

CHARLOTTE M. ANDERSON: LONG-TERM SURVIVAL WITH SIX INCHES OF SMALL INTESTINE

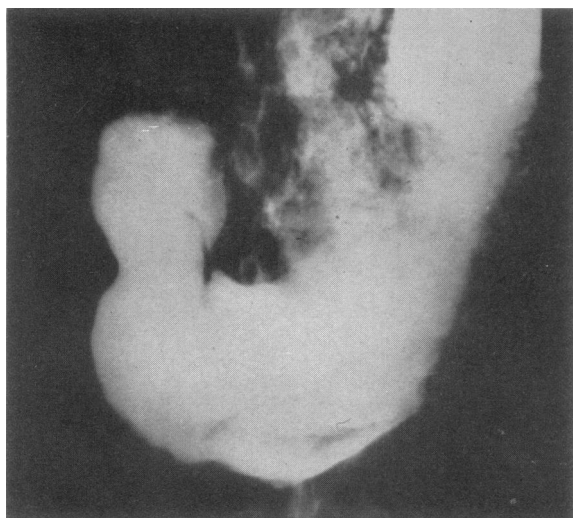


FIG. 1

FIG. 1.—Barium x-ray film at zero time, showing large stomach.

FIG. 2.—Barium x-ray film after 15 minutes, showing extent of small intestine and barium already in caecum.

