FIBROCYSTIC CHANGES OF THE PANCREAS IN JAPANESE INFANTS*

BY

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ABSTRACT

Several cases of fibrocystic disease of the pancreas or meconium ileus in Japanese children were reported recently. However, the majority of the cases in Japan were diagnosed by similar clinical symptoms of the disease and by typical histological findings, but not by satisfactory data.

In order to know the incidence of the fibrocystic changes of the pancreas, the pancreas of 422 autopsy cases of infants were examined histologically.

Results were as follows:

1. Inspissated material in acini was found in 14.7% of 422 autopsy cases.
2. Inspissated material in pancreatic ducts was found in 24.2%.
3. Cystic dilatation of pancreatic ducts or acini was found in 6.4%.
4. Fibrosis of the pancreas was seen in 14.7%.

Cystic and fibrotic changes of the pancreas in autopsy cases among Japanese children, were seen in extremely high incidence, not only in cases of the group who expired under conditions similar to fatal complications of fibrocystic disease of the pancreas, but also of cases who expired from other causes.

INTRODUCTION

Fibrocystic disease of the pancreas is geographically widespread, but rarely seen in the Negro and never in Oriental races. Several cases of fibrocystic disease of the pancreas or meconium ileus in Japanese children, however, were recently reported, as shown in Table 1.

Examinations of cases in Japan were unsatisfactory, from the view point of the diagnostic criteria presented by di Sant'Agnese.

The fibrocystic change of the pancreas in this disease is not only a part of the diagnostic findings, but is also not specific to the disease.

Fibrocystic disease of the pancreas is not a disease of the pancreas, but a generalized disorder, as is well known. Then it is very important to find whether Oriental races actually have such a typical generalized disorder and why there is such a difference in the incidence of the disease among races.

The majority of the cases in Japan which were reported as fibrocystic

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Table 1. Meconium ileus and ficrocystic disase of the pancreas in Japan

No. of Cases	Author	Year	Age	Sex	Family History	Examination					Histological Finding		
						Sweat			D.F.	X-P.	Cast	Cystic	Fibrosis
						Na	Cl	Na/Cl					
①	Masuno	1951	15D.	M.	(+)	mFg/L	mFg/L				(−)	(−)	(±)
②	〃	〃	6D.	M.	〃						〃		
③	〃	〃	11D.	F.	〃						〃		
4	Kokawa	1955	37Y.	F.								(+)	(+)
5	Fuchigami	1956	4Y.	F.	Consanguinity					(+)			
6	〃	〃	3Y.6M.	F.						(+)			
⑦	Yamamoto	1957	13D.	M.							(+)	(+)	(+)
8	Ito	1958	11M.	F.	Consanguinity						(−)	(+)	(+)
⑨	Yamauchi	1960	3D.	F.	(+)						(+)	(+)	(+)
⑩	〃	〃	3M.	F.									
11	Hamamoto	1961	5M.	M.	Consanguinity	210	82	2.65	(+)	(+)			
12	Kobayashi	1961	1Y.7M.	F.	Consanguinity					(+)		(+)	(+)
⑬	Kadowaki	1963	2D.	M.	(+)							(+)	(+)
14	Ikai*	1964	2M.	M.							(+)	(+)	(−)
⑮	〃	〃	3D.	F.							(−)	(−)	(+)
16	〃	〃	1.5M	F.							(−)	(+)	(−)
17	〃	〃	3M.	F.							(+)	(+)	(−)
18	〃	〃	1M.	F.							(+)	(+)	(+)
19	Tomisawa	1964	40D.	F.	(+)					(+)	(+)	(+)	(+)
⑳	Furuya*	1965	2D.	F.	(−)	74.1 101.1	33.5	2.21	(−)	(−)	(+)	(+)	(+)
21	Kato*	1965	2Y.	M.		112 109	144 141	0.78 0.77		(+)	(+)	(+)	(+)
22	Seki*	1967	2M.	F.		88	94	0.88		(+)	living		
23	Ishida*	1967	5Y.6M.	F.	(+) ?					(+)	(−)	(+)	(+)

Notes; Blank: not stated, (−): Examined, but negative, (+): Positive finding, D.F.: Duodenal fluid, X–P: Chest X–ray picture, ①……Meconium ileus, *Personal communication.

disease of the pancreas were diagnosed by similar clinical symptoms of the disease and by typical histological findings, but not satisfactory data.

