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# Ocular Manifestations in Multi-Transfused Children with Beta Thalassemia Receiving Chelating Agents

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**Abstract**
**Purpose:** *To determine prevalence of ocular abnormalities in children with beta thalassemia and assess the ocular side effects of blood transfusion and iron chelating agents.*
**Methods:** *Cross sectional study undertaken over a period of one year among 30 β Thalassemia major patients from age group of 6 months to 18 years were taken. Full medical history, thorough physical examinations were done to all patients groups, and ophthalmological examination to determine the prevalence of ocular manifestations for all patient groups and to correlate these manifestations or changes with iron chelating agents. Ocular examination including near and distance visual acuity assessment with Snellen's charts, colour vision testing, external examination with diffuse illumination, slit-lamp examination, direct and indirect ophthalmoscopy, schrimer test for tear break up time and ocular biometry will be performed.*
**Results:** *In 30 patients (16 females and 14 males) with age ranging between 2 years to 18 years. The prevalence of ocular abnormalities was 40%, (12/30). Ocular changes seen included decreased visual acuity 50% (6/12), ocular surface disorder 23%(7/12), lenticular opacity 16.5% (2/12), blurred optic disc margins 16.5% (2/12) and dilatation and tortuosity of retinal vessels 33% (4/12), Retinal pigment epithelium mottling 8%(1/12). The mean serum ferritin and the number of blood transfusions received were higher in children with ocular findings than in those with no ocular manifestations.*
**Conclusion:** *Ocular findings in beta-thalassemia may correlate to the disease itself, iron overload or the chelating agents used.Children with beta-Thalassemia develop various eye problems and Ocular changes are attributed to the course and severity of the disease. Therefore, beta thalassemia children should be screened periodically to enable early detection and delay of ocular changes.*

## Introduction

Thalassemias are the most common single gene disorder worldwide. Mutations involving the beta globin gene in beta-thalassemia cause disruption in red blood cell maturation leading to ineffective erythropoiesis and multi-system involvement. Ineffective erythropoiesis leads to severe anemia which is treated by regular blood transfusion to maintain hemoglobin at or above 10mg/dl. Multiple/repeated blood transfusions lead to siderosis[1]. More than 42,000 newborns are affected by Beta- thalassemia every year world-wide. Without blood transfusions, beta-thalassemia major causes death amongst affected

children before the age of three years[2]. Ocular changes like ocular surface disorders, cataract, optic neuropathy, retinal pigment epithelial degenerations, retinal venous tortuosity, vitreo - retinal haemorrhages are noted. This study was conducted to assess the ocular abnormalities in multi-transfused beta- thalassaemic Indian children and to determine their relationship with iron overload, chronic hypoxia, age, the dosage and duration of treatment with chelating agents.

**Aims and Objective**

- Study the ocular abnormalities in beta-thalassemia major patients.
- Assess the ocular side-effects of blood transfusions and iron chelating agents.

**Method of Study**

We conducted a cross sectional over a period of one year among 30 children with beta-thalassemia from age group of 6 months to 18 years attending the thalassemia clinic of the paediatric department of a tertiary care centre.

**Inclusion Criteria**

- Children diagnosed with Beta thalassemia major with or without Blood transfusion.

**Exclusion Criteria**

- Children with previous ocular morbidity.
- Trauma
- Active ocular infections
- Haemoglobinopathies other than beta thalassemia

An informed consent for participation was obtained from parents or legal guardians. The diagnosis of beta-thalassemia major will be confirmed by clinical and hematological studies.Data was collected in form of detailed medical history, Ironchelating agents: dose, duration and serum ferritin levels, thorough physical examination & Ophthalmological examination by an ophthalmologist included:

Visual acuity

Refraction testing

Slit-lamp examination

Colour vision testing

External examination with diffuse illumination and slit-lamp examination

Fundoscopy by both direct and indirect ophthalmoscope

Schirmer test and Tear break up time

**Results**

In 30 patients (16 females and 14 males) with age ranging between 2 years to 18 years. The prevalence of ocular abnormalities was 40% (12/30). Ocular changes seen included decreased visual acuity (5/30), ocular surface disorder (7/30), lenticular opacity (2/30), blurred optic disc margins (2/30) and dilatation and tortuosity of retinal vessels (4/30), Retinal pigment epithelium mottling (1/30).

**Table 1:** Serum Ferritin and ocular Manifestations

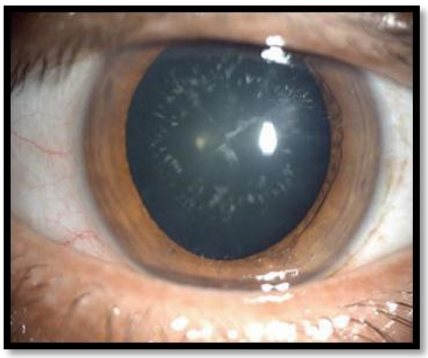
| SERUM FERRITIN | NO. OF SUBJECTS (N=30) | OCULAR INVOLVEMENT |
|---|---|---|
| LESS THAN 1000 | 4 | 0 (0%) |
| 1000- 3000 | 10 | 3(30%) |
| 3000-5000 | 9 | 5(55.4%) |
| 5000 OR MORE | 7 | 4(57.2%) |

