Know About ITP

Contents

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What is ITP?

Immune Thrombocytopenia is a condition which affects the platelets in the blood. Platelets contribute to the blood clotting process – for example, when someone has an injury or accident which causes bleeding. In ITP the platelets are attacked and destroyed by the immune system. ITP was previously called Idiopathic Thrombocytopenic Purpura*; sometimes the names may be used interchangeably.

In most people there are between 150,000 and 400,000 platelets in every cubic millimetre of blood (i.e. 150-400 x 10^9l.). However in the UK we simplify this by describing a platelet count of, say, 150 rather than 150,000. In an ITP patient the platelet count may be significantly lower, even as low as 1 or 2. In typical ITP the rest of the blood count is normal.

Features associated with ITP are large multicoloured bruises, many small reddish-brown bruises, nosebleeds, mouth bleeds and petechiæ* – a rash of red, pin-prick sized spots caused by leaking capillaries (tiny blood vessels near the surface of the skin). Women may experience heavy or more troublesome periods. Bruising may appear spontaneously or may be larger than would be expected after a simple knock or injury. Although symptoms are individual to each patient, in general the lower the count the more apparent the symptoms become.

According to current understanding ITP is not considered to be a genetic condition and it can arise in any person, adult or child, at any stage in their life.

Known causes of ITP

In many instances the cause is unknown, but for some it can be preceded by a viral infection (e.g. a cold, 'flu, glandular fever, chicken pox, etc.). Less frequently ITP can be secondary to an existing autoimmune condition, most commonly systemic lupus erythematosus (SLE*#). In children the MMR# (measles/mumps/rubella) vaccine can precipitate the onset of ITP in children, although this is very rare (about 1 case in 24,000 immunisations). When ITP does occur it is usually of very short duration.

Certain drugs are known to cause a low platelet count#; in particular, heparin, teicoplanin, quinidine and quinine. Quinine is an ingredient of many malaria tablets and some over-the-counter products, such as Anadin, or cramp remedies. Tonic waters and bitter lemon drinks may also contain quinine, which although in low concentration is sufficient to cause severe thrombocytopenia in some sensitive individuals. Thrombocytopenia so caused is iatrogenic* and known as 'drug dependent immune thrombocytopenia' (DDIP#). The platelet count will

*See Glossary #Booklet or factsheet available – see page 18

return to normal when the product or drug is no longer taken. (For more information see the Association's booklet *Drugs that cause or aggravate thrombocytopenia*#.

Some studies have shown that bacterial infections may trigger ITP: Helicobacter pylori# (a stomach bacterium known to cause peptic ulcers) and also 'strep throat' (bacterial infection in the throat). Food intolerance# has also been associated with thrombocytopenia on rare occasions and novel research in this area has being funded by The ITP Support Association

ITP in adults

In the UK the incidence of ITP is approximately 6 per 100,000 adults (120 new cases each year) and may often arise insidiously, without the apparent trigger of a virus or other illness.

ITP in adults is likely to be a long-term condition, though not necessarily severe, and is observed more often in younger women (possibly because low platelets are being noticed during routine pregnancy blood tests) and older men, which can sometimes be due to side effects of prescribed drugs taken for other conditions.

ITP in children

Childhood ITP can be broadly divided into two types: the 'acute' type which often arises suddenly and may last only a few days or weeks, and the less common 'chronic' form which persists for longer than 6 months. In medical use 'chronic' refers to the duration of a condition rather than its severity.

It is estimated that in the UK about 400 children are diagnosed with ITP every year (i.e. 1 in approximately 25,000 children). Of these 75% will recover completely within 3 months, and 90% will return to a normal platelet count within 12 months. The remainder (10%) will be classed as chronic sufferers, but even in these the ITP may remit or become less severe as they grow up.

ITP in babies

ITP can occur in very young babies, even as early as 3-4 months of age. When it occurs, it does tend to be severe, with very low platelet counts and sometimes bleeding in sites other than the petechiae and bruises in the skin (e.g., bleeding from the mouth and rectum). It is treated just like in older patients—i.e., IVIg and/or steroids when the bleeding is significant.

The important thing is to be as certain as possible to exclude other diagnoses, given that there is no "test" for ITP. If the baby has bleeding signs and low platelets from birth or just days thereafter or if there is an enlarged spleen or other abnormalities in the blood count, then other diagnoses would need to be considered (infection, low platelets in the mother, hereditary thrombocytopenia, leukaemia, etc).

The good news is that ITP in babies, when it occurs, is usually transient, resolves within weeks to several months just as in older children.

