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THE ANALYSIS OF CLINICAL AND SOCIAL PROFILE OF CONGENITAL RUBELLA SYNDROME SEEN AMONG UP-PGH PATIENTS FROM THE YEARS 1993 - 2002 (A 10 YEAR PREVALENCE REVIEW)

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Objectives :

1. To describe the socio-demographic profile of patients with Congenital Rubella Syndrome (CRS) seen in UP-PGH for the last 10 years.
2. To compare the clinical profile of patients with Congenital Rubella Syndrome (CRS) seen in UP-PGH to that of foreign published data.
3. To describe the frequency distribution of occurrence of clinical findings of patients with Congenital Rubella Syndrome (CRS).
4. To describe maternal factors such as antenatal maternal viral infection and MMR vaccination history among siblings with Congenital Rubella Syndrome (CRS).

Design: A descriptive study design.

Methodology: There were total of 58 cases of Congenital Rubella Syndrome (CRS) classified and retrieved in the Biostatistics and Epidemiology Section of the UP-PGH, OPD Division. The 58 case numbers of identified diagnosis in the charts were tracked down and analyzed if they fit in to the inclusion criteria of the study.

Results: In the wide spectrum of clinical manifestations of Congenital Rubella Syndrome (CRS), the youngest patient brought in for consult is within the1st month with the mean age of 8 months, with slight male preponderance.

Baseline anthropometrics showed 30% of infants classified as severely wasted although 20% of study population had normal weight . Majority (69%) had no stunting. Congenital catarract ranked the most common clinical presentation of patients with CRS, accounting for 24 cases (49%) followed by patent ductus arteriosus of 15 cases (31%). Most clinical findings that co-exists were: ocular, cranial, growth retardation, cardiac and sensorineural findngs. There is a high occurrence of antenatal maternal viral infection in this review – accounting to about 37 cases (71%) of study population.

Conclusions: This research study documented the continued existence of CRS and majority of the patients were delivered primarily by an unvaccinated women. The disease continued to be dynamically prevalent despite the illness is a vaccine preventable occurrence.

INTRODUCTION:

Congenital Rubella Syndrome is a serious multi-systemic disease with a wide spectrum of clinical expression and sequelae. This is comprised of constellation of birth defects which may occur when the mother acquires the infection during pregnancy , especially on her 1st trimester . In Congenital Rubella Syndrome (CRS) virtually every organ system maybe involved , singly , multiply , transiently or progressively and permanently.

The risk of congenital defects of the disease is greatest with the primary maternal infection during the first trimester which may lead to abortion , stillbirth,, or a child with anatomic defects. The risk is about 90% if the infection occurs before 11th week of pregnancy, and this decreases by 10-20% at the end of the 1st trimester, with an overall risk of 70% and maternal infection after the 16th week of pregnancy poses low risk of congenital defects although fetal infection may still occur.

From the records of Philippine General Hospital from the years 1993-2002, there are still 56 cases of congenital rubella syndrome (CRS) despite the introduction of MMR vaccination by our DOH since 1983. What are the factors that may have contributed to the Congenital Rubella Syndrome profile that is still prevalent in our community at present times?

In the study of Elias, Erik on Rubella (August 2002) states that after licensing the live attenuated vaccine in the US in 1969, a dropped has occurred in the number of cases of Rubella and Congenital Rubella Syndrome (CRS). As shown in the statistics of 1969 (from the total of 57,686 cases of rubella and 62 cases of CRS). From 1992-1998, the cases of rubella

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recorded annually decreased by the range of 128-364 cases and CRS decreased by 2-9 cases per year, although 15 cases of isolated outbreaks were still reported. The median cases per outbreak was 21 and most recent cases occurred in New York (1997-1998), Kansas(1998) and Nebraska (1999).

Likewise , CRS continuous to occur despite 25 years of immunization program in Canada and still requires further measures to eliminate this preventable disease. Between the years 1986-1995, a mean of 3 cases of congenital rubella syndrome (CRS) per year were reported to the Notifiable Diseases Reporting System in Canada. (Infectious Disease and Immunization Committee; Canadian Pediatric Society).

