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Experience with Surgical Treatment of Congenital Defects of the Cardiovascular System in Nigeria

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Summary. The salient features of the anaesthetic and general pre- and postoperative care underlying the successful management of six children with Fallot's tetrad, thirteen with persistent ductus arteriosus and one with coarctation, during the period 1968–70 are presented. The postoperative survival in these twenty children is a marked improvement over that achieved during the 1964–66 period when seven children including four with Fallot's tetrad and three with persistent ductus arteriosus, were operated upon. It is concluded that the better result is due to general improvement in surgical, anaesthetic and nursing care as well as to better understanding of the concept of the micro-circulation, hypoxia and metabolic acidosis; and the prevention and prompt treatment of these derangements. The need for improved facilities in Africa to undertake total cure of patients with cyanotic heart defects has been stressed.

Résumé. Les traits saillants de l'anesthésie et des soins pré et postopératoires relatifs à un traitement favorable entre 1968 et 1970 de 6 cas infantiles de Tétrade de Fallot, de 13 autres avec ductus arteriosus persistant et 1 avec coarctation, sont présentés. La survie opératoire chez ces 20 enfants manifeste une amélioration significative par rapport à celle obtenue en 1964–66 avec 7 enfants opérés dont 4 présentant la tétrade de Fallot et 3 un ductus arteriosus persistant. On conclut que cette réussite est due à une amélioration des soins chirurgicaux, anesthésiques et généraux, ainsi qu'à une meilleure compréhension du concept de la micro-circulation, de l'hypoxie et de l'acidose métabolique; et enfin à la prévention et au traitement rapide de ces troubles. La nécessité de meilleures facilités pour entreprendre la guérison compléte des sujets atteints de défauts cardiaques cyanotiques est soulignée.

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TABLE 1. Age, sex, type of defect, clinical status and surgical procedure on twenty children with congenital heart defect

No.	Age at operation (years)	Sex	Type of defect	Presenting symptoms	Procedure	Complications
1	2⅓	F	Persistent ductus arteriosus	Recurrent respiratory infections	Division and suturing	Nil
2	9	M	Persistent ductus arteriosus	Recurrent respiratory infections	Division and suturing	Nil
3	2	F	Persistent ductus arteriosus	Recurrent respiratory infections and failure to thrive	Division and suturing	Pneumonitis
4	3½	M	Persistent ductus arteriosus	Recurrent respiratory infections and failure to thrive	Division and suturing	Nil
5	2¾	F	Persistent ductus arteriosus	Heart failure and failure to thrive	Division and suturing	Nil
6	2½	F	Persistent ductus arteriosus	Failure to thrive	Division and suturing	Nil
7	8¾	F	Persistent ductus arteriosus	Recurrent respiratory infection	Division and suturing	Nil
8	7	F	Persistent ductus arteriosus	Recurrent respiratory infection	Division and suturing	Nil
9	3½	F	Persistent ductus arteriosus	Recurrent respiratory infection. Failure to thrive	Division and suturing	Nil
10	1	F	Persistent ductus arteriosus	Recurrent respiratory infection. Failure to thrive	Division and suturing	Nil

11	4	F	Persistent ductus arteriosus	Recurrent respiratory infections	Division and suturing	Pneumonitis
12	15	M	Persistent ductus arteriosus	Decreased exercise tolerance	Division and suturing	Nil
13	1	M	Persistent ductus arteriosus	Recurrent respiratory infections and failure to thrive	Division and suturing	Nil
14	14	M	Coarctation of the aorta	Abdominal cramps, intermittent claudication and hypertension	Excision and end-to-end anastomosis	Nil
15	5	M	Fallot tetrad	Marked reduction in exercise tolerance, very high haematocrit	Blalock-Taussig shunt	Nil
16	3	M	Fallot tetrad	Marked reduction in exercise tolerance, very high haematocrit	Blalock-Taussig shunt	Nil
17	4	F	Fallot tetrad	Cyanotic spells	Cooley-Waterston shunt	Nil
18	9	M	Fallot tetrad	Cyanotic spells, extremely poor exercise tolerance	Cooley-Waterston shunt	Convulsions, loss of consciousness, surgical emphysema
19	7	M	Fallot tetrad	Poor exercise tolerance	Cooley-Waterston shunt	Nil
20	2	F	Fallot tetrad	Numerous cyanotic spells, failure to thrive	Cooley-Waterston shunt	Ventricular fibrillation successfully defibrillated during surgery

