PRESCRIBING GUIDELINE CONTROL PAGE	
Title	Title: Suspected or known Immune Thrombocytopenia Management Plan (Children) Version: 1 Reference Number: MMC-G117
Supersedes	Supersedes: Nil Changes:
Minor Amendment	Date January 2015 Notified To Date Summary of amendments – New contact numbers, new referral form, new patient record. Treatment with anti-D removed as no longer available
Author	Originated / Modified By: Dr J D Grainger Designation: Consultant Paediatric Haematologist
Ratification	Ratified by: Paediatric Medicines Management Committee Date of Ratification: February 2015
Application	All Paediatric Staff
Circulation	Issue Date: March 2015 Circulated by: Paediatric Haematology Team Dissemination and Implementation: All clinicians – RMCH, Nursing staff – RMCH, All pharmacists
Review	Review Date: January 2020 Responsibility of: Dr J D Grainger (ITP Lead)
Date placed on the Intranet: March 2015	Please enter your EqIA Registration Number here: 106/15

1. Introduction

This care pathway is designed for use in children over 6 months of age who present with bruising or bleeding but are otherwise well with no history of bone pain. Apart from bruising or petechiae, examination should be otherwise normal with no significant lymphadenopathy or hepato-splenomegaly. The full blood count (FBC) should show depression of the platelets only with an otherwise normal blood film.

The absence of these features does not exclude Immune Thrombocytopenia (ITP) but warrants discussion with regional paediatric haematologist and may need a bone marrow aspirate to confirm the diagnosis. Please contact Dr John Grainger/ Paediatric Haematology Consultant colleague at RMCH on 0161-701 8416 during office hours or consultant paediatric haematologist on call via RMCH switchboard (0161-2761234).

