



ÇOCUK GÖĞÜS
HASTALIKLARI
3. KONGRESİ



OLGU SUNUMU

Ankara Çocuk Sağlığı ve Hastalıkları
Hematoloji-Onkoloji Hastanesi

Dr. Gökçen Dilşa TUĞCU

OLGU 1: N. B. 15 yař, kız hasta

- ▶ Term, NSVY ile dođmuř
- ▶ Sık tekrarlayan akciđer enfeksiyonu, malnütrisyon
- ▶ Anne-Baba 1. derece akrabalık+ (hala-dayı çocukları)
- ▶ 4 yařında KF tanısı almıř.

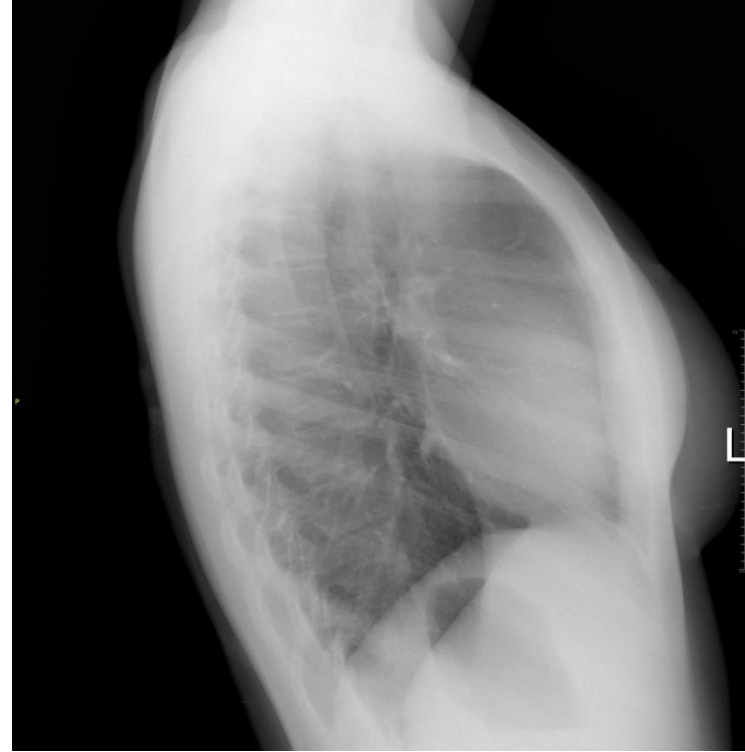
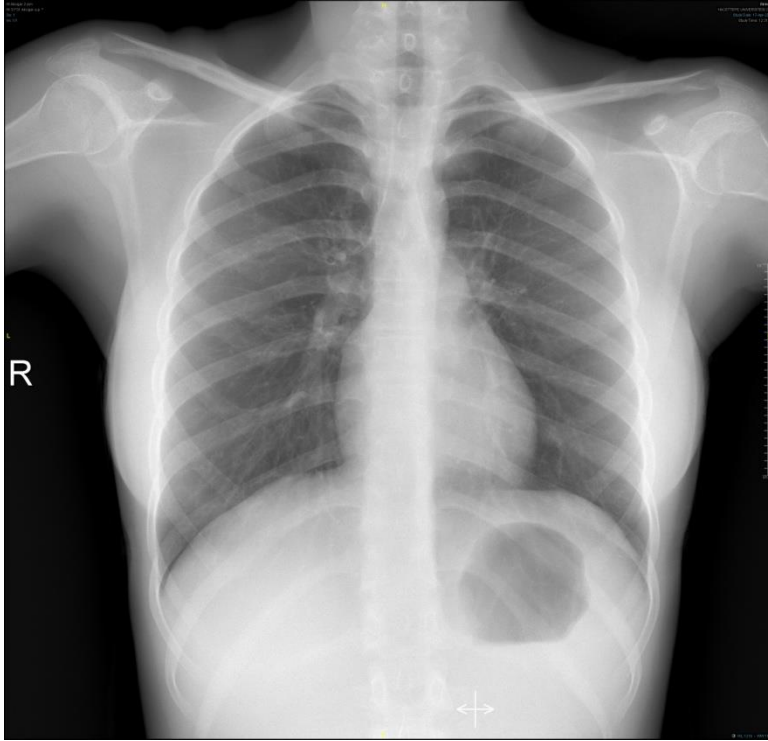


Fizik İnceleme Bulguları:

- ▶ SpO2: %98
- ▶ Nazal polip+
- ▶ Bilateral bazallerde daha belirgin kaba krepitan raller+
- ▶ Orta derecede çomak parmak
- ▶ Karaciğer subkostal yaklaşık 1 cm ele geliyor.



