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TETRALOGY OF FALLOT
Surgical Considerations

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Surgical treatment for patients with Tetralogy of Fallot has been available for about thirty years and corrective surgery has recently celebrated its twenty fifth birthday. Now seems an apposite time to take stock, to review what has been learnt and to see what questions remain and how we may progress towards answering those questions.

Kirklin and Karp in the introduction to their monograph on Tetralogy of Fallot describe the early years and comment on how difficult it was to get a child to survive after the corrective operation, but say that they were encouraged to proceed by the relatively better results which they were obtaining with other congenital heart deformities. This early experience suggested, and most would agree, that tetralogy is a disease which continues to pose challenges and problems. This presentation concentrates on the surgical aspects of the disease.

Tetralogy of Fallot is defined as a condition with concordant atrioventricular connexion and a characteristic abnormality involving the infundibular septum which is displaced anteriorly, rotated and malaligned with the muscular interventricular septum (Van Praagh 1970). This displacement creates a subaortic ventricular septal defect. The degree of displacement furthermore dictates a variable degree of overriding of the aorta to the right of the muscular ventricular septum. There is a variable degree of obstruction of the outflow tract of the right ventricle involving the infundibulum. The pulmonary annulus and valve, main pulmonary artery and right and left pulmonary arteries are variably developed but usually reduced in size (Becker et al 1975).

Various further abnormalities may be superimposed upon this basic defect. Double outlet right ventricle is defined as a condition in which more than one and a half great arteries arise above the right ventricle and we consider that double outlet right ventricle and tetralogy of Fallot may thus co-exist in the same patient, but of course not all patients with double outlet have tetralogy.

Tetralogy of Fallot, functionally and morphologically, is a progressive disease (Lev et al 1964; Bonchek et al 1973). Patients may present with significant symptoms in very early infancy or may be seen and evaluated purely because of the demonstration of abnormal cardiac physical signs but without significant symptoms at an older age. Symptoms are largely determined by morphology and it is, therefore, not surprising that the more complex the morphology the earlier in life presentation occurs.

In recent years various groups have reported successful results with corrective surgery as a primary procedure in symptomatic infants and young children and we shall refer in detail to this work later. Not all groups, however, have attained such results and it is important to analyse this (Barratt- Boyes and Neutze 1973; Castaneda et al 1977; Pacifico et al 1973).

Pre-operative Evaluation

It is not the aim of this paper to discuss pre-operative evaluation in detail, but it must be emphasised that detailed angiocardiographic analysis is essential to the decision making process regarding treatment and to the choice and conduct of an operative procedure. Selective right and left ventricular injections in the right and left anterior oblique projection provides information regarding the anatomy of the atrioventricular valves, the ventricular muscular and infundibular septa, the presence, site, size and number of ventricular septal defects, the anatomy of the outflow tracts of the ventricles, the connexions of the great arteries, the anatomy of the semilunar valves and the anatomy of the aorta and pulmonary artery. Although reasonable information regarding the anatomy of the right and left pulmonary arteries and their origins can usually be obtained from these views, we also perform studies with the patient in the 45° head up tilt position if more precise information about the origins of the right and left pulmonary arteries and their major branches is needed. This is indicated following previous shunt operations and when tetralogy co-exists with pulmonary atresia with abnormal aorta/pulmonary artery collaterals. The anatomy of the coronary arteries requires definition if corrective surgery is planned in infants or if previous intrapericardial operative procedures have been performed (Blackstone et al 1979; Calder et al 1979).

Surgical Treatment

The results of operation, of whatever type, are largely determined by events which occur in the operating room. This is especially true with open intracardiac operations, but equally applies to the results obtained with palliative shunt operations. Precise operative technique, meticulously planned and executed, is essential.

Shunt Operations

The Blalock, modified Blalock (end to side interposition of Gore Tex graft) and Waterston shunts are the techniques in general use, the classical subclavian artery to pulmonary artery shunt on the side of the innominate artery being the procedure of choice. Although many groups welcomed the development of the ascending aorta anastomosis because of unsatisfactory results from the Blalock shunt in small children, recently better techniques have been used and excellent early results for the Blalock in infants have been published. How infants treated in this way will progress and whether adequately increased pulmonary blood flow can always be obtained and maintained with growth is less certain.

