In-hospital Outcome of Children referred for Cardiac Surgery Abroad from a Developing Country

EN Ekure, CN Okoromah

Abstract

Ekure EN, Okoromah CN. In-hospital Outcome of Children referred for Cardiac Surgery Abroad from a Developing Country. Nigerian Journal of Paediatrics 2009; 36: 80

Background: Cardiac surgery is the definitive intervention for structural heart diseases particularly of congenital aetiology in children. Unfortunately, most developing countries currently lack adequate capacity for paediatric cardiac surgery often necessitating referral to foreign countries with appropriate facilities and capacities. We hereby report the in-hospital outcome of Nigerian children referred from Lagos University Teaching Hospital to undergo cardiac surgery abroad.

Methods: Nigerian children referred from the Lagos University Teaching Hospital, Lagos State, to centres abroad for cardiac surgery were reviewed. Data obtained from records for analysis were sex, pre-surgical diagnosis, age at surgery, year surgery was performed, surgical centre location, post-operative echocardiographyreport, length of hospital stay and in-hospital case fatality.

Results: In the five year period 68 children underwent heart surgery. Age at surgery ranged from 3 months to 14 years (mean = 41.71±43.88 months) while their average duration of hospital staywas 25 days. The surgeries performed were ligation of patent ductus arteriosus (PDA), repairs of atrial and ventricular septal defects, tetralogy of Fallot, atrioventricular canal defects, arterial switch operations, mitral valve repair and replacement. Three (4.4percent) in-hospital deaths occurred (all in children aged <2 years). Major post operative complications related to death were respiratory problems and low cardiac output state.

Conclusion: The in-hospital case fatality rate at 4.4 percent is low for Nigerian children undergoing cardiac surgery abroad with low cardiac output state common to all the deaths.

Keywords: In-hospital case fatality, Heart disease, Cardiac surgery, children.

Introduction

THE outcome following heart surgery in children with heart diseases has improved greatly with technological advancements in diagnostic and intervention procedures, especially in developed countries. Cardiac surgeryremains the major remedy for heart diseases in children especially those with congenital heart diseases. Any kind of surgery has its risks, but cardiac surgery has even additional peculiar risks. Reports have indicated that certain

factors such as age, sex, hospital case volume, other associated malformations, type of initial operative procedure, and use of cardiopulmonary bypass can affect the outcome of cardiac surgery.¹⁻³ The type of surgical procedure performed has also been identified as the most important risk factor in children.⁴

The advanced technology required for paediatric cardiac surgery is lacking in many developing countries, especially in Africa. This leaves millions of children with untreated correctable congenital heart diseases. ⁵ In Nigeria, though the first open heart surgery took place more than three decades ago, many challenges hinder the growth of that health sector, especially in children. ⁶⁻⁷ A few medical missions have come to the country but operate mostly on adults, with only a limited number of children benefitting. Consequently, minimal capacity

College of Medicine, University of Lagos, Nigeria

Paediatric Cardiology Unit Department of Paediatrics

Corresponding author: Ekure, EN. E-mail: ekaekure@yahoo.com exists in Nigeria at the time of this report for children with heart diseases that require cardiopulmonary

The reported case fatality rates following cardiac surgery in children vary from 0.3 to 29.8 percent depending on type of surgery. To improve the chances of survival in children, referral abroad to health institutions with facilities for cardiac surgery has been an option for the majority of cases in recent times. The patients who have been able to avail themselves of this opportunity provide an opportunity to study the outcome following cardiac surgery in Nigerian children.

Subjects and Methods

The studywas done in the Paediatric Cardiology unit of Lagos University Teaching Hospital (LUTH). This is a tertiary referral centre with facilities for evaluation of cardiac patients such as plain chest radiography, electrocardiography (ECG) and echocardiography. Cardiac catheterization is however not offered and cardiac surgery beyond drainage of pericardial effusion is not available for children. Intensive care support is available but with limitations in the capacity for paediatric support.

The study subjects were enrolled from the Paediatrics unit (out-patients and in-patients) and were from a data base of children aged below 18 years of age diagnosed with heart diseases over a 5-year period from January 2004 to December 2009. The diagnosis of the heart diseases was based on clinical evaluation supported by chest radiography, ECG and echocardiography. Heart disease was defined as a structural abnormality of the heart such as defects in cardiac septation, abnormalities of ventriculoarterial connections, rudimentary or absent chambers, abnormalities of ventricular inflow and outflow, and abnormal vascular connections and structures. Others were cardiovascular inflammatory disease, myocardial disease or cardiac tumours. Multiple cardiac lesions were allocated to the dominant lesion associated with haemodynamic changes.

