

Autonomic Testing

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BACKGROUND

The conceptual framework for autonomic testing began at the turn of the 19th century with a number of experiments in basic neurophysiology. These original tests were developed over time into a rigorously defined, standardized series of autonomic tests that are useful in the clinical assessment and care of patients with suspected autonomic disorders. Autonomic testing has been widely used in clinical practice for 50 years, with decades of extensive experience and thousands of studies published on its use. Comprehensive textbooks have been published on the purpose and methodology of autonomic testing.¹⁻⁴

Autonomic testing is an umbrella term that covers testing of the various branches of the nervous system: the sympathetic, parasympathetic, and enteric. It should be noted that the autonomic nervous system extends to nearly every organ system in the body; so many organ specific tests are in fact tests of autonomic function (such as urodynamic studies, gastric motility testing, pupillometry, tests of lacrimal and salivary gland production, etc.). For the purposes of this

policy, we will focus on the standard tests of sudomotor (sympathetic cholinergic), cardiovagal (parasympathetic) and sympathetic adrenergic system function.

As is true for other accepted tests of neurophysiologic function and clinical monitoring technologies, randomized controlled trials establishing the efficacy of autonomic testing have not been done. However, current data, accumulated through scientific research and clinical experience and published in peer-reviewed journals over the past 50 years, have defined the role of autonomic testing in the diagnosis and management of autonomic disorders in clinical practice and established the value of autonomic testing in the longitudinal study of disease.

Autonomic testing is an integral component of the clinical evaluation of patients with autonomic disorders. Autonomic Disorders is an established subspecialty recognized by the United Council on Neurologic Subspecialties (UCNS), which certifies physicians and laboratories with training and expertise in this discipline.

COMMON AUTONOMIC TESTING TERMS

Autonomic Nervous System: The part of the nervous system that controls involuntary visceral actions.

Cardiovagal: The parasympathetic response measured via cardiac function, which is under control of the vagus nerve, which influences heart rate variability.

Heart Rate Variability: A test of parasympathetic function in which an individual undergoes a standard series of breathing exercises and the variability in the heart rate response is measured. Diminished heart rate variability (diminished respiratory sinus arrhythmia) is a sign of parasympathetic dysfunction.

Quantitative Sudomotor Axon Reflex Test: A test to evaluate the integrity of postganglionic sudomotor nerves along the axon reflex to define the volume and distribution of sweat loss. This is accomplished by releasing acetylcholine into the skin, which activates distal postganglionic sudomotor nerves, when then activates receptors on the eccrine sweat

gland. The sweat response is typically recorded from four sites (one forearm and three lower extremity sites) and the waveforms generated are assessed for deficits.

Sudomotor: The sympathetic cholinergic component of the autonomic nervous system is responsible for sweat gland function and the production of thermoregulatory sweating.

Sympathetic Skin Response: A test to measure a provoked change in the electrical potential of the skin.

Thermoregulatory Sweat Test (TST): A test of sweat function and its neurologic regulation in which a generalized thermoregulatory sweating response is elicited by raising the core body temperature and monitoring sweat production by an indicator dye applied to the skin of the whole body (typically the anterior aspect). When the core temperature rises beyond a hypothalamic thermoregulatory set point, sweating occurs. TST investigates the integrity of the central and peripheral thermoregulatory sympathetic pathways, from

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the hypothalamus to the eccrine sweat gland, by use of an indicator powder mixture (typically alizarin red or iodinated corn starch). When a core temperature of 38°C is achieved, normal sweat production is visualized by a change in color in the indicator powder. Digital photography is used to document the sweat distribution, which can be characteristic of neuropathy, ganglionopathy, or generalized autonomic failure.

Valsalva Maneuver: An autonomic testing maneuver in which the patient exhales against resistance and the blood pressure and heart rate are recorded, typically on a beat-to-beat basis. This test evaluates the complex sympathetic adrenergic and parasympathetic responses to the transient reduction in cardiac preload caused by an increase in intrathoracic pressure.

