PROBLEMS ASSOCIATED WITH TOTAL CORRECTION FOR TETRALOGY OF FALLOT IN OLDER PATIENTS A COMPARATIVE STUDY

GHUFRANULLAH KHAN, AKHTAR HUSSAIN AND MUHAMMAD REHMAN

Department of Cardiac Surgery,
National Institute of Cardiovascular Disease, Karachi.

SUMMARY

During the year Jan 1995- Dec 1995, we performed total correction for tetralogy of Fallot in 80 patients. These patients were divided into two groups according to age in order to study potential problem of doing total correction in older age. Group I, which consisted of patients less than 12 years of age, had a total of 42 patients (24 males, 18 females). Group II which comprised of patients more than 12 years of age, had 38 patients (22 females, 18 males). The main symptom in group I was cyanosis of hyperkinetic spells. Patients with hyperkinetic spells underwent modified Blalock-Taussig shunts. !5 (36%) of group I patients had preliminary systemic pulmonary shunt carried out as opposed to only 2 (7%) for group II. In group II cyanosis was seen in only 75% and the rest 25% give history of cyanosis on exertion only. Their main clinical presentation was effort intolerance (90%). All patients had preoperative echocardiography and cardiac catheterisation. Mean haemoglobin and haematocrit was higher in group-II (9% Vs 5%). Complication like low cardiac output and acute RV failure were more common in group-II. Excessive bleeding was also more common in group-II. Older patients generally required heavier inotropic support and also their requirement for ventilatory support was more prolonged. In conclusion, total correction should be done early as repair in older patients carries more mortality and has potential for serious complications.

INTRODUCTION

Total correction is the ultimate goal one should strive to achieve in a patient with tetralogy of Fallot. Although early total correction in infancy is being advocated in an attempt to avoid late sequelae of cyanosis on the heart, brain and lung,[1,2,3] two stage repair is the preferred alternative with us. We routinely offer total correction to patients who are older than 4 years and prefer to do systemic pulmonary shunts if the patients becomes symptomatic before that age. This together with late referral and ignorance of the general population about their disease, has resulted in older patients coming for total correction. In our experience 30-50% of our patients are more than 12 years of age at the time of total correction.

The purpose of the present report is to highlight special problem of doing total correction in this age group.

MATERIAL AND METHODS

During the year Jan 1994-Dec. 1995, a total of 80 patients underwent total correction. As opposed to the 52 systemic pulmonary shunts were performed.

Patients were divided into 2 groups for the sake of comparison.

GROUP-I : Patients $\leq$ 12 years of age.

GROUP-II: Patients > 12 years of age.

Group-I consisted of 42 patients. Their ages ranged from 5-12, and the median age was 8 years. There were 24 males and 18 females.

Group-II comprised of 38 patients. Age ranged from 13-30 and the median age was 16 years. There were 22 females and 16 males.

All the patients were thoroughly investigated. Apart from routine investigation, echocardiography and cardiac caheterisation was carried out in all the patients. Every effort was made to bring the patients to their best physical shape before the operation. In those patients having hyperkinetic spells, a palliative shunt was preferred, and if the Hb level exceed 20 gm/dl, plasmapheresis was carried out. For those patients having high PCV plasma was kept ready.

In group-I, the main symptoms was cyanosis and hyperkinetic spells. Those with cyanotic spells underwent preliminary modified Blalock-Taussig shunt. A total of 15 out of 42 patients (36%) underwent systemic pulmonary shunts. The shunt was functioning in all except one at the time of total correction. In group-II, only 2 patients had preliminary systemic pulmonary shunt, one of which was Wasterston.

In group-II, the main symptoms was effort intolerance (90%) followed by cyanosis (75%). In the rest (25%), cyanosis appeared on exertion only.

Both the group were compared with regard to mortality and potential for different complications. Following total correction, excessive bleeding was recorded, and need for inotropic and ventilatory support was also assessed. Low C.O. was defined when inotropic support was required for more than 48 hours and ventilatory support considered prolonged if it extended for more than 12 hours.

Surgical technique

Median sternotomy was performed routinely, and autologous pericardial patch was harvested carefully and kept in cold normal saline. Cardio pulmonary bypass (CPB) was established with bicaval venous plus and ascending aortic cannulation. Temperature was drifted to 28°C. Left Ventricle (LV) was vented routinely through RSPV (right superior pulmonary vein). Cold crystalloid cardioplegia was delivered antegradely, after cross clamping the aorta. The intracardiac repair which consisted of closure of VSD dacron patch and relief of infundibular and valvar obstruction, was performed through the right ventriculotomy. The decision to extend the right ventricular outflow tract (RVOT) patch across the annulus was made at the time of operation by taking into account the size of the pulmonary valve ring and comparing it with the minimum required in that age group. After relief of right ventricular outflow obstruction, RVOT reconstruction was done with autologous pericardiac patch except in few where the infundibulum was large and

