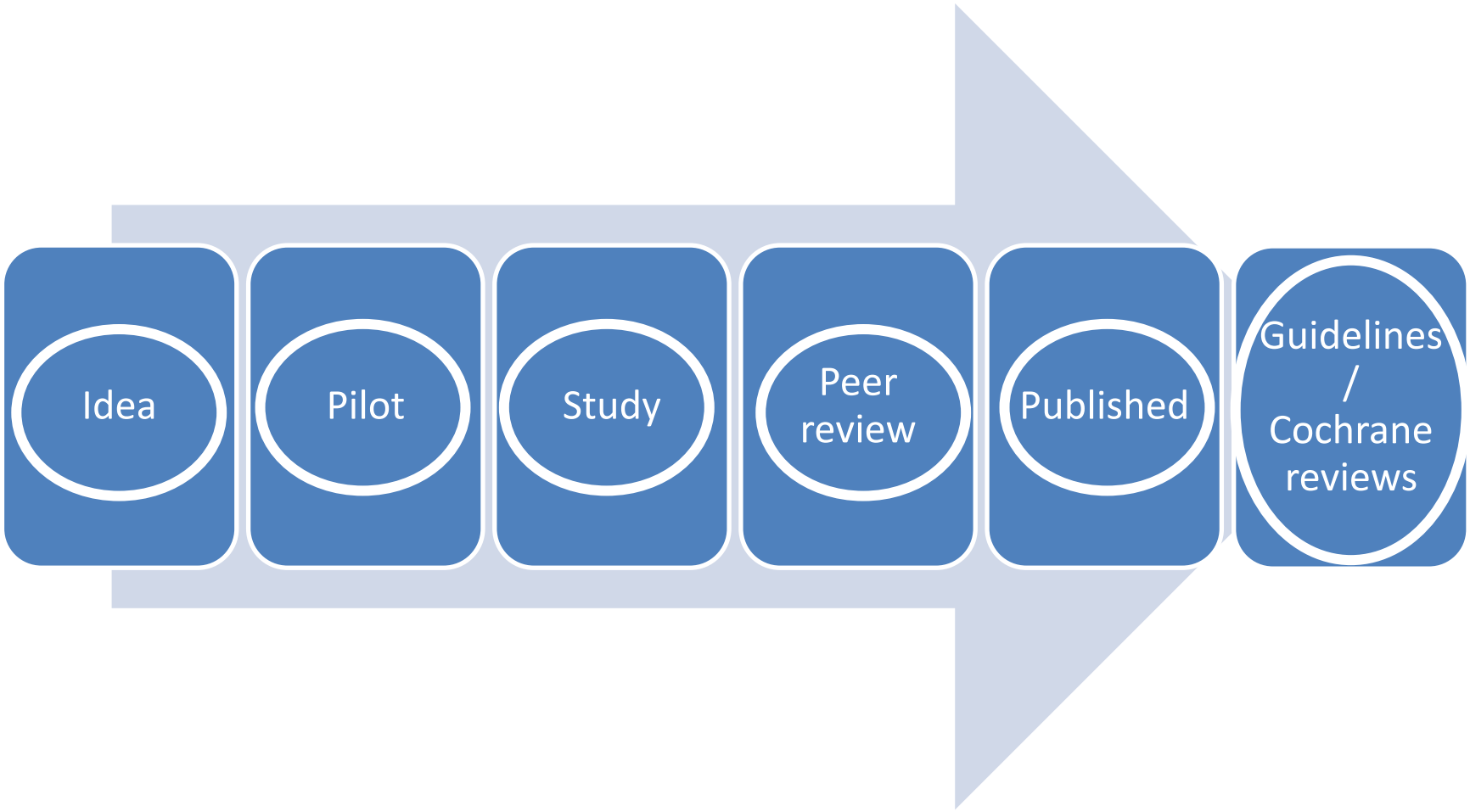
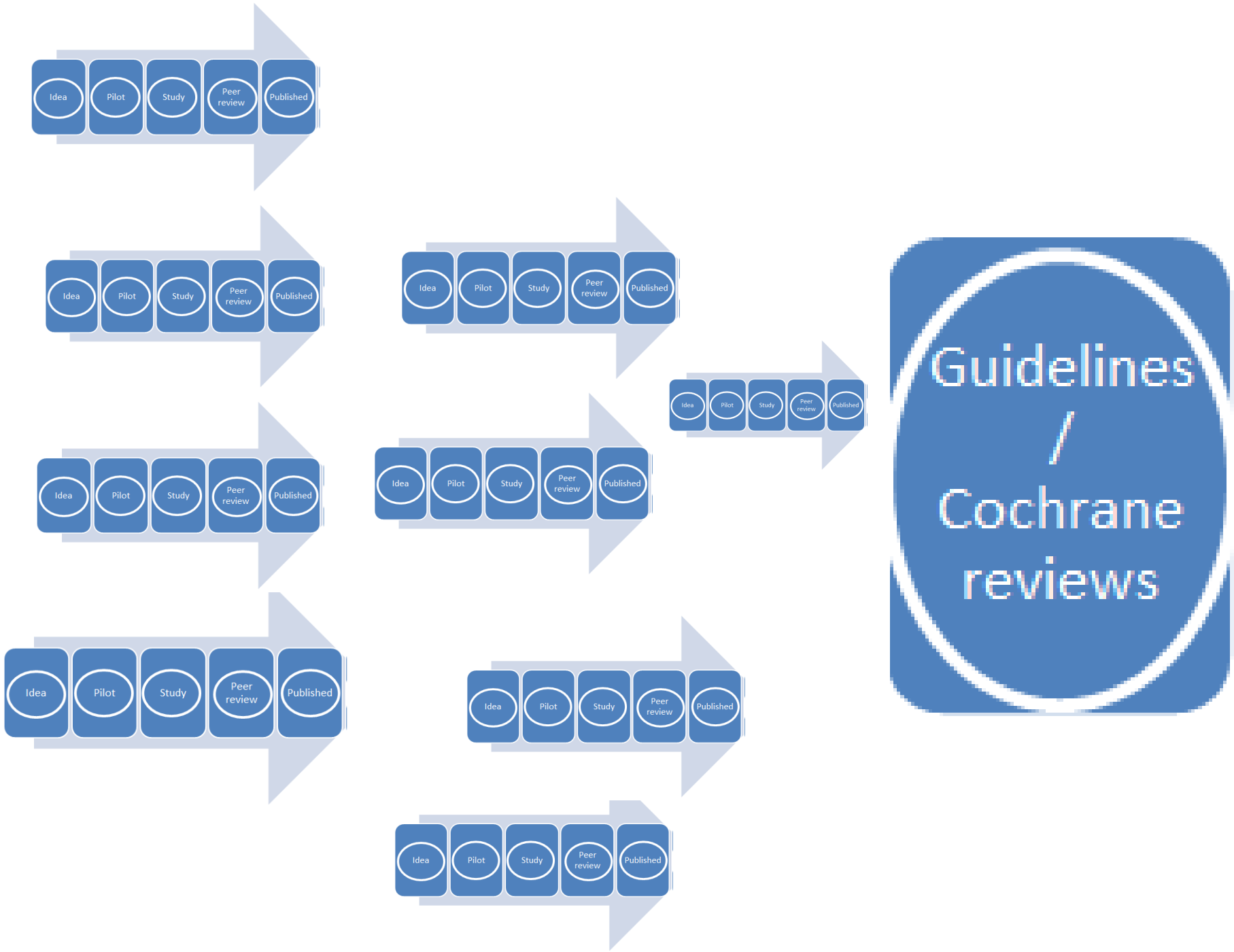
UPDATE ON NUTRITION AND GASTROINTESTINAL ISSUES