VARIATIONS IN METHODS OF TESTING, INCLUDING A DISCUSSION ABOUT AUTOMATED DEVICES

Autonomic testing using automated devices, in which software automatically generates an interpretation, has not been validated. Automated autonomic testing devices perform a simplified battery of autonomic tests—typically the heart rate response to deep breathing, the heart rate response to a Valsalva maneuver, and the blood pressure response to standing. Many devices are severely limited in the validity of the data they generate because they do not measure or control for expiratory pressure or include beat-to-beat blood pressure measurement, both of which are requirements, not only for scientifically accurate assessment, but also for billing of autonomic testing.^{5,6} For example, by failing to measure the expiratory pressure and blood pressure responses to a Valsalva maneuver, these devices generate meaningless heart rate data, since it is not possible to interpret the heart rate without information about its stimulus. Most of these devices generate reports automatically and do not allow physician interpretation of the raw data, which is a serious design flaw when evaluating patients who have, for example, cardiac rhythm abnormalities that mislead the testing results.

In contrast to state-of-the-art autonomic testing (as discussed in detail below), a review of the scientific literature reveals that autonomic testing by automated devices has not been validated by any presentations to our knowledge at scientific sessions of the American Academy of Neurology or the American Autonomic Society, nor does a search of the literature on PubMed discover any published data demonstrating that such automated testing is clinically meaningful.

Automated testing devices do not satisfy the conditions that are required for using the billing codes that were developed for autonomic testing. The CPT code 95922 requires a five-minute tilt table test and continuous beat-to-beat blood pressure monitoring in order to be billed. It is clear that the vast majority of bills from code 95922 using automated devices do not include beat-to-beat blood pressure testing and do not include a five-minute tilt table test. Furthermore, the new autonomic testing codes also require beat-to-beat blood pressure monitoring and a tilt table test. There are no automated devices currently on the market that, when used

alone, are sufficient to bill for autonomic testing using 95921–95924. Billing code 95943 is the only code appropriate for autonomic testing using automated devices.

Autonomic testing (CPT codes 95921–95922 and 95924) not using automated devices combines a battery of tests that typically includes the heart rate response to paced breathing, the heart rate response and continuous blood pressure response to a Valsalva maneuver, the heart rate response and beat-to-beat blood pressure response to a five-minute stand, and the beat-to-beat blood pressure response to a passive tilt table test of five minutes or longer. The data is rigorously reviewed and interpreted by a physician with expertise in understanding and interpreting the data in the appropriate clinical context. A vast number of medications may influence autonomic test results, and physicians who perform autonomic testing must be aware of the potential for misinterpretation of results in the context of polypharmacy. All of the extensive data in the literature about autonomic testing is based on these testing methods; extrapolating this literature to automatic devices that omit essential elements of autonomic testing as described would be scientifically unjustifiable.

In regard to the clinical utility of the well-established methods of autonomic testing discussed above, there has been no debate until recently. With the advent and increasing use of automated autonomic testing devices, physicians specializing in autonomic disorders are concerned that erroneous results obtained from devices of unproven scientific validity could adversely impact patient care. Additionally, payers have taken notice of increased billing for autonomic testing using these devices. Cost considerations have generated debate over autonomic testing in general. Whereas autonomic testing by the well-established methods in accordance with autonomic CPT codes is performed under carefully controlled conditions, can take 90–120 minutes to perform correctly, and requires interpretation by a physician familiar with autonomic nervous system physiology; automated testing devices are designed to generate data after approximately 10–15 minutes of testing and without physician interpretation. The automated testing devices are often utilized and billed for by physicians with no training in autonomic testing and no specialized expertise in

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autonomic nervous system physiology. This has resulted in misuse of the autonomic billing codes. Code 95922 requires both a five-minute tilt table test and beat-to-beat blood pressure monitoring in order to be billed. It is clear that the vast majority of bills from code 95922 using automated devices do not include beat-to-beat blood pressure testing

and do not include a five-minute tilt table test. Furthermore, the new combined autonomic testing code (95924) also requires beat-to-beat blood pressure monitoring and a tilt table test for use.

All autonomic testing using automated devices should be billed using code 95943 effective January 1, 2013.

