

# MANAGING DYSAUTONOMIAS

## OVERVIEW

Successful management of a dysautonomia involves more than obtaining a correct diagnosis and then instituting curative treatment. Even in the most sophisticated and knowledgeable centers, the diagnosis remains uncertain, especially for functional disorders. Even an agreed upon diagnosis, such as postural tachycardia syndrome, does not necessarily carry with it agreed upon ideas about the mechanism of the condition, the most appropriate treatment, or the long term outcome.

Dwelling on finding “cures” for dysautonomias is unrealistic.

On the other hand, there are many treatments for dysautonomias, including non-drug and drug treatments, and there are many coping tactics. This section focuses on these aspects of autonomic medicine.

# TREATMENT OF DYSAUTONOMIAS

## The Most Effective Treatments

Often the most effective treatment for a dysautonomia is time. For instance, postural tachycardia syndrome that comes on soon after a viral infection in an otherwise healthy person may “melt away” over many months or years.

Another often effective treatment is exercise, for a few reasons. First, in patients with chronic orthostatic intolerance, maintaining excellent muscle tone in the anti-gravity muscles of the buttocks, thighs, and calves maximizes the efficiency of muscle pumping to maintain venous return to the heart during orthostasis. Second, exercise training improves the ability to increase cardiac output. Third, it is important in chronic, debilitating disorders for the patient to regain a sense of at least some control over the situation.

**The most effective treatment of dysautonomias is education.**

Effective management includes learning about situations likely to worsen or improve symptoms. Joining a support group, one can compare notes with others in the same situation and “flip the clinic” by educating clinicians about individual experiences. One also benefits fellow patients and humanity in general by participating in research and in helping train physicians.

## Non-Drug Treatments

Several non-drug treatments are used for different types of dysautonomias. The reasons for a treatment depend on the particular dysautonomia. Sometimes the responses of a patient to a treatment help the doctor determine the diagnosis. Patients with dysautonomias can feel differently from day to day, even without any clear reason why. This means that if a treatment is tried, it may take several days to decide on whether the treatment has helped or not.

### Elevation of the Head of the Bed

In patients who have a fall in blood pressure every time they stand up (orthostatic hypotension), elevation of the head of the bed on blocks at night improves the ability to tolerate standing up in the morning.

### Salt Intake

High salt intake tends to increase the volume of fluid in the body. A small percent of this volume is in the bloodstream. Doctors usually recommend a high salt diet for patients with an inability to tolerate prolonged standing (chronic orthostatic intolerance) or a fall in blood pressure during standing (orthostatic hypotension).

Normally when a person takes in a high salt diet, the kidneys increase

the amount of salt in the urine, and this limits the increase in blood volume. After a few days of the same salt intake, the rate of sodium excretion equals the rate of intake. Drugs that promote retention of sodium by the kidneys, such as fludrocortisone, are usually required for high salt intake to increase body fluid volume effectively.

## Water Drinking

A relatively recently described tactic to increase blood pressure in patients with autonomic failure is to drink 16 ounces of water or other fluid. Why water drinking should increase blood pressure in patients with autonomic failure, when doing so does not affect the blood pressure of healthy people, remains unclear. Researchers have proposed the existence of an “osmopressor response,” in which ingested water without solute acts in the gut or liver to increase sympathetic noradrenergic system outflow at the level of the pre-ganglionic neurons in the spinal cord. The sensors evoking the response are still unknown. The osmopressor response may improve orthostatic tolerance in patients with baroreflex failure or autonomically mediated syncope.

Patients with chronic orthostatic intolerance, autonomically mediated syncope, or POTS often keep a water container with them and sip from it repeatedly during the day. This habit might indicate a tendency to dehydration and low blood volume, but the pathophysiologic meaning of the “water bottle sign” remains unclear.

## Meals

Eating a big meal leads to shunting of blood toward the gut. In people with dizziness or lightheadedness when they stand up (orthostatic intolerance), it is usually advisable to take frequent small meals.

Reducing the amounts of sugars or other carbohydrates in meals may help manage symptoms.

A substantial proportion of patients with chronic orthostatic intolerance have gastrointestinal symptoms and signs leading to a diagnosis of gastroesophageal reflux, slow gastric emptying, or irritable bowel syndrome. Gastroenterologists managing these patients should be aware that recommending a high fiber diet might worsen orthostatic intolerance by augmenting shunting of blood to the gut.

## Compression Hose

Compression hose or other compression garments tend to decrease the amount of pooling of blood in veins when a person stands. This can decrease leakage of fluid from the veins into the tissues and decrease swelling of the feet. In patients with veins that fill up or leak excessively during standing, compression garments can improve toleration of prolonged standing. In POTS patients a “step-in”

abdominal binder may be more efficient than compression stockings, by limiting orthostatic blood pooling in the abdomen and pelvis.

In patients with a fall in blood pressure during standing (orthostatic hypotension), the problem may be less with the veins than with the arteries and arterioles, the blood vessels that carry oxygen-rich blood under high pressure to the organs and limbs. Wearing compression hose therefore may be disappointing in the management of orthostatic hypotension.

## Coffee

Some patients with dysautonomias feel better drinking caffeinated coffee frequently. Others feel jittery or anxious and avoid caffeinated coffee. Still others notice no effect.

## Temperature

Patients with dysautonomias often have an inability to tolerate extremes of external temperature. When exposed to the heat, patients with failure of the sympathetic cholinergic system may not sweat adequately to maintain the core temperature by evaporation of the sweat. Patients with chronic orthostatic intolerance, such as from postural tachycardia syndrome (POTS), can have heat intolerance because of loss of blood volume by sweating or shunting of blood away from the brain. When exposed to cold, patients with

sympathetic noradrenergic system failure may not constrict blood vessels adequately in the skin, so that the body temperature falls (hypothermia).

## Exercise

Patients with dysautonomias sometimes benefit markedly from an individualized exercise training program. Often, however, the training does not eliminate the sense of fatigue. It might help to have small amounts of exercise daily, even for only 5-10 minutes. A formal online program was available at the University of Texas Southwestern Medical Center as part of a clinical research study, but the study ended.

As a person exercises, the blood vessels carrying oxygen-rich blood to the exercising muscle (arteries and arterioles) tend to relax, due at least partly to the accumulation of byproducts of metabolism. The sympathetic noradrenergic system normally counters this tendency, by increasing the tone of the blood vessel walls. The blood flow to the exercising muscle therefore is in a dynamic state of balance.

Activation of sympathetic nerves to the heart during exercise increases the force and rate of the heartbeat, and the total amount of blood pumped by the heart in one minute (cardiac output) increases. Meanwhile, like squeezing a tube of toothpaste, pumping of muscle during exercise increases the movement of blood from the limbs back to the heart. The increased metabolic activity tends to increase body



temperature, and sweating, which is stimulated importantly by sympathetic cholinergic nerves to sweat glands. Sweating increases the loss of heat by evaporation, helping maintain the core temperature.

If a patient had failure of the sympathetic nervous system, excessive production of byproducts of metabolism, or a form of heart disease where there were an inability to increase the force or rate of the heartbeat, then the blood pressure would fall during exercise, producing a sense of fatigue or exhaustion.

After exercise, when muscle pumping ceases, the blood can begin to pool in the legs or abdomen, while the rate of sympathetic nerve traffic falls to the resting rate. If the decline in sympathetic nerve traffic did not balance the decline in production of byproducts of metabolism, then the blood pressure would fall after exercise. At the same time, loss of body fluid via evaporative sweating would decrease the blood volume. Patients with a dysautonomia therefore can feel bad not only during exercise but also after exercise. It is important to stay hydrated and to avoid activities like eating a large meal after exercise, because this can divert already limited blood volume to the gut.

Perhaps surprisingly, even vigorously healthy, muscular, lean people can have a susceptibility to faint, and it is unclear if exercise training in general helps them. On the other hand, some patients can improve by isometric calf muscle training, where the patient learns to tense calf

muscles when standing. This tends to decrease the amount of pooling of blood in the legs. At the time of an acute episode, counter-maneuvers such as leg crossing and tightening the buttocks can temporarily maintain consciousness.

## Pacemakers and Sinus Node Ablation

Whether insertion of a pacemaker helps patients with autonomically mediated syncope is controversial. Having a pacemaker inserted may not be a cure, because the low pulse rate at the time of fainting might not cause the low blood flow to the brain that results in the loss of consciousness. On the other hand, a sudden absence of electrical activity in the heart (asystole) produces loss of consciousness within seconds, and in patients with chronic orthostatic intolerance and tilt-evoked asystole, a pacemaker could be curative.

