Acute Immune Thrombocytopenia Purpura (ITP)

Background

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.

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 focussed 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 $<10 \times 10^9/L$
- Significant enlargement of lymph nodes, liver or spleen should prompt consideration of an alternative diagnosis
- 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% resolve spontaneously within three months of onset
- 5-15% becomes persistent (lasting 3-12 months from diagnosis) or chronic (lasting > 12 months) – more common if presenting > 10 years of age
- 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 have menstruating
- Advise to avoid contact sports or activities with a high risk of trauma or head injury
- **All patients should have open access to CAU until their platelet count has recovered**
- All families should be given an information leaflet and the contact details for the ITP Support Association (http://www.itpsupport.org.uk)

Specific treatment:

The decision to treat should be based upon the severity of bleeding symptoms and not on the platelet count or cutaneous signs 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:

- Refer ALL patients via email to Dr Kotecha/Dr Bhuller who will arrange follow-up
- All patients with a platelet count <10 should have a repeat FBC in 1 week (arrange through Paediatric Day Unit)

- All follow up will be with the Paediatric haematology team

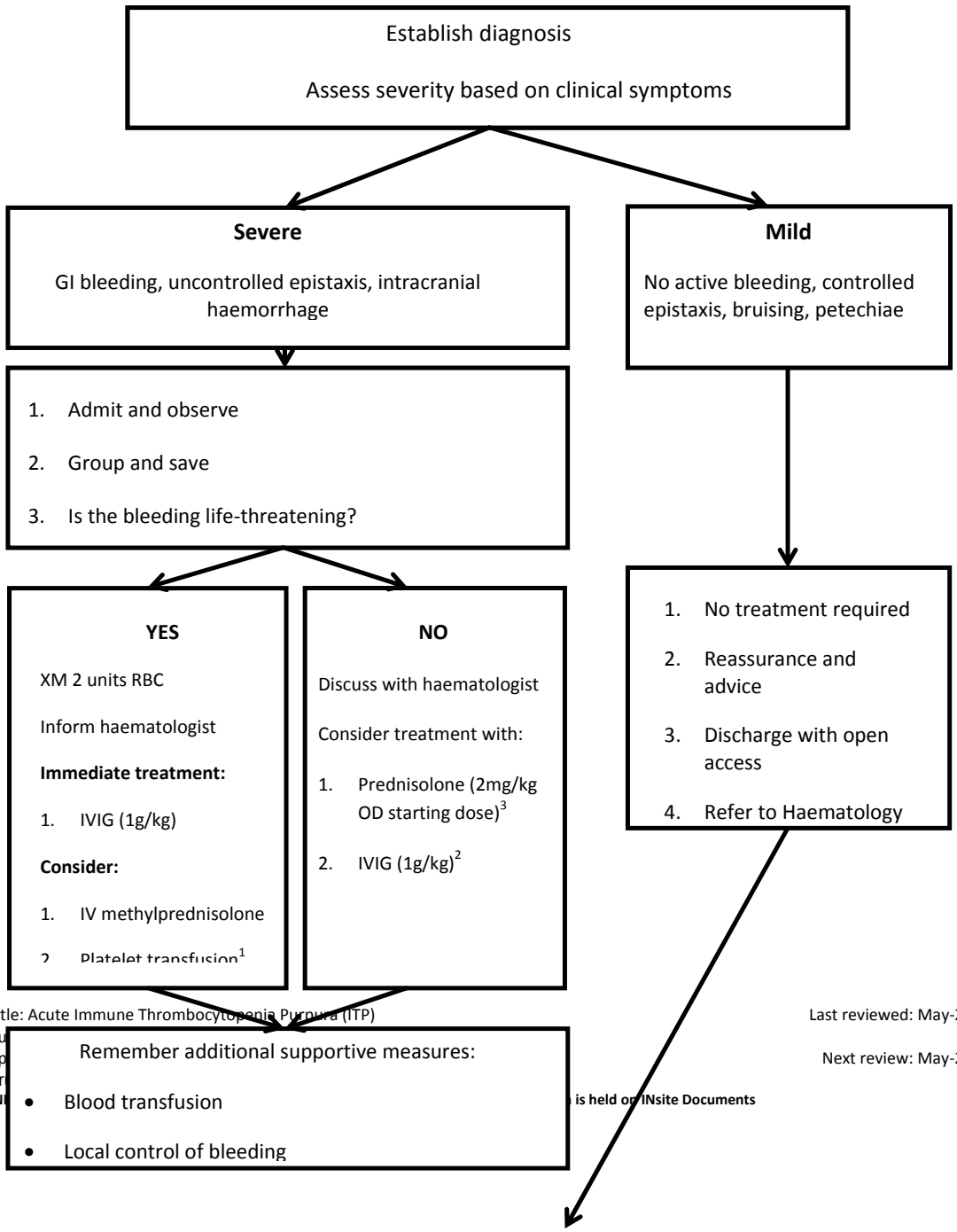
References:

1. Provan D, Stasi R, Newland AC et al. International consensus report on the investigation and management of primary immune thrombocytopenia. Blood 2010; 115:168-186

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

Flow Chart:

Management of acute ITP



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

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