QUALIFICATIONS OF PHYSICIANS WHO PERFORM AUTONOMIC TESTING

A number of automated testing devices have been developed over the past several years and advertised directly to non-neurologists and general practitioners who do not have training or expertise in the autonomic nervous system. Some of these automated devices may also generate patient-specific recommendations for treatment. As a result, physicians who do perform full autonomic testing have seen a large increase in the number of patients erroneously diagnosed with an autonomic disorder (unpublished data from the authors' practices). Prescribing unnecessary medications on the basis of an incorrect autonomic diagnosis could potentially harm patients because of the potential for serious adverse reactions. We have seen, for example, some patients treated with immune globulin for years for a diagnosis of autoimmune autonomic failure when all they had was a false positive automated autonomic test. Further, we have seen some patients who were told they have autonomic failure that is linked to a fatal neurologic disease (such as multiple system atrophy) when, in reality, they simply had an automated test that gave an incorrect result. Most physicians

without training in the autonomic nervous system do not know enough about many of the rare autonomic disorders to integrate the information supplied by automated devices into the clinical history of the individual in front of them.

Therefore, we strongly recommend that only physicians with the appropriate training perform and interpret clinical autonomic test results. Recommendations for required training are beyond the scope of this policy, but should follow the same guidelines for other tests of neurophysiology such as EMG, EEG, and evoked potentials. The United Council of Neurologic Subspecialties has established a board examination as one potential method to certify those individuals with expertise in autonomic disorders. In addition, some subspecialty societies are developing guidelines on what constitutes appropriate training for interpretation of autonomic test results. We also recommend that health insurance plans and exchanges allow for their patients to be covered for autonomic testing and consultation at centers where such expertise exists when it is not available within their network.

TECHNIQUES USED IN AUTONOMIC TESTING

Cardiovascular Autonomic Testing: Autonomic nervous system function testing for cardiovascular innervation has an enormous amount of clinical data supporting its use. It is the only reliable way to measure the function of the parasympathetic, or cardiovascular, nervous system.^{5, 7}

The American Diabetes Association (ADA) recommends that autonomic testing (including cardiovascular testing) be performed for all patients with type 2 diabetes mellitus at the time of diagnosis and five years after diagnosis in individuals with type 1 diabetes.^{8, 9} Those recommendations are based on evidence showing that individuals with diabetes that have evidence of cardiac autonomic neuropathy have significantly higher rates of mortality and silent myocardial ischemia. Guidelines for anesthesia, surgery and medical therapies to affect outcomes have been established for diabetic patients based on autonomic test findings.^{8, 10, 11} Cardiovascular testing has been demonstrated in a number of disease states to be an early marker of autonomic parasympathetic

dysfunction.¹²⁻¹⁵ Some disorders, such as amyloidosis and autoimmune autonomic ganglionopathy, preferentially affect autonomic nerve fibers and may not exhibit abnormalities of somatic nerve fiber tests (the latter detectable by nerve conduction tests and electromyography).^{16, 17} Heart rate variability is a simple and reliable test of cardiovascular function. It has a sensitivity of 97.5 percent for detection of parasympathetic dysfunction in diabetes when age-adjusted normative values are used.^{10, 18} The heart rate response to deep breathing, tilt table test, and the Valsalva maneuver are considered standard clinical tests of autonomic function and are sensitive, specific, and reproducible methods for grading the degree of autonomic dysfunction.⁵

Evaluation of parasympathetic function through cardiovascular testing has been firmly established for clinical use for decades.

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Vasomotor Adrenergic Autonomic Testing: Testing sympathetic adrenergic function is the primary method for evaluating patients with syncope, orthostatic hypotension, postural tachycardia syndrome, and postural dizziness.^{14, 19-26} Such testing is sensitive, specific, and clinically useful across diseases to diagnose patients with autonomic dysfunction. Sympathetic adrenergic testing (in conjunction with cardiovagal and sudomotor function testing) has been shown to aid in diagnosis, management, and outcomes in patients with autonomic dysfunction or syncope of unexplained cause.^{6, 26} Sympathetic adrenergic testing, when normal, is clinically useful also in ruling out autonomic failure when the history and bedside examination alone are diagnostically insufficient.