Table I

Tetralogy of Fallot
'Blalock' 1 Year

Authors	Year	No	% Mortality
Aberdeen	— 1975	13	—
Alvarez	— 1977	12	—
Arciniegas	— 1975	28	—
Bianchi	— 1974	10	—
Pontius	— 1974	32	8%
Selmonosky	— 1973	43	5%
PBD/'HSC'	— 1975	25	8%

Late complications after the Waterston anastomosis are not uncommon and the published early mortality is very variable and probably less predictable (Reitman et al 1974; Tay et al 1974).

Table 2

Tetralogy of Fallot
'Waterston' shunt

Authors	Year	No	% Mortality
Alvarez	– 1977	181	4.9%
Arreiniegas	– 1975	37	2.7%
Bianchi	– 1975	67	9%
Castaneda	– 1974	81	14%
Daicoff	– 1972	44(1)	41%
Waldhausen	– 1972	23	—

If a two stage surgical approach is used, we urgently need to know the total cumulative mortality and morbidity of the two operations and the period between them, and whether the first stage procedure produces any late post total correction problems eg. residual right pulmonary arterial stenosis after correction and disconnection of a Waterston anastomosis.

Corrective Operation

Open intracardiac operations are performed using one of the described perfusion techniques (Kirklin and Karp 1970; Barratt-Boyes and Neutze 1973).

Table 3

Correction of Tetralogy of Fallot
Determinants of Operative Result

Diagnosis
Myocardial Protection
Surgical Procedure
I VSD
Conduction Tissue
II ASD
III Outflow Tract
IV Aorta Pulmonary Connections
Post Operative Care

The technique of myocardial preservation is of vital importance, but there is no data proving the advantages of eg. cardioplegic ischaemic arrest with associated hypothermia as opposed to intermittent ischaemic cardiac arrest at 30° C interposed with periods of coronary perfusion. There is data, eg. with coronary artery disease, which suggests that the cardioplegic technique, if precisely applied, causes less myocardial damage. There are specific features with the tetralogy, however, to which attention must be paid in choosing the technique of perfusion and myocardial protection, quite apart from the dictates of patient size, etc. The right ventricle is hypertrophied as is the interventricular septum and there is often increased aorta to pulmonary artery collateral flow. Each may make adequate application of the cardioplegic technique more

demanding. The conduct of this facet of the correction is one of the major determinants of the outcome and should be controllable and predictable.

In general, many groups are using the cardioplegic/hypothermic technique, but the results obtained by McGoon, Starr, Malm and others have followed the use of the intermittent moderate hypothermia ischaemic technique. Whichever technique is chosen, it is essential for the technique to be analysed fully so that its potential and inadequacy be fully appreciated.

The corrective procedure consists of septation of the heart and relief of outflow tract obstruction. Its facets are coinsidered individually.

Atrial Septal Repair

Defects of the inter atrial septum, whether they be of the so-called primum (rarely), secundum or patent foramen ovale type, occur in about 30% of patients with tetralogy. The interatrial septum is explored routinely and usually this requires a short, right atriotomy. (In larger hearts it may be possible to repair the atrial septum via the tricuspid valve exposed through the right ventriculotomy).

There is frequently a period of relative right ventricular dysfunction after correction and right ventricular end endiastolic causes mean right atrial pressure to exceed mean left atrial pressure. Right to left shunting develops at atrial level. Analysis and treatment of the resultant system arterial desaturation may prove difficult.

Right Ventriculotomy

A transverse incision at the junction of the infundibular and sinus portions of the right ventricle avoiding major coronary arterial branches, is more than probably the least traumatic approach. Recently there has been a tendency to use vertical ventriculotomies; the incision subsequently being closed by the insertion of a small patch of Dacron or pericardium. If necessary, the incision can be extended through the pulmonary valve and along the pulmonary arteries if patch enlargement of the outflow tract is necessary.

The least traumatic ventricular incision should be used if possible and thus the transverse incision is used unless pre-operative evaluation and/or external inspection of the heart suggests that patch enlargement of the outflow tract across the pulmonary valve is probably necessary. The need for patch enlargement should be predictable from the pre-operative angiocardiographic data (Blackstone et al 1979; Calder et al 1979).

Ventricular Septal Repair

The interventricular septum is repaired by patching the sub-aortic ventricular septal defect and rarely closing further defects. It seems though, without objective proof, that a more haemostatic closure can be obtained using a continuous suture technique, but a very satisfactory repair using an interrupted suture technique is possible.

