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Dimitri Declercq / CF Centre

Nutrition in Cystic Fibrosis

The needs through different age groups

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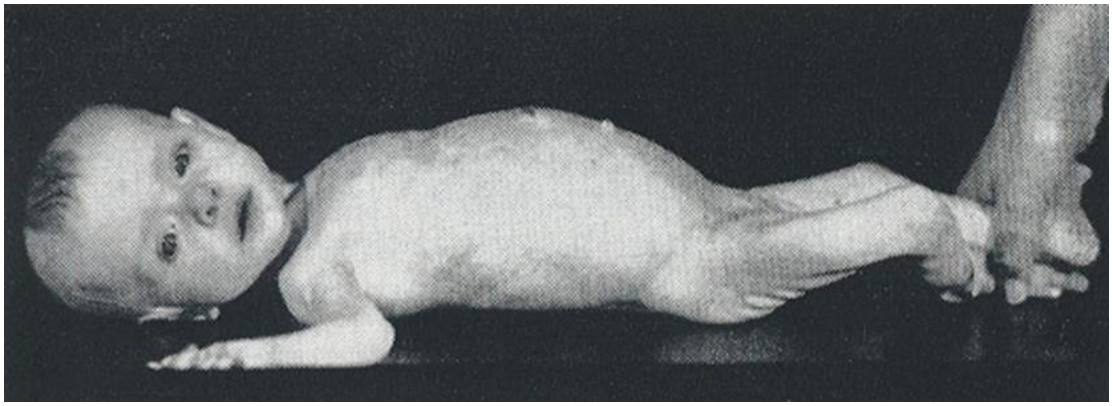
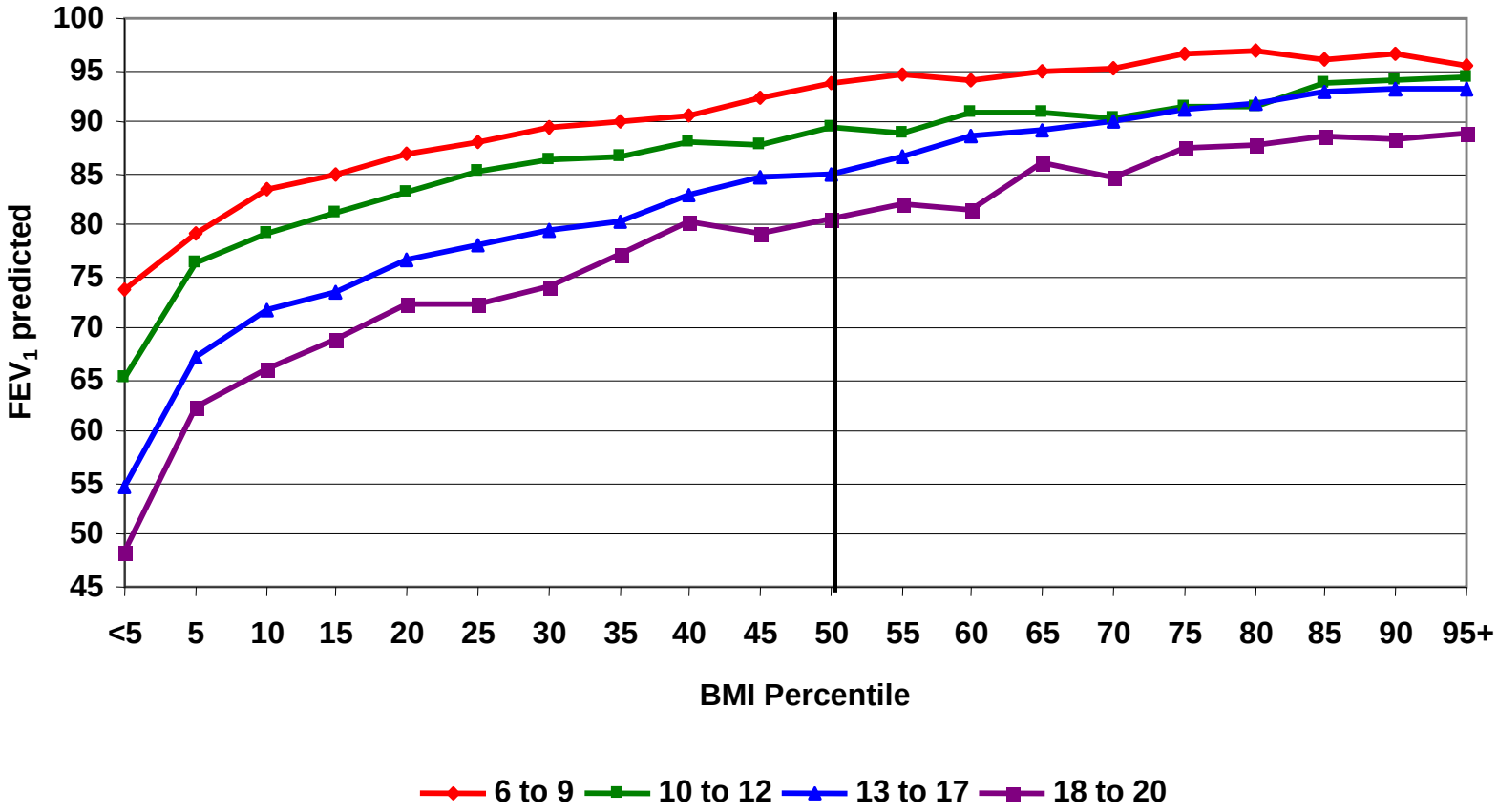
Cystic Fibrosis

