Surgical correction of tetralogy of Fallot has proven very successful in the short term, although from longer term follow up it is increasingly apparent that many patients are haemodynamically compromised and their condition has not been cured.

Tetralogy of Fallot (TOF) is a complex congenital cardiac abnormality representing approximately 10% of all cardiac malformations. It has a pathophysiology characterised by a ventricular septal defect (VSD) and right ventricular outflow tract obstruction (RVOTO) causing a right-to-left-shunt with low pulmonary blood flow and cyanosis. The extent of the RVOTO is highly variable and may include hypoplasia and dysplasia of the pulmonary valve as well as obstruction at the subvalvar and pulmonary artery level. The RVOTO is progressive, as is the compensatory right ventricular hypertrophy that adds to the obstruction. Without specific surgical management up to 35% of children will die within the first year of life, 50% by the third year, and only few will survive into adulthood. With this dismal natural history, interventions to change the outcome are much needed.

In 1944, TOF was the first congenital heart lesion to be palliated and 10 years later also the first complex cardiac lesion to undergo successful open repair. Over the last decades, advances in paediatric cardiology, surgery, and intensive care have transformed the outlook for patients with TOF. Currently in the UK, surgical correction carries a 30 day mortality of less then 3% (Central Cardiac Audit Database, personal communication, 2000–2001) and many operative survivors are expected to reach adulthood. Unfortunately, at long term follow up it is becoming increasingly clear that many patients are haemodynamically compromised and what was originally thought to be a definitive repair is in fact not a cure.

The principles of surgical correction of TOF involve closure of the VSD and relief of the RVOTO. To create an unobstructed way out of the right ventricle, pulmonary valvotomy, the insertion of an outflow tract patch or a transannular patch are often required. Consequently, most patients acquire pulmonary regurgitation as a result of the repair. While this is apparently well tolerated in the early postoperative years, it is now well established that in the long term chronic pulmonary regurgitation is associated with reduced exercise capacity, right ventricular dilatation, ventricular arrhythmias, and sudden death. Although recently the benefits of late pulmonary valve replacement for improving ventricular function and stabilising cardiac rhythm have been demonstrated, the indications and timing for this procedure remain to be defined.

**ACHIEVING PULMONARY COMPETENCE**

For many years cardiac surgeons have recognised the importance of achieving lasting pulmonary competence at the time of TOF repair, but various attempts to achieve this have so far failed. Construction of a biological or synthetic monocusp valve in the reconstructed right ventricular outflow tract proved disappointing because of early structural valve deterioration with subsequent pulmonary regurgitation. Also, the insertion of a pulmonary or aortic homograft at the time of repair provides only a temporary solution because of calcification and lack of growth of the homograft, leading to recurrent RVOTO. Lately, biological valve conduits have been marketed with the promise of “no calcification”, but clinical trials are still at an early stage.

If normal pulmonary valve function cannot be achieved, then the second best option may be to try to minimise the extent of the pulmonary regurgitation by accepting the smallest possible annulus size that still gives adequate right ventricular unloading. This option is explored by Uebing and colleagues in this issue of Heart. In a study of patients with a median follow up time of 4.8 years after TOF repair they confirmed the well known facts that more extensive pulmonary regurgitation is associated with a larger postoperative pulmonary annulus and right ventricular size. More interestingly, no correlation was found between the right ventricle/left ventricle systolic pressure index and pulmonary annulus diameter indexed for body surface area. Even for patients with a pulmonary annulus index close to the fourth lower standard deviation, the right ventricular pressure load was not significantly elevated. This suggests that in many patients the annulus was larger than strictly necessary for adequate right ventricular unloading. The authors recommended that at the time of surgical repair the pulmonary annulus should be enlarged to only the second lower standard deviation of normal.

A problem with the study by Uebing and colleagues is that a model of pulmonary incompetence and right ventricular dilation is used to define the minimum adequate pulmonary diameter for surgical correction. At the time of the

**Abbreviations:** RVOTO, right ventricular outflow tract obstruction; TOF, tetralogy of Fallot; VSD, ventricular septal defect
early redo surgery. On the other hand, the hypertrophied right
necessitate revision of the repair during the same operation or
worse by the use of inotropes. Residual RVOT is poorly
removal of the stimulus for right ventricular hypertrophy and
fibrosis, alleviation of systemic cyanosis and, in symptomatic
infants, avoidance of initial palliation with an arteriopulmo-
tary shunt. Recent series of patients operated on in the first
few months of life have shown excellent early and five year
survival of many complex cardiac abnormalities in neonates and
infants, there has been a move towards earlier primary correc-
tion of TOF. Arguments in support of this strategy include
infants, there is often moderate annulus hypoplasia with obstruction
at the level of the valve only. In these circumstances it is to be
expected that valvotomy with conservative annular extension
will provide adequate right ventricular unloading with little
pulmonary regurgitation. In contrast, patients with pulmo-
ary atresia type TOF often have a long segment infundibular
stenosis with a small, dysplastic pulmonary valve. In these
cases extensive infundibular resection and transannular
patching may be required. The extent to which the RVOT has
to be relieved may be difficult to judge in the paralysed heart
at the time of the operation, and residual obstruction may only
manifest itself when the heart has regained its tone. To com-
plicate matters further, this obstruction often has a dynamic
component to it, which diminishes over time but is made
worse by the use of inotropes. Residual RVOT is poorly
tolerated in the immediate postoperative period and may
necessitate revision of the repair during the same operation or
can easily be performed during the same operation. On the other hand, the hypertrophied right
ventricle copes well with pulmonary regurgitation. Therefore
if in doubt many surgeons will err on making the right
ventricular outflow tract just a little bigger.