Avoiding the conduction tissue is essential, and is most reliably obtained by placing sutures to the right of the crest of the septal defect and through the base of the septal and anterior tricuspid valve leaflet tissue. The anatomical studies of Lev and Anderson and colleagues have shown that the most dangerous area where the conduction system is most at risk is the area where the fibrous trigone of the heart, the tricuspid valve and the aortic valve meet, where the His bundle penetrates through to the ventricle. Beyond this point, the left bundle branch usually runs on the left side of the inferior margin of the septal defect, and it is safe to place sutures into the crest of

the interventricular septum. The occasional heart may, however, have conduction tissue along the inferior crest of the defect and it is safer to place the sutures to the right of the crest (Lev 1959; Anderson et al 1976).

The late results of correction may be significantly influenced by even temporary damage to the conduction system. There is evidence that the combination of right bundle branch block and left anterior hemi-block may be a dangerous prognostic feature, particularly if this abnormality occurs in a patient in whom there has been transient heart block in the early post operative period. There is an argument that such a patient should have a permanent demand pacemaker inserted (Steeg and colleagues 1975).

Repair of the Right Ventricular Outflow Tract

It is sometimes obvious that transannular patching of the outflow tract is, or is not, needed, but in many patients with tetralogy the decision is less clear cut.

Table 4

Minimum acceptable pulmonary valve ring diameter employed in 1975 and 1976

Weigtht (Kg.)	Minimum ring size	
	Diameter (mm.)	Area (sq. mm.)
3	6	28
4	7	38
5	7.5	45
6	8	50
7	9	63
8	9.5	72
9	10	81
10	11	90
12	12	113
14	13	126
16	13.5	144
18	14	162
20	15	177
25	17	225
30	18.5	270
35	20	314
40	20	314

In such hearts correction without transannular patching, discontinuation of bypass and measurement of right and left ventricular pressures is standard practise. If RV/LV pressure ratio is .75 or above, bypass is resumed and a transannular patch inserted. This technique is acceptable in older patients, but is inappropriate when circulatory arrest techniques using deep hypothermia are used and, in general, is contra-indicated when cardioplegic methods are utilised.

An alternative is to try and predict those patients who will or will not require transannular patching before operation. Kirklin and colleagues, utilising their own and Lev's data, have sought — on the basis of pre-operative angiocardiographic and intra-operative measurements — to create numerical rules which determine the need for trasannular patching.

Table 5

Estimated probability of requiring patch-graft enlargement across pulmonary valve ring

BSA (sq. M.)	Probability*					Highest value within 95% CL‡
	15% (mean normal)†	35% (<normal but within 50% CL)	50% (<normal but within 72.5% CL)	75% (<normal but within 95% CL)	90% (small, <than 95% CL)	
0.15	5.9	5.1	4.6	3.6	< 3.6	8.1
0.20	7.3	6.5	6.1	5.1	< 5.1	9.6
0.25	8.4	7.6	7.2	6.2	< 6.2	10.7
0.30	9.3	8.5	8.1	7.1	< 7.1	11.6
0.35	10.1	9.3	8.8	7.8	< 7.8	12.3
0.40	10.7	10.0	9.5	8.5	< 8.5	13.0
0.45	11.3	10.6	10.1	9.1	< 9.1	13.6
0.50	11.9	11.1	10.6	9.6	< 9.6	14.1
0.55	12.3	11.6	11.1	10.1	<10.1	14.6
0.60	12.8	12.0	11.5	10.5	<10.5	15.0
0.65	13.2	12.4	11.9	10.9	<10.9	15.4
0.70	13.5	12.7	12.3	11.3	<11.3	15.8
0.75	13.9	13.1	12.6	11.6	<11.6	16.1
0.80	14.2	13.4	12.9	11.9	<11.9	16.4
0.90	14.8	14.0	13.5	12.5	<12.5	17.0
1.0	15.3	14.5	14.0	13.0	<13.0	17.6
1.2	16.2	15.4	14.9	13.9	<13.9	18.5
1.4	17.0	16.2	15.7	14.7	<14.7	19.2
1.6	17.6	16.8	16.4	15.4	<15.4	19.9
1.8	18.2	17.4	16.9	15.9	<15.9	20.5
2.0	18.7	18.0	17.5	16.5	<16.5	21.0

Legend: Based on data of Rowlatt, Rimoldi and Lev,[1] described by the equation:

$$\text{Diameter (mm.)} = 10/\pi[3.5869 \cdot \log_{10}(\text{BSA}) - 9.5431]$$

CL, Confidence limits.

