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Original Article

Management of complex CHD at the National Cardiothoracic Center of Excellence, University of Nigeria Teaching Hospital, Enugu: the role of foreign cardiac missions in 3.5 years

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Abstract *Background:* CHD is defined as structural defect(s) in the heart and proximal blood vessels present at birth. The National Cardiothoracic Center of Excellence, University of Nigeria Teaching Hospital (UNTH), Enugu, through the aid of visiting Cardiac Missions has managed a significant number of patients within the last 3.5 years. *Aim/Objective:* The objective of this study was to review surgical options and outcome of complex CHD among patients attending UNTH, Enugu, Enugu. *Materials and Method:* During the period of 3.5 years (March, 2013 to June, 2016), a total of 20 cases of complex CHD were managed by cardiac missions that visited UNTH, Enugu. Their case notes and operating register were retrieved, reviewed, and analysed using SPSS version 19 (Chicago). *Results:* There were eight females and 12 males, with a ratio of 2:5. The age range was from 5 months to 34 years with a mean of 1.7. Among all, five patients died giving a mortality rate of 25%. The operative procedures ranged from palliative shunts to complete repair. The outcome was relatively good. *Conclusion:* Complex CHD are present in our environment. Their surgical management in our centre is being made possible by periodic visits of foreign cardiac missions.

Keywords: Complex; National Cardiothoracic Center; foreign cardiac mission

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CHD ARE THE MOST COMMON TYPE OF DEFECTS IN THE United States of America with an incidence of 1% of live births.[1] Complex heart disease consists of certain cardiac malformations that involve parts that are necessary for maintenance of the patient's life, and they are classified as follows: total anomalous pulmonary venous drainage, hypoplastic left heart syndrome, single ventricle, mitral atresia, pulmonary atresia with intact ventricular septum, tricuspid atresia, double right ventricular outflow tract, double left ventricular outflow tract, tetralogy of Fallot, tetralogy of Fallot with pulmonary atresia, tetralogy of Fallot with absence of pulmonary valve syndrome truncus arteriosus, and transposition of the great vessels.[2] The incidence of complex forms – that is, moderate and severe forms of CHD – is about 6/1,000 live births.[3] Others include coarctation of the aorta with ventricular septal defect, interrupted aortic arch, hypoplastic left heart syndrome, Ebstein's anomaly of the tricuspid valve, and severe aortic valve stenosis.

Patients with such defects are at variable age groups ranging from the neonatal period to adulthood, and in these groups they remain at risk of premature deaths. Complex CHD affect about 0.6% of newborns with stable incidence over time.[4,5] Unexpectedly, some affected patients now survive to adulthood. Complex CHD and unrepaired cyanotic lesions complicated with Eissenmenger's syndrome

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remain at risk for long-term complications along with risk of death as young adults.[6–9]

Carrying out corrective congenital heart surgery in resource-limited countries such as Nigeria and other sub-Saharan countries is a major developmental challenge;[10] however, the National Cardiothoracic Center of Excellence at University of Nigeria Teaching Hospital, Enugu, Nigeria, has remained a referral centre for cardiovascular diseases since 1984, but is ill-equipped both in skills and facilities to undertake repair for complex CHD. Such patients suffer and die unattended to, as they cannot afford the cost of corrective surgery abroad. Recently, Nigerians in Diaspora have used cardiac missions to visit home and operate on patients with the disease.[10]

These missions appear to be saving many children with moderate-to-complex CHD, as the surgeries are conducted locally by visiting teams alongside skills transfer to build local capacity.[11,12]

The present article thus aimed at reviewing surgical options and outcome of complex CHD among patients attending University of Nigeria Teaching Hospital, Enugu, Enugu.

Materials and method

We carried out a retrospective study at the National Cardiothoracic Center of Excellence, University of Nigeria Teaching Hospital, Ituku-Ozalla Enugu, Enugu, Nigeria. For about 3.5 years (February, 2013 to June, 2016), the National Cardiothoracic Center of Excellence, University of Nigeria Teaching Hospital, Enugu, Enugu partnered with cardiac missions from the United States of America (VOOM), the United Kingdom (Save-a-Heart Nigeria), and India (Innova Children Heart Hospital, Hyderabad), referred to as international collaborators for the management of complex congenital heart among other heart diseases. During the period, a total of 20 complex CHD were managed involving children and adults. The hospital provides care for children and also receives referrals from different parts of Enugu, the rest of Enugu State, and surrounding states. Enugu State of Nigeria has a population of about 3.3 million people according to the national census of 2006; the surrounding states of Abia, Anambra, Benue, Ebonyi, Delta, Imo, and Kogi have populations ranging from 2.2 to 4.2 million people.

