Height and weight in cystic fibrosis: a cross sectional study

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Abstract

Cross sectional data reporting the height, weight, and body mass index of UK patients with cystic fibrosis are presented. During the first decade of life height and weight in patients with cystic fibrosis are maintained at about 0.5 SD below those of the general population, which reflects an improvement over earlier published observations. Postpubertal stature and weight maintenance in the cystic fibrosis population still show substantial deficits which may be related to treatment.

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Keywords: cystic fibrosis; height; weight; body mass index

The growth of children with cystic fibrosis may be an indication of the effectiveness of treatment. There is well documented evidence that the survival of patients with cystic fibrosis in the UK has improved over several decades and that it may vary considerably between centres,[1] although the reasons for this variation are not known. There also appears to be an important positive association between survival and body weight.[2 3] The factors which lead to the slowing of growth or failure to maintain body weight are complex, but there is a widespread impression among paediatricians who direct cystic fibrosis clinics that these complications can be postponed by effective treatment of the disease, so that growth in childhood may be virtually indistinguishable from normal. For these reasons, the UK Cystic Fibrosis Survey (UKCFS) obtained cross sectional data on height and weight in a large sample of the cystic fibrosis population to document their basic anthropometric data.

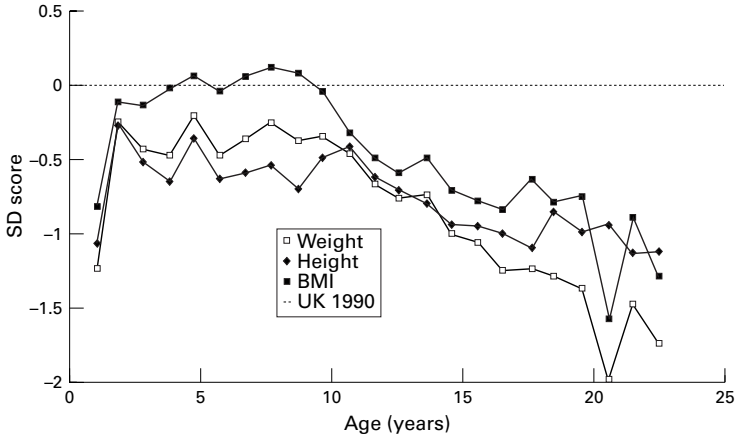

Figure 1 Mean weight, height, and BMI by age in male patients with cystic fibrosis expressed as SD scores relative to the British 1990 growth reference.[4 5]

Subjects and methods

The study was a cross sectional survey of patients attending recognised centres for the treatment of cystic fibrosis. During the period in question there were 36 such large clinics, with a minimum of 40 patients in each, distributed throughout England, Wales, Scotland, and Northern Ireland. Approximately 65% of the total cystic fibrosis population had some contact with these clinics. The centre may have provided all medical care, or conducted an annual review with day to day management being given at a peripheral hospital through a shared care arrangement. Careful measurement of height and weight, with minimal clothing, was recorded for each patient on one occasion only. Where more than one measurement was reported, only one, randomly chosen, was used. The data obtained were entered into a computer along with the date of the measurements and the patients' date of birth. The data were then analysed according to a previously published programme used for the derivation of general population centiles.[4 5] The results have been calculated as centiles and as SD scores.

Body mass index (BMI) was calculated as weight/height^2 (kg/m^2) and the data were then analysed in two ways. Firstly, height, weight, and BMI up to the age of 23 years were expressed as SD scores relative to the British 1990 growth reference.[4 5] Secondly, cystic fibrosis reference centile curves were constructed for data in the age range 0–30 years using the LMS method and maximum penalised likelihood.[6]

Results

Data were obtained from 31 of the 36 large clinics. Measurements were made between June 1994 and March 1995, on one occasion in each case, when patients attended the cystic fibrosis clinics for routine follow up. The total number of subjects measured was 3056, comprised of 1604 male and 1452 female patients. This represents 48.5% of the then current total UK cystic fibrosis population and 75% of patients treated in those centres. Measurements of 1338 male and 1232 female patients less than 23 years old were used to derive SD scores relative to the British 1990 reference (figs 1 and 2), whereas 1502 male

