28 juin 2024

Journée des diététiciennes
Vaincre la mucoviscidose

Présentation du congrès européen à Glasgow juin 2024

Lignes directrices sur les soins nutritionnels pour la fibrose kystique

Susan Ramirez et Hélène Attew

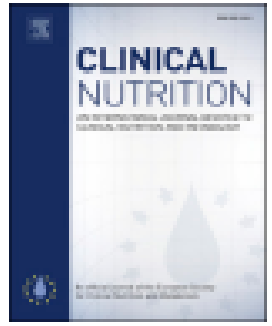
Lignes directrices sur les soins nutritionnels pour la fibrose kystique

Clinical Nutrition 43 (2024) 413–445

Contents lists available at ScienceDirect

Clinical Nutrition

journal homepage: http://www.elsevier.com/locate/clnu



ESPEN Guideline

ESPEN-ESPGHAN-ECFS guideline on nutrition care for cystic fibrosis

Michael Wilschanski [a, *], Anne Munck [b], Estefania Carrion [c], Marco Cipolli [d], Sarah Collins [e], Carla Colombo [f], Dimitri Declercq [g], Elpis Hatziagorou [h], Jessie Hulst [c, i], Daina Kalnins [j], Christina N. Katsagoni [k, l], Jochen G. Mainz [m], Carmen Ribes-Koninckx [n], Chris Smith [o], Thomas Smith [p], Stephanie Van Biervliet [q], Michael Chourdakis [r]

Michael Wilschanski and al. ESPEN-ESPGHAN-ECFS guideline on nutrition care for cystic fibrosis, Clinical Nutrition 43 (2024) 413e445

Introduction

- Ancienne version **2016**
- Groupe de travail **multidisciplinaire international**
- Mise à jour des **directives nutritionnelles**, y compris l'évaluation et la prise en charge à tous les âges.
- La supplémentation en vitamines et en enzymes pancréatiques reste en grande partie la même.
- Chapitres détaillés sur la grossesse, les maladies du foie liées à la mucoviscidose, le diabète lié à la mucoviscidose, les maladies osseuses, les suppléments nutritionnels et minéraux et les probiotiques.
- Il y a de nouveaux chapitres sur la nutrition avec des thérapies modulatrices très efficaces et la nutrition après une transplantation d'organe.
- **86 recommandations**