AKCİĞER GRAFİSİ

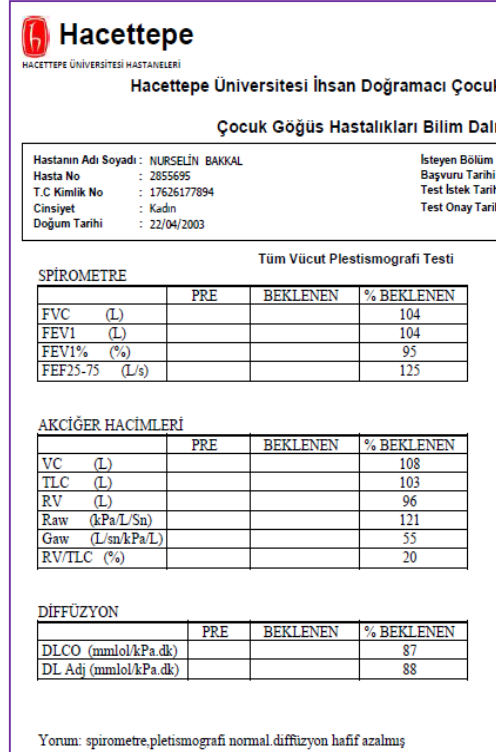
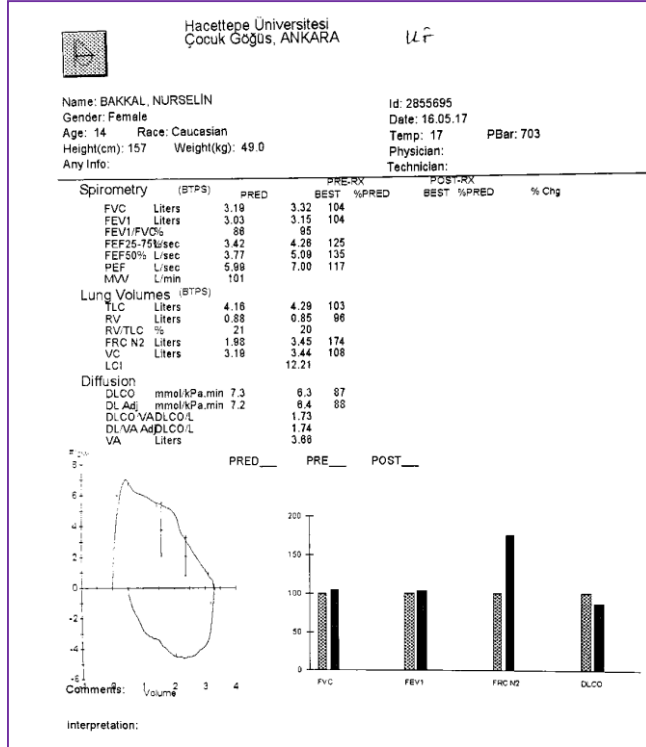


Tetkikleri:

- ▶ **Ter testi:** Cl 136 mmol/L
- ▶ **Ig:** Yaşa göre normal
- ▶ **Balgam kültürü:** *S. aureus*
- ▶ **CFTR sekans analizi:**R117H/L206W
- ▶ **EKO:** Hafif AY, MY, İAS'da anevrizma
- ▶ HbaIc, koagülasyon testleri ve vitamin düzeyleri normal
- ▶ Abdominal USG de karaciğer parankimi heterojen, kontrakte safra kesesi+



Solunum Fonksiyon Testi



FEV1: % 104

FVC: %104

FEV1/FVC:%95

FEF25-75: % 125

TLC: %103

DLCO: %87

LCI: 12.21

OLGU 2: D.D.,11 Yaş, Erkek Hasta

- ▶ Term, NSVY ile doğmuş, malnütrisyon, elektrolit imbalansı
 - ▶ Sürekli balgamlı öksürük
 - ▶ Tekrarlayan sinüzit ve tekrarlayan otit
 - ▶ Anne-Baba 1. derece akrabalık+ (kuzen evliliği)
 - ▶ 18 aylıkken KF tanısı almış.
-

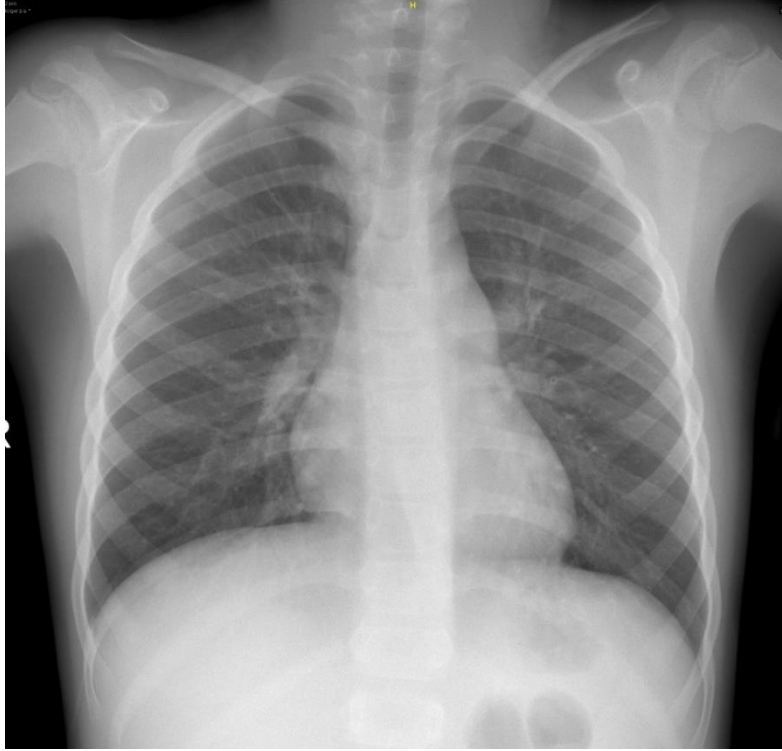


Fizik İnceleme Bulguları

- ▶ SpO₂: %96
- ▶ Bilateral bazallerde belirgin kaba krepitan raller+
- ▶ Hafif çomak parmak
- ▶ Tonsiller hipertrofik/hiperplazik
- ▶ Karaciğer subkostal 2 cm ele geliyor.



