If you would like further information on essential thrombocythaemia please contact:
Faye Armstrong, Haematology Specialist Nurse
Tel 0191 5656256 ext 47443

A Haematology Support Group is held on a monthly basis.
For further details please contact:
Faye Armstrong Haematology Specialist Nurse
Tel 0191 5656256 ex 47443

Useful websites
www.nhs.uk
www.sunderland.nhs.uk/chs
www.cancerbacup.org.uk

This information was correct at the time of printing. While the Trust makes every reasonable effort to keep its information leaflets up to date, very recent changes may not yet be reflected in the guidance and you should discuss this with the clinical staff at the time of your appointment.

Date of publication: December 2006
Review date: December 2009
CGSG approval March 2007
Ref: 050/07

City Hospitals Sunderland NHS
NHS Foundation Trust

ESSENTIAL THROMBOCYTHAEMIA (ET)

PATIENT INFORMATION LEAFLET

Haematology Department

City Hospitals Sunderland

The following information is about essential thrombocythaemia (ET), also known as primary thrombocythaemia. This is one of a group of conditions, known as the chronic myeloproliferative diseases (CMPDs).

The chronic myeloproliferative diseases

The myeloproliferative (pronounced mi-low-pro-liff-er-a-tiv) diseases are a group of conditions affecting the function of the bone marrow. They involve an over-production of one or more of the blood cells that are produced in the bone marrow. A small number of people with a CMPD will go on to develop a form of leukaemia, though this is quite rare.

The main types of chronic myeloproliferative disease are:

- essential thrombocythaemia (throm bo si theme ia).
- polycythaemia vera (polly si theme ia).
- chronic idiopathic myelofibrosis (i dio path ic my low fi bro sis).

The bone marrow

The bone marrow is the spongy material in the centre of some of our bones. It produces cells known as stem cells. Stem cells are immature cells that develop into three different types of blood cells:

- **Red blood cells:** carry oxygen to all cells in the body.
- **White blood cells:** fight infection and form part of our immune system.
- **Platelets:** help the blood to clot, to prevent bleeding.

Essential thrombocythaemia

Essential thrombocythaemia (ET) is a chronic myeloproliferative disease affecting the cells in the bone marrow that produce platelets. ET is a rare condition that can affect people at any age, including children, although it is very rare in people under 50. The cause is unknown.

Follow-up

Your treatment is likely to be life-long, and you will need to have regular check-ups and blood tests. These will probably be ongoing. If you have any problems, or notice any new symptoms in between these times, let your nurse or doctor know as soon as possible.

Your feelings and support

The need for practical and emotional support will of course be individual. Some people with ET may find that their life is largely unaffected; for others the condition may be a source of great fear and distress. You may have many different emotions, including anger, resentment, guilt, anxiety and fear. These are all normal reactions, and are part of the process many people go through in trying to come to terms with their condition and its treatment.

You don't have to cope with these feelings on your own; people are available to help you.

Interferon-alpha

Is a protein that occurs naturally in the body. It can also be made as a drug. It is given to stimulate the body's defence system, and can help to slow down the production of platelets. Interferon is given as an injection under the skin, usually three times a week. Side effects include flu-like symptoms, dizziness, and extreme tiredness (fatigue).

Anagrelide

Anagralide specifically reduces the number of platelets. It is given as a capsule. Side effects can include severe headaches and palpitations (sensation of a fast heartbeat), ocassionally it can affect an individual's blood pressure.

Anagrelide does not seem to affect fertility or increase the risk of the ET developing into leukaemia. The exact role of anagrelide in the treatment of ET is still being established.

Clinical trials

Research into new ways of treating essential thrombocythaemia is ongoing. Doctors use clinical trials to assess new treatments. In the UK, there are two research trials looking at the best way of treating ET. One trial compares the sole use of aspirin with a combination of aspirin and hydroxyurea. A second trial, called the MRC PT1 trial, is comparing aspirin with aspirin and hydroxyurea, or aspirin and hydroxyurea with aspirin and anagrelide. You may be asked to take part in either of these trials.

