Clinical Care Guidelines for Cystic Fibrosis

Outpatient and Inpatient

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Formation of the Clinical Care Guidelines

- The Cystic Fibrosis Care Workgroup Association was founded in 1997 in the context of the 1.German Cystic Fibrosis Conference in Lahnstein
- The foundation of the workgroup allowed for the first time nursing staff to be included in the continued educational program of the multidisciplinary team working in the field of CF
- Since 1997 nursing staff meets in Germany, Austria and Switzerland twice a year, exchanging experiences and to further develop their skills

Formation of the Clinical Care Guidelines

Tasks of the care workgroup:

- to improve the support of CF-patients by their family members
- to professionalize and scientifically further develop CF-spezific care

To create Clinical Care Guidelines with the goal of standardized caring guidelines

- to enable an interdisciplinary exchange
- to stabilize multidisciplinary collaboration
- to ensure national and international conference attendance
- to offer public statements regarding care guidelines
- to provide advice within the Mukoviszidose e.V.

Formation of the Clinical Care Guidelines

- Result of the first exchange of experiences

The quality of care depends heavily on the knowledge of caregivers, relatives and the patients themselves

- Creation of the first clinical care guideline in 1998

Compilation of CF-specific care measures in accordance with scientific insight and in close connection with securing high quality standards of care.

What is the goal of the Clinical Care Guidelines?

Clinical Care Guidelines and its goals

- Effective, high quality, good and coherent working standards
- Improved patient care
- Quality standards of care
- Guidelines for new employees
- Accompanying study material for patients and their relatives
- Highest possible treatment security in relations to different forms of therapy
- Current care guidelines in accordance with scientific literature

Clinical Care Guidelines and its goals

- Emphasize the professional conduct in care
- Independence of care givers in multidisciplinary teams
- Emphasizing the growing importance of the area of expertise.

Structure of the Clinical Care Guidelines

Structure of the Clinical Care Guidelines

- As workbook
- Without numerical structure or page count
- Colored segmentation of topics
- Introduction of by the Chair and medical board of advisors
- detailed description of the symptoms

Structure of the Clinical Care Guidelines

The most important topics were split into 7 chapters:

- Hygiene
- Nutrition
- Inhalation therapy
- Ambulant intravenous antibiotics therapy
- Stationary intravenous antibiotics therapy
- Oxygen treatment
- Non-invasive ventilation

Structure of the Clinical Care Guidelines:

Example of the stationary inhalation therapy

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Clinical care guideline: Inpatient inhalation therapy using an aerosol device/wall mounted automatic dispenser

Inhalation is a respiratory physical therapy procedure used to treat pulmonary disorders. The procedure employs antibiotic, secretolytic, antiinflammatory and bronchodilatory drugs.

Care objectives:	Nursing interventions:
Avoid infections:	use aseptic technique
Promotion of gas exchange:	proper inhalation technique and medication sequence
Administration of pharmaceuticals:	proper inhalation technique, functional aerosol device and nebuliser component
Avoid incompatibilities:	mixing of medications only on the order of a physician
Mobilisation of bronchial secretion:	Assisted expectoration
Independent inhalation:	Explain the procedure and training

Material:
Hand disinfectant
Inhalation solution according to prescription
Nebuliser components
Mask or mouthpiece with exhalation valve, nose clip and baby bend as needed
Compressor or wall mounted dispenser
Personal vaporiser or egg boiler as needed
Sterile disposable syringes, fill needles
Sputum cup, cleaning cloth
2 fresh cotton cloths washed at 60°C and ironed
Disposable gloves

Procedure:

1. Educate and instruct the patient appropriate to his/her age as well as the parents
2. Nasal inspection and cleaning of the nose if necessary
3. Hygienic hand disinfection
4. Assemble the dry nebuliser components and fill the inhalation solution into the medication cup
5. Inhalation is performed in a sitting position; bring infants into an elevated upper body position
6. Put on the mask without applying pressure but ensure it is tightly sealed or grip the mouthpiece with the teeth and tightly enclose with the lips
7. Instruct the patient to use proper rhythm of breathing:
   deep inhalation – breathing pause (3 seconds) – exhalation through mouth with pursed lips or slowly through the nose, discontinue nebulising during breathing pause and exhalation as needed
8. Breathing rhythm: slow and deep
9. Following mask inhalation, wipe moisture from the facial skin
10. Help the patient expectorate sputum

Processing of the used inhalation utensils

Procedure:

