

# Respiratory Measures ALS

## ENCALS SOP

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### **Background**

Vital capacity (VC) is one of the core outcome measures routinely used in ALS clinical trials. Therefore, accuracy and repeatability of VC measurements are essential to obtain reliable efficacy data in randomized ALS clinical trials. VC is defined as the volume of air expelled during expiration following a full inspiration and completed while the patient is as relaxed as possible while completing the expiration [1, 2]. Acceptable VC test quality requires optimal patient performance. Optimal patient performance requires adequate coaching.[3, 4]

During this training we will focus on Forced vital capacity (FVC), Slow vital capacity (SVC) and Sniff nasal inspiratory pressure (SNIP).

## **SVC and FVC TESTING**

### **Clarification of used terms:**

FVC; The volume change of the lung between a full inspiration to total lung capacity and a maximal expiration to residual volume. The measurement is performed during forceful exhalation; the preceding maximal inhalation need not be performed forcefully.[2, 5, 6]

SVC; This test resembles the FVC. The difference is that the expiration in the spirometer is done slowly. The patient inspires fully and then slowly expires all the air in his/her lungs (Inspiratory Vital Capacity) or the other way around: the patient expires fully and inspires slowly to a maximum (Expiratory Vital Capacity).[6, 7]

### ***Step 1) Preparation***

Ensure that the turbine transducer is plugged into either one of the first two sockets, on the right-hand side of the instrument.



### ***Step 2) Calibration***

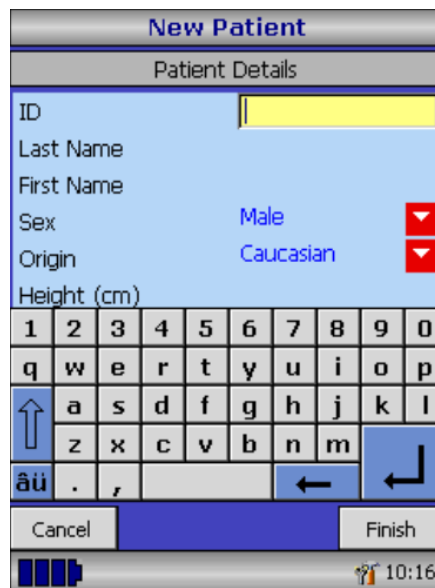
The calibration should remain stable at all times, unless the transducer is damaged. It is recommended that the calibration is checked once every three months, although the unit should not require re-calibration.

### Step 3) Adjusting the set-up menu

Your test conditions need to be set on the spirometer.

- Turn the machine on by pressing the on/off button.
- Select the test you would like to perform: SVC (shown as Relaxed Spirometry) or FVC (shown as Forced Spirometry).
- Report the patient's:
  - sex, select by dropdown menu: male or female
  - race\*, select by dropdown menu.
  - height, fill in the height, units are country-specific
  - weight, fill in the weight, units are country-specific (optional)
  - age, fill in the age in years

After you have filled in the patient's information, press 'finish'.

The image shows a 'New Patient' form. At the top, it says 'New Patient' and 'Patient Details'. Below this are fields for 'ID', 'Last Name', and 'First Name'. There are dropdown menus for 'Sex' (set to 'Male') and 'Origin' (set to 'Caucasian'). Below these is a 'Height (cm)' field followed by a numeric keypad (0-9) and a QWERTY keyboard. At the bottom, there are 'Cancel' and 'Finish' buttons. A status bar at the very bottom shows a battery icon and the time '10:16'.

### Step 4) Explaining the test

General: *“Your breathing capacity (how much air you can push in and out of your lungs) will be tested using the vital capacity test (VC). The VC measures the maximum amount of air you can exhale (blow out) following a deep breath. The spirometer will measure how much air you have exhaled. This test will be done 3 times to ensure that we have measured your best attempt.”*

- SVC specific; *“Please start with normal breathing. After a few breaths, I want you to fill your lungs completely, then blow out gently all the way until you are empty.[3]”*
- FVC specific; *“Please start with normal breathing. Then I want you to take a huge breath in until your lungs are completely full, and blast it out as hard and as fast as you can until you feel you are completely empty and cannot blow out further.[3]”*

**Make sure the subject has a complete understanding of the test!**

### **Step 5) Instructions for the subject during the test:**

General: During testing, the Research(er) (Nurse) should exhibit enthusiasm, allay the patient's anxiety, convey simple instructions, demonstrate each test, give vocal encouragement and provide feedback on performance. Observe the patient's nonverbal cues, such as facial expressions and body language, and use your own body language effectively. This can enhance the patient's test performance [3, 4].

The patient could be distracted by people surrounding him/her during performance of the VC test. Before you start the measurement, agree with the patient if he/she want(s) to perform the test alone so that he cannot be distracted by people around him.

Positioning; Make sure the patient sits upright with the legs next to each other, shoulders pulled back and eyes wide open.