*That is, probability of $Pav/Lv > 0.65$ without patch across valve ring.

†Phrases in parentheses describe size of pulmonary valve ring relative to patient's body surface area (BSA); the numbers are the measured internal diameter of the pulmonary valve ring (mm.) after valvotomy.

‡The woven Dacron tube from which the outflow patch is made can be this size.

The distal extent of patching depends on individual anatomy. It seems that more patients with relative stenosis of the origins of the right and/or left pulmonary arteries are seen when correction is carried out in infants. Second stage correction patients may have stenosis of the right or left pulmonary artery. Recognition of such stenoses and their relief is important.

The indications for the insertion of a valve mechanism when transannular patching is performed are not clear. A variety of techniques exist, eg. the insertion of a complete valve homograft to the utilisation of monocusp or bicusp patches (Asano and Eguchi 1970; Hawe et al 1970). A valve mechanism should be inserted if resistance to blood flow through the lungs is increased, eg. when previous shunts have created degrees of main branch pulmonary artery stenosis or if peripheral pulmonary stenoses exist. In 1960, Ferencz demonstrated the presence of obstructive thrombotic lesions within the small pulmonary arteries of cyanotic patients with tetralogy and showed that such obstructive lesions could disappear following the creation of an effective shunt. This may be relevant if primary total correction is planned in such patients.

There has been a relative lack of further information on the vasculature within the lungs in tetralogy. Recently, more information has become available in patients with tetralogy and pulmonary atresia showing the presence of anatomical abnormalities of intrapulmonary vascular architecture, which, if demonstrable, would represent an indication for the insertion of a valve mechanism to prevent or minimise pulmonary regurgitation at least in the early postoperative period.

Closure of previous shunts and control of aorta/pulmonary collaterals

Blalock, Waterston or, rarely, Potts shunts are closed as part of the corrective operation. When possible, the subclavian artery involved in the Blalock shunt is dissected at its origin in the neck, rather then dissecting it within the mediastinum close to the pulmonary arteries (Kirklin and Karp 1970; Elkins and colleagues 1972). If the Blalock has been performed on the side of the inominate artery dissection of the subclavian at its origin is simple, quick and less likely to be associated with bleeding.

In closing a Waterston shunt the right pulmonary artery is separated from the back of the aorta, the two arteries being repaired independently, rather than closing the shunt via a transaortic approach. It is possible to adequately repair the right pulmonary artery by the transaortic root if it is absolutely certain that no stenosis of the right pulmonary artery exists, but generally definitive repair is preferable.

Patent ductus arteriosus is uncommon with tetralogy but in young infants patent ductus is present frequently and duct closure is necessary as part of the corrective operation.

The control of other major aorta/pulmonary collaterals is more difficult. Frequently, the aorta/pulmonary collaterals are small and inaccessible and cannot be closed. In patients in whom multiple, large aorta/pulmonary collaterals exist, there is doubt as to whether an attempt should be made to close these or not. Macartney and colleagues have demonstrated and discussed in detail the concepts of unifocal and multifocal pulmonary blood supply in patients with pulmonary atresia. Sometimes segments of lung receive their sole blood supply from collaterals and closure may be contraindicated (Alfieri et al 1978). Similarly, such patients may have only limited pulmonary vessels connected to the central pulmonary arteries and hence available to accept right ventricular output.

DISCUSSION

These, then, are the fundamentals of surgical treatment of tetralogy of Fallot. Detailed reviews and discussions of results from different groups are readily available to us all in the literature, and it is not the aim here to present and analyse such results in detail. My own series of patients with tetralogy receiving corrective surgery is described in the Tables. Is is obvious that the results of primary total correction in infants are not satisfactory as compared to the results published by the groups in Auckland, Birmingham, Stanford and Boston (Table 6, Table 7).

Why the difference? Are the events in the operating room or the post operative care area the more important? Techniques of perfusion and myocardial protection are of major importance, but should be predictably reproducible. The technique of correction should be consistent in different hands as far as the septal defects are concerned with no damage to the conduction system.

