# Congenital Rubella Syndrome: Dental Manifestations and Management in a 5 year Old Child

Sarabjot K Bhatia * / Ashima Goyal ** / Mohit Dubey *** / Aditi Kapur ***** / Priyanshi Ritwik *****

*Congenital Rubella Syndrome is a rare disorder comprised of a constellation of physical abnormalities that develop in infants as a result of maternal infection and subsequent fetal infection with rubella virus. The congenital lesions involve vital organs such as heart, eye, ear, brain and endocrine system and less frequently, teeth. The severity of systemic involvement depends on the stage of gestation at which maternal rubella infection occurs. With the implementation of immunization programs worldwide, its incidence has been dramatically reduced during the past half century. This article provides an insight into the prolonged effect of the virus on ameloblasts by highlighting the presence of hypoplastic enamel in primary teeth and erupting permanent teeth in a female child diagnosed with congenital rubella syndrome.*

***Keywords:*** *congenital rubella syndrome, enamel hypoplasia, primary teeth.*
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## INTRODUCTION

Congenital Rubella Syndrome (CRS) was first documented in 1941 by Norman McAllister Gregg, an Australian ophthalmologist who made apparent its true nature and severity potential.[1] Subsequent studies confirmed the teratogenic potential of rubella virus and established the classic triad of rubella embryopathy, namely, cataract, perception deafness and congenital heart disease. Other associated systemic features include mental retardation, encephalopathy, diabetes mellitus and thyroid disorders. Children with CRS may also present with low birth weight, failure to thrive, signs of meningitis with central nervous system damage, microcephaly, bulging fontanelles, hepatosplenomegaly, intestinal pneumonia, petechiae, purpura, enamel hypoplasia and micrognathia. The association between dental anomalies such as enamel hypoplasia and congenital rubella syndrome, first described bySwan,[2] and Evans,[3] was challenged by others due to the inability to accurately diagnose prenatal rubella infection.[4,5] Guggenheimer et al. reported hypoplastic defects of enamel involving 28.8% of the erupted teeth in 12 out of 14 children with documented manifestations of CRS.[6] Hall, in a retrospective study on the effect of different medical conditions on overall prevalence of developmental defects of enamel, reported a high prevalence of enamel defects (81.8%) in rubella embryopathy.[7] Musselman evaluated 50 children with rubella embryopathy and found that 90% of children with CRS exhibited enamel hypoplasia, 78% had tapered teeth and 18% had notched teeth.[8] This case report highlights the presence of dental defects in the primary dentition and erupting permanent molars and describes management of the defects in a 5-year-old child who presented with CRS.

### Case report

A five-year-old North-Indian female child was referred from Advanced Pediatric Centre at the Postgraduate Institute of Medical Education and Research (PGIMER) in Chandigarh, India with the chief complaint of decayed and discoloured teeth with sensitivity to hot and cold. The child had been diagnosed with CRS and was undergoing treatment for cataract and perception deafness. The first-born child, her mother had developed high grade fever with macular rash during the third month of pregnancy. No diagnosis of severe illness was made by the treating physician at that time and no treatment was instituted. The child was born full-term with a birth weight of 4.5 pounds. At 3 months of age, her parents reported to the Advanced Pediatric Centre of the

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institute with the chief complaint of interrupted feeding, episode of cyanosis while crying, recurrent cough and fever and white patches in her eyes. The child was diagnosed with microcephaly, bilateral congenital cataract, congenital heart disease (acyanotic with Patent Ductus Arteriosus) and perception deafness. The prenatal history, clinical features and laboratory tests (positive ELISA for Rubella IgG, IgM) were consistent with the pathophysiologic effects of congenital rubella syndrome.

Examination showed the weight, height, state of nourishment and cognitive ability of the child to be within the normal range for a 5 year old child. Extra-oral examination revealed a convex profile and frontal bossing. Intraorally, the child had a full complement of primary teeth affected with varying degrees of hypoplasia, ranging from minor pitting to complete loss of enamel. In the maxilla, the incisors exhibited more loss of tooth structure than the molars, whereas in the mandible, the molars were more severely affected than the incisors. (Figure 1). The maxillary incisors exhibited loss of tooth structure down to the gingiva. However, it was difficult to discern whether pre-existing hypoplasia lead to such severe loss of tooth structure or if dental decay was the predominant causative factor. Parents reported that onset of caries after enamel had started chipping off spontaneously when the child was 3 years old. Other contributing etiological factors for enamel hypoplasia