Placing mask or mouthpiece:

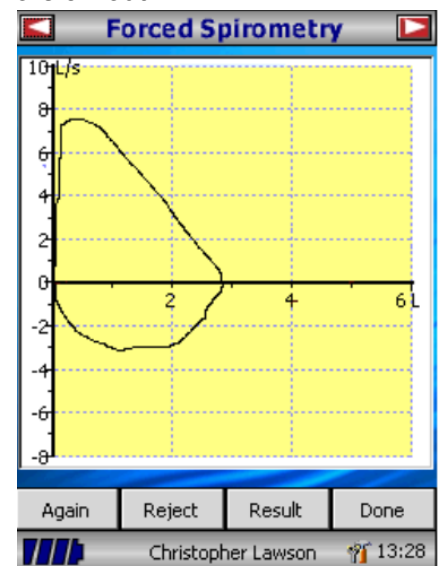
Clear of any saliva and place the mouthpiece in the patient's mouth and place nose clip on the nose making sure the patient can seal their lips around the mouthpiece and that no air is leaking. Or place the mask over the nose and mouth of the patient, supporting it firmly behind the head.

- SVC specific;
  - *'Please start with normal breathing. After a few breaths, I will ask you to take as big a breath in as possible to fill your lungs.'*
  - Say, *"I want you to breath in as deeeeeep as possible."* Then say, *"Deeper."*
  - *"blow out gently, all the way until you are empty."* A tactile cue, such as placing a hand gently on the shoulder of the patients and telling them to continue their expiration until the hand is lifted, can help. Alternatively, a time cue, such as asking them to continue their effort for *"another two seconds"* (or some other arbitrarily short and achievable duration) is used, once they have almost reached a plateau.
- FVC specific;
  - *'Please start with normal breathing. After a few breaths, I will ask you to take as big a breath in as possible to fill your lungs.'*
  - Say, *"I want you to breath in as deeeeeep as possible."* Then say, *"Deeper."*
  - Yell, clap your hands, or click your fingers near the patient's ear so as to startle the patient into delivering his or her fastest possible peak flow. And say, *"blast it out as hard and as fast as you can"*
  - Tell the patient quietly to *"keep going, I can see you're still getting more air out."* until a plateau is reached on the volume-time spirogram.

### **Step 6) Processing of the measurement outcomes**

After performing the test you can select different options on the device:

- Again, *if the previous maneuver was abnormal and you want to perform a new maneuver.*
- Reject, *if the performed maneuver is not within the range of 5% or the machine gives you the feedback that the test wasn't performed correctly.*
- Result, *If you want to see the results of the maneuvers performed so far.*
- Done, *If three sufficient maneuvers have been performed.*



Other options:

- Select 'done' if the patient performed 3 correct maneuvers (the machine will select automatically the best performed maneuver).
- Select 'print' to print the results of the maneuvers.
- Select 'save' to save the results.

### Additional Guidelines

Perform the Vital Capacity test at the beginning of each study visit to avoid fatigue from other testing.

Research(ers) (Nurses) often say, "Okay, take a deep breath and blow out fast. . . *Blow, blow, blow, blow, keep going, keep going, keep going, keep going.*" Emphasis should be on the first phase of the spirometry maneuver (maximum inhalation). This can obtain much larger vital capacities [4].

Facial expressions and body language are much more important than telling the patient what to do. If you recite all of the steps of the procedure (like flight attendants do before each take-off), by the time you have finished, the patient will probably have forgotten the first step, which is the most important. Showing enthusiasm, being a cheerleader, using body language, and observing the patient's body language are very important in order to obtain good spirometry results.[3, 4]

It is crucial that everyone in a multicenter study is using the same predicted values and making racial adjustments the same way and every time.

\*The following factors are recommended when using ECCS normal values[8]:

Hong Kong Chinese	100%
Japanese American	89%
Polynesians	90%
North Indians and Pakistanis	90%
South Indians and those of African descent	87%

### Care and cleaning of device

- Don't touch the screen with fingers; use only the stylus provided with the device.
- Don't direct the transducer holder towards a strong light source whilst operating the spirometer.
- Clean the transducer on a regular base with Perasafe. Alcohol and chloride solutions should be avoided

## **SNIP TESTING**

### Clarification of used terms:

SNIP; The SNIP is measured from functional residual capacity (FRC) in the sitting position using a catheter inserted through a plug occluding one nostril during 10 maximal sniffs through the contralateral nostril. For each test, the largest pressure measured in cm H<sub>2</sub>O is taken into account. The SNIP correlates well with invasive and non-volitional tests of diaphragmatic strength but also with the function of the sternocleidomastoid muscle.[2, 5, 9]

### ***Step 1) Calibration***

The calibration is factory set and should remain stable indefinitely.

### ***Step 2) Explaining the test***

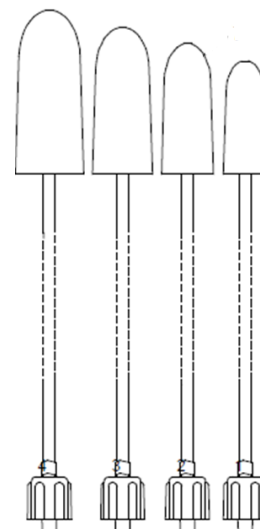
An example of an instruction: Your breathing capacity will be tested using the sniff nasal inspiratory pressure test (SNIP). The SNIP measures the amount of air you can inhale (via your nose) following a sharp short sniff, a total sniff duration of less than 0.5 second[9]. First I will select a popper nasal probe. If the nasal probe fits properly I will ask you to Sniff as hard as you can. This test will be done 10 times to ensure that we measured your best attempt.

