

**ADDENDUM TO THE RECOMMENDATIONS FOR XLA
RESPIRATORY COMPLICATIONS IN HUMORAL IMMUNE DEFICIENCY**

RECOMMENDATIONS FOR DIAGNOSIS AND TREATMENT

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AIM

The diagnostic protocol of respiratory complications in humoral immune deficiency is an addendum to the previous recommendations defining the diagnostic criteria and treatment recommendations for X-linked agammaglobulinaemia (XLA) and common variable immunodeficiency (CVID).

The present document stems from the need for an in-depth assessment of respiratory disorders which are currently the main complication encountered in these immunodeficiencies despite IVIG replacement therapy.

The aims of the protocol are to:

- define the **diagnostic criteria** for chronic upper and lower airway complications;
- assess the degree of **impairment at the time of patient enrolment**, also in relation to different factors (age at diagnosis, infections before diagnosis, adequacy of replacement therapy, etc.);
- **monitor the progression** of complications through standardized follow-up;

Recommendations for medical management and physiotherapy (prevention and rehabilitation) are also provided.

The *first part* presents the state-of the-art on the problem of respiratory complications in XLA. The *second part* outlines the annual diagnostic protocol whose results will be reported at the end on each year on the specific form. The *third part* gives treatment recommendations. The *fourth part* contains explanatory attachments to facilitate application of the protocol and cooperation with other specialists on technical aspects, including the principles of respiratory physiotherapy, for subsequent in-depth practical analysis by the Centres involved.

The ultimate aim of the protocol is to improve the prognosis of the primary immunodeficiencies, currently linked mainly to respiratory complications, and hence improve the quality of life of these patients.

INTRODUCTION

An abnormal susceptibility to infections is the common denominator shared by all of the primary immunodeficiencies. In particular, pyogenic bacteria like *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis* and *Staphylococcus aureus* are responsible for most infections in patients with a humoral immune deficiency.

The respiratory apparatus is the organ most often involved, followed by the gastrointestinal tract and the skin. Infections of the upper and lower airways are the most commonly reported illnesses in patients with XLA and common variable immunodeficiency (CVID).

High dose intravenous immunoglobulin replacement therapy (IVIG) introduced in the 1980s has drastically reduced the number of acute sinupulmonary infections, namely severe forms of pneumonia in patients with hypogammaglobunaemia.

Unfortunately, even in patients receiving appropriate treatment the reduction in the number of acute infections has not prevented the onset of chronic respiratory complications like chronic sinusopathy, chronic bronchitis, bronchiectasias and chronic respiratory failure, probably because IgA and IgM cannot be replaced in the mucous membranes. These conditions may also be exacerbated by infections favoured by a vicious circle of infection and inflammation secondary to an inability to eradicate certain pathogens in the absence of opsonization and phagocytosis.

These complications are less severe than they used to be and seldom result in respiratory failure. Nonetheless, they must be addressed using appropriate diagnostic tools as clinical presentation may be insignificant and progression occur without further episodes of acute infection.

The importance of early diagnosis and prompt institution of replacement therapy was recently confirmed by a multicentric Italian study (Plebani et al., 2002) showing that the cumulative risk is correlated to age at diagnosis in patients with chronic lung disease, whereas the risk is correlated to the length of follow-up monitoring in patients with later onset disease. Although this is an important issue, few studies to date have assessed the incidence and severity of respiratory complications, especially in children, and guidelines for diagnosis and treatment are lacking.

CLASSIFICATION OF THE MAIN RESPIRATORY COMPLICATIONS

Chronic sinusitis

Inflammation of the paranasal sinuses lasting > 90 days

Diagnosis is primarily clinical and based on the persistence of nasal or retropharyngeal discharge (mucus and purulent mucus) and daily cough. Patients may also present nasal congestion and headache.

If there is no nasal or retropharyngeal discharge and the cough is suspected to be due to chronic bronchitis **rhinofibroscopy** must be performed using a fibre optic fibroscope to disclose mucopus in the middle meatus and/or ostia. Rhinoscopy is also useful in patients failing to respond to treatment to confirm the original diagnosis and diagnose any complications such as nasal polyposis.