**Table 2:** Chelation dose and ocular manifestations

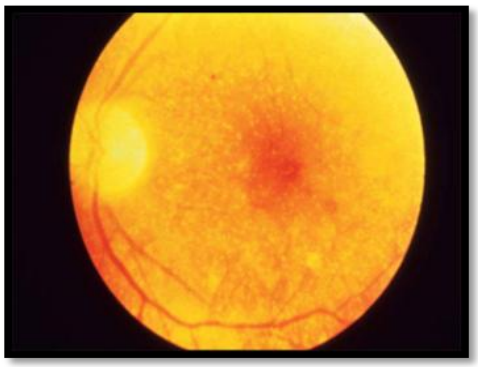
| Dose of chelating agent | NO. OF SUBJECTS (N=30) | OCULAR INVOLVEMENT |
|---|---|---|
| <20mg/kg/day | 8 | 2(25%) |
| 20-30 mg/kg/day | 10 | 4(40%) |
| >30mg/kg/day | 12 | 6(50%) |

**Table 3:** Age Relation with Ocular Manifestations

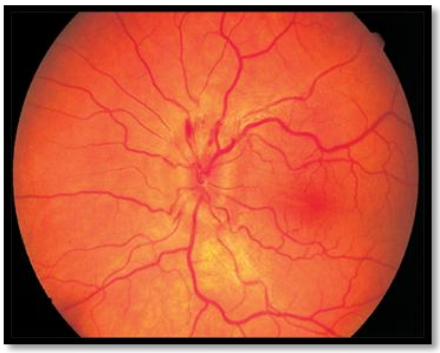
| AGE | NO. OF SUBJECTS (n=30) | OCULAR INVOLEMENT |
|---|---|---|
| Less than 5 years | 4 | 1(25%) |
| 5 – 10 years | 14 | 4(28.5%) |
| 11 – 18 years | 12 | 7(58.3%) |



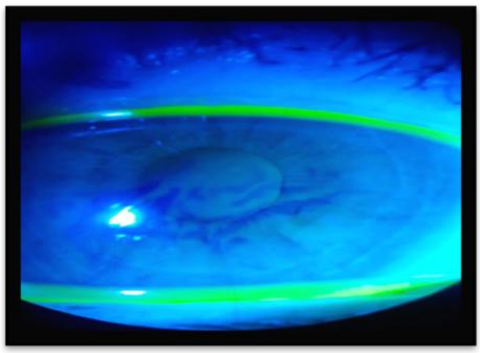
**Lenticular opacity**



**RPE mottling**



**Disc hyperemia**



**TBUT**

**Table 4:** Thalassemic Children with Ocular Findings

| AGE (years) | GENDER | MEAN SERUM FERRITIN (ng /ml) | MEAN DAILY DOSE (mg/kg) | VISUAL ACUITY RE | VISUAL ACUITY LE | SLIT LAMP | FUNDOSCOPY |
|---|---|---|---|---|---|---|---|
| 6 | M | 1420 | 9.8 | 6/9 | 6/6 | Normal | Normal |
| 9 | F | 2978 | 16.5 | 6/6 | 6/6 | OSD | Normal |
| 9 | F | 2887 | 18.8 | 6/6 | 6/6 | OSD | Normal |
| 10 | M | 4657 | 10.7 | 6/6 | 6/6 | Normal | RPE mottling |
| 10.5 | F | 2800 | 31.8 | 6/12 | 6/18 | Lenticular opacity | Blurred optic disc margins, vessels tortousity |
| 11 | M | 3589 | 21.3 | 6/6 | 6/6 | Normal | Normal |
| 12 | F | 3890 | 30.5 | 6/12 | 6/9 | OSD | Normal |
| 13 | M | 5400 | 32.4 | 6/6 | 6/6 | OSD | Blurred optic disc margins |
| 13 | M | 4595 | 24.6 | 6/6 | 6/6 | OSD | Normal |
| 15 | F | 5200 | 32.8 | 6/9 | 6/9 | OSD | Vessels tortousity |
| 18 | F | 5560 | 35.6 | 6/12 | 6/9 | Lenticular opacity | Normal |
| 18 | F | 5774 | 38.5 | 6/18 | 6/12 | OSD | Normal |

## Discussion

In a resource poor country like India, definitive therapy like stem cell therapy is beyond the reach of majority of the children. These children are on multiple blood transfusions, giving rise to iron over load. Iron overload affects all the organs in

the body including the eyes. Frequency of ocular involvement in our study was 40%, Gartagantis et al. reported figures of 41.3%, Gaba A et al. reported ocular involvement in 71.4% while Taneja et al. reported figures of 58% of subjects in their respective studies[3,4]. Visual acuity was affected in 50% patients in our study while in Gaba et al. study, the figure was 70% and Taneja et al. study, this figure was 67%.

Prabhjot Kaur Sekhon et al conducted an study among 75 Beta thalassemia children and showed that the most common eye changes were seen in Retina (n=24), Iris (n=26) and len s(n=24). Corneal dryness, lenticular opacities, disc atrophy and thinning were observed in children with Beta Thalassemia and on blood transfusions. These changes increased with duration of disease[5]. Gosai DK et al conducted a study in which most of the ocular changes of beta thalassemia are attributed to the course and severity of the disease. Reduction in serum iron and serum Ferritin levels by iron- chelating agents and regular ocular examination to look for side-effects of such agents can aid in preventing or delaying ocular complications[6].

**Limitations**

- Small sample size
- It cannot conclusively establish whether ocular changes are a result of the disease or due to iron- chelating agents. This requires stoppage of chelation therapy.

**Conclusion**

Children with beta-Thalassemia develop various eye problems and Ocular changes are attributed to the course and severity of the disease. Therefore, beta thalassemia children should be screened periodically to enable early detection and delay of ocular changes.

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