Staff Meeting Bulletin Hospitals of the » » » University of Minnesota

Cystic Fibrosis of Pancreas

Volume XVI | Friday, May 25, 1945 | Number 28

STAFF MEETING BULLETIN
HOSPITALS OF THE . . .
UNIVERSITY OF MINNESOTA

Volume XVI | Friday, May 25, 1945 | Number 28

INDEX

Published for the General Staff Meeting each
week during the school year, October to May.

Financed by the Citizens Aid Society,
Alumni and Friends.

William A. O'Brien, M.D.

I.

UNIVERSITY OF MINNESOTA MEDICAL SCHOOL
CALENDAR OF EVENTS
May 28 - June 2, 1945

No. 73

Monday, May 28

9:00 - 10:00 Roentgenology-Medicine Conference; L. G. Rigler, C. J. Watson and Staff; Todd Amphitheater, U. H.

9:00 - 11:00 Obstetrics and Gynecology Conference; J. L. McKelvey and Staff; Interns Quarters, U. H.

12:30 - 1:30 Pathology Seminar; Pulmonary Embolism; J. S. McCartney; 104 I. A.

Tuesday, May 29

9:00 - 10:00 Roentgenology-Pediatrics Conference; L. G. Rigler, I. McQuarrie and Staff; Eustis Amphitheater, U. H.

11:00 - 12:00 Urology Conference; C. D. Creevy and Staff; Main 515 U. H.

12:30 - 1:30 Pathology Conference; Autopsies; Pathology Staff; 104 I. A.

12:30 - 1:30 Physiology-Pharmacology Seminar; The Expression of the Inherited Hormonal Influence in Castrated Mice; Miss Fern Smith; 214 M. H.

4:00 - 5:00 Physiological Pathology of Surgical Diseases; Physiology and Surgery Staffs; Todd Amphitheater, U. H.

4:00 - 5:00 Obstetrics and Gynecology Conference; J. C. McKelvey and Staff; Station 54, U. H.

4:00 - 5:30 Pediatrics Grand Rounds; I. M. Quarrie and Staff; W-205 U. H.

4:30 - 5:30 Ophthalmology Ward Rounds; Erling Hansen and Staff; E-534, U. H.

5:00 - 6:00 Roentgen Diagnosis Conference; Solveigh Bergh and Harry Mixer, 515 U. H.

Wednesday, May 30 -- MEMORIAL DAY

Thursday, May 31

9:00 - 10:00 Medicine Case Presentation; C. J. Watson and Staff; Todd Amphitheater, U. H.

12:30 - 1:30 Physiological Chemistry; Intermediary Metabolism of Carbohydrates; M. F. Utter; 116 M. H.

4:00 - 5:00 Pediatric Journal Club; Review of Current Literature; Staff; W-205 U. H.

4:30 - 5:30 Ophthalmology Ward Rounds; Erling Hansen and Staff; E-534, U. H.

4:30 - 5:30 Roentgenology Seminar; Spondylolisthesis Cases of the Familial Origin; Captain Daniel Fink, M-515, U. H.

Friday, June 1

9:00 - 10:00 Medicine Grand Rounds; C. J. Watson and Staff; Todd Amphitheater, U.H.

10:00 - 12:00 Medicine Ward Rounds; C. J. Watson and Staff; E-214 U. H.

10:30 - 12:30 Otolaryngology Case Studies; L. R. Boies and Staff; Out-Patient Otolaryngology Department, U. H.

11:45 - 1:15 University of Minnesota Hospitals General Staff Meeting; Erythrocyte Protoporphyrin in the Anemias; C. J. Watson; Powell Hall Recreation Room.

1:00 - 2:30 Dermatology and Syphilology; Presentation of Selected Cases of the Week; Henry Michelson and Staff; W-206 U. H.

1:30 - 3:00 Roentgenology-Neurosurgery Conference; H. O. Peterson, W. T. Peyton, and Staff; Todd Amphitheater, U. H.

Saturday, June 2

8:00 - 9:00 Surgery Journal Club; O. H. Wangensteen and Staff; M-515 U. H.

