

Management of Acute Immune Thrombocytopenic Purpura (ITP)

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Intended Audience

This document contains information and clinical guidelines for management of children attending the Sheffield Children's Hospital (SCH) or designated shared care centres. It is to be used by staff within SCH, the shared care Trust or the community whenever they are caring for these children either in hospital or at home.

Purpose

This guideline is designed to guide general paediatricians in the investigation and management of children with suspected acute Immune Thrombocytopenia (ITP) aged 6 months to 16 years.

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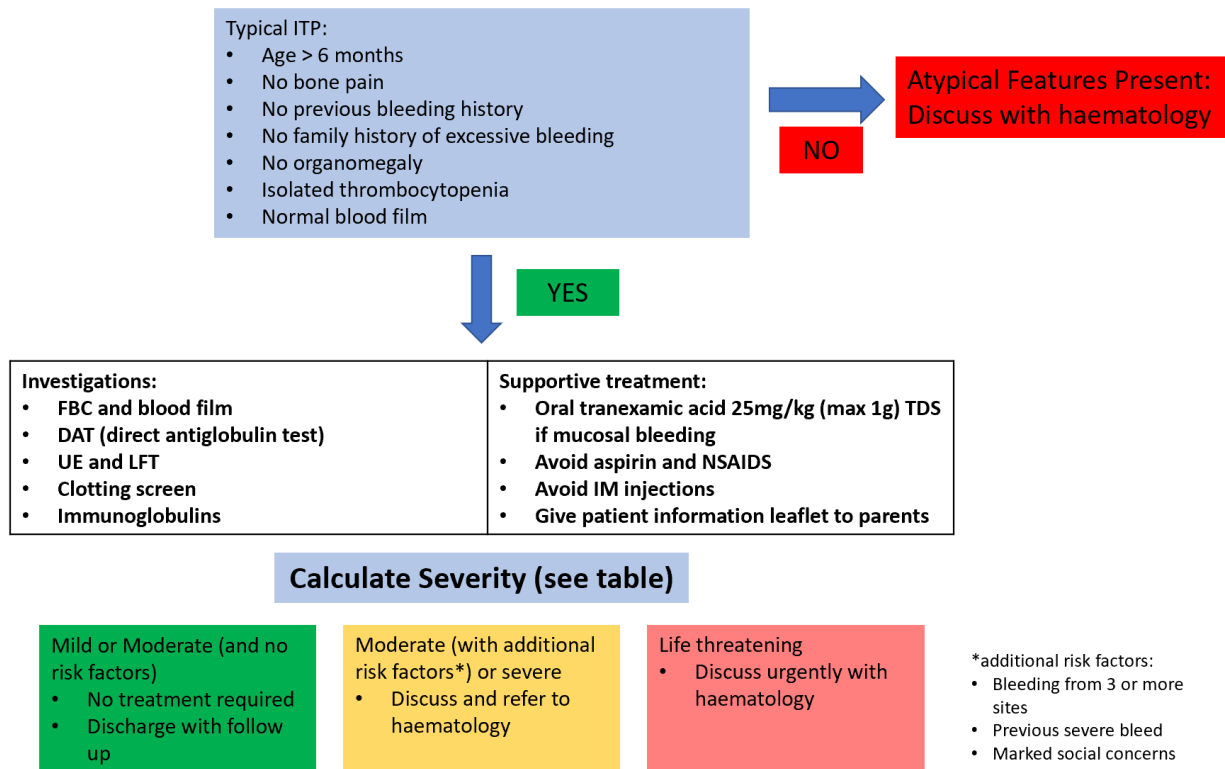
1. Introduction

ITP may present with bleeding, bruising or a petechial rash. Children often have a platelet count of $<10 \times 10^9/L$ and it often follows a self-limiting viral infection.

A diagnosis of ITP can be made when **ALL** of the following criteria are fulfilled:

1. Isolated thrombocytopenia with an otherwise normal full blood count (FBC)
2. A normal blood film other than thrombocytopenia
3. Absence of atypical features, including
 - Bone pain or limp
 - Abnormal Lymphadenopathy
 - Hepatosplenomegaly
 - Persistent fever
 - Macrocytosis
 - Family history of excessive bleeding
 - Personal history of excessive bleeding, prior to this presentation

2. Assessment



Management of Acute Immune Thrombocytopenic Purpura (ITP)

