Cystic Fibrosis Related Diabetes: Patient Information Part I

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Introduction:

People with CF have a unique form of diabetes called Cystic Fibrosis Related diabetes (CFRD). It is not the same as the commonly known type 1 and type 2 diabetes mellitus, but it is the most common extra-pulmonary complication of CF. In fact, few people with CF have normal glucose levels. One study found that up to 75% of adults with CF have some form of glucose intolerance. This condition worsens with age and CFRD affects 20% of adolescents and 40-50% of adults. It is important to screen for it because it is associated with worsening pulmonary function, increased mortality, and microvascular complications. The decline starts even before the actual diagnosis, so early intervention is key.

Pathophysiology:

In CF, the abnormal CFTR gene results in thick viscous secretions building up in the pancreas, similar to the process in the lungs. This leads to chronic infection and inflammation which eventually results in scarring of the pancreatic tissue. This affects insulin release and causes elevations in blood glucose levels. Insulin resistance is another feature of CFRD and is due to chronic inflammation, infections and medications.

The process shares some similarities with the typical type I and II diabetes but remains different and progression is slower and more gradual.

Signs/Symptoms:

Most patients with CFRD have no obvious signs or symptoms. This emphasizes the importance of the annual oral glucose tolerance test (OGTT) starting at age 10 years (or earlier if CFRD is suspected). Some individuals with CFRD, however, may experience fatigue, inability to gain weight, weight loss, declining lung function, and increased respiratory infections.

Prognosis:

Untreated CFRD can worsen the progression of CF through poor nutritional status, declining lung function, increased pulmonary exacerbations, and decreased survival. Uncontrolled diabetes can also lead to microvascular complications meaning nerve damage, small vessel damage in the retina and kidneys and autonomic dysfunction such as gastroparesis. These do not progress to the same level of severity as your typical type I and II diabetes. Cardiac events, in contrasts to type I and II diabetes, are rare.

For More Information:

https://www.cff.org/managing-cf/cystic-fibrosis-related-diabetes

https://www.cysticfibrosis.org.uk/what-is-cystic-fibrosis/how-does-cystic-fibrosis-affect-the-body/symptoms-of-cystic-fibrosis/cf-related-diabetes

https://cystic-fibrosis.com/cfrd