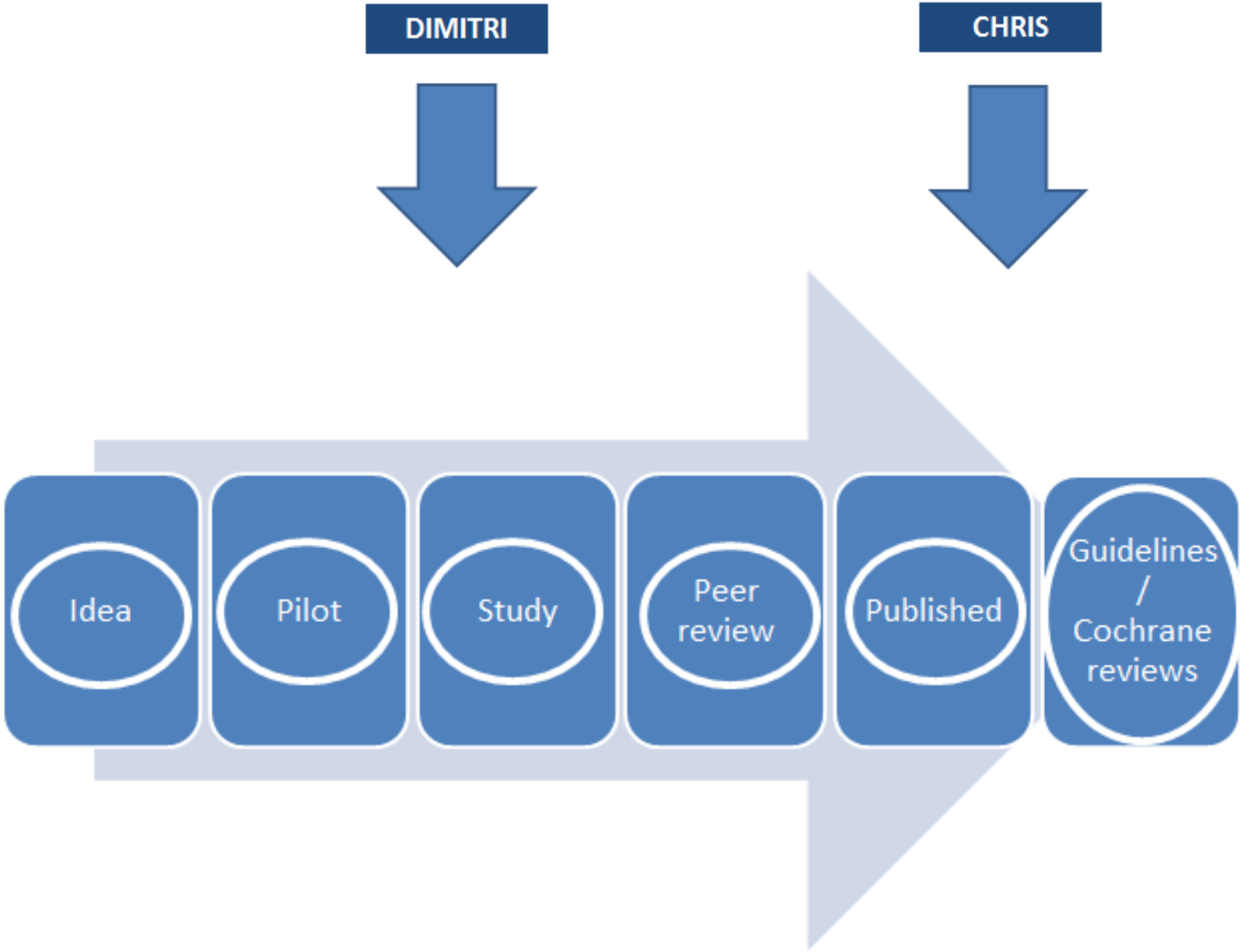
CHRIS SMITH, UK

DIMITRI DECLERCQ, BE

Co Chairs of European CF Nutrition Group


Idea
Pilot
Study
Peer review
Published
Guidelines / Cochrane reviews


Idea
Pilot
Study
Peer review
Published
Guidelines / Cochrane reviews


DIMITRI
CHRIS
Idea
Pilot
Study
Peer review
Published
Guidelines / Cochrane reviews

