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Fall-Winter 2001

A PUBLICATION FOR, BY, AND ABOUT PEOPLE WITH EHLERS-DANLOS SYNDROME.

EDS Today

Dear Readers,

2001 has been a frantically busy and incredible journey for EDS Today. We have soared into issue after issue, only to find more information about EDS waiting for us to share with the world.

Our lead article, "The Prevalence of Non-Surgical Complications in the Ehlers-Danlos Syndrome" really is an excellent and interesting article. I would like to thank Dr. Pamela-Popken-Harris for granting reprint permission. We encourage you to share this with your physician and other medical staffers. Awareness is the key and every little bit of information we can retrieve on EDS saves not only our lives but also opens the doors for those we love that are afflicted

Speaking of awareness, our Editor, Barb, is heading up the EDS Awareness Campaign that will kick off in February of 2002. If you have not signed-up to help with this campaign, may I remind you that it is not too late to get involved. Visit http://members.home.com/uggenb/ or write to EDS Today, PO Box 88814, Seattle, WA 98138-2814. Every organization involved is working one way or another to create awareness, now we are all pulling together our creativity to insure that EDS will have a month and a day that it is known for. Pulling together is all we have to let the world know how real this syndrome is.

As you can see, this issue is once again packed with informative and useful information. EDS TODAY is in flight and it is only because of you, our readers, that we can fly beyond our expectations. Thank you for your dedication and remember, - "STAY WELL, STAY WARM and always SMILE!"

— Christine Phillips, Publisher

Inside this issue:

Featured Topic:
EDS and Job Hunting

Letter from the Editor

It's been a busy summer and early fall for all of us at EDS Today. We've been working hard to spread the word about EDS and EDS Today and to bring you the most information possible.

First, we've filed our 501(c)3 application. It was a lengthy project, but well worth the effort. I hope to have good news to report about our non-profit status by next issue.

Second, we've acquired a domain name for our website. Our new home on the web is:

http://www.EDSToday.org/

Third, we've been working with several other groups to start an EDS Awareness Campaign. For more details about the campaign, please see the announcements on page 16.

Finally, with an initial donation from Stephen and Robin Polischeck, we have established a research fund. Over time, we hope to watch the fund grow and begin distributing the funds to various EDS research projects. Thank you so much Stephen and Robin! Your contributions are a wonderful first step towards advancing our knowledge of EDS. If you would like to contribute to the research fund, please send a check payable to EDS Today to:

EDS Research Fund

C/O EDS Today

PO Box 88814

Seattle, WA 98138-2814

— Barbara J. Uggen-Davis, Editor

APOLOGIES

Our sincere apologies to Jill Douglas-Hand for the missing photo in last issue. There was an error at the printing place.

IMPORTANT RENEWAL NOTICE

Check the address label on the back of the newsletter. If the date on your label is 10/15/01 then it's time to renew! Don't wait, send your check and renewal form today or renew online at:

http://www.EDSToday.org/onlineorder.htm

This is your newsletter, so we encourage you to get involved. We are currently seeking articles on Exercise and EDS. Articles may be submitted to EDS Today, PO Box 88814, Seattle, WA 98138-2814 or via e-mail at info@edstoday.org or through our online form on our web-site at:

http://www.EDSToday.org/submissions.htm

This issue is dedicated those affected by the September 11, 2001 terrorist attacks. Our hearts go out to the families of the victims and to the brave heroes in uniform. We will never forget.

Members of the EDS Today staff have donated their time, money and blood to the New York Fire and Police Departments and the Red Cross. We encourage everyone to do whatever they can to help in our nation's time of need. To learn more on ways you can help, please see the last page of the newsletter.

Want to read more about something you saw in EDS Today? Visit our links page on our web-site at:

http://www.EDSToday.org/resource.htm

For every issue of EDS Today, we have links and resources for more information on the web. For this issue, we feature information on the awareness campaign and much more.

The Prevalence of Non-Surgical Complications in the Ehlers-Danlos Syndrome

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[1]Pamela Popken-Harris, Ph.D., [2]Scott Ketover, M.D., and [3]Mary Ella Pierpont, M.D., Ph.D. [1]David F. Hickok Memorial Cancer Research Laboratory, Abbott Northwestern Hospital, and the University of Minnesota Cancer Center, Minneapolis, MN. [2]Minnesota Gastroenterology, Minneapolis, MN. [3] Department of Pediatrics, University of Minnesota, Minneapolis MN.

Address correspondence to: Pamela Popken-Harris, Ph.D., David F. Hickok Memorial Cancer Research Laboratory, Abbott Northwestern Hospital, Minneapolis, MN 55407; telephone (612) 863-4439, fax (612) 863-4936.

Abbrviations used in this Manuscript:

Cerebral Vascular Accident (**CVA**)

Ehlers-Danlos Syndrome (**EDS**)

Gastroesophageal Reflux Disease (**GERD**)

Mitral Valve Prolapse (**MVP**)

Non-Steroidal Anti-Inflammatory Drug (**NSAID**)

1-deamino-8-D-arginine vasopressin (**DDAVP**)

Extracellular Matrix (**ECM**)

Inflammatory Bowel Disease (**IBD**)

Mixed Connective Tissue Disease (**MCTD**)

Systemic Lupus Erythrematosus (**SLE**)

EDS Today wishes to thank Pam Popken-Harris Ph.D for granting reprint permission for this article. To learn more about the author, please see our Memorials and Tributes section on page

Introduction

The Ehlers-Danlos Syndrome (EDS) is a heterogeneous group of inherited connective tissue disorders, which share in common, mutations in the synthesis, secretion, or processing of various collagens. Current estimates place the prevalence of EDS at between 1/5,000 to 1/20,000, but recognition of the syndrome can be difficult, and this difficulty is often compounded by phenotypic overlap between the various types (14). In order to reduce confusion, a new nosology (Table 1), corresponding to known defects in specific types of collagen has been proposed. However, as many of the participants in this study were classified to type according to the previous nomenclature (Table 1), we still refer to this, and place the new designation behind in parenthesis when appropriate.

Although surgical complications can be associated with invasive procedures, particularly in those individuals diagnosed with type IV (vascular type of EDS)(2, 40, 60, 65), the prevalence of non-surgical complications in this syndrome are much less well known. Therefore, we performed a study to determine the prevalence of other types of medical conditions or complications, primarily in individuals diagnosed with EDS types I, II, and III as they comprise approximately 90% of those diagnosed with this syndrome. A detailed questionnaire was mailed out to EDS support groups world-wide to evaluate the types of conditions that may be common to this syndrome including those from the gastrointestinal, musculoskeletal, respiratory, cardiac and genitourinary systems. Questions about the types of medications commonly prescribed in this population were also in-

cluded. Only those results where a diagnosis of EDS confirmed by a physician are presented. Our findings indicate that arthritis, cardiac and skeletal thoracic abnormalities were common as were symptoms related to gastrointestinal system. We were surprised to receive 2 reports of inflammatory bowel disease (IBD), specifically Crohn's disease in our respondents. Our findings are compared to those in the literature.

Patients and Methods

In compliance with institutional review board guidelines and through the cooperative efforts of the Ehlers-Danlos National Foundation (EDNF), Canadian Ehlers-Danlos Association (CEDA), and Ehlers-Danlos Support Group in the United Kingdom, a call for participants to complete a detailed questionnaire to determine the prevalence of cardiac and or skeletal abnormalities, medication use, and medical conditions including the various forms of arthritis, gastrointestinal ulceration, post-operative infections, general infections, respiratory type complications, allergy, asthma, autoimmunity and cancer was issued. Demographic questions as to age, sex, and EDS type were also included. In the period in which we collected data, 140 responses were received, including those from single individuals and sometimes extended families diagnosed with EDS types I-IV, VI, and unknown. We could verify through medical records that 43 of our survey respondents were diagnosed with EDS by a geneticist or rheumatologist, and report only on these. Only those responses to individual questions that were definitely positive were included in the analyses, and any response that was considered questionable was scored as negative. In those instances where an abnormal response may have been indicated, such as a report of a coagulation defect, a recurrent or suspicious infection such as pyelonephritis or endocarditis, an autoimmune disease, inflammatory bowel disease or cancer, written verification in the form of medical records of the abnormal response was requested. Statistical analyses were also performed on some data sets to determine an exact two-tailed p-value (95% confidence level) using Fisher's exact test for unpaired samples with the aid of a computer program Graph Pad Prism, version 2.01 (Graph Pad Software, San Diego CA).