The incidence of congenital heart disease in Nigeria has been reported to be of the order of 3·5/1000 of all births (Gupta & Antia, 1967). From clinical and autopsy studies carried out at the University College Hospital (UCH) Ibadan, Caddell & Morton (1967), Antia & Williams (1971) have shown that various types of congenital heart disease occur as frequently in Africans as in non-Africans. The conclusion from these studies is that with the control of more common and preventable conditions such as malnutrition, infections, and parasitic diseases in many developing countries in Africa, congenital heart disease will engage the attention of the medical profession more than at present. This communication reports our early experience in the surgical management of congenital heart disease at the University College Hospital, Ibadan.

PATIENTS

Over the past 6 years (1964–70) twenty-seven children with congenital heart disease underwent surgery for correction of their malformations. Operation was carried out on seven of the twenty-seven children in the period 1964–66 and on twenty in the period 1968–70. Of the seven children who underwent surgery in the period 1964–66, there were four cases of Fallot's tetrad and three of persistent ductus arteriosus. All the four patients with Fallot's tetrad died shortly after various palliative surgical procedures carried out on them (Antia & Williams, 1971). The three patients with persistent ductus survived the surgical operation.

Twenty patients including six with Fallot's Tetrad, thirteen with persistent ductus arteriosus and one with coarctation of the aorta were operated on in the period 1968–70. All of these children survived. Details of the surgical management including anaesthesia and postoperative follow-up are described below. The age, sex, mode of presentation, diagnosis, surgical procedure and complications are summarized in Table 1. Diagnosis of the defect was made clinically in each case and confirmation by angiocardiography was only required in five instances.

ANAESTHETIC MANAGEMENT

Details of the anaesthetic management of these twenty patients have been described elsewhere (Oduntan, 1971) and only a brief account is given here. The children were admitted 5–7 days before operation. During this period, pre-operative management included skin preparation in the form of cetrimide scrubs to the trunk twice daily for the 3 days preceding operation and the administration of prophylactic antibiotics (penicillin 20,000–40,000 units/lb/day, and streptomycin 30 mg/lb/day) 24 hr before surgery. Haemoglobin genotype was determined by electrophoretic method in every patient and arterial blood gases ($Paco_2$, Pao_2 and pH) were estimated in those with cyanotic heart disease. This period also provided an opportunity for the patients to get used to the environment and the staff, an essential pre-requisite for co-operation in the critical postoperative period.

Premedication for patients with cyanotic defects consisted of half the usually recommended dose of atropine (0·005 mg/lb) and pethidine (1 mg/kg).

Induction of anaesthesia was with slow intravenous thiopentone sodium (4–5 mg/kg body weight) in a majority of the children. For the over-anxious child who would not accept thiopentone injection, anaesthesia was induced with halothane in oxygen. After induction, the patient was given suxamethonium chloride intravenously (1–1·5 mg/kg) and artificial ventilation performed with oxygen. Subsequently, laryngoscopy and endotracheal

intubation were carried out and the respiration controlled manually by compressing the reservoir bag, delivering 0·5% halothane in oxygen until there was recovery from the depolarizing block.

The anaesthetic circuit used in every case was the Ayre's T-piece with a high flow of oxygen. All the patients were paralyzed with D-tubocurarine (0·5 mg/kg body weight) and the respiration was controlled manually. Light anaesthesia was maintained with either 0·5–0·75% halothane in oxygen or 50% nitrous oxide in oxygen, and intravenous phenoperidine which is a potent analgesic drug.

A Cotel-Keating pulsemeter, with the sensing device attached to the thumb, was used to give a visual indication of the pulse rate. The central venous pressure was constantly monitored via a catheter inserted through an external jugular vein during the operation and in the immediate postoperative period especially in patients with Fallot's tetrad. Care was taken to avoid hypoxia, hypercarbia, or sudden hypotension as these situations could lead to serious arrhythmias.

In the patients with Fallot's tetralogy, sodium bicarbonate (2 mEq/kg body weight) was given soon after induction of anaesthesia, and repeated as indicated by the results of blood gas studies undertaken during the operation. This group of patients also received low molecular weight dextran (Rheomacrodex) during the operation in an effort to enhance tissue perfusion.

