NATIONAL DYSAUTONOMIA RESEARCH FOUNDATION

NDRF CHAT

National Dysautonomia Research Foundation **Spring 2003**

NDRF Conference 2002 Highlights

Inside this issue:

Patients, caregivers, doctors, elected officials, and federal agency representatives were all in conversation about Dysautonomia when the NDRF hosted its second Patient/Caregiver Conference in Washington, D.C. July 18-20, 2002. Thanks to the enthusiastic yet thoughtful participation of all involved, the three day conference was a wonderful success.

Inside this issue are highlights of the conference. Physician speakers included:

⇒ David Goldstein (NIH)

⇒ Cecil Coghlan (Univ. of Alabama)

⇒ Blair Grubb (Med. College of Ohio)

⇒ Phillip Low (Mayo Clinic)

⇒ David Robertson (Vanderbilt Univ.)

⇒ Julian Stewart (NY Medical College)

⇒ Roy Freeman (Beth Israel Deaconess)

⇒ Ken Davis (PhD., Ohio)

Topics were wide-ranging, from understanding the role of neurotransmitters in Dysautonomias, to coping and management strategies.

Over 275 people came together for the NDRF Conference this past July.

Inroads at the National Capitol

At this past summer's conference, Dan Smith, the NDRF President, summarized the week's contacts with congressional representatives. He, along with Executive Director Linda J. Smith, Drs. David Goldstein, Blair Grubb, MSA patient Bob Miller, and Orthostatic Intolerance patient Lynn Adams - met with key Congressional leaders, including the late Senator Paul Wellstone from Minnesota and Senator Jeff Sessions from Alabama. Both sit on the Senate Health Committees that make recommendations for health care funding.

Four key issues the group brought forward are:

- The lack of research dollars for Dysautonomias
- The lack of physician knowledge about Dysautonomias, resulting in inadequate local care as well as unnecessary tests and costs
- Difficulties getting managed care to allow referrals to knowledgeable physicians

("Congressional Inroads," Continued on page 2)

Congressional Inroads (cont. from p. 1)

- The lack of an adequate number of physicians for on-going care

The group requested funding for efforts to address these needs. Those requests included funding for Centers of Excellence and for research efforts. NDRF representatives pointed out that no federal funding agency has dysautonomias as a specific research category. That fact, along with the interdisciplinary nature of autonomic conditions, means no single specialty group owns it, which reduces the chances of proposals receiving funding.

The senators were informed that dysautonomias are primarily a women's health issue, and that misdiagnosis is common and that many patients are told they have "Psychological Disorders". They also discussed the difficulties obtaining Social Security Disability Insurance (SSDI) which can occur because the rules do not include a classification for autonomic disorders. Children's research lacks direction, and is extremely limited.

The enormous lack of educational material and support for people with Dysautonomia was brought forward as an issue, and contrasted with the opportunities for people with better-known conditions such as cancer or heart disease.

The group was successful in gaining a commitment from Wellstone and Sessions to help develop legislation to request funding to address research needs. Sessions also made a commitment to contact others for support. garner his support. This is a huge step forward, but only one of many that the NDRF, with your help, will need to take to assure the needs of the dysautonomia community are better met in the future.

Audrey Carpenter, who has lobbied for a variety of public health care programs and spent several years at the National Institutes of Health as a health care provider, discussed the importance of patient involvement, and the important role that "Fundraising" can give to this enormous group of patients. She encouraged people to do their part to support efforts. Her suggestions included doing local fundraisers, asking legislators to speak on health care issues at civic organizations, churches and businesses, and helping to raise awareness about dysautonomia at the NIH.

To express you feelings to our government, please contact the NDRF Congressional Aide, Audrey Doyle, at audrey.doyle@ndrf.org. Alternatively, write to the NDRF and share the story of the impact that Dysautonomia has had on you and your family.

"...there is such a thing as perfection...and our purpose for living is to find that perfection and show it forth....Each of us is in truth an unlimited idea of freedom. Everything that limits us we have to put aside."

~ from **Jonathan Livingston Seagull**, by Richard Bach

Conference Q/A

Q/A with Dr. Philip Low

The following are Dr. Low's responses to questions at some of the question/answer sessions at the July 2002 NDRF conference.

1. In MSA with OH, what causes stuffy nose and what can be done about it?
It's common for people with MSA to have prominent mouth secretions – the amount of secretions is normal but they have trouble evacuating it. Dr. Low sometimes uses botulism toxins to treat this problem. The toxin is injected into the parotid and submandibular glands to paralyze the autonomic nerves that cause salivation.

2. What is included in a good baseline POTS assessment? Is a tilt table test alone sufficient?
The most important aspect is what the patient tells you about their symptoms; the tilt only provides information on blood pressure and heart rate. Dr. Low feels there are three aspects to consider: a) presence of orthostatic intolerance, b) presence of spontaneous episodes, and c) associated fatigue.

> *"The most important aspect is what the patient tells you…"*

3. What do you make of complex situations with multiple organ impairment, but with a negative autoimmune antibody test?
According to Dr. Low, this type of situation could still be due to autonomic neuropathy, assuming that autonomic tests are abnormal. A negative antibody test doesn't rule out this diagnosis because a number of antibodies exist (many of which we don't know how to measure), and because there are sometimes cellular/immune changes without antibodies.

4. Do you have ideas for managing GI symptoms?
It's important to find out what is truly going on: is there nausea, bloating, or gastroparesis? If the symptoms are apparent only when standing up, they are probably related to sympathetic over-activity and can be treated as a part of OI. If gastroparesis exists, it is evidence of upper GI denervation, which has a different set of options for treatment, include a modified diet, gastric pacing, and medications.

> *"If gastroparesis exists, it is evidence of upper GI denervation,…"*

5. Is there any evidence of an increased number of infections in people with Dysautonomia?
Dr. Low's group hasn't found that to be the case in the 500 POTS patients in their database. However, a small subset has limited neuropathy associated with the immune system.

