The Value of Exercise in the Cystic Fibrosis Clinic

Dr Patrick J Oades


Miller
Lite
RBEL
YELL

WHO Global recommendations for physical activity and health 2010.

Geneva: WHO Library Cataloguing-in-Publication Data. [ISBN: 978 924159 9979]

Participation in physical activity on a regular basis has important health benefits.

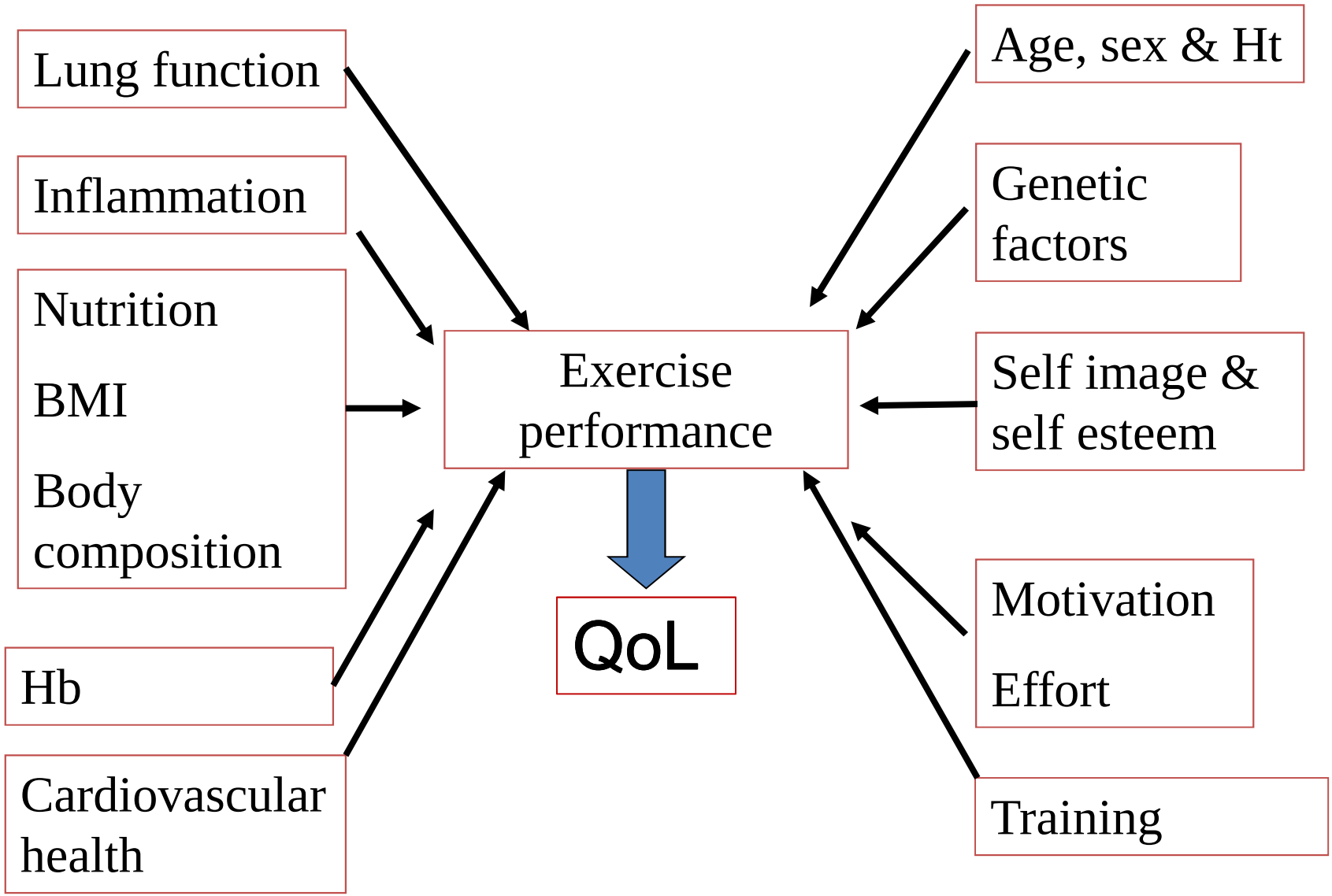
In healthy adults at least 150 minutes of physical activity each week (moderate-vigorous intensity)

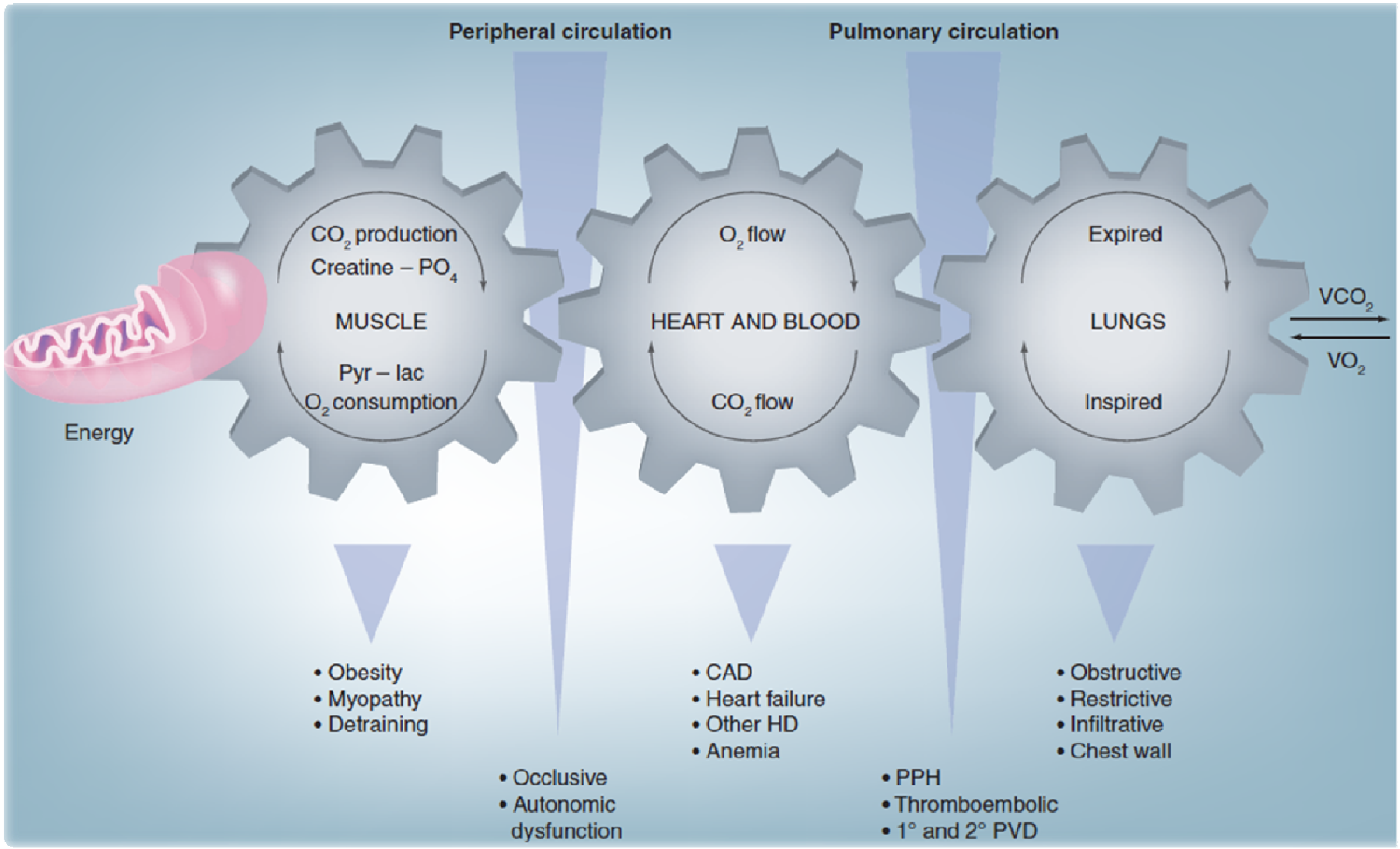
Improves:

- Cardio-respiratory fitness
- Bone health

Reducing the risk of non-communicable diseases:

- Obesity
- Heart disease
- Depression


Lung function
Inflammation
Nutrition
BMI
Body composition
Hb
Cardiovascular health
Age, sex & Ht
Genetic factors
Exercise performance
Self image & self esteem
QoL
Motivation
Effort
Training


Peripheral circulation
Pulmonary circulation
CO_2 production
Creatine – PO_4
MUSCLE
Pyr – lac
O_2 consumption
O_2 flow
HEART AND BLOOD
CO_2 flow
Expired
LUNGS
Inspired
VCO_2
VO_2
Energy
• Obesity
• Myopathy
• Detraining
• CAD
• Heart failure
• Other HD
• Anemia
• Obstructive
• Restrictive
• Infiltrative
• Chest wall
• Occlusive
• Autonomic dysfunction
• PPH
• Thromboembolic
• 1° and 2° PVD

Manifestations of

Cystic Fibrosis

General
-Growth failure (malabsorption)
-Vitamin deficiency states
(vitamins A, D, E, K)