Trimetaphan (arfonad), a short-acting hypotensive agent administered in a continuous intravenous infusion in a concentration of 0·5 mg/ml in 5% glucose was used to produce hypotension during surgery in the one patient with coarctation of the aorta. A careful check on the blood loss was maintained throughout, and in any patient from whom blood loss was considered to be excessive, transfusion of warm blood was carried out. Sodium bicarbonate and calcium gluconate were given with blood transfusion to maintain homeostasis of hydrogen and calcium ions.

SURGICAL MANAGEMENT AND POSTOPERATIVE CARE

The clinical presentation, surgical procedures and postoperative complications are summarized in Table 1.

(a) *Persistent ductus arteriosus*

Thoracotomy was performed through the fourth intercostal space in the left posterolateral positions. In every patient a consistent but transient elevation in blood pressure and a compensatory decrease in pulse rate were observed beginning at the moment the ductus was cross-clamped.

Postoperatively, effort was made to achieve early mobilization. The chest drainage tube was removed 24 hr after operation. Antibiotics, started before operation were continued for 8 days. No patients required postoperative digitalis, even though a few of them had been on digitalis up to 2 weeks prior to admission.

(b) *Coarctation of the aorta*

This 15-year-old boy (Case 14, Table 1) presented with colicky abdominal pains and distentions, cold lower extremities and intermittent claudication for 2 years. Physical examination revealed considerably reduced pulse volume in the lower extremities. The

blood pressure measured 170/80 mmHg in both arms and 110/80 in the legs. The lower extremities were cold compared to the upper ones. An aortogram confirmed the diagnosis of post-ductal type of coarctation (Fig. 1).

Thoracotomy was performed through the fourth intercostal space in the left postero-lateral position under hypotensive anaesthesia. An aortic segment, 2 cm long, just below the left subclavian artery take-off was resected. During the procedure one pair of intercostal arteries was sacrificed and the ligamentum arteriosum which was connected to the upper portion of the resected segment was divided. An end-to-end anastomosis measuring 1·5 cm in diameter was carried out with minimal difficulty. The resected segment contained a

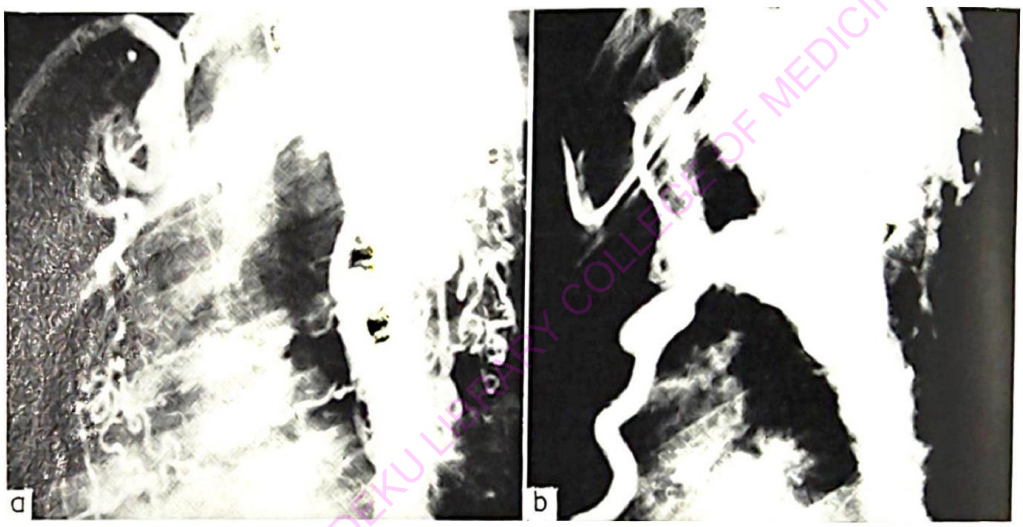

FIG. 1. (a) Aortogram showing numerous anastomoses and the segment of the coarctation. (b) Aortogram showing dilated internal mammary artery and the upper segment of the coarctation.

diaphragm with a pinhole opening across it. The postoperative course was uneventful, with no abdominal pain. At a recent follow-up the blood pressure in the arms and legs measured 140/80 and 150/80 respectively. All the symptoms had disappeared.

(c) *Fallot's tetrad*

Transpericardial side-to-side aortopulmonary anastomosis (Cooley-Waterston shunt) was performed in four patients. The anastomosis, measuring approximately 4 mm in diameter, was created between the posterior aspect of the ascending aorta and the right pulmonary artery. The Blalock-Taussing procedure, in which the right subclavian artery is anastomosed to the right pulmonary artery, was performed on the remaining two patients with Fallot's tetrad.

