



GROWTH AND NUTRITION IN SOUTH AFRICAN CHILDREN WITH CYSTIC FIBROSIS

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Objectives. To study nutritional status and its dietary correlates in a South African cystic fibrosis (CF) population.

Design. Cross-sectional survey.

Population. Thirty-eight children and adolescents attending the CF clinic at Red Cross War Memorial Children's Hospital, Cape Town.

Methods. Standard anthropometry and a 3-day weighed food record.

Results. Median percentage expected weight for height (WFH) was 93 (interquartile range 84 - 101). Sixteen per cent of patients were below the 5th percentile for height. The proportion of patients who were malnourished (WFH less than 90) was greater among those over 10 years of age (47% v. 14.3%, $\chi^2 = 4.33$, $P = 0.037$). Sixty-eight per cent of patients consumed less than the recommended daily intake of energy. There was no correlation between WFH and energy intake. Fat intake represented 29.6% (interquartile range 27.5 - 33%) of daily energy intake.

Conclusions. Young South African children with CF are growing well despite relatively low intakes of energy and fat. Greater attention needs to be given to overcoming malnutrition among older children.

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Cystic fibrosis (CF) is the most common life-threatening genetic disease among Caucasian groups. It is characterised by chronic lung disease and nutrient malabsorption, both of which significantly compromise growth.¹ The lifespan of individuals with CF improves with nutritional status,² probably as a result of a reduction in the rate of progression of the lung disease in better-nourished patients.³ Nutritional management is one of the central tenets of CF care.

In the past four decades major strides have been made in the nutritional management of patients with CF. It has been

recognised that high dietary energy intakes may be required to achieve optimal growth and nutrition. An average energy intake exceeding 120% of the recommended daily intake (RDI) of non-CF patients is advised.⁴ Fat should constitute 35% of dietary energy intake.⁴ The development of efficient and effective pancreatic enzyme replacement formulations has made it possible to minimise malabsorption. Coinciding with better survival rates among CF patients are recent reports from Western countries which indicate that in the first decade of life at least, normal growth indices can be achieved in almost all young CF patients.^{5,6}

Survival of CF patients in South Africa lags a decade behind survival in Western countries.⁷ In a clinical and epidemiological study of CF conducted at Red Cross War Memorial Children's Hospital, Cape Town, in 1986, 36% of patients were below the 3rd percentile for weight and 24% below this level for height.⁸ Henley and Hill's report⁹ at around the same time showed that there was widespread ignorance of the nutritional principles of CF care among families dealing with the disease. If the survival of CF patients in South Africa is to improve, careful attention will need to be given to their nutritional care.

The present study was undertaken to provide up-to-date and detailed data on nutritional status and its dietary correlates in our CF clinic population. Such data will equip CF management teams to intervene more effectively at the individual and clinic levels in order to improve the nutritional status, and therefore the prognosis, of CF patients in South Africa.

METHODS

The study population consisted of children and adolescents who received regular care from the CF clinic at Red Cross War Memorial Children's Hospital in Cape Town. This clinic is attended by almost all children and adolescents with CF in the Western Cape. Patients under 2 years of age, those on continuous home oxygen therapy, and for logistical reasons those living more than 60 km from Cape Town were excluded.

Weight was measured using a scale accurate to 0.1 kg. Height was measured with a stadiometer (Seca, Germany) accurate to 0.5 cm. Mid-arm circumference (MAC) and triceps skin-fold thickness (TST) were measured using a plastic tape measure and a Harpenden calliper respectively. The weight and height measurements were transformed to percentage of ideal weight for age (WFA), height for age (HFA) and weight for height (WFH) using National Center for Health Statistics percentile charts.¹⁰ WFH less than 90% of expected was taken to indicate malnutrition.⁴ Percentiles for the upper arm indices were obtained from Frisancho.¹¹

Each family was supplied with an electronic scale to weigh all the food and drink the patient consumed over 3 consecutive days (2 weekdays and 1 weekend day). From this 3-day weighed food record each patient's daily dietary intake of

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energy, protein, fat and carbohydrate was calculated using the Foodfinder computer program (Version 1.10, National Research Programme for Nutritional Intervention, 1992). Stool was collected over the same 3-day period for stool fat estimation.¹² A coefficient of fat absorption less than 93% defined steatorrhea.⁴

Statistical analysis was undertaken using the Statistica computer package (Version 5.1 Tulsa, USA, 1997).

RESULTS

Of the 45 eligible patients, 38 (ages 2 - 18 years, median age 10 years) completed the 3-day weighed food record. Table I shows their anthropometric indices. TST and MAC measurements were taken for 21 of the 38 patients. Classification of nutritional status is in accordance with the recommendations of the Consensus Committee of the CF Foundation.⁴ One patient did not have pancreatic insufficiency. Median WFH was 93% (interquartile range 84 - 101%). Forty-seven per cent (8/17) of patients 10 years and older were below 90% of expected WFH. In contrast, 14.3% (3/21) of those under 10 years of age were below 90% of expected WFH. The difference was significant ($\chi^2 = 4.33$, $P = 0.037$). Mean HFA was 96% (standard deviation (SD) 4.7%).

