Cystic Fibrosis and nursing

Kristine Colpaert
CF Nurse, UZ Leuven Belgium
Teamwork
Education and knowledge
Tasks
Long term care for children and adults with CF
Cystic Fibrosis (CF) = Complex disease

- Disease progression
- Severe psychosocial impact
- Multidisciplinary team
TEAMWORK IN CARE

- Doctor
- Nurse
- Physiotherapist
- Psychologist
- Social worker
- Dietician
- Secretary

CF patient
Cystic Fibrosis / course of disease
Genetics
Medication
Physiotherapy
Nutrition
Pathogens / infection controle
Additional diagnoses (diabetes – osteoporosis)
New insights / developments in CF care
**Outpatient clinic**

vital signs, bloodtests, pulmonary function tests, chest x-ray etc

**Hospitalisation**

diagnosis, exacerbation, pre-transplant investigation...
at hospitalisation: plan for discharge, home iv therapy

**Post – clinic patient review**

Patient contact by mail/phone e.g. change in therapy, additional tests

Support, counseling and referring to other team members/healthcare workers
Diagnosis

Information about CF

Introducing team to patient / parents

Nebulizer - use of / maintenance

Hygiene

Medication - indication, how to take and preserve
Continuous care

Support in understanding CF and how to insert therapy in daily life

Adherence

Coping with CF diagnosis
Transition from pediatric to adult care

Dependency of parents
Farewell to pediatrician
Future?

Self care & independency
Other hospital / CF team

Pride & satisfaction
Care specific for CF-adult
Sexuality – Fertility – Pregnancy
Additional Diagnosis
Increasing disease severity
Transplantation
End of life
Sexuality & Fertility

- Stress
- Malnutrition
- Tough mucus
- Irregular or anovulatory menstrual cycle

Less fertile?

Congenital Bilateral Absence of Vas Deferens

Obstructive azoospermia 98%
Health care professionals should initiate discussions with patients and their parents/partners.

Provide information during early adolescence.

Repeat information over time, perhaps at set stages within child/adolescents development.

Parents need help to fulfill their role to inform and educate their children.

Possible misconceptions need permanent attention, e.g., men mixing up infertility and impotence.
Fertility and Pregnancy

- CF inheritance
- Burden on healthy partner when caring for ill parent
- Ethical aspects of severe illness / untimely death of CF parent
- Child caring for ill parent
- **ICSI** IntraCytoplasmic Sperm Injection

**Female:**
- Medical condition
- Invasive treatment
- Risk to mother and child

**Male:**
- Treatment female partner
Additional diagnosis – Diabetes

**Screening** from age 12

30% of CF adults

Diagnosis - a **shock**

Poor **adherence** to insulin

**Diet** measures
Increasing disease severity

- Increasing daily therapy / burden
- School / work
- Friends and family life
- Anxiety
- Adaptation to restricted possibilities
- Future?
Transplantation

Increasing illness severity: infections, complications

Choice of the patient

Preparing for transplantation: time to transplantation, anxiety, life after, financiel aspects

After: rehabilitation, medication, hygiene, nutrition, fear for infection / rejection.
End of life

Not delay talk with patient and family about possibilities and expectations

Importance of good communication among healthcare workers

Respect for patients’ choice

Organising comfort care

Care for bereaved relatives
IS MULTI TASKING .... With TLC
(Tender Loving Care)