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Cystic Fibrosis

Patient name: _____ Admission: _____

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I. The client/caregiver can define cystic fibrosis.

- A. It is an inherited disease that affects breathing and digestion.
- B. Cystic fibrosis affects the movement of sodium (salt) into and out of certain cells.
- C. This malfunction results in formation of thick, sticky mucus.
- D. It affects multiple systems of the body, such as
 - 1. Lungs (most common)
 - 2. Pancreas (also common)
 - 3. Gastrointestinal tract
 - 4. Salivary glands
 - 5. Reproductive tract
- E. It is usually diagnosed in early infancy or childhood. Cystic fibrosis rarely occurs in blacks and Asians.

II. The client/caregiver can recognize signs and symptoms.

- A. Coughing or wheezing
- B. Repeated lung infections
- C. Shortness of breath
- D. Greasy, foul-smelling, pale stools
- E. Excessive appetite but poor weight gain and growth
- F. Intestinal blockage in newborns
- G. Rectal prolapse
- H. Salty sweat, tears, and saliva
- I. Infertility
- J. Clubbing of the fingers and toes

III. The client/caregiver can list measures to manage cystic fibrosis.

- A. Prevent infection.
 - 1. Avoid persons with respiratory infections.
 - 2. Take antibiotics as needed.
 - 3. Obtain immunizations as recommended.

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B. Provide respiratory care.

- 1. Be aware of postural drainage and percussion (respiratory care treatments).
- 2. Take oxygen as needed.
- 3. Use nebulizer treatments.
- 4. Take medications as ordered.
- 5. Do breathing exercises.
- 6. Avoid known irritants such as smoke and air pollution.

C. Provide a well-balanced diet.

- 1. Eat food that is high in calories and protein but low in fat.
- 2. Increase fluids.
- 3. Eat salty foods, and add salt to food at the table as tolerated.
- 4. Take pancreatic enzyme supplements. Take supplements with foods especially carbohydrates.
- 5. Discuss the use of vitamins A, D, E, and K and zinc with physician.

D. Do daily exercise with rest periods. Use respiratory treatments before exercise. Modify exercise on warm days, and drink plenty of fluids.

- E. Keep follow-up appointments.
- F. Provide good oral hygiene.

IV. The client/caregiver can list possible complications.

- A. Pneumothorax
- B. Cor pulmonale
- C. Dehydration
- D. Salt depletion
- E. Rectal prolapse
- F. Damage to the eye
- G. Osteoporosis

(Continued)

RESOURCES

March of Dimes
www.marchofdimes.com

Cystic Fibrosis Foundation
800-344-4823
www.cff.org/

National Heart, Lung, and Blood Institute Health Information
Center
301-592-8573
www.nhlbi.nih.gov/health/infoctr/index.htm

Support groups

Dietician

Counseling

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