Akciğer Grafisi



Tetkikleri:

- ▶ **Ter testi:** Cl 142 mmol/L
 - ▶ Kantitatif immunoglobülinler yaşına göre normal
 - ▶ **Balgam kültürü:** *A. Baumannii*, *P.aeroginosa* ile kronik kolonize
 - ▶ Hba I c, koagülasyon testleri ve vitamin düzeyleri normal
 - ▶ Abdominal USG de karaciğer parankimi heterojen
 - ▶ EKO: Normal
 - ▶ **CFTR sekans analizi:**L299F/D1152H
-



Solunum Fonksiyon Testi

Hacettepe Üniversitesi İhsan Doğramacı Çocuk Hastanesi
Göğüs Hastalıkları Ünitesi

Hastanın Adı Soyadı : DAĞHAN DAĞ
Hasta No : 2875971
T.C Kimlik No : 10877627200
Cinsiyet : Erkek
Doğum Tarihi : 23/08/2007
Başvuru Tarihi : 13/02/2017

Tüm Vücut Plestismografi Testi

SPIROMETRE

	PRE	BEKLENEN	% BEKLENEN
FVC (L)			104
FEV1 (L)			92
FEV1% (%)			83
FEF25-75 (L/s)			88

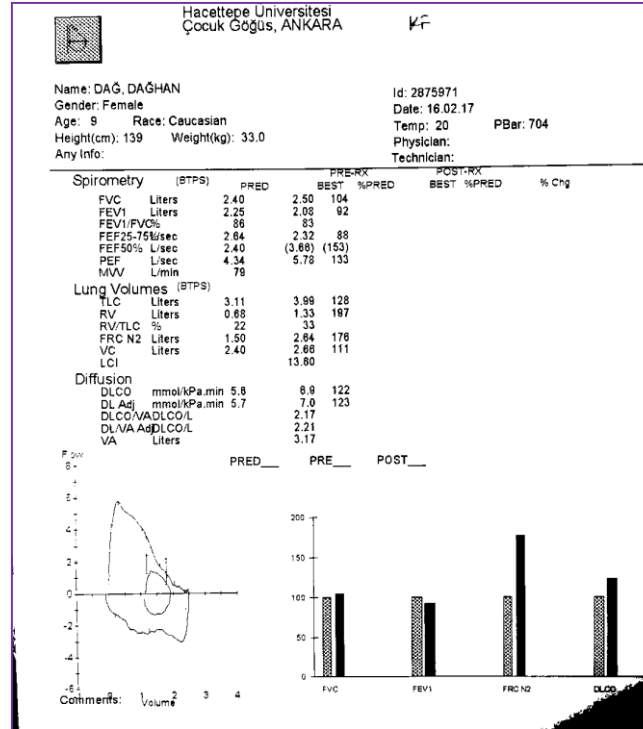
AKCİĞER HACİMLERİ

	PRE	BEKLENEN	% BEKLENEN
VC (L)			111
TLC (L)			128
RV (L)			197
Raw (kPa/L/sn)			126
Gaw (L/sn/kPa/L)			35
RV/TLC (%)			33

DİFFÜZYON

	PRE	BEKLENEN	% BEKLENEN
DLCO (mmolol/kPa.dk)			122
DL Adj (mmolol/kPa.dk)			123

Yorum: Spirometre normal Hipereraasyon. Difüzyon normal



- ▶ FEV1: % 92
- ▶ FVC: %104
- ▶ FEV1/FVC:%83
- ▶ FEF25-75: %88
- ▶ TLC: %128
- ▶ DLCO: %122
- ▶ LCI: 13.6

Lung Clearance Index

- ▶ MBW tekniđi ilk defa 1950'lerde kullanılmaya başlanmıř.
- ▶ 2000'li yıllardan itibaren yenidođan taraması ve yeni tedaviler ile erken dönem hastalık tutulumunu göstermede kullanılmaya başlanmıř.
- ▶ KF hastalarında periferik hava yolu hasarını erken dönem göstermede FEV1'den daha hassastır.
- ▶ İnfantlar ve okul öncesi yař grubunda hafif akciđer hasarını göstermede etkindir.





Journal of Cystic Fibrosis 13 (2014) 123–138



Review

Lung clearance index: Evidence for use in clinical trials in cystic fibrosis

L. Kent ^{a,b}, P. Reix ^c, J.A. Innes ^{d,e}, S. Zielen ^f, M. Le Bourgeois ^g, C. Braggion ^h, S. Lever ⁱ,
H.G.M. Arets ^j, K. Brownlee ^k, J.M. Bradley ^{a,b}, K. Bayfield ^l, K. O'Neill ^m, D. Savi ⁿ, D. Bilton ^o,
A. Lindblad ^p, J.C. Davies ^{l,o}, I. Sermet ^{g,q},
K. De Boeck ^{r,*}, On behalf of the European Cystic Fibrosis Society Clinical Trial Network
(ECFS-CTN) Standardisation Committee

İlk defa 2013'te KF de LCI ölçümü uygun bulunmuş.

Lung Clearance İndex

- ▶ KF de LCI yüksek deęerleri havayollarında bölgesel endoluminal obstrüksiyonu gösterir
- ▶ 1. Sekresyonlar
- ▶ 2. İnlamasyon
- ▶ 3. Remodelling/fibrozis
- ▶ Akut alevlenmelerden sonra normal deęerlere inebilir.