Dorothy Andersen, 1938, ***American Journal of Diseases of Children***

- *Cystic Fibrosis of the pancreas and its relation to celiac disease: a clinical and pathological study*



Cystic Fibrosis



CFF Patient Registry

Evidence?

Contents lists available at ScienceDirect

Clinical Nutrition

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

e-SPEN guideline

ESPEN-ESPGHAN-ECFS guidelines on nutrition care for infants, children, and adults with cystic fibrosis

Dominique Turck [a], Christian P. Braegger [b], Carla Colombo [c], Dimitri Declercq [d], Alison Morton [e], Ruzha Pancheva [f], Eddy Robberecht [g], Martin Stern [h], Birgitta Strandvik [i], Sue Wolfe [j], Stephane M. Schneider [k, l], Michael Wilschanski [l, *, 1]

- Update
- Expert opinions
- New era

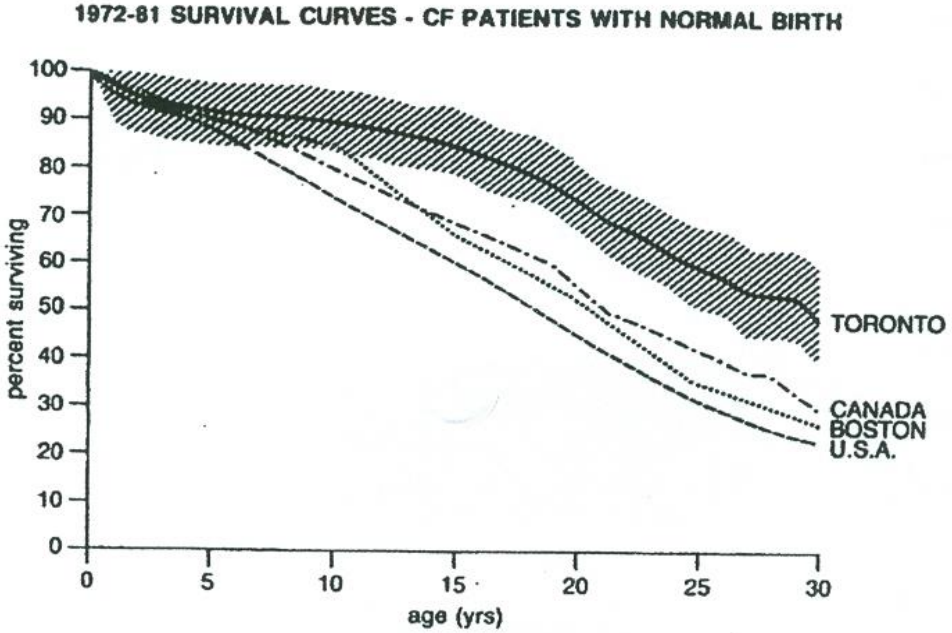
Nutritional requirements: age, length, weight, gender, physical activity and disease stage

