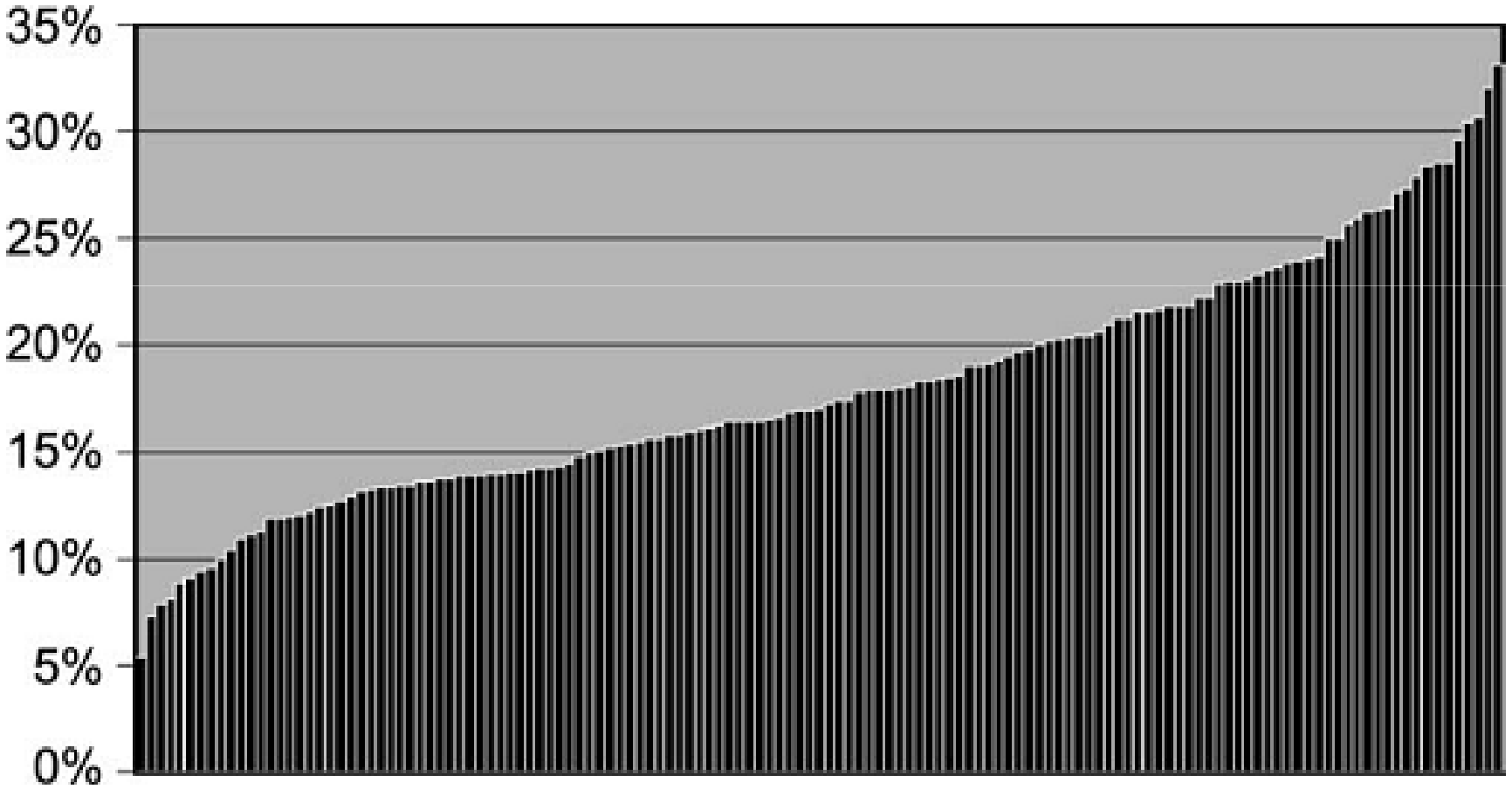
16:15 *Il bambino con fibrosi cistica: la dieta per respirare meglio?*
Giovanna Pisi, Giuseppe Magazzù

