TABLE 1. Forms of α-thalassaemia.

α-thalassaemias	Genotype (chromosome 16)	Designation	Clinical
Silent carrier (silent trait)	-α/αα	α^{+}/α	MCV normal, MCH normal Hb H inclusions (-) Hb A2 normal
α-thalassaemia trait	--/αα (cis or Asian type)	α^{0}/α	MCV normal/low, MCH normal/low
	-α/-α (trans or African type)*	α^{+}/α^{+}	Hb H inclusions + (present in a small percentage of red cells) Hb A2 normal/low
Hb H disease	--/-α (deletional) (75%)	α^{0}/α^{+}	MCV low, MCH low
	--/$\alpha^{T}\alpha$ (deletion/non-deletion) (25%)	α^{0}/α^{T}	Hb H inclusions +++
	--/α^{CS}a (deletion/non-deletion)	α^{0}/α^{CS}***	Hb A2 normal/low

* *The African type of α^{+}-thalassaemia (also known as $/\alpha^{3.7}$) is the mildest α-thalassaemia gene.*

** *The* **Hb H inclusions** *(β4) are made visible by methyl violet stain (like Heinz bodies). They are present in a very small percentage of red cells and they are more likely to be found in α^{0}/α than in α^{+}/α^{+} thalassaemia.*

*** **Hb Constant Spring** *is the product of a non-deletional α-thalassaemia gene ($/\alpha^{CS}\alpha$) typically found in South-East Asia. The occurrence of Hb CS is usually limited to the geographic area which includes Southern China and South East Asia. In 1968, Hb CS was also found to occur in the Mediterranean area where it was originally described as Hb Athens. It is caused by a point mutation in the stop codon of α2 gene (α2 is dominant over α1 in the production of α chains) resulting in the production of an elongated α chain which binds β chain to form Hb Constant Spring (which is very slow on Hb electrophoresis). The abnormal mRNA is very unstable so that only a small amount of Hb CS is produced; also Hb CS is itself unstable. The characteristic finding in Constant Spring disease (Hb H - Constant Spring, i.e. $\alpha^{0}/\alpha^{CS}\alpha$) is a moderate to severe thalassaemia with near normal MCV for example, MCV 72 fl versus 59 fl in deletional Hb H disease (α^{0}/α^{+}). The blood film in haemoglobin Constant Spring trait (Hb Constant Spring heterozygocity) shows prominent coarse basophilic stippling unlike other α-thalassaemias.*