CD Part 9

Hormone Disorders

A. Definitions

The following definitions are for words used in this chapter and during the SSA disability process. If you need additional definitions, consult a good medical dictionary, available in most bookstores and libraries. You can also look at online medical dictionaries like the one at www.medlineplus.gov.

Acidosis. Abnormal condition, characterized by an increase in the blood's acidity. See *ketoacidosis*.

Addison's disease. A form of adrenal insufficiency, characterized by a decreased production of cortisol.

Adrenal glands. Hormone-producing glands sitting on top of each kidney. The outer layers of the adrenal glands (cortex) secrete the natural steroid hormone cortisol. The adrenal glands also secrete aldosterone, epinephrine, norepinephrine, and normally small amounts of sex hormones.

Adrenal insufficiency. Decreased hormone output by the adrenal glands, usually in reference to decreased secretion of cortisol.

Adrenalectomy. Removal of part or all of an adrenal gland.

Adrenogenital syndrome. A disorder characterized by increased adrenal gland production of hormones that increase male sex characteristics (adrenal androgens). In boys, the effect is the production of early and excessive early (precocious) male sex characteristics; in girls, the effect is the production of both male and female sex characteristics (pseudohemaphroditism).

Aldosterone. Adrenal gland hormone important in maintaining blood pressure and blood volume by causing the kidneys to retain sodium and water. The right amount of aldosterone is important, but excess amounts can lead to high blood pressure. Excess aldosterone production is called hyperaldosteronism and a deficiency of aldosterone is known as hypoaldosteronism.

Androgens. Hormones that induce male sex characteristics.

Antidiuretic hormone. Hormone released from the pituitary gland that causes the kidneys to save body water. Also known as *vasopressin*.

Bone densitometry. Several possible x-ray methods of measuring bone density to determine if there is osteoporosis.

Calcitonin. A hormone secreted from the thyroid gland whose actions decrease blood calcium levels. It has the opposite effect as parathyroid hormone.

Cataract. Degeneration of the lens of the eye so that light cannot easily pass through it. Most cataracts are related to aging, but some date from birth (congenital cataracts) or from the use of medication, such as the chronic use of steroid drugs. Cataracts are sometimes described by doctors as lens opacity.

Congestive heart failure. Failure of the heart as a pump—the heart cannot keep up with pumping out the blood flowing into it. Consequently, the heart is enlarged and there is fluid congestion of organs such as the liver and lungs.

Convulsions. Involuntary contractions of muscles, related to abnormal electrical activity in the brain. Also known as *seizures*.

Corticosteroids. Hormones secreted by the outer layer (cortex) of the adrenal glands. Corticosteroids are divided into glucocorticoids such as cortisol and mineralocorticoids such as aldosterone. Glucocorticoids influence the metabolism of carbohydrates, fats, and proteins. Mineralocorticoids are important in maintaining the body's water and electrolyte (especially sodium and potassium) balance. Hormones with sexual activity are usually excluded from the classification as corticosteroids although they are also normally produced in small amounts by the adrenal cortex (see *adrenogenital syndrome*).

Cushing's syndrome. A group of physical abnormalities associated with the effects of glucocorticoid hormones such as cortisol. Cushing's syndrome can be caused either by increased output of cortisol from diseased adrenal glands or by excessive long-term intake of glucocorticoid drugs like prednisone.

Diabetes insipidus. A condition associated with excessive loss of body water through the kidneys as a result of decreased antidiuretic hormone (ADH). ADH is normally secreted by the back part of the pituitary gland (neurohypophysis). Also known as *neurohypophyseal insufficiency* and *neurogenic* or *central diabetes insipidus*. (Another condition, nephrogenic diabetes insipidus, is not due to lack of ADH, but is caused by kidney disease that makes the kidneys nonresponsive to ADH. Most kidney diseases

are potentially capable of causing nephrogenic diabetes insipidus.)

Diabetes mellitus. A disorder of glucose metabolism caused by insufficient insulin; may also involve the cellular resistance to insulin that is present.

Diabetic necrosis. Death of tissue as a result of uncontrolled diabetes.

Dysgenesis. Defective development.

Endocrine. Refers to the hormone system of the body.

Exophthalmometry. Measurement of the amount of exophthalmos, measured with an instrument called an exophthalmometer.

Exophthalmos. A bulging outward of the eyes, a possible complication of hyperthyroidism.

Fasting blood glucose (FBG). Blood glucose measured after at least an overnight fast. In nonpregnant adults FBG should be less than 140 mg/dl. Also called fasting blood sugar (FBS).

Glucose. A simple sugar that is the body's main source of energy.

Goiter. An enlarged thyroid gland sufficient to cause a visible swelling in the front of the neck.

Gonad. Reproductive organ—ovary or testis.

Gonadal dysgenesis. See *Turner's syndrome.*

Heart failure. See *congestive heart failure.*

Hirsutism. Abnormal hairiness—an effect of excessive male sex hormones on children or women.

Hormone. Any of a number of chemicals produced naturally in the body that regulate the activity of glands, organs, or cells.

Hypercalcemia. Abnormally high blood calcium. Normal serum calcium is about 8–10 mg/dl.

Hyperglycemia. Abnormally high blood glucose.

Hypernatremia. Abnormally high blood sodium.

Hyperparathyroidism. Disorder in which there is excessive production of parathyroid hormone (PTH).

Hypertension. High blood pressure. Hypertension usually means systemic hypertension, or high blood pressure in the arterial system of the body other than the lungs. Hypertension in adults is defined as any pressure of 140/90 or greater. In children, normal expected blood pressure varies with age.

Hyperthyroidism. Disorder in which there is excessive production of thyroid hormone.

Hypocalcemia. Abnormally low blood calcium. When blood calcium falls to about 7 mg/dl, a life-threatening situation is present.

Hypoglycemia. Abnormally low blood glucose. Hypoglycemia is present with blood glucose levels of less than 50 mg/dl in adult males following a prolonged fast, but in women, children, and infants glucose levels may drop below this value without symptoms, and in these instances the diagnosis of hypoglycemia based on blood sugar alone might be inappropriate.

Hypoparathyroidism. Disorder in which there is a deficiency of parathyroid hormone (PTH).

Hypothalamus. An area at the bottom of the brain with a number of functions, including the production of various types of *releasing hormones* that trickle down the pituitary stalk into the pituitary gland and cause it to in turn release other hormones. For example, thyrotropin-releasing hormone (TRH) is made in the hypothalamus and stimulates the pituitary gland to release thyroid stimulating hormone (TSH), which in turn stimulates the thyroid gland in the neck to release thyroid hormone.