Di cosa parliamo

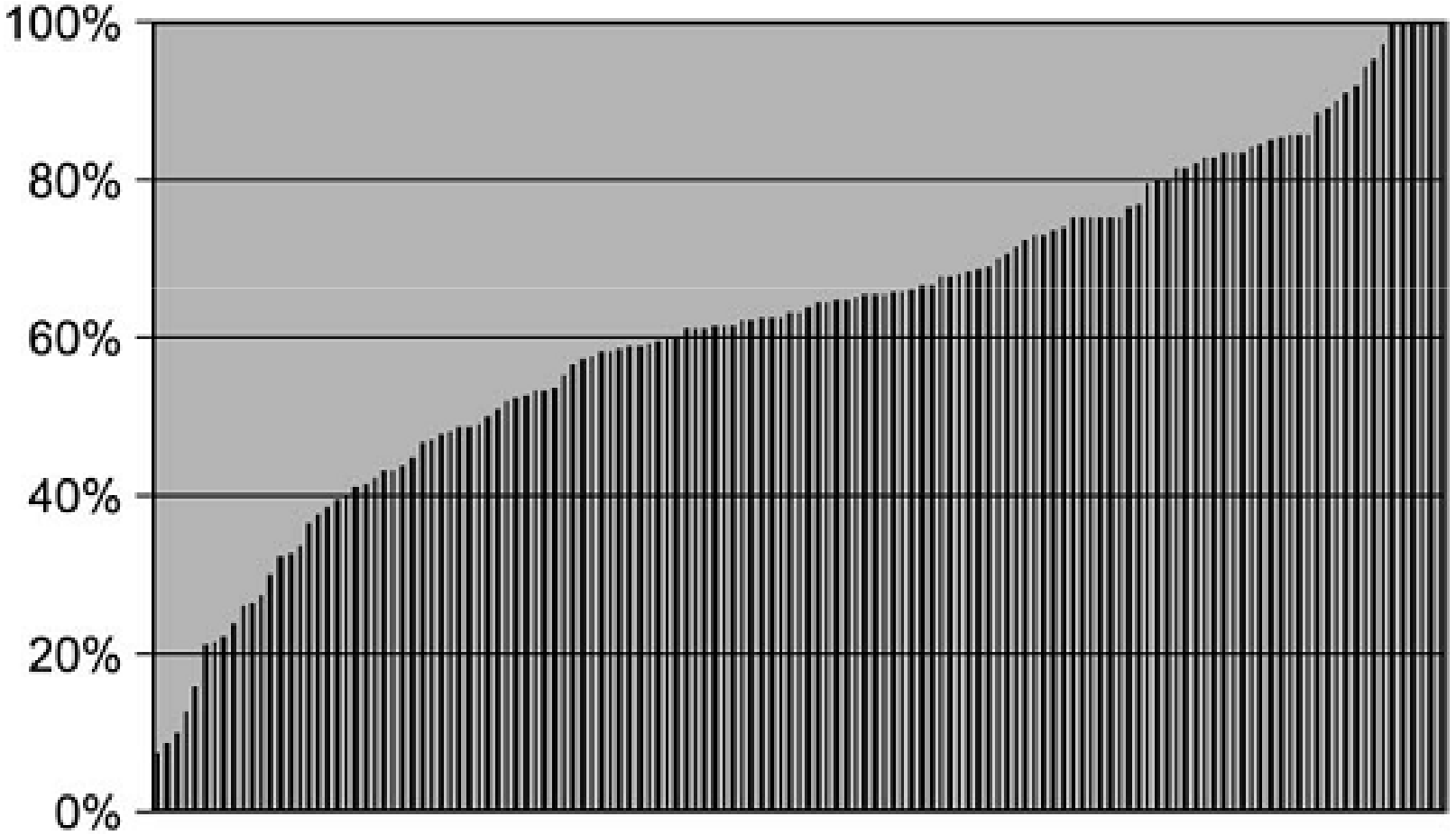
- L'importanza delle diverse attitudini dei Centri verso lo stato nutrizionale e l'alimentazione
- (Esempi di) Come le diverse attitudini possono influenzare il comportamento alimentare e lo stato di nutrizione dei bambini
- I fattori da prendere in considerazione nella riabilitazione nutrizionale