Autonomic testing (including adrenergic testing) is recommended for all patients with type 2 diabetes at the time of diagnosis and five years after diagnosis in individuals with type 1 diabetes.⁸ Individuals with diabetes that have cardiac autonomic neuropathy have a significantly higher mortality, and guidelines for anesthesia, surgery, and medical therapies to affect outcomes have been established.^{8, 10, 11}

Vasomotor adrenergic autonomic testing has been firmly established for clinical use for decades.

Sudomotor Testing: There are more than 60 years of data supporting the use of sudomotor testing in clinical practice. Following World War II, investigations of injuries to the nervous system involving sudomotor pathways led to the development of the quinizarin sweat test. The thermoregulatory sweat test (TST) is a sophisticated modification of the quinizarin sweat test that is widely used in the diagnosis of peripheral neuropathy. A large body of evidence exists to support its role in detecting and distinguishing among specific types of central nervous system disorders and peripheral neuropathies that impair sudomotor pathways, particularly when those disorders do not involve somatic sensory or motor nerve fibers.²⁷

The QSART device was first reported in detail in 1983 and has been in widespread clinical use ever since.²⁸ There are well established normative values and clinical guidelines for the use of this test.²⁷ The sympathetic skin response is another widely utilized test of sudomotor function.²⁹⁻³¹ The overwhelming amount of data supporting the use of QSART and SSR define these tests as medically necessary for the evaluation of autonomic dysfunction. QDIRT and silastic sweat imprint methods are also widely used, but they do not have the same level of clinical data supporting their use and at this time would be considered investigational.³² All of these tests measure only post-ganglionic sudomotor function, whereas the TST also assesses central nervous system sudomotor pathways.

Data from a considerable body of literature indicate that sudomotor testing may be the most sensitive means to detect a peripheral small fiber neuropathy.³³ Sudomotor testing is also the only way to detect isolated damage to sudomotor nerves, as it can occur in a number of specific disease states, including Ross syndrome, Harlequin syndrome, diabetes mellitus, multiple system atrophy, Parkinson's disease, autoimmune autonomic ganglionopathy, lepromatous leprosy, amyloid neuropathy, Sjögren syndrome, Fabry's disease, Lambert-Eaton myasthenic syndrome, pure autonomic failure, chronic idiopathic anhidrosis, and a host of other disorders that frequently come to the attention of the neurologist.^{28, 34-37} The clinical implications of testing and outcomes are reviewed in detail in a plethora of studies across numerous diseases.

Autonomic testing (including sudomotor testing) is recommended for all patients with type 2 diabetes at the time of diagnosis and five years after diagnosis in individuals with type 1 diabetes.^{8-10, 18, 38} Individuals with diabetes that have autonomic neuropathy have a significantly higher mortality, and guidelines for anesthesia, surgery and medical therapies to affect outcomes have been established.^{8, 10, 11}

WHAT TYPES OF PATIENTS WILL BENEFIT FROM AUTONOMIC TESTING?

Examples of the many situations where autonomic testing is of clinical utility include:

- Patients with syncope: Autonomic testing is necessary to differentiate neurally mediated syncope from neurogenic orthostatic hypotension and other causes of syncope.^{26, 39-43}
- Patients with diabetes mellitus: All patients with diabetes are recommended to have autonomic testing (sudomotor, cardiovagal and adrenergic) at diagnosis (type 2 diabetes) or five years after diagnosis (type 1 diabetes).^{8-10, 18, 38, 44} There is a high prevalence of cardiovascular autonomic

neuropathy in the diabetic population.⁴⁵⁻⁴⁷ The relationship between autonomic dysfunction and cardiovascular risk has been well documented and is important to monitor for patients planning major surgical procedures or considering moderate to high intensity physical exercise. This is the reason that the ADA recommends autonomic testing for all patients with type 2 diabetes at the time of diagnosis, and all patients with type 1 diabetes five years after diagnosis. The increased perioperative mortality in cardiovascular autonomic neuropathy is linked to greater blood pressure instability and hypothermia.⁴⁵⁻⁵⁰ This information may prompt high-risk patients to forgo an elective procedure or allow the anesthesiologist to

