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# BEYOND THE CLASSIC TRIAD: CONGENITAL RUBELLA SYNDROME PRESENTING WITH NEUROLOGICAL DYSFUNCTION AND BILATERAL BASAL GANGLIA–THALAMIC ABNORMALITIES IN A NEONATE — A CASE REPORT

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**ABSTRACT** **Background:** Congenital rubella syndrome (CRS) is a preventable multisystem disorder resulting from maternal rubella infection during pregnancy. Although sensorineural hearing loss, ocular abnormalities and congenital heart disease are classical manifestations, neurological involvement may dominate the neonatal presentation and delay recognition.**case Presentation:** A term male neonate weighing 2.75 kg was delivered by caesarean section for breech presentation. He developed a high-pitched cry, jitteriness, poor feeding, hypotonic suck and absent Moro reflex. Examination revealed craniofacial dysmorphism, hepatomegaly, stridor, excessive oral secretions, palmoplantar desquamation, central hypotonia with appendicular hypertonia, hyporeflexia and weak primitive reflexes. Neurosonography demonstrated bilateral subdural collections. Magnetic resonance imaging showed T1 hyperintensity involving the bilateral basal ganglia and thalami, mild subdural haemorrhage and no diffusion restriction. Echocardiography revealed a small patent ductus arteriosus with a left-to-right shunt. Auditory brainstem response testing demonstrated bilateral moderate-to-moderately severe sensorineural hearing loss, while ophthalmological examination was normal. Rubella serology showed elevated rubella-specific IgG with positive rubella-specific IgM, supporting CRS.The neonate received antiseizure treatment, nutritional support and neurodevelopmental stimulation, with multidisciplinary follow-up.**conclusion:** CRS should be considered in neonates with unexplained neurological abnormalities accompanied by hearing impairment or congenital heart disease, even when the complete classical triad is absent. Careful interpretation of neonatal serology and comprehensive multisystem evaluation enable timely diagnosis and early intervention to reduce preventable long-term sensory and developmental disability.

**KEYWORDS :** Congenital rubella syndrome; sensorineural hearing loss; patent ductus arteriosus; basal ganglia; neonatal neurological dysfunction.

## INTRODUCTION

Congenital rubella syndrome (CRS) results from transplacental transmission of the rubella virus following maternal infection during pregnancy. The risk and severity of fetal injury are greatest when infection occurs during the first trimester, particularly during the initial 8–10 weeks of gestation, when multiple fetal abnormalities may develop in up to 90% of affected pregnancies (1).

The classical clinical spectrum includes sensorineural hearing loss, ocular abnormalities such as cataract, congenital glaucoma or pigmentary retinopathy, and congenital heart disease, particularly patent ductus arteriosus and pulmonary artery stenosis. However, CRS is a multisystem disorder and may additionally cause growth restriction, hepatosplenomegaly, thrombocytopenia, jaundice, microcephaly, meningoencephalitis and neurodevelopmental impairment. The manifestations vary according to the timing of maternal infection, and the complete classical triad may not be evident during the neonatal period (2).

Despite the availability of effective rubella-containing vaccines, CRS remains an important preventable cause of childhood morbidity in India. Sentinel surveillance conducted in India during 2016–2018 found laboratory-confirmed CRS in approximately one-fifth of infants evaluated for suspected disease, with cardiac defects, ocular abnormalities and hearing impairment forming the principal clinical manifestations (3). Neurological presentations may be diagnostically challenging because abnormalities of tone, feeding, primitive reflexes or seizure-like activity are not specific to rubella and may initially suggest hypoxic-ischaemic, metabolic or other congenital infectious disorders.

We report a term neonate with prominent neurological dysfunction, bilateral sensorineural hearing loss, patent ductus arteriosus and rubella-specific serological evidence. Neuroimaging demonstrated bilateral basal ganglia and thalamic T1 hyperintensity with mild subdural haemorrhage, while ophthalmological examination was normal. This case highlights the variable clinical expression of CRS and the importance of multisystem evaluation when the classical triad is incomplete.

## CASE PRESENTATION

A term male neonate with a birth weight of 2.75 kg was born by lower-segment caesarean section for breech presentation to parents in a second-degree consanguineous marriage. Apgar scores were 7/10 at 1 minute and 9/10 at 5 minutes.