9:00 - 10:00 Pediatrics Grand Rounds; I. McQuarrie and Staff; Eustis Amphitheater, U. H.

9:15 - 10:30 Surgery Roentgenology Conference; O. H. Wangensteen, L. G. Rigler and Staff; Todd Amphitheater, U. H.

9:00 - 10:00 Medicine Case Presentation; C. J. Watson and Staff; M-515 U. H.

10:00 - 12:00 Medicine Ward Rounds; C. J. Watson and Staff; E-221 U. H.

11:30 - 12:30 Anatomy Seminar; The Intrahilar Anatomy of Broncho-Pulmonary Segments; E. A. Boyden; 226 I. A.

II. CYSTIC FIBROSIS OF THE PANCREAS

T. B. Mercer
J. F. Bosma

1. Introduction

The purpose of this presentation is to call attention to a clinical entity which, though adequately described only recently, is encountered with surprising frequency in the practice of pediatrics. Eight typical cases of this disease, cystic fibrosis of the pancreas, have been selected from those observed at the University of Minnesota Hospitals during the past few years to illustrate its known clinical and roentgenological characteristics.

Since one of the principal characteristics of the disorder is the occurrence of an excess of fat in the stools, it is to be distinguished from the more familiar "celiac disease," or ideopathic steatorrhea, with which it was formerly confused. Children suffering from either of these pathological conditions present the same clinical characteristics of bulky stools, enlargement of the abdomen, and signs of nutritional deficiency. Those with cystic fibrosis, however, actually suffer from a generalized disease, the most striking manifestation of which is chronic disease of the respiratory tract.

The coexistence in certain children of severe respiratory infection and a celiac-like picture was first described in 1913 by Garrod and Hurtley[7]. The first observation of pancreatic disease in these children was made in 1919 by Passini[10], who described the typical findings of cystic fibrosis of the pancreas in a child who had had abnormal stools since birth, and who died of broncho-pneumonia at the age of 9 months. A similar pancreatic lesion was found in a sibling of this child who died during its second month of life.

Sporadic reports of this coincidence of steatorrhea with respiratory and pancreatic disease continued to appear in the literature until 1938, when Anderson[1] published a series of 49 pathologically proven cases of cystic fibrosis of the pancreas, 20 of which she had found among the material of 605 autopsies at Babies' Hospital in New York City. Anderson classified these into 3 groups according to the period of their survival. In ten per cent of the children death occurred within a few days of birth due to obstruction of the intestine by an unusually viscid meconium. Sixty per cent of the infants developed respiratory infections, and died before the age of 6 months. The remaining 30 per cent survived for longer periods with a uniform picture of chronic respiratory disease and malnutrition. The surprising incidence of this supposedly rare condition was later verified by Farber[6], reporting the autopsy findings of 87 cases. Smaller series were reported by Snelling[13], Daniel[5], and Shohl[12].

2. Etiology

The etiology of this condition remains obscure. Accumulated evidence is much in favor of its congenital origin in the great majority of children. In many instances the condition is obviously present at birth. It frequently occurs in several siblings. Kennedy and Bagenstoss[9] described a typical patient having cystic fibrosis of the pancreas, 5 of whose 8 siblings died before the age of 8 months with symptoms of chronic cough and diarrhea.

The atrophy and cystic degeneration of the exocrine parenchyma of the pancreas, and the increase in its interstitial fibrous tissue, are found to be proportionate to the degree of occlusion of the duct system of the pancreas, and are probably secondary to it. Anderson interprets this occlusion as an anomaly of development. She believes that the nutritional deficiency incident to the loss of pancreatic function is the cause of the other manifestations of this disease. The severity and chronicity of the respiratory infections are attributed particularly to a deficiency of vitamin A.

It has been found, however, that though the absorption of vitamin A is impaired in these patients, it is not distinctively less than in those children with the classical celiac disease. This latter category of patients has a greater than

normal susceptibility to minor respiratory disease as it occurs in each of these two conditions. Anderson has placed much emphasis upon the hyperkeratoses which she has found in some of these patients as an evidence of a deficiency of vitamin A. Farber was able to find such changes in but a few instances in his larger series of postmortem examinations of these patients.