Results

Gastrointestinal Complications

Out of 43 respondents, there were 10 who reported at least one prior gastrointestinal ulceration. Only one of these could be positively associated with Helicobacter pylori infection; however, only the six gastric ulcers (6/10) were tested. Esophageal ulcers, previously associated with non-steroidal anti-inflammatory drug (NSAID) use (6), were the second most common type of ulcer reported (3/10), with only one of the ten reported ulcers (1/10) left as unspecified. In this regard, we also found that 14/43 or 32.6% of the population also indicated the need for one or more types of daily anti-reflux medications (Table 2) including one or more of those agents designed to increase gastric motility as well as those that decrease or neutralize gastric acid production.

Of relevance to the incidence of non-Helicobacter related gastric ulceration in the adult EDS population, and the prevalence of various arthritic signs and symptoms, was the observation that 19/43 or 44.2% of our respondents also indicated the need for the daily use of NSAIDs or aspirin for pain relief (Table 2). Accordingly, approximately half of our respondents (5/10), who indicated a prior gastrointestinal ulceration attributed their ulcer directly to the use of NSAIDs. Possibly in relation to this, two individuals also reported a diagnosis of Crohn's disease.

Medication Use

Table 2 is a summary of the types of medications commonly prescribed to our respondents. The most common type of medications used by our survey respondents were those designed to ameliorate or reduce pain. These included NSAIDs (44.2%), non-NSAID pain relievers such as acetaminophen and tramadol (18.6%), and narcotic type pain relievers

such as codeine or slow release morphine (7.0%). Overall, the next two most common types of daily medication were those from the anti-reflux and the anti-depressant/anti-anxiety/tranquilizers categories with 14/43 or 32.6% of our respondents reporting the use of these two types of agents. It was also evident that for women over the age of 21, that hormone replacement therapy was frequently reported (35.3%), indicating a high level of hysterectomy. Daily allergy and asthma medication were common, while somewhat surprisingly, relatively few individuals (11.6% overall), indicated the need for a daily cardiac type medication. Types of cardiac related medications that were reported as being used on a daily basis included b-blockers (1/43), calcium channel blockers (2/43), and blood thinners (2/43). Additionally, while almost one third of the respondents indicated the occasional need for oral prednisone (13/43 or 30.2%), we found that the actual indication for use was often for the relief of allergic and asthmatic type symptoms (7/43), or joint pain (3/43), as there were only 3 reports of prednisone use for an actual autoimmune disease. Quite a number of respondents (44.2%) also indicated the need for at least one steroid type injection into a joint for pain relief. We also found that while there was only one respondent who reported requiring an osteoporosis modifier, that there were three reports of mesalamine use for the treatment of inflammatory bowel disease. There were two reports of methotrexate use, including one report from a type II female who also reported Crohn's disease, and the other, also from a type II, to treat chronic costochondritis. Finally, 5/43 or 11.6% of our respondents indicated the use of prophylactic antibiotics for a joint replacement.

Cardiac and Skeletal Thoracic Abnormalities

As shown in Table 3, reports of cardiac defects such as mitral valve prolapse either alone or in combination with certain other types of skeletal thoracic abnormalities including scoliosis, straight backs, and/or pectus deformities were also common. Overall, these types of abnormalities were reported by 22/43 or 51.2% of the total respondents with 12/43 or 27.9% reporting scoliosis, 3/43 or 7.0%, a straight back, and 3/43 or 7.0% the presence of kyphosis or lordosis. Cardiac abnormalities reported included mitral valve prolapse (MVP) or regurgitation (9/43 or 20.9%), and less commonly tricuspid valve abnormalities (2/43 or 4.6%). Mild pectus deformities including pectus carinatum and pectus excavatum were also common and reported by 6/43 or 14.0% of our total respondents. We also found from inspection of the data presented in Table 3 that none of these abnormalities appeared to be EDS type specific.

All of the respondents with a cardiac valve defect also indicated the use of prophylactic antibiotics (Table 2). Two of these respondents with a cardiac valve defect also had at least one episode of bacterial endocarditis. One respondent, had both endocarditis and subsequent cerebral embolization associated with a tricuspid valve replacement. The other respondent had recurrent endocarditis associated with the presence of a MVP (twice due to Streptococcus viridans, and once due to Staphylococcus aureus). In the last patient, a final episode of endocarditis was the most serious, as it was also associated with a cerebral vascular accident (CVA). In all of these episodes of infection, the respondent improved with standard antibiotic therapy and recovered.

Coagulation Factor Abnormalities

In agreement with other reports which indicate that mild coagulation factor deficiencies can sometimes occur in EDS (3, 21, 56), we also received two verified reports of a coagulation factor deficiency, including a mild factor VII deficiency, and a mild factor XI deficiency from type unknown and type III individuals respectively.

Arthritis

The most common arthritic signs and symptoms included dislocations occurring in one or more joints (67.4%), bursitis (62.8%) and temporoman-

(Continued on page 30)

EDS and Job Hunting

Our featured topic is EDS and Job Hunting. Next issue will feature articles on EDS and Exercise.

We encourage readers to submit articles, stories, and topic suggestions. Just send them to: EDS Today, PO Box 88814, Seattle, WA 98138-2814 or info@edstoday.org.

The following is part two of "On the Job with Arthritis" from the Mayo Clinic.com (www.mayoclinic.com). Part one was published in EDS Today, Fall-Winter 2000. Since arthritis and EDS have many symptoms in common, these articles may be useful for people with EDS as well.

For copies of part one, please order a back-issue of EDS Today, Fall-Winter 2000. Supplies are limited to stock on hand or visit our links page online at: www.EDSToday.org/resource.htm.

On the Job with Arthritis -

Multiply Your Career Options

http://www.mayohealth.org/home?id=HQ01145

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Despite all that you and your employer do to accommodate for your arthritis, you may have to cut back on the number of hours you work. You might even choose to find a new job or career.

If your job requires heavy physical labor, for example, your doctor may refer you to an occupational or physical therapist in a work rehabilitation center. These people can help you build your strength and determine how much weight you can safely lift. If their evaluations reveal that there is no job you can do with your current employer, there are state and private vocational rehabilitation agencies that can help you prepare for and find a new job.

There also are certified rehabilitation counselors and rehabilitation psychologists specifically trained to address the vocational concerns of individuals with disabilities. These professionals can provide vocational testing that objectively assesses your vocational aptitudes, interests and work personality. The data resulting from such an assessment may provide fresh ideas and a reliable new direction.

Find job openings

When looking for a new job, don't lean heavily on the classified ads.

Competition for the positions listed there often is fierce. Many people turn there first instead of learning more effective techniques for job hunting.

One of the best ways to get a job — often before it's advertised — is to draw on your network of contacts. Three out of four jobs are obtained through personal contacts. Start with family and friends. Let them know you are looking for a job, tell them what kind of position you want and ask if they know of any openings. Ask former employers and coworkers, professionals you work with (physicians, accountants) and even casual acquaintances.

In addition, try these strategies:

Expand your network. Attend a career-planning workshop. Join a civic organization. Join a national association in your chosen field and attend

workshops at its convention. Take a community college course relating to the field you want to enter and meet others with similar career goals.

Call companies that interest you. Ask to speak to one of their employees and schedule an information interview. This meeting may open the door to employment.

Speak to a person in charge of hiring, even if there are no job openings at the moment. You'll make an important contact for the future.

Call recorded messages about job openings. Many companies have such a line or a Web site with links to career opportunities.

Attend job fairs. Get a list of employers who will attend, choose several that capture your interest and research them. If you speak to a recruiter, get that person's business card. Then follow up with a letter and a phone call.

Contact state and federal employment centers. Some local Arthritis Foundation chapters also have a job placement service.

Read the classified ads in the newspaper, on the Internet and in trade journals.

