PATIENT-HELD RECORD

United Kingdom Thalassaemia Society
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Version: 1.1

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About This Booklet.

This is a booklet for a patient's personal use. We recommend however that you take it with you on every hospital appointment so that you can fill it in with your doctor or nurse specialist.

It is hoped that it will help you to participate in the care and treatment given to you by the Health Professional. It will act as a reminder as to which tests and medication you should be receiving.

This booklet is meant to act as a general guide only. Individual treatment regimes differ and you should always follow the advice of your thalassaemia doctor.

The UKTS is your Society and is here to help patients receive the best possible treatment. The Society apart from funding research is also actively supporting awareness amongst the at risk population.

Your input and support is vital if we are to succeed in our efforts. Please do not hesitate to contact the UKTS should you require any further information.

Thank you,

The U. K. Thalassaemia Society would like to express its grateful thanks and appreciation to **Dr Paul Telfer** and **Sister Emma Prescott**, for their commitment and dedication in writing and editing this booklet. Additionally we would like to thank all the doctors who have contributed to and supported this project.

Dr Vasili Berdoukas
Dr Philip Darbyshire
Dr Bernard Davies
Dr Sally Kinsey
Dr Melanie Pollitzer
Prof John Porter
Dr Beatrix Wonke
Dr Christine Wright
Dr Anne Yardumian

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APOTEX RESEARCH INC.,
NOVARTIS PHARMACEUTICALS.

Tips for Parents

We hope that the following tips will help you to cope with your child's treatment.

Ask for information to be repeated – Don't be afraid to ask if you are unsure about something or can't remember what you had previously been told. When you visit the hospital with your child make a list of things you want to ask your doctor or nurse specialist. If in doubt, ASK AGAIN – your doctor and nurse specialist would prefer you to ask rather than be anxious because you are unsure about something.

Help your child to understand – Talk to your child about their treatment so that they understand what is happening and why. If you want help or advice on how to do this, talk to your doctor or nurse specialist.

Help your child to relax – Some children may become anxious about visiting the hospital. Find ways of helping your child to relax. It may be helpful if you can include a fun/enjoyable activity with the end of the hospital visit.

Maintain consistency and discipline – Parents often find it hard to discipline their thalassaemic child. However, being consistent with "rules" and discipline is important for your child and the rest of the family. Don't forget to give lots of praise and encouragement for times when your child has coped well with their treatment.

Supporting Siblings

Sometimes brothers and sisters may feel that the child with thalassaemia always “comes first” and that their needs are ignored and rejected. They may sometimes feel worried about their sibling but be afraid to ask questions. You may notice changes in their behaviour – younger children may become clingy and reluctant to be away from you; older children may have difficulty concentrating at school. Sometimes siblings can act in a jealous way and become more demanding of your attention. They may become angry and argue more frequently. Here are a few suggestions on how to help your other children cope.

Quality time – brothers and sisters may feel that the thalassaemic child gets all the parents’ attention. Try to have special times for the other children in the family so that they can sometimes have your undivided attention.

Help brothers and sisters to understand – tell your thalassaemic child’s brothers and sisters about the treatment, what is happening and why. Sometimes children can fill in gaps in their knowledge by imagination and come to the wrong conclusions. Ask your doctor or nurse specialist if you need help or advice in helping your children to understand.

Parents – Don't Forget Yourselves!

Parents of thalassaemic children may experience unwarranted feelings of guilt, which can cause them to feel that they can never leave their child even for a short time. However it will help you to cope with your child's treatment if you have the occasional "time out". Give yourself permission to take a break, relax and do the things you enjoy. Your child will manage without you providing you have a competent person who can look after them while you are out. If you are feeling depressed and finding it difficult to cope with your child's treatment speak to your doctor or nurse specialist.

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PERSONAL DETAILS

Personal Details

Name	
Address	
Address	
Address	
Address	
Tel No	
Mobile No	
E-mail	
Date Of Birth	
Thalassaemia Doctor	
Thalassaemia Doctor Tel. No	
Thalassaemia Nurse	
Thalassaemia Nurse Tel. No	
Cardiologist	
Endocrinologist	
Other Specialist	
GP	
GP's Tel. No	

In Case of Emergency please contact

Name	
Relationship	
Address	
Address	
Address	
Tel. No	
Mobile No	

Other useful contacts

In case of an emergency

Occasionally, medical problems occur suddenly when you are far from home and from your own clinic. You may have to attend a hospital emergency department and be seen by a doctor who doesn't know you, and has very little experience of managing thalassaemia.