During the neonatal period, the child developed a high-pitched cry, persistent jitteriness, poor oral feeding, hypotonic suck and an absent Moro reflex. Examination showed craniofacial dysmorphism in the form of frontal bossing, a depressed nasal bridge and a high-arched palate. Systemic findings included hepatomegaly, stridor, excessive mucoid oral secretions and desquamation of the palms and soles. Neurological examination revealed central hypotonia with appendicular hypertonia, hyporeflexia and weak primitive reflexes.

Laboratory evaluation showed mild anaemia without thrombocytopenia. Liver function testing demonstrated mild hyperbilirubinaemia, with normal aspartate aminotransferase, alanine aminotransferase, albumin and globulin levels. Serum electrolytes and arterial blood gas parameters were within normal limits. TORCH IgG serology showed a positive rubella IgG level of 54.8 IU/mL, positive cytomegalovirus IgG of 19.3 AU/mL, positive herpes simplex virus 1 and 2 IgG with a ratio of 1.543, and negative toxoplasma IgG. The available clinical records also documented rubella-specific IgM positivity.

**Table 1. Torch Serology Profile**

| Test parameter | Result | Unit | Laboratory interpretation |
|---|---|---|---|
| Toxoplasma IgG | 0.01 | IU/mL | Non-reactive |
| **Rubella IgG** | 54.8 | IU/mL | Positive |
| Cytomegalovirus (CMV) IgG | 19.3 | AU/mL | Positive |
| Herpes simplex virus 1 and 2 IgG | 1.543 | Ratio | Positive |

Neurosonography demonstrated bilateral subdural collections with thickening over the cerebral hemispheres. Magnetic resonance imaging of the brain showed T1 hyperintensity involving the bilateral basal ganglia and thalami, with mild subdural haemorrhage and no diffusion restriction. Echocardiography revealed a small patent ductus arteriosus with a left-to-right shunt, normal biventricular function and no pulmonary hypertension.

Auditory brainstem response testing demonstrated bilateral moderate-to-moderately severe sensorineural hearing loss, with reduced response amplitude in the right ear. Ophthalmological examination was normal.

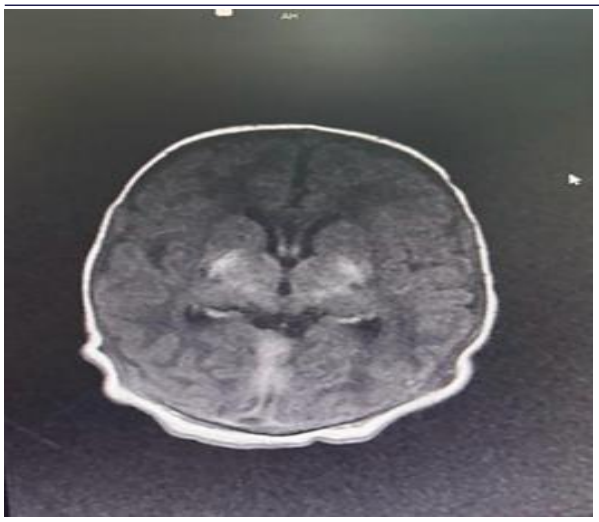

**Figure 1. Brain MRI showing T1 hyperintensity involving the bilateral basal ganglia and thalami. Mild subdural haemorrhage was also documented, with no diffusion restriction.**

The neonate received levetiracetam and clonazepam for seizure control, with monitoring for breakthrough seizures. Feeding support consisted of exclusive breastfeeding or expressed breast milk administered through an orogastric tube or by spoon, together with continued oromotor exercises. Physiotherapy and sensory-stimulation exercises were advised. Vitamin $D_3$ supplementation was provided. Follow-up was planned with paediatric neurology, the neurodevelopmental clinic, audiology, ophthalmology and paediatric cardiology.

