Proceso diagnóstico de enfermedad quística pulmonar

Hallazgos de PFR

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Introducción

- Patogénesis de los quistes
- Papel de las PFR en el diagnóstico de las enfermedades quísticas difusas
- PFR en el diagnóstico diferencial con otras entidades
- Las PFR en el seguimiento de los pacientes
- Correlación de las PFR con las pruebas de imagen
- PFR en las 4 principales
  - Histiocitosis
  - LAM
  - NIL
  - BHD
- Otras consideraciones
- Conclusiones

Patogénesis de los quistes

- 1 obstrucción valvular
  - Inflación distal vía aérea

  Bronquiolitis folicular
  Metástasis
  Neumatoceles
  LAM
  Histiocitosis

- 2 isquemia
  - Infiltración y obstrucción vascular bronquiolos terminales

  Amiloidosis
  Histiocitosis

- 3 Degradación tejido conectivo y remodelado por MMPs

  LAM
  Histiocitosis
  Depósito cadenas ligeras

RESPIRATORY CARE • JANUARY 2020 VOL 65 NO 1
AmericanJournalofRespiratoryandCriticalCareMedicineVolume192Number1|July12015
Breathe | June 2020 | Volume 16 | No 2

Papel de las PFR en el diagnóstico de las enfermedades quísticas difusas

- Las PFR en los algoritmos diagnósticos

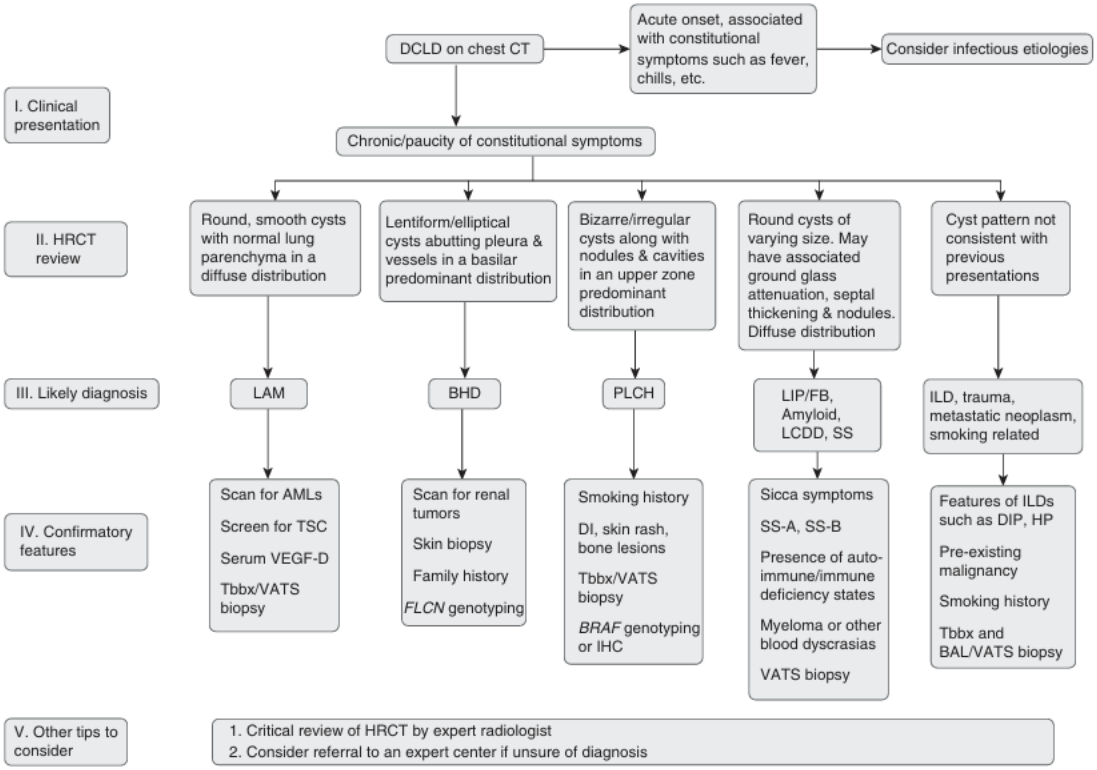

Figure 4. Algorithm to guide approach to the diagnosis of diffuse cystic lung diseases. AML = angiomyolipoma; BAL = bronchioalveolar lavage; BHD = Birt-Hogg-Dubé syndrome; *BRAF* = v-Raf murine sarcoma viral oncogene homolog B; CT = computed tomography; DCLD = diffuse cystic lung disease; DI = diabetes insipidus; DIP = desquamative interstitial pneumonia; FB = follicular bronchiolitis; *FLCN* = folliculin; HRCT = high-resolution computed tomography; HP = hypersensitivity pneumonitis; IHC = immunohistochemistry; ILD = interstitial lung disease; LAM = lymphangioleiomyomatosis; LCDD = light-chain deposition disease; LIP = lymphoid interstitial pneumonia; PLCH = pulmonary Langerhans cell histiocytosis; SS = Sjögren syndrome; Tbbx = transbronchial biopsy; TSC = tuberous sclerosis complex; VATS = video-assisted thoracoscopic surgery; VEGF-D = vascular endothelial growth factor-D.

American Journal of Respiratory and Critical Care Medicine Volume 192 Number 1 | July 1 2015

Table 4. Summary of Clinical and Diagnostic Features of Selected Diffuse Cystic Lung Diseases

	LAM	PLCH	BHD	LIP/FB	Amyloid	LCDD
Personal history	Pneumothorax, angiomyolipomas, chylous effusions, and cortical tubers, seizures, skin lesions if TSC	Pneumothorax, smoking	Pneumothorax, skin lesions, renal tumors	HIV, autoimmune diseases, sicca symptoms, Raynaud's phenomenon	Sicca symptoms, autoimmune diseases	Lymphoproliferative disorders
Family history	TSC	Not relevant	Pneumothoraces, skin lesions, renal cancers	Not relevant	Not relevant	Not relevant
Extrapulmonary manifestations & other associations	Renal angiomyolipomas, chylous effusions, TSC manifestations	Diabetes insipidus, cutaneous & osteolytic bone lesions	Renal tumors, skin fibrofolliculomas	SS & other CTDs, HIV, EBV, CVID	SS & other CTDs, systemic amyloidosis	Lymphoproliferative disorders, renal failure
Laboratory testing	Serum VEGF-D	Serum & urine studies for diabetes insipidus	Genetic testing for *FLCN* mutations	Polyclonal dysproteinemia	Monoclonal dysproteinemia	Lymphoproliferative disorders, renal failure
Diagnostic yield of bronchoscopy (BAL, TBBx)	>50%	30–50%	0	Low yield	Low yield	Low yield
Consider surgical lung biopsy	Yes	Yes	No	Yes	Yes	Yes
Genetic testing	TSC mutations, but usually not clinically indicated	BRAF mutation	*FLCN* gene mutation	No	No	No
Treatment	Sirolimus	Smoking cessation, immunosuppression, cladribine	None available	Corticosteroids & other immunosuppressive agents for LIP	None available	None available