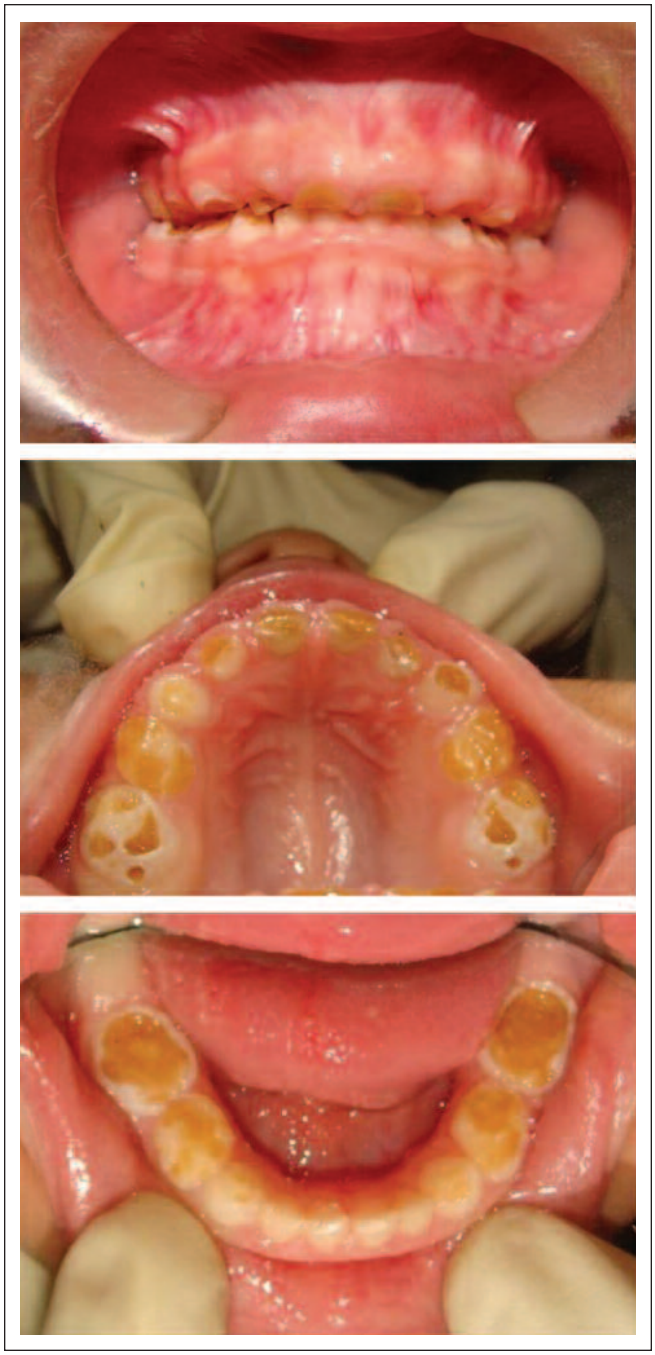

**Figure 1.** Pre-operative view. Note the presence of hypoplasia on all the primary teeth

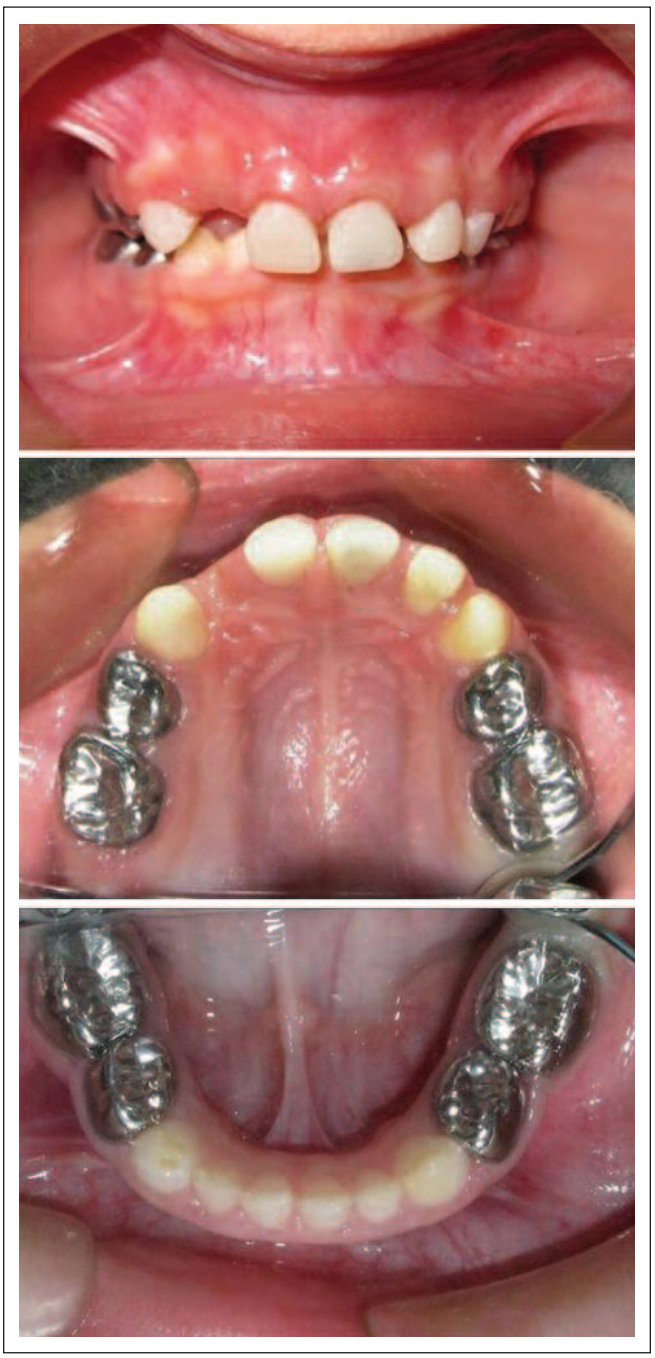

**Figure 2.** Complete rehabilitation: Immediate post operative

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were ruled out. The child was still bottle-fed at the time of initial examination at age 5 years. Orthopantomogram (OPG) showed an irregular pattern of generalized enamel wear and loss of tooth structure with no congenitally missing primary or permanent teeth. Taurodontism was seen in the maxillary primary second molars.

The child and her parents were taught about the importance of maintaining at–home oral hygiene, use of fluoridated toothpaste, daily application of CPP-ACP™ (RECALDENT) as home care measures and the need to eliminate bottle feeding. Treatment was initiated in the dental office and full mouth rehabilitation was planned. Posterior teeth received full coverage restorations with preformed stainless steel crowns. The maxillary right lateral incisor was extracted as more than half of its root had resorbed. The maxillary central incisors and the left lateral incisor received pulpectomy along with short fiber posts (3M ESPE) for intracanal reinforcement and enhanced retention of the final restoration. The final restorations were

