Nutritional risk in children with cystic fibrosis is associated with reduced lung function, pancreatic insufficiency and gender *

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BACKGROUND

Nutritional status is correlated with lung health and disease severity in cystic fibrosis patients and is an independent predictor of mortality. Cystic fibrosis patients may have impaired pancreatic and intestinal function resulting in malabsorption of fat-soluble vitamins and reduced nutritional status. No previous data has been published pertaining to the Western Australian cystic fibrosis population. As Princess Margaret Hospital is the sole paediatric cystic fibrosis care centre for Western Australia, it represents the entire cystic fibrosis population of Western Australia. This study describes the cystic fibrosis population of Princess Margaret Hospital in terms of nutrition risk, lung function, rate of hospitalisation and fat-soluble vitamin status and examines the associations between these factors.

METHOD

A cross-sectional, retrospective medical record audit was undertaken of 162 children diagnosed with cystic fibrosis in Western Australia and admitted to Princess Margaret Hospital. Gender; age; height (cm); weight (kg); number of hospitalisations over 12 months; serum fat-soluble vitamins levels (vitamin A: serum retinol; serum 25 hydroxy vitamin D; vitamin E: serum α-tocopherol, vitamin K: serum prothrombin); pancreatic function (sufficient/insufficient); and lung function (forced expiratory volume [FEV1%]) were collected. Patient nutritional risk was calculated based on BMI percentile and change in weight and categorised as High Nutrition Risk, At Nutrition Risk, Not At Nutrition Risk. χ^2 analysis and one-way ANOVA were used to determine the relationship between risk category and categorical variables (gender and pancreatic sufficiency) and continuous variables (lung function, age, fat-soluble vitamins) respectively.

RESULTS

We determined that 20% (*n*=32) of the Princess Margaret Hospital cystic fibrosis population were at High Nutrition Risk, 9% (*n*=15) were At Risk and 71% (*n*=113) were Not At Risk. There was a significant difference in distribution of gender across the nutrition categories (χ^2 =8.37, P=0.015). Patients classified At Nutrition Risk or High Nutrition Risk (29.4%) were more likely to be female (21%) than male (13%). Seventy-five per cent (*n*=24) of the subjects in the High Risk category and 66% (*n*=10) of the subjects in the At Risk category were female. The number of males and females in the Not At Risk category was not significantly different. The mean ages of patients in the High Risk and At Risk categories were greater than those in the Not At Risk Category (F=5.032, P=0.008).

The mean FEV1% was 97.3% (SD 16.1). Patients in the High Risk (87±13%) and At Risk (89±13%) categories had a significantly reduced score when compared to the patients in the Not At Risk category (102±15%, F=10.68, P=0.001). Those in the High Risk or At Risk categories were more likely to have pancreatic insufficiency and reduced lung function when compared to those in the No Risk category. There was no significant difference in the mean levels of fat-soluble vitamins between nutrition risk categories. A χ^2 test of the mean number of hospitalisations over the past 12 months showed no significant difference between nutrition risk categories.

Serum vitamin retinol levels were within the reference range for 87% of subjects, below range for 5% and above range for 8%. Serum 25 hydroxyvitamin D levels were below the cut off (>75 nmol/L) for 57% of subjects and within range for 43%. Levels of α-tocopherol were above the reference range (7–30 nmol/L) in 29% of subjects, within range for 71% and below range for 6%. Prothrombin times were within range (12.3–18 seconds) for 89% of subjects, with 9% below range and 2% above.

CONCLUSIONS

These data confirm an association between nutrition risk and gender, age, pancreatic insufficiency and reduced lung function in the Western Australian cystic fibrosis population. The prevalence of nutrition risk was lower than anticipated but higher among females and those who were older. Those categorised as High Nutrition Risk had a lower lung function and more likely to have pancreatic insufficiency. Further exploration into the reasons for these differences should be undertaken. Patients in the High Nutrition Risk and At Nutrition Risk populations may benefit from increased provision of dietetic intervention.

No association was found between fat-soluble vitamin status and the health outcomes measured, although the

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high prevalence of vitamin D deficiency should be acknowledged and additional vitamin D supplementation should be commenced. This could be achieved in addition to the current supplementation regimen or may be grounds for the reformulation of the current supplement used. Further exploration of vitamin K status may be indicated as combined vitamin D and K deficiency may increase the risk of poor bone mineralisation and osteoporosis in the population. Further study on the bone density of this population with regard to vitamin D and K status is recommended.