As far as the operation is concerned, therefore, the most variable and most unpredictable element is repair of the outflow tract. I suspect, but cannot prove, that it is not simply a question of transannular patching or not, but also how big a patch, what material, and valved or not. The precise, subtle details of transannular patches are much

more variable than the technique of repair of the septal defect. It is for this reason that the attempt to quantitatite patches numerically and the creation of rules for or against transannular patches will produce more equality in operative results. Which material to use for outflow tract patching is an open question.

Table 6

Tetralogy of Fallot 1970-1979

	No. of patients	Deaths
Total Correction	206	(24)* 11.6%
Primary	151	(19) 11.8%
Secondary	55	(5) 9.1%
— Waterston	27	(1) 3.8%
— Blalock	24	(4) 16.6%
— Other	4	—

* 2 of 3 patients with complete A-V canal died.

Table 7

Tetralogy of Fallot
Total correction

Age	No	Mortality
Under 3 months	1	(1) 100%
3-6 months	7	(3) 42%
6-12 months	8	(3) 37%
All Infants	16	(7) 43%
1-5 years	80	(11) 13.7%
5-15 years	96	(5) 5.2%
over 15 years	14	(1) 7.1%
	206	24

Preoperative assessment should be a reproducible facet of care. Post operative care, especially in infants, is not predictable and reproducible since there are real differences in terms of personnel, expertise and equipment. Good post operative care may recover ground lost in the operating room. Less good care increases mortality and morbidity regardless of the operation (Kirklin and Karp 1970).

What are the major questions when assessing the results of surgery for the tetralogy? Do we perform one-stage correction, or two-stage correction? In recent years excellent results have been reported from primary correction of all patients with tetralogy. The groups in Birmingham, Auckland, Boston and Stanford have all quoted a mortality of well below 10%. Studies have been published whereby patients operated on as infants have been reinvestigated after two to three years and found to have excellent haemodynamics. The mortality for primary correction in other centres has been less satisfactory with mortality rates between 20% and 60% being quoted.

Table 8

Tetralogy of Fallot
Correction % Mortality

Authors		Primary	Secondary *
Bahnson	(1969)	11.4%	11.1%
Ebert	(1967)	14.3%	10.8%
Karp	(1970)	3.4%	2.4%
Malm	(1970)	6.4%	6.5%
Shumway	(1978)	2.1%	0%

* Single Blalock

If primary correction is the procedure of choice, what is the ideal perfusion technique? What are the late effects of circulatory arrest? What are the precise indications for the insertion of an outflow tract patch, how big should that patch be and what materials should be used, and even more debatably should that patch contain a valve mechanism? Are there absolute contraindications to one stage repair eg. the presence of anomalous coronary arteries.

Inevitably, transannular patching results in a degree of pulmonary regurgitation (Calder et al 1979). Pulmonary valvotomy without a transannular patch may similarly produce pulmonary regurgitation. There is no absolute data comparing the degree of regurgitation occuring in these two groups. Regurgitation may be more significant if a patch has been placed and particularly if pulmonary artery endiastolic pressure is elevated. What are the long term consequences of pulmonary regurgitation? Although a valve mechanism may function early post operatively, does this function persist? Late post operative studies have shown that the cardiac output at rest is normal and responds relatively normally to exercise. Heart size is increased — this affecting the right heart structures. Right ventricular endiastolic pressure and right atrial pressure are above normal and become more elevated on exercise.

Patients with the syndrome of tetralogy and absent pulmonary valve do well without the insertion of a pulmonary valve for some years after operation, but late right heart failure has now been described.

There is, therefore, clinical and haemodynamic evidence of adverse late effects from chronic pulmonary regurgitation (Epstein et al 1973).

Whereas in older patients and assuming normal pulmonary vascular resistance transannular patching does not increase the risk of correction, there is data to show that transannular patching does increase the immediate risk of correction under two years of age (Kirklin et al 1979). Thus acute pulmonary regurgitation is an adverse factor under some circumstances.

