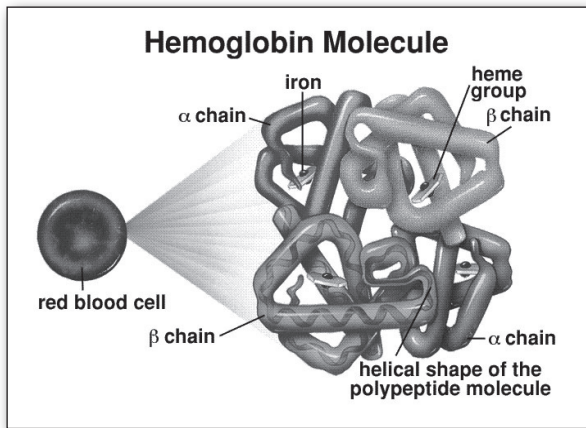




# ธาลัสซีเมียอินเตอร์มีเดีย : ปัจจัยทางพันธุศาสตร์ต่อเบต้าธาลัสซีเมีย Thalassemia Intermedia Genetics modifier of $\beta$ -thalassemia)

(red blood cell) (Hb) (Hb) (hemoglobin E Hb E)  
 (polypeptide) (heme group) (unstable)  
 2 (beta chain) (alpha chain) 1 (F chain)  
 (balance F and globin chain) (0-thal) (+-thal)

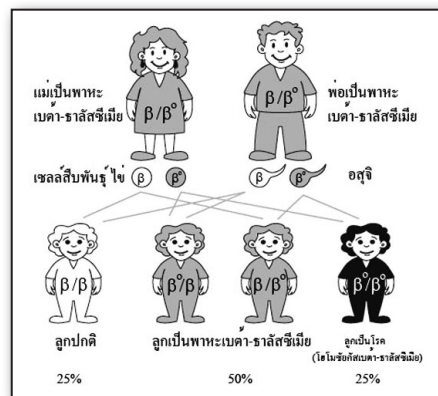


1 ( )

(Hb E)  
 (autosomal recessive) 2  
 (alleles) ( 2)  
 / ( 3)

(thalassemia)

thal  
2



2

(F-thalassemia)

(F<sup>0</sup>)

1(F-thal1)

(F<sup>+</sup>)

2

(-thal 2)

(-thalassemia)

(<sup>0</sup>-thal)

(<sup>+</sup>-

thal)



3

/



**5.6.6.2.1**

assaemia trait) 270 (carrier thal- 80 (coinheritance of F-thalassaemia)  
 ( -gene mutations) (Hereditary persistence of hemoglobin F, H<sub>PFH</sub>)

200 (malarial region)

fusion-dependent) (trans- (thalassaemia (severe) trait) (°)

major) (thalassaemia (homozygous be- ta-thalassaemia) / (°) 6 20

2 810 /

(+ ++)  
 1. (+/ + +/ ++ ++/ ++ ) /  
 2 (+/ E ++/ E)

(°) CBC  
 MCV (mean cell volume) 63±34 (femtoliter, fL)  
 MCV (+) 69.3±56 fL  
 ( ) HbA<sub>2</sub> 35-55%  
 ( HbA<sub>2</sub> <35%)  
 ( )

(genetic counseling)

(°)  
 Codons 41/42 (-TTCT), 17 (A-T),  
 71/72 (+A), 35 (CA)  
 (□+) -2BATA (A-G) Codon 19  
 (A-G)

1		
-	( )	<2
-	( / . )	67
- -		-
-	(%)	>50
-	HbA <sub>2</sub>	
-		° + ++
-	(H <sub>PFH</sub> )	
-	-	
-	G XMN1 Polymorphism*	

\* (G )

/ (+/ + +/ ++)  
 / (+/ E ++/ E)  
 + ++

1. Thain SL. Genetic modifiers of beta-thalassaemia. *Hematologica* 2005;90(5):649-60
2. Cappellini MD, Cohen A, Eleftheriou A, Piga A, Porter A and Taher A. Guidelines for the Clinical Management of thalassaemia 2<sup>nd</sup> edition, Nicosia-Cyprus Teamup Creations Ltd 2007 P 121-31.