Table 2 summarizes the results of the arterial blood gas studies carried out on one (Table 1, Case 18) of these four patients. Similar results were obtained in the other three patients. The pre-operative arterial oxygen tension (Pao_2 = 26 mmHg) was considerably reduced in all the patients. Because of the high concentration of oxygen in the inspired mixture used

during the operation there was marked rise in the arterial oxygen tension even before the shunt procedure.

The postoperative course in the two patients who had Blalock-Taussing operation and in three of those who underwent the Cooley-Waterston procedure was uneventful. All the six patients were digitalized after the operation. The changes in the haematocrit and Pao_2 in all the six patients are illustrated in Table 3. There was a significant fall in the haematocrit and an equally significant rise in the Pao_2 in all the patients. A continuous shunt murmur appeared in all the patients between the second and third postoperative day.

TABLE 2. Summary of blood gases—Case 18

Blood gases	Pre-operative values	During operation	
		Pre-anastomose	Post-anastomose
Pao_2	26	77·5	68·0
$Paco_2$	39·5	48·5	47·0
pH	7·325	7·260	7·36

TABLE 3. Changes in PCV and PaO_2 following shunt

Case no.	PCV		Pao_2 (mmHg)	
	Pre-operative	Postoperative	Pre-operative	Postoperative
15	74	52	26·0	34·0
16	72	48	27·0	41·0
17	70	47	21·0	40·0
18	80	58	26·0	48·0
19	85	69	30·0	50·0
20	66	47	28·0	47·0

As shown in Table 1 serious complications occurred in two patients (Cases 18 and 20). Case 18 underwent a Cooley-Waterston shunt. On the first postoperative day the patient developed a temperature of 104°F. He also had a residual right pneumothorax. Despite administration of antibiotics, chloroquine and Dispirin the pyrexia persisted. On the second postoperative day the patient developed generalized convulsions which were difficult to control with repeated administration of paraldehyde. On the same day the residual pneumothorax previously noted developed into extensive emphysema involving the neck and face.

The management consisted of administration of paraldehyde, a total of 40 ml of 50% glucose in two divided doses, intramuscular furosemide and full digitalization. Oxygen was administered continuously. On the third postoperative day the convulsions ceased and thereafter the patient's condition gradually improved. He was fit to be discharged on the twenty-fifth postoperative day. He has remained remarkably well since discharge.

Ventricular fibrillation occurred during surgery in Case 20 and defibrillation was successfully carried out.

COMMENTS

Persistent ductus arteriosus

Shapiro & Keys (1943) have shown that without surgical obliteration, a persistence of ductus arteriosus is associated with premature death. Other complications of this condition include congestive heart failure, subacute bacterial endocarditis, recurrent respiratory infections and rarely right ventricular failure secondary to pulmonary hypertension. Therefore, elective surgical obliteration of the ductus should be performed in every case. Most surgeons would not operate on asymptomatic persistent ductus in infants because of a high operative mortality (Bernhard & Norman, 1967). Even in the presence of complications, it is often wise to delay surgical treatment and to employ appropriate medical management in these infants. It must, however, be emphasized that in certain cases a carefully performed operation is the only means of salvaging these very ill infants (Nadas, 1963). For example, an operation is strongly indicated when a ventricular septal defect co-exists with a persistent ductus arteriosus and consequently there is further increase in pulmonary blood flow and volume load on the right ventricle. Banding of the main pulmonary artery may also be required in such cases (Bernhard & Norman, 1967). Complete division and suture of the ductus as advocated by Gross (1947) is technically easy; this procedure fully guarantees against recanalization, a problem which sometimes occurs when ligation is the procedure carried out (Keith, Rowe & Vlad, 1967). In terms of technical feasibility Mustard (1955), has shown that division and suturing is preferred to ligation where the duct is short and wide. In the present series all the ducts were divided and sutured since most of them were of the short and wide type.

Postoperative complications are uncommon with proper timing of surgery and good anaesthetic management. However, surgical complications such as recurrent laryngeal nerve injury, haemorrhage and pneumonitis still occur. In patients with substantial left-to-right shunt through the ductus, systemic hypertension sometimes occurs postoperatively and congestive heart failure may follow surgery. This did not occur in the present series. There were, however, two patients who developed postoperative pneumonitis.