Some patients who have a very fast pulse rate undergo destruction of the sinus node pacemaker cells in the heart (sinus node ablation). The doctor must be sure that the fast pulse rate results from a problem with the heart and does not result from a compensation by the sympathetic nervous system for another problem, such as low blood volume, because eliminating the compensation could make the patient worse rather than better. Sinus node ablation is not thought to help patients with POTS.

## Neurosurgery

Some patients with chronic orthostatic intolerance have a type of change in the brainstem called Chiari malformation. This is an anatomic abnormality where part of the brainstem extends below the hole in the skull between the brain and spinal cord. Neurosurgery can correct the malformation, but the orthostatic intolerance does not necessarily disappear. This is a controversial topic, and patients should seek a second opinion before agreeing to this procedure.

## **Constipation or Urinary Retention**

Patients with failure of the parasympathetic nervous system can have problems with constipation and retention of urine in the bladder. The constipation is treated non-specifically, with stool softeners, bulk laxatives, and if needed milk of magnesia, magnesium citrate, senna, or cascara.

Urinary retention can be associated with urinary urgency and incontinence. Drugs that stimulate receptors for acetylcholine, such as urecholine, might be tried. Often patients with autonomic failure must learn to self-catheterize to empty the bladder, by inserting a plastic or rubber tube into the urethra and then into the bladder, in order to obtain relief.

## **Drug Treatments**

Several drug treatments are used for dysautonomias. Some of them are powerful or can produce harmful side effects. Patients should take medications only under the supervision of a doctor with expertise and experience in the treatment of dysautonomias.

Different centers use different drugs from a long “menu” to treat dysautonomias.

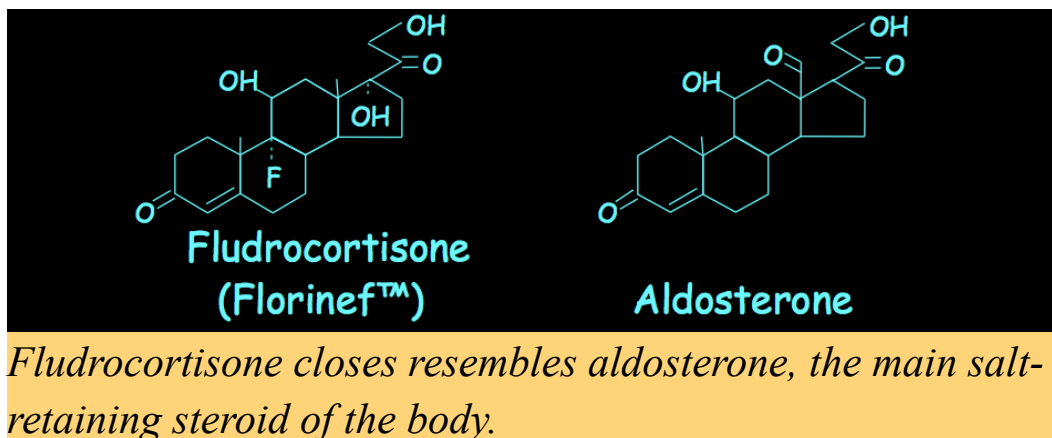
The following is a summary of some of the drugs used to treat dysautonomias.

<b>Drug</b>	<b>Goal of Treatment</b>
Fludrocortisone (=Florinef™)	Increase blood volume Increase blood pressure
Midodrine (=Proamatine™)	Tighten blood vessels Increase blood pressure Prevent fainting
Beta-Blocker	Decrease heart rate Decrease blood pressure Decrease adrenaline effects Prevent fainting
Erythropoietin (=Procrit™)	Increase blood count Increase blood pressure
Amphetamines	Tighten blood vessels Increase alertness

Desmopressin (=DDAVP™)	Tighten blood vessels
“NSAID”	Tighten blood vessels
Somatostatin (=Octreotide™)	Tighten blood vessels in gut
Selective Serotonin Reuptake Inhibitor	
	Improve mood, allay anxiety (=SSRI)
“Tricyclic”	Improve mood
Alprazolam (=Xanax™)	Increase sense of calmness Improve sleep
Clonidine (=Catapres™)	Decrease blood pressure Improve sleep
Bethanechol (=Urecholine™)	Increase salivation Improve gut action Improve urination
Yohimbine	Increase blood pressure
Pyridostigmine (=Mestinon™)	Increase blood pressure
L-DOPS (=Droxidopa, Northera™)	Increase blood pressure

## Fludrocortisone (Florinef™)

Fludrocortisone (Florinef™) is a man-made type of drug called a salt-retaining steroid, or mineralocorticoid. The drug closely resembles the body’s main salt-retaining steroid, aldosterone.



Fludrocortisone (Florinef™) forces the body to retain salt.

Fludrocortisone must be taken with a high-salt diet to work. It forces the kidneys to retain sodium, in exchange for potassium. Water follows the sodium, and so Florinef™ is thought to increase the blood volume. The patient gains “fluid weight,” and blood pressure increases. Because of the tendency of Florinef™ to waste potassium, Florinef™ can cause a fall in the serum potassium level, which if severe can be dangerous. Patients taking Florinef™ should have periodic checks of their serum potassium level, and if it is low take a potassium supplement.

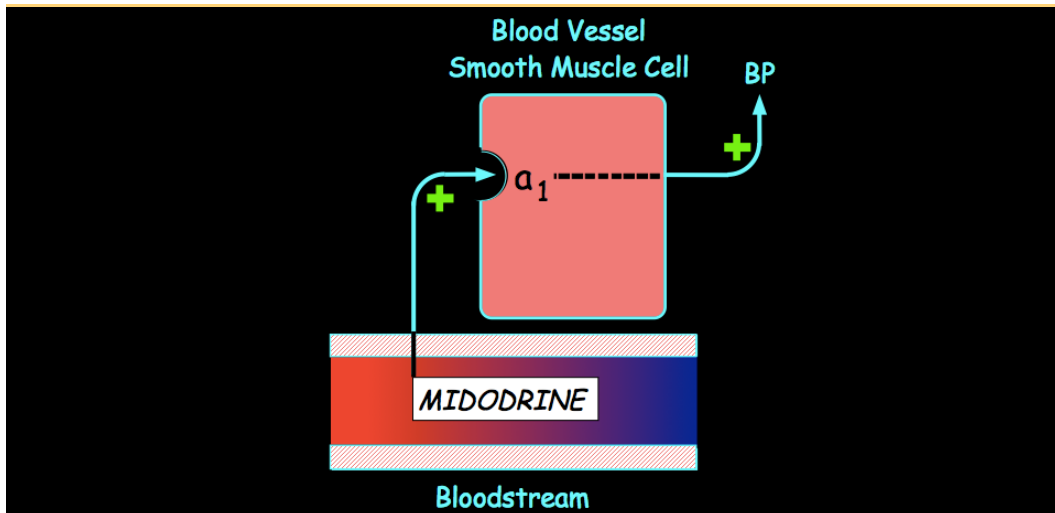
Florinef™ treatment increases the blood pressure regardless of the patient’s posture. The increased blood pressure when the patient is standing may be large enough that the patient does not have lightheadedness or other symptoms of orthostatic intolerance.

Florinef™ given to patients with chronic autonomic failure can cause or worsen high blood pressure when the patient is lying down. Sometimes the doctor faces a difficult dilemma, balancing the long-term increased risk of stroke, heart failure, or kidney failure from high blood pressure against the immediate risk of fainting or falling from orthostatic hypotension.

In some patients with chronic orthostatic intolerance such as POTS, Florinef™ can produce improvement; however, in other patients there is no improvement by Florinef™ treatment. Perhaps treatment with a salt-retaining steroid is effective only in POTS patients who have low blood volume. This is a matter for future research.

## **Midodrine (Proamatine™)**

Midodrine (Proamatine™) tightens blood vessels throughout the body. That is, it is a vasoconstrictor. Midodrine works by stimulating alpha-adrenoceptors in blood vessel walls. When a person stands up, the sympathetic noradrenergic system is normally activated reflexively, the chemical messenger norepinephrine is released from the sympathetic nerves in blood vessel walls, the norepinephrine binds to alpha-adrenoceptors in the blood vessel walls, and the stimulation of the alpha-adrenoceptors causes the blood vessels to constrict (vasoconstriction), increasing the blood pressure.