Cure variabili = esiti variabili



% DI PAZIENTI CON PESO< 5 C. NEI VARI CENTRI

Cure variabili = esiti variabili



% DI PAZIENTI CON P < 5 C. CON SUPPLEMENTAZIONE NUTRIZIONALE

Category	Target/group	Goals
Routine management	All CF patients	Nutritional education Dietary counseling PERT Vitamins
Anticipatory guidance	CF patients at risk to develop energy imbalance	Further education for increased energy needs Dietary intake monitoring Increased caloric density Behavioral counseling
Supportive intervention	Decreased weight velocity %W/H 85-90	All above plus oral supplement
Rehabilitative care	%W/H < 85	All above plus NG or PEG enteral supplement
Resuscitative/ palliative care	%W/H < 75	All above plus continuous EF or TPN

Ricoverato in TIN per ittero colestatico
Screening positivo per FC
Diagnosi genetica di Gilbert e FC
Avviata terapia sostitutiva pancreatica. Diagnosi bioptica di epatopatia FC >>>> UDCA
Offerta formula semielementare, supplementi proteici e latte materno a richiesta
A 83 giorni, PESO: 4.460 gr. centile: 5.9; lunghezza 55.0 cm. centile: 1.3; % del peso ideale per altezza 101.6
A 7 mesi la colestasi da FC si attenua ma peso e lunghezza al 3^ centile (inferiore al target genetico).
Assume ancora idrolisato con aggiunta di MCT
Da 1 settimana svezzamento con la raccomandazione di cibi ricchi in grassi da aggiungere a un elenco di creme e omogeneizzati che il b. "sputacchia", con proibizione dell'uovo fino a 12 mesi. Il b. è molto attratto dalle pietanze che vede sulla tavola della famiglia che vengono proibite

Category	Target/group	Goals
Routine management	All CF patients	Nutritional education Dietary counseling PERT Vitamins
Anticipatory guidance	CF patients at risk to develop energy imbalance	Further education for increased energy needs Dietary intake monitoring Increased caloric density Behavioral counseling
Supportive intervention	Decreased weight velocity %W/H 85-90	All above plus oral supplement
Rehabilitative care	%W/H < 85	All above plus NG or PEG enteral supplement
Resuscitative/ palliative care	%W/H < 75	All above plus continuous EF or TPN

Corey 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

- Marked difference in median age of

Il bambino deve mangiare tutto quello che desidera e vede sulla tavola, incluso l'uovo, che può mangiare già nelle polpette al sugo

- Patients in Toronto were taller than those in Boston

Ricoverato in TIN per ittero colestatico
Screening positivo per FC
Diagnosi genetica di Gilbert e FC
Avviata terapia sostitutiva pancreatica Diagnosi bioptica di epatopatia FC >>>> UDCA
Offerto formula semielementare, supplementi proteici e latte materno a richiesta
A 83 giorni, PESO: 4.460 gr. centile: 5.9; lunghezza 55.0 cm. centile: 1.3; % del peso ideale per altezza 101.6
A 7 mesi la colestasi da FC si attenua ma peso e lunghezza al 3^ centile (inferiore al target genetico).
Assume ancora idrolisato con aggiunta di MCT
Da 1 settimana svezzamento con la raccomandazione di cibi ricchi in grassi da aggiungere a un elenco di creme e omogeneizzati che il b. "sputacchia", con proibizione dell'uovo fino a 12 mesi. Il b. è molto attratto dalle pietanze che vede sulla tavola della famiglia che vengono proibite

Journal of Pediatric Gastroenterology and Nutrition
35:246–259 © September 2002 Lippincott Williams & Wilkins, Inc., Philadelphia

Consensus Report on Nutrition for Pediatric Patients With Cystic Fibrosis

*Drucy Borowitz, *Robert D. Baker, and †Virginia Stallings

TABLE 2. *Definition of nutritional failure in patients with CF and those at risk*

