

Doctors discover first effective treatment for sickle cell anemia

BETHESDA, Md. (AP) — A cancer drug has proved to be the first effective treatment for sickle cell anemia, a disabling blood disorder affecting 72,000 black Americans.

The drug hydroxyurea reduced the excruciating attacks of sickle cell so dramatically that the National Institutes of Health ended drug trials four months early, and on Monday notified 5,000 doctors of the treatment.

"Patients must understand hydroxyurea is a treatment, not a cure," cautioned Dr. Samuel Charache of Johns Hopkins University, who led the study funded by the National Heart, Lung and Blood Institute. And the drug, which carries some risks of its own, is not for all sickle cell sufferers.

Still, "it's very exciting," said Ralph Sutton of the Sickle Cell Disease Association of America. "This means a significant improvement in the quality of life for people with sickle cell disease."

Sickle cell anemia, an inherited disease common among people with

ancestors from Africa, the Middle East and the Mediterranean, causes hemoglobin inside red blood cells to clump together. That changes the normally round cells into a sickle shape that can't squeeze through tiny blood vessels.

Patients, whose tissue doesn't get adequate blood, suffer pain so severe it frequently requires long hospitalization. The disease eventually causes organ damage, and patients frequently live only into their 40s.

About 8 percent of black Americans carry the gene. There is no cure, only treatment for pain, and until now, no way to prevent symptoms.

In the study released Monday, doctors tested 299 adults with severe sickle cell beginning in 1992. Half took hydroxyurea every day, the others a placebo. Hydroxyurea reduced by 50 percent patients' pain episodes, hospitalizations, need for blood transfusions and cases of a life-threatening complication called acute chest syndrome.

Ruthie Abney, 40, of Washington

once endured weeklong hospitalizations at least eight times a year. Since beginning the drug in 1992, she has suffered only six pain crises, none severe enough to require hospitalization. "I can't begin to relate to people what it means to be pain-free and live a normal life like anyone else," she said Monday.

Hydroxyurea appears to work by stimulating the body to produce a type of hemoglobin found in fetuses, a kind that resists sickle cells' clumping. "It's like the chaperone at a dance, keeps the molecules from getting too close together," Charache explained.

After birth, fetal hemoglobin's gene becomes dormant and an adult type susceptible to sickle cells' stickiness forms. Monday's study found the cells of hydroxyurea takers contained 20 percent fetal hemoglobin — enough to battle the disease.

Hydroxyurea is already on the market as a cancer drug, and doctors can legally prescribe any drug for any purpose.

Nuclear accidents, radioactive pollution plagues Russian village

MUSLIUMOVO, Russia (AP) — The shallow creek runs beneath an abandoned mill. Cows wander knee-deep in the water. In the summer, it is where the village's children swim.

This pastoral scene is deceptive, however. The Techa River is radioactive and has been for almost half a century.

The nearby Mayak nuclear complex, also known as Chelaybinsk-65, began dumping raw nuclear waste into the Ural Mountains river in 1949, when it built the Soviet Union's first reactor to produce plutonium for atomic bombs.

By the mid-1950s, radiation at the top-secret plant affected 124,000 people living along the Techa, which flows through a pretty forest and lake region.

About 20 villages around Musliumovo, with their 8,000 to 9,000 residents, were evacuated because radiation levels were considered too dangerous.

Musliumovo was not, even though radiation in the village often exceeded that at the evacuated sites. Many villagers suspect they were left behind as human guinea pigs.

"For 40 years, they've been checking how a living being can survive in a radiation zone,"

said Valentina Kaidaneyeva, a teacher. "A lot of professors studying us must be dead by now, but we are still alive," she told a visiting group of foreign scientists, politicians and reporters this fall.

Officials are at a loss to explain why Musliumovo, 930 miles east of Moscow, was not relocated.

"I don't think it was done on purpose, but probably because the village was too big and too expensive to evacuate," said Mira Kosenko, an expert on radiation medicine from Chelyabinsk, the regional capital.

Whatever the case, thousands of people remained in Musliumovo, using the river water for their households and letting cattle graze in contaminated fields, unaware of the poison creeping into their bones.

The former Soviet Union zealously guarded its nuclear secrets, and public health hardly mattered. So the villagers were not told anything about strontium-90 and cesium-137. Instead, they were told to keep out of the river because it was dirty.

For them, the Techa was a source of life. If they fell sick, medical personnel were under orders to keep silent about radiation, Kosenko said.

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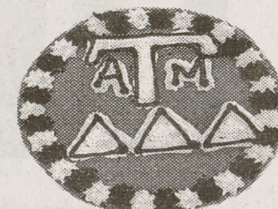
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