Diagnosis

On presentation the doctor will arrange a blood test (FBC*). Diagnosis of ITP is based on the exclusion of other causes of thrombocytopenia and is suggested by bleeding or bruising in association with a low platelet count. Mild ITP may also be detected from a blood test before surgery or following an investigation into another illness or condition.

Tests may show increased levels of antibodies* directed against the platelets and if a bone marrow aspiration* is given an increase may be observed in the cells producing platelets. In straightforward cases, particularly in children, a bone marrow test is usually considered unnecessary.

ITP and associated diseases

Simple ITP does not result from a weakened immune system. ITP does not leave the patient more exposed to infections or illness (although some treatments do), neither is it associated with an increased incidence or risk of cancer or leukaemia. It is possible for someone to have more than one autoimmune disease concurrently. Occasionally, some forms of leukaemia, such as chronic lymphoid leukaemia (which is seen mainly in elderly people) may be associated with thrombocytopenia, but the diagnosis is usually obvious and the thrombocytopenia does not predate the leukaemia. ITP can be secondary to CVID (common variable immune disease) in which repeated infections, particularly chest infections and pneumonia, are a feature.

Familial thrombocytopenias

These are extremely rare conditions and are different from ITP because they are typically the result of an inherited defect of platelet production. There is not the increase of platelet destruction that is seen in ITP. In most familial thrombocytopenias there are rarely problems of bleeding or associated risks.

Management and Treatment

At present no treatment is guaranteed to cure# ITP, however it can raise the platelet count and ease symptoms when problems with bleeding arise. Only a small proportion of patients will require treatment. The aim is to maintain the platelet count at a level which enables the patient

to lead a normal life without troublesome symptoms, but it may not be possible to achieve this without side effects from the treatment.

Management should be based more on bleeding problems, if any, than on the level of the platelet count. For many, particularly children, this will be a 'watch and wait' policy in which no treatment is given but regular check-ups are arranged to monitor the child's condition.

On occasions it may be necessary to raise the platelet count to cover surgical procedures#, e.g. for some dentistry and during childbirth.

Treatment options

Treatment is traditionally directed toward controlling the destruction of platelets, which the following listings indicate. All work by dampening the immune system although some are specifically classed as immunosuppressive drugs. However a class of new drug, see page 8, is coming into use which stimulates the production of platelets.

Corticosteroid#: usually prednisolone. This is typically given orally in a daily dose of 1-2 mg. per kilogram of patient body weight, then reduced to a level which will maintain an absence of significant bleeding. Alternatively, if a swift response is necessary, methylprednisolone can be administered intravenously but its effect is relatively short-lived.

Short-term oral steroid use is associated with significant weight gain and a moon-shaped face, increased appetite, insomnia, mood swings, anxiety, irritability and, infrequently, bleeding from the intestines. All of these will be reversed on discontinuing the treatment, but if side effects become severe, medication may be prescribed to be taken on alternate mornings. Enteric coated tablets can be given if stomach irritation arises.

There is also an increased danger from some viral infections whilst on steroids, particularly chickenpox. This can be severe and life threatening in rare cases and patients on steroids should always seek medical attention immediately.

Long-term steroids should be avoided, or used with caution since they can reduce bone density and cause diabetes, cataracts, peptic ulcer (rarely) and in children are known to inhibit growth.

Intravenous Immunoglobulin#* Known as IVIg, immunoglobulin blocks platelet destruction and is typically used when a rapid increase in the platelet count is needed. Manufactured from up to 10,000 pooled donations of human blood plasma it is administered in hospital by intravenous drip over a period of two consecutive days. Additional treatments are given intermittently as required. Reactions can occur in some patients (shivering, nausea, joint pain and headaches) which can be reduced with the use of antihistamine and cortisone injections given immediately before IVIg is administered. Immunoglobulin is available under various brand names.

Other treatments

Anti-D immunoglobulin#. This is an alternative immunoglobulin, prepared from 1000 screened donors, injected rather than infused, which is simpler and more convenient to give. It cannot be used in patients whose blood group is rhesus negative, nor is it recommended for asplenic patients (i.e. those who have had their spleen removed).

As with all blood products immunoglobulin preparations used in the UK are subject to rigorous screening and anti-viral treatments.

Dexamethasone. An oral steroid used as an alternative to prednisolone. This is given in up to six 'pulses' (three days treatment once a month) of very high dosage. Initial studies have been variable, steroidal side effects are common, but some patients have shown a good response.

