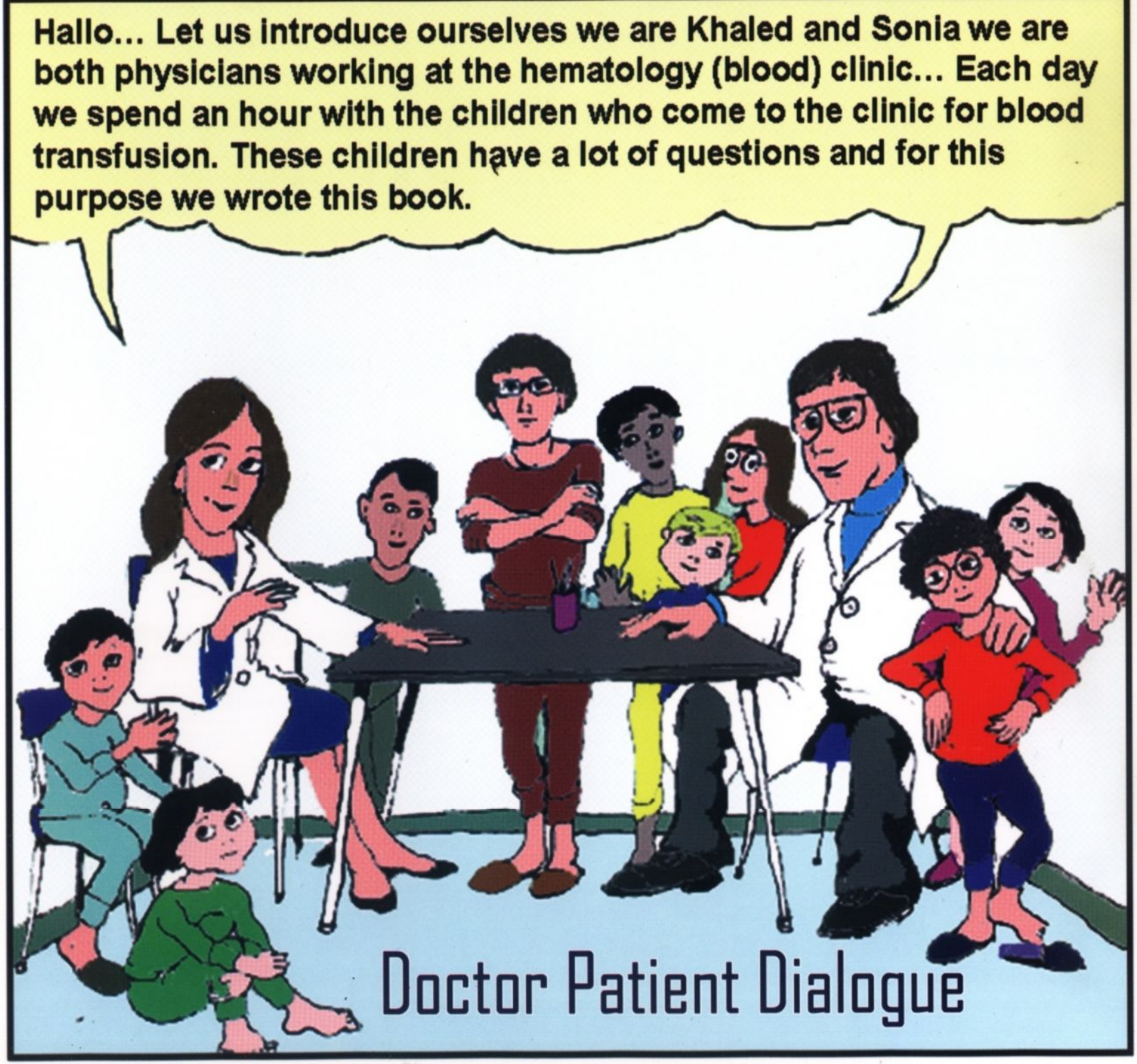
Hallo... Let us introduce ourselves we are Khaled and Sonia we are both physicians working at the hematology (blood) clinic... Each day we spend an hour with the children who come to the clinic for blood transfusion. These children have a lot of questions and for this purpose we wrote this book.
Doctor Patient Dialogue

Especially for those whom
I love the most

My dearest thalassaemia patients in Egypt and all over the world, I would like to dedicate this book to you, a hand-out that explains in a simple way what is thalassaemia, how to treat it; and how to prevent it.

The many years that I have spent with you so closely, living your ups and downs; made me only certain that you deserve nothing less than happiness and accomplishment of your goals in life. You have taught me that the key to reach out to your aspirations is Hope and Commitment to therapy.

