CFRI community
48 Years of Research, Education, Advocacy, & Support
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Aging With CF: Special Considerations

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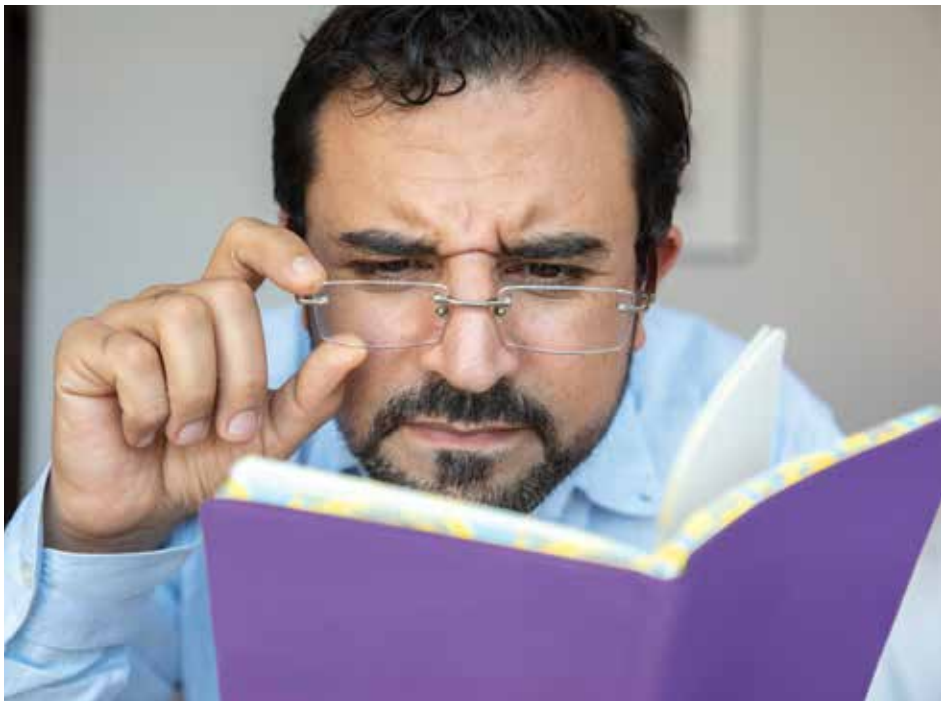
I spent my entire life wanting to grow older. And now I'm over 50 and my body is showing normal but challenging signs of aging with CF. Menopause, aches and pains, bone loss, poor eyesight... I try to remember these are all the benefits of still being alive!"

— Isabel Stenzel Byrnes, LCSW, MPH,
living with CF and lung transplant recipient



Advances in cystic fibrosis (CF) therapies have steadily increased survival for those living with the disease, and many members of our CF community are now reaching middle age. Until recently, issues that impact all people as they age – cancer, heart disease, diabetes, arthritis – were often not discussed by medical care providers with their CF patients. The improved health of many people with cystic fibrosis – especially among those benefiting from highly effective modulator therapies – has moved these age-related issues to the forefront. Growing old with CF presents its own unique blessings and challenges. The resilience required to live with CF is further tested with age.

Pulmonologists at adult CF centers are now encouraging broader health screenings and the integration of preventative care. Says Dr. Ahmet Uluer, director of the Adult CF Center at Brigham and Women's Hospital and Boston Children's Hospital, "Current guidelines do not adequately address the changing landscape of aging during the highly effective modulator era. We urgently need a modified lens to expand and adjust the age of screening, identify and prevent other complications, and support age-related CF research."

CF and Cancer

Individuals with cystic fibrosis have five to ten times the rates of colon cancer as the general population. For those post-transplant, who are dependent upon

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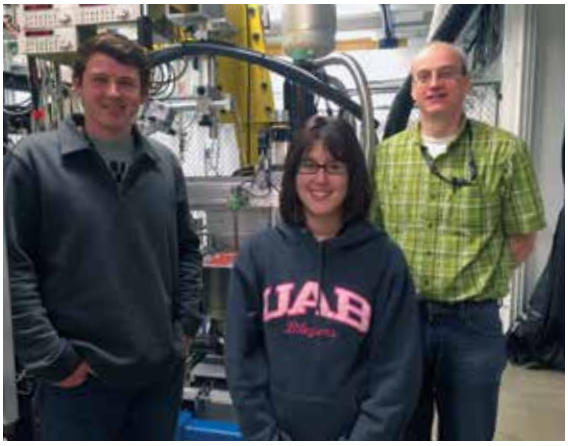

Cole Martin, Dr. Christina Le and Dr. Stephen Aller

Role of CFTR Arginine-933 in Channel Function and Drug Potentiation

Wei Wang, Lianwu Fu, Cole L. Martin, Steven M. Rowe and Stephen G. Aller
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Cystic fibrosis is caused by protein defects in the CF transmembrane conductance regulator (CFTR). CFTR is an inherently difficult protein to study because: 1) it is located in the cell membrane; 2) folding of CFTR from non-CF individuals is still inefficient compared to other membrane proteins; 3) many mutations in CFTR that cause disease fold extremely inefficiently, and 4) it is very difficult to achieve sufficient folded/functional protein for biochemical studies. Even today, very rare mutations of CFTR, such as changes to Arginine-933 (Arg933),

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immunosuppressants to prevent organ rejection, the risk increases to 25 to 30 times that of the general population.

