EVOLUTION AND PROGNOSIS IN B-THALASSAEMIA MAJOR IN ROMANIA

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The thalassaemias are a group of heterogeneous hemoglobin disorders in which the production of normal hemoglobin is partly or completely suppressed as a result of defective synthesis of one or more globin chains.

The homozygous or double heterozygous for thalassarmia – major thalassaemia – have been primarily described by Thomas Cooley, in 1925. The anaemia developed during the first month of life is becoming progressively severe, imposing chronicle blood transfusion. In the absence of blood transfusion severe complications occurred followed by premature death (mostly in the first decade of life).

Iron overload – a frequent complication in major thalassaemia – occurs as a result of blood transfusion or because the increased absorption of iron from the digestive tract. In the absence of an adequate and constant chelation therapy severe organ disorders and even premature death will follow, most frequently in the second decade of life).

In Romania, since 1995 blood transfusion are scheduled and since 1996 the Ministry of Health implemented the National Program for Thalassaemia (and also for Haemophilia), providing chelation therapy for all the registered patients.

Our Institute is monitoring the registered patients' evolution since 1995, taking into consideration different parameters, such as: organs disorder, different complication, iron loading level, causes of death, quality of life and life expectation.

The evolution in time is strictly connected to both transfusion program observation and compliance to chelation therapy. More than 70% of the patients comply with the treatment, having an evolution without complications, a better quality of life and a significant increasing of life expectation.

Conclusion: An adequate transfusion program and a correct chelation therapy (doses and timing) the evolution of our patients with β-Thalassaemia Major was favorable, without complications and a quality of life close to normal.