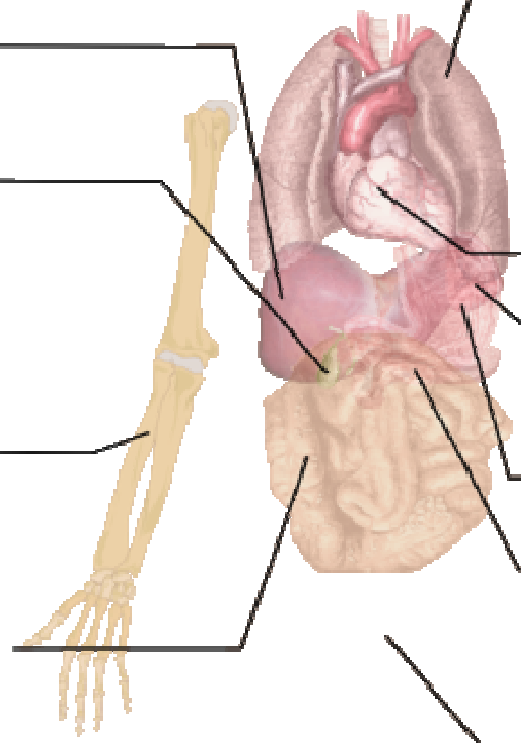
Nose and sinuses
-Nasal polyps
-Sinusitis

Liver
-Hepatic steatosis
-Portal hypertension

Gallbladder
-Biliary cirrhosis
-Neonatal obstructive jaundice
-Cholelithiasis

Bone
-Hypertrophic osteoarthropathy
-Clubbing
-Arthritis
-Osteoporosis

Intestines
-Meconium ileus
-Meconium peritonitis
-Rectal prolapse
-Intussusception
-Volvulus
-Fibrosing colonopathy (strictures)
-Appendicitis
-Intestinal atresia
-Distal intestinal obstruction syndrome
-Inguinal hernia



Lungs
-Bronchiectasis
-Bronchitis
-Bronchiolitis
-Pneumonia
-Atelectasis
-Hemoptysis
-Pneumothorax
-Reactive airway disease
-Cor pulmonale
-Respiratory failure
-Mucoid impaction of the bronchi
-Allergic bronchopulmonary aspergillosis

Heart
-Right ventricular hypertrophy
-Pulmonary artery dilation

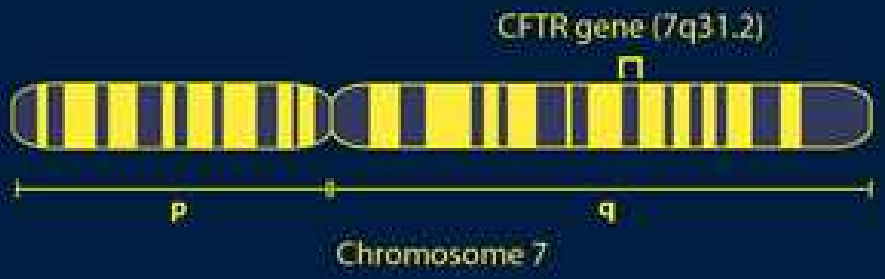
Spleen
-Hypersplenism

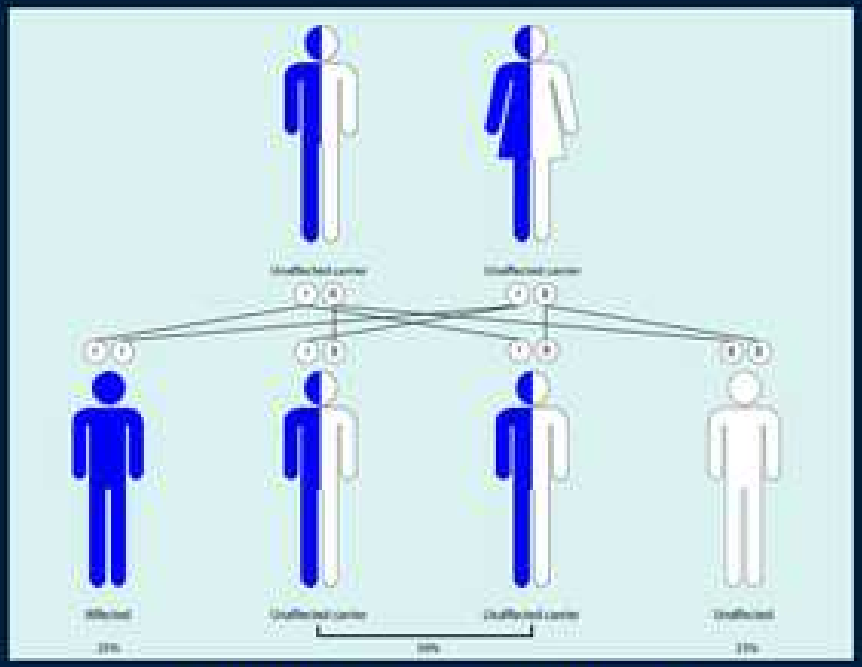
Stomach
-GERD

Pancreas
-Pancreatitis
-Insulin deficiency
-Symptomatic hyperglycemia
-Diabetes

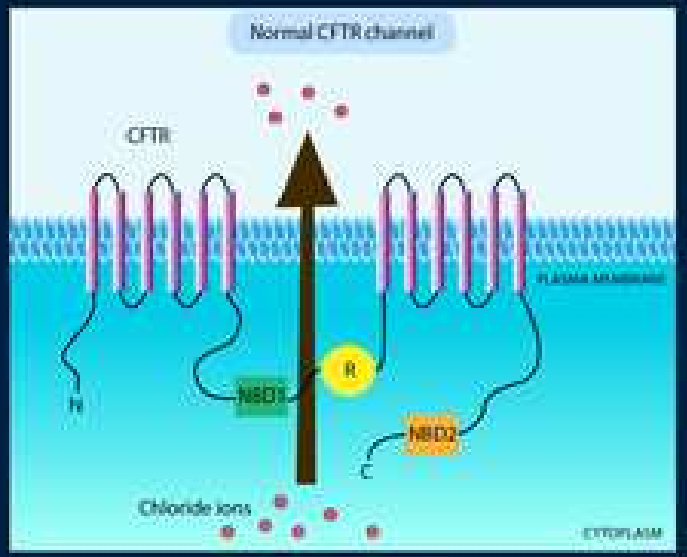
Reproductive
-Infertility
(aspermia, Absence of vas deferens)
-Amenorrhea
-Delayed puberty

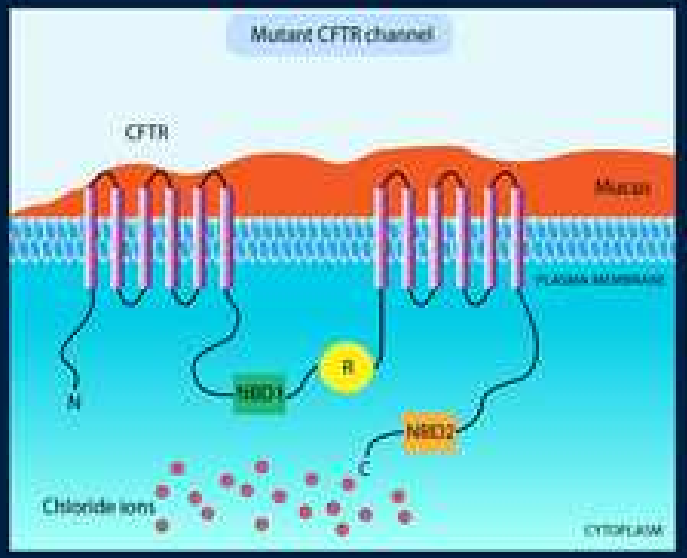
Cystic fibrosis





Autosomal Recessive Inheritance





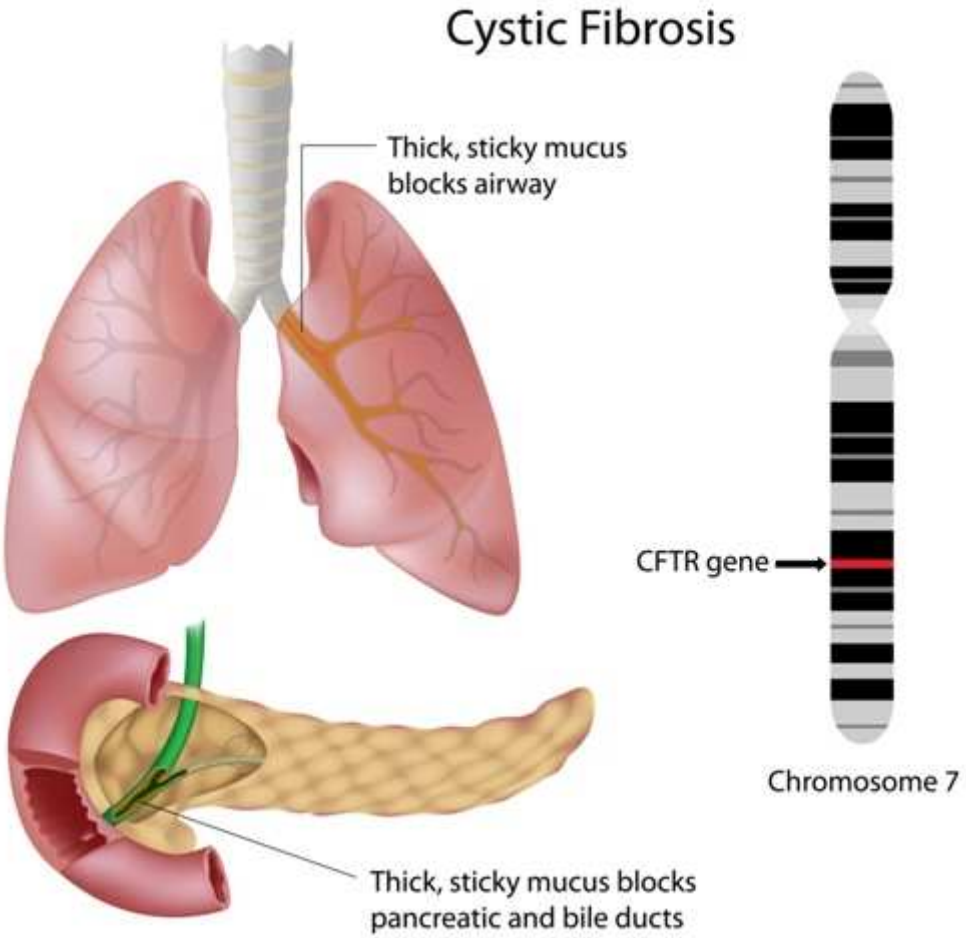
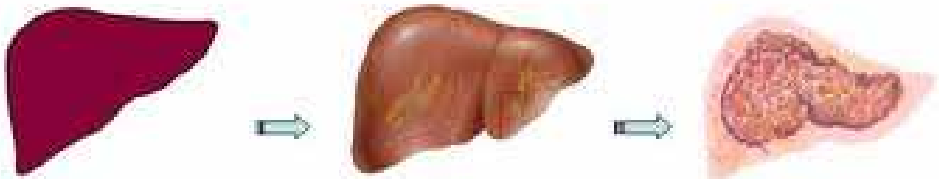