Splenectomy#. The spleen is responsible for removing platelets damaged by the body's immune system and removal of the spleen is an option for the patient with chronic severe ITP. The success rate is thought to be 65–75%. There is an increased risk of severe infection arising after splenectomy, and in the UK patients will be advised to take a daily antibiotic (usually penicillin) for the rest of their lives. This has to be carefully balanced against the problems of ITP itself. Splenectomy is rarely performed in children, since the risk of infection is higher than in adults.

A few specialist hospitals in the UK can perform an indium-labelled platelet spleen scan (see *Splenectomy – a guide for ITP patients*#) to assess whether splenectomy is likely to be successful. This test is available on the NHS, but patients must be referred by their doctor or consultant.

Before splenectomy patients will need to be vaccinated against hepatitis B, pneumococcal infections, meningitis and hæmophilus influenzæ, repeated as necessary.

Asplenic patients are strongly advised to carry with them the ITP Emergency card, or similar, in case of accident or emergency. An application form is available on request from The ITP Support Association.

Platelet transfusions. These are only used in life threatening emergencies or during surgery (usually in conjunction with other treatment) owing to the rapid destruction of transfused platelets in a patient with ITP. It can cause fever and shivering, also the rare possibility of transfusion transmitted infection.

Immunosuppressive drugs. These are slower acting treatments used in ITP for suppressing the production of antibodies. The three most commonly prescribed are azathioprine, cyclophosphamide and cyclosporin A. They have significant side effects, particularly if taken for a year or more and treatment will have to be carefully monitored. They should not be used during or before pregnancy.

These drugs increase the risk of infection and are linked to infertility. They should be used for children only in exceptional circumstances and always with extreme caution.

Their side effects are as follows:- Azathioprine is given orally and is known to cause nausea, liver toxicity, hair loss and joint pain. Patients will require regular liver function tests. Cyclosporin A is administered orally and may cause acne, headache, nausea, leg cramps, hair growth and shakiness. Cyclophosphamide is given intravenously and if used long term is associated with increased risk of malignancies in later life, hæmorrhagic cystitis caused by damage to the bladder lining and mouth ulcers.

Vinca-alkaloids. These drugs suppress the immune system and include Vincristine and Vinblastine. Given as an injection (usually 3 or 4 treatments) they increase the risk of infection and may produce hair loss, muscle pain, numbness in the hands and feet, pins and needles.

Danazol can be used as an alternative to high dose steroids and sometimes post splenectomy when long-term treatment is required. Taken orally, its side effects may include liver abnormalities and it can also encourage the growth of facial hair in women.

Rituximab# can be an effective therapy for chronic ITP with some patients enjoying lasting remission. Known as a monoclonal antibody rituximab is used in ITP on an 'off-licence'* basis only. Treatment consists of 4 intravenous injections one week apart in each case. Common side effects include fever, chills and rash, with a possible increased risk of infection. In rare cases patients have developed allergic arthritis, kidney failure, severe allergic reaction and (very rarely) PML, a viral infection of the brain.

Dapsone is taken orally and can be used in cases of less severe ITP. It is less effective in asplenic patients. Side effects are anæmia and skin rash.

Thrombopoietin receptor stimulators

In recent years a new class of drug known as TPOs#, have emerged which work by encouraging the production of platelets in the bone marrow and do not suppress or disable the immune system as with the treatments listed above.

Two drugs have been approved by NICE for use in chronic ITP for adult patients who meet certain criteria. These drugs are expensive and not all hospital authorities are willing to fund them but patients will have to discuss this with their consultant for further information. These drugs are not currently licensed for pregnant womenTPOs were intended as a continuous therapy to maintain the platelet count but evidence is emerging that some patients achieve a sustained remission on tapering off these drugs.

Romiplostin (known as Nplate) is intended for use in simple ITP only (where no other disease is associated with ITP) and is given subcutaneously (under the skin) in weekly doses. Other than headache at the recommended dosage, and marrow fibrosis at higher than recommended dosage, no significant side effects have yet been identified. As with any new drug some may

emerge once use in ITP becomes more established. It is currently licensed for adults and (at the time of writing) may soon be licensed for children.

Eltrombopag (known as Revolade in the UK and Promacta in the US) is taken orally, and foods with calcium must be avoided for four hours before and after the once daily dosage. No significant side effects have been recorded, but a theoretical risk* of cataracts and bone marrow fibrosis has been recognised. It is licensd for adults and children.

Further options

Some patients who require treatment do not respond to standard therapies, listed on the previous pages. In such cases other options are available, but these are limited and results are less well documented.

