Effect of transfusion, iron chelation and splenectomy therapies in HbE/β-thalassaemia individuals in Malaysia

Wai Feng Lim[1,2], Logeswaran Muniandi [1], Elizabeth George[3], Jameela Sathar[4], Lai Kuan Teh[5] and Mei I Lai[1,6*]

[1]Department of Pathology, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, 43400 UPM Serdang, Selangor, Malaysia
[2]Integrative Pharmacogenomics Institute (iPROMISE), Universiti Teknologi MARA Selangor, 42300 Bandar Puncak Alam, Selangor, Malaysia
[3]Assunta Hospital, 46990 Petaling Jaya, Selangor, Malaysia
[4]Department of Haematology, Ampang Hospital, 68000 Ampang, Selangor, Malaysia
[5]Department of Biomedical Science, Faculty of Science, Universiti Tunku Abdul Rahman, 31900 Kampar, Perak, Malaysia
[6]Genetics & Regenerative Medicine Research Centre, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, 43400 UPM Serdang, Selangor, Malaysia

HbE/β-thalassaemia is a compound heterozygous mutation with a vast clinical phenotype [1]. To improve quality of life, HbE/β-thalassaemia individuals receive different treatment strategies, either individually or in combination with therapy(ies), including blood transfusion, iron chelation and splenectomy [2-3]. Thus far, there are limited studies conducted regarding the effect of treatments in HbE/β-thalassaemia individuals. We hereby investigated the effect of treatments with respect to red blood cell indices, haemoglobin subtypes and gene expressions among 30 HbE/beta-thalassaemia individuals. Statistical analyses were carried out using SPSS 17.0. As compared to single therapy (transfused only individuals) and double therapies (transfused-chelated only individuals), individuals receiving triple therapies (transfused-chelated-splenectomised individuals) showed significantly high mean cell volume (MCV), mean cell haemoglobin (MCH) and reticulocytes count (Fig.1).

These findings suggest that triple therapies are the most effective in ameliorating the severity of the disease in terms of microcytosis and hypochromia [3-5]. The high reticulocyte count in triple therapies also allows the bone marrow to actively produce red blood cells suggesting that these therapies have clinical benefits by suppressing the ineffective erythropoiesis and improving the erythropoietic environment significantly among HbE/β-thalassaemia individuals in our studied group [6-7].

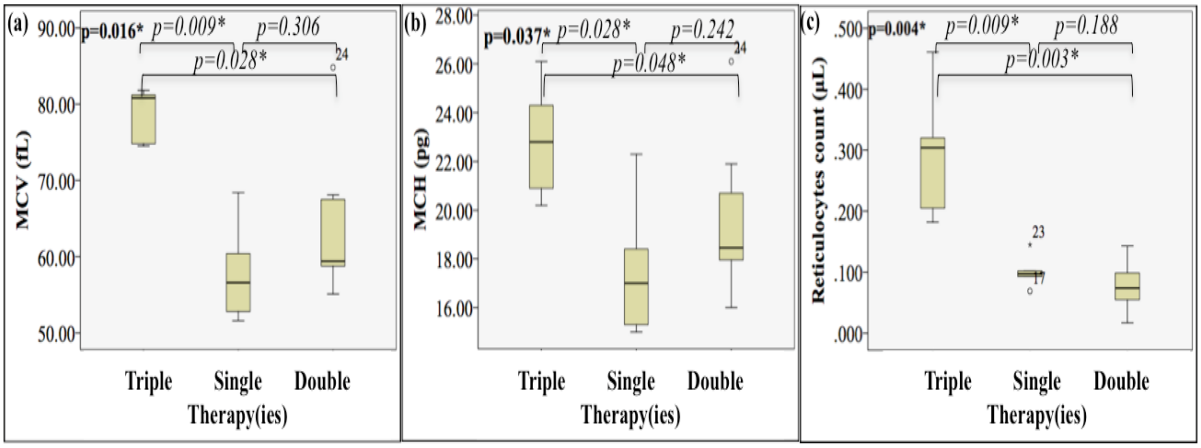

Fig. 1: Correlation of therapies received to clinical and laboratory data in HbE/β-thalassaemia individuals. Abbreviation: Single therapy (transfused only), double therapies (transfused-chelated only) and triple therapies (transfused-chelated-splenectomised). Correlation of (a) mean cell volume, MCV (p=0.016*); (b) mean cell haemoglobin, MCH (p=0.037*); and (c) reticulocyte count, (p=0.004*). Other variables showed no significant differences (data not shown).

The effectiveness of these treatments is different among each HbE/β-thalassaemia individual whereby clinical variabilities among them could be a contributing factor. Triple therapies giving the best advantage to the HbE/β-thalassaemia patient in this study.

Keywords: transfusion, iron chelation, splenectomy, HbE/β-thalassaemia

*** Correspondence:** lmi@upm.edu.my

Acknowledgements
This study was funded by the Fundamental Research Grant Scheme, Ministry of Higher Education, Malaysia (03-1-07-324FR and 04-04-10-837FR) awarded to Mei I Lai.

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