The most acceptable explanation so far offered is that of Farber[8], who points out that the secretions of all of the mucous-secreting organs of these patients are characteristically viscid, and that the mechanical obstruction caused by these secretion is a prominent factor in the development of pneumonia, and is probably primary to the cystic changes in the pancreas.

The loss of various dietary factors which occurs in steatorrhea of any form is summarized in Table I. Comparative studies of the nutritional loss in ideopathic steatorrhea and that associated with cystic fibrosis of the pancreas have yielded variant results. It may be stated that in general there is an increased fecal content of partially digested lipids, proteins, and lesser quantities of carbohydrates, in this latter entity.

The steatorrhea found in these patients is analogous to that described by Vermeulen, Owens, and Dragstedt[14], and by Coffey and Mann[4] in de-pancreatized dogs. In carefully controlled metabolism experiments it was found that these animals suffered an increased loss of fecal fats, both whole and hydrolized, with a smaller and more variable loss of protein and carbohydrate.

TABLE I.
NUTRITIONAL LOSS IN STEATORRHEA

Ingested Food	Fecal Loss	Clinical Manifestation
Neutral fat . . . *Fatty acids	+	Large, odorous stools
and soaps	+	Protruberant abdomen
		Caloric deficiency
Carbohydrates . . *. . . . .) Significant loss only in exacerbation		
Proteins . . . . . *. . . . .)		
Fat-soluble vitamins		
Vit. A.	+ -	Keratoses (infrequent)
D.	+ -	Osteoporosis, rickets
K.	?)	
E.	?)	
Water-soluble vitamins	) No evidence of deficiency	
Vit. B.	?)	
C.	?)	
Minerals		
Calcium . . Soaps (insoluble)	+	Osteoporosis, rickets
Iron . . . . " "	+	Hypochromic anaemia
Sodium . . . " (soluble))		
Potassium. . " ") Significant loss only in exacerbation.		
Water . . . . .)		

*Digestion impaired in cystic fibrosis of the pancreas, with loss of unsplit fats, carbohydrates, and proteins.

TABLE II

Patient Age at Observ.	Nutrition-State	Steatorrhea			Respiratory Infections	X-ray Findings in Chest	Duodenal Aspir. (Trypsin Activ.)	Sibling History
		Size of Abdomen	Char. of Stools	Fecal Fat (% Dry Wt.)				
Group I								
1. .* 3-4 day.	"Good"	---	---	---	---	(-)	---	2d.pneumonia *(1.) in infancy
Group II								
2. .* 4 mo.	"Well nourished	---	Bulky, odorous	---	Pneumonia at 3 mo.*	+	---	1-d.pneumonia in infancy* 1-normal
Group III								
3. . 5-11 mo.	"Markedly thin"	Large	Bulky, odorous	35	Pneumonia at 5, 8, 10 mo. Chronic cough	+	Absent	1-d.pneumonia in infancy 1-chronic cough 1-celiac disease 1-rickets 3-normal
4. . 3-12 mo.	"Emaciated"	Large	Bulky, odorous	---	Persistent "colds"	-	Absent	3-normal
5. .* 3 yr.	"Markedly undernourished"	Large	Bulky, odorous	38	Pneumonia at 6 mo., 2 yr.* Chronic cough.	+	---	2-d.pneumonia in infancy
6. . 4-5 yr.	"Undernourished"	Large	Bulky, odorous	45.1	"Colds," pneumonia at 2 yr.	+	Absent	2-normal
7. . 5-6 yr.	"Emaciated"	Large	Bulky, odorous	53	Bronchitis at 1½ yr. Chronic cough	+	Absent	2-normal
8. .... 6-7 yr.	"Emaciated"	Large	Bulky, odorous	++++ (stain)	Pneumonia at 6 yr. Chronic cough	+	Absent	No siblings

*Autopsy findings characteristic of cystic fibrosis of the pancreas.

3. Clinical Characteristics

A part of the relevant data on the 8 patients selected to illustrate this discussion is summarized in Table II. The classification suggested by Anderson has been followed in this table.