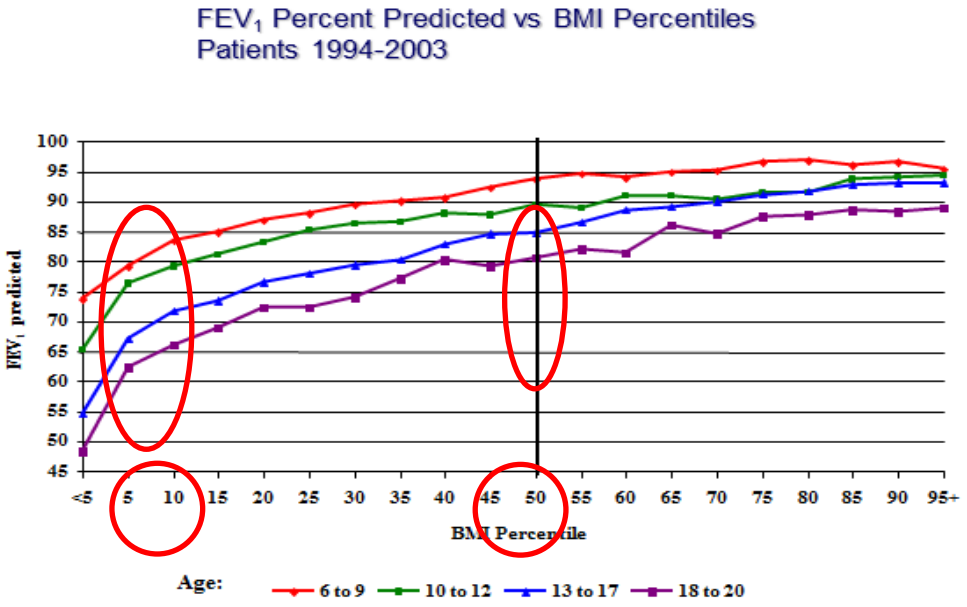
Review

European Cystic Fibrosis Society Standards of Care: Best Practice guidelines

Journal of Cystic Fibrosis 13 (2014) S23–S42

