Management of Transposition of the Great Arteries in Infants

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The principal aim of any operation in an infant with a congenital cardiac anomaly is to improve hemodynamic function. To this end, palliative and corrective operations must be selected and timed to attain maximum benefit with minimal risk. In recent years, palliative operations in infants have been progressively supplanted by corrective ones, because mortality rates for the latter have declined substantially, and because the late hemodynamic results of early correction have proven equal to those obtained by delayed repair. Nonetheless, areas of controversy still remain.

Transposition of the Great Arteries (TGA) is an anatomic deformity of such severity that the hemodynamic derangement usually produces symptoms that require intervention early in life. As a result, the most critical decisions about patients with TGA must usually be made in infancy. This report will briefly review the anatomy and physiology of TGA, the various means of treatment, techniques of operation and perfusion, and our current feelings about the timing and selection of operative approaches in infants.

ANATOMY AND PHYSIOLOGY

TGA consists essentially of a reversed connection between the heart and the great arteries, so that right ventricular blood (unoxygenated) is ejected into the aorta, and left ventricular blood (oxygenated) is ejected into the pulmonary artery. This arrangement is inherently incompatible with life, since the oxygenated blood circulates entirely in the pulmonary circuit and never reaches the peripheral tissues. Survival is possible only if a defect between the two sides of the heart such as an atrial or ventricular septal defect (ASD or VSD) permits mixing of oxygenated and unoxygenated blood, thus allowing some oxygenated blood to reach the periphery. If the only defect present is a small patent foramen ovale (a naturally occurring opening in the atrial septum), severe cyanosis is usually present from birth.

PALLIATIVE OPERATIONS

These are designed to create a larger ASD, usually by the method Blalock and Hanlon described in 1950.[1] This operation creates an ASD by excising the posterior portion of the interatrial septum in an ingenious closed-heart procedure. Open-heart techniques of excising the septum under direct vision with inflow (caval) occlusion have had a higher operative mortality in our hands.[2]

Unlike the case with Tetralogy of Fallot, creation of a high pressure shunt between the aorta and pulmonary artery is not advantageous in uncomplicated TGA, because there is no pathway by which this shunted blood can readily return to the systemic circulation. Most infants with TGA already have greatly increased pulmonary blood flow, and congestive heart failure is common; a shunt merely adds to these problems.

In 1965, Rashkind and Miller described a non-operative technique of creating an ASD with a balloon-tipped catheter.[3] The catheter is inserted through the femoral vein, the deflated balloon is passed through the foramen ovale into the left atrium, and the balloon is then inflated and withdrawn forcibly into the right atrium, thereby rupturing the flap valve of the foramen ovale. The procedure is relatively safe, and leaves the pericardial space unviolated, thus simplifying subsequent operations. It is now universally accepted as the initial procedure of choice in symptomatic infants, and may even be repeated later if subsequent deterioration occurs. The improvement provided by balloon septostomy, though frequently substantial and durable, may be brief and of unsatisfactory

quality. In some cases, this may be because of failure to adequately rupture a particularly tough or elastic atrial septum. (Indeed, in our own experience, at the time of total correction of TGA in infants who have previously undergone balloon septostomy we have commonly found stretching of the foramen ovale with no distinct tear and no disruption of the septum.) In other cases, however, there is little improvement even though subsequent direct inspection of the septum at operation reveals an adequate defect. In these cases it is likely that balanced pressures in the two atria have prevented good mixing despite an adequate defect.

PALLIATIVE VS. CORRECTIVE OPERATIONS

When severe cyanosis, exercise intolerance, congestive heart failure or marked retardation of growth and development persist after balloon septostomy, surgical intervention must be considered, and the choice made between a palliative or a corrective operation. The Blalock-Hanlon operation has had a mortality of approximately 20% in most reported series, and in certain cases provides only modest improvement, probably for the same reasons cited earlier in the discussion of balloon septostomy. Since correction of TGA with current techniques can now be carried out with a mortality as low or lower than that for the Blalock-Hanlon procedure, we have for several years advocated the Mustard operation as the best initial choice in infants with uncomplicated TGA.[2] When we last reviewed our experience, 10 infants with uncomplicated TGA had undergone corrective operations under two years of age with two operative deaths.[4] With increasing experience, the mortality should decline further.