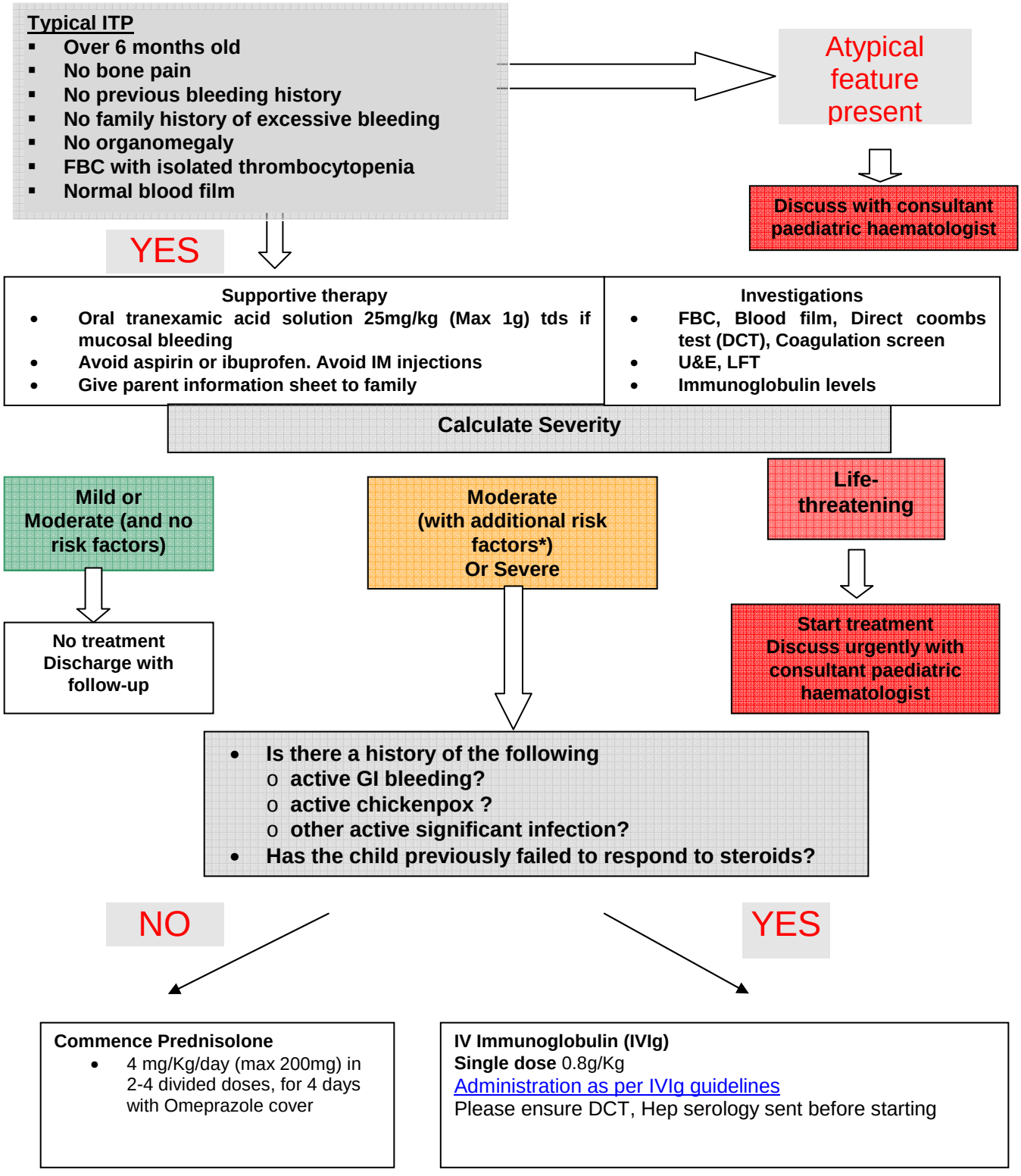
2. Purpose

The following guidelines are provided for all members of trust staff involved in clinical care of children. They provide information on the management of paediatric patients with suspected ITP. They should be used in conjunction with the individual patient's history.

3. Roles and Responsibilities

- The Divisional Clinical Effectiveness Manager will ensure that Consultant staff are informed of new and reviewed documents, and that teams responsible for induction of Junior Medical staff are informed of current Divisional documents (to allow incorporation into induction)
- Consultant Medical staff are responsible for reinforcing guidelines appropriate to their specialty with junior medical staff
- The Deputy Director of Nursing – Children and the Head of Nursing (Children) will ensure that guidelines are available in every ward/department.
- Lead Nurses will ensure that all nursing staff within ward and specialty based teams within their Directorate are aware of and adhering to all Divisional guidelines and policies
- Modern Matrons, Ward Managers and Specialist Nurses will promote the guidelines and ensure adherence to them by nursing staff
- All nursing staff are responsible for ensuring:
    - they are familiar with nursing guidelines and policies
    - their practice follows nursing guidelines and policies

4. Assessment:



***Additional risk factors include three or more bleeding sites (eg skin, Oral, GI), previous severe bleed, marked social concerns**

Severity grade	*Bleeding*	*Management*
Mild	• **Few petechiae and small (<5 cm) bruises** • **Epistaxis, stopped by applied pressure within 20 minutes**	**Watch and monitor**
Moderate	• **Numerous petechiae and large (> 5 cm) bruises** • **Epistaxis longer than 20 min.** • **Intermittent bleeding from gums, lips, buccal, oropharynx, or gastro-intestinal tract.** • **Hypermenorrhagia, haematemesis, macroscopic haematuria, melaena- without hypotension and falling Hb<2 g/dL**	**Watch and monitor OR Treatment for selected cases**
Severe	• **Epistaxis requiring nasal packing or cautery** • **Suspected internal haemorrhagia (lung, muscle, joint, others)** • **Hypermenorrhagia, haematemesis, haematuria, melaena leading to hypotension or falling Hb>20 g/L**	**Treatment**
Life threatening or ICH	• **Intracranial Haemorrhage (ICH) or** • **Continuous or high volume bleeding resulting in** ▪ **hypotension <u>OR</u> prolonged capillary refill <u>AND</u>** ▪ **Requiring fluid resuscitation <u>OR</u> blood transfusion (>10ml/Kg)**	**Urgent treatment**

Management

1. In the presence of atypical features (bone pain, failure to thrive, lymphadenopathy, additional cytopeniae) or severe/ suspected life-threatening bleeding then discuss urgently with local consultant and consultant paediatric haematologist (Dr John Grainger in day or consultant on call)

2. Obtain history/ examination and FBC. You should speak with the local haematologist on call so the blood film is examined to exclude leukaemia. The child should stay on the unit until blood film is reported (may require overnight admission).

3. Document bleeding sites and severity of bleeding from each site.

4. Additional investigations should include Direct Coombs Test DCT and Immunoglobulin levels. Further investigations such as clotting studies may be necessary based on the severity of bleeding and clinical history.

5. The majority of children will have only mild or moderate bleeding symptoms and will not require treatment other than supportive therapy

6. Children with complex social backgrounds or additional risk factors for severe bleeding (bleeding from multiple sites, previous severe bleed) may require extended observation or treatment even with mild or moderate bleeding problems.

