

AN ANALYSIS OF 46 AUTOPSY CASES OF JAPANESE
CHILDREN, SHOWING FIBROCYSTIC CHANGES OF
THE PANCREAS AND ONE CASE OF FIBRO-
CYSTIC DISEASE OF THE PANCREAS*

BY

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ABSTRACT

More than twenty cases of meconium ileus or fibrocystic disease of the pancreas in Japanese children were reported, only four of which were examined for sweat electrolytes.

Two cases out of the four, including our case, showed abnormally high sweat electrolyte levels with a reasonable ratio of sodium to chloride.

The majority of cases reported, in Japan, as fibrocystic disease of the pancreas was diagnosed on the basis of clinical symptoms of the disease and on the findings of fibrocystic changes of the pancreas.

According to the analysis of the 46 cases, selected from 422 autopsy cases, fibrocystic changes of the pancreas were found. It is known that fibrocystic changes of the pancreas is not a specific change in fibrocystic disease, but is the result of miscellaneous unknown causes.

Therefore, the results of the sweat test should be recorded, besides the histological findings of the pancreas, in cases reported as fibrocystic disease of the pancreas in Oriental races.

A case of fibrocystic disease of the pancreas diagnosed by clinical symptoms, findings of the chest X-ray, and the results of abnormally high sweat electrolyte levels with reasonable ratio of sodium to chloride, was also reported.

INTRODUCTION

Although it is believed that fibrocystic disease of the pancreas is never seen in Oriental races¹⁾, more than 20 cases of meconium ileus or fibrocystic disease of the pancreas in Japanese children, have recently been reported²⁾.

The majority of cases reported as meconium ileus or fibrocystic disease of the pancreas were diagnosed from clinical symptoms and fibrocystic findings of the pancreas, without sweat test results.

Fibrocystic changes of the pancreas were seen in high incidence in autopsy cases of Japanese infants and children³⁾, but it should be emphasized

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that fibrocystic changes of the pancreas cannot be considered a diagnostic finding of this disease.

Purpose of this paper is to analyze the 46 cases which showed fibrocystic findings and to report one case which was diagnosed as fibrocystic disease of the pancreas by clinical symptoms, findings of chest X-ray and the results of abnormally high sweat electrolytes.

AN ANALYSIS OF THE 46 CASES

These 46 cases were selected from 422 autopsy cases, because they showed histological findings of the pancreas in fibrocystic disease.

The age distribution of these 46 cases is shown in Figure 1. The majority was less than 30 days old. The ratio of male and female was 26 to 20.

AGE DISTRIBUTION of 46 CASES

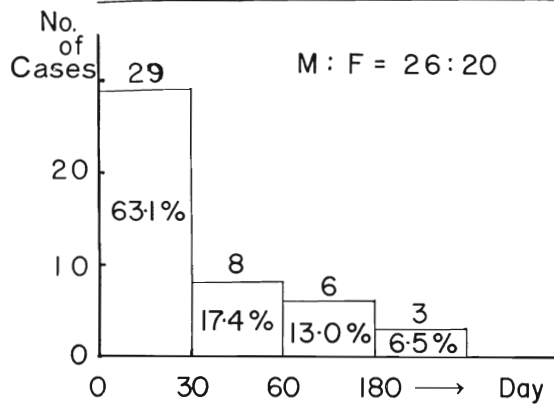


Figure 1.

BODY WEIGHT at BIRTH (42 Cases)

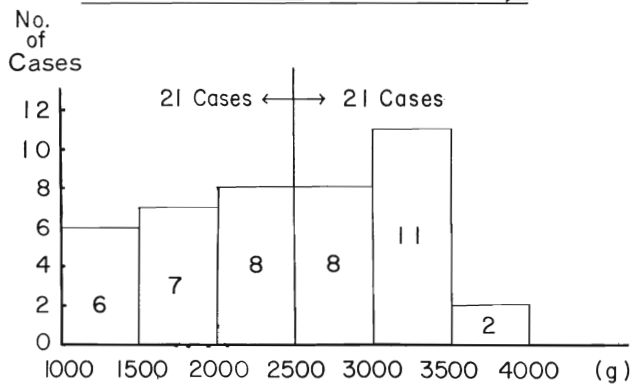


Figure 2.

