

가

CT

cys -

tic fibrosis transmembrane conductance regulator (CFTR)

가 1

가

CT

(Fig. 1).

theophylline 10 μ g/ml

28

40

38

가

CT

(Fig. 2A).

(2, 3).

가

(Fig. 2B),

(Fig. 2C).

CFTR

CT

1

Amylase 60 U/dL

CT

CF transmembrane conductance regula -

tor (CFTR)

가

60

1

50

(subcostal retraction)

1:8

CFTR

Cl -

가

2002 8 8

2002 11 13

693

가

(4).

CT

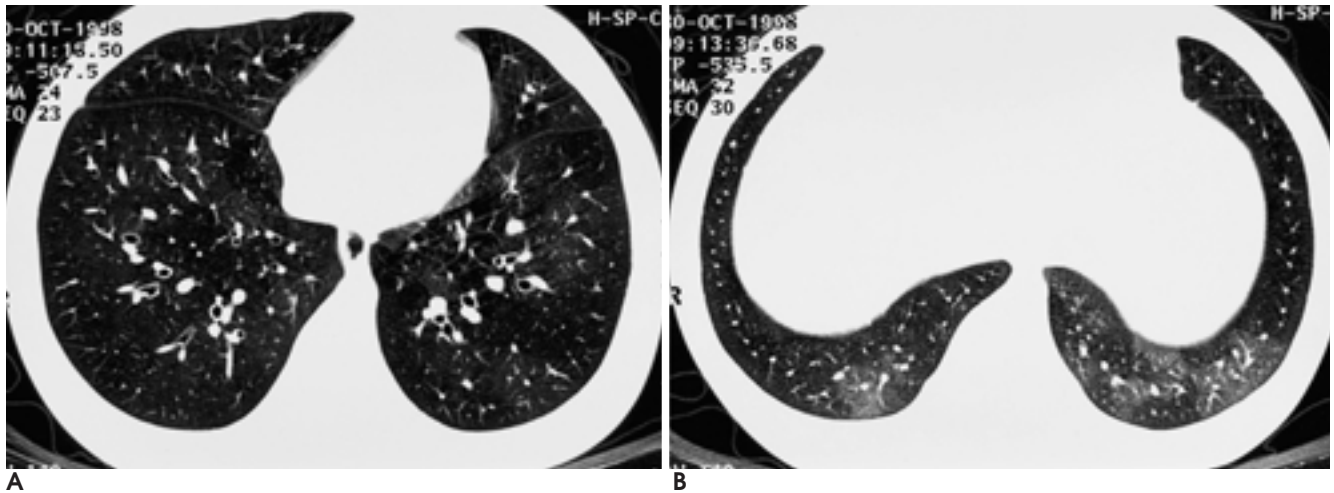


Fig. 1. A 6-year-old boy with cystic fibrosis.

A, B. High-resolution CT scans at the level of the lower lobes show a poorly defined heterogeneity of lung density.

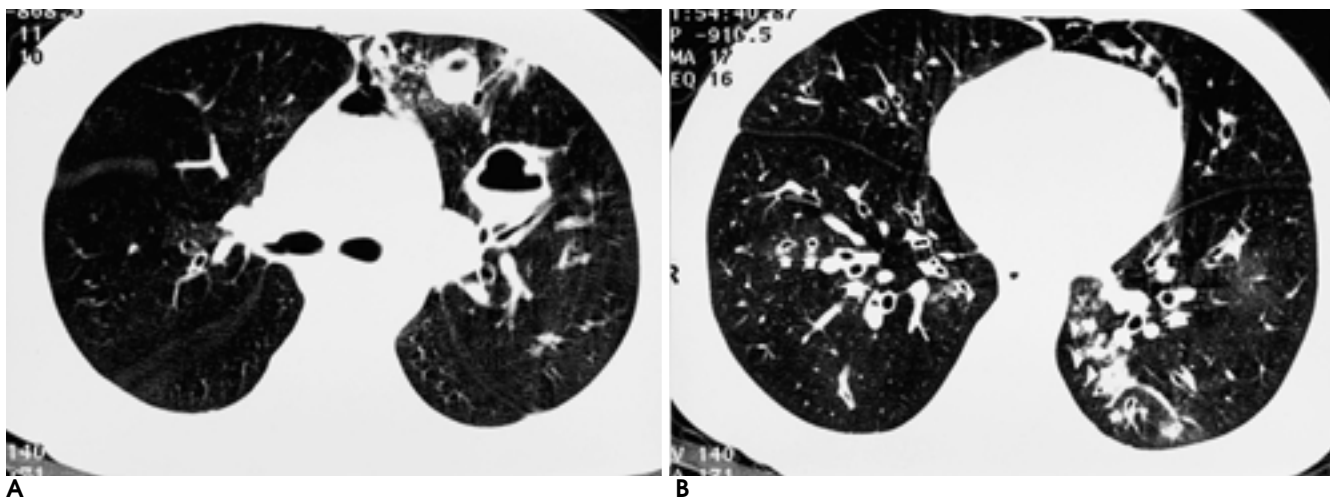
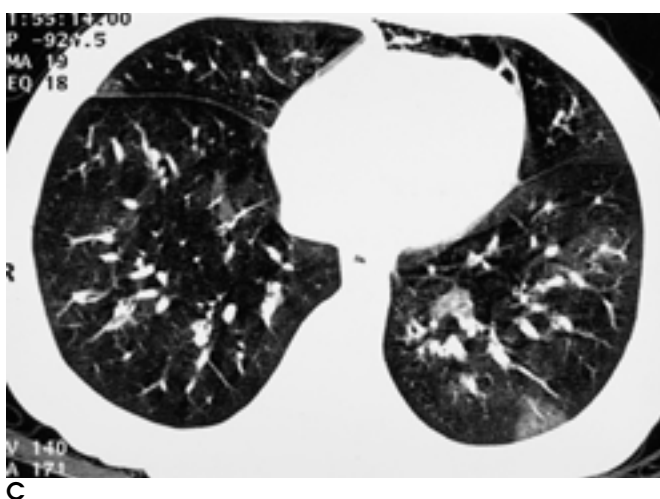


Fig. 2. High-resolution CT scan obtained 34 days after figure 1.

