Immune Thrombocytopenia (ITP)
**ITP - What is it?**

ITP is a blood disorder affecting platelets in the blood. Platelets are small cells in your blood that help your blood to clot. In ITP the body’s **immune** system produces antibodies which destroy platelets.

**Thrombocytopenia** refers to a reduced number of platelets. The normal range of the platelet count is usually expressed as between **150-400**. People with ITP have a reduced platelet count but the way they function is normal.

**Purpura** refers to the purple bruising under the skin and mucous membranes, such as the lining of the mouth.

**Who does it affect?**

ITP can affect both children and adults. It can be both acute and chronic. It may affect both men and women.

**Adults with ITP**

**How is it diagnosed?**

- A medical history will be taken looking at a history of bleeding.
- A drug history is also taken, as some drugs may affect your platelet count.
- A physical examination is carried out.
- A blood count is taken and a blood film (slide) is examined under the microscope.
- A test for the presence of Helicobacter Pylori (an infection that may be found in the gut) may be taken and if positive, medication may be given to help resolve the problem.
- A bone marrow test may be performed.

Symptoms may vary from people with mild bruising and bleeding to those who have more serious bleeding episodes.

Head injuries or ongoing headaches should always be treated seriously and medical help sought urgently.

**What treatment is available?**

- In general, people with platelet counts exceeding 30 require no treatment unless they need any interventions that may be associated with blood loss, such as surgery, dental extraction or childbirth.

- **Prednisolone** (oral steroid) is usually the initial treatment. Two thirds of patients will respond to prednisolone at a dose of 1mg/kg/day for 2-4 weeks reducing the dose gradually over several weeks. Side effects may include: weight gain, gastric irritation, disturbed sleep patterns and possibly increased risk of infection.
You should always let your doctor know if you are unwell and taking steroids. Long-term complications may include thinning of the bones and the development of diabetes.

- **Intravenous immunoglobulin** (IVIG) may be used and is effective in three quarters of cases. Response may be short-lived and the platelet count may start drifting back down in 3-4 weeks.

  It is useful to know if people respond to IVIG as it can be used to cover surgical procedures. How it works is not fully understood. It is a blood product. Several steps have been taken by the manufacturer to reduce the risk of blood-borne viruses to extremely low levels.

- **Splenectomy** (removal of the spleen) is an option and 2/3rds of people with ITP who undergo it will achieve a normal platelet count with no additional treatment. Some people will receive a partial or passing response.

  The spleen plays an infection-fighting role and as a result there is a need for pre-operative vaccinations and post-operatively life-long antibiotics. Predicting the response to splenectomy is individual and your doctor will discuss this with you. Your surgeon will discuss the actual surgery with you.

- **Dexamethasone** (steroid) may be used as a four-day cycle at a dose of 40mg orally a day. The cycle may be repeated every 28 days for 6 cycles.

- **Methyprednisolone** (steroid) is also used. This is given as an intravenous infusion over 30-60 minutes as an outpatient.

- **Nplate** (Romiplostim) is a drug used to treat adult patients with chronic ITP, who are resistant to other therapies. It works by stimulating the bone marrow to produce more platelets. It is given by a weekly subcutaneous (under the skin) injection and requires regular monitoring of platelet counts and possible dose alteration. If Nplate is stopped your platelet count is likely to fall. This drug is not used as first line therapy.

- **Eltrombopag** (Revolade) belongs to a group of medicines called *thrombopoietin receptor agonists*. It is used to help increase the number of platelets in your blood. It is given orally and has some dietary restrictions.

- **Rituximab** is a drug that has proved useful for some patients. It is given as an infusion weekly for 4 weeks. A pre-medication is given to help reduce possible side-effects.

- **Immunosuppressive therapies** may be used in people who do not respond to other therapies. Mycophenolate mofetil, azathioprine or cyclophosphamide may be used.

**Signs and symptoms that need action:**

- Head injury or other severe injury.
- Nose/mouth bleeds that do not stop with usual first aid.
- Headaches that do not resolve.
Seek advice:

- If you notice blood in your urine or stool.
- If your periods are heavier than normal.
- If you notice that you are bruising for no reason.
- If you notice petechiae – a rash of red, pin-prick sized spots.

You need to have a full blood count taken and then you will be referred appropriately, depending on the result. Do not take aspirin or non steroidal anti-inflammatory drugs, such as “Nurofen” ibuprofen, unless discussed with your doctor first.

Children with ITP

Acute ITP is often seen in young children, sometimes following a viral infection. The onset is usually sudden. About 80% of children with ITP will recover spontaneously within 6-8 weeks without any intervention.

Chronic ITP refers to an episode which lasts longer than 6-8 weeks. Many children settle with an adequate platelet count i.e. more than 20 and have no symptoms unless injured. Children who are under 10 years of age at the time of diagnosis are likely to have a spontaneous remission.

Children over 10 years of age at diagnosis, and in particular adolescent females, are more likely to sustain a chronic course and are managed similarly to adults with the condition.

How is it diagnosed?