Nutritional requirements

Age

Pathogenesis of Malnutrition in CF

M. Corey *et al.*

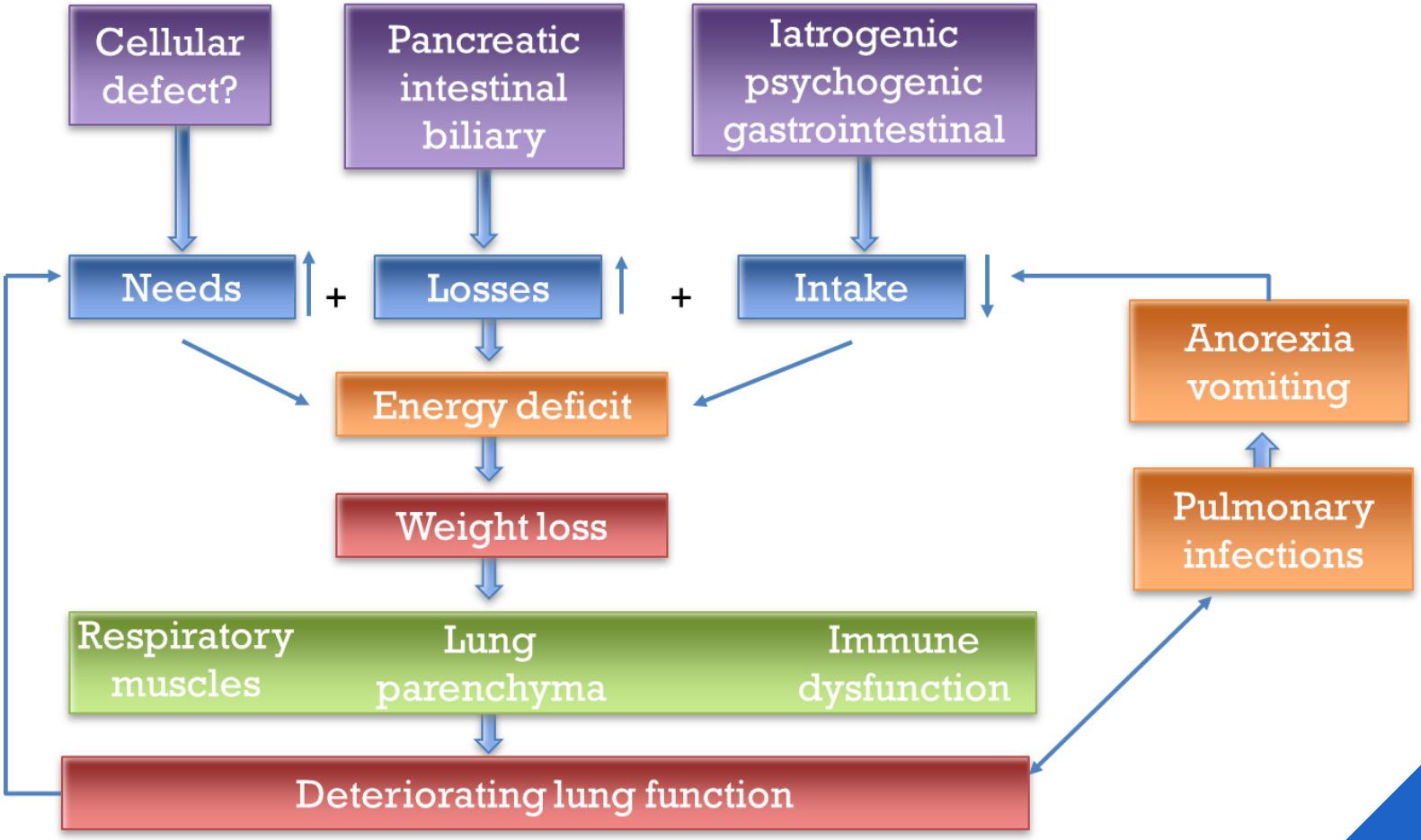


Survival curves 1972–81 for patients without meconium ileus at birth in Toronto and Boston, and in all CF care centres in Canada (including Toronto) and U.S.A. (including Boston).

