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C/ β 0 thalassemia

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► **To cite this version:**

Barbara J Bain. C/ β 0 thalassemia. American Journal of Hematology, 2009, 84 (11), pp.749.
10.1002/ajh.21534 . hal-00510307

HAL Id: hal-00510307

<https://hal.science/hal-00510307>

Submitted on 18 Aug 2010

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C/ β 0 thalassemia

Journal:	<i>American Journal of Hematology</i>
Manuscript ID:	AJH-09-0542
Wiley - Manuscript type:	Morphology Update
Date Submitted by the Author:	11-Aug-2009
Complete List of Authors:	Bain, Barbara; Imperial College Campus of Imperial College Faculty of Medicine, St Mary's Hospital, Praed Street, W2 1NY., Haematology
Keywords:	Hematology- general
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<p>Fig. CBetaZero7</p>	



Morphology Update

C/ β^0 thalassemia

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The blood film illustrated is from a 60-year-old Afro-Caribbean woman with compound heterozygosity for haemoglobin C and β^0 thalassemia. She had required a cholecystectomy for gall stones at a young age but otherwise had had only minor symptoms. Her spleen was palpable on inspiration. Hemoglobin concentration (Hb) and red cell indices were: red cell count $4.96 \times 10^9/l$, Hb 7.8 g/dl, hematocrit 0.25 l/l, mean cell volume 62 fl, mean cell hemoglobin 19.1 pg and mean cell hemoglobin concentration 30.9 g/dl. The reticulocyte count was $173 \times 10^9/l$, bilirubin 56 $\mu\text{mol/l}$ (normal range 3–17) and serum ferritin 431 $\mu\text{mol/l}$ (normal range 15–300).

The blood film shows a haemoglobin C crystal (centre right) and a poorly hemoglobinized erythroblast (centre left). In addition there are target cells and numerous irregularly contracted cells. Pappenheimer bodies were also present. Haemoglobin C crystals are very distinctive because of their straight parallel edges. They are often in an erythrocyte that is otherwise empty of hemoglobin. They can be seen in hemoglobin C disease and in compound heterozygous states when hemoglobin C is co-inherited with either hemoglobin S or β thalassaemia. The combination in this patient of features of hemoglobin C (crystals, target cells and irregularly contracted cells) with features of thalassemia (microcytosis, poor hemoglobinization and Pappenheimer bodies) points to the correct diagnosis.