*Midodrine works like artificial norepinephrine, increasing blood pressure (BP) by stimulating alpha-adrenoceptors in blood vessel walls.*

In patients with orthostatic hypotension related to a loss of sympathetic noradrenergic nerves, there is little decreased norepinephrine to release. In this situation, the blood vessels become supersensitive (denervation supersensitivity), perhaps by the alpha-adrenoceptors accumulating on the surface of the cells in blood vessel walls. In patients with denervation supersensitivity midodrine can be very effective in raising the blood pressure.

Midodrine works like artificial norepinephrine, increasing blood pressure (BP).



In using midodrine to treat elderly men with orthostatic hypotension, the doctor should be aware that stimulation of alpha-adrenoceptors can worsen symptoms of prostate problems, such as urinary retention, urgency, and decreased urinary stream.

Alpha-1 adrenoceptor blockers are effective in treating benign prostatic hypertrophy (BPH), and alpha-1 adrenoceptors blockers interfere with midodrine's effects.

In patients with neurogenic orthostatic hypotension, the symptoms are often worst in the morning. As the day goes on, the blood pressure increases. One may not need an alpha-adrenoceptor agonist throughout the day, and in patients with sympathetic denervation taking midodrine around the clock might desensitize the alpha-adrenoceptors. It is reasonable to try taking midodrine early in the morning before getting up and then perhaps at lunchtime to avoid post-prandial hypotension but not to take it later in the day, so that by the next morning the drug has worn off and the alpha-adrenoceptors are super-sensitive again.

## **Beta-Adrenoceptor Blockers**

Beta-adrenoceptor blockers are drugs that interfere with some effects of norepinephrine and adrenaline. Norepinephrine and adrenaline produce their effects by binding to specific receptors on the target cells, such as heart muscle cells. Beta-blockers interfere with this

binding.

There are two types of receptors for norepinephrine and adrenaline, called alpha-adrenoceptors and beta-adrenoceptors. Adrenaline stimulates both types. Adrenaline tightens blood vessels in most parts of the body, such as the skin, due to stimulation of alpha-adrenoceptors in blood vessel walls. Vasoconstriction of skin blood vessels decreases local blood flow, and the skin becomes pale. This is why pallor can be a sign of high adrenaline levels. In skeletal muscle, adrenaline generally relaxes blood vessels, due to stimulation of beta-2 adrenoceptors. By this action adrenaline tends to shunt blood toward skeletal muscle. This makes sense in terms of the need for abundant blood flow to skeletal muscle in emergency situations. Adrenaline also stimulates beta-adrenoceptors in the heart, and this increases the force and the rate of the heartbeat. Because of the effects on the heart, the amount of blood pumped by the heart per minute (cardiac output) increases.

Beta-1 adrenoceptors and beta-2 adrenoceptors are abundant in the human heart, and stimulation of these receptors produces about the same effects.

**All beta-blockers decrease the rate and force of the heartbeat.**

In contrast, beta-2 adrenoceptors are much more abundant on skeletal muscle blood vessels and in the lungs than are beta-1 adrenoceptors.

Stimulation of beta-2 adrenoceptors on smooth muscle cells of the airways relaxes the airways. This is a reason that beta-2 adrenoceptor stimulants are used to treat asthma.

Drugs that act at beta-adrenoceptors are often grouped in terms of whether they are “selective” for beta-1 adrenoceptors or are “non-selective,” meaning they block both types of beta-adrenoceptors. Beta-2 adrenoceptor stimulants relax the airways and are used to treat asthma. There are no approved drugs that block beta-2 adrenoceptors selectively.

In patients with autonomically mediated syncope and high levels of adrenaline in the bloodstream, the adrenaline stimulates beta-2 adrenoceptors on blood vessels in skeletal muscle. This relaxes the blood vessels and decreases the resistance to blood flow. Blood may then be shunted away from the brain and towards the skeletal muscle, contributing to lightheadedness or loss of consciousness. In such patients, non-selective beta-adrenoceptor blockers might be preferable to selective blockers.

Beta-adrenoceptor blockers decrease the pulse rate, the force of heart contraction, and the systolic blood pressure. In patients with rapid pulse rates, associated with a sense of pounding or irregular beating of the heart (palpitations) or chest pain, beta-adrenoceptor blockers decrease the heart rate and can help relieve the pain and prevent abnormal heartbeats or heart rhythms. These drugs are also

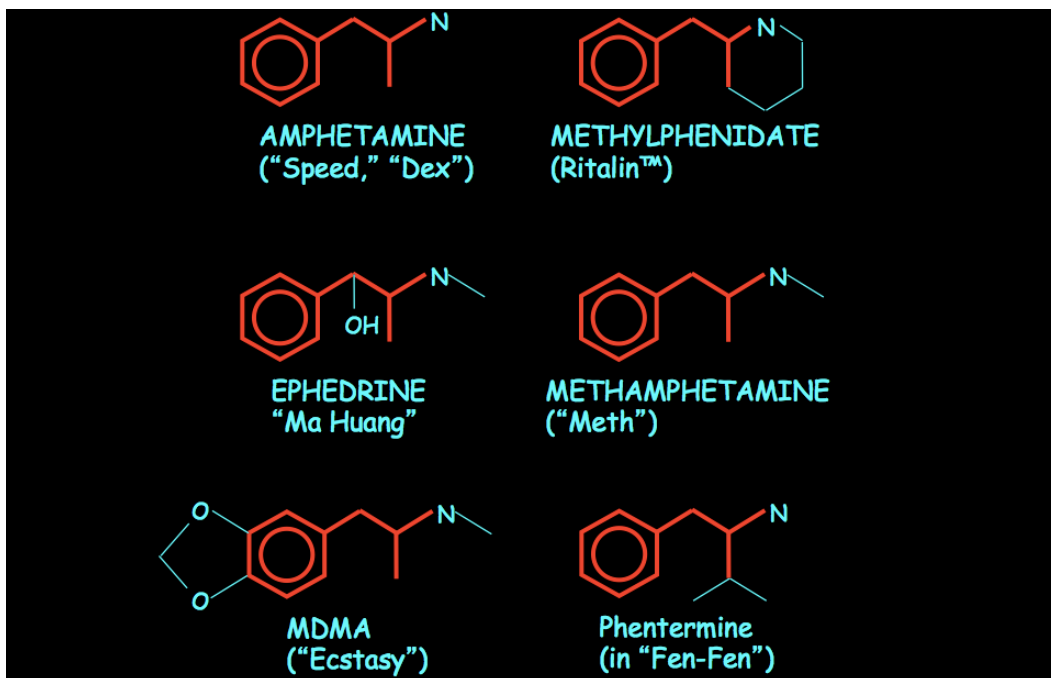
commonly used to treat long-term high blood pressure (hypertension). Because of decreased systolic blood pressure and heart rate, the rate of consumption of oxygen by the heart decreases, and this can help patients with coronary artery disease.

In patients with postural tachycardia syndrome (POTS) the value of treatment with beta-adrenoceptor blockers may depend on whether the rapid pulse rate when the patient stands up reflects a primary or compensatory response. If the rapid pulse rate were a compensation for another problem, such as low blood volume due to bleeding, then blocking that compensation would not help the patient. But if the rapid pulse rate were the result of an inappropriate, excessive rate of sympathetic nerve traffic to the heart, then blocking the effects of the excessive nerve traffic would help the patient.

## Amphetamines

Amphetamines are chemicals that resemble the drug, dextro-amphetamine, or d-amphetamine.

Amphetamines are in a class of drugs called indirectly acting sympathomimetic amines. They produce their effects at least partly by increasing delivery of norepinephrine to its receptors, both in the brain and outside the brain.



*Amphetamines share a particular chemical structure (alpha-methyl-phenylethylamine).*

By way of effects in the brain, amphetamines increase the state of arousal and attention, prevent or reverse fatigue, decrease appetite, and at high doses increase the rate and depth of breathing. By way of effects on the sympathetic noradrenergic system, they increase blood pressure.

Pseudoephedrine (Sudafed™) is structurally a mirror image (stereoisomer) of ephedrine. This difference changes the properties of the drug, producing much less central nervous system stimulation. By releasing norepinephrine from sympathetic nerve terminals in the

mucous membranes of the nasal airways, pseudephedrine tightens blood vessels, making them less leaky and thereby relieving nasal congestion. Because in a laboratory pseudephedrine can be converted easily into other amphetamines that are drugs of abuse, over-the-counter sales of pseudephedrine are now restricted.

Methylphenidate (Ritalin™), another sympathomimetic amine, is used commonly to treat attention deficit-hyperactivity disorder.

Phenylpropanolamine (PPE) was used in over-the-counter diet pills until the discovery of serious adverse effects such as severe high blood pressure and stroke. PPE was taken off the non-prescription drug market.