Definition of abbreviations: BAL = bronchioalveolar lavage; BHD = Birt-Hogg-Dubé syndrome; BRAF = v-Raf murine sarcoma viral oncogene homolog B; CTD = connective tissue disease; CVID = common variable immune deficiency; EBV = Epstein-Barr virus; FB = follicular bronchiolitis; *FLCN* = folliculin; LAM = lymphangioleiomyomatosis; LCDD = light-chain deposition disease; LIP = lymphoid interstitial pneumonia; PLCH = pulmonary Langerhans cell histiocytosis; SS = Sjögren syndrome; TBBx = transbronchial biopsy; TSC = tuberous sclerosis complex; VEGF-D = vascular endothelial growth factor-D.
Data from References 18, 19, 45, 63, 69, and 97–104.

Table 3. Summary of Clinical, Radiographic, and Pathologic Features of Common DCLDs

	LAM	PLCH	BHD	LIP/FB
Cyst characteristics				
Distribution	Diffuse, random	Upper lung zone, spares costophrenic angles	Basilar, subpleural	Diffuse, random
Size	2 mm to 2 cm	2 mm to > 2 cm	Usually < 1 cm	3 to 1 cm
Shape	Round, uniform	Bizarre, irregular	Elliptical, lentiform	Round, variable, may contain internal structure
Pathology	Cyst walls containing HMB-45+ LAM cells with smooth muscle phenotype	Cystically dilated airways associated with aggregates of S100+ and CD1a+ Langerhans cells	Intraparenchymal and subpleural cysts abutting interlobular septae and lacking neoplastic or significant inflammation	LIP: diffuse interstitial lymphocytic infiltrate FB: peribronchiolar follicular lymphoid infiltrate with germinal centers
Inheritance pattern	Autosomal dominant (TSC-LAM) or sporadic	Not heritable	Autosomal dominant	Not heritable
Genetic mutation	*TSC*	*BRAF*, *MAP2K1*, other mutations in the MAP kinase pathway	*FLCN*	N/A
Gender tendency	Women >> men	Women = men	Women = men	Women > men
Clinical features	Dyspnea on exertion, fatigue, spontaneous pneumothorax	Pneumothorax, diabetes insipidus, skin and osteolytic lesions	Pneumothorax, skin fibrofolliculomas, renal tumors	Sicca symptoms, arthralgias, skin rash, Raynaud symptoms
Other radiologic findings	Pleural effusions, AMLs, lymphadenopathy, lymphangioleiomyomas. MMPH in patients with TSC-LAM	Lung nodules with or without cavitation, lytic bone lesions	Renal tumors, often multiple and bilateral	Ground-glass attenuation, lung nodules
Diagnostic yield of bronchoscopy with TBBx	> 50%	30–50%	0	Low yield
Treatment	Sirolimus/everolimus	Smoking cessation, cladribine, MAPK/BRAF inhibitors	None available	Immunosuppression

Data from References 1, 31, 37, 47, 48, 51, 66, 70, 72, 76, 86, 87, 94, 98, 99, 102, 117, 118.
DCLD = diffuse cystic lung diseases
LAM = lymphangioleiomyomatosis
PLCH = pulmonary Langerhans cell histiocytosis
BHD = Birt-Hogg-Dubé syndrome
LIP = lymphoid interstitial pneumonia
FB = follicular bronchiolitis
HMB-45 = human melanoma black-45
TSC = tuberous sclerosis complex
BRAF = v-Raf murine sarcoma viral oncogene homolog B
MAP2K1 = mitogen-activated protein kinase 1
MAP = mitogen-activated protein
FLCN = folliculin
N/A = not applicable
AML = angiomyolipoma
MMPH = multifocal micronodular pneumocyte hyperplasia
TBBx = transbronchial biopsy

Table 2 *Characteristics of different diffuse cystic lung diseases (Continued)*

	LAM	PLCH	BHD	Lymphocytic interstitial pneumonia	Amyloidosis	Light chain deposition disease	Infection
Extrapulmonary manifestations	Renal angiomyolipomas Chylous effusions (pleural or ascites) Lymphangioleiomyomas TSC: skin lesions, seizures, hamartomatous lesions in various organs	Adult PLCH is generally isolated Rarely: bones (lytic lesions), pituitary (diabetes insipidus), skin rash	Skin fibrofolliculomas Renal tumours	Autoimmune diseases and associated symptoms including sicca symptoms and Raynaud's	Renal, cardiac, gastrointestinal, neurological and skin manifestations	Renal failure Lymphoproliferative disorders	PJP: immunosuppression, constitutional features RRP: upper airway involvement
Supporting investigations	VEGF-D >800 pg·mL^{-1} Tbbx or VATS biopsy	*BRAF* mutation testing Tbbx or VATS biopsy	Genetic testing for *FLCN* mutations Imaging for renal tumours Skin biopsy	Autoimmune panel HIV testing VATS biopsy	Presence of monoclonal protein VATS biopsy	Investigation for lymphoproliferative disorder Renal biopsy VATS biopsy	HIV status, CD4^{+} cell count PCR of induced sputum, BAL fluid or nasopharyngeal aspirates
Treatment	mTOR inhibition: everolimus, sirolimus	Smoking cessation	Nil	Corticosteroids and immunosuppression	Treatment of underlying disease	Treatment of underlying disease	PJP: trimethoprim-sulfamethoxazole RRP: cidofovir

PLCH: pulmonary Langerhans cell histiocytosis; BHD: Birt–Hogg–Dubé syndrome; Hx: history; *FLCN*: folliculin; PJP: *Pneumocystis jirovecii* pneumonia; RRP: recurrent respiratory papillomatosis; HPV: human papilloma virus; FHx: family history; VEGF: vascular endothelial growth factor; Tbbx: transbronchial biopsy; VATS: video-assisted thoracoscopic surgery; BAL: bronchoalveolar lavage; mTOR: mammalian target of rapamycin.

Papel de las PFR en el diagnóstico

- En general no son diagnósticas
- PFR completas útiles en el momento del diagnóstico (pero no específicas)
- Espirometría normal no excluye enfermedad
- Mejor con pletismografía
  - detectar aumento de volúmenes por atrapamiento aéreo
- Patrón obstructivo más común
  - a veces con sorprendente respuesta broncodilatadora
- Patrón mixto a veces con neumotórax y pleurodesis.