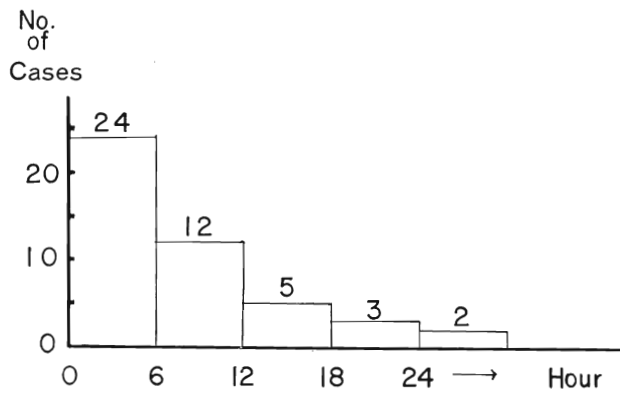
LENGTH of TIME after DEATH at AUTOPSY

Figure 3.

Body weight is shown in Figure 2. 21 out of the 42 recorded cases were premature babies of less than 2500 gm.

Length of time after death before autopsy is shown in Figure 3. Autopsies were performed within 12 hours after death in the majority of cases, while two cases were more than 24 hours after death.

Direct cause of death, one from each case, was listed and classified as shown in Table 1. Complications of the respiratory system including pneumonia were seen in 23 cases.

Classification of the main patho-anatomical diagnosis after autopsy is shown in Table 2. Although there were only 46 cases, a total of 55 is listed because some cases had more than two important diseases.

21 had diseases related to the digestive system, 9 to the respiratory system out of which three cases were listed as pneumonia since it was a primary disease and not a complication.

Histological findings of lung, liver and kidney were re-checked.

Table 3 shows that more than half of the 46 cases had pneumonitis and pulmonary hemorrhage.

Table 4 shows that 29 or 63% of the 46 cases had degeneration or necrosis of the liver. Congestion of the liver was seen in a high percentage of cases.

Table 5 shows the findings of the kidney. Important findings are: congestion—32.6%, nephrosis—23.9%, and degeneration—15.4%.

CASE REPORT

This 2-month-old baby girl (body weight 3,980 gm.) was admitted to the hospital, because of her persistent stridor, tachypnea and fever. The symp-

toms were noted from the time the infant was a few days old. Pregnancy was uncomplicated.

Table 1. Cause of death (46 cases)

Respiratory	23
Pneumonia	16
Others	7
Hepatic	7
Cirrhosis	3
Hepatitis	3
Jaundice	1
Cardiovascular	3
Nervous system	3
Malignant tumor	2
Others	8
Tot.	46

Table 2. Patho-anatomical diagnosis

Gastrointestinal	21
Atresia or Stenosis	5
Congenital biliary atresia	4
Chronic ulcerative colitis	4
Hepatitis	3
Ileus	2
Jaundice	2
Tracheoesophageal fistula	1
Respiratory	9
Pulmonary dystelectasis	3
Pneumonitis	3
Cyst	1
Abnormal vein	1
Abscess	1
Cardiovascular	8
Nervous system	3
Others	14
Prematurity	5
Multiple malformation	2
Sepsis	2
Carcinoma	2
Omphalocele	1
Mongolism	1
Pseudohermaphroditism	1
Tot.	55

Table 3. Findings of the lung (46 cases)

Findings	No. of cases	(%)
Pneumonitis	24	(52.2)
Hemorrhage	24	(52.2)
Aspiration	16	(34.8)
Congestion	11	(23.9)
Tracheo-bronchitis	8	(17.8)
Dyslectasis	7	(15.5)
Atelectasis	6	(13.0)
Edema	4	(8.7)
Mucus in bronchus etc. etc.	4	(8.7)

Table 4. Findings of the liver (46 cases)

Findings	No. of cases	(%)
Degeneration or Necrosis	29	(63.0)
Fatty	19	
Vacuolar	3	
Irregular	2	
Necrosis	5	
Congestion	23	(50.0)
Hematopoiesis	17	(36.9)
Jaundice	14	(30.5)
Kupffer stellate cell change	9	(19.6)
Inflammatory cell infiltration	8	(17.4)
Bile cast or stasis	8	(17.4)
Hemosiderosis	6	(13.6)
Fibrosis	6	(13.6)
etc.		

Table 5. Findings of the kidney (46 cases)

Findings	No. of cases	(%)
Congestion	15	(32.6)
Nephrosis	11	(23.9)
Biliary	5	
Lower nephron	2	
Glomerulotubular	2	
Hydro-	2	
Degeneration	7	(15.4)
Fatty	4	
Hyaline	2	
Granular	1	
Claudy swelling	4	(8.7)
Cast formation	3	(6.5)
Nephritis	2	(4.4)
Calcium deposition	2	(4.4)
etc.		

