1A 2-year-old boy has been unwell with repeated chest infections since the age of six months. There is a family history of severe and recurrent infections in male relatives and a male cousin had died in infancy from an unknown cause. On investigation, the patient was found to be agammaglobulinaemic and B cells were absent. X-linked agammaglobulinaemia was confirmed on genetic studies. Intravenous immunoglobulin (IVIG) replacement therapy was commenced. Which of the following are constituents of IVIG and which are not?

(a) Blood products pooled from 100–500 donor
(b) IgD
(c) Stabilising agents such as sucrose
(d) Cytokines
(e) Replacement levels of IgA

2A 31-year-old woman recovering from a recent diarrhoeal illness presented with ascending symmetrical weakness which had started to affect her trunk. Cerebrospinal fluid analysis demonstrated raised protein with normal cell numbers and an EMG was consistent with Guillain–Barré syndrome. Her condition deteriorated, with worsening respiratory function and the development of autonomic dysfunction. High-dose IVIG therapy was commenced. Which of the following are true mechanisms of action of IVIG and which false?

(a) Neutralisation of a wide range of pathogens by IgM
(b) Effects on cell adhesion
(c) Effects on IgG catabolism and recycling via FcεR1
(d) Interference with antibody-dependent cellular cytotoxicity
(e) Inhibition of deposition of activated complement

3A 40-year-old man presented with an eight-year history of recurrent chest infections. He was a non-smoker. Severe bronchiectasis was demonstrated on high resolution computed tomography scan of the chest. On further investigation, he was found to have severe panhypogammaglobulinaemia. After exclusion of other causes, a diagnosis of common variable immunodeficiency was made and treatment with IVIG was discussed with the patient. Which of the following statements regarding the use of IVIG are true and which false?
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(a) Anaphylactic reactions are rare.
(b) There are documented cases of hepatitis C transmission.
(c) There are documented cases of prion disease transmission.
(d) When replacing Ig in primary antibody deficiency, the cheapest currently available IVIG preparation should be used.
(e) In primary antibody deficiency, it is best to rotate IVIG products regularly to give the broadest range of antibody binding specificities.

4 A 62-year-old man complained of several weeks of tiredness and muscle aches and three days of blurred vision. On suspicion of temporal arteritis, the erythrocyte sedimentation rate was checked and found to be markedly abnormal at 120 mm/1st hour (normal <25 mm/1st hour). Prednisolone was started at a dose of 1 mg/kg. On review in the ophthalmology clinic two days later, dilated, tortuous veins were seen and hyperviscosity syndrome suspected. Further blood tests showed a plasma viscosity of 2.8 mPa/s (normal <1.72 mPa/s) and an IgM monoclonal serum protein at a concentration of 29 g/l. Waldenstrom’s macroglobulinaemia was diagnosed; treatment with urgent plasmapheresis and cyclophosphamide was started. Which of the following statements are true and which false?
(a) Monoclonal IgG paraproteins lead to a higher plasma viscosity than equivalent IgM paraprotein concentrations.
(b) Plasmapheresis, removing the equivalent of one plasma volume (3 litre), will reduce the IgM paraprotein level to approximately 50% of the pretreatment level.
(c) Removal of one plasma volume (3 litre) from a 70 kg patient at the rate of 50 ml/kg/hour will normally take 8 hours.
(d) IgM will be completely removed in three daily procedures, removing one plasma volume (3 litre) each time.
(e) The cellular components of blood, such as erythrocytes and leukocytes, do not significantly affect whole blood viscosity.

5 A 68-year-old woman with a three-year history of a distal sensory neuropathy enquired about plasmapheresis treatment at her routine neurology clinic review appointment. She had read about plasmapheresis on the internet. Previous electrophysiological studies had been consistent with a demyelinating neuropathy. Repeat blood tests were performed. The full blood count, blood film and biochemistry screen were normal. The only abnormality was a low level of an IgM paraprotein in the serum. Bone marrow examination was judged unnecessary by a haematologist. Which of the following statements are true and which false?
(a) The IgM paraprotein could be an incidental finding, unrelated to the neuropathy.
(b) Detectable anti-myelin associated glycoprotein (MAG) would be an important finding in this patient.
(c) Anti-MAG is more likely to be IgM isotype than IgG or IgA.
(d) Anti-MAG antibody is typically associated with distal motor neuropathies.
(e) Plasmapheresis is not useful in neuropathies associated with IgA or IgG paraproteins.

