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Congenital Rubella Syndrome and Hearing Loss in Newborn: A Short Review

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ABSTRACT

Rubella (German measles) is a contagious viral infection caused by Rubella virus (RuV) and most frequently it occurs in the fetus during pregnancy. Congenital rubella infection is one of the most common causes of hearing loss in newborns. The aim of the present study is to systematically review the audiological findings in congenital rubella syndrome. Based on the audiological evaluation the degree, type and configuration of hearing loss could be identified for both ears separately. The studies reported that degree of hearing loss varied from mild to profound hearing loss and have bilateral symmetrical hearing loss. They do often exhibit sensorineural or neural hearing loss. Early intervention can diminish the consequences of hearing loss on child development. Rubella antigen vaccination to all the mothers during their gestation period must be essential in order to prevent congenital rubella syndrome. Screening of all neonates before discharge from hospital should be implemented in all the hospitals to identify congenital hearing impairment at the earliest possible.

Keywords: Rubella virus, Hearing loss, Congenital, Rubella syndrome

INTRODUCTION

Rubella (German measles) is contagious mild viral infection usually manifests with fever and rash caused by Rubella virus (RuV) occurs in fetus during pregnancy [1]. It is Togaviridae family of the genus Rubivirus which affects multiple organs in the system [1]. The effect of rubella infection varies depending on time of gestational period. Previous study reported that the frequency of occurrence of congenital infection is greatest (80%) during the fetus and reduces at the end of second trimester [2,3]. Congenital rubella infection causes multiple congenital abnormalities with wide spectrum of clinical manifestation in fetus, i.e., auditory deficits, vestibular deficits, cataracts, cardiac defects, nerve deafness and cerebral lesion. The complications in later stages include thyroid disease, diabetes, growth hormone deficiency and pan encephalitis in newborns [1,4]. Rubella infection in pregnant women may cause fetal death or congenital defects. Congenital deficits in children due to rubella infection are diagnosed as congenital rubella syndrome (CRS). Immunization is only treatment option for prevention of congenital rubella syndrome [5]. The countries without having rubella vaccination program are suffering with high prevalence of congenital hearing loss due to congenital rubella infection as a major leading environmental cause.

GLOBAL BURDEN OF CRS AND HEARING LOSS

WHO (World health organization) estimate that worldwide more than 100,000 children are born with CRS every year [5]. Hence, it may be assumed that around 300 children are born every day with congenital rubella syndrome with multiple disabilities. However, among these disabilities, the prevalence of hearing loss in congenital rubella syndrome was unexplained. In India, exact estimates of CRS are not available due to lack of studies in general population. However, few studies explained the prevalence of CRS based on clinical and laboratory conditions [5]. A large community based study in India reported that nearly 2.1% children had clinically suspected CRS, 0.58% had clinically confirmed CRS and 0.0009% had laboratory confirmed (IgM) CRS among 51,548 children [6]. As per world health organization (WHO) records, the countries containing first dose of rubella vaccination are depicted in the **Figure 1**.

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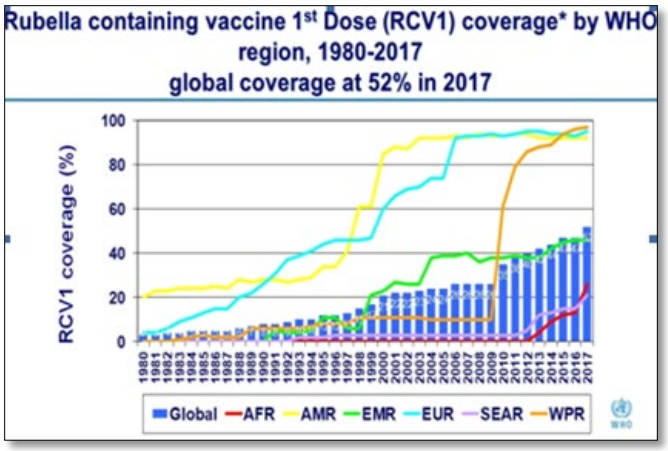

Figure 1. The immunization coverage of rubella vaccine by WHO [7].

