Energy and nutrient intakes in cystic fibrosis

R M BUCHDAHL, C FULLEYLOVE, J L MARCHANT, J O WARNER, AND
M J BRUETON

Paediatric Cystic Fibrosis Clinic, Brompton Hospital, London

SUMMARY The diets of 20 children with cystic fibrosis were analysed for energy and nutrient content with simultaneous measurement of energy losses in stools. Median energy intakes were in excess of the WHO estimated daily requirements (118.2%) when expressed as MJ/kg/24 hours, the excess almost accounted for by energy losses in the stools. When expressed as MJ/24 hours, however, median energy intakes were 98.7% of that estimated for normal children of median weight for age. Compared with recently published data for normal school children the fat content of the diet was reduced (30.0%) as were intakes of iron and zinc. Children whose whole milk intakes were high had the greatest amount of fat and energy in their diets and were able to absorb energy in excess of that recommended. We conclude that many children with cystic fibrosis are still on low fat diets and whole milk is the single most useful food for the provision of extra dietary fat and energy.

The traditional dietary advice given to families with children who have cystic fibrosis has been that they should be given diets low in fat in order that symptoms of steatorrhoea secondary to pancreatic disease may be alleviated.¹ In 1974, however, Crozier and his colleagues in the Toronto cystic fibrosis clinic reported that by increasing dietary fat and giving appropriate enzyme treatment, children with cystic fibrosis could benefit from the increased energy density of the diet. Early studies confirmed improved growth velocities and showed higher circulating blood cholesterol and triglyceride concentrations.² In the late 1970s many of their patients had weights and heights comparable with those of their normal adolescent counterparts.³ This improvement in growth and nutritional state is associated with an increased survival, recently published figures giving a median survival of 30 years.⁴

In the United Kingdom the change to higher fat diets has been slower⁵ but in the past three or four years as the more effective enteric coated microsphere enzyme preparations have become more widely available, cystic fibrosis clinics have begun to adopt this policy. During 1986 we undertook a dietary survey of a group of children with cystic fibrosis to find out how far this change had progressed and what effect it had had on the nutrient composition of their diet and absorption of energy.

Subjects and methods

Twenty three children with cystic fibrosis confirmed by sweat iontophoresis were selected at random from the cystic fibrosis clinic. These children and their families had all received previous advice on nutrition that had emphasised the importance of not restricting fat. Families were interviewed at each outpatient attendance and give a pamphlet detailing recommended foods. For the outpatient study all the children were in a clinically stable state. Clinical assessment included scoring using the Shwachman method.⁶ Weight and height were measured using standard techniques and expressed as standard deviation scores (measured value—expected value for age divided by the standard deviation of expected value).⁷

Expected values and standard deviations for normal children were derived from the growth tables of Tanner and Whitehouse and of the Department of Growth and Development, Hospital for Sick Children, Great Ormond Street.⁸ The study was approved by the Brompton Hospital ethical committee and informed consent was obtained from both parents and children.

Dietary and absorbed energy intakes were calculated from seven day prospective records of the weights of all food consumed and three day collections of faeces. All energy containing foods and

Table 1 *Clinical data and nutrient intake/24 hours of 20 children with cystic fibrosis*

	Case numbers								
	1	2	3	4	5	6	7	8	9
Age (years)	5.3	6.2	8.0	8.6	8.8	9.5	9.6	10.0	10.4
Sex (M/F)	M	F	M	M	M	M	F	F	F
Weight (kg)	18.3	16.3	21.6	35.0	20.7	27.1	28.3	23.0	23.1
Energy intake:									
MJ	7.19	5.01	6.81	11.9	6.91	7.91	9.55	7.89	6.74
per kg	0.393	0.307	0.315	0.340	0.334	0.293	0.337	0.343	0.292
Percentage energy intake:									
As % of WHO estimated average daily energy requirements:									
Median weight									
for age	95	70	79	137	80	88	122	100	75
Actual weight									
for age ¹³	102	89	98	106	104	97	130	132	123
Coefficient of									
energy									
absorption	0.85	0.97	0.96	0.88	0.96	0.68	0.86	0.74	0.82
Fat (g)	41	45	52	78	36	52	92	88	49
Protein (g)	69	32	43	90	46	92	74	68	49
Carbohydrate (g)	286	177	264	474	305	282	307	278	261
Zinc (mg)	9.1	4.5	6.1	12.6	5.8	14.3	8.9	12.6	7.4
Calcium (mg)	1066	637	673	1547	627	953	1513	809	653
Iron (mg)	8.4	3.4	5.5	17.3	9.8	13.2	8.4	7.0	8.3
Enzyme preparations:									
None taken	No	Yes	Yes	No	No	No	No	No	No
Non-enteric									
coated									
preparation	Yes	No	No	No	Yes	No	Yes	No	No
Enteric coated									
microsphere									
preparation	No	No	No	Yes	No	Yes	No	Yes	Yes