The clinical picture of the newborn infants having meconium ileus is entirely different from that of older children with cystic fibrosis of the pancreas, though the autopsy findings are similar. These infants have the typical clinical evidences of intestinal obstruction, with abdominal distention, vomiting, and absence of stools. Surgical exploration reveals an elongated mass of adherent gelatinous meconium occluding a portion of the lower intestine, which is collapsed distally to this site. These infants usually expire of meconium peritonitis or pneumonia within the first few days of life.

A summary of the essential data in a typical case illustrating this group follows:

Case 1. The patient was born spontaneously after a normal pregnancy. The birth weight was 8 lb. 7 oz. No meconium was released per rectum. Persistent vomiting developed shortly after birth, and abdominal distention appeared at 12 hours of age. The patient was admitted to University Hospitals on the second day of life.

This patient was the fifth child, and two of its siblings were living and well. One sibling had intestinal obstruction which was released spontaneously on the third day of life, but an alimentary disturbance, with episodes of vomiting, persisted. This child died of pneumonia at the age of 3 months. The fourth sibling also had an intestinal disturbance, with passage of bulky stools. She developed a chronic cough at the age of three months, and died of pneumonia at age of four months.

By digital examination, the rectum was found to have a small lumen. An x-ray film of the abdomen showed the stomach and intestine to be markedly distended. Laparotomy was performed on the third day of life. The distal ileum was found to be distended with viscid meconium, and the colon collapsed to a mean diameter of 5-7 millimeters. Ileostomy was performed. The patient developed persistent respiratory difficulty with cyanosis and died on the fourth day of life.

Autopsy revealed an increased amount of thick mucous in the bronchi and bronchioles. Microscopic study of the pancreas revealed a striking degree of atrophy of the exocrine parenchyma and an apparent increase in the amount of connective tissue. The small and large ducts were dilated and filled with homogeneous eosinophilic material which histologically had the appearance of inspissated secretin.

Anderson's differentiation of the second and third groups of these patients upon the basis of their survival for less or more than six months is more arbitrary. A typical example of the patients in each of these categories follows.

Case 2. This child was admitted to the University Hospitals at the age of four months with a history of chronic cough, poor feeding and weight loss of two weeks' duration.

The patient had one well sibling. A previous child had died at the age of seven months after a similar episode of chronic cough and digestive disturbance. Postmortem examination of this sibling revealed broncho-pneumonia and the characteristic findings of cystic fibrosis of the pancreas. The family history was otherwise non-contributory.

At the time of the patient's admission the physical findings were essentially negative except for the observation of a mild dyspnea and the presence of coarse rales distributed diffusely over the lung fields. During the three weeks of hospital care the chronic cough increased and became paroxysmal in nature. The respiratory difficulty was progressive, with the development of severe inspiratory retraction and per-

sistent cyanosis. Successive chest x-ray films revealed a persistent emphysema, with increasing bronchial markings suggestive of bronchitis or bronchiectasis. Direct laryngoscopy revealed no significant pathology. Sulfadiazine therapy, intravenous aminophyllin and steam were of no benefit. Despite continuous administration of oxygen, the child expired.

Autopsy revealed a large amount of muco-purulent material in the bronchial tree. Microscopic examination of the lungs demonstrated a moderate bronchiectasis and a severe peri-bronchial interstitial pneumonia. The pancreas showed an early typical cystic fibrosis.

Case 8. This patient had been chronically ill for several years. Episodes of diarrhea and recurrent alimentary difficulties had been present since the second year. The patient had been thin and pale and had enlargement of the abdomen for an indefinite period. He had a chronic non-productive cough since the age of 4 years. He was admitted to the hospital at the age of 6 years for therapy of a broncho-pneumonia which appeared as a complication of measles.

There were no siblings of this child. The family history was otherwise non-contributory, except for the mother's admission that her diet had been inadequate during her prognancy.

At the time of admission the boy weighed 34 pounds. His appearance was that of emaciation and chronic illness. He was slightly dyspnic and cyanotic. The chest was emphysematous and coarse rales were heard diffusely within it. The abdomen was protuberant and of a doughy consistency on palpation.