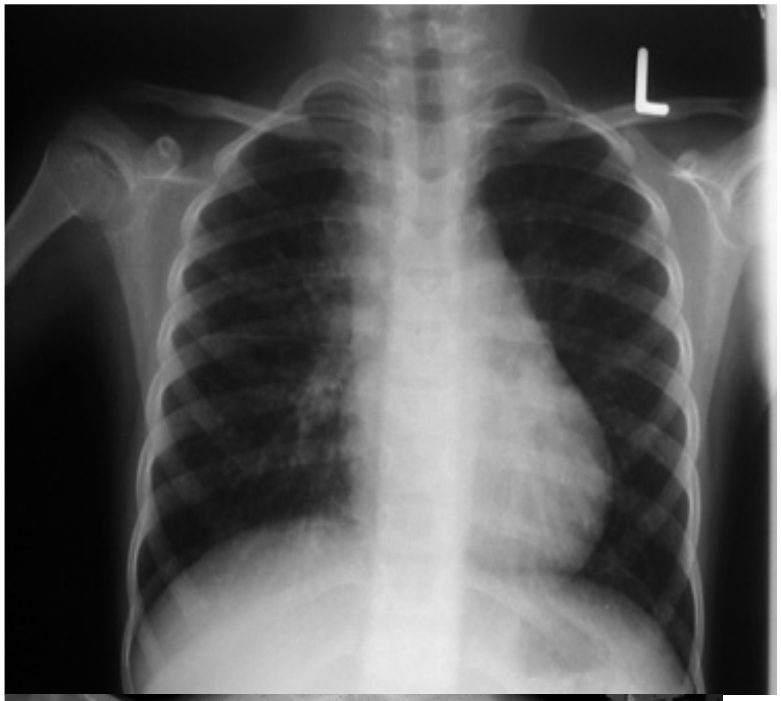
FIGURA 1



HIGADO NORMAL

HIGADO CON CICATRICES (FIBROSIS)

CIRROSIS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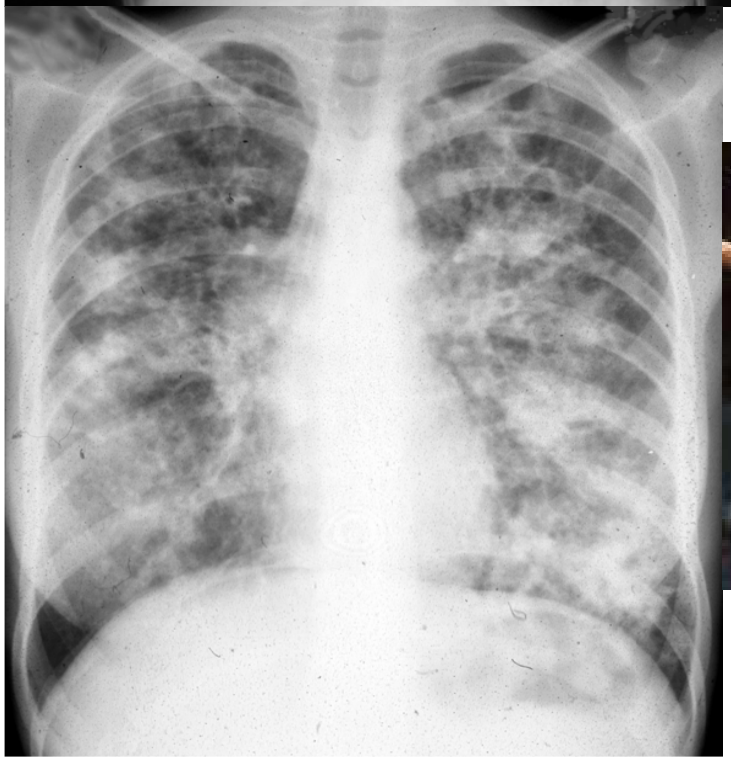
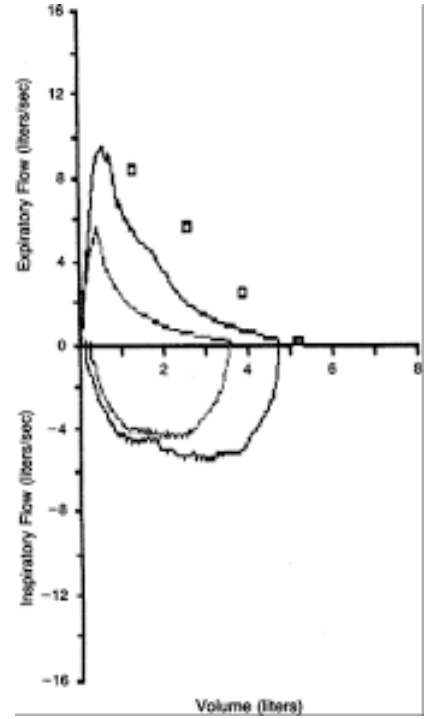
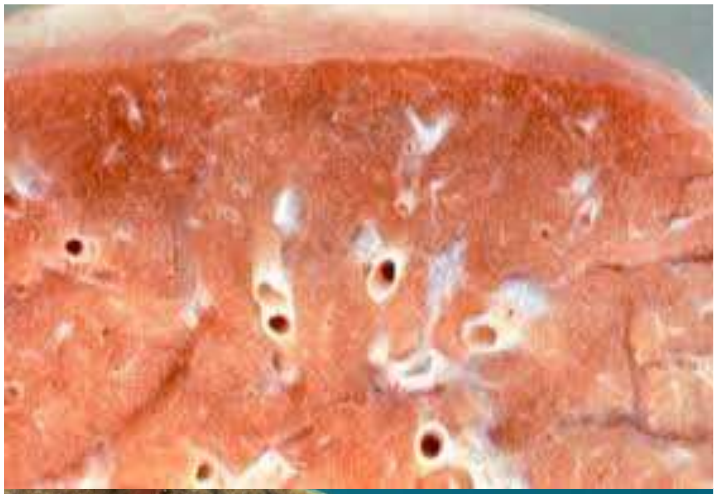
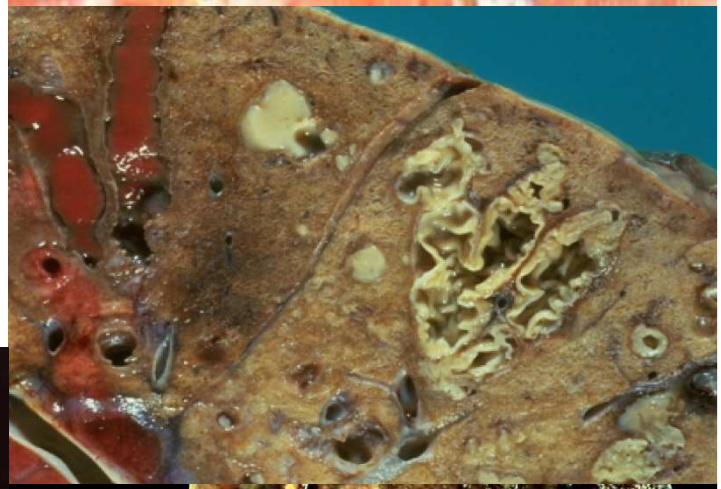
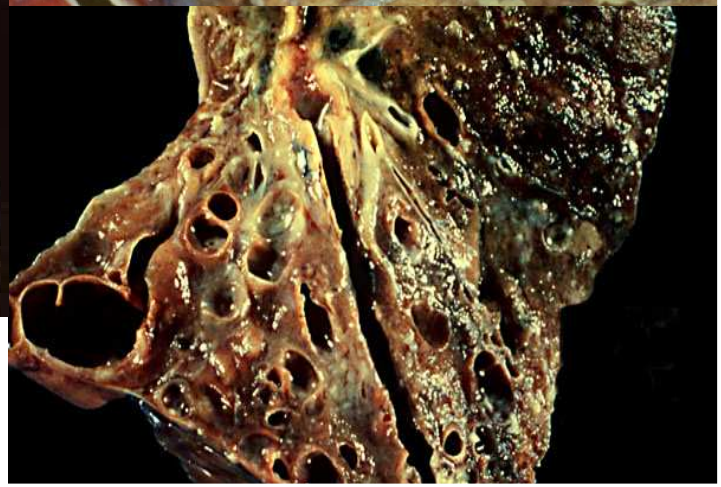
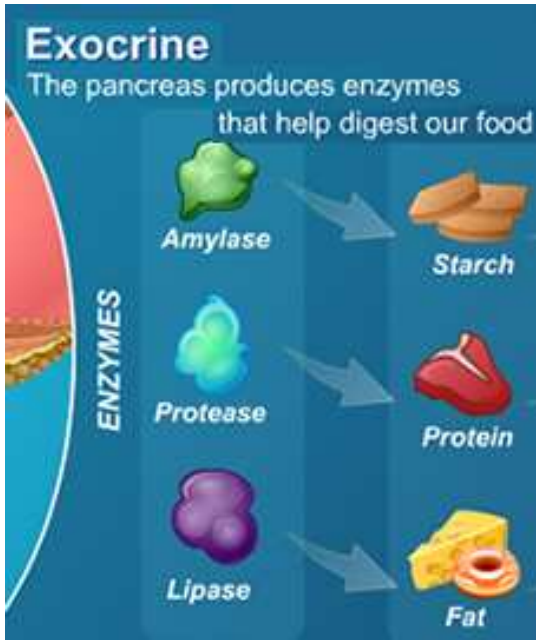


