mortality (6.4% mortality in neonatal repair vs 1.9% mortality in 31- to 90-day repair).[50] This paper aimed at identifying an elective primary complete repair of asymptomatic infants with TOF by excluding infants with pulmonary atresia, prostaglandin use, and prior shunt palliation, and emergency repairs by excluding patients with those codes from the analysis. It is worth noting that 3 of the 4 centres that had experience with performing neonatal repairs did not experience any neonatal TOF repair morality (although the morbidity was similar using a by-centre analysis). Moreover, important variables (prematurity, weight, degree of subpulmonary stenosis, pulmonary valve annulus size, and coronary and pulmonary artery morphology, TAP placement) were not reported.

In a paper comparing primary vs staged repair for symptomatic neonates with TOF weighing less than 2.5 kg, mortality and reintervention were independent of the treatment strategy. Death occurred in 12 patients (15.8%) in the staged repair group and in 8 patients (18.2%) in the primary repair group ($P = .735$). Early morbidity favoured staged repair, whereas cumulative morbidity favoured primary repair.[51]

Palliation Options

Different surgical and catheter-based options are available if an initial palliative procedure is decided on to stage the repair of TOF. An expert panel provided a weak recommendation for consideration of a transcatheter intervention for palliation over a surgical intervention in high-risk symptomatic neonates with TOF in whom a palliative strategy has been chosen.[48] A transcatheter palliative option that is considered in lieu of a surgical one (typically a modified Blalock-Taussig-Thomas shunt) should take into account the expertise of the treating institution and the implications of the palliative procedure on the future surgical correction. Transcatheter placement of an RV outflow stent, for example, had been shown to be a safe palliative procedure to defer corrective surgical repair in patients with TOF with high-risk features by relieving cyanosis and allowing pulmonary artery and somatic growth that is comparable to early surgical repair in standard-risk patients.[45] However, placing an RV outflow stent could commit the patient to a TAP.[52]

Transcatheter insertion of a ductal stent has been shown to be an appropriate alternative to surgical palliation for patients with ductal-dependent pulmonary blood flow with equal to superior outcomes to surgical shunts and success rates ranging from 80% to almost 100%.[53] Ductal stenting is reported to promote a more global pulmonary arterial growth with more balanced development and less procedure-related complications and distal branch pulmonary artery stenosis compared with a modified Blalock-Taussig-Thomas shunt procedure.[54–56] The individual patient's anatomy has to be considered (eg, high risk of ductal stent protruding into the pulmonary artery end and complicating the surgical correction procedure) as well as the expertise of the treating institution when deciding about the optimal palliative procedure for the patient.

Early Primary Repair of TOF

Excellent results have been demonstrated and reported with the early primary repair of TOF while avoiding the aforementioned morbidities associated with an initial palliative procedure.[57] In a paper looking at 155 patients with TOF with pulmonary stenosis who underwent early primary repair (median age of 75 days) without initial palliation, multivariate lineal regression analysis revealed that a younger age at the time of the procedure was not associated with increased ventilation time, intensive care unit stay, hospital stay, or increased hospital charges.[58] Using a technical performance score, younger age at TOF repair, and smaller patient size has also been shown not to affect the adequacy of technical repair.[59]

In an analysis of intermediate-term outcomes of elective early TOF repair, an age cutoff of 55 days (>55 days at repair regardless of size) was found to be safe for performing the procedure without any increased risk of reintervention rates, whereas a younger age (<55 days) was an independent predictor of overall intermediate-term reintervention. They hypothesize that the 55 days' cutoff might be related to continued evolution of the ductal tissue in the pulmonary artery end.[60] A systematic review of the literature from 1950 to 2008 identified 8 papers representing the best evidence to answer the question of whether early correction of TOF (<6 months) is comparable to surgery after 6 months of age. They concluded that the early primary repair of TOF is comparable to the later repair with similar freedom from reintervention and mortality with the observation that in patients who are repaired at an age younger than 3 months, the length of intensive care unit stay, the period of mechanical ventilation, and the need for inotropes are increased.[61]

In a paper looking at the optimal timing of TOF repair,[26] an age of <3 months at TOF repair had good outcomes but was found to be an independent risk factor for prolonged time

to lactate clearance, longer intubation, and increased total hospital stay, whereas infants aged 3-11 months were found to have the most rapid recovery from operative therapy (even compared with infants >12 months of age). Children >12 months at the time of repair had the least favourable outcomes with significantly higher hospital deaths. As a result, the authors of the paper concluded that an age of 3-11 months is the optimal age for elective TOF repair based on observed mortality and physiological outcomes.[26]