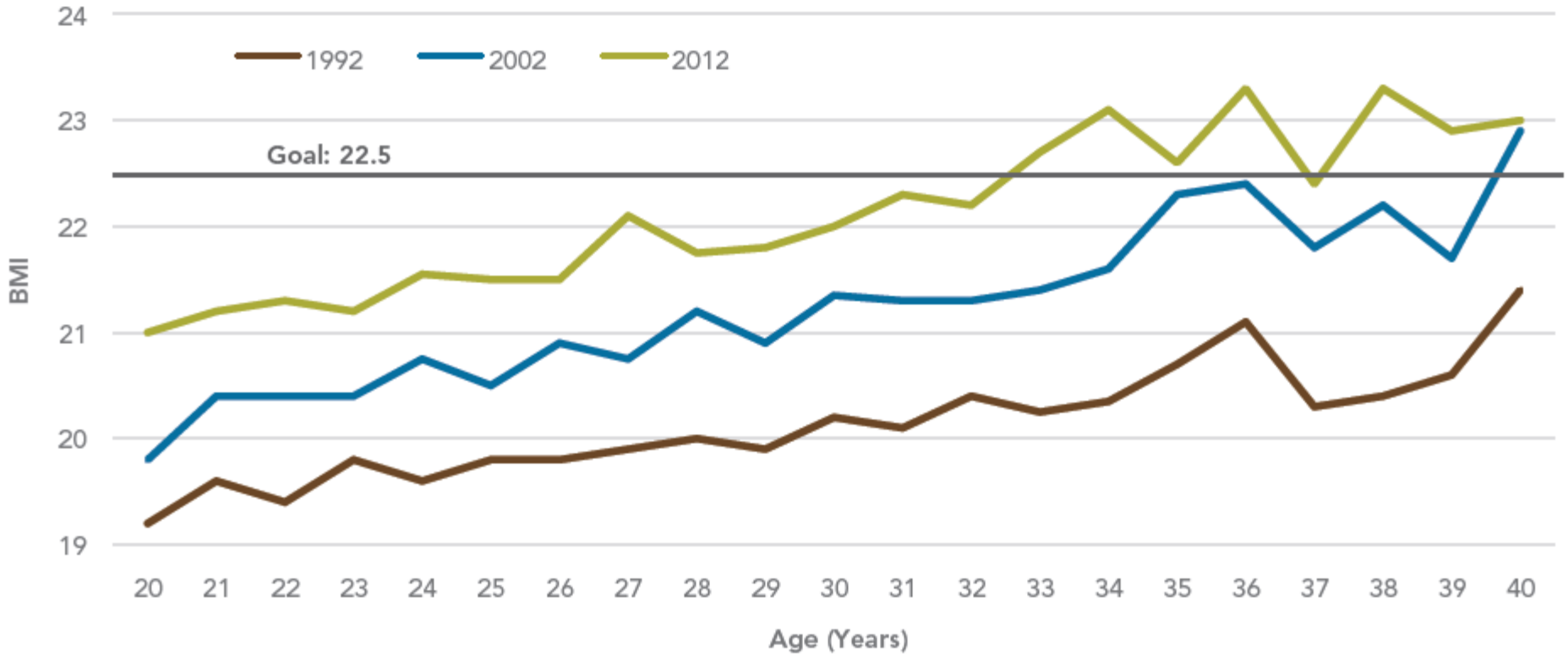
Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

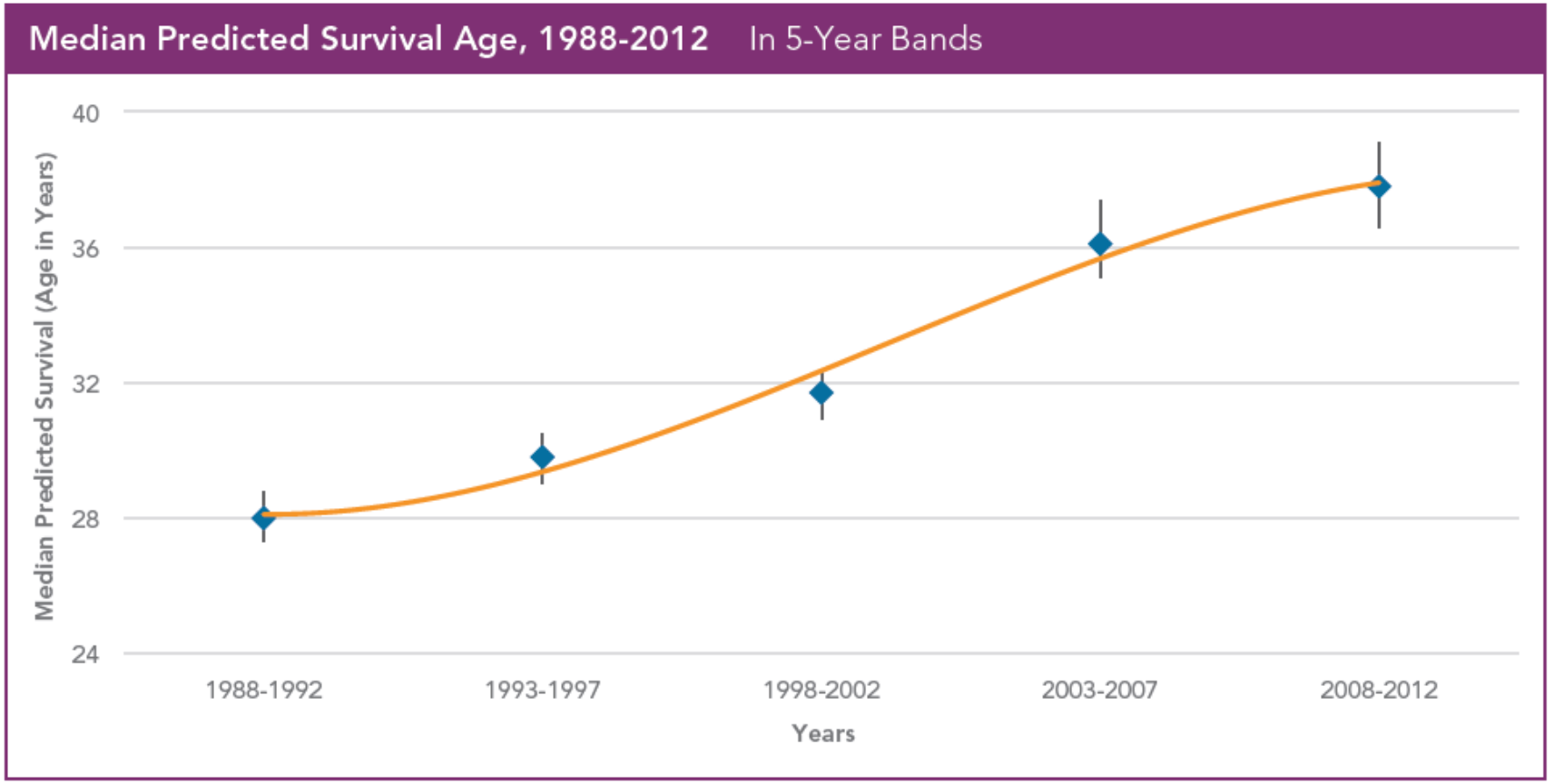
- What are the goals for nutritional status in patients with CF?