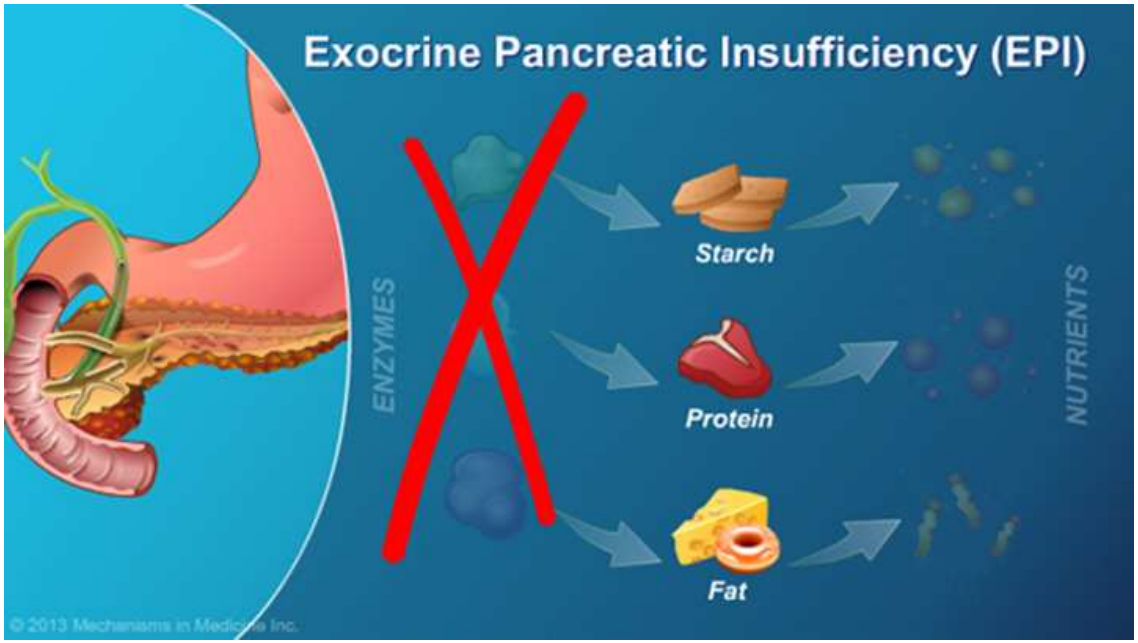

Expiratory Flow (liters/sec)
Inspiratory Flow (liters/sec)
Volume (liters)
16
12
8
4
0
−4
−8
−12
−16
2
4
6

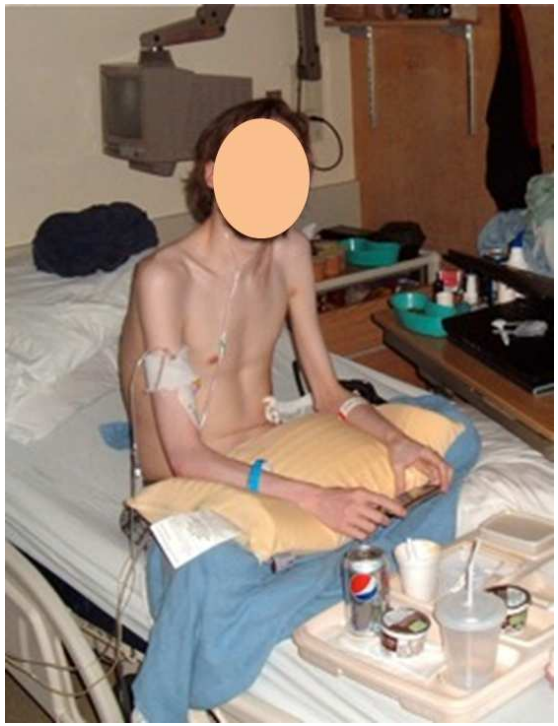







Exocrine
The pancreas produces enzymes that help digest our food
ENZYMES
Amylase
Starch
Protease
Protein
Lipase
Fat


Exocrine Pancreatic Insufficiency (EPI)
ENZYMES
Starch
Protein
Fat
NUTRIENTS
© 2013 Mechanisms in Medicine Inc.



Malabsorption

Poor appetite when unwell

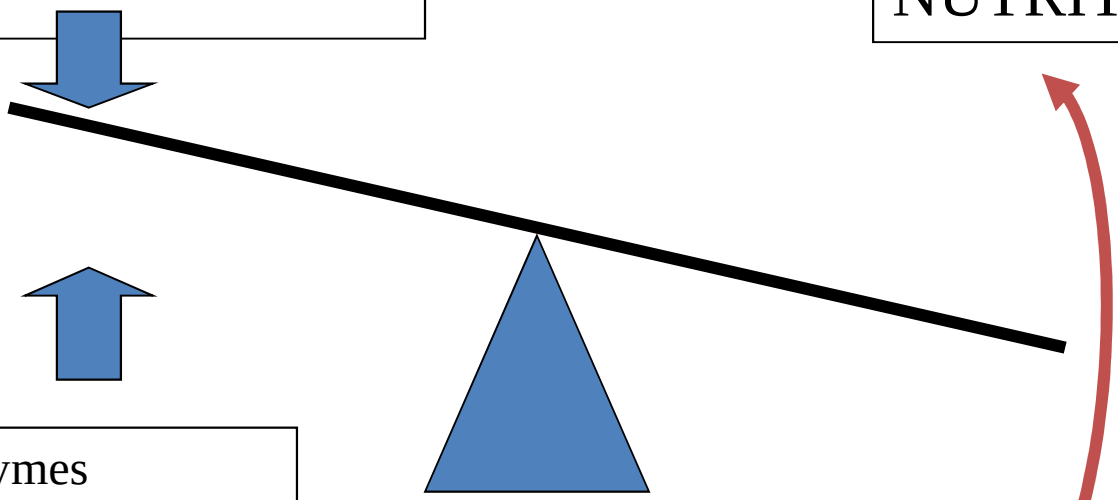
High energy demands of infection.

Intrinsic energy wasting defect.

Diabetes.

Eating disorders.