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prepare for potential hemodynamic changes, thereby reducing the risks of morbidity and mortality.^{49, 45-50}

- Patients with orthostatic dizziness: Patients with recurrent dizziness when standing may have autonomic dysfunction, postural tachycardia syndrome, or other autonomic neuropathy that can be treated if a diagnosis is made.^{17, 51-61} All autonomic tests (sudomotor, cardiovagal, and adrenergic) are appropriate to use in forming a differential diagnosis, defining the physiology of orthostatic intolerance in the individual patient, grading the severity of impairment, and directing appropriate therapy.
- Patients with disorders of sweating: Autonomic testing can provide a diagnosis which can lead to treatment of the underlying disorder and improvements in clinical outcomes.⁶²⁻⁷² Patients found to have global anhidrosis

may be at risk for heat exhaustion or heat stroke and can benefit from interventions to restore sweating, when a reversible cause is diagnosed, or otherwise from management strategies to avoid heat stress. Although sudomotor testing will provide specific information about the problem with sweating, cardiovagal, and adrenergic testing will narrow the differential diagnosis and are therefore integral parts of the autonomic test (i.e., does the patient have an autonomic ganglionopathy, an isolated autonomic neuropathy such as Ross syndrome, a peripheral neuropathy causing distal anhidrosis and proximal hyperhidrosis, how severe or anatomically widespread is the deficit, etc.).

- Patients with peripheral neuropathy from a number of different causes such as (but not limited to) amyloidosis, Fabry's disease, Sjögren's syndrome, and autoimmune neuropathies.⁷³

CLINICAL INDICATIONS FOR TESTING

The following indications are examples of appropriate situations in which to consider autonomic testing (note: this is not an exhaustive list):

- Evaluate orthostatic hypotension in a patient with dizziness, a drop in blood pressure, or syncope upon standing. Autonomic testing can assist the physician in distinguishing neurogenic from other causes of orthostatic hypotension, evaluate the severity of adrenergic failure, and assess for associated recumbent hypertension.
- In central neurodegenerative disorders, especially the synucleinopathies, distinguish multiple system atrophy from Parkinson's disease and diffuse Lewy body disease.
- Diagnose the presence of autonomic neuropathy in a patient with signs or symptoms suggesting a progressive autonomic neuropathy, including, but not limited to:
 - Diabetic neuropathy
 - Amyloid neuropathy
 - Sjögren's syndrome
 - Idiopathic neuropathy
 - Pure autonomic failure
 - Multiple system atrophy
- Evaluate the severity and distribution of a diagnosed progressive autonomic neuropathy.
- Differentiate certain complicated variants of syncope from other causes of loss of consciousness.
- Evaluate inadequate response to beta blockade in vasodepressor syncope.
- Evaluate distressing symptoms in the patient with a clinical picture suspicious for distal small fiber neuropathy in order to diagnose the condition when nerve conduction studies are normal.
- Diagnose and differentiate the cause of postural tachycardia syndrome.
- Evaluate change in type, distribution or severity of autonomic deficits in patients with autonomic failure.
- Evaluate the response to treatment in patients with autonomic failure who demonstrate a change in clinical exam.
- Diagnose axonal neuropathy or suspected autonomic neuropathy in the symptomatic patient.
- Evaluate and diagnose sympathetically maintained pain, as in reflex sympathetic dystrophy or causalgia.
- Evaluate and treat patients with recurrent unexplained syncope to demonstrate autonomic failure.

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DIAGNOSES WHERE TESTING IS INDICATED

The following disorders are known to impair the function of the autonomic nervous system. When clinically indicated, the following disorders may require testing of the autonomic nervous system (note: this is not an exhaustive list):

- Orthostatic hypotension
- Multiple system atrophy
- Lewy body disease
- Parkinson's disease
- Pure autonomic failure
- Cerebellar ataxia
- Syncope
- Orthostatic intolerance including postural tachycardia syndrome (POTS)
- Anhidrosis
- Autoimmune autonomic neuropathy/ganglionopathy
- Idiopathic autonomic neuropathy
- Paraneoplastic autonomic neuropathy
- Chagas disease
- Intestinal pseudo-obstruction (if generalized or cholinergic neuropathy is suspected)
- Autonomic dysreflexia
- Familial dysautonomia or Riley Day syndrome
- Baroreflex failure
- Hereditary sensory and autonomic neuropathies
- Ross syndrome
- Lambert Eaton myasthenic syndrome
- Morvan syndrome
- Guillain-Barré syndrome

