

CONGENITAL RUBELLA SYNDROME IN KORLE BU TEACHING HOSPITAL

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SUMMARY

Eight cases of Congenital Rubella Syndrome (CRS) were reported between 1st July and 30th September 1998 at the Department of Child Health, Korle Bu Teaching Hospital, Accra. All the cases were born at term, with six of them small for age. Five of the mothers were multiparous, while three were primiparous. All the cases had cataracts, and all were compatible CRS according to WHO guidelines for the diagnosis of CRS.

These eight cases of CRS recorded within three months in one hospital is alarming considering the fact that only 12 cases were reported in the United States of America in a three year period from 1994 to 1996. Further studies are needed to find the true burden of CRS in Ghana to ascertain whether preventive measures should be instituted.

Keywords: Congenital rubella syndrome, cataract, congenital heart disease, deafness.

INTRODUCTION

Rubella is a mild disease, characterized by a low grade fever, maculopapular rash and adenopathy. However, maternal illness during pregnancy may affect the fetus and lead to congenital rubella syndrome (CRS). This makes it a public health concern.

Maternal illness may not be apparent in at least fifty percent (50%) of cases. The risk of fetal infection is greatest in the first month of pregnancy even before the mother knows she is pregnant. This is within the period of organogenesis and the heart and eyes are maximally affected¹. According to Millar E. et al, the risk of fetal malformation detected by two years of age, was ninety percent (90%) of children exposed to the virus prior to eleven weeks gestation and thirty-three percent (33%) infected at eleven to twelve weeks gestation. No defects were detected among sixty-three (63) infants born to mothers infected after sixteen weeks gestations². However, a

study done in the USA by South MA and Sever JL found a small risk of CRS among infants born to mothers infected after sixteen weeks gestation³. CRS may be diagnosed by its classic triad of clinical signs: heart disease, cataract and deafness. However, many infants only have one of these manifestations. Hence the need to use the WHO guidelines⁴.

Confirmed CRS – Any one of the congenital defects listed below in addition to laboratory confirmation:

- a. Eye defects i.e. cataract/glaucoma
- b. Congenital heart disease
- c. Deafness: this is easily detected after 2 years and may be the only sign in up to 50% of affected children.
- d. Pigmentary retinopathy.

When laboratory data are not sufficient for confirmation, any two congenital defects listed above or any one congenital defect in addition to any one of the manifestations listed below:-

- Purpura, splenomegaly, microcephaly, mental retardation, jaundice in 24hours, radio lucent bone disease, meningo encephalitis

is adequate for diagnosis of compatible CRS.

Rubella causes severe disabilities in an estimated 300,000 babies every year worldwide⁵. Data on the incidence of CRS provide the most direct evidence of the burden of disease. Unfortunately, Ghana has no documented data on CRS⁶. This study was therefore undertaken to document cases of CRS, to demonstrate the need for a national surveillance system, with the ultimate aim of instituting preventive measures if necessary.

STUDY POPULATION AND METHODS

This was a descriptive study conducted over a period of three months. It was noted in the middle of 1998 that the incidence of infants with cataracts

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was unusually high. Therefore infants who presented to the Department of Child Health, Korle Bu Teaching Hospital, Accra with cataracts were studied. These cases were seen between 1st July and 30th September 1998. Blood samples were taken for rubella specific IgG, which is the only method available in Ghana. Patients with CRS were identified using the World Health Organisation (WHO) guidelines for the diagnosis of CRS. The antenatal history, birth weight, gestational age and clinical examination were documented.

RESULTS

Three thousand and fifty nine (3,059) new patients were registered at the OPD of the Department of Child Health, Korle Bu between 1st July and 30th September 1998. Out of these eight were classified as CRS.

or who live with deafness which we fail to associate with CRS.

The management of a pregnancy during which the mother is exposed to the rubella virus include laboratory testing for the Immunoglobulin G (IgG) antibodies in the mother. If it is reactive it means the mother has immunity already and the fetus is not at risk. IgG antibodies are permanent and protective. CRS after maternal reinfection is rare. If the test is not reactive, it should be repeated in three weeks time (since the incubation period of the infection is two to three weeks). A positive result means that new maternal infection has occurred and the fetus is at risk. In such a situation, the options available to the mother are either to terminate the pregnancy or continue with the pregnancy despite the risk of CRS. If the repeat test is non-reactive at three