POOR NUTRITION

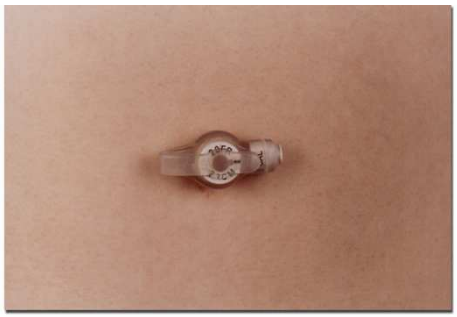


Pancreatic enzymes

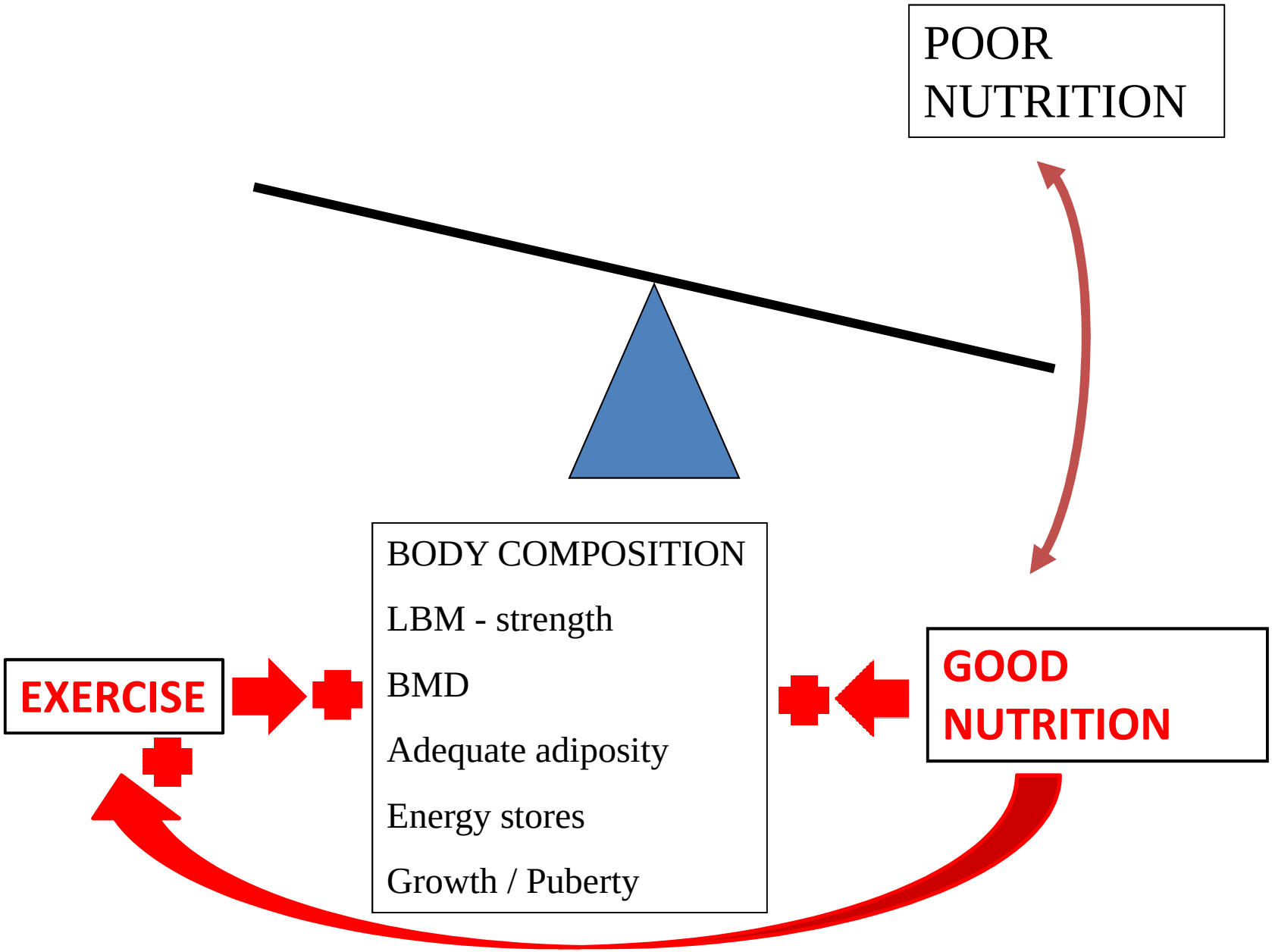
High energy diet

Supplements

Insulin



GOOD NUTRITION


POOR NUTRITION
BODY COMPOSITION
LBM - strength
BMD
Adequate adiposity
Energy stores
Growth / Puberty
EXERCISE
GOOD NUTRITION

EXERCISE

TREATMENT

TESTS

Treatment:
Benefits of exercise on lung disease in CF

- ↑Airway clearence (↑ ventilatory flows, ↓ sputum viscosity, vibration & impact).
- ↑ lung function / slows the rate of decline.
- ↓ Exacerbation rate and hospitalisations
- ↓ Reduced mortality
- ↑ Functional capacity
- ↑ QoL / self esteem

NEED TO PERFORM AIRWAY CLEARENCE TOO! Pausing for huff or coughing.

Exercise is medicine in CF. *Wheatley et al.* ***Exerc Sport Sci Rev. 2011;39:155.***
Effects of exercise on respiratory flow and sputum properties in patients with CF. *Dwyer et al,* ***Chest 2011;139:870.***
The role of exercise in maintaining health in CF. *Dwyer et al.* ***Curr Opin Pulm Med. 2011;17:455–460.***
Longitudinal relationship between physical activity and lung health in CF. *Schneiderman et al.* ***Eur Respir J. 2014;43:817–823.***
Effects of a supervised, outpatient exercise and physiotherapy programme in children with cystic fibrosis. *Urquhart D et al.* ***Pediatr Pulmonol. 2012;47:1235.***
Quality of life is associated with physical activity and fitness in CF. *Hebestreit et al.* ***BMC Pulm Med. 2014;14:26.***

Physical training for cystic fibrosis.

- Improves aerobic capacity (VO2max)

Gruber W et al. J Pediatr. 2011;158:71–76.
Gruber W et al. Eur Respir J. 2011;38:1336–1342.
Hulzebos H et al. Physiother Theory Pract. 2011;27(3):231–237.

- Improves strength, lean body mass and self concept.

Selvadurai H et al. Pediatr Pulmonol. 2002;33:194–200.
Orenstein D et al. Chest. 2004;126:1204–1214.

Bradley JM, Moran F. ***Cochrane Database of Systematic Reviews*** *2008.*

Regular physical activity may:

- Diminish dyspnoea
- Improve exercise tolerance.

Exerts a positive influence on:

- Bone mineral accretion
- Blood glucose regulation
- Posture
- Appetite

Other considerations

- Nutritional support: fluid, energy & salt intake.
- Infection control - cross-infection risk,
  - Patients must be segregated from their CF peers
  - Advised not to socialise or exercise together in rehabilitation programs.
  - Testing equipment sterile/environment.
- Psychological support, dealing with limitations

As perception of illness severity ↑:

- Participation in exercise decreases.
- Patients describe ↓ importance & ↑ burden of exercise activities when compared to other therapies.

Myers LB. Journal of Cystic Fibrosis 2009;8(6):425–7.

Adherence to prescribed exercise programs reported at around 50%.

White D et al. Journal of Cystic Fibrosis 2007;6(3):163–70.

EXERCISE TREATMENT

Training format and frequency? – Influences the pattern observed in outcomes.

Individualised (gender, age, maturity, interests)
Variety (to maintain interest)
Reinforced – Trainer.
Cost (time and money).
Adherence?
Sustainable?

Pull back intensity during exacerbations.

Exercise and Habitual Physical Activity for People with CF: Expert Consensus, Evidence based Guide for Advising Patients.

Swisher et al. Cardiopulm Phys Ther J. 2015;00:1-14

Lifelong regular physical activity should be part of daily care.

Recommendations:

Habitual activity: children 60min/day; adult 150- 300 min/week

Aerobic exercise - prescribe (7+) at an intensity relative to max aerobic capacity.

Children: 30- 60 min/day, moderate/vigorous devel. appropriate activity.

Adults: 30-60 min/day of moderate/vigorous activity.

Resistance training prescribe (13+) relative to max strength.

Repetitive bouts 2-3x/week.

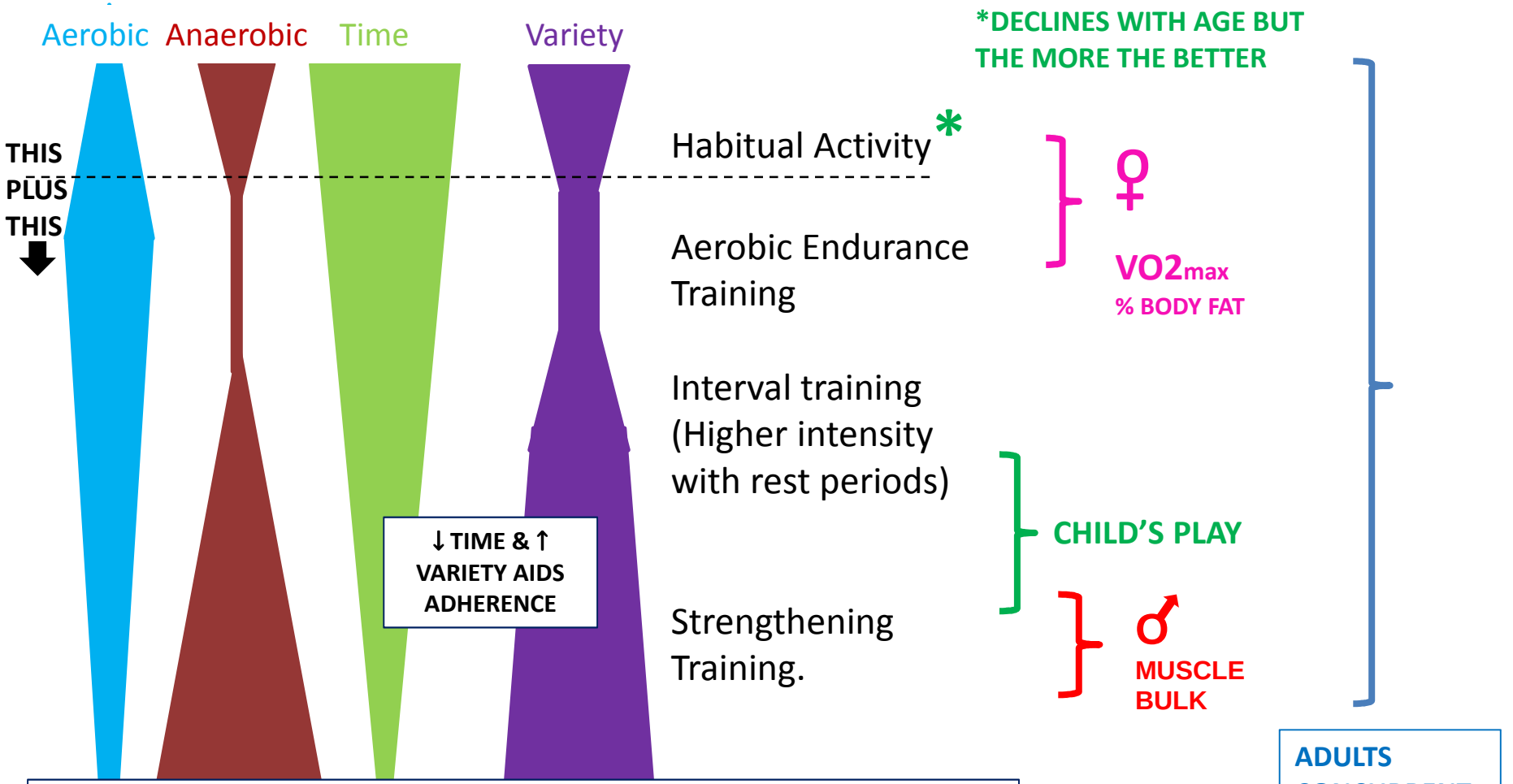
Avoid Valsalva and other breath-holding patterns.

