The ‘T’ in ITP remains

The “I” has changed its meaning, the “P” is not necessary to make the diagnosis, and the syndrome is not strikingly common in adults. But the disease formerly known as idiopathic thrombocytopenic purpura (ITP) remains important for internists to diagnose because thrombocytopenia is extremely common.

ITP has long been recognized in children (in whom it is often self-limited but severe) and in adults (in whom it is often more insidious and chronic, with a wider differential diagnosis).

The “I” used to stand for “idiopathic” but now it stands for “immune,” following a half decade of work by many investigators. The seminal experimental work of Dr. William J. Harrington and others while the former was a hematology fellow with Carl Moore at Washington University is the stuff of legend and ethical debate. Harrington had himself injected with a pint of serum from a patient with ITP and nearly died, demonstrating that the patient’s blood contained a substance or substances capable of inducing reversible profound thrombocytopenia in the recipient.1 Today, this classic experiment would probably not be performed, nor would the many more infusions that Harrington subsequently gave himself, other physicians, and support staff.2

Understanding the immunologic basis for ITP has led to treatments that are usually but not uniformly successful. Prednisone remains the main initial therapy, but its myriad side effects have led to the strategy of turning sooner to other therapies, such as intravenous immunoglobulin, splenectomy, rituximab (Rituxan), and, most recently, thrombopoietin agonists, in order to control the disease or even put it into remission.

Treatment decisions are often assigned to the hematologist or rheumatologist, but recognizing ITP and distinguishing it from other causes of thrombocytopenia remain the province of primary care providers, as discussed by Thota et al on page 641 in this issue of the Journal.3 The diagnosis of ITP does not require the presence of purpura, which most adults ITP patients probably do not have, nor does it always require a bone marrow biopsy. It is important that ITP be distinguished from thrombocytopenia that is induced by drugs (particularly heparin) and myelodysplastic and other marrow processes (potential clues being other cytopenias, an unexplained elevated mean corpuscular volume, or constitutional symptoms). Undiagnosed thyroid disease and HIV infection should be tested for routinely, once drug-associated and other obvious causes are excluded.

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REFERENCES

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