CT scanning of the face is only indicated when surgery is envisaged based on rhinofibroscopy after failure to respond to treatment: in this case the examination should also search for any nasal polyps and the reason for impaired patency of the osteomeatal complex. CT scan is required to confirm the diagnosis of chronic sinusitis (in the presence of chronic cough alone) only if rhinofibroscopy cannot be performed.

Chronic bronchitis

Productive cough present for more than four days a weeks for at least three months of the year for at least two years.

Diagnosis is clinical, but analysis of lower airway bacterial colonization, lung function tests and a CT scan are essential to establish the degree of pulmonary impairment and monitor the condition over time.

Culture of the expectorate is useful to disclose chronic bacterial colonization of the lower airways irrespective of acute infectious episodes and for the antibiotic treatment of acute infections **overlying bronchial or pulmonary infection**. Persistent lower airway bacterial colonization is a more severe form of chronic bronchitis.

Lung function tests are essential for to establish whether the chronic bronchitis presents obstructive and/or restrictive features and to define its severity. Determination of the flow-volume curve after forced expiratory manoeuvres under basal conditions and after bronchodilator administration is usually sufficient. Plethysmographic or gas dilution techniques may be useful in patients with moderate-severe broncho-obstruction (FEV1 < 65% of predicted) or restriction.

Lung CT scan will demonstrate areas of carnification and dysventilation, bronchiectasias and minor but important findings subject to improvement such as thickening of the bronchial walls and mucus plugs. The examination should be performed using a high resolution system in cooperative patients or a spiral technique in uncooperative subjects to identify bronchial disease. For the purposes of standardization it is useful to apply a score for areas of thickening and fibrosis, bronchiectasias, peribronchial thickening and mucus plugs.

DIAGNOSTIC PROTOCOL

1. Inclusion criteria

All patients with a diagnosis of humoral immune deficiency (XLA and CVID) established according to the criteria approved in previous recommendations will be enrolled.

2. Clinical, laboratory and instrumental tests in the last year

From 2003 the annual follow-up form includes specific fields for chronic respiratory complications.

2.1 Diagnosis of chronic sinusitis

An initial diagnosis of chronic sinusitis will be based on clinical assessment: persistent (> 90 days) nasal or retropharyngeal discharge (mucus and purulent mucus) + daily cough worsening at night.

If the patient has received antibiotic treatment the 90 days may not be consecutive and symptoms may not be present on some days.

If there is no evidence of nasal or retropharyngeal discharge but only persistent cough (> 90 days even if not consecutive following antibiotic) diagnosis will be established by:

a) rhinofibroscopy (preferably with a fibre optic fibroscope)

positive: presence of mucopus in the middle meatus and/or ostia of the affected sinuses

or, if rhinofibroscopy cannot be performed:

b) CT of the face

positive: partial or total opacification of one or more paranasal sinuses and obstruction of the osteomeatal complex (OMC).

In cases of chronic sinusitis, rhinoscopy, or CT scan if necessary, are essential for the diagnosis of complications, namely nasal polyps.

2.2 Diagnosis of chronic bronchitis and assessment of bronchial bacterial colonization

Diagnosis of chronic bronchitis will be established on the basis of: productive cough (cough with catarrh) on most days (4 or more days a week) for at least three months of the year for at least two years. If chronic bronchitis has been diagnosed, culture of the expectorate should be done at least three weeks after recovery from an episode of respiratory reactivation.

Culture of the expectorate is done on a sputum sample collected in a sterile container, preferably during physiotherapy. The criteria for assessment of specimen adequacy and seeding are listed in Attachment 1.

The following micro-organisms are considered potentially pathogenic: *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Pseudomonas aeruginosa*, *maltophilia* and *cepacea* and *Staphylococcus aureus* isolated in high loads (more than 10.000 cfu/ml if the specimen is processed by dilutions (to be preferred) or more than 2+ of processing is done using a semiquantitative technique. More than one potentially pathogenic micro-organism may be present in the same expectorate specimen.

The following micro-organisms found in the normal oropharyngeal flora are not considered potentially pathogenic as they do not usually give rise to respiratory infections: *Streptococcus viridans*, *Neisseria spp*, *Corynebacterium spp*, *Candida spp*.