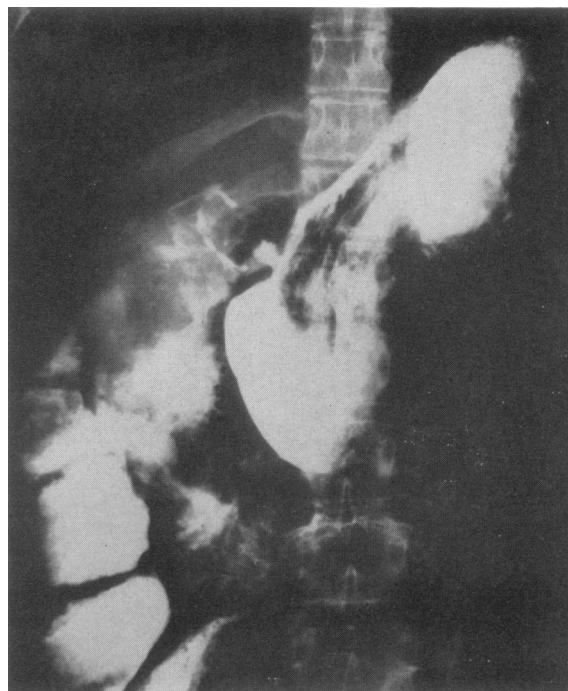


FIG. 2

H. GARLAND *ET AL.*: IATROGENIC VERTEBRAL ARTERIOVENOUS FISTULA



FIG. 1

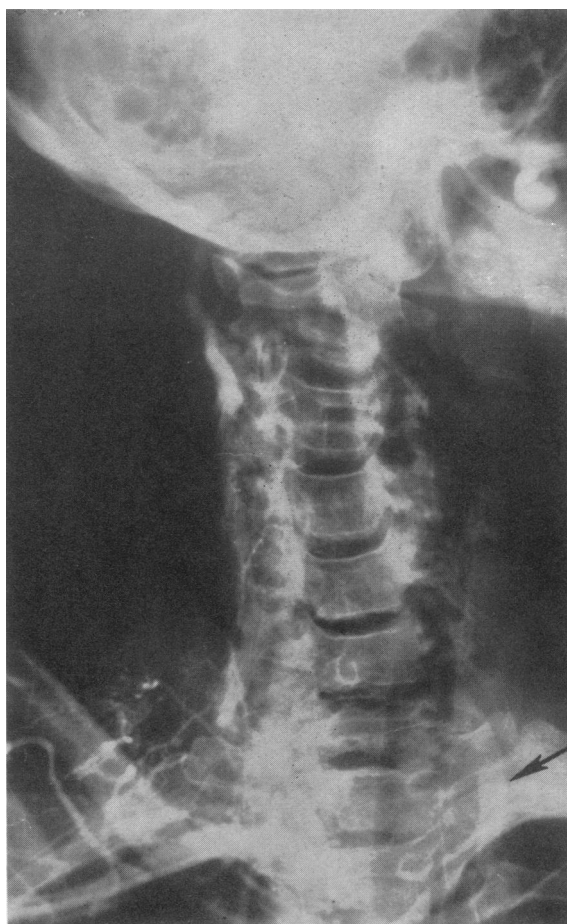


FIG. 2

FIG. 1.—Right vertebral arteriogram via axillary artery. Contrast has passed directly into greatly dilated vertebral veins and into the deep cervical vein. There is already some contrast in the vertebral veins of the opposite side. Note the dilated vertebral artery, which narrows suddenly at C 4 level, the probable site of the fistula.

FIG. 2.—Arteriogram taken one second after Fig. 1. Contrast is draining downwards through dilated vertebral and deep cervical veins on the right side. Note also the small veins extending across the midline at mid-cervical level and the outlining of the left vertebral vein (arrowed).

defective are more likely to volunteer than those who are untroubled by ocular symptoms. Again, in the publicity campaign it was reported that people whose relatives have glaucoma are more likely to be affected themselves, and I feel sure many have volunteered on this account. These factors would tend to exaggerate the true incidence of glaucoma; but equally there are other factors which might minimize it. We know that the incidence of glaucoma increases with age, and it is possible that fewer elderly people have volunteered because they are incapacitated in some way or even so blind from glaucoma that they have not read of the scheme in the local press. Again, some who are aware of defective vision may be afraid to have their suspicions confirmed.

Ideally we should enlarge the sample to include the whole population above the age of 40. Certainly we should see many thousands more and make special efforts to screen the elderly and infirm.

Our second aim was to assess the methods of diagnosis and investigation at present available. We have established that applanation tonometry can be done satisfactorily by a trained technician under the prevailing conditions and that it gives an excellent criterion for assessment. The level of 21 mm. Hg as a satisfactory criterion is supported by the mean values for the normal eyes of 15.2 mm. Hg (S.D. 2.516) for the right eyes and 14.9 (S.D. 2.533) for the left eyes. Two standard deviations above the mean give values of 20.2 and 19.96 mm. Hg for the right and left eyes respectively.

So far as other diagnostic methods are concerned, some new techniques are being evaluated at the moment.

From the organization point of view we can say that, with adequate publicity and good relations between the population and the local health services, volunteers are forthcoming for screening programmes, and such programmes can be carried out successfully from a public health clinic.

So far as research is concerned, we are collecting an interesting group of patients who have some anomaly of

aqueous dynamics but who, up to the present, do not show the classical picture of glaucoma.

Perhaps one of the most important things we have learnt is that the investigation of the borderline group is very time-consuming and difficult. Our experience suggests that as much time must be spent on investigating the 8% of patients referred for assessment as is spent in screening the rest. This must be borne in mind in any discussion of glaucoma-screening on a national scale. The investigation of 8% of the whole population over the age of 40 (perhaps some 20 million persons) would mean investigating over one and a half million people. As there are approximately 350 consultant ophthalmologists in the country, and assuming the burden was shared equally, each would have some 4,500 glaucoma suspects to deal with—a sobering thought. It seems clear that if a nation-wide screening programme is evolved special glaucoma clinics must be provided to assess doubtful cases and supervise the results of treatment.

Summary

The organization, method of screening, criteria for selection of cases for reassessment, and preliminary results of the current glaucoma survey at Bedford are described. Definite glaucoma was diagnosed in 1.6% of the first 2,000 cases. As much time must be spent on assessment of the 8% of doubtful cases as on screening the rest.

I should like to acknowledge here that much of the success of this survey has resulted from the smooth organization provided by Dr. C. Sharp, the Medical Officer of Health for Bedford, and his staff, and the high standard maintained by the ophthalmologist, Dr. J. E. Wright, and the technician, Miss E. Verkoczy.

