Diagnosis and treatment of cystic fibrosis in adults: NetWellness

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Just 20 to 30 years ago, most people with CF did not survive into adulthood. Fortunately, most children and teens with CF today will live to become adults thanks to improved medication, care and understanding of the disease. Most will even be healthy enough to pursue the same goals that those without CF dream of.

Today's children may be able to live until middle age or even to a normal life expectancy. Healthy adults with CF now pursue careers, raise families and enjoy most leisure activities.

Adults with more advanced illness do have fewer opportunities, a shorter expected lifespan, and a more restricted lifestyle. But these adults can still benefit from improved therapies that reduce the frequency of severe health problems. All patients can expect further scientific advances to better their lives over the next several years.

Nonetheless, it is important to be realistic. Progressive lung disease, breathing difficulty and relatively early death are still the eventual outcomes...
Is CF ever diagnosed in adults?

CF is usually diagnosed in children or teens. But one or more of the following conditions may be seen in adults diagnosed with the disease:

- Chronic asthma
- Sinusitis
- Allergies
- Diarrhea
- Low weight problems

A sweat test or genetic testing will confirm the diagnosis of CF. Confronting the prospect of a new, life-shortening disease can be challenging. It can be psychologically difficult and it involves adding a complex and time-consuming new treatment regimen into an already-busy life. Often, the person will be admitted to the hospital to receive initial IV antibiotics and training.

It can also be a relief to get a CF diagnosis, identifying why someone has been ill. In the long run, improved health and energy usually offset the new difficulties.

How is cystic fibrosis treated in adults?

Treatment of CF in adults is similar to that of children and teens. In adults, sticking to therapies is at least as important, if not more so, than when younger. Inevitably, lung damage is greater in adulthood than earlier in life. Therefore, it is critically important to maintain the best possible health. Steps you should take include the following:

- Maintain excellent nutrition
- Maintain a normal weight
- Keep lungs as clear of mucous and secretions as possible
- Treat infections promptly

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These requirements are difficult to fit into a busy schedule, especially one that involves school, parenting or work. Thus, you should clarify with the care team which activities are crucial, and which can slide a little. Those that are important must be fit in, or your health and ability to enjoy other life pursuits will suffer.

Treatments may consist of very few medications and therapies. More likely, treatments involve a half-hour of inhaled medication and lung-clearance activities once or twice a day. Oral medication before meals or snacks is also needed to improve food absorption. Other medications and treatments depend on the level of care required.

It is critical to monitor problems early before they worsen and cause permanent damage. Oral or intravenous antibiotic medication may be needed at times to decrease the level of lung or sinus infection. This will sometimes require admission to the hospital.

What other kinds of problems occur in adults along with CF?

Diabetes: It is estimated that 15 to 45 percent of those older than 30 in the CF population have CF Related Diabetes Mellitus (CFRDM). The primary source of the CFRDM appears to be an inability of the pancreas to produce enough insulin. The usual treatment involves the use of injected insulin. Some people can be treated with oral medications, at least early in the process. Often, these people will eventually require the use of insulin.

Osteoporosis: Thin bones are almost universally common in adults and even teens with CF. It is still unclear exactly why this occurs. Adults with CF should have routine bone-density evaluation. If needed, adults should be treated with weekly or monthly medication for treatment of osteoporosis to prevent broken bones.

Liver problems: About 25 percent of children and adults with CF have some degree of liver abnormality, although it is mild for most. Only a small number in this group will have problems that affect their health. Occasionally, these problems will require daily medication. Even less commonly, they will cause cirrhosis and the need for liver transplantation. The possibility of liver problems makes it a necessity to keep alcohol intake at a moderate level, at most.

Joint pain/arthritis: A significant minority of people with CF will have some degree of joint pain, and sometimes actual arthritis. This can range from mild, occasional discomfort to chronic arthritis pain. This is always best treated with the usual arthritis medications, and sometimes with the assistance of an arthritis specialist. It is best not to be treated with any habit-forming or narcotic medications.

Can people with CF have families?
Yes, but some CF-related factors can have an effect. Both men and women need to consider their health and life expectancy in their decisions about caring and providing for a family.

Women with CF may have greater difficulty conceiving. Before conceiving, women should consider both their ability to stay healthy during pregnancy and while caring for a demanding infant.

Most men (95 percent) with CF cannot conceive through usual intercourse due to a blockage or absence of the tubes that carry sperm from the testicles. However, men should NOT assume infertility until tested for fertility to avoid surprises. Men can have biological children through the use of alternative methods involving sperm harvesting and artificial insemination.

**Can people with CF have careers?**

Almost all teens and adults with CF can pursue educational and work goals, and should plan to do so. Keep in mind that some career paths require careful consideration and may be difficult to follow. For example, health care fields involve contact with ill individuals, which can be risky for a CF patient. Your CF team can help sort out these issues.

**What are the problems facing adults with advanced illness?**

Virtually all adults with CF will face increasing lung disease and possibly the need for lung transplantation. This can improve life expectancy and the quality of life. For those who are either unable or unwilling to get a transplant, legal, family and personal issues should be resolved in advance. It is important for individuals to experience the end-of-life period according to their wishes.

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