TABLE –I

RVOT RECONSTRUCTION

	GROUP I (%)	GROUP II (%)
TRANSANNULAR PATCH	80	73
INFUNDIBULAR	20	17
DIRECT CLOSURE		10

TABLE – II

ASSOCIATED ANOMALIES

	GROUP I/n (%)	GROUP II/n (%)
ASD/PFO	10 (23)	12 (31.5)
LSVC	3 (7.1)	1 (2.6)
RIGHT AORTIC ARCH	8 (20)	4 (10.5)
ABNORMAL CONAL ARTERY OR LAD CROSSING RVOT	5 (11.9)	4 (10.5)
AORTIC INCOMPETENCE	0	2 (5.26)
TRICUSPID REGURGITATION	0	5 (13.5)
PATENT DUCTUS ARTERIOSUS	1	0

direct closure appeared feasible. The detail of RVOT reconstruction is given in Table-I. Transannular patch was used in 80% of group-I as opposed to 73% of group-II.

Direct closure of infundibulotomy could be achieved only in group-II patients. These patients had large infundibular chamber (third chamber) and so-called midcavity RV obstruction. Additional cardiac defects encountered in both the groups are listed in table-II.

The atrial septum was routinely inspected and PFO or ASD was closed when found. After completion of the intracardiac repair, temporary pacing wire was attached to RV.

RESULTS

The mortality in group-II was higher 9% Vs. 5%. Important cause of mortality in group-II was low cardiac output secondary to acute RU failure. Those patients in group-II who had preoperative TR fared badly and all of them went on to develop acute RV failure in the postoperative period. Cause of death in group-I was respiratory tract infection and intractable arrythmia.

Morbidity

The complication which were seen are listed in table II.

Complications

While chest infection and transient arrhythmia were equally distributed, excessive bleeding (leading to reopening) was more prevalent in group-II. Low cardiac output was noted in 18% of group as opposed to 7% in group-I. Acute RV failure was seen 13% of group-II and only 2% of group-I.

Inotropic requirement

Average dobutamine requirement for group-II was 13ug/kg/min and 7 ug/kg/min in group-I.

Ventilatory requirement

60% of group-II as opposed to 30% of group-I required ventilatory support for > 12 hours.

DISCUSSION

The optimal management of tetralogy of Fallot is still controversial. Although the safety of total correction in infancy is well established.[1,2,3] Some group still adhere to

TABLE – III
COMPLICATIONS

	GROUP I/n (%)	GROUP II/n (%)
EXCESSIVE BLEEDING	2 (4.76)	6 (15.78)
CARDIAC TAMPONADE	2 (4.76)	2 (5.26)
VENTRICULAR ARRHYTHMIAS	2 (4.76)	3 (7.89)
LOW CARDIAC OUTPUT	3 (7.14)	7 (18.47)
ACUTE RV FAILURE	1 (2.38)	5 (13.15)
SUPERFICIAL WOUND INFECTION	1 (2.38)	0
CHEST INFECTION	3 (7.14)	2 (5.24)

the two stage total correction.[3] Definitive correction should be undertaken early after preliminary systemic pulmonary shunt procedure. Our policy is to undertake total correction as soon as possible after 4th birthday (after palliative systemic pulmonary shunting) provided that no contraindications exist. The only contraindication which precludes total correction is moderate to severe LV dysfunction. This LV dysfunction is usually seen as a result of prolonged LV volume overload imposed by preliminary systemic pulmonary shunting or by abnormal uncontrolled aortopulmonary collaterals. Aortopulmonary collaterals are rare in tetralogy with pulmonary stenosis. Volume overload produced by shunts resulting in dysfunction of LV rarely appears before 5 years after shunting procedure, and was specially common after Waterson and Potts Shunts.[4,5]

The risk of cerebral complication, bacterial endocarditis and pulmonary thrombosis place the life of any uncorrected individual with a functioning shunt in constant jeopardy. Simply observing the patient through childhood, adolescence and adult life not only subjects them to the hazards attendant with a functioning shunt and uncorrected defect, but adds unwarranted risks when subsequent total correction is undertaken.[6]

Non palliated adolescents and adults should be corrected as soon as possible after the diagnosis is documented, provided that adequate pulmonary bed is present. In the experience of Beach and Bowman,[6] correction of nonshunted patients has never been associated with clinical morbidity related to small LV noncompliant right ventricle (RV) or diminutive pulmonary vascular bed. However, in our experience a small LV is a strong predisposing factor for postoperative low cardiac output. These patients have a propensity to ago into pulmonary edema and need prolonged inotropic and ventilatory support. The other problem to which these adult patients are predisposed is acute RV failure leading to low cardiac output in the perioperative period. This is because the RV with has been the seat of prolonged of RV function produced by ventriculotomy and myomectomy could well precipitate acute. RV failure in these patients. We fell that in patients in whom RV is dilated and preoperative tricuspid regurgitation (TR) is present, definitive repair should be withheld as they fare very badly after total correction.