OPTIMAL TIMING AND APPROACH

The optimal timing and surgical approach for correction of
TOF remain controversial. With increasing experience in the
repair of many complex cardiac abnormalities in neonates and
infants, there has been a move towards earlier primary correc-
tion of TOF. Arguments in support of this strategy include
removal of the stimulus for right ventricular hypertrophy and
fibrosis, alleviation of systemic cyanosis and, in symptomatic
infants, avoidance of initial palliation with an arteriopulmo-
nary shunt. Recent series of patients operated on in the first
few months of life have shown excellent early and five year
survival.1 A serious concern, however, was an increase in the
use of transannular patches and right ventricle to pulmonary
artery conduits; in one study 42% of patients required
pulmonary incompetence.4 Careful long term follow up is
required with emphasis on arrhythmias and right ventricular
function to define the indications and benefits of early repair.
In conclusion, the outcome after surgical repair of TOF is far
superior to the natural history of the disease, but the timing of
surgery and operative techniques remain controversial. At
present many patients with “repaired” TOF are left with
haemodynamic defects that are poorly tolerated long term and
may require further intervention. Therefore life long follow up
at a congenital heart centre is mandatory.

REFERENCES

1 Campbell M, Deuchar DC, Brock RC. Results of pulmonary valvotomy
and infundibular resection in 100 cases of Fallot’s tetralogy. BMJ
2 Block A, Taussing HB. The surgical treatment of malformations of the
heart in which there is pulmonary stenosis or pulmonary atresia. JAMA
1945;128:189.7
3 Lillehei CW, et al. Direct vision intracardiac surgical correction of the
tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects:
4 Discigil B, Dearani JA, Puga FJ, et al. Late pulmonary valve replacement
after repair of tetralogy of Fallot. J Thorac Cardiovasc Surg
2001;121:344–51.
5 Therrien J, Siu SC, Harris L, et al. Impact of pulmonary valve
replacement on arrhythmia propensity later after repair of tetralogy of
annulus diameter on pulmonary regurgitation and right ventricular
7 Wensley DF, Karl T, Dearfield JE, et al. Assessment of residual right
ventricular outflow tract obstruction following surgery using the response
8 Hirsch JC, Mosca RS, Bose EL. Complete repair of tetralogy of Fallot in
9 Pigula FA, Khalil PN, Mayer JE, et al. Repair of tetralogy of Fallot in

WEB TOP 10

www.heartrnl.com
These articles scored the most hits on Heart’s web site during August 2002.
1 Acute myocardial infarction: reperfusion treatment
F Ribichini, W Wijns
September 2002;88:298–305. (Education in Heart)
2 Antiplatelet treatment in unstable angina: aspirin,
clopidogrel, glycoprotein IIb/IIIa antagonist, or all three?
SA Harding, NA Boon, AD Flapan
July 2002;88:11–14. (Viewpoint)
3 Cardiovascular and coronary risk estimation in hypertension management
EJ Wallis, LE Ramsay, PR Jackson
September 2002;88:306–12. (Education in Heart)
4 Adherence to statin treatment and readmission of patients after myocardial infarction: a six year follow up study
L Wei, J Wang, P Thompson, S Wong, AD Struthers, TM MacDonald
September 2002;88:229–33. (Cardiovascular medicine)
5 Off-pump coronary artery bypass surgery
PP Th de Jaegere, WIL Suyker
September 2002;88:313–18. (Education in Heart)
6 New insights into the mechanism of neurally mediated syncope
MA Mercader, PJ Varghese, SJ Potolicchio, GK Venkatraman, SW Lee
September 2002;88:217–21. (Cardiovascular medicine)
7 Clinical outcomes of patients with diabetes mellitus and acute myocardial infarction treated with primis angioplasty or fibrinolysis
LF Hsu, KH Mak, KW Lau, IL Sim, C Chan, TH Koh, SC Chuah, R Kam, ZP Ding, WS Teo, YL Lim
8 Heart failure in the young
M Burch
August 2002;88:198–202. (Education in Heart)
9 Long term efficacy and safety of atorvastatin in the treatment of severe type III and combined dyslipidaemias
M van Dam, M Zwart, F de Beer, AHM Smelt, MH Prins, MD Trip, LM Havierek, PJ Lansberg, JP Kastelein
September 2002;88:234–8. (Cardiovascular medicine)
10 Arteriosclerotic renal artery stenosis: conservative versus interventional management
C Haller
August 2002;88:193–7. (Education in Heart)
Visit the Heart website for hyperlinks to these articles, by clicking on “Top 10 papers”
www.heartnl.com