
Nutritional supplements in cystic fibrosis

A new study suggests they're ineffective, but concordance may have been poor

People with cystic fibrosis and their caregivers know that the better the patient's nutritional status, the better their survival, lung function, wellbeing, and mental capabilities.¹⁻³ Poor nutrition is also associated with delayed puberty, a higher risk of pneumothorax, and a worse outcome after lung transplantation.^{4,5}

The increased demand for energy in cystic fibrosis is well recognised.⁶ Despite this, almost 20% of children with cystic fibrosis fall below the fifth centile for weight set by the Centers for Disease Control and Prevention,⁷ and their height and weight z scores often decline with age. Combating nutritional failure to ensure normal growth and weight gain is therefore a key challenge for

the entire team—doctors, nurses, dietitians, psychologists, and social workers.

Nutritional management in cystic fibrosis is difficult for several medical and psychosocial reasons. One priority is the optimal medical management of malabsorption from exocrine pancreatic insufficiency and deranged enterohepatic circulation of bile. Patients continuously have to adjust the dose of exogenous pancreatic enzymes they need to take to allow assimilation of ingested food. Meanwhile, patients often need lung clearance techniques and antibiotic treatments to reduce the increased energy demand from chronic pulmonary infection and inflammation and breathing difficulties. These toddlers, children,

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adolescents, and adults have to integrate into “normal” life this huge burden of lifelong daily treatment.

Mealtimes can be psychological battlefields for such families, with eating often reflecting a child’s first experience of power struggles. Difficulties may recur in puberty, an age that may be critical for the further course of the disease; they are often exacerbated by the need for patients to conform to today’s ideal that “thin is beautiful.” Just as treating patients with anorexia nervosa needs more than simply prescribing supplements, efforts to clarify the underlying psychodynamic processes in cystic fibrosis may be critical for success.

In this issue (p 632) Poustie and colleagues present an important, well planned, and carefully conducted randomised multicentre study of oral supplements in cystic fibrosis patients.⁸ Concordance with the intervention was reinforced continuously by staff making regular home visits. Nevertheless, the prescription of extra oral supplements failed to improve the nutritional status of the patients.

The trial was conducted to detect a 10 point difference in centile for body mass index within one year of the intervention, comparing patients receiving dietary advice with patients who were also prescribed oral energy supplements. This means that the trial was powered to detect, for example, a difference in weight gain of 2 kg between two 9 year old girls allocated to the different treatment groups. This was a noble goal, but an over-optimistic end point for a clinical trial in cystic fibrosis.

The authors say that the intervention failed because of inconsistency or inaccuracy in the dietary records and, perhaps, because patients did not take the supplements they were prescribed. In addition, the appropriately used statistical method, intention to treat analysis, may have led to underestimation of the effects of the intervention, but a per protocol analysis was not possible in this trial because adherence to treatment was unknown. Therefore conclusions from the trial must be drawn very carefully.

Treatments for cystic fibrosis are time consuming, generally unpleasant, and potentially anxiety provoking, and this can be particularly problematic when patients are feeling generally healthy. Professionals often have to “sell” such medical options to patients. Powers and colleagues have reported how they successfully influenced the behavioural aspects of nutrition in children with cystic fibrosis by using a multidisciplinary approach.⁹ The “intention to feed” is certainly not enough.

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