Niveau d'évidence et grade de recommandation

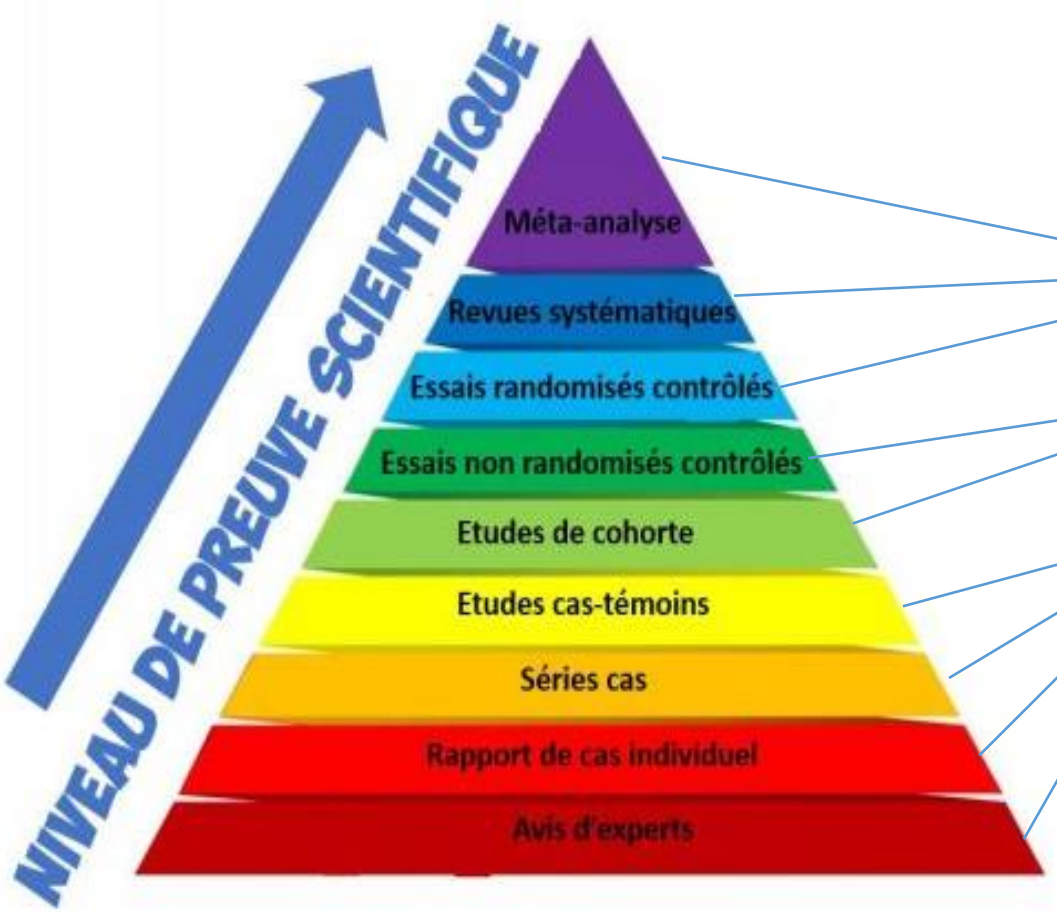


Tableau 1. Grade des recommandations

A	**Preuve scientifique établie** Fondée sur des études de fort niveau de preuve (niveau de preuve 1) : essais comparatifs randomisés de forte puissance et sans biais majeur ou méta-analyse d'essais comparatifs randomisés, analyse de décision basée sur des études bien menées
B	**Présomption scientifique** Fondée sur une présomption scientifique fournie par des études de niveau intermédiaire de preuve (niveau de preuve 2), comme des essais comparatifs randomisés de faible puissance, des études comparatives non randomisées bien menées, des études de cohorte.
0	**Faible niveau de preuve** Fondée sur des études de moindre niveau de preuve, comme des études cas-témoins (niveau de preuve 3), des études rétrospectives, des séries de cas, des études comparatives comportant des biais importants (niveau de preuve 4).
GPP	**Accord d'experts** En l'absence d'études, les recommandations sont fondées sur un accord entre experts du groupe de travail, après consultation du groupe de lecture. L'absence de gradation ne signifie pas que les recommandations ne sont pas pertinentes et utiles. Elle doit, en revanche, inciter à engager des études complémentaires.

Evaluation nutritionnelle - Générale

Recommendation 1

Standard assessment and monitoring of nutritional status in people with CF (pwCF) should be accurately performed regularly and should include as minimum the following parameters which need to be interpreted together and longitudinally:

- **Infants: z-scores for weight-for-age, length-for-age and weight-for-length (WFL) as well as head circumference-for-age every one to two weeks until evidence of adequate nutrition and ideal nutritional status is established, then monthly through the first year of life.**
- **Children >1 and ≤ 2 years: z-scores for weight-for-age, length-for-age and WFL as well as head circumference for age every two to three months.**
- **Children >2 years: z-scores for weight-for-age, height -for-age and BMI-for-age at least every three months.**
- **Adults: BMI at least once every three to six months.**

Patients with poor nutritional status and/or growth should have more frequent monitoring than patients with adequate nutritional status.

Grade of recommendation GPP – Strong consensus 100 % agreement

Surveillance régulière de l'état nutritionnel

→Nourrissons : poids, taille et PC toutes les 1-2 semaines

→Enfants de 1-2ans : poids, taille et PC tous les 2-3 mois

→Enfants > 2 ans : poids, taille et IMC tous les 3mois

→Adultes : IMC tous les 3-6mois

Evaluation nutritionnelle - Générale

Recommendation 4

Longitudinal assessment of body composition to obtain estimates of fat mass (FM) and fat-free mass (FFM), rather than sole reliance on BMI, should be performed in pwCF because their association with (respiratory) outcomes is stronger than for BMI alone and because a low or normal BMI can mask high FM or low FFM. The choice in body composition method should be guided based on availability, resources, technical factors and clinical factors such as age and hydration status.

