Acute immune thrombocytopenic purpura

To treat or not to treat?

P. Bolton-Maggs
Department of Clinical Haematology, Central Manchester and Manchester Children’s University Hospitals, Manchester Royal Infirmary, UK

Summary
Immune thrombocytopenia in children is usually a self-limiting illness, but in adults the disease is likely to be chronic, and may be associated with other pathology which predisposes to bleeding. Despite very low platelet counts serious bleeding is rare in both adults and children. More than 80% of children have mild clinical manifestations. Intracranial haemorrhage is rare at all ages, is unpredictable and can occur at any time when the count is very low. Currently recommended therapies for both adults and children are associated with significant side effects and occasional deaths. Treatment may interfere with quality of life more than the illness itself. Drugs can be withheld in the majority of children with appropriate advice to child and family. Treatment can be individualised, taking into account the person’s needs and lifestyle as well as bleeding. In chronic ITP many need no active therapy. The situation with adults is more complex but those with a platelet count above $30 \times 10^9/l$ usually need no treatment as bleeding is rare, and these adults with refractory ITP unresponsive to treatment live with very low counts for years without significant bleeding suggesting the need to re-evaluate the balance of risks of treatment versus bleeding. It is notable that adults with ITP may die from infection, probably related to therapy.

Keywords
ITP, thrombocytopenia

ITP management
In many European countries children with ITP have been managed without platelet-enhancing therapy because of the short duration of the illness and rarity of bleeding (4, 19). In the USA normal practice has generally been to treat the count so that the ASH guideline (11) indicated that all children with ITP many need no active therapy. The situation with adults is more complex but those with a platelet count above $30 \times 10^9/l$ usually need no treatment as bleeding is rare, and these adults with refractory ITP unresponsive to treatment live with very low counts for years without significant bleeding suggesting the need to re-evaluate the balance of risks of treatment versus bleeding. It is notable that adults with ITP may die from infection, probably related to therapy.

Minimum treatment
If treatment is required, either for more troublesome bleeding or for quality of life issues (activities, anxiety) then the minimum treatment should be given. Recent studies suggest that very short courses of high dose prednisolone (4 mg/kg/d for 4 days) are effective without running the risk of serious side effects (7). It is notable that in children with severe bleeding, the response to therapy was less effective than expected in raising the count, although bleeding ceased (16). Emergency treatment of serious bleeding is usually with IVIG together with steroids and platelet transfusions which may be more effective in the context of immune suppression.

It has been customary to treat all adults with ITP because of the impression that the risk of bleeding is higher. However, many adults are minimally symptomatic, similar to children, even with very low counts. A recent analysis of 152 adults with chronic ITP studied over a 10 year period demonstrated that 85% did not require treatment as the counts settled out at an acceptable level ($>30 \times 10^9/l$). The mortality in this group...
was no different to the normal population. Among those with lower counts (9%) the mortality risk was increased but as many died from infection (probably related to therapy) as died from bleeding (17). There is a strong case for tailoring the treatment to the individual patient, avoiding the use of toxic medication in those with counts above $30 \times 10^9/l$. Analysis of patient views indicates that steroids are very unpopular because of the side effects (20). The selection of alternative agents is not straightforward as each works for some but not other patients, and it is very useful to discuss the side effect profile as well as potential benefits with individual patients.

**Splenectomy**

Splenectomy may be indicated for those whose count is persistently below $30 \times 10^9/l$ and who have bleeding symptoms, but accepting that about 25% will relapse. The place of splenectomy has been questioned in the light of success with newer agents such as rituximab (8, 9), and thrombopoietic agents (6, 14) which may provide long term treatment with minimal side effects.

**References**


Correspondence to:
Poula Bolton-Maggs, DIA, FRCP, FRCPath
Department of Clinical Haematology, Central Manchester and Manchester Children’s University Hospitals
Manchester Royal Infirmary, Oxford Road
Manchester M13 9WL, England
Tel. +44(0)161/276 48 11.
E-Mail: paula.bolton-maggs@manchester.ac.uk