In Kuala Lumpur, Malaysiathe incidence of CRS from 1993 - 1998 varies from 19-93 per 100,000 deliveries.[13]

In Canada, the epidemiologic pattern of rubella underscored the importance of the collection and analysis of information on demographic, vaccination history, source of infection in relation to transmission and outbreaks.[2]

In a local study done, one of the variables considered was the educational attainment of parents or guardians on how they perceive the disease entity itself and the benefits of vaccination.[17]

In a GENEVA conference , held November 28, 2001, the UNICEF (United Nations Children's Fund) hailed the massive immunization of individual between 15 months to 25 years old , to significantly reduce the cause of birth defects in children which is now estimated to be about 110, 000 cases of CRS per year worldwide.

The intent of this paper is to have a wide panoramic view of the demographic and psychosocial profile of Congenital Rubella Syndrome among Filipino children to guide us in early diagnosis and subsequently pave way for the policy makers to formulate a timely medical intervention and prevention programs to abort further spread of virus and hinder specific disabilities for progressive deterioration.

GENERAL OBJECTIVE

To describe the clinical and social profile of patients with Congenital Rubella Syndrome (CRS) seen in UP-PGH from the years 1993-2002.

SPECIFIC OBJECTIVES:

- To describe the socio-demographic profile of patients with Congenital Rubella Syndrome (CRS) seen in UP-PGH for the last 10 years .
- To compare the clinical profile of patients in terms of type and number of clinical signs of Congenital Rubella Syndrome (CRS) seen in UP-PGH versus foreign published data.
- To describe the frequency distribution of occurrence of clinical findings of patients with CRS seen in UP-PGH.
- To describe maternal factors such as frequency of antenatal maternal viral infection and MMR vaccination history among siblings with Congenital Rubella Syndrome.

METHODOLOGY

Study design : Descriptive study design

Sample size/Population:

Population considered are all charity pediatric patients seen and/or admitted at Pedia ER, OPD, Wards 9 and 11, PICU, and NICU of UP-Philippine General Hospital from years 1993-2002. Excluded in the study were those admitted at paywards and other departments who were not referred nor co-managed by pediatric residents.

Inclusion criteria:

1. patients aged 1 month to 18 years old.
2. patients consulted and / or admitted at UP-PGH: PER, OPD, Wards 9 and 11 and special areas like PICU and NICU.
3. patients admitted in other wards but seen or co-managed by pediatric residents.
4. patients whose charts met the clinical case definition and classification of Congenital Rubella Syndrome (CRS) in APPENDIX E and with pertinent data needed in the study.

Exclusion criteria:

1. patients seen at paywards. or other departments not referred or co-managed by pediatric residents.
2. patients more than 18 years old at the time of consult.

Actual Methodology:

There was total of 58 cases of Congenital Rubella Syndrome (CRS) classified and retrieved in the Biostatistics and Epidemiology Section of the Record Division, 5th floor OPD, UP-PGH which were tracked down and analyzed if they fit in to the inclusion criteria.

PIDSP Journal, 2005 Vol. 9 No. 2

However, only a total of 52 charts were reviewed. The rest were missing due to the typhoon that has embarked Metro Manila in the past years destroying considerable amount of old documents in the years 1993-1994 in the attic of the record section.

STATISTICAL ANALYSIS

In this research study , the test of 2 proportion was used and the frequency distribution for descriptive data was likewise adapted .

RESULTS and ANALYSIS

Eight-year incidence of CRS cases and sociodemographic profile

A total of 52 patient charts from 1995 to 2002 were reviewed. The frequency distribution of congenital rubella syndrome cases from years 1993-2002 is summarized as figure 1-1. A gradual increasing trend was seen in 1998 up to the last two previous years.