The incidence of transannular patches in young patients is higher than the incidence of transannular patches in older patients with one or two-stage correction. Alfieri et al have demonstrated that the pulmonary annulus and main pulmonary artery increase in size after the performance of an ascending aorta to right pulmonary artery anastomosis — true growth occurs. There is no similar data with the Blalock anastomosis, but at least inferentially one would expect this to be so. It is likely, therefore, that a shunt may lessen the need for outflow tract patches by producing relative growth of the pulmonary annulus and main pulmonary artery. Ferencz's data suggests that a shunt may 'prepare' the pulmonary vasculature for correction but her data concerned chronically cyanosed patients and not infants.

Is it age, size or anatomy that is important in determining the higher mortality in infants in most centres? There is evidence that extracorporeal perfusion produces organ dysfunction which is related to trauma to blood constituents during bypass and to

changes in body composition and to the distribution of components. The general experience is that the relative amount of organ dysfunction is greater in smaller subjects. Although improvements have taken place in recent years in the design of extracorporeal perfusion systems, and the introduction of membrane oxygenators may prove significant biologically, perfusion remains more traumatic in small subjects and remains a determinant of mortality and morbidity.

The process of natural selection in patients with tetralogy causes those with the severely abnormal morphology to present in early infancy. Primary correction in infants will select out patients who may well require the most radical corrective surgery, but these patients will be subjected to the greatest biological insult. On the other hand, however, tetralogy is a progressive disease. Right ventricular myocardial hypertrophy progresses with age. The extent of muscle resection in the right ventricle becomes greater, in general, with increasing age and fibrosis and hypertrophy of the left ventricle become evident.

Table 9

Tetralogy of Fallot
Primary Correction in Infants

Authors	Year	No	% Mortality
Barratt Boyes	— 1979	43	7%
Matsumoto	— 1974	16	38%
Pacifico	1975 — 1976	11	9.1%
Parenzan	— 1977	16	18.7%
Malm	1967 — 1975	11	27%
Starr	— 1977	34	9%
Subramanian	— 1977	22	32%

If results favour the two-stage approach to treatment, which shunt is best? Does the performance of a shunt increase the risk of the subsequent repair and what is the total cumulative mortality and morbidity resulting from the two-stage approach as compared to the one-stage procedure? The Blalock anastomosis can be constructed with low risk and does not increase the risk of correction. The incidence of complications at pulmonary artery level is low, and closure of the shunt relatively straight forward. The shunt may cause true growth of the pulmonary annulus and main pulmonary artery.

The Waterston shunt is quicker to construct but, in most centres, seems to have a higher initial mortality and certainly causes late complications. Its closure is more complex and, even with patch reconstruction of the rigth pulmonary artery, significant residual haemodynamic changes may persist. Why the results in the two-stage group (Table 10) having a previous Waterston are so much better (apparently) is not clear. In this series the total data of cumulative risk is not available but probably shows the apparently better 'Waterston'data to be fallacious.

Table 10

Tetralogy of Fallot
Mortality of Correction After Previous Waterston

Alvarez	57	6	10.5%
PBD	27	1	3.7%
Parenzan	26	1	3.8%
TAY	10	4	40%

There is, unfortunately, little precise data answering the question: what is the cumulative mortality of two stage as opposed to one stage correction *in comparable groups of patients?* There has been no controlled trial of this. The encouragingly low mortality of the Blalock operation in infants is good, but there is no data about morbidity or mortality in these patients between the time of primary shunting and secondary total correction, no data about the long term subsequent correction in *these same patients.*

No attempt has been made here to assess economic, social and psychological factors.

The total sum of available data at this stage does not permit a definite answer to the questions posed. Continued, careful data analysis, follow up and late clinical and haemodynamic evaluation is now required.

RESUMO

TETRALOGIA DE FALLOT

A cirurgia paliativa, da tetralogia de Fallot iniciou-se há pouco mais de 30 anos e a sua correcção intracardíaca tem 25 anos de existência. Os progressos técnicos da cirurgia do coração aberto permitiram com alguns grupos fazerem a correcção cirúrgica no 1.º ano de vida como indicação electiva. A finalidade deste artigo é analisar a situação actual da cirurgia, especialmente no que se refere à posição dos shunts paliativos.

A Tetralogia de Fallot é uma cardiopatia congénita em que existe conexão atrio ventricular concordante e uma anomalia característica do septo infundibular que se encontra deslocado anteriormente, rodado e mal alinhado em relação ao septo interventricular. Esta alteração anatómica é responsável pela existência de uma comunicação interventricular, e da sua intensidade, depende o grau de dextroposição da aorta. Verifica-se também uma estenose infundibular do ventrículo direito de grau variável. O anel e a válvula pulmonar principal e seus ramos têm desenvolvimento variável mas normalmente reduzida.