2.3 Lung function tests

To be done at least three weeks after recovery from an episode of respiratory reactivation

Spirometry: determination of the flow-volume curve after forced expiratory manoeuvres under basal conditions (suspension of any bronchodilator treatment for at least 10h) and after bronchodilator administration (salbutamol) if FEV1/FVC (Tiffeneau index) < 85 %

Acceptance criteria:

Operator's impression of satisfactory cooperation;

Absence of artefacts (cough, delayed start of expiration, insufficient expiratory effort);

Duration of forced expiration: at least three seconds in children and at least six seconds in adolescents and adults;

At least three tests should be done satisfying the acceptance criteria + forced vital capacity (FVC) and expired volume at the first second of forced expiration (FEV1) with a variability of less than 5 %.

Spirometric indices to be assessed under basal conditions:

FVC in litres

FEV 1 in litres

FEV1/FVC * 100 (% vital capacity expired at the first second, or Tiffeneau index)

PEF (peak expiratory flow) in litres/s

FEF 25-75 (average forced expiratory flow between 25% and 75% of the forced expiratory curve) in litres/s.

In case of bronchial obstruction (FEV1/FVC < 85%) the increase in FEV1 will also be assessed in % after bronchodilator administration.

2.4 High resolution or spiral lung CT scan

To be done every four years in patients with stable clinical status, hence not to be repeated if done in the last four years.

If the clinical status is not stable (increase in the number of months/year of cough and catarrh, drop of FEV1 > 10% in at least two successive spirometric tests carried out some time after episodes of respiratory reactivation) or if the patient has presented at least one episode of severe pneumonia CT scan can be brought forward.

CT scan should be done at least three weeks after recovery from an episode of respiratory reactivation.

Technical details and assessment criteria for lung CT scanning (modified Bhalla score) are listed in Attachment 2.

TREATMENT RECOMMENDATIONS

The aims of treatment are to control infection and delay or block the evolution of pulmonary damage. For this reason the backbone of treatment is prompt effective antibiotic administration to treat acute infections and chronic sinusitis followed by respiratory physiotherapy.

It is important not only to monitor medical management and especially support treatment and respiratory physiotherapy but also to enhance the compliance of patients and their families.

Antibiotic treatment

Antibiotic treatment for *acute respiratory infections* (otitis, bronchitis/bronchopneumonia, sinusitis) is the same as that administered to patients without an immunodeficiency since the respiratory pathogens involved are the same with the same sensitivity to antibiotics. Likewise, treatment is often empirical.

Given the likelihood of more severe disease, it is particularly important to establish an aetiological diagnosis of the agent responsible in order to devise a targeted treatment and to institute prompt therapy by intravenous antibiotic administration.

Antibiotic treatment for *chronic sinusitis* is also the same as that administered to patients without an immunodeficiency. The few data available show that the pathogenic agents involved are largely the same (*Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Staphylococcus aureus* and anaerobic bacteria). An empirical choice of antibiotic against the most common pathogens implicated and their antibiotic sensitivity appears to be effective.

Amoxicillin associated with clavulanic acid (50 mg/kg/daily in 3 divided doses) is the first choice treatment.

The recommended duration of treatment is 2-3 weeks. If the response is partial or there is a good response but a relapse of symptoms in the following two weeks a second or third cycle of treatment may be indicated associating an antibiotic against anaerobic bacteria (clindamycin or metronidazole).

The choice of antibiotic against anaerobic bacteria (to replace amoxicillin and clavulanic acid) should be guided by culture information only in patients failing to respond to empirical treatment (no improvement of symptoms following antibiotic and topical treatment) when the infection is suspected to be caused by drug-resistant bacteria. The gold standard for aetiological diagnosis of the causative agent is puncture of the maxillary sinus. In addition, the culture of material aspirated from the middle meatus under ultrasound guidance is reliable and can be considered the first choice investigation given the invasiveness of maxillary puncture.

The bacteria present in the expectorate presumably play a role in episodes of respiratory reactivation in patients with *chronic bronchitis* and lower airway bacterial colonization. For this reason, initial antibiotic treatment during reactivation must take into account the type(s) of bacteria usually present and their sensitivity to antibiotics. The bacteria most commonly involved include *Haemophilus influenzae*, *Streptococcus pneumoniae* and *Moraxella catarrhalis*. Chronic colonization of the lower airways by *Pseudomonas aeruginosa* presumably occurs later and is associated with a more severe condition of obstructive chronic bronchitis.

