Palliation for transposition of great arteries

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Abstract

Background: At the University College Hospital Ibadan we have no facility for total surgical correction of transportation of the great arteries (TGA). This prospective study reviews the palliative procedures we have used in the management of TGA.
Method: Patients with the diagnosis of TGA were evaluated for morphological type. The choice of palliative procedure was made in some of the patients with morphological type in mind. No fixed criteria were used for allocating patients to Blalock-Hanlon (B-H), atrial septectomy while pulmonary banding (PB) and Blalock-Taussig (B-T) shunt have definite indications.
Results: Fourteen consecutive patients with TGA were palliated. The ages of these patients ranged between 3 to 11 months (6.8 ± 2.4 months), there were 8 males to 6 females (1.3:1). Six patients had B-H atrial septectomy and 2(33.3%) died within 48 hours, 4 patients had B-T shunt and there were no mortality, 4 patients had PB and 2 (50.0%) died within 72 hours. The overall operative mortality was 28.6%. All the 10 survivors had improvement of their clinical features and fall in packed cell volume during the period of follow-up, which lasted 5 to 13 months (mean 9.3 ± 1.2 months). All patients had delayed wound healing.
Conclusion: Appropriate and timely palliative surgery has a place in patients with TGA as an interim care.

Key words: Palliative surgery, transposition, great arteries

Introduction

Transposition of the great arteries (TGA) was first observed by Steno in 1672[1] and was subsequently described by Morgagni (1761) [2] and Baille (1797). [1,2] In this severe congenital cyanotic heart disease, the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle in the patient with atrio-ventricular concordance. The physiologic effects are acute, with cyanosis and distress usually obvious at birth. Survival depends on mixing of blood between pulmonary and systemic circulation, at atrial level chiefly via a patent foramen ovale, and to a lesser extent at arterial level via the patent ductus arteriosus.

These may sometimes coexist with ventricular septal defect. [2] Before the development of effective palliative surgery for TGA 50% of infants died in the first month and 85% in the first six months of life. [3] Cyanosis, hypoxemic deterioration, or heart failure with early death is the usual clinical course in the untreated infant with complete TGA.[4]

Initial palliation and prolongation of life was achieved by atrial septectomy described by Blalock and Hanlon in 1950 [5]. Subsequently with the advent of cardiac catherisation, the procedure of balloon atrial septostomy (BAS) became popular. [6-7] When BAS is successful in improving the clinical status of the patient and in reducing left atrial pressure, corrective operation can be deferred until a later date when the child is older and larger; with an improved operative risk. [1]However, there are several anatomic and functional factors that have been integrated to form four clinical types of TGA. In clinical type 1 TGA the patient has intact ventricular septum (IVS) or small ventricular septal defect (VSD) with increased pulmonary blood flow (PBF) and small intercirculatory shunting (ICS). Type 2 TGA is characterized by large VSD with increased PBF and large ICS. Patients with type 3 TGA have VSD and left ventricular outflow obstruction (LVOTO), they have restricted PBF. In 4th clinical type, there is a VSD and pulmonary vascular obstructive disease (PVOD) with restriction of PBF. The later type is not amenable to meaningful palliation. The clinical manifestations and course of these variants are influenced essentially by the extent of intercirculatory mixing. [4] This is a report of our experience with the various palliative options in a situation where no facility exists for definitive correction.

Materials and Methods

Fourteen consecutive patients with TGA whose cardiac morphology had been defined by Paediatric Cardiologist were referred to the Cardiothoracic Surgery Unit of the University College Hospital, Ibadan between 1985 and 1996. Diagnostic investigations done included chest radiograph, electrocardiogram, angiocardiography and/or echocardiography.

Initial resuscitation included correction of metabolic acidosis, maintenance of normothermia, treatment of congestive heart failure when present and treatment of hypoglycemia. Adequate ventilatory care was provided for the profoundly distressed infant. The choice of procedure was made after joint evaluation of morphological type of TGA by the cardiologists and the cardiothoracic surgeons. The surgical options were Blalock-Hanlon (B-H), pulmonary banding (PB), and modified Blalock-Taussing (B-T). No fixed criteria were used for allocating a patient to B-H atrial septectomy. The patients earmarked for pulmonary banding had pulmonary plethora, large VSD and refractory heart failure. Any patient who had pulmonary oligaemia had modified Blalock-Taussig to "improve" pulmonary blood supply.

Results

The ages at presentation ranged between 3 to 11 months (6.8 $\pm$ 2.4 months). There were 8 males to 6 females (1.3:1). All patients were deeply cyanosed while six of them were admitted with congestive cardiac failure (CCF).