PFR en el diagnóstico diferencial

Table 1. Fleischner Society Definitions of Air-Space Lucencies on CT Scan

Lesion	Definition
Cysts	Thin-walled (< 2 mm wall thickness), spherical, air-filled lucencies interfaced with normal lung
Cavities	Irregular, thick-walled, air-filled structures within lung mass, consolidation, or nodule
Bullae	Thin-walled focal lucencies that are usually > 1 cm in diameter and are typically associated with emphysematous changes
Blebs	Thin-walled, air-filled structures that are usually < 1 cm and are typically adjacent to the visceral pleura
Pneumatoceles	Round, air-filled structures that are surrounded by a thin wall and are usually caused by infections, aspiration, or trauma

Data from Reference 2.
CT = computed tomography

Table 2. Classification of DCLDs

Figure 7 – *The mimics of cystic lung disease along with their radiographic and clinical features. PFT = pulmonary function test.*

PFR en el diagnóstico diferencial

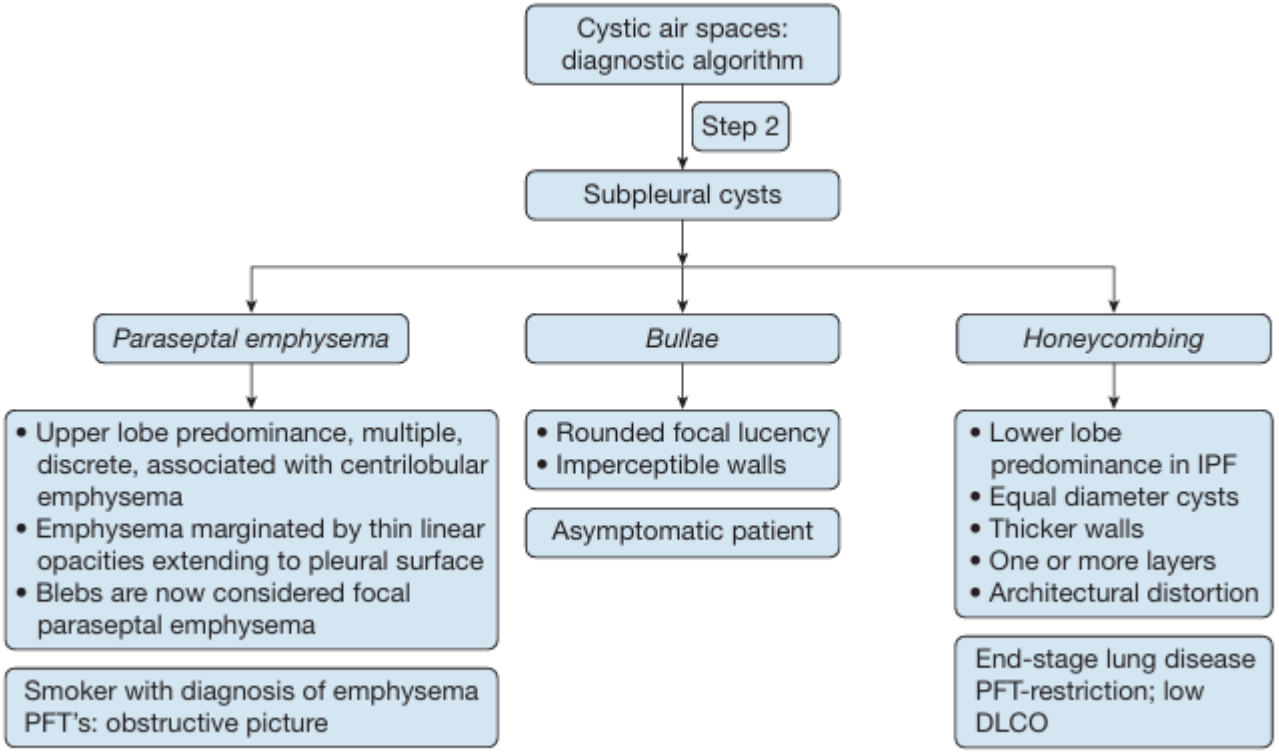

Figure 11 – *The major etiologies of cysts that are present predominantly in the subpleural areas. Of note, Birt-Hogg-Dubé (BHD) syndrome may also give rise to cysts in the subpleural area. See Figure 7 legend for expansion of abbreviation.*

CHEST 2016; 150(4):945-965

Papel las PFR en el seguimiento de las enfermedades quísticas difusas

Table 6. Respiratory Care Implications for DCLD Patients

Topic	
Supplemental oxygen	
Pulmonary rehab	
Positive pressure ventilation	
PFTs	

Data from References 125–128.
6MWT = 6-min walk test
DCLD = diffuse cystic lung diseases
LAM = lymphangioleiomyomatosis
PFT = pulmonary function test
RT = respiratory therapist
BD = bronchodilator response

Table 4. Pneumothorax in DCLDs

	LAM	PLCH	BHD
Prevalence of spontaneous pneumothorax	55–73	10–20	25–75
Ipsilateral pneumothorax recurrence rate	73	56	73
Contralateral pneumothorax occurrence rate	74	29	48
Ipsilateral recurrence rate after conservative management	66	58	63
Ipsilateral recurrence rate after chemical pleurodesis	27	Unknown	30
Ipsilateral recurrence rate after surgical pleurodesis	32	0–20	35

Data are presented as %. Data from References 26, 34, 54, 79, 85, 92, 119.
DCLD = diffuse cystic lung diseases
LAM = lymphangioleiomyomatosis
PLCH = pulmonary Langerhans cell histiocytosis
BHD = Birt-Hogg-Dubé syndrome

nts pulmonary
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following PFT.
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CLD patients

Correlación imagen - PFR en las enf quísticas difusas

Objetivo: correlacionar score cuantitativo de lesiones por HRCT density mask (DM) software y datos PFR.