**Table 2. Clinical And Investigative Profile of the Neonate**

| Domain | Documented findings |
|---|---|
| Perinatal details | Term male neonate; birth weight 2.75 kg; LSCS for breech presentation; Apgar 7/10 and 9/10 |
| Family background | Second-degree consanguineous marriage |
| Presenting features | High-pitched cry, jitteriness, poor feeding, hypotonic suck and absent Moro reflex |
| Dysmorphic features | Frontal bossing, depressed nasal bridge and high-arched palate |
| Systemic findings | Hepatomegaly, stridor, excessive mucoid oral secretions and palmoplantar desquamation |
| Neurological findings | Central hypotonia with appendicular hypertonia, hyporeflexia and weak primitive reflexes |
| Haematology and biochemistry | Mild anaemia without thrombocytopenia; mild hyperbilirubinaemia; normal AST, ALT, albumin, globulin, electrolytes and ABG |
| Serology | Rubella IgG 54.8 IU/mL; CMV IgG 19.3 AU/mL; HSV-1/2 IgG ratio 1.543; toxoplasma IgG negative; rubella-specific IgM positivity documented |
| Neurosonography | Bilateral subdural collections with thickening over the cerebral hemispheres |
| Brain MRI | Bilateral basal ganglia and thalamic T1 hyperintensity; mild subdural haemorrhage; no diffusion restriction |
| Echocardiography | Small PDA with left-to-right shunt; normal biventricular function; no pulmonary hypertension |
| Auditory assessment | Bilateral moderate-to-moderately severe sensorineural hearing loss; reduced response amplitude in the right ear |
| Ophthalmology | Normal examination |
| Management | Levetiracetam, clonazepam, feeding support, oromotor exercises, physiotherapy, sensory stimulation and vitamin D3 |
| Follow-up | Paediatric neurology, neurodevelopment, audiology, ophthalmology and paediatric cardiology |

## DISCUSSION

Congenital rubella syndrome is characterised by considerable phenotypic variability, and affected neonates may not demonstrate the complete classical triad of hearing impairment, ocular abnormalities and congenital heart disease. In the present case, bilateral sensorineural hearing loss and a patent ductus arteriosus provided two characteristic manifestations of CRS, despite a normal ophthalmological examination. The associated hepatomegaly, mild hyperbilirubinaemia and neurological abnormalities further supported multisystem involvement. Recent reviews emphasise that the absence of cataract or another component of the classical triad does not exclude CRS (2).

The neurological presentation was prominent, with a high-pitched cry, jitteriness, poor feeding, hypotonic suck, absent Moro reflex, central hypotonia, appendicular hypertonia, hyporeflexia and weak primitive reflexes. Neurological involvement in CRS may include microcephaly, meningoencephalitis, developmental impairment, abnormalities of tone, seizures and later behavioural or cognitive difficulties. Because these findings are not specific to rubella, they should be interpreted together with cardiac, auditory, ophthalmological and laboratory evidence rather than used independently to establish the diagnosis (4).

Neuroimaging demonstrated bilateral basal ganglia and thalamic T1 hyperintensity, mild subdural haemorrhage and absence of diffusion restriction. Congenital infections can produce a broad range of neuroimaging abnormalities, including intracranial calcification, white-matter injury, ventriculomegaly, cortical malformations and destructive lesions. Nevertheless, no single imaging pattern is diagnostic of CRS, and imaging findings must be correlated with the clinical phenotype and laboratory results (5). Bilateral basal ganglia and thalamic abnormalities also have a wide differential diagnosis that includes hypoxic-ischaemic injury, bilirubin-related injury, metabolic or toxic disorders, infection and haemorrhage. The absence of diffusion restriction in this neonate is therefore relevant, but the observed T1 hyperintensity should not be attributed exclusively to rubella without clinical and biochemical correlation (6). Similarly, the mild subdural haemorrhage may represent an associated perinatal finding rather than a recognised defining manifestation of CRS.

The small patent ductus arteriosus with a left-to-right shunt was clinically important because patent ductus arteriosus and pulmonary artery abnormalities are among the most characteristic cardiovascular defects associated with CRS. Indian surveillance data have shown that congenital cardiac abnormalities are frequent among laboratory-confirmed cases, with patent ductus arteriosus forming a substantial proportion of the documented lesions. Cardiac assessment and continued follow-up are necessary even when the initial shunt is small and ventricular function is preserved (7).

Auditory brainstem response testing demonstrated bilateral moderate-to-moderately severe sensorineural hearing loss, with reduced response amplitude in the right ear. Hearing impairment is one of the most frequent manifestations of CRS and may occur in isolation or in combination with cardiac and ocular abnormalities. Its severity may be variable, and early audiological surveillance is important because delayed intervention can adversely affect speech, language and neurodevelopmental outcomes (8). The normal ophthalmological examination in this child does not eliminate the need for continued monitoring because some ocular and developmental complications may become apparent later.