Table I. Anthropometric indices in CF patients

	Frequency (%)
WFH > 110% (overweight)	5.4
WFH 90 - 110% (normal)	62.2
WFH 85 - 89% (underweight)	5.4
WFH 80 - 84% (mild malnutrition)	13.5
WFH 75 - 79% (moderate malnutrition)	5.4
WFH < 75% (severe malnutrition)	8.1
HFA < 5th percentile*	16.2
MAC < 5th percentile*	38.1
TST < 5th percentile*	28.6

* N = 21.

WFH = weight for height; HFA = height for age; MAC = mid-arm circumference; TST = triceps skin-fold thickness.

Daily energy intake exceeded 120% of the RDI in 8.1% of cases, while 68.6% of patients consumed less than 100% of the RDI. There was no correlation between WFH and daily energy intake ($r = 0.034$, $P = 0.84$). Fat intake represented 29.6% (interquartile range 27.5 - 33%) of daily energy intake. Protein and carbohydrate constituted a median of 16% (interquartile range 13.9 - 18.2%) and 51.7% (interquartile range 49.9 - 56.8%) of daily energy intake respectively.

Nineteen pancreatic insufficient patients (51.4%) completed the 3-day stool collection. The median coefficient of fat absorption was 89.09% (interquartile range 81.7 - 96%), with 7 patients (36.8%) achieving absorption coefficients above 93%.

DISCUSSION

Nutritional management is a cornerstone of CF care. The Consensus Committee of the CF Foundation advises regular monitoring of growth and energy intake as well as regular expert dietary guidance for CF patients.⁴ WFH is the recommended growth index for the assessment of nutrition in CF.⁴

In this study one-third of patients 18 years or younger had low WFH. That this deficit was nutritional in origin is supported by the similar proportion of patients with upper arm indices below the 5th percentile. In 16% of patients malnutrition had led to stunting of growth, although the mean height of 96% of expected was almost that of the normal population. Malnutrition was mainly found in patients 10 years of age and older. Only 3 of the 20 patients aged between 2 and 10 years (all of whom came from poor social circumstances) had WFH less than 90% (81%, 89%, 73%). It is important to note that infants and patients with very severe lung disease, in whom nutritional deficits are common, were absent from this analysis.

The nutritional status in CF patients reported here is surprisingly similar to that in reports from better-resourced centres. Using body mass index (BMI) as a measure of nutritional status, the most recent report from the UK CF survey⁵ shows that after the first 2 years of life mean BMI remains in the normal range until the age of 10 years, after which it declines. Likewise, North American 5 - 10-year-olds with CF have WFHs in the normal range.⁶ Reports from other centres published in the 1990s show a similar proportion of stunted patients to our cohort (Morrison *et al.* 14%,¹³ Wootton *et al.* 17%¹⁴). However, in common with comparable studies,^{5,6} we have shown that for the majority of young patients with CF, stature in the normal range can be expected.

In our study, nutritional status after the first decade of life deteriorated to the point where 47% of patients were malnourished. In contrast, similar patients in the UK have a BMI less than 1 SD below population norms.⁵ Declining adherence to therapeutic and dietary regimens among adolescents is common. Compared with the UK, South African CF clinics have fewer clinic and community resources to counteract this tendency. Given the poorer prognosis of CF in South Africa,⁷ adolescents are likely to have more severe lung disease than their UK counterparts. This will also make it more difficult to achieve adequate nutrition and growth.

Logically it would seem probable that those with the lowest relative energy intake would have the greatest degree of malnutrition. We were unable to show this. This may reflect the small sample, although two studies with similar numbers of patients were able to show some correlation between energy intake and measures of nutritional status.^{14,15} This lack of correlation may also cast doubt on the accuracy of the weighed



food records. However, other results based on these records were in accord with previous studies of CF patients. For example, the finding that most patients did not reach the normal RDI for energy or fat, let alone the higher intakes recommended for CF patients, is not unique to Cape Town.^{14,15}

Despite the relatively low dietary energy and fat intakes, 67.6% of our patients had achieved WFH above 90%. This apparent paradox may be because on average CF patients weigh less than their peers.¹³ As Wootton *et al.*¹⁴ point out, since RDI is calculated on an individual's age rather than weight, energy needs will be overestimated in CF patients.

Our results indicate that dietetic advice needs to be individualised, with specific attention being targeted at those with declining WFA, HFA or WFH, and those approaching their teenage years. The emphasis will need to be on increasing fat intake, as patients with higher fat intakes absorb higher amounts of energy.¹⁵

Only half the patients completed the 3-day stool collection. Despite pancreatic enzyme replacement therapy, and in common with their counterparts in developed countries,^{6,13,14} the majority of patients tested were malabsorbing fat. This will have contributed to energy deficits and consequent malnutrition. Further education and closer attention to the detail of enzyme use will be necessary to improve fat absorption.

The gains made in the prognosis of CF in recent decades have largely been due to the greater attention given to the details of conventional care, i.e. dietary management, pancreatic enzyme replacement therapy, physiotherapy, and treatment of respiratory infections. Our study has shown that with regard to nutrition, the majority of our young CF patients compare well with patients from better-resourced countries. However, it is also plain that for some patients, and especially for adolescents,¹ greater attention to nutritional care and enzyme use is needed, both of which require ongoing expert medical and dietetic care. The data in this study were collected, in part, to assist in tailoring advice to the individual and family. The recognition of persisting dietary energy and fat deficits and the need to maintain gains made in the first decade of life have led to the publication of a South African book of nutritional advice for families dealing with CF.¹⁶

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