6. Is there any relationship between Dysautonomia and Bell's palsy?
Not that Dr. Low is aware of.

7. Can a person have POTS and PAF?
In the early stages of PAF, the heart will increase to compensate, but that compensation disappears as the disease progresses. There are usually enough differences between the two conditions to allow their separation.

8. Is there recent research that has had an impact?
The identification of autoimmune antibodies to acetylcholine, and the identification of a genetic defect related to abnormal release of NE are both significant. The latter is important because it is a door to understanding an exact mechanism. Parkinson's' research has also been important: Some of these patients have Dysautonomia and have loss of sympathetic nerves to the heart -the view of the disease may change from that of a brain disease to that of a type of Dysautonomia.

10. Is there any evidence that deep massage to the neck and upper back on a regular basis help in keeping the head more erect in MSA patients?
No scientific evidence to date.

11. How does a physician differentiate between Pure

("Low," Continued on page 4)

Low, continued

(Continued from page 3)

Autonomic Failure (PAF) and Multiple Systems Atrophy (MSA) both in terms of observation/clinical findings and testing?
There is usually no confusion because the two conditions are so different. In general, neurological features are more reliable than tests in separating the two. Rarely, a patient will start as PAF for a number of years then develop neurological features of MSA.

12. Have you seen much evidence that Dysautonomia can progress from Orthostatic Intolerance to Orthostatic Hypotension to Pure Autonomic Failure to MSA in the same patient?
Nothing is impossible in life but in practice this is rare. With a full autonomic laboratory, I have never seen POTS evolve into PAF.

13. What is the next step if you have increased salt, water and have done resistance training to the lower extremities for over 4 months and still not feeling much better?
Unfortunately this happens in some patients, especially those where fatigue is a major problem. Depending on the situation, a change in medications is an option. Some patients move from the non-responsive part of the curve to the responsive following a three week program of multidisciplinary rehab program.

"Numbness is usually part and parcel of neuropathy."

14. Do you recommend using a cholinergic agent for urinary frequency associated with Dysautonomia?
You mean an anticholinergic medication. It depends. Frequency alone is not an indication for anticholinergic medication, but urgency is (when you have to go you have to go).

15. Is peripheral neuropathy progressive, to the point where you become numb?
Numbness is usually part and parcel of neuropathy. Most neuropathies are progressive, but to varying degrees.

16. Are there cases of dysautonomia as a prelude or co morbid presentation with ALS?
Dysautonomia can occur in ALS, although it is usually mild.

17. How do you treat hypertensive POTS? What is the risk of strokes with this?
Depends on the situation. Most times this occurs because of medications the patient is taking, especially drugs like midodrine and Erythropoietin. Some patients have wild swings in BP with autonomic storms. Those patients sometimes respond to clonidine. Milder cases might respond to Phenobarbital or an SSRI. Stable hypertension will often respond to a nonspecific beta blocker.

18. Why would one bite of food or drink cause severe shortness of breath?
I don't know. Anxiety is one mechanism.

19. Why do problems go from system to system?
When many systems are involved and evaluation of these systems show no disease, somatic vigilance is a possible mechanism.

Low, continued

(Continued from page 4)

20. For people who have beta-sensitive POTS, but who are allergic to beta-blockers, what do you do?
POTS symptoms are real and need better individualized treatment. A problem has been that we keep finding apparently different mechanisms at work between patients, and thus we can't apply results universally to everyone - it's necessary to look at all aspects of each individual's situation. In POTS, there is an interplay with psychological issues that can sometimes make the POTS symptoms worse. In particular, symptom vigilance can interact with autonomic nervous system activation. Because of this, we now incorporate biofeedback and other techniques into our treatments. We look at symptoms more comprehensively and treat more comprehensively to get the best results.

"How do you treat hypertensive POTS?"

21. Have you seen problems with people having too much urine?
I have seen this, but haven't found any abnormality when I have studied these patients. They don't seem to have a kidney defect or an endocrine problem. People sometimes drink too much. If the volume of urine is more than 1.5-2.0 liters, the person is probably drinking too much.

22. What might cause a 15-20 pound fluctuation in weight? Is it possible the body is not using water correctly?
Some people will gain 2-5 pounds pre-menstrually, and it affects their symptoms. Otherwise, the standard way to study whether the body is using water correctly is a fluid balance study in the hospital to look at fluid/salt regulation. This involves a study of kidney function and the endocrine study of fluid and electrolytes. This sounds different than typical POTS and may be something else, which might be better addressed by a renal specialist.

23. What does it mean when a person has small fiber sensory neuropathy plus POTS plus hypertension?
Some people do have neuropathy with a burning sensation of their feet. It can be part and parcel of POTS and is called a distal small fiber neuropathy (DSFN). Regarding treatment, the approach is similar and involves volume expansion. Midodrine can be used carefully, but could be a problem if the patient is hypertensive. It is important to check if the hypertension is real or simply due to unstable BP, when it is due to the autonomic nervous system trying to compensate.

24. What would a person have who has severe gastroparesis, POTS, sicca complex, hypertension and other symptoms, but does not have MSA?
This person probably has a high ganglionic antibody titer with this complex. It is probably not progressive, and can be treated with gamma globulin. Among the factors that are predicators of a high titer are gastroparesis, sicca complex, and bladder problems.

25. How should I start an exercise program?
Start with physical counter-maneuvers, which are not complicated to learn, and are published elsewhere. These include crossing your legs and squeezing for 30 seconds. Add resistance training, which will increase the effectiveness of the counter-maneuvers. You can also contract thighs and buttocks when you walk.

26. How should a personal trainer adapt for a person with POTS?
Focus on leg lifts and other exercises for the legs and the waist and below. Work with the heaviest weights you can. Try to exercise with them 2-3 sessions per week, in 2-3 series of 12 repetitions. If you can't do 12 at a time, rest for a few minutes and then come back and finish.

Low, continued

(Continued from page 5)

27. Do people with POTS tend to have additional problems over time?
Regarding the prognosis for POTS, the information is limited. In our studies, conducted over 52 months, most people improved and could go back to work. However, they still needed treatment and could be very tired when they came home - they weren't "normal."

28. Will people who are treated promptly recover better?

For those that have post-viral onset, recovery seems to be more complete. We don't have data on the effect of rapidity of treatment on outcomes.

