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Paola Sandroni and Phillip A. Low
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Introduction to Tests

Studies cannot be interpreted in the absence of clinical information and testing situations. One can describe the abnormalities, make assumptions, but many may look alike but have very different meanings and implications. Obtaining a detailed history of medications taken (including over-the-counter) prior to testing is crucial as it is vital to have a well-trained technician who can identify technical artifacts, effort or any other patient-related issue that can impact the test quality and meaning.

Note on Figure legends

The testing techniques and physiology have already been detailed in multiple publications and books. Here is simply a key point synopsis to aid the reader.

Deep breathing (a.k.a. metronomic breathing) test: test is performed supine at a breathing rate of 6/min, which generates the largest heart rate variability. The response is generated by the Hering-Breuer reflex (mediated by lung stretch receptors), Bainbridge reflex (mediated right heart filling receptors), and baroreflex activation. Afferent and efferent branches are both vagal, and the signals are processed in the nucleus of the solitary tract. Factors affecting the response include: age, rate of breathing, CO_2 level, presence of primary cardiac or lung pathology, conditions affecting the mechanics of respiration, influence of sympathetic outflow, medications.

Valsalva maneuver: a forced expiration at 40 mmHg for 15 seconds performed supine. The maneuver results in a fall of BP (early phase II) that activates baroreflex. This triggers a sympathetic surge with

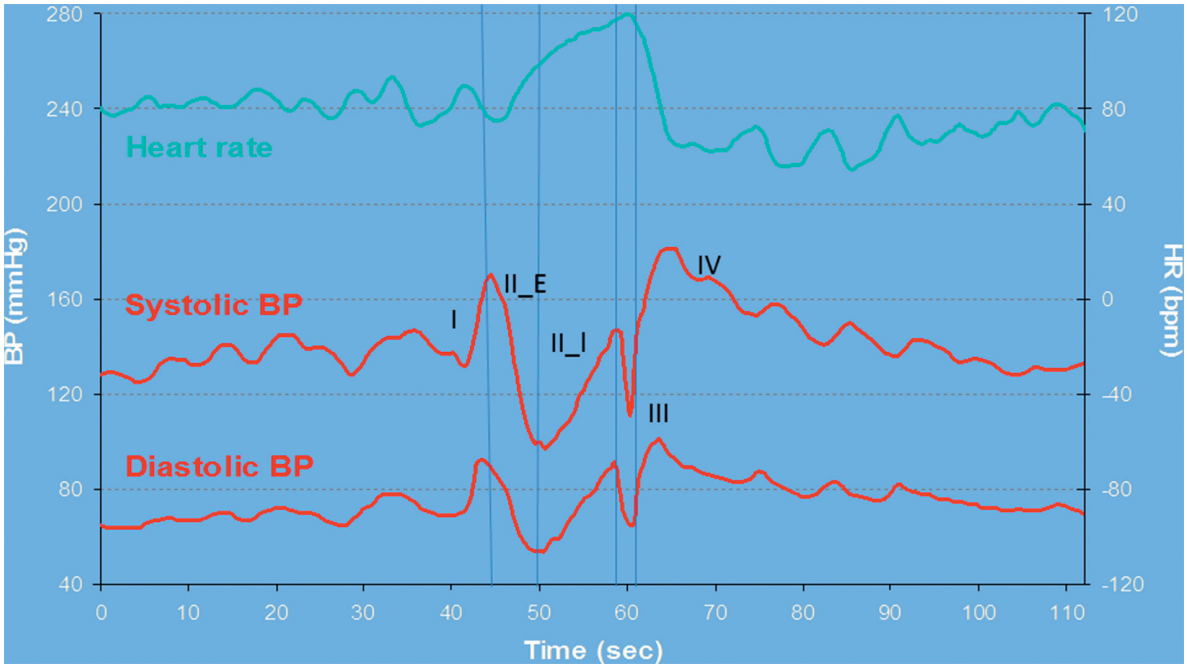

Fig. I.1

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peripheral vasoconstriction and tachycardia that generates a BP rise (late phase II), usually with mean BP back to baseline value. At the end of the maneuver, a brief drop in blood pressure occurs (phase III), followed by an overshoot (phase IV). Baroreflex activation blocks the sympathetic outflow and triggers vagal surge, resulting in bradycardia and release of peripheral vasoconstriction (Fig. I.1). Factors affecting the response include: position of the subject, expiratory pressure, duration of effort, age and gender, volume status, medications. Valsalva ratio = maximum/minimum HR. The blood pressure profile should be described, including recovery time (time from the bottom of phase III to return to baseline BP; normal value < 6 seconds) if abnormal.

Tilt test: passive tilt-up for 10 minutes after 20–30 minutes supine baseline. Blood pressure and heart rate are monitored beat-to-beat throughout the studies.

Quantitative sudomotor axon reflex test (QSART): after stable baseline is obtained, acetylcholine is iontophoresed by applying electrical stimulation for 5 minutes, recording continues for 5 minutes more. By activation of axon reflex, sweating occurs. Sweat volume is obtained by integrating the area under the 10 minute curve.

QSART tracings: red = forearm; blue = proximal leg; green = distal leg; yellow = foot. The marks indicate: start of recordings to obtain baseline, acetylcholine injection in capsule, stimulation starts, stimulation ends, recording ends.