Update your résumé. With job leads in hand, you're ready to update your résumé. On one page, outline your career goal, qualifications, experience and education.

Don't use a generic résumé for all of your job queries. Instead, adapt your résumé to highlight facts pertinent to each company. Make note of your job achievements, documented with facts and figures wherever possible.

Also write a cover letter to introduce your résumé. Address this letter to a person you have identified. Then write a single page — just three paragraphs:

In the first paragraph, mention the job you desire and the reason you're applying. If someone in the company suggested you apply, mention that person's name.

In the second paragraph, explain why you want to work for this particular company. Identify your top qualifications for the job. List the problems you can help the company solve.

Close the letter in the third paragraph by promising to call the employer for a meeting. Also thank the person for considering you.

Complete the application

Your research and résumé revision eventually may lead to another task: Completing a job application.

Before you begin filling in blanks, read all the directions. Have on hand all the information you need: names, addresses and phone numbers for references; schools you have attended and the dates; and a list of your previous jobs. Include this information on the application — even if it's also on your résumé.

If you find a blank that asks for your salary requirements, write "open to discussion." That gives you room to negotiate later. Also remember that employers are not supposed to ask questions about your age, race, religion and family. You may leave such questions blank.

Though employers are not allowed to ask if you have a disability, they may ask if you are able to perform job functions of the specific position you're applying for. Sample question: "Do you have any physical limitations that would hinder your performance?"

Questions like this can put you in an awkward situation. Some career counselors say that you shouldn't disclose that you have arthritis at this early stage in the job hunt. They suggest answering "no" and assuming that the employer will provide the legally required "reasonable accommodations" if this becomes necessary.

Another possible response if you're not sure how your arthritis will affect your job performance: "Will discuss."

Anticipate interview questions

After completing the job application, compile a list of questions that you expect to be asked during an interview. Then compose and rehearse your answers. Approach the interview as a performance — dress for the part and present yourself as someone who knows your lines.

Perhaps the most popular interview question is "Tell me about yourself."

Don't think of this as a pleasant icebreaker. It could make or break your interview.

Law prohibits employers from basing their hiring decisions on age, sex, race, religion, health or a record of non-felony arrests. But in describing yourself you might accidentally reveal such information — including information about your arthritis.

Instead, plan what you'll say. Briefly summarize assets you bring to the job. If your arthritis is obvious, mention it briefly. But don't shift the focus to your limitations. Talk about adjustments that allow you to stay productive.

For instance, you could say: "Since walking isn't my strong point, I've learned to organize and plan carefully to save steps." Or a broader approach: "I know you're legally prohibited from asking about my arthritis, except for questions about how I would do specific tasks required on the job. But I'd be happy to offer relevant information, because I'm certain I can do the work."

Discussing accommodations

If your arthritis will be obvious to the interviewer, consider dropping a hint about your needs over the phone before your interview. Do this only after you schedule the interview — and only if you're speaking to the person who will interview you. Possible hint: "I sometimes have trouble with stairs. Do you have an elevator?"

If your arthritis is not obvious but will require job accommodations, you face a dilemma. Should you say nothing until you have a job offer? That means your disability won't rule you out. Even so, the employer might feel misled, and this could create hard feelings. One approach is to describe the minor accommodations that you will require immediately after the job offer is made. You can add you did not want to be inappropriately screened out by mentioning this at the outset.

Another approach is to tell the employer about your arthritis, especially if you know you'll need some accommodation. Employers aren't obligated to provide accommodations until you tell them that you have a disability. If you choose this option, emphasize that the accommodations usually are inexpensive and will boost your productivity.

Follow up and find out more

After the interview, send a short letter of appreciation. Thank the interviewer for talking with you. Then mention something about the interview to refresh the person's memory. Repeat your interest in the job and express your desire to work for the company.

With thoughtful preparation at each step of career planning and job hunting, you have every right to be hopeful about your future in the workplace.

Need help with finding a job? Check out Just One Break: www.justonebreak.com. Just One Break is the nations oldest not-for-profit employment placement service for people with disabilities.

Getting Started

By Vicki Lansen

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It's been 65 years since The League for the Physically Handicapped staged a sit-in in New York City protesting the government's refusal to refer people with disabilities to a government work program. But many people with disabilities still encounter difficult - yet surmountable - hurdles on the road to gainful employment.

Ideally, everyone with a disability who desires to work and is qualified for a job should be able to get to that job. For many, there are the logistics of getting to and from a traditional job interview; the transportation arrangements for those who are not able to drive and must use public transportation. There are job accommodations to think about. First and foremost, people with disabilities seeking to enter (or reenter) the workplace need to be familiar with the ADA and be aware of employment opportunities that match their academic credentials and lifestyle needs.

If you've had little actual job experience before, you might think you aren't employable. Not so. People who have not had workforce experience often have very measurable skills. List expertise in home finance, organizational skills, or volunteer work and classes you have attended. Life skills not only count for college credit these days, properly phrased, they can highlight skills your prospective employer is looking for.

Prepare a resume

Major headhunters and resume services know that prospective employers spend less than a half-minute reviewing a single resume. Several on-line sites offer resume preparation & filing services for a fee. Before you spend money with one of these services, ask friends or family members to share their resumes. Find a simple one-page resume and follow the style when preparing yours. Ask several people to review it for form and clerical errors. Centers for Independent Living (CIL) also offer resources for assistance in preparing a resume and employment placement counseling. Halftheplanet.com has a complete listing of all CIL's in the United States.

Find an interested employer

Every major city and most, smaller metropolitan areas have Job Centers that post jobs ranging from private sector, to government positions. Job positions include everything from the dogcatcher to senior city attorney to clerical and entry-level jobs. Centrally located and federally funded, this is a good place to start for entry level searchers and for people interested in employment in their area. The offices are generally accessible and many host web sites for at-home online searching. For a complete listing of state employment sites, see:

http://usgovinfo.about.com/blstjobs.htm

Start Getting your Resume out there

With on-line resume sites and major corporations such as Johnson & Johnson (www.jnj.com) and government agencies like the Central Intelligence Agency (www.odci.gov/cia) actively recruiting people with disabilities, getting your resume to interested employers is quicker and more efficient. You can also search jobs and find employment career opportunities by posting your resume and searching the job databases at some of the many different job search sites like Employers Online or Career Mosaic.

It's still important to provide a cover letter in short form when sending a resume by U.S. mail. A short

(Continued on page 21)

Christine Phillips
Publisher

Fall is finally here and I promised you the secret behind that smiling face in the upper left hand corner of Coping Strategies.

For over 15 years now, my husband and I have taken coenzyme Q10 and Vitamin E on a regular basis. We both notice when we miss a day because of the sluggishness and short-tempered attitudes we corner. I can usually notice a difference in my behavior within an hour or so after taking my vitamins. It may be in my head but I know I do feel better physically!

These incredible supplements have been around forever, and are becoming more and more valuable.

I will cut to the chase and get right to the reasons these supplements will help you storm through those winter months.

—Christine Phillips-Publisher

Editor's notes: The views and opinions expressed are those of the author and are not to be construed as an endorsement by EDS Today. Always consult your physician before beginning any alternative therapy treatment.

Coenzyme Q10

My knowledge comes from being raised by a grandmother that read health guides for entertainment and used her natural remedies on her grandchildren. I managed a nutrition center and studied all new information on health foods and supplements as I tried to learn what was going on with my body and what EDS was. My Uncle, Mike, owned several health food stores throughout the Chicago area and he always encouraged us to try new things. One supplement stood out for me and I have been on it ever since reading that first sentence, "Coenzyme Q10 is one of the ten common substances found in the human tissue. This substance plays a critical role in the production of energy in every cell of our bodies." Wow, pretty powerful words aren't they? Well, my research had begun and this is what I learned along the vitamin road.

Coenzyme Q10 is a substance found in all the cells of living organisms. It is the spark that ignites the energy at the cellular level. Coenzyme Q10 is like vitamin E but has been proven to be a more powerful antioxidant. It aids circulation, stimulates the immune system, increases tissue oxygenation, and has a vital anti-aging effect. Some deficiencies of coenzyme Q10 have been linked to periodontal disease, diabetes and muscular dystrophy.