29. Do some people with dysautonomias tend to have insulin resistance?
There is interest in that some people have hyperglycemia seem to develop neuropathy quite early and the relationship between insulin resistance has been claimed to be better than that of hyperglycemia. Although insulin resistance isn't routinely studied in POTS, I don't feel it is a major mechanism.

30. Is it possible to have POTS and yet pass a tilt-table test?
The tilt response can vary depending on the medications a person is taking, their blood volume, and other factors. So, yes, the status of a tilt exam for a person with POTS can vary. That's why we look at other autonomic tests beyond the tilt exam to help diagnose someone.

"When one door closes another door opens; but we often look so long and so regretfully upon the closed door that we do not see the ones which open for us."

~ Alexander Graham Bell (1847-1922)

Note how good you feel after you have encouraged someone else. No other argument is necessary to suggest that never miss the opportunity to give encouragement.

~ George Adams

Q/A with Dr. Julian Stewart

The following are Dr. Stewart's responses to questions at some of the question/answer sessions at the July 2002 NDRF conference.

1. *Does blood pooling cause permanent damage?*
   No evidence of permanent damage exists.

2. *In POTS, what causes flushing and dry eyes?*
   Mastocytosis can cause both flushing and orthostatic intolerance (OI). In some cases, a tumor may cause flushing in people who flush frequently. It's possible that a cholinergic blockade is involved.

3. *Are there suggestions for hydration beyond water?*
   Water and salt have a limited effect; IV infusion seems to help more. Two cups of water will increase BP noticeably and increase sense of well being for several hours in pure autonomic failure (PAF), although the same effect is not seen in normal people. People can get osmotic dieresis if drinks have too much sugar and water, and so Dr. Grubb encourages use of water instead.

4. *Are there any medications that can help the venous pooling in the lower extremities that are safe to use?*
   "Pooling" remains largely unexplained. It does not to seem to result from changes in veins or their ability to contract but may instead relate to artery properties. Also it seems more associated with low blood flow and increased venous resistance than with collections of blood. General measures include support stockings. Midodrine can occasionally prove helpful.

5. *What can you do for severe nausea and why does it happen so much?*
   Nausea may result from processes local to the gut circulation or its local nervous system. The use of local receptor antagonists for serotonin have been quite useful, but I believe side effects preclude regular usage. Increases in vagal tone may also have a role. Improvement in overall circulation seems to be of greatest help.

6. *Is it ok just to drink salt water? Or do you need plain water as well?*
   Man cannot subsist on salt water, which is hypertonic. Increased salt of a lesser degree can be helpful in retaining fluid and increasing blood volume. Recently Jordan and others at Vanderbilt have shown a beneficial effect of drinking 16 oz. of plain water.

7. *What is your opinion regarding a stomach stapling in a POTS patient?'*
   I have no opinion.

8. *What are the long-term effects of repetitive inadequate/insufficient sleep in relation to NCS and POTS. Could sleep problems cause Dysautonomia?*
   Past literature (I forget the author) failed to show a relationship between sleep and orthostatic intolerance. I believe studies looked particularly at NCS.

"Pooling" remains largely unexplained. It does not to seem to result from changes in veins or their ability to contract but may instead relate to artery properties.

9. *I bruise easily, is it because of these leaky capillaries?*
   Perhaps, but this may be a common finding in types of Ehlers-Danlos syndrome. It may also relate to endothelial cell activation.

10. *What medications are to be avoided when you have POTS as well as NCS.*
    Most medications that lower the blood pressure are to be avoided. This includes nitrates, hydralazine, ethanol, diuretics. Also medications that increase heart rate are often poorly tolerated. Recent data suggest that beta blockers are harmful in POTS and of equal efficacy as placebo in NCS.

Research Briefs

The following research abstracts were taken from PubMed. Please do NOT contact NDRF for the full articles; instead, go to the PubMed website for more information. As always, talk with your physician about studies you think may have particular relevance to your situation.

"Cerebrospinal fluid levels of catechols in patients with neurogenic orthostatic hypotension." Clin Sci (Lond) 2003 Jan 22; [epub ahead of print]. Holmes C, Patronas N, Kopin IJ.

"http://www.ncbi.nlm.nih.gov/entrez/query/egifs/-PMGifs-Toolbar-button_cs.gif"

Background.
In multiple system atrophy (MSA) and pure autonomic failure (PAF), orthostatic hypotension (OH) results from deficient noradrenaline release from sympathetic nerves during standing. Postmortem findings have indicated loss of central noradrenergic cells in both diseases. This study sought in vivo neurochemical evidence for central noradrenergic deficiency in MSA+OH and PAF.

Methods.
Twenty-eight patients with OH (18 with MSA, 10 with PAF), had cerebrospinal fluid and blood sampled for levels of noradrenaline and its neuronal metabolite dihydroxyphenylglycol. Fifty-one control subjects included 10 elderly normal volunteers, 10 patients with Alzheimer's disease, 16 with dysautonomia (postural tachycardia syndrome or neurocardiogenic syncope), and 6 with MSA lacking OH.

Results.
Patients with OH had lower cerebrospinal fluid concentrations of noradrenaline (0.53 +/- 0.07 nmol/L) and dihydroxyphenylglycol (6.52 +/- 0.46 nmol/L) than did control subjects (0.90 +/- 0.09 and 9.64 +/- 0.46 nmol/L, p=0.0001). The MSA+OH group had higher plasma levels of both catechols (1.31 +/- 0.16 and 5.08 +/- 0.43 nmol/L) than did the PAF group (0.38 +/- 0.08 and 2.53 +/- 0.30 nmol/L, p<0.001), despite similarly low cerebrospinal fluid values. In MSA patients, those with OH had lower cerebrospinal fluid levels of noradrenaline and dihydroxyphenylglycol than did those without OH (1.71 +/- 0.64 and 10.41 +/- 1.77 nmol/L, p=0.006).

"The findings are consistent with central noradrenergic deficiency in MSA + OH and PAF.."