ELSEVIER

Contents lists available at [ScienceDirect](#)

Paediatric Respiratory Reviews



Review

Ventilation heterogeneity and the benefits and challenges of multiple breath washout testing in patients with cystic fibrosis

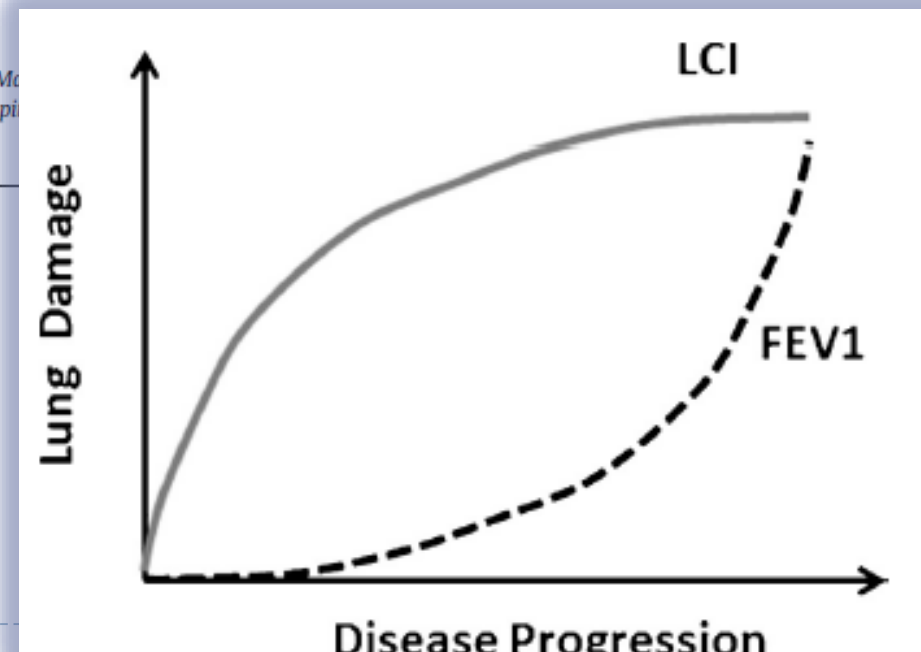


Alex Horsley^{1,2,3,*}, Jim M. Wild³

¹ Respiratory Research Group, Institute of Inflammation and Repair, University of Manchester, UK

² Manchester Adult Cystic Fibrosis Centre, North West Lung Centre, University Hospital of Manchester, UK

³ Academic Radiology, Cardiovascular Sciences, University of Sheffield, UK



WORKSHOP REPORT

Multiple-Breath Washout as a Lung Function Test in Cystic Fibrosis A Cystic Fibrosis Foundation Workshop Report

Padmaja Subbarao¹, Carlos Milla², Paul Aurora³, Jane C. Davies⁴, Stephanie D. Davis⁵, Graham L. Hall⁶, Sonya Heltshe⁷, Philipp Latzin⁸, Anders Lindblad⁹, Jessica E. Pittman¹⁰, Paul D. Robinson¹¹, Margaret Rosenfeld¹², Florian Singer¹³, Tim D. Starner¹⁴, Felix Ratjen¹, and Wayne Morgan¹⁵

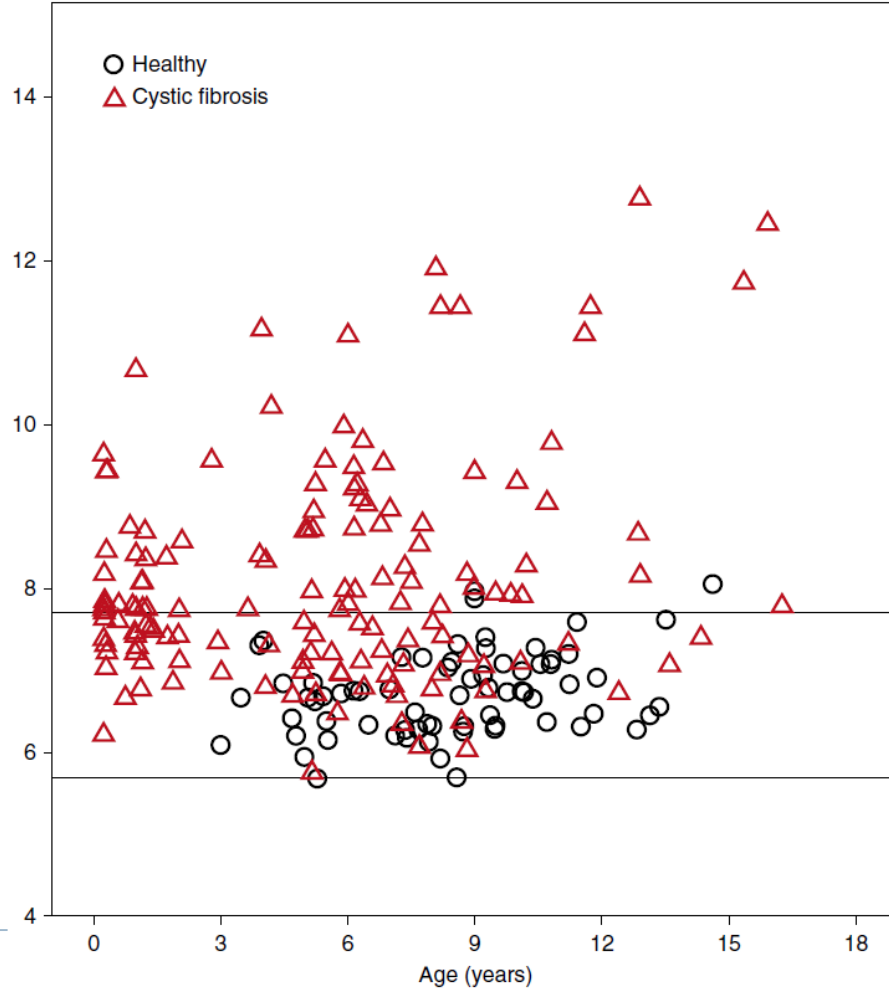
¹Division of Respiratory Medicine, Department of Pediatrics, Physiology, and Experimental Medicine, The Research Institute, Hospital for Sick Children, University of Toronto, Toronto, Ontario, Canada; ²Department of Pediatrics, University of California, San Francisco, San Francisco, California; ³Great Ormond Street Hospital, London, United Kingdom; ⁴Riley Children's Hospital, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana; ⁵Riley Children's Hospital, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana; ⁶University of Washington, Seattle, Washington; ⁷University of Washington, Seattle, Washington; ⁸University of Gothenburg, Gothenburg, Sweden; ⁹University of Gothenburg, Gothenburg, Sweden; ¹⁰University of Washington, Seattle, Washington; ¹¹University of Washington, Seattle, Washington; ¹²University of Washington, Seattle, Washington; ¹³University of Iowa, Iowa City, Iowa; and ¹⁴Arizona State University, Phoenix, Arizona