Hypothyroidism. Disorder in which there is a deficiency in thyroid hormone. Also known as *myxedema.*

Iatrogenic. Reference to disorders caused by treatment.

Insulin. A pancreatic hormone that circulates in the bloodstream to the cells of the body. Taken into cells, insulin is necessary for the metabolism of glucose for energy.

Insulin dependent diabetes mellitus (IDDM). Diabetes mellitus is that which requires treatment by insulin injection for proper control.

Intracranial. Inside the head.

Ketoacidosis. Abnormal acidity of the blood resulting from poorly controlled Type I diabetes mellitus.

Ketosis. See *ketoacidosis.*

Metabolism. The total chemical and physical activity of the body associated with the production and maintenance of life.

Myopathy. Any disorder affecting muscle tissue; myopathy results in muscle weakness.

Neovascularization. New blood vessel growth. The word is most frequently used to refer to new blood vessel growth in the retina of the eye as a result of uncontrolled diabetes mellitus. See *proliferative retinopathy.*

Nephrogenic diabetes insipidus. See *diabetes insipidus*.

Neurohypophyseal insufficiency. An abnormal condition associated with excessive loss of body water through the kidneys as a result decreased antidiuretic hormone (ADH). ADH is normally secreted by the neurohypophysis. Also known as *diabetes insipidus*.

Neurohypophysis. The back part of the pituitary gland.

Neuropathy. Any disease of nerves, usually taken to mean peripheral nerves. Peripheral nerves are those connecting the spinal cord to the various organs and tissues of the body. Neuropathy is best demonstrated by weakness, decreased reflexes, loss of sensation, and decreased nerve conduction velocity (NCV). Motor neuropathy means affecting the motor nerves that carry impulses away from the spinal cord to stimulate muscles. Sensory neuropathy means affecting the sensory nerves that carry touch, pain, vibration, limb position, heat, and cold sensations from the tissues of the body to the spinal cord for transmission to the brain. Not every type of sensation need be affected by the neuropathy.

Non-insulin dependent diabetes mellitus (NIDDM). Diabetes mellitus that does not require control by treatment with insulin injections. NIDDM is most likely to apply to adult-onset (Type II) diabetes mellitus.

Obesity. The excessive accumulation of body fat significantly beyond what is necessary for health. Some authorities attempt to define overweight, obesity, morbid obesity, etc., in terms of specific weights related to a person's sex and height. However, such naming systems are completely arbitrary and not universally accepted.

Osteoporosis. Loss of bone mass—that is, a thinning of bone substance.

Pancreatitis. Inflammation of the pancreas.

Parathyroid glands. Small glands located inside of the thyroid gland that produce parathyroid hormone.

Parathyroid hormone (PTH). A hormone important in the way the body regulates the use of calcium. PTH tends to raise blood calcium levels and has the opposite effect as calcitonin. Also known as *parathormone*. Normal PTH values are about 10–50 or 60 picograms/milliliter (pg/ml), increasing with age.

Peripheral arterial disease. Any disease of the arteries in the arms or legs.

Peripheral neuropathy. See *neuropathy*.

Pitressin. Trade name for a synthesized drug that is exactly like human antidiuretic hormone.

Pituitary gland. Pea-sized gland that hangs down from the bottom of the brain on a stalk. It produces a wide range of important hormones, including sex hormones, adrenocorticotropic hormone that stimulates the adrenal glands to release cortisol, growth hormone, thyroid stimulating hormone, antidiuretic hormone that causes the kidneys to conserve water, and oxytocin, which is important in uterine muscle contraction during delivery and expression of milk from the breast during suckling.

Precocious puberty. See *adrenogenital syndrome* and *virilization*.

Proliferative retinopathy. Retinal disease resulting from poorly controlled diabetes mellitus, with excessive new blood vessel growth (neovascularization) and an increased risk of retinal detachment and bleeding into the eye.

Pseudohemaphroditism. See *adrenogenital syndrome* and *virilization*.

Pseudohypoparathyroidism (PHP). Genetic disorder in which the cells for receiving parathyroid hormone contain defective receptors.

Purple striae. Purple abdominal skin markings that may be associated with Cushing's syndrome.

Random blood glucose (RBG). Blood glucose measured without consideration of what the person ate before the test or when it was eaten. In a nonpregnant adult or child, the RBG should not be greater than 200 mg/dl. Also known as random blood sugar (RBS).

Standard deviation. A statistical calculation expressing the amount of deviation of a value from average.

T3 resin uptake. A test of thyroid function; should not be confused with a thyroid scan/uptake.

Tetany. Tetany refers to muscular twitching, cramping, and spasms caused by hypocalcemia. There may be sudden violent contractions of muscles in the larynx (laryngospasm) with difficulty breathing and spasm of muscles in the hands and feet (carpopedal spasm).

Thyroid gland. Gland in the front part of the neck that produces thyroid hormones under the stimulation of thyroid stimulating hormone (TSH) released from the pituitary.

Thyroid hormones. The major thyroid hormones are *thyroxine (T4)* and *triiodothyronine (T3)*. Synthetic forms of these hormones are available to treat hypothyroidism and can be taken as small tablets once a day. Thyroid hormones stimulate the DNA in the nuclei of cells to increase protein synthesis and oxygen consumption and are therefore vital to the body's metabolic activities.

Thyroid scan/uptake. A test of thyroid function by administering radioactive iodine and scanning the thyroid gland. The amount of radioactivity emitted by the thyroid indicates how well it takes up iodine.

Thyroid stimulating hormone (TSH). Hormone released by the pituitary gland to stimulate the thyroid gland to release thyroid hormones. Thyroid hormones in turn feed back onto the pituitary gland and decrease TSH release. High TSH levels indicate hypothyroidism.

Thyroid ultrasound. A method of making images of the thyroid gland using high-frequency sound. It is a harmless and painless test.

Thyroidectomy. Partial or total surgical removal of the thyroid gland.

Trachea. Windpipe.

Turner's syndrome. An inherited disorder of females involving abnormal combination of sex chromosomes.

Vasopressin. See *antidiuretic hormone.*

Virilization. Abnormality associated with the adrenogenital syndrome and refers to the development of male sexual characteristics—such as hirsutism, development of a deep voice, male hair patterns on the scalp, and acne, as well as accelerated bone, muscle, and genital development. Accelerated male sexual characteristics in boys results in precocious puberty. In girls, the development of male sexual characteristics can result in genital abnormalities, such as enlargement of the clitoris to appear more like a small penis. The development of male sexual characteristics along with female characteristics is known as pseudohemaphroditism. Virilization is also known as masculinization.