Conclusions.
The findings are consistent with central noradrenergic deficiency in MSA+OH and PAF. In MSA, central noradrenergic deficiency seems to relate specifically to OH.

PMID: 12540289 [PubMed - as supplied by publisher]

"Autonomic nervous system testing may not distinguish multiple system atrophy from Parkinson's disease." J Neurol Neurosurg Psychiatry 2003 Jan;74(1):56-60. Riley DE, Chelimsky TC. Department of Neurology, University Hospitals of Cleveland and Case Western Reserve University .

Background.
Formal laboratory testing of autonomic function is reported to distinguish between patients with Parkinson's disease and those with multiple system atrophy (MSA), but such studies segregate patients according to clinical criteria that select those with autonomic dysfunction for the MSA category.

Objective.
To characterize the profiles of autonomic disturbances in patients in whom the diagnosis of Parkinson's disease or MSA used criteria other than autonomic dysfunction.

Methods.
47 patients with parkinsonism and autonomic symptoms

("MSA and Parkinson's" Continued on page 8)

who had undergone autonomic laboratory testing were

Research, continued

MSA and Parkinson's (*continued from p. 6)*

(Continued from page 8)
identified and their case records reviewed for non-autonomic features. They were classified clinically into three diagnostic groups: Parkinson's disease (19), MSA (14), and uncertain (14). The performance of the patients with Parkinson's disease was compared with that of the MSA patients on five autonomic tests: RR variation on deep breathing, heart rate changes with the Valsalva maneuvre, tilt table testing, the sudomotor axon reflex test, and thermoregulatory sweat testing.

Results. None of the tests distinguished one group from the other with any statistical significance, alone or in combination. Parkinson's disease and MSA patients showed similar patterns of autonomic dysfunction on formal testing of cardiac sympathetic and parasympathetic, vasomotor, and central and peripheral sudomotor functions.

"Current clinical criteria for Parkinson's disease and MSA that direct patients with Dysautonomia into the MSA group may be inappropriate."

Conclusions.
This study supports the clinical observation that Parkinson's disease is often indistinguishable from MSA when it involves the autonomic nervous system. The clinical combination of parkinsonism and Dysautonomia is as likely to be caused by Parkinson's disease as by MSA. Current clinical criteria for Parkinson's disease and MSA that direct patients with Dysautonomia into the MSA group may be inappropriate.
PMID: 12486267 [PubMed - in process]

"Dysautonomias: clinical disorders of the autonomic nervous system." Ann Intern Med 2002 Nov 5;137(9):753-63. Goldstein DS, Robertson D, Esler M, Straus SE, Eisenhofer G. Clinical Neurocardiology Section, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Building 10, Room 6N252, 10 Center Drive MSC-1620, Bethesda, MD 20892-1620, USA.

The term Dysautonomia refers to a change in autonomic nervous system function that adversely affects health. The changes range from transient, occasional episodes of neurally mediated hypotension to progressive neurodegenerative diseases; from disorders in which altered autonomic function plays a primary pathophysiologic role to disorders in which it worsens an independent pathologic state; and from mechanistically straightforward to mysterious and controversial entities. In chronic autonomic failure (pure autonomic failure, multiple system atrophy, or autonomic failure in Parkinson disease), orthostatic hypotension reflects sympathetic neurocirculatory failure from sympathetic denervation or deranged reflexive regulation of sympathetic outflows. Chronic orthostatic intolerance associated with postural tachycardia can arise from cardiac sympathetic activation after "patchy" autonomic impairment or blood volume depletion or, as highlighted in this discussion, from a primary abnormality that augments delivery of the sympathetic neurotransmitter norepinephrine to its receptors in the heart. Increased sympathetic nerve traffic to the heart and kidneys seems to occur as essential hypertension develops. Acute panic can evoke coronary spasm that is associated with sympathoneural and adrenomedullary excitation. In congestive heart failure, compensatory cardiac sympathetic activation may chronically worsen myocardial function, which rationalizes treatment with beta-adrenoceptor blockers.

"...changes range from transient….to progressive neurodegenerative diseases...."

("Dysautonomias, continued on p. 10)

Research, continued

Dysautonomias (continued from page 7)

(Continued from page 9)

A high frequency of positive results on tilt-table testing has confirmed an association between the chronic fatigue syndrome and orthostatic intolerance; however, treatment with the salt-retaining steroid fludrocortisone, which is usually beneficial in primary chronic autonomic failure, does not seem to be beneficial in the chronic fatigue syndrome. Dysautonomias are an important subject in clinical neurocardiology.

PMID: 12416949 [PubMed - indexed for MEDLINE]

"Clinical and physiological effects of an acute alpha-1 adrenergic agonist and a beta-1 adrenergic antagonist in chronic orthostatic intolerance." Circulation 2002 Dec 3;106 (23):2946-54. Stewart JM, Munoz J, Weldon A. Department of Pediatrics, New York Medical College, Valhalla, NY 10595, USA. stewart@nymc.edu

Background.
Adrenergic agents are commonly used in the treatment of chronic orthostatic intolerance with postural tachycardia syndrome (POTS). POTS may be associated with increased limb blood flow ("high flow") and defective orthostatic vasoconstriction or decreased limb blood flow ("low flow") and potentially with small blood volume.

Methods and Results.
We investigated the consequences of short-term intravenous administration of an alpha-1 adrenergic agonist, phenylephrine, and a beta-1 adrenergic antagonist, esmolol, in 14 patients with POTS aged 13 to 19 years. Indices of heart rate and blood pressure variability, peripheral blood flow, and arterial resistance were assessed, and the capacitance relation was computed for every subject using venous occlusion plethysmography. Patients were tilted to 35 degrees upright while medicated and while unmedicated, and measurements were repeated.

Phenylephrine improved orthostatic tolerance and normalized hemodynamics and indices of heart rate/blood pressure variability while supine and while upright, producing significant peripheral vasoconstriction and venoconstriction (20% capacitance change). Esmolol did not improve orthostatic tolerance or hemodynamics. A subgroup of low-flow POTS patients had exaggerated venoconstriction to phenylephrine (50% capacitance change) but others had no response.

