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Review

Neonatal complete correction of tetralogy of Fallot versus shunting and deferred repair: is the future of the right ventriculo-arterial junction at stake, and what of it?

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THE OPTIMAL SURGICAL APPROACH AND TIMING for repair of tetralogy of Fallot remains one of the more controversial issues in the treatment of congenital cardiac malformations. Central to the controversy is the fate of the entire right ventriculo-arterial junction, and particularly that of the pulmonary valve. Historically, the approach to patients with tetralogy and most cyanotic cardiac diseases has been a staged one, involving an initial construction of a palliative systemic-to-pulmonary arterial shunt, followed by takedown of the shunt and complete repair at an older age. This strategy involves at least two operations, but appears to result in a lesser need for transjunctional patches,[1,2] and a higher percentage of patients keeping their native functioning pulmonary valve. Owing to advances in surgical technique, perfusion technology, and peri-operative care, the general trend in recent years has been to perform one-stage complete repair of most congenital defects in the neonatal period, including tetralogy of Fallot. Theoretical advantages include the need for only one operation, a shorter interval of right-to-left shunting with the deleterious effects of cyanosis and polycythemia, diminished right ventricular hypertrophy, and no left ventricular volume overload during the interval following construction of a systemic-to-pulmonary arterial shunt.

Although the immediate results of both approaches are encouraging, the mid and long-term outcomes are often marred by progressive failure and insufficiency of the pulmonary valve. Clear-cut surgical indications may pertain to certain anatomic and physiologic situations, whereby either approach is justified or, on the contrary, unnecessary, but the gray zone is ever expanding. In other words, what is the best approach to asymptomatic patients? How should we now deal with the "pink Fallot's", diagnosed early in the neonatal period? Hard evidence in favor of one or the other approach is difficult to find, and more importantly, difficult to define. Beyond the functional aspects of the pulmonary valve, is there really a difference for the patient in a broader and long-term sense? The central aspect of the pulmonary valve may have multiple consequences on the entire right heart, ranging from dilation and failure of the right ventricle, tricuspid valvar insufficiency with resultant right atrial dilation and atrial arrhythmias, pulmonary arterial aneurysm, and ventricular arrhythmias. But which approach, staged or early complete repair, leads to better quality of life, less health-related hazard, and what are the implications in decision making for the physicians caring for these patients? If the overall outcome of a surgical modality is to be judged on the basis of freedom from continuous medical treatment, or the need for reoperation, rather than on operative mortality, does one or the other surgical strategy lead to an overall better outcome?

With these questions in mind, we present an overview of the various surgical results in the treatment of tetralogy of Fallot, taking into account the long-term outcome and surgical implications of each approach.

Historical background

The first surgical treatment for tetralogy of Fallot was performed by Blalock and Taussig[3] in 1945. They attached the left subclavian artery in end-to-side fashion to the left pulmonary artery, thereby beginning a

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new era in the palliation of cyanotic heart defects. A number of other palliative systemic-to-pulmonary shunts soon developed, including those described by Potts in 1946,[4] Waterston in 1962,[5] and de Leval in 1981,[6] to name but a few. Direct relief of pulmonary stenosis by closed valvotomy had been introduced by Brock[7] in 1948. The first successful complete correction was performed by Lillehei and Varco[8] in 1954, using "controlled cross-circulation", with another human being as the oxygenator. The first successful repair using a pump oxygenator took place in 1955, and was carried out at the Mayo Clinic by Kirklin[9]. Warden and Lillehei[10] were the first, in 1957, to insert a patch to enlarge the right ventricular outflow tract, and placement of the patch across the ventriculo-pulmonary junction, the so-called "annulus", was first reported in 1959 by Kirklin.[11] The use of a conduit placed between the right ventricle and the pulmonary arteries was introduced by Rastelli and Kirklin[12] in 1965, and insertion of a valved aortic allograft to reconstruct the outflow tract was introduced by Ross and Somerville in 1966.[13]

The staged approach

Despite the current trend for complete neonatal correction, many centers still prefer a staged approach, involving an initial construction of a systemic-to-pulmonary arterial shunt, followed by deferred complete repair. As the Waterston and Potts shunts have generally been abandoned by most surgical centers, we will not dwell on their increased morbidity and mortality, nor the relative difficulty encountered in their takedown prior to complete correction. When considering palliative shunts, we will confine our attention to the more commonly used modified Blalock-Taussig and central shunts.

