A Study to Develop and Standardize Tool to Assess the Quality of Life among Children Suffering with Thalassemia: A Systematic Review

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Abstract

Background: Beta thalassemia is a major chronic blood disorder that has a great impact on the affected child. The great expense and difficulties in providing optimal treatment for patients causes fatal conditions among untreated cases **Objectives:** The objective of this study is to review the relevant literature to find out the various associating factors which will indicate the effect of thalassemia on different domains of quality of life of children affected with thalassemia. **Methods:** A comprehensive data base search from national & international Journals, E. Library, Pub med, Google search & Email contact with Authors carried out. A systematic review of the published literature 2004 - 2014 has been used.**Results:** Initial search started with 120 studies. After eliminating unwanted articles based on inclusion & exclusion criteria, only 5 studies were selected and explained in this review, which depicts specific information about cause of thalassemia, effect of thalassemia on growth pattern & sexual maturation rate, cognitive abilities, mood changes & adaptive functioning, literacy status, and specific causative factors affecting the quality of life. **Discussion:** Systematic review reveals various factors responsible for the occurrence of disease & its effects on various systems of the body. The findings also reveal that the thalassemic children have 3 times as much depressive manifestations, perform significantly poorly on measures of cognitive & intellectual functioning and shows poor adjustment in the social, occupational & communication domains. It affects their self confidence and give rise to emotions and thoughts which negatively impact their Quality of life. There is an urgent need for the prenatal diagnosis of thalassemia for healthy future generation.

Keywords: Thalassemia, HRQOl (health related quality of life) prevalence, cause, trends in management, treatment modalities.

Background

Thalassemia syndromes are a heterogeneous group of usually inherited chronic disorders that are characterized by an absence or decreased synthesis of one of the normal globulin chains of hemoglobin. Thalassemia is an autosomal recessive disorder which may also be the result of spontaneous mutation **(Marlow R, Dorothy and Barbara A. 1998)**[1] Originally thought Disease was limited to Mediterranean countries (Greece, Italy & Cyprus), but in 1938 Mukerjee published "the first recorded case of thalassemia on east side of suez". The case was observed by Dr M. Bose of Campbell Medical School & Hospital, Calcutta (new Kolkata) in a Hindu boy aged 30 months **(Iswar C. Verma et al 2011)**[2]. Worldwide approximately 15 million people were estimated to be suffering from Thalassemia with 240 million carriers. India recorded 30 million. The Carrier rate for í thalassemia varies from 1-17% with an average of 3.2%, this means that on an average 1 in every 25 Indian is a carrier of thalassemia **(Gosh Sharmila)**[3]. Maximum number of carriers for thalassemia have been reported in Gujarat (10.0 to 15.0 %) followed by Calcutta (10.2 %), Punjab (6.5%), Delhi (5.5%), Tamil Nadu (4.0%), Bengal (3.5%), Mumbai (2.6%), Maharashtra (1.9%) & Kerala (90.6%) **(G.Shanti, D. Balasubramaniam, and Srinivasan R.2013)**[4]. About 10,000 children are suffering from thalassemia in Madhya Pradesh.**(Sajid Khan, 2008)**[5]

A clinical and demographical study of 122 í-thalassemia revealed that 84 (68.58%) í-thalassemia children were

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