It is important to give the doctor the information needed to treat you quickly and correctly. If you bring this file with you, and explain what it is, it will give the doctor a lot of vital information. Your file should contain the number for your thalassaemia doctor, and you should point it out and ask he/she is contacted. This will ensure good communication so that the best plan of treatment can be discussed with the emergency doctor.

Thalassaemia Record: Basic Details

			COMMENTS
Your thalassaemia mutation/DNA profile			
Blood Group			
Red cell antibodies	**YES / NO**		
Splenectomy	**YES / NO**	when	
Cholecystectomy	**YES / NO**		
Heart problems	**YES / NO**		
Hepatitis C infection	**YES / NO**		
Diabetes	**YES / NO**		
Insulin:	**YES / NO**		
Dosage:			
Hypothyroid	**YES / NO**		
Hypoparathyroid	**YES / NO**		
Hypogonadism	**YES / NO**		
Osteoporosis	**YES / NO**		
Other medical conditions			
Usual medication			
ALLERGIC TO:			

BIOCHEMISTRY BLOOD TEST RECORD.

FILL EVERY TIME YOU HAVE BIOCHEMISTRY TESTS

Date	ALT	AST	Y-GT	Alkaline phosphatase	Phosphate	Calcium	Albumen	Bilirubin	Creatinine	Urea	Urate	Vit. D	Comments

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BLOOD TRANSFUSION

BLOOD TRANSFUSION

In thalassaemia, there is a deficiency of haemoglobin, the oxygen carrying protein. As a result, the red blood cells cannot carry sufficient oxygen to the body. This situation is called anemia, and results in poor growth, tiredness, lethargy and susceptibility to infection. Ultimately, there is severe strain on the heart. Another result is expansion of the red bone marrow leading to enlargement, deformity and weakness of the bones, and enlargement of the spleen and liver.

The main aims of transfusion are

- To correct the anemia
- To inhibit the expansion of the bone marrow

If transfusions are given according to the recommended guidelines, we should expect a good level of health and fitness, together with normal growth and development during the early years of childhood.

The following guidelines for transfusion should be followed:

- The hemoglobin level should always be kept above 9.5-10 g/dl
- Transfusions can be given every 2-4 weeks
- The amount of blood transfused should be 10-20 milliliters per kilogram body weight
- Over the course of a year, the amount of blood transfused should not be more than 200 milliliters per kilogram body weight of UK blood
- You should keep a card (issued by the blood transfusion center) with details of your blood group and any red cell antibodies previously detected
- Transfusion reactions should be taken seriously and investigated by your doctor so that measures can be taken to avoid recurrence

You should keep a record of dates of transfusion, amount of blood given and your haemoglobin level before the transfusion, any side-effects of transfusion are helpful in ensuring the guidelines are adhered to.

THALASSAEMIA TRANSFUSION RECORD.

Fill in every time you have a transfusion

Date	Weight	Hb level before transfusion	Blood volume (mls) or weight (mgs)	Desferal IV/ dose	Reaction YES/NO	Comments

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CHELATION

IRON CHELATION

Red blood cells contain iron. Regular transfusion will result in iron loading, which eventually will cause serious damage, particularly to the heart, liver, and hormone glands. Iron can be removed from the body by treating with 'chelating' drugs. The standard treatment is desferrioxamine (Desferal), by subcutaneous infusion using a syringe pump. If the treatment is taken as recommended most of the damage done by the iron can be avoided. There is an alternative oral drug, deferiprone (Ferriprox), which can be used in older children and adults if it is not possible to take desferrioxamine. Chelation treatment is very hard, and to keep it going, you may need help at times from staff, support groups as well as your family.

Recommended dosage

Desferrioxamine

The standard dose in childhood is 20-40 milligrams per kilogram body weight over 8-12 hours 5-6 nights per week, and up to 50 mg/kg in adults. In very young children (<3 years), the dose and frequency should be even lower. The dose will be adjusted by your doctor depending on age and degree of iron overload

Deferiprone

75mg per kg body weight per day taken in three doses. Occasionally a higher dose may be recommended under very close supervision.

Combination: Desferrioxamine/ Deferiprone

Research is currently taking place into treatment using desferrioxamine and deferiprone in combination.

Is the chelation working?

This should be assessed at least every three months

1. Check on how many treatments have been given and problems with the treatment
2. Medical check to detect any signs of iron damage
3. Blood test for serum ferritin (see page 19).
4. Your doctor may request extra tests such as liver biopsy, Magnetic resonance scan (MRI), SQUID scan

Are there any side effects?

