

**In Memoriam**  
**Dr. Deron Arnold**  
**“Here In Time”**  
**August 18, 1971 – November 8, 2009**



“Further up and further in” came the cry upon  
the wind,  
“Leave the shadow-land so dim, my friend”  
----“The Last Battle” by C.S Lewis

On November 8, 2009, Dr. Deron Arnold passed away at the University of Minnesota-Fairview hospitals due to complications of a double lung transplant. He was diagnosed with cystic fibrosis at the age of four and made it his goal to become a doctor at a young age.

Dr. Arnold joined the staff of MPC on July 7, 2003. He had been working as a locum tenens off and on for approximately the previous year and a half prior to joining the pathology group. Dr. Arnold received his undergraduate degree in Chemistry from South Dakota State University in Brookings, SD and attended medical school at the University of South Dakota-Vermillion, receiving his MD in 1998. He completed his pathology residency at the University of South Dakota- Sioux Falls in 2003.

During his tenure at MPC, Dr. Arnold served as medical director at Appleton Area Health Services, Appleton, MN and at Johnson Memorial Health Services, Dawson, MN. He also served as clinical pathologist for hematology and coagulation and as anatomic pathologist for post-mortem services for Rice Laboratory. His areas of special interest were dermatologic, gastrointestinal, gynecologic, and hematologic pathology. Dr. Arnold wrote several “Path News” articles for the laboratory newsletter *Reachout*, and in honor of him the article that he wrote on cystic fibrosis from August 2005 is reprinted on the back of this memoriam.

Dr. Arnold enjoyed his profession so much that he often requested medical books as well as various study Bibles as gifts. From a young age, he used his treatment time to read and study. He loved to read the Bible, especially the Psalms and various books about Christianity. C. S Lewis

and J.R.R. Tolkien were among his favorite authors. During his later treatment time, he searched the internet for various tidbits of interest-many of which he incorporated into his blog “Here In Time.” Being a husband and father was one of his life’s greatest desires- one not often fulfilled for someone with cystic fibrosis. His wife Jan and sons Maxwell and Mark gave him a great deal of happiness, joy and fulfillment; like no other experience in his life.

### Remembrances:

“I will always remember how incredibly positive he was in the face of all his health issues. He set such a great example of how keeping the faith can spur us to be positive people in all areas of life.” --Joel

“He always greeted all staff whether in the lab or at break/noon meal. His routine greetings for the lab staff included “how are my fellow laboratorians?” and “Good morning fellow laboratorians.”—Connie, Junell, Steph

“He didn’t like his ice cream hard and would put it (ice cream bar) into his shirt pocket to warm up—when it was warm enough; that was the right time to eat it.” --Dawn

“My memories of Dr. Arnold always bring a smile to my face. He would always stop me as I was walking by his office and one never really knew what he was going to ask! Just random questions and I would think ‘where does he come up with some of these things?’ Dr. Arnold was a very knowledgeable man and therefore liked to share it with others. His positive attitude and fun stories about his life, his work, and especially his family always showed in his laugh and smiling face even when we all knew he was not feeling well. He will be missed by all who knew him.”--Lisa

## **PATH NEWS-Reprinted from the August, 2005, *Reachout***

### **CLINICAL SCENARIO:**

Five year-old Billy is brought to your laboratory for sweat chloride testing. He has been having repeated "colds" and has failed to gain weight appropriate for his age. A sweat chloride test is performed which shows a value of 81 mEq/L. The test is later repeated and the result is 78 mEq/L. This is considered positive for cystic fibrosis (CF).

Billy's father previously died in a boating accident. Billy's mother recently remarried and would like to have another child with her new husband, Ron. The mother knows she is a carrier for CF, but she is wondering if Ron is also a carrier. She asks you if there is any test to evaluate Ron's carrier status before she gets pregnant. Furthermore, she asks you if Ron is a carrier, what are their chances for having a child with CF?

### **WHAT IS CYSTIC FIBROSIS?**

Cystic fibrosis is an inherited disease that is fairly common and affects approximately 30,000 Americans. One in every 2500 Caucasian newborns is affected. It is caused by an abnormal gene called CFTR. The disease has numerous manifestations but most commonly attacks the lungs, plugging them up with thick mucus. In addition, the disease can prevent proper absorption of food thereby causing malnutrition. The typical presentation is a skinny, sick kid who "always has a cold".

Affected males ejaculate no sperm due to absence of the vas deferens. (The sperm is normal, however, and with *in vitro* techniques, CF males can now father children; however, the children will get the abnormal CF gene from the dad and will be CF carriers. But provided that the mom does not also give them an abnormal CF gene, the child will not develop CF.)

