# ROLE OF SURGERY IN THALASSEMIA

**ESSAY SUBMITTED IN PARTIAL FULFILLMENT OF MASTER DEGREE IN GENERAL SURGERY**

By

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بسم الله الرحمن الرحيم

قالوا سبحانك لا علم لنا إلا ما
علمتنا إنك أنت العليم الحكيم .

صدق الله العظيم
البقرة - ٣٢ -

بسم الله الرحمن الرحيم

**To my Mother**
**To my Father**
**To my Friends who Support me by all means**

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## Abbreviations

| | |
|---|---|
| **Hb** | Hemoglobin |
| **Hb.F.** | Fetal Hemoglobin |
| **Hb.A.** | Adult Hemoglobin |
| **R.B.Cs** | Red Blood Corpuscles |
| **W.B.Cs** | White Blood Cells |
| **M.C.H.** | Mean Corpascular Hemoglobin |
| **M.C.H.C.** | Mean Corpascular Hemoglobin Concentration |
| **M.C.V.** | Mean Corpuscular Volume |
| **B.M.** | Bone Marrow |
| **B.M.T.** | Bone Marrow Transplantation |
| **P.A.M.S.** | Peri-Arterial Macrophage Sheath |
| **P.A.L.S.** | Peri-Arterial Lymphatic Sheath |
| **D.F.** | Desferrioxamine |
| **Ca DTPA** | Calcium Diethylene Triamine Pentacetic Acid |

## Definitions

| | |
|---|---|
| **M.C.H.** | Average weight of Hemoglobin / each cell. |
| **M.C.H.C.** | Average amount of hemoglobin in 100 ml R.B.Cs. |
| **M.C.V.** | Average volume of one R.B.C. |
| **Heinz Bodies** | Denaturated hemoglobin containing ferric iron. |
| **Howell-Jolly Bodies** | Abnormal inclusion bodies in R.B.Cs consists of nuclear reminants |
| **Hemosiderin Granules** | Aggregated form of ferritin |
| **Anisocytosis** | Variation of the size of R.B.Cs than what is present normally (non specific as it is present almost in all blood diseases) |
| **Poikilocytosis** | Abnormal variation of the shape of R.B.Cs. |

# Normal Values

| | |
|---|---|
| **1 milligram** | = 1000 microgram (Mg) |
| **1 microgram** | = 1000 nanogram (ng) |
| **1 nanogram** | = 1000 picogram (pg) |
| M.C.H. | = 27 - 32 pg |
| M.C.H.C. | = 31 - 35 gm/dL |
| M.C.V. | = 77 - 93 fL |
| Hb | = 14 - 16 gm/dL |
| Number of R.B.Cs. | = 4.500.000 - 5.250,000 /Cmm. |
| Number of W.B.Cs | = 5.000 - 11,000 /C.m.m. |
| Number of platelets | = 150,000 - 300.000 / C.m.m. |
| Hematocrite value | = 42 - 47 % |
| Reticulocytes | = 0.5 - 2.0% of R.B.Cs. |
| Fragility of R.B.Cs | = Begins at 0.45% Saline & complete at 0.35% saline. |
| Packed cell volume | = 40 - 54% |
| Diameter of R.B.C. | = 6.7 - 7.7 mM |
| Serum ferritin level | = Male (25 - 250 Mg/L)<br>Female (10 - 130 Mg/L) |

# INTRODUCTION

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Thalassemia was not recognized as clinical entity until 1925, when Thomas Cooley, a Detroit pediaterician, described a syndrome among children of Italian descent characterized by profound anemia, splenomegaly, and bony deformities (Lukens, 1993).

The thalassemias are hereditary anemias that occur because of mutations that affect the synthesis of hemoglobin. In B-thalassemia there is deficient synthesis of B-globin, whereas in $\alpha$–thalassemia there is deficient synthesis of $\alpha$–globin, Reduced synthesis of one of the two globin polypeptides leads to deficient hemoglobin accumulation, resulting in hypochromic, microcytic red cells. These red cell abnormalities are the most constant and characteristic features of the group of disorders (Nienhuis, 1993).

Growth retardation and delayed or absent puberty have been reported to be a common problem in transfusion-dependant thalassemia. (Borgana, et al., 1985). Such retardation includes height, weight and head circumference being more obvious with the height and weight. There is decreased growth velocity after 12 years old with absence of the normal adolescent growth spurt in most of these patients (Behrman & Kliegman, 1992).

Multiple factors share in such retardation of growth such as severe anemia due to hypersplenism, iron overload in the older children and zinc deficiency in most cases of thalassemic children (Alan, 1987).

The different lines of treatment for thalassemia are to support the patients and prevent the side effects of the disease. Also to prevent the decompensating changes of most tissues which usually appear in the second decade threatining the life of the patient. These lines include, early frequent blood transfusion, continuous iron chelation therapy, splenectomy if needed, with the supplementation of zinc, vitamin E and folic acid (Behrman & Kliegman, 1992).

# REVIEW OF LITERATURE

## BONE MARROW

Hematopoiesis is, to a large extent, outside the bone marrow (Extramedullary) in the fetus. During the first two months of fetal life, blood cells are formed in yolk sac and thereafter in the liver and spleen until the bone marrow progressively takes over this function during the last three months before birth. Extramedullary hematopoiesis may persist until shortly after birth if there is excessive blood destruction in utero, also extramedullary hematopoiesis may occur in later life if the bone marrow is unable to meet an increased demand as in hemorrhage or hemolysis (Pearson & Benz, 1984).

The marrow cells consist of a "Multiplication Pool" of dividing cells such as pro-erythroblasts, myeloblasts, promyelocytes and early myelocytes, and a "Maturation Pool" of non-dividing cells such as late normoblasts, late myelocytes, band cells and polymorphs. The healthy marrow is able to compensate for a six folds increase in the rate of red cell destruction before significant anemia develops. This is especially so in the case of chronic hemolysis, since an increase in erythroid marrow, at the expense of fatty marrow takes place. This is not possible, however, in infants since the marrow spaces are filled with erythroid marrow. Compensation for hemolysis is then less efficient, being confined largely to extramedullary hematopoiesis (Hann, 1992).

Differential counts of bone marrow (Behrman & Kliegman, 1992)

| Age | Blasts (%) | Promyelocytes (%) | Myelocyte & Metamyelocyte (%) | Bands & polymorph-nuclears % | Eosinophils (%) | Lymphocytes (%) | Nucleated R.B.C. (%) | Myeloid/ Erythroid ratio |
|---|---|---|---|---|---|---|---|---|
| Birth | 1 | 2 | 5 | 40 | 1 | 10 | 40 | 1.2:1 |
| 6 months 2 years | 0.5 | 0.5 | 8 | 30 | 1 | 40 | 20 | 2.0:1 |
| 6 years | 1 | 2 | 15 | 35 | 1 | 25 | 20 | 2.7:1 |
| 12 years | 1 | 2 | 20 | 40 | 1 | 15 | 20 | 3.2:1 |
| Adults | 1 | 2 | 21 | 44 | 2 | 10 | 20 | 3.5:1 |