Family history: No fibrocystic disease of the pancreas in any relatives. Parents are healthy.

Figure 4 shows emaciation and remarkably distended abdomen.

Physical examination revealed respiratory murmurs in chest.

Figure 5 shows shadow in the upper right field of lung.

Slight steatorrhea was noted. Roentgen film test for fecal trypsin was slightly abnormal, but the result of enzyme activity in the duodenal fluid was within normal limits.

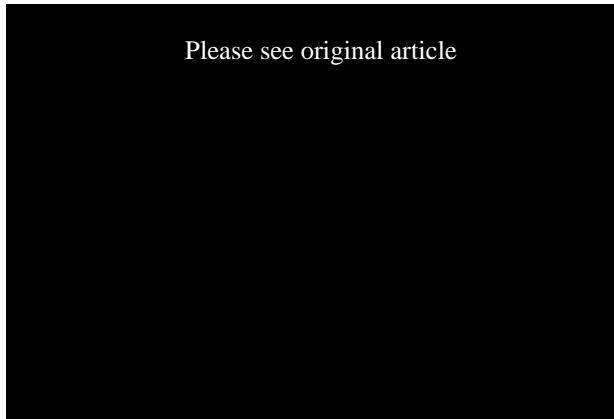


Figure 4. Emaciation and remarkably distended abdomen



Figure 5. Chest X-ray findings

Liver function was slightly disturbed, S-GOT 64 U., S-GPT 34 U.

Results of sweat test: Sweat sodium level 88 mEq/L, sweat chloride level 94 mEq/L. Ratio of sodium to chloride about 0.88.

This female infant is still living. She has respiratory difficulty, so naturally no histological examination of the pancreas has been made.

DISCUSSION

More than 20 cases of meconium ileus or fibrocystic disease of the pancreas in Japanese children, were reported. Only four cases, including our case, were given the sweat test for diagnosis of the disease.

The majority of cases reported in Japan were diagnosed by clinical symptoms similar to that of fibrocystic disease of the pancreas and by the fibrocystic findings of the pancreas after autopsy.

Positive sweat test is extremely reliable in the diagnosis of the disease and for that reason our case reported here, is a most important one.

Sweat sodium levels of the four cases were 210, 74.1, 112 and 88 mEq/L. Chloride were 82, 33.5, 144 and 94 mEq/L. The ratios of sodium to chloride were 2.65, 2.21, 0.78 and 0.88 respectively. Ratios in the first two cases are higher than in the latter two.

It is not clear why the ratio of sodium to chloride in these two cases is so high^{4,5)}, but Kato's case⁶⁾ and ours show a reasonable ratio of sodium to chloride when compared with the results of sweat tests in fibrocystic disease, reported among Caucasian children.

These two cases are very important, because the results of the sweat test were included in the diagnosis of fibrocystic disease of the pancreas in Japanese children. According to the diagnostic criteria presented by di'Sant Agnese⁷⁾, there are no doubtful points in this method of diagnosis.

Typical fibrocystic findings of the pancreas, after autopsy, were seen in Kato's case. Our case is still living, in poor condition, so we have not examined the pancreas yet.

Fibrocystic changes of the pancreas from autopsy cases of Japanese children seem very prevalent but these findings should not be considered as diagnostic findings.

It would be interesting to know why fibrocystic changes of the pancreas are in such high incidence, even among infants. Out of 422 autopsy cases of infants and children less than five years of age, 46 cases showed fibrocystic findings of the pancreas.

Although the actual causes of the fibrocystic changes of the pancreas were not clarified by the analysis of these 46 cases, we know, however, that the fibrocystic changes were the results of miscellaneous unknown causes and not specifically by fibrocystic disease of the pancreas.

CONCLUSION

1. Out of 422 autopsy cases of Japanese infants and children less than five years of age, 46 cases showing fibrocystic changes of the pancreas were selected and analyzed.

2. No specific cause for fibrocystic changes of the pancreas was found. The changes resulted from miscellaneous unknown causes and not, specifically, from fibrocystic disease of the pancreas.

3. A case of fibrocystic disease of the pancreas in a Japanese infant was presented.

4. Two cases of fibrocystic disease, including ours, were diagnosed using the results of abnormally high sweat electrolytes with reasonable ratio of sodium to chloride. These two are epoch making.

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