First and foremost, it is important to emphasize the anatomical and clinical situations where early complete correction is undesirable or unfeasible. These include generalized critical illness, hypoplastic pulmonary arteries, peripheral pulmonary arterial stenosis, prematurity with worsening cyanotic spells, and coronary arterial anomalies characterized by passage of a prominent arterial branch across the right ventriculo-arterial junction.[14,15] Other reasons are more institutional or surgeon-dependent, and may include potentially lower rates of morbidity and mortality, a preference to avoid placement of conduits from the right ventricle to the pulmonary arteries that will invariably lead to multiple reoperations,[16] initially diminished periods of time in both intensive care and hospital, as well as cost considerations per hospital stay.[17]

The mortality of the initial palliative procedure when the modified Blalock-Taussig shunt is chosen ranges between zero and 4%.[1,2,18–20] To this must be added the mortality which ensues during the palliated state before deferred repair. This may account for an additional 5–8% of attrition, which may or may not be shunt-related.[2,21]

Additional concerns in relation to construction of the modified Blalock-Taussig shunt include a 35–65%[19,21,22] incidence of iatrogenic deformation of the pulmonary arteries at the site of construction of the shunt. In their series of 141 children, Vobecky et al.[2] found that the shunt failed in one quarter, and half of those undergoing complete repair required pulmonary arterioplasty at the site of insertion to the pulmonary arteries. There is evidence suggesting a higher incidence in pulmonary arterial deformation when the shunt is performed at an earlier age,[23] particularly in neonates.[21] Nonetheless, in a series of 49 patients with diminished flow of blood to the lungs who underwent palliation at a median age of 14 days, Bove et al.[19] reported no pulmonary arterial distortion or hypertension. Their early and late mortality rates were 4% and 9%, respectively. Univariate analysis revealed age below 3 months, and weight less than 3.6 kg, to be risk factors for early failure, defined as complete occlusion of the shunt, progressive decrease in oxygen saturation, progressive increase in hemoglobin, need for a second shunt, or complete non-elective correction.[19]

Unequal flow between the two lungs, kinking, stenosis, and even occlusion of the pulmonary arteries after asymmetric construction of shunts, have led some authors to prefer the central shunt between the aorta and the pulmonary trunk.[24–26] Potapov et al.[26] reported on a series of 22 neonates with diminished pulmonary flow in which a central shunt was constructed with an operative mortality of 13.6%. They found no difference in growth of the arteries supplying the lower lobes of the right and left lungs, no incidence of pulmonary edema or pulmonary hypertension from shunt overflow, and a lesser degree of pulmonary arterial deformation on follow-up angiography. They proposed the central shunt as the procedure of choice in newborns with diminished flow of blood to the lungs, since this procedure avoids unwanted manipulation of the right and left pulmonary arteries, more often spares cardiopulmonary bypass, and allows easier access for takedown of the shunt before complete correction.[26] Their reported mortality, nonetheless, is disturbing.

After initial palliation, the median age and weight of patients are obviously greater at the time of complete deferred repair, but is the need for transjunctional incisions decreased? Earlier studies revealed the need for transjunctional patches in about two-thirds of children undergoing repair below the age of 12 months.[1,15] Upon analysis of the various sites of

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obstruction to flow of blood to the lungs according to age, Pozzi et al.[14] found a constant incidence of obstruction at the infundibular level, but a decreasing degree of hypoplasia of the right ventriculo-arterial junction as the patients got older. In their series, this lead to a lesser incidence of transjunctional incisions in the older patients.[14]