Mycophenolate mofetil is a powerful immunosuppressant, usually reserved for severe refractory* ITP. It is taken orally and may cause nausea and diarrhoea. In common with all immunosuppressants it increases the risk of infection.

Plasmapheresis. This removes anti-platelet antibodies by intravenous blood filtration. There are no observed side-effects but hospitalisation is required for treatment and the possibility exists for local infection or even thrombosis from the surgical line. (Its use is not recommended in published ITP Guidelines.)

Interferon is very rarely used in ITP. It is administered subcutaneously and may produce 'flu-like symptoms, fever, muscle aches, weakness, depression, anxiety and may also reduce the platelet count. Its use is not recommended in ITP Guidelines.

Tranexamic acid can be administered topically for dental extractions and severe nosebleeds, but is not otherwise recommended for ITP because of its tendency to promote thrombotic conditions. It is not a treatment for ITP. Its role is simply to increase clotting and decrease the risk for bleeding.

Alternative therapies

Some ITP patients may wish to seek help from alternative, or complementary, therapies but none has yet shown verifiable clinical improvement in ITP. It should be remembered that the majority of ITP children remit spontaneously within the first year, some adults also, and an improved platelet count or ITP remission has yet to be substantiated by such therapies.

When taken from a constitutional point of view, some complementary or alternative therapies may be said to improve a patient's sense of well-being. However, certain exclusions for ITP conditions should be carefully observed: for example, patients on cyclosporin should not take

St. John's Wort (hypericum), Ginkgo biloba and ginseng (herbal supplements) can act as anti-coagulants and may worsen bleeding in ITP patients.

Some over-the-counter products, such as Echinacea, are advertised to, 'boost the immune system'. However these are contra-indicated* for patients with autoimmune disease. Most patients with ITP have a robust immune system that functions well, but has become confused in destroying platelets. To take such products may worsen bleeding problems and lower the platelet count.

Vitamin C. Some studies in a small group of patients have shown that vitamin C may improve the platelet count slightly or lessen the symptoms of ITP. Although vitamin C is generally considered harmless in the RDA (recommended daily amount), higher doses can increase the risk of kidney stones and patients are strongly advised to discuss such doses with their doctor.

Dentistry

Patients should always inform their dentist if they suffer with ITP, or are asplenic, also stating their latest platelet count, if possible. Further information for dentists may be obtained in The ITP Support Association leaflet, *Protocol for Dentists*#. Where treatment involves the possibility of bleeding the dentist is advised to discuss the procedure with the patient's consultant beforehand.

For patients with a platelet count of 50 or below tranexamic acid can be used to assist normal clotting and healing after tooth extractions.

Asplenic patients should note that if a dental procedure carries a risk of infection (e.g. tooth extraction) a course of antibiotic cover should be given.

Over-the-counter drugs

Aspirin#, and products which contain aspirin, should not be taken by those with ITP. Aspirin has anti-coagulant properties which will affect the platelets and aggravate problems of bleeding. Non-steroidal anti-inflammatory drugs (NSAID), such as ibuprofen (Nurofen, etc.) should also be avoided for similar reasons. Paracetamol or codeine can be recommended for adults.

For young children, Calpol is considered a safe alternative to aspirin, but Calprofen is not advised since it contains ibuprofen. Before giving or taking any medicine always read the directions on the label and never exceed the recommended dose without specialist advice. If in doubt always ask your pharmacist.

Injections & vaccinations

Intramuscular injections (into the muscle) should be avoided, but most can be given subcutaneously (under the skin). Intravenous injections (into the vein) are permissible.