LIMITATIONS

Autonomic testing evaluates the physiologic responses to various stimuli. It is not a test for a specific disease; instead it investigates the degree of dysfunction of this part of the nervous system. This can aid in localization of the dysfunction, narrowing of the differential diagnosis, and in some cases making a clear diagnosis. Autonomic testing may not be able to differentiate between similar disorders that both cause autonomic dysfunction, but can quantify the severity of autonomic dysfunction, in other cases exclude autonomic dysfunction, and thereby lead to specific recommendations for treatment.

Autonomic testing can make a diagnosis of orthostatic hypotension, delayed orthostatic hypotension, neurally mediated syncope, or postural tachycardia syndrome. However, autonomic testing cannot always differentiate between the various disorders that cause these syndromes. The role of autonomic testing in this situation is to define, clarify, and gauge the severity of the problem so that the appropriate work-up and treatment can be instituted.

INDICATIONS FOR WHICH DATA IS LIMITED

The following are a list of indications for which data is limited at the time of writing this template policy (note: this is not an exhaustive list):

- Myofascial pain syndrome/fibromyalgia
- Raynaud's phenomenon
- Predicting foot ulcers
- Flushing syndrome
- Chronic fatigue NOS
- Generalized hyperhidrosis
- Palmoplantar hyperhidrosis
- Irritable bowel syndrome
- Somatization disorder
- Anxiety disorder

CPT CODES

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Code 95924 should be reported only when both the parasympathetic function and the adrenergic function are tested together with the use of a tilt table.

To report autonomic function testing that does not include beat-to-beat recording, or for testing without use of a tilt table, use 95943.

95921* Testing of autonomic nervous system function; cardiovagal innervation (parasympathetic function), including two or more of the following: heart rate response to deep breathing with recorded R-R interval, Valsalva ratio, and 30:15 ratio

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95922 Vasomotor adrenergic innervation (sympathetic adrenergic function), including beat-to-beat blood pressure and R-R interval changes during Valsalva maneuver and at least 5 minutes of passive tilt

Do not report 95922 in conjunction with 95921

95923 sudomotor, including one or more of the following: quantitative sudomotor axon reflex test (QSART), silastic sweat imprint, thermoregulatory sweat test, and changes in sympathetic skin potential

95924 combined parasympathetic and sympathetic adrenergic function testing with at least 5 minutes of passive tilt

Do not report 95924 in conjunction with 95921 or 95922

95943 Simultaneous, independent, quantitative measures of both parasympathetic function and sympathetic function, based on time-frequency analysis of heart rate variability concurrent with time-frequency analysis of continuous respiratory activity, with mean heart rate and blood pressure measures, during rest, paced (deep) breathing, Valsalva maneuvers, and head-up postural change

Do not report 95943 in conjunction with 93040, 95921, 95922, 95924

DISCLAIMER

AAN's Model Coverage Policies must be used by AAN members in an appropriate manner and not for engaging in anti-competitive activities. Antitrust laws involve complex legal and economic analysis beyond the scope of Model Coverage Policies; but they are integral in the overall consideration of building payer relationships and advocating for improving payer policies. No material contained in AAN Model Coverage Policies should be construed as legal advice. Neurologists are encouraged to seek advice from attorneys with expertise in payer relations.

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APPENDIX A—DIAGNOSES THAT SUPPORT MEDICAL NECESSITY

Note: All ICD-9-CM and ICD-10-CM codes listed below may be viewed as medically necessary; however, there may be other diagnostic codes not included in this list that are deserving of consideration for coverage. Such instances may require individual consideration.

