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ORIGINAL ARTICLE

Studies On Blood Profile In Thalassaemia Patients Before And After Blood Transfusion In And Around Rahata, Ahmednagar

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Abstract:

The present study was undertaken to evaluate the effect of blood transfusion in thalassaemia patient in urban and rural area of Rahata during July 2011 to February 2012. Thalassaemic fifty patients were selected and regularly blood transfusion was carried out. Pre- and post-transfusion Hb, RBC, WBC, HCT, platelets, MCV, MCH, MCHC, urea, sugar, creatinine and cholesterol were evaluated. The obtained results are discussed. Also symptoms, epidemiology, and possible physiology discussed.

KEYWORD:

Thalassaemia, heamatology, blood indices, blood biochemistry.

INTRODUCTION

Thalassaemia is an inherited autosomal recessive blood disease. It is the genetic defect, which could be either mutation or deletion, which reduces rate of synthesis or no synthesis of one of the globin chains that abnormal hemoglobin molecules. Thalassaemia is a quantitative problem of few globin synthesized of an incorrectly functioning globin. It is under production of normal globin proteins by mutations. Hemoglobinopathies gives structural abnormalities in the globin proteins. The two conditions may overlap that cause abnormalities in globin proteins and affect on their production.

The alpha- and beta- thalassaemia are prevalent invarious geographical clusters around the world. It is associated with malarial endemicity in ancient times. Alpha is prevalent in Western African, South Asian, Africa, Americas, Nepal and India [1] and is reported lower incidence of morbidity and mortality [2-4]. Beta thalassaemia is prevalent among Mediterranean, Europe, Greece, Turkey, South Italy, West Asia and North Africa. Maldives are also affected, with the world's highest concentration of carriers (18%) [5]. Thalassaemia resemble another genetic disorder affecting hemoglobin, sickle-cell disease [6].

Hemoglobin is composed of four protein chains (two α- and two β-globin) arranged into a heterotetramer. Suffered patients showed a deficiency of either α or β-globin, which produces a specific mutant form of β- globin. The β- globin chains are encoded by a single gene on chromosome 11 [7], α-globin chains are encoded by two closely linked genes on chromosome 16. In a normal person there are two loci encoding the β- chain, and four loci encoding the α -chain. Deletion of one of the α- loci has a high prevalence in African or Asian

The α-thalassaemia involves the genes HBA1 and HBA2. There are two gene loci and four alleles. It is also connected to the deletion of the 16p chromosome. α- Thalassaemia result in decreased α-globin production and produced few alpha-globin chains, resulted an excess of β- chains in adults and excess γ chains in newborns. The excess β- chains form unstable tetramers which have abnormal oxygen dissociation curves [7].

Beta thalassaemia are due to mutations in the HBB gene on chromosome 11 [7]. Mutations are characterized by either βo or β- thalassaemia major which prevent any formation of β- chains. They are characterized as β+ or β- thalassaemia intermediate; allow some β- chain formation. They bind to the RBC membranes, producing membrane damage, and toxic aggregates. As well as alpha and beta chains present

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in hemoglobin. Just as with beta thalassaemia, mutations that affect the ability of this gene to produce delta chains can occur.

Both α- and β-thalassaemia are often inherited in an autosomal recessive. Cases of dominantly inherited α- and β- thalassaemia have been reported. The autosomal recessive forms of the disease, both parents must be carriers in to a child to be affected. If both parents carrier there is a 25% chance with each pregnancy for an affected child. Genetic counseling and genetic testing is recommended for families that carry a thalassaemic trait [8]. However, 60-80 million people in the world are carrier of beta thalassaemia. India and Pakistan are increasing of thalassaemia patients due to lack of genetic counseling and screening. It may become a serious problem in the next 50 years.

Generally, thalassaemia is prevalent in populations in humid climates where malaria was endemic. It affects all races, as thalassaemia protected these people from malaria due to the blood cells' easy degradation. Thalassaemia are associated with people of Mediterranean origin, Arabs, and Asians. The Maldives (18%) has the highest incidence of thalassaemia in the world. The estimated prevalence is 16% from Cyprus, 1%, in Thailand, and 3-8% from Bangladesh, China, India, Malaysia and Pakistan. A very low prevalence has been reported from in Northern Europe (0.1%) and Africa (0.9%) [9]. It is also particularly common in populations of indigenous ethnic minorities of Upper Egypt [10]. People diagnosed with heterozygous (carrier) β- thalassaemia have some protection against coronary heart disease [11].

The most severe form of alpha thalassaemia major causes stillbirth. Children born with thalassaemia major are normal at birth, but develop severe anemia during the first year of life [12]. The other symptoms are bone deformities, face fatigue, growth failure, shortness of breath and jaundice. The minor form of alpha and beta thalassaemia has small RBC, abnormal shape of RBC and swollen spleen [13].

Health is basic need of nation and should provide facilities to each and every national person. But thalassaemia is another type of abnormalities in patient. It requires regular blood transfusion. Indian Government passed out the resolution free of cost blood because it required regularly and is the only medicine for thalassaemia. The ratio of thalassaemia in India is low but condition is critical. Considering such situation present study was assigned.

