Reviewer's report

**Title:** CANDLE SYNDROME: unreported orofacial features and dental implications

**Version:** 4  
**Date:** 12 March 2015

**Reviewer:** THASCHAWEE TA ARKACHAISRI

**Reviewer’s report:**

The authors report their observation of oro-facial and dental abnormalities in a South African girl who was claimed to have CANDLE syndrome. It, indeed, is the first time of these manifestations was reported. Certain clarifications are needed to increase the reliability and accuracy of the case report. I have concerns requiring modification or clarification as follows:

**Major Compulsory Revisions**

1. The case used in this paper will need to be more solid. The age of the patient is important along with onset of what symptoms or signs that brought the patient to medical attention. This is a congenital genetic inflammatory disease; I do expect a full manifestation of inflammation including recurrent fever, failure to thrive (provide weight and height percentile), lipodystrophy, recurrent neutrophilic dermatitis rash among others less commonly described including CNS, endocrine and rheumatologic manifestations. The phenotype (both physical signs and typical laboratory results) of this girl must be laid out clearly in order to increase the diagnostic accuracy.

2. Since genetic test to determine the genetic mutation of the PSMB8 gene is available, the authors will need to confirm the diagnosis with this investigation to make the case stronger.

3. Medical management since diagnosis may have certain effects on the dentition. The effect of chronic use of corticosteroids on oro-facial development and dentition will need to be discussed in detail.

4. Not only CANDLE and JMP (Joint contractures, Muscle atrophy, microcytic anemia, and Panniculitis-induced childhood-onset lipodystrophy) syndrome, but also Nakajo-Nishimura syndrome (NNS) are considered to be the same disease with continuum spectrum of the overall same immunoproteasome-associated autoinflammatory disorder. The authors will need to add this comment and references accordingly.

**Minor Essential Revisions**

1. Multiple typos were noted including the title itself and throughout the paper, please correct accordingly.

**Level of interest:** An article of limited interest
Quality of written English: Needs some language corrections before being published

Declaration of competing interests:

I declare that I have no competing interests