B. General Information

Endocrine disorders affect the body's structure or functions due to too much or too little hormone production. If the hormone disorder affects the function of other organ systems, evaluation should be done under the appropriate listing. For example, increased thyroid hormone could result in an abnormally fast heart rate and heart failure. That part of the impairment would be evaluated under the listings dealing with heart disease (CD Part 4). Lack of insulin could cause retinal disease with loss of vision. That disorder would be evaluated under the listings for visual impairments (CD Part 2). Many hormonal disorders respond to medical treatment, particularly if the only action necessary is replacement of the deficient hormone.

To qualify under the listings, hormonal abnormalities of the required severity must have persisted or be expected to persist, despite therapy, for at least 12 months. The SSA will require a description of the hormone disorder, physical findings, and diagnostic laboratory tests. Because of the way labs test for hormone levels, they generally have a wider range of normal for hormones than for other substances. To make up for this variation, the SSA considers the results abnormal if they are outside the normal range or greater than two standard deviations from the average of the testing laboratory. Because of this, the lab report should include information provided by the testing laboratory as to its normal values for the test.

When a listing requires an abnormal test result, such as an elevated hormone level, a number of tests over three months is necessary to establish an impairment likely to last 12 months.

C. Specific Listings and Residual Functional Capacity

The listings that follow are in the federal regulations. They have been interpreted and commented on for greater ease of understanding while explaining their requirements. It is impossible to discuss here all of the medical possibilities related to every kind of disorder, and you may need to seek help from your

treating doctor to more fully understand how your particular impairment relates to these listings. The discussion of residual functional capacity does not apply to children.

1. Listing 9.02: Thyroid Disorders (Adults)

The thyroid gland is located in the lower front of the neck, on both sides of the trachea. The major thyroid hormones are thyroxine (T4) and triiodothyronine (T3). Thyroid hormones stimulate the DNA in the nuclei of cells to increase protein synthesis and oxygen consumption and are therefore vital to the body's metabolic activities. Thyroid hormones are released from the thyroid gland into the bloodstream under the influence thyroid-stimulating hormone (TSH) from the pituitary gland.

There are many types of specific thyroid disorders, but this listing concerns hyperthyroidism and hypothyroidism. The most common form of hyperthyroidism is Graves' disease, also known as diffuse toxic goiter, and this disorder is frequently seen by the SSA. Hyperthyroidism can almost always be controlled with drugs, radioactive iodine to suppress the function of the gland, or with surgery. Hypothyroidism is also a common disorder, but is almost always easily treatable with a tiny once-a-day tablet of synthetic human thyroid hormone; such hormone replacement is very safe if the proper dose is given.

In hyperthyroidism, TSH tends to be low and thyroid hormones elevated. In hypothyroidism, TSH tends to be elevated and thyroid hormones low. However, it is possible to have abnormal thyroid tests and actually have normal thyroid function, if a patient is physiologically stressed such as by surgery or trauma.

Possible abnormalities occurring with hyperthyroidism include nervousness, tremors, goiter, abnormal heart rhythms such as a fast heart rate, heart failure, increased sweating, weight loss, increased reflexes, weakness, fatigue, diarrhea, and exophthalmos. Exophthalmos can result in blurry or double vision.

Possible abnormalities occurring with hypothyroidism include slowed thinking, decreased reflexes, slowed heart rate, weight gain, cold intolerance, heart failure, weakness, fatigue, physical sluggishness, hair loss, enlarged tongue, coarse thick skin, puffy facial features, and enlarged thyroid gland. In extreme cases, coma may result.

In adults, total T4 is about 4–11 micrograms per deciliter (4–11 mcg/dl), but the SSA uses the normal values reported by the testing laboratory to confirm the diagnosis or determine the adequacy of control by treatment.

a. Listing Level Severity

Impairments resulting from thyroid disorders should be evaluated under whatever listings are appropriate for the body system affected.

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the thyroid abnormality on other body organs and your response to treatment. Also see the description of possible complications under the comments about the listing above and discussion of RFC under whatever other listing is being considered. It should be pointed out that the majority of cases of thyroid disorder can be treated so well that RFC restrictions are not required.

2. Listing 109.02: Thyroid Disorders (Children)

The comments under Listing 9.02 apply here, especially regarding clinical manifestations of thyroid disorders.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have the thyroid disorder that satisfies Ⓐ or Ⓑ, below.

Ⓐ Hyperthyroidism with clinical manifestations despite prescribed treatment. Once such a clinical manifestation of uncontrolled hyperthyroidism is established—such as through abnormal heart rhythms or weight loss—laboratory tests must verify increased thyroid hormone levels.

Additionally, 1, 2, or 3, below, must be present. The normal reference values of the particular testing laboratory should be used regarding the tests mentioned by the listing. This is particularly

important in children because the normal values of thyroid hormone levels vary with age.

1. Elevated total serum thyroxine (T4) and either elevated free (T4) or resin (T3) uptake.
2. Elevated thyroid uptake of radioactive iodine.
3. Elevated serum triiodothyronine (T3).

Ⓑ Hypothyroidism, with 1, 2, or 3, despite prescribed treatment.

1. IQ of 70 or less. Infants with hypothyroidism who are not treated may have permanent brain damage with resulting mental retardation, a condition known as cretinism. However, hospitals routinely test the thyroid function in infants, and cretinism is rare in the U.S. Note: Some IQ tests have more than one IQ score. In these cases, the SSA is obligated by its own rules to use the lowest score in deciding if part Ⓑ1 is satisfied.
2. Growth impairment as described under the criteria in Listing 100.02Ⓐ and Ⓑ. Part Ⓑ2 recognizes that growth impairment can result from hypothyroidism and refers evaluation to the growth impairment listings (CD Part 1).
3. Precocious puberty. Part Ⓑ3 is satisfied by the presence of abnormally early sexual development. Such precocious puberty can occur in boys or girls and is usually associated with severe hypothyroidism over a prolonged period of time.

3. Listing 9.03: Hyperparathyroidism (Adults)

There are two little parathyroid glands, one each buried in the right and left part of the thyroid gland. Although they are within the thyroid gland, the parathyroid glands are separate glands that produce parathyroid hormone (PTH). PTH is important in regulating the way the body uses calcium. About 90% of cases of hyperparathyroidism are caused by a benign tumor in a parathyroid gland.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have hyperparathyroidism that satisfies Ⓐ or Ⓑ, below.

Ⓐ Generalized decalcification of bone found with an x-ray and an elevation of blood calcium levels to 11 mg per deciliter (11 mg/dl) or greater. Part Ⓐ deals with the fact that severe and uncontrolled hyperparathyroidism can have serious consequences regarding dangerous hypercalcemia. Parathyroid hormone causes the absorption of calcium from bones into the bloodstream and therefore tends to raise blood calcium levels. Excessive resorption of calcium from bones (decalcification of bone) can lead to osteoporosis. Even coma and death can result when levels are persistent and extremely high.

