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INCIDENCE OF RUBELLA IN PATIENTS PRESENTING WITH CONGENITAL OCULAR DEFECTS

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ABSTRACT

Objectives: To find out the incidence of rubella in children with congenital ocular defects. **Design of Study:** Co relational, quantitative & prospective study. **Settings:** Department of Ophthalmology, Allied Hospital, Faisalabad in collaboration with Departments of Pediatrics & obstetrics **Period:** June 1999 to July 2001. **Materials & Methods:** Total 100 children of five year or lesser age who presented or referred with some ocular problems of congenital origin were studied. The data was collected on special proforma. The relevant investigations were carried out and the correlation between rubella and ocular / systemic defects of congenital origin were studied. **Results:** The incidence of Congenital Rubella in children with congenital ocular defect was 16% while incidence of Rubella in various individual congenital ocular defect was Cataract 19.1%(13in 68), Glaucoma 11.5%(6 in 52), Retinopathy 42.3% (11 in 26), Microphthalmia 25% (4 in 16),Corneal Haze 11.3% (5 in 44), Microcornea 17.14% (6 in 35), Buphthalmos 8.82% (3 in 34), Corneal Scarring 10.52% (4 in 38), Squint 16.6% (4 in 24) and Keratoconus 100% (2 in 2). The associated systemic lesions in the patients of Congenital Rubella Syndrome included CNS lesions 28.75%, Cardiovascular abnormalities 25% and Biliary tract abnormalities 6.2% (one patient). **Conclusion:** The Congenital Rubella Syndrome (CRS), a preventable condition, presents with potentially blinding & visually handicapping problems and its incidence in children with congenital ocular defects in Pakistan is still very high i.e. 16% therefore responsible for a considerable visual morbidity in Pakistan whereas in most of developed countries, it has been controlled by rubella vaccination of mothers in routine.

KEY WORDS:- Congenital Rubella Syndrome (CRS), Microphthalmos, Cataract, Glaucoma, Pigmentary Retinopathy.

INTRODUCTION

The congenital rubella syndrome (CRS) is considered to be a major cause of the congenital ocular defects which poses a great challenge to the ophthalmologists in addition to being of a grave concern for the parents of the affected child. This is a preventable condition and has been almost eliminated or at the verge of elimination in the developed countries with the organized efforts. No such strategy has been adopted in Pakistan partially due to our limited resources and mostly due to lack of knowledge about the gravity of problem.

The German authors first of all recognized the rubella commonly known as German measles in the late eighteenth century. The Rubella was not considered to be of any clinical importance until 1941, when Australian Ophthalmologist Norman McAllister Gregg for the first time observed the association of Rubella with the congenital ocular defects during the follow up period of an epidemic of Rubella in Australia. Later on its strong association with the other congenital defects like congenital heart disease, deafness and many other systemic defects was established[1,2,3]. These defects secondary to rubella were termed CRS. The frequency of

their occurrence were found to be related to the gestational age at the time of maternal rubella infection[4].

Rubella is caused by Rubivirus, which is an RNA virus of Togavirus Family[5,6] and has only one serotype. It is a moderately contagious disease. The congenital infection spreads to fetus from infected mother through transplacental route while postnatal infection spreads as droplet infection from acute infection sufferer or a child with CRS who is shedding viruses. IgG is produced one week after the rash, positive for life, elicits a booster response and so imparts a life long immunity while IgM is produced one week after rash, disappears within one year, does not elicit a booster response and so a specific parameter to detect primary rubella infection. The placenta can transfer virus to infect the fetus but can not transfer maternal IgM and hence IgM antibodies in the new born infant are totally of fetal origin and are strong parameter to judge the congenital Rubella infection in first year of life[5].

The rubella affected fetal tissues show Hypoplasia, necrotizing angiopathy, tissue necrosis and inflammatory changes. If the infection occurs in the period of organogenesis, this results in either transient manifestations like hepatospleenomegaly, Jaundice, Thromocytopenia, cloudy cornea etc or permanent manifestations like cataract, Microphthalmia, Microcornea, Glaucoma, Retinopathy, Squint, optic nerve lesions, Iris Hypoplasia, Nystagmus, Uveitis, Lid defects, Cardiac defects, CNS abnormalities and delayed onset manifestations like keratoconus and subretinal neovascularization.