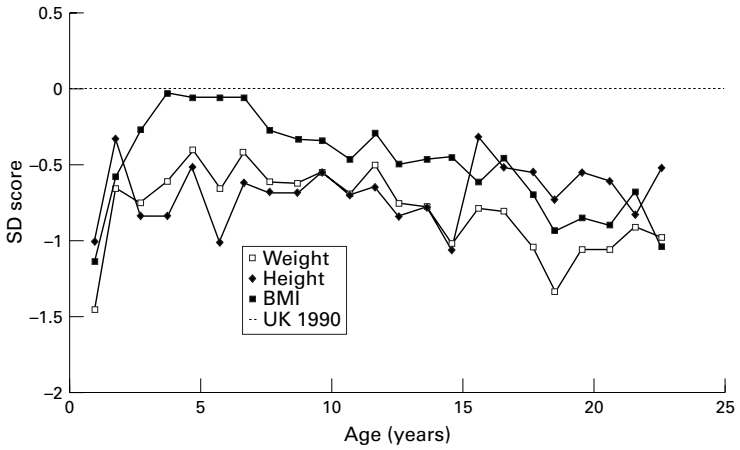

Figure 2 Mean weight, height, and BMI by age in female patients with cystic fibrosis expressed as SD scores relative to the British 1990 growth reference.[4 5]

and 1381 female patients less than 30 years old provided the cystic fibrosis reference centiles for height, weight, and BMI in figs 3 to 8.

Discussion

The sample of patients reported here was not drawn randomly from the whole UK population, so caution is required when extrapolating these results to other groups. We have no reliable information which would allow us to infer that patients obtaining all of their care from non-specialist clinics are similar to those seen in the participating cystic fibrosis centres.

The data confirm that the height and weight attained by patients with cystic fibrosis, as judged from a single 'snapshot' assessment, shows an improvement relative to similar measurements made in previous decades. Thus the weight SD measurements show a pattern similar in shape to that reported in American children with cystic fibrosis by Berry in 1975,[7] and similar to that in an unpublished British study of about the same time.[8] The present study shows important improvements: the plateau between ages 1 and 10 years is approximately 0.5 SD below the population mean, compared with a full SD in the American data of 1975.[7] Furthermore, the apparent decline from the population norm in older patients is slower. Nevertheless, the data suggest substantial continuing deficits in both weight and height during the first year, although this could in part reflect poor ascertainment of the milder

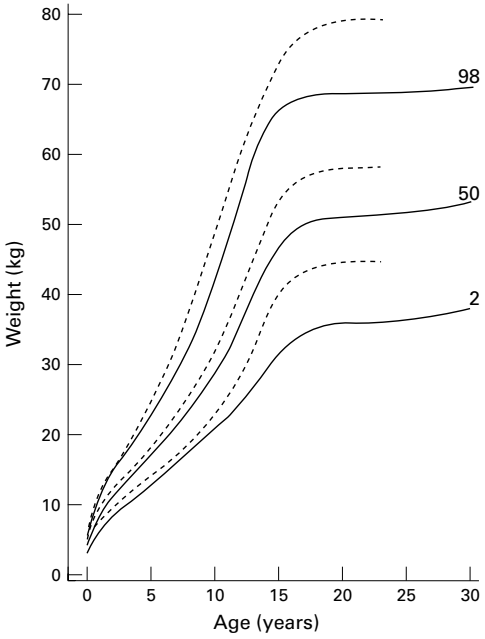

Figure 4 Reference centiles for weight in female patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