This was a cross-sectional, retrospective study in which a review of the records of all children and adults who underwent surgery for complex CHD was carried out. Factors such as demographic features, the types, the duration of both bypass time and aortic cross-clamp time, the duration of stay in the ICU and the hospital, as well as the immediate outcome, that is, 30 days postoperative morbidity and mortality, were determined.

Data were analysed using SPSS 20 (Chicago). Rates and proportions were calculated with 95% confidence intervals. The proportions were compared using Student's t-test. The level of significance was set at $p < 0.05$ (Table 1).

Results

A total of 20 patients underwent repair for complex CHD during the envisaged period. The male-to-female ratio was 5:2. The age range was 5 months to 34 years with a mean of 1.7 years. Repairs were divided into palliative (4, 33.3%) and definitive (16, 72.7%) procedures. Of the 20 patients, five (25%) died either intraoperatively or within a few days in the ICU. The 30 days postoperative morbidity/mortality for Tetralogy of Fallot with pulmonary atresia and major aortopulmonary collaterals, Tetralogy of Fallot with hypoplastic pulmonary artery, truncus arteriosus type 3, and common atrioventricular canal defect as well as one isolated TOF were eventful, as they all died. During the 3.5 years period of the cardiac missions, the centre managed 20 cases of

Table 1. Types of complex CHD.

Sl. No.	Types of complex CHD	Number	Percentage
1	Tetralogy of Fallot	9	45
2	Double-outlet right ventricle	2	10
3	Common atrioventricular canal defect with pulmonary hypertension	2	10
4	Truncus arteriosus	2	10
5	Ruptured sinus of Valsalva aneurysm	2	10
6	Pulmonary atresia with MAPCAS	1	5
7	Absent posterior and anterior leaflets of the tricuspid valve, with severe TVR	1	5
8	Mitral atresia with regressed left ventricle and pulmonary hypertension	1	5
Total		20	100

MAPCAS = multiple aortopulmonary collateral arteries; TVR = tricuspid valve regurgitation

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Table 2. Operative procedures for complex CHD.

Sl. No.	Indications	Sex	Age	Procedures
1	C-AVCD with PAH	M	4 years	Pulmonary artery banding
2	Truncus arteriosus type 1	M	18 months	Total correction double banding
3	DORV, hypoplastic LV, severe subvalvular PS	M	2 years	Bidirectional Glenn without cardiopulmonary bypass
4	Aortic valve disease with ruptured sinus of Valsalva aneurysms	F	34 years	Aortic valve repair + repair of sinus of Valsalva aneurysm
5	DORV, TOF-like	M	3 years	Modified Blalock Taussig shunt
6	Absent posterior and anterior leaflet of TV	M	10 months	TV repair + annular stabilisation
7	Truncus arteriosus, type 3	M	5 months	Total correction
8	VSD + ruptured sinus of Valsalva aneurysm	F	34 years	Intracardiac repair of VSD and RSOVA
9	Mitral atresia + regressed LV + severe PAH	F	5 months	Surgical septostomy + pulmonary artery banding
10	TOF with Pulmonary atresia + MAPCAS	F	3 years	Unifocalisation + aorto-neoPA shunt
11	C-AVCD	F	19 months	Intracardiac repair
12	TOF + hypoplastic PA	M	8 months	Augmentation of MPA + VSD closure
13	TOF	M	8 years	Total correction
14	TOF	M	1.4 years	Total correction
15	TOF, small PAs	M	12 years	Total correction
16	TOF	F	10 years	Total correction
17	TOF	M	15 years	Total correction
18	TOF	M	6 years	Total correction
19	TOF + small PDA	F	14 years	PDA ligation + total correction
20	TOF	F	1.3 years	Total correction

DORV = double outlet right ventricle; MAPCAS = multiple aortopulmonary collateral arteries; PAH = pulmonary arterial hypertension; TOF = Tetralogy of fallot; VSD = ventricular septal defect

Table 3. Average number of days stayed in the ICU and hospital by patients with complex CHD treated by surgery.