Ⓑ Any resulting impairment should be evaluated under whatever listings are appropriate. Part Ⓑ is a reminder that any disorders resulting from hyperparathyroidism should be evaluated under the appropriate listing. Although hypercalcemia may not produce any symptoms when mild, very high calcium levels can cause mental disturbances, abnormal heart rhythms, nausea, vomiting, pancreatitis, weakness, loss of appetite, and kidney stones with associated renal failure. For example, kidney stones would be evaluated under the listings dealing with renal failure (CD Part 6). Similarly, heart problems would be evaluated under the cardiovascular listings (CD Part 4).

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the parathyroid abnormality on other body organs and response to treatment. Also, see the description of possible complications under the comments about the listing above and the discussion of RFC under whatever other listing is being considered. Note that the majority of cases of parathyroid disorder can be treated so well that RFC restrictions are not required.

4. Listing 109.03: Hyperparathyroidism (Children)

See the comments under adult Listing 9.03. Note that this child listing is much more lenient than the adult listing, since only abnormal laboratory tests are required. However, it is still rare for a child to have

uncontrollable hyperparathyroidism. Allowances under this listing are very rare.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have hyperparathyroidism that satisfies Ⓐ or Ⓑ, below.

Ⓐ Repeatedly elevated serum calcium levels.

Ⓑ Elevated serum parathyroid hormone levels (PTH).

5. Listing 9.04: Hypoparathyroidism (Adults)

Hypoparathyroidism means abnormally low parathyroid hormone (PTH) output from the parathyroid glands, which are inside the thyroid gland. A frequent cause of this disorder is damage to the parathyroid glands during thyroidectomy, a complication that might not become apparent for years after surgery. When PTH levels are too low, blood calcium levels fall. The risks of hypoparathyroidism are those associated with this hypocalcemia.

a. Listing Level Severity

For your condition to be severe enough to meet the listing, you must have hypoparathyroidism that satisfies Ⓐ, Ⓑ, or Ⓒ, below.

Ⓐ Severe, recurrent, tetany. Part Ⓐ deals with a serious consequence of hypocalcemia: the uncontrollable muscle spasms known as tetany. When tetany develops, calcium levels are dangerously low and affect muscle function. It is not expected or reasonable that tetany be present as a daily occurrence, since such a life-threatening condition would receive medical treatment. Rather, recurrent episodes of tetany would be sufficient to qualify under Part Ⓐ. In the context of this listing, "severe" means "marked" or more than moderate. The SSA does not define how often "recurrent" has to be, leaving it to medical judgment by the person making the disability determination. However, it would be reasonable to accept fewer episodes of recurrent tetany if they are more prolonged and difficult to control.

Ⓑ Recurrent generalized convulsions. Part Ⓑ is similar to part Ⓐ, except that it requires recurrent convulsions affecting the whole body. By their very nature, such convulsions would be "severe"—indeed, life-threatening.

Ⓒ Cataracts, evaluated under the criteria in listings for visual impairments; see CD Part 2. Such cataracts are a sign of long-standing hypoparathyroidism.

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the parathyroid disorder on other body organs and response to treatment. For example, if cataracts are a problem, see the discussion of RFCs under the visual impairment listings.

6. Listing 109.04: Hypoparathyroidism and Pseudohypoparathyroidism (Children)

See the comments under adult Listing 9.04. In pseudohypoparathyroidism (PHP) the problem isn't a deficiency in parathyroid hormone, but that the body's cell receptors for the hormone are defective. The result is the same: the parathyroid hormone can't do its job.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have hypoparathyroidism or pseudohypoparathyroidism that satisfies Ⓐ or Ⓑ, below.

Ⓐ Severe, recurrent, tetany or recurrent convulsions. See comments under Listing 9.04.

Ⓑ Growth retardation as described under the criteria in Listing 100.02 Ⓐ and Ⓑ (CD Part 1).

7. Listing 9.05: Neurohypophyseal Insufficiency (Diabetes Insipidus) (Adults)

One of the hormones secreted by the neurohypophysis of the pituitary gland is antidiuretic hormone (ADH).

ADH is a hormone that acts on the kidneys to prevent excessive water loss from the body. A deficiency can usually be treated with a replacement form of the hormone delivered as a nasal spray. When the word "diabetes" is used, reference is usually being made to diabetes mellitus (sugar diabetes), which

is a common disorder. However, diabetes insipidus (DI) and diabetes mellitus are completely different diseases. One possible cause of DI is intracranial tumors that damage the pituitary gland by pressure.

With diabetes insipidus, the lack of ADH causes abnormally large volumes of water to be urinated away and as a result the concentration of sodium left behind in the blood increases—a condition known as hypernatremia. Also, the increased volume of urine causes the concentration of dissolved wastes in urine to decrease and such abnormally dilute urine can be detected with a simple test known as the specific gravity (Sp. Gr.). Urine specific gravity is normally in the range of about 1.016–1.022, but falls as the urine becomes diluted. If the Sp. Gr. decreases to 1.005, there is very severe and uncontrolled diabetes insipidus. To see how low your specific gravity is, all you have to do is look at the report of your routine laboratory analysis of the urine (urinalysis).

As excessive water is lost from the body in urine, a person with DI must drink large volumes to replace it. However, dehydration is still likely to occur. This is especially true if the person has an ineffective thirst center in the hypothalamus of the brain, a condition most likely in children and the elderly.

Note: If the pituitary gland is being damaged by pressure from a tumor, there may also be pressure on the nearby optic nerves that carry visual information from the eyes. Therefore, all claimants with pituitary tumors should be considered to have possible visual losses, especially loss of peripheral vision (visual fields). Special tests are needed to measure visual field losses and those are described under the listings for visual impairments (CD Part 2).

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have neurohypophyseal insufficiency causing your urine specific gravity to be 1.005 or below, persistent for at least three months, and recurrent dehydration.

The SSA does not define how often "recurrent" must be. Medical judgment has to be applied on an individual basis, depending on how much the disorder affects ability to function. The more severe the dehydration and associated symptoms, the fewer it would take to be disabling.

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the diabetes insipidus on other body organs and response to treatment. For example, DI could worsen cardiovascular (CD Part 4) or other disorders. On the other hand, DI could be so well controlled with hormone replacement that no RFC restrictions are needed. If dehydration is sometimes a problem, there should be restriction from heavy work or hot environments that would increase water loss through sweating. If the ADH is difficult to control, but not quite at listing-level severity, then weakness and dehydration could justify an RFC for no more than sedentary work.