- ▶ Kesitsel olarak görüntüleme, klinik ve spirometre ile LCI korele
- ▶ İnfant ve okul öncesi dönemde LCI yükseklikleri sonraki yıllarda spirometri değerleri ile korele
- ▶ Akciğer enfeksiyonunda sağlıklı kontrole göre yüksek (AREST CF)
- ▶ P.Aeroginosa izolasyonu ile LCI yüksek

ORIGINAL ARTICLE

Lung Clearance Index and Structural Lung Disease on Computed Tomography in Early Cystic Fibrosis

Kathryn A. Ramsey^{1,2*}, Tim Rosenow^{1,3*}, Lidija Turkovic¹, Billy Skoric^{4,5}, Georgia Banton¹, Anne-Marie Adams^{4,5}, Shannon J. Simpson¹, Conor Murray⁶, Sarath C. Ranganathan^{4,5,7}, Stephen M. Stick^{1,8†}, and Graham L. Hall^{1†};



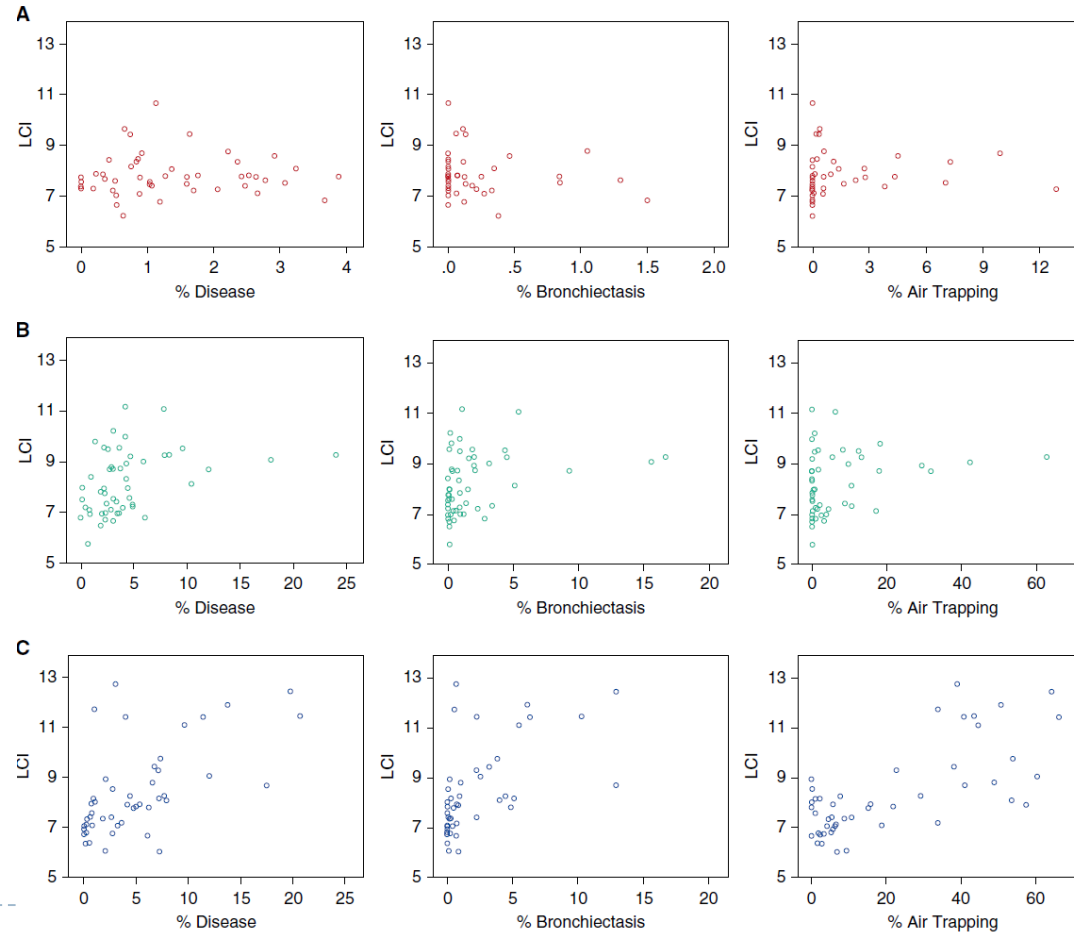
42 infant (3-24 ay), 39 okul öncesi (3-6 yaş), 38 okul çağı (7-16 yaş) KF hastası kontrol grubu ile karşılaştırılmış.

