Life Expectancy and Survival Analysis of Children with Spinal Muscular Atrophy in Thailand

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Abstract

Background

SMA is degeneration of motor neurons in the spinal cord. Recently, the introduction of specific treatments lead to an improved survival rate but survival of SMA patients in Thailand have not yet been reported.

Objectives

To study about the life expectancy, survival analysis and factors associated with the life expectancy and survival analysis.

Methods

This is single-center retrospective study. Inclusion criteria is patient with genetically diagnosed SMA before the age 18 years at Siriraj hospital during 1999 – 2022. Probability were evaluated by Kaplan-Meier survival method by using Stata 17.

Results

Of the 113 SMA patients, the SMA type 1, 2 and 3 patients were 37, 53, and 23 patients and the median life expectancy were 22.8 months, 14.3 years, and 21.7 years respectively. The significant factors were age at onset and sex in SMA type 2, which age at onset < 1 year, the median survival

time was 15.86 years and 19.01 years in ≥ 1 year onset. The median survival time of male was 19.01 years while female could not be identified due to inadequate death rate.

Conclusion

Age at onset of SMA type 1 and 2 had significant relation with survival time. The increasing of aging of onset 1 month, the yearly probability of death decreases for 17% and 20% respectively. In SMA type 2, the male gender was increase rate of death 12 times. The survival analysis data and life expectancy of each SMA type can be used for genetic and prognostic counseling.