Laboratory studies revealed a persistent leukocytosis and elevation of the sedimentation rate. The stool fat was found to be much increased by qualitative test. Duodenal aspiration revealed an absence of trypsin activity in the pancreatic juice. The glucose tolerance test revealed a low type of curve. X-ray examination of the chest showed moderate emphysema, a marked increase in bronchovascular markings, and evidences of broncho-pneumonia.

The response to sulfadiazine and penicillin therapy during the three months period of hospitalization was very slow. The dyspnea was slowly relieved, and the roentgenological findings of pneumonia gradually resolved. The chronic cough and the finding of rales scattered diffusely throughout the lung fields persisted.

Subsequent observation in the Out-patient service reveal that the condition of this patient is essentially unchanged, despite parenteral vitamin A therapy, oral vitamin B, high protein diet and periodic postural drainage. He remains thin and pale, and has a persistent cough which is productive of a little muco-purulent sputum. He continues to have intermittent episodes of fever, and is confined to bed much of the time.

The presenting complaints made by the parents of these patients may refer primarily to either the nutritional disturbance or to a respiratory infection, although both alimentary and respiratory disease are commonly present. In the case of younger infants, the failure of growth is neglected, and the usual principal complaint is that of chronic cough, or of the symptoms of an intercurrent pneumonia. During later infancy and the preschool years physical regardation is more apparent and these children are brought for medical attention because of marked dwarfism, with their chronic cough occupying a secondary place in the parents' interest.

Data regarding the physical retardation of these patients are recorded in Table II, with comments on the nutritional state of each patient at the time of admission to the hospital. The only patient who had not suffered a marked loss of physique was S.S., who appeared "well-nourished". Further review of her record however reveals that her weight had increased but two pounds above birth weight in her four months of life, despite an apparently adequate diet, and her weight of ten pounds at the time of hospital admission was four pounds less than the average for her age.

Much of the reason for the failure of

these children to grow normally is revealed in the column "Steatorrhea" of Table II. The observations of distinctively bulky and odorous stools in all of these patients and that of abdominal enlargement in six of the seven would lead one to suspect the presence of steatorrhea. This suspicion was verified quantitatively in four patients and qualitatively in one by the observation of a greater than normal loss of lipids in the feces.

These children are peculiarly susceptible to chronic respiratory infection, with progressive secondary emphysema and bronchiectasis. These infections are highly resistant to therapy, and a febrile state may persist for months, during which the patient coughs up quantities of thick mucopurulent sputum. The accumulation of the typical viscid bronchial secretions often causes episodes of dyspnea and wheezing, as reported in six of our eight patients. In two of these the mistaken diagnosis of pertussis had been made upon the basis of these symptoms. The respiratory obstruction and the increasing emphysema may combine to cause a persistent respiratory deficiency with cyanosis.

Episodes of bronchopneumonia may be superimposed upon this chronic respiratory disease, and are usually responsible for the death of the patient.

A great deal of emphasis has been placed upon the family history as an aid in the diagnosis of this condition. With the exception of a few rare instances in which a maternal intolerance to fat was suspected, the only significant coincidences found by others were in the occurrence of like disease in the siblings of the patients. This coincidence was found in siblings of four of the eight patients described here.

We summarize the clinical picture as one consisting of a chronic intestinal disturbance, with secondary growth failure and an associated persistent respiratory infection. We would like to emphasize the commonness of each of these complaints in pediatric practice. In clinical examination, without the aid of laboratory procedures, the presence of this systemic entity of fibrocystic disease could easily go undetected.

4. Laboratory Examinations.

A. Evidence of steatorrhea.

The presence of steatorrhea is confirmed by the finding of an increased amount of lipid in the dried stool. The most valid data in this determination is that obtained on the total stool collected over a period of several days, during which the patient is on a normal diet.

The distinctive proportion of neutral fat in the stools, which has often been thought diagnostic of cystic fibrosis of the pancreas, in distinction from iedopathic steatorrhea, has not been prominent in our patients. Neutral fat comprised but 28 per cent of the total fecal lipid in the patient E.S. It is the conclusion of Anderson[1] that the proportion of neutral fat in the total fecal lipid is highly variable, and cannot be used as a basis for differentiation of ideopathic steatorrhea from that associated with pancreatic disease.