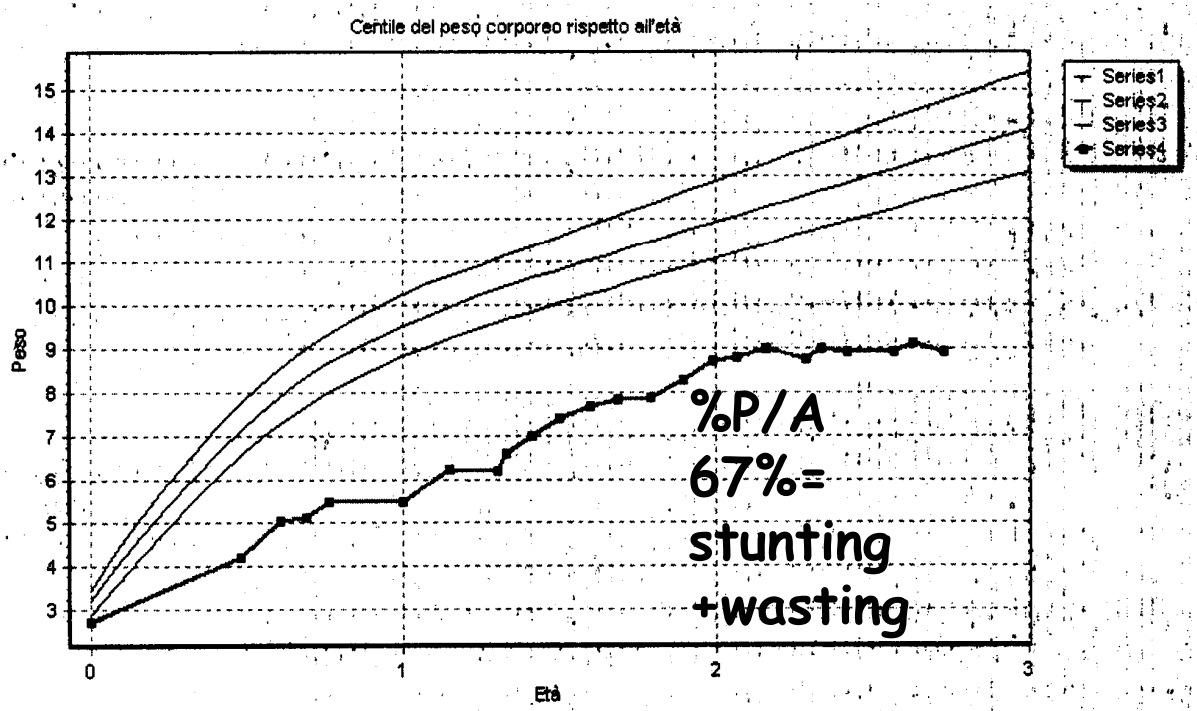
Nutritional status	Length or height	Percentage IBW[1] All ages	Weight-for-length percentile[2] 0 to 2 years	BMI percentile[3] 2 to 20 years	Action
Acceptable	Normal growth	≥90%	>25th	>25th	Continue to monitor with usual care
At-risk[4]	Not at genetic potential	≥90%, with weight loss or weight plateau[5]	10 to 25th	10 to 25th	Consider nutritional and medical evaluation; some but not all patients in this category are at risk for nutritional failure
Nutritional failure	<5%ile	<90%	<10th	<10th	Treat nutritional failure