8. Listing 109.05: Neurohypophyseal Insufficiency (Diabetes Insipidus) (Children)

See the comments under adult Listing 9.05. Documentation of a child's neurohypophyseal insufficiency requires that one of two possible tests be abnormal. These tests determine the body's ability to release antidiuretic hormone (ADH) from the neurohypophysis of the pituitary gland, which can then be measured in the blood. The tests must be done in a hospital and cannot be ordered by the SSA. However, if they have been done they should be in the child's hospital records with a full interpretation by the treating doctor. (The required tests are actually quite a bit more complex than discussed below and can involve various additional tests on urine and blood as well as administration of ADH to observe its results. But those details are beyond the scope of this book.)

- *Water deprivation test.* This test is simply based on the fact that when a person is deprived of water, the pituitary normally releases ADH to make the kidneys retain water and pass less of it out as urine.
- *Hypertonic saline test.* An intravenous infusion of hypertonic saline (concentrated sodium chloride salt solution) is a powerful stimulus to the pituitary gland to release ADH. Therefore, a normal pituitary gland should release ADH to make the kidneys retain water and pass less of it out as urine.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have neurohypophyseal insufficiency, documented by an abnormal hypertonic saline or water deprivation test. Additionally, the child's condition must satisfy Ⓐ, Ⓑ, Ⓒ, Ⓓ, or Ⓔ, below.

Ⓐ Intracranial space-occupying lesion, before or after surgery. Part Ⓐ is satisfied if there is a tumor ("space-occupying lesion") inside the head that is responsible for damaging the neuroyhypophysis by pressure against it. Such a tumor could arise from the pituitary gland itself or from outside the pituitary. Part Ⓐ can still be satisfied, even if surgery for the tumor has been performed.

Ⓑ Unresponsiveness to Pitressin. Part Ⓑ is fulfilled if the diabetes insipidus is not corrected by Pitressin, an injectable form of synthetic ADH exactly the same as ADH produced by the body. Other forms of ADH delivered by intranasal spray cannot satisfy part Ⓑ. The purpose of part Ⓑ is to identify the presence of nephrogenic diabetes insipidus. In these instances, kidney disease is the problem rather than lack of ADH, and there will be no improvement with the injected ADH.

Ⓒ Growth retardation as described under the criteria in listing 100.02Ⓐ and Ⓑ. Part Ⓒ recognizes that growth impairment can result from diabetes insipidus and refers evaluation to the growth impairment listings (CD Part 1).

Ⓓ Unresponsive hypothalamic thirst center, with chronic or recurrent hypernatremia. Part Ⓓ involves some type of abnormality in the thirst center of the hypothalamus of the brain. When concentrated salt solution in the form of the hypertonic saline test mentioned above is given, a normal thirst center will signal the pituitary gland to release ADH in order to conserve body water. If the child has a defective thirst center, there will be a weak awareness of the need to drink water and episodes of dehydration may occur. With dehydration, the sodium concentration in the blood will increase and this hypernatremia must be documented to satisfy part Ⓓ. The SSA does not define how often "recurrent" must be. Medical judgment has to be applied to claimants on an individual basis, depending on how much the disorder affects the child's ability to function. The more severe the dehydration and associated symptoms, the fewer episodes of hypernatremia it would take to be disabling.

Ⓔ Decreased peripheral vision (decreased visual fields) caused by a pituitary abnormality. Part Ⓔ concerns the fact that abnormalities such as pituitary tumors can put pressure on the optic nerves and thereby decrease peripheral vision. A more detailed discussion of peripheral vision can be found in the discussion of listings dealing with visual impairments (CD Part 2).

9. Listing 9.06: Hyperfunction of the Adrenal Cortex (Adults)

Hyperfunction of the adrenal cortex refers to the increased output of hormones from the outer layers of the adrenal glands. This adrenal gland overactivity can occur either because of an abnormality within an adrenal gland itself (primary disorder) or because of excessive stimulation of the adrenal glands by hormones released from the pituitary gland (secondary disorder). For example, the adrenal cortex normally produces just enough cortisol hormone for the body's needs, under the stimulation of a pituitary hormone known as ACTH. But an adrenal gland tumor can release excessive cortisol on its own, without needing ACTH stimulation—a primary disorder. More often, the adrenal glands have no tumor, but are overstimulated to produce too much cortisol by excessive ACTH released from a pituitary tumor—a secondary disorder. The listing can be satisfied either way—if there is a primary adrenal disorder of the glands themselves or a secondary disorder causing normal adrenal glands to hyperfunction.

Excessive secretion of cortisol can produce a large number of problems which grouped together are referred to as Cushing's syndrome. People with Cushing's syndrome tend to have a rounded facial appearance (called "moon facies" by doctors), purple striae, high blood pressure, obesity concentrated on the trunk of the body, thick skin, prominent fat pads on the upper back, easy bruisability, poor healing of wounds, osteoporosis, poor growth in children, irregular menstruation, diabetes mellitus,

kidney stones, myopathy, and in some cases mental impairment.

While abnormally increased cortisol production is the most frequent type of adrenal hyperfunction, there are other hormones produced in the adrenal cortex that can be secreted in excessive amounts. For instance, there are disorders classified as hyperaldosteronism, in which the adrenal cortex releases too much of the hormone aldosterone. The result can be high blood pressure. There are also disorders of the adrenal cortex involving release of excessive amounts of hormones with activities affecting sexual development, resulting in the adrenogenital syndrome. The adrenogenital syndrome is of particular interest in children (see Listing 109.11), because it can result in abnormal early sexual development in boys (precocious puberty) and development of male sex characteristics in girls (virilization or masculinization).

Because of the large number of possible effects of various excessive adrenal cortex hormones on different organs of the body, this listing has no specific criteria. Rather, it refers evaluation to whatever listing would be most appropriate. For example, high blood pressure would be evaluated under the cardiovascular listings (CD Part 4) while the effects of osteoporosis would be considered under the musculoskeletal listings (CD Part 1) and kidney disease would be evaluated under the listings dealing with renal disease (CD Part 6). If high blood pressure resulted in a stroke, evaluation would be done under the neurological listings (CD Part 11).

a. Listing Level Severity

To determine whether the required level of severity is met, the SSA would evaluate the resulting impairment under the criteria for the affected body system.

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the disorder on other body organs and response to treatment. See the discussion of RFC under whatever other listings are appropriate to the affected body system. The additional limiting effects of obesity in cases of Cushing's syndrome should be remembered, as well as possible decreased muscle strength as a result of excessive cortisol production.