Severity Grade	Bleeding	Management
Mild	<ul style="list-style-type: none"> Few petechiae and bruises < 5cm Nosebleed that stops within 20 mins with pressure 	Watch and monitor
Moderate	<ul style="list-style-type: none"> Numerous petechiae and large bruises > 5cm Intermittent bleeding from gums, lips, buccal, oropharynx, GI tract Hypermenorrhagia, haematemesis, haematuria, melaena without hypotension and Hb dropping by < 20g/L 	Watch and monitor, or treatment for selected cases
Severe	<ul style="list-style-type: none"> Epistaxis requiring nasal packing or cautery Suspected internal haemorrhage (lung, muscle, joint, others) Hypermenorrhagia, haematemesis, haematuria, melaena leading t hypotension or Hb dropping by > 20g/L 	Treatment as per haematology
Life threatening	<ul style="list-style-type: none"> Intracranial haemorrhage Continuous or high volume bleeding resulting in: <ul style="list-style-type: none"> Hypotension or prolonged capillary refill, AND Requiring fluid resuscitation or blood transfusion (>10ml/kg) 	Urgent treatment, as per haematology

3. Management and Follow Up:

If the patient has been assessed as suitable for watch and monitor:

- The patient should have a method of open access to their local hospital – this could be through the emergency department or directly to the paediatric ward, as per local policy
- A patient information leaflet should be given prior to discharge – see appendix 1
- Information should be given to parents regarding avoidance of NSAIDs, Intramuscular injections, contact sports and presentation and management of head injuries. This conversation should be documented in the notes
- Oral Tranexamic acid (15-25mg/kg, 2-3 times a day, max 1.5g per dose) can be given for patients with mucosal bleeding.
- A repeat full blood count should be performed at 1 week, and then every 2-4 weekly depending on symptoms and platelet trend
- Patients can attend school but should be advised to avoid any activities that carry a risk of falls or head injuries. Schools should be advised of the need to seek urgent medical attention for bleeding or head injuries.
- If the patient remains thrombocytopenic at 3 months, then a referral should be made to haematology at Sheffield Children's Hospital to discuss possible treatment options for persistent or chronic ITP.
- Patient's whose platelet count completely normalises on two consecutive FBCs can be discharged.

4. Indications for discussion with haematology at initial presentation

- Patient with severe bleeding (as per above table) or requiring treatment (other than tranexamic acid) due to bleeding symptoms
- Patients with any other abnormalities of the FBC or investigations performed at initial investigation (other than co-existing iron deficiency which should be appropriately managed)
- Patients with atypical features on history or examination

Any patient with a major or life threatening bleed, including a suspected intracranial bleed should be discussed urgently with the haematology registrar or consultant on-call at Sheffield Children's Hospital.

5. References

D Provan, D Arnold, J Bussel et al. Updated international consensus report on the investigation and management of primary immune thrombocytopenia. *Blood Advances* 2019;3(22):3780-3817

Grainger J. Suspected or known Immune Thrombocytopenia Management Plan (Children). North West Guideline. http://www.uk-itp.org/docs/ITP/suspected_or_known_immune_thrombocytopenia_management_plan_children.pdf [date of access 28/03/2020]

Appendix 1**Patient Information Sheet**Sheffield Children's Sheffield Children's Hospital
Western Bank
Sheffield
S10 2THTelephone: 0114 271 7000
www.sheffieldchildrens.nhs.uk**Acute ITP: Information for Patients and Parents****Background**

The blood contains tiny cells called platelets, which help to prevent bruising and bleeding. If there are not enough platelets in your blood, cuts in your skin can carry on bleeding longer than usual, your gums can bleed when you clean your teeth and you can bruise without even being aware that you have knocked yourself. You may also develop little red dots on the skin.

Platelets are produced in the bone marrow and travel around in the circulation until they are needed at a site of injury (such as a cut or bang). If they are not needed in this way they normally survive for about 10 days before being removed from the bloodstream and replaced with new ones from the 'Platelet factory' in the bone marrow.

What is ITP?

ITP is an acquired illness (you are not born with it) in which the body's own immune system removes normal platelets from the bloodstream by mistake. The immune system usually protects you by finding, labelling and getting rid of things that don't belong in your body – such as germs. In ITP the immune system mistakenly gets rid of your own platelets by attaching a special protein called an antibody to them so that they are attacked and destroyed. Although you are making plenty of platelets, they are being 'used up' more quickly than usual and you become 'thrombocytopenic' which just means you have a low number of platelets. Why the body begins to produce antibodies against its own platelets is not known. It may be that an infection such as a cold or sore throat a few weeks beforehand may trigger the immune system to behave in this way.

The problems you may notice are bruises, also known as 'purpura', tiny red dots on the skin, known as 'petechiae', nose bleeds, and sometimes other bleeding such as gum bleeding when you clean your teeth. Girls can have heavy, prolonged periods.

