CF MANAGEMENT – SUMMARY

CF is a multisystem, chronic disease that is best managed within the framework of a multidisciplinary team – don't be slow to seek the expertise of other health professionals in managing these patients

Infective exacerbations

Key assessment points

- FEV1 (relate to "recent best"/ "RB")
- Weight, body-mass index/ BMI (relate to "RB")
- symptoms (well-being, sputum production and purulence)
- Progress during treatment is indicated by improvement in these 3 parameters, so measure weight and FEV1 at least twice weekly

Key management points:

- Reduce pulmonary bacterial load - IV antibiotics (use appropriate antibiotics directed at organisms that the subject is colonised with, usually 2 anti-pseudomonal antibiotics of which one should be an aminoglycoside – eg tobramycin and cefipime)
  - while we consider the sensitivity patterns of *Pseudomonas aeruginosa* detected on sputum culture and sensitivity testing, clinical response is often not consistent with the sensitivity pattern seen due to substantial variability in sensitivity patterns
- Facilitate optimal sputum clearance – chest physiotherapy (at least twice daily)
  - consider nebulised salbutamol, potentially nebulised hypertonic saline
  - encourage exercise as form of chest physio ?visit gym during admission
- Optimise nutrition – additional nutritional supplementation must be prescribed or at least considered; subjects with poor baseline nutrition despite oral supplementation must be considered for more aggressive maintenance nutrition (eg NGT/ gastrostomy buttons)

Details of management:

- aminoglycoside monitoring – 1 hour post and 8-10 hours post levels, performed following the 3rd dose and thence depending upon initial levels
- CXR – not necessary with each admission unless suggestion of pneumonic or other process (eg pneumothorax) involved
- bloods – FBC, U/E, Mg++, LFT's, CRP should be performed with initial drug levels
- sputum culture – performed on admission

Additional investigations:

- only if "annual review" bloods/ investigations indicated during admission (these can be obtained form "annual review" sheet

Procedures (ports, gastrostomies, sinus surgery, etc):

Principles

- Ensure pulmonary function is adequate for procedure; in most case, a period of intravenous antibiotic therapy pre-procedure is appropriate
- Ensure appropriate liason with the associated team (eg vascular surgery for ports, ENT for sinuses, etc) so that miscommunication and mishaps are limited

Follow-up

- all CF patients should be seen in an outpatient "setting" every 3 months for monitoring, etc, although this does not need to be physically within the outpatients department

Appropriate infection control is imperative for all subjects with CF. Patients with CF should not be in the position to "associate" with other patients who are colonised with different organisms (eg subjects colonised only with Staphylococcus aureus should not be reviewed in conjunction with patients with Pseudomonas or Burkholderia cepacia, etc). Thus, patient review in clinics, etc, should be grouped according to colonising organisms.

Infection control measures must include attention to handwashing between all CF subjects.