Beyond the right ventriculo-arterial junction, there remain other sources of concern in the interval between palliation and deferred repair. There is evidence that right ventricular hypertrophy increases with age,[14,27,28] which may imply permanent damage to the myocardium, with longer standing obstruction to the right ventricular outflow. Pozzi and his colleagues[14] reported an increase in fibrosis of the right ventricular outflow with age, found in two-fifths of patients undergoing surgery below the age of 6 months of age, compared to nearly four-fifths of those submitted to complete repair beyond 2 years of age. This finding was shown to be statistically significant ($p < 0.04$). Whether an increase in fibrous tissue within the sub-pulmonary infundibulum correlates to a higher incidence of late ventricular arrhythmias and sudden death is speculative, although this mechanism has been suggested by others.[29] Volume overload of the left ventricle imposed by the shunt is another potential cause of deteriorating heart function, sometimes indicating the need for earlier complete correction.[30,31] The occurrence of hypoxic spells and loss of consciousness, the effect of chronic hypoxia and its secondary damaging effects on other organs, in particular the brain, is an argument in favor of removing the anatomic substrate for right-to-left shunting, in other words performing complete correction at an earlier age.[30,32] Reactive polycythemia with an increased hematocrit is another frequent indication for operation.[33] Recent series, nonetheless, have reported excellent results with the staged repairs, mostly in terms of operative rates of mortality (Table 1).

Complete early correction

Early complete repair, particularly during the neonatal period, may have several advantages over a staged approach, providing it can be performed with the same low rates of mortality and morbidity. These include prevention or avoidance of severe right ventricular hypertrophy and infundibular fibrosis, reduction in arrhythmias, and a more harmonious development of the pulmonary vasculature.[14] Furthermore, the interval with the deleterious effects of hypoxia on the brain, as well as cyanosis and secondary polycythemia, may be reduced. Controversially, it is argued that asymptomatic neonates with the morphology of "pink Fallot", who present in good general condition, have the lowest operative risk, and may be the optimal candidates for early correction, best to avoid the aforementioned sequels of deferred repair.[14,28]

It is a generalized, yet unproven, impression amongst various surgical institutions that complete repair at an earlier age, particularly in the neonatal period, may lead to a higher incidence of transjunctional incisions.[2,14,30,34] This is largely due to the perception that the dimensions of the pulmonary valve indexed to body surface index are lower at lower ages.[28,35] Pozzi et al.[14] needed transjunctional incisions in nine-tenths of their patients younger than 6 months, as compared to less than half of those older than 2 years of age, although this difference was not statistically significant. Di Donato et al.[30] similarly reported the need for transjunctional incisions in over nine-tenths of neonates. From the same institution, when reviewing a more recent series of repairs performed under 90 days of age, Pigula et al.[34] again found a need for transjunctional patches in more than four-fifths of the patients. Conversely, Ungerleider et al.[17] found only a need for transjunctional repair in only one-third of their series of young infants aged from 3 days to 9 months. Despite this trend, we must ask what consequences, if any, are there to be expected in the long term after incisions across the right ventriculo-arterial junction?

Transjunctional incisions potentially damage two cardiac structures, namely the anatomic right ventriculo-arterial junction, along with the leaflets of the pulmonary valve, and the right ventricular myocardium. Each leads to different pathophysiology, with dire long-term consequences of its own,

Table 1. Results of shunting and deferred repair.

Author	Shunt* (months)	Repair* (months)	Combined mortality (%)
Pozzi et al.[14] (2000); n = 132	2.9	23.2	0
Vobecky et al.[2] (1993); n = 237	–	37.1	7.6
Kirklin et al.[1] (1983); n = 112	–	74.5	1.8
Daily et al.[15] (1977); n = 33	4.3	–	5.7

* Indicates median age at time of surgery

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Table 2. Results of complete one-stage correction.