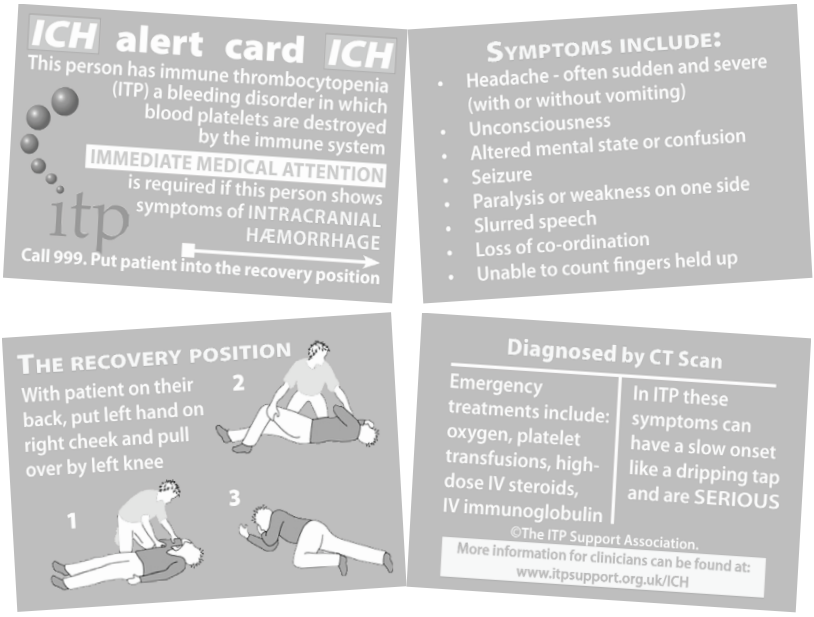
For individuals treated with systemic corticosteroids at high dosage (e.g. for children, prednisolone at 2mg per kilo of body weight daily for longer than one week and for adults, 60mg or more daily) live vaccines should be postponed until at least three months have elapsed following treatment. Live vaccinations such as polio, measles and BCG pose a risk of disseminated infection in the recipient if administered within this period, and non-live vaccines may be ineffective owing to suppression of the immune system by the steroid.

Likewise, live vaccines (with the exception of yellow fever vaccine) should not be given during the three month period following immunoglobulin as the immune response may be inhibited here also. Immunoglobulin derived from UK residents is unlikely to contain the yellow fever virus antibodies which would inactivate the vaccine.

ICH Alert Card

Having an ICH alert card is rather like taking out travel insurance. You'll probably never have to use it but it is there to give you peace of mind in case the worst happens.

It is very rare for anyone with ITP to have a brain haemorrhage (ICH), but if it should happen to you or your child, fast action by you or those around you plus prompt scans and specific treatment by emergency doctors can help prevent a serious situation becoming a disaster. The ITP Support Association developed the ICH alert card, available free on request, to alert patients about the early warning signs of ICH and alert paramedics or A&E staff that these signs in ITP patients must be taken seriously, demanding immediate medical attention.



General Outlook

Mortality in ITP children is very low. A survey into childhood ITP in the United Kingdom suggested that the incidence of intracranial hæmorrhage (bleeding into the head) was as low as 0.1% (1:1000) of the total number of children with ITP. Intracranial hæmorrhage seldom occurs spontaneously and there is usually an additional reason to the ITP for the bleeding to arise. It is not always fatal – most children make a complete recovery.

Mortality in adults is also recognised as low. Although ITP can be troublesome for some, for the majority it can be successfully managed, sometimes with treatment, sometimes without. Where ITP symptoms prove intractable hope can be drawn from the new approaches which are being introduced as a result of new research and development, not only into ITP but autoimmune diseases generally.

Emergencies

Fortunately, most ITP sufferers will not experience an emergency relating to their ITP, although some patients may be worried about knowing when – or when not – to call their doctor. Patients (or parents of ITP children) are advised to discuss this with their doctor. The NHS helpline may be useful for non urgent medical queries, but ITP can require prompt action on very rare occasions.

As a general guide, if bleeding cannot be stopped, or an unusually severe headache occurs (with nausea and/or loss of vision), fits, a serious blow to the head, or fever in asplenic patients, are good reasons to seek medical advice urgently. The medical staff should be informed immediately of the patient's ITP and no one should be afraid to make a fuss if the situation is not dealt with promptly.

Lifestyle and leisure

Many patients with ITP live a normal life, although a significant number report increased tiredness# and fatigue in relation to a reduced platelet count. There is no clinical explanation for this, as yet, but for those with no active bleeding there need be no unnecessary restrictions upon lifestyle apart from a general caution over some activities, for example, if the count is below 20 contact sports and occupations with a risk of injury should be avoided.

Holidays# may be taken as normal, but for those likely to suffer bleeding problems holiday destinations should be chosen where there is easy access to a hospital if necessary. Asplenic patients should take extra care in countries where malaria# is endemic.

Some doctors believe that there may be a theoretical risk of bleeding during flights* if the platelet count is below 20, but medical advisors to the ITP Support Association agree unanimously

that perceived risk is very low. It is a good policy for patients to discuss all holiday concerns with their consultant and it is also important to remember that holiday insurance will not cover claims relating to ITP unless specifically stated on the proposal form. For further information see *Holiday Insurance and Travel Guide* from the ITP Support Association.