drinks were weighed on Salter electronic scales and weights recorded in a diary (after subtracting plate waste).⁹ Children who were attending school recorded items of food and drink in a separate diary from which weights were later estimated. Compliance and technique were checked on home visits by a trained nurse technician. The available energy and macronutrient contents of the dietary records were calculated using a computerised version of the food tables of McCance and Widdowson.¹⁰ From these tables the zinc, iron, and calcium intakes were also calculated and compared with the recommended amounts published by the Department of Health and Social Security (DHSS) (for calcium and iron) and the United States National Academy of Sciences (for zinc).^{11 12} Energy intakes expressed as either MJ/24 hours or MJ/kg/24 hours for children with cystic fibrosis were compared with normal values derived from the WHO estimated average daily energy requirements for normal children on the basis of median weight for age and for kilogram of body weight.¹³

During the seven days of dietary recording, faeces were collected for three days using carmine dye capsules to mark a 72 hour sample. Three carmine

capsules (300 mg) were swallowed with the evening meal, and another three capsules exactly 72 hours later. Stools were collected in polythene bags from the time of appearance of the dye in the faeces until the second appearance of the dye. The bag was kept in a sealed plastic container at room temperature until the collection was completed. It was then delivered to the laboratory and stored at 4°C until analysis. The gross energy content of the faeces was determined by bomb calorimetry using a ballistic bomb calorimeter (Gallenkamp Ltd) after freeze drying a sample of the homogenate.¹⁴ Homogenisation was achieved by suspending the faeces in a known volume of water using Stomacher Lab-blender (AJ Stewart).

The coefficient of energy absorption was calculated using the formula: 1 – (gross energy of faeces divided by the gross energy of the diet). Gross energy of the diet was calculated from the available energy by using the factors 5.65 × g protein, 9.3 × g fat, and 3.75 × g carbohydrate and fibre.¹⁵ Absorbed energy was calculated by multiplying energy intake with the coefficient of energy absorption. Dietary records were completed and faecal collections were successful in 20 of the 23 children.

10	11	12	13	14	15	16	17	18	19	20
10.9 F 39.0	11.1 M 29.6	11.2 M 35.0	11.7 F 26.9	12.8 M 26.2	13.3 F 45.7	13.8 F 31.7	13.8 M 43.3	14.5 F 37.2	16.2 M 65.7	17.3 M 43.7
8.38 0.215	9.73 0.329	10.8 0.309	10.35 0.385	8.15 0.312	9.73 0.213	8.88 0.280	13.32 0.307	8.64 0.232	10.74 0.163	8.00 0.183
105	104	115	125	84	110	100	130	96	92	67
91	129	121	180	132	117	154	141	132	87	99
0.97 88 68 260 6.3 429 7.4	0.88 68 78 372 12.1 1286 15.5	0.92 106 85 342 11.3 1405 13.4	0.83 127 97 251 10.0 1851 8.9	0.91 57 66 309 9.6 1055 15.9	0.86 75 61 385 6.7 1018 7.7	0.81 76 76 301 9.0 676 10.4	0.91 133 95 424 12.3 1587 12.8	0.82 68 71 313 8.4 702 11.1	0.78 76 75 422 9.7 1012 10.9	0.86 45 73 324 8.0 874 13.0
Yes	No	No	No	No	No	No	No	No	No	No
No	No	No	Yes	No	Yes	Yes	No	Yes	Yes	Yes
No	Yes	Yes	No	Yes	No	No	Yes	No	No	No

Data was analysed by the Minitab statistics programme.¹⁶ Mean values for data were calculated (in most cases close to median values). Associations between variables were assessed by Spearman's rank and simple product correlation.

