

# Salt and Pepper Pigmentary Retinopathy in Congenital Rubella Syndrome: A Case Report

RAINA JAIN<sup>1</sup>, SACHIN DAIGAVANE<sup>2</sup>

## ABSTRACT

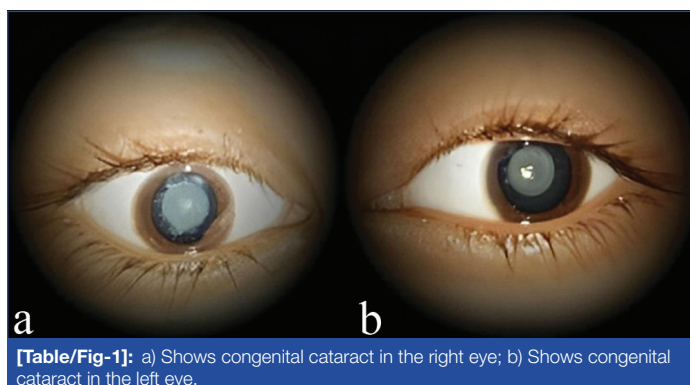
A fatal viral infection called rubella, often known as German or three-day measles, is spread by the Rubivirus genus of the *Togaviridae* family. The disorder known as Congenital Rubella Syndrome (CRS) has the potential to cause a wide range of multisystem problems. Every organ in the developing foetus can be damaged; the most prevalent involvements are cardiac, ophthalmic, and auditory abnormalities. This is a case report of an 11-year-old patient who was hospitalised at a tertiary care facility with severe signs and symptoms of CRS, which were identified at birth. Ocular abnormalities included congenital cataracts and salt and pepper retinopathy. The lens aspiration and Intraocular Lens (IOL) implantation were done. This case serves as an example of the significance of the rubella immunisation program.

**Keywords:** Congenital cataract, Salt and pepper retinopathy, Sensorineural hearing loss

## CASE REPORT

An 11-year-old boy presented to the Ophthalmology Outpatient Department (OPD) with his parents at a Tertiary Care Centre in Central India with complaints of progressive diminution of vision since birth. During the antenatal period, the mother suffered from fever for a long duration during the first trimester of pregnancy. The child was diagnosed with CRS at birth through raised IgG and IgM antibodies. In this case, the patient's CRS was identified at birth. Congenital rubella infection was confirmed with an increasing serum level of rubella-specific IgG over the first year; Anti-Rubella IgG is more than 360 IU/ml (positive), and Anti-Rubella IgM is more than 2.08 IU/ml (positive). The child had a history of Neonatal Intensive Care Unit (NICU) admission due to a low birth weight of 1.8 kg. The patient has several disease-related issues, including bilateral cataracts, salt and pepper retinopathy, speech difficulties, and sensorineural hearing loss in both the ears during examination with Brainstem Evoked Response Audiometry (BERA) since he was one year old. The parents gave a history of developmental delay in the child. No cardiac abnormalities were detected at birth.

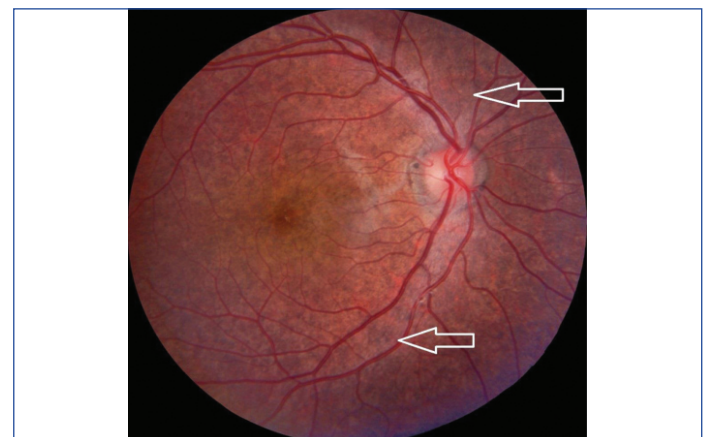
The patient had the best-corrected visual acuity of 6/36 in the right eye and 6/24 in the left eye. On examination, the Hirschberg test [1] was orthophoric. On slit lamp examination, the patient had a clear cornea, pupils normal in size and reactive to light, and congenital cataracts in both eyes [Table/Fig-1]. Intraocular pressure was 19 and 15 mmHg in the right and left eye, respectively, as measured by Goldmann Applanation Tonometry. Fundus evaluation could not be done in both eyes due to hazy media caused by dense cataracts. The patient was diagnosed with congenital cataract.



