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ANALYSIS OF CLINICAL FEATURES AND COMPLICATIONS IN PATIENTS WITH β-THALASSEMIA IN THE REGION OF STRUMICA

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Thalassemias are hemoglobinopathies and are hereditary deseases which are characterized by disturbance in synthesis of one or more polypeptide chains of globin. Thalassemias are type of anemia of quantitative nature, where synthesized chain of hemoglobin are with normal structure, but not in adequate quantities. β-thalassemia is a desease characterized by production of large quantities of α-chains which exceedes the production of β-chains, whereas β-chains accumulate and precipitate into precursors of red blood cells and change their permeability, which leads to lysis of red blood cells. Within Research Center for Genetic Engineering and Biotechnology (RCGEB) is operating national reference laboratory for hemoglobinopathies, founded in 1970, where over the past 40 years examined more than 30,000 individuals from our country. The analysis revealed that the average frequency of beta-thalassemia in Macedonia is 2.6%, of alpha thalassemia is 1.5%, delta-beta-thalassemia is 0.2%, while the share of Swiss type of hereditary persistence of fetal hemoglobin (HPFH) is 0.3%.

AIM OF THE STUDY

The objective of this study was to show the prevalence, or the presence, of a certain type of thalassemia in the region of Strumica, R. of Macedonia, the complications that arise in specific cases, and the diagnosis and treatment of this inherited disease, not only for successfully identifying the extent of the presence of thalassemias in this region, but also to display the patient status in this region.

MATERIAL AND METHODS

The target group for this research were registered patients with thalassemia from the region of Strumica. For that purpose, we processed medical histories of 29 subjects suffering from some type of thalassemia. All subjects which entered in this study were respectively coded in order to avoid violation of their privacy and were randomly selected.

As a material for the research we used data from medical histories of the subjects of interest, and subsequently analyzed those data. Of all data we selected those who are significant for our research like geographical location, sex, age, type of thalassemia, blood tests values, comorbidities and complications from the desease.

RESULTS AND DISCUSSION

Analysis of the available data shows that the most of the patients had or have β-thalassemia type minor, followed by patients with β-thalassemia type major, then α-thalassemia, and only 2 patients with β-thalassemia intermedia (Figure 1).
As far as geographical location we can conclude that the most of the patient came from the city of Strumica, while other patients are from rural suburbs of Strumica (Figure 2).
As suspected, all significant blood parameters which are characteristic for thalassemias, such as Hgb, Er, MCV, MCH, HbF, HbA_2, AST, ALT, and others, shows deviation from normal ranges.
Also we found a lot of comorbidities and complication associated with β-thalassemias (Figure 3).
All patients were treated with standard therapeutical agents and procedures, such as iron chelating agents, iron supplements and therapy with washed and filtered red blod cells.

FIGURE 1.
Patients with some type of thalassemia in the region of Strumica

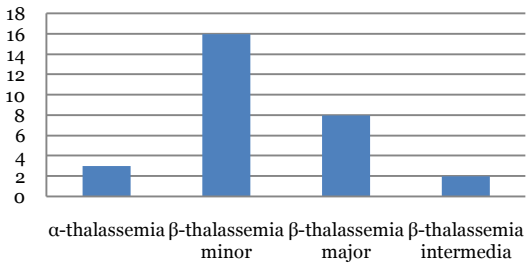


FIGURE 2.
Geographical distribution of patients with some type of thalassemia

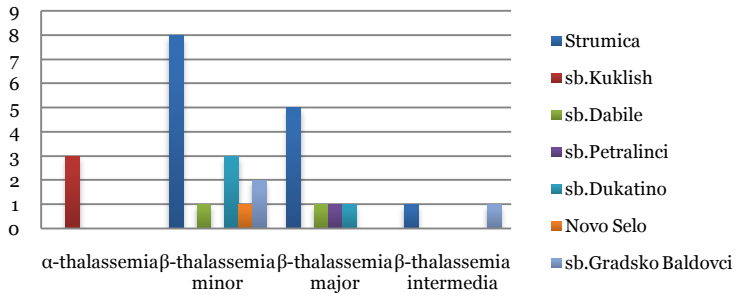
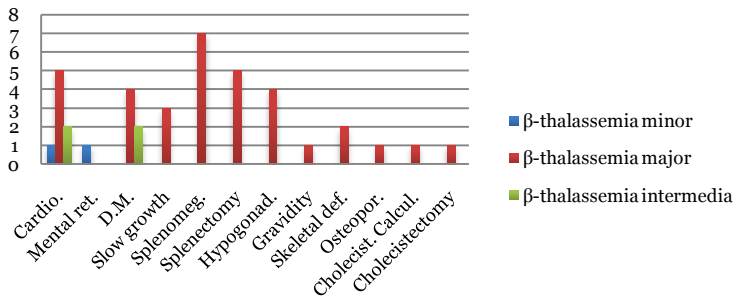


FIGURE 3.
Comorbidities and complication from β-thalassemias



CONCLUSION

The data analysis indicates the presence of many characteristic sympthoms and clinical manifestation of many features of thalassemias, which confirms the theoretical framework for this hemoglobinopathy.
In many patients it is observed decline in values of hemoglobin, reduces number of red blood cells, and disruption of liver function, followed by decreased function of liver enzymes.
Therapy that is administered to the patients is of basic character, a continuous blood transfusion in patients with milder form of thalassemia, and excess iron chelators, whilst in the patients with severe forms of thalassemia spelenctomy was performed.
The results shows that the experimental group of patients present many comorbidities and complications thay complays with the medical theory about thalassemias.