Ventilatory and Cardiovascular Responses to Hypercapnia and Hypoxia in Multiple-System Atrophy

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**Sudomotor subscore**

1. Any of the following alterations:
   a. Single QSART site abnormal or
   b. Length-dependent pattern (distal sweat volume less than one-third of forearm or proximal leg values) or
   c. Persistent sweat activity at foot (TST anhidrosis present but <25%)

2. Any of the following alterations:
   a. Single QSART site <50% of lower limit of normal
   b. ≥2 QSART sites reduced (TST anhidrosis 25%-50%)

3. ≥2 QSART sites <50% of lower limit of normal

**Cardiovagal subscore**

1. HR\textsubscript{DB} mildly reduced but >50% of minimum
2. HR\textsubscript{DB} reduced to <50% of minimum
3. Both HR\textsubscript{DB} and Valsalva ratio reduced to <50% of minimum

**Adrenergic subscore**

The adrenergic subscore is based on beat-to-beat BP alterations during the VM and head-up tilt testing. A score of 1 is given for any of the following changes in VM:

a. Phase II\textsubscript{e} reduction >20 but <40 mm Hg mean BP or
b. Phase II\textsubscript{l}, does not return to baseline or
   c. Pulse pressure reduction to <50% of baseline

If VM is normal, a score of 1 can be assigned if the following changes occur at head-up tilt testing:

a. Excessive oscillations in mean BP (>20 mm Hg occupying at least 50% of head-up tilt time)
   b. Pulse pressure reduction to <50% of baseline
   c. Transient decrease in systolic BP >20 mm Hg with recovery within 1 min
   d. Systolic BP reduction >20 mm Hg beyond 1 min
   e. Diastolic BP reduction >10 mm Hg beyond 1 min
   f. Overshoot at head-up tilt >20 mm Hg systolic BP

A score of 2 is assigned if the mean BP decrement in phase II\textsubscript{e} of VM is >40 mm Hg. If a score of 1 is determined from the VM, it can be increased to 2 if the following changes on head-up tilt testing occur:

a. Transient systolic BP decrease >30 mm Hg with recovery within 2 min
   b. Systolic BP reduction >20 mm Hg beyond 1 min
   c. Diastolic BP reduction >10 mm Hg beyond 1 min

A score of 3 is assigned if the following change occurs in the VM:

Phase II, mean BP reduction >40 mm Hg plus absent phase II; and phase IV

An additional point is assigned if a reduction in manual systolic BP >30 mm Hg occurs beyond 2 min and is sustained for at least 2 min during head-up tilt testing

Abbreviations: BP, blood pressure; HR\textsubscript{DB}, heart rate response to deep breathing; QSART, quantitative sudomotor axon reflex test; TST, thermoregulatory sweat test; VM, Valsalva maneuver.

*Phases refer to components of the VM: II\textsubscript{e}, and II\textsubscript{l}, are early and late proportions, respectively.