7. If treatment is required Steroids (Prednisolone 4mg/Kg/day, max 200mg, in divided doses for 4 days) are the preference providing there is no active infection or GI bleeding. IVIg causing a more rapid platelet recovery but is associated with a higher risk of side effects and should be reserved for severe bleeding or patients previously unresponsive/ intolerant of steroids. A bone marrow aspirate is not required prior to steroids in the absence of atypical features.

8. Platelet transfusions do not work unless administered with IVig and should be reserved for severe bleeding or if the thrombocytopenia is not thought to be ITP.

9. A patient information sheet is provided in Appendix A and should be copied for parents.

10. On discharge a management plan should be completed and filed in notes and a copy given to the family (Appendix B)

11. Local follow up should be with Dr Grainger (01617018416). Please Fax referral form (Appendix C) to 01617018410.

    a. Child should have open access to ward and be advised to return urgently in the presence of any of the following:

        i. A prolonged (over 20 minutes) nosebleed which will not stop despite pinching the nose

        ii. Prolonged gum bleeding

        iii. Blood in the poo or urine

        iv. Following a heavy blow to the head, particularly if the child is stunned or sickly

        v. Persistent or severe headache

        vi. Vomiting or drowsiness

        vii. Children on steroids are at a greater risk of a severe form of chickenpox. If your child has not had chicken pox then contact the hospital If your child is in direct contact with someone who has chicken pox or who develops chickenpox within 7 days of being with your child.

5. Equality Impact Assessment.

5.1 This guideline has had an initial Equality Impact Assessment (please refer to Document Control Sheet for details).

6 Consultation, Approval and Ratification Process

6.1 Consultation Process, Consultation and Communication with Stakeholders

Consultation on this document has been undertaken through the ITP Clinical Forum:
Dr Phil Connor (Cardiff)
Dr Keith Sibson (GOS)
Dr Jayshree Motwani (Birmingham)
Dr Liz Chalmers (Glasgow)
Mrs Shirley Watson (ITP Support)
SN Rachel McDermott (RMCH, ITP Nurse Specialist)
Also sent to regional hospitals including:
Jon McViety (Bolton)
Vanessa Holmes (Blackburn)
Reviewed by Mike Wilkinson (Pharmacy CMFT)

6.2 Policy Approval Process

The Paediatric Medicines Management Committee has approved the medicine-related components of this guideline

6.3 Ratification Process

The Paediatric Medicines Management Committee has ratified the medicine-related components of this guideline.

7 Dissemination and Implementation

7.1 Dissemination

7.1.1 Medical Staff
Email from Divisional Clinical Effectiveness Team to Consultant groups identified within circulation
Email from PGME to all junior medical staff in groups identified within circulation, inclusion of document on Junior Medical Staff induction CD

7.1.2 Intranet
Document to be placed on the intranet under pharmacy guidelines and previous versions removed.

7.2 Implementation of Procedural Documents

Lead Nurses are responsible for identifying arrangements for training and support for nursing staff.
Consultant medical staff are responsible for reinforcing the guidelines to junior medical staff in their team.

8 Monitoring Compliance of Procedural Documents

8.1 Process for Monitoring Compliance.

As a minimum include the review and monitoring arrangements:-

Compliance with the guideline will be monitored during daily clinical ward visits. No formal audit of compliance is required.

9 Appendices:

Appendix A- What is Immune Thrombocytopenic Purpura (ITP)? –parent leaflet

Appendix B: ITP patient record

Appendix C: ITP New Referral

Appendix A

What is Immune Thrombocytopenic Purpura (ITP)? –parent leaflet

Introduction

This leaflet explains about immune thrombocytopenia purpura (ITP), this is sometimes referred to as idiopathic thrombocytopenia purpura. ITP is a blood disorder causing the number of platelets to drop. This leaflet explains what to expect when your child is diagnosed with the condition.

What are platelets?

Platelets are one of the three types of blood cell, along with red and white blood cells. Platelets are small and sticky and their job is to prevent bruising and stop bleeding after an injury. Platelets, like red and white blood cells, are formed in the bone marrow. A rough idea of how many platelets are circulating in the bloodstream (platelet count) can be made using a sample of blood. The normal platelet count is between is 150 to 400 x 10^9/l. In most cases of ITP the platelet count is less than 20 x 10^9/l. A low platelet count is called 'thrombocytopenia'.

What is ITP?

Immune thrombocytopenia is a medical term for a condition in which there is bruising (purpura) because there are fewer platelets in the blood than usual (thrombocytopenia) and is usually caused by something going wrong with the immune system (the body's defence against infection) or an allergic reaction of some kind.

