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# Growth assessment in Egyptian children with cystic fibrosis: A single center study



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#### ABSTRACT

Background: Poor clinical outcomes in cystic fibrosis are often associated with undernutrition. Nutritional status is an important prognostic marker, regardless of the level of respiratory function in the patient. Careful and repeated nutritional assessments allow for early detection of nutritional deterioration.

Objectives: To assess the growth of cystic fibrosis children using different parameters as body mass index percentiles (BMIP) and standard anthropometric indexes, including height-for-age percentile (HAP), weight-for-age percentile (WAP) and compare between them as measures for nutritional failure in children with cystic fibrosis (CF).

Methodology: This was a cross sectional study including fifty children of both sexes with CF below 12 years of age, recruited from the cystic fibrosis clinic, Specialized Children's Hospital, Cairo University. Patients were subjected to full history taking including age at diagnosis, frequency of hospital admissions (per year), current medications and presence of complications. Thorough clinical examination was performed with special emphasis on anthropometric measurements including body weight, length/height, BMI calculation.

Results: This study included 30 boys and 20 girls between 2 and 11 years old with a mean of  $4.25\pm2.30$  yrs. Pancreatic insufficiency was found in 36% of patients, 28% had failure to thrive as the main complaint while 72% had recurrent chest infection as the main complaint. Growth assessment revealed that 21 patients (42%) were below -2 SDS for weight. As for BMI, 14 patients (28%) were below the 10th percentile. A highly significant negative correlation was reported between the weight SDS and the frequency of hospital admission (r = -0.412, p = 0.003). Furthermore, there was a highly statistical significant difference between the malnourished patients and the patients with average weight regarding the frequency of hospital admission (p < 0.01). As for BMI, there was a statistically significant negative correlation between the BMI and the frequency of hospital admission (r = -0.350, p = 0.013), with the different categories of BMI (malnourished, those who need nutritional reassessment, normal and obese) having highly statistical significant difference as regards the frequency of hospital admission (p < 0.01). Conclusion: Malnourished CF patients had the highest frequency of hospital admission. BMIP predicts nutritional failure more sensitively and accurately than conventional anthropometric indexes (WAP and HAP) in children with CF.

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#### **Background**

Cystic fibrosis (CF) is an autosomal recessive genetic disease characterized by chronic pulmonary infection, exocrine pancreatic insufficiency, and high concentrations of sweat electrolytes. Respiratory system changes constitute the primary cause of morbidity and mortality. CF is caused by mutations in the cystic fibrosis

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conductance regulator (CFTR) gene and usually presents with multiorgan involvement.<sup>2</sup>

Malnutrition seen in this patient population is a result of the energy losses and nutrition deficits caused by malabsorption, CF-related liver disease (CFLD), CF-related diabetes (CFRD), and various psychosocial issues, including stress and noncompliance.<sup>3</sup>

Exocrine pancreatic insufficiency (PI) is the most common gastrointestinal complication associated with CF and is seen in 85%–90% of the population. This is due to a mutation of the CFTR protein, which results in abnormal sodium and chloride transport. This causes the pancreas to have abnormally viscous secretions as

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well as an acidic lumen. The accumulation of mucous leads to ductal obstruction which causes tissue degradation, fibrosis, fatty replacement and ultimately, pancreatic insufficiency. The exocrine function of the pancreas is disrupted and the pancreas is unable to release the necessary enzymes critical to the digestion and absorption of fat, protein and fat-soluble vitamins. Pancreatic enzyme replacement therapy (PERT) is used to manage PI and associated malabsorption.

Current preparations of PERT consist of enteric coated microspheres that are designed to decrease enzyme deterioration by stomach acid and improve the efficacy of enzyme release in the duodenum, when pH exceeds 5.8. Despite optimizing PERT, CF patients continue to experience fat malabsorption, displaying 85%–90% of intestinal fat absorption as opposed to the >95% fat absorption of non CF individuals.<sup>5</sup>

The early growth pattern of infants with CF is dependent on both the age at diagnosis and the quality of the subsequent treatment they receive. The majority have pancreatic insufficiency and experience early gastrointestinal symptoms, which if not adequately treated can lead to subnormal weight gain. Even some infants diagnosed by neonatal screening have subnormal growth throughout the first year if the start of treatment is delayed by more than a few weeks after birth. The nutritional status of individuals with CF tends to decline during childhood. Data from (CFF) shows that the body mass index (BMI) percentile typically begins to decline at about five years of age but does not cross the 50th percentile (defined as "below BMI goal") until about nine years of age.