Sucessivamente se faz referência aos vários estádios desde o diagnóstico até ao tratamento cirúrgico e cuidados intensivos pós-operatórios.

Realçam-se o papel do diagnóstico pré-operatório completo e perfeito usando todos os métodos existentes. O que se passa no Bloco Operatório é grandemente responsável pelo resultado obtido. Discutem-se os vários tipos de anastomoses sistémico-pulmonares paliativos.

Para se poder optar pelo tratamento cirúrgico em dois tempos é necessário conhecer a mortalidade e morbilidade conjunta das duas operações. O tratamento cirúrgico correcto tem pasado por várias fases e deve realçar-se o papel da técnica de preservação do miocárdio e da sua evolução. Na correcção intercardíaca, há vários tempos operatórios: sutura da comunicação interauricular, ventriculotomia direita, encerramento da comunicação interventricular, alargamento do infundibulo do ventrículo direito e, finalmente, encerramento e contrôlo dos «shunts» prévios de colaterais aortopulmonares.

Finalmente discutem-se os vários problemas enunciados admitindo-se que diversos centros tenham atitudes cirúrgicas diferentes. O somatório dos dados actualmente existentes não permite uma resposta exacta em todos os problemas enunciados. É necessário continuar a análise cuidadosa de todos os dados, muito especialmente no que se refere aos resultados à distância tanto clínicos como hemodinâmicos.

REFERENCES

ALFIERI O, BLACKSTONE EH, KIRKLIN JW, PACIFICO AD, BASFERON LM: Surgical treatement of Tetralogy of Fallot with pulmonary atresia. *J Thorac Cardiovasc Surg* 76: 321, 1978.

ANDERSON RH, MONRO JL, YEN HO S, SMITH A, DEVERALL Ph B: Les voies de conduction auriculo-ventriculaires dans la Tetralogie de Fallot. *Coeur — Vol. VIII — No 3, Mai-Juin 1977. Numéro spécial: Journées internationales de cardiologie Pédiatrique. Octobre 1976. Bendor J Thorac Cardiovasc Surgery.*

ASANO KEN-ICHI, EGUCHI SHOJI: A new method of right ventricular outflow reconstruction in corrective surgery for Tetralogy of Fallot. The Application of the valve-retaining pulmonary artery graft as a patch. 59: 512, Apr. 1970.

BARRATT-BOYES BG, NEUTZE JM: Primary repair of Tetralogy of Fallot in Infancy using profound hypothermia with circulatory arrest and limites cardiopulmonary bypas. *Ann Surg* 178: Oct. 1973.

BECKER AE, CONNER M, ANDERSON RH: Tetralogy of Fallot: A morphometric and geometric study. *The Am J of Cardiol* 35: 402, Mar. 1975.

BLACKSTONE EH, KIRKLIN JW, PACIFICO AD: Decision-making in repair of Tetralogy of Fallot based on intraoperative measurements of pulmonary arterial outflow tract. *J of Thorac Cardiovasc Surg* 77: 526, Apr. 1979.

BONCHEK L, STARR A, SUNDERLAND CO, MENASHE VD: Natural history of Tetralogy of Fallot in Infancy. Clinical classification and therapeutic implications. *Circulation* 43: Aug 1973.

CALDER AL, BARRATT-BOYES BG, BRANDT PWT, NEUTZE JM: Postoperative evaluation of patients with Tetralogy of Fallot repaired in infancy. Including criteria for use of outflow patching and radiologic assessment of pulmonary regurgitation. *J of Thorac and Cardiovasc Surg 77: 1979.*

CASTENADA AR, FREED MD, WILLIAMS RG, NORWOOD WT: Repair of Tetralogy of Fallot in infancy. Early and late results. *J Thorac Cardiovasc Surg* 74, 372, 1977.

DEVERALL PB: Tetralogy of Fallot — pallisation VS correction in infancy. *Paediatr Cardiol* 1977. Churchill Livingstone p 283.

EGUCHI SHOJI, ASANO KEN-ICHI, NIIGATA: Homograft of pulmonary artery or ascending aorta with valve as a right ventricular outflow. *J Thorac Cardiovasc Surg* 56: Sep. 1968.