A normal platelet count is between about 150 and 450 depending on age. Often people with ITP have platelet counts of less than 20 when they are first diagnosed with the condition. Experience has shown us that although there may be easy bruising, children with platelet counts around 20 rarely have any serious bleeding.

What are the symptoms to expect in ITP?

Most children seem perfectly normal, happy, active and well apart from the bruises and sometimes little red spots on the skin. The child will not have any pain or fever and will usually just be their normal selves. Some children may have nose bleeds which may take quite a while to stop, or they may have bleeding or 'blood blisters' in the mouth. Serious bleeding is very rare.

Is it very dangerous to have a low platelet count?

Although the numbers of platelets are reduced, the bone marrow is working hard supplying new platelets to replace the ones being removed by the immune system and the platelets that are present work very well. Although the symptoms described above can be alarming at first, they are usually not dangerous and the safest thing to do once the correct diagnosis has been made, is to just wait for the platelet count to recover on its own.

One important and serious problem that can occur very rarely is bleeding inside the head. This is called an **intracranial haemorrhage**. It is important to contact your hospital if your child has a head injury whilst their platelets are low so that they can be assessed and observed on a ward if necessary. It is also important to contact a doctor if you feel your child is not themselves, with drowsiness, vomiting weakness or fits.

Although intracranial haemorrhages are very rare, it is important to avoid activities that carry a risk of head injury, until the platelet count begins to recover. Depending on the age of your child, these might include climbing frames, rollerblading, horse riding, trampolining, contact sports etc. Individual activities can be discussed with your doctor.

Is ITP common?

ITP affects about 4 in every 100,000 children each year. It happens most frequently in both boys and girls aged between 2 and 6 years but any age can be affected.

How is the diagnosis made?

This is done by listening to your description of your child's symptoms examining your child and taking a blood test to exclude other causes of low platelets. Occasionally if the platelet count is taking longer than usual to recover or if there are any symptoms that are unusual a bone marrow test may need to be done. The reasons for this and the procedure itself would be explained fully by your doctor.

How do you treat ITP?

Mostly ITP just gets better on its own, so the most important thing to do is to reduce the risk of any serious bleeding whilst waiting for recovery to happen. Medicines such as Ibuprofen and aspirin should be avoided as they stop the platelets working effectively. It is safe to use Paracetamol if it is needed.

Intramuscular injections should be avoided – usually they can wait until the child has recovered, if not they can be given subcutaneously (just under the skin, not deep into the muscle). Activities should be restricted as described earlier and the child may temporarily need closer supervision, depending on their age. Any head injuries should be reported to the hospital. The child can continue attending school or nursery as previously but the staff should be made aware of the information on this fact sheet.

You will be given a 24hr/day contact phone number to ring if you ever need advice or reassurance.

If there is no important bleeding we usually just wait for your child's platelets to come back up to normal on their own. Minor mouth and nose bleeding can be helped by a medicine called **tranexamic acid** which makes any blood clots that do form stronger and less likely to dissolve away. It doesn't have any effect on the platelet count itself.

If there is important bleeding, or your child needs an operation (for some unrelated condition) it may be necessary to give specific treatment for the ITP to try to raise the platelet more quickly. If this was required, your doctor would discuss the treatments available with you.

How long does ITP last?

Most children have acute ITP which lasts for less than 3 months. In fact most are showing signs of recovery within a month and a few within a week. Children who have platelets which remain low between 3-12 months have persistent ITP. There are some children (10-20%) who still have low platelets longer than 12 months after diagnosis. This is called chronic ITP. The chronic form is commoner in girls and in older children but it is not possible to tell from the outset who will recover quickly and who will not. A very few children make a full recovery but then their platelet count falls again weeks or months later, often following a viral infection, and then the platelets usually return to normal again. This can be repeated several times and is known as relapsing ITP.

Eighty to ninety percent of children will have a normal platelet count a year after diagnosis without requiring treatment. Any child who still has low platelets 3 months after the initial diagnosis, will be referred to the haematology team at Sheffield Children's Hospital.

Can you catch ITP?

No. No one will catch ITP from your child and your child did not catch it from anyone.

What should I do if I'm worried?

At home watch your child for new bruises and bleeding. These should gradually improve and the old bruises and petechiae will completely disappear. If your child has a head injury or develops bleeding that will not stop with simple measures you should contact the hospital via the telephone numbers you were given. There is a space to write these numbers at the bottom of this leaflet.

Contact Details:

Ward: _____

Telephone Number: _____