



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

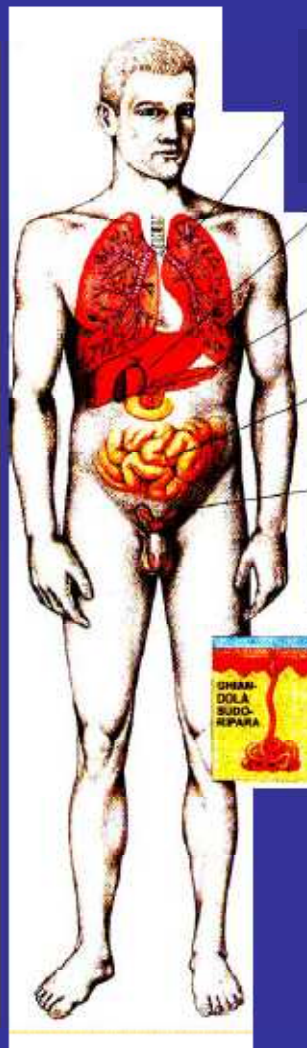
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CFTR: a multisystemic protein



airways

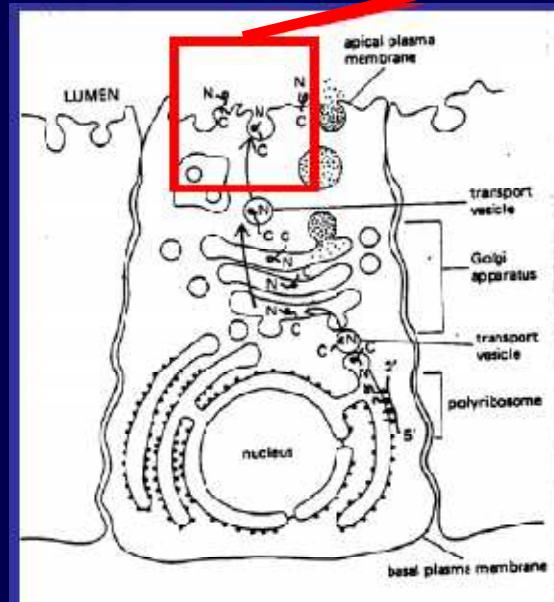
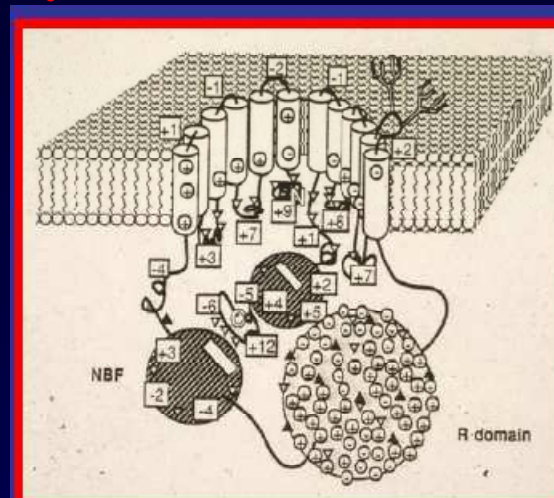
liver

pancreas

gut

testicle

sweat glands



from Riordan JR et al., Science 1989

Similarities between GI & respiratory tract

Interaction between nutritional status and lung function

How does nutritional status affect lung function?