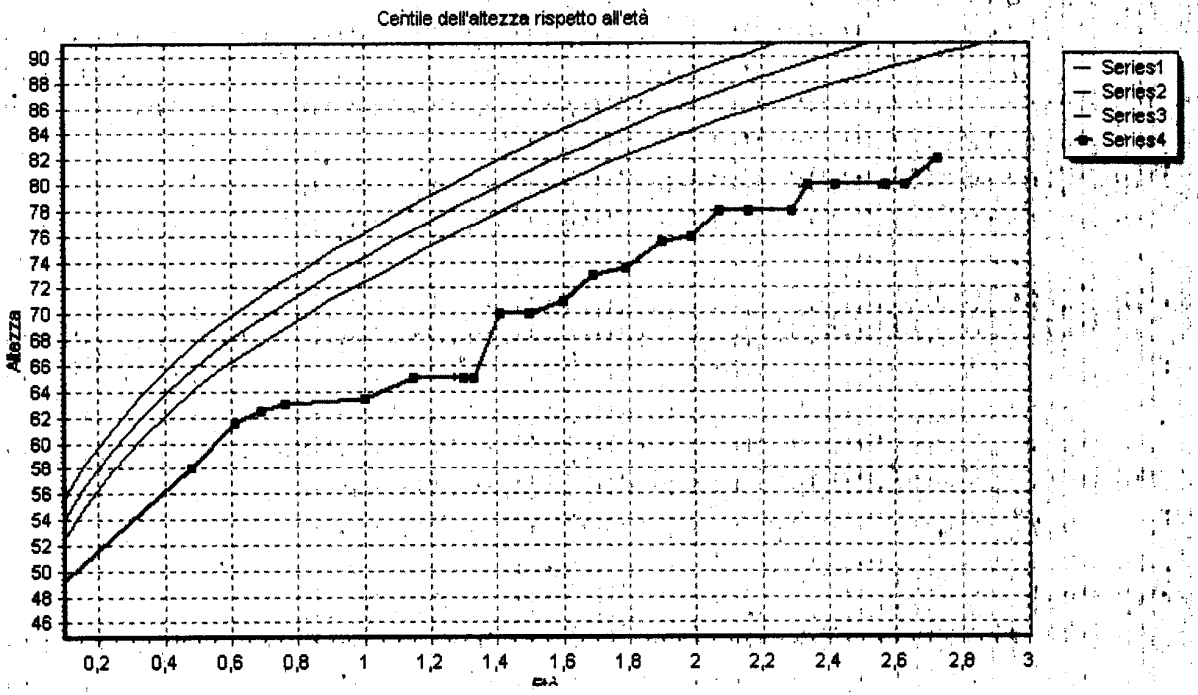
Comparison of the use of body mass index percentiles and percentage of ideal body weight to screen for malnutrition in children with cystic fibrosis[1–3]

Zhumin Zhang and HuiChuan J Lai

Am J Clin Nutr 2004;80:982–91.

Compared with BMIp, %IBWCFF underestimated the severity of malnutrition in children with short stature and overestimated the severity of malnutrition in children with tall stature. Am J Clin Nutr 2004;80:982–91.





Diagnosticata per sintomi a 6 mesi con grave insuff. respiratoria

Staf. Aureo

Persistente steatorrea nonostante PERT

Escluse altre cause GI

Enterale x NG con semielem. a 1 cal/ml

Peso: 800g in 8 gg

x 2 mesi e poi PEG?

Stature as a prognostic factor in cystic fibrosis survival

LEILA T. BEKER, PhD, RD; ESTELLE RUSSEK-COHEN, PhD; ROBERT J. FINK, MD
J Am Diet Assoc. 2001;101:438-442.

Table 1
Males with a height-for-age below the fifth NCHS percentile are more likely to die[a]

Age	Log relative hazard (±standard error)	Relative hazard (95% Confidence Interval)
5 years	−1.07* (±0.44)	2.9[a] (1.23, 6.91)
7 years	−1.84*** (±0.56)	6.3 (2.10, 18.87)

[a]Based on analysis of 1,170 subjects.
[b]Relative hazard of 2.9 implies shorter children are 2.9 times more likely to die given they have survived to a given age.
*$P<.02$.
***$P<.001$.

Table 2
Females with height for age below the fifth NCHS percentile are more likely to die[a]

Age	Log relative hazard (±standard error)	Relative hazard (95% Confidence Interval)
5 years	−1.46*** (±0.27)	4.3[b] (2.54, 7.31)
7 years	−1.75[d] (±0.42)	5.8 (2.53, 13.11)

[a]Based on analysis of 1,103 female subjects.
[b]Relative hazard of 4.3 implies shorter children are 4.3 times more likely to die given they have survived to a given age.
***$P<.0001$.
[d]$P>.0001$.

Article in Press
Stunting is an independent predictor of mortality in patients with cystic fibrosis

Clin Nutr

Body composition and mortality in chronic obstructive pulmonary disease[1,2]

Annemie MWJ Schols, Roelinka Broekhuizen, Clarie A Weling-Scheepers, and Emiel F Wouters