LCI, bütün yaş gruplarında kontrole göre yüksek saptanmış.
(5,7-7,7 normal)

ORIGINAL ARTICLE

Lung Clearance Index and Structural Lung Disease on Computed Tomography in Early Cystic Fibrosis

Kathryn A. Ramsey^{1,2*}, Tim Rosenow^{1,3*}, Lidija Turkovic¹, Billy Skoric^{4,5}, Georgia Banton¹, Anne-Marie Adams^{4,5}, Shannon J. Simpson¹, Conor Murray⁶, Sarath C. Ranganathan^{4,5,7}, Stephen M. Stick^{1,8†}, and Graham L. Hall^{1†}; on behalf of AREST CF[‡]

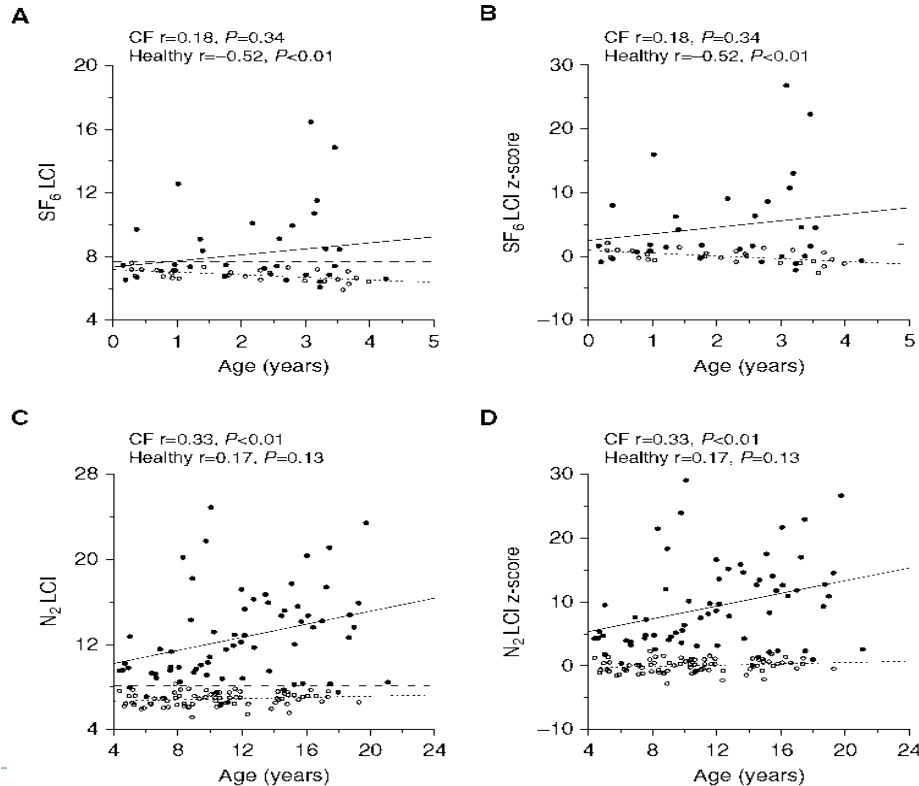


- ▶ İnfantlarda yapısal hasar ile korele değil
- ▶ Okul öncesinde hastalık yaygınlığı ile uyumlu
- ▶ Okul çağında ise hava hapsi ve bronşiektazi ile de korele
- ▶ İnfantil dönem dışında LCI bronşiektazi için hassas ancak nonspesifik

ORIGINAL ARTICLE

Comparison of Lung Clearance Index and Magnetic Resonance Imaging for Assessment of Lung Disease in Children with Cystic Fibrosis