Sincerely, I have learned that these are your tools for the life you are looking for; whatever your choice is, being a doctor, an engineer, an artist, a mother or a husband.

May you blossom in a prosperous future!

By you, and with you always.

Prof. Amal EL Beshlawy
Professor of Haematology – Cairo University
President of the Egyptian Thalassaemia Association ETA

Table of Contents

This Book is Designed and Prepared by

Prof. Amal EL Beshlawy

Professor of Pediatric Haematology – Cairo University
President of the Egyptian Thalassaemia Association ETA

&

Dr. Hisham Sedky Abdou

The Egyptian Thalassemia Association (ETA)

Introduction

Thalassemia is a common problem in Egypt. Carrier rate varies between 6 to 10%. It was estimated that more than 1000/1.5 million live birth /year are born with thalassemia in Egypt.

The number of patients in pediatric Hematology clinic of Cairo University hospitals the biggest hematology clinic in Egypt (Abou- El - Rish) is more than 2500.The Egyptian Thalassemia Association (ETA) was established in 1990 motivated and supported by The Thalassemia International Federation (TIF) in the Pediatric Hospital of Cairo University.

The members are the medical staff of our center together with doctors and professor from other Egyptian centers - patients and their parents .

Founder and president of ETA is Professor ***Amal El-Beshlawy,*** ***Prof. of pediatric Hematology in Cairo-University***.

Honorary president Her Excellency
Mrs Suzan Mubarak
First Lady of Egypt

Goals of ETA

- To improve quality of life and survival to thalassemia patients and support of their families in Egypt.
- To increase the population awareness and doctors awareness to thalassemia in Egypt through lectures, scientific meetings and conferences.
- Help in creating thalassemia centers in Cairo and other governerates in Egypt.
- Social and medical support to the patients and their families.
- Support the medical and research activities in the domain of thalassemia

Resources :

Mainly personal and non governemental organizations donations.

Achievements and activities

- Medical support to the new and follow up patients with financial cost of 200,000 US Dollars/ year.
- Social and financial support to the patients and parents in the religious and social events.
- Delivery of certificates to support the education and employment of our patients (50 / years).
- Held the annual international thalassemia conferences in the 8th of May (International Thalassemia day) which are attended by more than 900 Physicians from Egypt and the Arab countries also attended by our patients in a special session with the international experts.
- Support the sending of our thalassemia patients for bone marrow transplantation in Italy (Dr. Lucarelli center). According to agreement which support the BMT for 50 patients from Egypt free of charge.
- Initiating and supporting the training programs on thalassemia for Egyptian and Arab countries physicians (Iraq and Jordan...etc).
- Initiation and establishment of the Bone Marrow Transplantation Center in the Pediatric Hospital of Cairo University for cure of Thalassemia patients.
- Participating in the international trial for the new oral chelating drug .

Special achievements

- Motivate and support the issue of the Ministerial Decree in 1998 for the employment and education of the patients.

Future Plans

- Activates Bone Marrow Transplantation for our thalassemia patients who has a donor.
- Prevention of Thalassemia by premarital screening and prenatal diagnosis.
- Increasing the population awareness to thalassemia and its prevention.
- Promoting patients compliance to chelation therapy by the availability of the oral easy taken drugs.

Our Mission

- To prevent the increasing No. of new cases of thalassemia in Egypt.
- To secure the cure and good quality of life for our patients.

The Egyptian Thalassemia Association Board of Directors

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Mobile: 2012-312-4674

Hallo… Let us introduce ourselves we are Khaled and Sonia we are both physicians working at the hematology (blood) clinic… Each day we spend an hour with the children who come to the clinic for blood transfusion. These children have a lot of questions and for this purpose we wrote this book.

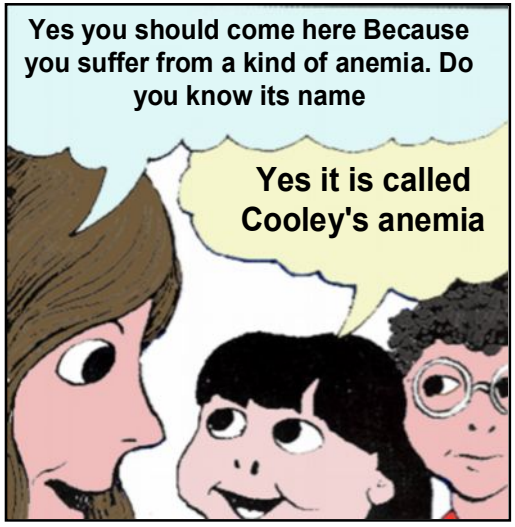
Khaled! I was not intending to come to the hospital today, I want to go to the school.

