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Original Article

CHALLENGES ON THE MANAGEMENT OF COMPLEX CONGENITAL HEART DISEASE IN CHILDREN IN NIGERIA: THE IRRUA SPECIALIST TEACHING HOSPITAL EXPERIENCE

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ABSTRACT

Back ground: Complex congenital heart disease (CCHD) is a group of life threatening heart defects that are present at birth. Children born with CCHD in Nigeria have inadequate cardiovascular services leading to increased morbidity and mortality. This study was conducted to document the challenges encountered and outcomes in the management of children with complex congenital heart disease seen in Irrua Specialist Teaching Hospital (ISTH), in Edo State.
Method: The records of children seen by the Paediatric Cardiology unit of ISTH Irrua from August 2023 to October 2024 were reviewed. The biodata, diagnosis, investigations and outcome of interventions were obtained. Data was analyzed using Excel spreadsheet.

Results: A total of 177 children had congenital heart disease. Thirty one (17.5%) of these children had complex congenital heart disease. Their ages ranged from 2 days to 16 years with an M: F of 2.4:1. The challenges encountered in their management were lack of resources for life saving surgical interventions within and outside Nigeria, lack of facilities to make accurate diagnosis in some cases and shortage of full complement of experienced skilled personnel when resources were available. Five children (16.1%) died over the period under review while 7 (22.6%) have not returned for follow up.

Conclusion: There is a need for government funding and support in the treatment of CCHD in children. The establishment of regional cardiac centers with trained specialized manpower (foreign and local) and highly subsidized cost of treatment will mitigate the challenges encountered in the management of children with CCHDs and improve outcome.

Key words: Congenital heart disease, Tetralogy of Fallot, Cor triatriatum dexter, Outcome, Irrua Specialist Teaching Hospital, Nigeria.

Introduction

Congenital heart disease (CHD) is a structural abnormality of the heart and/or great vessels that is present at birth (Santiago *et al.,* 2024). It is the most frequent cause of severe birth defects, affecting 6–8/1,000 live births each year (Van *et al.,* 2021). Complex congenital heart disease (CCHD) is a group of potentially life-threatening heart defects present at birth from which infants can die or need to undergo invasive procedures (surgery or cardiac catheterization) early in life. (Goldstein & Krasuski., 2024) Children with CCHD are at approximately 12 times higher risk of mortality in the first year of life (Otaigbe & Tabansi., 2014). It is estimated that about 15% of patients with CHD have a defect that is categorised as CCHD. (Santiago *et al.,* 2024)

In developed countries, prenatal diagnosis is used to detect CHD and CCHD before birth, but in developing countries, this is not common. After birth only a few children in developing countries are detected and a minimal number benefit from surgical treatment (Ana *et al.,* 2011). This is a very sad reality. More than 90% of children born with moderate and complex congenital heart defects are expected to survive into adulthood and life expectancy is almost at par with the general population, in developed countries (Reid *et al.,* 2006). This cannot be said of developing countries (Otaigbe & Tabansi., 2014). Children with CCHD often require palliative or definitive surgical interventions to restore cardiopulmonary function. Lack of early cardiac intervention contributes to large numbers of preventable deaths and sufferings among them (Ujunwa *et al.,* 2021).

Due to the heterogeneity of CHD, the classification of the complexity of CHD has been repeatedly revised (Stout *et al.,* 2019). They were previously classified by their anatomic complexity and the presence or absence of cyanosis. The 2018 American College of Cardiology (ACC/ American Heart Association (AHA) Guidelines created the Adult with CHD Anatomic and Physiological Classification System (Ujunwa *et al.,* 2021). This system includes the native

anatomy, stage of surgical repair and the current physiology and functional state of the heart (Stout *et al.,* 2019). Using this resource the scope of CCHD includes tetralogy of Fallot (TOF), hypoplastic left heart syndrome, pulmonary atresia with intact ventricular septum, Ebstein anomaly, tricuspid atresia, transposition of the great arteries (TGA), univentricular heart, interruption of the aortic arch, anomalous pulmonary venous connection, atrioventricular septal defect, multiple simple defects, and Eisenmenger syndrome (Stout *et al.,* 2019, Congenital heart disease. 2022). The heterogeneity and complexity of congenital heart disease require deep knowledge of congenital heart diseases, expertise in the diagnosis of the defects using cutting-edge technology and a multimodality/multidisciplinary approach (Festa *et al.,* 2023). Cardiac imaging plays a critical role in the diagnosis, treatment, and surveillance of CCHD. The introduction and dissemination of cutting-edge multimodality imaging techniques are instrumental in understanding the structure and function of the heart in such patients (Sachdeva *et al.,* 2024). Furthermore, advanced imaging enables periprocedural planning for surgical and catheter-based interventions and improves patient outcomes (Sachdeva *et al.,* 2024). The recent addition of artificial intelligence has increased the speed and flexibility of handling complex imaging data. It helps in patient selection and protocoling, image acquisition, signal denoising, image registration and rendering, quantification, and interpretation (Dey *et al.,* 2023).