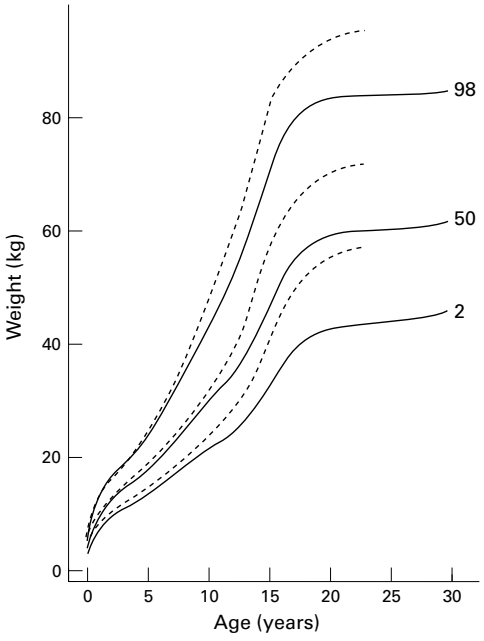

Figure 3 Reference centiles for weight in male patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

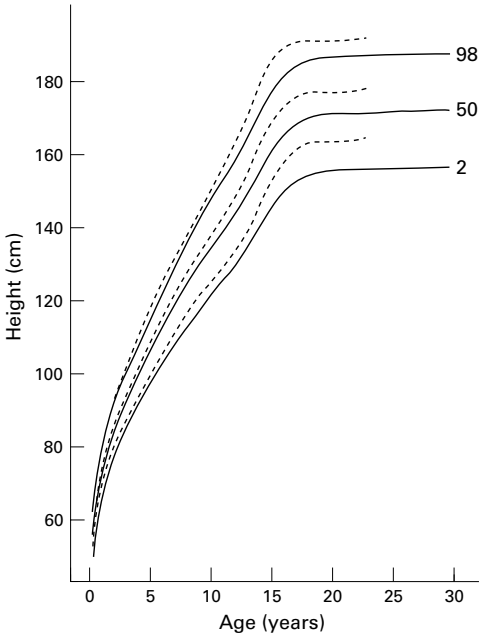

Figure 5 Reference centiles for height in male patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

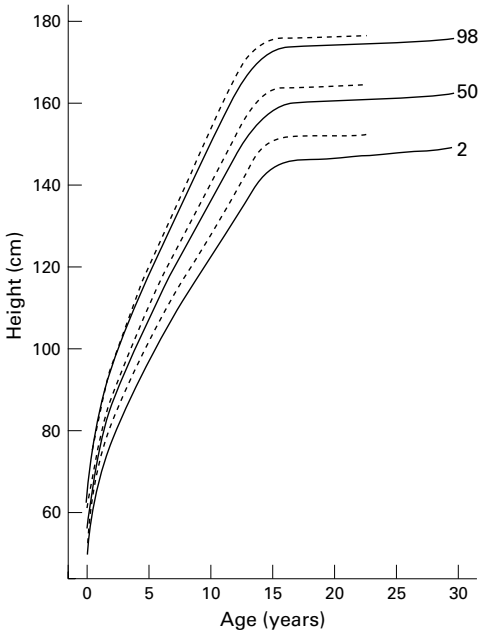

Figure 6 Reference centiles for height in female patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

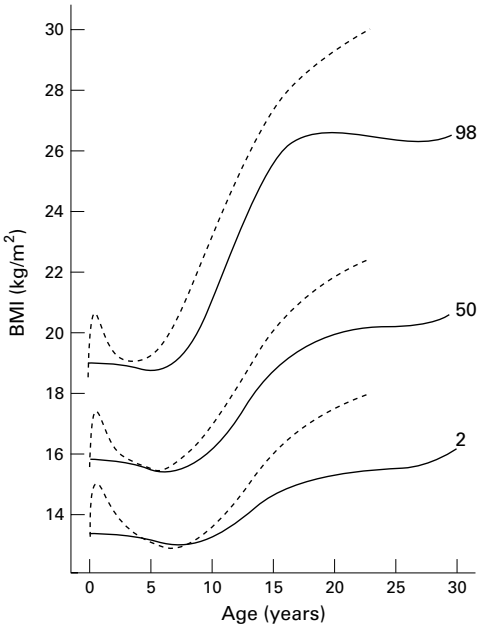

Figure 8 Reference centiles for BMI in female patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

cases in the early years of life. Reversion of SD scores towards the mean takes place by 2 years of age and does not further improve thereafter. This improvement may be partly explained by the inclusion of late diagnosed mild cases in the older cohorts, but as yet unpublished data from the UKCFS show that currently about 70% of patients in the UK are diagnosed by 1 year of age. The better status of 2–3 year olds will therefore probably also reflect 'catch up' growth which follows effective treatment.