REFERENCE

- Sorsby, A. (1956). *Blindness in England, 1951-1954*, pp. 6, 13. H.M.S.O., London. (Ministry of Health Report.)

Long-term Survival with Six Inches of Small Intestine

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[WITH SPECIAL PLATE]

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Much has been written about the fate of patients who have had considerable lengths of small intestine removed surgically, and it seems that survival without severe malabsorption and malnutrition can certainly be expected when up to 50% has been removed. Pietz (1956) offers this opinion from an appraisal of the literature to that date, but states that fair health can be maintained if only 2 ft. (60 cm.) of intestine remains. However, long-term survival when less than 2 ft. (60 cm.) of small intestine has been left behind is very uncommon (Levin, Zamcheck, and Gottlieb, 1961). The purpose of this paper is to record the survival in reasonably good health and nutrition of a girl, now 22 years of age, who had all but

approximately 6 in. (15 cm.) of her small intestine removed seven and a half years ago.

Case Report

In March 1957 the patient, a girl aged 14½ years, had a severe attack of abdominal pain associated with vomiting. She had suffered from mild attacks of a similar nature since the age of 10, but otherwise had been well and had grown normally. The pain and vomiting continued for some days and she became very ill, being admitted to hospital in a state of dehydration and circulatory failure. General peritonitis was diagnosed and after resuscitation an emergency laparotomy was performed. Volvulus of the small intestine had occurred and the whole of the bowel concerned in the volvulus was gangrenous. All but 3 in. (7.5 cm.) of the jejunum beyond the duodeno-jejunal flexure, and 3 in. (7.5 cm.) of ileum proximal

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to the ileocaecal valve was removed and an end-to-end anastomosis performed. The surgeon stated that he measured the remaining pieces of intestine.

Early Post-operative Course

The immediate post-operative course was not abnormal considering the nature of the operation. The child was discharged from hospital after three weeks, weighing 30 kg., whereas two months before the operation her weight was known to be 44.6 kg. She was passing 9 to 12 small watery bowel actions shortly after the bowel resection, but these changed to large porridgy bowel actions after a few weeks and gradually decreased to two a day. She was weak and lethargic, being inclined to feel faint, to have slurred speech, and to drop off to sleep if she did not eat frequently. Instructions to eat at hourly intervals and to have a high-protein diet had been given. Several weeks after discharge from hospital a regime of fortnightly injections of 100 µg. of vitamin B₁₂ was started.

In September 1957, six months after operation, she came under our care for the first time. She was a tall emaciated girl and still weighed only 30 kg. (Fig. I). Her abdomen was somewhat distended, had a tympanic percussion note, and numerous borborygmi could be heard. Reflexes were active and blood-pressure was 90/60. She was admitted to hospital for assessment of small-intestinal function and nutritional state. While in hospital she was weak, apathetic, lethargic, and subject to weak turns. Each day she passed one or two very large bowel actions, which were frothy, pale, porridgy in consistency, and 1 to 2 litres in volume.



FIG. I



FIG. II

FIG. I.—Patient aged 15 years, six months after resection of all but 6 in. (15 cm.) of small intestine. Weight 30 kg. FIG. II.—Patient aged 22 years, seven years after resection of all but 6 in. (15 cm.) of small intestine. Weight 50.8 kg.; height 164 cm.