Similarities between GI & respiratory tract

Interaction between nutritional status and lung function

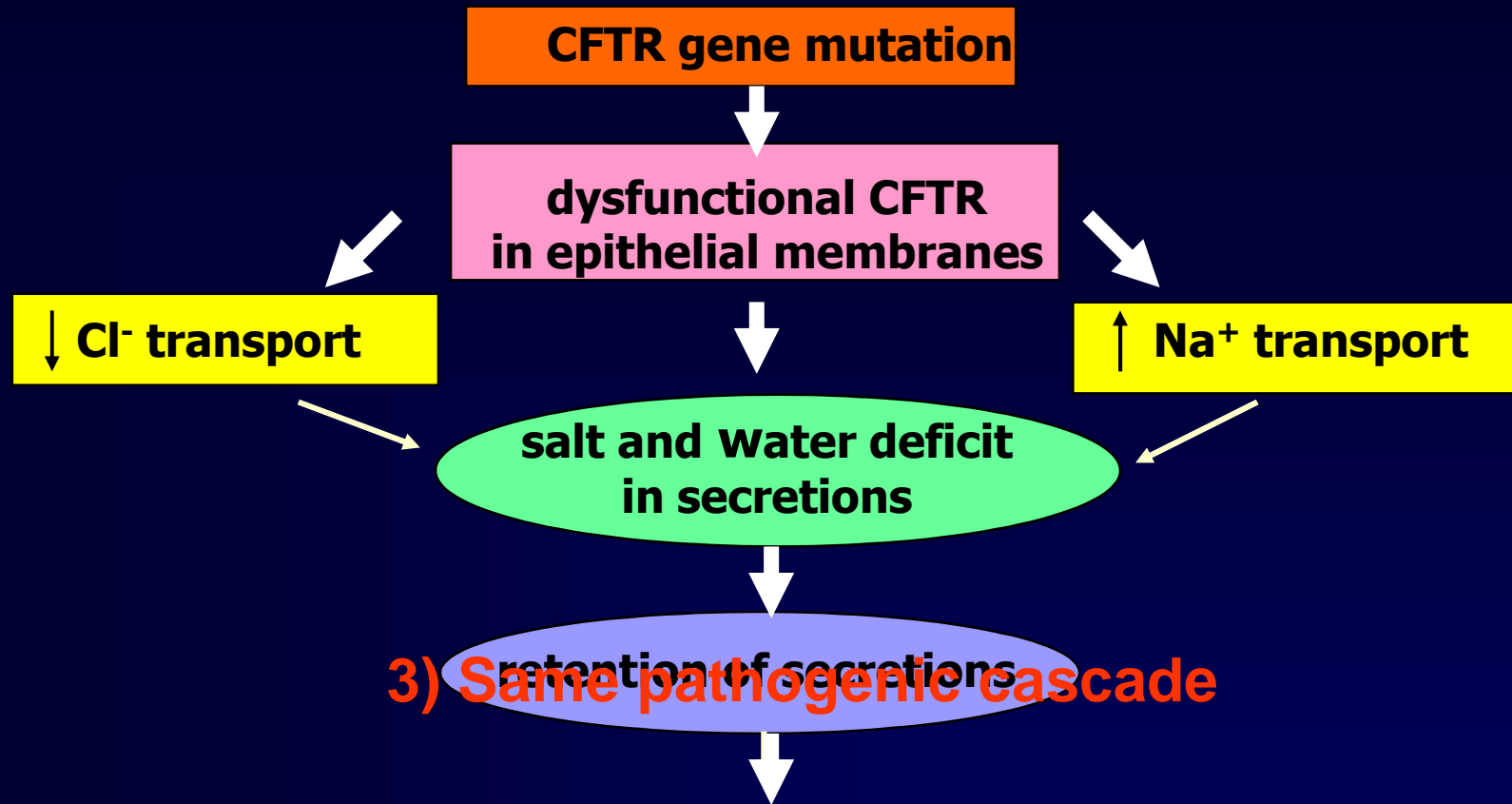
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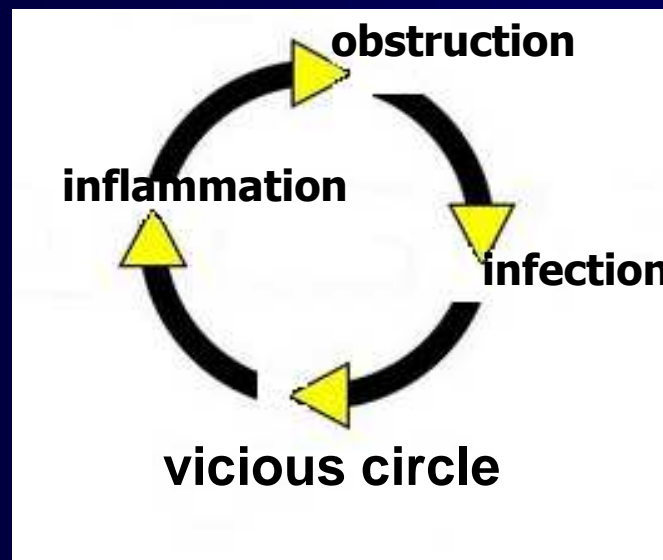
1) Same embryological origin

