**Idiopathic Thrombocytopenic Purpura (ITP)**

## Background

ITP (also known as primary immune thrombocytopenia) is an acquired thrombocytopenia (platelet count $<100x10^9$) due to immune mediated shortened circulating platelet survival in the absence of other disturbances of haemostasis or coagulation. In most childhood ITP platelet autoantibodies are absent. It can occur at any age, but is rare below 2y, and extremely rare <6 months.

Patients fall broadly into two categories:

1. Acute (> 70%): self limiting disease (sometimes preceded by a viral syndrome) with spontaneous resolution within 6 months (usually within 2 months).

2. Chronic (< 30%): does not remit within 6 months.

## Presentation

- Most children present with bruising and petechiae alone. In some instances there is oral bleeding, epistaxis, rectal bleeding or haematuria.

- Some have a history of recent viral infection or vaccination

- Morbidity in ITP is usually minimal.

- The incidence of intracranial haemorrhage is less than 1%.

## Assessment

- The clinical diagnosis of ITP depends on there being manifestations of thrombocytopenia without other abnormal findings, in particular no pallor, lymphadenopathy or hepatosplenomegaly.

- Confirmation rests on the adequate exclusion of other causes of thrombocytopenia. The most important conditions to exclude are acute leukaemia, other marrow infiltrative conditions and aplastic anaemia. An FBC (including blood film) will usually confirm the diagnosis. Check coagulation screen, & send group & save sample if significant bleeding.

- A bone marrow aspirate is an invasive procedure with some morbidity in children who bruise easily, and is only necessary if the diagnosis is uncertain.

- Bleeding is graded from “mild” to “life threatening” (see Bleeding Severity Grade document)

## Management

Initial treatment options include no treatment, supportive treatment (e.g. tranexamic acid,oral contraceptives) and oral steroids. Without active treatment, most patients platelet counts will return to a level at which normal activity can be recommenced within 4-6 weeks.

**a) Conservative Outpatient Management**

Most patients can be managed as outpatients with no specific treatment. The following criteria must be met:

- The diagnosis is unequivocal. No pallor, hepatosplenomegaly or lymphadenopathy; isolated thrombocytopenia without abnormalities in Hb or WCC, or blood film changes (**blood film** may not be available out of hours – **if you take the sample it is <u>your</u> responsibility to ensure that this is chased & appropriate action taken the following day**).

- Bleeding severity "mild" (see bleeding severity grade)

- The child is otherwise well & over 12 months of age

- Social circumstances allow confidence about the degree of parental supervision and relative safety of the home environment, particularly for younger children.

- Parents are given written information (see 'parent info' document)

- You can arrange outpatient follow-up in 2 weeks time supervised by the Children's Emergency Department Consultant.

**During normal working hours:** d/w ED Consultant

**Out of Hours:** child can be discharged if clinically appropriate (see above) but it is <u>your</u> responsibility to ensure that the blood film is chased the following day with any abnormality discussed with the ED Consultant on duty, & to ensure that 2 week OP follow-up is booked.

**b) Conservative Inpatient Management**

a. If the diagnosis of ITP is not certain or any other of the above criteria are not met, then admission & observation is necessary.

c) **Treatment As An Inpatient**

a. Any patient with ITP who has active bleeding apart from resolved epistaxis lasting <20mins (Bleeding severity "moderate"/ "severe") should be admitted, and given oral prednisolone (for dose see "management summary"). Consider tranexamic acid if ongoing mucosal bleeding/menorraghia (not if massive haematuria/concomitant severe renal failure).

b. Any patient with ITP who has Bleeding Severity in the category "severe"/"life-threatening" should be resuscitated as per clinical requirement, admitted to HDU, have

ENT/surgical referral to manage ongoing bleeding & be discussed with the responsible Consultant to consider one (or more) of the following platelet-raising therapies:

(i) oral prednisolone/IV methylprednisolone (time to platelet response 2-7d)

(ii) Normal human immunoglobulin 0.8-1g/kg IV as single dose, to be repeated if necessary. Can be used in combination with steroids. (Time to platelet response 1d; order from pharmacy)

(iii) IV Anti-RhD immunoglobulin IN RhD +ve CHILDREN ONLY (time to platelet response 1-2d; order from pharmacy)