Coarctation of the aorta

The indication for surgical treatment in the single patient with coarctation of the aorta was development of systolic hypertension and ischaemic symptoms in the legs and the abdomen. Aortic resection and anastomosis without graft replacement was all that was required to restore aortic continuity.

Fallot's tetrad

Since the first report of successful palliative operation in this malformation by Blalock & Taussig (1945), other palliative procedures have been developed. The basic technique in these various palliative procedures is anastomosis of a systemic vessel to pulmonary artery. Numerous reviews have been reported on the results of these procedures (Campbell, 1958; Shumacher & Mandelbaum, 1960; Taussig *et al.*, 1962; Kaplan *et al.*, 1968; Kirklin & Karp, 1970). In most centres total intracardiac correction of the malformations is being carried out, and the lower age limit for such correction is being gradually reduced, thus restricting the need for palliative shunts to cases of symptomatic infants and children under 4 years old. In these the mechanical problems of cannulation and the non-availability of oxygenators designed for small blood volumes are current limiting factors. In most develop-

ing countries there are at present no facilities for total correction, therefore palliative procedures continue to be the treatment of necessity.

The choice of the palliative procedure depends on such factors as the safety of the procedure, the size of the subclavian artery to be used in the anastomosis and the ease with which the anastomosis itself can be closed during subsequent total repair of the malformations. Kirklin & Karp (1970) have stated that the Blalock-Taussig procedure is the safest. However, in this type of procedure adequate dissection of the subclavian artery is necessary and the anastomosis must also be performed on the opposite side to the direction of the aortic arch in order to prevent tension or kinking of the subclavian artery. Furthermore, the size of the anastomosis is limited by the size of the subclavian artery. Therefore, the procedure is not very useful in patients below the age of 2 years (Woodson *et al.*, 1969). In the present series the procedure was carried out in only two patients.

The other procedure carried out on the remaining four of our patients was the aorto-pulmonary artery anastomosis (Cooley-Waterston shunt). This procedure has certain advantages over the subclavian-pulmonary artery anastomosis of Blalock-Taussig. First, a satisfactory anastomosis can be performed in very young patients. Secondly, the anastomosis is created on the right side irrespective of the direction of the aortic arch. A serious disadvantage of this procedure is the possibility of creating too large a shunt which would permit excessive pulmonary blood flow from the systemic circulation. In fact, this occurred in one of our patients. Hallman *et al.* (1967) have reported the occurrence of this complication in 26% of their patients.

Apart from general improvement in surgical, anaesthetic and nursing care over the last decade, there are other factors which contribute significantly to the improved surgical survival of severely cyanotic children. These factors include better understanding of the concept of micro-circulation, hypoxia and metabolic acidosis in these cyanotic patients, and the prevention or prompt treatment of these derangements (Bernhard & Norman, 1967). Achievement of satisfactory haemodilution by the use of plasma or low molecular weight dextran to replace blood loss minimizes sludging of the blood and improves tissue perfusion. The administration of anaesthetic gases in such concentration as to ensure the highest possible concentration of inspired oxygen eliminates or minimizes hypoxia. Hyperbaric oxygenation has been used for this purpose by Bernhard *et al.* (1964). By monitoring blood gases (pH, Pao_2 and $Paco_2$) as well as blood pyruvate/lactic acid ratio before and during operation, the acid–base equilibrium can be closely followed. Data obtained from these measurements are extremely useful indicators of any derangement in the acid–base equilibrium. Any such derangement would be promptly treated with sodium bicarbonate or THAM (2-amino, 2-hydroxy methyl 1, 3, propanediol). Postoperatively, it is equally important to avoid respiratory impairment and/or cardiac failure either or both of which tend to aggravate the effect of hypoxia and acid–base imbalance.

In our experience, the above factors which contribute to the surgical survival of patients with cyanotic heart defect were not fully appreciated initially and hence our high mortality in the period 1964–66. Operations on these patients were carried out by different surgeons with no special interest in cardiovascular surgery. Since 1968, however, all the operations have been carried out by one of us (MAB) and the anaesthesia given by one person (SAO). There has also been better medical and nursing management of these cases in the post-operative period, thus contributing to the survival of all the cases operated on during the period, 1968–70.

At best, all paliative operations for Fallot's tetrad can only give 3–10 years of satisfactory function, after which a repeat shunt or total correction must be undertaken. It is our hope that in the near future, facilities will be available for complete repair of congenital heart defects encountered in this part of the world.

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