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Trikafta Update

The U.S. Food and Drug Administration has approved the use of Trikafta® (elexacaftor/tezacaftor/ivacaftor) for children ages 6 through 11 who have at least one copy of the F508del mutation or certain mutations in the CFTR gene that are responsive based on lab data.

Nearly 1,500 children will now be eligible for a CFTR modulator. Highly effective CFTR modulators, such as Trikafta, are proven to have a positive effect on the health and well-being of many people with CF. For instance, studies have shown that some young children taking CFTR modulators have had their pancreatic function restored. However, these therapies do not reverse established lung damage. This is one of the reasons it is important to start on CFTR modulators at a young age. Research suggests that beginning treatment with a modulator early could help slow or even prevent the irreversible progression of cystic fibrosis. Despite the significant progress that has been made in helping people with CF live longer and healthier lives, there is still important work that needs to be done to help all people living with this disease.

For more information, please see the Vertex press release at:

<https://investors.vrtx.com/news-releases/news-release-details/vertex-announces-us-fda-approval-trikafta>

CF Foundation's "Path to A Cure" Initiative Update

The Cystic Fibrosis Foundation has recently announced three new research awards as part of its "Path to a Cure" initiative. The funding includes up to \$2.6 million to Elox Pharmaceuticals to identify compounds that could potentially treat individuals who have nonsense mutations, as well as awards to Hunterian Medicine and Metagenomi to advance genetic therapies for all people with CF.

What is the CF Foundation's "Path to A Cure" initiative?

The CF Foundation launched its \$500 million "Path to a Cure" initiative in October 2019. This initiative is focused in developing new treatments for the underlying cause of CF. It centers around three core strategies:

- Repairing broken CFTR protein
- Restoring CFTR protein when none exists
- Fixing or replacing the underlying genetic mutation to address the root cause of CF

For more information on "Path to a Cure," please visit cff.org.

Sodium Chloride

What is Sodium Chloride?

Sodium and chloride are minerals that together are known as salt. They are “essential” which means they can only be obtained through diet.

Salt helps maintain the body’s fluid balance – or keeping the body’s water within healthy ranges. Salt also helps muscles and nerves function correctly. Not getting enough salt can cause a variety of issues including:

- Interfering with growth
- Reducing appetite
- Stomach pain, weakness, nausea, and headache

How much Salt is needed?

It has not been determined exactly how much salt children & adults who have CF need. The recommendation is to eat salty foods and use the salt shaker freely at meals and snacks. People with CF, especially if they play or exercise outside in hot weather, lose extra salt in their sweat. They may want to add 1/8 teaspoon of salt to 1 ½ cups (12 ounces) of a sports drink, such as Gatorade or PowerAde.

Best Sources of Salt

The best sources of sodium chloride are table salt and processed foods like bacon and pickles. Fresh foods such as lean meats, fruits, vegetables, rice and pasta naturally have very little salt. However, these foods become high in salt when they are processed into canned and boxed soups, vegetables, pastas, and frozen dinners. It is important to read food labels and use the salt shaker to add extra salt. Food labels tell you how much sodium (or salt) is in food.

North American Cystic Fibrosis Conference Update

Registration for the Virtual Event is Now Open!

Due to rising cases of COVID-19 infection across the United States, the CF Foundation has decided to cancel the in-person, 2021 North American Cystic Fibrosis Conference (NACFC). This was originally scheduled to take place September 30th – October 2nd in San Antonio, TX. NACFC 2021 will now be virtual-only, and is rescheduled for November 2nd – 5th, 2021. For more information on the conference, please visit www.nacfconference.org

Important Dates:

Cystic Fibrosis Clinic Dates

September 16th, 2021
October 21st, 2021
November 18th, 2021
December 16th, 2021

Patient Advocacy Group Dates

November 11th, 2021
January 13th, 2022

Your CF Team Members:

Dr. Monica Paulo
Dr. Jane Gorud
Dr. Parveen Wahab
Deb Fueller FNPC
Heidi Shannon FNPC
Leanne Saville PharmD
Stephanie Friedt RRT.RPFT
Kim Chumley RN
Amber Burck, LPN
Samantha Snow, RDN, LRD
Christina Bond, LMSW

35th North American Cystic Fibrosis Conference

November 2nd – 5th, 2021 (Virtual only)

More information is available at <https://www.nacfconference.org/Home/>