Consider platelet transfusion

Consider tranexamic acid

**Avoid aspirin and non-steroidal anti-inflammatory drugs, and intramuscular injections.**

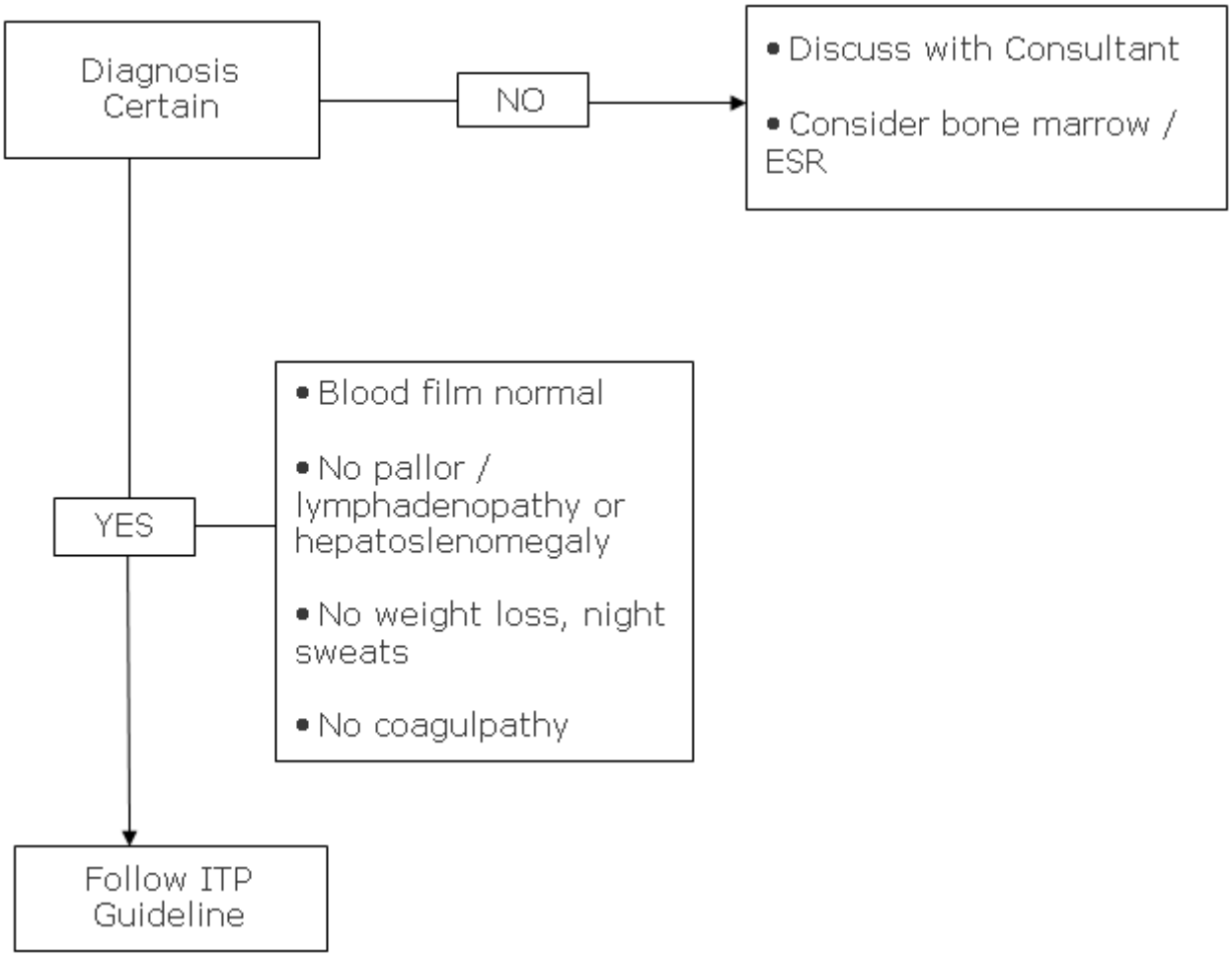
## Acute, relapsing ITP

In some cases, thrombocytopenia will redevelop months or years after the first episode has resolved. These relapses or recurrences are usually precipitated by viral infections. Provided the first episode remitted spontaneously without complication and the patient has been well with a documented normal platelet count between episodes, these cases can be managed as for acute ITP.

## Chronic ITP

Ongoing thrombocytopenia after a 6 month period denotes chronic ITP. A history of bruising from infancy should prompt suspicion of one of the rare congenital thrombocytopenias. Careful inspection of the blood film and tests of platelet function will serve to exclude other diagnoses. Bone marrow examination is helpful in confirming chronic ITP.

Rarely, splenectomy is required (success rate 70-80%).




## REFERENCES

1. Idiopathic Thrombocytopaenic purpura. PHB Bolton-Maggs,

*Arch Dis Child* 2000:83; 220-222

2. Queens Medical Centre, Nottingham guideline ITP 1997

3. Royal Childrens' Hospital, Melbourne ITP guideline 2000

4. Changing Trends in the UK Management of Childhood ITP. John D. Grainger, Joanne L. Rees, Marie Reeves, Paula H. B Bolton-Maggs *Arch. Dis. Child* 2012; 97: 8-11

5. International Consensus Report on the investigation and management of Primary Immune Thrombocytopenia. Proven *et al Blood* 2010; 115:168 -186

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