Tentative diagnostic criteria for Nakajo-Nishimura Syndrome

[Kanazawa N., Allergol Int 2012;61:197-206. Accessible at:
http://ai.jsaweb.jp/fulltext/061020197/061020197_index.html
Kanazawa N. et al., Jpn J clin Immunol 2011;34 :388-400. Accessible at:
https://www.jstage.jst.go.jp/article/jsci/34/5/34_5_388/_article
(in Japanese)]

A clinical diagnosis of Nakajo-Nishimura Syndrome can be made if at least 5 of the following 8 features are positive:

1. Autosomal recessive inheritance (parental consanguinity and/or familial occurrence)
2. Pernio-like purplish rash in hands and feet (appearing in winter since infancy)
3. Haunting nodular erythema with infiltration and induration (sometimes circumscribed)
4. Repetitive spiking fever (periodic, not necessarily)
5. Long clubbed fingers and toes with joint contractures
6. Progressive partial lipomuscular atrophy and emaciation (marked in the upper part of body)
7. Hepatosplenomegaly
8. Basal ganglia calcification