Currently, the Cystic Fibrosis Foundation recommends that those with CF have their first colonoscopy at age 40 (30 for those post-transplant), but there are many who believe that this age should be lower. "I was diagnosed with stage 4 colon cancer at the age of 34," says Anna Payne. "Colon cancer symptoms mimic CF-related GI symptoms. I would never have survived until 40 to get that first screening. I strongly believe the age must be lowered." Adds Dr. Uluer, "People with CF live with the expectation the other shoe is going to drop. Health screening such as colonoscopy can not only detect but prevent the other shoe from dropping by potentially preventing colorectal cancer."

Skin cancer is another common cancer, particularly for those post-transplant. Regular screening by a dermatologist and a daily commitment to the use of sunscreen and protective clothing are the best defense. In addition to colon and skin cancers, rates of pancreatic, small bowel and other GI cancers are elevated for those with CF. For those post-transplant, the risk of developing cancer increases dramatically, with studies finding increased rates of lip, cervical, testicular, kidney, lung, and bladder cancers, and lymphomas. The importance of regular screenings cannot be overstated.

Heart Disease

Heart disease among those with CF was historically associated with chronic hypoxemia (low levels of oxygen in the blood), and was most often seen in those with advanced lung disease. This is beginning to change since the approval of the triple-combination modulator Trikafta. Many who struggled for decades to keep weight on now find themselves with significant weight gain and obesity, leading to a higher risk of hypertension or high blood pressure.

CF-Related Bone Disease

It is very common for osteopenia to develop in adolescence, and adults with CF have higher rates of osteoporosis. Reasons for this include CFTR dysfunction that leads to lower bone mineral density, poor absorption of calcium, vitamins D and K, the use of glucocorticoids, CF-related diabetes, and

delayed nutrition. Individuals with CF should have their first DEXA scan (bone density scan) by age 18 to detect bone loss and prevent fractures.

Hearing Loss

The use of IV antibiotics to treat pulmonary exacerbations has played a key role in improving survival. If not appropriately monitored, and even when they are, some of these antibiotics may be associated with ototoxicity, inner ear damage that develops as a side effect of the medications. Often beginning with a ringing in the ears (tinnitus), ototoxicity can advance to severe hearing loss and balance issues. Hearing loss is associated with depression, social isolation, and cognitive decline. Hearing tests should be routinely conducted to monitor the impact of specific IV and inhaled medications.

Liver Disease

As our CF community ages, the prevalence of CF-related liver disease is growing. The mucus that is the hallmark of CF may also block bile ducts in the liver. When bile cannot exit the liver, inflammation and scarring occurs, and the liver cannot function properly. Long-term use of IV antibiotics can also damage the organ. While there is no specific test for CF-related liver disease, clinical assessment of symptoms, blood tests and ultrasounds are used to make a diagnosis.

Menopause

It has long been understood that hormones can play a role in lung function and exacerbations. As more women with CF reach the age of menopause, a new field of research has launched to assess the impact of menopause upon women with CF. The CF Foundation notes that women with CF may enter menopause two to three years before women in the general population. Menopause has impacts upon bone loss, lung function, muscle strength and mental health, and the management of symptoms should be explored with one's CF care team. The Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC) has an excellent resource on CF and menopause which can be found at cfreshc.org.

Dental Health

Studies have found that individuals with CF are more likely to have a lower pH in their saliva, and reduced saliva secretion. This can be worsened by medications, and can lead to oral infections, gum disease and cavities. While addressing dental health may have been a lower priority for many in light of addressing lung infections and other CF complications, as CF community members age, there is growing recognition of the need to monitor and support dental health.

Mental Health

The TIDES study demonstrated the higher rates of depression and anxiety experienced by those living with cystic fibrosis. Growing old with CF can create an entirely new source of fears, anxiety and depression. Financial concerns may increase for people who never expected to survive to an age where a retirement fund would be needed. New age-related health complications and the isolation created by cross-infection risks exacerbate anxiety. Lower energy or cognitive changes can make one's self-care management overwhelming, leading to weariness. The correlation between mental and physical health is direct. It is as important to address one's mental health as one's physical health. Counseling, support groups, social engagement, mindfulness practices, and physical activity are all strategies that can support improved mental health. Having access to disability resources and affordable insurance programs as people with CF age is critical as well.

As we celebrate the medical advances that have led to an aging cystic fibrosis community, we must also recognize and address the interplay of CF with the challenges of age-related health issues. Shares Isabel Stenzel Byrnes, "When I was 22, my 46-year-old friend and mentor with cystic fibrosis said, 'Aging ain't for sissies.' Wasn't he correct? But it sure beats the alternative. Aging gracefully with CF invites us to remain tenacious, informed, and grateful."

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