CASE 4.2
Cystic Fibrosis | Level 2

Hanna Phan

LEARNING OBJECTIVES

1. Identify the presenting signs and symptoms of cystic fibrosis (CF) acute pulmonary exacerbation in a pediatric patient.
2. Apply pharmacokinetic differences noted in pediatric patients with CF in drug therapy selection and dosing.
3. Recommend a pharmacologic regimen for treatment of an acute pulmonary exacerbation in a pediatric patient with CF.
4. Develop a discharge plan for a pediatric patient with CF including patient/caregiver education.
5. Identify a medication-related problem and develop a plan for resolution of the problem.

CHIEF COMPLAINT: "I have been coughing more often than usual and coughing up more 'green-yellow stuff.' I don't want to be sick right before my birthday."

History of Present Illness: An 11-year-old boy came to the CF clinic with his mother for a routine quarterly visit and reported a 1-week history of increased cough and sputum production. The patient stated he has been more tired in the last few days and had to miss school the last 2 days. His mother stated the household has been fighting off a "cold" for the past few weeks, and her son was the last to "catch it." She also reported the patient has had less of an appetite, has been eating fewer snacks, and has difficulty finishing any of his meals while he has not been feeling well. The patient reports no changes in bowel movements with the exception of occasionally floating with greasy stools, and the last incident about 3 days ago. His mother started increased airway clearance therapy with his vest for the past 3 days, which has increased from once daily to twice daily, without much improvement in his symptoms. He denied fever, vomiting, hemoptysis, and abdominal pain or discomfort. The attending physician asks the pharmacist to develop a pharmacologic regimen for his admission for CF acute pulmonary exacerbation.

Review of Systems: Positive for increased cough and sputum production and respiratory distress

Birth History: Born at 38 weeks via NSVD, no complications except meconium ileus that initiated an evaluation for CF

Past Medical History: Patient was diagnosed with CF at 1 month of age through combination of newborn screen, workup, and borderline sweat test, followed with confirmation by genotyping (delta-F508 homozygous). Pancreatic insufficiency secondary to CF; asthma and allergic rhinitis was diagnosed at age 4 years; GERD was diagnosed 3 years ago; and ADHD was recently diagnosed 1 month ago. Patient's baseline FEV_1% predicted is 98. MRSA and *P. aeruginosa* has grown from sputum cultures in the past 12 months.

Past Surgical History: None

Development History: In kindergarten, no history of developmental delay

MEDICATION HISTORY

Medication	Sig	Start Date	End Date	Taking	Authorizing Provider
Albuterol 0.083% inhalation solution	2.5 mg nebulized bid	Since diagnosis		Currently taking	Dr. Lee
Dornase alfa inhalation solution	2.5 mg nebulized daily	Since 3 yr of age		Nonadherent	Dr. Lee
Tobi Podhaler®	112 mg (4 of the 28 mg capsules) inhaled bid—28 days on/off	12/01/2013		Currently taking	Dr. Lee
Lansoprazole capsule	15 mg po daily	02/01/2011		Currently taking	Dr. Lee
Creon® 12,000 capsules	5 caps po with meals, 3 caps po with snacks	Dose adjusted 6 mo ago		Currently taking	Dr. Lee
Cetirizine tablet	10 mg po daily	06/01/2010		Currently taking	Dr. Smith
Fluticasone 44 mcg HFA MDI inhaler	2 puffs inhaled with spacer bid	03/01/2010		Nonadherent	Dr. Lee
Hypertonic saline 7% inhalation solution	4 mL nebulized daily	06/01/2010		Nonadherent	Dr. Lee
AquADEK® MVI capsule	1 cap po daily	Since age 9 yr		Currently taking	Dr. Lee
Azithromycin tablets	250 mg po MWF	Since age 6 yr		Currently taking	Dr. Lee
Ibuprofen tablets	400 mg po bid	Dose last adjusted 6 mo ago		Currently taking	Dr. Lee
Methylphenidate LA capsules	40 mg po daily	1 mo ago		Currently taking	Dr. Smith

Social History: Lives with his mother, father, and his 14-year-old sister; pets at home include a golden retriever (Sunny)

Family History: 2nd cousin on paternal side has CF; maternal history of asthma

Diet: High-calorie diet, BOOST® Kids (two cans/day) supplements for additional calorie intake

Immunizations: Family previously declined all immunizations, but patient caught up with routine childhood schedule in the last 3 months, except annual influenza

Allergies: Sulfamethoxazole–trimethoprim (hives), penicillin (rash, tolerates cephalosporins)

PHYSICAL EXAM

BP 100/60 mm Hg | Pulse 90 beats per min
Temp 37.1°C (temporal) | RR 24 breaths per min
Wt 26 kg (lost 1.5 kg from 4 months ago)
Ht 132 cm | SpO_2 87% on room air

General Appearance: Alert, actively coughing throughout exam

HEENT: Normocephalic, PERRLA, EOMI, moist oral mucosa, nasal polyp on right

Lungs: Crackles and rhonchi on RLL and LUL, no retractions or wheeze

Heart: RRR, no murmurs or irregularities noted

Abdomen: Soft, nontender, nondistended with positive bowel sounds; no hepatosplenomegaly

Musculoskeletal: Minor clubbing of upper extremities

Skin: No exudates or erythema

Neurological: Neurologically appropriate; nonfocal exam; cranial nerves II–XII intact