Investigations

Table I shows the levels of various serum and blood constituents at this stage and at later periods. The electrolytes and total proteins

were substantially normal, but serum cholesterol was low (64 mg./100 ml.) and there was no evidence of anaemia. Liver-function tests were normal. Table II shows daily excretions of fat and protein in the stools, indicating gross malabsorption of both substances, but particularly fat. As fat intake was increased protein absorption lessened. Duodenal intubation revealed that levels of the pancreatic enzymes trypsin, lipase, and amylase in duodenal fluid were normal and the bacterial flora was considered within normal limits. Examination of the stool under the microscope showed numerous fat droplets and undigested meat fibres. Random blood-sugar estimations did not reveal hypoglycaemia, but during a blood-sugar curve the rise in blood sugar was not more than 10 mg./100 ml. X-ray studies of the intestine with barium (Special Plate, Figs. 1 and 2) showed the very short length of small intestine stretching across the abdomen from the duodeno-jejunal flexure to the caecum, which was situated somewhat above the right iliac fossa. The length of intestine appeared to be about 6 in. (15 cm.). The stomach was large, and the large intestine very dilated with much fluid content. Barium was seen to mix with this, and some was still present in the 24-hour film.

TABLE I.—Serum Electrolyte, Protein, and Haemoglobin Levels at Intervals During Seven Years After Intestinal Resection

Time After Operation	Serum Electrolytes (mEq/litre)						Total Protein	Haemoglobin	Blood Film
	Sodium	Potassium	Chloride	Calcium	Magnesium	Phosphate			
							g./100 ml.		
6 months	148	3.8	106	6.0	—	2.2	6.4	14.9	Normal
18 months	156	2.2	115	5.25	0.5	1.4	5.46	9.6	
3 years ..	148	4.2	107	3.2	—	2.1	6.8	14.3	Mild anisocytosis and macrocytosis
5 years ..	152	4.1	105	4.6	—	2.0	6.5	13.7	Mild anisocytosis
7 years ..	144	3.0	100	4.1	2.1	3.6	8.0	10.8	

TABLE II.—Daily Fat and Protein Output and Percentage Absorption During Seven Days, Estimated Six Months After Resection of All But Six Inches (15 cm.) of Small Intestine

Fat in Diet (g.)	Total Fatty Acid in Stool (g.)	3-day Mean % Absorption of Fat	Protein in Diet (g.)	Protein in Stool (g.)	3-day Mean % Absorption of Protein
70	27.7		54	16	
74	64.9	40	50	30.4	61
96	51.2	31	67	20.9	54
100	69.0	30	76	37.2	52
81	73.2	11	61	38.9	29
98	106.7	3	75	74.4	21
99	89.3		78	55.0	

Treatment and Progress

The patient was discharged from hospital and closely followed. During the next 12 months her weight remained stationary at 30 kg. despite attempts to improve the absorption of foodstuffs with pancreatic extract, papain, and emulsifying agents, none of which made any difference to the stools. She continued to eat frequent meals, but these were kept low in fat and high in protein. She did not find this type of diet very palatable, and gradually reverted to a diet containing more fat—in fact, she finally ate what she wanted, as often as she wanted. She avoided those things which increased flatulence and diarrhoea, but was encouraged to take fruit juices for their potassium content. Injections of 100 µg. of vitamin B₁₂, and 2 ml. of vitamin B complex and ascorbic acid (Nicholas) were continued every fortnight as well as a multivitamin preparation orally. She had been given some cortisone before admission to our unit but this was gradually discontinued without any change in her condition.

In September 1958, 18 months after the operation, her general condition was very poor. She had not gained weight, was tired, depressed, and had an occasional episode of dizziness and twitching and could not go to school. Her parents thought she had had one fit, as she had fallen out of bed and appeared semiconscious. She was extremely sleepy, and spent most of the day lying on her bed. At times her behaviour was irrational. She was admitted to hospital as her parents felt they could not cope with her mental state at home. On examination she was emaciated but not dehydrated. Her appetite in hospital was not good, and she would lie in bed, her

condition fluctuating between a state of unrousable drowsiness for several days at a time and a rather more alert condition. She seemed very depressed. It was felt that she was probably entering the terminal phase of her illness and that survival could not be expected with so little small intestine.

