Echocardiography Findings in Clinically Confirmed Congenital Rubella Syndrome Cases seen at the University of Port Harcourt Teaching Hospital, Nigeria

ABSTRACT
BACKGROUND: Congenital rubella syndrome (CRS) is known to affect thousands of children in the developing world because rubella vaccination is not available routinely in most of these countries. Among its many manifestations only congenital heart disease is life threatening. This study was undertaken to ascertain the cases of echocardiographic determined congenital heart disease in clinically confirmed CRS cases.

METHODS: Data of patients with clinically confirmed CRS seen over a period of 5 years in the Paediatric cardiology clinic of the University of Port Harcourt Teaching Hospital was retrieved and analysed.

RESULTS: Seven cases (2.8 % of total cardiac cases) were seen. 6 (85.7%) cases had at least one murmur on auscultation. Patent ductus arteriosus was the commonest cardiac defect seen either in isolation or in combination with a VSD or ASD. Only one child had no cardiac defect. 4 (57.1%) of them had been admitted for heart failure at least once and 2 (28.6 %) were on anti-failure regimen, one of whom had cardiac surgery one month ago.

CONCLUSION: In view of the fact that 6 (85.7%) of the patients with CRS had at least one congenital heart defect, we advocate routine echocardiography on patients with CRS to ensure early treatment and reduce mortality and morbidity. We also advocate that rubella vaccination be given routinely in developing countries. WAJM 2012; 31(2): 135–138.

Keywords: Congenital Rubella Syndrome, Echocardiography, Port Harcourt.
INTRODUCTION

The birth of a baby with congenital rubella syndrome is both a personal and a community tragedy. The Congenital rubella syndrome (CRS) is seen in children whose mothers are infected with rubella during pregnancy especially in the first trimester. It is estimated that between 100,000 and 238,000 children are born with CRS worldwide – most in the developing countries where incidence rates range from 0.4–4.3 per 1,000 live births. The World Health Organisation (WHO) developed definitions for diagnosing CRS in 1998. The clinical case definition of CRS is any defect(s) or laboratory data consistent with congenital rubella infection. The specific signs and symptoms include a) cataracts/congenital glaucoma, hearing impairment, congenital heart disease, b) microcephaly, hepatosplenomegaly, jaundice, developmental delay, meningencephalitis and radiolucent bone disease. Laboratory criteria for diagnosis are isolation of rubella virus or presence of IgM antibody. The case classification include Probable CRS which is a case that is not laboratory confirmed but has 2 complications in a) or 1 each from a) and b). Suspected CRS is one in which some compatible clinical findings are present but do not meet the criteria for a probable case and Confirmed case of CRS is a clinically consistent case which is laboratory confirmed.

Cardiovascular anomalies are reported in 45–50% of children with CRS. These are the most life threatening complications of CRS resulting in increased morbidity and mortality. They commonly are a patent ductus arteriosus (PDA), pulmonary artery or valvular stenosis, ventricular septal defects (VSD) and complex cardiac defects involving a PDA and any of these other cardiac lesions – atrial septal defects (ASD),pulmonary stenosis and VSD. There has been no previous report on echocardiographic findings of cardiovascular malformations in CRS in Nigeria, necessitating this study.

SUBJECTS, MATERIALS AND METHODS

Data of all cases of congenital heart disease seen at the paediatric cardiology unit of the University of Port Harcourt Teaching Hospital (UPTH) from January 2006 to November 2010 were retrieved and all cases of CRS (using the WHO criteria) seen were analysed. The UPTH is the main tertiary health care facility in Rivers State located in the Niger-Delta area of Nigeria and is a major referral centre in the region with catchment areas from neighboring states like Abia, Imo, Bayelsa, and Akwa-Ibom.

Information obtained from the case folders included the patient’s biodata, birth weight, history of a rash in pregnancy and prior maternal immunisation with rubella vaccines. Physical examination and Echocardiography were performed by the Consultant Pediatric Cardiologist.

All patients were assessed by the Ophthalmologists and Otorhinolaryngologists. The Echocardiography was done using a Sonosite Micromaxx machine.

A diagnosis of congenital rubella syndrome was made based on the case definitions by the W.H.O.

