INTESTINAL OBSTRUCTION AS A LATE COMPLICATION OF FIBROCYSTIC DISEASE OF THE PANCREAS (MUCOSIS)

BY

O. D. FISHER

From the Royal Belfast Hospital for Sick Children

(RECEIVED FOR PUBLICATION FEBRUARY 8, 1954)

The features of fibrocystic disease of the pancreas are well recognized following the description by Andersen in 1938 of three main clinical forms.

The first is that of intestinal obstruction in the first few days of life due to meconium ileus. About a fifth of all cases present with failure to pass meconium which is followed by vomiting and abdominal distension. It results from inspissation of meconium in the small bowel and at operation or necropsy the upper part of the small bowel is seen to be distended with gas and fluid. The lower part is filled with hard inspissated meconium and the bowel wall hypertrophies and tapers into an unexpanded ribbon-like colon. Perforation of the bowel wall may occur causing meconium peritonitis while volvulus and sometimes atresia of the small bowel may result. Bodian (1952) describes three instances of meconium retention in which spontaneous recovery occurred a few days after birth although all finally succumbed to the disease.

The second form is that of respiratory disorders which is marked by a distressingly persistent and spasmodic cough which simulates pertussis and is associated with episodes of respiratory infection usually due to Staphylococcus aureus. Respiratory distress with an increased respiratory rate, signs of infection and emphysema may be noted. Bronchiectatic changes are common and finger clubbing and even cor pulmonale may develop.

The third form is that of the coeliac syndrome in which failure to thrive in spite of an excellent appetite is followed by loose offensive stools, gaseous abdominal distension, wasting and stunting.

Meconium ileus is usually fatal though immediate survival has followed surgical intervention but later features of the disease soon supervene. Numerous variations and combinations of symptoms may confuse the clinical picture but, despite fluctuations, the course of the disease is invariably fatal.

In fibrocystic disease of the pancreas, intestinal obstruction after the neonatal period due to inspissated small bowel contents with recovery is considered worth while reporting.

Case Report

John C. was first admitted to hospital on December 8, 1951, aged 13 months, with a persistent cough and failure to thrive. Birth weight was 6 lb. and after two months of breast feeding, dried milk and later raw cow’s milk feeds were substituted. At 3 months a spasmodic but persistent cough resembling pertussis developed. At 6 months mixed feeding was started, and in spite of an excellent appetite the child did not thrive, having loose, offensive and frequent stools.

Of the family, the parents and three brothers are alive and well, two sisters and a brother died at 2 months, 6 months and 4 months; each child failed to thrive and developed loose, offensive stools and a cough like that of pertussis which led to a terminal respiratory illness.

On examination, the child was marasmic, weighing 12½ lb., with abdominal distension and a loose cough associated with scattered ronchi in the chest. Investigation showed no tryptic activity in the stools or in three specimens of duodenal juice. The Mantoux reaction was negative. Staphylococcus aureus was cultured from the nasopharynx and a radiograph of the chest showed emphysema and heavy hilar shadows. Treatment with aureomycin and pancreatin resulted in marked improvement and a gain of 2 lb. in weight in 3 weeks. The child was discharged on January 4, 1952, on a high-protein diet with pancreatin granules, 45 gr., before each feed and aureomycin, 250 mg. daily.

Progress was satisfactory until February 4, 1952, when the supply of pancreatin ran out and could not be replenished. The next day the child vomited and later developed abdominal pain. On February 9, when readmitted to hospital aged 15 months, vomiting was continuous and, except for one small stool two days before, constipation was complete.

The child was irritable with bouts of abdominal pain causing the legs to be drawn up on to the abdomen. The latter was grossly distended but resonant. Bowel
sounds but no visible peristalsis were present. An
indefinite mass was present in the right iliac fossa, the
rectum was empty and no blood or mucus was present
or had been passed.

Acute intestinal obstruction was diagnosed and
immediate laparotomy undertaken. On opening the
peritoneal cavity a small amount of clear fluid was seen;
the whole of the small bowel was found to be inter-
mittently obstructed by masses of putty-like material in
the bowel lumen. It was so firm that it could not be
expressed through the ileocaecal valve. The large bowel
was unobstructed. A portion of the small bowel was
packed off and an incision made through the bowel wall;
from the lumen a tenacious material like liquid rubber
was extracted. The incision was repaired and as the
whole of the small bowel was affected no further opera-
tive procedure was undertaken.

Post-operatively hydration was maintained by intra-
venous and subcutaneous fluids. After gastric aspiration
45 gr. of pancreatin granules freshly dissolved in a small
quantity of water were given at four-hourly intervals.
A daily bowel wash-out was given and on February 16
a large bowel action followed with relief of abdominal
distension and vomiting.