Deep breathing (DB), Valsalva maneuver (VM) and tilt tracings: blue = chest strap indicating breathing effort; green = heart rate; red = systolic blood pressure; pink = mean BP; yellow = diastolic BP and expiratory pressure generated during VM. During DB the marks indicate each inspiratory and expiratory act. During VM the marks indicate: patients take deep breath, expiratory effort start, expiratory effort ends. During tilt the marks indicate tilt-up and tilt-down.

Thermoregulatory sweat test (TST): the normally sweating areas are in purple, while the areas with reduced or absent sweat are in yellow. The test utilizes alizarin red as an indicator, which turns from yellow to purple when wet. Actual photos are shown, not the drawings. Reduced sweat is often seen over bony areas, callused skin, stretch marks, scars, and various skin conditions.

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Normal examples

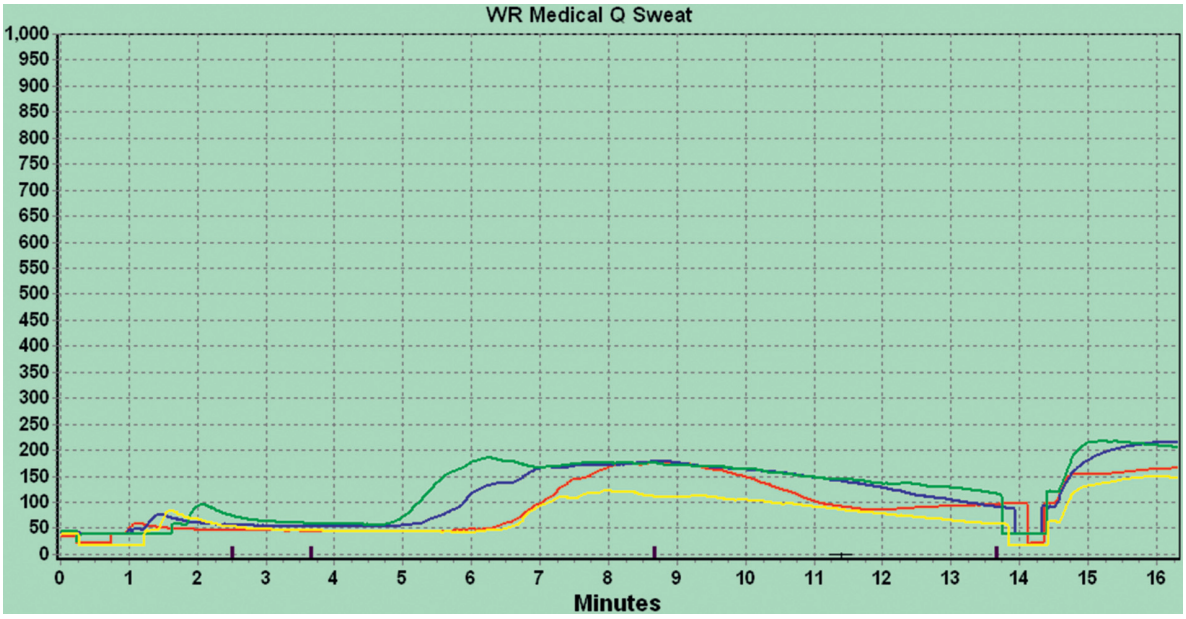

Fig. I.2(a). Normal sweat responses in a female and a male case. Generally, males have larger output.

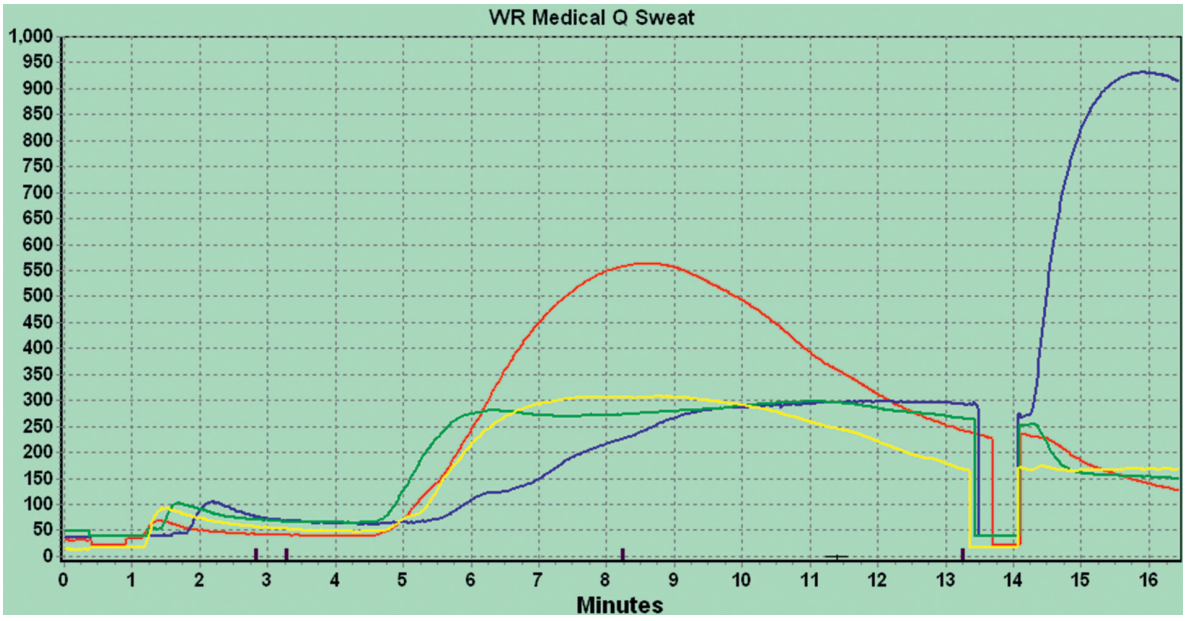

Fig. I.2(b).