The data was then analyzed using Epi Info 6.

RESULTS

There were 250 cases of echocardiography-confirmed congenital heart defects seen in the Paediatric cardiology unit during the study period. Of these, six were clinically confirmed cases of congenital rubella syndrome and one was a clinically and laboratory confirmed case – this was a male with rubella IgM positive and rubella Elisa of 11.6 iu/ml which was highly elevated. The total of seven cases among the cardiac cases seen in the paediatric cardiology unit gives the incidence rate of CRS as 2.8%.

Of these, 3 were males (42.8%) and 4 females (57.1%) giving a male: female ratio of 0.75:1. Age ranged from 3 to 32 months with a median of 15 months.

None of the mothers had received rubella vaccination and none recollected any history of rash in their pregnancy. Two cases were low birth weight (28.6%) and one (14.3%) was normal weight, while the weights of the others were unknown by the mothers. All 7 cases had microcephaly and murmurs were heard in all. Two of the patients with cataract had cataract extraction within one year of life and could see with the aid of glasses as adequate sized intraocular lenses were unavailable. Table 1 shows the clinical features and cardiac anomaly present in each child.

The cardiovascular manifestations seen were solitary PDA in 3 (42.9%) cases (Fig 1), PDA with ASD in 1 (14.3%) and PDA with VSD in 2 cases (28.6%). One child had a structurally normal heart. Of the 7 cases, 4 had been on regular follow up at the clinic and one of them, an only child of couple after 14 years infertility had surgery for PDA/ASD closure in India a month ago. Two were lost to follow up and one of them is the child with a structurally normal heart. One patient died from a septicaemic illness in a private clinic.

Table 1: Clinical Features and Echo Findings of CRS Cases seen in UPTH

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MC, Microcephaly; MO, Microphthalmia; HSM, Hepatosplenomegaly; LBW, Low birth weight
Current knowledge puts 2005; 2009; Robertson SE, Featherstone DA, Gacic-Namaei MH, Ziaee M, Naseh N. Sadighi J, Eftekhar H, Mohammed K. Br J Ophthalmol 2006; 13: 135–41. Congenital Heart Disease 91: 2007; Indian J Pan Am J Public Health Congenital Rubella Syndrome 15: Lambert SR. Congenital rubella: the end 17 17 Congenital Kyaw-Zin-Thant, Win-Mar-O In Cardiology and Optometry Otaigbe BE, Brown T, Esu R. Confirmed congenital heart disease with either an ASD or a VSD. However, ventricular septal defect (VSD) was the commonest cardiac defect in a Myanmar study12 while more patients were found to have pulmonary stenosis in Ghana.13 A case of CRS with Ebstein’s anomaly has been reported.14 Other cardiac abnormalities, such as peripheral arterial stenosis, and myocardial abnormalities have been reported rarely. Valve abnormalities include valvular pulmonic stenosis, polyvalvar dysplasia, and valvular aortic stenosis. As noted in our study, there is a relative absence of conotruncal, atrioventricular-canal, laterality, and single-ventricle defects.17 The frequency of cardiac abnormalities in CRS (>50%) is sufficiently high to warrant a focused cardiac examination of all patients suspected of having CRS.15 This should include auscultation and palpation over the precordium and peripheral arteries, as well as a blood pressure measurement. The murmurs of peripheral pulmonic stenosis and patent ductus arteriosus are distinctive; however, they are not always diagnostic and can mask underlying cardiac pathology. If a murmur is present, a cardiology consultation with echocardiography is indicated.

An alternative approach is to use prospective baseline echo-cardiography for all patients suspected of having CRS. Patients with CRS and congenital heart defect are managed medically and surgically in the same manner as non-CRS patients.

Conclusion
The study shows that CRS exist in Nigeria and is easily diagnosed using the WHO case definitions. That 87.5% of the cases in this study have congenital heart disease which is the most life threatening feature with its associated increased morbidity and mortality makes us recommend routine echocardiography on all cases of CRS. We also advocate the introduction of rubella vaccine into the national immunization program for all children in Nigeria to avoid the unnecessary socio-economic, psychologic and physical burden associated with care of affected children.

REFERENCES


