Common Names for Disorder:

- CF, Cystic fibrosis of pancreas, Fibrocystic disease of pancreas, Mucoviscidosis

Causes/Etiology of CF:

- Mutations in the CFTR gene cause CF.
- The CFTR gene provides instructions for making a channel that transports chloride ions into and out of cells. Chloride ions help control the movement of water in tissues, which is necessary for the production of thin, freely flowing mucus.
- Mutations in the CFTR gene disrupt the function of the chloride channels, preventing them from regulating the flow of chloride ions and water across cell membranes. As a result, cells that line passageways of the lungs, pancreas, and other organs produce mucus that is unusually thick and sticky which clogs airways and various ducts.

Incidence:

- The most common, life-limiting recessive genetic disorder in Caucasians
- Approx. 30,000 people have CF in the US and 1,000 new cases are diagnosed each year, with males and females affected equally
- Typically diagnosed by the age of 2

Characteristics of CF:

- Very salty-tasting skin, persistent coughing, frequent lung infections including pneumonia or bronchitis, wheezing, poor growth or weight gain in spite of a good appetite, frequent greasy, bulky stools or difficulty with bowel movements, nasal polyps, chronic sinus infections, clubbing or enlargement of the fingertips and toes, rectal prolapse, male infertility
- The median age of death in CF patients is 25, but life expectancy has improved to 46 years with advancements in medicine

Diagnostic Category/Classification: DSM and IDEA

- DSM: Major Depressive Disorder Unspecified with recurrent episodes 296.30(F33.9) and Specific Learning Disorder with impairments in Reading 315.00(F81.0), Writing 315.2(F.81.81), or Math 315.1(F81.2)
- IDEA: Other Health Impairment

Deficits:

- Overall, deficits in verbal and spatial memory, processing speed, and cognitive flexibility

- In preschool, developmental issues related to language skills, understanding causality and assessment of ability to control the environment, and emotional/behavioral regulation development may be delayed. Challenges surrounding meal time are more common in this age group.
- During school age, children with CF may become more attune to negative peer interactions, teasing, and bullying. This may continue to cause problems with emotional/behavioral regulation abilities.
- In adolescence, a time when students are typically thinking about their futures and gaining more independence, children with CF may be thinking about their shortened life span or the course their disease is taking

Strengths

- Resilient as their survival rates continue to improve
- Vigilant as they have to stay on top of their care routine
- Vibrant and cheerful because they're children and they Like to see the best in things

Long Term Developmental Outcomes:

- Poor growth and weight gain
- At greater risk for psychopathology such as depression
- Cystic Fibrosis-related diabetes (CFRD) typically occurs during late adolescence and affects 10-15% of adults with CF – associated with early mortality
- Long-term chronic pain
- End stage lung disease can begin around 25

Assessment Approaches:

- Cognitive Assessment – WISC
- Social/Emotional/Behavioral – BASC, CDI-3, RCMAS, Piers Harris
- Other: The Cystic Fibrosis Impact Questionnaire
    - Semi-structured qualitative interviews are conducted with the CF patient and their caregiver for patients <18. This measure collects data across domains such as activity limitations (physical, social, leisure); functional limitations (school, work); vulnerability/lack of control; emotional impact; treatment burden; and future outlook.
- Clinical Interviews with parents and teachers
- Classroom observations

Interventions and Treatments:

- Airway clearance therapy, inhaled medicines, pancreatic enzyme supplement, individualized fitness plan to help improve energy and lung function, CFTR modulators.
- In schools we can implement the fitness plans prescribed by doctors, encourage all students to wash their hands and use sanitizer to avoid lung infections. Students with

CF should know they can leave the classroom as needed to go to the restroom or to cough, eat in class without asking, and always carry a water bottle.

Contributions of the School Psychologist:

- As a school psychologist we should always be checking in with the parents to see how the child is doing. We may also decide to find the child eligible for an Other Health Impairment and write in the IEP the specifics listed above. We should also include that if there are two students with CF, they are not to be within 6 feet of each other to prevent the spread of infections that only kids with CF contract.
- We should also monitor social/emotional/and behavioral functioning as these students are at greater risk for psychopathology such as depression.
- We can also help teachers understand that these students may have deficits in verbal and spatial memory, processing speed, and cognitive flexibility and consult with them about ways to help the student.

Resources for Parents, Teachers, and Professionals:

- https://www.cff.org/intro-cf/about-cystic-fibrosis
- https://www.cff.org/intro-cf/teachers-guide-cystic-fibrosis
- https://kidshealth.org/en/parents/cf-video.html

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