Coenzyme Q10 has been researched to find that it has the ability to counter histamine, and therefore is beneficial for people with allergies, respiratory diseases, and asthma. It is even used by many health care professionals to treat anomalies of mental functions such as those associated with schizophrenia and Alzheimer's. It is also beneficial in fighting obesity, candidiasis, diabetes and multiple sclerosis.

More importantly, coenzyme Q10 has taken a giant step forward in the treatment and prevention of cardiovascular diseases. Coenzyme Q10 is widely used in Japan. According to the *Nutritional Healing, Balch and Balch;* more than 12 million people in that

country are reportedly taking it at the direction of their physicians for treatment of heart disease (it strengthens the heart muscle), high blood pressure, and to enhance the immune system. Research in Japan has shown that Coenzyme Q10 also protects the stomach lining and duodenum, and may help heal duodenal ulcers.

I started by taking one a day, this was a mere 50 mgs. I now take 3-4 a day. Comparison shopping is the best way to keep costs down and product superiority up. When I forget to take my coenzyme Q10, I seem to dive as far as energy and immune system. Every bug that walks through that door has my name on it! I seemed to have found a natural fly-swatter, so to speak.

There are also food sources in which we can find Coenzyme Q10 if you are not into supplements. Mackerel, salmon, and sardines contain the largest amounts of coenzyme Q10. It is also found in beef, peanuts and spinach.

Coenzyme Q10 is oil soluble and absorbed best when taken with oily or fatty foods, such as fish. I know some of you are wondering, "what is the difference between oil soluble and water-soluble?" Water-soluble vitamins must be taken into the body daily. They cannot be stored and can be excreted within one to four days. The best-known water-soluble vitamins are Vitamin C and the B-complex vitamins. Oil-soluble vitamins store longer in our bodies, because they are stored in our liver and fatty tissues. Vitamins E, D, A and K are oil-soluble.

This leads me into Vitamin E, which I also take. Vitamin E is also an antioxidant that helps prevent cancer and cardiovascular diseases. It improves circulation and is necessary for tissue repair.

For women, vitamin E is useful in treating premenstrual syndrome and fibrocystic disease of the breast. It promotes normal blood clotting and healing, reducing scars from some wounds.

It strengthens capillary walls, provides healthy nerves and muscle. In addition, it promotes healthy skin and hair. As an antioxidant, vitamin E prevents cell damage.

When we were small we would break open a Vitamin E capsule and put it on our scratches and bites. My scarring is really limited as I also use it after surgeries. I have found it helpful in relaxing leg cramps and lowering blood pressure.

Vitamin E deficiciency may result in damage to red blood cells and destruction of nerves. There are several signs of deficincy, including: infertility, menstrual problems, neuromuscular impairment, shortened red blood cell life span, and uterine degeneration.

Vitamin E

Vitamin E, like Coenzyme Q10, can be found in foods like dark green leafy vegetables, legumes, nuts, seeds, cold pressed vegetable oils and whole grains, brown rice, milk, oatmeal, soybeans, sweet potatoes, cornmeal and eggs.

I have shopped several places and found the best buys on vitamin E are brands that have been around longer. I spend a little more on the vitamin E and Coenzyme Q10 than any other vitamin I take because I want to make sure that the two supplements most important to my body are the highest quality.

This is not an in depth study, but these supplements have worked for me and many other people with EDS and other chronic illnesses. Everywhere I go, I always tell people about these two vitamins and how they make a difference in my body and my life. We are so different from one another and yet so much alike. We have to try little combinations of everything to see what works best for our bodies. Experimenting can be a good thing. Do some research on your own and talk to your doctor to make sure this particular supplement is right for you. Take Uncle Mike's advice and try new things, they may work for you too.

Hidden Disabilities

Bek Oberin

Columnist Bek Oberin wrote a regular column on Invisible Disabilities for the former website Themestream. She has generously granted reprint permission for the series to EDS Today.

What's An Invisible Disability?

IDs - Invisible Disabilities. What's an invisible disability?

"Invisible" is a pretty easy word to define. The Webster's dictionary says:

> *Invisible (a): Incapable of being seen; not perceptible by vision; not visible.*

But in this case we don't necessarily mean completely invisible. By "invisible", I mean things that can't be seen by a casual observer the way you will notice somebody in a wheelchair, or somebody with a seeing-eye dog. For example I class hearing impairment as an invisible disability, even though hearing aids can be marginally visible.

First glance is important because in the society we live in, it's the first look that makes the biggest impression on people. If people's first glance classes you as a non-disabled person, you will have all the problems that people with invisible disabilities face even if your disability *can* be seen if people look harder.

"Disability" is a little harder to define. Looking at the Webster's again we see:

> *Disability (n): State of being disabled; deprivation or want of ability; absence of competent physical, intellectual, or moral power, means, fitness, and the like.*

I don't like that definition though, it doesn't feel good. I checked out the definition that the Americans with Disabilities Act uses:

The term "disability" means, with respect to an individual -

1. a physical or mental impairment that substantially limits one or more of the major life activities of such individual;
2. a record of such an impairment;
3. or being regarded as having such an impairment.

"Substantially limits ... major life activities" is a nicely broad definition and seems a lot less judgemental than what the dictionary says.

So an invisible disability is something not immediately noticeable but which substantially limits your major life activities.

Not surprisingly, living with an invisible disability can be a huge challenge! Perhaps it's even more of a challenge than living with a visible disability because people with invisible disabilities have to deal with society's indifference and misunderstandings as well as dealing with their disabilities.

It's this "extra" challenge that this column is all about.

I am always thrilled to get feedback about the articles I have written, or about future articles you'd like to see. Email me at gossamer@tertius.net.au with comments.

EDS Today sends special thanks from EDS Today to Applied Mechanical, Rick Burkhead and Family and to Barb Tuttle R.N. for everything.

Laura Hague

Laura Hague is a history instructor at Austin Community College. She has a Ph.D. in history from the University of Texas and is currently doing research on changes in the perception of disability at the end of the 19th century. She is currently having quite a bit of a row with her hands and feet but at least has, as of this writing, her head (physically, at any rate) back on her shoulders. She also has a grown daughter with HEDS, Sarah Meador.

Gallagher, Hugh Gregory. *Black Bird Fly Away: Disabled in an Able-Bodied World*. Vandamere Press, 1998, PO Box 5242, Arlington, VA 22205.

Hugh Gregory Gallagher relates his experiences as a man who acquired his disability through infection with the polio virus in 1952. This work is primarily a collection of his writings, some previously published elsewhere, that provides a window into his struggle for self-acceptance.

Gallagher has been at the forefront of disability activism even before he was willing to accept himself as disabled. For decades, he shunned the "professional disabled," those who, having found themselves disabled then found themselves work revolving around the creation of a disability community. Indeed, he avoided even socializing with other disabled persons. From the 1980s on, however, Gallagher has accepted membership in this community, and accepted the mantle of leadership. *Black Bird Fly Away* is ultimately a chronicle of that change in his own attitudes.

While Gallagher now self-identifies very strongly as "a polio," his experience of disability will likely ring a bell with those with other disabilities, including EDS. The frustration and anger that he felt, and repressed, as his youthful dreams withered along with his legs is familiar. Equally familiar is the depression that came, twenty years later for him, when he finally admitted to himself that he was crippled. The depression, in Gallagher's estimation, was far more painful than even the active disease of polio, and far more lonely. He at last faced the choice to either accept himself as a disabled person, or to die, emotionally if not physically. His answer was to stop being "Super Crip," to stop being concerned about whether he was a burden to others, to stop punishing himself for being crippled.

The autobiographical tales that he tells, such as his wrestling with himself over whether to make the change to an electric wheelchair from manual one, will seem very familiar. His speeches and brief papers on the history of the disabled, most published for the first time in this volume, examine how far the disability rights movement has come, and what dangers still lurk. Gallagher draws a telling parallel between the cost analysis of modern day HMOs and government-sponsored medicine and that of the Nazi health care system, in which hundreds of thousands of handicapped and mentally ill persons were euthanized. Anyone who has ever had a wheelchair denied or needed therapy curtailed will feel a chill of recognition as Gallagher explains how the Nazis rationalized murders in the name of rationing scarce health care resources.

