Idiopathic thrombocytopenic purpura

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ITP , also known as immune and autoimmune thrombocy - topenic purpura, results from antibodies to specific platelet membrane glycoproteins (antiplatelet IgG autoantibodies) that result in isolated thrombocytopenia in the presence of - normal bone marrow and the absence of other causes of thrombocytopenia. Two distinct clinical types are evident: an Acute ITP often follows an acute infection and usually resolves spontaneously . Chronic ITP persists longer than 6 months without a specific cause being identified. Clinical features The adult form normally a ff ects females between the ages of 15 and 50 years, although it can be associated with other conditions, including systemic lupus erythematosus, chronic lymphatic leukaemia and Hodgkin’s disease. The childhood form is distributed equally between males and females and commonly presents before the age of 5 years. Purpuric patches (ecchymoses) occur on the skin and mucous membranes. Following trauma or pressure, examina tion often reveals number s of petechial haemorrhages in the skin. There is a tendency to spontaneous bleeding from mucous membranes (e.g. epistaxis); menorrhagia in women and pro longed bleeding of minor wounds are common. Haemorrhage from the urinary and gastrointestinal tracts and haemarthrosis are rare. Although intracranial haemorrhage is also uncom mon, it is the most frequent cause of death. The diagnosis is indicated by the pr esence of cutaneous ecchymoses and a pos itive tourniquet test. The spleen is palpable in fewer than 10% of patients. The pr esence of gross splenic enlargement should raise suspicion of an alternative diagnosis. Investigations Coagulation studies are normal and a bleeding time is not helpful in diagnosis. Platelet count in the peripheral blood film 9 is reduced (usually <60 /uni00A0 ×/uni00A0 10 /L). Bone marrow aspiration reveals a plentiful supply of platelet-producing megakaryo cytes. Treatment The course of the disease di ff ers in children and adults. The disease usually regresses spontaneously in paediatric cases. Short courses of corticosteroids in both adults and children are usually followed by recovery . Steroid therapy should not be prolonged. Splenectomy is usually recommended for refractory or relapsing ITP . Up to two-thirds of patients will be cured by surgical intervention and 15% will be improved, but no ben efit will be derived in the r emainder. The response to steroids predicts a good response to splenectomy if the disease relapses. Laparoscopic splenectomy is rapidly becoming the main stay of treatment since the size of the spleen is usually normal or slightly enlar ged and the spleen is not friable. It is important to identify and remove splenunculi as they have the potential to enlarge with time and lead to recurrence of symptoms. In the acute setting, fresh blood transfusion or transfusion with platelet concentrates before operation may be necessary , although these are generally withheld until the splenic vessels have been controlled.

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