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## IDIOPATHIC THROMBOCYTOPENIC PURPURA

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- Natural History
  - History & Physical Examination
  - Investigations
  - Management
  - Indications for Admission
  - Indications for Treatment
  - Intravenous Gammaglobulin
  - Oral Prednisone / Prednisolone
  - Treatment Failure or Relapse
  - Life Threatening Haemorrhage
  - Indications for Bone Marrow Aspiration
  - Follow-up
  - References
- 

The most common age of diagnosis is between 2 and 10 years. The usual presentation is an acute onset of bruising, purpura and petechiae. Less commonly there is mucosal bleeding from gums, nose and rectum. Almost two-thirds of children have a history of a viral infection within the preceding 3 weeks.

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### Natural History

50% of children recover in 3-4 weeks, 75% in 6 months, 90% within 12 months. Chronic ITP is arbitrarily defined as thrombocytopenia that persists for 6 months or more. There is some evidence that treatment may increase the risk of chronicity. Intracranial haemorrhage occurs in 0.1 - 0.5% of cases of acute ITP, and overall mortality rates are of the order of 0.3%. There is no evidence to date that therapy reduces the risk for ICH.

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### History & Physical Examination

These should aim at excluding other causes of thrombocytopenia e.g. drug induced, emerging autoimmune disease. A family history of bleeding is suggestive of von Willebrand's disease whilst a history of recurrent infections raises the possibility of an immunodeficiency (eg Di George's or Wiskott-Aldrich syndromes).

The presence of pain (limb or abdominal), a limp, lymphadenopathy or hepatosplenomegaly should lead to the consideration of more serious pathology. Short stature or skeletal malformations occur in Fanconi's anaemia or thrombocytopenia absent radius syndrome.

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### Investigations

A full blood count and platelet count is always required. All blood films are to be discussed with a Paediatric Haematologist. At least two separate platelet counts should be performed to exclude artificially low counts e.g. due to clotting.

Bone marrow examination is only indicated if the history, physical examination (lymphadenopathy, hepatosplenomegaly), blood count (neutropenia, anaemia) or blood film (macrocytosis) is atypical. Such cases require consultation with a Paediatric Haematologist.

An auto-immune screen should be performed in chronic cases, as ITP may be a presenting feature of SLE (particularly adolescent girls).

APTT and INR should not be requested routinely.

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### Management

Most children can be managed as outpatients. The risk of serious bleeding, in particular intracranial haemorrhage is probably as small at home as in hospital.

Avoid drugs that reduce platelet adhesiveness (e.g. salicylates, antihistamines, *NSAIDs*). Paracetamol is satisfactory if required, but always suspect intracranial haemorrhage in the presence of headache. IM injections are contraindicated. Rough play and high risk contact sports should be discouraged.

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### Indications for Admission

Indications for admission include:

- Active bleeding
- *Mucous membrane bleeding*
- If parents do not understand the nature of the disease, require further education, live a long distance from medical care or are unable to observe the child closely at home
- Diagnostic uncertainty

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### Indications for Treatment

Treatment should only begin after blood films have been reviewed.

Some advocate treatment to prevent significant clinical bleeding in children with very low platelet counts (e.g less than  $20 \times 10^9/L$ ). There is no evidence that such treatment prevents intracranial haemorrhage. Others advocate "watchful waiting" and argue that therapy exposes some children to undesirable side-effects.

Treatment should be given to children who have:

- evidence of mucosal bleeding (e.g. petechiae of soft palate or buccal mucosa)
- haematuria or retinal haemorrhages.
- a need for urgent increase in platelet count (e.g. emergency surgery or suspected intracranial haemorrhage - see below).

The two alternatives for treatment outlined here are either intravenous immunoglobulin or high dose oral Prednisone / Prednisolone. A minority of children may not respond to either therapy. In those who do respond, there is a risk of relapse in up to 25% of cases with either therapy.

The risks and benefits of the two treatment options must be fully explained to the parents, and written consent for Intragam will be required.

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### Intravenous Gammaglobulin

Intravenous immunoglobulin is the treatment of first choice. It can be given as a single dose. There is good evidence that the platelet count rises more quickly and more predictably than Prednisone / Prednisolone or no specific therapy, thus resulting in earlier discharge from hospital. It avoids the potential side effects associated with high dose oral steroids.

Transient side effects of headache, fever and nausea may be seen during the course of the infusion (see the guidelines on Intragam in this folder). The Blood Transfusion Centre has produced a consent form for all blood products.

**Dosage:** 0.8 gm / kg as a single dose by IV infusion

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### Oral Prednisone / Prednisolone

These should never be used if there is diagnostic uncertainty.

High dose Prednisone / Prednisolone can be used in a dose of 2-4 mg / kg / day (maximum daily dose is 60 mg) for 4-7 days. Some have used a 4-day course, but a 4-day course may have a greater risk of rebound thrombocytopenia.

With this duration of therapy, there is no need to taper the dose and virtually no risk of significant HPA axis suppression

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### Treatment Failure or Relapse

For those patients given Intragam, if there is inadequate rise in platelet count after 72 hours, a further dose of Intragam 0.8 gm / kg may be given. If there is still no response, discuss the child with a Paediatric Haematologist before deciding to give oral Prednisone / Prednisolone.

If there is no effect within 7 days of beginning Prednisone / Prednisolone therapy, therapy with Intragam should be offered. Discuss with a Paediatric Haematologist.

Intragam is recommended for treatment of relapse in those who have previously responded to Intragam. Those who relapse after initial response to Prednisone / Prednisolone should be discussed with a Paediatric Haematologist. They may be offered another course of Prednisone / Prednisolone.

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### Life Threatening Haemorrhage

The extremely small risk of intracranial haemorrhage persists throughout the period of profound thrombocytopenia and does not diminish with time. The presence of severe headache or neurological signs in any patient with a very low platelet count should be treated as an emergency.

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These children require:

- Discussion with Acute Paediatric Consultant on call and Paediatric Haematologist on call
- Admission to PICU
- Urgent CT head scan
- Platelet transfusion (20 units or more may be required if CNS haemorrhage is confirmed, or there is a strong clinical suspicion of CNS haemorrhage)
- High dose IV methyl prednisolone (30 mg/kg/day)
- I.V. Immunoglobulin
- Simultaneous emergency splenectomy and craniotomy.

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### Indications for Bone Marrow Aspiration

- Clinical suspicions of malignancy
- Abnormal blood film, e.g. neutropenia, anaemia, macrocytosis
- Treatment failure. All these patients require discussion with a paediatric haematologist
- Chronic ITP

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### Follow-up

Children who have been successfully treated should have regular FBC until their platelet count has returned to  $150 \times 10^9/L$  or above. They may be followed up by their GP.

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### References

Blanchette V, Maggs–Bolton P. Childhood ITP: diagnosis and management (Review). Paed Clinics Nth Am 2008;55(2) :393-40

Current options for treatment of ITP Arnold DM. et al Seminars in haematology. 2007; 44(4Suppl 5) S12 S23