

# Cardio-Pulmonary Bypass During Tetralogy of Fallot Repair

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## INTRODUCTION

Among congenital abnormalities, Tetralogy of Fallot is one of the more difficult cases for the perfusionist. Special detail must be given to priming for these cases since the patient is usually cyanotic and had an elevated hematocrit before bypass. During the past three years we have done one hundred seventeen tetralogy repairs at The Johns Hopkins Hospital. Outlined below is the technique we have found, has given us the best results.

From an anatomical stand point in Tetralogy of Fallot the right ventricular outflow tract is narrowed to various degrees by hypertrophied muscle and a variable amount of fibromuscular tissue in the infundibular area. A large interventricular septal defect is located just below the aortic annulus. The direction and the volume of the shunt across the defect are entirely dependent upon the amount of resistance at the right ventricular outflow tract compared to aortic (systemic) resistance.

Cyanosis is usually not present at birth or in early infancy. It may not appear until later in childhood. Affected children are usually cyanotic and poorly developed. The development of cyanosis and polycythemia depends upon the amount of right-to-left shunting and the reduction in pulmonary perfusion.

## TECHNIQUE

Because of the polycythemia the hematocrits of some patients preoperatively was as high as seventy-two per cent. The prime selected therefore should take into account the increased volume of red cells. In the adult patients (over 50 kilograms) and with a hematocrit over thirty-two per cent, the standard prime of 2000cc lactated Ringers with five per cent dextrose and 8000 units sodium heparin, were used.<sup>1,2</sup> In the small adult, pediatric, and infant patients the prime varied from case to case. After calculating the patients blood and red cell volumes and considering the pump circuit volume, a prime is calculated so that a total circuit hematocrit of thirty per cent is obtained.<sup>3,4</sup> In the infant (below fifteen kilograms) fresh heparinized red cells are used for the prime. In the small adult and the pediatric patients, red cells stored in CPD solution are used. Citrate Phosphate Dextrose solution USP has replaced Acid Citrate Dextrose at this institution due to advantages such as increased pH levels and better maintenance of 2,3-DPG and ATP levels during storage. In cases where the hematocrit is high it is sometimes possible to prime using no additional red cells. The prime is then made up of Ringers Lactate and fresh plasma or Plasmanate.\* Lactated Ringers may be added in amounts up to the daily fluid intake requirements of the patient. Fresh plasma or Plasmanate is used for the remainder of the volume. Dextrose fifty per cent USP is added to all primes in an amount sufficient to make a five per cent volume in the prime. The dextrose provides substrate for the myocardial metabolism and also aids in the diuresis of the patient. In the primes that use CPD blood or Ringers, heparin is added at the rate of 2000 units per 500cc prime volume.

Prior to the placement of the perfusion cannulae, the patient is heparinized with an initial dose of 400 units (4mg) per kilogram of body weight. Because of the lesser sensitivity of children to heparin, one third of the initial dose is repeated at the end of two hours of bypass and then at hourly intervals till the end of bypass.<sup>5,6</sup> The ascending aorta is cannulated for the return of the arterialized blood to the patient. Venous blood is returned to the pump via cannulae placed in the superior and inferior vena cavae. In the small infants undergoing deep hypothermia the blood is returned using a single atrial basket cannula. Flow rates range from 50 cc per kilogram in the adult patients

\*Manufactured by Cutter Laboratories, Inc., Berkley, California

to as high as 200 cc per kilogram in the infants. In the adults the arterial pressure is maintained at 60 mmHg or above. If during the bypass the optimum flow was reached for the size of the patient and the pressure still fell below the lower limit, phenylephrine (0.10-1.0mg) or mephenteramine (3-15mg) were given. We find that in adult and larger pediatric cases that an arterial pressure of over 60 mmHg, gives the best results both in adequate tissue and cerebral oxygenation and renal function as reflected by normal blood gas determinations and adequate urine output.<sup>7</sup>

Central venous pressure should range from 0-3 cm. of water and any sudden decrease of venous return should be checked immediately as this could indicate a blockage of one of the vena cava cannula which could lead to severe physiological damage. Blood gas evaluation of acid base should be done routinely on all cases. In the adults a sample is obtained one-half hour after starting bypass and subsequent samples were done at one hour intervals.

In the pediatric and infant cases more frequent sampling was done with samples done as often as once every fifteen minutes. The arterial O<sub>2</sub> partial pressure should be adjusted between 150-225 mmHg and the CO<sub>2</sub> partial pressures between 30-40 mmHg. The use of high O<sub>2</sub> partial pressures has a toxic effect<sup>8</sup> on the blood and has the negative effect of causing CO<sub>2</sub> washout which leads to severe respiratory alkalemia and an upward shift of pH to as high as 7.7. When the CO<sub>2</sub> washout exceeds the metabolic CO<sub>2</sub> production respiratory alkalemia will lead to increased cerebral vascular resistance and increased lactate levels.<sup>9, 10</sup>

Previous surgical shunts should be taken down if possible before going on bypass. This will help eliminate excess amounts of pulmonary blood flow during bypass, and will reduce the amount of cardiotomy suction return.

