



No. 28 – The Relationship between ITP and

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Title: **The Relationship between ITP and lupus**

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Most people with immune or idiopathic thrombocytopenic purpura (ITP) are otherwise healthy. They may have other medical conditions unrelated to ITP, but the antibody against their platelets that causes ITP generally attacks only the platelets and not other parts of the body. However, there are exceptions to this rule (aren't there always?!).

First, some people with ITP (more so in adults than children) have other symptoms and problems (for instance joint pains, low grade fever, and malaise) that suggest an immune-mediated condition. Some of these patients have demonstrable antibodies against other components of their blood or other tissues. For instance, a small percentage of persons with ITP have antibodies attached to their red blood cells. This results in a positive "Coombs test" and may result in anemia, which is called Evans Syndrome. The abnormally regulated immune system in ITP (remember that we really do not know what causes the immune imbalance in ITP) can also lead to production of autoantibodies against the thyroid gland, stomach cells, and, in fact, the nucleus in all cells in the body. The nucleus is the part of every cell (except red blood cells) that carries the DNA, the genetic code. These latter antibodies are called anti-nuclear antibodies (ANA). A person with them is described as having a positive "ANA test". A positive ANA test is common in persons with ITP, occurring in up to 20-30% of adults and children with chronic ITP. In general having a positive ANA shouldn't cause concern even though a positive ANA is also seen in virtually everyone with lupus. Lupus (also called systemic lupus erythematosus or SLE) is a serious autoimmune disease which sometimes begins with just a low platelet count that may seem to be ITP. Patients with SLE often have arthritis, skin rash, kidney problems, and may also have complications involving the brain, heart, and other organs. These problems are due to multiple autoantibodies that cause damage. Most patients with lupus also have reduced blood counts, including platelets.

Because of the apparent overlap of ITP with lupus, it is important to distinguish the two conditions. Fortunately, there are several quite specific tests for lupus. They should always be done in persons with apparent ITP who either have a positive ANA test or other problems (with the joints, skin, kidneys, etc.) that suggest the possibility of lupus. On rare occasions what appears to be "regular" ITP occurring in otherwise well patients evolves into lupus years later. However, this is very uncommon, and the vast majority of children and adults with ITP, even those with a positive ANA test, never develop any signs of lupus or other serious autoimmune diseases.

Another type of antibody that persons with ITP occasionally develop is an anti-phospholipid antibody. Phospholipids are the lipid (or fat) molecules that form the structure of the membranes surrounding all of the cells of our body. Although these antibodies can be harmless, they may cause excessive blood clotting (so called thrombosis), which can occasionally be serious. When such anti-phospholipid antibodies are also encountered in lupus, they are often responsible for thrombosis. However, persons with ITP who have anti-phospholipid antibodies infrequently have either lupus or problems with thrombosis.

The "bottom line" is that ITP is one condition in a large spectrum of poorly understood immunologic or autoimmune disorders where a person makes an antibody against part of themselves. When it is limited to the platelets, we consider it to be ITP. When other cells or tissues are affected by autoantibodies (for example, antinuclear antibodies or anti-phospholipid antibodies) then specific testing should be undertaken for lupus or similar conditions. Whether a person has "regular" ITP or instead low platelets due to lupus its treatment is similar. Steroids, IVIG, and splenectomy can all be helpful in raising the platelet count. Of course many patients with lupus and thrombocytopenia, just like persons with ITP and thrombocytopenia, do not need any treatment at all and can lead a quite normal life.