10. Listing 109.06: Hyperfunction of the Adrenal Cortex (Children)

See the comments under adult Listing 9.06. However, this child listing concerns only excessive cortisol hormone secretion from the adrenal glands. Such increased cortisol can result in Cushing's syndrome, just as it can in adults. Adrenal cortex hyperfunction resulting in the adrenogenital syndrome is considered under Listing 109.11.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have hyperfunction of the adrenal cortex (primary or secondary) that satisfies both parts Ⓐ and Ⓑ, below.

Ⓐ Elevated 17-hydroxycorticosteroids in the urine (or elevated 17-ketogenic steroids). Part Ⓐ deals with the fact that compounds known as 17-hydroxycorticosteroids (17-OHCS) are found in the urine in increased amounts when the adrenal glands secrete excessive cortisol. This happens because 17-OHCS are a metabolic breakdown product of cortisol. Normal values of 17-OHCS depend on the normal reference values used by the testing laboratory. However, as a general guide, normal results are:

17-Hydroxycorticosteroids Levels

Age	Normal Values of Urine 17-OHCS
less than 8 years	less than 1.5 mg/24 hours
8–12 years	less than 4.5 mg/24 hours
over 12 years (boys)	4.5–12 mg/24 hours
over 12 years (girls)	2.5–10 mg/24 hours

If normal values are exceeded, then part Ⓐ is satisfied. All you have to do is read the normal values off of the laboratory testing report, which should be in the medical records of the child's treating doctor or in hospital records.

Measurement of 17-ketogenic steroids (17-KS) in the urine is an obsolete test, but mentioned here because it is still in the listing. 17-KS are actually 17-OHCS compounds altered by a laboratory procedure, and have the same meaning as increased 17-OHCS regarding diagnosing increased cortisol output from the adrenal glands.

Ⓑ Failure of 17-hydroxycorticosteroids (or 17-ketogenic steroids) to fall with administration of a low-dose dexamethasone suppression test. Part Ⓑ requires a dexamethasone suppression test. When the drug dexamethasone is given, there will normally be a fall in cortisol levels in both the blood and urine, as well as a fall in 17-OHCS in the urine. In people with abnormally increased cortisol production, dexamethasone will not suppress the release of cortisol; levels of cortisol will remain high, along with elevated urinary 17-OHCS. The same is true for 17-KS levels.

Since the dexamethasone suppression test involves the administration of a drug, the SSA will not authorize such testing. Test results should be obtainable from the treating doctor, since testing would be required to reach an accurate diagnosis.

11. Listing 109.07: Adrenal Cortical Insufficiency (Children)

Adrenal cortical insufficiency refers to decreased cortisol hormone output by the outer layers of the adrenal glands. Primary adrenal insufficiency (Addison's disease) is caused by disorders involving abnormal adrenal glands, such as infections, trauma, drugs, adrenalectomy, autoimmune diseases, and hereditary diseases. In secondary adrenal insufficiency, the adrenal glands are normal but the pituitary gland doesn't release the ACTH needed to stimulate them to produce cortisol.

Cortisol is a steroid hormone necessary to maintain life. Without enough cortisol, blood pressure falls (circulatory collapse) and death may follow. In many instances, the adrenal insufficiency can be treated with steroid drugs that replace the function of cortisol, usually cortisone or hydrocortisone.

The child's adrenal insufficiency must be diagnosed in two steps.

1. Either of the following:
   - persistently low blood cortisol levels, or
   - persistently low urine levels of the metabolic breakdown product of cortisol known as 17-hydroxycorticosteroids (17-OHCS). (Federal regulations also accept the measurement of low 17-ketogenic steroids (17-KS) in the urine; however, measurement of 17-KS is generally considered obsolete.)
2. The adrenal glands must be unresponsive to stimulation by injected ACTH. In other words, the child must have abnormal adrenal glands—that is, primary adrenal insufficiency.

a. Listing Level Severity

For the child's condition to be severe enough to meet the listing, the child must have adrenal cortical insufficiency with recent, recurrent episodes of circulatory collapse. Any episodes of circulatory collapse (adrenal crisis) related to the listing would require hospitalization. The SSA does not define recent or recurrent. In general, most events over three months past would not be considered recent. Certainly, events more than six months old would not be recent by reasonable medical judgment. Recent should be interpreted in light of the fact that disability is based on problems present at the time of disability determination and expected to last a minimum of 12 months. A child with an episode of circulatory collapse many months before application for disability should be considered stable with treatment, rather than having an ongoing problem. Recurrent is a matter of medical judgment, depending on how much the disorder affects the child's ability to function. The more severe the episodes of circulatory collapse and associated symptoms, the fewer it would take to be disabling.

12. Listing 9.08: Diabetes Mellitus (Adults)

Diabetes mellitus is a common and serious disorder and also a frequent impairment seen in disability determination cases. Diabetes mellitus should not be confused with diabetes insipidus, a completely different and much less common disorder. Diabetes mellitus is associated with insufficient insulin, but

also can be worsened by the resistance of cells to the effects of the insulin that is present. There are some very basic facts you should know about diabetes mellitus to understand this listing.

Glucose is the basic source of energy for cells, and insulin is required for glucose to enter cells. With insufficient insulin, hyperglycemia results because glucose cannot be utilized.

If the diabetes is mild enough, it can be treated with diet, exercise, and oral medications that either increase insulin secretion from the pancreas or increase the effectiveness of the insulin that remains. Diet and exercise help because obesity and lack of exercise increase insulin resistance. When diabetes can be treated without insulin, it is called non-insulin dependent diabetes mellitus (NIDDM).

However, when severe diabetes is present, treatment by insulin injection is required. Diabetes that requires treatment with insulin is called insulin dependent diabetes mellitus (IDDM).

There are two basic types of diabetes mellitus:

Type I. In Type I diabetes, onset is earlier in life and it is also referred to as juvenile diabetes. Most cases of Type I diabetes are now known to be caused by an autoimmune disorder in which the person's own immune system destroys the pancreatic cells that produce insulin. Type I diabetics usually require insulin.

Type II. Type II diabetes is extremely common, because obesity and lack of physical exercise increase the resistance of the body's cells to the utilization of insulin. It is more likely in middle-aged and overweight people who do not exercise regularly. Therefore, it is also known as adult-onset diabetes. Type II diabetes can sometimes be controlled with oral drugs, but progression to insulin dependence is not unusual.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have diabetes mellitus that satisfies Ⓐ, Ⓑ, or Ⓒ, below.

