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Alfred Health

INFECTION PREVENTION IN PEOPLE WITH CYSTIC FIBROSIS

- Nothing to declare

Overview

- Cystic fibrosis
  - Effects on the body
  - Medical management
- Infection prevention issues
- Emerging pathogens
- Revised US CF Foundation Guidelines 2013

Cystic Fibrosis (CF)

- Most common genetically acquired, life-shortening illness affecting young Australians
- 1 in 2,000 people CF disease
- 1 in 25 carry defective recessive CFTR gene

- CFTR protein is absent or not functioning properly, disturbing the salt/ water balance of epithelial fluid transport in the lung, pancreas and other organs

Cystic Fibrosis Federation Australia

Effects CF Disease

- Frequent pulmonary infections
  - Bronchiectasis
  - Reduce lung function
  - Resp failure
- Pancreatic insufficiency
  - Malnutrition and diabetes (25%)
- Liver disease (10%)
- Osteoporosis (40%)
- Male infertility

Life Expectancy

Year	Median Life expectancy
< 1960	Few months
1980's	17 years
2005	26 years
2016	41 years

UK Cystic Fibrosis Trust 2016

Reid D et al, *Changes in cystic fibrosis mortality in Australia, 1979-2005*. Med J Aust 2011;195:392-5

17th Annual Report
Australian Cystic Fibrosis Data Registry 2014

U.S National Patient Registry 2012.
Cystic Fibrosis Foundation

Progressive Lung Damage

- Excessive, thick lung mucous:
  - Perfect breeding ground for bacteria, fungi and viruses
- Cycle of inflammation and infection causes progressive lung tissue damage
  - Reducing quality of life
  - Early death

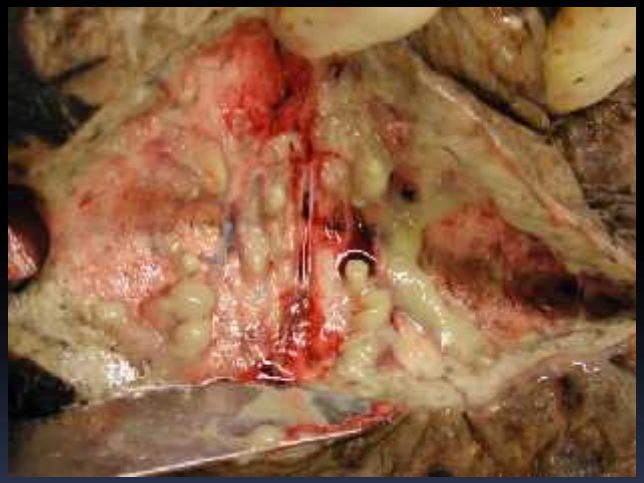
- Thick, sticky mucous overlying cilia

Pulmonary Treatment

- CF treatments have improved over the decades
- Potentiators and correctors for CFTR gene
- Most treatment is symptomatic
- Airway clearance
  - Autogenic drainage, PEEP
  - Nebulisers
    - Hypertonic saline
    - Mucolytics
  - Inhaled bronchodilators, steroids

Antibiotic/Healthcare Exposure

- 94% antibiotic treatment- exacerbations
  - Mostly empiric for Gm negatives
- > 69% adults inhaled abx
- Continuous oral abx
  - Very young children (53%)
  - Adolescents (41%)
- Severe exacerbations infection
  - 10-14 days abx treatment
  - 45% adults at least 1 episode/year -mean LOS 24 days



17th Annual Report 2014
Australian Cystic Fibrosis Data Registry

Lung Transplant

- Lung transplant considered as pulmonary failure progresses

Vs

- Immunosuppressive treatments
  - Increase chance of infections
  - Chance of organ rejection
- 5 year survival post Lung Tx currently 67%
  - CFTR gene not present in Tx lung
  - Existing colonising organisms can infect Tx lungs
  - CF disease progression in other organs