AFR: African Region; AMR: American Region; EMR: Eastern Mediterranean Region; EUR: Europe Region Southeast Asian region; WPR: Western Pacific Region

Note: *In some of the countries rubella vaccination is not introduced, namely, Algeria, Bangladesh, Botswana, Burundi, Democratic People's Republic of Korea, Eritrea, Gambia, Ghana, Indonesia, Lesotho, Malawi, Myanmar, Pakistan, Rwanda, Swaziland, United Republic of Tanzania, Vietnam and Zambia [8]*

CONGENITAL RUBELLA SYNDROME AND HEARING LOSS

Congenital rubella infection is one of the most common cases of hearing loss in newborns [2]. In children with CRS sensorineural hearing loss is the second most commonly noted among the various congenital abnormalities in CRS [4,9]. Previous studies reported that the type of hearing loss generally exhibited by CRS are profound degree with sensorineural hearing loss usually bilateral than unilateral hearing loss [10]. Some studies also reported that the degree of hearing loss may vary in range from mild to profound and audiometric configuration is generally flat but few may exhibit sloping pattern and trough pattern audiogram. The type of hearing loss is depending on site and extent of lesion in cochlea and trimester of congenital rubella infection [10,11]. The mixed type hearing loss is uncommon in children with congenital rubella syndrome [11]. Recent study on profiling the audiological characteristics in early infancy (before 6 months of age) by Sao and Navya [12]. In this study, the infants underwent detailed audiological evaluation including otoscopic examination, behavior observation audiometry (BOA), immittance audiometry, otoacoustic emission (OAE) and auditory brainstem response (ABR) test. The otoscopic examination revealed no structural abnormalities of ear canal and tympanic membrane. In behavior observation audiometry, the auditory behavior responses ex. eye blinking, startle responses etc. varies depending on degree of hearing loss. Since, the BOA has poor sensitivity (66.7%) and specificity (86.9%), the objective diagnostic test were adopted [13,14]. The immittance audiometry findings suggestive of intact functioning of middle ear. The absent of OAE suggestive of abnormal outer hair cell functioning in both ears in infant with congenital rubella syndrome. The ABR test was used for threshold estimation, suggestive of infant with CRS have mild to profound degree of hearing loss. In the same study it was also concluded that in CRS may have present OAE and absent of ABR, suggestive of intact functioning of sensory hair cell, i.e., outer hair cell but dysfunctioning of auditory nerve indicative of neural hearing loss in both ears. Based on the audiological evaluation the infants with CRS were diagnosed as having bilateral symmetrical sensorineural hearing loss which may range from mild to profound hearing loss. Thus in this study, all administrated objective test batteries will give valuable information regarding symmetry and severity of hearing loss in infant with CRS before 6 months of age and audiological outcomes in newborn with CRS by using state of art standardized technological instruments for better understanding which aids in channelizing to accurate diagnosis [12].

The newborn have high risk of congenital hearing loss if rubella infection occurs before the 18th week of gestations [10]. Histopathological studies reported that inner ear is most susceptible to damage during sixth to twelve weeks of pregnancy [9]. Thus sensorineural hearing loss in newborns with CRS can be attributed to hemorrhagic damage of sensory end organ of hearing, i.e., organ of corti [15] and interruption in further development of different parts of inner ear and auditory nerve [16]. Bhandary et al. [17] reported that virus affects the blood supply and cystic dilation of the stria vascularis and partial collapse of reissner's membrane of inner ear. It was also reported that the effect of hearing loss resulting in delayed development of speech and language milestone mental retardation and cerebral palsy seen in newborns with CRS [18,19].

INITIATIVE OF GOVERNMENT OF INDIA FOR PREVENTION AND IDENTIFICATION HEARING LOSS IN CRS AND MANAGEMENT

The Government of India has launched the national program, i.e., National Program for Prevention and Control of Deafness (NPPCD) and Rashtriya Bal Swasthya Karyakram (RBSK). This program is also integrated with state and district level program, i.e., National rural health mission in India for early identification and early management of hearing loss. In addition, Government of India has assign work to Anganwadi (AWW) workers and Accredited Social Health Activist (ASHA) workers for door to door survey of deafness maintain records and refer the cases of hearing loss in district hospital/state medical colleges for detail audiological evaluation and treatment [19]. The newborn screening program is also started by All India Institute of speech and hearing funded by Ministry of Health and Welfare. This program focuses on the early identification and rehabilitation of hearing loss in newborns. The early identification of the hearing loss with CRS in newborns will lay a path for the efficient intervention program for the welfare of the child as early as before 6 months of age itself [21].

CONCLUSION

Congenital rubella syndrome is one of prenatal high risk factor causing congenital deafness in newborns. Proper care should be taken by the mother during her gestation period in order to avoid the negative impact of CRS in hearing system and other health problems. Rubella antigen vaccination to all the mothers during their gestation period must be essential in order to prevent congenital rubella syndrome. Screening of all neonates before discharge from hospital should be implemented in all the hospitals to identify congenital hearing impairment at the earliest possible. No drug therapy exists for sensorineural hearing loss but early identification and intervention can facilitate development of communication. Finally, it is important to get sensitized to the effect of congenital rubella infection on hearing system and refer every newborn with congenital rubella syndrome for detail audiological evaluation.

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