LRI Children's Hospital

Acute Immune Thrombocytopenia Purpura (ITP)

Staff relevant to:	Healthcare professionals involved looking after children who present with a low platelet count
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1. Introduction and Who Guideline applies to

Primary immune thrombocytopenia (ITP) is an acquired immune mediated disorder characterised by isolated thrombocytopenia, defined as a peripheral blood platelet count of less than 100 x 10^9/L in the absence of any obvious initiating or underlying cause.

This guideline is for any healthcare professional involved looking after children who present with a low platelet count. For the majority of cases, this will be for the general paediatric and the childrens' emergency team.

Management of Acute ITP

1. Establish diagnosis – if unclear discuss with Haematology registrar
2. Assess severity based on clinical symptoms

Severe
GI bleeding, uncontrolled epistaxis, intracranial haemorrhage

1. Admit and observe
2. Group and save
3. Is the bleeding life-threatening?

Yes
- XM 2 Units RBC
- Inform haematologist

Immediate treatment:
1. IVIG (1g/kg)

Consider:
2. IV methylprednisolone
3. Platelet transfusion[1]

No
- Discuss with haematologist

Consider treating with:
1. Prednisolone (2mg/kg BD starting dose)
2. IVIG (1g/kg)[2]

Remember additional supportive measures:
- Blood transfusion
- Tranexamic acid
- Local control of bleeding

Mild
No active bleeding, controlled epistaxis (stops in less than 20 mins with gentle pressure and Hb normal), bruising, petechiae.
NB careful clinical assessment is required. If mucosal bleeding or blood spots in mouth are present consider admission +/- treatment. These patients have a higher risk of haemorrhage

No treatment required

Before discharge:
1. Educate, reassure & advise (see in management section)
2. Give contact details for ITP support association (http://www.itpsupport.org.uk)
3. Weekly follow up FBC should be booked in Childrens daycare for 4 weeks on a Monday between the hours of 10am-12pm or Thursday 1.30pm-3.30pm & email haematology nurses to follow result uho-tr.leicshaemliasionnurses@nhs.net
4. General Paediatric Consultant to arrange clinic follow up 6-8 weeks' time with open access via ED

For weekly FBC in Childrens Daycare:
- Blood test to be booked for Monday morning or Thursday after 1.30pm so haematology liaison nurses can review patient.
- If bleeding, Hb and/or neutrophils falling – needs reassessment
- FBC monitoring in daycare continues until OPD clinic appointment

If low platelets persist at 6 months, refer to paediatric haematology

2. Guideline Standards and Procedures

Pathophysiology

Immune mediated thrombocytopenia is primarily a disease of increased peripheral platelet destruction with most patients having autoantibodies directed against specific platelet membrane antigens. Antibody coated platelets are rapidly cleared by tissue macrophages predominately in the spleen leading to a substantial fall in the platelet count. Impaired platelet production and T cell mediated effects also play a role.

Epidemiology:

- 4/100 000 children in the UK
- Typically less than 10 years of age

Clinical Manifestations

ITP typically presents with the sudden appearance of a petechial rash, spontaneous bruising and/or bleeding in an otherwise well child.

History: History should be focused on assessing the risk or extent of bleeding and excluding other causes of thrombocytopenia

- Type, severity and duration of bleeding
- Presence of systemic symptoms such as fever, anorexia, bone or joint pain and weight loss which may be indicative of an underlying disease such as malignant or autoimmune disease
- A history of viral infection within the preceding month is present in about 60% of cases
- Prior history of significant disease or exposure to relevant drugs (e.g. Phenytoin, Valproate, Carbamazepine, Vancomycin, Septrin)
- Family history of thrombocytopenia or bleeding disorders
- The possibility of non-accidental injury should be considered in young children presenting with bruising

Examination: Physical examination should be normal aside from bleeding manifestations:

- Cutaneous bleeding such as petechiae, purpura and bruising
- Mucosal bleeding involving nasal passages, oral mucosa, gastro-intestinal or genitourinary tracts

- Mucosal bleeding is significantly more common if the platelet count is <10 x10^9/L

- Significant enlargement of lymph nodes, liver or spleen should prompt consideration of an alternative diagnosis (See section below)

- Neurological signs which may be indicative of intracranial haemorrhage particularly in those patients with a history of head trauma

