

Maintaining Nursing Care Quality

—Clinical Care Guidelines for Cystic Fibrosis: Outpatient and Inpatient

Melanie Koeller¹, Cornelia Meyer²

¹Fachklinik Satteldüne für Kinder und Jugendliche, Nebel/Amrum, Germany

²College of Medicine, University of Tübingen, Tübingen, Germany

Email: meyer.cornelia@gmx.net

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Abstract

The Care Committee of the Mukoviszidose e.V., headed by Cornelia Meyer, has drawn up a completely revised version of the German Guidelines to Care in Cystic Fibrosis. It addresses all nursing staff concerning children, adolescents or adults with cystic fibrosis (CF) either in an inpatient and outpatient setting or in rehab clinics. The guidelines evolve along with the increased life expectancy of cystic fibrosis patients and improved quality of life which is described by CF patients and can be observed by the therapeutic CF team. For example, the treatment of secondary diseases like CFDR (CF related diabetes) which comes along with an increased life expectancy has been added over the years. It is essential that within such a complex disease pattern, professional experience of care staff who worked in specialized CF care centers (50+ patients per year) for many years has to be transferred to other nursing staff or beginners. This is especially important in the context of the heterogeneous characteristics of this disease. Often it can be seen that the quality of the care for CF patients depends on the know-how and professional experience of the interdisciplinary CF therapists team. Moreover, it depends on the knowledge which the patient has gathered by himself by experience or learning. And, of course, it depends also on the knowledge of the relatives especially parents or partner respectively.

Keywords

Cystic Fibrosis, Clinical Nursing, Guidelines

1. Introduction

Cystic fibrosis (CF) is one of the most common life-shortening inherited diseases. In Germany about 8000 people are affected [1]. Even though there is still no cure for the disease, improvement in symptomatic therapy has enhanced life expectancy to around

40 years. Cystic fibrosis is caused by the mutation of a gene called cystic fibrosis transmembrane conductance regulator (CFTR). This gene defect results in the production of thick mucus in the internal organs causing inflammation, frequent lung infections and digestive problems. Long-term issues include difficulty breathing and coughing up mucus as a result of frequent lung infections. Other signs and symptoms include sinus infections, poor growth, fatty stool, clubbing of the fingers and toes, and infertility in males, among others. Different people may have different degrees of symptoms. Lung transplantation may be an option if lung function continues to worsen. Lung problems are responsible for death in 80% of people with cystic fibrosis. Although technically a rare disease, CF is ranked as one of the most widespread life-shortening genetic diseases [2].

Life expectancy of CF patients has been doubled to nearly 40 years of age within the last 20 years. This is mostly based on newly developed medication and a profound knowledge of the physiological and chemical processes in the human CF cell; furthermore, through dense treatment within interdisciplinary teams like medicine, care, physical therapy, sports therapy, psychotherapy, nutrition therapy and social educators; and furthermore through the empowerment of the patient him-/herself. An important key issue of the therapy is the education and support of the autonomy of the patient aside from medication management, physical therapy and sports.

Patients' life expectancy has been steadily increased over the years (and hopefully will do in future), but causes simultaneously that the patient and the nursing staff have to face new challenges. Nowadays, to live with cystic fibrosis still means to maintain a disciplined daily lifestyle for more than 40 years, maintain motivation and handle the disease in a salutogenesis way by strengthening the sense of coherence [3]. Despite all the positive developments we still see difficult ethiopathology in which the CF patient still dies, in some cases as a child, or its survival depends on the last line of treatment, the lung transplantation. This holds also for patients in the early teenage years. This revised version of the guidelines offers updated and helpful support for optimal patient care. The guidelines show in a practical manner which benefit might arise by using it for the caring of cystic fibrosis patients and which advantages come along for the other departments.

2. Guidelines in Cystic Fibrosis

The guidelines, as a kind of handbook, aim at comprehensive support for all those involved in CF nursing for children and adults. The guidelines address primarily nursing professionals, close relatives and all others concerned. The 5th revision of the German Guidelines to Care in Cystic Fibrosis are a loose-leaf edition. The page numbers and numerical structure have intentionally been left out. This allows for uncomplicated additions of updates. Individual subject matters are highlighted. The authors have carefully compiled cystic fibrosis specific measures of action and presented them after scientific evaluation and emphasis on quality and safety. Strong emphasis is also taken on appreciation of the complex work of nursing. The new edition of the guidelines offers comprehensive up-to-date support in nearly all caring situations.

“Furthermore, patient care is facing increasing challenges because the patients are fortunately increasing in age but also becoming highly complex patients at the same time. These patients may develop additional complications such as osteoporosis, high blood pressure, depressive episodes, pain, diabetes mellitus and many other disorders which patient care must be able to handle. Adaption to this new situation is necessary for a good patient care and appropriate specialised training have to be implemented.” (Dr. C. Schwarz, Medical Advisor, Charité Berlin, Preface [4]).