In order to know the incidence of the fibrocystic changes of the pancreas, the pancreas of autopsy cases of infants were examined histologically.

Materials and Methods

The pancreas of 422 autopsy cases, under 5 years of age, were examined histologically. The age distribution of these 422 cases is shown in Fig. 1.

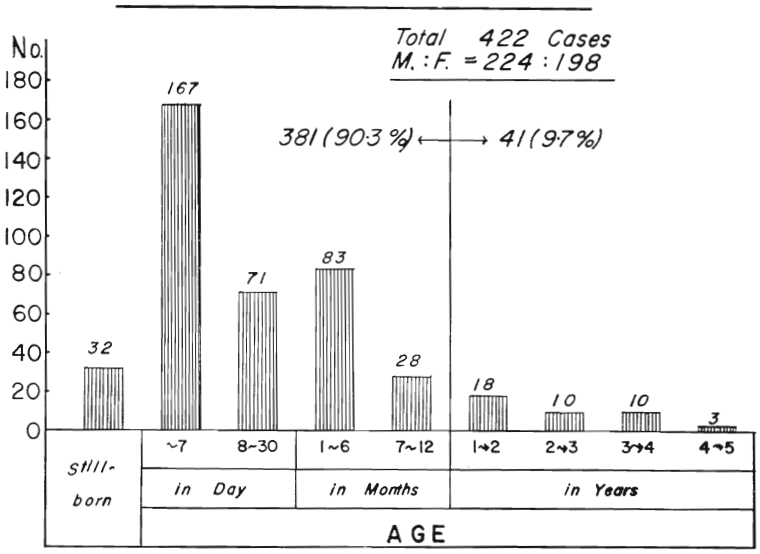

Fig. 1. Age distribution of autopsy cases under 5 years of age

The majority of the cases is less than 1 year of age, 381 cases (90.3%) out of 422. The ratio of male and female is 224 to 198.

Classification of clinical and pathoanatomical diagnosis of the 422 cases is shown in Table 2. Pulmonary disorders were found in 163 cases and gastrointestinal disorders in 66 of 422.

Histological findings of the pancreas stained by hematoxylin and eosin are classified as follows:

1. Inspissated material in pancreatic ducts and acini as shown in Figs. 2 and 3.
2. Cystic dilatation of ducts and acini as shown in Figs. 4 and 5.
3. Fibrosis of the pancreas as shown in Figs. 6 and 7.

The 422 cases were divided in 2 groups; Group 1, those showing similar clinical symptoms to fatal complications of fibrocystic disease of the pancreas

Table 2. Clinical or pathoanatomical diagnosis of 422 cases

Diagnosis	Number of Cases
Pulmonary	163
Pneumonia	116
Atelectasis or Dystelectasis	30
Hyaline membrane disease or aspiration of amniotic fluid	14
Others	3
Gastrointestinal	66
Atresia, Stenosis etc.	24
Jaundice	18
Billiary atresia	7
Dyspepsia or Ekiri	6
Hepatitis	4
Others	7
Cardiovascular	52
Nervous system	37
Sepsis etc.	17
Hematologic	6
Genito-urinary	6
Tumor	6
Others	69
Total	422

and Group 2, the balance of the cases. The percentages of the histological changes of the pancreas in both groups were compared.

Results

The results of histological findings in the 422 cases are shown in

Table 3. Histological findings of the pancreas of 422 cases

		Grade			Total
		(−)	(±)	(+)	
		Cases (%)	Cases (%)	Cases (%)	Cases (%)
Inspissated material (Cast)	in acini	340 (80.6)	20 (4.7)	62 (14.7)	422 (100)
	in ducts	287 (68.0)	33 (7.8)	120 (24.2)	422 (100)
Cystic dilatation of acini or ducts		381 (90.3)	14 (3.3)	27 (6.4)	422 (100)
Fibrotic changes		280 (66.4)	60 (18.9)	62 (14.7)	422 (100)