Corey M, McLaughlin F J, Williams M et al. A comparison of survival, growth, and pulmonary function in patients with cystic fibrosis in Boston and Toronto. J Clin Epidemiol 1988; 41: 583– 591

Pathogenesis of Malnutrition in CF

Pencharz et al., 2000



Evidence?
(Turck et al., 2016)

Age	Energy target	Detail
Infants and children ≤ 2 years	110% - 200% of energy requirements healthy peers	Adapt energy intake according to evolution
Children 2 – 18 years	110% - 200% of energy requirements healthy peers	Adapt energy intake to BMI percentile
Adults > 18 years	110% - 200% of energy requirements of healthy peers to maintain BMI targets	Adapt energy intake to BMI percentile

- Carbohydrates: 40 – 45%
- Protein: 20%
- Fat: 35 – 40%

Evidence?

Estimating energy requirements
(Lahiri et al. (2016); Ramsy et al. (1992))

- <u>BMI at targets:</u>
  - CF children: 2 - 5 years
    - ≥ 90 – 110 kcal/kg per day
    - 2 – 3 year old: ≥ 13 g protein/day (13,5 kg (B)/13 kg (G))
    - 4 – 5 year old: ≥ 19 g protein/day (17,7 kg (B)/16,8 kg (G))
- <u>Daily Energy Requirements:</u>
  - BMR x (AF + DF) x 1.094 (0,93/CFA)
  - Activity Factor (AF): 1.3 – 1.5 – 1.7
  - Disease Factor (DF): 0 – 0.2 – 0.3 – 0.5

Evidence?

Estimating energy requirements (Trabulsi et al. (2007))

- Aim: to establish the most suitable formula to calculate TER in CF children
- Subjects: 6 – 8 years (n=53); PI; FEV1%: 98 (19)(55-133)
- Methods: comparing 6 different formulas with doubly labeled water

	Calculated TER	Difference: Calculated TER − TER-CF[2]	%TER-CF[3]	Indexes of model fit[4]		
				RMSE	P	R^2
	kcal/d	*kcal/d*	%			
Formulas for healthy children						
$EER_{low\ act}$	1542 ± 182 (1305–2158)[5]	−199[6]	90	224	< 0.01	0.41
EER_{act}	1755 ± 206 (1504–2452)	13	102	221	< 0.01	0.42
RDA	1799 ± 385 (1264–2860)	57	104	246	< 0.01	0.28
Formulas for children with cystic fibrosis						
RDA × 1.2	2158 ± 462 (1517–3431)	416[6]	125	246	< 0.01	0.28
$CFCR_{med}$	1876 ± 596 (1344–4413)	136[6]	108	235	< 0.01	0.35
$CFCR_{act}$	2122 ± 673 (1523–5001)	380[6]	122	234	< 0.01	0.35

Evidence?