Laboratory results

- FBC/blood film – thrombocytopenia should be the ONLY abnormality in the 3 main cell lines (atypical lymphocytes may be present in viral infections)

- Anaemia may be present IF there has been significant blood loss

- Coagulation screen – normal

- Bone marrow examination

    - Bone marrow examination is unnecessary in children with typical features of ITP and no other worrying signs or symptoms

    - Contrary to previous guidance it is no longer necessary prior to initiation of treatment with steroids

    - It is recommended in patients who do not respond to treatment or prior to splenectomy

Diagnosis

Diagnosis is one of exclusion. The presence of atypical features determines whether further investigation is needed to rule out specific causes of thrombocytopenia including:

- Bone marrow disease including leukaemia and other malignancy or aplastic anaemia

- Inherited thrombocytopenia including TAR syndrome, Wiskott Aldrich syndrome, Bernard Soulier syndrome, type IIB von Willebrand disease

- Autoimmune disease such as SLE or other rheumatoid disorders

- Immunodeficiency such as common variable immunodeficiency and Di George syndrome

- Viral infections including HIV, Hepatitis or CMV

Prognosis

ITP is typically benign and self-limiting.

- 75% will have improved by 6 months after the start of ITP
- After 6 months, about 25% will fully recover over the following year
- Intracranial haemorrhage is a rare but serious consequence (0.1-0.5%)

Management

General advice:

Reassure patients and parents – most patients can live comfortably and safely with petechiae and a low platelet count

- No IM injections
- Stop/do not prescribe NSAIDs and review any other medication
- Consider Norethisterone or the OCP in girls who are menstruating
- Advise to avoid contact sports or activities with a high risk of trauma or head injury including trampolining and soft play centers.
- Helmets should be warn when riding bikes and scooters etc
- PE should be avoided until platelet count Is >30
- Monitor mouth twice daily for blood blisters

All patients should be advised to attend ED for review under the care of general paediatrics if:

- any nosebleeds lasting longer than 20 minutes
- any prolonged gum bleeding/ new mouth blood blisters
- any blood in stool or urine
- **Give Haematology Liaison Phone number to parents: 07866 002304**
- All families should be given the contact details for the ITP Support Association (http://www.itpsupport.org.uk) and an information leaflet can be downloaded from this site

Specific treatment:

The decision to treat should be based upon the severity of bleeding symptoms and not on the platelet count alone.

In general specific treatment to raise the platelet count should only be considered in those patients with severe bleeding including GI bleeding, uncontrolled epistaxis or intracranial haemorrhage (see flow chart).

Follow up:

- All patients with a platelet count <10 should have weekly FBC monitoring under the care of the general paediatric team for the first four weeks after diagnosis on a Monday between hours of 10am-12pm or Thursday 1.30pm-3.30pm Please arrange this through the Childrens' day care unit and email the paediatric haematology specialist nurses who will follow up the results uho-tr.leicshaemliasionnurses@nhs.net
- Refer patients to the Paediatric Haematology team who have a persistent thrombocytopenia > 6 months post diagnosis

Prescribing and ordering IVIG:

IVIG is indicated in life-threatening bleeds or intracranial haemorrhage. It is a RED classification and therefore does not require approval from the Trust Immunoglobulin Assessment Panel prior to issue.

1. Complete new patient request form (available on InSite) and email to immunglobulins.mailbox@uhl-tr.nhs.uk
2. Prescribe immunoglobulin on drug chart as per IV policy
3. Send prescription and a printed copy of the request from to pharmacy who will issue

3. Education and Training

None

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Follow up blood tests after initial presentation with ITP	Audit 1 month period of children with ITP having blood tests in childrens daycare	Kaljit Bhuller	Annual	Childrens Audit Meeting

5. Supporting References

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

2. Neunert C, Lim W, Crowther M et al. The American Society of Hematology 2011 evidence based practice guideline for immune thrombocytopenia. Blood 2011; 117:4190-4207

6. Key Words

Haematology, ITP, Platelets, Petechial rash

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Kaljit Bhuller - Consultant	**Executive Lead** Chief medical officer
Details of Changes made during review: • Follow up blood tests under general paediatrics and only refer to haematology if persistent after 6 months • Day care management updated • Prognosis amended from 75% resolve spontaneously within 3 months from inset to 6 months, after 6 months 25% recover in the following year • Added indications for ED review • Amended follow-up FBC monitoring	