Table 4. Histological changes of the pancreas in the 2 groups A and B*

			Grade			Total
			(−)	(±)	(+)	
			Cases (%)	Cases (%)	Cases (%)	Cases (%)
Inspissated material (Cast)	in acini	A	145 (77.6)	11 (5.9)	31 (16.5)	187 (100)
		B	195 (83.0)	9 (3.8)	31 (13.2)	235 (100)
	in ducts	A	127 (68.0)	13 (6.9)	47 (25.1)	187 (100)
		B	160 (68.1)	20 (8.5)	55 (23.4)	235 (100)
Cystic dilatation of acini or ducts		A	167 (89.3)	3 (1.6)	17 (9.1)	187 (100)
		B	214 (91.0)	11 (4.7)	10 (4.3)	235 (100)
Fibrotic changes		A	141 (75.4)	19 (10.2)	27 (14.4)	187 (100)
		B	139 (59.2)	61 (25.9)	35 (14.9)	235 (100)

* A: 187 cases=Selected cases showed similar clinical symptoms of C.F. fatal complications.
B: 235 cases=others.

Table 3. Inspissated material in pancreatic ducts was found in 24.2%. Inspissated material in acini was found in 14.7%. Cystic dilatation of the ducts or acini was found in 6.4%. Fibrosis of the pancreas was seen in 14.7%.

The percentages of the histological changes of the pancreas in the two groups were compared in Table 4. There was almost no difference in percentages of the pancreatic changes in the two groups.

The protocols of the autopsy cases that showed fibrocystic findings of the pancreas were carefully checked all over again, from the view point of the diagnostic criteria of fibrocystic disease of the pancreas, but no case of fibrocystic disease could be seen which had been diagnosed by laboratory data including sweat test.

Summary and Conclusion

1. Cases diagnosed and reported as fibrocystic disease of the pancreas or meconium ileus in Japan were reviewed.

2. Cystic and fibrotic changes of the pancreas in autopsy cases among Japanese children, were seen in extremely high incidence, not only in cases of the group who expired under conditions similar to fatal complications of fibrocystic disease of the pancreas, but also of cases who expired from other causes.

3. Fibrocystic changes of the pancreas should not be a diagnostic

finding for fibrocystic disease. Laboratory data, especially the results of sweat test, are most important in the correct diagnosis of fibrocystic disease of the pancreas.

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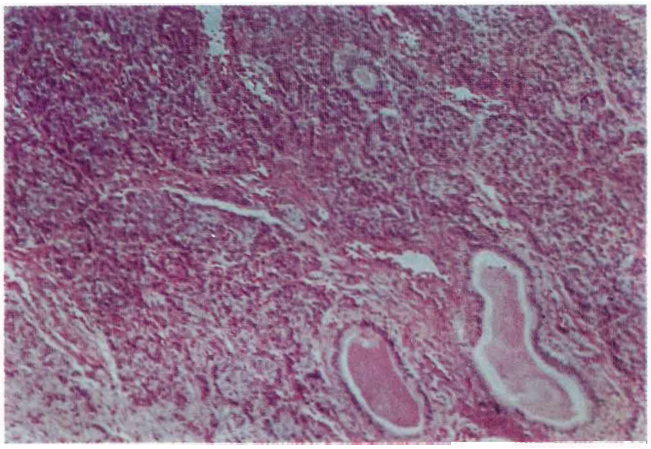

Fig. 2. Inspissated material in pancreatic ducts. H.E. ×100.

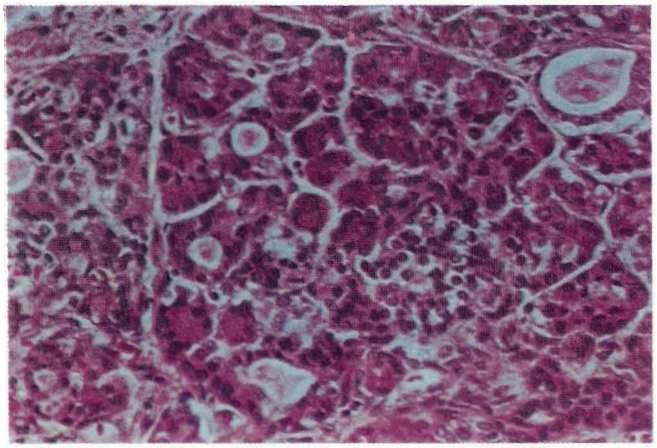

Fig. 3. Inspissated material in ducts and acini. H.E. ×270.