But why should you come to the hospital Aly?

Because we are sick and affected by a kind of anemia

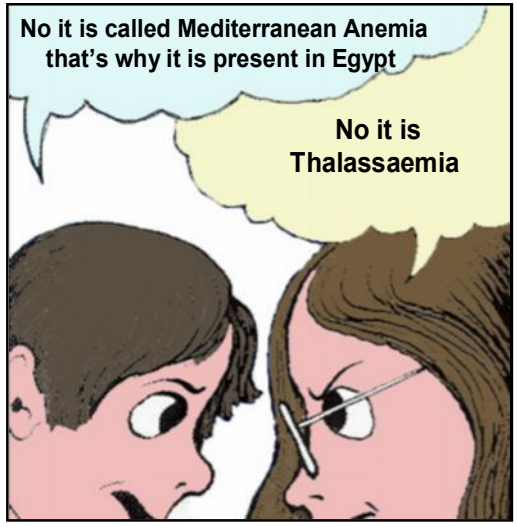
Yes you should come here Because you suffer from a kind of anemia. Do you know its name

Yes it is called Cooley's anemia



No it is called Mediterranean Anemia that's why it is present in Egypt

No it is Thalassaemia

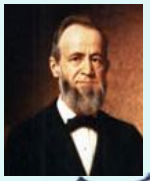


So it has many names, but what does that mean

It is called Mediterranean Anemia as it is present in the countries around the Mediterranean sea

it is present in the countries around the Mediterranean sea as Egypt, Syria, Greece, Italy, Lebanon, and others
That is why it is called Thalassaemia
Haemia = Anemia Thalass = Sea

It is also called Cooley's anemia as it was discovered by an American Scientist called Thomas Cooley 1925. as he was treating children who came from Italy. This Anemia is also present in Africa and Asia



But what is the meaning of anemia

It means that we do not have enough blood

Or that we need blood transfusion

But what is blood made of

lasma Red Blood Cells White Blood Cells Platelets

Let us carefully see what the blood is formed of. Today at 9 am we took a blood sample from Howayda and we put it in this tube

As you see it is no more one color.
The upper part is bright yellow and it is called plasma
and the lower part is dark red as it contain red blood cells

What about the white blood cells?

The no of white blood cells is small in comparison to the red blood cells that is why it is difficult to see them, the same situation is for the platelets.

Omar.. What is the color of the transfused blood to you?

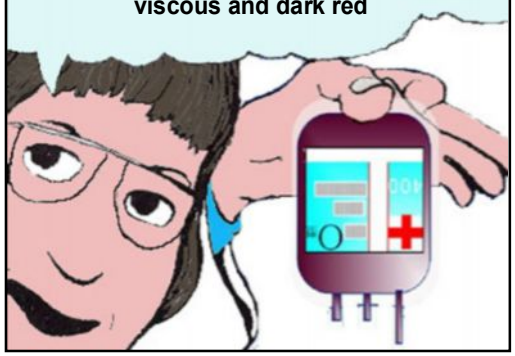
Dark red

Surely it contain red blood cells only

Sonia how blood is available ?

There are people who donate their blood

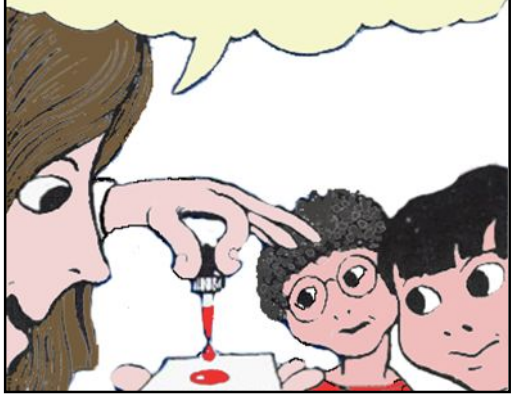
They are known as blood donors, we separate the red blood cells from the blood then we pack it in the blood transfusion bags. This is why it is viscous and dark red



Let us see from what are the red blood cells formed of and what is its function

These are tiny red balls, they are too small that we can not see them by our eyes

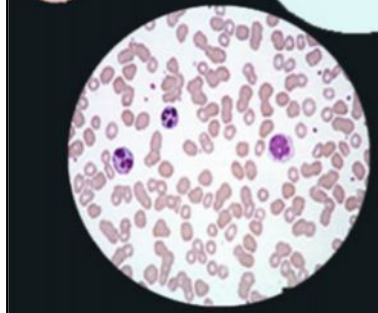
To see these cells we must take a drop and place it on a glass plate