Results

Dietary records were completed and collections of faeces successfully carried out in 20 out of the 23 children (11 boys and nine girls). Their mean age was 11.1 years (range 5.3 to 17.3) and their Shwachman scores ranged between 57 to 94. Weight for age standard deviation scores ranged between -2.8 to +2.3 with a mean of -0.8. Seventeen out of the 20 children were taking enzyme replacements. In eight cases microsphere enteric coated preparations (either Pancrease or Creon) were used. There was no significant association between energy intake or absorption, and the type of enzyme replacement used. The three children not taking enzyme replacements had coefficients of energy absorption above 0.92.

ENERGY AND NUTRIENT CONTENTS OF DIETS

On average (SD) total 24 hour energy intakes were

Fig 1 Energy contributions from fat, protein, and carbohydrate as percentage of the total dietary energy intake in 20 children with cystic fibrosis compared with normals.⁹

close to the WHO estimated requirements for normal children of median weight for age ((98.7% (20%)). Median (SD) energy intakes expressed/kg body weight/24 hours were on average 118.2 (24%) above those of normal children. On average 30.0% of the total daily dietary energy intake was from fat (2.25 g/kg), 13.2% (2.21 g/kg) from protein, and 56.3% (10.63 g/kg) from carbohydrate. The balance of macronutrients compared with recently published values for normal children is shown in fig 1.⁹ When energy losses in the faeces were taken into account the mean absorbed energy intake was either 85.3% of the estimated normal requirement (for children of median weight for age) (SD 19.8%), or 101.9% (expressed as energy/kg) (SD 20.2%). There were significant correlations between absorbed energy intakes expressed as percentages of the normal estimated amount both for median weight for age ($r=0.52$, $p<0.05$) and energy/kg ($r=0.56$, $p<0.05$), and the proportion of fat in the diet, implying that children with a higher proportion of fat in their diets ate and absorbed proportionately more energy (fig 2).

ZINC, IRON, AND CALCIUM INTAKES

Seventeen out of the 20 children had inadequate zinc intakes compared with the recommended amounts,¹² and 13 out of the 20 had low iron intakes.¹¹ Only one child had a calcium intake less than that recommended for age.

MILK AND DIETARY SUPPLEMENTS

All 20 children drank whole milk and some also drank skimmed or semiskimmed milk. Fourteen children drank mainly whole milk, although in five cases skimmed milk was also consumed (usually with supplements). Six children drank mainly skimmed milk, all six also taking whole milk at

Table 2 Energy and macronutrient contributions from different milks and supplements

	Total No	Mean % daily dietary intake			
		Energy	Fat	Protein	Carbohydrate
Whole milk	20	10.9	18.2	17.2	5.5
Skimmed milk	11	3.2	0.3	9.4	4.1
Total milk	20	12.6	18.7	22.4	7.8
Supplements	7	3.9	0	5.3	6.9

times—for example, in coffee or tea. The energy and macronutrient contributions to the diet by whole milk, skimmed milk, and both combined are shown in table 2. For example, whole milk accounted for (on average) 10.9% of the daily dietary energy intake and 18.2%, 17.2%, and 5.5% of the fat, protein, and carbohydrate intakes by weight, respectively.

There were seven children who also took dietary supplements (Build-up, $n=6$; Maxijul, $n=1$; Polycal, $n=1$; Nesquik, $n=1$). Their standard deviation scores for weights and heights for age were not significantly different from those who were not taking supplements. The fraction of the total energy intake from supplements in the seven children was 3.9%. This compared with 10.9% from whole milk in all 20 children.

Discussion

The results of this survey suggest that many children with cystic fibrosis are still on low fat diets, but overall they seem to maintain energy intakes close to the estimated energy requirements for normal children. Using the WHO energy requirements tables¹³ it is possible to estimate the daily energy needs of a population of children of average (median) weight for age. The tables also allow adjustment for children of different weights at particular ages by listing the energy requirements/kg body weight. In this study we have included both comparisons because there is no accepted method for estimating the true requirements of a group of undernourished children with a wasting disease like cystic fibrosis. In so far as the energy requirements of an individual subject are determined by the active tissue mass of that individual, it would perhaps seem more reasonable to compare the energy intakes of children with cystic fibrosis with those of normal children of the same weight—that is by energy/kg. When this is done, many of the children with cystic fibrosis have above average energy intakes (mean

Fig 2 Absorbed energy expressed as a percentage of estimated daily energy requirement (energy/kg), and the percentage of the total dietary energy intake from fat.