2) Mechanisms of clearance of obstruction and infection

Lung	GI tract
ASL hydration +MCC	Luminal hydration + microvilli
Cough	Peristalsis
Antimicrobial function BALT	Antimicrobial function GALT
Submucosal glands	Submucosal glands



3) Same pathogenic cascade



4) Respiratory & Gut Microbiome



Serial Analysis of the Gut and Respiratory Microbiome in Cystic Fibrosis in Infancy: Interaction between Intestinal and Respiratory Tracts and Impact of Nutritional Exposures

J. C. Madan et Al.

The majority of the bacteria present in the gut and in the respiratory tract were represented by 8 distinct genera and the core microbiota was dominated by *Veillonella* and *Streptococcus*.

Bacterial diversity increased significantly over time, more rapidly in the respiratory tract.

There was a high degree of concordance between the bacteria in both compartments. For some bacteria, gut colonization presages their appearance in the respiratory tract.

Some profiles of respiratory bacteria are associated with breast-feeding, and intestinal microbiota with introduction of solid foods .

Conclusions: nutritional factors and gut colonization patterns are determinants of the microbial development of respiratory tract microbiota in infants with CF and present opportunities for early intervention in CF with altered dietary or probiotic strategies.

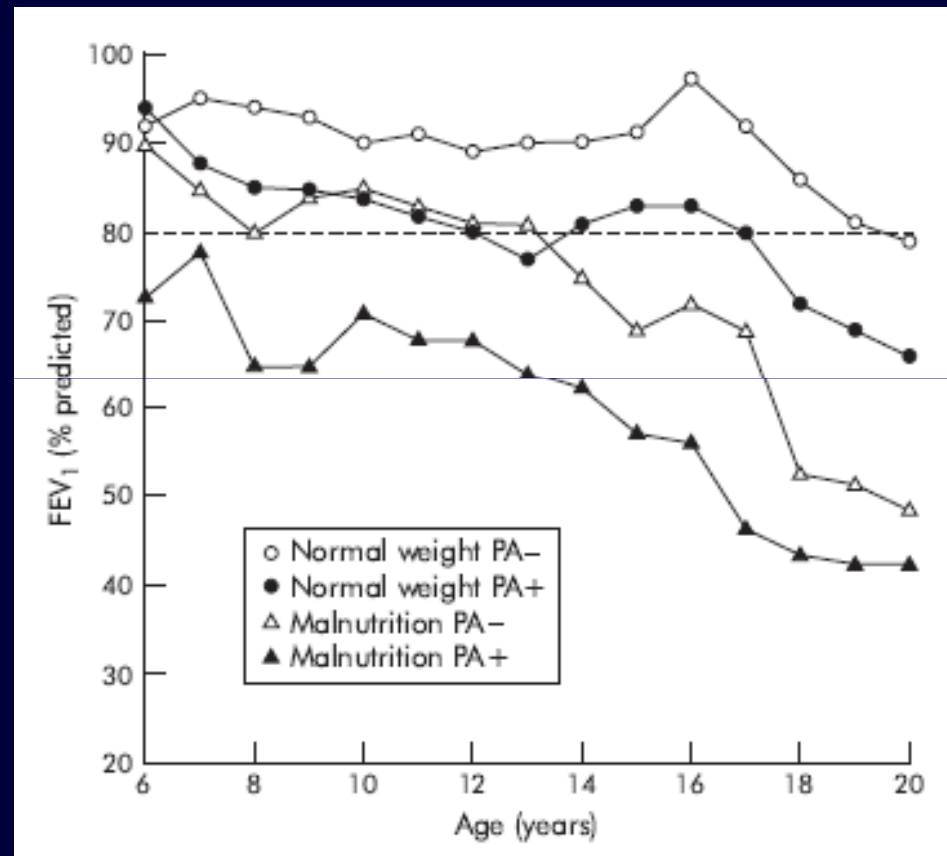
Similarities between GI & respiratory tract

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How does nutritional status affect lung function?