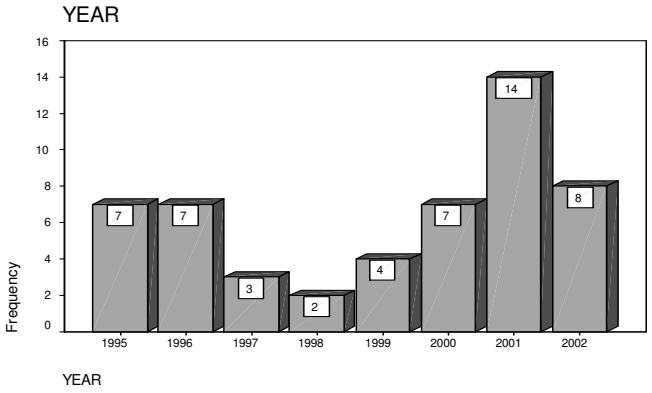

Figure 1. Frequency distribution of confirmed and probable CRS cases from 1993-2002, UP-PGH Data

The socio-demographic characteristics are summarized in Table 1. The mean age was 8 months. The youngest patient on the time of consultation was 1 month old while the oldest was 60 months old. There was a slight male preponderance 29 (56%) as opposed to females. Mean weight on consult was 5 kg while the mean height was 65 cm. Thirty percent (30%) of these children are classified as severely wasted while 20 % had normal weights. Majority (69%) had no stunting.

Much of our registered patients come from the greater Manila and Southern Tagalog area (65% & 30% respectively), There were 9 patients identified living in Paranaque City, 6 patients in Las Pinas City, while Mandaluyong, Taguig and Quezon City had 4 each, and Pasay, Caloocan, Makati and Metro Manila had 1 patient each. Majority of the parents of congenital rubllea syndrome (CRS) finished high school (26%), as compared to those who finished collegiate level (19%), and no formal education (13%).

Antenatal maternal infection and vaccination

The number of women who had a history of rubella (confirmed by history alone) is summarized in Figure 2. There is a high occurrence (37 out of 52) of antenatal maternal rubella in this review, while 12 mother's charts did not indicate the presence or absence of rubella exposure. Two siblings underwent MMR vaccination and this was seen in the years 2001 and 2002 respectively.

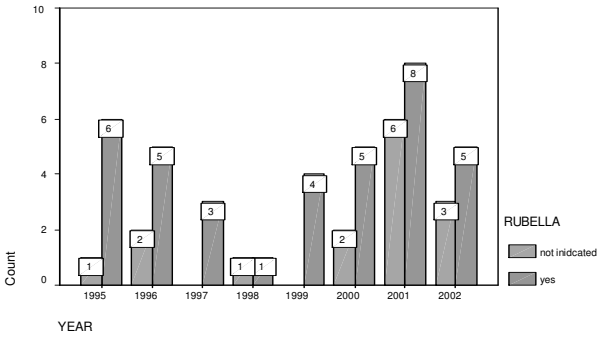

Figure-2. Antenatal history of maternal rubella in CRS cases from 1995-2002 , UP-PGH Data

Table-1. Socio-demographic characteristics of patients with Congenital Rubella Syndrome seen from 1995-2002 , UP-PGH.

UP-PGH Characteristics	Patients with CRS N= 52	Percentage %
Age in months (mean ± SD)	8 ± 10	
Sex		
Males	29	56
Females	23	44
Baseline Anthropometrics		
Length (mean ± SD)	65.3 ± 16 cm	
Weight (mean + SD)	5.7 + 3 kg	
Waterlow Classification		
Wasting		
Normal	15	28.8
Mild	11	21.2
Moderate	10	19.2
Severe	16	30.8
Stunting		
Normal	36	69.2
Mild	5	9.6
Moderate	7	13.5
Severe	4	7.7
Geographic Distribution		
Central Luzon	1	19
Southern Tagalog	16	30.7
Western Visayas	1	1.9
NCR*	34	65.3
Educational Background of Parents•		
No formal education•	7	13.5
Elementary•	6	11.5
High school•	14	26.9
College•	10	19.2
Vocational•	2	3.8
Not stated	13	25