Rarely, desferrioxamine can cause damage, particularly to the bones, ears and eyes. This happens if the dose is too high and/or the iron stores are relatively low. These investigations should identify any problems:

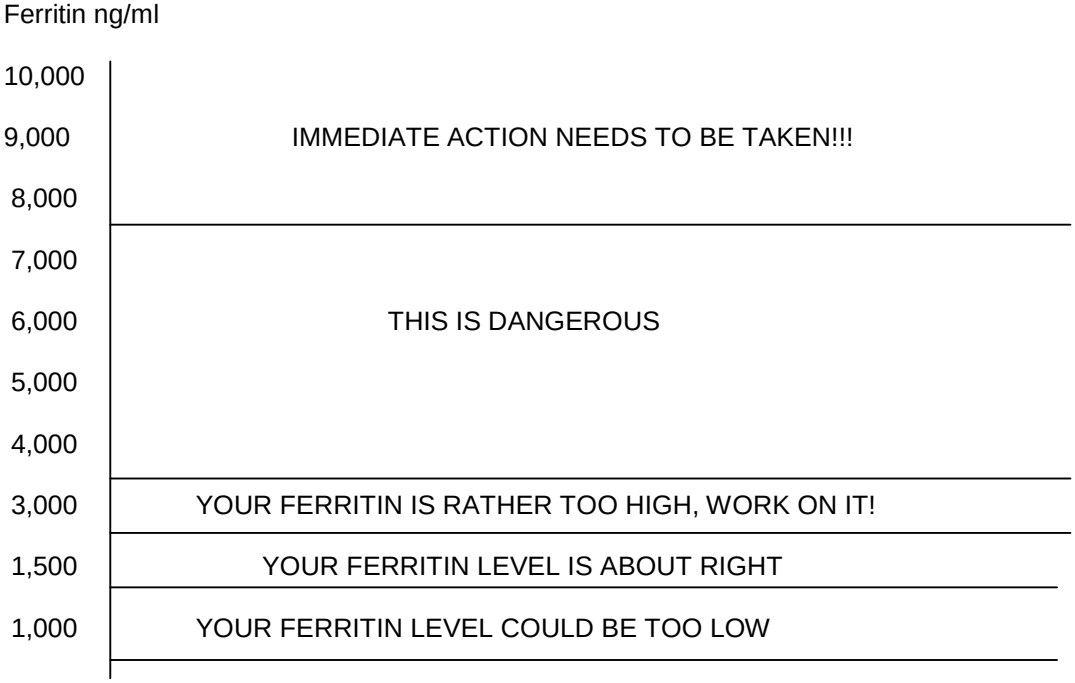
- Standing height, sitting height, growth velocity during childhood
- Hearing and Eye tests
- Also it is important to remember that desferrioxamine can encourage a serious infection called Yersinia. For this reason, you should not take Desferal if you have a fever, or a bad stomach upset (abdominal pain, diarrhoea). **CONTACT YOUR HAEMATOLOGIST IMMEDIATELY.**

With deferiprone, the side effects are different. The white blood cell count can fall; joints can become painful and swollen, stomach upset, or weight gain. Blood tests are needed at least once per month (more frequently at the beginning of treatment)

- Full blood count including white blood count. Weekly neutrophil count for first 6 weeks, then monthly. In case of fever, stop taking deferiprone immediately (this could be due to a drop in white blood cells). **CONTACT YOUR HAEMATOLOGIST IMMEDIATELY.**
- Liver tests
- Zinc level (every 3 months)

WHAT DOES YOUR SERUM FERRITIN MEAN?

Serum ferritin levels do not always reflect levels of iron in the heart and should not be relied upon on its own. There are other tests that your doctor will use to monitor your iron stores, e.g. SQUID, liver biopsy, MRI and others.



Chelation Calendar

Month	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
JAN																															
FEB																															
MAR																															
APR																															
MAY																															
JUN																															
JUL																															
AUG																															
SEP																															
OCT																															
NOV																															
DEC																															

Use the above calendar to keep a record of your chelation.