Conclusions.
Phenylephrine, but not esmolol, improves orthostatic tolerance and hemodynamics in POTS. This lends support to the use of oral alpha-1 agonists in the treatment of patients with chronic orthostatic intolerance.

PMID: 12460877
[PubMed - indexed for MEDLINE]

"The findings are consistent with central noradrenergic deficiency in MSA + OH and PAF.."

Research, continued

"Midodrine in neurally mediated syncope: a double-blind, randomized, crossover study." Ann Neurol 2002 Sep;52(3):342-5. Kaufmann H, Saadia D, Voustianiouk A. Department of Neurology, Mount Sinai School of Medicine, New York, NY 10029, USA. horacio.kaufmann@mssm.edu

Neurally mediated syncope is the most frequent cause of syncope in patients without structural heart disease. Its most common trigger is a reduction in venous return to the heart due to excessive venous pooling in the legs. We conducted a double-blind, randomized, crossover trial to investigate the efficacy of midodrine, a selective alpha-1 adrenergic agonist that decreases venous capacitance, in preventing neurally mediated syncope triggered by passive head-up tilt.

Twelve patients with history of recurrent neurally mediated syncope, which was reproduced during head-up tilt, were randomized to receive a nonpressor dose of midodrine (5mg) or placebo on day 1 and the opposite on day 3. One hour after drug or placebo administration, patients underwent 60-degree head-up tilt lasting 40 minutes (unless hypotension or bradycardia developed first).

In the supine position, midodrine produced no significant change in blood pressure or heart rate. The responses to head-up tilt were significantly different on the midodrine and the placebo day: on the placebo day, 67% (8/12) of the subjects suffered neurally mediated syncope, whereas only 17% (2/12) of the subjects developed neurally mediated syncope on the midodrine day ($p < 0.02$).

These results indicate that midodrine significantly improves orthostatic tolerance during head-up tilt in patients with recurrent neurally mediated syncope.

PMID: 12205647 [PubMed - indexed for MEDLINE]

"...midodrine significantly improves orthostatic tolerance…"

When we honestly ask ourselves which person in our lives mean the most to us, we often find that it is those who, instead of giving advice, solutions, or cures, have chosen rather to share our pain and touch our wounds with a warm and tender hand. The friend who can be silent with us in a moment of despair or confusion, who can stay with us in an hour of grief and bereavement, who can tolerate not knowing, not curing, not healing and face with us the reality of our powerlessness, that is a friend who cares.

~ Henri Nouwen, Out of Solitude

Coping Strategies

Maintaining Self-Esteem

Many of the sources of your self-esteem can be threatened by your role and identity within your family or in your work life when you have Dysautonomia. Don't underestimate your courage or what you are learning because you live with this illness. Take pride in the way you are coping with your illness. Gain a new appreciation for how much you are loved—not because of what you do but because of who you are.

Your illness does not suddenly define you as a Dysautonomia patient, as if that is your new identity. Individuals need to manage the Dysautonomia and respect the limitations it brings, but also "keep it in its place." It is important not to become the illness, but to stay who you are as a balanced individual with many talents, interests, and qualities that only you possess. By focusing on the positive qualities of who you are, you will feel differently. Your overall emotional well-being will be enhanced when you discover or develop new sources for positive self-regard.

"...respect the limitations it brings, but also "keep it in its place."

Finding a Positive Meaning

While living with Dysautonomia can feel like an awful experience, it can be an opportunity for positive change. For many, illness inspires individuals to pay more attention to what matters most. This could mean spending more time with family and close friends, showing more appreciation for all that you have and are, bringing forth aspects of your personality that have been suppressed, taking better care of your physical and emotional needs and seeking new hobbies and creative endeavors. Your illness can become an impetus for positive change. This is often called the ``enlightenment" or ``gift" that comes with illness, or the ``wake-up call". Individuals who embrace this aspect of their illness' experience have been found to be especially well adjusted and better able to deal with the many trials and disruptions caused by their illness.

"For many, illness inspires individuals to pay more attention to what matters most."

Andrew Kneier, PhD, Ernest Rosenbaum, MD, and Isadora R. Rosenbaum, MA .

Tips for Parents

Attitude

Try to develop an attitude where you think of your children as kids first, with the same mental and emotional needs as all other children. They are much more like other children than they are different. As part of this attitude, I've found it so important not to coddle them. Their everyday life should be, as much as possible, filled with opportunities to learn and experience life, with the goal of growing up just as other children expect to. These kids will be often called upon to have great courage – about dealing with frightening tests and procedures, about enduring pain, and about handling the stares and ignorant behavior of others. Because of this, they need to learn self-discipline and be challenged by our high expectations, which will help them manage their own fears and hurts. I don't mean that adopting a "stiff upper lip" attitude is useful--they need to have our support but also our confidence that they have what it takes to manage the difficulties they face. Humor helps. When the body just won't work right or when it makes a mess, sometimes the best thing to do is have a little running joke about it. Try to avoid that "oh, you poor little thing" attitude at all costs!

"...think of your children as kids first…"

~ Carol Balliet, Band-Aides and Blackboards

Things to Remember...

- ✯ Recognize that life is more than the chronic illness
- ✯ Recognize the positive aspects of your child and his or her contribution to your family.
- ✯ Accept your kids even if they cannot do what you dreamed they would
- ✯ You can't eliminate your loved one's condition, but you can understand the illness and your options, and then take steps to adapt along the way.
- ✯ This is not your fault or your child's fault.
- ✯ Cherish the good times.
- ✯ Modify activities. Instead of going to a busy amusement park, go to a park to feed the ducks, soak up the sunshine, and get fresh air.
- ✯ Focus on the fun of the day rather than on what you may be missing out on.
- ✯ Watch what you say to or about your children. One child writes: "Here are some things that used to made me feel so guilty for being sick and needing to go the doctor. My parents would say, and I certainly will never forget: · 'If I had a nickel for every time I took you to the doctor I'd be a rich person.'…..'Doctors, doctors, doctors, that is all I ever do.'….'She is always convalescing.'…. No wonder I feel awful about being sick so much." If your children are sick a lot, please don't take it out on them."