Stephenson L et al *Clinical and demographic factors associated with post–lung transplantation survival in individuals with cystic fibrosis*. The Journal of Heart and Lung Transplantation, 2015

Infection Prevention

Factors in pulmonary colonisation/ infection

- Thick sticky lung mucous medium for m/o growth
- Damaged lung tissue & Tx ⬆ risk of fungal infection
- Chronic rhinosinusitis
- Coughing, sputum clearance practices cont environment
- Frequent abx exposure
- Exposure to other CF patients clinic, hospital

Prevention of acquisition of particular m/o can be critical:

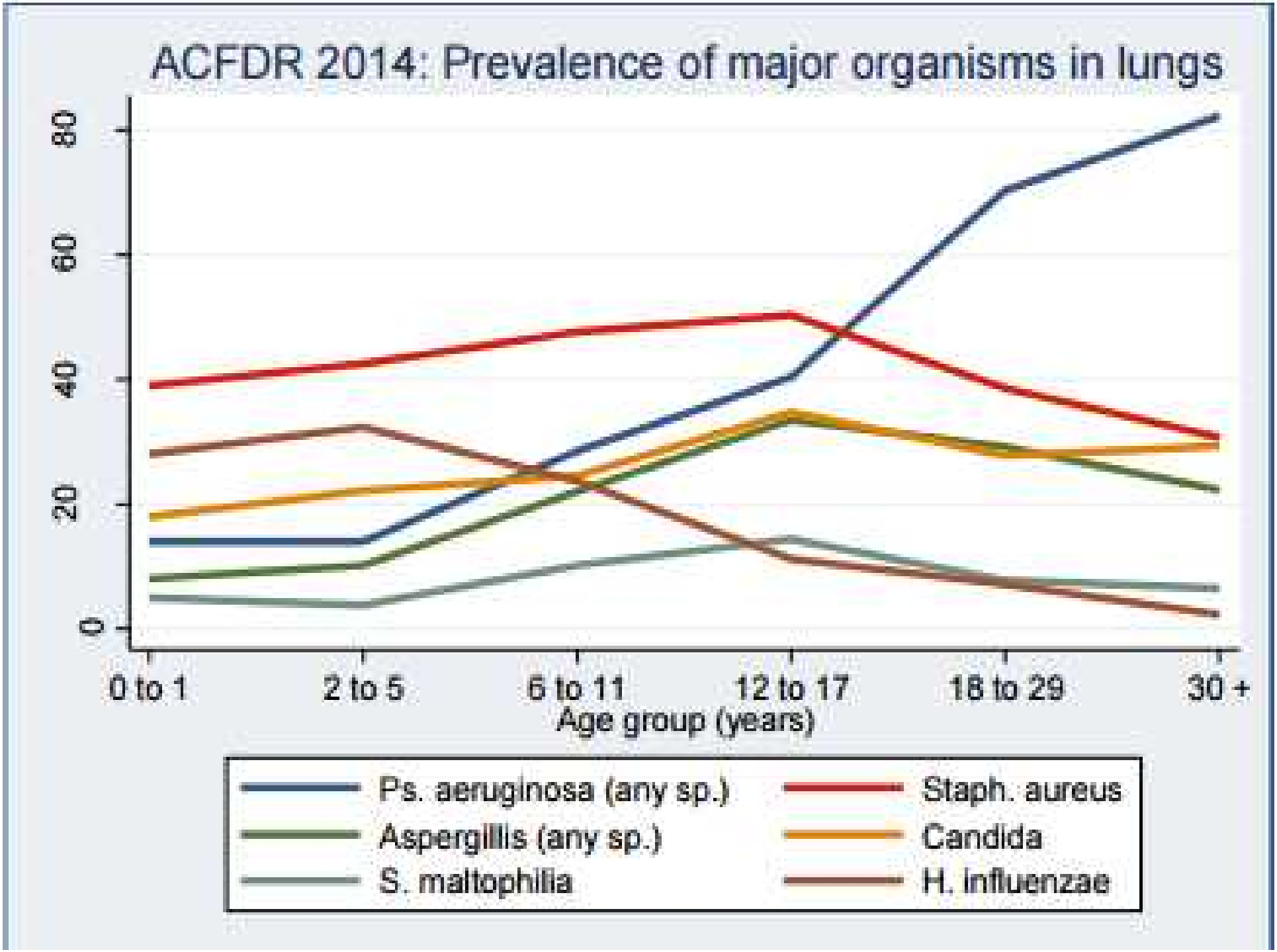
- Lung function
- Lung Tx consideration

Pulmonary microorganisms

- *Pseudomonas aeruginosa* (70% adults)
  - *matt* → *mucoid variant*
- *Burkholderia cepacia complex* (2.7%)
- *Aspergillus fumigatus* (22%), *Scedosporium* (esp lung Tx)
- *Stenotrophomonas maltophilia* (8.5%)
- *Achromobacter xylosoxidans*
- *Ralstonia spp*
- NTM, mostly MAC (13%)
- MRSA (6.5%)
- Respiratory viruses
  - Influenza, RSV, Picornavirus, HMPV, Parainfluenza

Colonisation/ infection generally increases with age except for MSSA/MRSA

Alfred Health data


ACFDR 2014: Prevalence of major organisms in lungs
0
20
40
60
80
0 to 1
2 to 5
6 to 11
12 to 17
18 to 29
30 +
Age group (years)
Ps. aeruginosa (any sp.)
Aspergillis (any sp.)
S. maltophilia
Staph. aureus
Candida
H. influenzae

Changes to prevalence over time

Stenotrophomonas maltophilia

- Improved micro identification
- Increasing patient longevity
- Antimicrobial selection pressure

Pseudomonas aeruginosa

Burkholderia cepacia complex

- Eradication strategies
- Improved infection prevention

Burkholderia cepacia complex: Leeds Regional Adult and Paediatric Cystic Fibrosis Units, St James's University Hospital, Leeds, UK.

IP Considerations