And then to put them under a microscope to see

This is what we will see

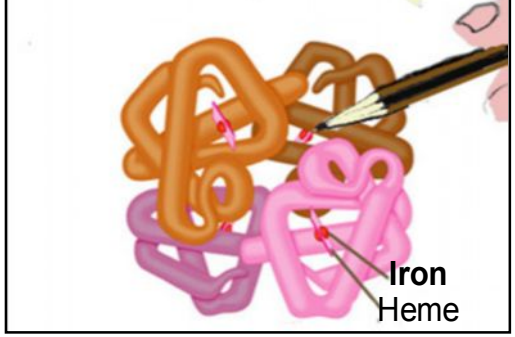
In this drop of blood there is more than 200 million red blood cell

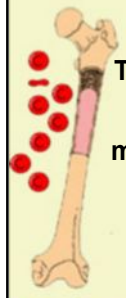


What about Hemoglobin

The hemoglobin is inside the red blood cells which act as a bag that contain a no of hemoglobin molecules

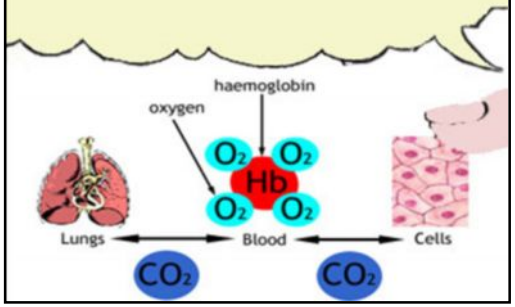
Each molecule of hemoglobin has a molecule of iron attached to it





The red blood cells are formed by the bone marrow and it moves to the blood to circulate in the whole body

The red blood cells move to the lung where oxygen combines with hemoglobin to be transported to the whole body, and give the energy



And anemia as we said means that there are no enough red blood cells. Your bone marrow is unable to manufacture potent red blood cells

Anemia means that there are few red blood cells and that this red cells are not fully build. They are smaller in size like balls empty from hemoglobin so unable to carry oxygen that is why it is important to give blood transfusion.

Khaled is it a must to come every month for blood transfusion? Why is it every month?

The red blood cells perform a tedious job in the body and it is always moving so after a while it gets tired

In your case the red blood cells received by transfusion live for one month after which it must be replaced by blood transfusion

Yesterday khaled told me that my hemoglobin level is 8 gm and that I am late for transfusion

Yes you feel tired and exhausted because you are late for transfusion

khaled what should be my hemoglobin level ?

A person of your age Hassan his hemoglobin level should be 13 gm. To keep your activity and vitality the hemoglobin level must not be less than 9- 11 gm.

To maintain a good health you must come every month at the transfusion time and never to be late

But if we eat a lot we will not need to come here frequently

NO

This is not true !

Ahmed is right. You do not need to eat a lot as the food quantity does not affect the blood transfusion

But does the effort and frequency of playing affect the number of transfusions?

No it does not . change whether you are moving or playing much more or less

Two months ago I had a fever and when I came to the clinic they found that my hemoglobin level was low

When a child has fever the transfused red cells do not last a month and the child must come to the clinic few days earlier than the time of his monthly transfusion

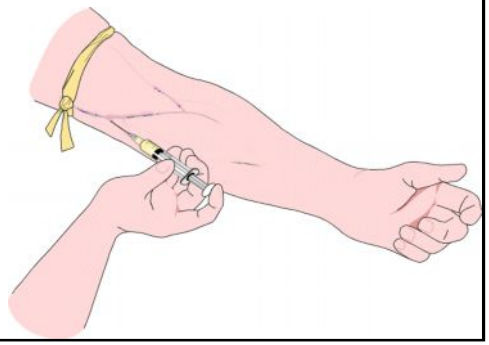
Sonia when I grow older , I will need blood transfusion?

Yes certainly as your bone marrow is unable to manufacture enough potent red blood cells

