Children With Tetralogy of Fallot in an Urban Centre in Africa

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Introduction: Tetralogy of fallot (TOF) is the most common cyanotic heart lesion beyond the neonatal period and it accounts for a third of all congenital heart disease in patients less than 15 years of age. It occurs in 10% of all congenital heart disease. The prevalence of TOF is approximately 3.9 per 10000 live births in the United States and 10%-26.2% of all congenital heart diseases in Nigeria. Diagnosis of TOF is confirmed with echocardiography. Definitive diagnostic features and other associated cardiac abnormalities can be identified. In advanced countries, diagnosis can be made in as early as 12 weeks of gestation with fetal echocardiography. Management is both medical and surgical and depends on the degree and type of right ventricular outflow tract obstruction and the centre’s protocol. There is a dearth of literature on TOF in children in Sub-Saharan Africa. This study aim to describe the prevalence, clinical profile and associated cardiac anomaly of children diagnosed with TOF documented over an eight year period in a tertiary hospital in South Western Nigeria.

Materials and Methods: A prospective review of all consecutive cases of TOF diagnosed with echocardiography at the Lagos State University Teaching Hospital (LASUTH) between January 2007 and December 2014. Data were analyzed using SPSS version 20. Tables and charts were used to depict those variables. Descriptive statistic are presented as percentages or means and standard deviation. Means of normally distributed variables were compared using the student t test and proportions using chi-square test. Skewed distribution were analyzed using appropriate non-parametric tests. Level of significance set at P < 0.05.

Result: The prevalence of TOF among children presenting at LASUTH at the study period was 4.9 per 10000 while its prevalence among those with congenital heart disease was 16.9%. There was a male predominance and most children presented within 1-5 years of age. Chromosomal abnormalities such as Down syndrome, Turner syndrome and CATCH 22 syndrome were documented in some subjects. Some of the subjects had atypical presentation.

Conclusion: TOF is as common in Nigeria as other parts of the world, there is a need to established cardiac centers to salvage these children. Collaboration from developed countries will be helpful in this resource limited region.

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TOF was made with echocardiography.\textsuperscript{3} The data were analyzed using SPSS version 20. The prevalence of TOF was calculated from all children who presented in the hospital as in-patient and out-patient during the study period. The prevalence of TOF was also calculated amongst those with congenital heart lesions and those with cyanotic congenital heart disease. Tables and charts were used to depict those variables. Descriptive statistics are presented as percentages or means and standard deviation. Means of normally distributed variables were compared using the student t-test and proportions using chi-square test. Skewed distribution were analyzed using appropriate non-parametric tests. Level of significance set at $P < 0.05$

**Results**

**Prevalence of Tetralogy of Fallot**

A total of 155 patients with echo diagnosis of TOF were documented between January 2007 and December 2014 with a total of 315,150 patients seen as out-patient and in-patient at the study centre during the study period hence the prevalence of TOF amongst the children who were seen in the hospital during the study period was 4.9 per 10,000. A total of 983 had congenital heart disease while 311 of the 983 patients with congenital heart disease had cyanotic congenital heart disease therefore, TOF accounted for 15.8% and 49.8% of congenital heart disease and cyanotic congenital heart disease respectively. Table 1 depicts the prevalence of TOF in the study subjects. Table 2 illustrates the prevalence of TOF in other countries.

**Clinical Presentation**

The male to female ratio was 1.7:1. The mean age of the children was 51.6 ± 52.5 (months). Table 3 describes the age groups of the patients. The most common indication for cardiac evaluation in the study subjects was cyanosis with a suspicion of a cyanotic congenital heart disease. 110 (71%) children were clinically cyanosed on presentation. Record of oxygen saturation was documented with a pulse oximeter in 89 subjects. 69 of the 89 (77.5%) of those with cyanotic congenital heart disease, 71% of our subjects presented with cyanosis. Pulse-oximeter is a vital tool in evaluation of patients with congenital heart disease, we were able to document cyanosis using pulse oximeter in up to 91% of the subjects who were cyanosed. Few patients, 5 (3.2%) presented with atypical presentation like gangrene of the right hand and forearm, stroke, cerebral abscess and congestive cardiac failure. Onset of clinical manifestation in patients with TOF depends on the severity of right ven-

| Table 1. Yearly Prevalence of TOF at the Study Center |
|---------------------------------|---------|---------|-----------|------------|------------|
| Year   | CHD (n) | CCHD (n) | TOF (n) | % of TOF in CHD | % of TOF in CCHD |
|--------|---------|---------|---------|-----------------|-----------------|
| 2007   | 87      | 22      | 13      | 14.9            | 59.1            |
| 2008   | 119     | 29      | 10      | 8.4             | 34.5            |
| 2009   | 90      | 23      | 18      | 20              | 86.9            |
| 2010   | 103     | 36      | 12      | 11.6            | 33.3            |
| 2011   | 153     | 48      | 27      | 17.6            | 56.3            |
| 2012   | 143     | 47      | 26      | 18.2            | 55.3            |
| 2013   | 180     | 65      | 27      | 33.7            | 41.5            |
| 2014   | 108     | 41      | 12      | 11.1            | 29.3            |
| Total  | 983     | 311     | 155     | 15.8            | 49.8            |

Abbreviations: CHD, congenital heart disease; CCHD, cyanotic congenital heart disease; TOF; tetralogy of Fallot.
tricular outflow tract obstruction. Patients may present in neonatal period when there is severe obstruction to right ventricular outflow. The youngest patient was a 2 days old female who was part of a conjoint twin with TOF and pulmonary atresia. Two-thirds of the patients were under 5 years at diagnosis and 60.4% of those were between 1 and 5 years. One-third of the subjects were more than 5 years at presentation. The mean age of the subjects at presentation was 51.6 ± 52.5 in months (4.3 ± 4.4 in years). This mean age is almost similar to that reported by Kennedy and Miller in Malawi. Other authors have presented earlier age at diagnosis of TOF. However diagnosis of TOF and other congenital heart diseases are usually made between 1-5 years of age, especially in resource poor countries. Possible reasons for the trend of late presentation and diagnosis includes difficulty in assessing specialized care, poverty and poor health seeking behaviour. Chromosomal anomalies have been documented in up to 25% of patients with TOF, the commonest being trisomies and 22q11.2 micro deletion syndromes. We documented in this study 11 (7.1%) patients with TOF and chromosomal anomalies. There were 7 (4.5%) patient with Down syndrome, 3 (1.9%) of Turners syndrome and 1 (0.65%) case of CATCH 22 syndrome. Risk of recurrence rate for TOF is 3%, in this study we had two cases of TOF from the same family, a younger and other sibling. Patients with TOF may have other associated cardiac defects. In this study, we documented the other associated cardiac lesions in our subjects with TOF. The most common was pulmonary atresia followed by atrial septal defect (ASD). All the patients with ASD had secundum ASD.

**Conclusion**

We report a prevalence of 4.9 per 10 000 children in this hospital base study. The prevalence of TOF in congenital heart disease is 15.8% and this was similar to other studies within Nigeria and the sub-region. There was a male predominance and most children presented within 1-5 years of age. Chromosomal abnormalities such as Down syndrome, Turners syndrome and CATCH 22 syndrome were documented in some subjects. Some of the subjects had atypical presentation. TOF is as common in Nigeria as other parts of the world, there is a need to established cardiac centers to salvage these children. Collaboration from developed countries will be helpful in this resource limited region.

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**Ethical Issues**

Ethical clearance was obtained from the ethical committee of the hospital.

**Competing Interests**

None to be declared.

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