The preface is followed by a detailed description of the CF disease pattern [5]. This is particularly helpful for beginners who have been given a complete overview over the complexity of the disease. The following seven chapters contain the central themes:

- Hygiene.
- Nutrition.
- Inhalation treatment.
- Outpatient IV antibiotic treatment.
- Inpatient IV antibiotic treatment.
- Long-term oxygen therapy.
- Noninvasive ventilation.

Each chapter contains aspects being used practically or are of great importance, especially in combination with hygiene. The main subject of hygiene had been excluded from the other chapters and had been included in a separate chapter. Specific hygiene subjects, for example, nebuliser sterilisation, can be found in the corresponding chapter. The topic of hygiene will become more and more an important issue throughout the progression of the disease and it is particularly important for the hospital setting especially for disinfection reasons and in cases of patient isolation. In future more work must be done to cover the rapidly increasing knowledge due to germs and drug resistance. Further, the guidelines describes CF-relevant lung bacteria like *Pseudomonas aeruginosa* and how to deal with it in the clinical setting. Also, procedures and processes of bacterial isolation are being described together with up-to-date references to multiresistant *Pseudomonas* sp. (MRGN) [6] [7].

3. Using the Guidelines

By creating a fictional case report we will demonstrate how the guidelines are to be used. We picked out the issue of nutrition which is another key issue in the treatment of CF [8]. In a cross sectional and longitudinal analysis from Steinkamp *et al.* (2002) there find evidence for the hypothesis that a near normal weight is associated with a better lung function in CF [9]. In consequence a higher life expectancy can be assured.

Julia, a nine years old child with CF, is receiving inpatient treatment. Her weight is within the 10th percentile and her height is within the 25th percentile. She has had recurrent infections which has caused the hospital stay. She is concerned about her diagnosis and would like most to “get rid of CF”. Her mother cooks additionally hypercaloric meals and supervises the enzyme dosage over day in the hospital. The moth-

er-daughter interaction is limited to Julia's treatment (eating, inhalation, physical therapy). The attending nursing staff is aware that the mother is well-informed and that she "only want the best for Julia".

What do the guidelines say? In this case the guidelines (Ch. 2: Nutrition) emphasize on the encouragement of the patients' self-reliance. This should be in Julias' own interest. Considering her age she should be able to know about the correct dosage of enzymes by estimating how much fat the food contains. Also, she should carry the enzymes by herself and disperses them autonomously over the meals. She should be able or should be encouraged to ask questions to determine the correct dosage of the enzymes. The guidelines offer helpful informations for the nursing staff about correct dosage or derivation of an adequate formulas depending on the meals amount of fat. As she is still a growing child she requires a higher energy density of the food. Assistance for choosing the right food items is been given by using the "nutrition cube" as recommended by the study group "Nutrition" of the Mukoviszidose e.V. A nutrition protocol should be drawn up regularly during the inpatient stay to check for Julias' adequate energy supply. This have to be done by the nursing staff. Furthermore, the nursing staff suggest recommendations which raise the appetite level of the patient and finally discusses them with Julia and her mother. Further, the possibility to get additionally calories by using hypercaloric meals has to be offered. A daily blood sugar profile might also be of use to elucidate why Julia has increased frequency of infection and persisting malnutrition. For an extremely reduced nutrition status the guidelines suggest the use of a stomach tube (PEG). In future, Julias' height and weight need to be continuously monitored and readjusted. At least, the contact to an educated dietician have to be established.

It is important to realize that there are things Julia can handle by herself (by the mean of salutogenesis) in order "to move towards health", *i.e.* to avoid the "get rid of cf" way of thinking. She have to take responsibility for her own disease and not despair due to the diagnosis. This is a problem especially evolving during puberty. As mentioned before, CF means a disciplinary lifestyle—forever! Therefore it is important to get into action as soon as possible and to counteract negative developments in time.

4. Conclusions

Since the share of inpatient stay has drastically decreased, it has become increasingly important that the possibility for the nursing staff to get in close contact with the patient is upheld in order to have a positive effect on the patient and his/her relatives. The consultation of the patient has to start before the patient starts to ask questions. Especially for chronic diseases, it holds the fact that the patient and their relatives have become a kind of specialist for CF and this has in any case to be taken into consideration. But this does not implicitly mean; however, there is no room for improvement or there are no possibilities for improvements within the individual daily therapy management. The guidelines are supposed to give us also a sense of safety by dealing with CF.

Since the first edition of the German Guidelines to Care in Cystic Fibrosis which had

been published in 1998 the guidelines are being revised regularly. In 2015 all chapters have been revised completely by several authors who have worked with CF patients for several years. New developments in treating CF, inpatient and outpatient, have been taken into account. For example, especially the chapter of hygiene (MRGN: multi resistant gram negative bacteria), IV antibiotic treatment (picc line catheter: Peripher Inserted Central Catheter), inhalation treatment (new hygienic standards) and nutrition (CFRD: CF related diabetes) have been updated. The 5th completely revised and amended version of the Clinical Care Guidelines for Cystic Fibrosis is available on request by Mukoviszidose e.V. (Adress: In den Daunen 6, 53117 Bonn-Germany. E-mail: info@muko.info).

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