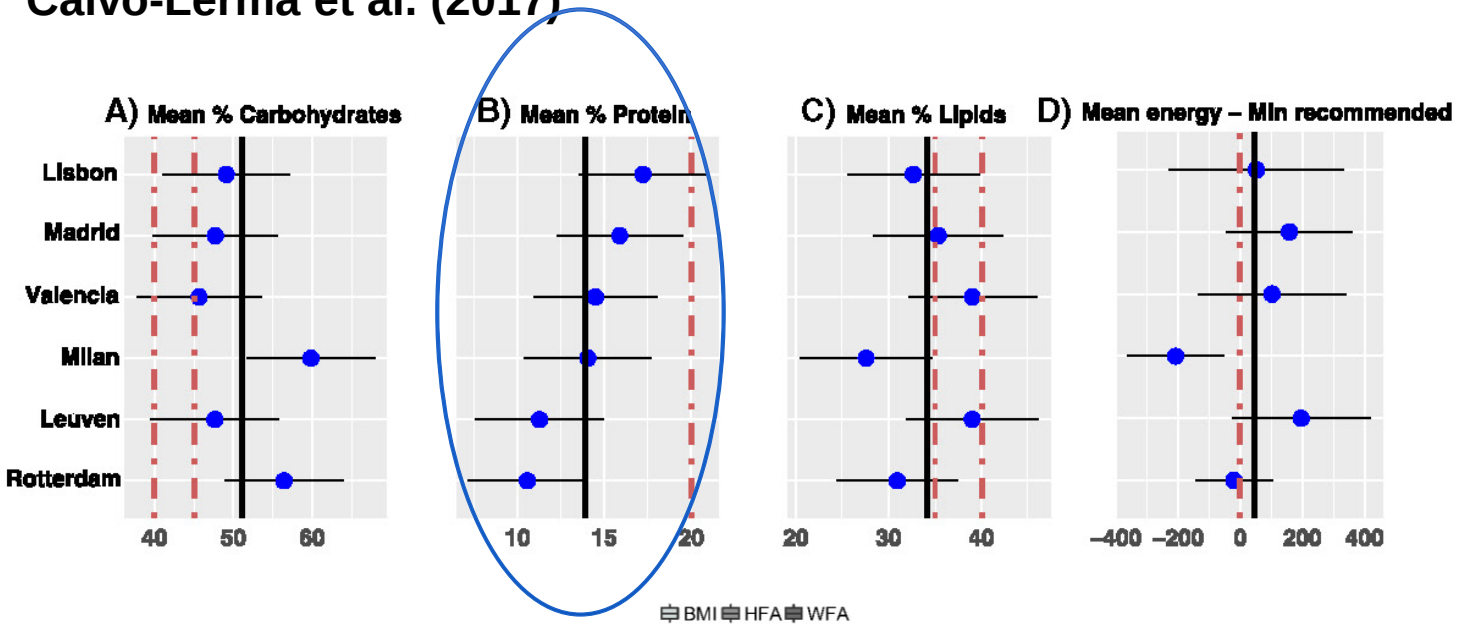
Estimating energy requirements (Davies et al. (2002))

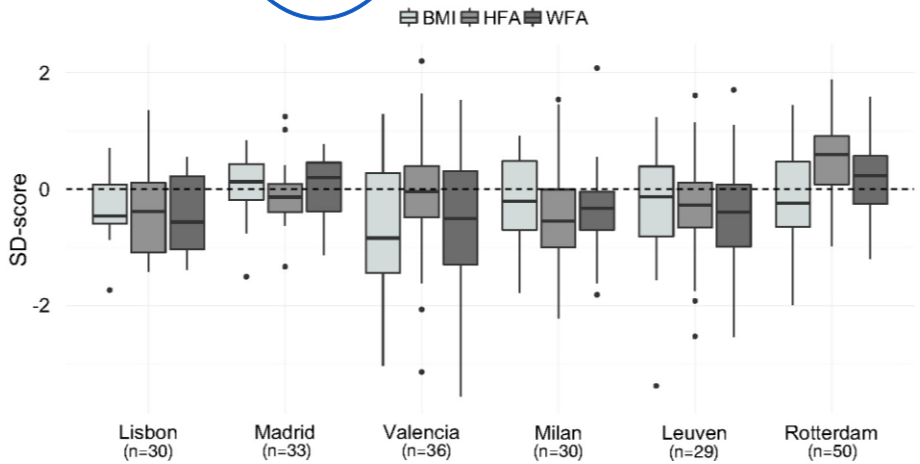
- Aim: longitudinal measurement of TEE
- Subjects: infants (n = 12) age 2,1(0,8)
- Methods: TEE and LBM determined by doubly labeled water at 2 – 6 – 12 months

