CONTINUED FROM P. 12

to muscle weakness and wasting in the hands, feet, and lower legs, which is why patients are often confined to a wheelchair within 10 to 20 years of the appearance of symptoms. Typically, symptoms begin in patients ages 5 to 15. Difficulty with walking is usually the first symptom to appear. As the disease progresses, patients may experience a gradual loss of sensation in the extremities; rapid, involuntary eye movements; slow, slurred speech; spinal curvature (scoliosis); and heart disease and heart failure.

The rate of progression and the life expectancy varies from person to person. Most people with Friedreich's ataxia die in early adulthood if they have significant heart disease (the most common cause of death in these patients). Some people with less severe symptoms of Friedreich's ataxia live into their sixties or seventies.

In order to diagnose Friedreich's ataxia, neurologists perform a careful clinical examination. They may also perform tests such as an electromyogram (EMG), which measures the electrical activity of muscle cells; electrocardiogram (EKG), which shows the beat pattern of the heart; magnetic resonance imaging (MRI) or computed tomography (CT) scan, which show a picture of the brain and spinal cord; a spinal tap to examine the cerebrospinal fluid; nerve conduction studies, which measure the speed with which nerves transmit impulses; blood and urine tests to look for high glucose levels; and genetic testing to identify the affected gene.

Adaptive devices (such as canes or walkers), therapies (such as speech therapy to improve speech and aid swallowing), and medications for accompanying complications (like heart problems) can assist patients.

Although there is no effective treatment or cure for Friedreich's ataxia, researchers are hard at work.

"Idebenone has shown some efficacy in preventing or reversing the associated heart disease, but even idebenone has failed to reverse or slow progression of the neurological symptoms," Dr. Gottesfeld says. " Our compounds—called histone deacetylase inhibitors—target the loss of frataxin protein, which is the cause of the disease. They do this by reactivating the gene that is silenced in Friedreich's ataxia."

Dr. Gottesfeld will continue to study how the enzyme blocked by 4b controls frataxin production and its relationship to inactivating the frataxin gene. Repligen Corporation, which was involved in Dr. Gottesfeld's study, is doing all of the preclinical testing necessary to initiate a human safety trial.

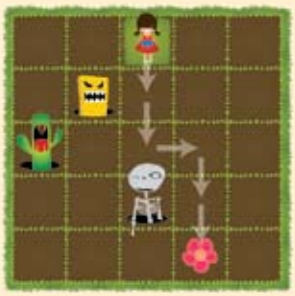
"Erythropoetin has also been found to increase the levels of frataxin protein, and this drug is in clinical trials," Dr. Gottesfeld adds.

Genetic testing assists with prenatal diagnosis, clinical diagnosis, and carrier status determination. Psychological counseling and support groups may also help patients and their families cope. —*Elizabeth Stump*

MEMORY | LANGUAGE | VISUAL | SPATIAL | LOGIC | NUMBER

NEUROBICS

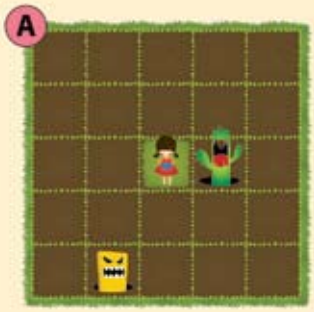
Monster Garden



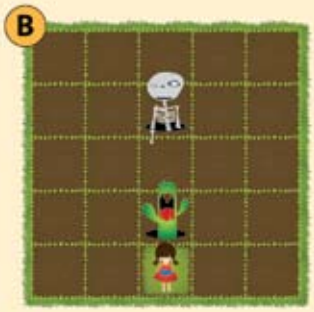
The garden at right is divided into squares. The gardener in the green square wants to find the shortest path to the flower that avoids all the monsters. In this case she can get there in just five steps by following the arrows.

Now it's your turn. Your job is to find the shortest path from the gardener to the flower. But this time you'll have to use your memory. Look at garden A below. Memorize the positions of the gardener and the two monsters. Got it? Once you have the positions memorized, turn the page and continue reading.

GO TO P. 15

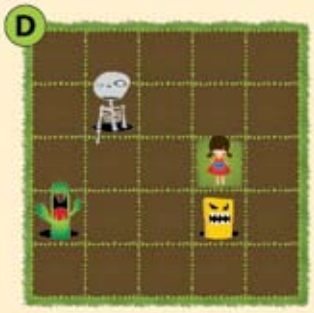


Remember these positions



Remember these positions

Remember these positions



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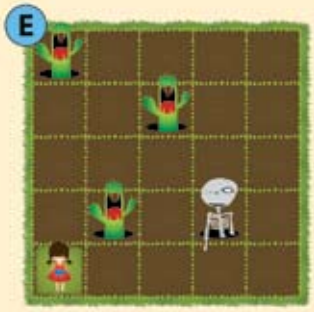
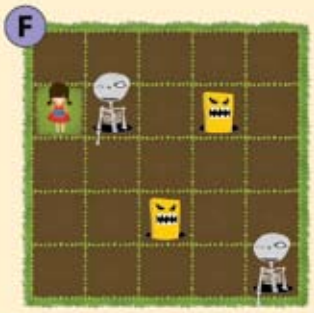


Remember these positions



Remember these positions