Sherief it is time to take your blood sample

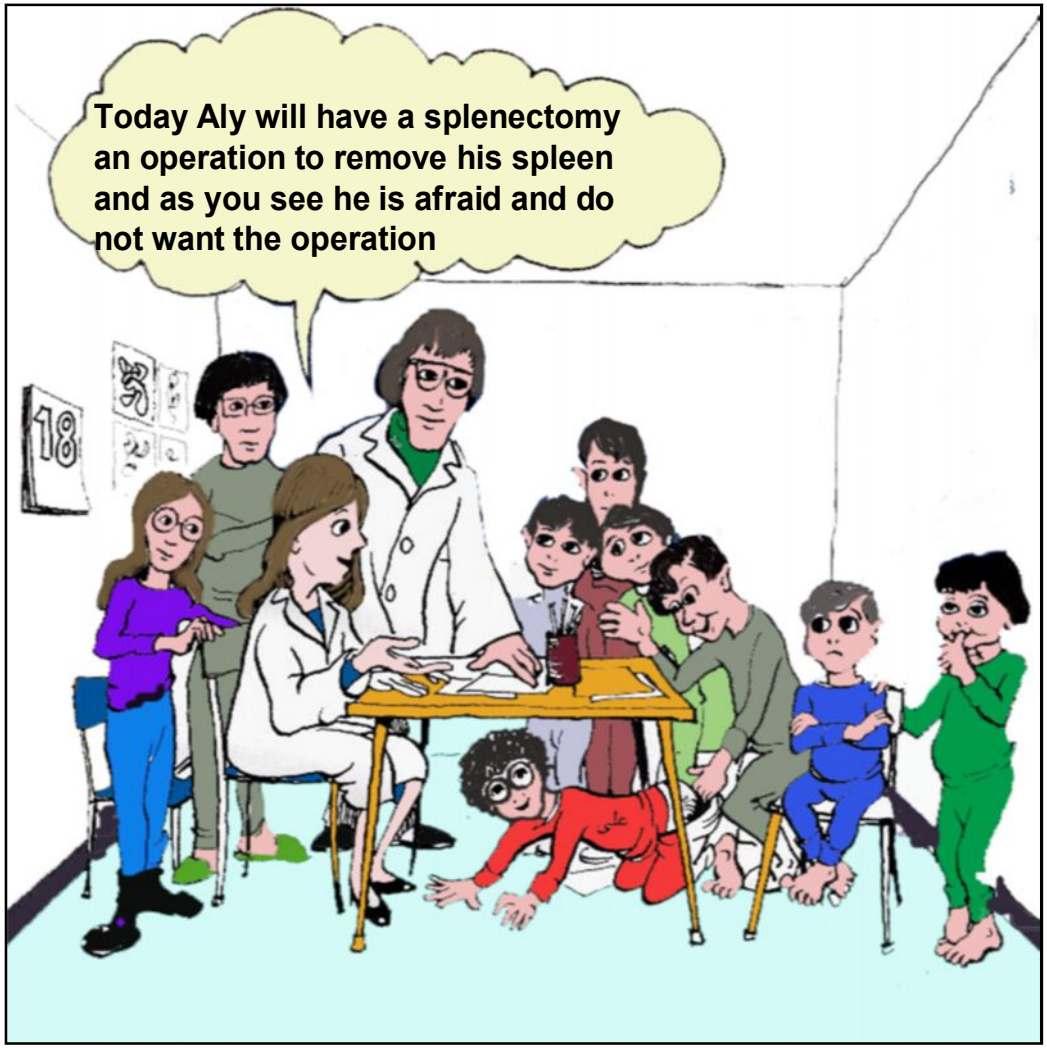
Oh my God..! Why should I give a blood sample Sonia?

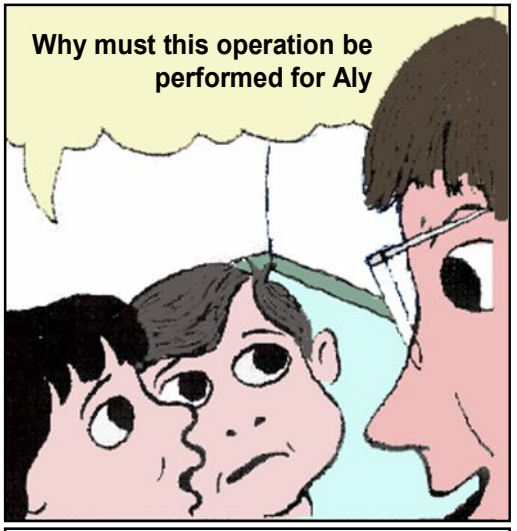
We take a blood sample to measure your hemoglobin level to decide whether you need blood transfusion or not.

