MCG PEDIATRIC PODCAST

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Season 2, Episode #12

Cystic Fibrosis-

Nutrition and Exercise Considerations

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I. Cystic Fibrosis, or CF, is an autosomal recessive inherited disease caused by a homozygous mutation in the CFTR gene.
   a. This disease is most common in Caucasians – approximately 1/25 Caucasians are carriers of the CF gene, and approximately 1/2500 Caucasians have CF.
   b. This gene is located throughout the body which contributes to many different phenotypes associated with CF.

II. If prenatal testing or newborn screening identifies a patient who may have CF, that patient undergoes sweat chloride testing.
   a. The sweat chloride test is a safe, painless, and effective way to diagnose CF. This test measures the amount of a chemical element called chloride in the sweat. Patients with CF have high levels of chloride in their sweat compared to those without CF. The sweat chloride test has a 99.7% specificity for CF

III. Most notably, CF affects the lungs. This is why people with CF develop thick mucus which is difficult to clear, leading to increased risk of harboring for infection.
   a. Due to the localization of CFTR, patients with CF are characterized by pulmonary dysfunction, pancreatic insufficiency, decreased exercise capacity, and other systemic abnormalities such as meconium ileus or risk of intestinal obstruction. Many of these abnormalities cause these individuals to be at risk for malnutrition.

IV. Children with CF often have pancreatic insufficiency which can lead to maldigestion, malabsorption, and may even lead to CF-related diabetes.
   a. Symptoms of pancreatic insufficiency resulting in maldigestion and malabsorption may include: Poor weight gain despite a good appetite, Abdominal pain, gas, and bloating, and frequent loose, foul-smelling stools or fatty appearing stool known as steatorrhea.
   b. Pancreatic insufficiency is present at birth in about 60 to 80 percent of children with CF. This means that the majority of these children may never have the chance to maintain adequate nutrition without supplementation.

V. Children with CF often require higher calorie diets to maintain adequate nutrition, and that their growth needs to be monitored more closely that children their age who do not have CF.
   a. These children also may need vitamin and mineral supplementation, specifically sodium chloride, zinc, and essential fatty acids.

VI. If children with CF are not able to maintain a good body composition on their own (i.e. appropriate ratio between fat mass and fat-free mass), enteral nutrition may be needed.

VII. Exercise is also extremely important for people with CF, and exercise capacity is a marker of disease-associated morbidity and mortality.
   a. The mechanism behind exercise intolerance in these patients is multifactorial, but regular exercise has been associated with improvement in clinical symptoms of CF. Studies have shown that individuals with CF have decreased exercise capacity, independent of lung function, indicating that poor pulmonary function is not the driving force for exercise intolerance.

References:

1. Gajbhiye, R., et al., *Cystic fibrosis transmembrane conductance regulator (CFTR) gene abnormalities in Indian males with congenital bilateral absence of vas deferens & renal anomalies.* Indian J Med Res, 2016. 143(5): p. 616-23. AND Elborn, J.S., *Cystic fibrosis.* Lancet, 2016. 388(10059): p. 2519-2531.)
2. (Kuk, K. and J.L. Taylor-Cousar, *Lumacaftor and ivacaftor in the management of patients with cystic fibrosis: current evidence and future prospects.* Ther Adv Respir Dis, 2015. 9(6): p. 313-26.)
3. (Farrell et al, Siret el al, Sims et al).
4. Sullivan, J. S., & Mascarenhas, M. R. (2017). Nutrition: Prevention and management of nutritional failure in cystic fibrosis. *Journal of Cystic Fibrosis, 16*. doi:10.1016/j.jcf.2017.07.010
5. Committee On Practice And Ambulatory Medicine, & Workgroup, B. (2020, March 01). 2020 recommendations for Preventive Pediatric health care. Retrieved March 22, 2021, from https://pediatrics.aappublications.org/content/145/3/e20200013
6. Cystic Fibrosis Foundation, et al. Cystic Fibrosis Foundation evidence-based guidelines for management of infants with cystic fibrosis, J Pediatr 2009; 155(6 Suppl):S73-93.)
7. (Nutrition Prevention and Management Article)
8. (Mattar AC, Leone C, Rodrigues JC, Adde FV. Sweat conductivity: an accurate diagnostic test for cystic fibrosis? J Cyst Fibros. 2014 Sep;13(5):528-33. doi: 10.1016/j.jcf.2014.01.002. Epub 2014 Jan 31. PMID: 24485874.)
9. Ameet Daftary, James Acton, James Heubi, Raouf Amin, Fecal elastase-1: Utility in pancreatic function in cystic fibrosis, Journal of Cystic Fibrosis, Volume 5, Issue 2, 2006, Pages 71-76,ISSN 1569-1993,
10. J. Walkowiak, D. Sands, A. Nowakowska, R. Piotrowski, K. Zybert, K.H. Herzig, *et al.* Early decline of pancreatic function in cystic fibrosis patients with class 1 or 2 CFTR mutations J Pediatr Gastroenterol Nutr, 40 (2) (2005), pp. 199-201
11. Witt, H. (2003). Chronic pancreatitis and cystic fibrosis. *Gut, 52*(90002), 31ii-41. doi:10.1136/gut.52.suppl_2.ii31
12. Fielding, J., Brantley, L., Seigler, N., McKie, K. T., Davison, G. W., & Harris, R. A. (2015). Oxygen uptake kinetics and exercise capacity in children with cystic fibrosis. *Pediatric Pulmonology, 50*(7), 647-654. doi:10.1002/ppul.23189
13. Orenstein, D. (2018). The Relationship between CFTR Genotype and Exercise Tolerance in Cystic Fibrosis.. *AnnalsATS, 15*(2), 166.
14. 14. Marcotte JE, Canny GJ, Grisdale R, Desmond K, Corey M, Zinman R, Levison H, Coates AL. Effects of nutritional status on exercise performance in advanced cystic fibrosis. Chest. 1986 Sep;90(3):375-9. doi: 10.1378/chest.90.3.375. PMID: 3743150.
15. 15. Pastré, J., Prévotat, A., Tardif, C., Langlois, C., Duhamel, A., & Wallaert, B. (2014). Determinants of exercise capacity in cystic fibrosis patients with mild-to-moderate lung disease. *BMC pulmonary medicine, 14*(1), 1-8.
16. 16. Hulzebos, Erik H. J.1; Bomhof-Roordink, Hanna1,3; van de Weert-van Leeuwen, Pauline B.2; Twisk, Jos W. R.3; Arets, H. G. M.2; van der Ent, Cornelis K.2; Takken, Tim1 Prediction of Mortality in Adolescents with Cystic Fibrosis, Medicine & Science in Sports & Exercise: November 2014 - Volume 46 - Issue 11 - p 2047-2052 doi: 10.1249/MSS.0000000000000344
17. 17. Ding S., Zhong C. (2020) Exercise and Cystic Fibrosis. In: Xiao J. (eds) Physical Exercise for Human Health. Advances in Experimental Medicine and Biology, vol 1228. Springer, Singapore. https://doi.org/10.1007/978-981-15-1792-1_26
18. 18. Moorcroft AJ, Dodd ME, Morris J, Webb AK. Individualised unsupervised exercise training in adults with cystic fibrosis: a 1 year randomised controlled trial. Thorax. 2004 Dec;59(12):1074-80. doi: 10.1136/thx.2003.015313. PMID: 15563708; PMCID: PMC1746905.
19. 19. Pianosi P, Leblanc J, Almudevar A. Peak oxygen uptake and mortality in children with cystic fibrosis. *Thorax.* Jan 2005;60(1):50-54.

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