

Congenital Cardiac Malformations: A Serious Health Problem in Nigeria?†

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Summary

Jaiyesimi F. Congenital Cardiac Malformations: A Serious Health Problem in Nigeria? *Nigerian Journal of Paediatrics* 1982; 9: 67. Congenital heart disease is a serious health problem in Nigeria and, with its incidence at 3.5 per thousand births, approximately 12,000 babies with various types of cardiovascular malformations are born in the country annually. Because of gross inadequacy of medical facilities, only a minority of the cases are detected, commonly at a late stage after complications have occurred. In a majority of children with these malformations, the aetiological factors are unknown. However, recognized factors such as congenital rubella and perinatal asphyxia are preventable. A plea is made for the provision of adequate medical and surgical facilities at existing centres which care for children with heart disorders.

Incidence and Prevalence

It seems appropriate to begin this communication by asking the question, 'Are congenital malformations of the heart a serious health problem in Nigeria?'. This question is necessary considering the fact that only two decades ago, there were no reports of these malformations among Nigerian children. Even when the first reports appeared, one of them dealt with only 67 cases seen in Ibadan by Caddell and Morton¹ during an 18-month period. It might therefore have been permissible then, to doubt the significance of cardiac malformations as a cause of morbidity

and mortality in Nigerian children. This doubt no longer exists. One of the earliest pointers to the serious problem posed by these malformations came also from Ibadan in 1967 when Gupta and Antia,² in a study of 4,220 births, estimated the incidence of congenital heart defects in Nigeria to be 3.5 per thousand. This estimated incidence was close to those of between 3 and 9 per thousand reported from Europe and America.³⁻⁵ However, this incidence becomes more meaningful when it is related to the number of babies who are born annually in this country.

According to the 1977 World Population Data Sheet,⁶ the crude birth rate in Nigeria is 49 per thousand. Assuming a population of 70 million, this means that about 3.5 million babies are born annually. With the incidence of cardiac malformations at 3.5 per thousand, it means that about

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12,005 children with malformed hearts are born in this country every year.

Hospital data also attest to the high prevalence of cardiac malformations. In the University College Hospital (UCH), Ibadan, 72% of children who attend the paediatric cardiology clinic have cardiac malformations, and an average of 100 such new patients are seen every year. Since about 80% of Nigerians reside in rural areas and do not have routine access to the teaching hospitals, it is evident that children with congenital heart disease seen in a centre such as the UCH, represent only a minority of the patients.

Aetiology

In our recent study of 635 children with cardiac malformations,⁷ aetiological factors were identified in only 72 cases (11%). This was not surprising because it accorded with the well-known fact that in most cases of cardiac malformations, the aetiology is unknown. But the few recognized aetiological factors (Table I) deserve mention in that some of them are preventable. In this regard, it is worth noting that congenital rubella and perinatal asphyxia, both largely preventable conditions, accounted for 57 (79%) of the 72 cases.

TABLE I
*Aetiological Factors in 72 Children with
Congenital Malformations of the Heart*

| Factor | No. of Cases | % of Cases |
|--------------------------------|--------------|------------|
| Intrauterine rubella infection | 43 | 60 |
| Neonatal asphyxia | 14 | 19 |
| Chromosomal aberrations | 13 | 18 |
| Genetic | 2 | 3 |
| Total | 72 | 100 |

One question which is often asked by parents of affected children is the probability of subsequent siblings being affected. Of 631 families in our study,⁷ there were only four families from each of whom two siblings were affected. However, studies from other centres have estimated the incidence of congenital heart disease to be ten times higher in siblings of patients than in the general population.⁸

Type of Defect

Table II summarizes the prevalence of the major malformations in 635 children. Ventricular septal defect was the commonest type and occurred

TABLE II
*Prevalence and Sex Ratio of Major Cardiac Malformations in 635
Children*

| Type | % of Cases | M/F Ratio |
|---------------------------------|------------|-----------|
| Ventricular septal defect | 35 | 1 : 1 |
| Persistent ductus arteriosus | 22 | 1 : 2 |
| Fallot's tetralogy | 10 | 2 : 1 |
| Pulmonary stenosis | 9 | 1.3 : 1 |
| Atrial septal defect | 7.5 | 1 : 2 |
| Transposition of great arteries | 4.5 | 1.3 : 1 |
| Coarctation of aorta | 2 | 1.4 : 1 |
| Others | 10 | 1 : 1 |
| Total | 100 | 1 : 1 |

in over a third of the patients, followed by persistent ductus arteriosus (PDA). Fifty-seven (41%) of the 138 cases of PDA were attributable to congenital rubella and perinatal asphyxia. Fallot's tetralogy, the commonest cyanotic malformation beyond infancy, occurred in 10% of the patients. Aortic stenosis and coarctation of the aorta were relatively uncommon, the former occurring in only four (0.6%) of the 635 cases.