Relationship between nutritional status and lung function in cystic fibrosis: cross sectional and longitudinal analyses from the German CF quality assurance (CFQA) project

G Steinkamp, B Wiedemann, on behalf of the German CFQA Group Thorax 2002;57:596–601



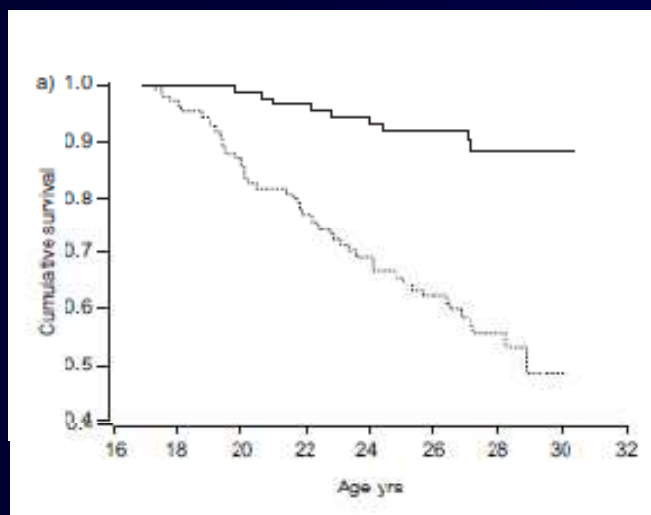
In malnourished adolescents yearly \downarrow weight for height $>5\%$ pred. associated with \downarrow FEV₁ of 16.5% pred.



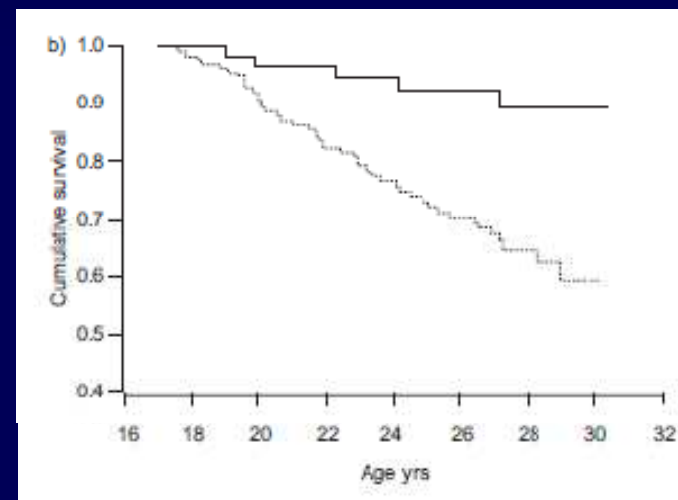
From registry to quality management: the German Cystic Fibrosis Quality Assessment project 1995–2006

M. Stern*, B. Wiedemann[#] and P. Wenzlaff[†] on behalf of the German Cystic Fibrosis Quality Assessment group

Cumulative survival (1995–2005) vs age in CF patients in 1995 in relation to nutritional status and respiratory function