No information is available on the efficacy of antibiotic prophylaxis which tends to be avoided, especially in Northern Europe, to prevent the emergence of drug-resistant bacteria. Prophylaxis is nonetheless widely implemented and controlled studies are required.

Support treatment

Chronic sinusitis. Nasal irrigations with saline or hypertonic solution aid the mechanical removal of secretions and reduce oedema of the mucous membranes improving mucociliary clearance (Attachment 3).

Topical corticosteroids are recommended in addition to nasal irrigations to reduce oedema of the mucous membranes and boost patency of the osteomeatal complex and drainage of secretions from the affected sinuses, even though randomized controlled studies are currently lacking. In our experience chronic sinusitis is never cured without adequate support therapy even in non immunodepressed patients.

There are no specific indications on the duration of support treatment. In our experience both nasal irrigations and topical corticosteroids must be administered at least twice daily for at least four weeks.

Patients should be shown how to administered topical treatment and warned that this treatment is essential in addition to antibiotics to ensure a successful outcome. Correct topical application and patient compliance should also be monitored during the course of treatment.

If nasal polyposis fails to respond to topical treatment, systemic administration of steroids is indicated (prednisone 1 mg/kg/die, up to 60 mg/die) for three weeks.

Surgery is indicated in cases of chronic sinusitis which fail to respond to medical management.

If the symptoms of sinusitis persist after 2-3 cycles of antibiotic + topical treatment, particularly if nasal polyps is detected by fibroendoscopic examination, endoscopic surgery must be entertained.

Chronic bronchitis. The treatment of chronic bronchitis is mainly based on respiratory physiotherapy. Respiratory physiotherapy is a curative and preventive treatment which aims to replace and/or enhance the tracheobronchial clearance mechanisms, remove airway obstruction, expand and/or re-expand hypoventilated and atelectasic areas, improve the distribution of inspired air and recondition and boost physical activity (Attachment 4).

Eliminating excess secretions helps to prevent infections and reduce bronchial obstruction and re-expand collapsed parts of the lung.

Until ten years ago postural drainage accompanied by percussion and oscillation was the only means known and available in Italy. There is now scientific evidence of the effectiveness of other techniques to improve and/or stabilize lung function in non immunodepressed patients with chronic bronchitis with or without pulmonary obstruction.

Current consensus is that the most appropriate treatment is that best suited to the individual patient. A key issue is ongoing treatment compliance at home. The treatment protocol must therefore be customized, properly explained and easy to perform.

In cases of bronchial obstruction (spirometry: FEV1/FVC < 85 for children and < 82 for adolescents and adults) and response to bronchodilator administration (increase in FEV1 ≥ 12%) bronchodilator treatment via aerosol may also be useful (short or long-acting beta 2-stimulant). When possible, bronchodilator treatment must be administered before the physiotherapy session.

ATTACHMENT 1

ASSESSMENT OF EXPECTORATE ADEQUACY

Having checked the fluidification of the specimen, it is useful to judge the quality of the expectorate. This is done by microscopic analysis of specimen composition on a slide stained with methylene blue. Smears must be done for all specimens examined (expectorate and bronchial aspirate). Slides are initially examined at low magnification (100x) to disclose squamous epithelial cells and leucocytes. Specimens can be classified into six groups on the basis of the ratio between the two cell components:

GROUP	No. cells/microscopic field x 100	
	Leucocytes	Squamous epithelial cells
1	<25	<25
2	>25	<10
3	>25	10-25
4	>25	>25
5	10-25	>25
6	<10	>25

Only specimens belonging to groups 1, 2 and 3 are suitable for seeding and there is a good correlation with specimens obtained by transcrucoid puncture.

Expectorate specimens belonging to groups 4, 5 and 6, characterized by >25/CM squamous cells, are considered highly contaminated by the bacterial flora of the upper airways and hence not representative of the lower airways.

SEARCH FOR POTENTIALLY PATHOGENIC MICRO-ORGANISMS

A search should be made for the following micro-organisms by seeding on the appropriate growth media: *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Pseudomonadaceae*, *Staphylococcus aureus* and *Enterobacteriaceae*.