118.2%) lending credence to the traditional observation that clinically stable children with cystic fibrosis often have voracious appetites.¹ This apparent increased energy intake presumably reflects increased energy demands either through energy losses in the stools or as a result of possible increased energy expenditure.¹⁷ Interestingly, after adjusting for energy losses in the stools the absorbed energy was close to the average estimated energy requirement (mean 101.9%). Because the cost of catch up growth for both weight and height in the clinical context of malabsorption and pulmonary disease is unknown, however, it could be argued that energy requirements for a group of children with cystic fibrosis would be better estimated on the basis of a group of normal children of median weight for age. When this is done the apparent energy intakes of the children with cystic fibrosis are less than those calculated on the basis of energy/kg (mean 98.7%). After faecal energy losses were taken into account many children fell short of the estimated energy requirements (mean 89.1%).

We found no significant difference in the coefficients of energy absorption between children taking enzyme replacements as encapsulated microsphere preparations and those on other preparations, a surprising finding in view of cross over studies that have shown that the recently developed microsphere encapsulated preparations significantly reduce steatorrhoea when compared with the older pancreatic preparations.¹⁸ It should be appreciated, however, that the subjects in the current study were unselected and examined cross sectionally. There was, perhaps, a tendency for children with less steatorrhoea and malabsorption to remain on the older preparations.

Though fat was often reduced in the children's diets there were increased fractions of carbohydrate and protein: indeed, mean protein intakes (2.21 g/kg/day) were over twice those recommended by WHO, not an uncommon finding in other nutritional surveys of patients with cystic fibrosis.^{1 19} The associations between the proportions of fat and carbohydrate in the diet and energy intake suggest that diets with higher fat and lower carbohydrate fractions result in improved energy intakes. Fat has about twice the energy density (39 kJ/g) compared with carbohydrate (17 kJ/g), or protein (23 kJ/g). In addition, if the goal of nutritional rehabilitation in cystic fibrosis is the synthesis of fat stores from which energy can be used at times of infection, thus sparing lean body tissue breakdown, then the most economical source of this body fat is dietary fat. This is because the metabolic cost of converting dietary fat to body fat is low compared with that of converting carbohydrate or protein, and in the

conversion of protein up to 30% of the energy value may be lost.²⁰

Deficiencies of the elements calcium, zinc, and iron have in the past been suggested as having a role in the pathogenesis of some of the clinical features of cystic fibrosis.²¹ In particular, low concentrations of iron have been associated with anaemia and low concentrations of zinc with failure of growth. Few studies, however, have looked at the dietary content of these elements. Though Bell *et al*¹⁹ found the calcium contents of their diets for cystic fibrosis adequate, they noted that some patients had low iron concentrations; our study confirms their findings. The results should, however, be interpreted with caution as iron intakes as low as these have recently been measured in apparently healthy British school children.⁹ Difficulties also arise with interpretation of the zinc intakes. Recommended intakes for zinc do not exist in the United Kingdom, and elsewhere in the world they vary widely; the United States¹² tends to recommend up to twice as much zinc as, for example, Canada.²²

The study was not specifically designed to find out why children were on low fat diets, but it was our impression that in some cases the importance of increasing the fat content of the diet had not been sufficiently emphasised by medical staff or dietitians. Many families were eating what they regarded as normal diets, which included a variety of foods with reduced fat contents—in particular skimmed milk products. In the last few years such families have become more aware of the need to reduce fat in their diets to lessen the risks of atherogenesis.

We found that the children who had the highest fat and energy intakes were those taking more milk, in particular more whole milk. Those children who consumed mainly skimmed milk derived less nutritional benefit from this constituent of their diet (table 2). This is hardly surprising when the nutritional compositions of whole and skimmed milk are compared (whole milk energy/100 g—272 kJ; energy proportions—fat 52.6%, protein 20.3%, and carbohydrate 27.1%; skimmed milk energy/100 g: 138 kJ; energy proportions—fat 2.7%, protein 41.2%, and carbohydrate 56.8%).

In some cases milk was taken with a dietary supplement but the nutritional contribution from the supplements themselves was low. Examination of the dietary records showed that in most cases the supplements were taken irregularly. Nutritional intervention studies have suggested that dietary supplements can improve energy intakes.²³ The object of the present study, however, was not to test the value of such supplements but merely to survey their use and contribution to the diets of a random sample of children attending the cystic fibrosis

clinic. To this extent we did not find them especially beneficial. We believe that whole milk has more to offer in terms of nutritional benefit, and perhaps more time should be spent in clinics emphasising the importance of whole milk in the diet instead of prescribing dietary supplements that can be expensive and are less readily available.