CCF was refractory in two patients who exhibited profound respiratory distress. Among the remaining 4 patients with CCF, two subsequently had minimal respiratory difficulty from recurrent chest infection, while the other 2 had improved breathing preoperatively. The preoperative packed cell volume (PCV) ranged from 68% to 78% (mean 72%). All of the patients had metabolic acidosis; this was severe (HCO_3 < 15 mmol/L) in the six patients with CCF.

Six patients aged between 3 to 6 months had Blalock-Hanlon performed of which two died (33.3%) within 48 hours of surgery. Four patients between ages 6 and 11 months had modified Blalock-Taussig. There was no mortality. Four patients with refractory CCF had pulmonary banding, 2 of which died (50%) within 72 hours of operation. The overall operative mortality was 28.6%.

The ten survivors had improved respiratory status. There were no post-operative neurological deficits. The preoperative irritability and difficulty in feeding were considerably reduced. None of the patients had phrenic nerve palsy post-operatively. There was delayed wound healing in all patients. Serial packed cell volume showed reduction in polycythaemia. PCV range was 59-67% (mean 65%) at last follow-up. The period of follow-up of the patients in the outpatient was from 5 months to 13 months (mean 9.3 ± 1.2 months) before they were all lost to follow-up.

Discussion

The number of patients with TGA that were admitted and referred for surgical intervention may well be the tip of the iceberg with respect to the incidence within the Nigerian population. Gupta and Antia [8] indicated that TGA is the commonest cardiac cause of neonatal death in Nigerian children and also the commonest cardiac cause of stillbirths in Nigeria. It therefore follows that neonatal blue babies should be referred early to capable centers for full evaluation, resuscitation and palliation prior to definitive surgery.

The basic principle of most palliation is to improve mixing between the pulmonary and systemic circulation by creating or enlarging an atrial septal defect. B–H atrial septectomy still remains the most effective, widely used means of achieving this, even after Rashkind BAS. [2] B-H atrial septetomy, though technically demanding, if done carefully, produces excellent results. Although B-T shunt offers the best palliation in our hands Blalock-Hanlon, [5] Cooley, [6] Spencer [7] and Hamilton [9] have corroborated the low mortality rate after B-H. Artrial septectomy. Though we had few patients who had this procedure, it still proved an acceptable palliation for TGA in our hands. Extensive excision of the atrial septum should be avoided to preserve the artery to the Sino-atrial (SA) node and internodal pathway. [10]

Infants with TGA and a large VSD (without left ventricular outflow tract stenosis) require treatment before they are 6 months old to prevent pulmonary vascular disease. They also need to be operated on earlier to relieve congestive cardiac failure. [11, 12] PB (or surgical constriction of the of the pulmonary artery) provide effective palliation in these patients. [11, 12] Now, early repair is preferred in most canters but we have used PB in this series in the absence of facility for early total repair. Where there facilities for arterial switch, there specific indications for PB in palliation for TGA: complex, multiple VSD; coexisting medical condition e.g., cerebrovascular accident [12] .PB results in acute reduction in volume load on the left ventricle with immediate geometric changes of the ventricle including an increase in wall thickness and decrease in cavity dimension. [13] The net effect of the PB apart from the reduction of PBF is to encourage intercirculatory mixing through the available shunts and improved oxygenation of the systemic circulation. [13]However when there is hypoplastic left ventricle or muscular VSD, PB and B-H septectomy should be combined. [11] In the current series the option was to do the least possible because of the clinical condition of the patients at presentation. B-H septectomy allows for more adequate PB. The operative mortality of PB in the newborn period historically should be in the range of 5% or less. [12] Mortality in our series is very high

because of the older age group we operated on. It is also of note that PB in infants with TGA is done more loosely than in normally related great arteries because the pathophysiology in the former requires higher pulmonary blood flow for intercirculatory mixture. [12]

In the literature, infants with TGA plus a VSD and left ventricular outflow tract obstruction (LVOTO) were palliated before they were old enough for repair. [7, 10] The patients do not present in heart failure and usually benefit from a shunt [11,12], classical or modified Blalock-Taussig are the easiest shunt to close at the time of definitive repair and are associated with fewest complication. [11,14] The few cases we did corroborate the specific place of this procedure in TGA with this morphology. Combination of Blalock-Hanlon and Blalock-Taussig has a definite place in TGA with severe LVOTO when associated with VSD. [2]

We believe there is still a place for well-chosen palliative procedures in this sub-region for the temporary management of TGA after morphological classification. Early diagnosis at birth and adequate resuscitative measures will improve the initial survival before palliation.

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