MATERIAL & METHODS

A total of 100 children who presented at Eye OPD and referred from Paediatrics and Obstetrics Departments, Allied Hospital, PMC, Faisalabad were examined. The children with 1 day to five years of age presenting with congenital ocular defects were included. Those not included in the study were the children who were more than five years of age or have undergone vaccination for rubella or they have suffered from postnatal rubella. The children were admitted in Ophthalmology Department and detailed history of ocular problems of congenital origin in children was taken. The age, parity, vaccination status of the mother and rubella like illness during the pregnancy with special relation to the gestational age were recorded. The general, physical and system examinations i.e. CVS and CNS were done in the department and also by the paediatrician.

Routine Laboratory investigations and "TORCH" test was done. Rubella specific IgG & IgM titer assessment was made to confirm the diagnosis. Visual acuity was checked with preferential looking, persuit movements, picture charts, E-chart and snellen chart while papillary reaction, Ocular motility and squint assessment was made. The children were examined either under general anaesthesia or under sedation for examination of ocular problems and or with slit lamp in cooperative children.

The cornea was examined for edema, haze, size, haabs stria, and scarring, keratoconus was tested with slit lamp and photokeratoscope. Anterior chamber, Iris was examined for AC reaction and Hypoplasia. Intraocular pressure was measured with Perken's hand held applanation tonometer. Gonioscopy was done where possible. The lens, vitreous and fundi were examined for cataract, optic nerve lesions and retinopathy using Microscope, direct and indirect Ophthalmoscope, 90 D Volks and Goldman lens. The cataract and glaucoma were managed with surgery while other lesions were treated according to their own merit.

RESULTS

Out of a total of hundred children of one day to 5 years presented with ocular defects of congenital origin, the ocular problems found were cataracts (68 patients), Glaucoma (52), Corneal edema or haze (44), Corneal scarring (38), Microcornea (35), Buphthalmos (34), squint (24), Retinopathy (26), Microphthalmia (16) and Keratoconus two patients. (Table-I).

Table-I. Incidence of various ocular defects (n=100)

Ocular condition	Total Cases	Incidence (%)
Cataract	68	68.0
Glaucoma	52	52.0
Corneal Haze	44	44.00
Corneal Scarring	38	38.00
Microcornea	35	35.00
Buphthalmos	34	34.00
Squint	24	24.00
Retinopathy	26	26.00
Microphthalmia	16	16.00
Keratoconus	2	2.00

Out of these hundred patients, 16 children were detected to be suffering from Rubella thereby making a gross incidence of rubella in ocular congenital defects to be 16%. Out of these Rubella positive children were 4 female and 4 male. At the time of conception, the mothers age ranged from 25 to 38 years.

Table-II. Incidence of Rubella in individual congenital ocular defects

Ocular condition	T. Cases	+ for Rubella	Incidence of Rubella (%)
Any congenital ocular defect	100	16	16
Cataract	68	13	19.1
Glaucoma	52	6	11.5
Corneal Haze	45	5	11.36
Corneal Scarring	38	4	10.52
Microcornea	35	6	17.14
Buphthalmos	34	3	8.82
Squint	24	4	16.66
Retinopathy	26	11	42.3
Microphthalmia	16	1	25

Table-III. Incidence of Ocular and systemic defects in 16 cases of CRS

Ocular/systemic condition	T. Cases	Incidence (%)
Cataract	13	81.25
Retinopathy	11	68.75
Microcornea	6	37.50
Glaucoma	6	37.50
Corneal Haze	5	31.25
Squint	4	25
Microphthalmia	4	25
Corneal Scarring	4	25
Buphthalmos	3	18.75
Karatoconus	2	12.5
CNS lesions	3	18.75
CVS lesions	4	25
Biliary system defects	1	6.2

Table-IV. Associated ocular and systemic lesions in 13 cases of congenital cataract due to congenital rubella syndrome