Similar findings were also reported in a retrospective multicentre study for a younger age of repair during the primary surgical correction of TOF; an age of <3 months was found to be significantly associated with postoperative complications and adverse events at follow-up.[62] Longer hospital stay and longer support times were also reported for patients younger than 3 months of age in a paper looking at a single-centre elective TOF repair with 277 patients younger than 6 months of age.[63] TOF correction at age <2 months was also found to be an independent predictor for reintervention.[64] Other authors echoed these findings for TOF correction at <3 months of age.[65,66] In a paper looking at over a thousand patients with TOF repaired during their infancy (<1 year of age), when comparing patients aged <1 month with older patients (1 month to 3 months), longer length of stay (28.9 ± 3.3 days vs 16.4 ± 1.7 days) and related interventions (18.92% vs 4.92%) were noted in the neonatal group.[66] Older age of repair (>3 years) should also be avoided as it has been associated with negative effects on late mortality related to prolonged exposure to the effects of hypoxia and right ventricle overload.[67] Achieving a good TOF repair during early infancy should not be precluded by the presence of coronary anomalies with proper modifications of the surgical technique.[68]

Repair at 3-6 Months vs 6-12 Months

Multiple publications have shown the feasibility of elective repair at 3-6 months compared with repair at 6-12 months with good outcomes in both; it has been noted, however, that TAP usage may be higher in older patients (>6 months) than younger patients (<6 months). This is possibly explained by the progressive right ventricle hypertrophy and the reduced pulmonary artery development with longer-standing RV outflow obstruction leading to a more aggressive nonpulmonary valve—sparing relief of RV outflow obstruction at the time of complete repair.[38] In a paper involving over a 1000 infants with the complete repair of TOF between 2005 and 2011 who were divided into groups based on their age (<1 month, 1-3 months, 3-6 months, and 6-12 months), the patients in the younger age group (<1 month) were found to have the highest risk of postoperative complications and longest hospital stay. When comparing the 3-6 months and the 6-12 months groups, similar hospital length of stay, rates of intervention (catheterization, extracorporeal membrane oxygenation, device implantation, and chest and pericardial drainage), blood transfusion, and postoperative complications were noted.[66] The recently published consensus document from the American Association for Thoracic Surgery reviewed 163 publications from the year 2000 or later and reached a consensus recommendation for elective complete surgical correction of asymptomatic TOF infants to be performed between 3 and 6 months of age to reduce the length of stay, rate of adverse events, and the need for a TAP.[48]

The authors' management strategy to the symptomatic neonate with TOF is simple and consists of primary repair. Asymptomatic patients are repaired between 3 and 6 months.

Comment

For over 30 years, we have been proponents of the primary repair of TOF. We currently advocate repair between 3 and 6 months of age in the asymptomatic patient or earlier if the patient is symptomatic. We do not palliate patients who present as neonates without high-risk features and opt for a neonatal complete repair of TOF without first-stage palliation. We are not convinced that a palliative procedure would greatly impact the decision to use a TAP. Patients who have a small enough "annulus" to warrant a non—valve-sparing repair, whether they present in the neonatal period or later for elective repair, would, in all likelihood, still require a TAP whether they are palliated with a shunt or not.

We hypothesize that pulmonary valve annulus growth would lag behind the patient's somatic growth, resulting in a smaller annulus size relative to the patient size and decreasing the ability of pulmonary valve preservation.

Conclusions

The optimal age for the elective repair of TOF with pulmonary stenosis appears to be between 3 and 6 months, closer to 3 months if pulmonary valve preservation is likely. Primary neonatal or early repair is our preferred approach for the treatment of symptomatic infants with TOF. Initial palliation (surgical or interventional) has a role for symptomatic neonates with TOF with high-risk features in mitigating the initial vulnerable period, but its role in improving the rates of pulmonary valve—sparing repair in TOF surgical correction is uncertain.

Ethics Statement

The research reported has adhered to the relevant ethical guidelines.

Patient Consent

The authors confirm that patient consent is not applicable to this article. This is a review article of the literature therefore did not require consent from the patients.

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