At this stage electrolyte, protein, and haemoglobin values were not as satisfactory as previously (Table I), serum potassium being only 2.2 mEq/l. Electrocardiograms supported potassium deficiency. Serum magnesium was estimated and was low, but the reliability of the method was in doubt. Although serum calcium was normal she did have some tingling in the hands and feet, and in view of the history of twitchings at home she was given calcium gluconate tablets by mouth. The administration of extra potassium did not improve her condition noticeably, although serum potassium levels came to normal. Unfortunately, magnesium was not administered until a few months later, when more thought was given to its possible importance. With the idea of making her feel less depressed and unhappy chlorpromazine, in the form of Largactil 25 mg. t.d.s. rising to 50 mg. t.d.s., was given, and to our surprise during the next two weeks she became more alert, ate better, and was able to get out of bed. During the next three months, although she remained most of the time in hospital, with occasional week-ends and days at home, she was alert and cheerful but still weak. However, she started to gain weight and when she was finally discharged in February 1959, had gained 2 kg.

At this time treatment included vitamin B₁₂, and vitamin B complex injections as before, extra potassium in the form of fruit juices, calcium gluconate, and multivitamin tablets daily, and courses of vitamin K intermittently. Chlorpromazine was continued (50 mg. t.d.s.). She was now eating well and continuing to gain weight. In August 1959 she weighed 39 kg. and in March 1960, at the age of 17½ years and three years after operation, 43 kg. However, serum calcium (Table I) at this time was low, and the bones showed some decalcification when x-rayed. She also had occasional tired lethargic spells. She was given 30 g. of calcium gluconate powder daily and 50,000 units of calciferol every second day. Injections of 5 ml. of 50% magnesium sulphate were given intermittently each time she complained of a "tired attack," and each time the symptoms disappeared coincident with its administration. Calcium and calciferol have been continued since and serum calcium has remained normal and the bones look more calcified.

At the age of 18 she started to menstruate, and breast development was obvious. About three months after this chlorpromazine was ceased. The menses became irregular again for about six months, but since then have continued normally. No other ill effect was observed after the cessation of chlorpromazine.

The patient had never excelled at school work and had not continued any studies since operation. She was now sent to a rehabilitation centre, where her education was continued and she was taught typing. She developed physically and emotionally while there, and in March 1962, five years after operation, weighed 51 kg., and was attractive in appearance and fashionable in dress. She was able to participate in social life, the only drawback to this being her embarrassment at her inability to prevent herself passing offensive flatus on occasions. Charcoal was found to best control this, but even now this disability remains to a certain extent. In August 1962 she was able to take a position as a typist and has held this well, losing only an occasional working day because of ill health.

Seven-year Reassessment of Intestinal Function

In December 1963 she complained of pins-and-needles and occasional stiffness in her hands and again felt rather tired. No additional magnesium had been given for about six months. As the time had come to transfer her to the care of a gastro-enterologist looking after adults she was admitted to hospital for a short period to reassess her intestinal function and nutritional state. In January 1964, almost seven years after operation, she appeared attractive (Fig. II), weighed 50.8 kg., and was 164 cm. in height. Physical examination did not reveal any abnormal neurological or other features, the abdomen was not distended, and the skin was normal. The electrolyte pattern (Table I) was essentially normal, except for potassium and magnesium values, which were a little low. Haemoglobin and serum iron (96 µg./100 ml.) were low normal, but serum folic acid activity (15 µµg./ml.) and a serum vitamin B₁₂ level (470 µµg./ml.) were within normal limits. Fat in stools on each of three days was 37 g., 134 g., 118 g. She was not on a strictly

measured intake, but appeared to put out in the stools slightly more than her estimated intake. The stools were voluminous but were passed only twice each 24 hours. They were frothy, strongly acid, and fermentative, and the lactic acid content of three 24-hour stools was 1.9 g., 3.6 g., 1.7 g.—a very excessive amount. Barium x-ray studies showed the small intestine to be still about 6 in. (15 cm.) in length and of a normal calibre. The stomach and colon were very large, and some barium entered the caecum within 15 minutes of leaving the stomach.

She was given an injection of magnesium sulphate intramuscularly and started on a regime of magnesium chloride 1.5 g. t.d.s. and increased potassium in her diet. The numbness and tingling sensations in her extremities disappeared and she has felt well since.

