The Pulmonary Manifestations
of Fibrocystic Disease of the Pancreas*

PAUL A. di SANT'AGNESE, M.D.
New York, New York

Cystic fibrosis of the pancreas is an hereditary disease of children
classified by the association of pancreatic deficiency and chronic pul-
monary disease. It was briefly recorded in 1936 by Fanconi in Switzer-
land, but its first complete description was given in 1938 by Andersen
in this country. Simultaneously in the same year, two smaller series of
patients were reported by two other groups, one in the United States
and one in Australia. Pancreatic enzymes are usually, but not always
absent at birth; the respiratory involvement has a variable onset from
the age of a few weeks to that of several years. Fibrocystic disease of the
pancreas occurs equally in all subdivisions of the white race; it is rare
in Negroes and has never been found in Mongolians.

It was therefore recognized from its first descriptions that this con-
dition involved both the pancreatic and pulmonary areas. It has recently
been shown that yet another organ system, the sweat glands, is consis-
tently affected in this disease; while less commonly hepatic cirrhosis
may also be present.

The pathologic lesions in the pancreas and the clinical manifesta-
tions of pancreatic deficiency first attracted attention and gave the disease its
name. However, a variable degree of bronchial and pulmonary involve-
ment is a virtually constant feature of the disease, usually dominates the
clinical picture and determines the fate of the patient.

The basic nature of this widespread disturbance is as yet not known.
It has been postulated that mucus secreted in various parts of the body
may be abnormal in its physico-chemical characteristics, thus causing ob-
struction of the pancreatic ducts with consequent dilatation of secretory
acini and parenchymal fibrosis. By the same token, failure to remove
bronchial mucus secretion leads to obstruction of the air passages and
secondary bronchopneumonia. This hypothesis thus gives a reasonable
explanation of the pulmonary and pancreatic lesions, but does not account
satisfactorily for the electrolyte abnormality of sweat.

There is an accumulating body of evidence to suggest that the secretory
activity of many and perhaps all exocrine glands, mucus-producing and
others, is affected in this condition. Therefore, it must be recognized that
so-called cystic fibrosis of the pancreas is in reality a generalized disease
of which the pancreatic and pulmonary lesions are only one expression.

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From the Department of Pediatrics, Columbia University and the Babies Hospital.
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pancreas, and in part by the Corning Fund for Medical Research.
I. Respiratory Involvement

In the 15 years from 1939 to 1954, 292 children with cystic fibrosis of the pancreas have been seen at Babies Hospital. Only 21 were hospitalized in the first five years, while 270, an average of 27 new patients per year, have been admitted in the last decade. The increase in the number of cases in the last 10 years is a reflection both of greater recognition of the disease and of the advent of effective antibiotic agents which greatly prolong the life span. In all of these patients pulmonary involvement of varying degree occurred at some time in the course of the illness. In the average case the respiratory disease was severe. In the whole group it accounted for 90 per cent of the 133 deaths.

The age of onset of the pulmonary component varies greatly and ranges from a few days after birth to the age of several years. In the great majority of cases it has its onset between six months and two years of age.

The general pattern of the respiratory disease is strikingly uniform, except for the degree of severity and the duration of single phases.

Initially, following a mild unresolved acute respiratory infection, a dry, non-productive, hacking cough sets in and persists despite treatment for weeks or months. Although the state of nutrition as a consequence of the pancreatic deficiency is variable, the patient frequently does not appear sick at this time. Temperature is within normal limits, on auscultation of the chest a few scattered rales may be heard and little may be seen in roentgenograms except for an increase in bronchovascular markings, and early signs of obstructive emphysema. Respirations are often wheezing, with a prolonged expiratory phase and a diagnosis of so-called "asthmatic bronchitis" is frequently made. The cough may at times become paroxysmal and simulate pertussis. Antibiotics have a beneficial, but only temporary effect on the as yet mild respiratory involvement.