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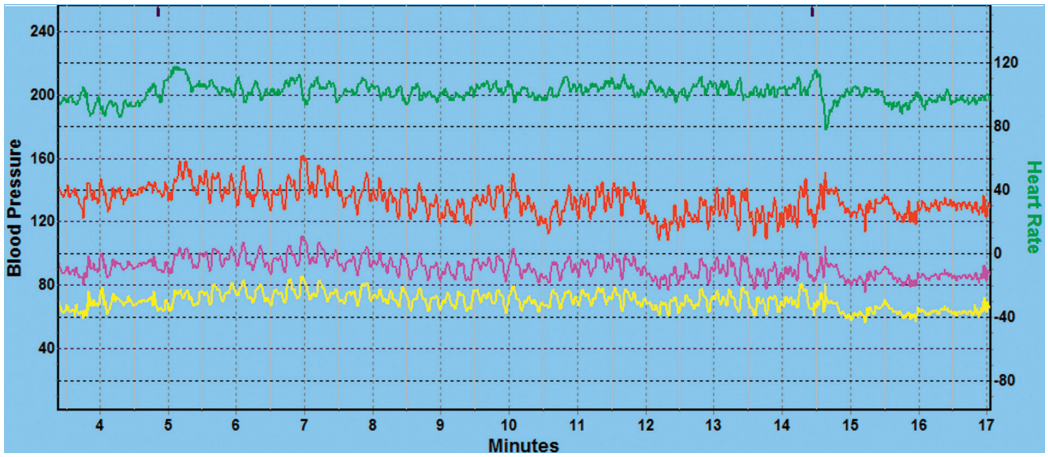
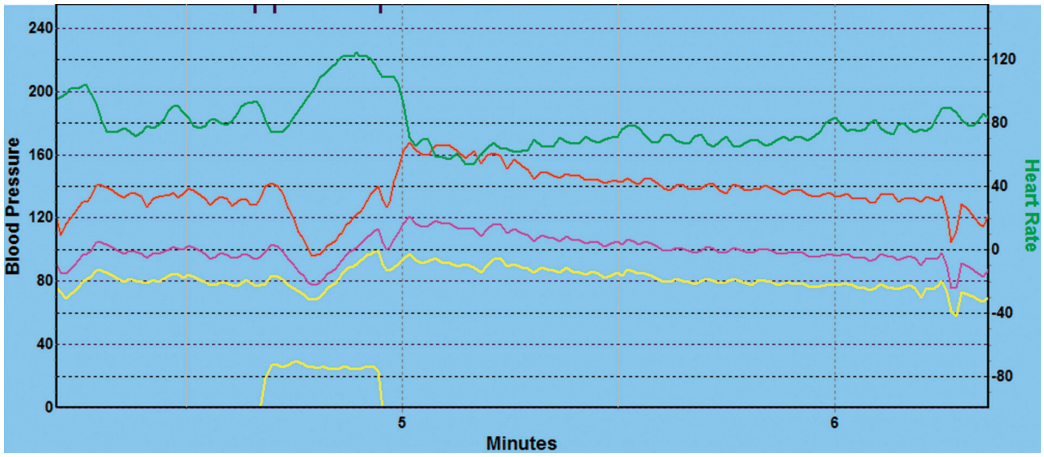