For Example:

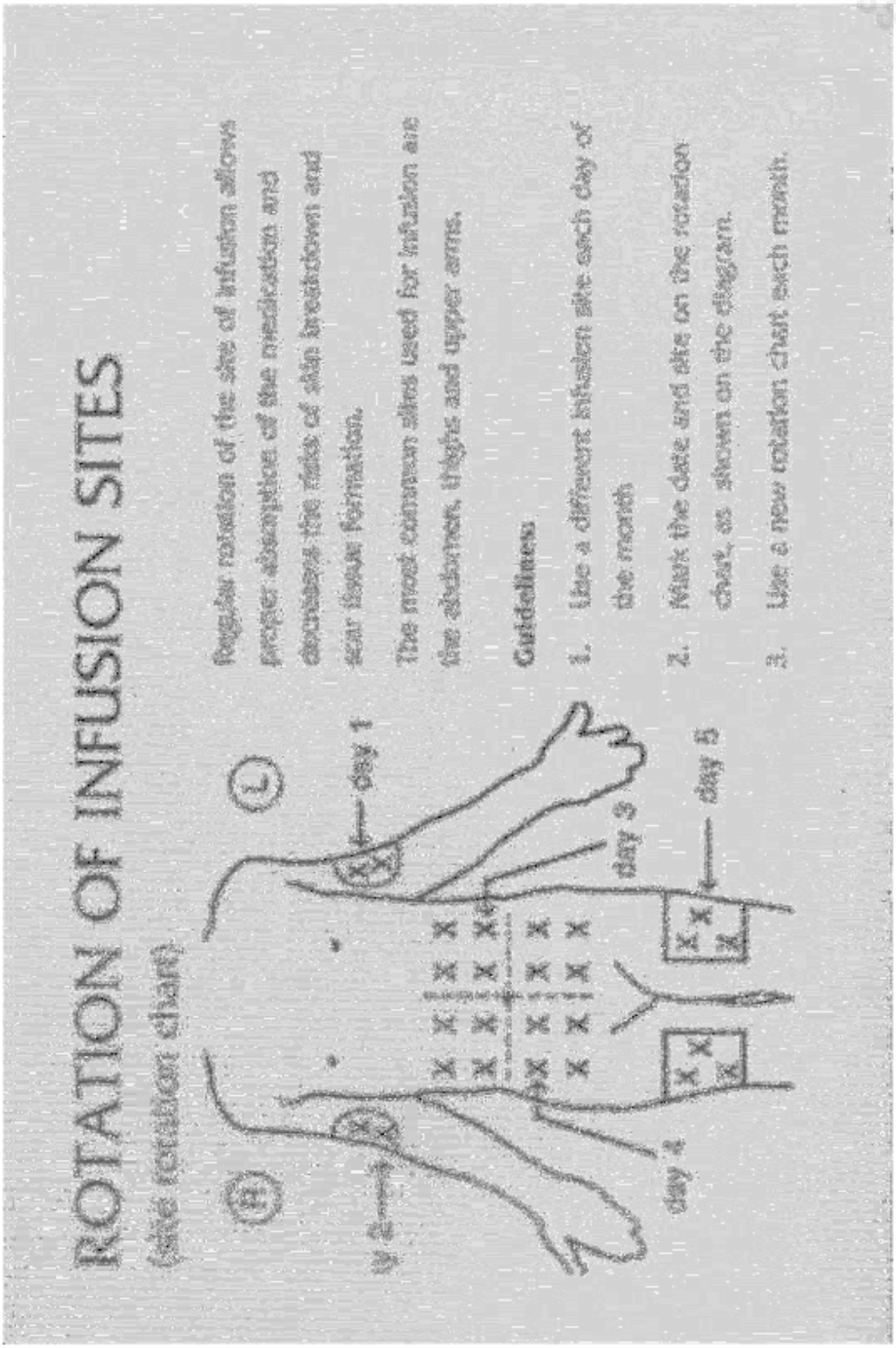
Use Red for Desferal days

Use Blue for Ferriprox / L1 / Deferiprone days

Use Green for combination days

Remember don't cheat ☺

ROTATION OF INFUSION SITES



DESFERAL IRON CHELATION RECORD

Fill in record every three months when you see your doctor, or if the prescription is changed

Date	Weight	Desferal dose (in milligrams or bottles used)	Desferal frequency (times per week)	Ferritin level	Hearing test (normal or abnormal)	Eye check (normal or abnormal)	Other tests and Comments	Side effects
				3 monthly	Annual	Annual		

DEFERIPRONE IRON CHELATION RECORD

Fill in record every three months when you see your doctor, or if the prescription is changed

Date	Weight	Deferiprone dose (number of tablets per day)	Ferritin level	Neutrophil count	Other tests and comments	Side effects

COMBINATION IRON CHELATION RECORD

Fill in record every month when you see your doctor, or if the prescription is changed

Date	Weight	Desferal dose (in milligrams or bottles used)	Desferal frequency (times per week)	Ferritin level	Hearing test (normal or abnormal)	Eye check (normal or abnormal)	Deferiprone dose (number of tablets per day)	Neutrophil count	Other tests and comments	Side effects
				3 monthly	Annual	Annual		Weekly		

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HEART PROBLEMS

HEART PROBLEMS

If iron chelation treatment is inadequate, iron levels can build up and cause damage to the heart. This is potentially very dangerous. Warning signs of heart problems include episodes of faintness, dizziness, and palpitations, getting unusually short of breath, swelling of the feet, ankles or face. If there is any sign of heart problems, urgent treatment is needed to prevent serious complications. This treatment will normally include specialist advice about increasing the chelation treatment, and use of heart drugs.