Grade of recommendation B - Strong consensus 92 % agreement

Evaluation de la composition corporelle pour obtenir une estimation de la masse grasse (FM) et de la masse maigre (FFM).

→Un IMC faible ou normal peut masquer une FM élevée ou une FFM faible

Consensus fort 92%

Thérapie enzymatique substitutive du pancréas

Recommendation 15

Pancreatic enzymes shall be started in all patients who have evidence of exocrine PI. The intake of pancreatic enzymes results in improved anthropometric outcomes and maldigestion related gastro-intestinal complications.

Grade of recommendation A - Strong consensus 100 % agreement

Recommendation 16

The pH in the small intestine determines the release of pancreatic enzymes from the enteric coated beads. Evidence is lacking around routine use of proton pump inhibitors (PPI) to improve enzyme efficacy.

Grade of recommendation GPP - Strong consensus 100 % agreement

→Mises en place chez tous les patients

→Amélioration des résultats anthropométriques et des complications gastro-intestinales liées à la maldigestion.

→Il n'y a pas de données probantes concernant l'utilisation systématique d'IPP

Pancreatic enzyme lipase replacement therapy: consensus guideline.

Age	Suggested supplementation
Infants (up to 12 months)	2000–4000 U lipase/120 mL formula or estimated breast milk intake and approximately 2000 U lipase/gram dietary fat in food
Children 1–4 years	2000–4000 U lipase/gram dietary fat, increasing dose upward as needed (maximum dose 10,000 U lipase/kg/d)
Children >4 years and adults	Consider starting at 500 U lipase/kg/meal, titrating upward to a maximal dose of: - 1000–2500 U lipase/kg per meal, or - 10,000 U lipase/kg/d, or - 2000–4000 U lipase/gram dietary fat taken with all fat-containing meals, snacks and drinks.

Vitamines liposolubles

Recommendation 21

For pwCF and exocrine PS, fat-soluble vitamins may be assessed annually using plasma levels.

Grade of recommendation 0 - Strong consensus 100 % agreement

Recommendation 22

25(OH)D should be measured annually. Individuals with levels of 25(OH)D <20 ng/mL (<50 nmol/L) should be considered deficient, and levels >30 ng/mL (>75 nmol/L) should be sufficient. The goal 25 (OH) levels should be 30–50 ng/mL (75–125 nmol/L) and levels should not exceed 100 mg/mL (250 nmol/L).

Grade of recommendation GPP - Strong consensus 100 % agreement

→Evaluation annuelle des vitamines liposolubles

→Les taux de vitamines D doivent être entre 30-50ng/mL et ne pas dépasser 100ng/mL

Zinc

Recommendation 53

Zinc supplementation for pwCF who have proven zinc deficiency or who are at risk of concerns of zinc insufficiency (e.g. sub optimal growth, assessed low dietary intake, increased susceptibility to infections, delayed sexual maturation, and acrodermatitis) may be considered. Routine additional zinc supplementation for pwCF may not confer any clinical benefit.

Grade of recommendation 0 - Strong consensus 100 % agreement

Recommendation 54

Assessment of zinc status should include biochemical markers (interpreted with caution) in combination with clinical examination and assessment of dietary intake

Grade of recommendation GPP - Strong consensus 100 % agreement

→Supplémentation en zinc si carence avérée

→Supplémentation systématique = pas de bénéfices clinique

→Evaluation du statut en zinc avec marqueurs bio + examen clinique + évaluation des apports alimentaires

Sel

Recommendation 57

In infants, children and adults with CF, specific levels of salt supplementation may be required depending on age and clinical condition, physical activity and climate situation (see Table 7).