Table 1 Congenital rubella syndrome case reports – July – September 1998

No	Age/sex	Maturity	Maternal illness in pregnancy	Weight (Kg)	Heart (Clinical Diagnosis)	Cataracts	Other Eye Findings	Minor Criteria	Laboratory Immunoglobulin (IgG) Mother	Child	WHO Guideline
1	DOD M	Term	Febrile illness at two to three months	1.95	PDA/VSD	Bilateral	-	-	Reactive	Reactive	Compatible CRS
2	DOD M	Term	Itchy rashes at two months	2.8	Pulmonary stenosis Tricuspid Regurgitation (ECHO)	Bilateral	-	-	Reactive	Reactive	Compatible CRS
3	One week F	Term	'Flu' at two months	2.2	VSD	Bilateral	-	-	Reactive	Reactive	Compatible CRS
4	Two weeks M	Term	Febrile illness at seven months	2.8	PDA	Bilateral	-	-	Reactive	Reactive	Compatible CRS
5	DOD M	Term	Fever at nine months	1.9	PDA/VSD	Left Eye	Bilateral Corneal Edema	-	Reactive	Reactive	Compatible CRS
6	Three months F	Term	'Measles' during first trimester	2.78 <3 rd Centile	PDA/VSD	Right Eye	Bilateral Microphthalmia	-	Reactive	Reactive	Compatible CRS
7	DOD M	Term	-	1.6	VSD	Bilateral	Keratitis Right Eye	-	Reactive	Reactive	Compatible CRS
8	Three months F	Term	'Measles' at three months	3.48 <3 rd Centile	Sub valvular pul Stenosis (ECHO)	Left Eye	-	Microcephaly	Reactive	Reactive	Compatible CRS

Abbreviations

VSD	-	Ventricular Septal Defect	ECHO	-	Echo Cardiography
PDA	-	Patent Ductus Arteriosus	DOD	-	Day of Delivery

All the cases were born at term. Seven mothers gave a history of febrile illness during pregnancy. Two of the cases had their heart diseases confirmed by echocardiogram. Table 1 summarises the findings based on the WHO guidelines for diagnosis of CRS. Since our patients were all below 6 months of age and we could only measure IgG antibodies our cases were all classed as compatible CRS.

DISCUSSION

The commonest clinical presentations of CRS in our patients were cataracts and congenital heart disease. This is not surprising since these clinical problem are more easily detected than some of the other features such as deafness. These eight patients therefore represent the tip of an iceberg of a larger group of children in our society who either die in utero or within the first month from severe disease

weeks, it should be repeated at six weeks post exposure. If it is still non-reactive it means no infection occurred and fetus is not at risk.

The burden of CRS is enormous. Thus the best option is to prevent it occurring. The could be accomplished bearing in mind that in any rubella control strategy, it is essential to protect women of child bearing age. Rubella vaccination is very effective because there is only one viral strain. In clinical trials, some studies have shown 99-100% seroconversion after a single dose of rubella vaccine⁷, and some studies found no difference in seroconversion when the vaccine is given at nine months or fifteen months of age⁸. Clinical efficacy and challenge studies indicate that more than ninety percent of vaccinees are protected against both clinical rubella and viremia for at least fifteen years⁹, and vaccine-

induced protection is generally assumed to be life-long.

Registers were kept in Sweden, United Kingdom, Germany and USA of women who were seronegative and who inadvertently received rubella vaccine within three months of conception and continued with their pregnancy¹⁰. None of the 515 infants had anomalies compatible with CRS, thus the observed risk of vaccine – associated CRS was zero. Screening women of child bearing age before vaccination is an option which could be considered. However the cost of screening should be balanced against the higher coverage achievable without serological screening.

Cases of CRS may be reduced with the following strategies:

1. Selective vaccination i.e. direct protection to women and/or school girls.
2. Childhood vaccination i.e. as part of Expanded Programme of Immunization.
3. Combination of the two strategies above.

However in order to prevent Congenital Rubella Syndrome (CRS), vaccination coverage has to be high, otherwise the pool of women who have naturally acquired antibodies from childhood rubella, may reduce as they are less likely to come into contact with rubella. If vaccination coverage is inadequate the number of susceptible women can actually increase. It is therefore essential to carry out a national survey to determine the prevalence of seronegativity among women of child bearing age.

The eight cases of CRS recorded within a three month period is alarming since only 12 cases were recorded in the whole of the United States of America in a three year period from 1994 to 1996⁵. The study was limited to infants with cataracts. Interestingly the author has since the study recorded two cases of compatible CRS in children aged more than one year, both of whom presented with Congenital Heart Diseases and Deafness, but without cataracts. There is therefore the need for collaborative work involving Paediatricians, ENT Specialist and Ophthalmologists to find the true burden of CRS. This may draw the appropriate authorities attention, to ascertain whether preventive measures should be instituted.

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