- M/O can be acquired from:
  - Non CF people and family (resp viruses)
  - Healthcare workers (resp viruses)
  - Other CF people
  - Natural environment (soil, organic matter, water)
  - Healthcare environment (sinks, surfaces)
  - Healthcare equipment (nebulisers, pulmonary function equipment)

  - Different Gm negs acquired different sources
    - *S. maltophilia*- environmentally acquired
    - *B. cepacia*- more evidence of transmission between CF

Consider Opportunistic Transmission

- Obligate transmission
  - Occurs under natural conditions
- Preferential transmission
  - One route is the usual route but another route has been described
- Opportunistic transmission
  - Pathogen usually transmitted by droplets
  - Can be transmitted by droplet nuclei under unusual environmental conditions

Roy et al, *Airborne transmission of communicable infection—the elusive pathway*. N Engl J Med 2004;350:1710– 1712.

IP Principles- General

- Early identification m/o and patient counselling
- Cough etiquette, management
- Spatial segregation from other CF people socially (unless live in same household)
  - No indoor CF camps, shared education programs
  - No shared fitness classes, prolonged car trips
  - No intimate contact, handshaking
  - No sharing eating and drinking utensils
  - No sharing respiratory equipment
  - Limit prolonged, repeated exposure to soil and organic matter

IP Principles- Healthcare

Outpatient:

- Clinic segregation (pathogen specific)
- Minimise time in common waiting rooms
- No sharing computers, toys
- Available cough cups with lids, tissues, waste receptacles
- Hand hygiene supervised on entry/ exit

Inpatient:

- Single room preference (or cohort with non CF)
- No bathroom sharing
- Cough cups with lids, respiratory therapy in room
- Dedicated equipment
- Gym sessions managed