Ⓐ Neuropathy that results in persistent difficulty standing and walking, or persistent difficulty using the hands, including both gross and dexterous movements of the fingers. Part Ⓐ recognizes the importance of neuropathy in diabetes. Sensory neuropathy causing numbness or confusion about the position of the legs can make standing and walking difficult or interfere with ability to work with the hands. Motor neuropathy can decrease strength in the arms and legs. Either sensory or motor neuropathy can qualify, or a mixture of the two. Two extremities must be involved, but it could involve two legs (most common), two arms, or one arm and one leg. If your legs are involved, the listing does not require that you need an assistive device like a cane or crutch to walk—but it should be severe enough that you cannot do even sedentary work requiring standing or walking a total of two hours during an eight-hour workday. "Gross" movements of the hands are the ability to handle larger objects regarding lifting, carrying, and grasping. For example, grasping a doorknob or other object about the size of a tennis ball or picking up a chair would be a gross movement. "Dexterous" movements are those that require more coordination and speed, such as manipulating small objects, and good separate control of each individual finger. For instance, playing a musical instrument, sewing, typing, picking up coins, and buttoning clothes are all dexterous movements. During physical examinations, you should be able to quickly and easily touch each of your fingertips to your thumb in rapid succession, if your dexterous abilities are intact.

Ⓑ Episodes of acidosis occurring on the average of once every two months. Documentation must be by appropriate blood tests. Part Ⓑ takes into account that severe, uncontrolled diabetes may result in ketoacidosis. In ketoacidosis, the body can't metabolize glucose for energy since lack of insulin prevents its entry into cells. Therefore, fats are used for energy, resulting in breakdown products called ketones. These ketones increase blood acidity. Diabetic ketoacidosis (DKA) is life threatening and must be treated in a hospital. If you've had such episodes they would be documented in your medical records.

Ⓒ Retinitis proliferans. Evaluate under the listings for visual impairments. Part Ⓒ deals with the

fact that poorly controlled diabetes is a major cause of blindness. If the severe retinal damage characteristic of retinitis proliferans is present, evaluation is done under those listings dealing with visual disorders (see CD Part 2).

Note: The fact that your disorder might not satisfy any part of this specific diabetic listing does not mean that you couldn't meet another listing. For example, severe diabetes can make heart disease much worse. If you have heart disease, you would also want to consider the cardiovascular listings (CD Part 4). Diabetes is also a major cause of kidney failure, which would be evaluated under the listings dealing with kidney disease (CD Part 6).

b. Residual Functional Capacity

Medical judgment must be applied on a case-by-case basis to determine RFC, taking into account the effect of the diabetes on other body organs and response to treatment. Some diabetics require no functional restrictions, because they either have mild diabetes mellitus or have excellent control with no significant organ damage. Others may have visual impairments, heart failure, difficulty walking because of neuropathy, or decreased blood flow in their leg arteries, etc. Make sure that the SSA is aware of your limitations in daily activities and how such restrictions relate to your diabetes. You can record your difficulties and symptoms on the forms the SSA will give you, but it is even better if your treating doctor discusses your problems in your medical records. For example, the comments about neuropathy under part Ⓐ of the listing also apply to RFC considerations when of a lesser degree of severity. If you have diabetes-related or other conditions not considered by this listing, also refer to the RFC discussions under whatever listing is appropriate.

13. Listing 109.08: Juvenile Diabetes Mellitus (Children)

See the comments about diabetes under adult Listing 9.08, keeping in mind that specific parts of the adult and child listings are not the same.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have insulin dependent juvenile diabetes mellitus that satisfies Ⓐ, Ⓑ, Ⓒ, or Ⓓ, below, despite prescribed therapy.

Ⓐ Recent, recurrent hospitalizations with acidosis. The SSA does not define recent or recurrent. In general, most events over three months past would not be considered recent. Certainly, events more than six months old would not be recent by reasonable medical judgment. Recent should be interpreted in light of the fact that disability is based on problems present at the time of disability determination and expected to last a minimum of 12 months. A child with an episode of acidosis many months before applying for disability should be considered stable with treatment, rather than as having an ongoing problem. Recurrent is a matter of medical judgment, depending on how much the disorder affects the child's ability to function. The more severe the episodes of acidosis, the fewer it would take to be disabling.

Ⓑ Recent, recurrent episodes of hypoglycemia. Part Ⓑ deals with episodes of low blood glucose. Although diabetes is associated with high blood glucose, a mistake resulting in insulin overdosage can cause hypoglycemia. Such hypoglycemia can be a serious problem in diabetics, resulting in brain damage, coma, and even death. Also, some cases of diabetes are difficult to control and are called "brittle" diabetes. These children are more susceptible to developing accidental hypoglycemia. Except for involving hypoglycemia rather than acidosis, the terms "recent" and "recurrent" have the same meaning here as for part Ⓐ.

Ⓒ Growth retardation as described under the criteria in Listing 100.02Ⓐ or Ⓑ (CD Part 1).

Ⓓ Impaired kidney function that satisfies the criteria of the listings that deal with kidney disease (CD Part 6).

14. Listing 109.09: Iatrogenic Hypercorticoid State (Children)

Iatrogenic hypercorticoid states are those caused by steroid drugs like prednisone when taken for a prolonged period of time, as might be necessary to

treat certain immune diseases, cancers, skin diseases, blood diseases, respiratory diseases, and eye diseases. The long-term side effects of such treatment can result in Cushing's syndrome. (See the definition of Cushing's syndrome and the discussion under adult Listing 9.06 for a description of the abnormalities associated with this disorder.)

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have been through iatrogenic glucocorticoid therapy resulting in Ⓐ, Ⓑ, Ⓒ, Ⓓ, or Ⓔ, below.

Ⓐ Osteoporosis. Part Ⓐ refers to osteoporosis, which only needs to be severe enough to be a definite diagnosis—the SSA's own regulation does not have any requirement that the osteoporosis be advanced (marked) in severity. If there is a chance the child has osteoporosis, bone densitometry x-rays should be done rather than plain x-rays. Osteoporosis is visible on plain x-rays only when a large amount of bone has been lost. Therefore, the child could possibly be deprived of a legitimate allowance if the SSA misses osteoporosis by relying on plain x-rays. Bone densitometry testing is widely available and could certainly be purchased by the SSA if necessary.

Ⓑ Growth retardation as described under the criteria in Listing 100.02Ⓐ or Ⓑ (CD Part 1).

Ⓒ Diabetes mellitus as described under the criteria in Listing 109.08.

Ⓓ Myopathy as described under the criteria in Listing 111.06 (CD Part 11).

Ⓔ Emotional disorder as described under the criteria in the mental disorder listings (CD Part 12). (In some children, steroids can produce anxiety, depression, insomnia, mood swings, personality changes, euphoria, and even psychosis; also, steroids might aggravate emotional problems already present.)