The mean body weight in boys with cystic fibrosis remains between −0.25 and −0.5 SD until the age of 10 years; the BMI is maintained until 10 years of age (figs 1 and 7). In girls the mean body weight approximates 0.5 SD below the population mean during the same period, but the BMI apparently decreases from the age of 5 years onwards (figs 2 and 8). There is a wide range, however, and the SD for measurements at different ages remain high, as will be apparent from the centile charts on which the UK population curves are superimposed. The BMI in men decreases further after the age of 20 years, but continues to increase in women. The reason for this is not clear, but it may reflect a bias towards survival in more mildly affected women, although overall female patients experience a higher mortality than male patients. In this context it must be recognised that all older patients are survivors from earlier cohorts in which the age/sex specific mortality was higher than for contemporary younger cohorts and the treatment antecedents were different from those offered now.

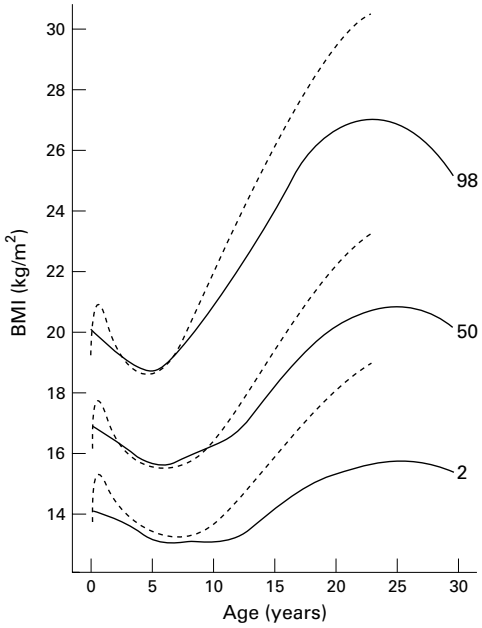

Figure 7 Reference centiles for BMI in male patients with cystic fibrosis showing 2nd, 50th, and 98th centiles (solid lines) and the British 1990 reference centiles for comparison (broken lines).

Several workers have documented low birth weight in patients with cystic fibrosis[8 9] and it is not clear whether this contributes to the initial weight and height deficit in the youngest patients reported, nor whether it indicates an intrinsic constraint on growth in cystic fibrosis. If this were so, then the normal mean BMI in the first decade might reflect optimum nutrition in this age group. It is also arguable that the severity and duration of the initial deficits might be reduced by earlier diagnosis—for example, by neonatal screening—but there are no published data to indicate whether this would have an effect on subsequent anthropometric patterns.

The decrease in BMI in boys after the age of 10 years may be in part a reflection of delayed puberty in boys with cystic fibrosis. Although the stature and body mass of patients with cystic fibrosis appears to be improving, in

association with their improved survival, growth still remains below that of the unaffected general population to an important degree, as is clearly shown in figs 1 and 2. A decrease from population norms occurs earlier in girls than in boys, but is less steep and more protracted. This may be associated with the clinical observation that women with cystic fibrosis who survive into their 20s have no greater risk of death than men with cystic fibrosis of a similar age,[10] but this may again reflect cohort survival bias.

The survival of the UK cystic fibrosis population continues to improve, with year on year reductions in the age/sex specific mortality.[11] If the growth/survival association is maintained, further improvements in these anthropometric measures may be anticipated.

Conclusions

Despite clinical impressions and demonstrated improvements during the last 20 years, the height achieved by patients with cystic fibrosis remains below that of the general population. There is also still a problem with the weight of adults which may be due to their treatment antecedents. Attention to these nutritional deficits might further improve survival.

This paper was only made possible by the cooperation of the directors of cystic fibrosis centres throughout the UK, who generously contributed their data to the UK Cystic Fibrosis Survey.

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