The patients included in this studywere those on the database who underwent surgery. Data obtained were sex, pre-surgical diagnosis, age at surgery, surgeryyear, surgical centre location, post-operative echocardiographyreport, length of hospital stay and patient mortality outcome.

Procedures performed on the children were used to group the cases with congenital heart disease using the Risk Adjustment for Congenital Heart Surgery version 1 (RACHS-1) method ⁷. RACHS-1 is a valid clinical research tool, which is widely applicable to the evaluation of differences in outcomes in both

existing and future data sets among groups of patients with congenital heart disease. It specifically adjusts for differences in in-hospital mortality rates among groups of children undergoing surgery for congenital heart disease. The risk categories range from 1 to 6 moving from simple procedures such as repair of atrial septal defect(ASD) or PDA in stage 1 to repair of hypoplastic left heart syndrome in stage 6. ⁷

The In-hospital outcome was assessed by fatality. The fatality rate was defined as death within the period of admission in hospital following cardiac surgery.

Results

Clinical profile of Nigerian children undergoing cardiac surgery abroad

In the five year period 1,820 new cardiac cases (364/year) were seen at LUTH and 1259 (69 percent) required cardiac surgery. Of this number, 68 (5.4 percent underwent cardiac surgery. In all these instances, the cardiac surgeries were performed at referral centres abroad (63 in India and one each in Israel, Germany, Italy, UK and South Africa). Fourteen health facilities were involved.

The cases comprised of 30 (44.1 percent) females and 38 (55.9 percent) males. All 68 children had congenital heart diseases except two who had rheumatic heart disease with severe mitral regurgitation. Fifteen patients (22.1 percent) had Downsyndrome. The heart diseases were diagnosed at various ages ranging from birth to 6 years. The peak in the number of patients who had surgerywas in 2007 with 21 cases. (Figure 1). The cardiac diagnoses of the 68 children who had surgery are shown in Table 1.

The age at which surgery was carried out ranged from three months to 14 years with 12 (17.6 percent) having surgery in infancy. Most (43=63.2 percent) of the children were aged two years and below. Whereas 47.1 percent of the children were aged between one and three years. 32 percent were older than three years.

Surgical procedures and outcomes including the in-hospital case fatality rate

Sixty three (92.6 percent) of the 68 surgeries occurred in India. The types of surgery performed on the 68 patients are shown in table 2 according to the RACHS-1 risk categories. Most (57.4 percent) patients had surgeries that belonged to risk category 2. Two patients with tetralogy of Fallot with pulmonary atresia underwent surgery twice. The duration of hospital stay for those who had surgery ranged from 3 days to 94 days with an average of 25

Table I

Cardiac diagnoses of the 68 patients who underwent cardiac surgery

Cardiac Diagnosis	Number of Cases(%) 19 (27.9)	
Ventricular Septal Defect		
Tetralogy of Fallot	19 (27.9)	
Atrioventricular canal defect	8 (11.8)	
Patent ductus arteriosus	9 (13.2)	
Pulmonary valve stenosis1	(1.5)	
Double outlet right ventricle	2 (2.9)	
Atrial septal defect with right pulmonary artery stenosis.	1 (1.5)	
Tricuspid Atresia	1 (1.5)	
Truncus Arteriosus	1(1.5)	
Transposition of great arteries	1(1.5)	
Partial pulmonary venous return	1 (1.5)	
Single ventricle	3 (4.4)	
Rheumatic heart disease with severe mitral regurgitation	2 (2.9)	
Total	68 (100)	

days. The peak of hospital staywas between 15 and 28 days. Within 28 days (four weeks) of admission, 47 (69.1 percent) of the children had been discharged following surgery while 16 (23.5 percent) of them spent 14 days (2 weeks) or less in hospital. There was at least one patient from all the reported surgical risk categories that stayed for more than six weeks in hospital. Post operative complications encountered were bronchopneumonia(4), pleural effusion (3), low cardiac output heart failure(2), multi organ failure (2), septicaemia(2), pneumothorax (1), pericardial effusion(1) and acute respiratory syndrome (1).

The in-hospital case fatality rate in this series was 4.4 percent i.e. 3/68. One case occurred in the youngest patient in the series, a 3 month old female

with Down syndrome, Ventricular Septal Defect (VSD), Patent Ductus Arteriosus (PDA) and moderate pulmonary hypertension. She repeatedly developed respiratory distress following extubation and therefore required reintubation. By the 20th day post operatively, she developed right pneumothorax and low cardiac output. Terminally she suffered from acute respiratory distress syndrome and multi-organ failure and died 27th day post op. The second mortality occurred in a year old male infant with Down syndrome, large VSD, muscular right ventricular outflow tract, small PDA and diaphragmatic hernia. The patient developed repeated cardiac arrest from a low cardiac output state post operatively and died 6 weeks post op. The

Table II

Duration of Hospital Stay by Surgical Procedures and Surgical Risk Category.