1. Hygienic hand disinfection
2. Rinse out the medication cup, mask and mouthpiece after each inhalation with warm running water
3. Thoroughly dry the accessories and envelope in a clean cloth
5. Replace the nebuliser components after each inhalation
6. Hand over the nebuliser components after each inhalation to the sterilisation unit or disinfect in the patient's own vaporiser (follow the manufacturer instructions; 70°C, water vapour for 10 minutes)
7. The nebuliser components remain with the patient
8. Use the compressor to blow out the connecting tube after each inhalation
9. Daily surface disinfection of the compressor; perform additional surface disinfection if the compressor is also used for other patients (including all cables and additional accessories)
14. The devices must be checked according to manufacturer's instructions (e.g. in medical supply stores)
15. Year pack prescription on an annual basis (nebuliser, filter, connecting tube)

Note:

- ⇨ Expectorated sputum is generally to be considered and treated as infectious and requires corresponding hygienic measures
- ⇨ Sufficient supply of liquid is important in facilitating expectoration.
- ⇨ Do not perform inhalations directly after administration of food (expectoration of secretions, abdominal press may lead to vomiting)
- ⇨ Arrange the inhalation sessions based on the physical therapy protocol
- ⇨ Do not perform inhalations in infants during sleep unless the patient's strong resistance makes inhalation impossible when awake
- ⇨ Inhalations are performed before or after physical therapy, depending on the objective
- ⇨ Inhalations must be performed as directed by a physician
- ⇨ The most therapeutically effective aerosol particle size is 2-5 µm
- ⇨ The nebuliser components must be completely dry prior to the next inhalation
- ⇨ If antibiotics/antimycotics are inhaled: use filter

Improper breathing technique may lead to hyperventilation.

Example for medication sequence: first inhale drugs that dilate the bronchi, then inhale mucus-liquefying inhalation solutions and finally inhale the antibiotic.

Inhalation may be combined with respiratory aids (e.g. LC Cornet). This shortens respiratory therapy.

The various nebulisers must be used based on the age of the patient and the drug to be inhaled.

Effecte/side effects:

Beta-2-mimetics	Bronchodilating effect, may cause tachycardias
Anticholinergics	bronchodilating effect, causes oral dryness
Antibiotics	antibiotic effect, may cause irritating cough and tightness, risk of candidiasis
Corticosteroids	antiinflammatory effect, risk of candidiasis
Hypertonic solutions	mucus-liquefying effect, may cause bronchospasms
Pulmozyme®	mucus-liquefying effect which may increase expectoration of mucus, may cause hoarseness, medication must be stored in the refrigerator **CAUTION:** if predisposition to hemoptysis exists

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Miscibility of inhalation solutions in the nebuliser

Miscibility	Tobramycin	Colistin	Salbutamol	Ipratropium bromide	Dornase alfa	Acetyl cysteine	Isotonic saline solution (0.9%)	Hypertonic saline solution (6%)
Tobramycin		No	Yes	Yes	No	No	Yes	No
Colistin	No		Yes	No	No	No	Yes	No
Salbutamol	Yes	Yes		Yes	No	Yes	Yes	No
Ipratropium bromide	Yes	No	Yes		No	No	Yes	No
Dornase alfa	No	No	No	No		No	No	No
Acetyl cysteine	No	No	Yes	No	No		No	No
Isotonic saline solution (0.9%)	Yes	Yes	Yes	Yes	No	No		Yes
Hypertonic saline solution (6%)	No	No	No	No	No	No	Yes	

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Literature:

Kamin W., Schwabe A., and Krämer. Inhalation solutions-Which one are allowed to be mixed? J.Cyst.Fibros 2006;5:205-213

Schwabe A., Kamin W., and Krämer. Physik.-chem. Kompatibilität von Inhalationslösungen in Verneblersystemen, Krankenhauspharmazie 2005;26:119-126

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What use has the regular editing of the Clinical Care Guidelines?

Changes

- Guidelines need to be revised and updated regularly
- The improvement of therapy requires new versions of supporting material
- Occurrences of secondary illnesses do to a continuous increase in life expectancy such as CFRD, Osteoporosis

How were the new Clinical Care Guidelines developed?

Development

- On the basis of latest care standards
- through highly qualified nursing staff
- literature review
- exchange and collaboration within the working group
- review by medical advisory board

How are the Clinical Care Guidelines distributed?

Distribution

- Through the working group members within their organizations
- as being part of the official study material for becoming a certified CF-trainer
- quality trait of certified CF-centres is the employment or training of specialized CF- nursing staff
- certified CF-centres need to work in accordance with the clinical care guidelines
- the guidelines are referenced to within the context of the german CF-congress Würzburg
- Mukoviszidose e.V. website

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Thank you very much for your attention