Author	Median age	Mortality (%)
Karl et al.[39] (1992)		
366 patients	15.3 months	0.5
Ungerleider et al.[17] (1997)		
18 patients	3.4 months	0
Pigula et al.[34] (1999)		
99 patients	27 days	3
Tchervenkov et al.[40] (2000)		
20 patients	5.5 months	0
Di Donato et al.[30] (1991)		
27 patients	3 days	18.5

both compromising late right ventricular function, and resulting in a higher risk of reoperation.[1,36] First, an incision across the right ventriculo-arterial junction will invariably lead to pulmonary regurgitation, progressing to a degree of severity that leads to right ventricular dilation and eventual failure. This may, in turn, produce tricuspid valvar regurgitation, with resultant right atrial dilation and atrial arrhythmias. Secondly, a large right ventriculotomy may produce immediate postoperative low cardiac output, and is incriminated in the long-term increased risk of ventricular arrhythmias and sudden death.[37,38]

Based on this evidence, a transatrial, or combined transatrial-transpulmonary approach, is deemed the ideal one for complete repair of tetralogy, avoiding in this way both the need for a transjunctional patch, placement of a conduit from the right ventricle to the pulmonary arteries, or a ventriculotomy. These approaches have given excellent results, as illustrated by the impressive series of 366 non-neonates reported by Karl et al.,[39] with an operative mortality of 0.5%, and an actuarial freedom from reoperation for any reason of 95% at 5 and 10 year follow-up. The results of other recent series are presented in Table 2.

Transjunctional incisions and valved homografts

Although intuitive, it remains unproven that transjunctional incisions lead to a higher incidence of reoperation for any reason pertaining to the right ventriculo-arterial junction. It is clear that transjunctional patching leads to a higher incidence and degree of long-standing pulmonary valvar insufficiency,[36,41] but whether, and/or when, this needs to be corrected by surgical re-intervention is still controversial. This disagreement arises from the difficulty in determining the presence or degree of right ventricular failure by current clinical and qualitative diagnostic tools.[42] More quantitative and reproducible diagnostic measures of right ventricular function, such as resonance imaging combined with dobutamine stress testing, or assessment of neurohormones in the plasma, may allow for better stratification of right ventricular dilation and impending failure, before the right ventricle becomes irreversibly damaged.[43]

Beyond the controversial issues concerning timing, it has been demonstrated that insertion of a pulmonary valve restores right ventricular function in patients with long-standing pulmonary valvar insufficiency after surgery for tetralogy of Fallot.[41,44,45] This is illustrated by regression of right ventricular dilation, a reduction in the incidence of atrial arrhythmias, and by improvement in exercise tolerance and functional class.[41,44,45] The risk of reoperation for pulmonary valvar replacement after repair, with or without other associated procedures such as closure of a residual septal defect, repair or replacement of the tricuspid valve, or repair of right ventricular or pulmonary arterial aneurysms, ranges between zero and 3%.[31,36,41,46]

For the reconstruction of the right ventriculo-arterial junction, mechanical prostheses or valved xenograft conduits have largely been abandoned, and most surgeons currently rely on valved allografts or homografts. Despite the favorable initial results in terms of valvar function and transvalvar gradients with homografts, there is increasing concern as to the long-term outcome and the need for reoperation. This may be required because of mismatch in the size of conduit and child with growth, or because of failure of the conduit. The current mortality of primary insertion of a homograft for any diagnosis is low, no more than 5 or 6%, and the risk of replacement of conduits has decreased in recent years to less than 3.5%.[16] Stark et al.[47] reported 84%, 58%, and 31% freedom from reoperation after replacement of homografts at 5, 10, and 15 years, respectively. They found that conduits used in reoperations fared worse than those implanted at an original operation, and that homografts inserted earlier in their series lasted longer. Niwaya et al.[48] reported a 90% freedom of failure of allografts at 5 years, and 82% after 8 years. Gerestein et al.[49] reported a similar freedom from reoperation for valve-related events, such as dysfunction of the homograft or endocarditis, of 91% and 87% at 5 and 8 years, respectively. They found the use of an aortic allograft ($p = 0.02$), and an extra-anatomic position of the allograft ($p = 0.03$) to be risk factors for accelerated failure of the graft.[49]

Despite their invaluable role in reconstruction of the right outflow, and in right ventricular remodeling, it is evident from the previously presented data that valved homograft conduits have a limited life expectancy. For this reason, many surgeons attempt to limit or avoid their use in smaller and younger patients, whereby the probability and number of reoperations would otherwise inevitably increase.