Fig. I.3. Normal tilt: very few hemodynamic changes occur in a normal subject



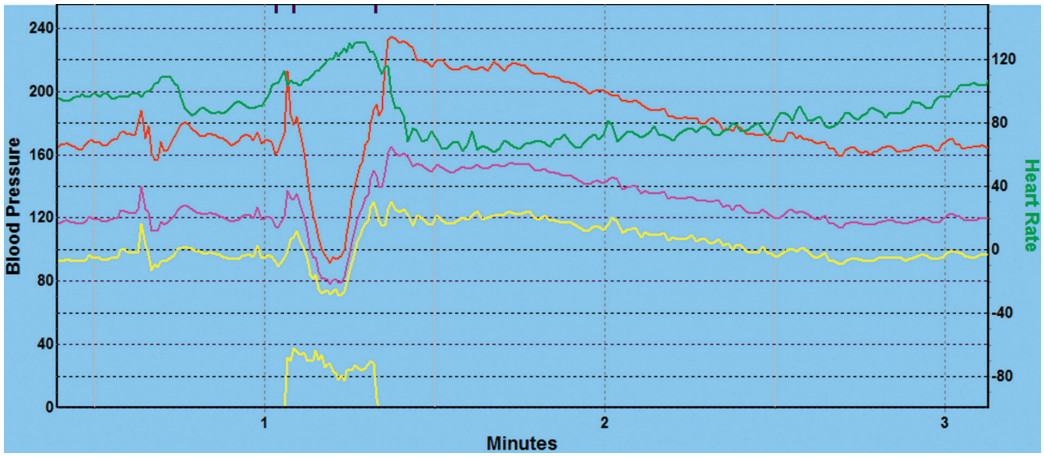


Fig. I.4. Two examples of normal Valsalva profiles: note the robust blood pressure rise during late phase II particularly in the second tracing, induced by a more prominent early phase II and pulse pressure compression suggesting volume contraction. Phase IV is also well demarcated. The heart rate shows a good acceleration during the maneuver followed by a rapid drop below baseline during phase IV.

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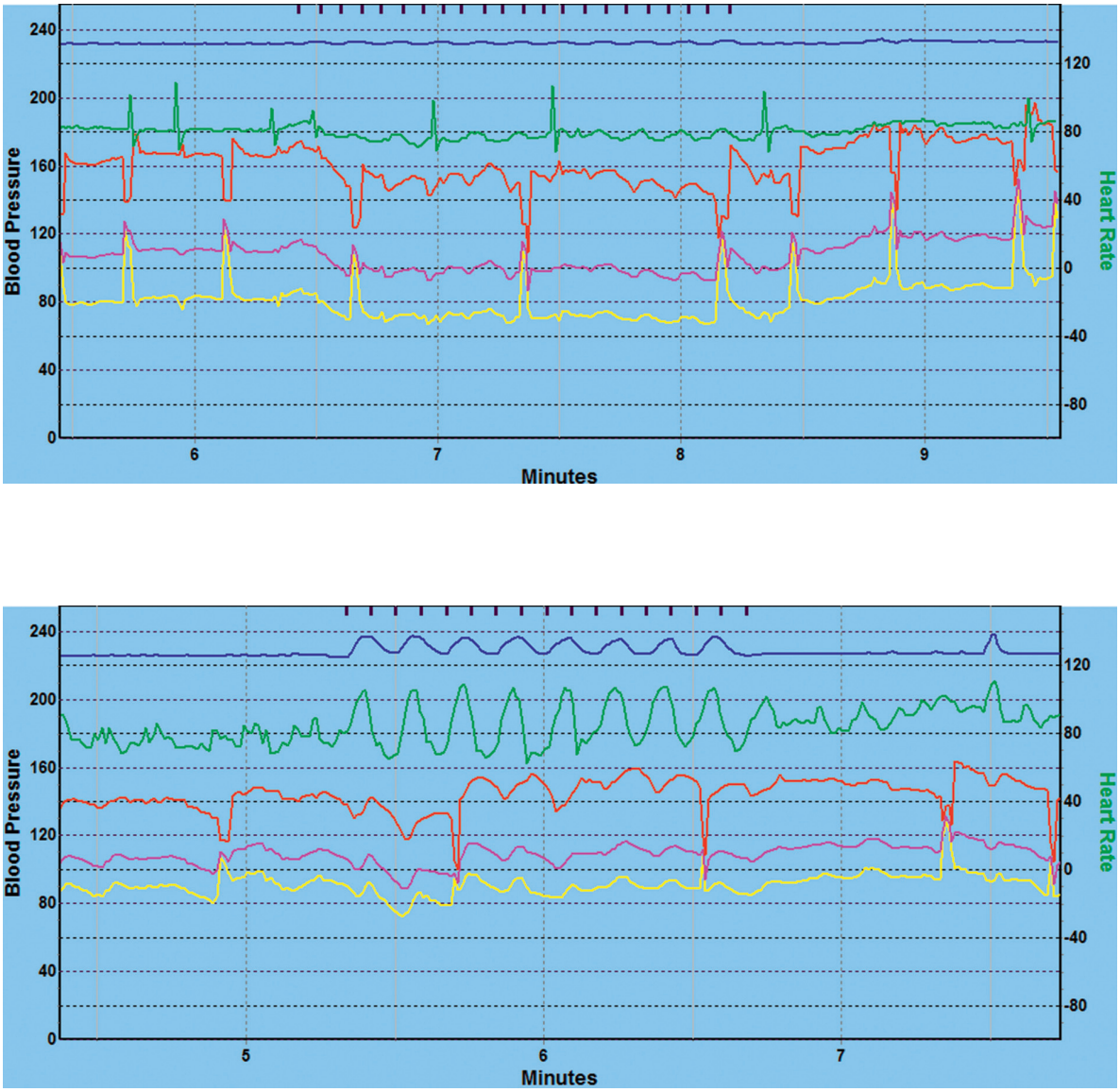

Fig. I.5. Deep breathing profile in an older subject (top) and a younger one: the amplitude of the heart rate oscillations diminishes with age. In comparison, in the young they can be > 40 beats per minute.