These cutting-edge multimodality imaging techniques and surgery are the recommended interventions in CCHD. They include echocardiogram, Cardiac Magnetic Resonance Imaging, Cardiac Catheterization and Angiogram, CT (Computerized Tomography) of the Heart, Transesophageal echocardiogram (TEE) (Sachdeva *et al.,* 2024) and surgery (Ujunwa *et al.,* 2021). In middle and low income countries (MLIC), like Nigeria, these interventions are not readily available (Lakshmanan & Mbanze., 2023).There is also the challenge of the dearth of specialized manpower (paediatric cardiac surgeons, interventional cardiologists, paediatric intensivists etc) and well-equipped intensive care units (Kelechi & Charles., 2016). The treatment and rehabilitation of children with CCHD is very specialized and expensive (Chinawa *et al.,* 2013). In Irrua, Nigeria, many parents cannot afford the treatment and rehabilitation of their children and this can be attributed to financial constraints, disparities in healthcare and technical infrastructure. This is made worse by the rapid depreciating value of the Nigerian

national currency (Naira) since 2023(World Food Programme, n.d.), Occasional short duration, foreign congenital cardiac surgery missions are inadequate in addressing complex heart defects in children in Nigeria and many patients cannot afford medical tourism to India, Ghana, Israel, Italy Sudan, Egypt, South Africa, UK and USA (Ikechukwu & John ,2019), Several advances have been made in developed countries regarding prenatal and post-natal diagnosis, treatment and follow-up of children with difficult and complex congenital cardiac diseases resulting in favorable outcomes in the affected children (Ana *et al.,* 2011, Reid *et al.,* 2006). This does not apply to the children seen in ISTH Irrua and probably other tertiary hospitals in Nigeria. This study is aimed at documenting the pattern of CCHD in children seen in ISTH, the challenges encountered in the management of these defects and possible mitigating measures to improve outcomes in the children.

Subject and methods:

The study was a retrospective review of the records of children seen in the Paediatric Cardiology unit of Irrua Specialist from 1st of August, 2023 to 31st October, 2024. The data retrieved were, age, gender, echocardiogram report, diagnosis, treatment and outcome. The echocardiogram was done with a Vivid T8 Cardiac ultrasound machine by GE Health Care. The scope of CCHD in this study includes tetralogy of Fallot (TOF), Ebstein anomaly, tricuspid atresia, univentricular heart, anomalous pulmonary venous connection, atrioventricular septal defect, Eisenmenger syndrome, unqualified complex heart defects, cor triatriatum, single atrium, simple but complex defects (Congenital heart disease.,2022) (VSD, ASD, PDA in a child less than 5kg) and aortopulmonary shunts. The children were categorized into 4 groups 0< 12 months, 12<60 months, 60< 144 months, and 144< 216 months. Data was analyzed using an Excel spreadsheet.

Results: A total of 188 records of children with structural heart diseases were reviewed, 177 (94.1%) had congenital heart disease and 11 (5.9%) had acquired heart disease. Thirty-one (17.5%) of the children with congenital heart disease had complex congenital heart disease. Their ages ranged from 2 days to 196 months (16 years) with a mean age of 40.5 months. There were 22 males (71%) and 9(29%) female with an M: F of 2.4:1.

Table 1 shows the age distribution of children with CCHD. Children less than 60 months were affected (71%) more than those above 60 months (29%). Among those less than 60 months, those in the 12-60-month age group (38.7%) were the most affected.

Table 1: Age distribution of children with complex congenital heart disease

AGE IN MONTHS	**NUMBER OF DEFECTS**	**AFFECTED MALES**	**AFFECTED FEMALES**
0-1	2	0	2
1-12	8	5	3
12- 60	12	9	3
60-144	5	4	1
144-216	4	4	0
TOTAL	**31**	**22**	**9**

Table 2 shows the types and frequency of occurrence of CCHD in the reviewed cases. Tetralogy of Fallot (TOF) was the most common complex congenital heart disease (16.1%), followed by cor triatriatum dexter (13%) and complete atrioventricular septal defect (12.9%).

Table 2: Types and frequency of occurrence of CCHD

COMPLEX CONGENITAL HEART DISEASE	**NUMBER (%)**
TOF	5 (16.1)
Univentricular heart	2 (6.5)
Ebstein anomaly	1(3.2)
Single atrium	2 (6.5)
Cor triatriatum dexter (CTD)	2(6.5)
Atrial septal defect(ASD)/ Cor triatriatum dexter (CTD)	2(6.5)
Complete Atrioventricular septal defect (AVSD)	4(12.9)
Transitional AVSD	2(6.5)
Partial anomalous pulmonary venous connection (APVC)	2(6.5)

ASD, Patent ductus arteriosus (PDA), Critical Pulmonary Stenosis (PS)	1(3.2)
ASD/PDA/Tricuspid regurgitation(TR)/Mitral regurgitation(MR)/complex anatomy	1(3.2)
ASD/Ventricular septal defect (VSD)/PDA	3 (9.7)
Tricuspid atresia	1(3.2)
TOF/ASD/PDA	1(3.2)
VSD/Aorto-Pulmonary (AP) shunt	1(3.2)
ASD/AP shunt	1(3.2)
TOTAL	31(100)

Six patients had AVSD (complete and transitional); five had Down syndrome while one was a non-Down AVSD.