Suma y media de los % de atrapamiento a 3 niveles del estudio DM (*arco Ao*, origen B LII y 2 cm del diafragma) vs. parámetros PFR y 6MM

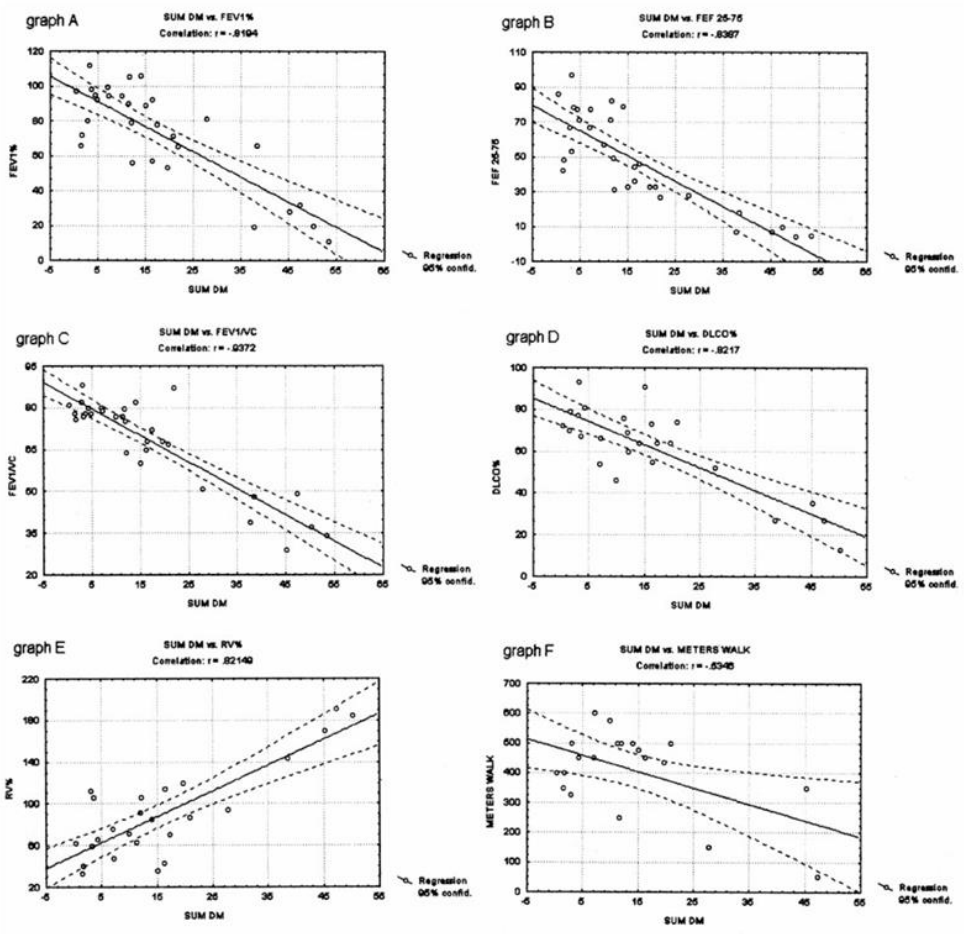


FIGURE 3. Scatterplot graphs of the correlation between the sum of DM values and important functional and exercise parameters. *Top left*, A: FEV_1. *Top right*, *B*: FEF_{25-75}. *Middle left*, *C*: FEV_1/VC. *Middle right*, *D*: DLCO. *Bottom left*, *E*: RV. *Bottom right*, *F*: meters walked (MW).

Resultados estudio cuantitativo se correlacionan con:

- FVC ($r = -0.56$; $p < 0.001$),
- FEV_1/VC ($r = -0.94$; $p < 0.002$),
- mesoespiratorios ($r = -0.84$; $p < 0.05$),
- FEV_1 ($r = -0.82$; $p < 0.05$),
- DLCO ($r = -0.82$; $p < 0.05$),
- sRaw ($r = 0.79$; $p < 0.05$)
- 6-MWT ($r = -0.53$; $p < 0.05$).

- No con gasometría.

PFR en Histiocitosis de células de Langerhans

- PFR normales/obstructivas/restrictivas/mixtas
- Normal/restrictiva en fases tempranas
  - 20% normal al momento del diagnóstico
- 70%-90% reducción DLCO: lo más frecuente
  - Algunos ↓DLCO y desaturación esfuerzo (afectación vascular)
  - Limitación al esfuerzo inicial
- ↓ VC, TLC y ↑VR/TLC es un perfil común
- Obstrucción en fases más avanzadas
  - Desproporcionada para el grado de tabaquismo (bronquiolar)
  - Obstrucción + atrapamiento + ↓DLCO + hipoxemia en enfermedad avanzada con extensa afectación quística.
  - Limitación al esfuerzo en fases más avanzadas
- PFR similares en pacientes con y sin neumotórax
- No hay correlación PFR con HP (vasculopatía primaria)

Suri et al. Orphanet Journal of Rare Diseases 2012, 7:16
Eur Respir J 2006; 27: 1272–1285

Limitación capacidad ejercicio en Histiocitosis

- Mecanismos de limitación ejercicio poco estudiados
- Estudio 62 pacientes con PECP
  - V'O2 pico no relacionado con disnea, ni Borg ni mMRC
  - V'O2 pico correlación independiente con 4 componentes:
    - PC1= alteración intercambio: DLCO, DLCO/VA, DA-aO2
    - PC2= volúmenes pulmonares: FEV1, FVC ↓
    - PC3= equivalentes ventilatorios
    - PC4= atrapamiento aéreo: RV, RV/TLC ↑
  - Disminución capacidad por alteración multifactorial respiratoria (orden):
    - Alteración del intercambio gaseoso
    - Atrapamiento aéreo
    - Pseudorrestricción
    - Hiperventilación inducida por ejercicio

PLOSONE|DOI:10.1371/journal.pone.0170035 January10, 2017

PFR en Histiocitosis de células de Langerhans

- Gasometría
  - A-aO2 al e
- PFR cada 3-
- Implicación
  - Si declina
  - Se ha repe
- Pronóstico
  - Difícil pre
  - Fisiología
  - DLCO grav
  - HP

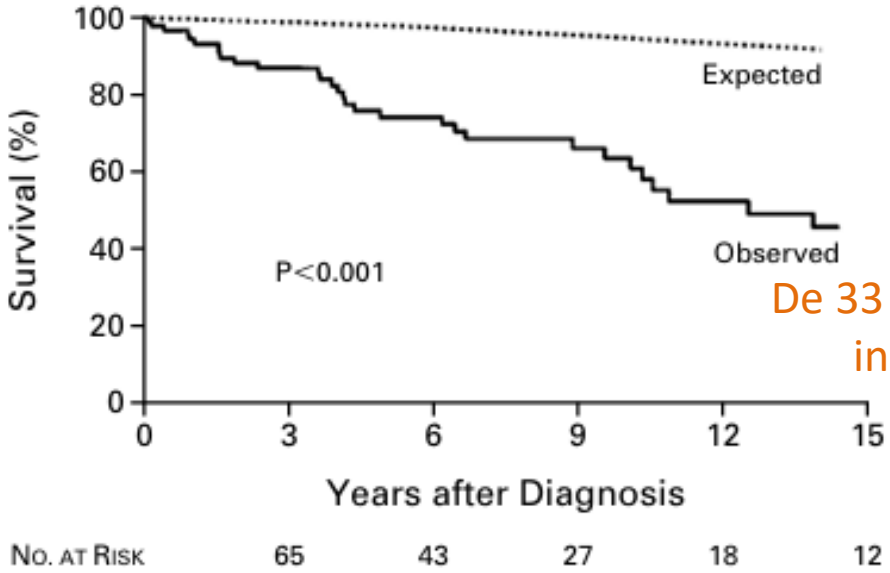


Figure 1. Kaplan–Meier Analysis of Expected and Observed Survival among 102 Adults with Pulmonary Langerhans'-Cell Histiocytosis.