	Age group											
	2 months				6 months				12 months			
	CF		Controls		CF		Controls		CF		Controls	
	Mean	s.d.	Mean	s.d.	Mean	s.d.	Mean	s.d.	Mean	s.d.	Mean	s.d.
Actual age (months)	2.1	0.8	2.5	0.01	5.8	0.5	5.9	0.2	12.4	1.4	12.0	0.1
Energy expenditure												
kJ/day	1686	384	1644	410	2803	481	2493	397	3585	1384	3372	740
kJ/kg body weight	348	76	297	75	435	95**	329	51	454	77	356	76
kJ/kg FFM	420	83	380	90	563	51**	451	65	562	110*	475	97
Z-Scores												
kJ/day	0.21	0.99			0.64	1.05			0.79	1.01		
kJ/kg body weight	0.67	0.96			1.63	1.45			1.31	0.97		
kJ/kg FFM	0.45	0.87			1.31	0.89			0.91	1.12		

Nutritient intake

Calvo-Lerma et al. (2017)





Nutritional requirements in CF

Protein (Engelen et al. (2014))

- Essential for building and restoring, plays a role in endocrine and immune functioning
- Adequate energy intake is essential to spare protein breakdown
- No evidence – based recommendations for daily protein intake
  - ≥ 20 EN%
- Dietary goals for protein intake -> recieve little attention

Invited Review

aspen LEADING THE SCIENCE AND PRACTICE OF CLINICAL NUTRITION
American Society for Parenteral and Enteral Nutrition

Protein Requirements of the Critically Ill Pediatric Patient

Jorge A. Coss-Bu, MD[1,2]; Jill Hamilton-Reeves, PhD, RD, CSO[3]; Jayshil J. Patel, MD[4]; Claudia R. Morris, MD[5]; and Ryan T. Hurt, MD, PhD[6]

Nutrition in Clinical Practice
Volume 32 Supplement 1
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journals.sagepub.com/home/ncp
SAGE

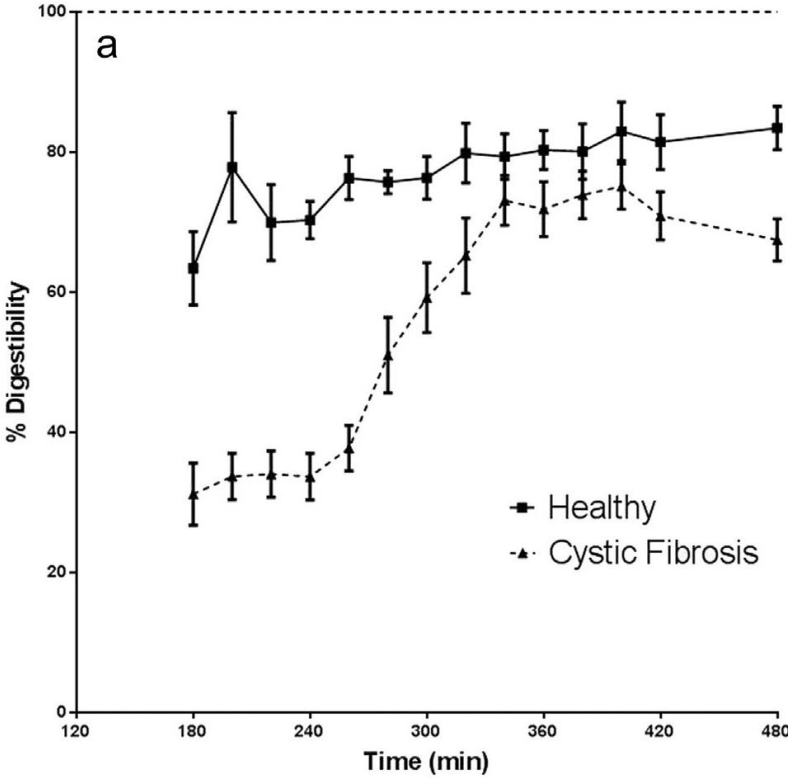
Age, y	Maintenance Requirement	Growth Requirement	Average Requirement	DRI
0.5	0.66	0.27	1.12	1.43
1	0.66	0.17	0.95	1.18
1.5	0.66	0.11	0.85	1.04
2	0.66	0.08	0.79	0.96
3	0.66	0.04	0.73	0.90
4–5	0.66	0.02	0.69	0.86
6–10	0.66	0.05	0.74	0.91
Girls				
11–15	0.66	0.03	0.71	0.88
16–18	0.66	0.01	0.67	0.83
Boys				
11–15	0.66	0.04	0.73	0.90
16–18	0.66	0.02	0.70	0.86