![FIGURE 1A](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21264/)

**FIGURE 1A**

*Figure 1—A & B: L. N., female, age four years—generalized obstructive emphysema and chronic bronchopneumonia.*
Then suddenly (Table I), following another more severe intercurrent respiratory infection, the failure to remove bronchial secretions leads to widespread and severe bronchial obstruction. The infection, at first mild and perhaps localized to the main divisions of the bronchi, suddenly becomes widespread and severe by invasion of the obstructed air passages down to their smaller subdivisions. The temperature is elevated and the usual laboratory signs of infection are found. Respiratory distress is present to a varying degree and may be severe with cyanosis and air hunger, and the patient is generally quite ill, exhibiting the picture of severe generalized pulmonary infection.
Massive lobar or multilobar atelectasis leading to death may occur at this time.\textsuperscript{11} It was also not infrequent in pre-antibiotic days for patients to succumb at this stage because of a spread of the infecting agents to the blood stream. If the patient survives, this cycle can be repeated again on the occasion of subsequent respiratory infections, thus explaining the characteristic course of these patients, punctuated as it is by relatively sudden attacks of markedly increased bronchial obstruction and secondary infection. Any one of these episodes may be fatal.

The physical examination and the roentgen picture are both quite characteristic at this stage. The chest is barrel-shaped and the note on percussion is markedly tympanic. On auscultation there are scattered rales which clear up temporarily after an episode of paroxysmal coughing. There is gradually increasing clubbing of fingers and toes. On roentgenographic examination the signs of marked obstructive emphysema can be seen (Fig. 1, A & B): Markedly increased antero-posterior diameter of the chest, flattening of diaphragmatic leaves, and a heart which is small in relation to the total thoracic diameter. In addition, the signs of chronic bronchopneumonia are present with a “honey-comb” appearance of the lungs and often marked peribronchial cuffing.

If permanent damage is done to the bronchial wall (Table I), the patients go on to a severe progressive bronchopneumonia, not checked by antibiotics. The x-ray film frequently shows diffuse focal “snowflake” shadows similar to those of disseminated tuberculosis (Fig. 2). Measurements of pulmonary function\textsuperscript{13} at this time show the vital and total lung capacity to be greatly reduced, in association with a marked increase in the volume of residual air and the index of intra-pulmonary mixing. There is profound arterial anoxia with marked increase in the CO\textsubscript{2} tension. The

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure2.png}
\caption{Figure 2—G. D., male, age 13 years: Severe chronic bronchopneumonia (patient died five months later).—Figure 3—R. B., female, age 16 years: No evidence of bronchiectasis on bronchography, despite moderately severe bronchopneumonia of many years duration.}
\end{figure}
distressing picture of pulmonary insufficiency thus develops and eventually leads to death in two or three years on the average. At any time, however, events may take a precipitous turn leading rapidly to fatal termination.

At autopsy in patients who have died of severe chronic bronchial and pulmonary infection, there is almost complete occlusion of the bronchial lumens by extremely thick, tenacious, muco-purulent material. Section of a fresh lung gives a striking picture, with pus welling from every bronchus to form whitish mounds against a fairly homogeneous, pale parenchyma. Multiple lung abscesses are not uncommon and bronchiectasis is a consistent finding.

On nose, throat, sputum cultures and from the lungs at autopsy Staphylococcus aureus hemolyticus is usually recovered. Indeed, the finding of this organism is so consistent as to suggest a possible metabolic factor to explain its striking association with this disease. While staphylococcus was frequently obtained in pure culture in pre-antibiotic days, with the advent of broad-spectrum agents it is not uncommon to find proteus, Pseudomonas aeruginosa, E. coli, and even Candida albicans in the nose and throat cultures or at autopsy.

Fortunately, however, some patients do not have quite as grim an outlook. The pulmonary component of fibrocystic disease of the pancreas...
Vol. XXVII  FIBROCYSTIC DISEASE OF THE PANCREAS

is an intra-bronchial disease, which does not primarily affect the walls of the respiratory passages. Secondary infection may never cause irreversible damage (Fig. 3), leading only to a mild chronic lung disease which is effectively kept in check by the prophylactic administration of broad-spectrum antibiotics (Table I). If patients reach late childhood without irreversible damage being done to their lungs, they seem to improve and the degree of clearing in the roentgen picture is surprising (Fig. 4). Pulmonary function at this time may be virtually normal. What the future holds for such patients we do not know, but suspect that at least many will go on to chronic pulmonary disease as adults.