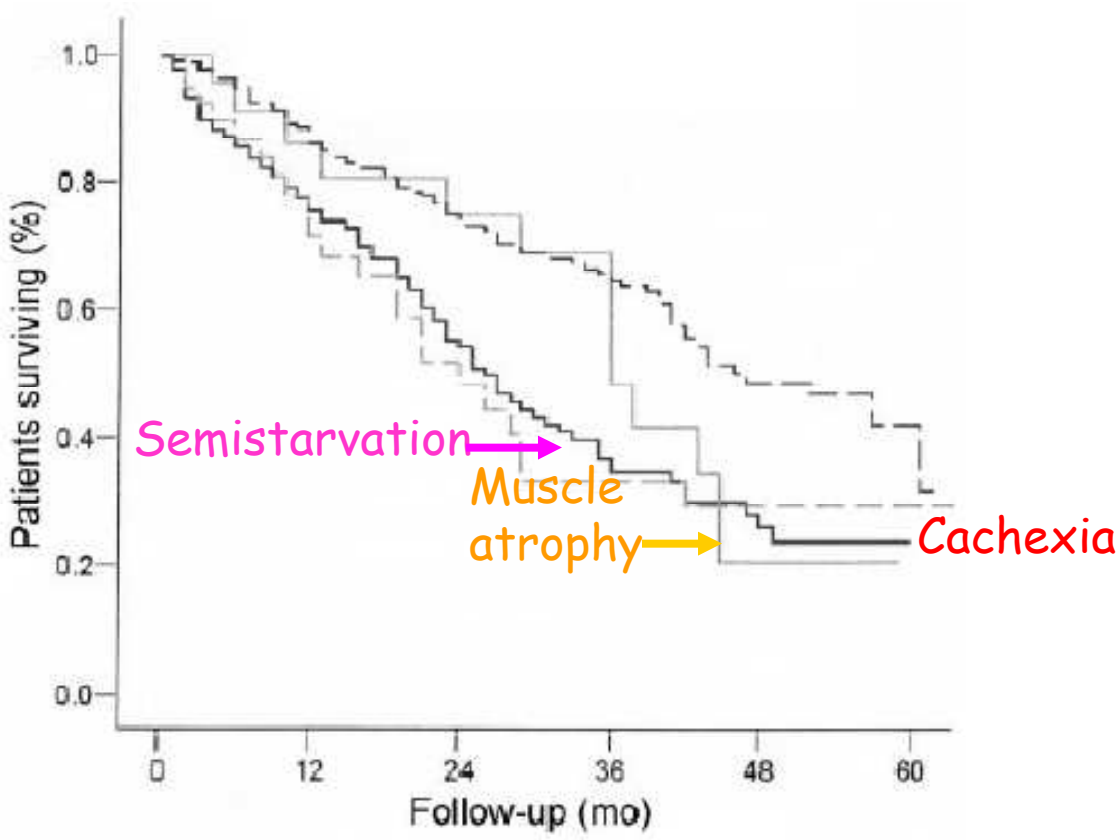
BIA Nutritional categories

Patients in category 1 (cachexia) had BMI < 21 and FFMI < 16 (men) or < 15 (women);

Patients in category 2 (semistarvation) had BMI < 21 and FFMI > 16 (men) or > 15 (women);

Patients in category 3 (muscle atrophy) had BMI> 21 and FFMI< 16 (men) or< 15 (women);

Patients in category 4 (no impairment) had BMI 21 and FFMI 16 (men) or 15 (women).



Fat-free mass depletion in cystic fibrosis: Associated with lung disease severity but poorly detected by body mass index

Susannah J. King Ph.D.[a,b,c,*], Ibolya B. Nyulasi M.Sc.[a,c], Boyd J.G. Strauss Ph.D.[a,e], Tom Kotsimbos M.D.[b,d], Michael Bailey Ph.D.[f], John W. Wilson Ph.D.[b,d]

Nutrition 2009

-Using a BMI <18.5 kg/m2 to define malnutrition fails to identify 58% of patients with CF and FFM depletion