The prompt resumption of normal growth and development after correction of TGA contrasts markedly with the course of infants who are followed for prolonged periods after balloon septostomy. (Fig. 1) The low mortality of total correction in infancy suggests that survival alone should not be deemed successful palliation following balloon septostomy, but that adequate relief of cyanosis and congestive heart failure as well as reasonable growth and development should be sought. If palliation is inadequate by these criteria, total correction in infancy should be carried out before there is irreversible physical retardation.

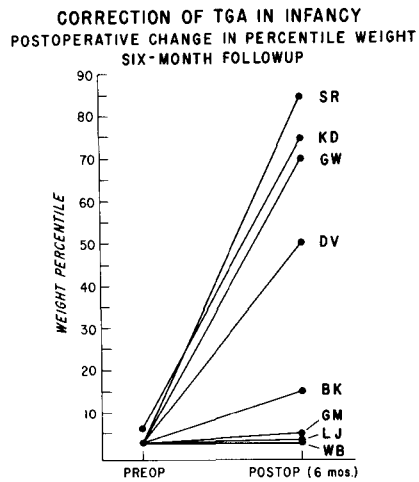

Fig. 1. Preoperative and 6-month postoperative weights of the 8 youngest survivors expressed as percentiles from standard growth charts. W. B., L. J., and G. M. were far below the third percentile preoperatively, and their substantial weight gain was not reflected in significant percentile changes 7 months postoperatively.

TECHNIQUES OF PREOPERATIVE AND OPERATIVE MANAGEMENT

Until very recently, all intracardiac operations in our clinic have been carried out with high flow cardiopulmonary bypass and moderate hypothermia.[4] This discussion will be confined to those techniques.

PREOPERATIVE PREPARATION AND ANESTHESIA:

Deficits in serum electrolytes are corrected meticulously prior to operation, but since nutritional deficits respond poorly to intensive management until the cardiac abnormality is corrected, their presence does not delay operation. Preoperative digitalization with digoxin is routinely carried out, but digoxin is withheld beginning 24 hours prior to operation. Premedication consists of atropine and morphine. Anesthesia is induced with intramuscularly administered ketamine and is maintained with nitrous oxide and supplemental ketamine given intravenously. Halothane was used early in our experience, but ketamine is now preferred because it is not a myocardial depressant and it is antiarrhythmic. Morphine is no longer used extensively because it is metabolized slowly in infants, and recovery is prolonged after high doses. Uncuffed nasotracheal tubes are used routinely and are maintained for postoperative respiratory support. Doppler ultrasonic transcutaneous flow probes in conjunction with standard manometer cuffs are routinely used for blood pressure determination. Arterial catheters have been eliminated completely in recent years to save time, to prevent catheter occlusion at inconvenient moments, and to avoid problems with flushing solutions that increase the fluid load in small infants.

OPERATIVE TECHNIQUES: In 1964, Mustard described a corrective operation that transposes venous return with an intraatrial baffle with techniques proposed by Albert in 1955.[5-6] Following excision of the atrial septum, a baffle of pericardium or synthetic fabric is inserted to create a new septum that directs caval blood through the mitral valve to the left ventricle, while pulmonary venous blood returns through the tricuspid valve to the right ventricle.

