



The 65th ASH Annual Meeting Abstracts

POSTER ABSTRACTS

112. THALASSEMIA AND GLOBIN GENE REGULATION

Evolving Changes in the Characteristics of Death in Transfusion Dependent Thalassemia in Greece

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Introduction: During the last decades, improvements in the treatment of transfusion-dependent thalassemia (TDT) with safer transfusion schemas, improved management of iron overload and of comorbidities, have resulted in significantly increased life expectancy. With increased survival, the pattern of characteristics of death is changing.

Aim: The aim of our study was to analyze the causes and other characteristics of death in transfusion-dependent thalassemia patients in Greece during the last 45 years.

Patients and Methods: A retrospective cohort study was conducted in our Unit, which is the largest one in country, where around 15-20% of Greek patients (pts) are followed. The study included all deaths in TDT pts occurring between 1979 and 2023. The observation period was analyzed in 5 years' segments. For each patient, data on the age of death, cause of death, age of splenectomy, age at first chelation treatment and mean ferritin values for the last two years before death, were captured. Patients were classified into two groups: (1) mild (ferritin < 2500 µg/L) and (2) moderate-severe (ferritin: <2.500g/L). Statistical analysis performed with RStudio v.3.6.2.

Results: A total of 172 deaths (male/female: 107/65) were recorded during this observation period. Pts' characteristics are summarized in table 1. The overall incidences of the main causes of death are heart disease (57%), malignancy (12%), infections (included HIV and sepsis) (12%), liver failure (4.6%) and accidental events (4.6%). Distribution of causes of death during the different intervals are shown in figure 1. Malignancies (20 pts) are the most prevalent cause of death for the last observation period 2019-2023 and include liver cancer (11 pts), renal cancer (2 pts), colorectal cancer (2 pts), lymphoma (2 pts), lung cancer (1 pt), pancreatic cancer (1 pt) and cholangiocarcinoma (1 pt). Malignancy has gradually replaced cardiac disease as the leading cause of death over the last years. Infections increased as a cause of death in the last 5 years and reached similar levels to the initial observation time segment. A gradual increase in the age of pts at the time of death was noted, starting from a mean (±SD) of 15.7±3.6 years at the period of 1979-1983 to 46.6±6.0 years for 2019-2023. The percentage of splenectomized pts varies significantly throughout the observation period. There is a positive correlation between ferritin levels and causes of death due to cardiovascular disease. The percentage of deceased pts on chelation therapy gradually increased while the age of starting chelation therapy gradually decreased during the observation period.

Conclusions: The present study shows that there is a shift in the main cause of death in transfusion-dependent thalassemia patients the last 45 years, from cardiovascular to malignancies, while there is variability in the distribution of all causes of death during the different 5-year intervals. This reflects the overall changes in the treatment of TDT, which has transformed TDT from a pediatric disease of guarded prognosis to a chronic disease of adulthood.

Disclosures Kattamis: Chiesi: Honoraria; Vertex Pharmaceuticals: Consultancy, Membership on an entity's Board of Directors or advisory committees; Amgen: Consultancy; Agios Pharmaceuticals: Consultancy; Bristol Myers Squibb/Celgene: Consultancy, Honoraria, Membership on an entity's Board of Directors or advisory committees, Research Funding; Novartis: Consultancy, Honoraria, Research Funding; Ionis Pharmaceuticals: Consultancy; Vifor: Consultancy.

Figure 1. Distribution of causes of death in 5 years' time periods
 BMT: Bone Marrow transplantation, others (acute disseminated encephalomyelitis, acute respiratory distress syndrome, multiorgan failure, unknown)

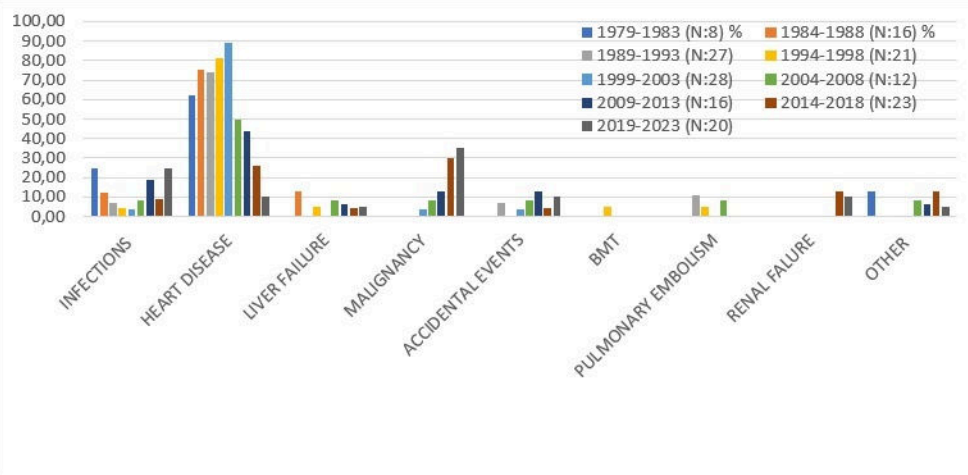


Table 1.: Patients' Characteristics

	1979-1983	1984-1988	1989-1993	1994-1998	1999-2003	2004-2008	2009-2013	2014-2018	2019-2023
Number of pts	8	16	27	21	28	13	16	23	20
Age of death; (years) Mean ±SD	15.7±3.6	19.3±2.8	21.0±3.3	24.6±3.5	28.1±4.8	31.6±6.1	34.9±5.9	46.3±5.5	46.6±6.0
(%) pts with Splenectomy	87	69	48	57	75	42	38	78	50
Ferritin levels (g/L) (Median-range)	4935 (1674-5597)	3690 (1295-9740)	3325 (755-9780)	1986 (260-5632)	4306 (1000-9285)	2771 (871-10534)	2658 (679-8100)	1348 (166-9540)	1752 (202-11076)
(%) pts on chelation	100	100	100	100	96.43	83.33	93.75	100	100
Age at first chelation; (years) mean ±SD	9.5±2.9	7.3±4.7	6.9±5.4	4.9±3.7	5.2±3.9	4.7±3.2	4.2±2.3	5.2±4.1	3.8±3.6

Figure 1

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