Her diet is to all intents and purposes normal. An estimate of her caloric intake indicates that it is in the region of 3,600 a day. Vitamin B₁₂ and vitamin B complex injections are given only about three-monthly. Calcium and calciferol are continued, and charcoal tablets reduce the offensiveness of her flatus, but this still remains somewhat of a social problem.

Discussion

The literature regarding survival after massive small-intestinal resection is voluminous. It is not the intention of this report to review the whole subject but just to record what seems to be a very unusual case—that of a girl, left with only 6 in. (15 cm.) of small intestine after surgery, who has been followed for seven and a half years, and who appears to have every chance of continuing to live a reasonably healthy and normal life, provided a certain regime is followed. No report of such long-term survival with so little small intestine has been encountered in the literature. There are several reports, including those of Pilling and Cresson (1957) and Althausen, Uyeyama, and Simpson (1949), describing cases followed for several years, who had 12 to 18 in. (30 to 45 cm.) of small intestine remaining. Jackson *et al.* (1951) recorded the progress of a man aged 29 who had survived for two and a half years with only approximately 6 in. (15 cm.) of small intestine, but who died a year later (Jackson and Linder, 1953). Lawler and Bernard (1962) describe the case of an infant who suffered a volvulus of the small intestine at 6 weeks of age. All but 10 cm. of jejunum and 5 cm. of ileum was removed. The infant had been followed until 18 months of age, and was then in good nutrition and passing five semi-formed stools each day.

Of course, survival after bowel resection depends on a number of factors, including the underlying disease, age and health of the patient, type of anastomosis, and region of small intestine resected. The present case offers some of the best circumstances—that of a young girl in good health whose bowel lesion was due to an anatomical anomaly, not to degenerative or neoplastic disease. However, she was severely ill with shock, circulatory failure, and dehydration at the time of the operation.

There are a number of interesting features in the patient's progress and treatment. Firstly, since shortly after operation she has not been troubled by actual diarrhoea. Although the stools are very large in volume they are passed only about twice a day. The stomach is large and the large intestine tremendously dilated, and this was so within six months of operation. The latter certainly seems to act as a reservoir, and probably allows absorption of fluid and electrolytes to be more complete. The large stomach probably assists in prolonging the period during which food enters the small intestine after a meal. Other authors have referred to dilatation of the colon and enlargement of the stomach (Jackson and Linder, 1951; Pilling and Cresson, 1957). It would be of some interest to know the absorptive capacity of the large intestine in such patients. Pilling and Cresson (1957) refer to greater absorption of water, salts, and protein from the large intestine. In our patient considerable fermentative activity was occurring in this region as the lactic acid content of the stool was extremely high, the stools were frothy, and much flatus was passed. Perhaps the large intestine may behave like the rumen of ruminants, where carbohydrate

is fermented by bacterial activity and volatile fatty acids are absorbed.

Whether the remaining small-bowel mucosa hypertrophies and the bowel dilates or elongates has been the subject of debate, with evidence put forward for both positive and negative viewpoints (Harrison and Booth, 1960). Much earlier, Flint (1912) suggested that the villi proliferated, and Clatworthy, Saleeby, and Lovingood (1952) found hypertrophy and dilatation in experimental puppies. We were unable to obtain evidence regarding the villi as an effort to obtain mucosa by peroral biopsy failed. However, x-ray films after seven and a half years were no different from the initial ones, and there was no dilatation of the short segment. There has also been no improvement in the absorption of fat. Nitrogen absorption has apparently been adequate, as serum proteins have remained normal and fairly adequate growth has occurred. It is remarkable that about 40% of the ingested protein can be absorbed by such a short small intestine. Is it possible that by bacterial activity in the large intestine amino-acids are formed and absorbed as happens in ruminants?

Haematologically no real trouble was encountered, but the patient was given vitamin B₁₂ replacement from the outset. Neither folic acid nor iron was given apart from that in the diet, yet these two substances appear to have been adequately absorbed, supporting the theory that their absorption is maximal in the upper small intestine.