From February 14, terramycin, 50 mg., was given orally
twice a day. The child made an uninterrupted recovery and
was discharged to continue on a high-protein diet
with pancreatin 45 gr. before each feed. On July 21,
1952, the child was again admitted with a further respi-
atory infection which responded to aureomycin
therapy. Investigations again confirmed the lack of
tryptic activity in stools and duodenal juice.

Discussion

This child’s illness illustrates the customary course
of fibrocystic disease of the pancreas with the early
onset of respiratory and alimentary symptoms. The
family history suggests that three siblings suffered
from the same disease which is known to be
inherited as a recessive characteristic. The diagnosis
was confirmed by the lack of tryptic activity in the
stools and duodenal juice on repeated examinations.

The development of intestinal obstruction due to
inspissated small bowel material has been reported
only once before as a late complication in fibro-
cystic disease of the pancreas.

Levy (1951) records such an instance in a child
at 7 months of age. Fibrocystic disease of the
pancreas had been diagnosed at 9 weeks and treated
with pancreatin, 60 gr. daily. When acute intestinal
obstruction developed a tentative diagnosis of
intussusception was made and laparotomy per-
formed. The small bowel was found to be dis-
tended and plum-coloured, with haemorrhages in
the root of the mesentery; the colon was empty and
contracted. No surgical procedure was undertaken
and the child died the following day. At necropsy
addition to the laparotomy findings, the small
bowel was filled with putty-like material. There
was no evidence of volvulus, intussusception or any
other organic obstruction present. The pancreas
was firmer than normal and showed histologically
the features characteristic of fibrocystic disease with
a great increase in fibrous tissue, cystic dilatation
of the ducts which contained desquamated epithe-
lium and homogeneous eosinophilic tissue.

These two cases closely parallel those of meconium
ileus with the inspissated gut contents causing
intestinal obstruction which affects the small bowel
leaving the large bowel collapsed but unobstructed.
In meconium ileus the primary cause of obstruction
is the abnormal physical character of the meconium,
suggested by Landsteiner as early as 1905 to be
due to lack of pancreatic ferments. It may be
followed by volvulus and may lead, according to
Lelong, Petit, Le Tan Vinh and Borniche (1950),
to secondary atresia of the small bowel. In fibro-
cystic disease of the pancreas the degree of involve-
ment of the pancreas and its ability to secrete
pancreatic ferments must influence the manifesta-
tions of the disease; absence of secretion in the
late intra-uterine period may lead to meconium
ileus, partial lack to meconium retention, while
deficient secretion in infancy may result in the
colic syndrome or obstruction due to inspissation
of bowel contents as in these cases.

It should be noted that vomiting unassociated
with coughing, which is not an uncommon symptom
of this disease, may be due to such an abnormality
of small bowel contents. This is supported by the
report of Rasor and Stevenson (1941) of a boy
suffering from fibrocystic disease of the pancreas in
whom vomiting was a persistent feature. At post-
mortem examination at 1 year of age, the intestinal
contents were found to be of a putty-like consistency.
As Levy (1951) suggests, the younger and more
debilitated children are liable to this form of
intestinal obstruction as older children with their
more powerful intestinal peristalsis can probably
propel the abnormal faeces through the ileocaecal
valve.

In the case reported the intestinal obstruction
followed closely the cessation of pancreatin therapy
and was possibly relieved by the oral administra-
tion of pancreatin. This suggests a further use for
pancreatin which has been advocated by Andersen
(1945) as a form of replacement therapy. Newer
preparations of enteric-coated pancreatin granules
of three times the strength of the B.P. preparation
are fairly well tolerated and effective in improving
the absorption of food as well as the consistency
and odour of the stools.

The occurrence of intestinal obstruction as a late
complication of fibrocystic disease of the pancreas must be fully recognized. Laparotomy will confirm the obstruction is due to inspissation of small bowel contents and an ileostomy should be performed because the site of maximal obstruction is in the region of the ileocaecal valve and as this procedure has proved successful in cases of meconium ileus reported by Bodian (1952) and Hiatt and Wilson (1948). By this route, freshly prepared pancreatin can be irrigated into the small bowel while pancreatin is also given by gastric tube after aspiration. By these means, the abnormal bowel contents may be softened and liquefied thus relieving the intestinal obstruction.

**Summary**

A case of intestinal obstruction due to inspissated small bowel contents at 15 months of age is reported. Occurring as a late complication of fibrocystic disease of the pancreas the similarity to meconium ileus is discussed. Attention is drawn to the condition which is best treated by ileostomy and pancreatin therapy.

I am grateful to Dr. A. A. H. Gailey and Mr. Ian Fraser for permission to publish this case.

**REFERENCES**

Intestinal Obstruction as a Late Complication of Fibrocystic Disease of the Pancreas (Mucosis)
O. D. Fisher

Arch Dis Child 1954 29: 262-264
doi: 10.1136/adc.29.145.262

Updated information and services can be found at:
http://adc.bmj.com/content/29/145/262.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/