It is important to put ITP into perspective. It can be a troublesome condition for patients with chronic severe ITP, but most sufferers manage to adapt ITP into their way of life once they understand the disease. Many discover that their lifestyle requires few changes. Although a small proportion of ITP sufferers may require a more aggressive approach to their treatment, the vast majority follow a full and meaningful life.

ITP and Pregnancy

Approximately 5% of all pregnant women experience a slight fall in their platelet counts (120–150) during the later stages of their pregnancy. This Is a normal adaptation of pregnancy. It is not a disease. It is not related to ITP. It causes no health problems for either the baby or the mother. The platelet count returns to normal after the birth.

However for a mother-to-be with ITP the platelet count may decrease below 100 (or much lower) during pregnancy. ITP, like other autoimmune disorders, can be more severe during pregnancy. Among most women with ITP, the risk of complications is no greater than those without ITP.

The mother with ITP

Although ITP varies from one individual to another, patients with a history of ITP may see their platelet count drop during their pregnancy, but it is unusual for it to fall so that it becomes dangerous for either mother or baby. Bleeding during pregnancy due to a lower platelet count is uncommon and serious bleeding (including vaginal bleeding) is most unlikely, especially if the platelet count is 50 or above.

During gestation the platelet count will be monitored and collaboration between hæmatologist, obstetrician and pædiatrician should determine whether treatment for the ITP is required. Mothers without bleeding symptoms and with a platelet count of 20 or above should not require treatment until the delivery, but this will depend on the proposed method of delivery and any other conditions associated with the ITP. If treatment is considered necessary the usual methods are steroids, intravenous immunoglobulin and platelet transfusion.

Steroids. Usually prednisolone or related treatments. These will slow down the destruction of the platelets.

Intravenous immunoglobulin. This is drip fed into the vein. Immunoglobulin helps to prevent the breakdown of platelets by the immune system and may be used several times during pregnancy.

Platelet transfusion. May be used in the rare instances where the platelet count is very low. In most cases the treatment is successful in raising the platelet count to a satisfactory level during pregnancy and labour.

Side effects of treatment

Steroids, although having an effect on the mother are broken down by the placenta and will not usually have any significant effect on the baby. If used for a prolonged period they may cause the mother increased weight, changes in blood pressure and changes to blood sugar levels. This will be monitored by the mother's doctor(s).

Immunoglobulin, in rare cases, can produce an adverse reaction. To prevent this it is usually administered slowly under close supervision. Temporary side effects, such as light headedness or fever are usually mild.

Precautions

There are no particular medical precautions to take during an ITP pregnancy, other than those recommended for normal conditions. Any undue strenuous or unusual activity should be discussed first with the patient's consultant who will advise as to its suitability.

As with any pregnancy, the taking of drugs should be avoided, other than those prescribed or recommended by a doctor.

The delivery

The obstetrician will advise on the best and safest method of delivery and pain relief, based on the platelet counts and taking into consideration any previous obstetric history. Cæsarian or vaginal delivery will be determined by obstetric considerations rather than the patient's ITP history.

Intravenous immunoglobulin may be given prior to labour. This is to ensure that the platelet count is as high as possible in preparation for the delivery or cæsarean section. For mothers on steroids, an increased dosage may be required to help with the stress of labour.

Epidural pain relief may be considered by the obstetrician who will balance its benefits against the bleeding risk, but is generally considered safe if the platelet count is 80 or above.

Ventouse (where a suction cap is placed on the baby's head to assist the birth) or forceps delivery will usually be avoided, especially if the baby also has a low platelet count. In other

respects the delivery should be normal.

Pain relief, post-delivery, under normal conditions may include non-steroidal anti-inflammatory (NSAIDs) drugs. These should avoided for a mother whose platelet count is 100 or below.

The baby

In some cases it is possible for the mother's platelet antibodies to cross the placenta, entering the baby's blood. The baby may have a lower platelet count during the first few weeks after birth, although this can take up to three months for the count to reach normal levels, when the baby's own immune system takes over.

A blood sample will be taken from the umbilical cord to ascertain the baby's platelet count and those with a subnormal count will be closely monitored. In the rare cases where treatment is required immunoglobulin can be given. Once a normal platelet count is reached it is very unlikely that the baby will experience further problems with ITP.

An asplenic mother with a normal platelet count may still have platelet antibodies in her body which may pass across the placenta to the baby. In this case the baby will require monitoring and treatment if necessary.

