TETRALOGY OF FALLOT

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This presentation is based on the total experience with Tetralogy of Fallot of the cardiac surgery group in South Australia—and includes all cases managed from the first shunting procedure in 1955 up until December, 1971.

Almost everything has been said which could possibly be said about Tetralogy of Fallot in many excellent monographs and publications. In spite of this the clinical entity is such a broad one and presents so many challenges to the cardiologist and the surgeon that further analyses are usually productive.

On this occasion the total South Australian experience is dissected to illustrate the incidence, the methods of management available, the changes that have taken place in our own attitude to management and reasons why the changes have been made. As the discussion develops the many treatment options which have to be considered will be assessed and some of the reasons for choosing one or other of these will be discussed. In particular some decisions will be specifically related to management in our part of the world in contra-distinc tion to management in the more experienced centres dealing with larger numbers of cases.

In this series there have been 175 operations, of these 119 were full correction on cardio-pulmonary by-pass, one Brock procedure on by-pass and 55 attempts to perform a palliative shunt. As 25 of the shunted cases were reoperated by the end of 1971 this means that 150 patients were treated surgically in this time.

South Australia is a relatively isolated community with 80% of the population of approximately 1 million people living within 50 miles of Adelaide and the next nearest cardiac units are 400, 700 and 1400 air miles distant. It so happens that 48 of the 150 patients operated came to Adelaide for treatment from outside the State boundaries, hence 102 cases of Tetralogy presented for treatment in 17 years from our own community or 6 cases per million per year.

To put Tetralogy of Fallot in some sort of surgical perspective we find that since the first cardiac operation in 1949 and up to the end of 1971, 1,286 operations for congenital heart disease were performed in South Australia and the 175 operations for Tetralogy represent 13.6% of this total. In the open heart program (1960 to 1971) there were 1,215 operations including 641 for congenital conditions, thus the 119 Fallot corrections represent 9.8% of this total.

Because of the anatomical and haemodynamic variants covered by the definition Tetralogy of Fallot, a number of options have to be considered in treatment and these must be looked at seriously and critically in conjunction with the facilities which are available and the skill and experience of the surgical team on hand. Remembering these factors of skill and experience it is encumbent on us to offer to each patient as they come into our care what may be their best chance for life and health.

It is dishonest to state that the risk of surgery for a certain operation is very low simply because we have read or heard such figures from the proven masters of surgery. Each unit and each surgeon carry their own mortality for a certain condition, often very different from that of the masters and certain to vary in time with the surgeon's own increasing experience. It is important to relate our management decisions to these factors. In this way, failures and disasters will be reduced to a minimum while time and experience are gained.

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Admittedly there are occasions when desperate situations require immediate intervention. Under these circumstances we must do the best we can. But in Tetralogy of Fallot deterioration is often slow and frequently operations can be deferred and performed at some time of election with advantage to the patient.

Furthermore, as will be shown, time can often be gained for the patient with relatively simple palliative procedures at low risk. Nobody doubts that full correction is the ideal form of management where all circumstances are favourable and all skills are available.

Up to now consideration has been given to what may be broadly termed philosophical options which relate to local skills, experience and attitudes. We must now relate these philosophies to specific problems:

(a) Time of election and age of election for operation.
(b) The "mild" Fallot.
(c) The "severe" Fallot.

and also to options of a purely technical nature, such as:

(a) Hypothermic arrest,
(b) Myocardial protection during repair,
(c) Out-flow tract management.

Then perhaps the most important option of all, the timing and place of palliative procedures in management.

In the matter of time of election for example, when we began our own open heart program in 1960, being aware of the limitations of our own ability and the evil reputation Fallot correction had at that time, we elected not to attempt any full corrections until our team had been working together for nearly a year.

In that first year our first six corrections had a mean age of 20 years with a 16% mortality. Up to that time (1961) the mean age of cases being shunted in the Unit was 5 years. In comparison over the last three years (1967-1971) 48 cases with a mean age of 8.7 years had full corrections with a mortality of 6% and in the same three year period the cases shunted had a mean age of 15 months. Thus with experience our ages of election have clearly changed but had we elected to attempt to correct all the Fallots below the age of six years who presented in 1960 the results may have been disastrous.