- A full blood count may be taken and examined. Other blood tests may be taken to exclude any other diagnosis.
- Occasionally a bone marrow biopsy may be required to assist with the diagnosis.
- A full drug history will be taken as some drugs may affect platelet production.

When is treatment required?

More than 80% of children with ITP do not have significant bleeding symptoms and they do not require specific interventions.

What treatment is available if needed?

Corticosteroids are generally first-line treatment. They may be given orally as prednisolone or intravenously as methylprednisolone. They are given at high doses but for short periods. Side effects may include:

- Increased appetite and weight gain
- Disturbed sleep pattern, mood changes.
Stomach irritation.

Increased susceptibility to infection. Always let your doctor know that you are taking steroids.

With long-term use, other side effects may develop such as diabetes and thinning of the bones. The use of long-term steroids is generally avoided.

Intravenous immunoglobulin (IVIG) may be used. This is given as an infusion in hospital over a few hours. It is a blood product and reactions may occur. During manufacture precautions are taken against blood-borne infections but they cannot be completely eliminated. Patients may receive a pre-medication in order to help prevent this. If patients respond to treatment IVIG may be used to cover surgical/dental procedures.

Splenectomy may be considered in some children.

Long-term responses may vary and the problem may return. Do not take aspirin or non steroidal anti-inflammatory drugs, such as ibuprofen (Nurofen), unless you have discussed it with your doctor.

ITP in pregnancy

Mild to moderately low platelets are not uncommon in pregnancy. Platelet count tends to fall in the later stages of pregnancy to 120-150 (normal range 150-400). The reason for this “gestational thrombocytopenia” is unknown. It seldom causes problems for either the baby or the mother and the count usually returns to normal once the baby is born.

The mother with ITP

How is it diagnosed?

A full blood count is taken and a blood film (slide) examined under the microscope. Various other blood tests are taken, to exclude other causes of low platelets.

Diagnosis is largely one of exclusion.

A bone marrow biopsy is not usually performed.

What treatment is available?

Treatment is based on the risk of bleeding. The platelet count may fall as the pregnancy progresses. Therefore careful planning is needed to ensure a safe count at the time of delivery.

If you have no bleeding symptoms and the platelet count is 20 or greater then no treatment is needed until delivery is imminent, but careful blood monitoring is required.

Platelet counts of 50 or more are regarded as safe for normal vaginal delivery.

Platelet counts of 50 are also safe for a Caesarean delivery, but epidural anaesthesia would be avoided. The platelet count should be 80 for this.
If the duration of treatment is likely to be short then prednisolone, a steroid may be used at a dose of 1mg/kg/day based on pregnancy weight with the dose gradually being reduced.

- Side effects may include: increased appetite and weight gain, mood changes, disturbed sleep pattern, possible increase in infection risk, blood pressure and blood sugars will be monitored. Diabetes and thinning of the bones may be a problem with long-term use.
- Steroids are broken down by the placenta and will not usually have any significant effect on the baby.

Intravenous immunoglobulin (IVIG) may also be used, doses may vary. It is a blood product and is usually given with pre-medication under supervision and over a few hours. The manufacturer takes precautions against blood borne infections but they cannot be completely eliminated.

What happens if I do not respond to treatment?

- High dose methylprednisolone (intravenous steroid) may be used in combination with IVIG and/or immunosuppression therapy.

How will my baby be delivered?

- The obstetrician will advise on the safest method of delivery and pain relief looking at platelet count and any previous delivery history.
- Caesarean or vaginal delivery will be determined by obstetric indicators rather than the ITP history alone.

The baby

- ITP is not an inherited condition. However it is possible that the mother’s antibodies have crossed the placenta, entering the baby’s blood and the baby may have a lower platelet count for the first few weeks or so after birth. A cord blood platelet count should be taken. If the platelet count is low, daily platelet counts should be taken from the baby until a rise is seen, usually day 2-5 post birth.

- Breastfeeding – there is no reason why you cannot breast feed your baby.
- Non steroidal anti-inflammatory drugs, such as “Nurofen”, ibuprofen, should be avoided whilst the platelet count is below 100.
Treatment Record

Name ........................................... Date of Birth ......................................
Normal platelet range: 150 - 400

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Clinic Discharge Advice

When you are discharged from the haematology clinic, the alert signs that you need to be aware of are:

- Increased or unusual signs of bleeding or bruising.
- A rash of pin prick sized spots (petechiae).
- If your menstrual periods are heavier or longer than usual.
- Increased nose or gum bleeds.
- Unresolved headaches.

In the first instance, a full blood count should be taken and depending upon the result your General Practitioner may refer you back to the Haematology Department.

Where to get information:

- **Haemostasis/Thrombosis Clinical Nurse Specialist**
  - Voicemail: (01803) 655270
  - E-mail: fiona.dooley@nhs.net

- **ITP Support Association:** send an A5 envelope with 2 first class stamps to:
  
The ITP Support Association  
  Synehurste  
  Kimbolton Road  
  Bolnhurst  
  Bedfordshire  
  MK44 2 EW

  [www.itpsupport.org.uk](http://www.itpsupport.org.uk)