**National Capital Region*

Clinical Profile of Congenital Rubella Syndrome (CRS) cases

The common clinical findings of CRS is tabulated in table-2. Only 5 clinical findings were mentioned in the 52 charts, namely cataract, PDA, hepatitis, jaundice, pulmonary artery stenosis, extrauterine growth retardation, glaucoma and hemolytic anemia. Congenital cataract ranked as the most common syndrome finding accounting for 24 (49%) of cases, followed by PDA in 15 cases (31%). Barring the differences survey design and geography, we roughly compared each proportion of findings with that of an 11- year US- based epidemiologic data previously published by Schluter et.al. Among all the mentioned CRS findings, PDA, hepatic derangement and pulmonary artery stenosis was statistically significantly higher in the US. This could be explained by the relatively early detection of abnormalities. *(p-value <.05).* Other socio-economic variables, such as cost for diagnostic work-up , short hospital stay and even parent preference may possibly explain the low sign/symptom detection rate of the above 3 findings in our setting.

Table 2. Clinical Findings of CRS

Clinical Findings (ranked)*	UP-PGH *N=122(%)	Foreign Census	*p*-value
Cataract	23(44)	55(45.1)	.628
Patent Ductus Arteriosus (PDA)	16(31)	62(50.8)	*.22^*
Hepatomegaly/ Hepatitis	5(10)	43(35.2)	*.0009^*
Jaundice	5(10)	18(14.8)	.3955
Pulmonary Artery Stenosis	3(6)	22(18)	*.04^*
Extrauterine Growth Retardation	2(4)	15(12.3)	.94
Glaucoma	1(2)	4(3.3)	
Hemolytic Anemia	1(2)	rare	NA***

**Only 5 clinical signs were identified in this study. Several signs co-exist*
***Data from the Schluter et.al., Changing Epidemiology of CRS in the US,1985-1996*
^ p-value significant at <.05
****Not applicable*

Syndrome Profile of CRS

In UP-PGH, we identified the frequency of combinations of organ systems affected. (See figure-3). From the years 1995 and onwards, the presence of cataract was a consistent syndrome finding. Cardiac findings, PDA, PAS and ASD were not that common. Jaundice and hepatosplenomegaly had been shown to be consistent, although less common.

The combinations of findings were not accurately determined in this review due to several reasons. Each patient came in at different periods of the follow-up, the presenting symptoms might have not incited a high index of suspicion so as to subject the patient to intensive work-up. So far, in this study , ocular plus cranial plus growth retardation plus sensorinueral findings and cardiac findings is still the most common combination.

Figure-3 Syndrome Profile of Congenital Rubella from 1995-2002, UP-PGH Data

Year	Findings n=52	Frequency
1995	Cataract alone	4
	PDA	4
	Microcephaly	2
	hepatitis/jaundice	2
	hepatosplenomegaly	2
	generalized lymphadenopathy	2
1996	Cataract	4
	EUGR	1
	PDA	3
	PAS	2
	VSD	1
	Microcephaly	3
	TTP	2
	Blueberry Muffin Syndrome	1
	Hepatitis/Jaundice	2
1997	Cataract	2
	EUGR	1
	PDA	2
1998	Cataract	1
	EUGR	1
	Hepatitis/Jaundice	1
1999	Cataract	3
	EUGR	2
	Sensorineural deafness	1
	PDA	1
	PAS	1
	ASD	1
	Microcephaly	1
	Mental retardation	1
	Hepatosplenomegaly	1
2000	Cataract	7
	Microphthalmia	1
	PDA	2
	Microcephaly	4
	Full anterior fontanelle	1
	Blue Berry muffin syndrome	1
	Thombocytopenic purpura	1
2001	Cataract	10
	sensorineural deafness	1
	PDA	6
	PAS	4
	microcephaly	1
	meningoencephalitis	1
	Hepatitis/ jaundice	1
2002	Cataract	5
	sensorineural deafness	1
	PDA	3
	PAS	2

DISCUSSION :

Since the introduction of MMR (measles, mumps, rubella) vaccine in the Philippines since the year 1983, there remains sporadic cases of CRS that occurs in the country and eradication of the disease seemed to be an elusive dream.