Serological interpretation requires particular care in neonates. The positive rubella IgG level indicates exposure to rubella-specific antibody but, when measured during early infancy, may reflect passive transplacental transfer of maternal IgG. In contrast, detection of rubella-specific IgM in the infant supports congenital infection because maternal IgM does not cross the placenta. Laboratory confirmation may also be established by detection of rubella viral RNA, virus isolation, or persistence of rubella IgG beyond the expected period of passive maternal antibody decline (1). The documented rubella-specific IgM positivity, together with sensorineural hearing loss and patent ductus arteriosus, supported the diagnosis in this case. However, the timing and source of the serological sample remain essential considerations when interpreting the results.

Management of CRS is supportive and directed towards the affected organ systems, as no antiviral treatment reverses established congenital injury. This neonate required antiseizure medication, assisted feeding, oromotor exercises, physiotherapy and sensory stimulation. Long-term care should include neurological and developmental assessment, repeated audiological evaluation,

ophthalmological surveillance and cardiac monitoring. The case highlights that CRS should remain a diagnostic consideration in a neonate with unexplained neurological dysfunction when hearing impairment or congenital heart disease is also present, even in the absence of the complete classical triad.

## CONCLUSION

Congenital rubella syndrome may present with prominent neurological dysfunction even when the complete classical triad is absent. In this neonate, the combination of bilateral sensorineural hearing loss, patent ductus arteriosus and documented rubella-specific serological findings supported the diagnosis, while the bilateral basal ganglia-thalamic abnormalities required careful differential interpretation. Early recognition and coordinated neurological, developmental, auditory, ophthalmological and cardiac follow-up are essential to reduce preventable long-term disability.

### Authors' Contributions

Dr GL and Dr DS conceptualized and conducted the study, collected data, and prepared the manuscript. Both authors contributed to data interpretation, manuscript revision, and approved the final version.

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**Data Availability Statement:** Data supporting the findings of this study are available from the corresponding author upon reasonable request.

## REFERENCES

1. World Health Organization. Congenital rubella syndrome: vaccine-preventable diseases surveillance standards. Geneva: World Health Organization; 2018. Available from: https://cdn.who.int/media/docs/default-source/immunization/vpd_surveillance/vpd-surveillance-standards-publication/who-surveillancevaccinepreventable-03-crs-r2.pdf
2. De Melo LC, Rugna MM, Durães TA, Pereira SS, Callado GY, Pires P, et al. Congenital rubella syndrome in the post-elimination era: why vigilance remains essential. J Clin Med. 2025;14(11):3986. doi: 10.3390/jcm14113986.
3. Murhekar M, Verma S, Singh K, Bavdekar A, Benakappa N, Santhanam S, et al. Epidemiology of congenital rubella syndrome in India, 2016–18, based on data from sentinel surveillance. PLoS Negl Trop Dis. 2020;14(2):e0007982. doi:10.1371/journal.pntd.0007982.
4. Fortin O, Mulkey SB. Neurodevelopmental outcomes in congenital and perinatal infections. Curr Opin Infect Dis. 2023;36(5):405–413. doi: 10.1097/QCO.0000000000000946.
5. Lucignani G, Guarnera A, Rossi-Espagnet MC, Moltoni G, Antonelli A, Figà Talamanca L, et al. From fetal to neonatal neuroimaging in TORCH infections: a pictorial review. Children (Basel). 2022;9(8):1210. doi: 10.3390/children9081210.
6. Van Cauter S, Severino M, Ammendola R, Van Berkel B, Vavro H, Van den Hauwe L, et al. Bilateral lesions of the basal ganglia and thalami (central grey matter): pictorial review. Neuroradiology. 2020;62(12):1565–1605. doi: 10.1007/s00234-020-02511-y.
7. Gunasekaran PK, Shanmugasundaram D, Santhanam S, Verma S, Singh K, Dwibedi B, et al. Profile of cardiac lesions among laboratory-confirmed congenital rubella syndrome infants: a nationwide sentinel surveillance, India, 2016–22. Lancet Reg Health Southeast Asia. 2023;16:100268. doi: 10.1016/j.lansea.2023.100268.
8. Korver AMH, Smith RJH, Van Camp G, Schleiss MR, Bitner-Glindzicz MAK, Lustig LR, et al. Congenital hearing loss. Nat Rev Dis Primers. 2017;3:16094. doi: 10.1038/nrdp.2016.94.