Breast feeding

There is no reason why mothers with ITP should not breast feed their baby. Steroids have not been found to hinder the production of breast milk and mothers on usual doses of steroids are unlikely to affect their baby as only small quantities will be found in breast milk. If special precautions are required for mothers on high doses of steroids the mother's doctor(s) will advise.

Future pregnancy

If problems with a low platelet count have been experienced during a pregnancy, special monitoring will be given in future pregnancies. Any treatment will be based on current or earlier experiences associated with ITP.

Menorrhagia (extra heavy periods)

Excessive blood loss can be very troublesome for some women and teenage girls with ITP#. Symptoms can be controlled in severe cases with tranexamic acid tablets (taken during menstruation only) or with the contraceptive pill.

The Mirena coil (a fitted intra-uterine device (IUD) which contains non-systemic progesterone) may be fitted after which the majority of women will find that periods will cease altogether.

Glossary

Acute: a disorder that arises suddenly and is of short duration.

Antibody: a protein produced by the immune system which binds to an antigen* and causes its destruction. (e.g. vaccination with the tetanus vaccine causes the body to produce antibodies directed against the tetanus bacteria, or germ. If the tetanus bacteria enters the body through a wound those antibodies are ready to bind to the bacteria and cause their destruction).

Antigen: any substance within the body which the immune system recognises as 'foreign'. These can be a virus, bacterium or a germ and the immune system responds by producing antibodies* to neutralise the antigen.

Asplenic: a person who has had their spleen removed.

Biopsy: a laboratory test conducted in hospital in which a small sample of tissue is removed from the body in order to make an accurate diagnosis.

Bone marrow aspiration: a fluid sample is removed from the bone marrow for analysis.

Capillaries: minute blood vessels located close to the surface of the skin.

Chronic: a disorder which persists for a long time, typically 6 months or longer. The term does not indicate the severity of any medical condition, which may range from mild to severe.

FBC (full blood count): a sample of blood is examined in the laboratory to measure the quantity and size of red cells, white cells and platelets in one millilitre of blood.

Iatrogenic: a condition caused by medication.

Idiopathic: a disease of unknown cause.

Immune system: the body's biological system of defence which protects it from infection and foreign organisms by the formation of antibodies* and specialised white blood cells.

Immunoglobulin: the protein from which antibodies* are made – found in the blood and in tissue fluids. Immunoglobulins are produced in the body by cells of the immune system known as B-lymphocytes. It is also manufactured by a number of drug companies from human donors under various brand names – for example, Alphaglobin.

Off-licence drug: a medicine licensed being prescribed for a medical condition different to the one for which it has been licensed.

Named patient basis: a medicine not licensed for any medical condition but which can be ordered by a doctor for a named patient.

Non-steroidal anti-inflammatory drug (NSAID): a drug such as ibuprofen (commonly sold as Nurofen or Brufen) which reduces inflammation.

Petechiæ: a rash of tiny red spots on the skin which do not disappear when a glass is pressed on them.

Prophylactic: a treatment or process used to prevent disease.

Plasma: the fluid remaining when the cells from the blood are extracted.

Platelet: a small blood cell (thrombocyte) which helps to maintain the integrity of blood vessels. Platelets are produced in the bone marrow by large cells called megakaryocytes. A bone marrow aspiration* is necessary sometimes to make a diagnosis in a patient with a low platelet count.

Purpura: a condition characterised by purple or reddish-brown spots on the skin or mucous membranes caused by leakage of blood from the capillaries.

Refractory: a condition that is resistant to treatment.

Simple ITP: where ITP is not affected by, or connected to, any other medical disorder.

SLE (Systemic Lupus Erythematosus): an autoimmune disorder which features inflammation of the connective tissues. It is diagnosed by a blood test or skin biopsy.

Steroids: compounds which resemble certain hormones found naturally in the body. They are frequently used to treat autoimmune disorders, such as ITP, in addition to other inflammatory conditions like rheumatoid arthritis. Oral contraceptives, usually derived from oestrogen and progesterone, are also part of the steroid family.

Theoretical risk: a recognised risk, unlikely in normal circumstances.

Thrombocyte: see **Platelet**.

Thrombocytopenia: an abnormally low number of platelets circulating in the blood.

Referral & Second Opinion on the NHS

Some patients with troublesome ITP may occasionally experience problems with the management or treatment of their ITP and wish to consult a specialist. Patients within the UK are entitled to request a second opinion or referral.

In Good Medical Practice published by the General Medical Council (the regulatory body for doctors) paragraph 2C states that good clinical care must include referring a patient to another practitioner, when this is in the patient's best interests, and paragraph 3e states that in providing care doctors must respect the patient's right to seek a second opinion. It is of course important that as far as possible doctor and patient agree and co-operate on this.