B. Evidence of pancreatic deficiency.

The single laboratory test, other than roentgenological studies, which provides a valid basis for the diagnosis of cystic fibrosis of the pancreas in the living patient is the analysis of the pancreatic juice, as obtained by duodenal aspiration. Adequate precaution must be taken in the procedure of obtaining this material, to insure that it is obtained from the duodenum. The aspiration tube should be perforated only within a few inches of its tip, and it must be shown by fluoroscopy to be in the second prtion of the duodenum. The most accurate index of the functional capacity of the pancreas is obtained when the organ is stimulated, as by the instillation of a small amount of hydrochloric acid into the tube.

The element of pancreatic secretion which is most commonly analyzed is its content of trypsin. This is evaluated by the lysis of a gelatin substrate by successive dilutions of the aspiration

material, after the method of Anderson.

c. Evidence of infection.

The white blood count and sedimentation rate are persistently elevated in these patients, manifesting the chronicity of their respiratory infections. Cultures of the sputum during the acute episodes of bronchopneumonia yield staphylococcus aureus with great consistency. This organism was found in six of the seven patients from whom throat cultures were taken.

5. Pathology

The essential pathology according to Anderson[1] is in the pancreas and the pulmonary changes are secondary, due to deficiency of vitamin A. The systemic nature of this disease, however, is very striking, and the pancreatic findings are only part of the whole picture. Farber[6], in recent review of the autopsy findings on eighty-seven cases of pancreatic fibrocystic disease, describes the uniform presence of a homogeneous sticky material of the mucous glands of the trachea, bronchi, bile ducts, salivary glands, and pancreas. He believes the inspissation of this material in the ducts causes the characteristic secondary changes.

A. Pancreas.

In gross examination, lobulation of the pancreas due to contraction of fibrosed areas below the surface of the gland was frequently found. Dilatation of the ducts and plugging of their lumina with the inspissated material previously described was common. Dilatation of the small ducts gave the pancreas a cystic appearance. Atrophy of the exocrine parenchyma of the organ was usually present, with an increase in its interstitial fibrosis. The degree of involvement of the pancreas varied greatly. In the minimal cases the only findings may be occlusion of the ducts and acini with no atrophy. It should be noted, however, that even these minimal cases usually had shown no evidence of trypsin in the duodenal contents. The islet of Langerhans are not involved.

B. Liver

The liver was often larger than normal, and contained varying amounts of fat. The degree of fatty degeneration seemed to vary directly with the amount of pancreatic destruction.

C. Respiratory tract

In each of the forty-nine cases in Anderson's original series there were changes in the respiratory tract. There was mild tubular dilatation of the small bronchi and bronchioles, with plugging of their lumina with tenacious purulent material. In some cases these bronchiectatic changes were very marked. Often there were multiple abscesses arising in the small bronchi, sometimes spreading to form multi-locular abscesses. Fresh lobular pneumonia arising in the parenchyma was not uncommon. The larger bronchi and the trachea also contained sticky muco-purulent material. At times the appearance suggested that the patient must have experienced suffocation due to the abundance of the exudate.

6. Roentgenological Findings.

A. Small intestine.

According to Golden[8], the small intestine of newborn infants frequently presents a different appearance on examination with barium tothat found in older children. Apparently due to a lack of nervous reflex control, areas of spasm alternating with areas of dilatation are present. A "segmentation" effect results; other terms used in respect to this appearance are "puddling" or "sausage" formation.

This effect is greatly accentuated in nutritional disturbances. Vitamin deficiency, especially lack of the vitamin B complex, apparently plays an important part in the production of the intestinal changes seen in steatorrhea. In the nutritional deficiencies, there is considerable thickening of the mucosal folds with elimination of the normal fine mucosal pattern. There may be merging of the folds due to the thickening, result-

ing in a smooth appearance of the mucosa. This effect is most noticeable in the jejunum because the folds are normally more prominent here than in the ileum.

Hypomotility is another prominent feature. Normally the barium passes thru the small bowel into the cecum in 3 to 6 hours. In many cases of steatorrhea, passage is delayed for as long as 8 or 10 hours.