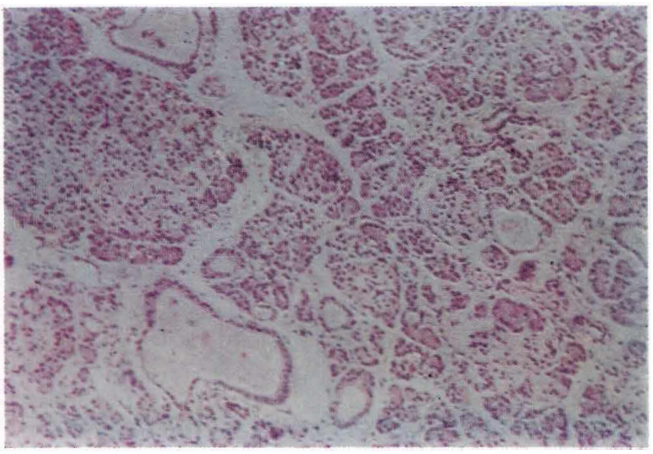

Fig. 4. Cystic dilatation of ducts or acini. H.E. ×100.

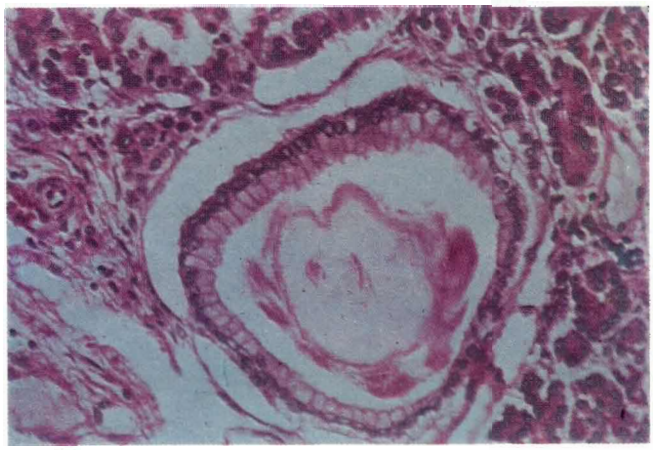

Fig. 5. Cystic dilatation of duct or acinus. H.E. ×270.

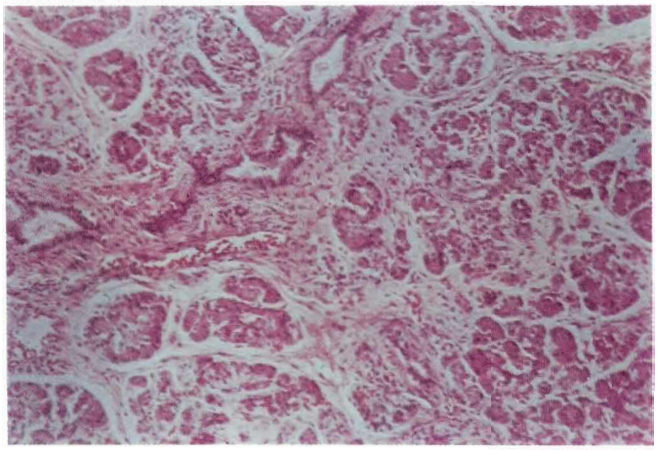

Fig. 6. Fibrosis of the pancreas. H.E. ×100.

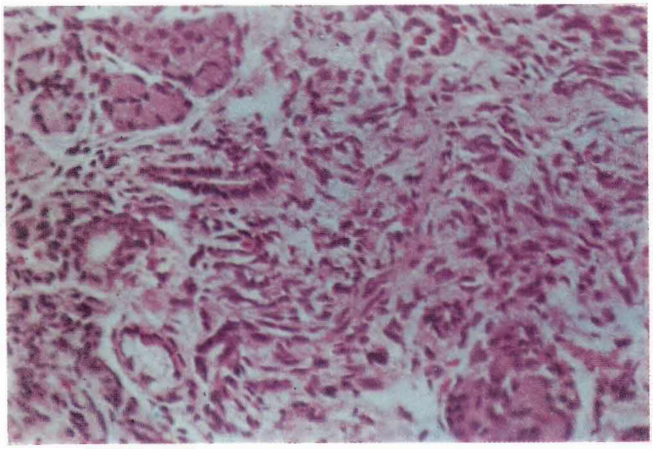

Fig. 7. Fibrosis of the pancreas. H.E. ×270.