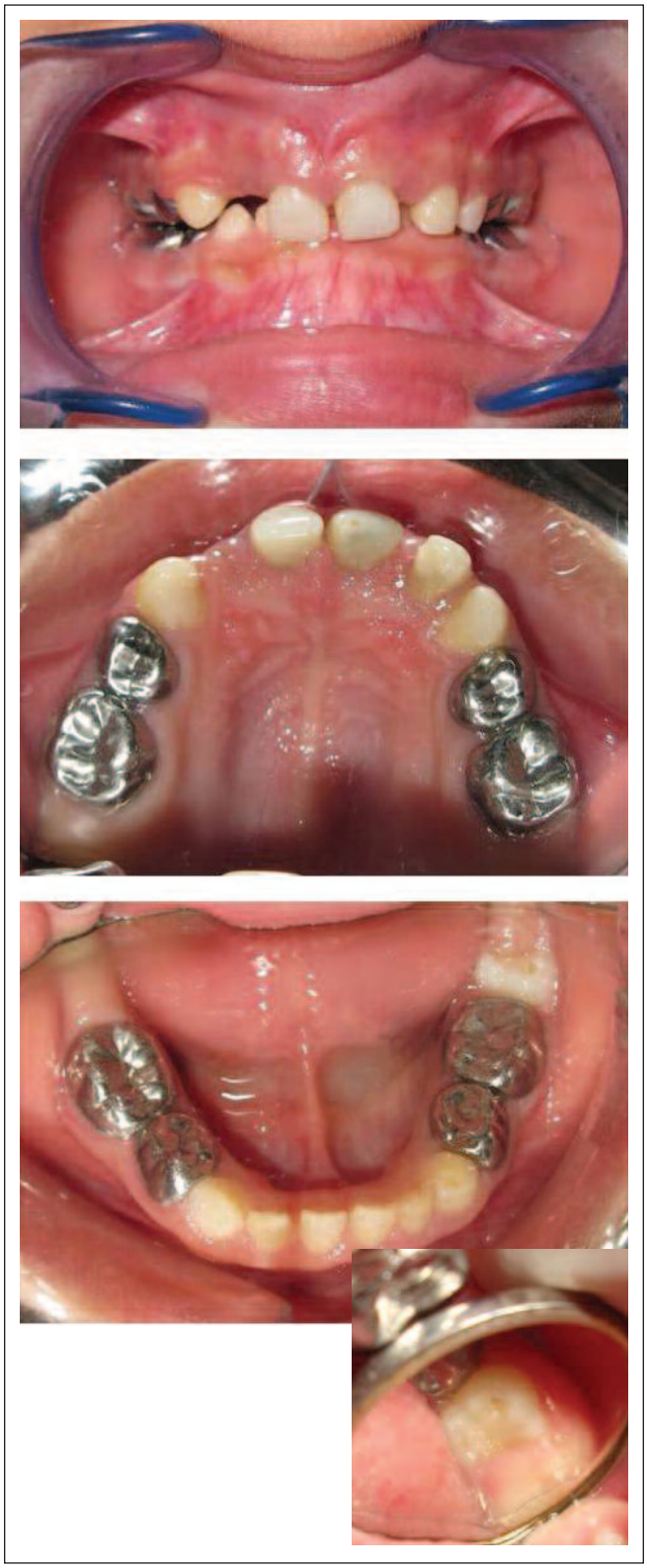

**Figure 3.** Follow up after 12 months. Note the hypoplastic erupting permanent mandibular first molar

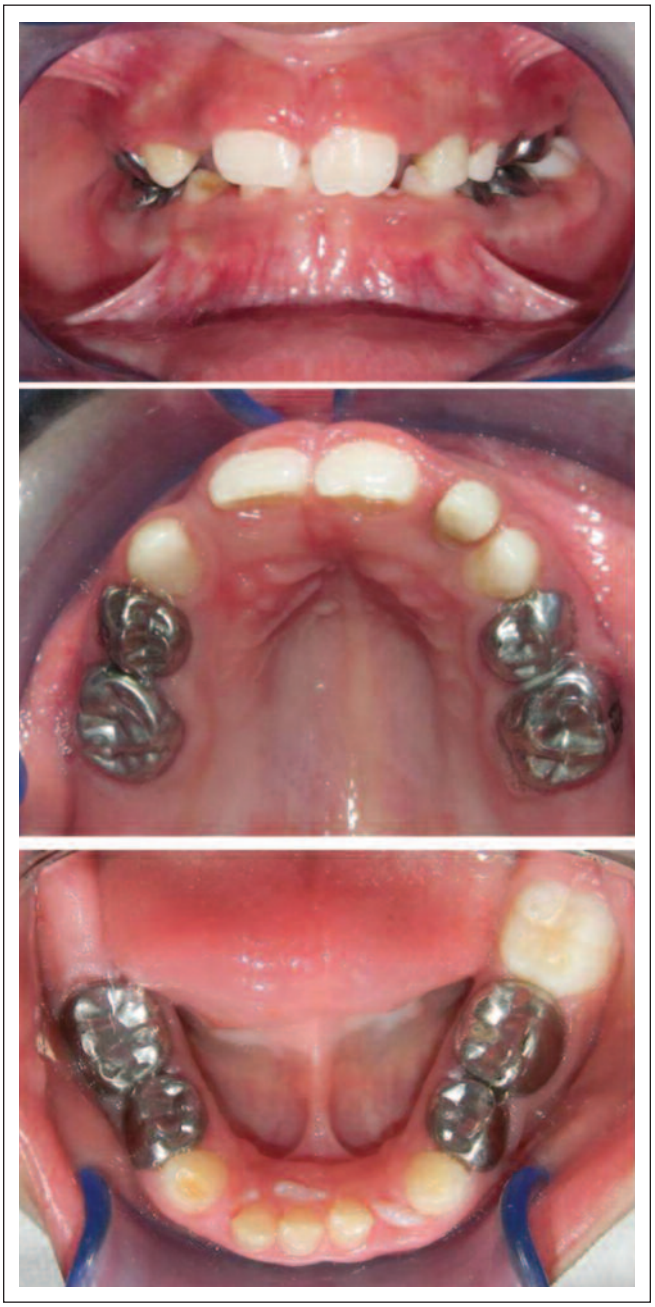

**Figure 4.** Follow-up after 24 months. Maxillary permanent incisors are unaffected

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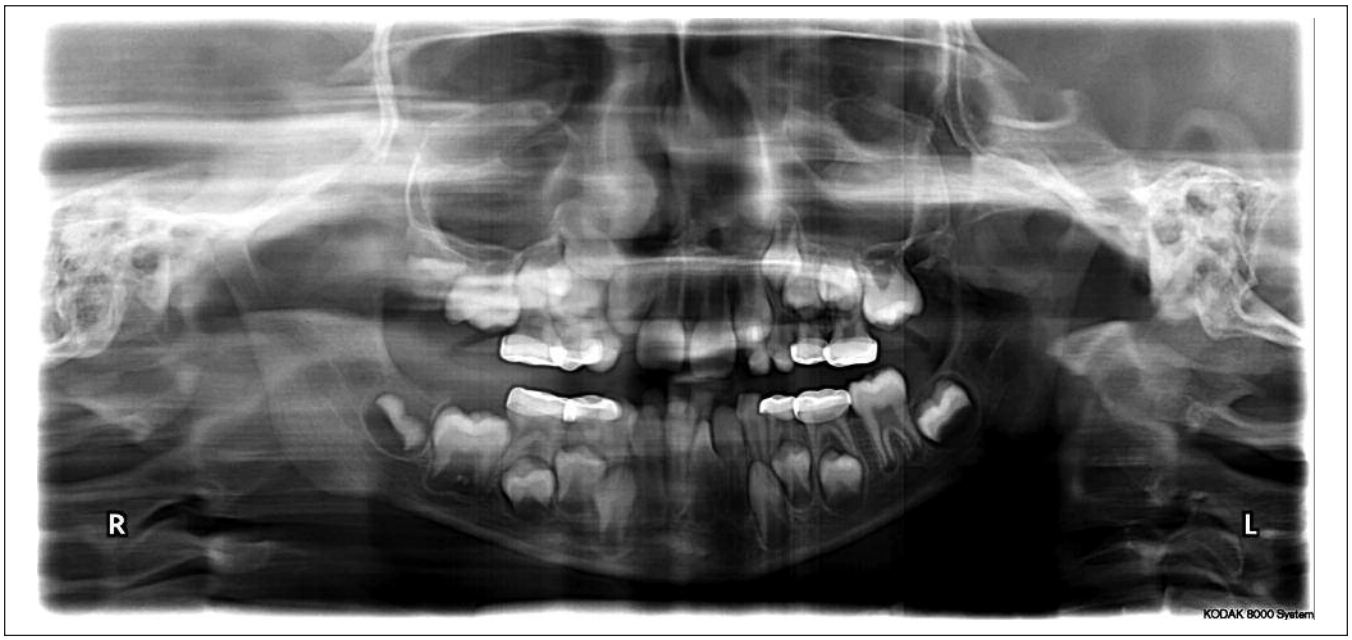


**Figure 5.** Orthopantomograph at 7 years of age

direct dental composite crowns with celluloid strip crowns (Pedoform strip crowns Unitek /3M ESPE). The maxillary canines and mandibular canines and incisors were restored with composite restorations (Z350 3M ESPE, shade A2). (Figure 2)

**Follow-up**

The child did not complain of any sensitivity or pain at follow-up visits every three months. At the 12 month follow-up, the mandibular permanent left first molar was erupting and exhibited enamel hypoplasia (Figure 3). Sealant and fluoride varnish were applied to the molar. The patient was then scheduled for periodic 6-month follow-up appointments to evaluate and manage erupting permanent teeth. At 24 months follow-up, the permanent incisors had erupted and were unaffected (Figure 4). The remaining three first permanent molars had not erupted. The OPG taken at this visit did not show any additional abnormalities (Figure 5).

## DISCUSSION

Since its discovery in the mid 19th century, the rubella epidemic has been reported to occur at 6 to 9-year intervals, and major pandemics have occurred every 10 to 30 years.[9] The initialization in the early 1970's of vaccination programs worldwide has led to a dramatic decrease in the incidence in developed countries, with an estimated incidence of less than 2 per 100,000 live births.[10] In developing countries however, the picture is dismal as only 28% of the population is routinely vaccinated against rubella and the disease continues to affect pregnant females and subsequently their offspring with an incidence rate of 0.4-4.3/1000 live births.[11]