Parent Tips continued

Free Medical Resources for Kids

The Medicine Program.com tells you how to apply for free prescription medicines by completing a form. Here are the rules: Your child must not have insurance coverage for outpatient prescription drugs; you must not qualify for a government program which provides for prescription medication like Medicaid; and your income must be at a level causing hardship when you need to purchase the medicine from your pharmacy at retail price.

RXHope.com is another website providing free prescription drugs...supported by the PhRMA (Pharamceutical and Research Manufacturers of America) and participating pharmaceutical companies. Your doctor or nurse practitioner can use this website in order to obtain the prescription drugs.

RXAssist.org is like RXHope.com in that it is used by health professionals who are applying for free prescription drugs on behalf of qualified, uninsured patients. There is a downloadable form that your doctor or nurse practitioner can fill out on your behalf.

PHRMA.org lists specific pharmaceutical companies which participate in free prescription drug programs.

Needy Meds.com is another website worth checking out. It is extremely easy to navigate and provides a list of drug companies participating in give-away programs as well as other useful information

Chronic Illness and Education

At school two federal laws may be relevant. Section 504 of the Rehabilitation Act of 1973 states:

"No otherwise qualified individual with handicaps in the United States… shall solely by reason of her or his handicap, be excluded from the participation in, be denied the benefits of, or be subjected to discrimination under any program or activity receiving federal financial assistance…" In other words, the law prohibits discrimination on the basis of disability in any program or activity receiving federal funds. Since public schools all receive federal funding, the law ensures that students with disabilities receive equal access to a public education.

Under the Individuals with Disabilities Education Act (IDEA), children with disabilities and their families have legal rights to early intervention services for their children with special needs birth to three and special education programs for children ages three through 21. For more information about IDEA, you might visit the Office of Special Education Programs (OSEP) within the US Department of Education at www.ed.gov.offices/OSERS/OSEP or www.ideapractices.org.

The major purposes of the IDEA are:

(a) to ensure that all children with disabilities have available to them a "free appropriate public education" that emphasizes special education and related services designed to meet their unique needs and prepare them for employment and independent living;
(b) to ensure that the rights of children and youth with disabilities and their parents are protected;
(c) to assist States, localities, educational service agencies, and Federal agencies to provide for the education of all children with disabilities; and
(d) to assess and ensure the effectiveness of efforts to educate children with disabilities.

For those who may need a distance program for a portion or all of their child's education, the University of Missouri offers excellent accredited distance courses for a variety of ages through grade 12. With cooperation with local schools, these classes can serve in place of local classes, too, for families who want to maintain local educational ties. Go to: http://cdis.missouri.edu/MUHighSchool/HShome.htm to take a look at the available high school classes.

Working with Your Doc

Prevent Medical Errors

A 1999 report, ***To Err is Human,*** estimated that between 44,000 and 90,000 people each year die as a results of medical errors that occur in hospitals alone, making medical errors one of the country's leading causes of death and injury.

Of course, not all bad outcomes are the result of error – injuries and illness are sometimes too serious to overcome, complications sometimes occur, and no treatment is 100% effective all of the time. Some errors involve medications, but misdiagnosis, incorrect treatment, inaccurate or misread lab tests, etc., are also possible sources of errors. And not all medical errors harm patients – for instance, a pharmacist might misread your physician's handwriting and give you an incorrect medicine; however, if you are vigilant you might catch that the medicine is incorrect.

Dysautonomias are frequently missed as a diagnosis, discovering the "best" treatment and management strategy is sometimes lengthy and trying, and the disorders are often chronic. Given this, it becomes very important to understand what we can do to help prevent errors in our care. This article presents information on actions to take, and is taken from the web site of the federal Agency for Healthcare Research and Quality.

1. The most important thing is to be an active member of your health care team. Take part in the decisions about your care. Research shows that patients who are more involved in their care tend to get better results.

"Make sure your doctors know about everything you are taking…"

To help prevent medicine errors:

2. Make sure your doctors know about everything you are taking, including over the counter medicines, vitamins, and herbs. That way, you can talk about whether there are any potential concerns about taking them together, and can help keep your records up to date.

3. Make sure to tell your doctor about any allergies or adverse reactions you have to medicines. This is important to help avoid getting a medicine that could be harmful in your particular case.
4. Make sure you can read the prescriptions your doctor writes: if you can't, your pharmacist may not be able to either.

5. Ask for information about your medicine so that you understand its role in your care. What is it for? How long are you supposed to take it? What side effects are likely? Is it safe to take with your other medicines and supplements? What food, drink, or activities, if any, should you avoid while taking this medicine?

"If you have any questions at all about your medicine, ask."

6. When the pharmacist gives you the medicine, check to see if it is the medicine your doctor ordered, in the right dose. A study by the Massachusetts College of Pharmacy and Allied Health Sciences found that 88% of medicine errors involved the wrong drug or the wrong dose.

7. If you have any questions at all about your medicine, ask. Make sure you understand the directions for taking it.

8. Ask your pharmacist for the best device to measure your liquid medicine, and how to use it. Many people do not understand the right way to measure liquid medicines. Special devices, like marked syringes, are accurate. Avoid using things like household teaspoons, as they often do not hold a true teaspoon.

9. Ask for written information about the side effects your medicine could cause. Many pharmacists automatically provide this in writing; if they don't, ask for it- it can help you recognize problem side effects so you

("Assistance," Continued on page 16)

Errors *(continued from page 12)*

can reports problems right away.

To help prevent hospital errors:

10. If you have a choice, stay at a hospital where many patients have the procedure or treatment you need. Patients tend to have better results when they are treated at hospitals that have a great deal of experience in their condition.

11. If you are in a hospital, consider asking all health care workers who have direct contact with you whether they have washed their hands. This is an important way to prevent the spread of infections in hospitals, but it is not done regularly or thoroughly enough.

12. When you are being discharged from the hospital, ask your doctor to explain the treatment plan you need to use at home. Learn about the medicines you need to continue taking, and find out when you can get back to your regular activities.

To help prevent surgery errors:

"You have a right to question anyone who is involved in your care."