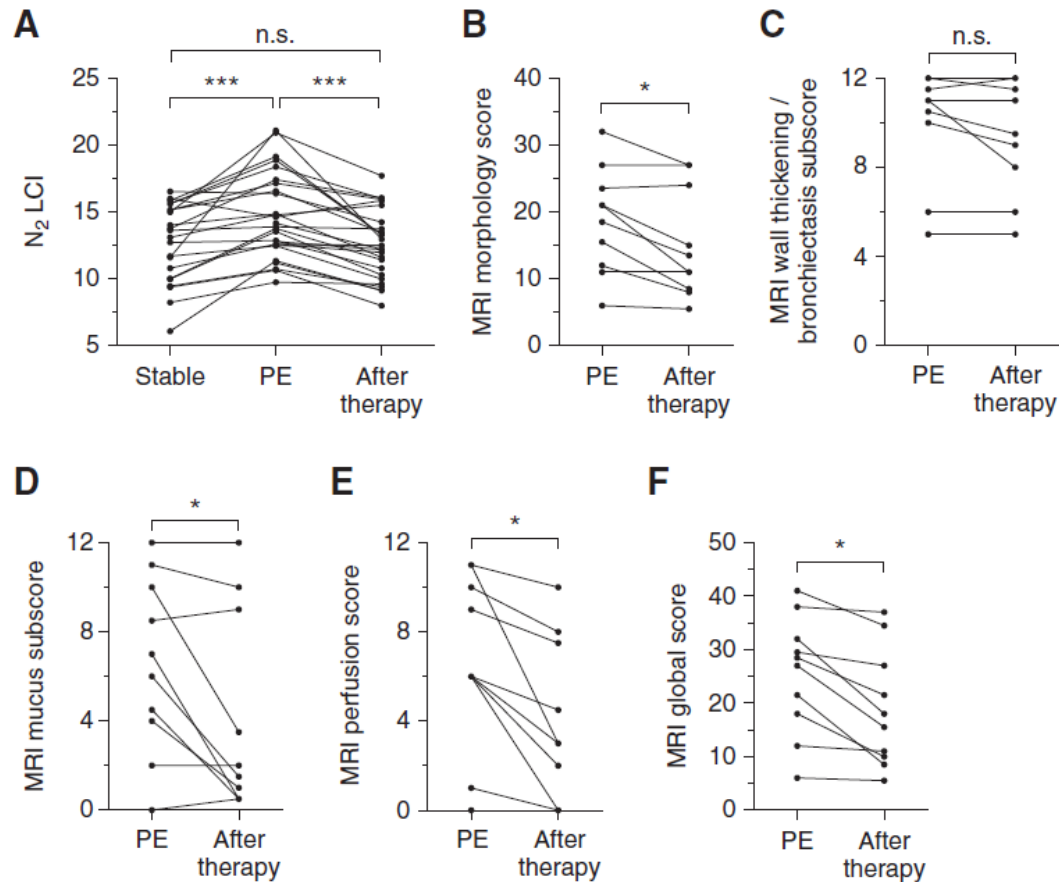
Mirjam Stahl^{1,2,3*}, Mark O. Wielpütz^{3,4,5*}, Simon Y. Graeber^{1,2,3}, Cornelia Joachim^{1,2,3}, Olaf Sommerburg^{1,2,3}, Hans-Ulrich Kauczor^{3,4}, Michael Puderbach^{5,6}, Monika Eichinger^{3,5,6}, and Marcus A. Mall^{1,2,3}



- ▶ KF tanılı , klinik stabil 97 çocuk
- ▶ MRG'da morfoloji, bronşiektazi, mukus birikimi ve perfüzyon için skorlama yapılmış.
- ▶ MRG bulguları ile LCI korele

Comparison of Lung Clearance Index and Magnetic Resonance Imaging for Assessment of Lung Disease in Children with Cystic Fibrosis

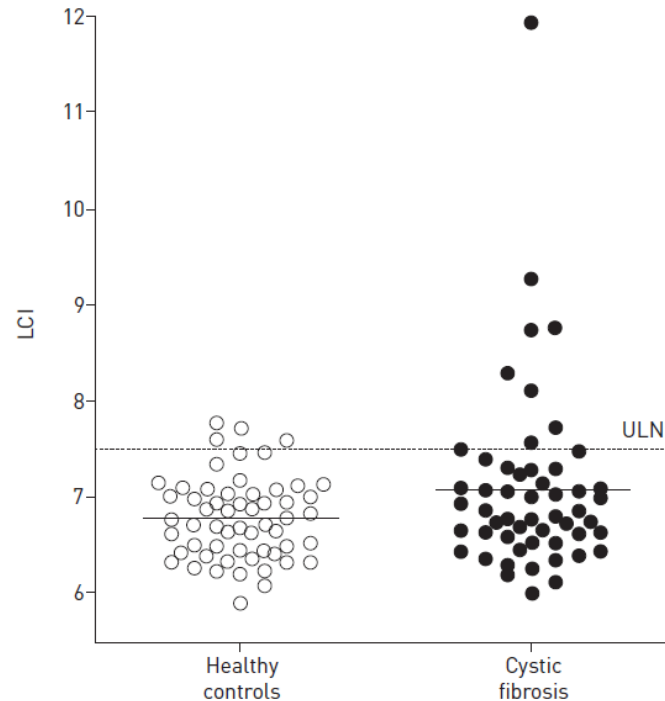
Mirjam Stahl^{1,2,3*}, Mark O. Wielpütz^{3,4,5*}, Simon Y. Graeber^{1,2,3}, Cornelia Joachim^{1,2,3}, Olaf Sommerburg^{1,2,3}, Hans-Ulrich Kauczor^{3,4}, Michael Puderbach^{5,6}, Monika Eichinger^{3,5,6}, and Marcus A. Mall^{1,2,3}



- ▶ 26 hastada akut alevlenme ve iv ab tedavisi sonrasında MRG ve LCI karşılaştırılmış.
- ▶ Tedavi yanıtını değerlendirme de korele

Elevated lung clearance index in infants with cystic fibrosis shortly after birth

Elisabeth Kieninger^{1,10}, Sophie Yammine^{1,10}, Insa Korten^{1,2},
Pinelopi Anagnostopoulou¹, Florian Singer^{1,3}, Urs Frey², Anne Mornand⁴,



- ▶ Yenidoğan taraması ile tanı alan 53 infant (ort. 7,8 haftalık) 57 kontrol ile prospektif olarak karşılaştırılmış.
- ▶ Uykuda, MBV TV ile yapılmış.
- ▶ KF hastalarında LCI yüksek saptanmış.
- ▶ LCI, erken dönem hafif tutulumda ventilasyon inhomojenitesini göstermekte.

