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Original Article

Nutritional considerations for a new era: A CF foundation position paper

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Overview

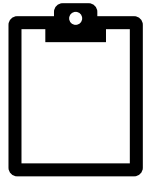
Objective:

- To provide interim advice and considerations to CF community around CF nutrition in the current era.
- Insufficient evidence to develop a formal guideline but need for guidance.

Scope: Covers children and adults

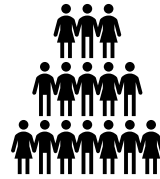
Methods

Survey



- Survey to pwCF/families and CF providers
- 20 questions on nutrition related issues

Responses

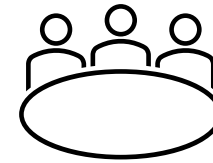


155 pwCF



422 CF providers

Workshops on 8 topics



Topics based on survey

- PubMed search conducted for each topic
- Committee members declared COI
- CFF summarises the evidence in this paper- provide reasonable clinical guidance.

Weight management

- Intake of energy dense foods is no longer a priority for some pwCF.
- Recommend a healthy, age-appropriate diet associated with positive health outcomes for general population (McDonald *et al*, 2020)
- Prevalence of overweight and obesity in CF has increased- CFF registry 2019 – 23% BMI \geq BMI 25kg/m² in 2021 up to 40% (same in UK registry).
- HEMT potentially increases weight by reduced REE, improving smell/taste, enhancing appetite, optimising fat absorption, increasing fat mass (Gelfond *et al*, 2017; Stallings *et al*, 2018; Edgworth *et al*, 2017)
- BMI remains an imperfect surrogate for nutritional status and potential metabolic risk.
- An MDT approach is helpful in management of patients with excess weight.

Obesity co-morbidities

- Data on cardiovascular outcomes is limited in pwCF- potential for increased prevalence.
- Arterial stiffness and inflammatory markers higher in children with CF (Eising et al, 2018; Buehler et al, 2012)
- Median cholesterol levels and prevalence of systemic hypertension higher in OW pwCF compared to underweight and healthy weight (Bonhoure et al, 2020, Gramagna et al, 2022).
- Evidence for elevation of BP after 12 months of ETI (Petersen et al, 2022).
- Median % FEV₁ higher in pwCF who are OW but there is a possible plateau at higher BMI values (Gramenga et al, 2022).

Medical and surgical management of obesity

- As in general population lifestyle modifications including diet, exercise and behaviour change management are typically the first recommendation
- Caution in extrapolating general population guidelines for management of obesity.
- Lack of data on optimal diet, effective lifestyle , pharmacological and surgical management of obesity in pwCF.
- Pharmacotherapy (orlistat, liraglutide, semaglutide)- may be associated with worsening malabsorption, exacerbation of underlying GI symptoms –no data on safety or efficacy but anecdotal reports of use in pwCF.
- Some case reports of bariatric surgery in pwCF - ? Safety and efficacy

Weight stigma/Weight neutral approaches

- Considering potential for weight stigma to be an issue in CF clinics – in other populations it is associated with disordered eating patterns and negative health behaviours (Puhl et al,2020; Tomiyama et al,2018)
- Weight neutral approaches- focus on optimising other health outcome measures rather than promoting weight loss – i.e., improvement in cholesterol, waist/hip ratio, physical activity , diet quality and self esteem (Mensing et al, 2016).
- CF teams should be aware of these methods and consider them for pwCF who struggle with body image issues (Darukhanavala et al 2021; Kass et al,2022)

Eating behaviours

- Is there an increased risk of ‘inappropriate compensatory eating behaviours’ in CF ?
- Conflicting evidence on whether there is increased prevalence of eating disorders in pwCF – common issues include misusing PERT, food restriction, binge eating and skipping meals (Kass et al, 2022)
- A CF-specific screening tool for assessing eating attitudes and behaviours has been developed for use but requires further testing for validity and reliability (Randlesome et al, 2013)
- ‘Need for an evidence based , brief and easy to use screening tool with low participant burden to better identify disordered eating attitudes and behaviours’

Body image

- Body image is an important topic that should be discussed comfortably and supportively in clinic – this may be more complex in CF given previous emphasis on BMI and weight gain (Tierney, 2012).
- HEMT may introduce new issues of concern for body image disturbance as some pwCF experience weight gain.
- Importance of constructive language- supportive and free of ‘blame and shame’ – can improve conversations between pwCF and CF HCP.