Chronic ITP is the term for ITP that has not gone away on its own after 12 months. Only 1 in 4 children with ITP will develop chronic ITP. The majority of children with "chronic" ITP will still have some recovery of the platelet count at a later date and the majority of younger children will still completely recover after a few years even if the ITP is still present at 12 months.

How common is ITP and who does it affect?

About four in every 100,000 children develop ITP each year. There seem to be two groups who develop ITP: young children and young adults. It is more common in girls than boys.

What are the symptoms of ITP?

Most children with a platelet count of under 20 x 10^9/l will have petechiae (pinprick blood spots under the skin) and limited bruising. Bruising most commonly follows minor knocks ("easy bruising") but may also occur spontaneously without trauma. Apart from the bruising/ bleeding the children are otherwise well. Common sites of spontaneous bleeding are the gums and nose. Girls may be troubled with heavy periods.

Less common and potentially serious are spontaneous bleeds occurring from the gut or brain. Data from international studies suggests that the risk of serious bleeds is about 3 in 100 children and the risk of brain bleeds is about 1 in 300 children. These bleeds most often occurred in the first week of ITP and most frequent in children with more significant bleeding. The risk of serious bleeding is much lower when the platelet count recovers to over 20 x 10^9/l.

What causes ITP?

ITP commonly results due to the immune system mistaking platelets as being foreign and attacking the platelets. In many cases this may follow a viral infection or vaccination during which time the immune system attacks the virus but the immune system then goes on to think that the platelets are viral material and starts to attack the platelets.

How is ITP diagnosed?

ITP is usually diagnosed using a blood test called a 'full blood count'. When a sample of your child's blood is examined under a microscope, a haematologist can examine each blood cell type closely. This is to rule out other conditions that may cause similar symptoms to ITP. If the platelets, red blood cells and white blood cells all look normal, this rules out leukaemia. If the low platelet count improves quickly and no treatment is needed, your child will not need any further tests.

If the platelet count is not showing signs of recovery by 3 to 6 months then a small sample of bone marrow may need to be taken and examined under the microscope. Additional blood tests may be taken at this time to exclude rare clotting or immune diseases that can mimic ITP. If the bone marrow looks normal, with the usual or higher number of platelet parent cells (megakaryocytes) and other blood tests are normal then the doctor will diagnose persistent ITP.

What is the outlook for children with ITP?

Many children, particularly younger ones, suddenly improve within six weeks, whether or not treatment has been given. Three out of four children will have improved by 6 months after the start of ITP. Even those who fail to recover completely usually reach a platelet count over 20 x 10^9/l and have fewer bleeding problems. After six months about 25% of children will fully recover over the following year and over half will recover over several years.

When ITP recovers about one in 20 children will have a further occurrence in the future.

How is ITP treated?

Most children do not need any treatment unless they have severe bleeding, and most children improve whether or not treatment is given. The type of treatment recommended depends on your child's symptoms rather than their platelet count. All the various forms of treatment aim to temporarily improve the platelet count and do not cure the condition itself. When treatments are considered, you will have the chance to discuss the risks and benefits of these, as opposed to no treatment, with the doctor. The options for treating ITP include:

1) No treatment

The majority of children with ITP have a low platelet count but do not have dangerous bleeding. If severe bleeding is not present at the time of diagnosis then it is very rare for dangerous bleeding to develop later. Without treatment most children will have a platelet count > 20 $x10^9$/l within 5 days and a normal platelet count by six months.

2) Tranexamic acid

Tranexamic acid does not increase the platelet count but does help the blood to produce clots. It is particularly useful for gum bleeds, nose bleeds or heavy periods and helps the blood to form clots without altering the platelet count. It is best taken as a liquid ("swish and swallow") three times per day. It must not be used if there is any blood in the urine.

3) Steroid treatment

Steroids are sometimes given to children with ITP on a short-term basis in an attempt to increase their platelet count. However, when the steroid dose is reduced, the platelet count will drop again after a few days. Steroids should only be given for a short period of between 4 to 14 days. Side effects such as weight gain and mood changes are common. Longer courses of steroids may dampen the immune system, weaken bones, cause diabetes or obesity and stunt growth.

4) Intravenous immunoglobulin

Immunoglobulins are antibodies which can reduce platelet destruction. They are a blood product produced from many donors and have a theoretical but very low risk of transmitting blood-borne infections. Treatment with immunoglobulin takes a full day at the hospital and the benefit will usually last about a month. Side effects such as fever and headaches are common.

