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Title	:	A case series of α-thalassemia intermedia due to compound heterozygosity for Hb Adana [HBA2: C179G>A (or HBA1); P.Gly60Asp] with other α-thalassemias in malay families
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Abstract	:	Hb Adana [HBA2: c179G>A (or HBA1); p.Gly60Asp] is a rare hemoglobin (Hb) variant due to a mutation at codon 59 of the α2- or α1-globin gene resulting in a glycine to aspartic acid substitution. Two siblings with a unique coinheritance of Hb Adana and Hb Constant Spring (Hb CS, α142, Term→Gln, TAA>CAA; HBA2: c.427 T>C) (α codon 59α/αCSα), were compared phenotypically with another two siblings carrying the Hb Adana mutation and a 3.7 kb deletion (αcodon 59α/-α3.7). Although they all had α-thalassemia intermedia (α-TI), the former were clinically more severe than the latter. The first pair of siblings presented at a much younger age than the second pair and showed lower Hb levels and significant extramedullay hemopoiesis. Another case of a hydropic fetus as a result of Hb H/Hb Adana is also described. Their clinical phenotypes and hematological parameters are all presented for comparison.
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