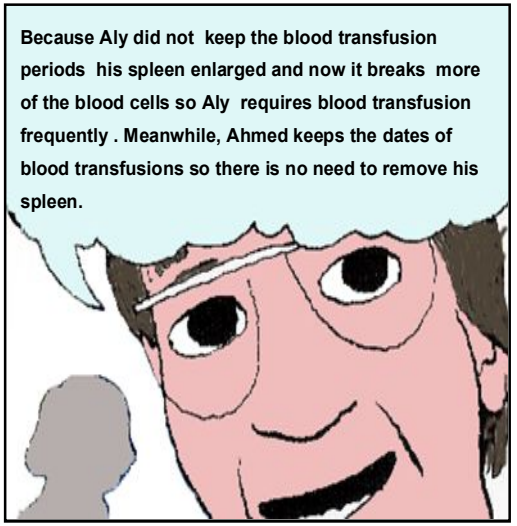


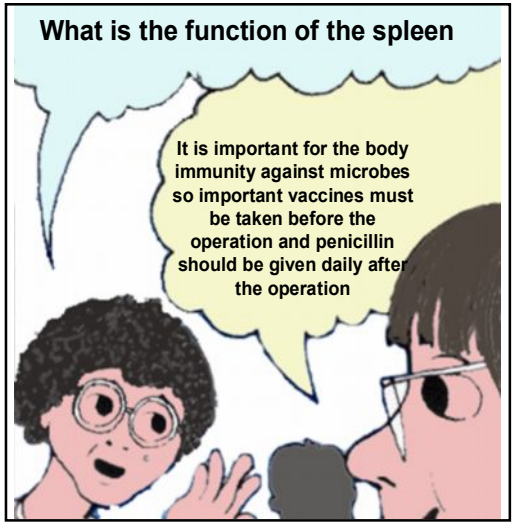
That is enough for today
Good bye Friends

Good bye ....See you later

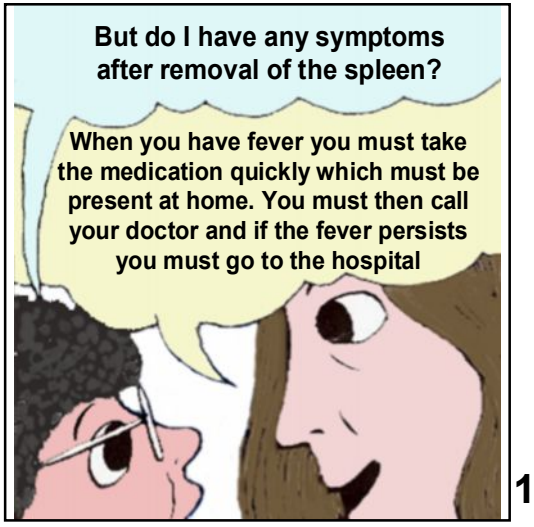

Today Aly will have a splenectomy an operation to remove his spleen and as you see he is afraid and do not want the operation


Why must this operation be performed for Aly


Because Aly did not keep the blood transfusion periods his spleen enlarged and now it breaks more of the blood cells so Aly requires blood transfusion frequently . Meanwhile, Ahmed keeps the dates of blood transfusions so there is no need to remove his spleen.


What is the function of the spleen
It is important for the body immunity against microbes so important vaccines must be taken before the operation and penicillin should be given daily after the operation

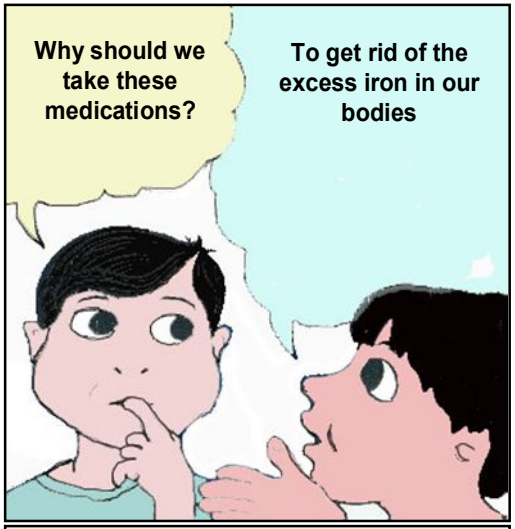

Aly Will need for transfusion after that ??
He will not be need for a while. But later we start monthly transfusion. The hemoglobin level will remain high


But do I have any symptoms after removal of the spleen?
When you have fever you must take the medication quickly which must be present at home. You must then call your doctor and if the fever persists you must go to the hospital

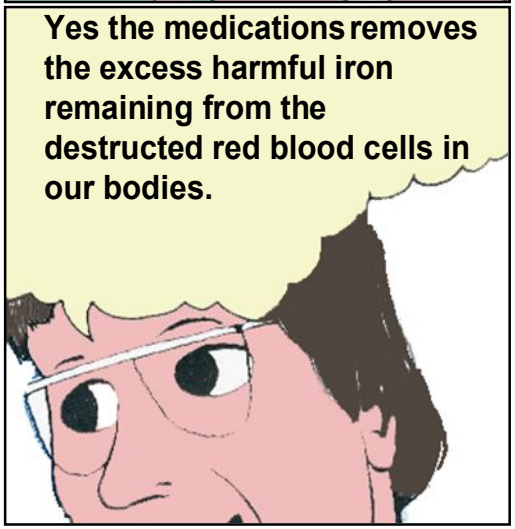
Today we have Salwa she takes the medication that make the body get rid of excess iron

Why should we take these medications?

