Percentile curves for red cell indices of \( \beta^0 \)-thalassaemia heterozygotes in infancy and childhood

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Abstract. This paper describes the percentile curves for red blood cell (RBC) count, Hb, mean corpuscular volume (MCV) and mean corpuscular haemoglobin (MCH) values of \( \beta^0 \)-thalassaemia heterozygotes during infancy, childhood and adolescence. Hb values were about 2 g/dl below those of normal controls with a progressive increase with age parallelling the normal developmental trend. The Hb increase with age was due to a progressive rise in the Hb content per cell, the number of RBC remaining nearly constant. MCV and MCH values also increased with age with a pattern parallel to normal control. Because of the high prevalence of \( \alpha \)-thalassaemia in the Sardinian population, to which all the subjects investigated belong, the 3rd MCH-MCV percentile curves of normal overlap the 97th curve of \( \beta^0 \)-thalassaemia heterozygotes. The HbA2 levels, however, were always increased as compared to normal. These results confirm in children than screening for heterozygous \( \beta^0 \)-thalassaemia with moderate microcytic anaemia. If the values lie outside the extreme limit of variability for \( \beta^0 \)-thalassaemia superimposed iron deficiency should be suspected and appropriately investigated.

Because few studies, on a limited number of subjects, are available on the developmental modifications of red blood cell (RBC) indices in \( \beta^0 \)-thalassaemia heterozygotes \([1, 4]\) we developed percentile grids for RBC number, Hb, MCV and MCH values versus age in a large group of children who are heterozygotes for \( \beta^0 \)-thalassaemia.

Methods

RBC indices were determined with a Coulter Counter model S standardized daily with 4 C commercial standard (Coulter Electronics, Hialeah FL, USA). Haemoglobin electrophoresis was carried out on Titan III acetate cellulose plates in Tris-EDTA-borate buffer at pH 8.6 (Helena Lab. Beaumont, Tx, USA). The haemoglobin A2
was quantified by microchromatography on DE-52 [5]. Serum iron and iron binding capacity were measured according to Lauber [7]. For zinc erythrocyte protoporphyrin determination we used the Esa P-4000 Haematofluorometer (Esa, Mass, USA).

The data were analysed and converted to percentiles by an IBM (New York, USA) computer system.

Results

Figure 1 depicts percentile curves for Hb in children heterozygous for β-thalassaemia compared to normal standards from the same population.

Thalassaemic 50th percentile values were at each age considered approximately 2 g/dl lower than those in age-matched controls. The 90th and 97th percentile curves for thalassaemic children overlapped the normal standard curves. Hb levels progressively increased in β-thalassaemia heterozygotes during childhood with a pattern parallel to that of normal controls.

Figure 2 demonstrates the age related percentile grid for the RBC count compared to normal standards. RBC counts were elevated without significant age-related modifications.

Figures 3 and 4 illustrate the age related changes of MCV and MCH values. The 97th thalassaemic percentile curve overlapped the 3rd normal curve. MCV and MCH values increased with age parallel to that of normal controls. In β-thalassaemia heterozygotes HbA2 levels were always increased (Fig. 5). There was a progressive increase until 6 years with nearly constant levels thereafter.

Discussion

This study presents percentile grids for Hb, HbA2, RBC counts, MCH and MCV values in β-thalassaemia heterozygotes from infancy to adolescence which may be very useful for evaluating a child with microcytic anaemia. The differences between the percentile curves of β-thalassaemia heterozygotes and normals are slightly obscured because of the high frequency of α-thalassaemia in our population, which on the one hand tends to lower the 3rd normal MCV and MCH percentile curves and on the other hand tends to raise the 97th β-thalassaemia curve with a resulting slight overlap between them. The shape of the 97th β-thalassaemia curve is, indeed, due to