BETTER BMI = BETTER LUNG FUNCTION

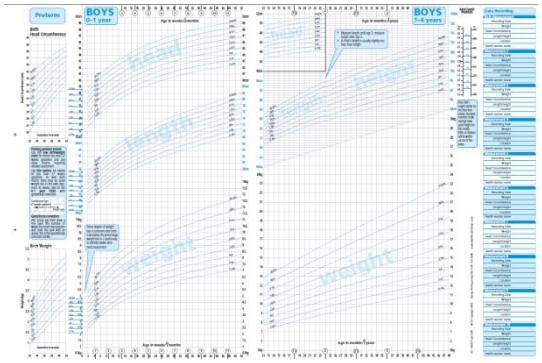
Median BMI for Adults by Age - 1992, 2002 and 2012

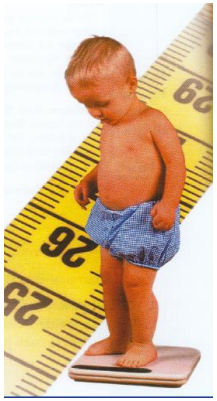




BETTER BMI's CORRELATE WITH BETTER SURVIVAL

MONITORING GROWTH





Comparison of WHO and CDC Growth Charts in Predicting Pulmonary Outcomes in Cystic Fibrosis

Machogu, Evans*; Cao, Yumei†; Miller, Tami‡; Simpson, Pippa†; Levy, Hara*; Quintero, Diana*; Goday, Praveen S.||

Journal of Pediatric Gastroenterology & Nutrition:
March 2015 - Volume 60 - Issue 3 - p 378–383

Additional recommendations:

1. Most EMRs now use WHO growth curves from Birth to 24 months of age. However, for those who still use and/or who prefer to plot on paper charts, the CDC growth curves may be used. Dietitians are encouraged to help families understand the differences between the WHO and CDC percentiles.

2. It is important to obtain accurate weight and length measurements. Ideally, these measurements will be done by a limited number of trained individuals and using precise equipment (e.g. routinely calibrated scales and length boards as opposed to measuring tapes).

- **GOOD NUTRITION IS A VITAL PART OF TREATMENT AND MANAGEMENT**

- **IMPROVING NUTRTIONAL STATUS IN INFANT IS A KEY PRIORITY**

Review

European Cystic Fibrosis Society Standards of Care: Best Practice guidelines

Journal of Cystic Fibrosis 13 (2014) S23–S42

- What are the options to improve nutritional care?

- Anticipatory guidance.

  Reinforcement of adherence to diet, sodium and enzyme recommendations, using behavioural modification or motivational interviewing

ENZYMES

REVIEW

Cystic fibrosis: An update for clinicians. Part 2: Hepatobiliary and pancreatic manifestations

Oren Ledder,* Wolfram Haller,* Richard TL Couper,† Peter Lewindon‡ and Mark Oliver*,§

*Department of Gastroenterology and Clinical Nutrition, Royal Children's Hospital Parkville, †Department of Paediatrics, University of Melbourne, Melbourne, Victoria, ‡Department of Gastroenterology, Women's and Children's Hospital, Adelaide, South Australia, and §Department of Gastroenterology, Royal Children's Hospital Brisbane, Brisbane, Queensland, Australia

JGH Journal of Gastroenterology and Hepatology JGHF

Accepted for publication 20 August 2014.