Since this book was published three years ago, it may not be easy to find. I think it worthwhile to look for it, though, especially if you are going through the process of accepting your own disability. That can be a very lonely, trying time, and this book offers a gentle reminder that others have been there, too, and you, like them will survive.

A Mom's Ramblings

By Jennifer Cameron

My Children have a rare inherited disorder called Hypermobile Ehlers Danlos Syndrome (EDS). Basically this means that their connective tissue or collagen does work properly. In daily living this translates to multiple dislocations daily, poor healing, increased risk for infection, asthma, gastro-intestinal problems, constant pain and fatigue.

It really is a struggle to deal with EDS and kids. A lot of choices need to be based on your individual situation and how affected your kids are. Here is a small picture of my kids and how we are dealing with eds.

Andew

Andrew will be seven next month. He was diagnosed with EDS when he was five. The first obvious sign was the difficulty and pain he had printing. His fingers dislocate daily. I never understood why he didn't color in pictures (it hurts!). Soon we noticed additional problems including: reflux and subluxing patellae, toes, mid-foot and ankles.

When Andrew started kindergarten he really struggled because he tried to keep up with his peers physically and couldn't. He would come home after the morning and be sick to his stomach because he was in so much pain. It took a long time and Counseling to get him to pace himself and take breaks. Part of the problem he faces is that he is allergic to pretty much all pain medication stronger than Tylenol. Andrew does not remember what it is like to have a day without pain. I think that as a parent this is one of the toughest things to deal with.

In the last year and a half Andrew has progressed to the point of requiring a scooter for distances; ankle, knee and wrist bracing; and ring splints for all fingers. He requires daily medication for reflux and sleeps with a bolster. On a good day he has about twenty to thirty dislocations but during bad weather it can be fifty or more. He can reduce fingers toes and mid-foot independently most days. He also subluxes his hips, elbows, shoulders and ribs. I can usually reset everything but ribs. I don't reset his ribs because I could easily risk puncturing a lung.

Andrew has been on home instruction from school since April 2000 due to an anaphylactic allergy to latex. His latex allergy has made bracing and splinting a challenge as so many companies are unable to assure us that their products are not contaminated. The one company that has helped us is Bio-Skin as their products are safe, custom or over-the-counter made, lighter and stronger than neoprene.

As of January 2001, Andrew will finally attend school in a regular classroom. He will have the support of his physio-therapist, occupational therapist, counselor and nurse case manager to facilitate his re-integration. He will be using an Alpha Smart 3000 to do his "written work" to conserve his joints.

On good days Andrew likes to be like any other boys his age — playing soccer, building snow forts etc. We let him and treasure these times as they come. He is a bright little boy with lots of interests which we do our best to encourage.

Joshua

Joshua turned four in August 2000 and started showing signs at a very early age. At three months he was diagnosed with asthma. Early on we noticed Joshua didn't heal well and got cellulitis within an hour of getting a mosquito bite. He had ten ear infections before his first birthday. Surgery for tubes at eleven and a half months went well and significantly reduced the frequency of the ear infections.

He has a history of choking to the point of complete obstructions requiring the Heimlich. He frequently aspirates resulting in bronchitis or pneumonia.

(Continued on page 15)

Joshua now recognizes the early signs of bronchitis. He averages a round of bronchitis or respiratory problems every six to eight weeks.

Currently his speech and language pathologist has speculated that his swallowing issues are worse due to a recent growth spurt which has resulted in a lack of coordination for swallowing.

Joshua dislocates and subluxes his toes, ankles, knees, hips, fingers, wrists, elbows, ribs, shoulders and jaw. He also averages twenty to thirty dislocations a day and significantly more on bad days. During storm systems his joints are like Jell-o and a simple touch can dislocate something.

Joshua also had dental surgery at two years of age due to a broken tooth and cavities.

In May 2000 he had his tonsils and adenoids removed at Sick Kids in Toronto. The local hospital and E.N.T. were not comfortable dealing with the risks of EDS and a metal sensitivity. He gets hives from his medic alert bracelet unless it has a protective coating.

Joshua is the dare devil of our household and has been blessed with an incredible mechanically inclined mind. The Occupational Therapist has marveled that she rarely has to teach his energy conserving techniques as he has already figured them out.

Joshua does require constant supervision especially on the stairs as patella and ankle dislocations often result in falls down the stairs. He has had several concussions due to falls.

Joshua uses a picture scale to help us understand his pain. He is on daily Tylenol for pain, Prevacid for reflux, puffers and Zaditen for asthma. He uses a Panda chair for energy conservation and pain management. He has ankle and knee braces, resting splints for his hands and wrists, and ring splints. Joshua also has contact allergic reactions to latex.

As a Mom, one of the biggest difficulties I have is remembering to stop and look after me! I have EDS too. I have been having major problems with my hands but was so busy looking after the kids needs that I waited two months before doing anything about it.

Although our lives are challenging, busy and some days just plain hard we are able to celebrate the good times and support each other during the rough times.

My Earth Angel

By Jil "Anjil" Manning

There's a good friend whom I met,
My best inspiration yet.
She inspires so many folks,
She gives such faith, love, and hope.

Troubles? She's got more than many…
Complaints, whining? You won't hear any.
She volunteers to help others.
She's friend, wife, wonderful mother.

An angel inspiration…
I have such love and admiration.
Her very challenging world,
Makes her *such* an amazing girl.

So this is my tribute to,
Such an extraordinary friend who,
Keeps on going when life's so tough,
Smiles and prayers, no matter how rough.

So now I will tell you who
This exceptional woman is, so true.
Rickie Murphey, my angel friend.
Love and hugs, with this poem I send.

Brace Yourself

Barbara Uggen-Davis
Editor

This installment of "Brace Yourself" looks at knee braces. There are a lot of different options out there. What works for one person may be all wrong for another. Hopefully this column will give an idea of the range of products available so that you may discuss various options with your physician and find the right choice for you.

The braces listed in this article come from personal research and discussions with other people who have EDS.

— Barbara J. Uggen-Davis

The views and opinions expressed here are those of the author and are not to be construed as an endorsement by EDS Today. Always consult your physician before trying any brace.

Knee Braces

Knee problems are common for many of us with EDS. Knees take a lot of stress from walking, standing, running, and even sitting. My knees are often most stressed from not using good body mechanics at my computer. Even as I write this, I am sitting in a position that I know to cause knee pain because it is more comfortable for my other joints.

I recently asked other people with EDS about their knee braces – what works, what doesn't, and how many different braces they tried before finding the right one. I was surprised by the variety of braces people use. What worked great for one person was not helpful to another. The severity and type of knee problem is a big factor in the decision whether or not to brace and what type of brace will work.

In 1984, the American Academy of Orthopedic Surgeons classified braces into three groups. [1]

- Prophylactic - to prevent or reduce the severity of knee injuries.
- Rehabilitative - to allow protected motion of injured knees or knees that have been treated operatively.
- Functional - to provide stability for the unstable knee.

Based on their ongoing research, the AAOS does not believe that prophylactic knee bracing is effective. In fact, findings show that prophylactic bracing may contribute to knee injury rather than prevent it. On the other hand, the AAOS has stated, "rehabilitative knee braces and functional knee braces can be effective in many treatment programs, and that this efficacy has been demonstrated by long-term scientifically conducted studies."

Functional knee braces, as defined above, are designed for unstable knees, characteristic of EDS. They range from over-the-counter elastic and neoprene braces to

custom-made orthotics. It is not surprising then that most of the respondents use bracing in this category.

Respondents reported varying types and degrees of knee problems including hypermobility, patella instability, medial/lateral instability, medial/laterial meniscus injury, osteoarthritis, tendonitis, and both torn and detached anterior cruciate ligaments (ACL). The types of braces they use are also quite varied. In their own words, they describe what works for them.

"I have an open patella brace and I love it."

[I use a] "Donjoy Defiance, which is a hinged, calf to mid-thigh brace that was custom-made. If needed, tape and tubigrip underneath for patella stabilizing because the brace does not work on the patella at all."