Considerations:

Rest / Posture / Joint flexibility

Drugs & co-morbidities (severe impairment of lung function, hypoxaemia, post transplant, recent haemoptysis / PnTx, pregnancy, low BMI, CFRD, low BMD).

Risks (Injury/arrhythmias/hypoglycaemia/pneumothorax), all very rare



ALL ACTIVITY UNDERPINNED WITH:

- **NUTRITION SUPPORT (inc. FLUID INTAKE AND NaCl)**
- **MAINTENANCE OF FLEXIBILITY, ESPECIALLY IN OLDER GROUPS**

The role of exercise and physical activity in optimising outcomes among patients with CF.

GD Wells. Johns Hopkins e Literature Rev. 2012

TESTING

USUAL

Ht & Wt, BMI

SaO2

FEV1

Symptoms

Signs

Respiratory microbiology

SOMETIMES

CXR

HRCT

Bronchoscopy

Measures of Exercise Performance?

Detailed, objective measures of physiological health.

STANDARDISATION

'OUTCOMES' used for

1. Monitoring
2. Response to intervention

What more does exercise testing tell us?

Lung function and exercise tests monitor different aspects of the disease. Poor relationship between these two tests, especially in mild/mod disease. Peak oxygen uptake (Vo2peak) and forced expiratory volume in 1 s (FEV1) are independent predictors of mortality in CF.

Exercise testing and prognosis in adult cystic fibrosis. *Moorcroft. Thorax 1997; 52: 291–293.*
Role of exercise in the evaluation and management of pulmonary disease in children and youth. *Nixon PA. Med Sci Sports Exerc 1996; 28: 414–420.*
Early Oxygen Uptake Recovery Following Exercise Testing in children with Chronic Chest Disease. *Stevens D, Oades P, Armstrong N, Williams C. Pediatr Pulmonology 2009;44:480-8.*

ATS/ACCP Statement on cardiopulmonary exercise testing.
Am J Respir Crit Care Med 2003; 167: 211–277.
'The use of CPET in patient management is increasing with the understanding that resting pulmonary and cardiac function testing cannot reliably predict exercise performance and functional capacity and that, overall, health status correlates better with exercise tolerance rather than resting measurements'.

Respiration

Guidelines

Respiration
DOI: 10.1159/000439057

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Statement on Exercise Testing in Cystic Fibrosis

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Frank Cerny[g] Erik H.J. Hulzebos[c] Chantal Karila[j] Larry C. Lands[k]
John D. Lowman[h] Anne Swisher[i] Don S. Urquhart[e]
for the European Cystic Fibrosis Exercise Working Group

International multi-discipinary consensus endorsed by ERS & ECFS.

Testing evaluates physical limitation and explores exercise related symptoms.

Gold standard is an incremental test to measure:

- SaO2,
- Ventilatory gas exchange,
- VO2max
- max WR

} Exercise capacity – prognosis / QoL

Cycle Ergometer - Godfrey Protocol.

Field tests (6 min walk, 3 min step, SWT) – cheaper set up, much less info.

Why use CPET?

ECG treadmill
- Ischemia/12-lead ECG
- Heart rate/HRR
- Blood pressure, double product
- Estimate of METs
- Symptoms/reason for stopping

CPET
- Ischemia/12-lead ECG
- Heart rate/HRR
- Blood pressure, double product
- Measured work rate
- Symptoms/ reason for stopping
- Oxygen saturation
- Maximal oxygen uptake (VO_2 max)
- Lactic acidosis threshold
- Carbon dioxide output (VCO_2)
- Minute ventilation (V_E), TV, RR/BR
- Ventilatory equivalents (V_E/VO_2, V_E/CO_2)
- VO_2/ work rate relationship ($\Delta VO_2/\Delta WR$)
- O_2 pulse (SV × C(a-v)O_2 difference)
- Respiratory exchange ratio
- End tidal O_2, CO_2
- Blood gases/COHb
- Vd/Vt
- P(A-a)O_2 → low V_A/Q
- P(a-ET)CO_2 → high V_A/Q
- Expiratory flow pattern

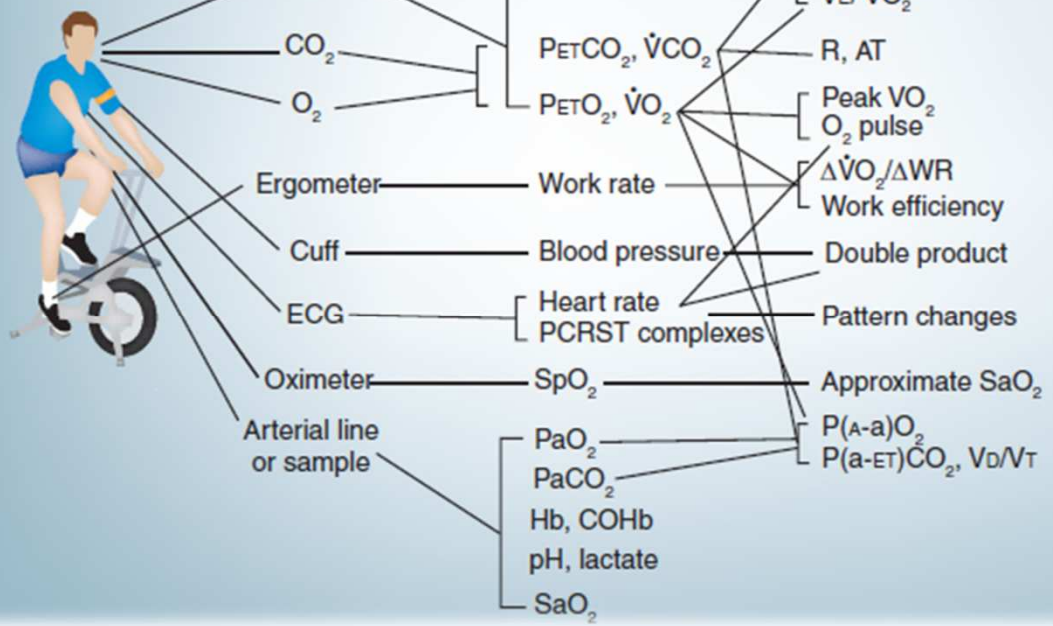


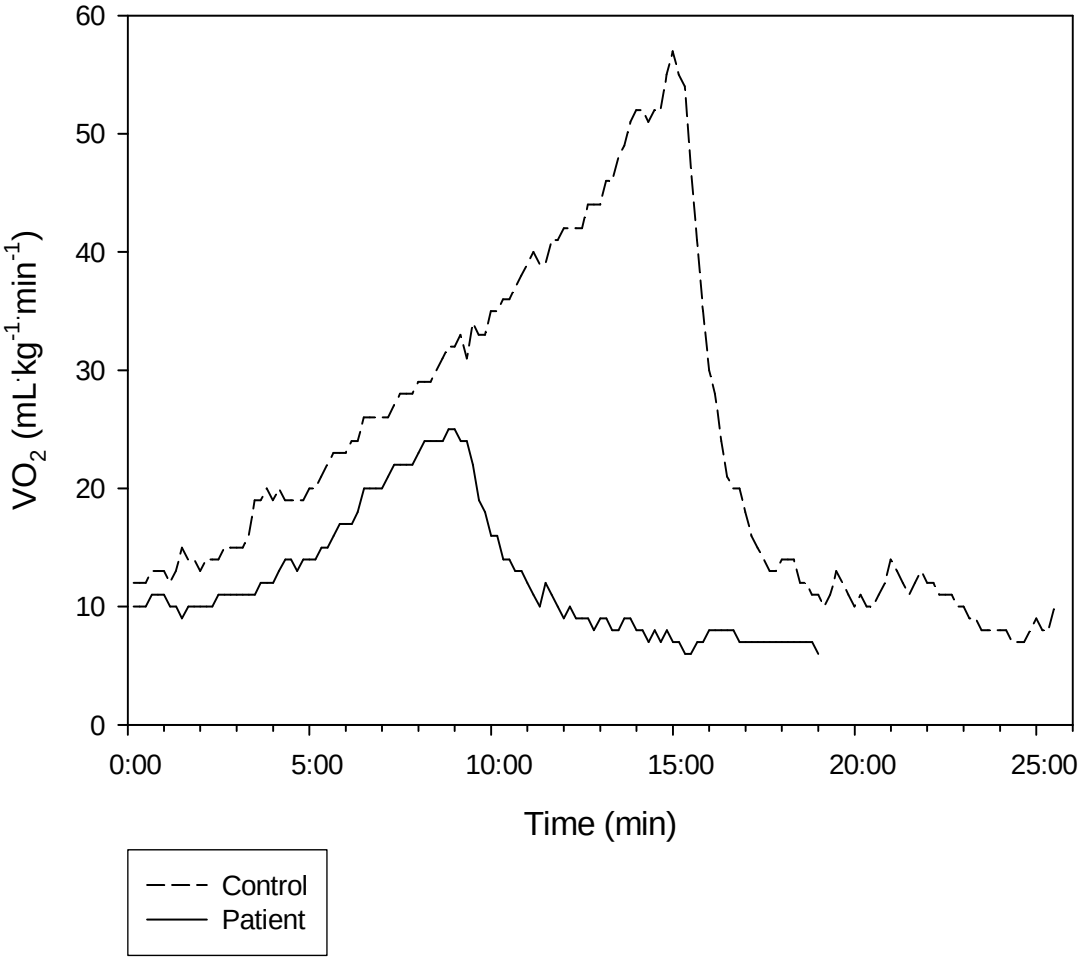
Table 2
Patients' physiological responses to CPET during the three visits.

