

START SMART CODING TIP

Cystic Fibrosis

Identify the type of CF and any associated manifestations or acute complications

RELEVANT CODING CLINICS

1Q 2021

October 2026

THE CONDITION

What Is Cystic Fibrosis?

A disorder of the exocrine glands that causes the accumulation of thick, tenacious mucus



PULMONARY

Although cystic fibrosis affects the body in a number of ways, **progressive respiratory insufficiency** is the major cause of illness.

The pulmonary manifestation results in mucus secretions that clog the airways and allow bacteria to multiply. **This can lead to complications such as:**

- Acute and chronic bronchitis
- Bronchiectasis
- Pneumonia
- Atelectasis
- Pneumothorax
- Hemoptysis



DIGESTIVE

In some glands involved in digestion, like the **pancreas**, the thick mucus may become an obstruction, preventing digestive enzymes from reaching the intestines.

Intra-abdominal complications such as meconium ileus, rectal prolapse, inguinal hernia, gallstones, ileocolic intussusception, and gastroesophageal reflux also occur.

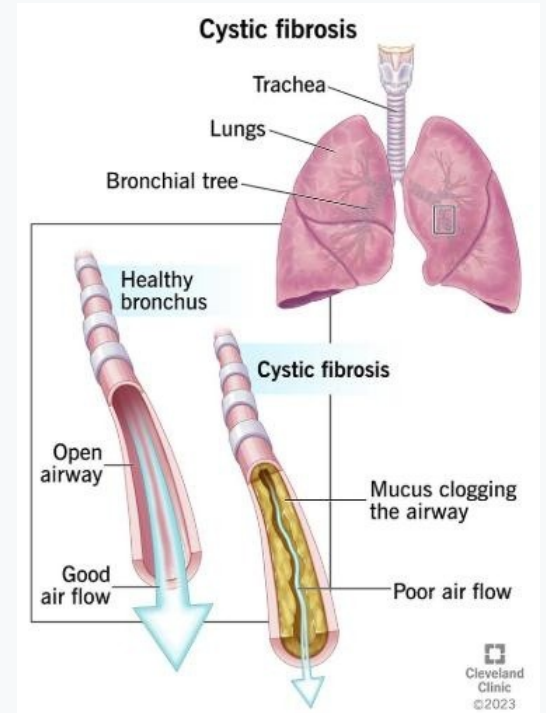


Illustration: Cleveland Clinic ©2023

SITE OF MANIFESTATION

Cystic Fibrosis Coding Tips

Specific ICD-10-CM codes identify the site of manifestation involvement



E84.0

PULMONARY INVOLVEMENT

Cystic fibrosis with pulmonary manifestations



E84.11

MECONIUM ILEUS

Meconium ileus in cystic fibrosis



E84.19

OTHER INTESTINAL MANIFESTATIONS

Cystic fibrosis with other intestinal manifestations



E84.8

OTHER SITE INVOLVEMENT

Cystic fibrosis with other manifestations



MANIFESTATION NOT SPECIFIED

E84.9

E84.9 (Cystic fibrosis, unspecified) should be used if the manifestation is not specified.



TREATMENT

Treatment of cystic fibrosis focuses on managing complications & maintaining adequate nutrition & respiratory function

Read the Notes at E84

Category and code level instructions shape what else gets reported with cystic fibrosis

E84	Cystic fibrosis INCLUDES mucoviscidosis Code also exocrine pancreatic insufficiency (K86.81) <i>DEF: Genetic disorder affecting the respiratory, digestive, and reproductive systems in infants to young adults by disturbing exocrine gland function and causing chronic pulmonary disease with excess mucus production and pancreatic deficiency.</i>	
E84.0	Cystic fibrosis with pulmonary manifestations <i>Use additional code to identify any infectious organism present, such as: Pseudomonas (B96.5)</i> AHA: 2021,1Q,23	MCC HCC
E84.1	Cystic fibrosis with intestinal manifestations	
E84.11	Meconium ileus in cystic fibrosis EXCLUDES1 meconium ileus not due to cystic fibrosis (P76.0)	MCC HCC N
E84.19	Cystic fibrosis with other intestinal manifestations Distal intestinal obstruction syndrome	CC HCC
E84.8	Cystic fibrosis with other manifestations	CC HCC
E84.9	Cystic fibrosis, unspecified	CC HCC



INSTRUCTIONAL NOTES

Code also exocrine pancreatic insufficiency (K86.81)

Use additional code to identify any infectious organism present, such as: Pseudomonas (B96.5)

Excludes1 meconium ileus not due to cystic fibrosis (P76.0)

What to Look For

Documentation should identify the type of CF and any associated manifestations or acute complications



REVIEW THE PROVIDER'S DOCUMENTATION FOR TERMS SUCH AS

- ✓ Pulmonary exacerbation
- ✓ Chronic sinus disease
- ✓ Bronchiectasis
- ✓ Colonization or infection with an identified organism
- ✓ Pancreatic insufficiency



FOLLOW THE INSTRUCTIONAL NOTES

When the documentation supports it, report the related condition too: **pancreatic insufficiency** pairs with the code also note for **K86.81**, and an **identified organism** with E84.0 gets an additional code, such as **B96.5** for Pseudomonas.