6 A 35-year-old woman presented with recent onset of aggressive rheumatoid arthritis. She had obvious bilateral synovitis affecting the metacarpo-phalangeal, proximal interphalangeal and metatarso-phalangeal joints. She was found to have a high titre of rheumatoid factor in the serum and evidence of erosive disease on imaging of the hands and feet. Which of the following treatment options are supported by a product licence for first-line therapy in these circumstances and which are not?
(a) Rituximab
(b) Adalimumab with methotrexate
(c) Etanercept
(d) Infliximab monotherapy
(e) Abatacept

7 A 38-year-old man with a history of Crohn’s disease presented as an emergency to the gastroenterology outpatient clinic with a painful perianal lesion that had been discharging for a week. His maintenance immunosuppression was azathioprine (AZT) 100 mg per day; he had also been taking oral prednisolone for five days. On examination, he was found to have a perianal fistula. Which of the following statements are true and which false?
(a) Antibiotics and drainage are appropriate first-line treatments.
(b) Infliximab should be considered if he fails to respond to a full course of conventional treatment.
(c) Etanercept is indicated.
(d) National Institute for Health and Clinical Excellence guidance is available to help in the treatment decision.
(e) Natalizumab would be the biologic of choice.

8 A 72-year-old woman who lived in a remote area and was a keen gardener was stung by a wasp on the foot. Within 5 min she developed itching and was described as feeling faint. Her husband called her general practitioner (GP) who noted generalised urticaria, swollen eyelids, hoarseness of the voice and a wheezy chest. Her pulse was 100 b/min and blood pressure was 110/70 mmHg. The GP administered intramuscular antihistamine and hydrocortisone. There was a gradual recovery over 30 min, with resolution of rash and respiratory symptoms. There was a medical history of hypertension controlled by a calcium channel blocker, but no other significant personal or family history. Which of the following statements are true and which false?
(a) The administration of antihistamine and hydrocortisone under the supervision of the GP was the correct management.
(b) Advice to avoid a further sting and prescription of a self-injectable adrenaline device is a reasonable long-term solution for this woman.

(c) Wasp venom immunotherapy is indicated in patients with large local reactions following a sting in view of the risk of anaphylaxis following a further sting.

(d) Wasp venom immunotherapy prevents anaphylaxis in more than 90% of cases on re-sting.

(e) Venom immunotherapy may afford long-term protection in anaphylaxis caused by stings.

9 A 35-year-old man with a renal transplant attended the emergency department complaining of a fever and shortness of breath. He said his medication had been changed recently. He knew it included Neoral but was unsure of the dose. Examination revealed signs of a left basal pneumonia. Investigations showed serum potassium 5.4 mmol/l, serum creatinine 94 µmol/l and alanine aminotransferase 82 u/l. Which of the following statements are true and which false?

(a) The pneumonia is unlikely to be associated with his immunosuppression
(b) The hyperkalaemia is likely to be related to his immunosuppression
(c) Changes in ciclosporin brand may be important
(d) Measurement of his serum ciclosporin level will indicate whether or not his renal transplant is rejecting
(e) The pneumonia is probably related to hypogammaglobulinaemia secondary to ciclosporin

10 A 47-year-old woman was advised to start AZT treatment for her systemic lupus erythematosus. She was concerned about taking an immunosuppressive drug. Which of the following statements concerning AZT are true and which are false?

(a) AZT may make her Ig levels fall, leaving her susceptible to infection
(b) Measuring thiopurine methyl transferase enzyme activity may be useful in predicting the likelihood of AZT toxicity
(c) Monitoring of blood counts is not required for this drug
(d) Corticosteroids should not be used during AZT therapy
(e) Changes to her hair may occur during AZT treatment