The average prognosis for CF used to be downright dismal but now it's just poor. Most patients can expect to make it to age 30. Like many other genetic diseases though, the severity of CF is variable. For some patients, the only symptom they'll ever notice is infertility or sinusitis. Although any ethnicity can be affected, Caucasians are affected much more frequently than any other group.

### **HOW IS CYSTIC FIBROSIS DIAGNOSED?**

In infants or children in whom the disease is suspected, a sweat chloride test is ordered. This is a strange little test that measures the amount of chloride in the patient's sweat. Basically, a small area of the skin is made to perspire. This perspiration is collected and the chloride ion is then quantified. The theory behind the test is that the CFTR protein is responsible for helping to transport electrolytes in and out of a patient's cells. A mutated protein doesn't transport ions into and out of the cell effectively and therefore certain ions (i.e. sodium and chloride) build up in the extracellular fluid. Because these ions can't get into the cell, they stay outside the cell and are eventually excreted in the sweat. The more sodium chloride in the sweat, the more likely the patient has CF. This is why a patient with CF has salty tasting skin.

The cut-off used here at Rice for the diagnosis of CF is 50 mEq/L. Any level above this should be retested for confirmation. The reported sensitivity and specificity of the sweat chloride test is roughly 94% and 99%, respectively. Most experts agree that the test is less reliable in infants less than 4 weeks old and in children/adults over 10 years of age.

### **WHAT ROLE DOES GENETIC TESTING PLAY?**

One major role of the genetic test is to determine CF carrier status. A CF carrier has one mutated CFTR gene and one normal CFTR

gene, while a patient with CF has two mutated CFTR genes. As CF is an autosomal recessive disorder, both parents of a child must be a carrier in order for the child to be affected. Each parent must give his/her abnormal CFTR gene to the child. If one parent gives a normal CFTR gene and the other parent gives an abnormal CFTR gene, the child will only be a carrier (i.e. asymptomatic).

If both parents are carriers, each child they conceive will have a 25% chance of developing the disease.

The mutated CF gene is present in 1 in every 29 Caucasians; therefore, many couples want to be tested for CF in the process of planning a pregnancy; or sometimes they desire the testing even during the pregnancy. Although over 1,000 mutations have been found to cause CF, most of these mutations are actually quite rare and therefore only a small fraction of the known genetic mutations are tested. The genetic test we routinely use here at Rice (via Mayo Medical Labs) will detect approximately 91% of CF carriers in the Caucasian population.

Genetic testing can also be utilized in the actual diagnosis of affected CF patients including *in utero* fetal diagnosis.

### **WHEN SHOULD TESTING BE PERFORMED?**

Genetic testing is best performed prior to a woman getting pregnant. If the father and mother are both found to be carriers, they are faced with several options, two of which are discussed here. One option would be for *in vitro* fertilization where the resulting embryos can be tested for the disease and only healthy ones are implanted in the mother's uterus.

Another option would be to simply go ahead and conceive naturally but utilize the nine months of pregnancy to learn more about CF and plan for raising a child with CF. An amniocentesis performed during the pregnancy can help establish the diagnosis and better help the parents to prepare.

*Genetic testing should only be performed in conjunction with genetic counseling.*

### **WHAT ABOUT NON-CAUCASIAN PATIENTS OR PARENTS?**

To diagnose CF, non-Caucasians can be tested the same way as their Caucasian counterparts with a sweat chloride test. Genetic testing can also be performed by Mayo Medical Laboratories. The Mayo genetic test will detect 58% of Hispanic carriers and 72% of African-American carriers. It should be stressed, however, that CF is very rare in these populations.

### **ARE THERE ANY OTHER TESTS FOR CYSTIC FIBROSIS?**

Sweat conductivity is a test similar to the sweat chloride test. It is sometimes used as a screening test and may be easier and less time consuming to perform but is not widely used as it may be less accurate. Other testing modalities such as serum trypsin immunoreactivity and nasal ion potential difference are not widely used in the U.S.

### **LITTLE KNOWN FACT ABOUT CYSTIC FIBROSIS:**

Because of their defective chloride transport protein, most CF patients are resistant to the toxin released by *Vibrio cholerae*. Thus those with CF can take some comfort in knowing that if there's ever a cholera epidemic, they won't get the runs. This is probably how the mutation was selected for in the past. CF carriers survived cholera epidemics so they were more likely to reproduce.

*Dr. Deron Arnold  
Minnesota Pathologists Chartered*