In your clinic, there should be a heart specialist who works closely with your doctor. Alternatively, you may be offered an appointment at a specialist thalassaemia heart clinic. From the age of 10, you should have a heart check-up every year this will include:

- Physical examination
- Electrocardiograph (ECG)
- Echocardiograph
- MRI scan of the heart is currently being evaluated in London. Ask your doctor about this

HEART ASSESSMENT RECORD.

Fill in every time you see the heart specialist(recommended once very 6 months) or have a heart problem

Date	Heart Clinic, (ECHO or MRI)	ECHO Results	MRI Results	Heart	Comments
		(normal or abnormal		medication prescribed	

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HORMONE PROBLEMS

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Iron overload causes damage to the hormone glands, and impairs the action of some hormones. This damage can occur even when chelation treatment is relatively good. In general, once this damage has occurred, it cannot be reversed, so it is important to detect the earliest signs if damage is to be prevented.

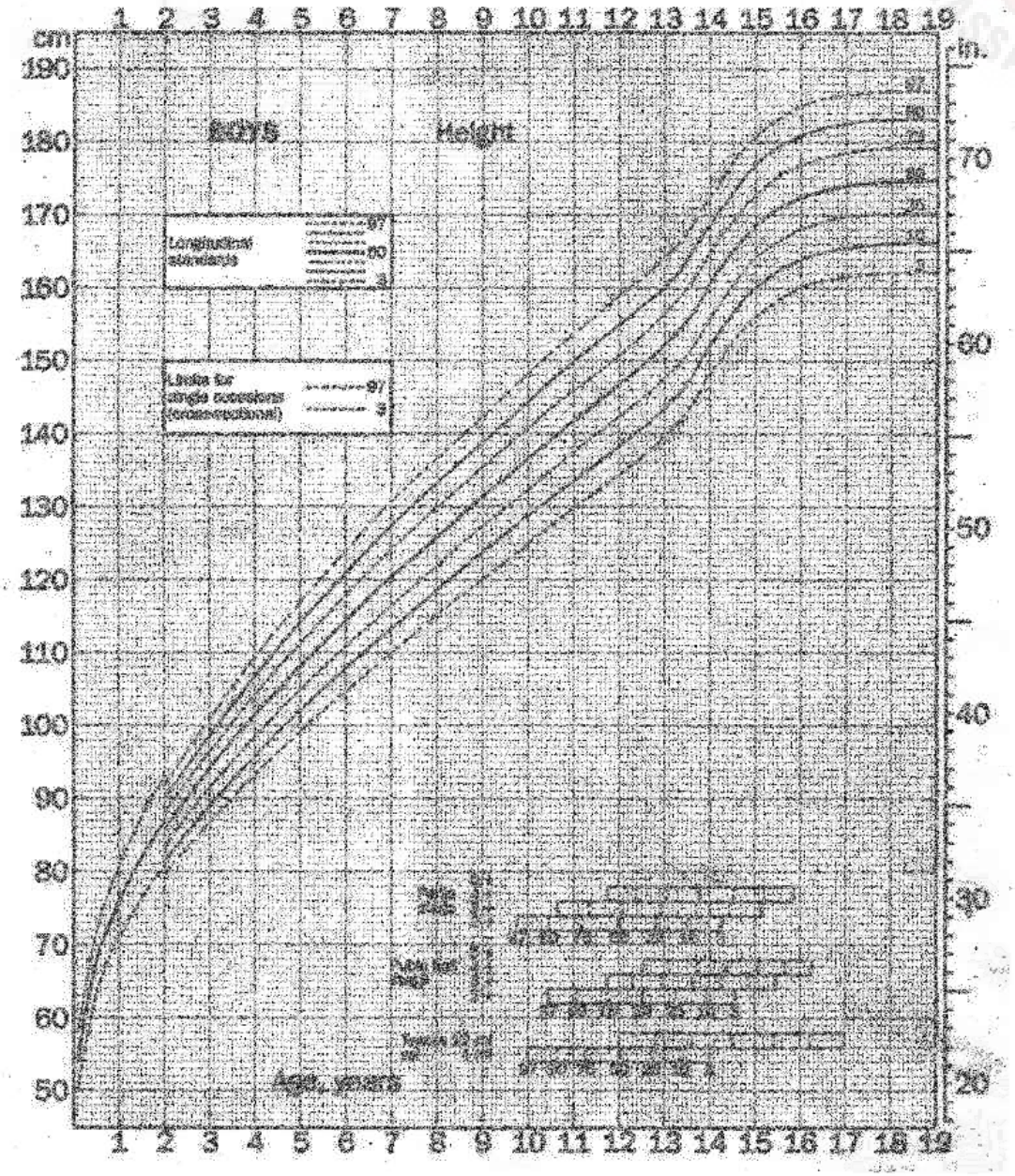
During childhood, growth and development should be assessed every 3 months with height, weight (plotted on growth chart), and in teenage years, assessment of puberty. Hormone tests are usually done every year from age 10. You should see a hormone specialist at least once every year, and more frequently if taking hormone treatments. The most frequent complications are:

