atory features of sickle cell disease in The Netherlands

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Abstract:

Out of about 200 patients with sickle cell disease (SCD) in the Netherlands, 6% are non-negroid patients from Turkey. 83 were assessed clinico-haematologically regarding the type of SCD, ethnic origin, concurrent alpha-thalassaemia (alpha-thal), and type of sickle cell gene (beta S-chromosome). 54 patients had homozygous sickle cell (SS), 1 sickle cell haemoglobin D (SD) Punjab, 5 sickle cell beta o-thalassaemia (S beta o-thal), 5 sickle cell beta +-thalassaemia (S beta +-thal) and 18 sickle cell haemoglobin C (SC) disease. 14% of the 83 patients were from Turkey, the others were of West Indian and African origin, most (73%) of whom were from Surinam. The Netherlands may be the only country in the world where non-negroid SCD patients are present in such a proportion to negroid SCD patients. alpha-thal was detected in 16 patients and in 14 of their relatives with sickle cell trait. Four main types of beta S-chromosomes were identified: Benin, Central African Republic, Senegal and Saudi Arabia types. SS and S beta o-thal disease ran a more severe course than S beta +-thal and SC disease. No clinical difference was ascribable to ethnic origin, alpha-thal or HbF-level but in each ethnic group there were some patients with a remarkably mild course of SS disease, which was related to the type of beta S-chromosome. These were the Senegal and Saudi Arabia beta S-chromosomes. Proper differentiation between genotypes is of prognostic and therapeutic relevance, especially in SC disease as it is sometimes discovered too late.(ABSTRACT TRUNCATED AT 250 WORDS)