

Recurrence of Kawasaki Disease: Look Before you Leap on the Bandwagon

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Verma and colleagues reported an interesting case of recurrent fever of immunological origin [1]. The first episode probably favors a diagnosis of Kawasaki disease because of the presence of coronary artery dilatation, which normalized with anti-inflammatory therapy. During the second episode, the child had high-grade fever with oral mucosal changes and cervical lymphadenopathy with no identifiable infective cause on basic laboratory evaluation. The coronary arteries were normal. The rise in acute phase reactants was not very impressive. It seems reasonable to consider Kawasaki disease among the differential diagnosis of this condition, but the following issues need reconsideration:

1. A lot of emphasis has been given to skin desquamation. Episodes of skin desquamation during febrile episodes have been reported even beyond 5 years from the initial episode of Kawasaki disease [2]. Hence caution should be exercised in considering skin desquamation in diagnosing disease recurrence.
2. Recurrent fevers with features of generalized inflammation after Kawasaki disease might represent patients with a genetic predisposition to inflammatory disorder [3].
3. Skin desquamation cannot be considered pathognomonic of Kawasaki disease. It is a feature of many infectious and immunological diseases, including Scarlet fever [4] and Leptospirosis. The skin desquamation in Kawasaki disease is generally limited to the palms and soles and severe peeling in sheets occurs only in a small proportion of patients.
4. The lower risk of disease recurrence in India more likely points to the lack of proper disease reporting systems. Experience suggests that recurrence commonly occurs in children who suffered a more severe disease with coronary artery involvement at initial presentation [3]. The disease recurrence frequently results in additional coronary involvement.

This essentially highlights that the lack of a confirmatory laboratory test leaves us reliant on clinical criteria for diagnosis of Kawasaki disease. Recent work on the pathogenesis of the vasculitides gives hope that a solution for this complex problem may be on the way sooner than later.

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Recurrence of Kawasaki Disease: Authors' Reply

We agree that during the second episode, coronary arteries were normal and rise in acute phase reactants was not very impressive in our patient. However, our case satisfied the criteria of incomplete Kawasaki disease during recurrence [1,2].

1. In our case, peeling of skin of sole occurred 2-3 weeks after the acute illness, that further supported the diagnosis, and does not indicate that skin peeling was mandatory for diagnosing the Kawasaki disease.
2. Broderick, *et al.* [3] reported recurrent fever in four patients with a history of Kawasaki disease. They had periodic fever occurring at regular intervals (2-6 weeks) and two of them were also having aphthous stomatitis. Fever was associated with rash in two children, but skin desquamation was missing in all these children with recurrent fever. In our child, there was no such history. However, we agree that recurrent fever syndromes should be considered, and needs to be excluded before labelling the case as recurrent Kawasaki disease.
3. In scarlet fever, the skin may start to peel even during the febrile stage. This peeling starts cephalo-caudally, and lastly palms and soles. In Kawasaki disease, sheet like skin peeling is characteristically limited to the palms and soles, as in our case. We agree that skin

desquamation is not pathognomonic of Kawasaki disease.

4. It is true that disease recurrence results in additional coronary artery involvement, but we had administered intravenous immunoglobulins early in the course of disease that probably prevented coronary artery involvement [4].

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Guaranteeing the Volume Guarantee Ventilation

Atelecto-trauma resulting from varying tidal volumes is the key factor resulting in ventilation associated lung injury and thereby causing chronic lung disease [1]. The Volume guarantee ventilation (Babylog), a form of volume targeted ventilation, has been documented to reduce chronic lung disease [2]. However, there is a caution while using this form of ventilation.

Volume guarantee ventilation is a time-cycled pressure – limited volume –targeted ventilation. The edge of Volume guarantee over other forms of ventilation is ensuring the target tidal volume, and auto weaning. The problem arises in this mode if the neonate has severe metabolic acidosis. The neonate is vigorous and has high spontaneous breath rates, due to acidosis, overshooting the back-up rates. As a result, the tidal volumes during these spontaneous breaths are higher or equal to the set tidal volume, and the working peak inspiratory pressure (PIP) is closer to the positive end expiratory pressure (PEEP) making the mode as ventilator continuous positive airway pressure (CPAP). The general condition would not allow to extubate to CPAP and the blood gases are also acceptable making it more difficult to raise the tidal volumes. We try to overcome this situation by switching off to synchronized intermittent mandatory ventilation (SIMV) mode keeping a difference between PIP and PEEP so that the delta P is maintained.

Allowing ventilator to auto-wean could lead to problems, if the flow sensor is blocked or is disturbed, resulting in higher tidal volume, VALI and inadvertent hypocarbia; hence, the flow sensor needs to be in efficient working condition. Auto-triggering owing to high leak may also result in hypocarbia.

Although, the Babylog recommends coupling the Volume guarantee with SIMV, Synchronized intermittent positive pressure ventilation (SIPPV) or Patient support ventilation (PSV); Volume guarantee should not be coupled with SIMV as the unsupported breaths would have varying tidal volumes, and could result in atelectotrauma.

Volume guarantee mode is a robotic mode which operates automatically rendering us interfering least, except for fraction inspired oxygen. The modality needs efficient understanding by the operator, and if used cautiously is a state-of-art. However, it could be double edged sword if the issues mentioned above are not looked into carefully, especially in a busy unit.

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