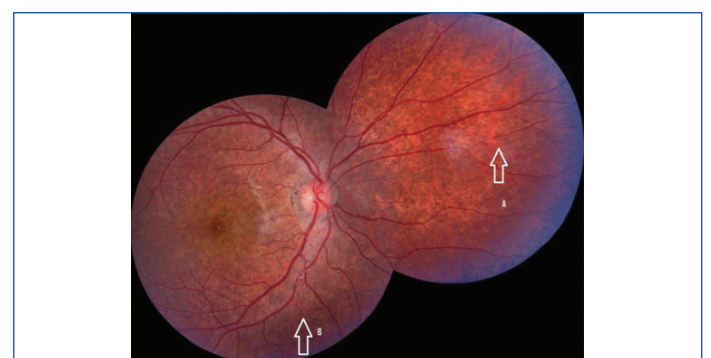
**[Table/Fig-1]:** a) Shows congenital cataract in the right eye; b) Shows congenital cataract in the left eye.

After obtaining informed consent and confirming fitness for surgery under general anaesthesia, the patient underwent cataract extraction by nucleus aspiration with foldable hydrophobic posterior chamber intraocular lens (ACRIOL Care Group) implantation in the right eye. Postoperative visual acuity was 6/12 in the right eye, and the patient was prescribed Tablet Ciprofloxacin 400 mg twice a day and eyedrop moxifloxacin 0.5% four times a day. Postoperative fundus examination by indirect ophthalmoscopy in the right eye revealed a disc within normal limits with multiple fine pigmentary changes in all four retinal quadrants extending from the posterior pole to the far periphery.

[Table/Fig-2] shows salt and pepper retinopathy in the right eye. [Table/Fig-3] shows a montage image of salt and pepper retinopathy in the right eye. Fundus evaluation could not be performed in the left



**[Table/Fig-2]:** Shows salt and pepper retinopathy in right eye.



**[Table/Fig-3]:** Shows a montage image of salt and pepper retinopathy in the right eye. Arrow A shows hypopigmentary and Arrow B shows hyperpigmentary lesions.

eye due to hazy media and dense cataract. Fundus photographs were taken using Zeiss Visucam 524.

The patient was discharged and advised to follow-up after one month for cataract extraction in the left eye during the next visit. On follow-up, vision in the right eye was recorded as 6/9, and the patient is willing to undergo left eye cataract surgery after one month.

## DISCUSSION

A fatal viral infection called rubella, often known as German or three-day measles, is spread by the Rubivirus genus of the *Togaviridae* family [1]. When a pregnant woman is exposed to the rubella virus in the first trimester, she acquires CRS. The virus spreads vertically through transplacental transmission, resulting in major congenital defects, abortion, and stillbirth [1]. It spreads through direct contact with the saliva or mucus of the infected individual, as well as respiratory droplets produced while coughing or sneezing [2]. CRS occurs when the developing foetus contracts the rubella virus from an infected mother, typically during the first trimester. A pregnant female with rubella during the first trimester of her pregnancy has a 90% chance of transplacental passage of the rubella virus to the foetus [3].

The aetiopathogenesis of rubella is due to aerosolised particles from an infected patient's respiratory secretions. The respiratory endothelium becomes infected with the virus, which then spreads haematogenously to local and distant lymphatics during the first viremic phase. Around 5 to 7 days after infection, the virus may be detected in the blood. The secondary viremic phase, lasting between six and 20 days, is when the virus spreads throughout the body, being found in the lymph nodes, urine, Cerebrospinal Fluid (CSF), synovial fluid, and lungs, among other bodily parts. After exposure of 3-8 days, an infected person can transmit the virus through droplet infection from the nasopharynx [2]. Rubivirus is the cause of CRS [4]. There is a significant probability that a pregnant mother who develops rubella during her first trimester will transmit it to her foetus, potentially leading to miscarriage due to the infection [4].

Ophthalmic anomalies such as cataract, pigmentary retinopathy (salt and pepper type), congenital glaucoma, chorioretinitis, sensorineural hearing loss, congenital heart defects (particularly branch pulmonary artery stenosis and patent ductus arteriosus), and in surviving neonates are the classic clinical triad of manifestations associated with CRS [5]. Blueberry muffin skin lesions, an enlarged spleen, developmental delay, meningoencephalitis, microcephaly, low birth weight, radiolucent bone disease, and jaundice within 24 hours of delivery are additional clinical markers [6,7].

In a case report by Nayyar M et al., a four-month-old infant was diagnosed with congenital cataract, and upon detailed evaluation, the patient was found to have CRS. A lensectomy with anterior vitrectomy was performed for cataracts, leaving the patient aphakic [8]. Khurana RN and Sadda S observed salt and pepper retinopathy in 39-year-old patients suffering from CRS [9]. The case report discussed by Damasceno N et al., presented a rare occurrence of unilateral rubella retinopathy in an adult, which has not been previously documented in medical literature. The patient, a 28-year-old healthy white man, experienced vasculitis and retinitis in his right eye, resulting in sudden vision loss. This condition followed an acute viral illness, and serology

tests confirmed positivity for rubella. Treatment involved corticosteroid therapy, particularly due to macular involvement, and the patient's final visual acuity improved to 20/60 from an initial 20/200. The study concludes that rubella was the aetiological factor, evidenced by positive serology results during the acute phase of the viral infection, even in the presence of viral retinopathy [10].

