

## Reversible sensorineural hearing loss in a girl with Kawasaki disease

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Kawasaki disease is an acute, self-limited vasculitis of infants and children that is now the most common cause of acquired heart disease in the pediatric age group in the United States and Japan. This report presents a case of classic Kawasaki disease with reversible sensorineural hearing loss that was treated with steroid. In conclusion, Kawasaki disease caregivers must be aware of possible sensorineural hearing loss, which is reversible by early intervention and thus improves quality of life.

*Key words:* Kawasaki disease, sensorineural hearing loss, steroid.

Kawasaki disease (KD) is an acute systemic self-limited vasculitis, which was first described in the 1960s in Japan by Tomisaku Kawasaki<sup>1</sup>. KD is characterized by fever, bilateral non-exudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash and cervical lymphadenopathy. The classic diagnosis of KD has been based on the presence of five or more days of fever and at least four of the five principle clinical features<sup>2</sup>.

Although coronary artery involvement is the most important organ manifestation of KD, a variety of other features are also characteristic of this syndrome, including aseptic meningitis, arthralgia and arthritis, urethritis, anterior uveitis, mild hepatobiliary dysfunction and gallbladder hydrops<sup>3</sup>. Furthermore, reports from Japan and the United States have documented the association of sensorineural hearing loss with acute KD<sup>3-6</sup>. These reports document more than 30 patients in whom hearing loss ranged from mild and transient to profound and permanent.

Diagnosis of KD relies on clinical criteria, and presence of associated organ manifestation could be a supporting feature. In this article, a case of KD with hearing loss that responded to dextran and steroid treatment is presented.

### Case Report

An eight-year-old female was transferred to Hacettepe University İhsan Doğramacı Children's Hospital with prolonged fever, rash and unilateral hearing loss. On the third day of fever, she had been admitted to a local health center with fever, sore throat and photophobia. She was initially diagnosed as pharyngitis and treated with amoxicillin. Because of continuous fever she was admitted to a local hospital on the fifth day of fever, and urine analysis revealed leukocyturia, and she was treated with ceftriaxone (intramuscular-im). The next day she developed polymorphous erythematous rash, non-exudative conjunctivitis, red lips and a strawberry tongue and erythema of her hands and feet with hearing impairment. A diagnosis of KD was made and the patient was given intravenous immunoglobulin (IVIG) and high-dose aspirin (80 mg/kg per day). After completing a full 2 g/kg dose of IVIG, her hearing impairment progressed to indiscrimination of speech. The patient was then transferred to Hacettepe University İhsan Doğramacı Children's Hospital on the eighth day of fever. At admission her temperature was 36.8°C, her speech discrimination was poor, and clinical examination showed normal

tympanic membranes and a positive Rinne test in both ears. An echocardiogram showed dilation on the right and left coronary arteries (4 and 6 mm, respectively). Laboratory studies showed a normal hemoglobin (13.1 mg/dl) and thrombocytosis 370,000/mm<sup>3</sup>. Pure tone audiometry showed sensorineural hearing loss which was confirmed by evoked response audiometry (Fig. 1). Her cranial magnetic resonance imaging study for sensorineural hearing loss was normal. She was given steroid (Delta Cortril 30 mg per day), dextran 40 10% (150 ml infusion over 6 hours/day), betahistine (8 mg, 3 times per day) and aspirin (2 mg/kg per day). Within five days her hearing improved and the steroid treatment was stopped gradually and the dextran treatment was discontinued. The patient remained afebrile over the hospitalization period and at the one-month follow-up, evoked response and pure tone audiometry had improved (Fig. 2) and echocardiography revealed left and right coronary enlargement with a distal 4.3 mm and 3.9 mm aneurysms. Betahistine therapy was stopped and she remained on aspirin.

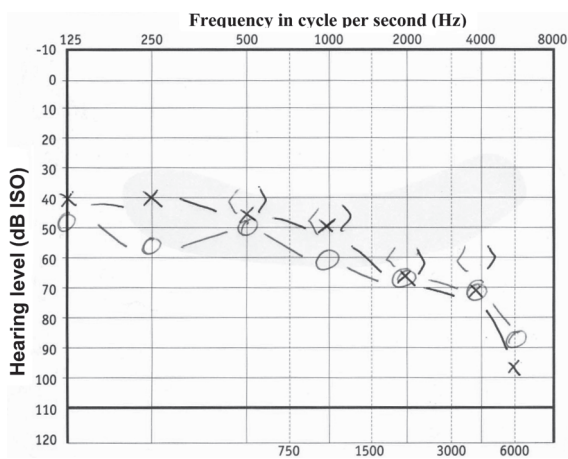


Fig. 1. Initial evaluation of pure tone audiometry.

