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THE SPECTRUM OF CONGENITAL RUBELLA

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Abstract

Twenty four children, all students at the deaf and blind children's centre, North Rocks, are reviewed. All have multiple defects as a result of congenital rubella. The types of defects are discussed with the prevalence of bilateral cataracts, nystagmus, psychomotor retardation and hearing loss being found to be significant. Emphasis is placed on the importance of immunisation against maternal rubella.

Key words: *Cataracts, nystagmus, retardation, hearing loss, immunisation.*

INTRODUCTION

The Congenital rubella syndrome in its most severe and complete form is characterised by signs of disseminated virus infection. Consequences of infection are determined primarily by the timing of the viral insult. Infection at any time during the first four or five months of gestation may result in widespread or multifocal inflammatory disease of varying severity. Infection in the first six to eight months presents the greatest hazard to organogenesis and life as a result of cell damage and necrosis.

The effects are widespread and severe including microcephaly, psychomotor retardation of varying severity, spastic quadriparesis and sensorineural hearing loss. Ocular abnormalities are cardinal manifestations of congenital rubella. They include unilateral or bilateral pearly nuclear cataract, frequently associated with microphthalmia, iris hypoplasia, atrophy synechiae and vascularization are common. Congenital glaucoma due to incomplete differentiation of the chamber also occurs and pigmentary retinopathy caused by a disturbance of pigmentation in the retinal epithelium. Optic nerve fibre damage and high refractive error are also common and these cause vision deficits, pendular ocular nystagmus and strabismus.

Sir Norman Gregg[1] first drew a correlation between maternal rubella and congenital cataracts in 1941 following an epidemic of german measles. In 1944 he reported further[2] on other defects which were also associated with maternal rubella including cardiac defects, deafness and mental deficiency.

The purpose of this review, of subjects attending the Deaf and Blind Children's Centre, North Rocks, was to assess the incidence of these associated defects in those diagnosed as having congenital rubella.

Information was gathered from the medical files of 24 subjects and by observation, since all are residents of the Centre. Age was considered, as well as the following eye signs — the incidence of cataracts, bilateral and unilateral, microphthalmia, vascularization of the cornea, congenital glaucoma, pigmentary retinopathy, optic atrophy, nystagmus, high refractive error. Other effects considered were microcephaly, psychomotor retardation, cerebral palsy, hearing

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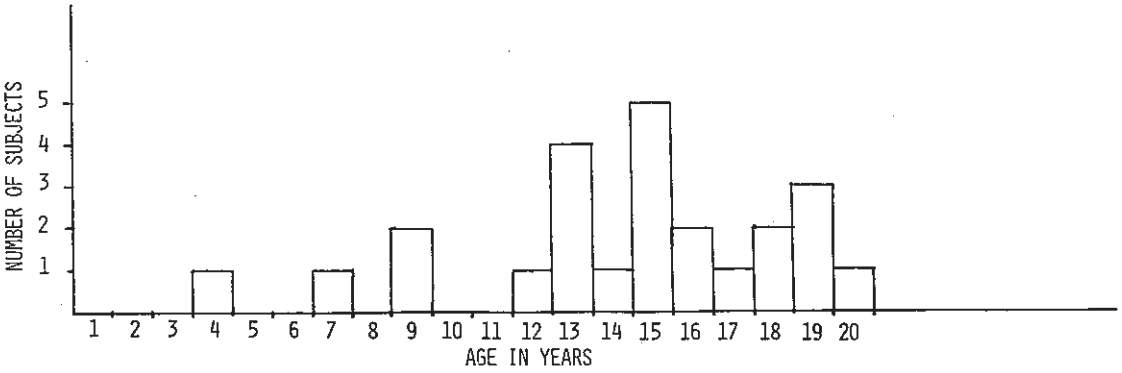

Figure 1: Age of subjects.

loss and congenital heart defects. Note was also taken of how many of the subjects were totally blind and if they had had one or both eyes enucleated.

RESULTS

The subjects' ages ranged between four and twenty years, with the mean age being 14.3. (Fig. 1).

Nystagmus, bilateral cataracts, psychomotor retardation and hearing loss were all found to be most frequently associated with congenital rubella.

One hundred per cent of the subjects in the study had psychomotor retardation ranging from moderate to severe. Eighty seven per cent had a hearing loss with some achieving good hearing with the help of hearing aids and others being classed as profoundly deaf. Seventy one per cent had bilateral congenital cataracts and 8% had unilateral cataracts. The cataracts associated with the congenital rubella syndrome are pearly white, involving all but the outermost layers of the lens, thus indicating that they had begun to develop early in the life of the embryo. Visual acuity was low in all subjects who had cataracts removed with the best assessable vision being 6/24. Twenty nine per cent of subjects were totally blind and nystagmus was present in the remaining 71%, the nystagmus varied between fine horizontal to the fairly gross wandering nystagmus associated with low vision. Thirty seven per cent had microphthalmia in either one or both eyes and glaucoma was present in 21%. Glaucoma is usually difficult to control in these children, two subjects had buphthalmos in one eye with one suffering from a chronic infection in that eye. One subject had a total anterior chamber hyphaema as a result of the glaucoma and another has had both eyes enucleated.