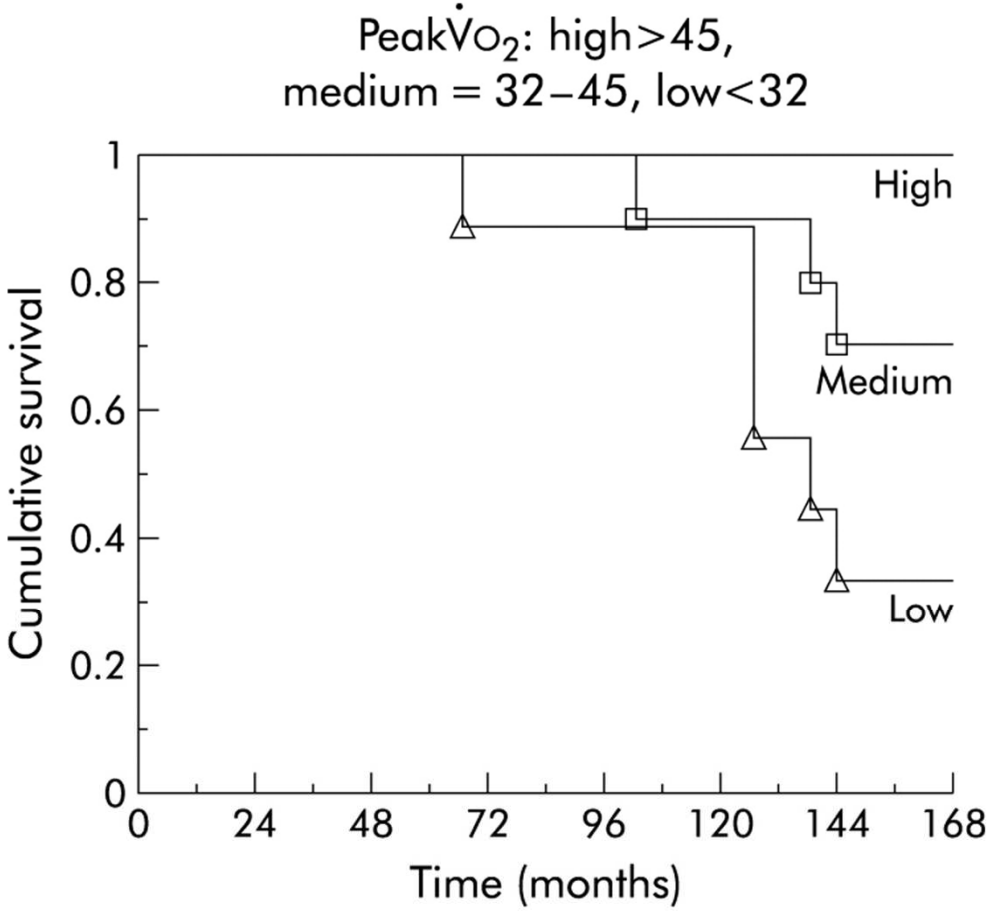
Variable	*n*	Test 1	Test 2	Test 3
Maximal exercise parameters				
$\dot{V}O_{2max}$ ($L \cdot min^{-1}$)	13	1.77 (0.57)	1.76 (0.56)	1.68 (0.55)
HR_{peak} ($b \cdot min^{-1}$)	11	190 (12)	186 (14)	186 (19)
SaO_2 (%)	13	95 (3)	96 (1)	96 (3)
RPE	13	9 (2)	9 (2)	9 (1)
RPD	13	7 (3)	6 (3)	8 (3)
Ramp peak power output (W)	13	157 (55)	148 (62)	145 (65)
Submaximal parameters				
GET ($L \cdot min^{-1}$)	12	1.00 (0.22)	0.93 (0.21)	1.05 (0.29)
MRT (s)	11	42 (15)	65 (17)	54 (26)
$\dot{V}O_2$ gain ($mL \cdot min^{-1} \cdot W^{-1}$)	12	8.01 (1.36)	8.11 (1.22)	7.73 (2.64)
$OUES_{100}$ ($mL \cdot min^{-1} \cdot logL^{-1}$)	12	803 (227)	789 (181)	799 (218)
$OUES_{GET}$ ($mL \cdot min^{-1} \cdot logL^{-1}$)	12	797 (223)	730 (188)	756 (389)
$\dot{V}_E/\dot{V}CO_2$-slope	12	34.13 (4.51)	33.26 (3.25)	32.14 (5.39)
$\dot{V}_E/\dot{V}O_2$ at the GET	12	28.57 (5.45)	28.63 (3.84)	28.09 (4.58)
$\dot{V}_E/\dot{V}CO_2$ at the GET	12	28.07 (3.96)	29.15 (5.43)	27.95 (5.51)

Values are means ± SD, with the range also displayed unless otherwise stated. $\dot{V}O_{2max}$, maximal oxygen uptake; HR_{peak}, peak heart rate; SaO_2%, end-exercise arterial oxygen saturation; RPE, end-exercise rating of perceived exertion; RPD, end-exercise rating of perceived dyspnoea; ramp; incremental ramp test; GET, non-invasive estimate of the lactate threshold which was verified by the ventilatory threshold; MRT, mean response time; $\dot{V}O_2$ gain, oxygen cost of exercise; $OUES_{100}$, oxygen uptake efficiency slope for the entire duration of the ramp test; $OUES_{GET}$, OUES to the GET; $\dot{V}_E/\dot{V}CO_2$-slope, ventilatory drive; $\dot{V}_E/\dot{V}O_2$, ventilatory equivalent for oxygen uptake; $\dot{V}_E/\dot{V}CO_2$, ventilatory equivalent for carbon dioxide.

PAST THE PEAK - RECOVERY

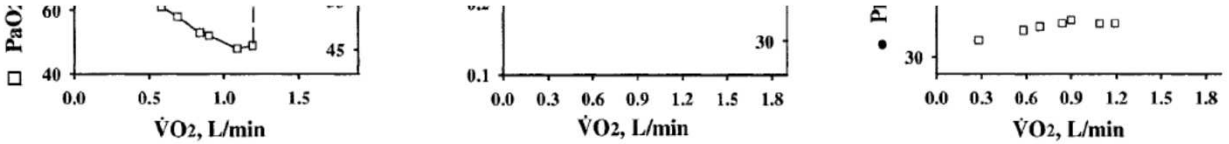
VO_{2peak} and VO_{2peak} recovery profile during and following maximal exercise in a young CF patient and control





28 patients (8-17 yrs) Maximal ETT

Pianosi, P et al. Thorax 2005;60:50-54



Cardiopulmonary exercise testing in children with cystic fibrosis: one centre's experience.

Weir et al. Arch Dis Child 2016. doi:10.1136/archdischild-2016-310651

- Demonstrated CPET as part of annual review in children aged >7 years is feasible
- In mild disease, there is no significant correlation between Vo2peak and FEV1 or body mass index.
- A decline in fitness can be used as a trigger for more intensive physiotherapy intervention.