Poor clinical outcomes in cystic fibrosis are often associated with undernutrition. Normal growth and development should be achieved in CF, and nutritional counseling is paramount at all ages. Prevention and early detection of growth failure is the key to successful nutritional intervention. The advance in nutritional management is certainly one factor that has contributed to the improved survival in recent decades.<sup>8</sup>

For children with CF, the BMI target range is above the 50th percentile. Children with BMIs between the 10th and 50th percentiles are generally considered at nutritional risk, and those with BMIs below the 10th percentile are in need of nutritional rehabilitation. For children younger than two years of age, the same percentile criteria are applied to weight-for-height rather than BMI. 9

#### Aim of the work

To assess the growth of cystic fibrosis children using different parameters as body mass index percentiles (BMIP) and standard anthropometric indexes, including height-for-age percentile (HAP) and weight-for-age percentile (WAP) and compare between them as measures for nutritional failure in children with cystic fibrosis (CF).

#### Patients and methods

Our study was a cross sectional study which included fifty children of both sexes with cystic fibrosis (diagnosed by clinical manifestations and confirmed by sweat chloride test) below 12 years of age, recruited from the cystic fibrosis clinic, Specialized Children's Hospital, Cairo University. Patients were recruited after obtaining an informed consent from their legal guardians. They were subjected to full history taking including personal data, main clinical presentations (respiratory symptoms, GIT symptoms, etc.), age at diagnosis, frequency of hospital admissions (per year), current medications and presence of complications. Thorough clinical examination was performed with special emphasis on anthropometric measurements including body weight, length/height, BMI

calculation. Weight was measured in kilograms using Seca delta Mod 707 standing scale, height was measured in centimeters using Harpenden stadiometer, BMI was calculated as weight in kilograms divided by the square height in meters [wt (kg)/ht (m) $^2$ ]. Growth assessment was done through plotting these results on the Egyptian Growth Charts. The data analyzed using SPSS. Descriptive analysis of data will include mean and the standard deviation for numerical data, frequency and percentage for qualitative data. Comparison of quantitative and qualitative data between groups will be performed using Student t test and Chi square test respectively. Pearson Correlation test will be used to correlate quantitative variables. P-value will be considered significant if less than 0.05.

#### Results

The current study included 30 boys (60%) and 20 girls (40%) with an age range between 2 and 11 years old and a mean value of  $4.25 \pm 2.30$  yrs. The history of NICU admission was reported in 16% of patients (2 of them due to pneumonia and 6 due to prematurity). Pancreatic insufficiency was found in 36% of patients, 28% of patients had failure to thrive as the main complaint while 72% had recurrent chest infection as the main complaint.

Descriptive data concerning hospital admission per year, weight (kg.), height (cm.) and SDS for weight and height as well as BMI is shown in Table 1.

Growth assessment in our study revealed that 42% of patients were <-2 SDS for weight while 58% were between -2 and 2 SDS, 50% of patients were <-2 SDS for height and 50% were between -2 and 2 SDS for height.

As regard the BMI, 28% of patients were below the 10th percentile (malnourished), 24% between the 10th and 50th percentile (in need for nutritional assessment), 22% between the 50th and 85th percentile (normal) and 26% above the 85th percentile (obese).

There was no significant correlation between the age of patients and the frequency of hospital admission (Table 2). A highly significant negative correlation was reported between the weight SDS and the frequency of hospital admission per year (r = -0.412, p = 0.003) (Fig. 1, Table 2). Furthermore, there was a highly statistical significant difference between the malnourished patients and the patients with average weight regarding the frequency of hospital admission per year with a mean value of  $2.62 \pm 1.04$  and  $1.49 \pm 0.70$  respectively (p < 0.01) (Table 3).

A statistically significant negative correlation was detected between the height SDS and the frequency of hospital admission (r = -0.307, p = 0.03) (Fig. 2, Table 2). However, no statistically significant difference was found between patients with short stature and the patients with average height regarding the frequency of hospital admission with a mean value of  $1.99 \pm 0.96$  and  $1.92 \pm 0.92$  respectively (p = 0.819) (Table 3).

As for BMI, there was a statistically significant negative correlation between the BMI and the frequency of hospital admission (r = -0.350, p = 0.013) (Table 2), with the different categories of

**Table 1**Descriptive data concerning hospital admission per year, weight (kg.), height (cm.) and BMI.

Descriptive data	Min.	Max.	Mean ± SD
Hospital admission per year	0.40	4	1.96 ± 1.02
Weight (kg.)	6.50	30	14.29 ± 5.08
Weight SDS	-3.72	0.9	$-1.180 \pm 1.120$
Height (cm.)	69	132	91.53 ± 14.33
Height SDS	-5.3	0.19	$-2.15 \pm 1.38$
BMI [wt./(ht.) <sup>2</sup> ]	12	24	16.46 ± 2.75

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