Risk Categor	Surgical Procedures		Duration of Hospital Stay				Total (%)
	up lage 'rogano'e strong 'college's lage to the college and th		<1-2weeks	3-4weeks	4-6weeks	>6weeks	1.3
1	PDA surgery at age > 30 d	9	3 are substit	6	o-surenti	pa Nel esti il	10(14.7)
	Secundum ASD repair & PAPVR	1	4000000		Hint ber	1	- 18 Tale 30
2	VSD repair	2	1 m years	6	5	g de sa	39(57.4)
	VSD repair and Subaortic stenosis resection	1	1	d - du 📑	.01-m-4		I Mare N
	VSD repair, pulmonary infundibular						
	resection and PDA closure	1	2 sedwh	1	1 1000	drobball	
	VSD repair & infundibular resection	2	1		generally	distant to	
	VSD repair and pulmonary valvotomy	1	1		1	11-217	
		2	4	8	3	2	
	THE PARTY OF THE P	7	2	1			
		3					
3	Repair of complete or transitional AV		2	2	1	2	15(22.1)
		7	# 10 TO	2		-	
	Repair of TOF with PA	2	41711 (419)		1	1	
	Repair of DORV	2	1	1	-		
	Primum ASD repair and Mitral						
	valvuloplasty	2	- 10/C L		2	The state of the s	
		2					
4	and the contract of the contra	1			1		4(5.9)
	Repair of TA	1		1			
	Repair of complex anomaly						
		2	<u> </u>			1	
Total ((%)	68	16(23.5)	29(42.6)	16(23.5)	7(10.3)	68(100)

^{*}No patient fitted RACHS-1 Risk Category 5 (Tricuspid valve repositioning for neonatal Ebstein anomaly at age d"30 d, repair of truncus arteriosus and interrupted arch) and 6 (Stage 1 repair of hypoplastic left heart syndrome (Norwood operation), Stage 1 repair of nonhypoplastic left heart syndrome conditions, Damus-Kaye-Stansel procedure).

PDA=Patent ductus arteriosus; ASD=Atrial septal defcet; PAPVR=Partial anomalous pulmonary venous return; TOF= Tetralogy of Fallot; AV=Atrioventricular; PA=Pulmonary atresia; TA= Truncus arteriosus.

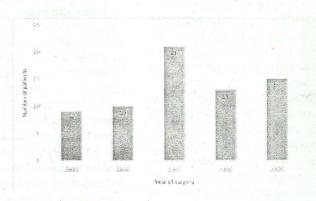


Figure 1: Total number of patients undergoing surgery by year

third mortality was also a year old male child with large VSD who also developed low cardiac output failure post operatively. The three in-hospital mortalities belonged to risk category 2 surgeries with 39 cases giving an in-hospital case fatality rate of 7.7 percent for that category. Sixty three (92.6 percent) of the 68 surgeries occurred in India and the three deaths occurred there. Two deaths occurred in one facility where 36 surgeries had been done for our patients giving an in-hospital case fatality rate of 5.6% for our patients treated in that that hospital while the third death occurred in a hospital where 9 surgeries had been done giving a rate of 11.1 percent.

Discussion

Our study involved a series of children undergoing cardiac surgical management abroad. We report the in-hospital fatality outcome of these children. The case fatality rate of 4.4 percent in this study falls within the rates of 4-6 percent reported from other studies in Africa and Asia. 8-11 The in-hospital death rates for Africa ranged from 4.3 percent to 6 percent for surgeries either by doctors within the continent or through programmes by visiting teams. 8-10 This contrasts greatly with the pre-cardiac surgery era in Africa where death of most cases with fatal congenital heart disease was the rule. A report by Gupta and Antia in 1967 showed that by age two years only 40 percent of children with congenital heart disease in Nigeria was still alive. 15 The low rates reported may also be because in those series, complex cases belonging to RACHS-1 risk categories 5 and 6 were not done.

Death following cardiac surgery is influenced by many factors. The type of surgical procedure carried out has been adjudged the most important risk factor for groups of children. ¹⁶ For in-hospital deaths, other reported factors are female sex, non-elective surgeries and case volume of the surgical center. ¹⁷ The fatality rate differed in the two hospitals were deaths occurred. Factors responsible for this difference may include difference in level of expertise of the operating team and quality of post operative care in each of the hospitals.