• ICD-9-CM

- 458.0 Orthostatic hypotension
- 780.2 Syncope
- 250.60–63, 357.2 Diabetic neuropathy
- 356.9 Idiopathic neuropathy
- 356.2 Hereditary sensory neuropathy
- 337.00 Idiopathic peripheral autonomic neuropathy
- 337.9 Unspecified disorder of the autonomic nervous system
- 333.0 Degenerative disease of the basal ganglia
- 785.0 Postural tachycardia

• ICD-10-CM

- I95.1 Orthostatic hypotension
- R55 Syncope and collapse
- E11.40 Type 2 diabetes mellitus with diabetic neuropathy, unspecified
- E11.41 Type 2 diabetes mellitus with diabetic mononeuropathy
- E11.42 Type 2 diabetes mellitus with diabetic polyneuropathy
- E11.43 Type 2 diabetes mellitus with diabetic autonomic (poly)neuropathy
- E11.44 Type 2 diabetes mellitus with diabetic amyotrophy
- E11.49 Type 2 diabetes mellitus with other diabetic neurological complication
- E11.610 Type 2 diabetes mellitus with diabetic neuropathic arthropathy
- E13.40 Other specified diabetes mellitus with diabetic neuropathy, unspecified
- E13.41 Other specified diabetes mellitus with diabetic mononeuropathy
- E13.42 Other specified diabetes mellitus with diabetic polyneuropathy
- E13.43 Other specified diabetes mellitus with diabetic autonomic (poly)neuropathy
- E13.44 Other specified diabetes mellitus with diabetic amyotrophy
- E13.49 Other specified diabetes mellitus with other diabetic neurological complication
- E13.610 Other specified diabetes mellitus with diabetic neuropathic arthropathy
- E10.40 Type 1 diabetes mellitus with diabetic neuropathy, unspecified
- E10.41 Type 1 diabetes mellitus with diabetic mononeuropathy
- E10.42 Type 1 diabetes mellitus with diabetic polyneuropathy
- E10.43 Type 1 diabetes mellitus with diabetic autonomic (poly)neuropathy
- E10.44 Type 1 diabetes mellitus with diabetic amyotrophy
- E10.49 Type 1 diabetes mellitus with other diabetic neurological complication

- E10.610 Type 1 diabetes mellitus with diabetic neuropathic arthropathy
- E11.40 Type 2 diabetes mellitus with diabetic neuropathy, unspecified
- E11.65 Type 2 diabetes mellitus with hyperglycemia
- E10.40 Type 1 diabetes mellitus with diabetic neuropathy, unspecified
- E10.65 Type 1 diabetes mellitus with hyperglycemia
- E08.42 Diabetes mellitus due to underlying condition with diabetic polyneuropathy
- E09.42 Drug or chemical induced diabetes mellitus with neurological complications with diabetic polyneuropathy
- E10.42 Type 1 diabetes mellitus with diabetic polyneuropathy
- E11.42 Type 2 diabetes mellitus with diabetic polyneuropathy
- E13.42 Other specified diabetes mellitus with diabetic polyneuropathy
- E08.40 Diabetes mellitus due to underlying condition with diabetic neuropathy, unspecified
- E09.40 Drug or chemical induced diabetes mellitus with neurological complications with diabetic neuropathy, unspecified
- E10.40 Type 1 diabetes mellitus with diabetic neuropathy, unspecified
- E11.40 Type 2 diabetes mellitus with diabetic neuropathy, unspecified
- E13.40 Other specified diabetes mellitus with diabetic neuropathy, unspecified
- G60.9 Hereditary and idiopathic neuropathy, unspecified
- G60.0 Hereditary motor and sensory neuropathy
- G90.09 Other idiopathic peripheral autonomic neuropathy
- G90.9 Disorder of the autonomic nervous system, unspecified
- G90.8 Other disorders of autonomic nervous system
- G23.0 Hallervorden-Spatz disease
- G23.1 Progressive supranuclear ophthalmoplegia [Steele-Richardson-Olszewski]
- G23.2 Striatonigral degeneration
- G23.8 Other specified degenerative diseases of basal ganglia
- G23.9 Degenerative disease of basal ganglia, unspecified
- G90.3 Multi-system degeneration of the autonomic nervous system
- R00.0 Tachycardia, unspecified

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