— BMI ≥ 19 kg/m²
..... BMI < 19 kg/m²

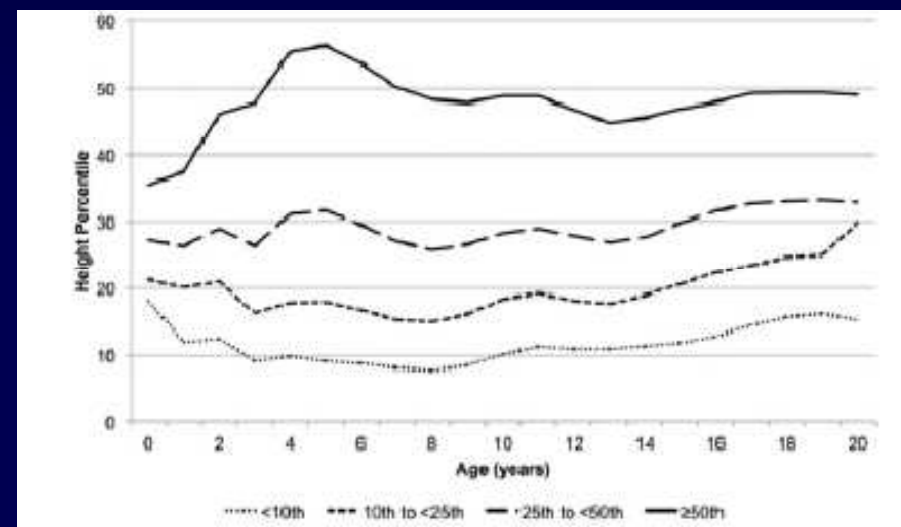
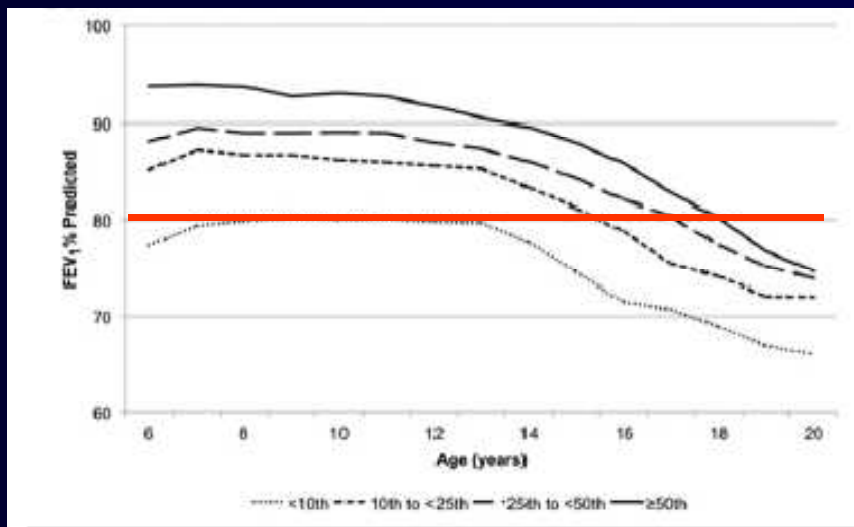


— FEV1 ≥ 80 %
..... FEV1 < 80 %

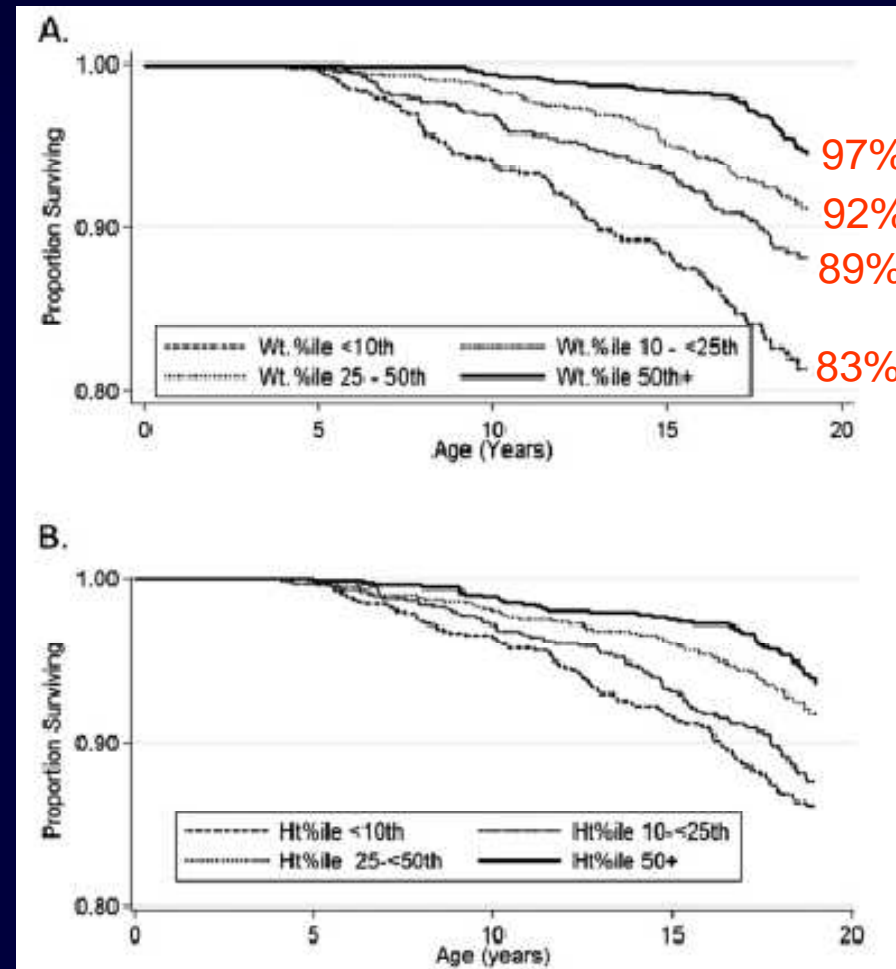
Better Nutritional Status in Early Childhood Is Associated with Improved Clinical Outcomes and Survival in Patients with Cystic Fibrosis