Ocular/systemic condition	T. Cases	Degree of Association (%)
Retinopathy	8	61.53
Microcornea	6	46.15
Microphthalmia	4	30.76
Corneal Haze	4	30.76
Glaucoma	3	23.07
Corneal Scarring	3	23.07
Squint	3	23.07
Keratoconus	2	15.3
CNS lesions	2	15.3
CVS Lesions	2	15.3

None of the mother had been vaccinated against rubella. Four mothers gave vague history of suffering from some

sort of skin lesions associated with feverish feeling during early pregnancy. Three mothers had no significant history of any ailment while one suffered from skin rashes only. None of the mothers whose children were included in the study was vaccinated for rubella at any stage of the life.

The incidence of Rubella in the individual ocular lesions in this study is shown is Table-11 while the order of frequency of ocular defects in CRS patients was tabulated in Table-lll. Since the cataract and pigmentary retinopathy were among the commenst findings in patients of CRS, their associated ocular / systemic congenital defects have been tabulated according to the degree of association in Table-IV & V.

Table-V. Various ocular and systemic defects associated with Prigmentary Retinopathy in congenital Rubella Syndrome

Ocular/Systemic condition	T. Cases	Degree of Association (%)
Cataract	11	100
Microphthalmia	3	27.27
Microcornea	3	27.27
Squint	3	27.27
CNS lesions	1	9
CVS lesions	1	9

DISCUSSION

It was observed during the course of study that among those children who presented with any congenital ocular defect regardless of the cause, the commonest ocular defect was cataract i.e. 68%. Moreover, it is the commenst finding amongst the congenital rubella syndrome children i.e. 81.25%. In the study of Ekstein et al, the incidence of cataract in infants affected with congenital rubella is 15%[7] which is probably due to the restriction of our cases to those who presented with some ocular / visual symptoms to eye OPD while excluding all others who either did not present to OPD or did not feel severe symptoms. The incidence of positive rubella in infants with congenital cataract was 19.1% while Tarin Shoaib Ahmad found the incidence to be 28.13% in a study at Mayo Hospital Lahore[8]. This is due to the age limit of 5 years in our study while no limit in Tarin SA cases indicating that the congenital cataracts, being a progressive lesion may become symptomatic at above 5years age and hence these children report to eye OPD which results in higher incidence. However, the rubella positive congenital cataract was 54% bilateral (7 out of 13) which is close to Jack S. Remington[4].

Jack J Kanski mentioned pigmentary retinopathy instead of cataract to be the commonest in CRS[4] while in our study, it is the second commonest lesion in'order of frequency in CRS. (Table-3). However, the incidence of rubella in infants of congenital pigmentary retinopathy is the highest i.e. 42.6% as shown in Table-2. The findings in pigmentary retinopathy of positive rubella infants was characteristically salt and pepper appearance mainly involving the posterior pole and obscuring the foveal reflex which is similar to that mentioned by Shelman J. Kieval[9].

Glaucoma incidence was found to be 11.5% while Jack J Kanski has mentioned it to be 10%[4]. The Glaucoma in CRS was uncomplicated similar to idiopathic congenital glaucoma or complicated with microcornea. It can be both open angle or angle closure type.

The corneal edema in CRS was found without any rise of intra-ocular pressure and this could be due to abnormal descemet's membrane, endothelitis or infilteration of corneal tissue by the virus. The persistant corneal edema and keratitis may lead to corneal scarring (Delus VP et al)[10]. The microphthalmos was found 25% in CRS in our study while Remington et al mentioned the incidence of microphthalmos to be 20%[1].

CONCLUSION

- Rubella is an important cause of potentially blinding and vision impairing congenital ocular defects.
- The incidence of CRS is still very high ie 16% in Pakistan.
- Congenital cataract & pigmentary retinopathy are the commonest defects in CRS.

- Therefore we recommend to include rubella in Expanded Programme of Immunization to make all women of child bearing age seropositive for rubella.

- Specialized paediatric ophthalmic units may be established to treat children with CRS to reduce the visually disabled citizens of our country.

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