In this study there is not much appreciable difference in the infection rates by sex distribution, a difference of 4 cases (8%) of slight male preponderance was noted. Only for the fact that most of the disease was primarily noted during "infancy period" where clinical features are clearly seen if not without complications, except for 3 isolated cases where patients presented beyond 3 years of life and were noted to reside outside Greater Manila Area (GMA), and sought consult at UP-PGH, for definitive surgical correction of either congenital catarract or cardiac anomaly.

The bulk of 34 cases (65%) out of 52 population were sporadically distributed along the metropolis and 16 cases (32%) in region IV and approximately (2%) in each of regions III and VI. A large number of cases were seen in Metro Manila where subjects are residing in the nearby vicinity of the studied institution (UP-PGH) and could avail the charity rates of medical consult and could take advantage of the laboratories and medical manpower the hospital could offer.

There is poor correlation of educational attainment of parents to having a child infected with congenital rubella syndrome (CRS). The study reflects that the disease affects equally different people of varying degree of literacy and levels of social strata, which should alert the public health officials and mass media to formulate infection control programs and strategies to heightened public awareness of the spread of infection since the disease targets the entire population.

In the study of Schluter in the most frequent clinical presentation of CRS is the sensorineural deafness which is about 80-90% of the total population followed by congenital catarract or glaucoma which is about 35% and cardiac anomaly of roughly 30%.[18]

In our institution (UP-PGH) most of our CRS cases sought consult due to ophthalmologic problem; about 23 cases (44%) of the study group population. Opacification of the lens from catarract are more common (22 (42%) than glaucoma 1(2%)) followed by congenital heart defects of 19 cases (36%) may which patent ductus arteriosus to be more common than pulmonary stenosis accounting for 16 cases (84%) and 3 cases (16%) respectively.

According to written by Baja-Panlilio, et all. The table of birth attendance , since 1990 documented that most of our mothers; (40%) delivered at home and attended by traditional birth attendant, and (30%) by midwives, (1%) by nurse or others and only (27%) by physician.[19]

And in most cases of non-institutionalized delivery of high risks mothers , there is a missed chance of the infant being evaluated by a trained health care providers and corollary be worked up and be seen by tertiary hospital subspecialties.

So, it follows that our congenital rubella syndrome cases would present at OPD/PER with the chief complaint of opacification of the eyes noted during infancy or in blatant cardiac failure which is probably due to inadequate anticipatory guidance from our traditional birth attendants (TBA) and/or midwives and which resulted to less preparatory movement on the part of the parents.

In rich countries, where most mothers delivered in a hospital, newborns have the benefit of being seen by a pediatrician or neonatologist prior to discharge and would present at ambulatory clinics later life for either a child with neurodevelopmental delay and/or for definitive management of hearing impairment.

CONCLUSION:

This research study documented the continued existence of CRS and majority of these born to mothers, who had history of antenatal maternal viral infection during pregnancy accounting for 37 cases (71%) of the 52 study population . The disease continued to be dynamically prevalent despite the illness is a vaccine preventable occurrence .

It is therefore recommended for public health officials to have high index of suspicion to the wide spectrum of clinical manifestations of CRS and subsequently send suspended infants for laboratory work-ups, thereby improving the country's surveillance system and in turn improve the Filipinos' quality of future generation.

RECOMMENDATIONS:

Serologic testing of unvaccinated person for rubella immunity before vaccination probably is not necessary because this would only postpone the opportunity for the vaccine to be administered.

Unless contraindicated, it is suggested that vaccine be offered to: primary health care clinics, STD clinics, travel clinics postpartum clinics, health care personnel, and to all unvaccinated person who lack the evidence of rubella immunity especially in child-bearing age and foreign born individuals.

The state health department along with the mass media should formulate programs and strategies to heighten public awareness and consequently educate its citizenry to bring patients to hospital for diagnostics and early intervention of the impact of associated disabilities and advice significant others on ways and means to deter the further shedding of virus in the community.

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