- Growth hormone deficiency. If not treated, the result can be a failure to achieve the expected growth in height.
- 'Gonadotrophin' deficiency. These hormones are secreted by the pituitary gland, and are important for progression through puberty and in maintaining fertility. If damage has occurred, hormone treatment can be given to stimulate puberty and fertility
- Diabetes. This can be due to damage to the liver and/or to the insulin-secreting cells in the pancreas. Iron overload is an important cause, but other factors may also be involved. Diabetes is diagnosed with an oral glucose tolerance test, or with a simple fasting glucose test. It can cause damage, for example to the eyes, nerves, kidneys, blood vessels. If diabetes develops, specialist treatment is needed to advise about diet, exercise and diabetic drugs. The aim of treatment is to keep sugar levels within an acceptable range and to avoid long-term damage.
- Hypothyroidism: Under active thyroid gland. The usual treatment is with regular thyroxin tablets
- Hypoparathyroidism: Results in abnormally low serum calcium level – causing tetany (muscle spasm). Treatment is with Vitamin D.

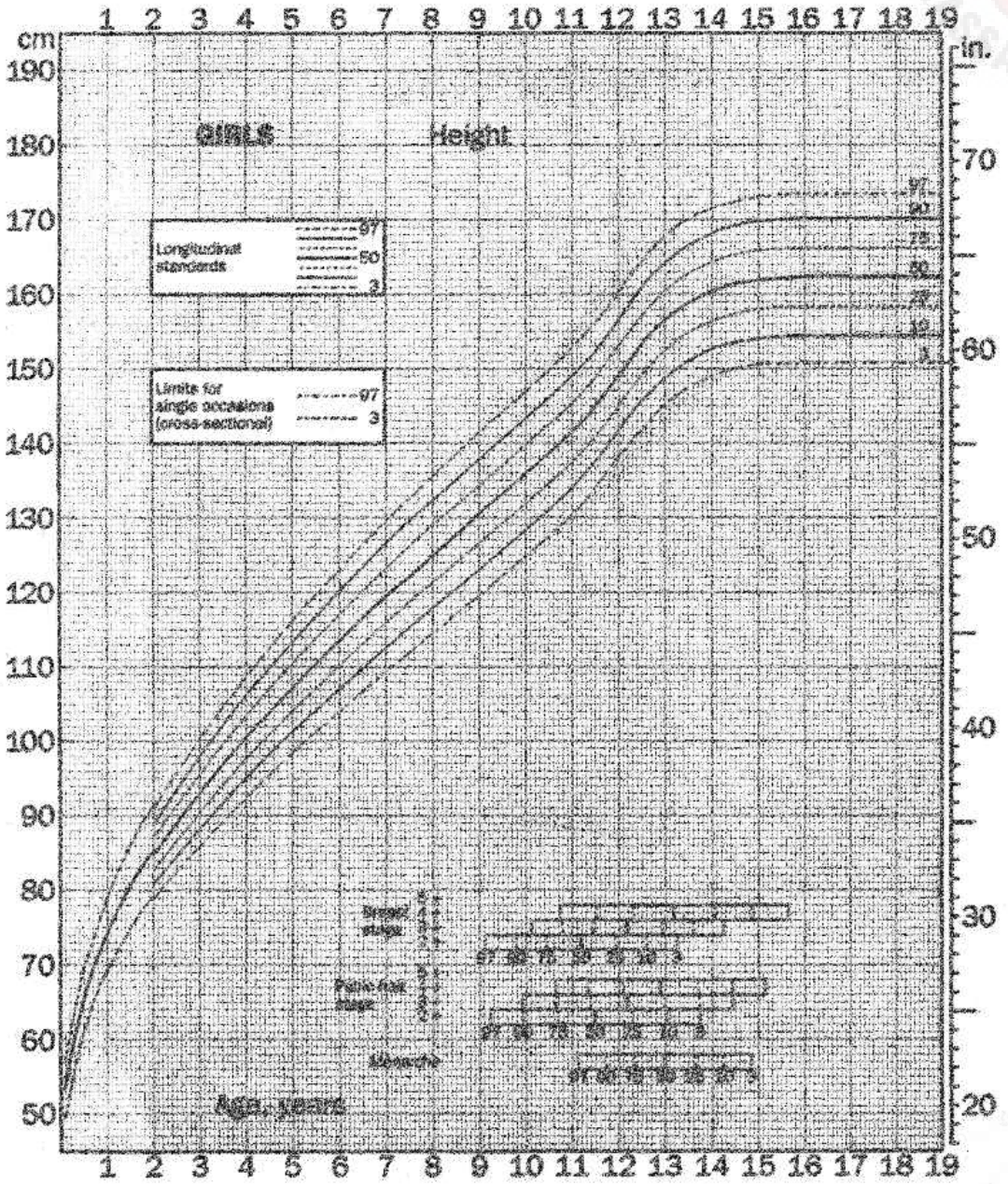
HORMONE RECORD

Date	Height	Weight	Calcium level (monthly)	Thyroid function (6 monthly from age 10)	Glucose tolerance test result (yearly from age 10/14)	Sex Hormones	Hormone treatment prescribed

Boy's growth chart



Girls growth chart



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OSTEOPOROSIS

OSTEOPOROSIS

Our bones support our weight and enable us to be mobile, do heavy exercise and lifting. Osteoporosis is the condition when the bones become excessively thin and fragile, leading to an increased risk of breakage or fracture. Osteoporosis is common in older women, especially affecting the bones of the lower spine, hips, and wrists. It is also common in thalassaemic adults. In thalassaemia the cause is not entirely clear, but several factors seem to play a role: diet low in calcium and vitamins, lack of exercise, hormone deficiency, insufficient transfusion, smoking and excessive alcohol. The best way of detecting osteoporosis is by measuring bone density with DEXA scanner (normally recommended annually from age 10), available in every major UK hospital. The result can be read as a 'Z score' which compares your result with the normal range for people of your age and gender. This scan should be done every year in adults.
