
Attention and Executive Function Impairment in Children with Beta-Thalassaemia Major

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Beta-thalassaemia major (b-th) is a chronic genetically haematological disorder due to defects in the synthesis of globin chains of haemoglobins [1]. Indonesia is among a group of countries with high prevalence of thalassaemia [2]. The most common treatment modality are transfusion, splenectomy, and iron chelation. Combination blood transfusion and iron chelation make significantly extend life expectancy but patients still suffer from many complications including neurological complications such as cognitive impairment [3]. Thalassaemia results in chronic hypoxic state due to chronic anaemia [4], iron overload due to chronic blood transfusions [5], and hypercoagulability which leads to susceptibility to vasoocclusion [6].

The aim of this study was to determine the characteristics of cognitive deficits such as attention and executive function in children with beta thalassaemia major. This study was an observational descriptive study, with cross-sectional study design, conducted in the outpatient clinic of the Pediatric Thalassaemia Unit of Hasan Sadikin Hospital, Bandung, Indonesia during August 2016 to January 2017. One hundred children with beta thalassaemia major age 8-14 years old participated in this study. All subjects performed vigilance, verbal fluency and block design test. The mean age of subject was 10.94 ± 1.72 years, predominantly male (52.0%). The mean frequency of transfusions per month was 1.30 ± 0.52 times. The mean haemoglobin level was 6.36 ± 0.80 g/dL, and the mean ferritin levels was 4164.42 ± 2238.60 μ g/L. Most of subject using deferiprone for iron chelation (78.0%). Attention impairment was found in 26% of children and executive function impairment was presented in 23% of children. Both impaired attention and executive function were found in 21% of children, the mean age of this group was 9.63 ± 1.29 years, the mean haemoglobin level was 6.40 ± 0.8

g/dL and the mean ferritin level was 3654 ± 1526 $\mu\text{g/L}$. The frequency of transfusion per month was 1.42 ± 0.74 times, and time from the first diagnosis was 9.87 ± 1.3 years.

Our study shows that almost quarter children with beta thalassaemia major suffer from impaired cognitive function, 26%, 23% and 21% for attention, executive function, and both attention and executive function respectively. Several studies show that cognitive function is significantly lower in patients with beta thalassaemia. Raafat et al studied children with beta thalassaemia using Wechsler Intelligence Scale for Children–Third Edition (Arabic version), showed that mean Full-Scale IQ and Performance IQ of patients were significantly lower than those of controls.[7] Monastero et al assessed cognitive function in 46 beta thalassaemia major patients and 46 controls and showed a significantly impaired cognitive function using all neuropsychological batteries [8].

Nemtsas mentioned that chronic hypoxia and iron overload contribute to neurological manifestation of beta thalassaemia.[3] Children with beta thalassaemia live with chronic anaemia condition which leads to chronic hypoxic state. Study from Ai et al stated that children who had low haemoglobin levels had significantly lower scores in Performance IQ, and group of children with high iron but low haemoglobin levels performed the worst score. [9] The mechanism of attention and executive impairment in beta thalassaemia children was never studied before but it was thought to be the results of chronic hypoxia [4], iron overload [5], and hypercoagulable state [6]. This study found that the haemoglobin level was low and ferritin level was high. Impairment of attention and executive function are important concern, because they comprise the ability of cognitive function.

According to this study, monitoring of cognitive function should be applied as early as possible to detect any kind of cognitive abnormalities in children with beta thalassaemia major so that the intervention can be done as quickly and accurately to improve their quality of life.

The limitation of this study is that we are not analysed others cognitive ability and external factors that may influence the whole of cognitive function.

Keywords: attention, cognitive, executive function, thalassaemia

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