In recent times, some centres have been able to achieve zero in-hospital mortalities in surgeries on some conditions like atrial septal defect, ventricular septal defect, atrio-ventricular canals and tetralogy of Fallot. ¹⁸ When other more complex cardiac conditions are added, the mortality increases. A study in California reported an in-hospital mortality of 5.9% and the significant risk factors were truncus arteriosus repair, total anomalous pulmonary vein repair, aortopulmonary shunt, and open valvotomy. ¹⁹ These are risk category 4 and above procedures. In

our series, the majority (65 percent) of the surgical procedures done belonged to risk categories 2. The three in-hospital mortalities occurred following risk category 2 surgeries. All three were less than two years of age with the youngest aged three months. No child in the series had risk category 5 and 6 surgeries which may have contributed to the low (4.4 percent) in-hospital case fatality.

Complications occurring during or after cardiac surgery could be a direct cause of death. In our series, although respiratory problems accounted for most (13.2 percent) of the post operative complications recorded, two out of the three case fatalities had complications related to low cardiac output. This is similar to findings by other authors. 20, 21 Ma et al 20 reported that out of 100 post cardiac surgery death cases, 52 had low cardiac output states. This included cases with inadequate postoperative physiology, ventricular failure, pulmonary hypertension, and severe atrioventricular valvar regurgitation. As in two of our cases, the majority of those deaths occurred because the overall cardiovascular physiology, even with a technically adequate repair and intensive support, was inadequate to meet physiologic demands.

Genetic abnormalities occur in 20 percent of children with congenital heart disease. In this series, 15 of the 68 cases had Down syndrome. Two (66.7 percent) of the three deaths had associated Down syndrome giving a case fatality rate of 13.3 percent among the children with Down syndrome. Neonates with genetic abnormalities have been reported to have a higher risk of postoperative complications and a longer hospital length of stay but no increase in hospital mortality.22 In our study, the patient with the longest hospital stay of three months had Down syndrome but was not one of the fatalities reported. Other studies have also found that associated Down syndrome is not independently associated with mortality risk in cardiac surgery.^{23,25} A report from Atlanta²⁶ showed higher mortality among black children with congenital heart disease and Down syndrome but linked it to a more difficult access to cardiac care in these patients rather than cardiac surgery.

Female sex has been reported to be associated with a higher mortality following paediatric cardiac surgery when compared with the male.^{27, 28} The reason for this is not very clear. In the current study, the three deaths occurred in males which could possibly be a reflection of the low study numbers and the preponderance (55.9 percent) of males among those who had surgery.

In conclusion, the in-hospital outcome is good with a low case fatality rate of 4.4 percent for heart surgeries among Nigerian children referred abroad for cardiac surgery. Young age, low cardiac output post operatively and associated syndrome were identified in the cases with mortality in this series.

References

- Seifert HA, Howard DL, Silber JH, Jobes DR.
 Female gender increases the risk of death
 during hospitalization for pediatric cardiac
 surgery J Thorac Cardiovasc Surg. 2007; 133: 66875.
- Curzon CL, Milford-Beland S, Li JS, O'Brien SM, Jacobs JP, Jacobs ML, Welke KF, Lodge AJ, Peterson ED, Jaggers J. Cardiac surgery in infants with low birth weight is associated with increased mortality: analysis of the Society of Thoracic Surgeons Congenital Heart Database. J Thorac Cardiovasc Surg. 2008; 135:546-51.
- Allen SW, Gauvreau K, Bloom BT, Jenkins KJ.
 Evidence-Based Referral Results in
 Significantly Reduced Mortality After
 Congenital Heart Surgery Pediatrics 2003;
 112;24-28
- Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI. Consensus-based method for risk adjustment for surgery for congenital heart disease. JThorac Cardiovasc Surg. 2002; 123:110–118
- 5. Pezzella AT. International aspects of cardiac surgery. Ann Thorac Surg 1998; 65:903–4.
- Anyanwu CH, Ihenacho HNC, Okoroma EO, Nwafo DC, Umeh BU, Okechukwu CC, Udekwu FA. Initial experience with open-heart surgery in Nigeria. Tropical Cardiology 1982; 8:123–7.
- 7. Eze J, Ezemba N. Open-Heart Surgery in Nigeria Indications and Challenges *Tex Heart Inst J* 2007; 34: 8–10.
- 8. Cohen AJ, Tamir A, Houri S, Abegaz B, Gilad E, Omohkdion S, Zabeeda D, Khazin V, Ciubotaru A, A. Save a child's heart: we can and we should. *Ann Thorac Surg* 2001; 71:462-8.
- 9. Tefuarani N, Hawker R, Vince J, Sleigh A, Williams GM. Surgical programme at Royal Alexandra Hospital, Sydney, for Papua New Guinea children with congenital heart disease, 1978-1994. J Paediatr Child Health 2002; 38:178-82.
- 10 Parry MM, Cooper DK, Barnard CN. Trends in cardiac surgery at the University of Cape Town, 1971-1981. *S Afr Med J* 1983; 63:189-93.
- 11 Xu ZW, Su ZK, Ding WX. Clinical analysis of surgical repair of congenital heart disease for 787 patients in less than 6 months of age. Chin J Clin Thorac Cardiovasc Surg 2005; 12: 69-72.