Grade of recommendation GPP - Strong consensus 100 % agreement

Approximately ¼ teaspoon salt contains about 25 mmol or 575 mg of sodium.

Recommendation 58

The need for salt/sodium supplementation can be assessed by measuring fractional excretion of sodium (FENa) and maintaining a FENa level between 0.5 % and 1.5 %. For routine practice, a urinary sodium:creatinine ratio is easier to measure and correlates with FENa.

Grade of recommendation GPP - Strong consensus 100 % agreement

→ Niveau spécifique de supplémentation en sel peuvent être nécessaire en fonction de l'âge et de l'état clinique, de l'activité physique et de la situation climatique

→ Besoins évalués en mesurant l'excrétion fractionnée de sodium *FE(U) = (urée urinaire x créatinine plasmatique)/ (urée plasmatique x créatinine urinaire)*

→ FE entre 0,5-1,5%

→ De façon plus courante, Na/créat plus facile à mesurer

Diabète

Recommendation 66

All pwCF should have annual screening for CFRD from the age of ten years.

Grade of recommendation GPP - Strong consensus 100 % agreement

Recommendation 67

In case of unexplained clinical decline or development of liver disease before the age of 10, screening for abnormal glucose tolerance should be performed.

Grade of recommendation GPP - Strong consensus 100 % agreement

→Dépistage annuel dès l'âge de 10ans

→Si déclin inexpliqué ou développement d'une maladie du foie avant 10ans -> dépistage

Foie

15. CF-associated liver disease

Recommendation 72

PwCF with cirrhosis and/or portal hypertension are at risk of malnutrition and should receive a thorough nutritional assessment every six months by a dietician experienced in CF in order to identify and/or prevent specific nutritional deficiencies and plan appropriate personalized interventions.

Grade of recommendation GPP - Strong consensus 100 % agreement

→Les patients atteints de cirrhose et/ou d'hypertension portale sont à risque de malnutrition

→Evaluation nutritionnelle approfondie tous les 6 mois

Nutrition et modulateur

17. Nutrition and CFTR modulator therapy

Recommendation 77

Appropriate dietary counselling should be provided for pwCF starting on CFTR modulator therapy, this should include advice about limiting and managing weight gain

Grade of recommendation B - Strong consensus 100 % agreement

Recommendation 78

The nutritional status and dietary intake of pwCF on CFTR modulator therapy including salt intake and fat-soluble vitamin status should continue to be regularly reviewed and modifications recommended according to changes observed

Grade of recommendation B - Strong consensus 100 % agreement

Recommendation 79

Gastrointestinal symptoms should be closely monitored following the initiation of CFTR modulator therapy and this should continue as part of routine CF care

Grade of recommendation B - Strong consensus 100 % agreement

→Conseils diététiques appropriés avec modulateurs pour limiter et gérer la prise de poids

→Suivi régulier des apports en sel et du statut vitaminique

→Les symptômes gastro-intestinaux doivent être surveillés

Nutrition et modulateur

Recommendation 80

In view of the fact that CFTR modulators may improve beta cell function and insulin secretion, blood glucose levels should be monitored regularly to avoid hypoglycemia when assuming the same insulin dose prescribed before starting modulators.

Grade of recommendation B - Strong consensus 100 % agreement

Recommendation 81

PwCF on CFTR modulator therapy should have their blood pressure routinely monitored three months after commencing therapy and at least annually and appropriate medical management started if clinically indicated.

Grade of recommendation B - Strong consensus 92 % agreement

Recommendation 82

PwCF on CFTR modulator therapy may have their lipid profiles checked annually and appropriate dietary advice and medical management should be given if required.

Grade of recommendation GPP - Strong consensus 92 % agreement

→Surveillance régulière des glycémies pour éviter les hypoglycémies chez les patients démarrant les modulateurs

→Surveillance systématique de la tension artérielle 3 mois après le début du traitement et au moins 1x/an

→Surveillance des profils lipidiques tous les ans.