ATTACHMENT 2

1- Conditions for high resolution CT lung scan (preferred modality):

Patient awake and cooperative

In deep inspiration, compatible with age

1 mm slices at 8-10 mm intervals

Reconstruction by means of specific algorithms for lung and mediastinum

2- Conditions for spiral CT lung scan:

Uncooperative patient (unable to maintain inspiratory apnoea) or sedated with chloral hydrate (50 mg/kg). 5 mm slices, pitch 1.5.

3- Assessment: Bhalla score (modified):

Severity of bronchiectasias: 0-3

0 absent

1 mild = Φ bronchial lumen slightly $>$ Φ adjacent vessel

2 moderate = Φ bronchial lumen $>$ 2-3x Φ adjacent vessel

3 severe = Φ bronchial lumen $>$ 3x Φ adjacent vessel

Extension of bronchiectasias: (= number of bp segments involved): 0-3

0 absent

1 1-5

2 6-9

3 $>$ 9

Consolidation or thickening zones (irrespective of number): 0-2

0 absent

1 subsegmental

2 subsegmental/lobar

Extension of mucus plugs (= number of bp segments involved): 0-3

0 absent

1 1-5

2 6-9

3 $>$ 9

Peribronchial thickening: 0-3

0 absent

1 thickening of the bronchial wall: $<$ 2x thickness of similar non pathological bronchial branches

2 thickening of the bronchial wall: = 2x " " " " " "

3 thickening of the bronchial wall: $>$ 2x " " " " " "

Notes:

- If bronchoectasias or peribronchial thickening are not uniformly severe in the different zones, report the score for the most common or most severe finding*
- Φ = diameter*

ATTACHMENT 3

REMOVAL OF NASAL OBSTRUCTION AND ADMINISTRATION OF TOPICAL CORTICOSTEROIDS

Nasal irrigation is done for the mechanical removal of dried secretions, crusts, dust and pollution debris obstructing the upper airways and leading to breathing impairment. Regular irrigation of the nasal cavities helps to maintain the mucous membranes and their structures in optimal anatomic-physiological condition for correct nasal function.

Material required:

- a 20 ml syringe
- saline solution
- a beaker

Instructions

- Fill the beaker with saline solution
- Heat the saline solution to around 37° C by immersing the beaker in hot water
- Aspirate the solution with the syringe
- Blow your nose well, one nostril at a time, with mouth closed
- Irrigate your nose over a washbasin with your chest bent forwards and head to one side below the shoulders
- Breathe through your mouth which should be kept open throughout the irrigation
- Insert the nozzle of the syringe into the higher nostril and **slowly** inject the whole content of the syringe
- Let the solution flow through the other nostril. If the second nostril is blocked, the solution will naturally come out through the mouth
- Blow your nose through both nostrils and then one nostril at a time
- Repeat the operation tilting your head to the other side.

Administration of topical corticosteroids

When cortisone nasal sprays are required some suggestions should be given to optimize the efficacy of treatment. The spray should be administered after thorough cleansing of the nasal fossae.

Material required:

- Nasal spray

Instructions:

- Irrigate the nose following instructions
- Shake the spray
- Insert the tip of the spray into the nasal fossa
- Tilt the spray upwards and then downwards keeping it as central as possible in the nostril to avoid damage to the mucous membrane
- Spray into the nostril under deep inspiration
- Repeat on the opposite side
- If 2 sprays per nostril are prescribed repeat as above.

ATTACHMENT 4

TECHNIQUES FOR REMOVAL OF BRONCHIAL OBSTRUCTION

POSITIVE EXPIRATORY PRESSURE MASK - ASTRA TECH MODEL PEP MASK (from M.Falk and S.Zuffo)

Patients required to undergo a respiratory physiotherapy programme to clear excess secretions can be trained to use a positive pressure mask for this purpose.

Treatment with a PEP mask is indicated to remove excess secretions, attenuate airway narrowing and uniformly expand atelectatic or hypoventilating areas.

The PEP kit includes a face mask, a unidirectional valve with a resistance set at the expiratory exit of the valve.