13. If you are having surgery, make sure that you, your doctor and your surgeon all agree and are clear on exactly what will be done. Doing surgery at the wrong site is rare, but occasionally happens.

Other steps to help:

14. Speak up if you have questions or concerns. You have a right to question anyone who is involved in your care.

15. Make sure someone, such as your personal doctor, is in charge of your care. This is especially important if your have many health problems or are in a hospital.

16. Make sure that all health professionals involved in your care have important health information about you. Do not assume that everyone knows everything they need to know.

"Ask your doctor if your treatment plan is based on the latest evidence."

17. Ask a family member or friend to be there with you and to be your advocate (someone who can help get things done and speak up for you if you can't.)

18. Know that "more" is not always better. It is a good idea to find out why a particular test or treatment is needed and how it can help you. You could be better off without it.

19. If you have a test, don't' assume that no news is good news. Ask about your results.

20. Learn about your condition and treatments by asking your doctor and nurse and by using other reliable sources. Treatment recommendations, for instance, are available form the National Guidelines Clearinghouse at www.guideline.gov. Ask your doctor if your treatment is based on the latest evidence.

To learn more about prevention of medical errors, visit the Agency for Healthcare Research and Quality Web site at http://www.ahrq.org/qual/errorsix.htm

Show your thanks to your local physician

We've all heard the war stories. People who spent years without a correct diagnosis or treatment. Even with a correct diagnosis, difficulties finding physicians who are knowledgeable (or willing to learn) and able to take on a dysautonomia patient on a regular, or even occasional, basis, seems all too common.

But mixed in amongst the war stories, we know there are also stories of physicians who have gone out of their way to help patients. The names of those who participated in the NDRF conference last summer, for instance, roll pretty easily off many people's tongues, and we are certainly grateful for the leadership, compassion, and dedication they show.

While the NDRF has always, and will continue, to recognize the efforts of these out-front leaders, the organization would also like to recognize those physicians who are in the trenches in our local clinics—the GPs, internists, and others, who on a day-to-day basis can help or hinder, support or set up barriers, to the on-going care of people with dysautonomias. Their assistance in developing personalized treatment plans, communicating with schools and employers, and sometimes just providing an empathic shoulder is essential if people with dysautonomias are to function as fully as possible.

If you would like to recognize your local physician or physicians for the support they have provided to you or a loved one, please e-mail Karen Harrington, the NDRF Chat Newsletter Editor, at:

KLHarr2000@aol.com.

Every nominee will receive a Certificate of Appreciation from the NDRF, as well as a letter thanking them for their efforts and the role they are playing in serving the needs of this community of people. In your e-mail, please provide a 1 or 2 paragraph summary of why you would like your physician recognized, so we may personalize their certificate.

Each nominee will receive a certificate of appreciation from the NDRF

Medical Resources for Kids

Families with children who have special health care needs may be eligible for financial support to help pay for medical treatment and other related services. For more information about family support services, you might visit Family Voices at **www.familyvoices.org**

The Healthy and Ready to Work initiative promotes a comprehensive system of family-centered, culturally competent, community based care for children with special health care needs who are approaching adulthood and may need assistance in making the transition from pediatric to adult health care and to postsecondary education and/or employment.
http://www.mchbhrtw.org/

Other resources include : Association for the Care of Children's Health, 3615 Wisconsin Avenue NW, Washington, DC 20016 (202) 244-1801

NDRF Actions

NDRF Focuses Efforts for 2003

Since its inception, the NDRF has always placed the needs of the Dysautonomia community at the forefront. Physician education, support group development, development and distribution of educational materials, conferences, and fundraising for research have all been priorities.

Several milestones have been reached. A network of support groups is in place. Key high-quality reference materials are now available, providing a basic foundation from which patients can discuss Dysautonomias with their physicians. Two very successful conferences have been held.

Two overarching concerns that continue to plague the community are the lack of funding for research and the lack of available, knowledgeable physicians. During the upcoming year, the NDRF leadership feels its efforts will have the most "bang for the buck" if it focuses attention more fully on these two concerns.

"Two overarching concerns that continue to plague the community are the lack of funding for research and the lack of available, knowledgeable physicians."

To that end, it will be intensifying its efforts in Washington, DC to reach legislators with the message that Dysautonomia research must be funded. In particular, the organization will be seeking funding for Centers of Excellence. NDRF also will be lobbying for recognition of Dysautonomias in rules for Social Security Disability. Finally, the organization will enhance its efforts to reach and teach physicians.

NDRF will Inform Physicians at Conferences

The NDRF is working closely with the Department of Health and Human Services to improve awareness within the medical community on disorders of the autonomic nervous system. Each year the Department of Health publishes an extensive report on hundreds of illnesses, this publication is distributed throughout the United States to thousands of physicians. Dr. Wanda Jones - Deputy Director for Women's Health at the Department, has assured the Foundation that Dysautonomias will now be added to this publication.

In addition, the NDRF will focus on the necessity to distribute literature at major medical conventions. By distributing information at these events, the medical community will increase their knowledge on these devastating disorders, minimizing or eliminating misdiagnosis and/or mismanagement – a major concern for patients suffering from Dysautonomias.

This increased knowledge will give patients a greater chance of finding a qualified and informed physician, increasing the patients chances to enjoy a greater "Quality of Life".

Resources

The Fainting Phenomenon: Understanding Why People Faint and What can be Done about It.. Dr. Blair Grubb, MD and Mary Carole McMann, MPH. This layperson's guide to fainting and the disorders that cause it is an excellent reference for anyone who wishes to learn more about fainting problems, OI, and Chronic Fatigue. It contains information on how the nervous system and cardiovascular system operate and influence orthostatic intolerance and hypotension, neurogenic syncope, POTS and CFS, and fainting. It describes basic diagnosis and treatments available. An extensive glossary of terms is included. Chapters include: The Normal Nervous System; The Normal Cardiovascular System; OI and Orthostatic (Postural) Hypotension; Neurogenic Syncope; POTS and CFS; Fainting in Children, Adolescents, and Older People, Diagnosing Fainting, and Treating Fainting

2000 NDRF Conference Video Set.

This three videotape set of the 2000 conference on OI and syncope include presentations by a host of speakers, including Drs.: D. Robertson, D. Goldstein, P. Low, B. Grubb, R. Mosqueda-Garcia, D. Streeten, J. Stewart, and C. Mathias. The presentations were geared to an audience of patients and family members impacted with Dysautonomia. Topics included the autonomic nervous system, OI, fainting/syncope, autonomic conditions in children and adolescents, chronic fatigue, and other autonomic disorders.