Elizabeth H. Yen, MD¹, Hebe Quinton, MS², and Drucy Borowitz, MD³

Studio prospettico, > 3000 paz. CFF registry , nati tra 1989- 1992



Kaplan-meyer survival curves of CF patients stratified by weight and height categories at age 4 years



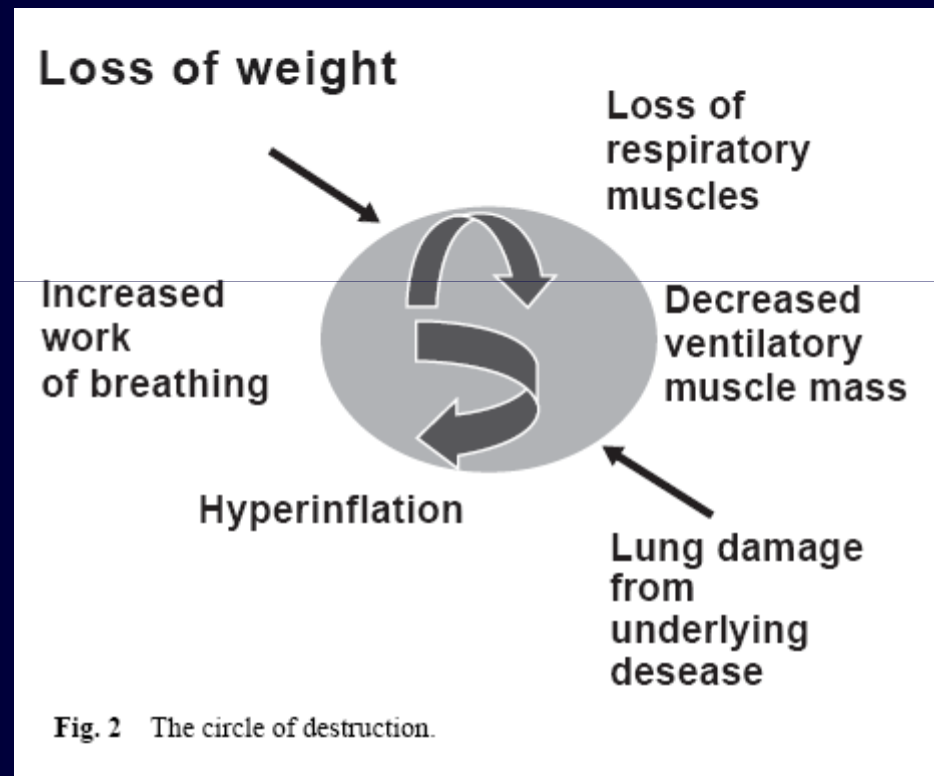
**Patients attaining higher age 4 years WAP and HAP
had a survival advantage throughout childhood**