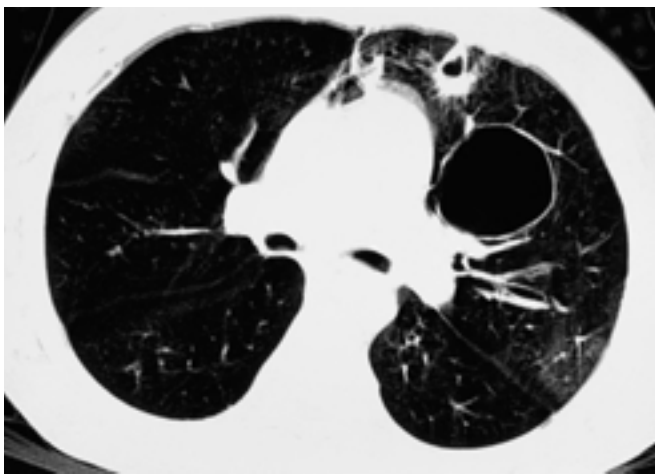
A. High-resolution CT scan at the level of the upper lobes demonstrates multiple cavitory lesions with air-fluid levels.

B. High-resolution CT scan at the level of the lower lung zones demonstrates bilateral bronchiectasis (small arrows), bronchial wall thickening (large arrows) and mucoid impaction (arrow-heads).

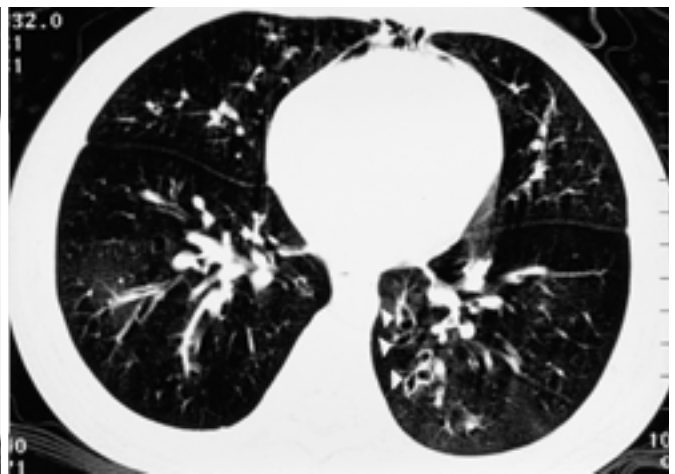
C. High-resolution CT scan at the level of the lower lobes demonstrates mosaic perfusion.



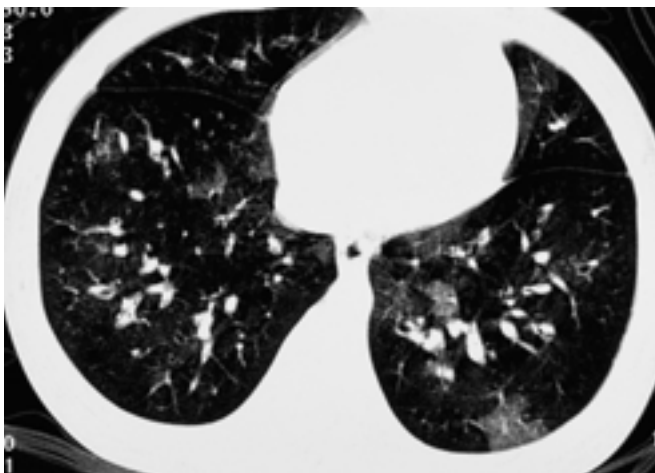
가 , 가 (8). CT CT
(Fig. 3A), (Fig. 3B).
(Fig. 3C).
CT (5), (severity) 가
(scorign)
(5). Santis 가 가
(6), Helbich , 0 - 5
, 50% 가 (5).
가 , 40% Lynch 12 가
CT (heterogeniety) 가 (7).
CT 가
, 가
CT (Fig. 2).
1 Cl- 가 Na+ Cl-
60 mEq/L (2).
95%
(1).
CT



A



B



C

Fig. 3. High-resolution CT scan obtained 70 days after Fig. 2.
A. High-resolution CT scan at the same level of Fig. 2A demonstrates a large thin-walled cavitory lesion with improvement of the air-fluid level.
B. High-resolution CT scan at the same level of Fig. 2B demonstrates improvement of the mucoid impaction and patent bronchial lumens (arrowheads).
C. High-resolution CT scan at the same level of Fig. 2C demonstrates geographic zones of varying lung attenuation representing mosaic perfusion.

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Cystic Fibrosis: Case Report¹

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Cystic fibrosis is a autosomal recessive genetic disease. Among Caucasians, it is the most common cause of pulmonary insufficiency during the first three decades of life. The prevalence of cystic fibrosis varies according to ethnic origin: it is common among Caucasians but rare among Asians. We report a case in which cystic fibrosis with bronchiectasis and hyperaeration was revealed by high-resolution CT, and mutation of the cystic fibrosis conductance transmembrane regulator gene (CFTR) by DNA analysis.

Index words : Lung, CT
Lung, infection
Children, genetic disease

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