



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

*Résultats de l'échocardiographie chez des cas de Syndrome de Rubéole Congénitale cliniquement confirmée à l'Hôpital Universitaire de Port Harcourt, Nigeria*

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#### 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.

#### RÉSUMÉ

**CONTEXTE:** Le Syndrome de Rubéole Congénitale (SRC) affecte généralement des milliers d'enfants dans le monde en développement du fait que la vaccination contre la rubéole n'est pas disponible dans la plus part de ces pays. Parmi ses nombreuses manifestations, seule la cardiopathie congénitale constitue une menace vitale. Cette étude a été réalisée pour évaluer les cas de maladie cardiaque congénitale mises en évidence par l'échocardiographie chez des cas de SRC cliniquement confirmés.

**METHODES:** Des données ont été collectées et analysées sur des patients suivis en cardiologie pédiatrique de l'Hôpital Universitaire de Port Harcourt présentant une SRC cliniquement confirmée sur une période de 5 ans.

**RESULTATS:** Sept cas (2.8 % des cardiopathies) ont été vues. Six cas (85.7%) avaient au moins un souffle à l'auscultation. La persistance du canal artériel était plus fréquemment observée de façon isolée ou associée à une communication inter ventriculaire (CIV) ou une communication inter auriculaire (CIA). Seul un enfant ne présentait pas d'anomalie cardiaque. Quatre (57.1%) d'entre eux étaient admis pour une insuffisance cardiaque au moins une fois et 2 patients (28.6 %) prenaient un traitement contre l'insuffisance cardiaque et l'un d'entre eux avait subi une chirurgie cardiaque datant d'un mois.

**CONCLUSION:** Compte tenu du fait que 6 des patients (85.7%) avec un SRC présentaient une malformation congénitale, nous plaidons en faveur d'une échocardiographie de routine chez les patients avec un SRC afin de donner un traitement précoce et ainsi réduire la morbidité et la mortalité. Nous plaidons aussi en faveur de la vaccination anti rubéole dans les pays en développement. *WAJM* 2012; 31(2): 135–138.

**Mots clés:** Syndrome de Rubéole Congénitale, Echocardiographie, Port Harcourt.

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Abbreviations:

**INTRODUCTION**

The birth of a baby with congenital rubella syndrome is both a personal and a community tragedy.<sup>1</sup> The Congenital rubella syndrome (CRS) is seen in children whose mothers are infected with rubella during pregnancy especially in the first trimester.<sup>2-5</sup> It is estimated that between 100,000 and 238,000 children are born with CRS worldwide – most in the developing countries<sup>2,6</sup> where incidence rates range from 0.4–4.3 per 1,000 live births.<sup>7</sup> The World Health Organisation (WHO) developed definitions for diagnosing CRS in 1998.<sup>8</sup> 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, meningoencephalitis 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.<sup>8</sup>

Cardiovascular anomalies are reported in 45–50% of children with CRS.<sup>7</sup> 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.<sup>8,9</sup> 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.<sup>8</sup>

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.<sup>10</sup> 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**

	MC	MO	HSM	LBW	Other	Defect		
1	Yes	Yes	–	–	Yes	Yes	Yes	PDA
2	Yes	Yes	–	–	–	–	Yes	PDA/VSD
3	Yes	–	–	–	–	–	Yes	NONE
4	Yes	Yes	–	–	–	–	Yes	PDA/VSD
5	Yes	–	Yes	–	Yes	–	Yes	PDA
6	Yes	–	Yes	Yes	–	–	Yes	PDA
7	Yes	Yes	Yes	Yes	Yes	–	Yes	PDA/ASD
<b>Total</b>	<b>7</b>	<b>4</b>	<b>3</b>	<b>2</b>	<b>3</b>	<b>1</b>	<b>7</b>	<b>6</b>

MC, Microcephaly; MO, Microphthalmia; HSM, Hepatosplenomegaly; LBW, Low birth weight



**Fig. 1: Shows an echo of a one-year old with a PDA.**

## DISCUSSION

Rubella is not a reportable disease in Nigeria and data of its epidemiology are extremely rare.<sup>10</sup> Most developing countries, including Nigeria do not have the Measles.mumps-rubella (MMR) vaccine as part of their National Programme of Immunization which implies that the incidence of CRS will be high in them compared to developing countries where it is given routinely. The defects caused by rubella infection are debilitating and irreversible. Of these, cardiac abnormalities – in particular, structural congenital heart defects are the classic and most life threatening of them. Over the almost 5 year period of review, only seven cases were detected from the Paediatric Cardiology clinic records accounting for 2.8% of children with congenital heart diseases referred for cardiac assessment and echocardiography. A previous retrospective study found that 30% of patients with CRS were identified from medical records in Cardiology and Ophthalmology clinics based on the finding of a cataract or congenital heart disease with either hearing impairment or mental retardation.<sup>11</sup> Congenital Heart Disease (CHD) was found in 85.7% of cases in this study, similar to a study done in Myanmar where 72% of CRS cases had a CHD.<sup>12</sup> Another review of prospectively ascertained CRS cases noted congenital heart defects in 40%–50%.<sup>10</sup> The commonest CHD in CRS as in our study is a PDA which may occur in isolation or with an ASD, VSD or pulmonary stenosis.<sup>5,6,9,12–15</sup> Current knowledge puts the incidence of PDAs in CRS patients

as ranging from 60–100%.<sup>8,12,13</sup> Similar observations were made in this study with 85.7% presenting with either isolated PDA or in combination with either an ASD or a VSD. However, ventricular septal defect (VSD) was the commonest cardiac defect in a Myanmar study<sup>12</sup> while more patients were found to have pulmonary stenosis in Ghana.<sup>15</sup> A case of CRS with Ebstein's anomaly has been reported.<sup>16</sup> Other cardiac abnormalities, such as peripheral arterial stenosis, and myocardial abnormalities have been reported rarely. Valve abnormalities include valvular pulmonic stenosis, polyvalvular dysplasia, and valvular aortic stenosis. As noted in our study, there is a relative absence of conotruncal, atrioventricular-canal, laterality, and single-ventricle defects.<sup>17</sup> The frequency of cardiac abnormalities in CRS (>50%) is sufficiently high to warrant a focused cardiac examination of all patients suspected of having CRS<sup>17</sup>. 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.

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