Treatment of osteoporosis is aimed at improving bone density and preventing future fractures. The following are important

- Diet rich in calcium and Vitamin D. Your doctor may also recommend Calcium and vitamin D supplements
- Regular exercise
- Hormone replacement therapy (if recommended by hormone specialist)
- "Bisphosphonate" drug therapy may be required: e.g. Pamidronate or Alendronate

BONE DENSITY RECORD – DEXA SCAN YEARLY FROM AGE 10

Date	Site of Bone pain/fracture	DEXA	DEXA	Hormone replacement treatment (Drug and dose)	Bisphosphonate treatment (Drug and dose)	Comments
		Spine bone mineral density (Z score)	Hip bone mineral density (Z score)			

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VACCINATION

VACCINATION AND PROTECTION AGAINST INFECTION

Hepatitis B virus which can be spread by blood transfusion is now a very rare cause of infection. However, all thalassaemic patients should be vaccinated and have a blood test (anti-HBs) every year to confirm that they are protected. They should have booster vaccinations when necessary. Blood is routinely screened to prevent infection with HIV and hepatitis B and C, and the risk of getting these infections from blood in the UK is remote, but it is still recommended that your blood is tested for hepatitis B, C and HIV every year

Remember, if you have had your spleen removed, your body has some weakness in its defence against infection. You should always carry a card with you indicating that you do not have a functioning spleen. You should have been vaccinated against Pneumococcus, Haemophilus Influenza type B, and Meningitis type C.

The pneumococcal vaccine needs to be repeated every 5-10 years. It is recommended that you take Penicillin V twice a day (adult dose 500mg twice a day, children 125mg-250mg twice a day depending on age). You may also need extra protection (including extra Meningitis vaccination and anti-malaria tablets) if you travel to certain parts of the world. You should consult your doctor or a travel clinic.