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Reoperations are required for failure of the conduit, or for mismatch relative to the size of the patient. As previously stated, it is strongly suggested by existing data that earlier complete repair increases the need for transjunctional incisions and patches.[1,2,14] If transjunctional patches are proven to increase the need for reoperation for insertion of valved homograft conduits, an argument could be made to discourage complete early correction, particularly in neonates.

There exist, however, anatomical situations in which it is difficult to avoid placing a conduit from the right ventricle to the pulmonary arteries, and where the use of a valved homograft should strongly be considered. These include an abnormal origin of the anterior descending coronary artery from the right coronary artery, or the finding of other major coronary arterial branches crossing the sub-pulmonary outflow tract; increased pulmonary arterial pressure/resistance, such as that present in patients with peripheral pulmonary arterial hypoplasia, or after a long-standing large aorto-pulmonary shunt; and right ventricular dilation and failure, which is usually associated with a severely regurgitant pulmonary valve.[16] Tchervenkov et al.[40] have recently reported a series of 20 infants with tetralogy or double outlet right ventricle in whom coronary arterial branches crossed the right outflow tract. In 18 of these patients, it proved possible to reconstruct the ventriculo-pulmonary junction without the use of a conduit, with no early or late mortality. During follow-up, only 2 reoperations were required after 8 and 11 years. They encourage the use of alternative surgical techniques to avoid the use of valved homografts in low-weight neonates and infants, and stress its feasibility even in patients with major coronary arterial branches crossing the sub-pulmonary outflow tract.[40]

Conclusions

The best approach to the repair of tetralogy of Fallot remains to be determined. In terms of early morbidity and mortality, both staged and early complete repairs give excellent results. Institutional and surgeon-based biases exist for either strategy, and ultimately are justifiable when they produce the best outcomes. Initial extreme presentation, anatomy and physiology, may in many instances give a clear-cut indication for one or the other surgical pathway. The management of asymptomatic pink Fallot patients, in contrast, where either solution may be considered at leisure, still provides for strong feelings and divided opinions.

Beyond considerations of rates of mortality, current standards of surgical outcome are increasingly set by the long-term results they produce. In the management of tetralogy of Fallot, these may be defined in terms of late functional status and exercise tolerance, right ventricular function, pulmonary and tricuspid valvar function, atrial and ventricular arrhythmias, the need for reoperation for any reason, and the general degree of morbidity, medical care, and close follow-up required. Accordingly, the fates of the right ventriculo-arterial junction and the pulmonary valve become paramount in regard to most of the long-term issues pertaining to the right heart.

It is intuitive that surgical repairs leading to a dysfunctional pulmonary valve, with resultant insufficiency, should be avoided when possible. In keeping with this philosophy, transjunctional incisions would be discouraged unless absolutely necessary. Current results of most series suggest that earlier complete correction, particularly in low-weight neonates, leads to an increased incidence of transjunctional incisions. Whether transjunctional incisions in themselves lead to a higher incidence of reoperation, most often for insertion of a competent pulmonary valve, remains to be proven. If so, the risks of multiple reoperations because of failure of the homograft, or mismatch in size between homograft and patient, not to mention the considerations of cost and the psychological implications for the patient, would strongly argue against repairs involving a transjunctional patch, and hence against neonatal complete correction. This needs to be weighed against the disadvantages of a staged approach, namely the definite need for two operations to achieve repair, the longer standing right ventricular hypertrophy and strain, cyanosis and polycythemia, the potential distortion of the pulmonary arteries at the site of insertion of the systemic-to-pulmonary arterial shunt, and the volume overload of the systemic ventricle during the palliated state.

Prospective randomized studies including both strategic arms, particularly in asymptomatic neonates and young infants, would give more insight as to the importance, or not, of maintaining a competent autologous pulmonary valve at initial complete correction. Knowledge of the long-term fate of the right ventriculo-arterial junction which would accrue from such studies would settle some of the remaining controversies as to the optimal timing and management of babies born with tetralogy of Fallot.

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