X — X Left ear hearing threshold (air).  
 O — O Right ear hearing threshold (air).  
 ] Left ear hearing threshold (bone).  
 [ Right ear hearing threshold (bone).

## Discussion

Kawasaki disease is now known to occur in both endemic and community-wide epidemic forms in Europe, Americas and Asia in children of all races. Neurologic symptoms are very rare, although extreme irritability, possibly due to

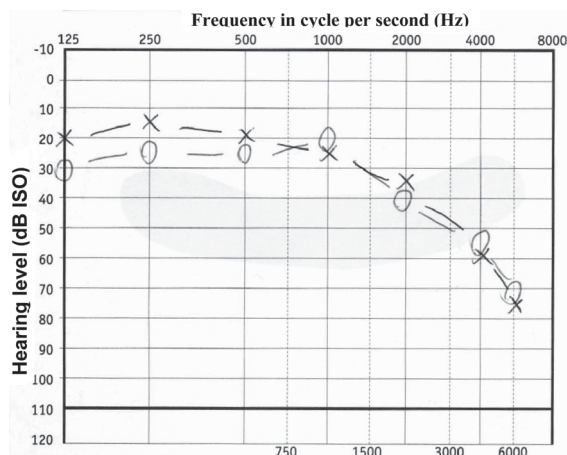


Fig. 2. Pure tone audiometry after treatment.

X — X Left ear hearing threshold (air).  
 O — O Right ear hearing threshold (air).  
 ] Left ear hearing threshold (bone).  
 [ Right ear hearing threshold (bone).

aseptic meningitis, is common in KD. Seizures, involvement of cranial pairs and hemiparesis caused by thrombosis or infarction are rarely reported in the literature<sup>4,5</sup>. We observed another kind of neurologic manifestation that was severe but likely reversible, sensorineural hearing loss.

Sensorineural hearing loss is a well-recognized condition of variable etiology, and it is also associated with systemic immune diseases such as rheumatoid arthritis, polyarteritis nodosa, Wegener's granulomatosis, giant cell arteritis and inflammatory bowel disease<sup>7</sup>. The etiology of KD remains unknown, although clinical and epidemiological features strongly suggest an infectious cause<sup>2</sup>. The sensorineural hearing loss associated with KD might then reflect infection of the inner ear, analogous to the direct cytopathic effects on the labyrinth and cochlea that occur with certain viral illness, such as mumps and rubella<sup>8</sup>. Alternatively, the sensorineural hearing loss may be associated with aberrant immune activation<sup>9</sup>. Hence, the autoimmune inner ear disease may occur due to the loss of immunological tolerance to self antigens, cross-reacting antibodies, self antigen modification, autoimmunity due to hidden self antigen or circulating immune complexes<sup>9</sup>. Though there is no doubt that association between sensorineural hearing loss and KD exists, the responsible mechanism remains unclear.

In the reported cases in the literature, there was speculation regarding salicylate ototoxicity in the genesis of hearing loss. Although most patients with hearing loss have been treated with aspirin, it is unlikely that the condition is caused by the use of this medication, since the serum levels of aspirin almost always remained under toxic levels<sup>3-6,10</sup>. In addition, in our case, hearing loss had developed before initiation of aspirin therapy.

In most of the reported cases, hearing loss was permanent, and the time between diagnosis of KD and the perception of hearing loss ranged from 10 days to 5 years<sup>4,5,10</sup>. However, Knott<sup>6</sup> reported that there were nearly 30 cases of sensorineural hearing loss in the first 30 days of fever, and two in the follow-up. In our case, the patient's age was the most important factor for early perception of hearing loss; the immediate treatment with steroid could be the key factor for our favorable outcome.

Spectrum of KD manifestation is so variable that in every stage of the disease, caregivers should be aware of unpredictable manifestations or complications. We would like to emphasize the importance of immediate detection of sensorineural hearing that is possibly treatable with steroid, with or without immunosuppressive therapy, to preserve auditory function, because sudden loss of hearing that may result in deafness, especially in a young patient, results in severe disability.

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