Phentermine prescribed with fenfluramine (“Phen-Fen”) was an effective combination to decrease weight, until serious adverse effects of this combination came to light, and this combination is no longer prescribed.

In treating patients with dysautonomias, amphetamines should be used sparingly because of the potential for tolerance and dependence. In patients with sympathetic neurocirculatory failure from abnormal regulation of sympathetic nerve traffic to intact sympathetic nerves, this type of drug releases norepinephrine from the terminals and increases the blood pressure. Some patients with chronic orthostatic intolerance can improve.

Amphetamines work both inside and outside the brain. They increase attention, decrease appetite, interfere with sleep, and often increase the blood pressure.

## Intravenous Saline Infusion

Inability to tolerate prolonged standing can result from low blood volume, excessive pooling of blood in the veins of the legs during standing, or exit of fluid from the blood vessels into the tissues during standing (extravasation).

In these situations, infusion of physiological saline solution can temporarily improve the ability to tolerate standing up.

Saline infusion temporarily increases the blood volume.

This is also useful for diagnostic purposes. Some patients with chronic orthostatic intolerance benefit from intravenous saline infusion given a few times per week by way of a permanent intravenous catheter. The clinician must weigh the potential benefit against the not insubstantial risks, such as of infection and intravascular clotting.

## Desmopressin (DDAVP™)

Desmopressin (DDAVP™) is a synthetic replacement for the hormone, vasopressin. Vasopressin tightens blood vessels and raises blood pressure. Vasopressin is also called anti-diuretic hormone (ADH), because it causes the kidneys to retain water and therefore decreases production of urine. Desmopressin taken nasally is used to treat orthostatic hypotension in patients with chronic autonomic failure.

### **Somatostatin (Octreotide™)**

Somatostatin (Octreotide™) is a hormone that inhibits the release of another hormone, growth hormone, from the pituitary gland at the base of the brain. Somatostatin can tighten blood vessels, especially in the gastrointestinal tract, and raise the blood pressure of patients with orthostatic hypotension. The drug must be injected to work, and it is expensive.

### **Pyridostigmine (Mestinon™)**

Pyridostigmine (Mestinon™) is a drug that works by blocking the enzyme that breaks down acetylcholine. Acetylcholine is the chemical messenger that is responsible for transmission of autonomic nerve impulses in ganglia. By inhibiting breakdown of acetylcholine, pyridostigmine is thought to increase activity of the sympathetic nervous system and improve orthostatic hypotension in patients with chronic autonomic failure.



Because pyridostigmine also increases activity of the parasympathetic nervous system, the drug can increase salivation and stimulate gastrointestinal or urinary bladder contractions. There may be psychological changes because of actions of the drug in the brain. By increasing activity of the sympathetic cholinergic system pyridostigmine can increase sweat production. The drug may also increase adrenaline release.

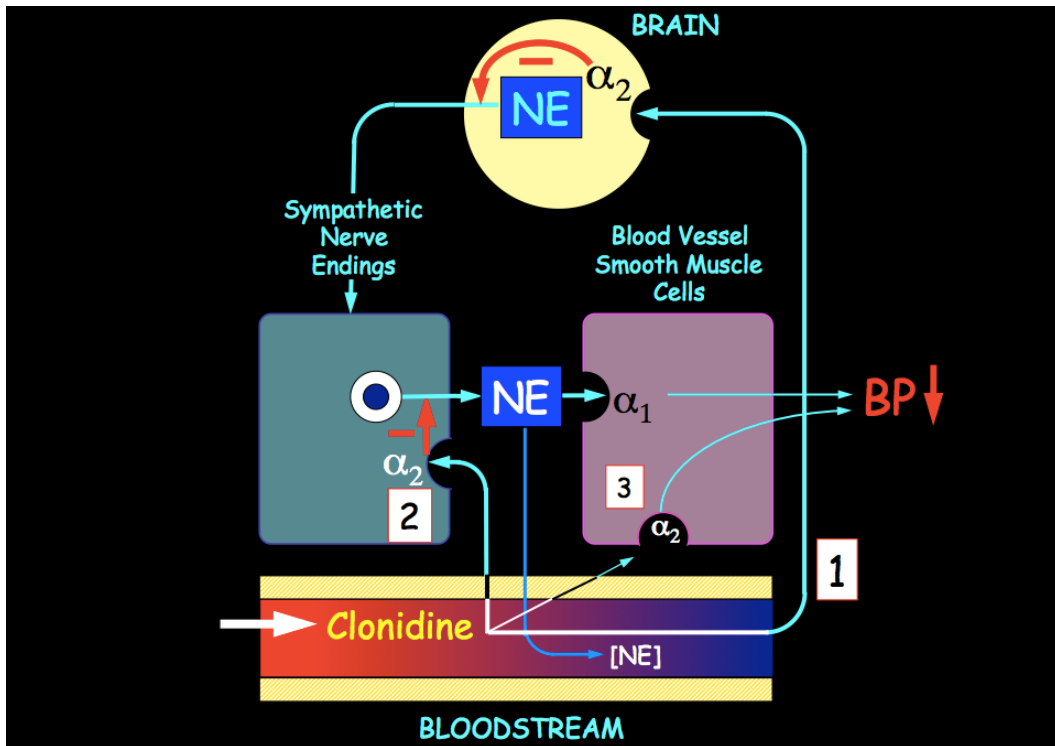
## Selective Serotonin Reuptake Inhibitors (SSRIs)

SSRIs inhibit a key process that is required for inactivating and recycling the chemical messenger, serotonin. The process is reuptake of released serotonin back into the nerve terminals that store it. SSRIs are widely used to treat depression, anxiety, and other psychiatric or emotional problems. They are also used to treat some forms of dysautonomias.

A word of caution is in order in the treatment of teen-aged dysautonomia patients who are depressed: Monoamine reuptake blockers have been statistically associated with a increased risk of suicide.

## Clonidine (Catapres™)

There are two types of alpha-adrenoceptors, called alpha-1 and



*Clonidine decreases sympathetic noradrenergic outflows and decreases norepinephrine release for a given amount of sympathetic nerve traffic.*

alpha-2. Stimulation of either type of receptor in blood vessel walls causes the vessels to constrict (vasoconstriction).

Clonidine stimulates alpha-2 adrenoceptors. Stimulation of alpha-2 adrenoceptors in the brain decreases the rate of sympathetic nerve traffic. Stimulation of alpha-2 adrenoceptors on sympathetic nerves decreases the amount of release of the chemical messenger,

norepinephrine, from the nerves. Even though clonidine stimulates a type of alpha- adrenoceptor, clonidine normally decreases the blood pressure.

Clonidine works both in the brain and outside the brain. It decreases the blood pressure and often causes drowsiness.

There are several uses of clonidine in the diagnosis and treatment of dysautonomias. In the clonidine suppression test, clonidine is used to separate high blood pressure due to increased sympathetic nervous system activity from high blood pressure due to a tumor that produces catecholamines—pheochromocytoma.

In patients with long-term high blood pressure (hypertension) due to excessive release of norepinephrine from sympathetic nerves (hypernoradrenergic hypertension), clonidine can be very effective in lowering the blood pressure. Clonidine is also effective in treating withdrawal from some addictive drugs.

Clonidine often causes drowsiness and dry mouth. The sedation from clonidine can limit its use.

Clonidine works both in the brain and outside the brain. It decreases the blood pressure and often causes drowsiness.

Erythropoietin (Procrit™)

Procrit™ (Erythropoietin) is a particular hormone that is used as a drug. Erythropoietin in the body is released into the bloodstream by the kidneys and acts on the bone marrow to increase the production of red blood cells. Procrit™ is helpful to treat low red blood cell counts (anemia), such as in kidney failure. Anemic patients look pale and feel tired.