Operation is performed at normothermia with high-flow cardiopulmonary bypass (2.4 liters per minute per square meter of body surface area) utilizing a disposable infant oxygenator primed with fresh heparinized or suitably buffered ACD or CPD blood. The cavae are cannulated at their junctions with the atrium as far to the patient's right as possible so that the atriotomy can be made to the left of the catheters to facilitate exposure. The ascending aorta is cannulated for arterial return. Electrical fibrillation is induced and maintained with the same wire that is used for ventricular pacing postoperatively, and no ventricular vents are used. The atrial baffle is made from a segment of pericardium cut in a generous rectangle with no preliminary sculpturing. This provides a baffle that is always adequate in circumference and that can be accurately and simply tailored during insertion if necessary. Suturing is begun at the left edge of the orifices of the left pulmonary veins (Fig. 2) and proceeds to the right and superiorly to exclude the superior caval catheter. (Fig. 3) Similarly, a suture is carried to the right and inferiorly to exclude the inferior caval catheter and the coronary sinus. (Fig. 4) Sutures are placed superficially

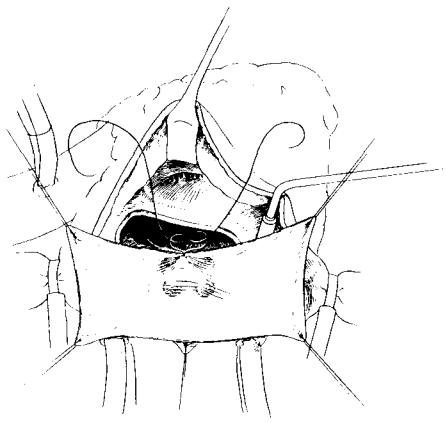

FIGURE 2.

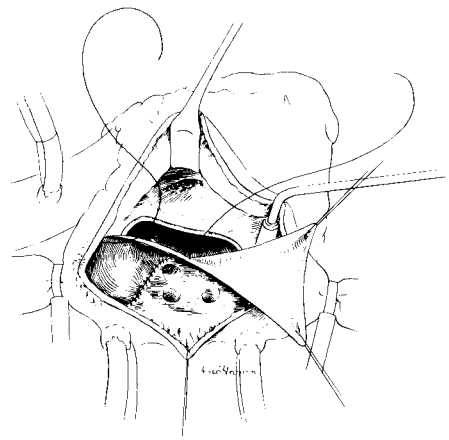

FIGURE 3.

Fig. 2. The rectangular pericardial patch is used without preliminary sculpturing. Suturing begins at the left edge of the orifices of the left pulmonary veins.

Fig. 3. Suturing proceeds to the right and superiorly to exclude the superior caval catheter.

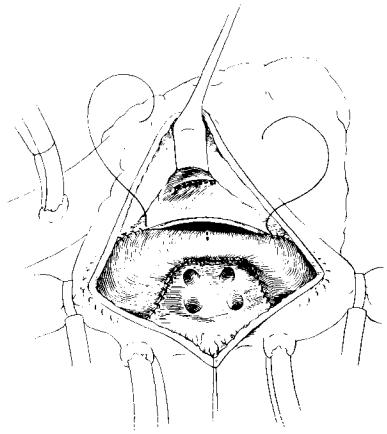

Fig. 4. Suturing then proceeds inferiorly to surround the inferior cava. Minor readjustments in patch circumference can easily be made by incision or excision at the dotted line.

around the coronary sinus, and it has been completely unnecessary to employ incisions of the sinus or the Aberdeen flap.

Intermittent aortic cross-clamping is used to facilitate exposure if needed. When 1 or 2 cm. of baffle remain to be sutured along the interatrial septal remnant, minor adjustments of the baffle's circumference can be made readily.

SELECTION OF THE OXYGENATOR SYSTEM FOR CARDIOPULMONARY BYPASS

A 6-inch disc oxygenator was used for infant patients before 1968, but this has now been completely discarded. The Temptrol bubble oxygenator is employed routinely at present because of its convenience and ease of assembly, disposability, small priming volume, integral heat exchanger, and effective bubble dissipation. A separate bubble trap has been unnecessary. High blood flow rates are sustained with no accompanying difficulty in complete oxygenation or dissipation of carbon dioxide. Metabolic acidosis has not occurred as a result of bypass, and buffers have not been administered during perfusion as they obscure an assessment of the adequacy of bypass. At a flow rate of 2.4 liters per minute per square meter of body surface area, tissue oxygen consumption is determined by oxygen needs and is not limited by oxygen supply.[7] This flow rate corresponds to more than 100 ml. per kilogram in infants. A normal buffer base concentration was present at the conclusion of bypass in all our patients without the addition of buffers to the pump although many infants were overtly acidotic at the beginning of bypass.

