

Supplementary-Table-3. Framework for the evaluation of global respiratory muscles and diaphragm function (adapted from (1,2)).

A. List of respiratory muscle variables assessed
<ol style="list-style-type: none"> 1. abdominal paradox on clinical examination (sitting or supine) 2. restrictive ventilatory defect on pulmonary function tests (PFT), defined as a vital capacity (VC) below 80% of normal predicted value (NPV) with normal forced expiratory volume in 1 second /VC ratio or a total lung capacity below 80% of NPV 3. VC drop above 15% in the supine position 4. maximal inspiratory pressure (MIP) and sniff nasal inspiratory pressure (SNIP) both lower than 80% of NPV 5. esophageal pressure drop in response to bilateral phrenic nerve stimulation below 11 cmH₂O 6. amplitude of diaphragm electromyographic motor responses to electric phrenic stimulation (M-waves) below 250 mV (micro-voltage), bilaterally 7. reduced diaphragm thickening fraction or excursion on ultrasound examination, bilaterally 8. reduced diaphragm excursion on dynamic magnetic resonance imaging, bilaterally 9. clustered episodes of hemoglobin desaturation and/or hypercapnia on nocturnal oximetry/capnography 10. hypercapnia (PaCO₂ ≥ 45mmHg) on arterial blood gases during unassisted breathing, in the absence of evident central hypoventilation and of obstructive ventilatory defect on PFT 11. presence of myositis/necrotic figures on diaphragm histopathologic description at autopsy.
B. Diagnosis of global respiratory muscle involvement
B.1. Certain
B.1.1. Restrictive ventilatory defect AND concomitantly low MIP+SNIP
B.1.2. <i>In the absence of either MIP or SNIP (technical issue or patient's inability to perform):</i> restrictive ventilatory defect AND low MIP or low SNIP AND hypercapnia
B.2. Probable
B. 2.1. Absence of restrictive ventilatory defect BUT low MIP+SNIP
B.2.2. Absence of spirometry AND MIP+SNIP (<i>technical issue or patient's inability to perform</i>) BUT hypercapnia OR diaphragm weakness considered certain and severe (see C)
C. Diagnosis of diaphragm involvement
C.1. Certain
C.1.1.at least two criteria among the following: <ul style="list-style-type: none"> - abdominal paradox; - supine VC drop >15%; - esophageal pressure twitch < 11 cmH₂O; - bilaterally micro-volted diaphragm M-waves; - reduced diaphragm excursion on ultrasound; - reduced diaphragm excursion on dynamic MRI; - clustered episodes of desaturation/hypoventilation on oximetry/hypercapnia.
C.1.2. complete diaphragm paralysis on diaphragm ultrasound or dynamic MRI
C.1.3. diaphragm myositis at autopsy
C.2. Probable
C2.1. Respiratory muscle weakness AND one of the criteria listed in C1
C2.2. Respiratory muscle weakness BUT none of the above.

Abbreviations: MIP: maximal inspiratory pressure ; NPV: normal predicted value; PFT: pulmonary function tests; SNIP: sniff nasal inspiratory pressure; VC: vital capacity

REFERENCES

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2. Laveneziana P, Albuquerque A, Aliverti A, Babb T, Barreiro E, Dres M, *et al.* ERS statement on respiratory muscle testing at rest and during exercise. *Eur Respir J* **2019**;53(6) doi 10.1183/13993003.01214-2018.