

Letter to the Editor

Management of transfusion-dependent thalassemia patients in emergency situations

Yoshihiro Aoki, MD,^a Chris Smith, MD, PhD,^{a,b} Koya Ariyoshi, MD, PhD.^{a,c}

^a School of Tropical Medicine and Global Health, Nagasaki University, Nagasaki, Japan.

^b Department of Clinical Research, London School of Hygiene and Tropical Medicine Faculty of Infectious and Tropical Diseases, London, UK

^c Department of Clinical Medicine, Institute of Tropical Medicine, Nagasaki University, Nagasaki, Japan

Corresponding author:

Yoshihiro Aoki, MD

School of Tropical Medicine and Global Health, Nagasaki University

1-12-4 Sakamoto, Nagasaki-city, Nagasaki 852-8523, JAPAN

Email: yaoki-hki@umin.ac.jp / TEL: +81- 95-819-7008 / FAX: +81-95-819-7892

Parakh et al. recently conducted a prospective observational study on the impact of the Covid-19 pandemic and the associated lockdown on the care of transfusion-dependent thalassemia (TDT) patients in North India.¹ They reported significantly lower mean pre-transfusion hemoglobin levels during and post-lockdown compared to pre-lockdown. They also described the countermeasures of the thalassemia outpatient clinic to address the various issues of Covid-19. Similar difficulties in clinic accessibility and declined donor blood can occur outside of the Covid-19 pandemic, such as in conflict settings.² However, the impact of these disruptions on long-term clinical outcomes of thalassemia patients is not clear. Further consideration should be given to potential harm as a consequence of treatment disruption for TDT patients.

Whether a pre-transfusion hemoglobin target of 9-10.5 g/dL is optimal for thalassemia treatment in emergencies is debatable. Maintenance of a certain hemoglobin level is vital in TDT patients because anemia contributes to extramedullary hematopoiesis, exacerbates splenomegaly, and increases hemolysis. On the other hand, hypertransfusion maintaining baseline hemoglobin at 10-12 g/dL contributes to iron overload and significantly affects delayed puberty, so moderate transfusion aiming for 9-10 g/dL has been considered better without causing extramedullary hematopoiesis.³ However, when considering population outcomes in situations of insufficient

transfusion or increased iron loading due to interruption of chelation therapy, it may be acceptable to lower the pre-transfusion hemoglobin to a certain degree. A prospective study should be considered to evaluate the association between even temporary lower hemoglobin targets and clinical outcomes after the Covid-19 era.

It was also shown that many patients were forced to receive transfusion treatment at alternative clinics. Under such circumstances, it may be helpful not only to develop telemedicine and inter-hospital collaboration but also to prepare a patient-held record with treatment information such as transfusion schedules, the dose of iron chelators, and complications. The patient would take the record consultations, thus withstanding interruption of electrical communication. Prior information sharing among hospitals, including knowledge of general thalassemia patient care and the possibility of emergency consultation, is essential.

Although the development and introduction of hematopoietic stem cell transplantation and gene therapy are increasingly expected, many patients live in areas with few resources, making it challenging to provide these advanced treatments.⁴ Since sudden artificial or natural disasters and epidemic diseases can occur worldwide, we should share ideas on maintaining TDT management in emergencies. Clinical research

on thalassemia management from resource-limited settings would also help prepare for a future crisis.

References

1. Parakh N, Pahuja S, Singh V, Kumar N, Chandra J. Covid-19 pandemic and care of transfusion-dependent patients of thalassaemia: Experience from a paediatric centre in North India. *J Paediatr Child Health*. 2022.
<https://doi.org/10.1111/JPC.16082>.
2. Foley D, Aoki Y, Roggeveen H, Carrion-Martin A I, Mullahzada AW, Hoetjes M. The forgotten frontline... Improving thalassaemia outcomes with iron chelation therapy in a conflict setting. 2019.
<https://doi.org/10.13140/RG.2.2.24015.53925>.
3. Cazzola M, Borgna-Pignatti C, Locatelli F, Ponchio L, Beguin Y, De Stefano P. A moderate transfusion regimen may reduce iron loading in beta-thalassemia major without producing excessive expansion of erythropoiesis. *Transfusion*. 1997; **37**: 135–40.
4. Kattamis A, Kwiatkowski JL, Aydinok Y. Thalassaemia. *Lancet* 2022; **399**: 2310–24.

Acknowledgements: Not applicable.

Conflict of Interests: The authors have no conflicts of interest to declare.

Funding sources: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethics approval: Not applicable.

Consent: Not applicable.