I. Herbert Scheinberg, Expert on Wilson Disease, Dies at 89

By WILLIAM GRIMES

I. Herbert Scheinberg, a specialist in rare hereditary diseases who helped develop a diagnostic test and treatment for Wilson disease, a potentially fatal buildup of copper in the body, died April 4 in Elizabethtown, N.Y. He was 89 and lived in Lewis, N.Y.

The cause was pneumonia, said his son, David.

Dr. Scheinberg devoted most of his career to studying, treating and raising awareness of Wilson disease, which affects about one in 40,000 people. It is a toxic accumulation of dietary copper in the liver, brain and other organs, leading to progressive neurological and behavioral problems and, if untreated, death. It is named after Dr. Samuel A. K. Wilson, who first described it in 1912.

Often, evidence of Wilson disease does not appear until adolescence or young adulthood. The symptoms, which include erratic or antisocial behavior, slurred speech and tremors, can be misinterpreted as schizophrenia or Parkinson’s disease. Because the disease is so rare, and because its symptoms resemble those of other diseases, it was misdiagnosed more often than not.

With Dr. David Gitlin at Harvard Medical School, Dr. Scheinberg developed a simple, inexpensive blood test in the early 1950s to detect abnormally low levels of ceruloplasmin, a protein that carries copper from the body. This red flag, when confirmed by tests for copper in the liver, made it possible to identify patients with Wilson disease in its early stages.

A modified version of the blood test is still used, in combination with a urine test and an eye examination to detect the distinctive golden-brown ring that forms around the corneas of those who have Wilson disease.

Israel Herbert Scheinberg was born in Manhattan and attended DeWitt Clinton High School in the Bronx. He graduated from Harvard in 1940 with a degree in chemistry and from Harvard Medical School in 1943, then interned at Peter Bent Brigham Hospital in Boston and served two years with the Army Medical Corps. He returned to Harvard as a junior fellow.

In 1957 he married Denise Mangravite, who survives him, along with his son, David, of Northville, Mich.; his daughters, Anne, of the Hague, and Cynthia, of Berkeley, Calif.; and four grandchildren.

Dr. Scheinberg became a professor of medicine at Albert Einstein College of Medicine on its founding in
1955 and from 1973 to 1992 led its division of genetic medicine. After developing the blood test for Wilson disease, he worked with Dr. Irmin Sternlieb and Dr. John M. Walshe of Cambridge University in England to treat the disease by using the oral drug D-penicillamine (or, later, trientine hydrochloride) to help the body excrete excess copper through the urine.

This approach is still used. After being purged of excess copper, patients keep the disease in check by taking low doses of the copper-leaching drugs, along with zinc to block absorption of copper, and by avoiding foods rich in copper, like organ meats, shellfish, nuts and chocolate.

In his later years, Dr. Scheinberg turned his attention to another genetic disorder, Menkes disease, which can be seen as a negative counterpart to Wilson disease, since those who have it cannot retain the small amounts of copper necessary for health.
Dr. Scheinberg around 1970.