Moderate hypothermia (28-29°C) was utilized for most patients and was found to provide better results than normothermia or slight hypothermia.

Immediately at the end of bypass, blood gases, hematocrit level, electrolytes are determined and necessary corrections are made. An attempt is made to transfuse as much blood as possible from the pump oxygenator reservoir to maintain venous pressure during the first fifteen minutes post bypass. This helps to decrease the amount of fresh bank blood required to maintain adequate blood volume. All cannulae and lines are then disconnected and all further blood losses are replaced with fresh bank blood. Heparin is converted using a 2:1 ratio of protimine, taking into consideration the active half life of all heparin used.

## DISCUSSION

Patients with Tetralogy of Fallot represents a reasonable challenge to the perfusionist because of the wide range of preoperative physiological parameters. In addition to the wide age range these patients can present with marked abnormalities in Hematocrit, acid base balance, and clotting parameters.

We have elected to use moderate hypothermia to protect the myocardium during aortic cross clamping. One must be aware of the changes in the solubility of oxygen and carbon dioxide in the blood and the resultant shift in blood gas parameters (see table 1). As the body temperature is reduced the oxygen requirements are substantially lowered giving greater tissue and myocardial protection (see table 2). In the pediatric and infant patients there was a tendency sometimes in the first five or ten minutes of bypass for the arterial pressure to drop as low as 20-25 mmHg. Sometimes this could be attributed to poor venous return resulting from a cannula malposition. Usually, however, it seemed to relate to a general systematic vasodilation. Increasing the perfusion rate to the maximum calculated flow sometimes would correct the problem. The increase of the blood flow beyond the maximum will not guarantee adequate tissue perfusion because of venous shunting found in patients with Tetralogy of Fallot. If within five minutes the increase in flow does not raise the pressure then small doses of vasopressor (usually 0.1-0.5mg phenylephrine) are given till the pressure reached 40mmHg. A sump vent is used because of the large amount of blood returning to the heart via collateral pulmonary circulation.

Over the past three years we have done one hundred seventeen tetralogy repairs. Overall mortality rate was 8.4 per cent. The cause of death in these patients included heart failure, heart block, postoperative hemorrhage, intratracheal hemorrhage, and pulmonary embolism. There was no evidence that any of the deaths were in any way related

to the cardiopulmonary bypass. In the surviving group, no evidence of complications resulting from perfusion were observed.

**TABLE 1**  
Effect of Hypothermia on Blood Oxygen and Carbon Dioxide Gases

	Oxygen	Carbon dioxide
Solubility of gas in fluids	Increase	Increase
Changes seen in blood gas analysis	Dissociation curve shift to left (less oxygen at the tissue)	Carbon dioxide tension decrease while carbon dioxide content remains constant

**TABLE 2**  
Decrease in Oxygen Requirement as the Temperature is Reduced

Degree hypothermia	Temperature	Oxygen Requirement
Mild	35°-28°C.	1/2 (30°C.)
Moderate	27°-21°C.	1/3 (25°C.)
Deep	20°-15°C.	1/5 (20°C.)
Profound	Below 15°C.	1/6 (10°C.)

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Ms. Emily P. Taylor  
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Dear Ms. Taylor:

Paula Steichen, the author of the short article which follows, is the daughter of Helga Crile who in turn is the daughter of the poet, Carl Sandburg. The ability to write well has obviously been passed on to her.

Somewhere in the hills of North Carolina, Paula's neighbor, Alvin was in renal failure as a result of longstanding diabetes. He was only 40 years old. There was no one in the family who was literate, and he lived about 40 miles from the nearest dialysis center. And so the granddaughter of Carl Sandburg undertook to learn home dialysis techniques and for eight months performed it on Alvin. When her only patient died, she wrote a letter to Drs. Crews and Moser to thank them for the help they provided all through Alvin's illness.

Unlike other centers, we have always been treating patients with diabetes and renal failure. It's difficult, but sometimes very rewarding. The general trend throughout the country now is to accept these patients rather than abandon them. From that point of view, Paula Steichen's letter is very opportune. Here is the letter:

"Even before Alvin died, I often thought of writing to both of you, to tell you some of our thoughts and feelings concerning dialysis. I realize dialysis is a relatively young venture—especially for diabetics—and it might be of some help to let you know how Alvin and I and his family felt about it. Everything that I will say in this letter, except that which concerns only me, I have discussed at length with Alvin and Geneva—we were all in accord on our thoughts.