The resistance is set individually for each patient according to age and clinical status choosing the resistance allowing the patient to maintain a stable expiratory pressure of at least 10 centimetres of water for two minutes without tiring.

The resistance diameter is set by the physiotherapist using a manometer.

USE IN COOPERATIVE PATIENTS

Instructions

The patient is in a sitting position with elbows resting on a table and the mask over the face covering the nose and mouth.

The patient **breathes** through the mask. Breathing must be active but calm at an amplitude slightly greater than the current volume level. Expiration must not be forced.

Breathing cycles consist of one minute's respiration through the mask then the mask is removed to perform different degrees of forced expiration followed by a cough to expel secretions. Breathing is then resumed through the mask as instructed for the indicated time.

Duration and number of daily treatments

1-2 daily treatment sessions are usually prescribed in 15-20 day cycles.

When a reacutezation of respiratory symptoms occurs and/or the secretions increase longer and/or more frequent treatments are indicated. If the treatment sessions become tiring a larger number of shorter sessions are recommended. It is important to arrange any treatment changes with the therapist.

USE IN UNCOOPERATIVE PATIENTS

Instructions

The mask should be fitted over the face covering the nose and mouth. Children should be held in the arms or placed in a sling so that an adult supports the head.

The child **breathes** through the mask and air should not escape from the mask edges.

Breathing cycles are: 1 minute's breathing through the mask then the mask is removed for 1-2 minutes for guided expirations and/or coughing to expel secretions (as instructed). Breathing is then resumed through the mask as instructed for the indicated time. At the end of the treatment session the PEP mask is dismantled for storage.

Duration and number of daily treatments

1-2 daily treatment sessions are usually prescribed in 15-20 day cycles.

If the child presents and exacerbation of cough or cold or secretions increase, a larger number of shorter sessions are recommended. It is important to arrange any treatment changes with the therapist.

Cleaning

It is essential to clean and periodically disinfect the PEP mask. The mask is personal and should not be shared with others.

AUTOGENOUS DRAINAGE TECHNIQUE

Autogenous drainage (AD) is a means of clearing secretions .

Aim:

The following procedure outlines the instructions for AD, a bronchial cleansing technique to mobilise secretions from the distal to proximal regions of the bronchial tree.

Indications:

AD requires the patient's full cooperation and is used to remove secretions located in the peripheral parts of the bronchial tree. AD can also be used in patients with severe bronchial irritability.

Instructions

1. The AD technique can be performed with the patient sitting upright with head slightly extended or in a supine position. Children can be held in the arms. The therapist's hands and those of the patient if sitting are placed on the chest to perceive the oscillations produced by the movement of secretions. The position can be changed during the treatment session to regionalize ventilation.
2. The patient must breathe the necessary volume in through the nose slowly and calmly to ensure uniform air distribution.
3. One volume, equal to the current volume, is ventilated at small, medium or high volumes depending on the location of secretions, i.e. at peripheral, middle or proximal levels. If untrained patients find it difficult to breathe at low lung volumes (**VRE**), breathing at their current volume can be suggested. This will start the flow and collection of secretions from the medium and large airways and low volume breathing can be inserted later on.
4. Inspiratory breath should be held for 3-4 seconds with the glottis open to fill all lung districts including those behind the obstructions.
5. Breath should be expired with glottis open through the mouth or nose. This technique must be carefully learnt beforehand by teaching the patient to breathe out without a

sound and mist over a mirror. If the glottis is kept open is easier to avoid airway compression.