II. Complications of Severe Respiratory Disease

1. Lobar atelectasis: In about 10 per cent of patients with cystic fibrosis of the pancreas, atelectasis of one or more lobes is present (Fig. 5). In the majority of cases it occurs during the first episode of pulmonary involvement and may lead rapidly to death. In others it accompanies a sudden worsening of long-standing respiratory infection and often represents a terminal complication. In patients who survive, atelectasis generally clears spontaneously within weeks, but at times months after its onset (Fig. 6, A and B). The atelectasis has generally involved the right middle or upper lobes; it has been seen only once in the left lung in the present series.

2. Mediastinal and subcutaneous emphysema: In three patients, massive mediastinal and subcutaneous emphysema developed spontaneously (Fig. 7). In all three it was an expression of a sudden exacerbation of the disease, but in only one did it lead to death. In the other two, admin-

![FIGURE 5—T. D., male, age 3 months: Atelectasis of right upper and middle lobes.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21264/ on 06/21/2017)
istration of antibiotics promptly initiated, both intramuscularly and by inhalation, checked the progress of the pulmonary infection.

3. *Cor pulmonale*: The pressure in the pulmonary vascular bed, high as a result of the chronic pulmonary disease, is markedly and suddenly increased during the repeated attacks of bronchial obstruction which occur during exacerbations of the respiratory infection. This sequence of events at times leads to a rapid dilatation of the heart, visible on x-ray films, and to the clinical signs of circulatory embarrassment. If the patient survives, the heart rapidly returns to its normal size and function, an expression undoubtedly of the intact myocardium in this young age group. Recently in a few patients with long-standing, severe progressive lung disease, there has been found a persistent dilatation of the pulmonary artery on x-ray film (Fig. 8). Polycythemia has generally not been present.

4. *Sudden death from asphyxia*: Sudden death due to coughing up of large amounts of thick, tenacious bronchial secretions which occlude completely the naso-pharynx and cause asphyxia has been seen on many occasions. Indeed it is not an uncommon mode of death in patients debilitated by a chronic wasting illness and unable to cope effectively with an added mechanical insult.

5. *Role of measles and pertussis*: It is not uncommon for the chronic bronchopneumonia in patients with cystic fibrosis of the pancreas to be initiated by measles or whooping cough. This is especially interesting in view of the time-honored observation that rubeola and pertussis are the initial insult leading in some patients to a chronic bronchopneumonia. Whoooping cough is less of a problem at the present time, because of the almost universal prophylactic inoculation of children against this disease. Rubeola, however, frequently leads to severe and at times fatal aggravation of a pre-existing respiratory infection in patients with fibro-

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**FIGURE 6A**

*Figure 6A*: A. H., age two months—Atelectasis of right upper lobe.

**FIGURE 6B**

*Figure 6B*: age 14 months—Clearing of right upper lobe after atelectasis had persisted unchanged for nine months.
cystic disease. Following subsidence of the fever and disappearance of
the rash, bronchial complications may set in.

Patients with cystic fibrosis of the pancreas usually weather without
complications the other common contagious diseases of childhood.

III. Diagnosis

In the average case the combination of symptoms of pancreatic insuffi-
ciency and of chronic pulmonary disease leads to the clinical suspicion
of cystic fibrosis of the pancreas. Definitive diagnosis usually rests on the
demonstration of absence of pancreatic ferments on duodenal drainage. Lung
tuberculosis can usually be reliably ruled out in children by a negative
intradermal tuberculin test.

There are a few patients, however, in whom the lung symptoms pre-
dominate and in whom pancreatic activity is not entirely absent on du-
odenal assay. In such patients the "sweat test" has proved of great
diagnostic assistance as it has been shown that the electrolyte content of
sweat is increased in fibrocystic disease of the pancreas above that of
almost any other disorder. In particular, other conditions characterized
by chronic pulmonary disease have a normal sweat chloride and sodium.

The diagnosis of cystic fibrosis of the pancreas, therefore, should be
based on the following criteria (Table II): 1) Pancreatic insufficiency
which leads to steatorrhea, large, bulky, foul stools and the absence of
pancreatic enzyme activity on duodenal assay. 2) Pulmonary pathology—
characterized by obstructive emphysema and chronic bronchopneumonia.
3) Abnormal sweat with abnormally increased sweat chloride and sodium
and leading to acute salt depletion and at times death in hot weather. 4) Family history—Siblings with the same condition. This last point is not
absolute as there may be no siblings or there may be several other children
who do not have fibrocystic disease. Their presence is therefore of diag-
nostic assistance, but their absence does not rule against the possibility.