RM Buchdahl and JL Marchant were supported by grants from the CF Research Trust.

References

- ¹ Chase PH, Long MA, Lavin MH. Cystic fibrosis and malnutrition. *J Pediatr* 1979;**95**:337-47.
- ² Crozier DN. Cystic fibrosis. A not so fatal disease. *Pediatr Clin North Am* 1974;**21**:935-50.
- ³ Gurwitz D, Corey M, Francis PWJ, Levison H. Perspectives in cystic fibrosis. *Pediatr Clin North Am* 1979;**26**:603-15.
- ⁴ MacClusky IB, Canny GJ, Levison H. Cystic fibrosis: an update. *Pediatric Reviews and Communications* 1987;**1**:343-89.
- ⁵ Bell L, Durie P, Forstner GG. What do children with cystic fibrosis eat? *J Pediatr Gastroenterol Nutr* 1984;**3**(suppl 1): 137-46.
- ⁶ Shwachman H, Kulczycki LL. Long-term study of one hundred and five patients with cystic fibrosis. *Am J Dis Child* 1958;**96**:6-26.
- ⁷ Cameron N. The methods of auxological anthropometry. In: Falline F, Tanner JM, eds. *Human growth*. Vol 2. New York: Plenum Press, 1978.
- ⁸ Tanner JM, Whitehouse RH, Takaishi M. Standards from maturity for height, weight, height velocity and weight velocity: British children 1965—part 2. *Arch Dis Child* 1966;**41**:613-35.
- ⁹ Wenlock RW, Disselduff MM, Skinner RK, Knight I. *The diets of British school children*. London: DHSS, 1986.
- ¹⁰ Paul AA, Southgate DAT. *McCance and Widdowson's the composition of foods*. 4th ed. London: HMSO, 1978.
- ¹¹ Report by the Committee on Medical Aspects of Food Policy 1979. *Recommended daily amounts of food, energy and nutrient intakes for groups of people in the UK*. London: HMSO (DHSS No 19): 1985 (3rd impression).
- ¹² Food and Nutrition Board Committee on Dietary Allowances. *Recommended dietary allowances*. 9th ed. Washington: National Academy of Sciences, 1980.
- ¹³ World Health Organisation. *Energy and protein requirements. Report of a joint FAO/WHO/UNU meeting*. WHO Tech Rep Ser no 724, Geneva: WHO, 1985.
- ¹⁴ Miller DS, Payne PR. A ballistic bomb calorimeter. *Br J Nutr* 1959;**13**:501-8.
- ¹⁵ Southgate DAT, Durnin JVGA. Calorie conversion factors. An experimental reassessment of the factors raised in the calculation of the energy value of human diets. *Br J Nutr* 1970;**24**:517-35.
- ¹⁶ Ryan BF, Joiner BL, Ryan TA. *Minitab handbook*. 2nd ed. Boston: Duxbury Press, 1985.
- ¹⁷ Buchdahl RM, Cox M, Fulleylove C, et al. Increased resting energy expenditure in cystic fibrosis. *J Appl Physiol* 1988;**64**: 1810-6.
- ¹⁸ Beverley DW, Kelleher J, Macdonald A, Littlewood JM, Robinson T, Walters MP. Comparison of four pancreatic extracts in cystic fibrosis. *Arch Dis Child* 1987;**62**:564-8.
- ¹⁹ Bell L, Linton WL, Corey ML, Durie P, Forstner G. Nutrient intakes of adolescents with cystic fibrosis. *Journal of the Canadian Dietetic Association* 1981;**42**:62-70.
- ²⁰ Pencharz PB. Energy intakes and low-fat diets in children with cystic fibrosis. *J Pediatr Gastroenterol Nutr* 1983;**2**:400-2.
- ²¹ Silk DBA. Future directions in supplemental nutrition. In: Lawson D, ed. *Cystic fibrosis: horizons. Proceedings of the 9th International CF Congress, Brighton*. Chichester: John Wiley, 1984:99-114.
- ²² Advisory Committee to Revise the Canadian Dietary Standard. *Recommended nutrient intakes for Canadians*. Ottawa: Health and Welfare Canada, 1983.
- ²³ Anonymous. Supplementary nutrition in cystic fibrosis. *Lancet* 1986;**i**:249-51.

Correspondence to Dr RM Buchdahl, Department of Child Health, King's College Hospital, London SE5 8RX.

Accepted 15 July 1988