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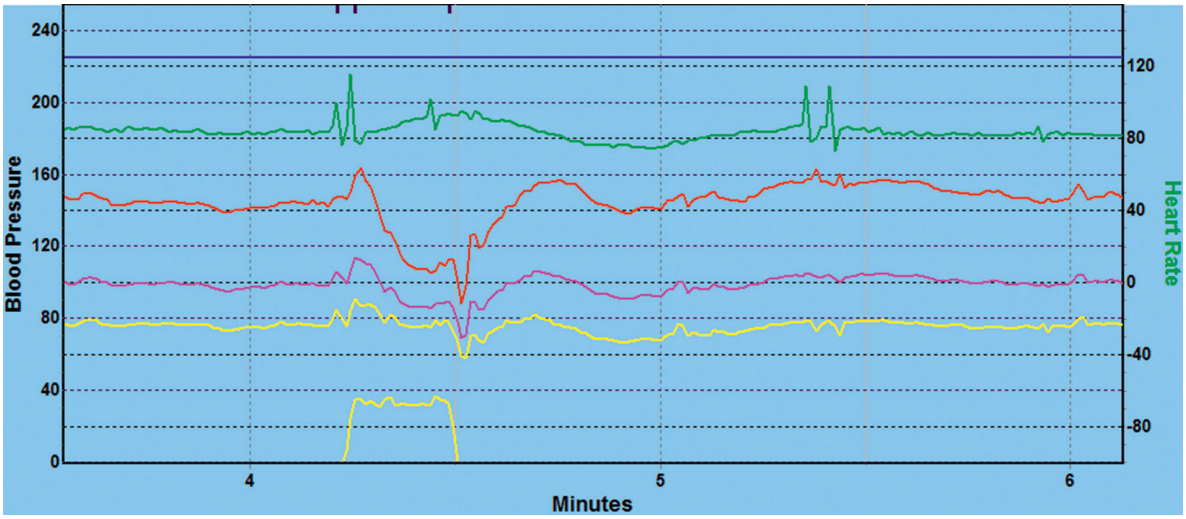

Fig. I.6. Valsalva maneuver in an older subject: late phase II rise may be blunted. It may be expression of mild dysautonomia of aging.

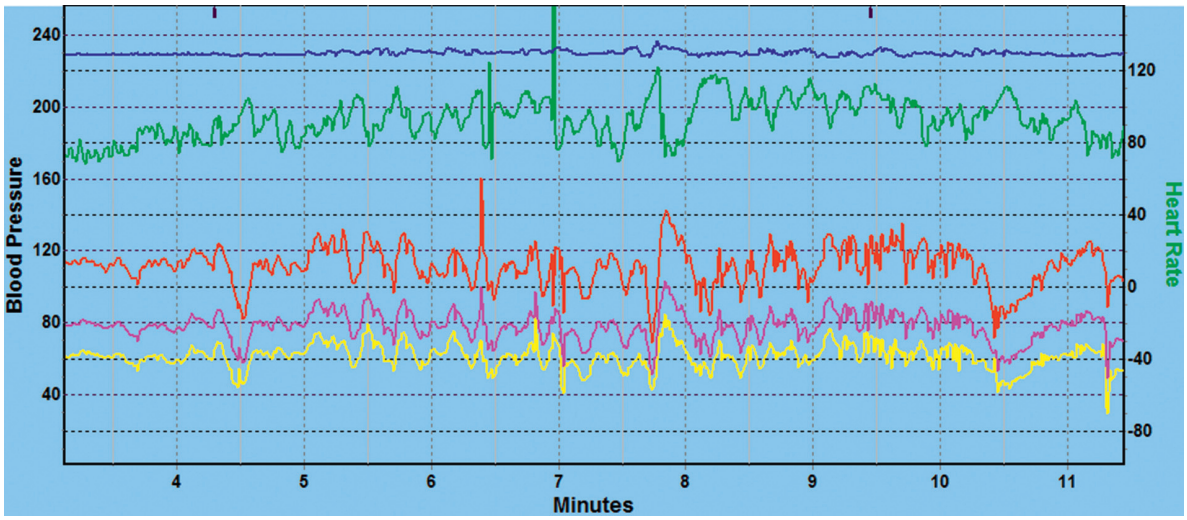

Fig. I.7. Tilt in a young subject: hemodynamic variations and oscillations are larger in the young when compared with adults.

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CASE 1 Prodromal possible Lewy body disease

History

A 44-year-old male presented with a history that began at the age of 37 years when he started to experience terrifying nightmares that he acted-out by thrashing his limbs or jumping out of bed. At the age of 42 years he began to complain of increasing light-headedness that worsened over time, resulting in multiple presyncopal episodes. Heat and activity worsen the symptoms. He noticed that he had stopped sweating over most of his body but reported increased sweating over his head. Except for mild constipation and bloating, he had no significant gastrointestinal symptoms. He felt he was not emptying his bladder fully and was experiencing erectile dysfunction not responsive to sildenafil. At the age of 43 years he began to have mild gait unsteadiness with a wide-based gait and frequent stumbling. Speech was unaffected as were his limb movements. There was no unequivocal cognitive difficulty. More recently he had been complaining of profound fatigue.

He had no significant past medical history except for a mild concussion at the age of 19 years. He worked as a welder and reported being exposed to various chemicals, including manganese. He also reported that two of his co-workers were diagnosed with multiple system atrophy.

The remaining history was unremarkable. His medications included midodrine, fludrocortisone, melatonin, and zolpidem.