Sl. No.	Complex CHD operated on	ICU stay (days)	Hospital (days)
1	TOFs	6	5
2	Pulmonary atresia	10	–
3	DORV, TOF variety	2	4
4	VSD + ruptured sinus of Valsalva aneurysm	3	3
5	DORV + hypoplastic LV + severe subvalvular PS	3	4
6	C-AVCD	3	5
7	TOF + hypoplastic PAs	4	–
8	Absent posterior and anterior leaflets of tricuspid valve	5	8
9	Mitral atresia + regressed LV + severe PAH	4	5
10	Aortic valve disease with ruptured sinus of Valsalva aneurysm	3	4

DORV = double outlet right ventricle; PAH = pulmonary arterial hypertension; TOF = Tetralogy of fallot; VSD = ventricular septal defect

complex CHD as seen in Tables 1 and 2. The patient with Tetralogy of Fallot with hypoplastic pulmonary artery died on table, whereas the others died within a few days in the ICU. The truncus arteriosus type 1 patient who survived had an unstable sternum, which was corrected surgically. This prolonged the total length of hospital stay (see Table 3). The Tetralogy of fallot with pulmonary atresia had a ventricular septal defect closure and a right ventricular to pulmonary artery conduit (see Figures 1 and 2).

An interesting case was a 10-month-old baby with absent posterior and anterior leaflets of the tricuspid valve with severe tricuspid valve regurgitation. A new valve was created, and a stabilisation ring was

Figure 1.
Intra cardiac repair for Tetralogy of fallot.

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left in situ, see Figure 3. The patient did well postoperatively in the ICU, see Figures 3 and 4.

Discussion

A few other medical disciplines have required for their development the degree of daring courage, tenacity, and drive that characterised the efforts of early pioneers in the field of congenital cardiac surgery.

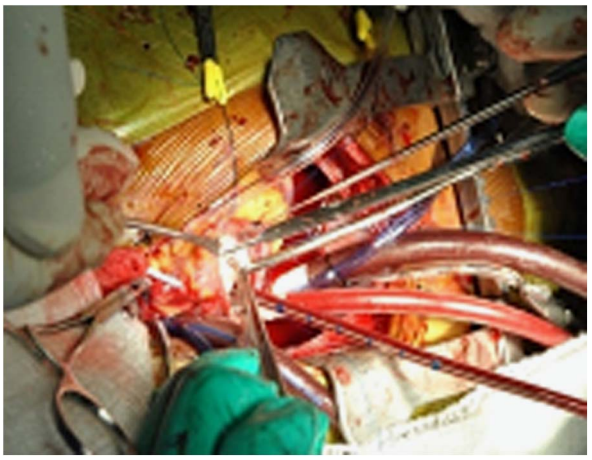

Figure 2.
Pericardium being harvested for creation of conduit.

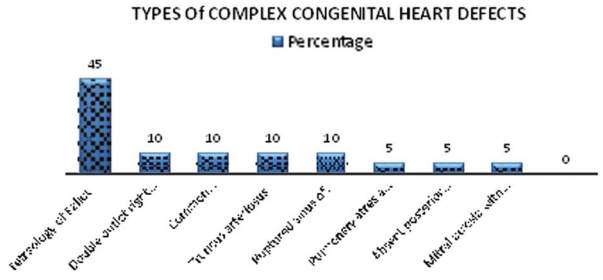

Figure 3.
Types of complex congenital heart defects.

Only a century ago, Theodore Billroth publicly condemned the dream of cardiac surgical interventions by stating that "Any surgeon who wishes to preserve the respect of his colleagues would never attempt to operate on the heart".[13] Over the last six decades, the specialty of paediatric cardiac surgery has evolved from a heroic effort with occasional success into a consolidated, sophisticated specialty with excellent outcome.[14]

As the defects are complex, many challenges are associated with the management, preoperatively, intraoperatively, and postoperatively.[15] Our centre has successfully managed simple defects such as patent ductus arteriosus, atrioseptal defect, and ventricluoseptal defects. Repair of complex CHD before the recent engagement of our international collaborators had not been carried out. During the envisaged period, 20 cases were managed. Notably, a truncus arteriosus type 1 was successfully repaired. Other complex CHD that had successful outcome were aortic valve disease with ruptured sinus of Valsalva aneurysm and congenital absence of both the posterior and the anterior leaflets of the tricuspid valve. An isolated TOF case performed by a visiting adult cardiac surgeon developed low cardiac output syndrome, acute renal failure, and septic shock. His total ICU stay was up to 14 days before he died. The cause of death could be due to inadequate relief of right ventricle outflow tract obstruction.