"My daughters and I between us have multiple knee 'symptoms.' They range from hypermobility to a knee cap that can be moved in a large circle to a constricture one daughter has in her right knee. The best brace we've found for all these problems and pain associated with them is an elastomag (solid magnetic surface – not spot magnets) wrap produced by a Japanese company called Nikken."

[I use a] "bilateral Versa from Breg."

"It's just a commercial neoprene open patella brace with extra reinforcements on one side. She also uses a Velcro knee brace. Also commercially available."

[I use] "the Donjoy knee brace like the football players use, due to an ACL tear/replacement."

"The only knee brace I have had was one of those full leg things you have to pull on, and I don't have the hand strength to pull it on!"

Many of the respondents tried several different bracing options before finding one that worked. Most respondents tried between 4-6, but some tried "virtually everything." Some respondents are still looking for the right brace or have given up trying. Some of the braces that did not work for respondents were:

- Elastic braces
- Neoprene braces of various styles
- Ace bandages
- Taping
- Custom-made braces

Allergic reactions to bracing materials were common, especially for neoprene. One respondent was allergic to the surgical steel used in a custom-made brace. Over-the-counter braces often did not provide adequate support or did not fit properly. Many braces failed to provide adequate patellar stability.

Although a few respondents did find the right brace on the first try, more often it was a matter of trial and error. Keep this in mind when you are looking for the right brace for your needs. Discuss all of the available options with your doctor. Be sure the braces you try address all of your knee problems. For example, if you have medial and lateral instability, as well as patellar instability, you don't want to get a brace what only stabilizes the patella. When you find a brace that works, remember that functional braces provide stability only in low-load conditions. They are not meant for athletic activities.[1] This does not mean that you should avoid exercise. Maintaining muscle strength is important, especially when wearing a brace as dependence on bracing can make the joint weaker over time.

As always consult your physician before deciding to use bracing and when choosing a brace for your needs. Your physician can refer you to specialists that can make custom bracing or help you determine which type of over-the-counter product is most likely to meet your needs.

1 http://www.aaos.org/wordhtml/papers/position/kneebr.htm, Position Statement, The Use of Knee Braces, American Association of Orthopedic Surgeons.

Announcements

Adolescent Chronic Fatigue Syndrome Study

Dr. Julian Stewart is seeking adolescents with CFS ages 13-19 for a new study funded by the National Institutes of Health. The study will be conducted at Westchester Medical Center, which is in Valhalla, New York, near New York City, the Hudson Valley, New Jersey, and Connecticut. If you know of any adolescents with CFS that might be interested in participating, please let them know about the opportunity. The primary goal of this study is to determine the cause of orthostatic intolerance (such as Neurally Mediated Hypotension and Postural Orthostatic Tachycardia Syndrome) in young persons with CFS. You do not have to have a diagnosis of orthostatic intolerance to participate in the study. The other goals of the study are to determine whether midodrine is a helpful medication for CFS in teens, and whether understanding the cause and subgroups of orthostatic intolerance in CFS can help predict who will benefit from treatment with midodrine. In addition, by understanding more about orthostatic intolerance when it is resistant to treatment, scientists may get ideas about how to treat it in the future. You can read more about this study and download consent and screening forms from the Home Page of Dr. Stewart's Center for Pediatric Hypotension: "http://www.syncope.org. Follow the link for the NIH-funded CFS study. You can also contact Dr. Stewart directly with questions at stewart@nymc.edu.

EDS and Hearing Study

This study, as announced last issue, is still accepting participants. For more information, please see our website at http://www.edstoday.org/research.htm or contact Marge Tamas at mtamas@mindspring.com or (770) 439-6037.

New Domain Name

EDS Today has a home on the internet. Check out our website at its new address: www.edstoday.org.

EDS Awareness Campaign

EDS Today is heading up a collaborative effort by several EDS groups worldwide to launch an EDS Awareness Campaign.

Volunteers are needed to assist with media exposure, educating the medical community, educating teachers, and increasing awareness in the general public. Your ideas and efforts will help make the campaign an international success.

Bumper stickers, awareness pins, and media kits will be available soon. To raise funds for the various EDS Groups participating in the event, Joyce Norris has offered to sell sterling silver awareness ribbon necklaces. A portion of the proceeds from each necklace will be donated to the EDS Group of your choice. If you wish to purchase the necklaces, you may use the order form at the back of this newsletter (proceeds will benefit EDS Today) or buy online through the campaign website at

http://members.home.com/uggenb/

To learn more about the campaign and how you can participate, visit the campaign website above or call EDS Today at (253) 835-1735.

T-Sirts

EDS Today has T-shirts with the EDS Today logo, as seen on the cover. Use the order form at the back of the newsletter to request yours. Sizes from S to 3X in cream, white, and gray. Prices $15 for S, $20 for M, L, XL, and $25 for 2X-3X.

Mail Bag

Dear EDS Today,

I want to thank you for the article on public school and EDS [A Mom's Rambling : When Traditional Schools and EDS Don't Mix (pg 23 vol. 2 issue 1 Spring-Summer 2001). I did not see a letter to editor section, but I had to respond to this article! I had terrible problems with fatigue, headache, and nausea when I was in grade school.

At the time, I did not have severe enough symptoms for anyone to suggest EDS. The fatigue was attributed to sleep problems... The headaches were either [labeled] "allergies" or "chronic sinus infections"... Now that I am "old enough" to have migraines, I must observe that I really cannot tell the difference between these and what I had as a child! I even got visual disturbance then, attributed to "sinus pressure". Only now I get to go home and lay down, and then I got a school nurse telling me I had no fever, and my normal temp didn't really run low!

I suffered nausea when I attended school and eventually when I awoke and CONTEMPLATED attending school. At the time, everyone said "school phobia," possibly aggravated by an allergy to school cleaning chemicals which was picked up after my 1 solitary "asthma" attack. In retrospect, though, I think this may have been primarily a manifestation of joint pain.

I had always felt "stiff" when I first woke up... and whenever I sat, stood, [or] lay still for more than a few minutes. Because of this, I was driven to engage in highly physical activities AT LEAST 2 hours a day… I never dared sit still for a whole 24 hr period because I "couldn't sleep"... if I did not use my muscles sufficiently before retiring... I told people I was "restless," and later thought that I might have some variation of "hyperactivity"… that drove me to "have" to DO something before bed. This probably began with my enrollment in baby gymnastics because I had a birth defect - partially formed hip joint… I must have immediately decided that life was more comfortable if very active! Because I grew up this way, I was accustomed to it and never thought of it as pain, or unusual for that matter. My mom and most of her family took it as a given that sitting still caused stiffness and this was a good reason for squirming. "Pain" was reserved for the headaches. I merely felt I was "uncomfortable" or that I "needed to move". I now think that my "nausea" when forced to sit still for 6 hrs in school was at least partially my way of describing the build-up of this "inactivity - pain". Probably my constantly moving, well used muscles prevented any one joint from standing out, but I noticed the "overall" effect of stomachache/nausea.

...I missed so much school and was late so often that my school threatened to fail me repeatedly… The only thing that stopped them was my high test grades, teacher support, and "honors" status. They required that I see the school psychologist regularly for "school phobia", then that I seek outside counseling. Eventually (and with my school psychologist's blessing, thank you) I dropped out and homeschooled. My school teachers were generally very supportive (this was end of 9th and all of 10th grade...), but the administration was outright MEAN! They had never had an honors student attempt to drop out before, and told me that I would surely fail AND have to repeat the year. Even when I got a 99% on 1 of my Regeant Exams that year, I was told that this exam… "did not reflect actual learning." So I gave up on my home district and transferred to a small, private boarding school (and spent all my intended college $!). This was an alternative school that allowed us to design our own curriculum,... use independent study, and had small, flexible classes. I have never regretted the lost college funds!

