



**PAEDIATRIC ACUTE CARE GUIDELINE**

**Immune Thrombocytopaenic Purpura**

<b>Scope (Staff):</b>	All Emergency Department Clinicians
<b>Scope (Area):</b>	Emergency Department

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# Immune Thrombocytopaenic Purpura

## Aim

To guide medical staff with the assessment and management of immune thrombocytopenic purpura (ITP).

## Definition

ITP is a common disorder in children aged 2 to 10 years of age, presenting with bruising, mucosal bleeding and petechiae. Thrombocytopenia is caused by immune destruction of platelets, often precipitated by viral infections

## Risk

Failure to follow this guideline will potentially result in unnecessary testing and incorrect management and follow-up of patients with ITP.

## Background

- ITP is the most common cause of thrombocytopaenia in childhood
- It is the result of immune mediated destruction of the platelets and there are no other coagulation problems.
- Presenting features include petechiae, purpura (bruising) and mucosal bleeding, particularly epistaxis and oral bleeding. Rarely there may be rectal bleeding or haematuria.

- There should be NO pallor, lymphadenopathy or hepatosplenomegaly. These findings are NOT consistent with ITP
- The risk of intracranial haemorrhage is < 1%, and seems to be greatest early on in the disease.
- The underlying cause is unknown, but it is often precipitated by intercurrent viral infections.
- Children present between the ages of 2 - 10 years with a peak incidence in the preschool years. A broad differential diagnosis should be considered for the older child.
- ITP can be divided into two clinical syndromes:
  - **Acute ITP**- 90% of children. These patients present acutely with spontaneous bruising, there is rarely mucosal bleeding, and they resolve within weeks to months of diagnosis.
  - **Chronic ITP**- 10% of children. This lasts more than 6 months, and often beyond 12 months. The presentation may be more insidious.



## Assessment

- Well children present with petechiae and purpura, mucosal bleeding is uncommon.
- The clinical assessment is aimed at **excluding** other causes of petechiae/purpura and thrombocytopaenia e.g. malignancies.
- The severity of the disease is determined by the clinical picture, **NOT** the platelet count.

## History

- Petechiae and purpura - type, severity, duration
- Mucosal bleeding - epistaxis, haematuria, rectal bleeding, severity and duration
- Previous haemostasis with blood tests, intravenous cannulae or other invasive procedures
- Systemic symptoms - especially any recent viral infections in the last 6 weeks
- Recurrent infections - suggestive of immunodeficiency
- Recent live virus vaccination (e.g. MMR)
- Medications - quinine, penicillin, digoxin, anti-epileptics, salicylates, heparin, warfarin
- Family history - thrombocytopaenia or other haematological or immunological disorders
- Co-morbid conditions that may increase the risk of bleeding
- Lifestyle factors that may pose a risk for trauma and bleeding
- Possible **systemic lupus erythematosus (SLE)** - can get isolated thrombocytopaenia. Suspect in **older children** from a **higher risk ethnic background** (e.g. Aboriginal, Asian, African, Maori), **family history** of SLE or rheumatoid arthritis. Ask about a photosensitive rash, arthritis, mglagia, oral ulcers, hair loss, dry eyes or mouth, fatigue, weight loss, fever.
- Possible **malignancy** - chronic pain, fevers, weight loss, pallor

## Examination

- Usually a well looking child with normal observations
- Bleeding signs – document location and size of pupura, areas of petechiae. Look for mucosal bleeding, check for retinal haemorrhages
- Pallor
- Lymphadenopathy
- Abdominal examination – hepatomegaly, splenomegaly
- Evidence of infection
- Weight loss
- Dysmorphic features – suggestive of a congenital syndrome e.g. Fanconi syndrome, Thrombocytopaenic-Absent Radius (TAR) syndrome

## Investigations

- Urinalysis – haematuria
- Full Blood Picture (FBP) – shows thrombocytopaenia, normal haemoglobin, normal white cell count, and normal blood film aside from large (left shifted – megakaryocytic) platelets and occasionally some atypical lymphocytes.
- The platelet count may be as low as  $< 20 \times 10^9/L$
- The blood film must be reviewed by a Laboratory Haematologist (preferably Paediatric) to *confirm the film features are in keeping with the clinical diagnosis of ITP*
- INR, APTT or clotting tests are **not** required unless significant haemorrhage or non-accidental injury (NAI) is suspected.
- A bone marrow aspirate is rarely required, and is only considered when the diagnosis is uncertain and a haematological malignancy needs to be excluded

## Differential diagnoses

- Systemic Lupus Erythematosus (SLE)
- Haematological malignancies (e.g. leukaemia)
- Aplastic anaemia
- Infections – viruses, meningococcal disease
- Drug induced thrombocytopaenia
- Haemolytic uraemic syndrome
- Other coagulation disorders (e.g. disseminated intravascular coagulation (DIC))

## Management

- ITP will spontaneously remit without any treatment within 6 months and most children can be followed up as outpatients
- Children should be admitted if they have significant bleeding

- No treatment is necessary unless there is significant bleeding
- Treatment (must be decided by a General Paediatric Team) may include oral steroids and intravenous gamma globulin. These drugs do not influence the natural history of the disorder but can acutely raise the platelet count.
- Significant bleeding needing immediate intervention includes:
  - Epistaxis for more than 1 hour.
  - Profuse oral or rectal bleeding.
  - Severe menorrhagia.
  - Any internal haemorrhage (including intracranial haemorrhage).
- Consider tranexamic acid for troublesome mucosal bleeding (NB Tranexamic acid is contraindicated in patients with haematuria).
- Discuss the management of these patients immediately with the on call General Paediatric Consultant and Paediatric Clinical Haematologist (and the General Surgical Team, Neurosurgical Team, ENT Surgical Team if required).
- Exceptional circumstances to consider treatment may be imminent overseas travel, an adolescent with self-image issues, menorrhagia or a high risk of trauma. In this group, oral steroids can be considered – prednisolone 4mg/kg once daily for 4 days.