To get rid of the excess iron in our bodies

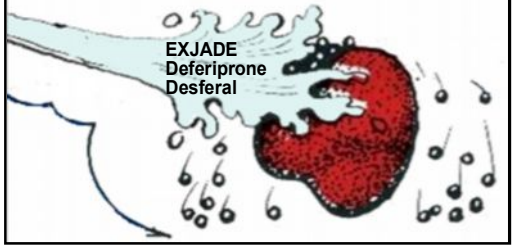


Yes the medications removes the excess harmful iron remaining from the destructed red blood cells in our bodies.

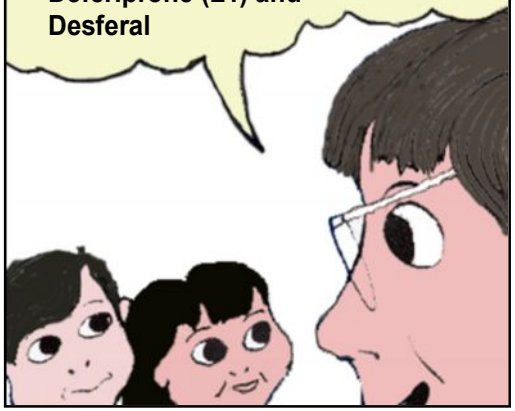


The solid particles of iron which are present with the hemoglobin remains inside our bodies then it accumulate and deposited in the heart, liver, glands and under the skin

And as we all know the iron particles are solid in nature mean while the body organs have to be flexible to perform their functions that is why we need a drug to remove these iron particles from our bodies.



The name of the drugs are
EXJADE,
Deferiprone (L1) and
Desferal



EXJADE and (L1) can be taken by mouth while Desferal is taken by injection under the skin by a pump for a period of 8-10 hours each day in five days of the week, many people find this commitment difficult, although very effective in getting rid of iron. .

What are the drugs taken by mouth? I can not regularly take the desferal and iron will accumulate in my body

There is the L1 but it can not be taken before the age of 4 years, it is in the form of tablets taken three times a day and follow-up of liver and white blood cell is important to prevent complications that might rarely occur.

What about the EXJADE which is taken by mouth

It is a new medicine very easy to use. Thlassemia patients two years old can take it

EXGADE has no taste and is taken once a day for half an hour before breakfast, different doses of 125 gm and 250 gm, 500 gm are present and is given according to weight of the patient and the quantity of iron accumulated in his Body.

•It is available in three concentrations 125 mg, 250mg and 500 mg tablets

It dissolves in water or orange juice using a glass cup and a plastic spoon

In 100- 200ml according to the drug concentration

Is the EXJADE effective in removing iron from my body?

Yes EXJADE is very effective it gets rid of excess iron in our bodies, it is taken by mouth once daily and its action lasts for 24 hours

EXJADE is active for 24 hours and get rid of the harmful iron which is present always in the blood and infiltrate important organs as the liver, heart and endocrine glands responsible forgrowth and thus these organs do not perform their functions to well.

Iron available in many foods and we must be careful in the quality of the food we eat for example, there is a lot of iron in the liver, red meat and some other foods. And we must know that drinking tea during eating reduces the absorption of iron

Did you hear that Salwa? There are many things you should do may be you do not like to do but this will keep you healthy and live a normal life

You need to do certain tests over a period of time to assess the stored iron, heart, liver functions and growth

Good Bye See you later

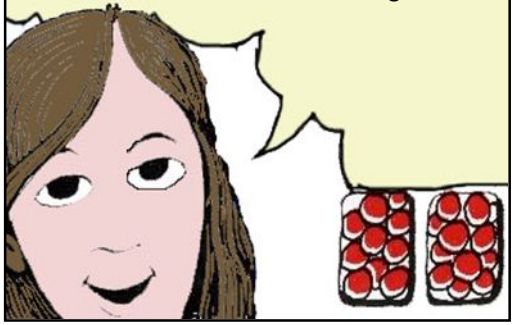
Salwa wants to ask a question

Why are we (I and my brother) sick while my sister and other brother are not?

Mediterranean anemia (Thalassemia) is a hereditary disease that means it is passed from the parents to their children

You have heard the word carrier as other children but you did not understand its meaning .

