HbA2 levels in β-thalassaemia carriers with the Filipino β0-deletion: are the levels higher than what is found with non-deletional forms of β0-thalassaemia?

Abstract

AIMS: Classical carriers of β-thalassaemia are identified by a raised HbA2 level. Earlier studies indicated that the Filipino β-deletion has high raised HbA2 levels. The introduction of automated high performance liquid chromatography (HPLC) for thalassaemia screening is an important advance in technology for haematology laboratories. The BioRad Variant II Hb analyser is a common instrument used to quantify HbA2 levels in thalassaemia screening. This study aimed to determine HbA2 levels in carriers of Filipino β-mutation using the BioRad Variant II Hb analyser.

METHODS: The Filipino β-deletion was identified using gap-polymerase chain reaction (PCR) in the parents of transfusion dependent β-thalassaemia patients who were homozygous for the Filipino β-deletion in the indigenous population of Sabah, Malaysia. Hb subtypes were quantified on the BioRad Variant II Hb analyser. Concurrent α-thalassaemia was identified by multiplex gap-PCR for deletions and amplification refractory mutation system (ARMS)-PCR for non-deletional mutations.

RESULTS: The mean HbA2 level for Filipino β-thalassaemia trait was 5.9±0.47 and with coinheritance of α-thalassaemia was 6.3±0.44 (-α heterozygous) and 6.7±0.36 (-α homozygous). The HbA2 levels were all >4% in keeping with the findings of classical β-thalassaemia trait and significantly higher than levels seen in non-deletional forms of β-thalassaemia.

CONCLUSION: The HbA2 level measured on the BioRad Variant II Hb analyser was lower than the level in the first description of the Filipino β-thalassaemia. β-thalassaemia trait with coinheritance of α-thalassaemia (-α) is associated with significantly higher HbA2 level.

Keyword: HbA2 levels; β-thalassaemia carrier; Filipino; β0-deletion; Non-deletional forms; β0-thalassaemia