The expected survival was defined as that for age- and sex-matched members of the general U.S. population.

De 33 pacientes 15 murieron por insuficiencia respiratoria

American Journal of Respiratory and Critical Care Medicine Volume 191 Number 12 | June 15 2015
ESPIRATORY CARE • JANUARY 2020 VOL 65 NO 1
N Engl J Med, Vol. 346, No. 7 · February 14, 2002

PFR en LAM

- Curso progresivo → 10 años:
  - 55% disnea
  - 20% requiere OCD
  - 10% éxitus
- Alteración DLCO en fases tempranas
- DLCO y FEV1 correlaciona con CT y con histología
- Obstrucción e hiperinsuflación lo más frecuente
- FEV1 declina 50-250ml/año
  - Más rápido S-LAM
  - Más si elevado D-VEGF
  - Más premenopáusicas, embarazo, estrógenos
  - PFR iniciales factor pronóstico → predice progresión
- MILES trial
  - Sirolimus estabililiza función pulmonar

RESPIRATORY CARE • JANUARY 2020 VOL 65 NO 1
American Journal of Respiratory and Critical Care Medicine Volume 191 Number 12 | June 15 2015

PFR LAM

- Recomendaciones ERS
  - Espirometría con test bd, DLCO y TLC en evaluación inicial
    - Respuesta bd muy significativa peor pronóstico*
  - Seguimiento
    - FEV y DLCO cada 3-6 meses si progresión
    - 6-12 meses si estable

- Gasometría arterial
  - Hipoxemia frecuente
  - Para definir necesidad OCD, excluir hipercapnia…
  - Realizar en evaluación inicial y para remitir a trasplante

- PEPC → Disminución V'O2max
  - Info adicional para discapacidad/progresión/respuesta a tto

Eur Respir J 2010; 35: 14–26
**Breathe | June 2020 | Volume 16 | No 2*

Función pulmonar, embarazo y LAM

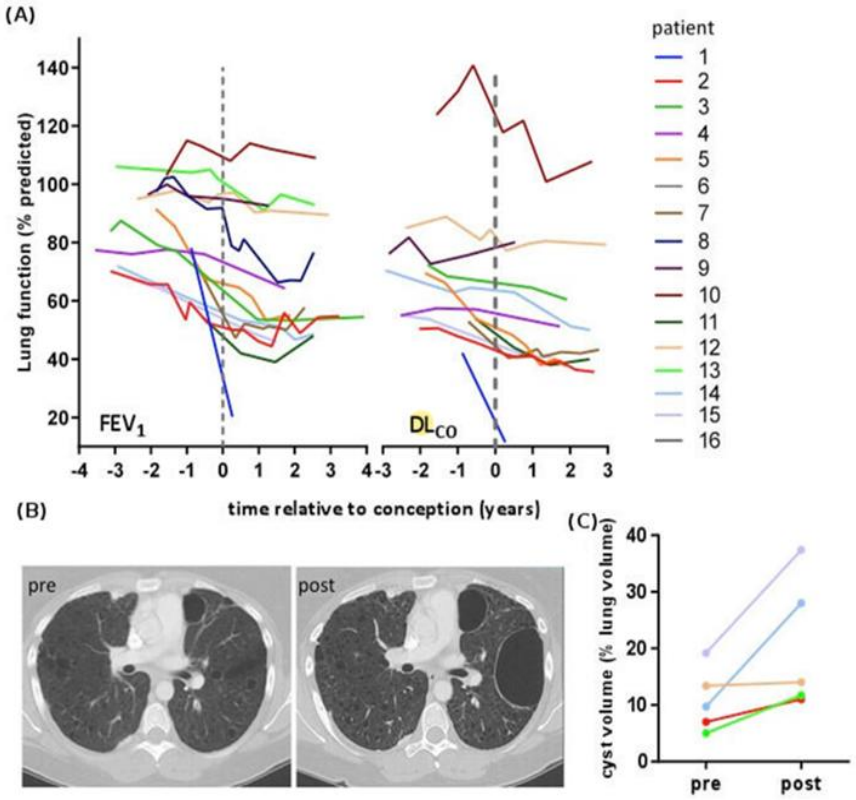


Figure 1 (A) Lung function trajectory in individual subjects before and after pregnancy for FEV_1 and DL_{CO}. The dashed line marks the start of pregnancy. Patient numbers correspond with tables 1 and 2 and (C). (B) CT scans of a LAM patient obtained 6 months before pregnancy (pre) and 6 months after delivery (post). Considerable progression of the cystic lung destruction was observed evidenced from an increase in cyst score from 9.8% to 28.8%. (C) Quantification of lung cyst scores as percentage of lung volume occupied by cysts for five subjects pre and post pregnancy. Two tail paired t-test p=0.06. FEV_1, forced expiratory volume in 1 s.

- FEV1
  - Pre 77% → Post 64%
- DLCO
  - Pre 66% → Post 57%
- Acelera ritmo pérdida
  - Aumenta tasa caída 50%
- Aumenta scores quistes

Taveira-DaSilva AM, et al. Thorax 2020;75:904–907. doi:10.1136/thoraxjnl-2020-214987

PFR en Birt-Hogg-Dubé

- Función pulmonar normal/ligera obstrucción
- DLCO/VA ligeramente disminuida
- FEV1 correlaciona inversamente con área quística medida por TC
  - Evidencia de progresión de quistes que se relaciona con ↓ FEV1, FEV1/FVC y CV ($p<0,0001$)*
  - *"stretch mechanism"*
- No progresión a insuficiencia respiratoria

Table 1 Proposed diagnostic criteria for BHD [2].

