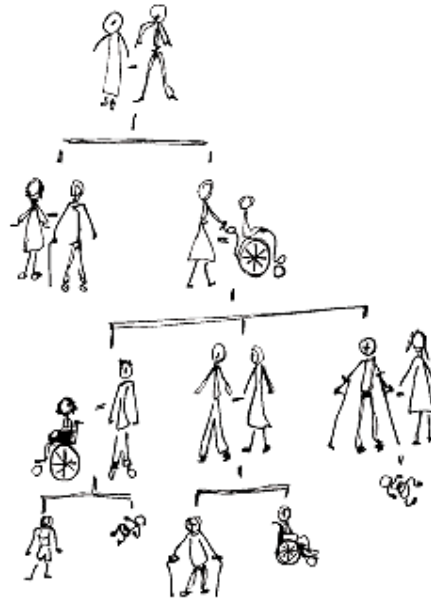


CROSS-CURRICULAR MATERIALS FOR KEY STAGE 4
(SCOTTISH CERTIFICATE OF EDUCATION - STANDARD GRADE)



Genes and You

Teaching about genetics from a human perspective

by Gill Mullinar

GENETIC INTEREST GROUP

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Published by the Wellcome Trust

The Genetic Interest Group is a registered charity, no.803424

The Wellcome Trust is a registered charity, no. 2107S3

GENETIC CHROMOSOMAL CONDITION CARDS

GENETIC CHROMOSOMAL CONDITION CARDS ON THE FOLLOWING:



THALASSAEMIA

Thalassaemia

WHAT IS THALASSAEMIA?

Thalassaemia (pronounced thal-er see-meeya) is an inherited blood disorder in which there is a defect in the structure of haemoglobin. Haemoglobin is a protein which is contained in the red cells of the blood. It picks up oxygen from the air in the lungs and carries it to the tissues of the body where it is needed. A person who does not have enough haemoglobin is anaemic (pronounced er-nee-mick). There are different types of Thalassaemia. This card is about beta (pronounced beet-er) Thalassaemia.

IS THALASSAEMIA PASSED ON?

The pattern of inheritance for Thalassaemia is autosomal recessive.

A person who inherits one faulty gene for Thalassaemia will be a carrier. Carriers are unaffected but can pass the faulty gene onto any children they may have. If one or both parents is a carrier, there is a 50% (1 in 2) chance that each child of theirs will also be a carrier. Carriers of Thalassaemia are sometimes said to have Beta-Thalassaemia trait (formerly referred to as thalassaemia minor). A child who inherits two copies of the faulty gene (one from each parent) will have Thalassaemia. If both parents are carriers, there is a 25% (1 in 4) chance of this happening.

THALASSAEMIA

A PERSON WITH BETA-THALASSAEMIA MAJOR MAY BE AFFECTED IN SOME, OR ALL, OF THE FOLLOWING WAYS:

- Children with Beta-Thalassaemia major appear healthy at birth but become anaemic between the ages of 3 and 18 months. They become pale, irritable and weak, do not sleep well, lose their appetite and may vomit feeds.
- Once Beta-Thalassaemia major has been diagnosed, the child has to have regular blood transfusions every four to six weeks for the rest of his/her life. Most children who have these transfusions grow normally and live happily into their teens, but to live longer, they also need other treatment (see below).
- Regular blood transfusions help by getting more red blood cells into the body, but these red cells contain iron and the body cannot easily get rid of excess iron. So treatment by blood transfusion must be followed by treatment with an iron-reducing (or chelating) drug such as Desferal to avoid damage to the heart, liver and other major organs. Desferal cannot be absorbed if taken orally and must be given by a slow infusion under the skin over 10 – 12 hours per day, at least 5 times a day.
- Thalassaemics are frequently affected by complications such as diabetes, osteoporosis and endocrine problems.

OTHER INFORMATION

Beta-Thalassaemia affects mainly people of Mediterranean, Middle Eastern or Asian origin. It is thought that Thalassaemia is more common in these parts of the world because people who carry the Thalassaemia gene are protected from the more severe forms of malaria. Thalassaemia is rare in North Europeans.

There are about 200,000 people in Britain who are carriers of Beta-Thalassaemia. They are healthy themselves but if their partner also carries the Thalassaemia gene, there is a 1 in 4 chance that each and every child of theirs will have Beta-Thalassaemia Major. At the moment, there are about 830 young people with Thalassaemia in Britain, but at least 100,000 children are born around the world with the disorder each year.

The outlook for people with severe Thalassaemia is improving as treatment improves. A bone marrow transplant is possible for some children with

Thalassaemia but is not without risk. It is best done when the child is still young and the donor must be a close family member (e.g. brother, sister, mother or father) who is an exact tissue match.

If you are interested in finding out more about Thalassaemia, you can contact the UKTS at UK Thalassaemia Society, 19 The Broadway, Southgate Circus, London, N14 6PH. Tel: 020 8882 0011 Fax: 020 8882 8618 email: office@ukts.org Website: www.ukts.org

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