DRI, dietary reference intake. Adapted from Garlick PJ. Protein requirements of infants and children. Nestlé Nutrition workshop series. *Paediatr Program.* 2006;58:39-47. Copyright 2006 Nestec Ltd., Vevey/S. Karger AG, Basel.

- Inflammatory conditions:
  - ↓ protein synthesis at the skeletal muscle level
  - ↑ protein degradation -> shift of AA and N to the tissues
  - Loss of LBM
    ➔ development of growth failure
  - Insulin ➔ effect on whole-body protein metabolism only with adequate availability of AA, related to glucose and energy homeostasis

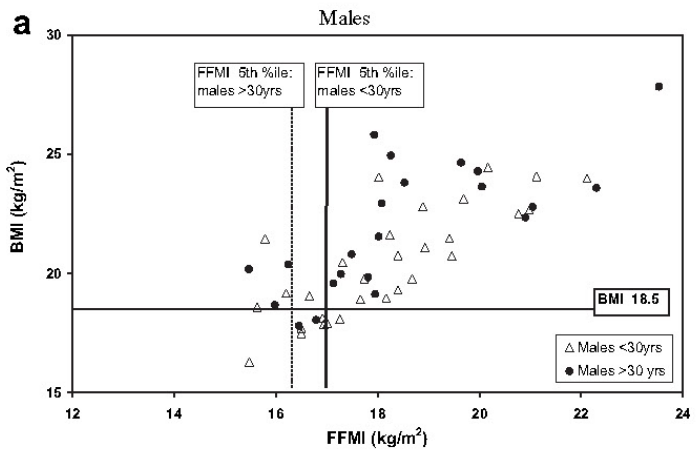
Nutritional requirements in CF

Engelen et al. (2014))



Nutritional assessment in CF

- Assessing body composition
  - LBM and BMC are more sensitive indicators of nutritional deficit than low BMI: low values predict impaired lung function in children and adults with CF



King S., et al. 2010 Nutrition

Independent variable*	Model 1, $\beta \pm$ SE (P value)	Model 2, $\beta \pm$ SE (P value)
Height, cm	1.11 ± 0.68 (0.12)	—
Body weight, kg	1.62 ± 0.30 (**<0.001**)	—
Body mass index, kg/m^2	4.89 ± 1.03 (**<0.001**)	—
Body fat, %	0.90 ± 0.63 (0.16)	−1.42 ± 0.64 (**0.04**)
Fat mass, kg	1.86 ± 0.29 (**0.004**)	−1.33 ± 1.09 (0.23)
Fat mass index, kg/m^2	4.71 ± 1.76 (**0.01**)	−6.44 ± 2.93 (**0.04**)
Fat-free mass, kg	2.67 ± 0.55 (**<0.001**)	1.70 ± 0.64 (**0.01**)
Fat-free mass index, kg/m^2	10.05 ± 1.99 (**<0.001**)	6.31 ± 2.93 (**0.04**)

Alvarez J., et al. 2016 Nutrition

Nutritional assessment in CF

- Need for salt/sodium supplementation
  - Fractional excretion of sodium (FENa) -> 0,5% - 1,5%
  - Routine practice: urinary sodium: creatinine -> 17 – 52 mmol/mmol

    (mmol to mg sodium x 23)
- Annually assessing calcium intake
- Annually all ages monitoring iron levels
  - Complicated by infection (ferritin and transferrin)
  - Serum transferrin receptors (sTfR) are not affected by inflammation
- At least annually and 3 – 6 months after a dosage change for fat-soluble vitamins

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