"The most important thing that I wish to say is that dialysis was for all of us completely worthwhile. I hope that there will never come a time when you will refuse diabetics on the dialysis program because of the briefness of their life expectancy. The only elements that I think are essential to accepting a patient on the dialysis program are that the patient have a distinct reason for living and that he have friends or family who are relatively strong and are devoted to him.

"From a doctor's point of view there must be some question as to whether or not it is worthwhile to prolong a patient's life by only six or eight months. I feel strongly that it is not just a question of time in such a situation, but, also of more intangible riches. From a practical point of view, these last months afforded Alvin the chance to verbally set forth certain goals for his children. Just one example of this is that in the fall Alvin made his family assure him that none of the children would quit school before graduating from high school. In a neighborhood where fully 90% of all adults and children either never finished school or never will, this was a tremendously important act. It is difficult to give you the full scope of all that Alvin did for his children, his relatives and friends in these last months. It was hard, I know, for his children to see him on the kidney machine, and, more so, to see him vomiting almost continuously during the last weeks—but they gained a great deal, too. He left them a legacy of quiet strength and endurance and determination. The day before he died he planted a 400 foot row of potatoes and his attendance to church, regardless of how he felt, has become something of a legend. The preacher at Alvin's church declared when Alvin was still living that he had done more for the church than all the preaching there had ever done.

“One of the most important things that the kidney machine did for Alvin and Geneva, his children and me, was to lend to us the element of hope. I cannot begin to tell you how important this was. I often thought how terrible it would have been if Alvin had been doomed to die of uremic poisoning and he had known that he had only a month to live. For even the most healthy individual it is of great importance to have some concept of a future, some feeling of hope. Many uninvolved onlookers thought of the kidney machine as a horrible device. But in this one way, at least, it came closer to being an angel and it enabled Alvin to live a much more normal life before death than he otherwise would have been able to live. At times Alvin thought he would die soon, but at other times (particularly before that last month) he felt he had years ahead—and we would plan for the steers, fruit trees and gardens of the springs and summers before us. I think I should tell you that even when Alvin was so miserable toward the end, even though he often said that he wanted to die then, he, however, never said that he wanted to stop dialysis. I had once told him that by stopping his treatments on the kidney machine, he could die within a month’s time.

“This element of hope, which the machine gave us, was of course, what kept me going, too. Realistically, I knew Alvin’s life span was limited—but there is something about operating the machine, about being so closely involved with keeping a person alive, that makes the idea of defeat seem an impossibility. In this sense I think Alvin’s death was more difficult for me to accept as a reality than for anyone else. Everyone who knew him had been expecting his death, but even after he died I could not rid myself of that feeling of hope that had become so firmly entrenched during all those months. I suppose it sounds unbalanced, but, even during the wake, when they brought Alvin back home for twenty-four hours, the thought constantly stirred within me that if I could only get Alvin out of that coffin and back into the garden or the fields, I could somehow bring him back to life. Geneva, who is a remarkable woman in many ways and who has the mountaineer’s acceptance of life and death, was close enough to me and understood so well that she finally persuaded me to touch Alvin’s cold, hard hands and feel the reality of his death.\*

“I am not doing a very good job of explaining—but what I am trying to say in essence is that the irony of dialysis is that the closer you are to it, the less terrible it becomes and the more of a blessing it becomes. I hated putting the needles in Alvin and I hated every bit of discomfort that he went through. It is impossible to tell you how hard it was for me to see him suffer. And yet, despite all this, I came to feel for that machine an eerie, almost human—directed love—for it was the thing that enabled Alvin and Geneva and me to have what time and happiness and hope there was.

“There is more to say, but this letter has gone on too long. More than anything, now, I want to tell you how much I appreciate the two of you. In many ways dialysis is very, very lonely and sometimes very frightening. The two of you, in your different ways, made things so much easier for me. I always felt badly about calling you on the phone, knowing how busy you were, and yet I would be so worried about Alvin that I finally would call—and you unfailingly gave me patience, reassurance, and, above all, sound medical advice. I am not exaggerating when I say that you made the difference between dialysis being an unbearable ordeal or an experience which we simply had to confront and work with. My admiration for the two of you is unbounded and I cannot thank you enough for all that you did.”

Not only is Paula Steichen’s article very well written, but I also believe that this is a beautiful example of help, love, and support she has given to a neighbor without any financial compensation.

Sincerely yours,  
W. J. Kolff, M.D., Ph.D.

\*(He died from abdominal vascular complications, certain hemorrhages, possible perforation, and peritonitis.)