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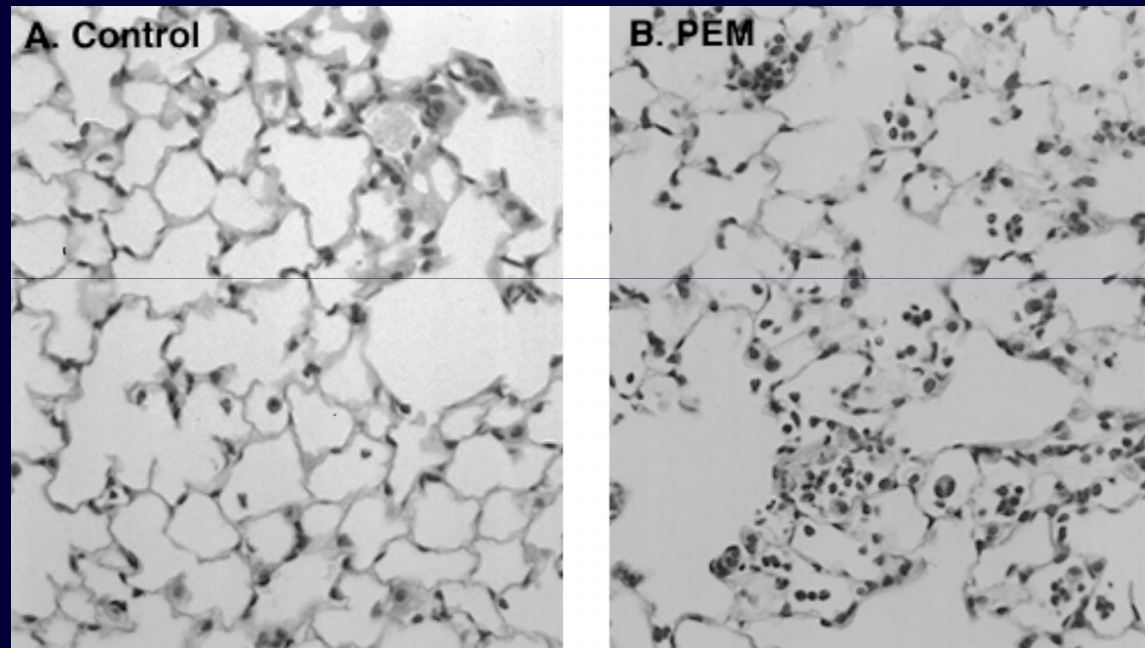
How does nutritional status affect lung function?

Effect of lean body mass depletion



Effect of protein energy malnutrition

Innate Lung Defenses and Compromised *Pseudomonas aeruginosa* Clearance
in the Malnourished Mouse Model of Respiratory Infections in Cystic Fibrosis
H. Yu, S. Z. Nasr and V. Deretic *Infect. Immun.* 2000, 68(4):2142.



Increased neutrophil and inflammatory cell infiltration in malnourished mice fed low-protein diet and exposed to *P. aeruginosa*.

Effect of Supplementing a High-Fat, Low-Carbohydrate Enteral Formula in COPD Patients

Baiqiang Cai, MD, Yuanjue Zhu, MD, Yi Ma, MD, Zuojun Xu, MD, Yi Zao, MD, Jinglan Wang, MD, Yaoguang Lin, MD, and Gail M. Comer, MD

Nutrition 2003;19:229–232.

Metabolic and ventilatory parameters (RQ, VCO₂, VO₂, VE) decreased significantly and FEV₁ increased significantly in the experimental group which received a high-fat, low-CHO oral supplement (17% protein, 55% fat, 28% CHO) in the evening as part of the diet.

Conclusion: pulmonary function in COPD patients can be significantly improved with a high-fat, low-CHO oral supplement as compared with the traditional high-CHO diet.

Conclusions

- 1) The association of better nutritional status and improved lung function is well documented.
- 2) Poor nutrition is a major risk factor for accelerated decline in lung function.
- 3) Poor nutrition and impaired lung function correlated with augmented mortality.
- 4) The association between nutritional status, respiratory outcomes, and survival starts early in life.
- 5) So gains were greatest when nutritional intervention and therapy initiated during infancy.