Examination

Neurologic examination was normal except for a mild deficit on the heel-to-shin test.

Pertinent tests

Complete blood count, routine chemistry analysis, neuroimmunology panel, and thyroid-stimulating hormone-sensitive testing were normal. Plasma norepinephrine level was 116 pg/mL when supine and 204 pg/mL when standing (an inadequate rise considering his orthostatic hypotension). The patient had a urine output of 1600 cm^3 over 24 hours with 203 mEq/L of sodium excretion (suggesting excellent fluid and salt intake).

Polysomnography revealed the presence of REM behavior disorder. Neuropsychometric testing was mildly abnormal, with deficits mainly in auditory verbal learning and memory that were not specific for any neurodegenerative disorder.

Autonomic testing showed that cardiovagal function was mildly reduced (Fig. 1.1), there was abnormal

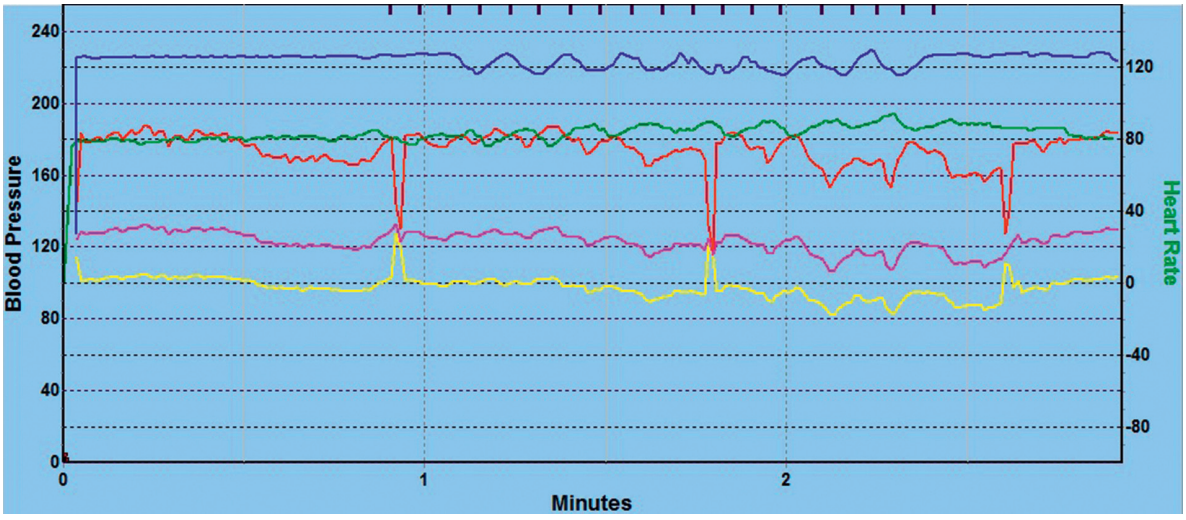

Fig. 1.1 Heart rate response to deep breathing was reduced for age.

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vasoconstriction during Valsalva with altered cardiac responses (Fig. 1.2), and identified the presence of orthostatic hypotension (Fig. 1.3). Sweating was only mildly abnormal on both QSART (Fig. 1.4) and TST (Fig. 1.5).

Comments

The case illustrates an example of what is most likely Lewy body disease [DLB] at an early stage. With such degree of abnormality on autonomic testing, multiple system atrophy [MSA] would have had more anhidrosis. DLB has more autonomic involvement than Parkinson's disease, but less than MSA as a group. Cognitive abnormalities are typically absent in MSA. There is no evidence that manganese or other toxins can cause DLB or MSA, while it can cause parkinsonism.

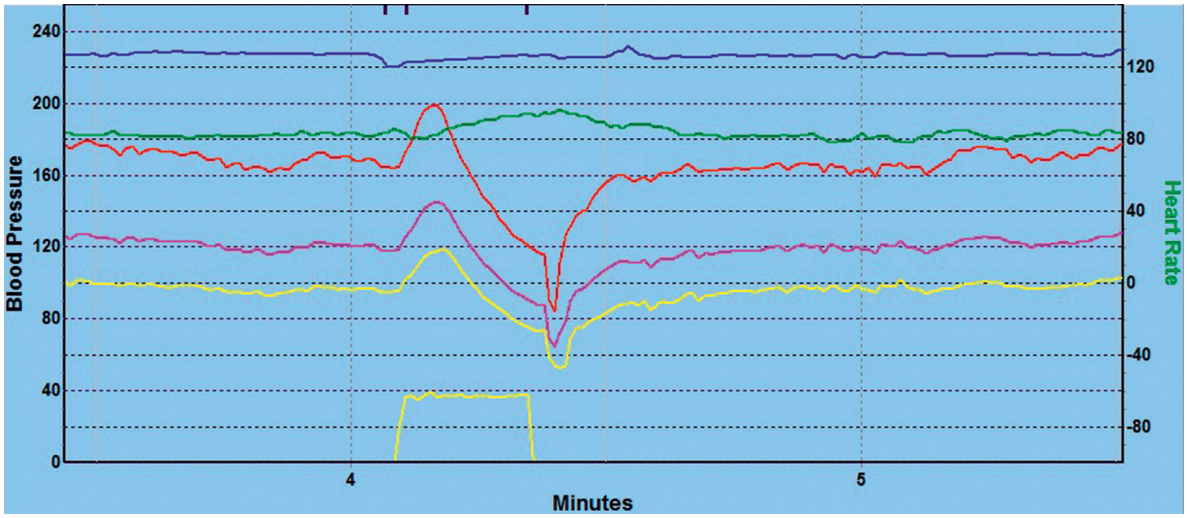

Fig. 1.2 Valsalva maneuver: there is reduced Valsalva ratio, indicating impaired cardiac responses, and the blood pressure profile is abnormal, with absence of late phase II and IV and prolonged recovery time.

