

Lung volume recruitment in patients with Duchenne Muscular Dystrophy in the early non-ambulatory stage

Reclutamiento de volumen pulmonar en pacientes con Distrofia Muscular de Duchenne en etapa no ambulatoria temprana

Annex 1. Description of evaluations

Evaluation of vital signs: user in a seated position (sitting in a wheelchair or chair, back resting on the backrest and feet on the footrest or box in its absence). Respiratory rate, axillary temperature, oxygen saturation, heart rate and blood pressure are measured. If at the time of the evaluation of vital signs the body temperature is greater than 37.5°C, the user will be referred to the health center of their network. If the oxygen saturation is less than 95%, 3 cycles of LVR with subsequent assisted cough stimulation will be performed, with a reevaluation right after. The patient will be referred to a health center and respiratory evaluation will be rescheduled.

Respiratory evaluation: fingertip pulse oximeter is maintained on the index finger of the dominant hand throughout the evaluation. Prior to each evaluation, the procedure is explained to the user and family, with a demonstration of the technique.

- **Vital Capacity (VC):** It is measured by means of a respirometer, a non-invasive device that, through an interface, allows determining the volume of exhaled air after maximum inspiration. The user is placed in a seated position. The evaluator holds the interface attached to the user's face and holds the respirometer. It is requested to perform 3-5 cycles of breathing at tidal volume, then request the performance of a maximum inspiration, followed by a maximum expiration within the interface. 3 acceptable and reproducible maneuvers are requested, with 1 minute of rest between them, of which the highest value will be recorded^{7,17,39}.

For the measurement of VC in the supine position, the same procedure as the previous measurement is performed, with the user located in the supine position.

- **Maximum insufflation capacity (MIC):** It is measured with the same device and in the same position as the VC, it allows us to know the maximum volume of air that the lungs can contain with the glottis closed after the maximum insufflation assisted with a manual resuscitation bag. The user is instructed to receive consecutive volumes of air, which will keep closing the glottis until the lungs are completely filled with air (patient must indicate the moment when they do not require further insufflation) and then proceed to perform a maximum expiration within the interface. 3 acceptable and reproducible maneuvers will be requested, with 1 minute of rest between them, of which the highest value will be recorded^{7,17,39}.
- **Peak cough flow (PCF):** It is measured through a Mini-Wright peak flow meter, which is connected to an interface. The user is in a seated position. For the measurement, maximum inhalation is requested, holding the breath while the evaluator positions the interface and then the user coughs vigorously within the interface. Then, the same maneuver (PCF) will be evaluated accompanied by abdominal compression. 3 acceptable and reproducible maneuvers are requested, with 1 minute of rest between them, of which the highest value will be recorded with a difference of less than 20 L/min between the two highest³⁹.
- **Peak cough flow from maximum insufflation capacity (PFT+MIC):** User is in a seated position. The professional set the interface to the user's face. The patient is asked for 3 breaths at tidal volume, after which must take maximum inspiration, at which time the evaluator or companion presses the manual resuscitation bag, delivering additional volume of air. Assisted breathing is repeated until the user gives the signal to stop. At that moment the companion removes the interface and installs the circuit with the Mini-Wright peak flow meter connected to the interface and the user must cough vigorously. 3 acceptable and reproducible maneuvers are requested, with 1 minute of rest between them, of which the highest value will be recorded with a difference of less than 20 L/min between the two highest^{7,17,39}.

Training Description:

The training consists of explaining to the user and caregiver the maneuvers to be performed, elements of the respiratory kit, use and dosage, which are:

1. **Active Lung Volume Recruitment Maneuver with Manual Resuscitation Bag and Manual Cough Facilitation:**

For the execution of this maneuver, a respiratory kit is needed that consists of: manual resuscitation bag, one-way valve, corrugated tube, adapter and nasobuccal interface; during this, the user is monitored with pulse oximeter monitoring in the dominant middle finger for HR monitoring and oxygen saturation, in addition to quantifying the level of fatigue through EPIInfant.

(*) If during training EPIInfant is > 7 , a 5-minute break should be taken to reevaluate and resume training⁴³.

Positioning: Seated user, caregiver with one hand sets the interface to the user's face, covering nose and mouth; with the other hand holds the manual resuscitation bag.

Procedure: 3 breathing cycles at tidal volume are requested slowly, followed by maximum expiration, and then staggered inspirations are performed simultaneously with insufflations performed by the caregiver, with subsequent glottic closure. At least 3 insufflations are performed until the MIC is reached, where the user performs a previously agreed signal, and the insufflations are stopped. The interface is removed, the user holds the air for 3 seconds and then the caregiver performs manual facilitation of the cough.

After hyperinflation, the caregiver should place both hands at the abdominal level and coordinate the expulsive phase of cough with compression in the anterior-posterior and ascending directions⁴⁴. (Figure 2).

Dosage: Begins with an adaptation period with 3 daily sets of 10 repetitions for the first 3 months, and then progresses to 2 daily sets of 20 repetitions for the remaining 9 months, ensuring 40 repetitions per day^{17,18}.

Annex 2. Follow-up Schedule

Information to collect during care:

- Patient's health status
- Difficulties presented in case of non-compliance with the indications (illness, family problems, etc.).
- Performance of the indicated breathing maneuvers
- Vital signs monitoring: if you have a pulse oximeter, consult for SpO2 and initial and final HR; and/or initial and final EPIInfant. (*) (43)
- Complete Match Schedule

	Active lung volume recruitment maneuver		Manual Cough Facilitation	
	YES	NO	YES	NO
It has all the implements of the respiratory kit (nasobuccal interface, adapter, corrugated tube, one-way valve, manual resuscitation bag)				
User correctly positioned (seated at 90° with feet on the floor or footrests, and back supported on the back of a chair).				
Caregiver with one hand fixes the interface to the user's face, covering nose and mouth; and with the other hand holds a manual resuscitation bag.				
User performs 3 breathing cycles at tidal volume slowly.				
The user takes inspirations simultaneously with insufflations performed by the caregiver, followed by glottic closure.				
User is able to perform at least 3 insufflations until reaching the MIC (previously agreed signal)				
User is able to hold air for 3 seconds				
The caregiver positions both hands at the abdominal level and coordinates the expulsive phase of coughing with compression in the anterior-posterior and ascending directions.				
Indicated dosage completed				

(*): If EPIInfant is >7, a 5-minute break should be taken to reevaluate and resume training⁴³.