Original Article

Respiratory rate in infants with cystic fibrosis throughout the first year of life and association with lung clearance index measured shortly after birth

Insa Korten ^{a,b,c}, Elisabeth Kieninger ^a, Sophie Yammine ^a, Giulia Cangiano ^d, Sylvia Nyilas ^{a,b}, ^e, ^f, ^g, ^h, ⁱ, ^j, ^k, ^l, ^m, ⁿ, ^o, ^p, ^q, ^r, ^s, ^t, ^u, ^v, ^w, ^x, ^y, ^z, ^{aa}, ^{ab}, ^{ac}, ^{ad}, ^{ae}, ^{af}, ^{ag}, ^{ah}, ^{ai}, ^{aj}, ^{ak}, ^{al}, ^{am}, ^{an}, ^{ao}, ^{ap}, ^{aq}, ^{ar}, ^{as}, ^{at}, ^{au}, ^{av}, ^{aw}, ^{ax}, ^{ay}, ^{az}, ^{ba}, ^{bb}, ^{bc}, ^{bd}, ^{be}, ^{bf}, ^{bg}, ^{bh}, ^{bi}, ^{bj}, ^{bk}, ^{bl}, ^{bm}, ^{bn}, ^{bo}, ^{bp}, ^{bq}, ^{br}, ^{bs}, ^{bt}, ^{bu}, ^{bv}, ^{bw}, ^{bx}, ^{by}, ^{bz}, ^{ca}, ^{cb}, ^{cc}, ^{cd}, ^{ce}, ^{cf}, ^{cg}, ^{ch}, ^{ci}, ^{cj}, ^{ck}, ^{cl}, ^{cm}, ^{cn}, ^{co}, ^{cp}, ^{cq}, ^{cr}, ^{cs}, ^{ct}, ^{cu}, ^{cv}, ^{cw}, ^{cx}, ^{cy}, ^{cz}, ^{da}, ^{db}, ^{dc}, ^{dd}, ^{de}, ^{df}, ^{dg}, ^{dh}, ^{di}, ^{dj}, ^{dk}, ^{dl}, ^{dm}, ^{dn}, ^{do}, ^{dp}, ^{dq}, ^{dr}, ^{ds}, ^{dt}, ^{du}, ^{dv}, ^{dw}, ^{dx}, ^{dy}, ^{dz}, ^{ea}, ^{eb}, ^{ec}, ^{ed}, ^{ee}, ^{ef}, ^{eg}, ^{eh}, ^{ei}, ^{ej}, ^{ek}, 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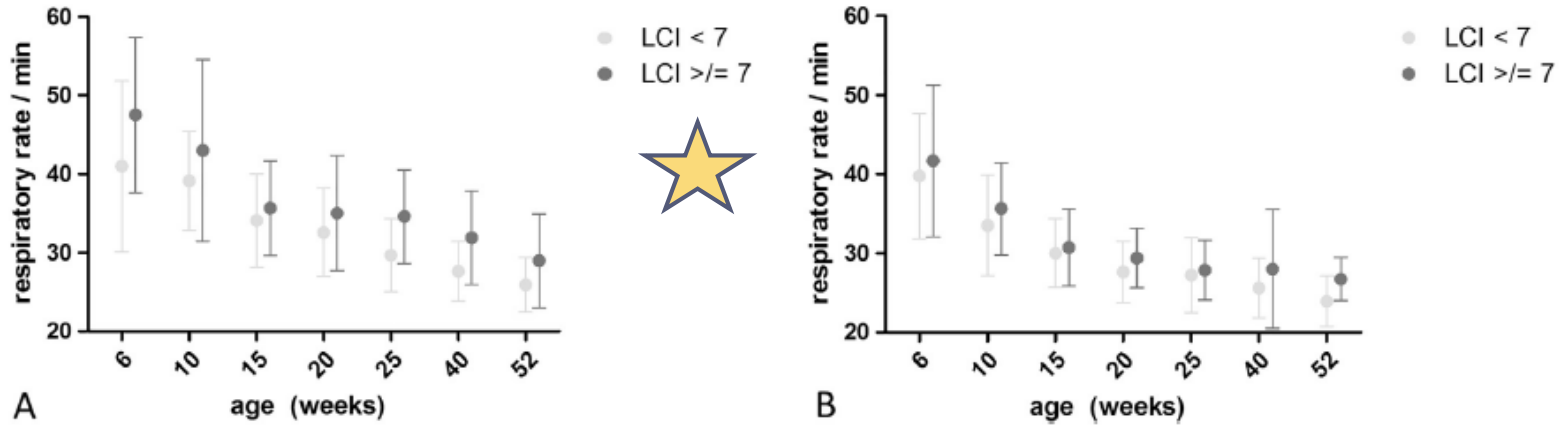


Fig. 2. Association of respiratory rate with lung clearance index. Respiratory rate measurements throughout the first year of life in (A) infants with CF and (B) healthy infants. RR measure

İnfanlarda LCI ölü boşluk ve solunum hızından çok etkilenebilir.

43 KF ve kontrol grubunda 50 hafta boyunca solunum hızı ile takip edilmiş.

İlk hafta ve enfeksiyonlar esnasında LCI ölçülmüş.

KF de solunum hızı ve LCI yüksek