- 12.O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C, Welke KF, Maruszewski B, Tobota Z, Miller WJ, Hamilton L, Peterson ED, Mavroudis C, Edwards FH. An empirically based tool for analyzing mortality associated with congenital heart surgery. J Thorac Cardiovasc Surg 2009; 138:1139-53
- 13. Karamlou T, Diggs BS, Ungerleider RM, Welke KF
 Evolution of treatment options and outcomes
 for hypoplastic left heart syndrome over an 18year period *J Thorac Cardiovasc Surg*2010;139:119-27
- 14. Billett J, Majeed A, Gatzoulis M, Cowie M. Trends in hospital admissions, in-hospital case fatality and population mortality from congenital heart disease in England, 1994 to 2004. *Heart* 2008; 94:342-8.
- 15. Gupta B, Antia AU. Incidence of congenital heart disease in Nigerian children. *Brit Heart J*: 1967, 29: 906-09.
- 16. Jenkins KJ, Gauvreau K, Newburger JW, Spray TL, Moller JH, Iezzoni LI, Consensus-based method for risk adjustment for surgery for congenital heart disease (RACHS-1) method. J Thorac Cardiovasc Surg 2002; 123: 110-8
- 17. Chang RK, Chen AY, Klitzner TS. Female sex as a risk factor for in-hospital mortality among children undergoing cardiac surgery *Circulation* 2002; 106:1514 22.
- 18. Ungerleider RM, Bengur AR, Kessenich AL, Liekweg RJ, Hart EM, Rice BA, Miller CE, Lockwood NW, Knauss SA, Jaggers J, Sanders SP, Greeley WJ. Risk factors for higher cost in congenital heart operations *Ann Thorac Surg* 1997; 64:44-8
- Chang RK, Rodriguez S, Lee M, Klitzner TS. Risk factors for deaths occurring within 30 days and 1 year after hospital discharge for cardiac surgery among pediatric patients. Am Heart J. 2006; 152:386-93.
- 20. Ma M, Gauvreau K, Allan CK, Mayer JE Jr, Jenkins KJ. Causes of death after congenital heart surgery. *Ann Thorac Surg* 2007; **83**:1438-45.
- 21.Parr GV, Blackston EH, Kirklin JW. Cardiac performance and mortality early after intracardiac surgery in infants and young children. Circulation 1975; 51:867–75.
- 22. Simsic JM, Coleman K, Maher KO, Cuadrado A, and Kirshbom PM. Do Neonates with Genetic Abnormalities Have an Increased Morbidity and Mortality following Cardiac Surgery? Congenit Heart Dis 2009; 4:160–165
- 23. Baciewicz FA Jr., Melvin WS, Basilius D, Davis JT. Congenital heart disease in Down's syndrome

- patients: a decade of surgical experience. *Thorac Cardiovasc Surg* 1989; 37:369-71.
- 24. Kabbani MS, Giridhar S, Elbarbary M, Elgamal MA, Najm H, Godman M. Postoperative cardiac intensive care outcome for Down syndrome children. Saudi Med J 2005; 26:943-6
- 25. ParvathyU, Balakrishnan KR, Ranjith MS, Saldanha R, Sai S, Vakamudi M. Surgical experience with congenital heart disease in Down's syndrome. *Indian Heart J* 2000; 52:438-41
- 26. Rasmussen SA, Wong LY, Correa A, Gambrell

_...

- D and Friedman JM. Survival in infants with Down syndrome, Metropolitan Atlanta, 1979–1998, *J Pediatr* 2006; 148:806–812
- 27. Klitzner TS, Lee M, Rodriguez S, Chang RK. Sexrelated disparity in surgical mortality among pediatric patients. *Congenit Heart Dis.* 2006; 1:77-88
- 28. Seifert HA, Howard DL, Silber JH, Jobes DR. Female gender increases the risk of death during hospitalization for pediatric cardiac surgery. *J Thorac Cardiovasc Surg.* 2007; 133:668-75