MATERIALS AND METHODS

The study was conducted in and around Rahata who regularly visit to the blood bank for the requirement of blood. In the study fifty patients were selected during July 2011 to February 2012. The following parameters were studied in the selected patients. The haemoglobin (Hb), peripheral court of red blood corpuscles (RBC), white blood corpuscles (WBC) were carried [14]. The packed cell volume (PCV) was determined according to ICSHC [15], platelets [16] and reticulocytes [17]. The following blood biochemical parameters were estimated as sugar, creatin and urea [18] and cholesterol [19].

Blood samples were collected from patient using heparinsed syringe (5000UI) in a sterilized vial on pre and post transfusion. Then the blood parameters were considered for the study blood indices before and after the transfusion of blood and presented in table 1.

PCV [packed cell volume] is the amount of centrifugation expressed percentage of total blood volume. Fall in haematocrit value observed in anemia and Hydremia. Increase in hematocrit observed in polycythemia, dehydration, congenital heart disease. Normal range was 42-52 % in male and 36- 48 % in female. When anti-coagulated blood is centrifuged in a haematocrit tube at high speed the erythrocytes sediment at the bottom the red cell column is called as PCV [packed cell volume] or hematocrit.

Blood indices

The erythrocytic indices were evaluated and are calculated from (1) hemoglobin concentration (2) PCV and total erythrocyte count. Three indices commonly calculated were (a) Mean Cell Volume (MCV), (b) Mean Cell Hemoglobin (MCH), and (c) Mean Corpuscular Hemoglobin Concentration (MCHC). These values give quantitative information about the red blood cells. RBC in by using blood sample determined (a) hemoglobin, (b) PCV, and (c) total erythrocyte count. The blood indices were calculated as :-

1)Mean Cell Volume (MCV) MCV = PCV *10 / RBC
Normal range: 82- 92 cumm.
2)Mean Corpuscular Haemoglobin (MCH), It is the amount of haemoglobin in average red cells. It is calculated as follows-
MCH = Hb *10 / RBC in million

Normal range: 27-32 pg, 1 pg = 10-12g

3)Mean Corpuscular Haemoglobin Concentration (MCHC). It is that portion of the average red cell containing hemoglobin. It is calculated as follows-

MCHC = Hb *100 / PCV

Normal range: 32- 36 %

All hematological parameters were estimated on fully automatic hematological analyzer (Erma pce 210 made In Japan) and biochemical parameters were done by colorimetric. The obtained data were presented in the table 1.

RESULTS AND DISCUSSION

Observations made on pre- and post- treatment on the diseases thalassaemia in and around Rahata and revealed blood properties, blood indices, blood serum biochemical alterations. It is noticed that some of the blood parameters were increased in the thalassaemia patient after regular blood transfusion. Also blood indices such as PCV, MCV, MCH and MCHC were noticed similar trends incline after transfusion of blood. In blood biochemical sugar and cholesterol increased while urea and creatinine remain same as pre and post transfusions in the patient.

Hb: The haemoglobin content of the thalassaemia post-transfusion person increased after each transfusion. It was inityialy 8.57 gm% and increased 9.68 gm% after transfusion. It was increased due to blood get transfused in the body of host. The mean Hb levels must be kept near 9 gm/dl before transfusion and the dose of desferrioxamine should be 20 mg/kg for children to avoid its toxic effect on bones.

RBC count: A slight increase was observed in total RBC count after transfusion in the patient. It was 3.8 m/cumm and inclined up to 4.07 m/cumm.

WBC count: A similar trend of increased was observed in WBC value after transfusion of desired blood. It was noticed 7989/cumm from 6924/umm.

Platelet: An incline in platelets value was noticed in the regularly treated patient. It was noticed initial 2.76 lakh/cumm and rise to 3.09 lakh/cumm.

Haematocrit (PCV): The regularly treated patient showed increased in haematocrit. It was initially 28.80 % and incline to 30.59%.

Mean Corpuscular Volume: MCV contents in the present study noticed that an increased in remarkable level. It revealed that the MCV was inclined 78.14 fl from 71.80 fl.

Mean Corpuscle Hemoglobin: MCH value was also found to be inclining in the blood of thalassaemia patient after regular transfusion of blood. Similar trend was noticed in the MCH was 21.31 pg of the patient before blood transfusion and showed 25.26 pg in the post treated. Mean Corpuscular Hemoglobin Concentration: The percent value of MCHC was also found to be inclined. It was noticed 31.44 gm/dl in the treated and 29.57 gm/dl in the diseased patient.

Blood Sugar (BSL): The blood serum sugar level was also increased in the body of diseased patient. It was noticed initially 92.64 mg % and increased up to 95.64 mg %.

Blood Urea (BUL): The blood urea level remain as similar as before and after transfusion of blood. That means the urea level in the patient body always showed increased if blood is not given the patient. It was revealed 33.42 mg % after and before the blood transfusion.

