

Difficult to control chest pain in patients with cystic fibrosis

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Objectives

- Understand the essentials, diagnosis and clinical manifestations of cystic fibrosis.
- Discuss the literature regarding pain in cystic fibrosis.
- Discuss options for therapy of pain in patients with cystic fibrosis.

Cystic Fibrosis Patients with difficult to control chest pain

3 CF patients since December:

21 yr old male with pleuritic chest pain

Deep chest pain worse with cough, nothing has helped so far

Palliative care consulted to start a trial of methadone therapy.

PFTs: FVC = 1.78L (34% predicted),

FEV1 = 0.82L (18% pred)

FEF₂₅₋₇₅ = 0.26 (5%)

Non cardiac chest pain

- GI related
 - GERD
 - Esophageal motility disorder
 - Visceral hyperalgesia
- Chest wall or musculoskeletal pain
- Pulmonary related
 - Venous ThromboEmbolic disease (VTE)
 - Pulmonary hypertension
 - Lung parenchyma related
 - Pneumonia / infection
 - Pleuritis / serositis
- Pain referred to chest

Cystic fibrosis - overview

- Most common fatal autosomal recessive disease among Caucasian population.
- A multi system disease affecting the digestive system, respiratory system, sweat glands, and reproductive tract
- Progressive lung disease is the main cause of morbidity and mortality.
- Abnormal transport of chloride and sodium across the respiratory epithelium

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

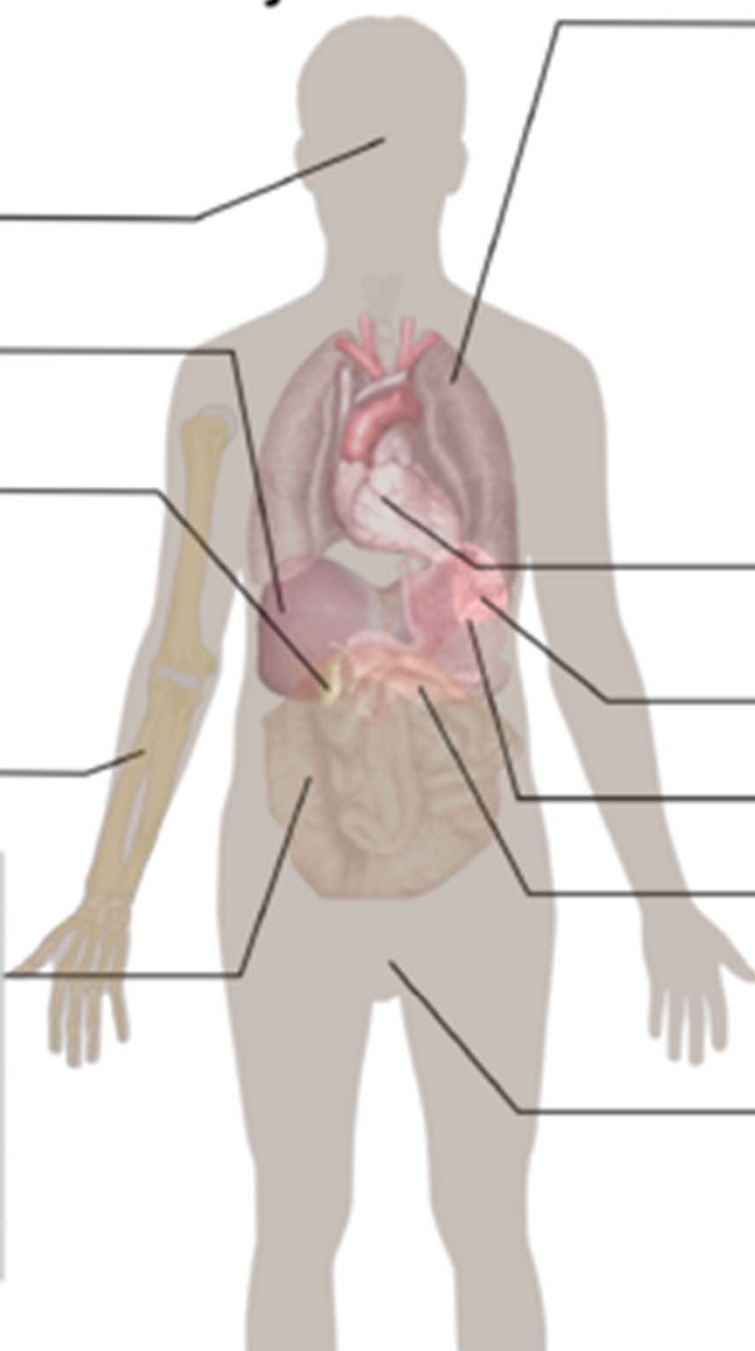
Heart
-Right ventricular hypertrophy
-Pulmonary artery dilation

Spleen
-Hypersplenism

Stomach
-GERD

Pancreas
-Pancreatitis
-Insulin deficiency
-Symptomatic hyperglycemia
-Diabetes

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



CF diagnosis

- Clinical symptoms consistent with CF in at least one organ system
- Evidence of CFTR dysfunction (cystic fibrosis transmembrane conductance regulator)
 - Elevated sweat chloride
 - Abnormal nasal potential difference.

