

When cystic fibrosis patients grow up

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Scott Johnson, now 39, was diagnosed with cystic fibrosis when he was three years old.

When Scott Johnson was diagnosed with cystic fibrosis at age three, most patients with the disease were dying in their teens.

Today, Johnson, 39, works as a clinical data manager in Wilmington, N.C. He's preparing for his sixth Ironman distance triathlon, which will take place in November.

Johnson is one of a growing number of American adults who are living with cystic fibrosis. Together, those adults are changing the way that doctors care for patients with the disease.

Cystic fibrosis is a genetic disease that causes the body's mucus, sweat

and digestive juices to be unusually thick and sticky. Those secretions can clog organs and airways, leading to dangerous infections.

The disease used to kill patients in childhood or adolescence. But today, nearly half of the 30,000 cystic fibrosis patients in the U.S. are adults, said Beth Anne Champion, MD, MPH. Champion is a clinical fellow in pediatric pulmonology for UNC Health Care.

Longer lifespans are the result of advances in treatments for cystic fibrosis patients. For example, Johnson received a double-lung transplant at UNC Hospitals in September 2001 after a bout with pneumonia.

Just a few decades ago, most cystic fibrosis experts were pediatricians. Today, more adult-medicine specialists are caring for patients. Centers for adult cystic fibrosis patients have sprung up around the country, including at UNC Health Care, which also operates a pediatric center.

Pediatricians are also learning to help their young patients transition to adult medicine.

Many cystic fibrosis patients receive care from the same team of doctors from birth through their teenage years, Champion said. Patients become attached to their doctors, and vice versa.

For patients, learning to work with a new team of adult specialists can be a difficult transition, Champion said: “You get used to seeing the same faces and seeing the same people.”

In addition, some cystic fibrosis patients have difficulty obtaining health insurance after they age out of their parents’ plans, Champion said.

Meanwhile, patients are dealing with the challenges that every teenager must confront: relationships, education, leaving home and choosing a

career, said Elisabeth Dellon, MD, MPH, assistant professor of pediatrics in the division of pulmonology.

“Those are all issues that – thankfully – our patients have to face,” Dellon said.

At UNC, doctors begin discussing transition with patients in early adolescence. Doing so can help patients understand that graduating to adult care is a normal part of growing up, Champion said. Doctors are also working to improve communication between pediatric and adult speciality teams and developing a program to orient teens to adult care.

TIPS

For cystic [fibrosis patients](#), Johnson, Dellon and Champion offered tips on preparing for transition.

- First, know your disease, Johnson said. “Learn as much as possible,” he said.
- For example, you should know how cystic fibrosis will affect your daily life and your future, Champion said. You should be able to recognize when your symptoms are getting worse and what to do about it, she said.
- You should also understand your therapy. Knowing why you need to take so many pills will help you stay motivated to stick to your treatment plan.
- Learn to navigate the health care system. For years, your parents or guardians have probably been finding doctors, scheduling your appointments and picking up your prescriptions. Mastering those tasks will help you stay healthy as an adult.
- Take ownership of your disease. Beginning in your early teen years, you can start to self-administer your home therapies, with the guidance of an adult.

- Ask questions. Your doctors will be happy to help you understand your disease and help you make good decisions. When it comes to trying new medications or adjusting doses, “the only person who knows if something is going to work or not is you,” Johnson said.
- Finish school and choose your career carefully. Because of the expense of therapies for [cystic fibrosis](#), you will likely need to find a job in which you will receive health insurance, Champion said. At UNC [Health Care](#), a clinical social worker can assist you in choosing a career path.
- Don’t forget that you will have a great team of doctors to care for you. “Yes, transition is a difficult time,” Champion said. “But adolescence and early adulthood are also a very exciting time.”

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