My joint flexibility apparently surpassed my ability to compensate fully when I was around age 20. Skin and organ symptoms appeared after, when I tried to put in a "full" day (1st 3/4 time work AND a few classes, then part-time work AND full-time school) despite the fatigue and joint objection. I was diagnosed 1st with "tissue dysplasia", then the following year with "EDS, probably Hypermobile type". As the soft tissue symptoms have become more pronounced and family history better explored, the likelihood of my "type" actually being a blend of paternal soft tissue (classical w/ gum involvement?) AND maternal hypermobility became likely. No one in either family has as severe or extensive/variable symptoms as I, though! None of my symptoms were severe enough to be picked up until after I left grade school, and then they progressed rapidly... I sill do not have [a] chronic fatigue diagnosis because my MDs have said either CFS is generally a problem in older people and also must have a definite "start date" or because they cannot diagnose CFS if a patient has a pre-existing condition (EDS). My episodes of fatigue have occurred on and off for as long as I can remember...

Despite this, I am currently an Honors student at university. I now plan my schedule to allow adequate "sick days", and schedule in "physical therapy" classes... into my school days. I use disability center for voice input/output computers (to spare my hand joints and eyes), and to help me arrange incomplete grades when I miss too much school and cannot recover... in time for catch up. I now know that if I ignore [my] "urge to move" more than 3 consecutive days, I risk painful subluxation (and eventually/occasionally dislocation), usually in bed at night. Probably my constant restlessness and activity level as a child and teen masked my loose joints. Ironically, this same activity made my adults skeptical of my fatigue and nausea when confronted with a long day of stillness at school. My absences/ lateness (which allowed extra sleep) probably allowed my body to cover most of the soft tissue symptoms!

I feel that getting out of the restrictions of public school schedule and learning to listen to my body and pace myself has allowed me to attend university. It has given me the energy to contemplate seeking a way around the unbearable 9-5 workday...

Thanks so much for a story about another child plagued with inability to maintain grade school attendance! Best of luck to Jayme Hobe's daughter!!! I hope she never stops listening to her body, the results of "ignoring" my body's plea for extra sleep and exercise… were the rapid progression of subluxations/dislocations and the onset of underlying soft tissue, skin, and bleeding problems! It led to the EDS diagnosis, but I would have preferred to learn gradually. Since EDS is not reversible, I suggest we all oblige our bodies' requests for rest and lack of immobility. they know what they ask!!!

Sincerely,
Sascha Horowitz
(23 yr old EDSer)
Las Vegas, NV USA

The Staff of EDS Today wishes to thank the following people. Thanks to all the Crystal Springs friends in Bolton MA, for their continued support and love. Thanks Diane Specht for your strength, endless energy and compassion — the fundraiser was so much fun! Thanks Tracey Wilkie for your friendship and enthusiasm for EDS Today. Thanks Florence Sutley for your endless stories of Atlantic City, you made our trip to Pottstown and back a joy! Thanks Cathy, Dave, Melissa and Donna for your love and kindness. Thanks Doug Phillips for believing in EDS Today as much as we do. Thank you Lorene for Doug. Thanks Joann, Larry and Johnny Sloan without you we could not do what we do.

(Continued from page 9)

form cover letter should outline the position you are applying for, how you feel your qualifications fit, and when you are available for an interview. Faxing resumes to perspective employers is a popular trend but don't discount the old fashioned snail-mail.

Following up and getting the first interview

Give the perspective employer at least a week to ten days before following up with a phone call or short note. Ask if they have received the resume and when they expect to interview. You may be informed that they have filled the position, or that they will be accepting resumes for several weeks.

When you receive a call back for a personal interview, don't drop all of your job-hunting efforts. For every twenty resumes a job hunter sends out, only five result in a call back. And on average, only one of those results in an actual offer. The most important part of looking for a job is getting started.

This means nothing if you can't get your foot, or wheels in the door. Because of the ADA you should be able to get in the door, but that isn't necessarily what happens. Unfortunately for a lot of people, the ADA is a big stack of paper that contains some meat somewhere. Where's the beef and how and when can you use it? We'll explore some real in-depth, real life stories in future articles. Things like "I'm qualified, I get through the first pass, but when I walk or roll in the door, that's where the brick wall is."

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Looking from the other side

Behind each smile there is a mask
Behind each mask there is a smile
Nobody except us knows which is which
We put on the mask each day,
To protect ourselves, and the ones we love

If our hurt shows, we hurt our love
If our love hurts, we show our love
It is the paradox of the love we have
To show our true selves can hurt beyond all measure
Not to show our true selves can hurt even more

To touch or not to touch?
Is a question begged
The slightest touch causes excruciating pain
The slightest touch soothes and brings us peace
Which is it today?

We try to keep our pain for you inside
We can not show the world that we are in pain
If we cry, the world asks why?
If we smile the world asks why?
Our answer is the same, because we care

For every step taken, we celebrate
For each obstacle tackled, we are in ore
The victory is yours, the valor is yours
For every tear you cry, we grieve
The pain too, is yours

Am I the carer? Or are you?
Or am I the sufferer? Or are you?
I am both, You are both
Who is who? Only you and I know
For we wear the same mask, you and I

By Jonathan Ashley

Service Dogs

EDS Today is proud to introduce a new series of articles on Service Dogs for people with EDS. This column will feature several authors affected by EDS who use service dogs.

This issue introduces three authors—two with EDS who are training their own services dogs and one dog trainer whose daughter has EDS.

The views and opinions expressed here are those of the author and are not to be construed as an endorsement by EDS Today.

Sherlock
Age 4 months

Meet Barbara and Sherlock

Last December, I adopted a 4-month old, Siberian Husky and German Shepherd mix. Sherlock is a smart boy with a big heart and warm, wet kisses. The woman we adopted him from told us she thought he would make an excellent police or service dog. Sherlock learns quickly. We had him potty trained in only 4 days. (Yes, really— 4 days!)

Although my EDS hasn't been severe enough lately to need a service dog, my husband felt it best to have a dog that can be trained for service work so we will both be ready when and if the time comes.

I didn't know anything about service dogs, but I've learned a lot over the past several months.

I've been an avid reader of service dog email lists. I quickly started recognizing names of people with EDS that I knew from EDS email lists. Pretty soon, the idea of a column on training service dogs for people with EDS evolved.

— Barbara J. Uggen-Davis, Editor

Paula Offutt
and Joella

Meet Paula and Joella

I am Paula Offutt, EDS Hypermobility Type. I currently use a power-chair to help with pain, subluxes, SI joint dys-function, and all the other usual stuff. I live in rural North Caro-lina, near Asheville. Last November, I started to seriously think about getting a Service Dog. I thought I had one picked out but he died of a puppy disease.

I had already contacted a dog trainer friend about helping me with Zeke. She called me about a month after he died and asked if I was still interested in a service dog. She had a dog that loved to retrieve so we went over to meet it. I did not like her at first. She was just under 7 months, scrawny, and a full breed. I am a mutt fan. But I slowly fell in love be-fore we left and the next day I brought her home. I named her Joella, Hebrew for "God is Willing". She is an adorable, sensitive, sweetheart of a clown.

She is also a Rottweiler which has caused some

trouble with various folks around me. But most who meet her are surprised by her quiet demeanor. Ha! That's how she is now — come home with me and see the REAL Joella!

Currently, I need a service dog to pick dropped items up for me. If I lean over to get something, I often start a spasm in my neck/shoulder areas. If I happen to be standing, I can reach the floor but my knees are now starting to buckle unexpectedly. So the first thing we taught Joella, after the usual obedience stuff, was to pick up my keys. Then we advanced to hats, bookbags, magazines, etc. If she sees or hears me drop something, she will automatically pick it up. I am now using a laser pointer to get her to pick up something specific. Sure, I could have gotten a reacher but a dog is much more fun! A reacher won't bring you a soggy fuzzy tennis ball when you are feeling depressed! A reacher won't get scared to death when the person dressed as the Easter Bunny waves at you at the mall!

— Paula Offutt

Meet Lynn Hoover

I live just north of Boston and am a medical family therapist and licensed independent clinical social worker (LICSW). I work as clinical director for K-9 Partners, Inc. a small, non-profit organization that provides trained dogs to serve children, teens and young adults with disabilities. I have a child diagnosed with Ehlers-Danlos and Fibromyalgia, and personal experience has taught me how assistance dogs help mitigate disabilities caused by these illnesses.