15. Listing 109.10: Pituitary Dwarfism (Children)

Pituitary dwarfism results from insufficient growth hormone released from the pituitary gland. Decreased growth hormone can be caused by damage to the pituitary gland or as a result of a tumor. Growth hormone deficiency can also be caused by damage to the hypothalamus of the brain, which releases another hormone—growth hormone releasing hormone—to stimulate the pituitary to release growth hormone. Either cause of dwarfism could potentially qualify under this listing.

With the advent of the genetic engineering of bacteria, human growth hormone is available to treat those children who are deficient. This listing refers evaluation to the growth impairment Listing 100.02Ⓑ (CD Part 1).

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have pituitary dwarfism with documented growth hormone deficiency and growth impairment as described under the criteria in Listing 100.02Ⓑ.

16. Listing 109.11: Adrenogenital Syndrome (Children)

Adrenogenital syndrome results from a genetic enzyme deficiency in the adrenal glands. That deficiency causes an increased secretion of adrenal hormones that produce male sex characteristics—a process known as virilization (see "Definitions" at the beginning of this chapter). In male children, the effect of virilization is precocious puberty. In female children, the effect is pseudohemaphroditism.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have adrenogenital syndrome that satisfies Ⓐ, Ⓑ, or Ⓒ, below.

Ⓐ Recent, recurrent salt-losing episodes despite prescribed therapy. Part Ⓐ is satisfied by excessive loss of sodium from the body. Events known as "salt-losing episodes" are present in the majority of cases and start soon after birth with weakness, vomiting, and dehydration. The SSA does not define how often "recurrent" must be. Medical judgment has to be applied on an individual basis, depending on how much the disorder affects the child's ability to function. The more severe the salt-losing episodes and associated symptoms,

the fewer it would take to be disabling. The SSA also does not define recent, but it is difficult to see how most events over three months in the past could be considered recent. Certainly, events more than six months old would not be recent by reasonable medical judgment. The word "recent" should be interpreted in light of the fact that disability is based on problems supposedly present at the time of disability determination and expected to last a minimum of 12 months. A child with a salt-losing episode many months before applying for disability should be considered stable with treatment, rather than as having an ongoing problem.

Ⓑ Inadequate hormone replacement therapy, as manifested by an accelerated bone age and virilization. Part Ⓑ applies to those children whose treatment is inadequate. The treatment is administration of the steroid hormone hydrocortisone, which must be given indefinitely. Hydrocortisone inhibits release of another hormone, ACTH, from the pituitary gland. Decreased ACTH means that there is less stimulation of the adrenal glands to produce the abnormal hormones causing the sexual development problems. However, if the problem is a tumor in the adrenal glands, this treatment won't work, and surgery is required to remove the tumor. Virilization can be determined through physical examination. Bone age can be determined from x-rays.

Ⓒ Growth impairment as described under the criteria in Listing 100.02 Ⓐ and Ⓑ (CD Part 1).

17. Listing 109.12: Hypoglycemia (Children)

In children, hypoglycemia is present if the fasting blood glucose (FBG) falls below 50 mg/dl and there are symptoms. In infants, hypoglycemia is not considered present by some authorities until the FBG falls under 40 mg/dl. The SSA does not specifically provide a blood sugar number to officially represent hypoglycemia, but it does say that laboratory tests can be considered abnormal if outside of the normal range used by the testing laboratory or if more than two standard deviations from the average normal value used by the testing laboratory. The severe physical abnormalities (convulsions or coma) required by the listing will not occur until blood glucose levels fall to values below the 40–50 mg/dl range as needed to affect the brain.

One cause of hypoglycemia in children is a condition known as ketotic hypoglycemia, which is a disorder of metabolism involving proteins. Frequent feedings with foods high in carbohydrates and protein are used to treat the disorder. Accidental insulin overdose, overdose with oral drugs used to treat diabetes mellitus, or overdose with some other drugs may result in hypoglycemia. Other possible causes of hypoglycemia in children include adrenal insufficiency, hereditary enzyme deficiencies, and pancreatic tumors that produce excessive insulin (insulinomas).

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have hypoglycemia with recent, recurrent hypoglycemia episodes producing convulsions or coma. The SSA does not define recent or recurrent. In general, most events over three months past would not be considered recent. Certainly, events more than six months old would not be recent by reasonable medical judgment. Recent should be interpreted in light of the fact that disability is based on problems present at the time of disability determination and expected to last a minimum of 12 months. A child with an episode of hypoglycemia many months before application for disability should be considered stable with treatment, rather than as having an ongoing problem. Recurrent is a matter of medical judgment, depending on how much the disorder affects the child's ability to function. The more severe the episodes of hypoglycemia, the fewer it would take to be disabling.

18. Listing 109.13: Gonadal Dysgenesis (Turner's Syndrome) (Children)

Cells normally have 46 chromosomes—44 somatic (nonsex) chromosomes occurring as 22 pairs and two X or Y sex chromosomes. The sex chromosomes in males are normally an XY combination and in females XX. A normal male's chromosome number would be written as 46, XY and a normal female as 46, XX. Gonadal dysgenesis, more commonly

known as Turner's syndrome, is a genetic disorder of females characterized by missing one of the two X chromosomes that are normally present in female cells. Such a condition would be written as 45, X. However, there are also variations of Turner's syndrome known as mosaics, in which cells in the same person's body have different genetic make-ups. A common mosaic in Turner's syndrome is that some cells are normal with two X chromosomes, while other cells are missing an X chromosome. Such conditions would be written 45, X/46, XX. In another type of mosaic abnormality, some cells are missing an X chromosome while others have a Y chromosome substituted for one of the X chromosomes (45, X/46, XY). Other combinations are also possible.

Children or adults with Turner's syndrome can have hormonal abnormalities, impairment of growth, hearing loss, and neurological problems involving the failure to integrate sensation and movement on a fine level. Reproductive abnormalities are typically present. However, intelligence is usually normal. Heart abnormalities may be present, as well as abnormalities of the kidneys and bony skeleton. In other words, evaluation of Turner's syndrome requires thorough physical examination and laboratory testing. This information should be available to the SSA. Evaluation of growth would be considered under the growth impairment listings (CD Part 1); hearing loss under the appropriate listings for hearing loss (CD Part 2); heart disorders under the cardiovascular listings (CD Part 4); and neurological problems under the nervous system listings (CD Part 11).

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have gonadal dysgenesis (Turner's syndrome), proven by chromosomal analysis. The SSA would evaluate the resulting impairment under the criteria for the appropriate body system. ■