The patient could be distracted by people who are around him while performing the VC test. Before you start the measurement, agree with the patient if he wants to perform the test alone so that he can't be distracted by people around him.

**Make sure the subject has a complete understanding of the test.**

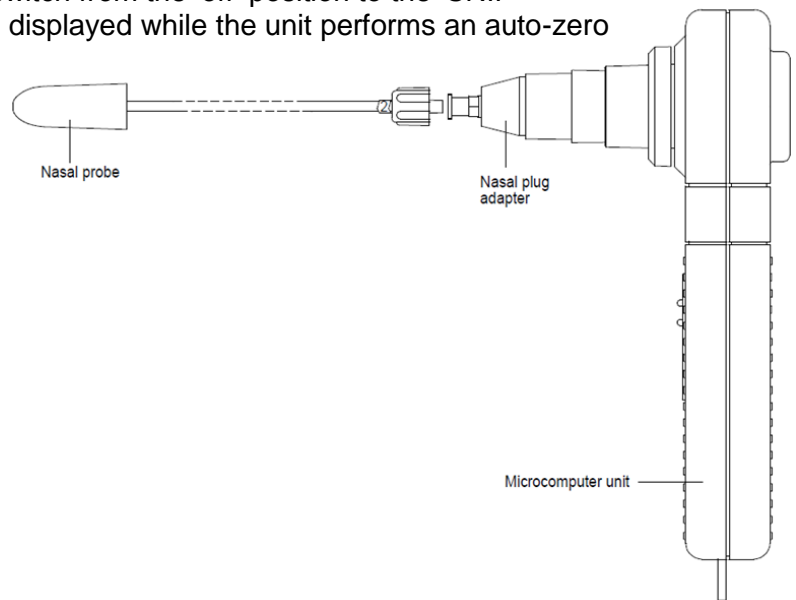
### ***Step 3) Asking for patient's information***

Ask the patient whether he/she has had any type of nasal surgery, is currently experiencing any nasal congestion, or if he/she has a deviated nasal septum (these items could possibly confound results). If the patient's response is positive to one of the items, this should be recorded as such.



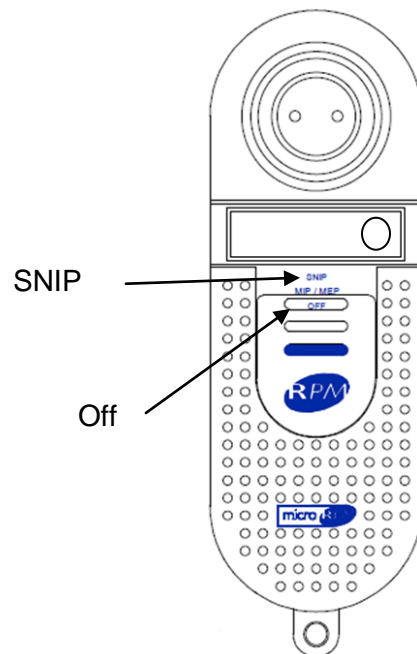
#### **Step 4) Preparing the test**

- **Select size:** The correct size (1-4) can be ascertained by fitting a Nasal Probe to the unit, then firmly inserting the Nasal Probe into a nostril.
- **Select nostril:** Select the nostril that seems most appropriate to obtain complete sealing of the nostril.
- **Apply the nasal probe to the selected nostril:** Check if the patient is able to in- and exhale through the nasal probe, by asking him/her to close the other nostril with a finger. Adjust if needed.
- **Attach the nasal probe to the device**
- Turn the device on by sliding the switch from the 'off' position to the 'SNIP' position (rotating segments will be displayed while the unit performs an auto-zero function).



### **Step 5) Performing the test**

- Instruct the patient to sit up straight with legs next to each other.
- In the nasal probe.
- After some tidal breathing, instruct the patient to inhale with as much effort as possible by saying 'Sniff as hard as you can through the free nostril with your mouth closed'.
- Note that the other nostril should NOT be occluded while performing the test.
- The total sniff duration is less than 0.5 second.
- The device will display the peak SNIP in cm H<sub>2</sub>O.
- Write down the measurement outcomes.
- Reset the device by sliding the switch from the SNIP position to the 'off' position and back to the SNIP position again.
- Make sure there are at least 30 seconds between each test.



### Additional Guidelines

Perform the Vital Capacity testing at the beginning of each study visit to avoid fatigue from other testing.

### Care and cleaning of device

The respiratory pressure meter requires no routine maintenance or servicing and is protected from contamination by the bacterial filter supplied. However, the nasal probes may be immersed in cold sterilizing solutions such as alcohol or chloride.



## **References**

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