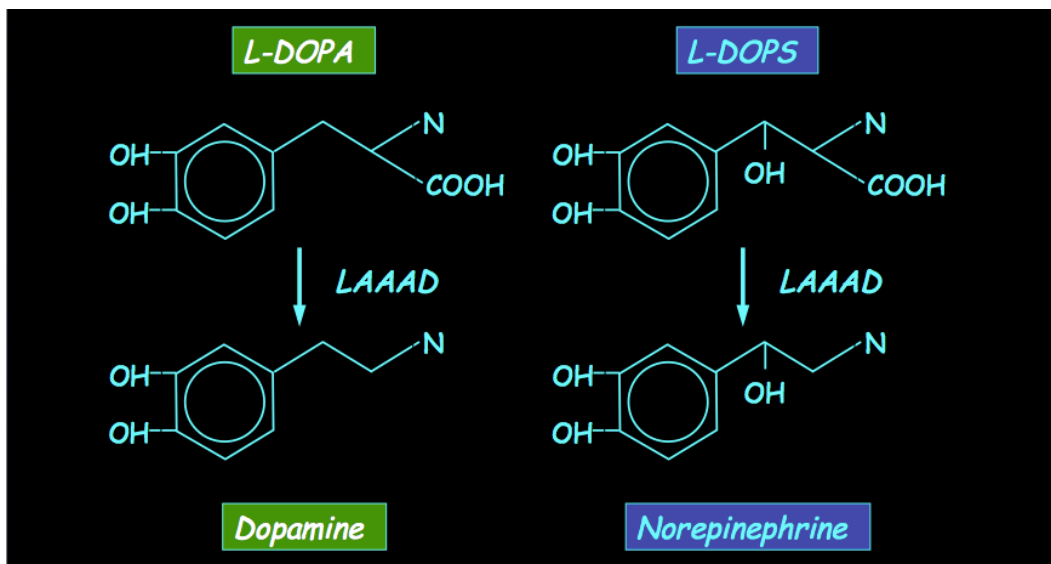
By mechanisms that remain incompletely understood, Procrit™ tends to increase the blood pressure. Some doctors prescribe Procrit™ to treat low blood pressure in patients with chronic fatigue syndrome who have a low red blood cell count.

## L-DOPS (Northera™)

L-Dihydroxyphenylserine (L-DOPS, droxidopa, Northera™) is a type of chemical called an amino acid. It is very closely related chemically to L-dihydroxyphenylalanine (Levodopa, L-DOPA), which is an effective drug to treat Parkinson's disease. L-DOPA works by being converted in the brain to the catecholamine, dopamine. L-DOPS works by being converted to the closely related catecholamine, norepinephrine, mainly outside the brain.

DOPS is converted to norepinephrine, like DOPA is converted to dopamine.

L-DOPS is a neutral amino acid and as such it is taken up into all cells



*As L-DOPA is converted to dopamine by L-aromatic-amino-acid decarboxylase (LAAAD), so is L-DOPS converted to norepinephrine.*

via the neutral amino acid transporter. In cells of the gut, liver, kidneys, and other organs that contain abundant L-aromatic-amino-acid decarboxylase (LAAAD), L-DOPS is converted to norepinephrine (NE). This means that L-DOPS can provide NE even in the absence of sympathetic nerves.

Because L-DOPS is a norepinephrine pro-drug, L-DOPS administration leads indirectly to stimulation of alpha-adrenoceptors in blood vessel walls, causing the vessels to constrict and increasing the blood pressure.

L-DOPS was recently approved by the US FDA for symptoms of orthostatic hypotension.

A potential problem with using L-DOPS to treat orthostatic hypotension in patients with Parkinson's disease is that the patients often are treated at the same time with Sinemet™. Sinemet™ is a combination of L-DOPA and carbidopa. The carbidopa interferes with the conversion of L-DOPA to dopamine. Since carbidopa does not enter the brain, the combination results in increased delivery of DOPA to the brain and increased production of dopamine. Carbidopa also interferes with the conversion of L-DOPS to norepinephrine. This might blunt the hoped-for increase in blood pressure by L-DOPS treatment; however, it appears that the dose of carbidopa in Sinemet™ is too small to interfere with the increase in blood pressure.

## Yohimbine

When alpha-2 adrenoceptors in the brain are blocked, this increases sympathetic nerve traffic and increases the amount of norepinephrine release for a given amount of sympathetic nerve traffic.

Yohimbine works both in the brain and outside the brain. The drug increases blood pressure and the state of alertness.

Yohimbine blocks alpha-2 adrenoceptors in the brain and on sympathetic nerves, and so it releases norepinephrine from the

terminals. The released norepinephrine binds to alpha-1 adrenoceptors in blood vessel walls. This causes the blood pressure to increase.

Even though yohimbine blocks alpha-2 adrenoceptors in blood vessel walls, the drug releases so much norepinephrine, and there are so many alpha-1 adrenoceptors in blood vessel walls, that normally yohimbine increases the plasma norepinephrine level and increases the blood pressure.

In patients with chronic autonomic failure and an inability to regulate sympathetic nerve traffic in intact nerves, such as in the Shy-Drager syndrome, yohimbine releases norepinephrine from the terminals and effectively increases the blood pressure. Theoretically, in patients with autonomically mediated syncope or POTS, yohimbine might attenuate sympathoadrenal imbalance and thereby prevent fainting episodes.

Yohimbine can cause trembling, paleness of the skin, goose bumps, hair standing out, an increase in salivation, or emotional changes.

Oral yohimbine was approved as a prescription drug to treat impotence from erectile dysfunction in men. Yohimbine in the form of yohimbe bark can be purchased in health food stores.

**Bethanechol (Urecholine™)**

Bethanechol is a drug that stimulates receptors for acetylcholine, the chemical messenger of the parasympathetic nervous system. The drug increases production of saliva, increases gut activity, and increases urinary bladder tone.

Bethanechol increases the muscle tone of the bladder, digestive motions of the gut, and salivation. It might be useful to treat urinary retention or constipation in patients with chronic autonomic failure.

Urecholine™ increases production of saliva, increases gut activity, and increases urinary bladder tone.



## **LIVING WITH DYSAUTONOMIAS**

Living successfully with a dysautonomia requires understanding about how the body's "automatic nervous system" (autonomic nervous system) functions and how changes in autonomic nervous system function cause symptoms. The first part of this book covers these topics.

This section, on living with dysautonomias, is focused on the patient, you.

Living successfully with a dysautonomia also requires understanding how chronic illness impacts patients, caregivers, and families—at home, at school, and at work. The changes you and your family may face can impose new emotional burdens. Coping with a form of dysautonomia almost certainly necessitates important changes in lifestyle. This section offers practical guidance for living successfully with dysautonomias.

### **Finding and Working with a Physician**

Because there are many different types of dysautonomias, and because disease mechanisms in dysautonomias often are not well understood in individual patients, your doctor and you will likely spend a lot of time trying to find reversible causes and devising a treatment

program. For these reasons the relationship between your physician and you is crucial.

Despite the fact that dysautonomias affect over a million Americans, you will probably find that very few people and surprisingly few doctors have ever heard of dysautonomias. It may be that no doctors in your area specialize in treating autonomic disorders.

### Few doctors have heard of dysautonomias.

Research over the last few years has increased awareness of the large number of people who are affected by dysautonomias. Dr. David Robertson, of the Autonomic Dysfunction Center at Vanderbilt University, has called this awakening an “epidemic of disease recognition.” With growing awareness, these disorders should become easier to recognize and treat.

The medical terminology can be confusing even to doctors. The same basic set of symptoms and signs can be called by a variety of names. For example, symptoms of a long-term inability to tolerate standing up—chronic orthostatic intolerance—have been labeled as “POTS” (Postural Orthostatic Tachycardia Syndrome, or postural tachycardia syndrome), “COI” (chronic orthostatic intolerance), Mitral Valve Prolapse-Dysautonomia Syndrome, Neurocirculatory Asthenia, Soldier’s Heart, neurally mediated hypotension, and other names in a long list. No wonder many patients feel frustrated and confused!

Finding a physician able to diagnose, treat, and follow patients with dysautonomias will likely take effort on your part. Unlike diseases or conditions that affect only one part of the body, dysautonomias can affect virtually every organ and system. Components of the autonomic nervous system play a variety of roles in regulating many largely automatic, involuntary, unconscious functions, such as breathing, blood pressure, heart rate, digestion, and urination. Because of the multi-dimensional aspects of dysautonomias, it is often difficult to determine which type of physician should manage the condition.

Your care will likely also require extra effort by your doctor. Since the cause of your symptoms may not be well understood, developing an effective treatment plan is likely to take time. People with dysautonomias must be both patient and persistent. Because of large differences among patients, and continuing mystery about mechanisms of dysautonomias, doctors need to learn from their patients about what works and what doesn't.

Your first priority should be to find a physician willing to work with you. Whether that physician is a cardiologist, neurologist, endocrinologist, psychiatrist, internist, or family practitioner is less important than his or her ability to work with you and other physicians on your behalf.

## Find a doctor who will work with and learn from you.

Because little is known about underlying mechanisms of many forms of dysautonomia, the physician will probably focus on treating symptoms without really knowing their exact cause. For this reason, much of what is done is through trial and error. Both you and your physician will need to understand that finding a program that works requires time, patience, and open and honest communication. Your relationship and ability to communicate with your doctor will make a big difference in putting together an effective therapy program.

