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CME article

Theme: A rare haemaglobinopathy

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A 53-year-old lady is referred to the haematology unit for further investigations of anaemia. She gives a history of blood transfusion 30 years back during her pregnancy. She is not on any medication since then. Her main complaint is fatigue. On examination she is pale and icteric. Her abdominal examination shows splenomegaly and hepatomegaly, 5 cm and 2 cm respectively. Other systemic examinations are unremarkable.

Her investigation results are given below

FBC – RBC 4.3 x10^{12} /L, Hb 8.3 g/dL, MCV 62.3 fL, MCH 19.2 pg, MCHC 30 g/dL

Retic count – 3.5% Ferritin – 1080 ng/mL

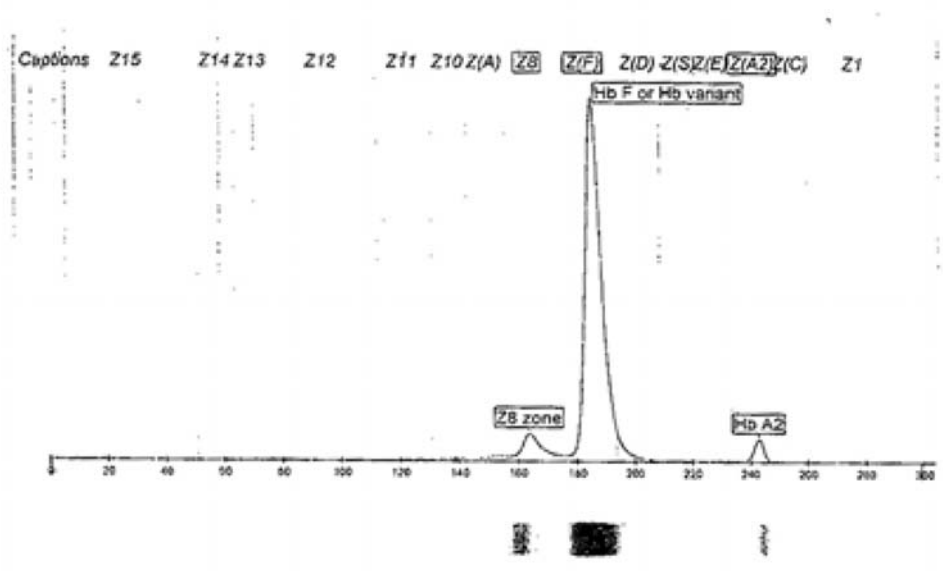
Bilirubin – Total 23 µmol/L, Direct 08 µmol/L, Indirect 15 µmol/L

A Hb capillary electrophoresis is done and the report is given below. The same sample is repeated with the HPLC to confirm the results.

1.1 Interpret the Hb capillary electrophoresis/HPLC results giving differential diagnosis?

1.2 Describe how you would arrive at a diagnosis?

1.3 What further tests to be done to confirm the diagnosis?



Haemoglobin typing

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Haemoglobin electrophoresis

Name	**%**
Z8 zone	5.3
Hb F or Hb variant	91.3
Hb A2	3.4

Results of the HPLC

Hb F	Hb A2	Hb A
91.3%	3.4%	Absent

CME points: 02

The answers and the discussion are given in page 39