Food insecurity

- FI defined as lack of consistent access to affordable , nutritious and healthy foods.
- 2019 US data – 33% of pwCF affected (3x national average) (Corbera-Hincapie et al,2022)
- Likely more of impact in countries with limited free healthcare as financial challenges to affording CF treatments.
- Paradox of higher risk of obesity in food insecure households?
- Suggest screening for FI within CF clinics (guidelines in paper)

Salt homeostasis and hypertension

- Despite high salt intake , hypertension was previously rare in pwCF.
- 7% of adults with CF had hypertension in 2021 CFF registry data (CFF registry, 2022)
- Moderate increases in SBP and DBP seen in clinical trials of ETI and case reports of newly diagnosed hypertension (Middleton et al, 2019; Gramenga et al, 2022;).
- ? Reasons – contribution of increased overweight and reduced salt losses
- Need to revisit salt recommendations in adults with CF, monitor BP and consider modifying salt intakes (especially in those with hypertension and/or normal sweat chloride after HEMT use)
- Extra care in post organ transplant patients (immunosuppression is a risk factor).

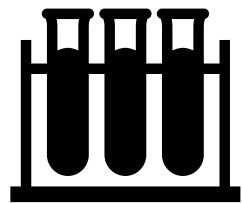
Pancreatic enzymes

- Emerging data that HEMT may improve or even reverse PI in some children with CF.
- Supported by physiological studies showing small intestinal luminal pH improves and improvement/normalisation of faecal elastase (Rosenfeld et al, 2019; Nicholls et al, 2020; Munce et al ,2020)
- Suggest checking faecal elastase after modulator initiation in children up to age 5 if change in pancreatic status is suspected.
- Recovery of pancreatic function may take several years after initiation of HEMT (Hutchinson and McNally, 2021)
- Insufficient new data on fat soluble vitamins in pwCF taking HEMT – supplementation should be monitored and adjusted as appropriate.

Key takeaways



✓ Although prevalence of overweight and obesity is increasing, undernutrition is still a concern for some pwCF.



✓ Blood lipid screening should follow guidelines in the general population until further CF specific data is available.

Key takeaways

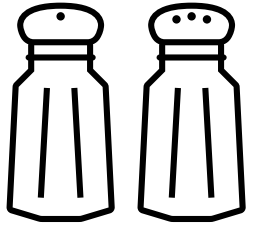


✓ 'Nutrition care should be individualised using clinical data and goals of people with CF



✓ Clinicians should be aware of the potential cardiovascular complications associated with the legacy, high fat/high calorie CF diet, particularly considering increasing life span.

Key takeaways



- ✓ There is a lack of data to make specific suggestions about salt intake in those pwCF and hypertension.



- ✓ Regular monitoring of BP is encouraged.

Key takeaways



- ✓ Food insecurity is a concern in pwCF from all socioeconomic sectors and screening should be part of standard care in CF.



- ✓ Since data suggest that there is the potential for return of pancreatic function in some children taking HEMT. Faecal elastase should be monitored if a change in pancreatic status is suspected.

Discussion points

What additional research/evidence will it take to be ready for new evidence based guidelines? What other methods could be used, e.g. Delphi consensus methods, given that there are very few recent RCTs of nutrition topics in CF

What are the change management counselling skills that dietitians are currently using and feel they need, to help patients with major transitions in diet?

How different and similar will paediatric CF dietetics look in the coming years?

Discussion points

How do we demonstrate to hospital funding/ resource allocation that we still need a similar amount of CF dietetics resourcing?

How can we ensure that we don't leave behind those who cannot access highly effective modulators?

QR code
linking to
paper



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