Salt and pepper retinopathy is observed due to atrophy and necrosis of pigment epithelium induced by the rubella virus. Rubella retinopathy can lead to subretinal neovascularisation, resulting in disciform maculopathy after scarred healing. This is a rare complication. It is advised to administer the Measles, Mumps, and Rubella (MMR) vaccine, which protects against measles, mumps, and rubella. The MMR vaccine should be given to infants between the ages of one year and 15 months and again between the ages of four and six years, as per medical advice. The MMR vaccine prevents rubella and provides lifelong immunity. Vaccination can prevent rubella from being transmitted to future pregnancies.

## CONCLUSION(S)

CRS often affects infants born to females from nations with inadequate immunisation programs. Early detection of rubella virus infections is essential to reduce the incidence of CRS. Typical issues in infants with CRS include congenital auditory and visual defects, developmental delays, and positive rubella serology in both the mother and newborn supports the diagnosis. Congenital cataracts can be managed through surgical intervention and IOL implantation, while there is no definitive treatment for pigmentary retinal changes. Vaccination remains the sole known preventive measure for CRS.

## REFERENCES

- [1] Seetoo K, Carlos MP, Blythe D, Trivedi L, Myers R, England T, et al. Three cases of congenital rubella syndrome in the postelimination era-Maryland, Alabama, and Illinois, 2012. *MMWR Morb Mortal Wkly Rep*. 2013;62(12):226-29.
- [2] Athira PS, Simon H, Sivakumar T. Congenital rubella syndrome- A case report. *Int J Res Rev* 2019;6(11):421-24.
- [3] World Health Organization. Measles. [cited 2023 Jul 31]. Available from: <https://www.who.int/news-room/fact-sheets/detail/measles>.
- [4] Boston Children's Hospital. Congenital Rubella | Boston Children's Hospital. [cited 2023 Jul 31]. Available from: <https://www.childrenshospital.org/conditions/congenital-rubella>.
- [5] WHO, CDC, International Clearinghouse for Birth Defects. Birth defects surveillance: Quick reference handbook of selected congenital anomalies and infections. Geneva: World Health Organization; [Internet]. [cited 2020 Dec 01]. Available from: <https://www.who.int/publications/i/item/9789240015418>.
- [6] Centers for Disease Control and Prevention. Congenital rubella syndrome (CRS). 2020 [cited 2023 Jul 31]. Available from: <https://www.cdc.gov/ncbddd/birthdefects/surveillancemanual/chapters/chapter-5/chapter-5-1.html>.
- [7] Parija SC. Infective syndromes: A system and organ-based approach. In: Parija SC, editor. *Textbook of microbiology and immunology*. Singapore: Springer Nature; 2023 [cited 2023 Aug 1]. Pp. 1039-77. Available from: [https://doi.org/10.1007/978-981-19-3315-8\\_71](https://doi.org/10.1007/978-981-19-3315-8_71).
- [8] Nayyar M, Dalvi M, Mohta A, Sabnis M, Kavthekar S. A case report & management of bilateral congenital cataract due to congenital rubella syndrome. *J Adv Med Dent Scie Res*. 2020;8(1):82-85.
- [9] Khurana RN, Sadda SR. Images in clinical medicine. Salt-and-pepper retinopathy of rubella. *N Engl J Med*. 2006;355(5):499. Doi: 10.1056/NEJMicm040780. Erratum in: *N Engl J Med*. 2006;355(15):1627. PMID: 16885553.
- [10] Damasceno N, Damasceno E, Souza E. Acquired unilateral rubella retinopathy in adult. *Clin Ophthalmol*. 2010;5:3-4. Doi: 10.2147/OPTH.S15273. PMID: 21311649; PMCID: PMC3032996.

### PARTICULARS OF CONTRIBUTORS:

1. Resident, Department of Ophthalmology, Jawaharlal Nehru Medical College, DMIHER, Sawangi, Wardha, Maharashtra, India.
2. Professor and Head, Department of Ophthalmology, Jawaharlal Nehru Medical College, DMIHER, Sawangi, Wardha, Maharashtra, India.

### NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Raina Jain,  
Resident, Department of Ophthalmology, Jawaharlal Nehru Medical College,  
DMIHER, Sawangi, Wardha-442001, Maharashtra, India.  
E-mail: rainajain93@gmail.com

### AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

### PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Aug 21, 2023
- Manual Googling: Nov 25, 2023
- iThenticate Software: Apr 08, 2024 (11%)

### ETYMOLOGY: Author Origin

EMENDATIONS: 6

Date of Submission: Aug 18, 2023

Date of Peer Review: Nov 18, 2023

Date of Acceptance: Apr 09, 2024

Date of Publishing: Jun 01, 2024