Definite pulmonary BHD

1. Characteristic[a] or compatible[b] lung HRCT and skin biopsy positive for fibrofolliculoma or trichodiscoma
2. Characteristic or compatible lung HRCT and confirmed family history of BHD in first or second degree family member
3. Characteristic or compatible HRCT and tissue confirmation of renal chromophobe adenoma or oncocytoma
4. Characteristic or compatible HRCT and tissue confirmation of genetic testing positive for BHD

Probable pulmonary BHD

1. Characteristic HRCT, exclusion of TSC and LAM, and personal or family history of pneumothorax
2. Compatible HRCT, exclusion of TSC and LAM, and any of the following:
   a. Family or personal history of renal tumors
   b. Skin angiofibroma
   c. Renal angiomyolipoma

Possible pulmonary BHD

Compatible or characteristic HRCT.

[a] Characteristic lung HRCT findings: Multiple thin-walled round, elliptical or lentiform well-defined air-filled cysts, without internal structure, in a basilar, medial and subpleural predominant distribution, with preserved or increased lung volume, and no other significant pulmonary involvement (specifically no interstitial lung disease).

[b] Compatible HRCT findings: Thin walled cysts without the more typical elliptical shape or subpleural distribution.

Cho et al. BMC Pulmonary Medicine 11
Breathe | June 2020 | Volume 16 | No 2

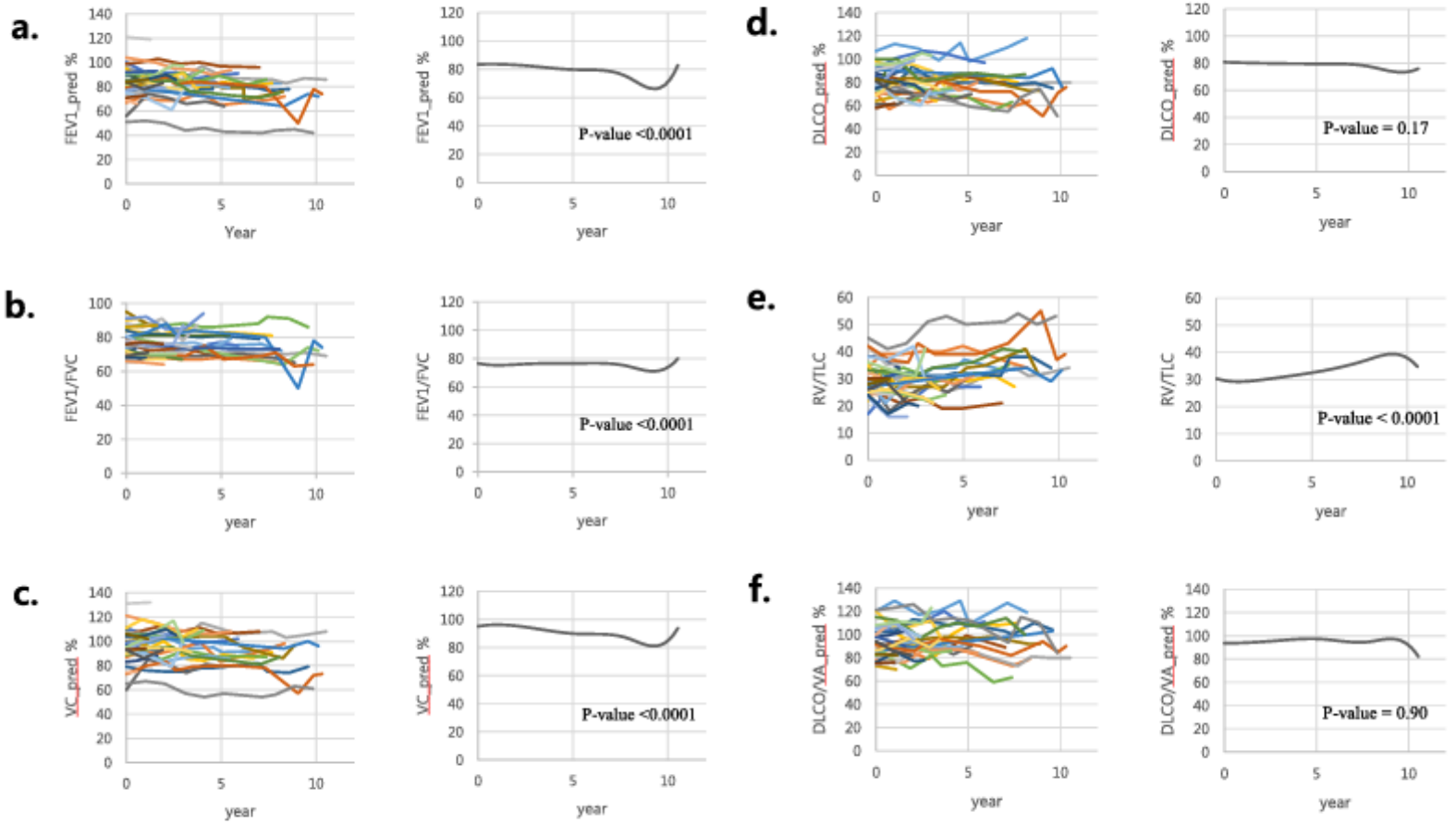


Fig. 3 Results of the evolution of the PFT. Evolution of the parameters of the PFT during serial follow up in patients with BHD syndrome, left: raw data, right: linear mixed model, **a**) FEV1, **b**) FEV1/FVC, **c**) VC, **d**) DLCO, **e**) RV/TLC, and **f**) DLCO/VA. FEV1_pred %, FEV1/FVC, and VC pred_% among the PFT parameters showed a statistically significant decreasing trend with time ($p < 0.0001$ for each) and RV/TLC values showed a statistically significant increasing trend over time ($p < 0.0001$). DLCO and DLCO/VA values showed no significant change over time. FEV1 forced expiratory volume in one second, FVC forced vital capacity, TLC total lung capacity, RV residual volume, DLco carbon monoxide transfer factor, DLco/VA carbon monoxide transfer coefficient, %pred percentage of predicted value capacity

PFR en NIL

- Trastorno restrictivo
- Disminución DLCO
- Obstrucción concomitante
  - Compresión bronquiolar y estrechamiento por el proceso infiltrativo
  - Bronquiolitis folicular
- Función pulmonar normal
- Gasometría arterial normal
- No hay datos sobre el efecto del tratamiento

