

Association of the degree of erythroid expansion and maturation arrest with the clinical severity of β^0 -thalassemia/HbE patients

Supplemental Table 1. β -Thalassemia clinical severity scoring system.

Supplemental Table 1.1 Clinical criteria, points scored and severity group according to Sripichai O, *et al*, 2008 [15].

Clinical criteria	Points Scored			
	0	0.5	1	2
Hemoglobin at steady state (g/dl)	>7.5		6 – 7.5	<6
Age at thalassemia presentation (year)	>10	3 – 10	<3	
Age at receiving first blood transfusion (year)	>10		5 – 10	<5
Requirement for blood transfusion	None/rare		Occasional	Regular
Size of spleen (cm)	<3		3 – 10	>10 or splenectomized
Growth and development	>25th percentile	3rd–25th percentile	<3rd percentile	
Severity category	Total score			
Mild	0 – 3.5			
Moderate	4 – 7			
Severe	7.5 – 10			

Supplemental Table 1.2 Clinical parameter and severity group according to Sripichai O, *et al*, 2008 [15].

Patients	Hemoglobin at steady state		Age at thalassemia presentation		Age at receiving first blood transfusion		Requirement for blood transfusion		Size of spleen		Growth and development		Total severity score	Disease group
	g/dl	Score	year	Score	year	Score	frequency	Score	cm	Score	percentile	Score		
BE1	8.2	0	6	0.5	50	0	Rare	0	2	0	>P50	0	0.5	Mild
BE2	8.1	0	10	0.5	Never	0	Never	0	0	0	>P50	0	0.5	Mild
BE3	8.2	0	4	0.5	Never	0	Never	0	8	1	P10-P25	0.5	2.0	Mild
BE4	9.1	0	12	0	12	0	Rare	0	0	0	>P50	0	0.0	Mild
BE5	9.3	0	1	1	Never	0	Never	0	5	1	P25-P50	0	2.0	Mild
BE6	8.2	0	<1	1	4	2	Occasional	1	12	1	<P3	1	6.0	Moderate
BE7	6.0	1	1	1	20	0	Regular	2	Splenectomized	2	P25-P50	0	6.0	Moderate
BE8	7.1	1	<1	1	Never	0	Never	0	10	1	<P3	1	4.0	Moderate
BE9	7.0	1	2	1	8	1	Rare	0	9	1	P25-P50	0	4.0	Moderate
BE10	6.7	1	7	0.5	13	0	Occasional	1	16	2	P25-P50	0	4.5	Moderate
BE11	5.2	2	2	1	4	2	Regular	2	5	1	>P50	0	8.0	Severe
BE12	6.2	1	<1	2	3	2	Regular	2	Splenectomized	2	<P3	1	10.0	Severe
BE13	6.0	1	1	1	1	2	Regular	2	Splenectomized	2	<P3	1	9.0	Severe
BE14	6.6	1	3	0.5	4	2	Regular	2	12	2	<P3	1	8.5	Severe
BE15	7.5	1	2	1	3	2	Regular	2	4	1	P10-P25	0.5	7.5	Severe

Supplemental Table 2. List of primer for quantitative RT-PCR analysis.

Gene name	PCR forward primer	PCR reverse primer
ACVR2A	5'-GATACCCATGGACAGGTTGG-3'	5'-TCATCTACAGGTCCATCTGC-3'
ACVR2B	5'-CAACTGCTACGATAGGCAGG-3'	5'-TGGCTCGTACGTGACTTCC-3'
FOXO3	5'-TCTGCCGGCTGGAAGAACTC-3'	5'-CCACGGCTCTTGGTATACTTGTTG-3'
GDF11	5'-GATCTTGAGCAAAGTGC GGC-3'	5'-GTACTGCTGGGTCCGTCTC-3'
HSP70	5'-AGCTGGAGCAGGTGTGTAAAC-3'	5'-CAGCAATCTTGGAAGGCC -3'
TGFBR1	5'-ACGGCGTTACAGTGTTTCTG-3'	5'-GCACATACAAACGGCCTATCTC-3'
RPS18	5'-GGATGAGGTGGAACGTGT-3'	5'-CTAGGACCTGGCTGTATTTTC-3'