Table 3 Pancreatic enzyme replacement therapy guidelines

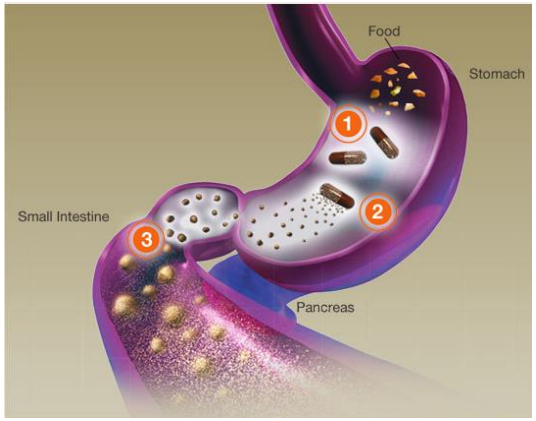
Dosing per dietary fat intake	
Age	Dose
Infants	500–1000 U/gram fat
	2000–4000 per breast feed/120 mL formula feed
Children	500–4000 U/gram fat
Dosing per bodyweight	
Age	Dose
< 4 years	1000 U/kg/meal
	Snack: half dose
> 4 years	500 U/kg/meal

Dose titration

Commence on minimal dose: increase dose based on weight gain, steatorrhea, and coefficient of fat absorption testing

Administration

- Swallow whole or sprinkle capsule on applesauce (or other non-alkali soft food)
- Rinse infant's mouth after administration
- Co-administer proton pump inhibitor therapy to maximize efficacy of therapy
- Assess fat-soluble vitamin (A, D, E and coagulation screen) and replace as necessary



Taking appropriate enzyme doses at appropriate times are a vital part of management for CF patients.

Review

European Cystic Fibrosis Society Standards of Care: Best Practice guidelines

Journal of Cystic Fibrosis 13 (2014) S23–S42

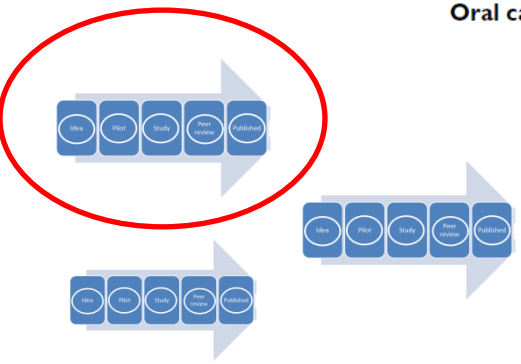
Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

- What are the options to improve nutritional care?

- Anticipatory guidance.
  Reinforcement of adherence to diet, sodium and enzyme recommendations, using behavioural modification or motivational interviewing

- Moderate malnutrition.
  Oral supplements or temporarily as meal replacement for ill patients.
  Temporary nasogastric (NG)/nasojejunal (NJ) feeds may be useful

ORAL SUPPLEMENTS



Oral calorie supplements for cystic fibrosis (Review)

Smyth RL, Rayner O

THE COCHRANE COLLABORATION®

This is a reprint of a Cochrane review, prepared and maintained by The Cochrane Collaboration and published in *The Cochrane Library* 2014, Issue 11

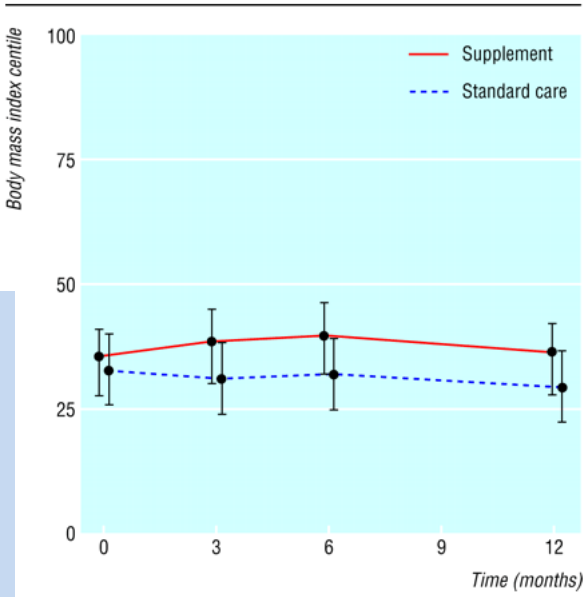
http://www.thecochranelibrary.com

Research

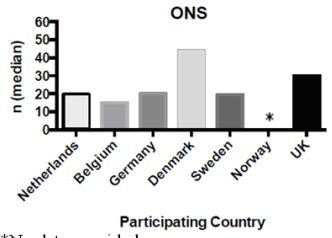
Oral protein energy supplements for children with cystic fibrosis: CALICO multicentre randomised controlled trial

Vanessa J Poustie, Jayne E Russell, Ruth M Watling, Deborah Ashby, Rosalind L Smyth, on behalf of the CALICO Trial Collaborative Group

BMJ, doi:10.1136/bmj.38737.600880.AE (published 8 February 2006)



Graphs showing median % prevalence of paediatric CF caseloads using ONS



*No data provided

"Supplements do not confer any additional benefits over standard advice"

HOWEVER

Many dietitians have experience where supplements have had significant impact on nutritional status. Consider use in individuals.

