QUESTION 36 ITP
A 30-year-old man has previously undergone splenectomy (six months prior) for chronic idiopathic thrombocytopenic purpura (ITP). Apart from occasional bruising, there are no haemorrhagic symptoms.

His full blood examination shows:

- haemoglobin: 135 g/L [128-175]
- mean cell volume (MCV): 96 fL [80-97]
- white cell count: 11.7 x 10^9/L [3.9-12.7]
- platelets: 44 x 10^9/L [150-396]

Repeted autoimmune and viral serology remain negative.
The most appropriate management of his thrombocytopenia is:

A. danazol.
B. immunoglobulin.
C. prednis(ol)one.
D. azathioprine.
E. observation.

1. Chronic Idiopathic thrombocytic purpura

**Etiology**
- Women age 20 – 40 commonly ratio 3:1
- Autoimmune disorder: Antibodies directed against target antigens on glycoprotein IIb/IIIa or less frequently Gp Ib/IX
- Low plt count may be the initial manifestation of SLE

**Ix**
- Normal Hb and WCC
- Plt < 20 x 10^9/L
- Bone marrow examination: normal or increased numbers of megakaryocytes
- PT and APTT are normal, fibrinogen is normal

ITP occurs in association with some malignancies (e.g. CLL, non HL, myelodysplasia), infections (EBV, HIV and malaria), connective tissue disorder (SLE).

Patients should be tested for ANF and anticardiolipin antibodies

**Treatment**
- Prednisolone
- IV immunoglobulin
  - obtaining a temporary rise in plt count
  - meningismus and headache, hepatitis C virus
- Splenectomy
  - For non responders with continuing symptoms and/or very low platelet counts
  - 2nd line after prednisolone
- Additional immunosuppressive therapy
  1. Azothioprine,
  2. Cyclophosphamide
  3. Cyclosporin A, Rhesus anti-D – cause hemolysis
  4. Rituximab (anti CD 20) * most commonly used*
     - preferable to long term steroids
     - eliminates normal B cells including those producing antiplatelet antibody
     - B cell depletion is transient
     - Few S.E or toxicities

1 Haematology at a glance
5.  ² Danazol
   - Attenuated androgen
   - Limited use, efficacy unknown

³ Persistant thrombocytopenia after splenectomy

Immunosuppressive drugs as above – should only be used to raise plt count temporarily and to support patients before surgery, labor and delivery
Not substitutes for splenectomy

If patient not bleeding and maintains plt count > 20,000/microL, withhold therapy

Answer D Observation
Plt count is acceptable at 44 X 10^9/L. Severe spontaneous bleeding were seen below platelet levels 10,000/microL = 10 X 10^9/L

² Up to date : Treatment and prognosis of ITP in adults
³ Harrisons – pg 675 ITP