If patients/parents require assistance to find a specialist in their region the Association may be able to help, and the UK medical advisors to the ITP Support Association are willing for patients to be referred to them. This will not be necessary for the majority of patients whose ITP is successfully managed at the local level. However, certain difficult conditions may require the attention of an ITP specialist and the patient's GP or hospital consultant should make the referral. Although the patient is entitled to request a referral, the clinician they wish to see cannot accept a direct approach from the patient since a full medical history from previous doctors is essential.

Doctors are usually willing to refer a patient for a second opinion or specialist consultation if it is felt that some benefit will follow, but may resist if it is thought that the condition does not warrant this step. Under certain circumstances a patient should feel able to challenge a refusal if it seems unreasonable. However, attempts should be made to resolve the matter amicably in order to maintain a good relationship with the local hospital should any bleeding emergency arise.

When things go wrong

In any vast organisation, such as the NHS, it is hardly surprising that occasionally a patient/parent feels they have cause for complaint. In the rare instance that it cannot be resolved at the time, and a formal complaint might even be envisaged, contact can be made initially with one of the bodies set up to oversee patient interests in the NHS. A visit or letter to the PALS (Patient Advice and Liaison Service) should be the first port of call, but if the issue remains unresolved a letter setting out the facts of the case, clearly but concisely, addressed to that hospital's general

manager or chief administrator should assist a balanced appraisal of the position. All GP practices should have a written complaints procedure, but as a first step speak to the practice manager.

In the unlikely event that the problem remains unresolved by these methods recourse can be made to the relevant ombudsman:

England - Health Service Ombudsman (www.ombudsman.org.uk)

Scotland - Scottish Public Services Ombudsman www.spso.org.uk

Wales - Public Services Ombudsman for Wales www.ombudsman-wales.org.uk

Ireland - Northern Ireland Ombudsman www.ni-ombudsman.org.uk

Some ITP Support Association publications

- *Protocol for Dentists.*
- *Drugs that cause or aggravate thrombocytopenia.*
- *Holiday insurance & travel guide (including 'Is air travel safe?)*
- *Guidelines for schools, clubs and playgroups*
- *What's it called (Adult ITP Q & A)*
- *What's it called (Childhood ITP Q & A)*
- *Employer's factsheet*
- *ITP 'n' Stuff. (for teenagers)*
- *Increasing platelet production.*
- *Intravenous immunoglobulin – the good news and the bad news.*
- *ITP and pregnancy*
- *ITP and tiredness.*
- *Helicobactor pylori & ITP*
- *MMR vaccine & ITP*
- *Rituximab for ITP.*
- *Splenectomy – a guide for ITP patients.*
- *Steroids - a boon and a bane.*
- *Surgery & the patient with ITP*
- *The relationship between ITP & lupus (SLE)*
- *Choosing your sport*

About the ITP Support Association

The ITP Support Association is a registered charity and non-profit-making organisation. Founded in 1995, it is run primarily by volunteers and exists to promote and improve the general welfare of adults and children with Immune Thrombocytopenia.

The Association co-operates with the medical profession to advance the knowledge and treatment of ITP and runs occasional medical seminars for health professionals. It funds ITP research, assists ITP data collection and conducts occasional surveys of its membership, the results of which are published to further educate health professionals about the problems of living with ITP.

Members will be sent an information pack, our quarterly journal, *The Platelet*, and a full list of available Association publications. ITP Support Association factsheets and special publications are free of charge to subscribing members, but we appreciate an SAE.

The Association depends on donations and fundraising activities in order to continue its work and fund much needed research (as there is no state funding for ITP research). Updates on the projects will be given periodically in *The Platelet* and full details of the research projects can be found on our website at *https://www.itpsupport.org.uk/index.php/en/itp-research*

The Association aims:

- to offer support and information to ITP patients and their family members
- to provide contact with other patients/parents and international ITP specialists at ITP conventions.
- to provide information and patient feedback to clinicians and other health professionals.
- to help patients/parents understand benefits and risks of treatment choices
- to provide specialist advice when necessary through its medical advisory team
- to answer questions on lifestyle, insurance, sickness benefits and travel issues.
- to assist the understanding of ITP within the medical profession by running educational seminars
- to make representations on behalf of people with ITP to leading ITP specialists, health departments and the pharmaceutical industry.
- to provide information to GPs, dentists and schools
- to fund research into the cause and best management of this frightening condition.

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