Your symptoms are likely to change over time. Keep your doctor informed about how you are doing and about changes you notice. For instance, a particular medication might make you feel better in one way but worse in another. Your doctor might be able to change your prescription or start you on another drug that would work the same way but with fewer side effects. If you notice major improvements, you should inform your doctor. It's possible you may not need as much medication to manage the problem.

## Keep your doctor informed.

You should develop a plan with your doctor about symptoms that require immediate attention and those that can wait for a return visit. A brief discussion about this will help give you peace of mind when your symptoms are of concern.

Talking with a physician about multiple symptoms can be a problem, if you've had unpleasant interactions at office visits in the past. You might be concerned about what your doctors might think: "What if they think I'm CRAZY?" Don't let this concern keep you from relaying everything the doctor needs to know. You can't expect your physician to put the puzzle together if you withhold half the pieces. Tell your physician about all your symptoms. Let your doctor decide what is important information.

Create a bullet list of questions to ask. Keep in mind that your doctor has limited time to discuss your condition and treatment. Before visiting your doctor, ask yourself, "If I could improve one symptom, which would it be?" This type of thought process will give you and your physician a better opportunity to work on the symptoms that cause you the most trouble.

You may want to consider having a family member or friend go with you. Having someone with you may make you feel more comfortable, and a family member or friend can also give your physician details you may not recall.

Keeping a daily journal can also be a useful tool, both for you and your doctor. This may allow your doctor an opportunity to diagnose your condition and see trends or patterns in your symptoms. You might include blood pressure, pulse rate, body weight, and the timing and circumstances of events that trigger symptoms, mood, activity,

external temperature, time of day, time of the month, fluid intake—even thoughts. Talk to your doctor about which information to record. Let your doctor review your journal, since what may seem insignificant to you may be significant to your doctor. It is part of the nature of dysautonomias that symptoms often have peaks and valleys, and patients have good days and bad days.

If your doctor starts you on a new medication, it is important to discuss potential side effects. It is helpful to identify which symptoms are triggered as side effects of drugs and not as a result of your condition.

## **Day by Day with Dysautonomia**

### **Chronic Illnesses**

By and large, dysautonomias are chronic. They can continue for long periods or even indefinitely. Many factors affect their courses, including heredity, environment, drug and non-drug treatments, and lifestyle. Living with a chronic illness poses continual challenges, marked by many ups, downs, and unexpected turns.

Living with a dysautonomia poses continual challenges.

### **Accepting Your Disorder**

With an acute illness, you know you will eventually feel normal again.

When you have a chronic illness, there often is no cure in the traditional sense. You may never return to your “normal” way of life. Adaptation and acceptance therefore become important in maintaining your quality of life.

The first step to accepting your condition is to understand it. Knowing the “details” (e.g., common symptoms) can alleviate uncertainty and help you learn how to manage life with a dysautonomia.

Understand your condition.

## Modifying Your Life

in the past you might have been able to work 8 hours and then do chores at home. Now doing so might put you in bed for a week! You may have to learn to pace your activities, take “baby steps.”

Coping successfully with a chronic illness requires significant lifestyle changes. Modifying your lifestyle to help you maintain as “normal” a life as possible can help you gain a sense of control over your illness, rather than feeling your illness controls you.

Making a weekly chart of activities/tasks can help. You might believe that you are not doing anything, yet when you review a list of your actual activities you might find you are trying to accomplish a great

deal. The list can help you set priorities about tasks that definitely need to be accomplished or can be put off or eliminated. This sort of chart can also help in decisions about how responsibilities can be shared among family members. For example, your spouse might take over the grocery shopping. Deciding on the right balance between overdoing it and doing too little will take time and a lot of trial and error.

Doing things you enjoy can distract you from your illness. Focus on hobbies and activities you can still do and look for new ones to replace those you no longer can pursue. An example would be avoiding noisy shows outdoors in the heat and instead attending quiet shows in the cool indoors.

Take an inventory of your interests. People often forget about things they had an interest in but have not thought about for years.

Know your limitations. Substituting one activity for another may become necessary to maintain your sense of well-being.

## Daily Life Tactics

Here are several basic tips to pace your life.

- Get adequate rest.
- Eat and drink right. Don't fast, and don't pig out.
- Try to keep a regular schedule.



- Get an appropriate amount of exercise, as prescribed by your physician.
- Avoid dehydration.
- Stay on your medication routine.

Mornings can be rough for people with orthostatic hypotension from chronic autonomic failure. Start slowly and use your knowledge. Studies have shown that autonomic failure patients can have a surprisingly large increase in blood pressure by drinking 2 glasses of water. You may find that drinking water about 15 minutes before getting out of bed in the morning helps you tolerate standing up. If your physician has advised you to increase your intake of fluid and salt, a glass of V8 or tomato juice might be helpful, as these drinks contain large amounts of sodium. Eating a large meal can shunt blood to the gut and decrease the ability to tolerate standing, and exposure to heat can decrease the blood pressure. If you attended a large church breakfast in the summer before standing still through a service, you could easily have a severe enough of a fall in blood flow to the brain to cause you to faint.

Exercise plays an important role in treating most chronic conditions, including dysautonomias. Staying in shape improves your sense of well-being. The veins in the legs contain one-way valves that allow blood to flow towards your heart without allowing it to back up into the legs. Muscle surrounds deep veins in the legs and compresses these veins when you contract your leg muscles. Muscle pumping

helps to keep blood moving towards the heart and upper body when you stand upright. You can do different types of exercise to assist your venous pump. You can tighten your calf, thigh, and buttocks muscles. Ask your physician about whether muscle pumping exercises would be appropriate for you.

Chronic illness, and especially chronic illness from an abnormality of the function of the autonomic nervous system, can increase the susceptibility to anxiety, panic, and depression. There is nothing wrong with asking your doctor if you might benefit from medication to help you cope.

Avoid triggers that worsen your condition. Some triggers to keep in mind are:

- Hot environment (e.g., hot shower, sauna, Jacuzzi)
- Dehydration (not getting enough fluids)
- Emotional distress
- Over-stimulation (i.e., amusement parks, concerts, sporting events, video games, loud telephone ringing)
- Large meals
- Skipping meals
- Alcohol
- Skipping medications

## Diet

Eating large meals tends to shunt blood toward the gut. This can worsen orthostatic intolerance and make a dysautonomia patient feel sluggish, tired, and worn out. Try eating smaller meals, more often. Check if sugary or starchy foods tend to worsen your symptoms. During eating, you might try elevating your feet to heart level and exercise your legs, to keep the blood from pooling. Just flexing your feet back and forth might provide a benefit.

For many patients with dysautonomias, a diet high in salt and fluids is necessary. Chicken noodle soup and V8 juice contain large amounts of salt. You should discuss salt intake with your doctor.

## **Environmental Temperature**

Patients with dysautonomia often have intolerance of heat or cold. If you have heat intolerance and plan on being outdoors during the summer, dress in cool, light clothes and limit the amount of time you spend in the heat.

You might feel faint taking a hot shower in the morning. Consider taking your shower prior to going to bed at night.

## **Compression Stockings/Abdominal Compression**

Compression stockings can help patients who have excessive blood pooling in the lower half of the body when they stand up. If you use compression stockings, it can take some time to take them on and off. You may find it easier to put your stockings on and off while lying in

bed. Lying down may also keep you from becoming symptomatic while taking them off. A small amount of baby power help when putting them on. Compression stockings may be ineffective in preventing a fall in blood pressure standing.

You can buy affordable pantyhose to reduce pooling of blood in veins during standing. Try two pairs, one size smaller than what you would normally wear, and wear both at the same time. Abdominal compression has also been used to help prevent blood pooling when you stand up. Depending on your particular condition, a girdle one size too small can make a difference in how you feel. If wearing girdles or compression stockings isn't your style, try wearing bicycle pants.

## **Medic-Alert Bracelets**

Patients with dysautonomias should wear a Medic-Alert bracelet. The back of the bracelet can state "See wallet." Inside your wallet you can have a piece of paper, laminated card, or electronic memory media about your condition, medications you take, allergies and sensitivities to medications, names and phone numbers of physicians, and emergency contact information for spouse or friend. For information on obtaining a Medic-Alert bracelet, visit <http://www.medicalert.com>.

## **Work**

Whether or not you keep working is an individual decision affected by a number of factors (e.g., severity of symptoms, type of work,

financial situation).

**It is likely your ability to work will be affected in some way by your illness.**

It may be that you can no longer work full days, or you may no longer be able to travel as part of your job. If your job requires you to be on your feet all day, this may not be possible any more.

