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Increasing survival of thalassemia children

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Abstract
This retrospective review study was conducted on 227 children registered to Thalassemia unit, Rajindra Hospital /Government College Patiala to study increasing age of survival. Demographic details of patients were recorded along with type of chelating agent being used on predesigned proforma. Data of these children was compared with that of year 2008 and 2009.In 2018, maximum number of children were in age group of 5-10 years. Mean age was 10.75 years and median was 10 years (range 1.0-29 years). Ten children were above age of 21 years. In 2008, no child was above age of 21 years. Males outnumber females overall in all years under comparison, but females had better survival above the age of 21 years in 2018.Maximum number of children were on iron chelating agents in 2018 as compared to previous years. With regular transfusion and use of chelating agents, there is increase in survival age of thalassemia children.

Keywords: Thalassemia, children

Introduction
Thalassemia represents a group of disorders resulting from impaired haemoglobin synthesis and ineffective erythropoiesis [1]. It is estimated that world over there are >200 million carriers of beta–thalassemia gene, 40 million of them are in India alone. The carrier rate for beta-thalassemia gene varies from 1-2% in southern India to 3-15% in Northern India [2]. For patients with more severe forms of Thalassemia, chronic life-long blood transfusions are the main stay of therapy. Untransfused children with severe Thalassemia often do not survive beyond 5 years. With transfusion and comprehensive care, birth cohorts followed from 1970 have shown life expectancy extending into the 4 Th decade of life and beyond [1]. But as survival increases so are the complications. Most common morbidities are endocrinologic (44.7%) followed by cardiovascular 41.3%, hepatic 40.5% and renal 4%. [3]. Cardiac complications remain leading cause of mortality in TM patients [2]. Low ferritin levels were associated with a lower probability of experiencing heart failure and with prolonged survival in the Italian study [4].

Subjects and method
The study was conducted on 227 Thalassemia children registered in Thalassemia Unit, Rajindra Hospital /Government Medical College Patiala. It was a retrospective review study. Demographic details of patients in form of age, sex, address, age of enrollment were recorded on a predesigned proforma. Type of chelating agents used were also noted. Data of these children was compared with that of year 2008 and 2009. Statistical analysis was done to calculate mean and median age of children on Microsoft Excel 2010 software.

Results
The study enrolled 227 Thalassemia Major (TM) children with mean age of 10.75years and median of 10 years (range 1.0years-29years). When stratification was done by age and sex, maximum children were in age group of 5-10years. Number of females out number males in younger (0-5 years) and last age (>21 years) category (Table 1). Above age of 21 years, one child each was of 22, 23, 24, 25 and 27 years old. Two children were of 26 years and 3 were of 29 years. In 2008 (Table 3), age distribution showed maximum number of children in age category of 5-10 years and there was no child above 21years. Overall though chelating agents were started in 2008, shift has occurred in choice of iron chelating agent from

Deferiprone in 2008 to Deferasirox in 2018 where maximum number of children (174) are using it.35 children now are also on combined use of Deferiprone and Deferasi rox (Table 2). Males outnumber females in year 2008, 2009 and 2018 (Table 4).

Table 1: Age and Sex Wise Distribution of Children in Year 2018

Completed Age	Total	Male	Female
0-5years	41	18	23
5-10years	73	50	23
10-15years	63	46	17
15-21years	40	30	10
>21years	10	4	6

Mean age-10.75years, Median age-10 years

Table 2: Distribution of Children Yearwise by Use of Iron Chelating Agent

Drug	2008	2009	2018
Deferiprone	35	22	18
Deferasirox	-	16	174
Deferoxamine		1	
Deferiprone &Deferasirox			35

Table 3: Distribution of Children by Age in Year 2008

Completed Age	Number of Children
0-5years	36
5-10years	54
10-15years	22
15-21years	7
>21years	0

Table 4: Sexwise Distribution of Children in Year 2008, 2009 and 2018

Year	Total	Male	Female
2008	119	82	37
2009	131	94	37
2018	227	148	79

Discussion

This study showed increasing survival of children with TM. No study was available from India for comparison. In our study, mean age was 10.75 years and median was 10 years (range 1.0-29 years). According to study by CK Li *et al* [5], median age was 15.5 years (range 1.4-30.3 years). The range in present study approximate with that of study done by CK Li *et al*. but median was lower. Three children in our study were of 29 years. Females did better in survival above age of 21 years as compared to males. In a study by Chali SC, oldest patient was 24 years old [2].

A report from 7 Italian centres showed that survival and complication free survival of patients with TM continue to improve especially for female patients born shortly before or after availability of iron chelation [6]. Same are conclusions drawn from present study that showed increasing survival in 2018 with maximum number of children on iron chelating agents and females doing better than males above age of 21 years.35 children in 2018 in present study are on 2 iron chelators contributing to better survival. Combination chelators tend to be more effective than monotherapy because chelators utilize different mechanisms for removing iron [7]. The prognosis for survival without cardiac disease is excellent for patients with TM who receive regular transfusion and whose serum ferritin concentration remain below 2500 ng/ml [8]. Survival was better for patients born in more recent years and for females who were less likely to die than males [9].

With improving survival, adverse effects of transfusions and chelation therapy also may impact quality of life. Having a child with thalassemia affects family, limiting family activities and negatively impacting parental time and emotion. Familial, social and psychosocial support are critical factors in improving quality of life (1). Job opportunities and concessions at job need to urgently consider.

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