

Multi-Sensory Sequelae of Congenital Rubella Syndrome: A Case Report on Ocular and Auditory Deficits

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Abstract: Congenital Rubella Syndrome (CRS) is a multi-systemic disorder characterized by a classic triad of ocular, cardiac, and auditory defects. This report evaluates an 8-year-old male with a history of maternal rubella and subsequent bilateral cataract surgery at age two. Clinical examination revealed horizontal pendular nystagmus, microcornea (9mm), and "salt and pepper" retinopathy. Beyond ocular sequelae, the patient exhibited sensorineural hearing loss requiring bilateral auditory amplification and developmental speech delay necessitating ongoing therapy. Visual acuity remained limited to 6/36 (OD) and 6/24 (OS) despite optimal refractive correction (+10.0D). This case highlights the necessity of a multidisciplinary approach in managing the complex neurosensory deficits associated with CRS. Early intervention is critical, as late-stage management is often limited by permanent structural and neurosensory anomalies.

Keywords: Congenital Rubella Syndrome, Salt and Pepper Retinopathy, Microcornea, Hearing Loss, Speech Therapy

1. Introduction

Congenital Rubella Syndrome (CRS) is a consequence of transplacental transmission of rubella virus, particularly when maternal infection occurs during early gestation, leading to permanent structural and functional abnormalities [1]. While ophthalmological findings like cataracts and retinopathy are hallmark features, the systemic impact often includes sensorineural hearing loss and neurological delays [2]. This case illustrates the long-term challenges of a patient navigating both severe visual impairment and auditory-speech deficits, emphasizing the limitations of late surgical intervention.

2. Literature Survey

The systemic burden of CRS is well-documented; hearing loss is reported in up to 80% of affected infants [1]. The pigmentary "salt and pepper" retinopathy is the most common ocular sign, resulting from irregular pigmentation of the retinal pigment epithelium (RPE) [3]. While the retinopathy is typically non-progressive, its association with microcornea (diameter <10mm) significantly increases the risk of secondary glaucoma and surgical complications like capsular phimosis [4]. Delayed speech acquisition is a frequent secondary effect of early-onset hearing loss, requiring specialized therapy [5].

3. Case Presentation

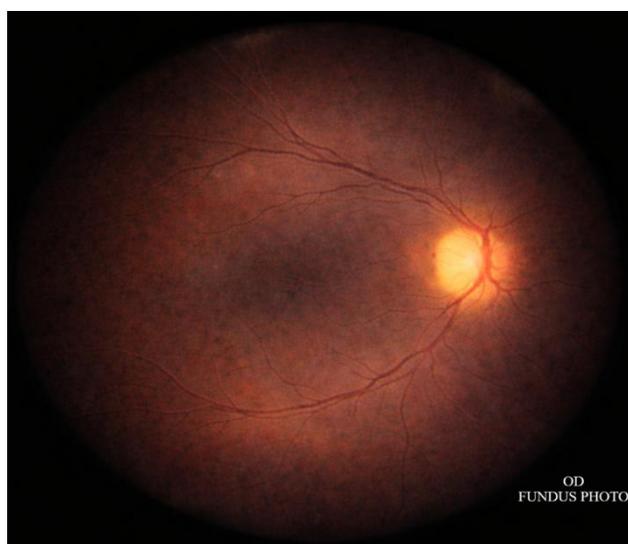
Clinical History: An 8-year-old male presented with a lifelong history of diminished vision. Maternal history was positive for rubella during the first trimester. The patient underwent bilateral cataract extraction at age 2. Systemically, the patient has significant auditory impairment, utilizing a hearing instrument for communication. Due to delayed language milestones, he is currently enrolled in speech therapy.

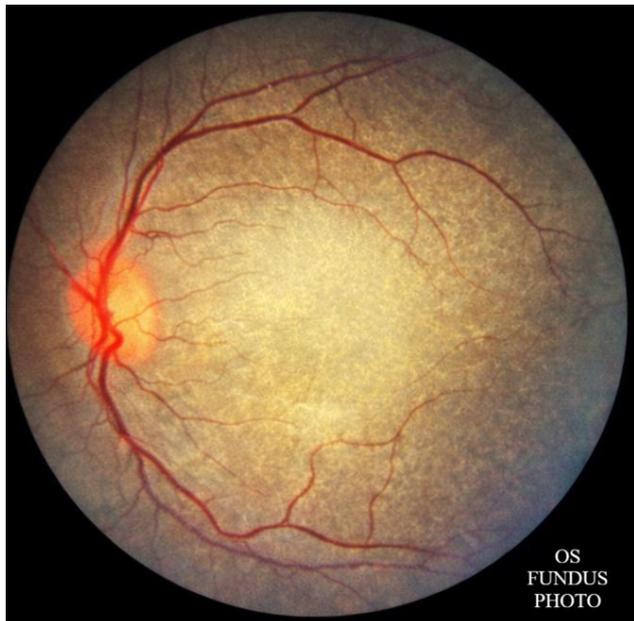
Table 1: Ocular Examination Data:

Parameter	Right Eye (OD)	Left Eye (OS)
BCVA (+10.0D)	6/36	6/24
Corneal Diameter	9 mm	9 mm
Anterior Segment	Aphakia, Capsular Phimosis	Aphakia
Nystagmus	Horizontal Pendular	Horizontal Pendular
Pupillary Dilation	5 mm	5 mm

Table 2: Fundoscopic Observations:

Fundus	Right Eye (OD)	Left Eye (OS)
Glow	Dull	Red
Disc	Oval disc with clear margins and healthy NRR	Oval disc with clear margins and healthy NRR
Cup to Disc Ratio	0.4	0.4
Vessels	Normal	Normal
Background	Dull background Mild Salt and Pepper pigmentation	Salt and Pepper pigmentation
Macula	Absent foveal reflex	Absent foveal reflex





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4. Discussion

The presentation demonstrates the classic multi-organ involvement of CRS. The 9mm microcornea and sensory nystagmus indicate profound early-life visual deprivation. Since cataract surgery was performed at age 2—well beyond the critical period of visual development—the patient developed permanent sensory amblyopia^[5].

The "salt and pepper" appearance and absent foveal reflex suggest foveal hypoplasia, which limits the potential for 6/6 vision even with refractive correction^[6]. Systemically, the patient's reliance on a hearing instrument and speech therapy highlights the high prevalence of sensorineural hearing loss. This dual sensory impairment poses a significant barrier to traditional learning, requiring a coordinated multidisciplinary approach between ophthalmology, audiology, and speech-language pathology.

5. Conclusion

This case underscores the irreversible multi-organ damage caused by the rubella virus. While surgical measures addressed the cataracts, the patient continues to struggle with permanent neurosensory deficits. Prevention through universal maternal immunization remains the only definitive strategy to mitigate the multi-systemic burden of rubella.

6. Future Scope

Long-term monitoring should focus on identifying late-onset glaucoma and potential subretinal neovascularization. Future research into the efficacy of early cochlear implantation versus traditional hearing aids in CRS patients may provide better developmental outcomes.

References

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