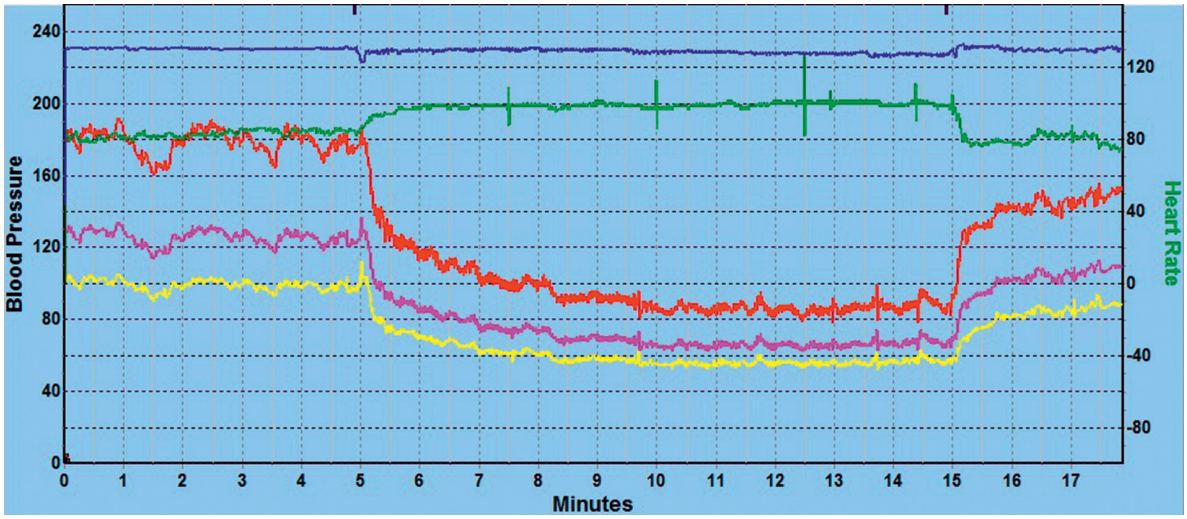

Fig. 1.3 Tilt study showed presence of orthostatic hypotension with attenuated cardiac responses.

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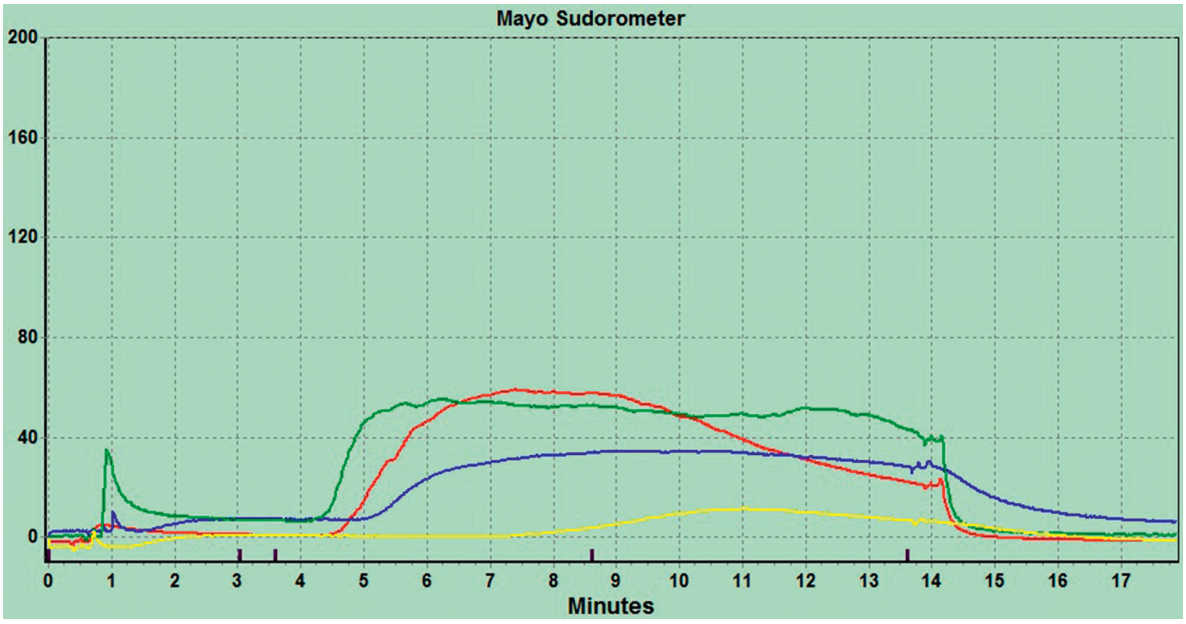

Fig. 1.4 QSART showed normal responses in all sites except in the foot, where it was reduced.