-Reduced FFMI is common in CF and is associated with low FEV1% predicted

-There is a role for body composition assessment in CF clinical practice

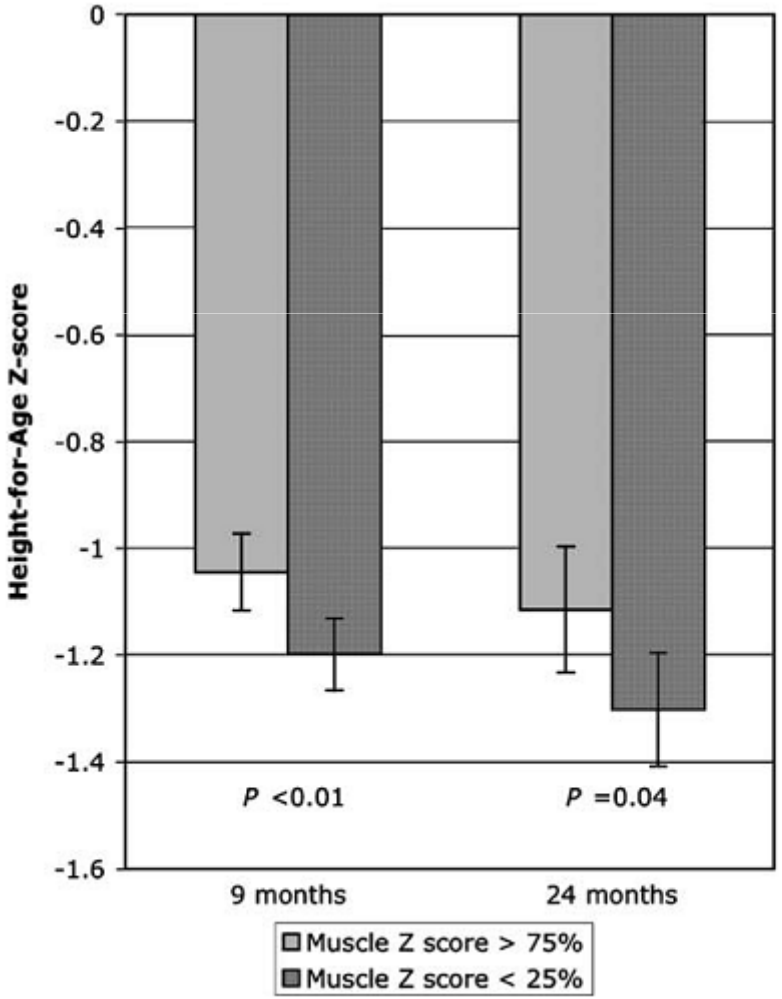
European Journal of Clinical Nutrition (2005) 59, 914–922

0954-3007/05 $30.00
www.nature.com/ejcn

ORIGINAL COMMUNICATION

Progression of stunting and its predictors among school-aged children in western Kenya

JF Friedman[1,2]*, PA Phillips-Howard[3,4], LB Mirel[3], DJ Terlouw[3], N Okello[4], JM Vulule[4], WA Hawley[3], BL Nahlen[3] and F ter Kuile[3,4,5]



Nutritional effects of long-term gastrostomy feedings in children with cystic fibrosis

MARGARET ROSENFELD, MD, MPH; SUSAN CASEY, RD; MARGARET PEPE, PhD; BONNIE W. RAMSEY, MD

J AM Diet Assoc 1999

Table 2
Nutritional status at baseline and after initiation of gastrostomy feedings among 18 subjects followed up for a minimum of 18 months after gastrostomy[a]

	No. of years before gastrostomy		6 to 18 mo after gastrostomy		18 to 30 mo after gastrostomy	
	Percentile	Range	Percentile	Range	Percentile	Range
Weight	2	1,6	12	1,28[b]	12	1,29[b]
Height	5	1,34	6	1,48[c]	10	2,34[c]
	%	*range*	%	*range*	%	*range*
Weight as % of ideal	88	80,92	90	85,99[b]	93	86,98[b]