ELKINS RC, FISHER RD, BENDER HW, BRAWLEY RK, GOTT VL, HALLER JA: Operative Management of the Blalock-Taussig anastomosis during total repair of Tetralogy of Fallot. *Ann Thorac Surg* 13: June 1972.

EPSTEIN SE, BEISER GD, GOLDSTEIN RE, ROSING DR, REDWOOD DR, MORROW AG: Haemodinamic abnormalities in response to mild and intense upright exercise following operative correction of an atrial septal defect or Tetralogy of Fallot. *Circulation* 47: May 1973.

FERENCZ C: The pulmonary vascular bed in Tetralogy of Fallot I. Changes associated with pulmonic stenosis. *Bulletin Johns Hopkins Hosp* 106: 81, 1960.

FERENCZ C: The pulmonary vascular bed in Tetralogy of Fallot II. Changes following a systemic-pulmonary arterial anastomosis. *Bulletin Johns Hopkins Hosp* 101: 100, 1960.

GERSONY WM, BATTHANY S, BOWMAN Jr FO, MALM JR: Late follow-up patients evaluation haemodynamically after total correction of Tetralogy of Fallot. *J Thorac Cardiovasc Surg* 66: Aug. 1973.

HAWE A, RASTELLI GO, RITTER DG, DUSHANE JW, McGOON DC: Management of the right ventricular outflow tract in severe Tetralogy of Fallot. *J Thorac Cardiovasc Surg* 60: 131, July 1970.

JARMAKANI JMM, GRAHAM Jr TP, CANENT Jr RV, JEWETT PH: Left heart function in children with Tetralogy of Fallot before and after palliative or corrective surgery. *Circulation* 46: Sep. 1972.

JONES EL, CONTI CR, NEILL CA, GOTT VL, BRAWELWY RK, HALLER Jr JA: Long-term evaluation of Tetralogy patients with pulmonary valvular insufficiency resulting from outflow-patch correction across the pulmonic annulus. *Circulation* Suppl. 3 47: July 1973.

KINSLEY RH, McGOON DC, DANIELSON GK, WALLACE RB, MAIR DD: Pulmonary arterial hypertension after repair of Tetralogy of Fallot. *J Thorac Cardiovasc Surg* 67: 110, Jan. 1974.

KIRKLIN JW, KARP RB: The Tetralogy of Fallot. *WB Saunders Co,* 1970.

LEV M: The architecture of the conduction system in congenital heart disease II. Tetralogy of Fallot. *Arch Pathol* 67: 572, 1959.

LEV M, RIMOLDI HJA, ROWLATT UF: The quantitative anatomy of cyanotic Tetralogy of Fallot. *Circulation* 30: Oct. 1964.

PACIFICO AD, BARGERON Jr LM, KIRKLIN JW: Primary total correction of Tetralogy of Fallot in children less than four years of age. *Circulation* 48: Nov 1973.

REITMAN MJ, GALIOTO Jr FM, EL-SAID GM, COOLEY DA, HALLMAN GL, McNAMARA DG: Ascending aorta to right pulmonary artery anastamosis. *Circulation* 49: May 1974.

STARR A, BONCHEK LI, SUNDERLAND CO: Total correction of tetralogy of Fallot in infancy. *J Thorac Cardiovasc Surg* 65: 45, Jan 1973.

STEEG CN, KRONGRAD E, DAVACHI F, BOWMAN Jr FO, MALM JR, GERSONY WM: Postoperative left anterior Hemiblock and right bundle branch block following repair of Tetralogy of Fallot. *Circulation* 51: June 1975.

SUNDERLAND CO, MATARAZZO RG, LEES MH, MENASHE VD BONCHEK LI, ROSENBERG JA, STARR A: Total correction of Tetralogy of Fallot in infancy. *Circulation* 48: Aug 1973.

TAY DJ, ENGLE MA, EHLERS KH, LEVIN AR: Early results and late developments of the Waterston anastomosis. *Circulatio* 50: Aug 1974.

VAN PRAAGH R, VAN PRAAGH S NEBESAR RA, MUSTER AJ, SINHA SN, PAUL MH: Tetralogy of Fallot: Underdevelopment of the pulmonary infundibulum and its sequelae. *Am J Cardiol* 26: July 1970.

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