A survey of Babies Hospital records shows that in the last 15 years,
chronic bronchopneumonia was entered among the discharge diagnoses in

| TABLE II |
|-------|-------|-------|-------|
| Criteria for Diagnosis | Clinical Manifestations | Test Indicated | Result of Test |
| 1. PANCREATIC INSUFFICIENCY | Steatorrhea Malnutrition | Duodenal Drainage | Absent Pancreatic Enzymes |
| 2. PULMONARY PATHOLOGY | Obstructive Emphysema Chronic Broncho-
                               pneumonia | X-ray film of Chest | --- |
| 3. ABNORMAL SWEAT | Salt Depletion in Hot Weather | Sweat Test | Increased Sweat Electrolytes |
| 4. FAMILY HISTORY | Siblings with Cystic Fibrosis of Pancreas | --- | --- |
244 children in addition to their basic disease. Three out of four of these 244 patients had cystic fibrosis of the pancreas, which accounted for the overwhelming number of cases of chronic lung disease in children. Pulmonary tuberculosis was excluded in all instances by repeated intradermal tuberculin tests.

In a previous study it was shown that fibrocystic disease was the leading cause of lobar atelectasis in infants under one year of age.11

IV. Treatment of the Respiratory Involvement

The treatment of patients with cystic fibrosis of the pancreas is directed to the pancreatic deficiency and the respiratory involvement.

The effects of pancreatic deficiency are well controlled by an appropriate diet,19 with a high protein, low fat content, adequate amounts of the liposoluble vitamins and added pancreatic extracts. Because these patients are prone to massive salt depletion through the sweat,5,6 liberal amounts of salt should be given in hot weather.

Antibiotic treatment is required by patients who have respiratory involvement.14,20,21 As the basic infection is due to Staphylococcus aureus hemolyticus,14 penicillin both by intramuscular injection and by inhalation is given, the latter to be considered a form of topical application. One of the wide-spectrum antibiotics— terramycin, aureomycin, achromycin or tetracyn—is added by mouth. If the cultures reveal the presence of gram-negative bacteria, streptomycin both by intramuscular injection and by inhalation is added. Intensive treatment in the hospital is usually given for a period of not less than seven days and not more than 15 days in a single course. Detailed dosages and routes of administration are listed.

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**FIGURE 7**

*Figure 7—E. F., male, age five years: Atelectasis of right lung with shift of trachea and heart. Marked tissue emphysema overlying right hemithorax and neck.*

**FIGURE 8**

*Figure 8—A. C., male, age seven years: Enlargement of heart and pulmonary artery shadows after years of severe chronic pulmonary disease. The patient had a right upper lobectomy for persistent atelectasis 3 years previously. The operation failed to influence progress of the disease.*
TABLE III
TREATMENT OF RESPIRATORY INVOLVEMENT IN CYSTIC FIBROSIS OF PANCREAS

A. THERAPEUTIC COURSE: 7-15 days

<table>
<thead>
<tr>
<th>AGENT</th>
<th>ROUTE</th>
<th>DOSAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. BROAD-SPECTRUM</td>
<td>orally</td>
<td>50 mgm. per Kg. body weight per 24 hours</td>
</tr>
<tr>
<td>ANTIBIOTIC</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(terramycin, aureomycin, achromycin, or tetracyn)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. PENICILLIN</td>
<td>intra-muscular</td>
<td>400,000 units twice a day</td>
</tr>
<tr>
<td>(procaine)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. STREPTOMYCIN</td>
<td>intra-muscular</td>
<td>1 Gram per 24 hours or 40 mgm. per Kg. body weight</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. PENICILLIN</td>
<td>inhalation**</td>
<td>100,000 units five times a day</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. STREPTOMYCIN</td>
<td>inhalation**</td>
<td>200 mgm. five times a day</td>
</tr>
</tbody>
</table>

B. PROPHYLACTIC ADMINISTRATION:
as long as there is respiratory involvement

1. BROAD SPECTRUM
   ANTIBIOTIC
   (terramycin, aureomycin, achromycin or tetracyn)
   orally
   10-20 mgm. per Kg. body weight per 24 hours

*All dosages for children.
**Penicillin and Streptomycin to be added together to 2 cc. of saline and nebulized (Vaponephrin nebulizer) with oxygen or compressed air flowing at 5 to 6 liters per minute.

in Table III. In recent times erythromycin (40-50 mgm. per Kg. of body weight per 24 hours) in association with gantrisin (0.1 gms. per Kg./24 hours) has been tried with some success.