6. The force exerted during the expiratory phase must be balanced to trigger the maximum possible flow without causing airway compression. Patients with moderate or severe impairment should breathe out blowing gently. Patients with very severe impairment should not breathe out more strongly than normal.
7. During correct expiration the oscillations produced by the secretions in movement can be perceived by **listening** to sounds at the mouth, placing a **hand** on the chest. The frequency of oscillations helps to locate the secretions: high frequency chest wall oscillations indicate secretions in the small airways, while low frequency oscillations are produced by secretions in the large airways. Tactile, auditory and proprioceptive feedback allows the patients to adjust the technique and establish AD times.
8. The cycle of inspiration– pause – expiration should be repeated until the secretions have collected in the large airways. The level of breathing is then gradually increased until the secretions are in the trachea.
9. The mucus is cleared by quick expiration or high volume expiration.
10. Coughing should be avoided at this stage and only used if necessary after expiration to bring the secretions into the mouth.
11. When the cycle is completed the patient must breathe calmly before repeating another cycle.
12. The duration of each session depends on the individual patient's quantity and viscosity of mucus and his/her ability to implement the technique.
13. If secretions are still present at the end of the session, AD should be repeated more often.
14. It is preferable to start AD by mobilizing secretions in peripheral airways, but if drainage is hampered by secretions in the upper airways these should be expelled first.
15. While training patients how to perform AD and in patients with abdominal muscle deficiency the therapist places a hand on the side of the abdomen and assists the manoeuvre. The therapist's other hand is placed on the side of the ribs for counter-resistance.
16. When the mobilized secretions have reached the upper airways the patient is spontaneously stimulated to expel them by forced expiration technique (FET) and coughing.
17. The manoeuvre should be repeated until the adventitious respiratory sounds perceived at the mouth and on chest auscultation have disappeared completely.

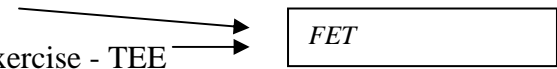
Assessment

The physiotherapist must check

- Correct performance of the technique
- Any change and disappearance of adventitious breath sounds
- The efficacy of the technique on bronchial toilette.

ACTIVE CYCLE OF BREATHING TECHNIQUES (Webber '88)

The physiotherapy technique is based on the alternate use of:

- Huff
 - Breathing control - BC
 - Thoracic Expansion Exercise - TEE
- 
- The diagram shows two arrows originating from the text 'Breathing control - BC' and 'Thoracic Expansion Exercise - TEE' respectively, both pointing towards a rectangular box containing the text 'FET'.

The exercises can be performed during postural drainage or in a sitting position with or without percussion, vibration or oscillation.

BREATHING CONTROL

Breathing control consists of taking a calm breath at current volume. The duration depends on the individual and the degree of bronchial hyperactivity.

CHEST EXPANDING EXERCISES

These exercises consist in deep breaths until the inspiratory reserve volume is reached followed by a pause at the end of inspiration. Expiration must be calm and not forced. The exercises can be combined with percussion, vibration and oscillation during expiration. The TEE should be repeated 4-5 times.

FORCED EXPIRATION TECHNIQUE (FET)

This technique serves to facilitate expectoration of bronchial secretions. FET consists of:

- one or two forced expirations called huffs, performed energetically but not violently keeping the glottis open
- followed by controlled breaths known as Breathing Control

ATTACHMENT 5

DOMICILIARY CLEANING AND DISINFECTION OF EQUIPMENT

The devices used by patients for respiratory treatment (aerosol vial, space maintainers, physiotherapy equipment) are readily contaminated by germs which may give rise to reinfection of the breathing apparatus. For this reason good hygienic measures are essential. Cleaning is the removal of surface dirt while disinfection is a physical (steam) or chemical (bleach) treatment designed to eliminate the majority of microbes.

CLEANING

All items should be carefully washed with washing-up liquid after use as follows:

- ❖ Soak all the dismantled parts of the device in detergent
- ❖ Carefully brush all surfaces paying attention to delicate parts
- ❖ Rinse with hot water
- ❖ Carefully dry the parts with a clean cloth made of filament-free material
- ❖ Store the dried dismantled items in a cloth bag
- ❖ Only reassemble the device when needed

DISINFECTION

It is important to disinfect devices regularly using one of the following methods depending on the type of material used, following the physiotherapist's instructions.

Clean all the dismantled parts as instructed then proceed to disinfection.

Cold disinfection

- Soak the items in a 10% bleach solution (1 part bleach in 9 parts tap water) for 20 minutes. Rinse with hot water
- Carefully dry the parts with a clean cloth made of filament-free material
- Store the dried dismantled items in a cloth bag
- **Only reassemble the device when needed**

Steam disinfection

- Place the items in a food basket or steam sterilizer for 10 minutes (the items must not be soaked in boiling water).
- Remove the items from the basket
- Carefully dry the parts with a clean cloth made of filament-free material
- Store the dried dismantled items in a cloth bag
- Only reassemble the device when needed.