Before any trial is allowed to take place, it must be approved by a research ethics committee, which protects the interests of those taking part. Before you enter a trial, your doctor or research nurse must discuss the treatment with you so that you have a full understanding of the trial and what it involves. You may decide not to take part, or to withdraw from the trial, at any stage. You will then receive the best standard treatment available.

Some people may have a faulty gene, called the JAK2 gene, that might have caused their condition.

Signs and symptoms

Having too many platelets in the blood may not cause any symptoms and some people are diagnosed with ET during a routine blood test, when they have no symptoms. A high number of platelets can cause the blood to clot more easily. It may also cause abnormal bleeding, because the platelets are immature and cannot function properly.

Symptoms can occur because a blood clot (thrombosis) has formed in a vein in the body. Clotting problems are more common in people over 60, or those who have already had a previous blood clot. The symptoms will depend upon where in the body a clot forms. Common sites for a clot to form are:

The deep veins of the leg This can cause symptoms such as pain, swelling, heat, and redness of a calf (known as deep vein thrombosis or DVT*).*

The brain This may cause mild symptoms such as headaches, sight disturbances, or dizziness. Blood clots in the brain may also cause symptoms that are more serious, such as a stroke or TIAs (transient ischaemic attacks or mini-strokes).

The heart A blood clot in the heart may cause chest pain and in severe cases, can result in a heart attack.

The lungs A blood clot in the lung may cause breathlessness and chest pain. This is known as a pulmonary embolism.

Problems caused by abnormal bleeding are less common than clotting problems. They can include:

- nosebleeds
- bruising

- abnormal vaginal bleeding
- bleeding gums

If you develop any of these symptoms in between your check ups, you should discuss them with your doctor or haematologist.

Diagnosis

ET is usually diagnosed by a haematologist (a specialist in blood disorders). Diagnosis is often made following a simple blood test called a full blood count. This counts the number of red cells, white cells, and platelets. In ET there will be a high platelet count.

Other conditions can cause the platelet count to be increased, such as infection, arthritis, chronic bleeding, or a lack of iron in the blood. These need to be ruled out before a diagnosis of essential thrombocythaemia can be made.

Bone marrow aspirate

This procedure involves taking a sample of bone marrow from the back of the hipbone (pelvis). The sample is then examined under a microscope to see if it contains any abnormal cells. The bone marrow sample is taken under a local anaesthetic. You are given an injection to numb the area, and a needle is passed gently through the skin and into the bone.

A tiny sample of the marrow is then drawn (aspirated) into a syringe. Sometimes a small core of marrow is also needed; this takes a few minutes longer. This procedure can be uncomfortable for a few seconds and you may need to take some mild pain killers, like paracetamol.

You can request a separate information sheet about bone marrow investigation:

Treatment

The aim of treatment is to control the number of platelets and so reduce the risk of abnormal clotting or bleeding and the associated complications.

If treatment is successful, the outlook for someone with ET can be very similar to that of someone of the same age who doesn't have the condition.

If you have been diagnosed with an ET which is not causing any problems, you may not need any treatment for a while. You will be seen by your doctor who will monitor your condition with regular blood tests.

Aspirin

Asprin is often used as it can affect the way the platelets 'stick' together, which can help to reduce the risk of clotting. However, aspirin will not reduce the platelet count. Side effects of aspirin include an increased risk of bleeding and ulcers in the stomach. You will be advised to use other simple painkillers, like paracetamol, should you need them.

Chemotherapy

Chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. It may be given to try and reduce the platelet count. The main drug used is Hydroxycarbamide (hi drox ecar ba mide, also called hydroxyurea*),* which is given as a tablet. It can cause **potential** side effects, although generally these will be mild. It can also affect **fertility,** and people taking hydroxyurea are advised not to get pregnant or father a child, as there may be a risk to the developing foetus.

It is therefore advisable to use adequate contraception while taking the drug and for a few months afterwards.

If used over a long period of time, hydroxyurea may increase the chance of the ET developing into a leukaemia**.** The chemotherapy drug busulphan (bu sul fan*)* may also be used. Busulphan is usually given as a tablet but can be given by injection into a vein. It can cause similar side efects to hydroxyurea.