You might have to struggle with what if anything to tell your employer. Do you maintain your privacy, or let your employer know, so special accommodations can be arranged? This is a personal decision, with no universal right or wrong answer. It may help to make a list of the pros and cons of disclosing your condition. Many things are going to affect your decision, including your specific work environment and job duties.

Work can involve episodes of emotional distress even in healthy people, so it's no surprise that it can worsen symptoms in someone with a dysautonomia.

You're probably going to have to make changes at work. This might mean setting more limits. It can be scary and frustrating to have to "slow down" at work. You might be afraid of what will happen and what people will think of you. You have to remember that if you don't slow down, you may be jeopardizing your health, which in the

long run will result in being able to do even less. If you are contemplating taking time off from work, be sure to investigate all your options regarding possible assistance. You might be able to telework.

There may come a time when you have to discontinue working altogether. The decision to leave the work world, whether temporarily or permanently, can be accompanied by a whole host of emotions, including anxiety, depression, guilt, or relief. To minimize anxiety associated with leaving work, structure your day (e.g., read books, listen to music, take a course over the internet, talk with friends), and try to learn something new. Make a list of your positive traits, to remind you that you are of value even if you're not working. Social networking with others in your situation can alleviate the sense of loneliness.

## Travel

Driving is one of the most important aspects of our independence and often a necessity of everyday life. Discuss driving with your doctor. Your doctor can help to determine if your condition puts you at risk. If you are not able to continue driving, you will have to find ways others can help with your travel needs. Besides family, friends, and neighbors, your community may have programs. Your local Chamber of Commerce or United Way can give you information about public transportation and other programs.

Wearing sunglasses when you travel can reduce stimulus overload. You may notice that your symptoms don't seem as intense when you travel in the evening than in the daytime, or vice versa. Wearing earplugs can also help reduce the impact.

Depending on your specific condition, wearing a girdle, compression stockings, or bicycle pants while traveling may be helpful. Have you ever noticed a change in your skin color when you stand upright? Rapid changes in the color of the skin are the result of blood. Compression garments may help you to keep blood in the upper part of the body when you are standing on line.

For many patients with dysautonomias, air travel can be a nightmare. It is best to discuss this with your physician. If your physician tells you it is all right for you to fly, discuss the following to see if they make sense for you:

- Drink extra fluids for at least a couple of days before departure.
- Eat a diet high in salt (V-8 juice, chips, pretzels, beef jerky, pickles).
- Avoid stressful, stimulating situations the day before or of departure. For instance, avoid going to the mall for last-minute shopping.
- Wear compression stockings and an abdominal compression garment.
- Wear earplugs or eyeshades.

- Ask your doctor about a medication to calm you and enable you to sleep during the flight.
- Fly with someone who knows your disorder.
- Request bulkhead seating, so you can elevate your feet to heart level during the flight.
- Request a wheelchair at your destination.
- Try to arrange a day of rest after your flight.

## When to Ask for Help

It is not easy to find the right balance between independence and seeking help. At different points, you may need practical, financial, emotional, or physical help.

**We all need help from others, whether we're healthy or not.**

People often feel guilty asking for help from family and friends. Think about how things would be if the shoe were on the other foot. If your spouse or best friend had a chronic illness that required your assistance, would you resent a plea for help?

Explaining exactly how someone can help can provide a sense of relief to the helper, who may not know what to do. Don't assume that others can read your mind. You need to be clear in relating how you feel and what you need. You can make a list of the areas where you do and do not need assistance. Your friends, family, and caregivers need to do the same. You may not be sure what you want.



## Social Activities

Staying involved in family and social activities as much as possible can help you cope with your illness. If you notice that these activities make your symptoms worse, then limit the time you spend on them. For example, if a family picnic were an all day function, you might plan on staying for only an hour or two.

Obviously, you do not experience your illness in a vacuum. Those close to you are also impacted. They won't experience the same physical effects you do, but they will share other struggles (e.g., emotional, financial). This is a time of heightened stress and anxiety for the entire family.

Try to arrange a quiet time to sit down and talk with your family about issues related to your health. Explain clearly, and speak directly. Ask if they understand what you're trying to say, and clarify what is not clear. Listen to what they have to say. Try to express yourself in a non-threatening manner. Statements like, "Why do you always avoid me?" will probably make your loved ones feel attacked and cause them to become defensive. Instead, try to phrase your statement in neutral terms, such as, "Help me understand what you are going through. I feel like you don't want to be around me anymore and that hurts me. I miss being around you." Remember that no one will be put off by your expressing how you feel.

Your loved ones should also be allowed to express their feelings. They may be experiencing some of the same emotions you are, including anxiety and guilt. Anger and other negative emotions are also likely and normal. You and your family members can expect to feel hurt at times. Try to remember that these negative emotions are reactions to the situation and not to you yourself.

## **Attitude is Half the Battle**

It is natural to have negative thoughts when your world seems to be crashing. People with chronic medical conditions are susceptible to experience emotional distress, fear, depression, anger, frustration, anxiety, or other negative emotions.

Keeping a positive attitude will help you move on with your life. You must meet your challenges with determination. Blaming or attacking your physician, family, friends, or even God won't improve your health. Having a positive attitude might make things easier on your family, friends, and neighbors.

This sounds rather platitudinous. What practically can be done? Talking to others with the same condition can help. There is nothing wrong with discussing your anger, frustration, concerns, and fears. A health psychologist may help you acquire coping strategies. Some psychologists emphasize the importance of a "family session," where all members of the family can relate the effects that the illness has had on them. Keep in mind that the entire family is impacted by your

illness.

It's been said that the key to happiness is appropriate expectations. Take time to recognize your abilities and what you can do. For example, you may need help with grocery shopping but not with putting the groceries away. It may take time to discover what you can still do despite your limitations. Make small goals. Your goal today might be to walk from the bedroom to the kitchen. Next month it might be to clean the kitchen.

## **Referral to an Autonomic Specialist**

Physicians in several fields of medicine specialize in dysautonomias. Testing in a specialized autonomic function laboratory can help identify what form of autonomic involvement you have and speed development of an effective therapy program.

### **Consider specialized testing.**

You should not feel reluctant to talk to your physician about going to another facility for testing. You will likely find that your physician will actually encourage you to do so, because the visit may provide valuable and otherwise unobtainable information that your doctor can use to help you.

Keep in mind, however, that there are relatively few autonomic function experts and testing laboratories, and an educated general

practitioner can take care of most of the management of dysautonomia patients. For a list of physicians and facilities in your area, try visiting the websites of the American Autonomic Society, at [www.americanautonomicsociety.org](http://www.americanautonomicsociety.org); Dysautonomia International, at [www.dysautonomiainternational.org](http://www.dysautonomiainternational.org); or the Dysautonomia Project, at [thedysautonomiaproject.org](http://thedysautonomiaproject.org).

### **Research Facilities - Should I Participate in a Study?**

There are a limited number of academic medical centers in the United States that conduct research on the autonomic nervous system, such as at Vanderbilt in Tennessee, the Mayo Clinic in Minnesota, the Harvard system in Massachusetts, NYU in New York, the University of Texas in Dallas, and at the National Institutes of Health (the NIH) in Bethesda, Maryland. Different centers study different types of dysautonomias. Patients are recruited to participate in research studies (also known as “protocols,” because the studies are designed, defended, approved, monitored, and reported according to pre-determined, detailed, written criteria). Each protocol has specific requirements, both for inclusions and exclusions. For a list of ongoing studies you can contact the NIH’s Clinical Trials web site at [www.clinicaltrials.gov](http://www.clinicaltrials.gov).

Participation in a research study may help; however, it is important that you investigate the study thoroughly and review the consent information prior to participation.

Some benefits of participating in research are:

- You are seen by people who specialize in this area of medicine. What may be unusual for your local physician may be routine for the investigators conducting the research.
- You have the opportunity to learn more about what may be causing your symptoms. The testing could reveal important information about your condition that may not be available to your personal doctor.
- The medical institution typically covers the costs of the research testing, which otherwise would be expensive if available at all.
- Even if you don't benefit personally from your participation, you help researchers understand the illness better, making it possible for them to devise better treatments.

If you decide to participate in a study, keep in mind some of the possible limitations of the research:

- You may be required to stop taking your medications, for the doctors to see how you function without them.
- You may have to pay for travel.
- Some tests can be painful, uncomfortable, or not directly related to your problem.
- You may have to spend several days in the hospital.
- You may need pre-certification from your insurance company.
- You have to meet the criteria for participation in the study. Not

everyone qualifies, and research patients may not be recruited once a quota is filled.

