

CYSTIC FIBROSIS AND OTHER PULMONARY PATHOLOGIES

AUTHOR: Eva Barón Castaño

Physiotherapist. Collegiate Member: 3821 . Oficial Physiotherapist Association of La Comunidad de Madrid. Collaborator of the Respiratory and Geriatrics Commission of the Association

INTRODUCTION

Cystic fibrosis or mucoviscidosis is a chronic hereditary and generalized illness which affects the exocrine glands, specially pancreas and the bronchial tree. In this essay, I am going to focus on its respiratory affection due to its significance in respiratory physiotherapy treatment.

- -In the pancreas, an obstruction in the pancreatic duct with amorphous eosinophil concretions with the following pancreatic enzyme deficiency is produced (digestion, fat absorption, etc. are affected), electrolytes level abnormally high with sweating and, finally, cirrhosis.
- -In the lungs, it causes a chronic pulmonary illness. There is an increase in secretions and in their viscosity, what produces obstruction in the airways, mechanical ventilation disorders, and it may become worse if a pneumonia, bronchitis, bronchiectasis, emphysema, extended fibrosis, etc. do appear. These illnesses may degenerate into a pulmonary hypertension and a right ventricular failure in severe cases.

A reduction in the exercise tolerance is also characteristic. The clubbing fingers is also an important discovery.

DX TESTS

In the pulmonary auscultation, it may show strong rales and rhonchus.

The thorax radiography is abnormal in the beginning of the illness, showing cystic changes, fibrotic areas. The previously mentioned health history is useful.

PULMONARY FUNCTION

The first thing to appear is the abnormal distribution in ventilation and an increase in the O₂ difference in the arterial alveolus.

Function tests in the small airways as, for example, the analysis of the speed flow with low pulmonary volumes, allow to detect the illness in the beginning.

There is a reduction in the FEV₁ and in the FEF_{25-75%}, which does not respond to bronchodilators. The RV and the FRC are increased and there may be elasticity loss with the following compliance.

KEY WORDS

Hereditary, obstruction, generalized, palliative treatment, respiratory physiotherapy.

MEDICAL TREATMENT

The person is born without the injury, which starts to show up from that moment. As we have previously said, gland hypersecretion colonizes with bacteria. In this case, the administration of antibiotics with bronchodilators is advised, whenever there is bronchospasm.

- Capacity to recover its normal shape when the pressure is removed without showing desorganization or expression of the capacity measure to do it (unity of pressure change x unity of volume change)
- Capacity of distension

PHYSIOTHERAPY TREATMENT

• OBJECTIVES:

Removal of secretions

To prevent deformities and imbalances between inspiration and expiration muscles.

Improve life quality.

• TREATMENT:

A) Removal of secretions

We do the auscultation to know where the secretions are located. If there are not areas which are affected by

their drainage, the auscultation must be done from the basal to the medial areas.

We won't work in an inclined position whenever there is right-heart failure.

Before the drainage, we will do aerosol-therapy, inhalers and bronchodilators. If the patient takes antibiotics, it will be done after the drainage.

It is also important to do it before the meals, and never immediately after.

If the secretion removal is difficult, we will use intermittent positive pressure ventilation during a maximum of 4 to 5 days and no more than 10cm de H₂O.

• **Techniques used:**

EDIC:

Technique which is used to remove secretions that are found in small caliber and peripheral airways.

We place the patient in lateral recumbent position, with the damaged lung above the position where the gravity affects the most the injured lobe.

We ask the patient to do a continuous expiration followed by a high volume inspiration and low flow, some seconds in apnea and, during expiration, he should keep his lips pursed. The physiotherapist brings towards the front part when the affected is the posterior lobe (and viceversa) whilst the patient does the inspiration. This technique combines ventilation and position.

ELTGOL:

Expiration –slow –total-glottis-open-infralateral
Technique which is used to remove secretions from medium caliber airways.

We place the damaged lung infralaterally.

We ask the patiente to do a normal inspiration, followed by the most slow possible expiration, keeping the glottis opened.

FET

Forzed expiration technique.

We ask the patient to do an inspiration followed by an expiration at high flow and low volume.

For FET, there is a protocole which is used in order to prevent risk situations:

1-Respiration at rest during one minute.

2-Thoracic expansion: ventilations focused in the 2 areas where the secretions are concentrated or with the aim to provide with the maximum air possible the area to work.

3-FET: three continuous as maximum.

When its done, the protocole must be repeated from the first point until all the secretions are removed.

We should applying FET in lateral recumbent position.

COUGH

It must be done like the FET, but in between inspiration and expiration, we introduce some seconds in apnea followed by a "cough stroke", leaning towards the front.

As it has been observed, I do not use postural drainage due to several reasons which have been scientifically proven:

-These positions have been deduced from a theoretical orientation (only a 10% of the people agree)

-The bronchial tree moves in inspiration-expiration in the three dimensions in space.

-Gravity acts in the bodies according to their mass, which is very small in the bronchii, mostly of small and medium caliber airways. Besides, these secretions have big adhesion force.

-It has been demonstrated an increase in the infralateral lung after the pulmonary drainage.

B) To prevent deformities and diaphragmatic reeducation:

We show the correct use of diaphragm through directed ventilation and correct costal respiration, and we will potentiate the expiratory abdominal muscles.

CONCLUSION

Cystic fibrosis is an illness which may be suffered within the 2 to 4 months in infants, and today with the treatment they may overcome 20 years. The treatment is palliative, but may help to improve life prognosis, as well as its quantity and quality.

Since it is a chronic illness, it is important the work of medical professionals, without forgetting parent collaboration.

Physiotherapy is becoming more important nowadays, because it may mean a dramatic improvement in many respiratory pathologies, like for example cystic fibrosis. And I think that is important to let people know that we can help them through information.

BIBLIOGRAPHY

- Dorland- Diccionario Médico –ED Mc Graw Hill. Interamericana.
- Harrison-Manual de Medicina -ed Mc Graw Hill. Interamericana.
- West “Fisiopatología Pulmonar”-ed panamericana.
- West “Fisiología Respiratoria”- ED panamericana.
- H. Rouviere “Anatomía Humana”-Ed Masson.
- Dr. Helmut Keudel “ Enfermedades infantiles”- ED Everest
- Review article à “EFFICACY OF DIAPHRAGMATIC BREATHING IN PERSONS WITH CHONIC OBSTRUCTIVE PULMONARY DISEASE “ Lawrence P. Cahalin, MA,PT, CCS; Malinda Braga, MSPT; Yoshimi Matsuo, PT; Edgar D. Hernandez, PT.