The above changes are not specific for the pancreatic type of steatorrhea. They are present also in ideopathic steatorrhea.

B. Chest

The appearance of the disease in the chest roentgenogram is somewhat characteristic, although it may be simulated by other conditions when still in its early form. Certain features are strongly suggestive of the presence of the disease.

The distribution is rather uniform and widespread, involving all lobes of both lungs equally[2]. Concentration of the lesions is greatest about the hilar regions with loss of the normal vascular markings. Mottled shadows fade out toward the periphery where they are gradually lost in the areas of peripheral emphysema.

Frequent acute episodes of bronchopneumonia occur which leave permanent damage to the lungs. Bronchopneumonia is shown on the films by consolidations of various sizes found in any part of the chest.

Bronchiectasis of an atypical type is one of the most characteristic features. It is very diffuse and involves the upper lung fields as well as the bases. A final diagnosis of bronchiectasis can only be made by lipiodol examination. The importance of the frequency of occurrence of bronchiectasis in this disease was stressed by Anderson. Of 8 children who died of bronchiectasis in the first year of life, 7 had definite cystic fibrosis of the pancreas.

Atelectasis is a frequent occurrence as a result of complete obstruction of a bronchus. A local area of homogeneous density with retraction of the mediastinum or elevation of the diaphragm is sometimes seen on the film.

Emphysema is quite marked and is much more apparent in the peripheral areas. The rib interspaces are widened and the diaphragm on both sides is depressed. Emphysema is due to partial bronchial obstruction.

C. Differential Diagnosis

The disease can be distinguished from bronchopneumonia by its lack of definite consolidation, by its diffuse character, and its extreme chronicity. Bronchiectasis in the ordinary form has a tendency to distribute itself in the lower lung fields. Bronchial asthma resembles this disease very closely in the chest film. The picture as seen in cystic fibrosis of the pancreas is distinguished by greater relative density of the hilar shadows.

In summary, the pulmonary changes in cystic fibrosis of the pancreas are fairly characteristic. The disease is diffuse, is of chronic nature, and appears to be progressive. Mottled densities are seen in both hilar regions extending in streaked fashion into the peripheral areas of emphysema.

7. Therapy

The therapy of this disease has so far been disappointing. The intestinal symptoms and the nutritional loss are improved by a diet low in fat and high in protein. The chronic pulmonary condition is benefited by periodic postural drainage. With the use of penicillin therapy these children now more often survive the intercurrent episodes of pneumonia to which they often succumbed. Upon the basis of Anderson's emphasis upon the importance of vitamin A in the etiology of the respiratory disease, therapy with massive doses of parenteral vitamin A has been attempted, but is not of specific value.

The greatest known age of survival at the present time is fourteen and a half years.

8. Summary

Cystic fibrosis of the pancreas is a systemic disease which is more prevalent than is generally recognized. It manifests itself clinically by steatorrhea, dwarfism, and chronic respiratory infection. Pathologically it is characterized by obstruction of the ducts of the pancreas, salivary glands, and of the trachea and bronchi, with resulting secondary changes of a serious nature. Its etiology has not been proven.

The clinician would be well advised to investigate any case in which steatorrhea and respiratory symptoms are present, with this diagnosis in mind. The roentgenologist can be of great assistance by recognizing the characteristic findings when these are present in the x-ray film.

