Systemic Lupus Erythematosus

Version of 2016

4. APPENDIX 1. Antiphospholipid antibodies

Antiphospholipid antibodies are autoantibodies made against a body's own phospholipids (part of a cell's membrane) or proteins that bind to phospholipids. The three best known antiphospholipid antibodies are anticardiolipin antibodies, antibodies against β2 glycoprotein I and lupus anticoagulants. Antiphospholipid antibodies can be found in 50% of children with SLE, but they are also seen in some other autoimmune diseases, various infections, as well as in a small percentage of children without any known illness.

These antibodies increase clotting tendency in blood vessels and have been associated with a number of illnesses, including thrombosis of the arteries and/or veins, abnormally low blood platelet counts (thrombocytopenia), migraine headaches, epilepsy and purplish mottled discolouration of the skin (livedo reticularis). A common site of clotting is the brain, which can lead to a stroke. Other common sites of clots include the leg veins and kidneys. Antiphospholipid syndrome is the name given to a disease when thrombosis has occurred along with a positive antiphospholipid antibody test.

Antiphospholipid antibodies are especially important in pregnant women, because they interfere with the function of the placenta. Blood clots that develop in the placental vessels can cause premature miscarriage (spontaneous abortion), poor foetal growth, preeclampsia (high blood pressure during pregnancy) and stillbirth. Some women with antiphospholipid antibodies may also have trouble getting pregnant.

Most children with positive antiphospholipid antibody tests have never had a thrombosis. Research into the best preventive treatment for such children is currently being carried out. At present, children with positive antiphospholipid antibodies and underlying autoimmune disease are

often given low dose aspirin. Aspirin acts on platelets to reduce their stickiness, and hence reduces the ability of the blood to clot. Optimal management of adolescents with antiphospholipid antibodies also includes the avoidance of risk factors such as smoking and oral contraception.

When the diagnosis of antiphospholipid syndrome is established (in children after thrombosis), the primary treatment is to thin the blood. Thinning is usually achieved with a tablet called warfarin, an anticoagulant. The drug is taken daily and regular blood tests are required to ensure that the warfarin is thinning the blood to the required degree. There is also heparin injected under the skin and aspirin. The length of anticoagulation therapy is highly dependent on the severity of the disorder and the type of blood clotting.

Women with antiphospholipid antibodies who have recurrent miscarriages can also be treated, but not with warfarin as it has the potential to cause foetal abnormalities if given during pregnancy. Aspirin and heparin are used to treat pregnant women with antiphospholipid antibodies. During pregnancy, heparin needs to be given daily by injection under the skin. With the use of such medications and careful supervision by obstetricians, about 80% of women will have successful pregnancies.