Clin Chest Med 37 (2016) 463–474

Otras consideraciones

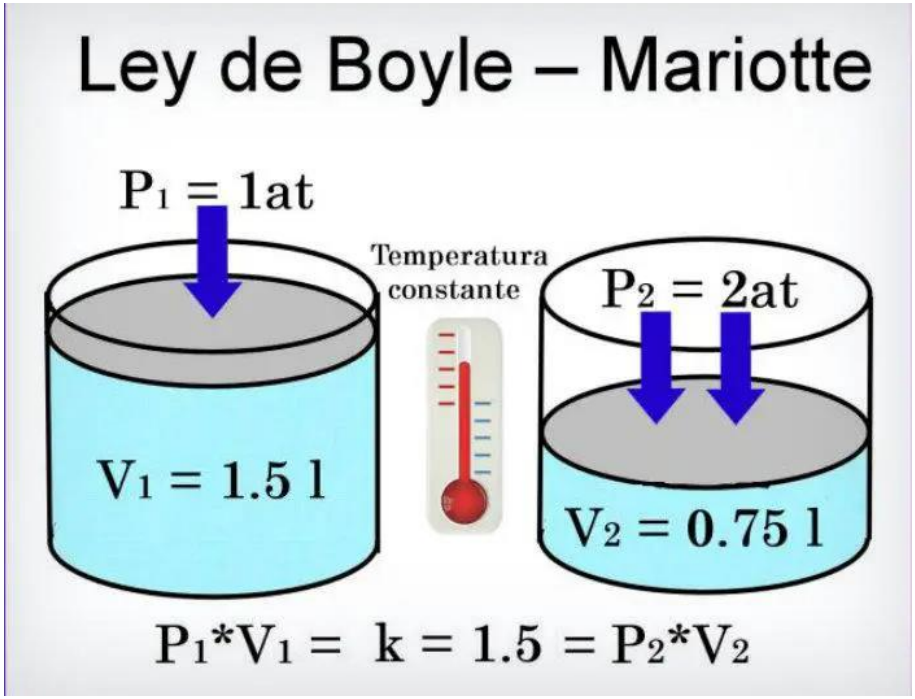
Table 5. General Recommendations Applicable to All Patients With DCLDs

1. Counsel to avoid smoking.
2. Stay up to date on vaccination including annual influenza vaccination and both pneumococcal (PPSV23 and PCV13) vaccines.
3. Air travel is safe for most patients with DCLDs. The risk of in-flight pneumothorax is approximately 1 per 100 flights. Patients should be educated about the typical symptoms of pneumothorax and instructed to seek medical attention if they have new-onset symptoms suggestive of a pneumothorax.
4. Advise against scuba diving due to the potential risk of spontaneous pneumothorax.
5. Patients presenting with spontaneous pneumothorax should undergo pleurodesis following the first episode of pneumothorax. Prior pleurodesis is not a contraindication for lung transplantation.

Data from References 120–124.
DCLD = diffuse cystic lung diseases

Otras consideraciones

- **Ley de Boyle-Mariotte** describe cómo cambia el volumen de un gas cuando se altera su presión, manteniendo constante su temperatura.



Otras consideraciones

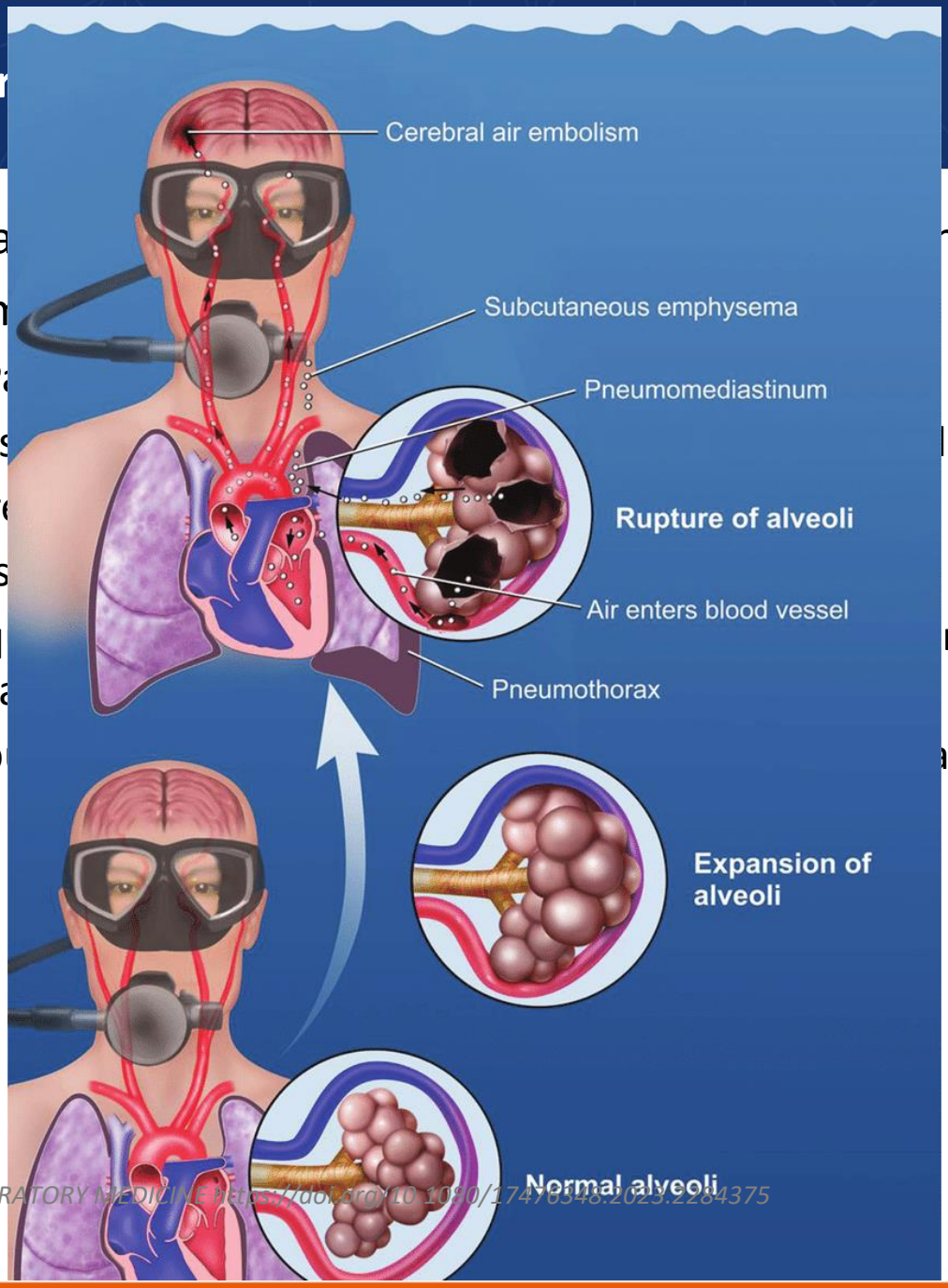
- Buceo y enfermedad quística
  - Reciente rápido aumento de interés por el buceo: 1 millón de certificaciones nuevas anuales en el mundo - Professional Association of Diving Instructors (PADI)
  - 2 muertes/100.000 inmersiones
    - Primera causa: ahogamiento
    - Segunda causa: embolismo gaseoso: 13-24% muertes
  - Ley de Boyle y ley de Dalton

    - La Ley Dalton, también llamada **de las presiones parciales** dice que, **a una temperatura dada, la presión total de una mezcla de gases es igual a la suma de las presiones parciales** ejercidas por cada uno de los gases que componen la mezcla.