CF GI disease

- Insufficiency of exocrine pancreas
 - Malabsorption of fat and protein
- Insufficiency of endocrine pancreas
- Focal biliary cirrhosis
- Meconium ileus (MI)– pathognomonic
- Distal intestinal obstruction syndrome (DIOS)
 - MI equivalent in older children
- Intussusception

CF - Reproductive system

- Infertility
 - 95% of men with CF are infertile
 - Defects in sperm transport
 - 20% of women with CF
 - Amenorrhea related to malnutrition
 - thick cervical mucous

CF – musculoskeletal system

- Reduced bone mineral content
- Hypertrophic osteoarthropathy

CF respiratory system

- Thick viscous airway secretions
- Chronic infection of respiratory tract
- Eventual infection with *Pseudomonas* species usually a mucoid phenotype

Respiratory symptoms

- Productive cough
- Chronic sinusitis
- Digital clubbing
- Progressive tissue damage – floppy airways leads to air trapping
- Airway hyper reactivity
- Bronchiectasis
- Obstructive pattern on PFTs
 - Decreased FEV1 and FEF25-75
- Decreased exercise tolerance
- Weight loss

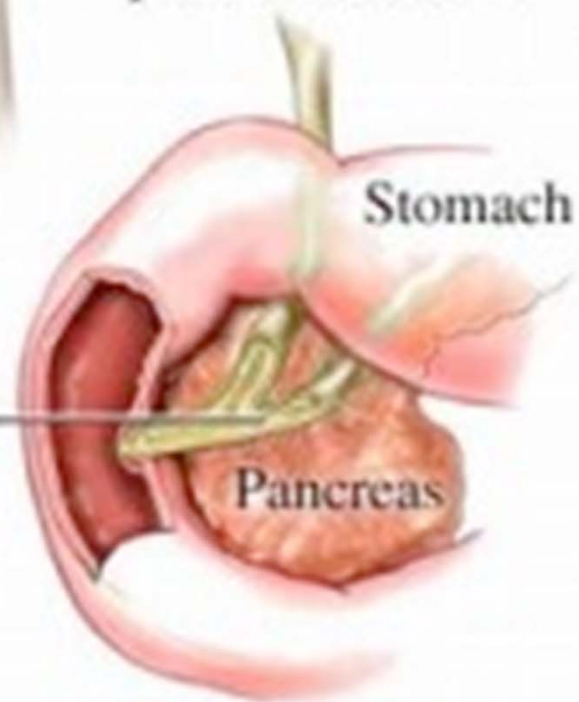


Mucus blocks
air sacs (alveoli)
in the lungs



Mucus blocks
pancreatic ducts

Pancreatic
duct



Pancreas

Digital clubbing in CF



Pain in CF?

Pain in children and adults with cystic fibrosis: a comparative study

- 73 children, 110 adults
- 59% of children and 89% of adults reported pain in past month
- No difference in patients with severe disease vs milder CF
- Abdomen in children
- Back, head and chest in adults
- Absenteeism in 15%
- 27% reported pain caused negative impact on family life
- Most common therapy – acetaminophen
- Even patients with mild disease had regular painful symptoms
- Chest physiotherapy reported as painful by 28% of children and 17% of adults

Prevalence of pain in adults with cystic fibrosis

- 250 adult CF patients in Italy
- 94.1% reported episodes of pain in the previous 2 months
 - Headache 63%, abdominal 51%, back 48%, “bones or muscular 44%, joints 41%
 - Chest pain #7 on list with 32%
 - 26% of patients with pain did nothing about it
 - 42% asked their CF center physician 3.5% asked PCP for help with pain
 - Affected QOL and every day activities

Chronic pain in cystic fibrosis

- 78 patients in a CF clinic in Boston with chronic pain 1984-93
- Chronic pain increased in the last 6 months of life
- Headache 55%, chest pain 65%, back pain 19%, abdominal pain 19%, limb pain 16%
- Chest pain – musculoskeletal, pleuritis, pneumothorax, rib fracture
- Pain therapy varied – NSAIDs, opiates, TCAs, epidural analgesia

Physiotherapy and massage to treat pain in CF

- 105 patients with CF in Victoria, Australia
- Assessed pain after one treatment “a combination of spinal joint and intercostal mobilization” “soft tissue therapy and remedial massage”
- 90% reported chronic pain
- Short lived reduction in pain intensity

Lee A, Holdsworth M et al. The immediate effect of musculoskeletal physiotherapy techniques and massage on pain and ease of breathing in adults with cystic fibrosis. *J Cystic Fibrosis*. 8(1);79-81:January 2009.

End of life care in CF

- End of life care differed compared to oncology patients
 - Many continued to receive antibiotics and other preventive and therapeutic therapy until death.
 - Many had blood draws for diagnostics on day of death
 - Out of 44 patients only 1 died at home on hospice

21 yr old male with uncontrolled chest pain – follow up

- Methadone did not seem to help
- Changed to oxymorphone ER also with little help
- Added Nortriptyline at hs
- What next?
- Three patients since December
 - 2 dead
 - 1 being evaluated for transplant

Therapeutic options for refractory pain

- The usual suspects
 - APAP, NSAIDs
 - Maximize dose of NSAIDs
 - Opiates
 - Adjunct medications
 - TCAs, anti-epileptic medications, steroids, topical therapy
 - Blocks, injection therapy
 - OMM?

Opiates at the end of life

- Opiates are safe at end of life and do not accelerate death
 - Morita T, Junichi T et al. Effects of high dose opioids and sedatives on survival in terminally ill cancer patients. J Pain Symptom Manage. April 2001;21(4):282-289.
 - Hawryluck LA, Harvey W et al. Consensus guidelines on analgesia and sedation in dying intensive care unit patients. BMC medical ethics 2002;3:3. www.biomedcentral.com/1472-6939/3/3
 - Mosenthal AC, Lee KF. Management of dyspnea at the end of life: relief for patients and surgeons. J Amer Coll Surgeons. Mar 2002;194(3):377-386

Other resources

- Palermo TM, Harrison D, Effect of disease related pain on the health related Quality of life of children and adolescents with cystic fibrosis. *Clinical J Pain*. 22(6) July / August 2006. 532-537.