HCWs with CF disease

- Receive counselling from CF care team:
  - Pathogen acquisition
  - Pathogen transmission
- Not care for others with CF
- Be assigned to care for patients on a case by case basis
  - Consider coughing
  - Resp secretion containment
  - PPE
  - Specific task duties

Emerging Pathogens: M abscessus

- Incidence ↑ worldwide
- Highly drug resistant
- Challenging to treat: 6-12 month course
- Previously absolute CI to Tx
- Rapid clinical deterioration and poor outcome after Tx
- *M. abscessus subsp massiliense* ? more infectious
- Evidence for and against person to person transmission

M abscessus

- UK: 168 isolates from 31 patients at Papworth Hospital -genome sequencing
  - *2* clusters near identical *M abscessus subsp massiliense*
  - Epidemiological links between patients of the same clusters in clinics and wards
  - Patient residences were not geographically grouped
  - Extensive hospital environmental sampling- neg

Bryant J et al, Whole-genome sequencing to identify transmission of *Mycobacterium abscessus* between patients with cystic fibrosis: a retrospective cohort study. *Lancet* 2013;381:1551–1560.

M abscessus

- UK: 27 isolates in 20 paediatric CF patients
  - Most strains were genetically unrelated
  - Two groups of closely related strains
    - The first cluster from 2 siblings
    - The second cluster was of 3 strains from patients with no epidemiological links

- US: . Olivier typed NTM in US hospitals and did not find evidence of within-centre clustering

Harris K et al,. Molecular fingerprinting of *Mycobacterium abscessus* strains in a cohort of peadiatric cystic fibrosis patients. *J Clin Microbiol* 2012;50:1758–1761.

Olivier K et al, Nontuberculous Mycobacteria: multicentre Prevalence Study in Cystic fibrosis AJRCCM 2002;167. No 6

M abscessus

- International guidelines
  - Single room with door closed, "air" room 1 hour post use → negative ventilation, contact precautions
- Major Aust centres:
  - IP single room, some droplet and contact precautions, Separate clinic
- National Dept Health TB guidelines released Sept 16
  - CF patients with *M. abscessus* complex requirements should be determined by local experts
- Alfred
  - Standard Precautions → single room, separate clinic, gym

Emerging Pathogens: Epidemic Burkholderia strains

- B. multivorans, dolosa, cenocepacia especially

  Cepacia Syndrome -fulminating pneumonic illness
- UK: 2 most predominant strains
  - B. multivorans & B. cenocepacia
  - Epidemic spread: 87.9% B. cenocepacia single strain type
- US: Predominant strain
  - B. multivorans
- Australia
  - B. cenocepacia is rare, relative CI to Tx
  - Most highly transmissible strain

Jones et al, *Burkholderia cenocepacia and Burkholderia multivorans: influence on survival in cystic fibrosis*. Thorax 2004;59:948-951

B.Cenocepacia

- International guidelines
  - Single room, contact precautions (organism specific) → Single room, contact precautions (regardless of sputum cultures)
- Alfred
  - Single room, door closed, mask patient out of room when on CF ward, or cared for on a non CF ward
  - Segregate gym and clinic

Infection Prevention and Control Guideline for Cystic Fibrosis 2013

Volunteer committee

- 4 ID physicians
- 4 Resp physicians
- 4 RNs
- 1 Resp therapist
- 1 ICP
- 3 parents CF children
- 1 adult with CF
- 1 social worker
- 2 CF foundation staff

- Systematic review
- 2 independent reviewers screened search results
- New recommendations:
  - >80% approval
  - Anonymous vote

Infect Control Hosp Epidemiol 2014; 35(S1): S1-S67

Local contentious issues

- Regardless of resp culture results:
  - Contact Precautions for all persons with CF
  - All persons with CF should wear a surgical mask when in a healthcare setting (not in clinic room or hospital room)
  - Segregate all CF people in clinics
- Potential role of droplet nuclei in CF pathogen transmission eg NTM

- Thank you