Review

European Cystic Fibrosis Society Standards of Care: Best Practice guidelines

Journal of Cystic Fibrosis 13 (2014) S23–S42

Journal of Cystic Fibrosis
The Official Journal of the European Cystic Fibrosis Society

- What are the options to improve nutritional care?

- Anticipatory guidance.
  Reinforcement of adherence to diet, sodium and enzyme recommendations, using behavioural modification or motivational interviewing

- Moderate malnutrition.
  Oral supplements or temporarily as meal replacement for ill patients.
  Temporary nasogastric (NG)/nasojejunal (NJ) feeds may be useful

- Severe malnutrition.
  Enteral feeding via NG or gastrostomy tubes

ENTERAL FEEDING

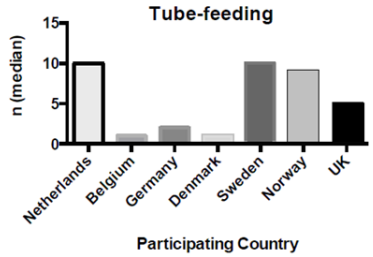
Cochrane Database Syst Rev. 2015 Apr 9;4:CD001198. [Epub ahead of print]

Enteral tube feeding for cystic fibrosis.

Morton A[1], Wolfe S.

Use with care

Graphs showing median % prevalence of paediatric CF caseloads using TF



PREVENTION OF NEED IS BETTER THAN TREATMENT

VITAMIN D

Original Article

Vitamin D deficiency is associated with pulmonary dysfunction in cystic fibrosis ☆

Journal of Cystic Fibrosis xx (2014) xxx–xxx

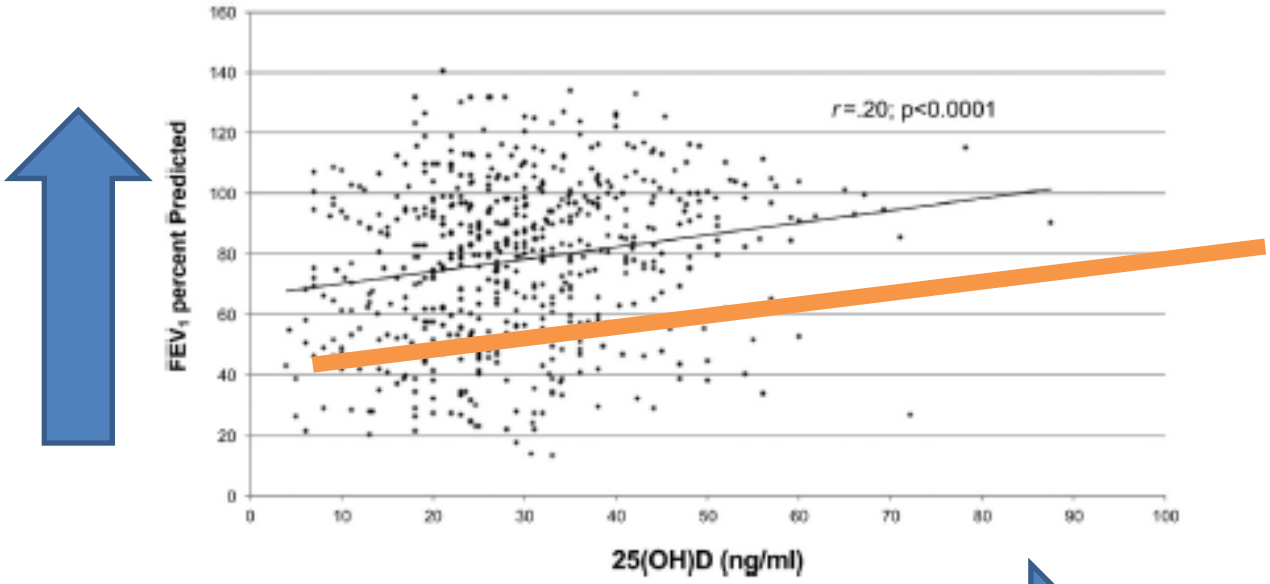


Fig. 1. Of the pulmonary function variables analyzed, 25(OH)D s… FEV_1 % predicted.