Creatinine: The creatinine after and before treatment revealed similar report that neither or nor incline or decline in post transfusion in patient. After transfusion it maintained kidney function normally.

Cholesterol: The cholesterol also known as is most important for body. The cholesterol was found to be 180 mg % in the patient body after the blood transfusion than the diseased patient as 175 mg %.

Endocrine complications in thalassaemia major are the result of iron deposition in the endocrine glands. The nature and the frequency of endocrinopathies differ between developing and developed countries. This showed hypogonadism complication.

The prevention of growth retardation is essential. A regular monitoring growth in all children by using growth charts is mandatory. The mean Hb levels must be kept near 9 gr/dl, before transfusion and the dose of desferrioxamine should be 20 mg/kg for children to avoid its toxic effect on bones. The iron chelation therapy prevents pituitary haemosiderosis, which is the main cause of growth hormone (GH) insufficiency. Therapeutic response of GH administration is not satisfactory. Growth acceleration is mainly promoted in children with sex steroids. Iron deposition in the pituitary cell is the mechanism of hypogonadism, which is manifested with sexual infantilism or failure to complete puberty [20].

Induction of puberty in boys is performed with the administration of testosterone (IM at the dose of 25-50 mg per monthly for 6 months), which resulted in penile and pubic hair development. Treatment is depending on clinical response and laboratory finding, where the final adult dose of 250 mg is reached after

a period of 2 to 3 years. Testicular enlargement and spermatogenesis can be achieved with the combined administration of HCG and HMG [20].

Induction of puberty in females is revealed with the oral administration of Ethinyl Estradiol, which resulted in breast and growth acceleration. This treatment followed gradually increased after one year. Transdermal administration of estrogens also be used. The combined administration of estrogens and progesterone is also used for induction of menarche and maintenance of the menstrual cycle. The transdermal use of Estradiol and Norethisterone is the ideal treatment in the hypogonadal female, because of its proven beneficial effect on bones.

Haemosiderosis of the thyroid gland is the cause of thyroid dysfunction in Thalassaemia. This complication is relatively rare and occurs after the age of 10 years. Measuring serum T4 and TSH levels easily make the diagnosis. The treatment of hypothyroidism is oral L-thyroxine at 25 µg daily for 2-3 weeks and then increased in 100 µg/m2 until thyroid hormones are normalized. This rare complication, which is mostly caused by iron deposition in the parathyroid glands presents after the age of 16 years equally in both sexes. The majority of the patients present with mild hypocalcaemia and very rarely with tetany and cardiac failure. The diagnosis is based on low serum calcium, high phosphate and low PTH levels. The oral administration of Vitamin-D and calcium is the treatment of choice. Patients usually present the impaired glucose tolerance, due to insulin resistance and develop insulin deficiency [5]. Diabetes mellitus is characterized with hyperglycemia is seen after the age of 10 years.

In cases of impaired glucose tolerance in the patients are advised to follow a proper diet and lose weight. Oral hypoglycemic drugs as Metformin and Glibenclamide are given when indicated. Insulin therapy is performed in the insulin deficiency. Insulin dose is adjusted based on glucose monitoring. The diabetic patient requires periodical eye examination and monitoring of renal function [5].

The reports of successful pregnancies provided strong evidence for the safety of the pregnancy in the thalassaemia woman. Spontaneous pregnancies in women with preserved

Hypothalamic–Pituitary–Gonadal axis, normal menstrual cycles and common ovarian function. Males have normal gonad function are able to the spermatogenesis and become fathers. In impaired spermatogenesis, a combination treatment with Gonadotrophins is best to reproductive capacity [12].

The thalassaemia woman to become a mother may needs to be special caution and medical care. It is sometimes impossible to materialize due to medical reasons. A specific criteria need to be met before a women are able to conceive. It is always need with special caution and sensitivity.

Thalassaemia minor usually does not require any specific treatment [12]. Consult to blood or oncology expert. Treatment for thalassaemia major includes chronic blood transfusion therapy, iron chelation, spleenectomy, and allogeneic hematopoietic transplantation [20]. Medical therapy for beta thalassaemia is primarily involves iron chelation. Deferoxamine is chelation agent. Deferasirox (Exjade), Deferiprone are an oral iron chelation drugs. The antioxidant indicaxanthin, Trolon and Vitamin-C reduce perferryl-Hb generated in met-Hb and hydrogen peroxide [21]. The indicaxanthin can be incorporated into the redox machinery of β-thalassaemic RBC and defend the cell from oxidation [10].

We were worked very hard on this disease. So far there has been a limited success and two distinct treatment strategies have been designed. However, this percentage is small it is infinitely better than what is available from conventional medicine where there are virtually no successes to show. We are handled only a handful cases that were being successfully treated and illustrate the point. In the present investigation effect on haematological and biochemical were evaluated and there is needed to take percussion. Based on present work authors would like suggest to people as- monitor blood properties regularly, do not marry thalassaemia minor with thalassaemia minor, treat thalassaemia patient properly and regularly, and transfuse blood regularly from recognized blood bank. Thalassaemia patient need to be special physician caution.

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