In our study, the mortality rate was 25% (five cases). Indeed, this is relatively high. In spite of working in a developing country, with technological challenges, the mortality rate in our study is comparable with the study by Bohumuil et al[16] in Croatia and Cavalcanti et al[17] in Brazil. Their studies recorded mortality rates ranging from 3.3 to 10.3% in all cadres of congenital heart surgery – simple to complex. Information on mortality rate for isolated complex CHD is lacking, due to inaccurate data from various centres.

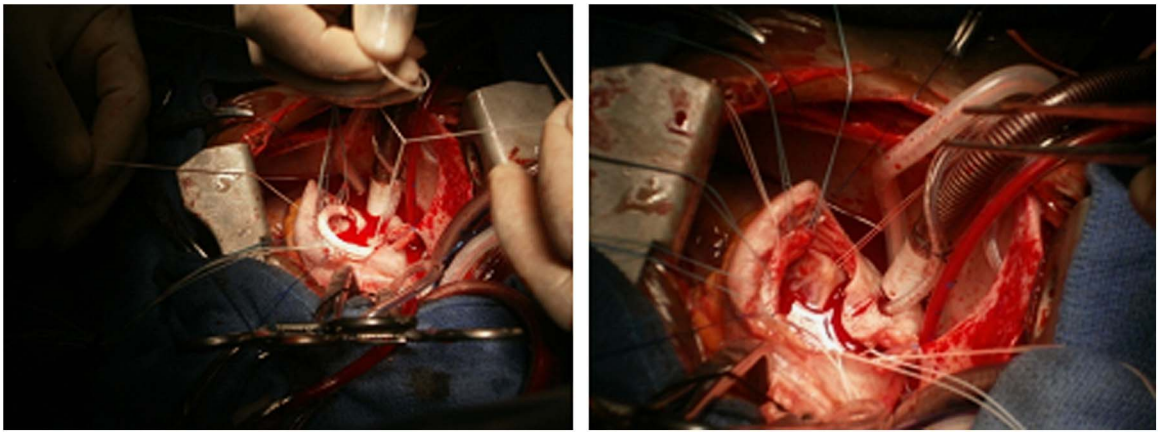

Figure 4.
Creation of neo-leaflets of tricuspid valve and annular stabilization ring in Epstein Anomaly.

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The operative time in complex CHD is usually prolonged relative to the simple or moderate counterparts because many of the repairs are performed in moderate-to-profound hypothermia. Invariably, there is prolongation of both the cardiopulmonary bypass time and aortic cross clamp time.[18,19] These findings agree with our study, which took average of 180 minutes for the Cardio Pulmonary Bypass and 120 minutes for the ACT.

The duration of ICU admission and hospital admission in days, starting from day 1 postoperatively, was taken into consideration in our study. We found that the more complex the intra-cardiac repair was, the more turbulent the postoperative management in ICU. This prolonged the ICU admission and consequently the total hospital stay. This finding is comparable with the study by Sanchez Andre's et al[20], who showed that patients undergoing repairs for complex CHD often develop storm postoperatively, and optimal management requires a thorough understanding of basic principles of tissue oxygenation, cardiovascular physiology, and anatomy, as well as the pathophysiology of complex congenital defects.

Surgical repair techniques for complex CHD were introduced several decades ago and subsequently have undergone continuous modifications. Surgical palliations such as modified Blalock–Taussig Shunt for double-outlet right ventricle and bidirectional Glenn shunt or hemiFontan procedures for tricuspid atresia and single ventricle pathology are performed for complex CHD. Other palliative procedures such as first-stage Norwood is performed for hypoplastic left heart syndrome. Of note are definitive procedures for unfortunate children with these rare but challenging defects. More recently the arterial switch operation for d-transposition of great artery, popularised by Jatene, replaced the cumbersome Senning and Mustard techniques. Completion Fontan procedure, total cavo-pulmonary connections, including its modifications such as lateral tunnel techniques and total extracardiac cavopulmonary connection, is a definitive procedure for univentricular physiology.[21]

Conclusion

Foreign cardiac missions are beneficial to developing countries in that they help curb the high morbidity and mortality associated with these complex cardiac disease as well as equipping the surgeons to take over these daring challenges in future.

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Conflicts of Interest

None.

Ethical Standards

All the patients had obtained consent and all the ethical standards were observed.

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