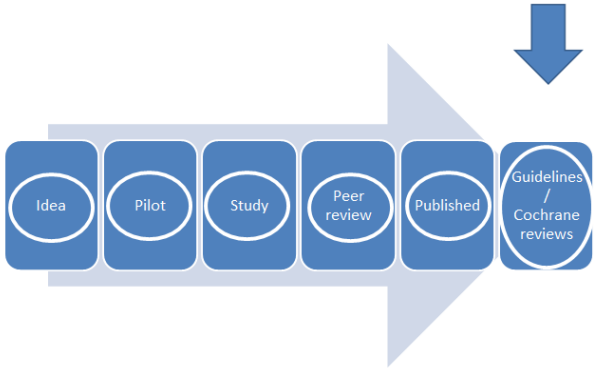
All aspects of treatment (including the seemingly small aspects such as Vitamin D) are important for the best care and contribute to better health

APPETITE STIMULANTS

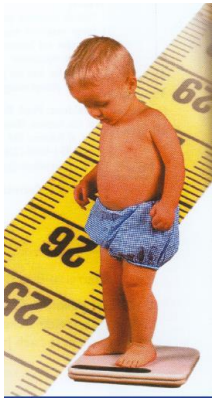
Evidence was systematically reviewed from 2002 to 2014.

Key Practice Point:

There is currently insufficient evidence to suggest that the routine use of appetite stimulants improves the nutritional status, including bone mineral density[4] , of patients with cystic fibrosis. Megestrol acetate[1 2] and growth hormone[3] may have a role in improving nutritional status in cystic fibrosis, however, further randomised, double-blind controlled trails are needed with increased numbers of subjects to improve the evidence and to determine their safety.



GROWTH HORMONE



Cochrane Database Syst Rev. 2015 May 20;5:CD008901. [Epub ahead of print]

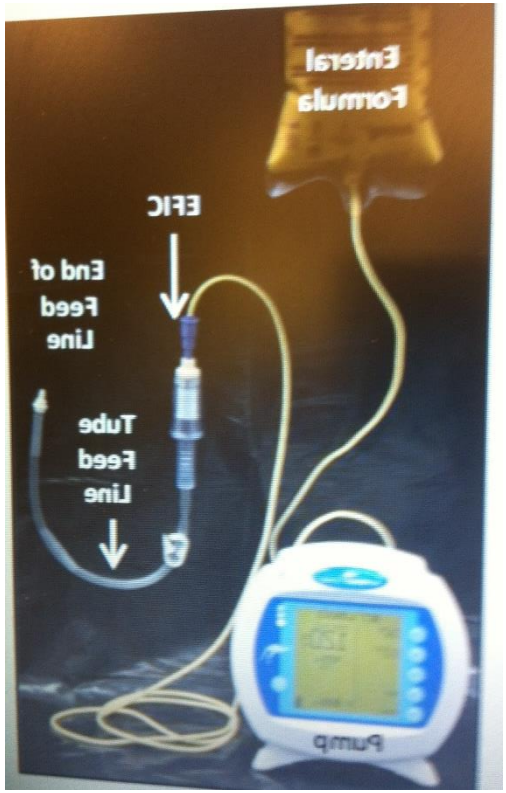
Recombinant growth hormone therapy for cystic fibrosis in children and young adults.

Thaker V[1], Haagensen AL, Carter B, Fedorowicz Z, Houston BW.

"Long-term, well-designed randomised controlled trials of recombinant growth hormone therapy in patients with cystic fibrosis are required prior to evaluation of human growth hormone treatment for routine use in patients."

PANCREATIC ENZYMES

- How much enzymes do we need to take?
- Enteral Feeding In-Line Cartridge (EFIC™)
  - Immobilized lipase
  - Current problem is administration of PERT
  - Most formulas up to 90% hydrolyses of FA especially EFA (EPA, DHA and AA)



OUTCOME MEASUREMENTS

- Current parameters:
  - Bodyweight
  - Length
  - BMI
- Progress away from BMI
- Body composition
  - BIA
- Patient reported outcomes
- Patient experience

CYSTIC FIBROSIS RELATED DIABETES

- CFRD
  - Might not only be a result of structural damage but also of the presence of the CFTR in beta cells
  - Therefore PS patients may also develop CFRD
  - Impaired insulin secretion starts at the age of 6
    - AGT: 6 – 9 years -> 41%
- ECFS registry data
  - 20% drop of FEV1%
  - 8,25% -1 z-score BMI around diagnosis

VITAMIN D

- Different cutoff values
  - 20 ng/ml European Guidelines (50 nmol/l)
  - 30 ng/ml US guidelines (75 nmol/l)

    => However in both situations difficult to achieve
- Different dosages used
  - Montreal: 2800 IU/day and 4400 IU/day during the dark months
    - 71,5% reached optimal Vitamin D levels
    - 6,5% still had levels < 10nmol/l
  - Stoss therapy (100000 – 600000 IU) Australia
    - However not all patients reached optimal levels
- Difficult in correcting the low values
  - Adherence
  - Other factors involved

CONCLUSION

- Continued work strengthening nutrition-health link
- Under and over nutrition
- More than just calories

Thank you!