ORDER INFORMATION

NDRF Patient Handbook *(NEW!)*	$27.95
The Fainting Phenomenon by Dr. Grubb	$17.95
2000 NDRF Conference Video Set:	$24.95
2 Book Set (Handbook and Dr. Grubb):	$43.95
Video and Patient Handbook:	$49.95
Video and Dr. Grubb's book:	$37.95
Video, Handbook, & Dr. Grubb's book:	$65.95
NDRF Chat Newsletter	$3.50

The new NDRF patient handbook now available:

The NDRF Handbook for Patients with Dysautonomias. David S. Goldstein, M.D. and Linda Joy Smith.

Sponsored by MEDTRONIC FOUNDATION, this 312 page hardcover handbook is written to give patients and their families a better understanding of the autonomic nervous system in both healthy and disease states. Mechanisms of diagnosis and management strategies are described, and readers are given information on caregiving, social security disability, children with Dysautonomia, support groups, and day-to-day management techniques.

If you would like to obtain a copy of the ***NDRF Patient Handbook on Dysautonomias,*** give us a call at 651-267-0525 or use the order information below.

Please note that electronic copies of the Handbook will be available to download for free from the NDRF website, as an alternative to purchasing a hard-copy of the book.

** Shipping: Prices include mass media shipping charges within the U.S. Please allow 4-6 weeks for delivery by mass media mail. **For first class shipping, please add $5.00.**

Foreign orders: An additional charge of $7.00 will be added to orders sent to Canada or Mexico, or an additional $12.00 to other countries outside the U.S. Please allow 6-8 weeks for delivery.

Order through www.ndrf.org on the web or send payments in US dollars, or credit card information, to:

NDRF, 1407 W. 4th St., Suite 160, Red Wing, MN 55066.

Contributions

The success of the NDRF hinges on the combined efforts and resources of its members. We would like to take this opportunity to recognize everyone who has contributed to the organization's mission over the past year! Every effort and donation, large or small, is greatly appreciated!

Karen would like to personally thank those who contributed articles and information for this newsletter, including Drs. Low and Stewart, Linda and Dan Smith, and Colleáyn Klaibourne.

Dan and Linda would like to give special recognize to the NDRF's corporate sponsors (Medtronic, Guidant and Shire), and to several individuals who went above and beyond the call of duty to raise awareness and funds for research. They include:

Bob & Judy Miller
MSA, Walk for Hope

Jane & Jack Mandula
Grape Harvest Wine Fesival

Church of the Bredthen
Fundraiser for Dysautonomia Members

Lynn Adams
Opelika Lion's Club

Kevin Wilkeson

Volunteers

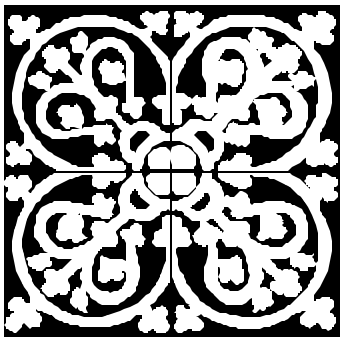
The NDRF would like to introduce a couple of wonderful volunteers for their contribution in helping those of us suffering from autonomic disorders. NDRF is honored to have the following individuals assisting:

⇒ Georgia K. Bergstrom, Conference Director

⇒ Karen Harrington, Newsletter Editor

With their combined efforts, the NDRF can continue in our missions to provide our readers with the most accurate and timely information available on Dysautonomias.

If you have ideas or suggestions that could help promote awareness or improve patient education on Dysautonomias, please let us know. Thank you!



New Giving Opportunity:

The NDRF Chat newsletter is one of our best ways to communicate with our members, and we would like to publish it at least semi-annually. However, increasing costs make the newsletter a rather expensive proposition. Because the NDRF charges no membership fees, generating the funds to cover the production and distribution costs of the newsletter is a challenge. If you would like to become a NDRF Chat sponsor and help defray the production and distribution costs, please call us at 651– 267-0525.

NDRF Support

NDRF relies upon your contributions to help us continue providing programs for education, support, and advocacy, and to continue serving the community of people impacted with disorders of the autonomic nervous system.

NDRF was able to achieve some important milestones in 2002:

√ Upgrades to the NDRF Web Site and continued implementation of the NDRF Discussion forum

√ Advocacy visits to congressional leaders in Washington, D.C.

√ Planning and hosting the 2002 conference

√ Moving into new office space, which will allow the NDRF staff and volunteers to better service you, our members

√ Publication of a new Patient Handbook, co-authored by Dr. Goldstein of the National Institutes of Health

√ Speaking at major medical conferences, including symposiums held at the Cleveland Clinic and the National Institutes of Health.

√ Working with the United States Department of Health and Human Services to increase awareness within medical teaching institutions.

The community of people impacted with Dysautonomia exceeds one million. Your contributions to NDRF will help us to continue and expand our mission. Won't you take a minute to lend your support for the coming year?

Yes, I want to help in the efforts to conquer Dysautonomia. My contribution will help NDRF continue with its mission for education, support, and advocacy:

Giving Levels

☐ Contributors $ 30.00

☐ Supporters $ 50.00

☐ Patrons $100.00

☐ Benefactors $500.00

☐ Sponsors $1000.00

☐ Other $_____

☐ *I would like NDRF to send a receipt for my contribution.*

Name: _____

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Donation Method: ___Check ___ Mastercard ___ Visa

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Suite 160
Red Wing, MN 55066-2108

Phone: 651-267-0525
Fax: 651-267-0524
Email: ndrf@ndrf.org

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