The pathogenesis of congenital rubella syndrome is not well-established. The rubella virus is generally described as non-cytolytic, allowing cell survival. However, it causes chromosomal breakdown and production of a protein that inhibits mitosis, thereby resulting in persistently infected cells that show reduced mitotic activity.[12] It has been suggested that failure of cell differentiation, disturbed morphogenesis and retarded growth in rubella infants may have been due to the lack of cellular nutrition which results from interference with vascular supply.[13] The defective enamel formation can be attributed to the failure of ameloblastic differentiation, interference with its secretory function or its total destruction. The evidence of necrosis of ameloblasts has been seen in electively aborted rubella fetus.[14] Kraus et al. cited that viruses affect the development of tooth buds and sometimes are responsible for their total disappearance.[15] Areas of clinical enamel aplasia or hypoplasia in the present case can be related to areas of complete absence of ameloblasts either caused directly by the virus or indirectly by the vascular changes. These could also be a result of total metabolic disturbance of the fetus or low birth weight.[16]

The risk of fetal infection varies according to the time of onset of maternal infection. The estimated risk of malformations is 90% for those infected between weeks 2 through 10 of gestation, 34% for those infected in weeks 11 through 18 and no malformations for those infected after 18 weeks.[17] In the present case, the maternal infection occurred between the 8th to 12th week of pregnancy and the child presented with most of the signs of CRS, i.e. congenital cataract, presence of patent ductus arteriosus, perception deafness, microcephaly and low birth weight. The dental manifestations included enamel hypoplasia in almost all of the primary teeth with symmetrical distribution of the defects, thus supporting the contention that "structures undergoing symmetrical stages of development while under the influence of a teratogen should show similar abnormalities."[6] Besides enamel hypoplasia, other oral findings have been micrognathia, cleft palate or abnormally shaped palate and delayed eruption of teeth.[6] In the present case, the eruption status

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appears to be normal as the parents reported that the first tooth erupted in the oral cavity at 7 months of age and the permanent mandibular first molar erupted at the age of 6 years.

Though maternal infection in the case described here preceded the onset of enamel calcification, which usually starts at around 13 weeks of intrauterine life, the severity of hypoplasia can be attributed to persistent infection, which could have influenced the calcification of primary dentition through its development (mineralization of the crowns of the entire primary dentition is not complete until about 12 months postnatal). Studies have shown that viral infection, once established, persists in fetal tissues and the infant keeps shedding rubella virus for months after birth. Rubella virus particles are retained in secluded sites such as crystalline lens and other target organs where they undergo recurrent periods of increased virus production and replication.[18] In the present case , the presence of hypoplastic defects in the erupting permanent teeth, , indicates persistent infection and presence of virus for a prolonged period that possibly led to hypoplastic lesions in permanent teeth, which begin to calcify after birth. Studies have shown that the rubella virus may persist for up to 3 years of age in severely infected infants. The virus has been isolated in adults as late as 28 years of age.[19] Our patient presented with hypoplastic permanent first molar, but not the permanent incisors. This further accentuates the time sensitive effect of the rubella virus. Development of the permanent first molars is initiated at 20 weeks in utero and calcification commences at birth. The initiation of the permanent incisors also occurs at 20 weeks in utero. However, calcification begins at 3-4 months post partum. It is possible that the difference of a few months in the commencement of calcification led to either inactivation of the virus or its complete clearance.

The management of children with CRS presents a significant challenge to the dentist because of physical and intellectual limitations and little or no parental concern about maintaining oral health.[20] Moreover, the presence of a varying degree of hypoplasia in these children can act as nuclei in the initiation and progression of dental caries, thus necessitating a long term preventive regimen tailored for the individual patient. Dental care is comprised of an assessment of diet cariogenicity and appropriate recommendations for dietary modifications, oral hygiene instructions, application of topical fluorides, home care measures such as daily use of fluoridated toothpaste and CPP-ACP, and placement of fissure sealants wherever required.[21]

## CONCLUSIONS

Despite massive vaccination programs throughout the world, children in developing countries continue to be affected by the congenital rubella syndrome due to inadequate vaccination programs and lack of routine surveillance for rubella infection. This case report has documented the manifestations of this syndrome in a five-year old female child with classical signs of CRS and enamel defects on almost all primary teeth and the first permanent molars. Enamel hypoplasia was the outcome of the persistent viral infection, which affected the ameloblasts for an extended period beyond the initial maternal infection. Regular follow-up of children with CRS is important until the eruption of permanent teeth to monitor the effects of the infection on the permanent dentition. Preventive dentistry should be the hallmark of dental management for these children.

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