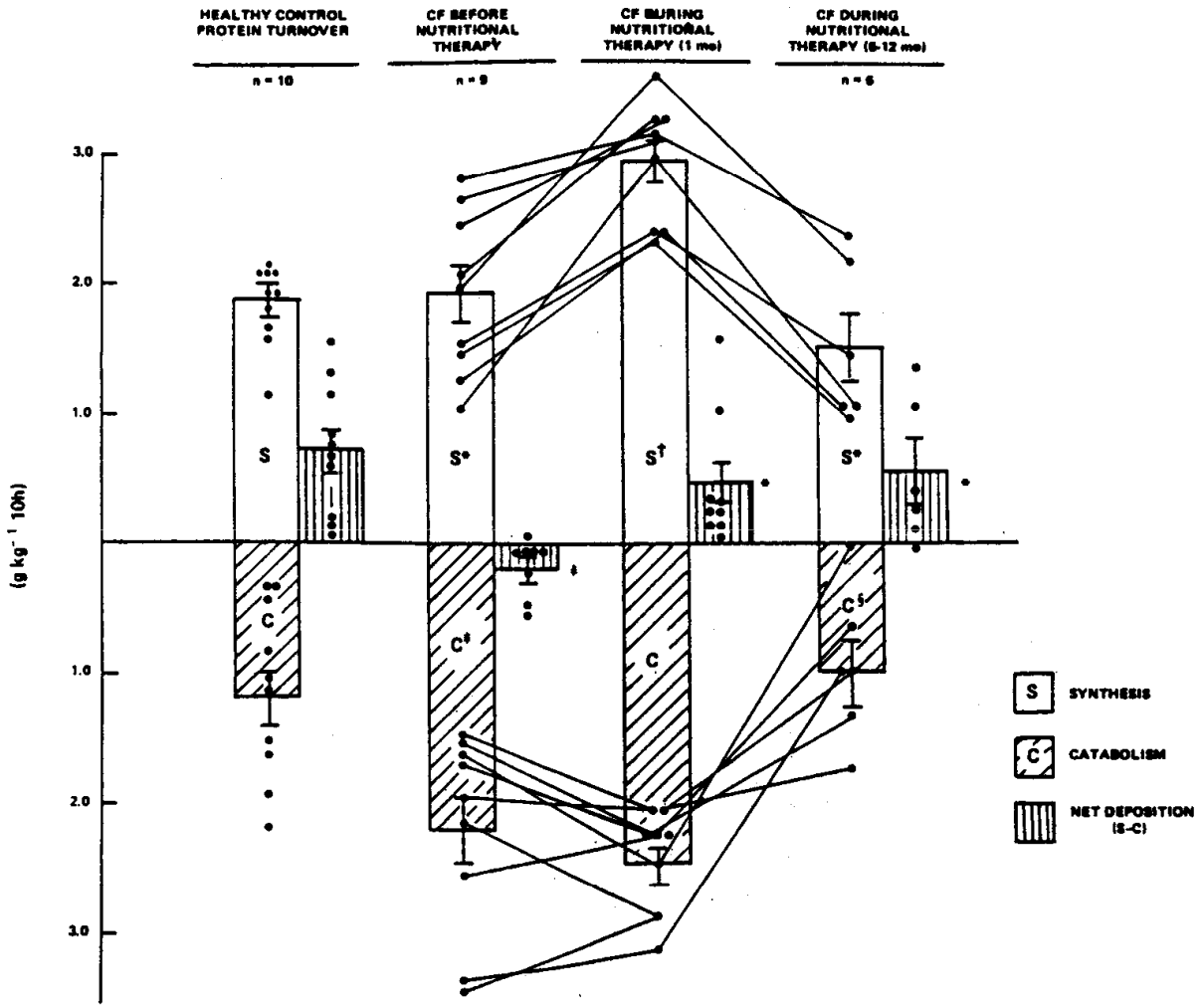


Fig. 3. Rates of whole-body protein synthesis *(S)*, catabolism *(C)*, and net deposition *(S-C)* showing individual data points, comparing data from healthy children (n = 10) with data from patients with cystic fibrosis (n = 9) before and during (1 month and 6 to 12 months) nutritional supplementation. *S and S-C vs control data, not significant. †S during therapy (1 month) vs S before and S control, P <0.001. ‡C before therapy vs C control, P <0.02. §C during therapy (6 to 12 months) vs C control, not significant; vs C at 1 month, P <0.001.

16:15 *Il bambino con fibrosi cistica: la dieta per respirare meglio?*

Giovanna Pisi, Giuseppe Magazzù

RISPOSTE E CONCLUSIONI

MIGLIORE NUTRIZIONE PER RESPIRARE MEGLIO

DIETA RICCA E NATURALE SIN DALLO SVEZZAMENTO

INTERVENTI NUTRIZIONALI PRECOCI TENENDO IN CONTO LA MASSA MAGRA E LA CRESCITA LINEARE

RCT NECESSARI PER INTERVENTI NUTRIZIONALI INVASIVI SUCCESSIVAMENTE