Let us see if this is the case in the thaalssemia.. To manufacture normal healthy red blood cells we must have the two normal signs



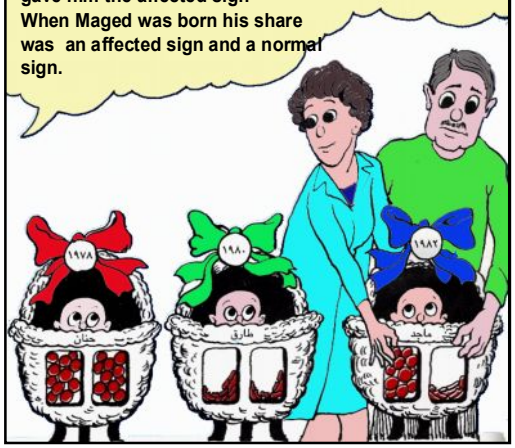
If the two signs are normal then the person manufacture normal red cells and pass to his child normal signs

But if one sign is normal and the other is defective that means that the person is a carrier. He manufacture normal blood cells but could pass the defective sign to his child

But if the parent is sick like you he can only give his son the defective sign

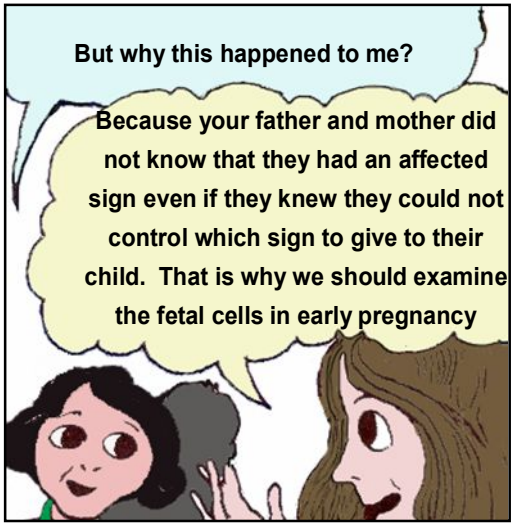
Let us see now how to answer Salwa's question. Since her father and mother are carriers of the disease character and they are both normal they can pass defective signs to their children.

When Hanan was born each parent gave her the normal sign. But when Tarek was born each parent gave him the affected sign
When Maged was born his share was an affected sign and a normal sign.



But why this happened to me?

Because your father and mother did not know that they had an affected sign even if they knew they could not control which sign to give to their child. That is why we should examine the fetal cells in early pregnancy

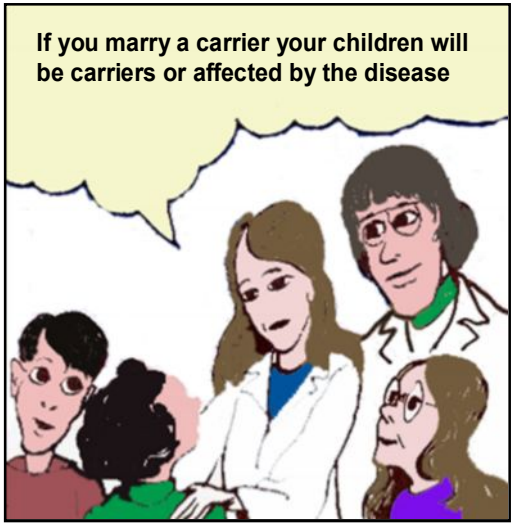


That is why I should not marry a carrier or an affected person , I should marry a normal non affected person

Yes if you marry an affected person all your children will be affected.

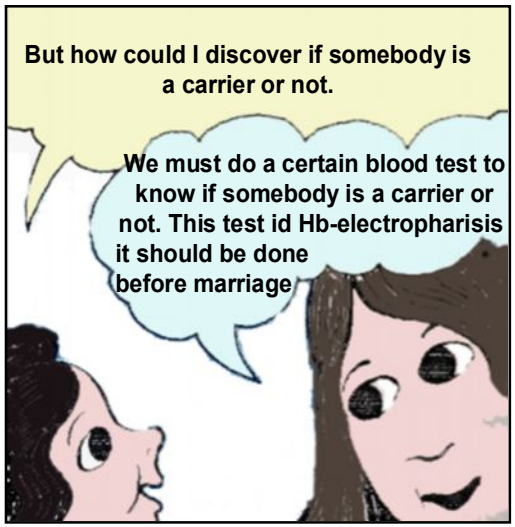
If you marry a normal person all your children will be carriers.

If you marry a carrier your children will be carriers or affected by the disease



But how could I discover if somebody is a carrier or not.

We must do a certain blood test to know if somebody is a carrier or not. This test id Hb-electropharisis it should be done before marriage



Good bye .. If you have any question you can visit your doctor at the hospital

Good Bye
See you later

With the Complement of
The Egyptian Thalassemia Association (ETA)