If patients show evidence of respiratory involvement, continued administration of one of the broad-spectrum antibiotics in prophylactic doses is advisable. This may have to be continued for months or years and it is only when all clinical and radiological signs of pulmonary infection disappear that the drugs can be discontinued.

The problem of resistance of bacteria to antibiotic agents has caused increasing concern in recent years. However, continued administration of these drugs is imperative for the welfare of most patients.

Therapeutic bronchoscopy is contra-indicated because of its difficulty in children, the adverse reactions by which it is often followed and the lack of improvement following the procedure. Lobectomy or partial pneumonectomy for bronchiectasis is clearly not indicated in a patient in whom a diagnosis of cystic fibrosis of the pancreas has been made, because of
the generalized nature of the pulmonary involvement. Lobectomy for persistent atelectasis should be considered if lobar collapse has persisted unaltered for more than one year. Spontaneous expansion of a collapsed lobe may appear even after months (Fig. 6). It should be kept in mind, furthermore, that lobectomy performed for this purpose has generally failed to modify the progressively down-hill course of the disease (Fig. 8). Postural drainage alone is usually ineffective.

In view of the frequency with which measles is followed by serious complications in fibrocystic patients, every effort should be made to prevent or modify this disease by the administration of gamma globulin if exposure is known. If the patient does come down with rubeola, antibiotic treatment is indicated prophylactically.

Immunity against pertussis should be kept up by regular booster injections of the appropriate vaccine.

Finally, administration of digitalis for brief periods may be of assistance in helping the patient overcome transient circulatory embarrassment secondary to an exacerbation of the chronic pulmonary infection.

V. Prognosis

Ten per cent of patients with fibrocystic disease of the pancreas present at birth congenital intestinal obstruction, so-called meconium ileus. About half of these succumb immediately and the other half is operated on successfully. The prognosis thereafter is that of all other children with this condition.

Pancreatic insufficiency with its consequent malnutrition and abnormal stools can be adequately controlled by appropriate dietary measures. Although patients have succumbed not infrequently in the past to acute salt loss in hot weather, this should not happen any longer now that the mechanism of this depletion through the sweat is clearly understood and the physiological needs for sodium and chloride balance in these patients are better known.

The variable degree of bronchial and pulmonary involvement, therefore, determines the fate of the patient. If untreated, the great majority of cases succumb to a progressive chronic bronchopneumonia leading through bronchiectasis and pulmonary insufficiency to death. The average survival subsequent to appearance of the respiratory disease has been two or three years, although any one of the exacerbations of the infection may lead to sudden and fatal increase in bronchial obstruction. The advent of effective antibacterial agents—first penicillin, then aureomycin and terramycin and later other wide-spectrum antibiotics—has greatly improved the outlook for these patients. There are increasing numbers of fibrocystics who with the help of antibiotics weather months and years of relatively mild bronchial and pulmonary infection without permanent damage to their respiratory passages (Table I). These children seem to improve after the age of 10 years, although there are not many who have been able to discontinue entirely the prophylactic administration of antibiotics. This improvement at a certain age may well be due to a purely
mechanical reason, the adverse and dangerous effects of bronchial obstruction being counteracted by the increase in size of these structures. It is not possible to predict the future of these youngsters (the oldest one followed in the present group is 18 years of age), but it would seem logical to assume that at least in some instances chronic lung disease will be present in adult life.

While a varying degree of severity in the involvement of various organ systems is inherent in this disease, there is little evidence that this variability can be influenced by extrinsic factors. Thus controlling the pancreatic deficiency by dietary measures does not seem to influence greatly the pulmonary involvement, except inasmuch as good nutrition increases a patient's ability to cope with infection. Severe and at times fatal lung involvement has often developed in patients in whom the disease has been recognized and who had been kept on an appropriate diet for months or years. Conversely, there are a few patients in whom the respiratory involvement has remained minimal despite the fact that complete pancreatic insufficiency had been present, unrecognized, for years. Another important argument has come to light in recent times with the recognition of children and probably adults in the families of patients with cystic fibrosis of the pancreas, who present the pulmonary manifestations and the abnormal sweat characteristic of this disease in the absence of pancreatic involvement.