VACCINATION AND VIRUS TESTING RECORD

Date	Hepatitis B immunity (anti-HBs)	Hepatitis B vaccine booster	Hepatitis C check	HIV check	Vaccine dates for those without spleens	Pneumococcal vaccine	Meningococcal vaccine	Other vaccinations and comments

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TREATMENT AWAY FROM HOME

Treatment away from home.

If you are temporarily moving house or going to a university away from home and this will involve having to receive treatment in the new area you are moving into, please contact the UKTS for advice. This advice will be valuable as the UKTS will be able to tell you which and where is the nearest hospital and Haematologist who are aware of how to treat thalassaemia.

The same applies if you are going abroad. On the next pages you will see a sample of treatment centres in the UK and local associations overseas. If you need advice about treatment available in a specific country, please contact the UKTS office for assistance.

ADDRESS DETAILS

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19 The Broadway, Southgate Circus, London N14 6PH
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Tel: 00357 22319129/319134 Fax: 00357 22314552
e-mail: thalassaemia@cytanet.com.cy website: www.thalassaemia.org.cy

British Liver Trust
Ransomes Europark, Ipswich IP3 9QH
Tel: 01473 276 326 Fax: 01473 276 327
e-mail: info@britishlivertrust.org.uk website: www.britishlivertrust.org.uk

Sickle Cell & Thalassaemia Association of Counsellors (STAC)
Woodenspoon House, 5 Dugard Way, London, Kennington, SE11 4TH
Tel: 020 7414 1339 Fax: 020 7414 1357
e-mail: jean.mullen@chsltr.sthames.nhs.uk website: www.stacuk.org

British Heart Foundation
14 Fitzhardinge Street, London W1H 6DH
Tel: 020 7935 0185 Fax: 020 7486 5820
Website: www.bhf.org.uk

Diabetes UK
10 Parkway, London NW1 7AA
Tel: 020 7424 1000 Fax: 020 7424 1001
e-mail: info@diabetes.org.uk website: www.diabetes.org.uk

National Osteoporosis Society
Camerton, Bath BA2 0PA
Tel: 01761 471 771 Fax: 01761 471 104
e-mail: info@nos.org.uk website: www.nos.org.uk

MAJOR THAL TREATMENT CENTRES AND SOCIETIES.

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University Hospital
Leeds LS9 7TF
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Manchester M13 9WL
TEL: 0161 276 4812

Dr Adrian Williams
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Bradford BD9 6RJ
TEL: 01274 364 516

SOCIETY: U.K. Thalassaemia Society,
19 The Broadway,
London N14 6PH.
TEL: 020-8882 0011

Centres Abroad

AUSTRALIA SOCIETY: Thalassaemia Assn of NSW, P.O.Box 80, Marrickville 2204, NSW. TEL: (02) 871 1701

CANADA SOCIETY: Ontario Thalassaemia Foundation, 75 Poplar Heights Etobicoke, Ontario M9A4Z3. TEL: 416-248 9275

CYPRUS SOCIETY: Cyprus Thalassaemia Assn, P.O.Box 8503, Nicosia. TEL: 22429141

GREECE SOCIETY: Panhellenic Thalassaemia Federation, Tzavella 1, 10681 Athens. TEL: 01-364 4682

ITALY SOCIETY: Lega Italiana Per La Lotta Contro Le Emopatiee, Tumori Dell'Infanzia, Via Giobert 27, 14100 Asti. TEL: 014-155 7034

INDIA SOCIETY: Federation of Indian Thalassaemics, P.O. Box 6658, New Dehli-110018. TEL: 550 1483

MALAYSIA SOCIETY: Pertubahan Thalassaemia Pilau Pinang, 56 Jalan Goh Guan Ho, 11400 Penang. TEL: 828 6412

PORTUGAL SOCIETY: Associato Portuguesade Paise Doentescom Hemoglobinopatias A.P.P.D.H. Inst. Nac de Saude Ricardo Jorge Av. Padre Cruz, 1699 Lisboa Codex. TEL: 011-757 7070

PAKISTAN SOCIETY: Pakistan Thalassaemia Welfare Society, 22 Atta ul Hag, Westridge – 1, Rawalpindi. TEL: 546 2084

SINGAPOREAN SOCIETY: TSS, 7th Floor, Children's Tower, KK Women's and Children's Hospital, 100 Bukit Timah Road, Singapore 229899. TEL: 63941862

TURKEY SOCIETY: TADAD, P.K. 403, Kadikoy, 81310 Istanbul. TEL: 01-358 0071

U S A SOCIETY: Cooley's Anemia Foundation,129-09 26th Avenue, Flushing, New York 11354 TEL: 2312-221 7525