Consider now a mild Fallot, over six years of age with a big pulmonary valve ring, normal pulmonary arteries and reasonab'e exercise tolerance. An operation in this situation is almost like a half-holiday for an experienced team. But this patient has a good life expectancy without operation and so should not be operated unless it can be offered at near zero mortality. This child can afford to wait for the surgical group to improve their rating. Under other circumstances and in every special surgical Unit a child with this sort of anatomy can be safely operated down to the neo-natal age period. This is a correct decision but **only** if it can be offered at a negligable mortality irrespective of age. It may be justifiable for a team with ten years experience and 2,000 by-passes behind them to say that they can offer a better percentage for the patient by doing a full correction as a neo-nate rather than shunting the chi!d and waiting, or even just waiting.

At the same time it does **not** mean that this plan is right for all of us and it is certainly not right for any of us early in our Fallot experience. Personally, we find doing full corrections under the age of three years very difficult both in operative technique and post-operative management, and **under** the age of one year we find them **very very** difficult indeed. Furthermore I believe that if I had not done more than a hundred full corrections irrespective of age I would be finding

full correction for neo-natal Fallots too difficult for me personally.

Leaving the "severe" Fallot aside for the moment this leads us to the first of the purely technical options—deep hypothermic arrest.

If the team is experienced enough to do Fallots as neonates, they will have to choose between core-cooling or deep surface hypothermia with ice bags followed by core control as is advocated by Shirotani and further developed by Sir Brian Barratt-Boyes. We believe that core-cooling is precise and effective and we intend to continue with core-cooling whenever total arrest is needed for this or any other condition irrespective of age.

To us deep hypothermia means adequate cooling of the central nervous system supported by monitoring to confirm that adequate temperature levels have been achieved to satisfy the time factor required. It seems logical to us to cool the central nervous system from within as long as the above criteria are satisfied. Reducing to these temperatures by surface cooling through a vasoconstricted, insulated peripherary aided only by a grossly reduced circulation seems to be time-consuming, and illogical. If it can be shown to be safer we may change but only then.

However, one other point could be made in relation to neo-natal repair stemming from Sir Brian Barratt-Boyes presentation of his results at the International Meeting in Auckland earlier this year; namely, that out flow patches were required in some 50% of his cases. This is perhaps due to the difficulty, in hearts of this size, in deciding whether out flow tract enlargement is needed or not. 50% is probably too high and time alone will tell whether this is a long-term disadvantage.

When conventional by-pass techniques are used in older patients there are further options to be considered. Namely, how the myocardium is to be protected during aortic cross clamping. The clearing of the outflow tract, the insertion of a V.S.D. path and perhaps with the addition of an out flow patch, takes us from 25 to 50 minutes. This is too long, in our opinion, for a single period af anoxic arrest. We have abandoned intermittent cross clamping and chosen to follow the iced saline technique, as used by Shumway with a left atrial suction vent in preference to venting through the apex of the left ventricle. Shumway's technique provides ideal conditions for patch placement and so reduces the risk of A-V block and patch leakage. This technique has been used by us in all cases for the last six years and because of the favourable conditions created for the surgery and the results we are getting at the present time we are unlikely to change.

In common with other series there has been virtually no choice in the method of closing the septal defect because there were only four direct closures in contrast with the remaining 115 who required dacron patches. The four direct closures were in cases where, even though the diameter of the defect was large enough to create a physiological single ventricle, the anatomical structure and fibrous margins of the defect warranted closure by direct suture.

It is difficult to lay down rules about the need for out-flow patches, and what to use for the purpose. As for the material used we have gradually changed over the years from using closely woven teflon in all cases to using pericardium in all cases. Teflon is perhaps easier to use when beginning this work, but because it is non-compliant it is more difficult to achieve a wide lumen with it at the level of the orifices of the pulmonary arteries when patching is necessary up to this level. Teflon may also be safer if the patch is mainly required in the out-flow region or just across the valve ring. All these decisions are very marginal and there are too many factors involved to attempt to generalise.

Having become used to using pericardium for Mustards operation, and being no longer driven mad by its cantankerous sewing qualities, it is clearly our first choice at the present time mainly because it moulds so well to natural contours.

The decision whether to use an out-flow patch at all can be a difficult one. It is impossible to generalise because of the anatomical variations encountered. It is dangerous to lay down a strict criterion of right ventricular pressure level relative to systemic pressure which should be judged as acceptable in any particular case. It must be left to the surgeon to achieve as good a technical result as possible in each case and then to accept what he gets. Going back on the by-pass and putting in longer and longer patches at second attempts usually doesn't improve the situation and costs the patient increasing time on by-pass, with more manipulation and more cross clamping all of which may add up to too much surgery with a resultant rise in mortality. With increasing experience the surgeon will fully understand the problem, he will do his technical best at the first attempt and with this will come his best results.