Atrial septal defect and PDA were more common in girls while ventricular septal defect occurred equally in both sexes and the other major defects were commoner in boys.

Peculiar Aspects of the Disease

Certain peculiarities of the disease in Nigeria are worth highlighting. The first is the rarity of aortic stenosis. In Europe and North America, the prevalence of this lesion varies between 5.5 and 7.1% and among South African whites, it accounts for 5.6% of cases.⁷ By contrast, in Nigeria and other black African countries, its prevalence is less than one per cent.⁷

It has long been recognized that there is an association between hypercalcaemia in infancy and aortic stenosis.^{9 10} On the basis of this, it has been suggested that foetal and infantile hypocalcaemia, resulting from maternal vitamin D deficiency and relatively low calcium content of breast milk, may be a factor that protects African children against aortic stenosis.⁷

Another peculiar aspect of the disease is the late presentation in hospital by our patients (Table II). The natural but unfortunate sequela of this delayed presentation was that many of the patients had developed complications before they sought medical attention. Such complications invariably result in increased morbidity and mortality.⁷

TABLE III

Age at Presentation of Children with the Three Most Common Cardiac Malformations

| Age | Percentage of Patients with | | |
|----------------|-----------------------------|-----|--------------------|
| | VSD | PDA | Fallot's Tetralogy |
| Below 1 month | 9 | 12 | — |
| 1 month-1 year | 55 | 46 | 9 |
| 1-5 years | 28 | 30 | 53 |
| Above 5 years | 8 | 12 | 38 |
| Total | 100 | 100 | 100 |

VSD = Ventricular Septal Defect
 PDA = Persistent Ductus Arteriosus

Occasionally, local customs and beliefs have prevented effective management. An example of this situation is provided by a 4-year old girl who was to be admitted for surgical ligation of a PDA. After discussing the proposed operation with her parents, they requested to be given some time to think it over. During the next visit to hospital, the child's father, a hospital porter, informed us that he would not consent to the operation because his own mother had decided that the patient's illness would be best treated at home and not in the hospital. The girl has since defaulted from hospital. Unfortunately such refusals of treatment are not limited to illiterate parents. Another example concerned a university lecturer who declined to have his child's PDA ligated because the family caucus had decided against it.

Prognosis and Prospects

The prognosis for the Nigerian child with congenital cardiac malformations depends largely on the type of defect. Relatively simple surgical procedures like ligation of ductus arteriosus and coarctectomy, both of which require no cardio-pulmonary by-pass and carry negligible risks, can be performed in a few centres in Nigeria.^{11 12} The outlook for patients with these defects is therefore, good. Similarly, effective palliation can be carried out on patients with Fallot's tetralogy. Hence, the short-term prognosis for this group of patients is fair.

However, the prospects for patients with complex lesions, the repair of which requires cardio-pulmonary by-pass, are dim. Because of limited facilities, it has not been possible to carry out open-heart procedures on young children. A small number of such patients had to be referred to better-equipped centres outside Nigeria. But the high cost of the treatment, about ₦4,000 in Abidjan, Ivory Coast, and up to £12,000 in the United Kingdom, is beyond the capability of most Nigerian families. Consequently, many of these unfortunate patients only hang on tenuously to life until they die of heart

failure, hypoxaemia or intercurrent infection. About 71 per cent of such deaths occur during the first year of life.

This is a distressing situation because a majority of these patients have remediable malformations and therefore, need not die. They can be salvaged if genuine efforts are made to equip a few centres in the country to undertake surgical repair of cardiovascular malformations. Indeed, the establishment of a single national Institute devoted to the study, prevention and treatment of cardiovascular diseases will significantly reduce the morbidity and mortality currently attributed to cardiac malformations.

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