The electrolytes most difficult to control have been calcium, potassium, and magnesium. During the early management of this case no attention was paid to magnesium replacements, but in the light of her later response to this electrolyte it seems that many of her symptoms during the first 18 months after operation might not have occurred if this had been given earlier. Perhaps her disturbed mental state, lethargy, and drowsiness might have been avoided. The man whose case was described by Jackson *et al.* (1951) showed a type of psychosis with some features resembling this girl's behaviour. There is no mention in their article of the giving of extra magnesium. Booth *et al.* (1963) have drawn attention to the importance of magnesium losses in patients with chronic steatorrhoea and diarrhoea, and Hanna *et al.* (1960) have described mental changes and even convulsions in cases with severe magnesium deficiency. Harrison and Booth (1960) suggest that a large excretion of fat in the stools encourages magnesium losses as magnesium soaps, and that a low-fat diet is advantageous. However, the patient would not keep to a low-fat diet for any length of time owing to its unpalatability and lack of hunger satisfaction.

The part played by chlorpromazine in improving the girl's general condition in the early stages is difficult to assess. Its administration certainly coincided with gradual and persistent improvement from what looked like a hopeless situation. It

was given to lessen misery. It certainly changed her mood and improved her appetite. Whether it had any other effect is open to conjecture. Extra magnesium was not given until the improvement was well under way.

If this case has taught anything it is probably that prompt and continuing attention to the intake of electrolytes such as calcium, potassium, and magnesium is a most important factor in ensuring survival. Vitamin B₁₂ therapy was also given early. With such a short length of bowel little seems to be achieved by dietary manipulation or additives such as pancreatic extract, emulsifying agents, protein hydrolysates, etc. The fact of survival with gain in weight indicates the tremendous reserves and ability for adaptation the human body possesses. It seems likely, from the cases recorded in the literature, that infants and young people adapt very much better to extensive losses of small intestine.

Summary

The clinical course of a girl who had all but approximately 6 in. (15 cm.) of her small intestine resected at the age of 14½ years has been described. She has now survived for over seven years and is in a state of good health and adequate nutrition. She is able to earn her own living and enjoy a reasonable social life. Prompt and careful attention to long-term electrolyte losses, especially of potassium, magnesium, and calcium, and the administration of vitamin B₁₂, seem the most important features of management. Dietary manipulation or additives were ineffective and difficult to maintain during a long period.

I am grateful to Dr. Ian Wood, and to Mr. Bruce Hallows, the surgeon who successfully resected the bowel, for referring this case to me for long-term management. Many other people have helped in her management over the years, particularly the clinical research ward registrars who inherited her care from each other.

REFERENCES

- Althausen, T. L., Uyeyama, K., and Simpson, R. G. (1949). *Gastroenterology*, **12**, 795.
 Booth, C. C., Babouris, N., Hanna, S., and MacIntyre, I. (1963). *Brit. med. J.*, **2**, 141.
 Clatworthy, H. W., Saleeby, R., and Lovingood, C. (1952). *Surgery*, **32**, 341.
 Flint, J. M. (1912). *Bull. Johns Hopk. Hosp.*, **23**, 127.
 Hanna, S., Harrison, M., MacIntyre, I., Fraser, R. (1960). *Lancet*, **2**, 172.
 Harrison, R. J., and Booth, C. C. (1960). *Gut*, **1**, 237.
 Jackson, W. P. U., Linder, G. C., and Berman, S. (1951). *S. Afr. J. med. Sci.*, **2**, 70.
 ——— (1953). *Metabolism*, **2**, 562.
 Lawler, W. H., and Bernard, H. R. (1962). *Ann. Surg.*, **155**, 204.
 Levin, H., Zamcheck, N., and Gottlieb, L. S. (1961). *Gastroenterology*, **40**, 818.
 Pietz, D. G. (1956). *Ibid.*, **31**, 56.
 Pilling, G. P., and Cresson, S. L. (1957) *Paediatrics*, **19**, 940.