## Management of significant bleeding

- Manage Airway, Breathing, Circulation
- Establish IV access (large bore cannula if possible)
- Obtain urgent Group and Hold +/- cross matched cells
- Consider blood transfusion to achieve homeostasis
- Seek surgical assistance to manage bleeding
- Discuss treatment with on-call clinical haematologist
  - Platelet transfusions should only be given for intra-cranial haemorrhage or other life-threatening bleeding.
  - In the presence of **significant bleeding causing clinical instability**, consider Intravenous immunoglobulin (IVIg) 0.8g / kg as it can raise the platelet count rapidly.
  - A short course of high dose methylprednisolone 5mg / kg six hourly may be given for intracranial haemorrhage after IVIg and platelets, with treatment titrated against the platelet count and with rapid tapering.
- Arrange admission to PMH/PCH
- Emergency splenectomy is rarely indicated in childhood acute ITP.

## Further management

- The platelet count should be monitored no more frequently than weekly.
- In chronic ITP, splenectomy and rituximab (an anti CD20 monoclonal antibody) can be considered.

## Rural Patients

- Discuss the diagnosis and management with the nearest local Paediatrician (if available)
- If unavailable, discuss with the General Paediatric Team at PMH/PCH – they can be phoned through switchboard 24 hours a day, 7 days a week
- Most patients can be managed at home, without the need to transfer to PMH/PCH
- See the links below for the GP letter and Health Fact Sheet for parents

## Admission criteria

- Significant bleeding, irrespective of the platelet count
  - Epistaxis > 1 hour
  - Hematemesis
  - Haemoptysis
  - Intracranial haemorrhage (ICH)
- Any suspicion of intracranial haemorrhage – needs urgent diagnosis and management
- If unsure after hours, consider admission to the ED Short Stay Unit and discuss with the Paediatric Emergency Department Senior Doctor in the morning.

## Discharge criteria

Patients can be sent home from the Emergency Department when:

- The diagnosis is definite and there is no active bleeding
- The child is well and the social situation is such that there is good parental supervision and safety in the home (with respect to the risk of trauma)
- There is the opportunity reassure and educate the parents in the Emergency Department
- There is appropriate follow up arranged with the General Paediatric Team within the next 1-2 weeks

## Referrals and follow-up

- Patients discharged with ITP require review by a senior doctor (Registrar or Consultant) to reassure parents and discuss outpatient management. If discharged by a registrar this should be discussed with the Consultant General Paediatrician of the day.
- The General Paediatric Team must review the patient within 2 weeks.
- Give the parent a form for a repeat FBP (to be done prior to the appointment).
- Referral to the Paediatric Haematology Outpatient Clinic can be done by the general paediatric team if the diagnosis remains uncertain, the white cell count is abnormal, the blood film is atypical, there is failure to respond to treatment or platelets have not resolved in 6 months (Chronic ITP).

- Rheumatology referral if history or examination suggestive of SLE, particularly a positive family history of SLE or rheumatoid arthritis, older child, higher risk ethnic background (Aboriginal, African, Asian, Maori).

### Additional considerations

- Avoid IM immunisations until resolution, although in chronic cases this may need to be on a risk-benefit assessment.
- Stop / do not prescribe NSAIDS
- Avoid contact sports until resolution

### Health information (for carers)

- The patient should avoid trauma – specifically no bicycles or trampolines
- The patient should not take any non steroidal anti-inflammatory medications eg: ibuprofen
- See the [ITP Health Fact Sheet](#)


### Management paperwork

- Referral to the General Paediatric Outpatient Clinic (within 2 weeks).
- The [ITP GP Letter](#) should be printed out and faxed/posted to the GP.
- The [ITP Health Fact Sheet](#) should be given to the family.
- A FBP request form should be given to the family to do prior to their outpatient appointment.

### References

1. Provan D SR, Newland A C, et al. International consensus report on the investigation and management of primary immune thrombocytopenia. *Blood*. 2010;115(2) pp168-86
2. Cole C. Rapid update on childhood immune thrombocytopenic purpura. *J Paediatrics & Child Health*. 2012;48 pp378-9
3. Haematology BSf. Guidelines for the investigation and management of idiopathic thrombocytopenic purpura in adults, children and in pregnancy. *British Journal of Haematology*. 2003;120 pp574-96.
4. Blanchette V & Bolton-Maggs P. Childhood Immune Thrombocytopenic Purpura: Diagnosis and Management. *Pediatric Clinics of North America*. 2008;55(2) pp393-420. Endorsed by Clinical Practice Advisory Committee September 2017

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