References

1. Anderson, D. H.
Am.J.Dis.Child. 56:344, '41.

2. Attwood, C. J. and Sargent, W. H.
Radiol. 39:417, '42.

3. Baylin, G. J.
Am.J.Roent. and Radium Ther., 52:303, '44.

4. Coffey, R. J. and Mann, F. C.
Am.J.Digest Dis. 7:41, '40.

5. Daniel, W. A.
Am.J.Dis.Child. 64:33, '42.

6. Farber, S.
Arch. Path. 37:238, '44.

7. Garrod, A. F. and Hurtley, W. H.
Quart. J.Med. 6:242, '13.

8. Golden, R.
Radiologic Examination of the Small Intestine.
J. B. Lippincott Co., '45.

9. Kennedy, R. L. J., and Baggenstoss, A. H.
Proc. Staff Meet. Mayo Clin. 18:487, '43.

10. Passini, F.
Deutsche med. Wchnschr. 45:851, '19.

11. Raub, S., Litvak, A. W. and Steiner, M. J.
J. Ped. 14:462, '39.

12. Shohl, A. T., May, C. D. and Schwachman, H.
J. Ped. 23:267, '43.

13. Snelling, C. E.
Arch. Dis. Child. 17:220, '42.

14. Vermeulen, C., Owens, F. M., and Dragstedt, L. R.
Am.J.Physiol. 138:792, '43.

- - -

III. GOSSIP

Last week the course in Ward Administration for Head Nurses at the Center for Continuation Study was given by Deborah MacLurg Jensen, of Washington University, St. Louis. She and her husband, Julius Jensen, Assistant Professor in Medicine, Washington University have written a book on Nursing in Clinical Medicine. Associate Editors are Richard S. Weiss, and Adolph H. Conrad, Dermatologists of Washington University, with diets by Howard A. Rusk. The disease is described first and then the nursing care. The Jensens are former Minnesotans who have gone far working together. This week the Center is presenting a Workshop in Hospital Administration to 30 administrators from the midwest. Studies are being conducted under the direction of Dr. Charles E. Prall, former Dean of Education, University of Pittsburgh, who is engaged in a three year study in educational practices in the training of hospital personnel for the College of Hospital Administrators, and the American Hospital Association. A grant of $90,000 has been made by the Kellogg Foundation for the study. Present workshop is one of the demonstrations in the study. Genevieve K. Bixler, Business Administration Consultant, from Mt. Ranier, Maryland, will hold a two day conference for Administrators and Assistant Administrators of Nursing Schools, May 28 and 29. June 11 through 16 there will be 2 courses at the Center for physicians. One group will study the Management of Infantile Paralysis, the second will be concerned with the Medical Aspects of Rehabilitation. No other medical or hospital courses are scheduled for June as the various workshops will move in at that time. Plans for next year include development of a larger number of medical courses to meet the greater demands...Commencement time is here and the good people of Morris, Chaska, Houston, and Gilbert have invited me to address their graduates. June 26 will be Medical Field Day at the University of Wisconsin Medical School. Students will present papers and demonstrations. Visiting speaker will address student body and faculty on a general subject after lunch. The afternoon will be given over to athletic events. In the evening there will be a special graduating ceremony for the medical students. This seems like a good idea and I am wondering if a similar project might not be developed at our University by the students....June 5 will be observed as Mayo Memorial Day. Surgeon General of the Navy will speak, the new President of the University is scheduled to appear. The importance of the Mayo Memorial project is growing as the future development of the University begins to take form. I am looking forward to June 21 when the first formal graduating exercise of the students in the Course for Funeral Directors and Embalmers will be held in the Auditorium of Natural History Museum. The University of Minnesota was the first to develop a course in this field. Once referred to in "Mercury" as the higher education at Minnesota, our relationships with this group have been developed on a sound basis through the years. Funeral Directors and Embalmers who have identified themselves with the University of Minnesota in their training program are among the first to encourage and develop the practice of general postmortem examinations. June 12 I will visit Glenwood, Minnesota to speak at the County Public Health meeting. Glenwood is the home of Dina Bremness, President of the Minnesota Hospital Association, whose little 26 bed hospital is a model of its kind. She has been grand prize winner in the small hospital group in the field of public relations in the national scene for years. She is quiet, efficient and pleasant, and has the universal support of her people. Another unusual Minnesota hospital is located at Breckenridge where I spoke recently at a cadet nurse induction ceremony. A group of the town's leading citizens met me at the train, to be sure that I understood how important was the place that the hospital occupied in the community Minnesota Cancer Society announces that upwards of $50,000 was collected in the state drive this year. Money will be devoted to education, research, care of the indigent, and future development of detection services. National drive to date has raised only 40% of desired amount. It is apparent that people give to such causes best when children are involved. It is interesting to note that more children died of malignant disease than of infantile paralysis in recent years. The Society thanks all who contributed in any way to the cause.