Fig. 1.5 TST showed a normal sweat pattern.

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CASE 2 Multiple system atrophy – cerebellar type

History

A 57-year-old female presented with an 18-month history of gait imbalance. She reported an insidious onset and evaluation at her local doctor's office revealed supine hypertension (new for her) as well as asymptomatic orthostatic hypotension. With the start of antihypertensive therapy, she began to experience severe light-headedness. Over the following months, she developed dysarthria, R > L upper-limb tremor, and worsening gait unsteadiness. Next she began to experience severe bladder urgency. She reported no change in her bowel pattern or sweating capacity. Although she

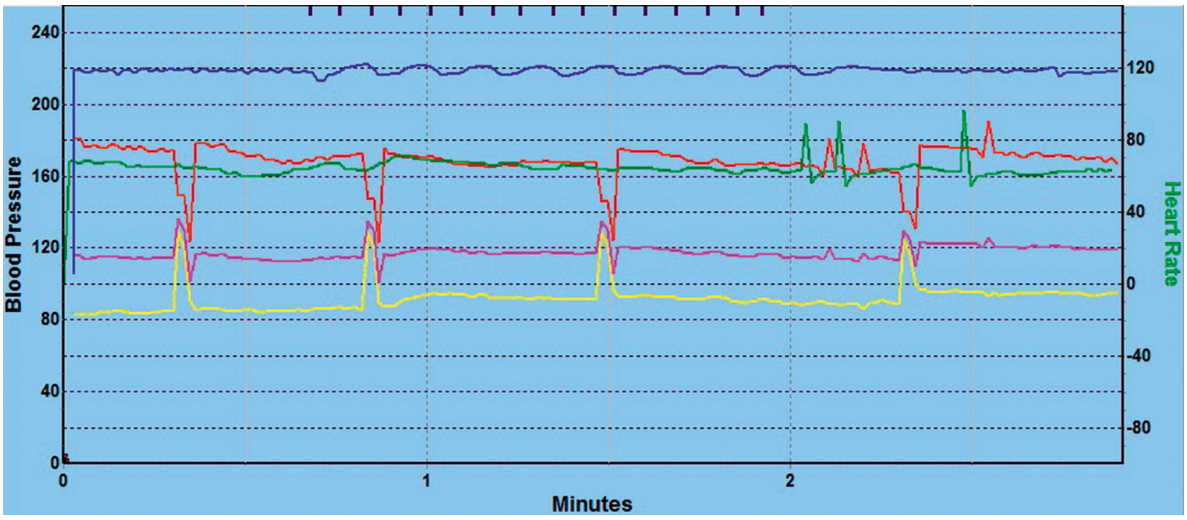

Fig. 2.1 Heart rate response to deep breathing was essentially absent.

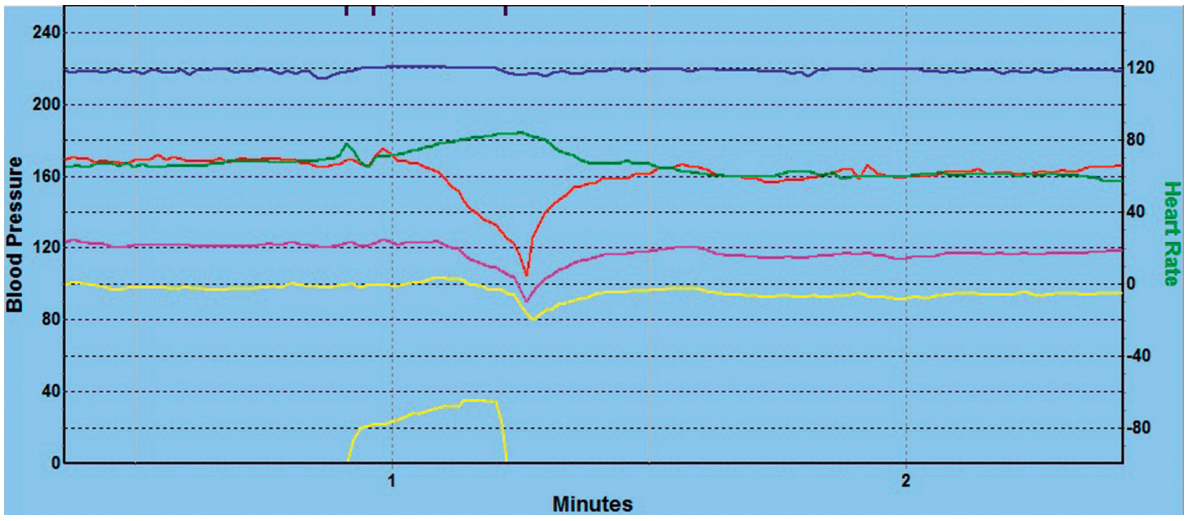

Fig. 2.2 Valsalva maneuver: there is reduced Valsalva ratio, indicating impaired cardiac responses, and the blood pressure profile is abnormal, with absence of late phase II and IV and prolonged recovery time.

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