5) Splenectomy and other treatments

In ITP the majority of platelets are destroyed in the spleen. Removing the spleen (splenectomy) is often effective in preventing early destruction of the platelets and allows the count to rise. In children however this is rarely

necessary unless the ITP persists longer than a year and the child has recurrent severe bleeds. Splenectomy is a major surgical procedure and carries a long term risk of severe infection. Other treatments to suppress the immune system (e.g. Rituximab, Ciclosporin) or stimulate platelet productions (e.g. Eltrombopag) are usually tried prior to splenectomy.

What about school, sport and holidays?

Most severe bleeds tend to occur in the first week and in children with a platelet count under 20 $x10^9$/l. In those children with a count over 20 $x10^9$/l they can return to school immediately after the head teacher has been informed about the ITP. In children with a lower platelet count school can resume after the first week and when the school have been informed. For primary school aged children with platelet counts under 20 $x10^9$/l it may be best if they take breaks inside if these can not be supervised. The ITP Support Association produces a document for schools, clubs and playgroups.

If your child is on steroids and has not had chicken pox then school will need to inform you if anyone in your child's class/nursery comes down with chicken pox.

At home it is best to take sensible precautions which all children should follow such only cycling with a helmet and if swimming no diving into the shallow end! It is sensible to avoid contact sports where there is a risk of head injury whilst the platelet count is below 50 $x10^9$/l. Make sure any sports teachers are aware. With a platelet count between 50 and 100 $x10^9$/l there may still be more bruising so encourage the use of shin pads etc. For further details discuss with your consultant.

It is best not to take any holidays abroad in the first three months of ITP as it may be difficult to get insurance. After this time most cases of ITP will have resolved. If the ITP does persist you will need to discuss further with your doctor and you will need specialist medical insurance. A list of recommended insurance companies can be obtained from ITP Support Association (details below)

What else can I do?

Your child should also avoid drugs like aspirin, ibuprofen or herbal medication which can increase the risk of bruising and bleeding. Finally, you should make sure that doctors and dentists know that your child has a low platelet count if they are due to have an operation.

When to seek help?

When your child is sent home you will be given a clinic appointment for review at the hospital and an emergency number (usually the phone number to the children's ward). You should contact the hospital in the following circumstances:

- A prolonged (over 20 minutes) nosebleed which will not stop despite pinching the nose
- Prolonged gum bleeding

- Blood in the poo or urine
- Following a heavy blow to the head, particularly if the child is stunned or sickly
- Persistent or severe headache
- Vomiting or drowsiness
- Children on steroids are at a greater risk of a severe form of chickenpox. If your child has not had chicken pox then contact the hospital If your child is in direct contact with someone who has chicken pox or who develops chickenpox within 7 days of being with your child.

Is there a support group?

The ITP Support Association
'Synehurste'
Kimbolton Road
Bolnhurst
Bedford MK44 2EW
Tel 0870 777 0559 Website: www.itpsupport.org.uk

Is there a UK registry?

To maintain accurate numbers of cases of childhood ITP and investigate possible markers for risk of severe bleeding a UK registry has been established (www.uk-itp.org) Families may be routinely asked to consent for anonymous data to be stored on the registry.

Appendix B: ITP patient record

PATIENT DETAILS

Surname: **NHS number:**

Other names: **DOB:**

Address: **GP Name & Address:**

Tel: **GP Tel:**

Weight (KG): **Allergies:**

Other medical problems: **Current Medication:**

Hospital contact Name(s) and number(s):

Date and location of next follow up:

Recent Blood counts

Date	Platelet count	Date	Platelet count

Previous treatments

Date	Treatment	Response	Comments

Treatment plan

In the event of severe bleeding the treatment plan for this patient is:

Sign/Date:

Appendix C: ITP New Referral

PATIENT DETAILS

Surname: **NHS number:**

Other names: **DOB:**

Address: **GP Name & Address:**

Tel: **GP Tel:**

Weight (KG): **Allergies:**

Past Medical History: **Current Medication:**

Referring Consultant: **Contact Details:**

PRESENTATION

Presenting Bleeding:

Bleeding Sites:

Bleeding severity: *circle as appropriate* Mild Moderate Severe

FBC at presentation: **Date** **Result**

Blood Film:

Platelet counts: Date **Result _____, Date** **Result**

Date **Result _____, Date** **Result _____,**

Date **Result _____,**

DCT test:

Immunoglobulin levels:

Additional investigations:

Treatment Plan: