CHAPTER IX

CONGENITAL STEATORRHŒA

THE anomaly now to be discussed differs from those which have gone before in that it is not an error of protein metabolism, but is concerned with the utilization of fats. From their birth onwards, the subjects of it pass with, and sometimes apart from, their motions, an oily material which solidifies into a crust or cake of solid fat on cooling. Even in infancy the peculiar greasy character of the stools, and the foul odour of rancid fat which they emit, attract attention and lead the mother to seek medical advice.

This phenomenon is well known as a symptom of disease, and was described in the seventeenth century by Tulpius [1] and others. Writing in 1824 Kunzmann [2] gave it the name of 'steatorrhœa', and recognized its association with disease of the pancreas. A few years later Richard Bright [3] arrived independently at a like interpretation of its significance, and later observations have tended to confirm this view. H. Salomon goes so far as to assert that such butter-stools indicate, with certainty, deficiency of pancreatic secretion.

The stools of normal adults contain fats, fatty acids and soaps, which together constitute some 21 to 28 per cent. of the dried fæcal material. Far larger amounts are passed by sufferers from diseases of the liver with biliary obstruction, of the intestinal walls and of the pancreas, but I know of no case in which butter-stools have resulted from hepatic

[1] *Observationum Medicarum Lib.* iii, cap. 18.
[2] *Journal der prakt. Heilkunde*, 1824, lix. 3, Stück 45.
[3] *Medico-chirurgical Transactions*, 1833, xviii. 1.

or intestinal disease. Moreover only a small proportion of patients with pancreatic disease, most of whom have the pancreatic duct occluded, pass such stools.

The pancreas, like other glands, is subject to a great variety of lesions, and it is not surprising that in cases of disease of this gland the amounts and proportions of the fatty substances excreted vary widely.

In some cases there is a large excess of fats, fatty acids and soaps, in others a great relative increase of neutral fats, and in yet others the fatty constituents do not exceed the normal quantities.

It is by no means clear what conditions determine the formation of butter-stools. Certainly excess of neutral fat is not the determining factor, for, as Vaughan Harley,[4] Gross,[5] and others have shown, analysis of such stools may reveal no such excess; and Harley, who analysed the actual oily substance, apart from the fæces, found it to consist for the most part of fatty acids, with only small quantities of neutral fat and soaps. The mere excess of fatty constituents undoubtedly plays a part, and patients who pass such stools may only do so when their diet is rich in fat. On the other hand stools exceptionally rich in fats may have no such character, as in a case, recorded by Whipple, of a remarkable intestinal disease not described previously, in which, although the fats and fatty acids constituted 80 per cent. of the dried fæces, the stools were white and creamy, and had a 'smooth, silky appearance'.

The study of fat excretion by sufferers from pancreatic disease has led to the conclusion that this gland exerts a powerful influence upon the absorption and utilization of fats, in which the lipase plays but a small part. It seems obvious that there is a second and little known factor concerned, possibly an internal secretion, although the

[4] *Journal of Pathology and Bacteriology*, 1896, iii. 245.
[5] *Deutsches Archiv f. klin. Medizin*, 1912, cviii. 106.

pronounced effect of obstruction of the duct lends no support to that view.

Up to now only three cases of congenital steatorrhœa have been recorded, of which two occurred in one family, and although two of these cases have been investigated with some thoroughness, any description of the anomaly based upon so few examples must needs be tentative.

The first case, that of a boy who came under observation in 1911, at the age of six years, was investigated by Hurtley and myself [6] at intervals during the two years following; the second, that of a boy aged three, was observed by R. Miller and H. Perkins [7] in 1914, and recorded by them in 1920. Both children were brought to hospital on account of the butter-stools, and in each case the evidence of an intelligent mother left no doubt that the anomaly had been present from earliest infancy. The parents of our patient were first cousins; he was the second of five children, three of whom were normal, whereas a younger brother, who passed similar stools, had died of measles at the age of eleven months. We were fortunate in being able to obtain information as to the patient's condition at the age of three, when he was, for a time, an inmate of the German Hospital.

Neither boy showed any arrest of growth or development, such as is apt to result from intestinal lesions associated with fatty stools. The patient of Miller and Perkins died at his home, of broncho-pneumonia, and no autopsy was made, but the other, when last seen in 1919, after an interval of seven years, was a well grown lad of fifteen, engaged in active work. Examination of his fæces, at that time, showed that the anomaly persisted, but he appeared to have so adjusted his diet that he suffered

[6] *Quarterly Journal of Medicine*, 1913, vi. 242.
[7] Ibid., 1920, xiv. 1.

little inconvenience, and butter-stools were then seldom passed.

Neither child had jaundice nor any other indication of hepatic lesion. To exclude a defect of hepatic secretion certain special examinations were carried out in our case. The fæces contained more obvious traces of cholalic acid than did those of a control boy; there was no disturbance of sulphur metabolism to suggest deficient formation of taurocholic acid, and Fel Bovinum, given by the mouth, did not improve the utilization of fats. No signs of an intestinal lesion were forthcoming, and when the fat in the diet was reduced to a minimum the appearance of the fæces was normal.

Apart from the steatorrhœa there was nothing to suggest disease of the pancreas in either case; no increase of urinary diastase, no adrenalin mydriasis, and, what is far more important, no signs of deficiency of tryptic digestion. The stools of our patient contained no undigested muscle fibres, and a diet rich in protein produced no noteworthy increase of non-fatty residue. Other pancreatic tests tended to confirm these findings. A dose of 100 grammes of glucose caused no glycosuria.

Many analyses of fæces were carried out in these cases, and in both the utilization of fats was investigated. Some of the analytical figures have been brought together, for purposes of comparison, together with some from a control boy, aged six years, convalescent from chorea. The differences of age, and consequent differences in the amounts of fat taken by the two patients, render the figures less strictly comparable than might be wished.

It will be seen that in both cases the proportion of fatty substances in the dried fæces was far above the normal. In Case 1 it varied little from 80 per cent. so long as the diet contained 177 grammes of fat, but the diet was one which produced but a small non-fatty residue, and the figure

is correspondingly high. With a diet containing 92 grammes of fat the proportion was 55·5 per cent. with nearly twice as much non-fatty residue, owing to the substitution of jam for dripping. In the case of the control boy the figure was considerably below the normal average for adults. In Case 2 the diet yielded more residue, but with 78 grammes of fat in the diet the percentage of fatty substances was 62.

In Case 1 when the diet contained less fat the proportion of neutral fat was not conspicuously large, but with the richer diet it rose to 61·4 per cent., and it is noteworthy that Katz considered that a lesion of the pancreas may be

		Percentages.					*Utilization.*		
	Fat in Diet.	*Fat in dry Fæces.*	*Neutral fat in total fat.*	*Fatty acids in total fat.*	*Soaps in total fat.*	*Total split fat in total fat.*	*Fat excreted, per cent. of fat taken.*	*Fat absorbed, per cent. of fat taken.*	*Total residue not fat grammes.*
Boy, 6–8 yrs. (Garrod & Hurtley.)	177 *	79·4	61·4	32·6	6·0	38·6	24·0	76·0	10·9
	92 †	55·5	32·0	45·5	22·5	68·0	25·9	74·1	19·1
almost fat free.		44·0	54·7	35·2	10·0	45·2	—	—	5·8
Boy, 3 yrs. (Miller & Perkins.)	78	62·0	21·1	55·0	23·9	78·9	42·1	57·9	20·2
	49	40·0	35·7	45·3	19·0	64·3	32·6	67·4	24·3
	22	31·2	10·8	54·6	34·6	89·2	15·9	84·1	7·7
Boy, 6 yrs. Control.	177	14·5	52·1	24·0	23·9	47·9	0·8	99·2	8·6

* Average of nine days. † Average of four days.

suspected when the proportion of neutral fat exceeds 30 per cent. In Case 2 the splitting of fats was far less

deficient, even when a diet rich in fat was taken. It is obvious that in these cases the steatorrhœa was not due to mere excess of neutral fat, for in Case 1 preparations, such as pancreon, which improved the splitting of fats, in no way improved their absorption.