EXPERT REVIEW OF RESPIRATORY MEDICINE https://doi.org/10.1080/17476348.2023.2284375

Otras consider

- Buceo recrea ntados
- Por cada 10m
  - 1ATA=10kPa
  - La presión s la mitad
  - Buceador re
  - Ascenso a s 100%
  - Es crucial q mente durante el a
  - Patología p éreo



Otras consideraciones

- Guías BTS 2003 contraindican buceo
  - Enfermedad bullosa o quística
  - Historia neumotórax
    - No hay consenso internacional si pleurodesis: desaconsejar buceo
- Screening todos buceadores con TC?
- BHD desaconsejar ⟶ TC baja radiación: desaconsejar si quistes
  - Y a familiares

EXPERT REVIEW OF RESPIRATORY MEDICINE https://doi.org/10.1080/17476348.2023.2284375

Otras consideraciones

- Yoga y LAM

HATHA YOGA
24 semanas

No diferencias significativas:

- FEV1
- V'O2 pico
- VEGF-D
- SGRQ

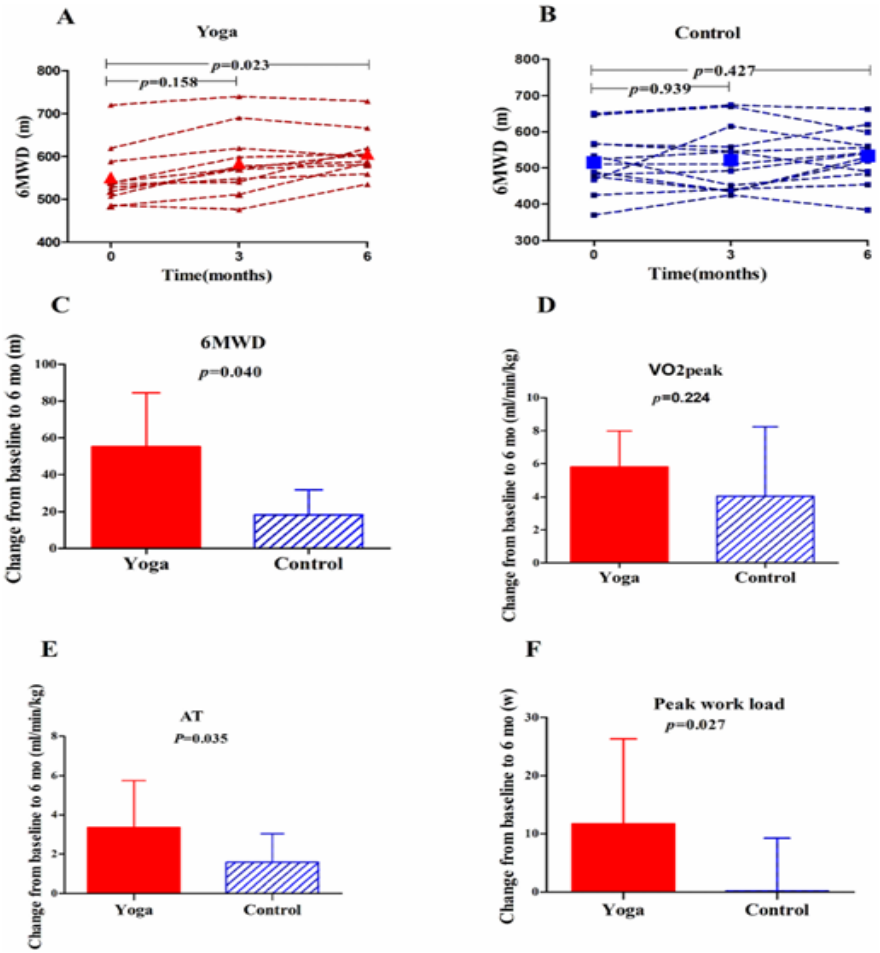


Fig. 2 Comparison of Six Minute Walking Test and Incremental Cardiopulmonary Exercise Test between Yoga and Control Groups. Six minute walking distance (6MWD) at baseline, 3 months and 6 months in yoga group (**a**) and control group (**b**). Changes of 6MWD (**c**), peak oxygen consumption (VO_2peak) (**d**), anaerobic threshold (AT) (**e**) and peak work load (**f**) from baseline to 6 months after yoga exercise or control

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Otras consideraciones

Table 6. Respiratory Care Implications for DCLD Patients

Topic	Guidance
Supplemental oxygen	Same as other chronic pulmonary disorders. Assess with 6MWT and pulse oximetry.
Pulmonary rehab	Symptomatic DCLD patients may benefit from pulmonary rehab. In one study of 40 LAM patients pulmonary rehab led to improvements in exercise capacity, dyspnea and health-related quality of life.
Positive pressure ventilation	Although there are no studies that specifically examined the effects of positive-pressure ventilation in patients with DCLDs, given the high risk of pneumothorax in this patient population, it is prudent to be aware of this risk, avoid positive-pressure ventilation unless absolutely necessary, and when utilized to use the lowest PEEP and tidal volumes possible to minimize the overall risk of pneumothorax.
PFTs	1. DCLD patients should undergo periodic longitudinal monitoring of PFTs to monitor the trajectory of disease progression and/or treatment response. 2. Although rare, RTs should be aware of the possibility of a pneumothorax during/immediately following PFT. 3. RTs should ask about pneumothorax history; recent pneumothorax (ie, within the past 30 d) is a relative contraindication to performing PFTs. 4. DCLD patients frequently exhibit a significant bronchodilator response to short-acting β agonists (eg, approximately one fourth of LAM patients). As such, unless contraindicated, spirometry on DCLD patients should include post-BD measurements.

Data from References 125–128.
6MWT = 6-min walk test
DCLD = diffuse cystic lung diseases
LAM = lymphangioleiomyomatosis
PFT = pulmonary function test
RT = respiratory therapist
BD = bronchodilator response

Conclusiones

- PFR no diagnósticas
- Importante volúmenes; valorar atrapamiento
- Útiles en seguimiento y pronóstico
- Tener en cuenta
  - Neumotórax
  - Vuelos en avión
  - Buceo
  - Ventilación con presión positiva

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