Transthoracic echocardiogram (TTE) was the investigation used in the study of the cardiac structure in the affected children. One (3.2%) patient with TOF who was referred to a cardiac centre in Nigeria for surgery had a cardiac CT and pulmonary angiogram done. He is yet to have surgery due to financial constraints. One (3.2%) patient with PAPVC had free surgical repair by a visiting cardiac surgery mission in Oraifite, Anambra state in September 2024. Five (16.1%) patients died within the period under review, they were:

1. ASD/PDA/MR/TR/complex anatomy, died on the 18th day of life
2. Complete AVSD (Down), died at 18 months
3. VSD/AP shunt, died intra-operation at 84 months
4. ASD/PDA/critical PS, died on the 3rd day of life
5. Single atrium, died at 3 months

Seven (22.6%) children were lost to follow-up while nineteen (61.3%) of the children in this study are on medical management. The 19 children on follow-up and medical management are financially incapable of paying for treatment both in Nigeria and abroad.

Discussion

In this study 17.5% of children with CHD seen over the period under review had CCHD. The reported incidence of CCHD among children with CHD is 15%-49.5 %. (Santiago *et al.,* 2024, Ujunwa *et al.,* 2021). The number found in this study falls within the reported range. (Ujunwa *et al.,* (2021) had a higher prevalence (49.5%) of CCHD among the children they studied. This may be because their center is a major referral center for cardiac surgery, so, many children with CCHD are referred to them for definitive surgery.

In this study, more males had CCHD than females (71% vs 29%), similar to the report by (Ujunwa *et al.,* 2021) (57.1% vs 42.9%) and Chinawa et al (87.5% vs 12.5%) (Chinawa *et al.,* 2013) The exact reason for this is not known, but this male preponderance has been reported in literature and the reason was not known (Pugnaloni *et al.,* 2023)

Complex congenital heart disease was noted mainly in children less than 60 months (71%), especially in the 12-60 month age group (38.7%). This may be due to the late presentation of the patients to the hospital. In the studies by other authors (Ujunwa *et al.,* 2021, Chinawa *et al.,* 2013), the incidence was also higher in children less than 60 months. Tetralogy of Fallot (16.1%) was the commonest CCHD in this study and this is comparable with the report by Ujunwa *et al.,* (2021) (55%) and Chinawa et al (37.5%) (Chinawa *et al.,* 2013) The other common CCHD found in this study was complete AVSD (12.9 %). Cor triatriatum dexter was seen as the only defect in 2(6.5%) children, and in combination with ASD in 2 (6.5%) children too. The combined incidence of 13% in this study is contrary to the rarity of this defect reported in the literature (Kalangos *et al.,* 2020). It can occur as an isolated cardiac anomaly or in association with other heart abnormalities (Simsek *et al.,* 2014) as in this study. A child with ASD/PDA/TR/MR/complex anatomy needed more investigations urgently to delineate the cardiac anatomy and show the cardiac function but this was not possible due to the unavailability of the needed equipment and financial constraints. The child died in the neonatal period. Five children (16.1%) died during the period under review, this is higher than the reported mortality in CCHD in developed countries (Çaylan *et al.,* 2024). Death from CCHD is expected to be higher in developing countries due to many factors and most patients die in the first 28 days of

life (Ujunwa *et al.*, 2021), one child with PAPVC had a free repair by a visiting cardiac surgery mission, 7 were lost to follow up and 19 are on medical management. All the patients reviewed could not afford treatment within or outside Nigeria. In some of the cases, advanced technology would have helped in the definitive diagnosis of the structural defects as TTE was not exhaustive. These were not readily available or out of reach in the few private facilities that have them. Some CCHD surgeries are performed in some centers in Nigeria with variable outcomes (Nwafor & Eze, 2019). Some parents would rather travel to India, hoping for better outcomes. It was cheaper and possible to appeal for funds to take children to India for surgery before 2023 in Nigeria but with the loss of value of the naira (World Food Programme, n.d.), the cost of surgery for one child with CCHD is enormous. The enormity of the cost of surgery for only one child with CCHD became a source of discouragement to donors, especially as the outcome of the surgery cannot be predicted.

The challenges faced in the management of the children in this study include the lack of facilities for diagnosis and treatment of the patients. When these facilities were available, they were limited to big cities and private hospitals where the cost of investigations and treatment was very high and inaccessible to the poor. Of note, is the fact that government-owned cardiac centers are also very expensive and inaccessible to the poor? There is also absence of health insurance cover for congenital heart disease and parents of these children have to make out of pocket payments. There is the added burden of the dearth of specialized manpower needed for the treatment and care of children with CCHD. This is worsened by brain drain as the few trained specialized man power leave the country for greener pastures.

Conclusion

There is need for government funding and support in the treatment of CCHD in children in Nigeria. The establishment of regional cardiac centres with trained specialized manpower (foreign and local) and highly subsidized cost of treatment will mitigate the challenges encountered in managing children with CCHDs and improve outcome.

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