Perhaps the major decision which faces most of us is when the clinical condition demands treatment at an age below the chosen age of election for full correction.

If the decision is against full correction because of age or for any other reason then a holding operation may be necessary. Here again there are a number of options dictated, by the age of the patient at the time the shunt is to be performed.

In our own experience and considering all factors, we have chosen the age of four to five years as a "convenient" age for full repair in non-shunted patients. In the last ten years no patient has been shunted in our Unit over the age of four years. This decision is clearly related to the age policy we follow for full correction. We can also show that our operative mortality on patients previously shunted is not significantly higher than on unshunted patients.

In the special circumstances where by-pass facilities are not available a shunt may be necessary above the age of four years and we have no doubt that Blalock's shunt is the operation of choice for older children. In the younger age group we firmly believe that Waterston's shunt is the procedure of choice.

As a corollary to the above, it is our policy to fully correct cases with favourable anatomy down to say 1½ years of age but with this exception children needing surgical help below the age of say three years are shunted. As we are only shunting very young children and more particularly as half the cases shunted are from one week to one year of age this is the reason we strongly favour the Waterston procedure. This has the same draw-backs as the Potts anastomosis but with meticulous attention to the technical details of the anastomosis, which is best performed with magnifying loupes as for coronary artery surgery, then uniformly good results can be expected. In the past Cooleys modification has been used; but the intra-pericardial adhesions resulting create problems at the subsequent full correction and this has convinced us that the extra-pericardial approach as described by Waterston is preferable.

The difficulties and the hazards associated with the closing of a Pott's shunt are enough to rule this out as an operation of choice. Pott's anastomosis should now only be used as an alternative to a planned left sided Blalock shunt which proves to be technically impossible. If a Pott's anastomosis has to be used the anastomosis should be placed as proximal as possible on the left pulmonary artery to facilitate subsequent closure. I am personally grateful that we have only four more Fallots with Pott's shunts requiring full correction and I hope there will be no more.

Of the 150 patients in the series only one has had a Brock's palliative procedure. This was done, on by-pass, in a boy with a "very severe" Fallot situation. Some years previously a shunt had been attempted but no pulmonary artery large enough for anastomosis was found. It is our firm conviction that even though this case has proved to be a signal success so far (and may even

come to full correction later) that this technique should be reserved for these very special circumstances. Brock's procedure should not be employed as an alternative palliative procedure to shunting.

This type of case represents the "extreme" end of the Fallot range.

Between this extreme case (and we have only had two such cases in the whole series) and the "mild" Fallot there are the "severe" Fallots. To an experienced team these are a challenge and provide most of the mortality in the big series. For an inexperienced team they should be palliated if possible either by shunting which will be technically difficult, or by sending them to somebody else!

In any event the risks are high but if successful the rewards are also high.

In summary the surgical treatment of Tetralogy of Fallot has much to offer. Because of our geographical situation we have been able to follow up all but one of our cases who have survived operation. This child was corrected in 1961, known to be well for some years and has since been lost. Analysis shows that of 119 cases fully corrected 8 (6.1%) died at operation, 4 (3.3%) died late deaths, (2 with heart block, 1 with peripheral pulmonary artery obstruction and 1 with digitalis overdosage for an atrial arrythmia). Six cases are classified as poor or only fair results and five of this group have proven peripheral pulmonary artery obstruction and the other one has tricuspid incompetence probably due to papillary muscle detachment at operation. In all, this leaves 100 (84%) patients from the original group submitted to full correction living normal or near normal lives, many engaging in strenuous competitive sport. In this same period of time there have been 15 so-called "Pink" Fallots operated but not included in the series, with no mortality and all with good post-operative results on follow-up.

Taking the original clinical condition into account this is a remarkable state of affairs. Figures with the same quantity and quality of survival have been shown in many other series, often larger than ours and all coming from experienced surgical groups. There is a clear message from the extensive literature on the surgery of the Tetralogy of Fallot that survival is almost synonomous with a good to excellent functional result. It is also clear, but for obvious reasons not well documented in the literature, that the surgery of this condition is not for the part time cardiac surgeon, because his mortality in this particular condition is too high. I have tried to show that there are many roads to be chosen for those setting out, each one has its special advantages and its pitfalls but when the correct path is selected the end result is well worth the considerable effort required.