In conclusion, while prognosis of a patient with fibrocystic disease of the pancreas is serious and at best guarded, it is not hopeless. Of the 292 children with this condition seen to date at Babies Hospital, 133 have died and 159 are living. Of the latter, 29 are from 10 to 18 years of age, some of them doing well.22 Early diagnosis and administration of antibiotic therapy before the respiratory disease causes permanent damage to the bronchi, in addition to appropriate dietary measures, offers the best hope of tiding these patients over into late childhood when the manifestations of this disorder seem to become milder.

ACKNOWLEDGEMENT

The author wants to thank Mr. Felix A. Battat for his assistance in the survey of the diagnostic files of Babies Hospital.

SUMMARY

Cystic fibrosis of the pancreas, despite its name, is a generalized hereditary disease of children in which the pancreas, the lungs, the sweat glands, and frequently the liver are affected. The basic nature of this widespread disturbances is not known, but it is probable that all exocrine glands are affected in this condition.

A variable degree of bronchial and pulmonary involvement is a virtually constant feature of the disease, usually dominates the clinical picture and determines the fate of the patient. Cystic fibrosis of the pancreas is the commonest cause of chronic non-tuberculous lung disease in the pediatric age group.

Cystic fibrosis of the pancreas differs from other diseases of the lungs
in that the basic phenomenon leading to respiratory involvement is a mechanical one: Failure to remove bronchial mucus, (perhaps abnormal in its physico-chemical characteristics), which results in widespread obstruction and predisposes the patients to secondary infection. Bronchial obstruction may thus be looked upon as the primary and cardinal manifestation of the pulmonary component of fibrocystic disease of the pancreas. The infecting organism is usually Staphylococcus aureus hemolyticus.

Early diagnosis and administration of antibiotics before the respiratory infection causes permanent damage to the bronchi, in addition to appropriate dietary measures, offer the best hope for such patients.

**RESUMEN**

La fibrosis quística del páncreas a pesar de su nombre, es una enfermedad generalizada hereditaria de los niños en los que el páncreas, los pulmones, las glándulas sudoríparas y a frecuentemente el hígado, está afectado. La naturaleza basada de este transtorno extenso no se conoce, pero es probable que esta condición afecte a todas las glándulas exócrinas.

Una característica virtualmente constante de esta enfermedad es un grado variable de compromiso bronquial y pulmonar, que habitualmente domina el cuadro clínico y determina el futuro del enfermo. La fibrosis quística del pancreas difiere de otras enfermedades pulmonares en que el fenómeno básico que conduce al daño respiratorio es mecánico: consistente de la falta de expulsión del moco bronquial, (el que probablemente es anormal en sus características físico-químicas) lo que produce una obstrucción diseminada y puede así considerarse como la manifestación primaria y cardinal de del compromiso pulmonar de la enfermedad fibroquística del páncreas. El germen infectante habitual es generalmente el estafilococo aureo hemolítico.

**RESUME**

La dégénérescence fibro-kystique du pancréas, malgré son nom, est une maladie héréditaire généalisée. Elle frappe les enfants, chez qui on constate l'atteinte du pancréas, des poumons, des glandes sudoripares, et fréquemment du foie. La nature véritable de ces troubles disséminés nous échappe, mais il est probable que toutes les glandes exocrines y participent. Les bronches et les poumons sont atteints d'une façon pratiquement constante, à un degré variable, et cette atteinte domine habituellement le tableau clinique, et tient sous sa dépendance l'évolution.

La dégénérescence fibro-kystique du pancréas diffère des autres affections pulmonaires, car dans ce cas, le phénomène essentiel qui détermine les troubles respiratoires est d'origine mécanique. Il s'agit de l'impossibilité d'évacuer le mucus bronchique dont les composantes physico-chimiques sont peut-être anormales. Le résultat est en une obstruction étendue, qui expose les malades à des infections secondaires. Ainsi l'obstruction bronchique peut être considérée comme la première et principale manifestation de l'atteinte pulmonaire de la dégénérescence fibro-kystique du pancréas. Le microbe qui est à l'origine de l'infection est habituellement un staphylocoque doré hémolytique.
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