Zoja and others have attached importance to scanty excretion of soaps as a sign of disease of the pancreas, but the proportions vary so widely in different cases that it is unsafe to place reliance upon this indication.

Special interest attaches to the utilization columns of the table. In Case 1 the proportion of the fats taken which reappeared in the fæces varied but little from 25 per cent. whether the diet contained 92 or 177 grammes, whereas, in Case 2, the percentage rose conspicuously when the fat in the diet was increased, and reached 42 with a diet of 78 grammes. This difference of behaviour, apparently so fundamental, loses much of its significance in view of the fact that like differences are observed in cases of serious disease of the pancreas. Thus Oscar Gross [8] investigated a case in which the waste of fat amounted to about 50 per cent. of that taken in diets with 84 and 375 grammes respectively, whereas in one recorded by Spriggs and Leigh [9] the waste of fat increased greatly when the fat in the diet was increased; with a diet containing 136 grammes 55 per cent. reappeared in the fæces, with a diet of 262 grammes 78 per cent. reappeared. It is clearly unsafe, in the light of our present knowledge, to draw any conclusions from the constancy or variations of the percentage of fats utilized, as to the seat or nature of the underlying cause of the steatorrhœa.

In some cases of pancreatic steatorrhœa the administration of pancreon has brought about conspicuous improvement, but in others it has failed to do so. Thus Gross found

[8] loc. cit., sub 5.

[9] *Quarterly Journal of Medicine*, 1915-16, ix. 11.

that in his cases pancreon had no effect upon utilization of fat, and Spriggs and Leigh remark that, in their case, it appeared that when pancreon and holadin were given less fat was absorbed than in control periods.

In our case pancreon and holadin were given, but we had not the opportunity of testing these preparations as thoroughly as we had intended, or of giving large doses with alkalies, which are said to increase their efficacy. Both preparations appeared, as in the pancreatic case of Spriggs and Leigh, to aggravate the failure to utilize fats, although they improved the splitting of fat.

It must be mentioned that steatorrhœa due to organic lesions of the pancreas may persist for years without grave impairment of the general health, provided that there be no attendant glycosuria, as in the remarkable case described by T. J. Walker,[10] of a medical man who died at the age of ninety, having passed butter-stools from his sixty-third year. At the autopsy his pancreatic duct was found to be blocked by calculi, and the gland substance was almost entirely replaced by fat. However, in such cases there are clear signs of impairment of tryptic digestion also.

As regards congenital steatorrhœa, the fact that the passage of butter-stools is so intimately associated with lesions of the pancreas suggests that in the subjects of this anomaly some factor of the pancreatic secretion is wanting, an hypothesis which brings this condition into line with other known errors of metabolism. Clearly a mere absence of lipase will not suffice, and we are driven to suppose that what is wanting is that unknown factor in the secretion of the pancreas which controls the utilization, as distinguished from the splitting, of fats.

We have seen that the two cases of congenital steatorrhœa hitherto investigated differ from each other in certain noteworthy respects; that in one the percentage of fat wasted

[10] *Medico-chirurgical Transactions*, 1889, lxxii. 257.

was constant in spite of changes of diet, in the other it varied with the amount of fat taken in the food, and that in the one case the splitting of fats was much more efficient than in the other. But we have also seen that such differences are met with in cases of gross disease of the pancreas. Moreover, the effect of the administration of pancreon and holadin in Case 1 can be matched in cases of pancreatic disease; and despite the lack of any failure of other functions of the pancreas in the congenital cases it is probable that they in no way impair the truth of Salomon's dictum, that true butter-stools indicate, with certainty, deficiency of pancreatic secretion.

Lastly, it is worthy of note that in the cases of congenital steatorrhœa the waste of fat is considerably less than in the recorded cases of steatorrhœa due to disease of the pancreas, which are as yet few in number, in which the utilization of fats has been estimated. It is tempting, but perhaps not justifiable, to connect this with an isolated failure of one single factor as distinguished from impairment or abolition of the functions of the pancreas as a whole.