Currently we recommend systemic pulmonary shunt as the only mean of possible palliation for these unfortunate patients.

Aortic incompetence is occasionally seen in older subject with uncorrected tetralogy of Fallot (2 in our series). The aortic root subjected to high flow over a prolonged period of years, dilates and renders the valve incompetent. Turbulence at the valve leaflets may result in marked calcification. The aortic incompetence hinder delivery of cardioplegia solution, might even need valve replacement which thus complicates the total correction and adds to the risks. We were fortunate that aortic incompetence in our patients was only mild and needed no attention.

Regarding palliative shunt surgery, our preference is for the modified Blalock Taussig (B-T) shunt. We tend to do modified BT shunt in patients who become symptomatic before 4 years of age. Although classical BT shunt has superior long term patency and growth potential than prosthetic shunt, their advantage has been neutralised by emergence of earlier complete intracardiac repair at an age before expected shunt failure.[7,8,9]

Moreover, technical difficulties add up when classical BT shunt is done in small babies because of small size of vessels which leads to early shunt failure and related mortality as well as pulmonary artery (PA) distortion particularly if constructed on the side of the aortic arch. Complete mobilisation of subclavian artery increases the risk of phrenic never injury which is poorly tolerated by an infant.[10,11,12,13,14,15] Differential upper extremity growth and arm ischemia are complications associated with sacrifice of the subclavian artery.

The Potts-Smith and Waterston-Cooley shunt are easier to construct with small PA, have good early patency but long term follow up is discouraging. Excessive or unilateral PA flow leading to pulmonary hypertension or congestive heart failure often occurs due to excessively large anastomosis. PA distortion or interruption of pulmonary arteries are more frequent late complications and difficulty of shunt disassembly at the time of complete repair adds significantly to the morbidity and mortality.[18,19,20,21,22]

The modified BT shunt can be constructed with equal ease and results on either side of the aortic arch, although subsequent shunt takedown is technically easier wit less risk of injury to the phrenic nerve, when the shunt is placed on the side of the superior vena cava. This is the reason we favour right modified BT shunt.[23,24]

For infants totally dependent on PDA, PA clamping and shunt construction is often better tolerated on the side opposite the ductus usually the right.

We feel, severely symptomatic older patients (those having frequent hyperkinetic spells), should first be palliated by modified BT shunt as a risk of total correction later are decreased. Preliminary shunting allows growth of pulmonary arteries and leads to amelioration of cyanosis and polycythemia. There is less bleeding at the time of total correction. Controversy still exists regarding the effect of preliminary shunt on the growth of pulmonary annulus. Although studies indicate that preliminary shunting may occasionally be associated with annular growth. This generally is not considered likely enough to justify delay of definitive repair for that reason alone. [25,26,27]

Transatrial transpulmonary approach although feasible in some patients, generally necessitates a ventricular incision and muscle excision especially when the pulmonary ring is small and the infundibulum is ling and narrow. For this reason we use ventriculotomy for both closure of VSD and relief of right ventricular outflow obstruction. We have modified the ventriculotomy incision in case and abnormal coval artery/ LAD crossed the RVOT.[28,29]

The pericardial monocusp is said to provide excellent early haemodynamic function and benefit, however, failure of the

monocusp in the form of pulmonary incompetence is the rule.[30,31]

Concerning RVOT reconstruction the ideal goal is annulus preservation with competent pulmonary valve which permits normal exercise capacity and similar life competency to general matched population. (Wessel)[32] However, hypoplasia of varying degree of RV infundibulum, pulmonary annulus and main pulmonary artery rarely permits this goal to be achieved, and a transannular patch is often necessary in upto 70% of patients. The prevalence of transannular patch in our patients is upward of 70%, and is because we do not trade for residual right ventricular outflow obstruction in place of incompetent pulmonary valve produced by transannular patch. The pulmonary insufficiency occasioned by a trans annular patch is tolerated very well acutely and is not deleterious in ling term studies. However, there does appear to be a deleterious effect of trans annules patch on late functional capacity and exercise tolerance. Wessel noted a reduction in exercise capacity irrespective of PA pressure, if a pulmonary regurgitation is present. Graham noted high RV end diastolic volumes late postoperatively in patients with a trans annular patch, as compared to those without. Late follow up studies at Mayo clinic suggest that trans annular patch is usually well tolerated, but appeared to be a major contributing factor to late postoperative cardiomegaly in only 5% of cases. This small subgroup usually need replacement of pulmonary valve later on.

CONCLUSION

The total correction in older age carries increased mortality and has potential for more serious complications. Patients with preoperative TR of moderate to severe degree should only be palliated with a systemic pulmonary shunt as total correction in this setting carries risk of postoperative low cardiac output and death.

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