Membrane oxygenators are improving rapidly, and may eventually supplant bubble oxygenators. At present, however, their gas exchange capacity is not as great. Urgent operations are commonly performed on cyanotic infants with profound respiratory and metabolic acidosis, and the bubble oxygenator system with its abundant reserve is able to restore normal blood gas concentrations and to correct metabolic acidosis during the bypass period. Although membrane oxygenators cause less deterioration of pulmonary function than do all oxygenator systems with an air-blood interface, this advantage is primarily apparent during long perfusions. Since the length of perfusion in infants is relatively short, the major advantages of the membrane oxygenator with regard to protein denaturation and destruction of formed elements in the blood are not as readily apparent.

Hemolysis during cardiac operations is primarily related to cardiotomy suction with its air-blood interface and mechanical trauma to blood[8] and membrane oxygenators thus cannot prevent hemolysis when suction is required.

RATIONALE FOR THE USE OF CARDIOPULMONARY BYPASS

When intracardiac operations in infants first became common in the early 1960's, standard cardiopulmonary bypass techniques were usually employed. Well designed mini-

aturized equipment rapidly became available, and with the introduction of infant-sized disposable bubble oxygenators, virtually all intracardiac operations in infants became technically possible. There has been increasing enthusiasm, however, for the technique of profound hypothermia with circulatory arrest during intracardiac repair, combined with limited cardiopulmonary bypass for a portion of the cooling phase and for all of the rewarming phase. This technique was introduced by the group in Kyoto, Japan,[9] and has been most extensively and successfully applied by Barratt-Boyes.[10]

Our experience thus far has been predominantly with cardiopulmonary bypass, because it avoids the potential for cerebral injury that is associated with circulatory arrest. Psychological and social developmental testing in our patients who have undergone correction of tetralogy of Fallot has revealed normal or above-normal intellectual and social development.[11] Similar studies after long-term follow-up are not yet available in infants who have had total circulatory arrest. Many infants have severe metabolic acidosis prior to bypass as a result of congestive heart failure, profound cyanosis, and low cardiac output. High-flow cardiopulmonary bypass corrects these abnormalities simply by restoring normal systemic flow, as noted above. Hypothermia may be induced during cardiopulmonary bypass when brief circulatory arrest is essential. Intermittent aortic cross-clamping is available to provide a quiet field and myocardial relaxation. Bronchial collateral return can be minimized by mild hypothermia and temporary reduction in the perfusion rate.

Nonetheless, in the smallest neonates, the caval catheters placed through the right atrium are often inconvenient, and the return of blood to the left atrium from mediastinal collateral vessels supplying the lungs can be distracting. In the youngest and smallest infants, the insertion of the caval catheters can be hazardous in itself.

In view of these problems, if long-term follow-up proves that cerebral function is unaffected by circulatory arrest under deep hypothermia for prolonged periods, the latter technique will be preferable in small neonates.

COMMENTS

It may perhaps be unduly optimistic to consider the Mustard operation "corrective," since "normal" pathways of blood flow are not created. In fact, late problems caused by shrinkage of the baffle and/or inadequate growth of the atrial walls have been reported. These include caval inflow obstruction (usually superior cava) and—less frequently—pulmonary venous obstruction. In addition, the long-term function of the tricuspid valve, which must withstand systemic pressure in the right ventricle, remains in doubt. In "congenitally corrected TGA," in which the valve between the left atrium and the systemic ventricle also has tricuspid morphology, there is a 15-20% incidence of tricuspid insufficiency, and it is reasonable to anticipate a similar incidence with surgcally corrected TGA.

Also, not all cases of TGA are uncomplicated. Many have an associated VSD, and some also have pulmonic (or subpulmonic) stenosis that impedes ejection of blood from the *left* ventricle into the pulmonary artery. In these complex cases, total repair in the smallest infants is not yet satisfactory, and palliative operations are usually wise.

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