Pigmentary retinopathy was present in 25% of subjects. This pigmentary retinopathy is caused by a disturbance of pigmentation in the retinal epithelium with discrete areas of pigment clumping and focal areas of atrophy and depigmentation of varying size and distribution. The pigment mottling and coarse "salt and pepper" changes are often most pronounced in the macular area and just posterior to the equator and the foveal reflex may be distorted. The retinopathy has minimal, if any, effect on visual functions; it is non-progressive and the ERG responses are generally normal. Vision deficits, pendular ocular nystagmus and strabismus are more commonly due to the cataracts, glaucoma, optic nerve fibre damage, or high refractive error.[3]

Optic atrophy had only been noted in 8%, and 4% of subjects had a high refractive error, e.g., — 10.00 DS. Congenital heart defects were present in 29% with the majority of these being cardiac murmurs. Twenty five per cent had cerebral palsy with 66% of those having spastic quadriplegia and the remaining 33% having hemiplegia. Twenty one per cent were diagnosed as having microcephaly and 17% had vasculari-

sation of the cornea with 75% of these being bilateral and associated with total blindness. Twenty five per cent were affected in one eye only with this eye being buphthalmic.

Other defects noted were hypertension, syndactyly of hands and feet, hare lip and cleft palate. Growth failure was also evident and most subjects had behaviour problems including hyperactivity and difficulty establishing normal sleep patterns.

DISCUSSION

The congenital rubella syndrome has been shown to cause many and varied developmental defects. Generally those affected tend to have several of the associated defects. The group used for this study would be expected to be more severely affected because they are residents in a school and activity therapy centre for multi-handicapped blind students. Therefore, it would be expected that the congenital defects would be found in higher percentages than groups reported by Sir Norman Gregg.[1,2] These two groups are used for a comparison, one with 99 cases and the other with 7. Deafness was present in 55% of the first group and 3% of the second, whereas the North Rocks group had 87%. Cataracts were not included in the first study; in the second they were present in 86% of the group with 57% being bilateral and 29% being unilateral. The North Rocks group had 79% with 71% being bilateral and 8% being unilateral.

Through this comparison it was found that the North Rocks group had a much higher percentage of deafness than Sir Norman Gregg's Group. This may be due to the fact that North Rocks caters for multi-handicapped blind children, so would therefore have a higher percentage of subjects with deafness or any other handicap. The prevalence of cataracts in both groups was in similar percentages indicating that it would be found to be significant in other studies. It is interesting to note that the North Rocks group had a much lower percentage of unilateral cataracts than Sir Norman Gregg's group. The majority of the North Rocks group, i.e., 90% having bilateral cataracts, whereas they were present in 66% of the group used for comparison.

The sample used for this study is small and limited in that all subjects were students in a special school for multihandicapped blind children, or an activity therapy centre which is a progression from the school. It must be assumed that not all babies born with the congenital rubella syndrome will be as severely affected as this sample. However, the risk involved is extremely high. Swan, Tostevin, Mayo and Black[4,5] found that in every instance in which the mother contracted rubella in the first two months of pregnancy she later gave birth to an infant with congenital abnormalities.

Of the subjects included in this study, several are state wards and others attend the centre on a full-time basis with occasional outings or weekends with their families. This is indicative of the difficulties families face in trying to care for these children. Behaviour problems include tantrums, scratching and biting and difficulty establishing normal sleep patterns. Many require full-time care with some being almost fully dependant until they are at least ten years of age. Most will always require some form of institutionalised care and if possible a sheltered workshop situation.

At this point, the advisability of vaccination against rubella cannot be overemphasised. The N.S.W. Department of Health visits all primary and secondary schools once a year and vaccinates all girls who have not yet been vaccinated, provided they have not reached the age of 15 years. Vaccination is still recommended if the child has previously had an attack of rubella. The Department of Health also recommends that all women, before starting a pregnancy, should check whether they are immune to rubella by having an H.A.I. blood test. This is advisable even if they were vaccinated at school.

CONCLUSION

This study has documented the vast range of disabilities that can result from the congenital rubella syndrome. It has shown that children are still being born with this syndrome despite the publicity and Department of Health warnings.

Therefore, a responsibility lies with all health professionals to educate the public as to the necessity for vaccination against rubella, even if it is only by having pamphlets available in their waiting rooms.

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