Conclusions: Exercise in CF

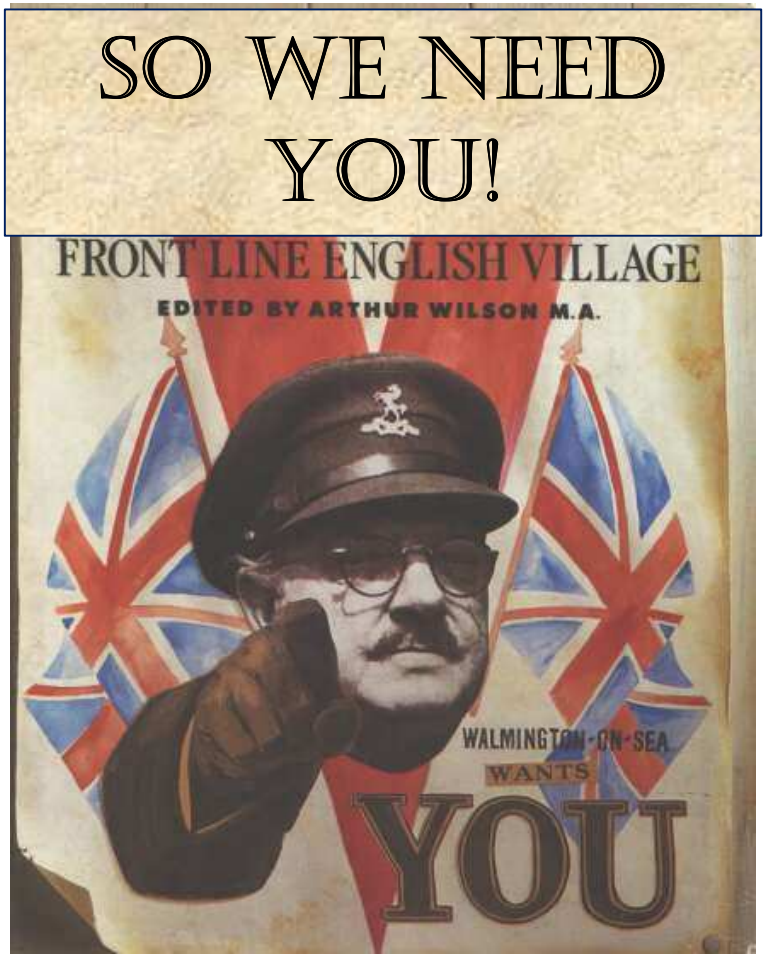
TREATMENT

- Exercise is good.
- Individualised.
- Improves outcomes, QoL & Self esteem.
- Needs nutritional support.

TESTS

- Need Standardisation
- Tell us additional information (to FEV1 and imaging)
- CPET measures are an independent prognostic indicator.
- Will be useful in monitoring and determining response to intervention.

Questions?


SO WE NEED YOU!
FRONT LINE ENGLISH VILLAGE
EDITED BY ARTHUR WILSON M.A.
WALMINGTON-ON-SEA
WANTS
YOU

£, ⌚, Motivation, Maintenance?

- One-off one-to-one counselling or advice.
- Self-directed or unsupervised participation in a prescribed physical activity programme.
- Supervised physical activity session in the home.
- Supervised physical activity session in a facility.
- On-going face-to-face counselling or advice.
- Telephone support.
- Written material.
- **Internet-based or tele-health advice and motivation.**
- Monitoring device for motivation, e.g. pedometer.

OUTCOME MEASURES USED FOLLOWING EXERCISE INTERVENTIONS

1. Exercise capacity (either maximal or submaximal where measured directly or by a standard field test)
2. Pulmonary function tests (change in per cent predicted or absolute measures from baseline, or rate of decline)
i) forced expiratory volume in one second (FEV1)
ii) forced vital capacity (FVC)
iii) forced expiratory flows between 25% and 75% of expired volume (FEF25−75)
3. Adverse outcomes (e.g. musculoskeletal injuries)
4. Body composition in terms of body mass index (BMI) and lean body mass
5. Bone mineral density (defined on dual energy X-ray absorptiometry (DXA) scans)
6. Adherence to the intervention programme
7. Compliance with other CF treatments, e.g. airway clearance techniques and nebulised medication; any measure of compliance such as pill counts, self-report diaries, electronic monitoring
8. Continued engagement in activity (quantified) beyond program – behaviour change.
9. Health-related QoL measures (generic +/or disease specific)
10. Cost evaluation

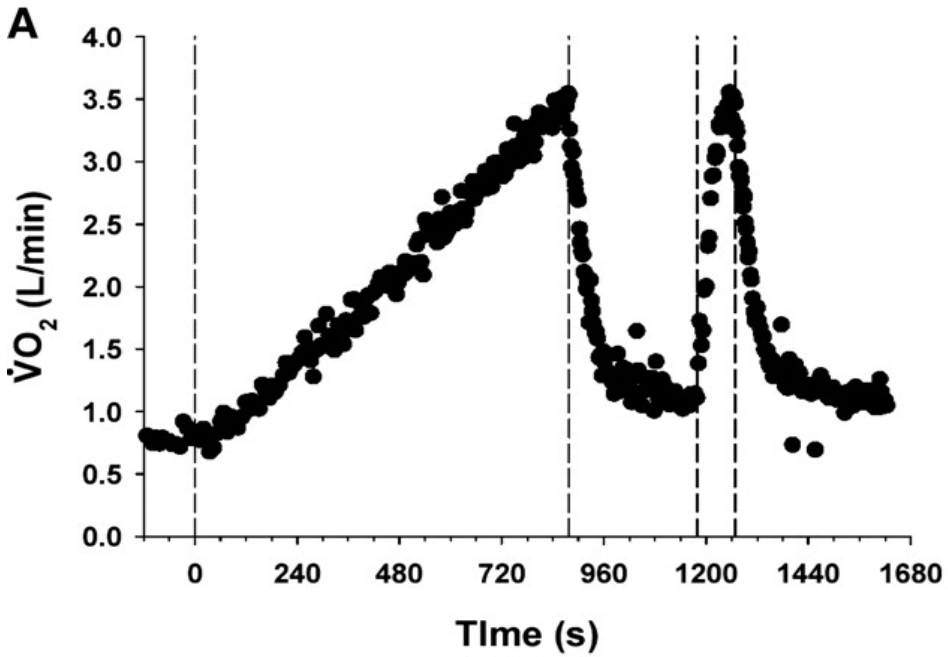
Valid CPET Protocol

Original research

A protocol to determine valid $\dot{V}O_{2max}$ in young cystic fibrosis patients

Zoe L. Saynor[a,b], Alan R. Barker[a], Patrick J. Oades[b], Craig A. Williams[b,*]

[a] *Children's Health and Exercise Research Centre, Sport and Health Sciences, University of Exeter, UK*
[b] *Royal Devon and Exeter NHS Foundation Trust Hospital, UK*

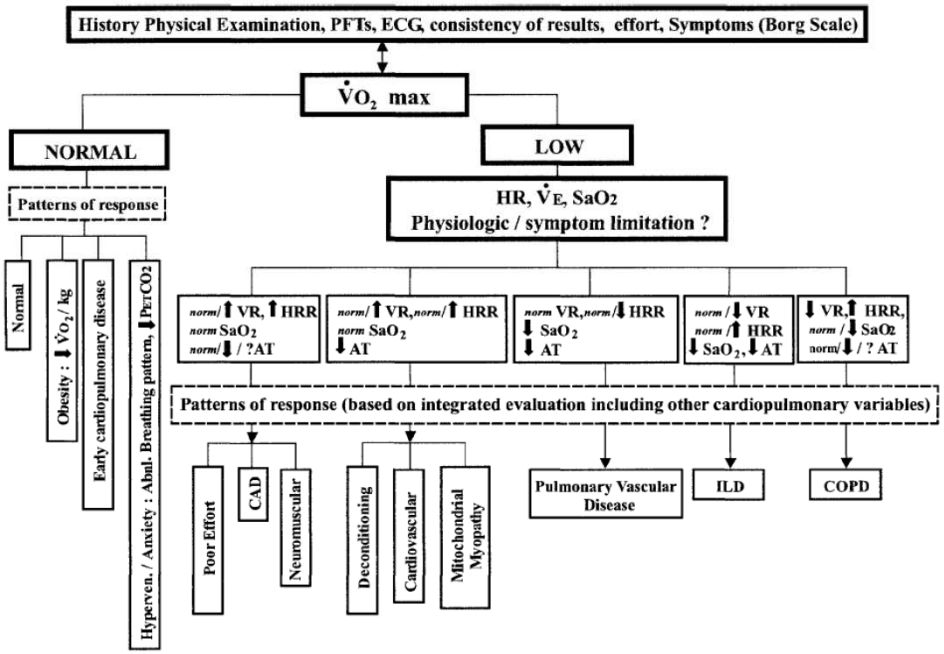


Saynor *et al.* (2013) J Sci Med Sport

Interventions for promoting physical activity in people with cystic fibrosis

Narelle S Cox, Jennifer A Alison, Anne E Holland

2011 Cochrane Database Systematic Reviews


History Physical Examination, PFTs, ECG, consistency of results, effort, Symptoms (Borg Scale)
V̇O2 max
NORMAL
LOW
Patterns of response
HR, V̇E, SaO2
Physiologic / symptom limitation ?
Normal
Obesity : ⬇ V̇O2 / kg
Early cardiopulmonary disease
Hyperven. / Anxiety : Abnl. Breathing pattern, ⬇ PETCO2
norm/⬆ VR, ⬆ HRR
norm SaO2
norm/⬇ / ?AT
norm/⬆ VR, norm/⬆ HRR
norm SaO2
⬇ AT
norm VR, norm/⬇ HRR
⬇ SaO2
⬇ AT
norm /⬇ VR
norm /⬆ HRR
⬇ SaO2, ⬇ AT
⬇ VR, ⬆ HRR,
norm /⬇ SaO2
norm/⬇ / ? AT
Patterns of response (based on integrated evaluation including other cardiopulmonary variables)
Poor Effort
CAD
Neuromuscular
Deconditioning
Cardiovascular
Mitochondrial Myopathy
Pulmonary Vascular Disease
ILD
COPD