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Title: What happens to adults with ITP

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In previous American Perspective essays, we discussed the outcomes of children who develop ITP. We examined the debate about whether or not children require specific drug treatment, for we know that most children will recover with normal platelet counts in several weeks or several months without it. Only about 25% of children will still have a low platelet count after 6 months (so are arbitrarily defined as having chronic ITP), but many, if not most of the children who have what is called chronic ITP will also eventually recover on their own.

We consider that ITP is a more chronic, persistent disorder in adults. But what does that mean? Does that mean that platelet counts will be low forever? Does that mean that drug treatment will be needed forever? Does that mean that there will be a risk for excessive bleeding forever? Since ITP is an autoimmune disorder, does this mean that other autoimmune conditions will eventually also occur? The answers to these questions have never been clear, because most published studies on adults with ITP have not followed large number of patients for many years. Or they have only followed the patients with problems, and we don't know what has happened to all of the rest of patients who did not have problems. To get a clear picture of the future for patients with ITP, it is important to assess and follow all patients who were diagnosed within a particular region or country on a long-term basis. If only selected patients with problems are monitored and described, we get a biased view, and we may think that ITP is worse than it actually is. We see a hint of this bias in our publication, *The Platelet*, when patients write in with their experiences. Patients describe their problems and therefore problems of ITP are what we read about. Patients who have no problems more likely have no story to tell. They have "forgotten" about ITP and gone on with their lives.

Two recently published studies give us a clearer picture of adults with ITP. From these studies, we not only learn what happens over many years, we also appreciate how the picture of ITP has changed over the past 30 years. The first study, by Johanna Portielje and her colleagues from Rotterdam described 152 consecutive patients who were diagnosed between 1974 and 1994 and carefully followed for an average of 10 years. The second study is by Annette Neylon and her colleagues from Newcastle in Staffordshire. They followed 245 adults with ITP diagnosed between 1993 and 1999 for an average of 5 years. Understanding their data can help us predict the future for patients with ITP.

The most important conclusion of these studies is that very few patients died from bleeding, and most of those who did had health problems in addition to ITP that could have made their bleeding worse. Only 2 of 152 patients from Rotterdam and only 3 of 245 patients from York died from bleeding. Of note, all 3 of the patients from York had other problems: one was a young man with severe hemolytic anemia (another autoimmune condition), another had lymphoma (lymph gland cancer), and the third was taking a blood thinner, warfarin, for thrombosis (blood clots). Further details are not available about the two patients from Rotterdam who died from bleeding. So the mortality from bleeding may be about 1% (5 patients among 497), but death from bleeding most commonly may occurs in patients with other serious problems that increase their risk.

What may be even more important is that in these studies, 5 additional patients died from complications of treatment: two from complications of splenectomy and 3 from serious infections related to the suppression of their immune system by steroids and other immunosuppressive drugs. These observations make a strong argument for conservative treatment of patients with ITP. A comparison of these 2 studies suggests that there is a trend for more conservative management of patients with ITP. Among the 152 patients from Rotterdam, about half (78 patients) had a splenectomy. In the more recent patients from York, only 30 (12%) of the 245 patients had a splenectomy.

Because platelet destruction in ITP is caused by autoantibodies, and since other serious disorders, such as systemic lupus erythematosus (SLE) are also caused by autoantibodies, patients with ITP are often concerned that a more serious autoimmune disorder, such as SLE, will eventually develop. Autoantibodies occur when the immune system gets confused and recognizes a cell or tissue of a person's own body as "foreign" and tries to destroy it. In ITP, the cell recognized as "foreign" is the platelet. In other disorders the targets of autoantibodies are cells other than platelets. For example, in autoimmune hemolytic anemia, autoantibodies cause red blood cell destruction. In thyroid diseases, such as hyperthyroidism (Graves' disease) or hypothyroidism, autoantibodies recognize thyroid tissue as "foreign". In SLE the autoantibodies may be directed against multiple tissues, such the kidney, lung, heart, joints, and skin. The good news is patients with ITP rarely develop other autoimmune disorders. For example, in these two studies from Rotterdam and York, only 9 (2%) of 428 patients developed lupus and these occurred many years after they had been diagnosed with ITP.

Therefore, although ITP in adults may persist for many years, almost all patients do well. That's good news!