— Most important, you should understand that the usual primary focus of a research study is not to help a single patient but to learn more about the condition in general. Research studies therefore may not provide for your long-term care or follow-up. This means that you will likely be returning to the care of your personal physician after participating in the research. Nevertheless, the researcher and the study results may help you and your doctor gain more knowledge about your condition and help devise an effective therapy program.

**Physicians conducting research should not take the place of your local physician.**

The research might give you immediate results, but alternatively it might take several months or even years before the research is completed and the results fully analyzed. You should have a clear understanding of what type of feedback to expect prior to your participation.

Keep educated about your condition. Passing along new information will help both you and your doctor. You will find that most physicians appreciate information provided them, especially if from a reliable source. Resource tools available today allow you a tremendous opportunity to stay abreast of new discoveries. You can find updates from a variety of sources (see the listing later in this section), patient conferences, books, and newsletters. The National Library of

Medicine's websites offer you easy access to medical search engines that can also help keep you informed of new research discoveries.

## **Caregiving and Support**

Caregiving is taking care of and feeling responsible for another person, loved one, or family member. Family caregiving is extremely important for coping with and successfully managing dysautonomias.

### **Family Caregiving**

A family caregiver is someone who feels primary responsibility for the well-being of another family member experiencing chronic limitations as the result of illness or injury. Caregiving has many facets, and each situation is different. The spectrum of caregiving responsibilities and capabilities may entail emotional, physical, social, practical, financial, logistical, and psychological care and support.

It is difficult to identify caregivers, because they don't feel that they are caregivers. Much of what caregivers do is out of love, respect, and being "family." The emotional and practical wear and tear on caregivers is real and needs to be understood. Caregiving doesn't come with a set of instructions, and after months or years caregiving can feel like a rut or trap. Without understanding the responsibilities of family caregiving many succumb to anger, resentment, confusion, and even physical ailments.

First and foremost is the need to recognize the role of being a caregiver. Not recognizing the caregiver role inherently prevents one from getting the understanding, help, support, and resources caregivers need.

## Family caregiving is hard.

Why is caregiving so hard?

— Family caregiving involves routine and repetitive day-to-day psychological and social issues, economics, and perhaps physical care needs, and ongoing balancing act of work, household, and other activities.

— Family caregiving is not intuitive. Your maternal/paternal instincts and childrearing experience are not substitute training for family caregiving.

— There are numerous role reversals, such as kids caring for parents.

— People tend to wait for a crisis rather than plan strategically.

— Family caregivers feel transparent, with everyone focused on the care receiver and not appreciating the caregivers' efforts. Family caregivers can feel lonely, like they are in this by themselves and that



no one understands what they are going through.

People rarely know what they don't know. Without instructions, planning, and clear understanding of the caregiver role, ongoing problems get harder to solve. Expectation management is a key ingredient in being a successful caregiver. A key to happiness is appropriate expectations.

### Caregiving for a dysautonomia patient is special.

Why is caregiving for someone with a dysautonomia different?

— People with a form of dysautonomia often don't look sick. Family, doctors, friends, schoolmates, and relatives have a hard time believing in the reality of the illness. Suspicions of malingering, psychosomatic illness, and “being lazy” are aroused continually.

— Dysautonomias typically are chronic illnesses. A chronic illness or disability such as congestive heart failure or stroke in an elderly person may mean 5-7 years of caregiving. When the onset is at birth or during adolescence, we may be talking about almost an entire lifetime. The younger the individual when illness strikes, the greater the scope of impact, including school, social life, relationships, future goals, responsibilities, work, and the entire family structure.

Kids don't think of themselves as caregivers, and they may be frightened and confused by the feelings they have. Most doctors and

teachers do not think about children in this sort of role. If your children have this role, they need special support and a trusted outsider to talk to as well as Mom or Dad.

## **Spousal Caregiving by Men**

For reasons that remain poorly understood, most patients with functional dysautonomias such as POTS are women. Spousal caregiving by men can be difficult. Seeing a wife or partner suffering and feeling inadequate to relieve the suffering can create a sense of emotional impotency. Physical sexual and other shared pleasures may be limited or lost, leaving the husband feeling lonely and unappreciated.

Lost opportunities for promotion, business travel, or increased responsibility add to the burden. The potential alteration or dissolution of plans, dreams, and expectations of life imposed upon by chronic illness must be faced. The loss of an anticipated future must be grieved. The process of grieving goes through stages from denial to acceptance and may last for years. The partners may be at different stages on the road to acceptance.

Unresolved issues from the past with family or with spouse may become overwhelming. The role of spousal caregiver may not always be possible. Some will leave. Often, however, one may find courage, strength, and renewed love in long-term commitment to stay in the relationship.

## Intimacy

Intimacy, which is important in a normal relationship, is greatly impacted and strained by the limitations of dysautonomias.

Intimacy is a major issue in caring for a spouse with a dysautonomia.

You can love someone and never be intimate or sexual with him or her.

You can have sex and never have intimacy with, or love for, the other person. You can love someone and have great intimacy without having physical sex. Whatever works for you is fine. The subject of intimacy is at the core of many of the issues couples face; it is inescapable for those dealing with chronic illness.

With dysautonomia you may look fine but feel awful. When you feel lousy, you don't feel sexy. That's a strain on any marriage or relationship.

## You Are Not Alone

Whatever your beliefs, or whether you have a formal religion, having a sense of spirituality, an awareness of a guiding creative force, or a sense of transcendence can be a comfort and a coping mechanism. Use this as it fits for you.

It is likely that for a relationship to work in the setting of a dysautonomia will require outside professional help. If you are a family caregiver, recognize you are not alone. Others have worked through similar life- changing events. You must recognize your problems and actively seek your own help. No one else is automatically coming to solve them for you.

Major organizations with family caregiver support create an opportunity for defining roles, outlining responsibilities, sharing information, and gaining better understanding. Just as important as knowing what doctor to go to and what medication to try is to recognize the major burden of family caregiving with the knowledge that you are not alone. Understanding this is not only helpful to those with chronic caregiving responsibilities but also to spouses, children, other family members, friends, and the community.

## **Support Groups**

Support groups are an invaluable tool to help deal with the consequences of dysautonomias. There can never be enough sharing thoughts, helping one another, learning, and listening.

One of the best sources of help is a support group. A support group is a regularly scheduled, informal gathering of people whose lives are affected directly by a chronic illness or by the caregiver role. Members benefit from the peer acceptance and recognition of their

common concerns and are grateful for the wisdom, insight, and humor of people in the same situation.

Learning coping techniques from others in a support group is extremely valuable. Patients with chronic illness need reliable guidance— understandable, clear, compassionate, and practical. Including the caregiver, significant other, or family members is especially important. Participants in support groups learn quickly from one another. Professional facilitators help accomplish even more.

Support groups are also a safe place to be heard and to listen and to understand symptoms and treatments. Support groups offer understanding on how to “reinvent yourself,” how to work with your healthcare team, how to communicate better with family and caregivers, and how to acquire effective strategies for daily living.

Today, physicians, social workers, rehabilitation specialists, neuropsychologists, and others refer patients to a recognized support group. Below is a listing of some dysautonomia support groups and their web addresses. The numbers of such support groups and sites are growing rapidly.

- Dysautonomia International ([dysautonomiainternational.org](http://dysautonomiainternational.org))
- The MSA Coalition ([multiplesystematrophy.org](http://multiplesystematrophy.org))
- The Dysautonomia Foundation, Inc. ([familialdysautonomia.org](http://familialdysautonomia.org))

- The Dysautonomia Project ([thedysautonomiaproject.org](http://thedysautonomiaproject.org))
- Dysautonomia Information Network ([dinet.org](http://dinet.org))
- National Dysautonomia Research Foundation ([ndrf.org](http://ndrf.org))
- Syncope Trust and Reflex Anoxic Seizures organization ([stars.org.uk](http://stars.org.uk))
- Dysautonomia Youth Network of America ([dynakids.org](http://dynakids.org))
- National Society For MVP and Dysautonomia ([mvprolapse.com](http://mvprolapse.com))
- Dysautonomics ([adiwebsite.org](http://adiwebsite.org))
- Fight Dysautonomia.org ([fightdysautonomia.org](http://fightdysautonomia.org))
- American Dysautonomia Institute ([dysautonomics.com](http://dysautonomics.com))

Taking the initiative to begin a support group and following through is a major commitment but with many rewards. It doesn't take special training, but it does take effort, dedication, and some ingenuity. You may also find it to be very rewarding.