K-9 Partners is unique because we are research-oriented. Our primary purpose is to contribute to the now-sparse literature on the benefits of service dogs for young people. Happily, along the way we meet human need. Our motto is "To Serve with Love is Perfect Freedom". I look forward to contributing to and learning from the EDS newsletter.

Lynn Hoover, LICSW

http://www.k-9partners.com/

Recipes for Health

Crock pot Veggie Stew (Ratatouille)

Submitted by Sharon Kay

For more recipes by Sharon, please visit her recipe website at http://www.sites4you.net/recipes/

Directions

Measure all ingredients into crock pot. Stir, and cook on low for 8-9 hours or high for 4- 4 1/2 hours. I serve this over rice.

Serves 4 or more

Ingredients

- 1- 19 oz. can tomatoes, with juice, broken up
- 1 small eggplant, with peel, cut into 1/2" pieces
- 1 cup finely chopped onion
- 1 medium green or red pepper, chopped
- 1/4 cup chili sauce or ketchup
- 2 teaspoons sugar
- 3 cups sliced zucchini, with peel, 1/4" thick
- 1 teaspoon parsley flakes
- 1/2 teaspoon salt
- 1/8 teaspoon pepper
- 1/4 teaspoon garlic powder
- 1/2 teaspoon dried oregano
- 1/2 teaspoon dried basil

Memorials and Tributes

I had an incredible summer this year. The best and most memorable time I had was touching base once again with my friend and inspiration, Cathy Bowen.

Walking into her home was as if I had lived there all my life, a place of love, joy and safety, yet also sadness and loss. Family members and neighbors greeted me as if I was their sister, their daughter, or their long lost friend just because of my association with Cathy. During this eventful stay I had the pleasure of meeting one of my newest and dearest friends. This beautiful young woman was a true joy and delight for me. We tried to spend as much time together as possible and the connection strengthened daily. I would like to proudly introduce to everyone the author of a very special tribute. Take it away my friend.

With Love,

Christine Phillips/Publisher

If you would like to honor someone close to you in our memorial or tribute pages, please e-mail me:

SJ-EDS-DAVID-MOM@ prodigy.net

-Cathy Bowen

Dedicated to the One we Love.

Hello everyone. I have been chosen to write an article for the newsletter my mother and friends brainstormed over for several years. I can only write what's in my heart and sealed in my soul. My new friend, Christine, informed me that this is what EDS TODAY was about, real people and real news. So my story begins.

One of the toughest times in my life was watching my family fall apart. We had always been a team, and I was the happiest when I was with my brother. We did everything together. He was a sports fanatic, and had an undying love for hockey. He could make everyone laugh at anything, at anytime.

Christmas Day, we always took turns opening gifts. Now as I watch our family videos, I notice how in awe I was of Dave, how happy I was when my brother received his gifts. I was happier yet because he was my "Big" brother, my best friend and someone I knew would always be there for me. I never realized what family meant until we lost David. As you all might know by now, I am Melissa Bowen and my brother was David Bowen. He had Ehlers Danlos Syndrome, Vascular Type [IV].

It was the summer of 1996 and I was only 10. My 14-year-old brother was in the hospital and I hardly got to see him. When my parents told me that David had died my life shattered, I was numb. Every tradition I had ever known and had done with our family was gone forever. All we have left now are our memories. All our goals for David were shredded, and we fight daily to keep David's memory alive.

My mother, Catherine Bowen, has done an impeccable and fascinating job at this task. My mother talks to people from all around the world, teaching them about Ehlers Danlos Syndrome. She works endlessly on awareness of this syndrome. She has contributed hours of selfless work to family members, neighbors,

Cathy Bowen With Daughter Melissa

and strangers. She is a regular contributor in numerous events, golf tournaments (3 years) and fundraisers for the New Jersey Chapter of EDNF. Mom packs her car up and heads to the malls around Atlantic City and hands out her flyers and talks about her billboard she made with pictures of EDSers, continually impacting lives through David's story. She has made a point of educating the medical society by being on two television shows, CN8 and Health Update (Channel 40). Her latest endeavor has taken her away from her normal routine and put her into researching for hours, not only on the Internet but medical libraries. Every year she gives scholarships in memory of David, I am so proud of mom, she is my hero.

Watching her has inspired so many people and most importantly, me. I honestly can say that I pray I will grow up to be half as wonderful and loving as my mother. Mother never stops sharing her energy and her strengths with so many people.

It has been 5 years. David would be 20 and shortly I will be turning 16. It has been so difficult watching the daily sadness and sorrow that will forever surround us, but somehow I have accepted that my life has forever changed.

We will always be a family, we will always have David's memory and spirit in our hearts forever, thanks to my mother…..CATHY BOWEN.

Melissa Bowen / Christine Philips.☺

Pamela Popken-Harris, Ph.D

Our featured article this month comes from Pamela Popken-Harris, Ph.D. What makes this article different from other EDS research articles we have published is that the author also has EDS.

I've known Pam for roughly five or six years. We first met through an email mailing list for people with EDS. We later met in person here in Seattle and again in California.

I've asked Pam to write a little bit about the research study in this issue and about her latest effort to generate research funding for EDS through the Arthritis Foundation. Here is what she had to say.

—Barbara Uggen-Davis

Dear EDS Today Readers,

Your editor, Barbara Uggen-Davis, has asked me to write and tell you a little bit about myself, and the research study being published in this issue of EDS Today. We first under took this study as anecdotal reports suggested that there may be an increased prevalence of certain immune related disorders including allergy and asthma in people with the Ehlers-Danlos syndrome (EDS). I was interested in this as I have a mild form of EDS and also Crohn's disease, a chronic inflammatory bowel disease. In order to determine whether or not any type of immune abnormality in EDS may exist, we mailed out approximately 300 surveys to members of EDS support groups world-wide. Out of a possible 300 responses, 140 were received. In addition to information about immune functioning in patients with EDS, we also attempted to gather other sorts of information including the types of medications used, and the presence or absence of other possibly EDS related conditions including the presence or absence of either a heart murmur, or a skeletal abnormality such as scoliosis. In the instance where any type of abnormal response was received, we also asked for medical record documentation of both diagnoses.

One of the main surprises was that in our 140 total

respondents that there seemed to be a statistically significant risk for allergy and asthma in those with either a cardiac or skeletal abnormality or both. Although not as statistically significant, this association seemed to hold true just in the 43 individuals with a diagnosis of EDS proven by medical record documentation. The other main surprise was that there were two respondents with both EDS and Crohn's disease. As we could not document any type of immune abnormality or immune deficiency in any of our respondents that would account for this association, this suggests that a gene that could sometimes also be associated with the connective tissue defect could be responsible. Interestingly, known susceptibility genes for allergy, asthma, Crohn's disease and a mild type of EDS associated with Tenascin-X deficiency are all located on a region of chromosome 6 known as 6p21.3. This is a chromosomal region that is also known as the major histocompatibility complex or MHC, and many of the immune response genes in our DNA are found here. Furthermore, genes within this region are often linked, and defects in one gene are often coinheritied with other genes on the MHC that are located relatively far away. Interestingly, this small region of the genome has also been associated with atrial septal defects and skeletal abnormalities such as scoliosis, and both of these two types of abnormalities have also been described in EDS.

Another possibility that could account for the suggested association between Crohn's disease and EDS is the observation that people with Crohn's disease often have an abnormally high ability to take up certain small compounds into their circulation through their gastrointestinal tracts. In fact, it has been suggested by Crohn's disease researchers that in order to develop Crohn's disease one needs both a "permeability defect that exposes an individual to a greater number of antigenic challenges as well as a particular genetically determined immune response." A genetic susceptibility for both diseases on the MHC would therefore fulfill this hypothesis as people with EDS would logically also be at risk for a permeability defect. At least a subset of people with EDS, including those of the vascular and a few of the classical type individuals have also been reported to have an intestinal perforation or rupture. Subsequent to surgery to repair these defects, many of these also reportedly also have poor wound healing at the site of anastomosis. This is also a characteristic of individuals with Crohn's disease.

In the meanwhile, I am hoping to raise money for the Arthritis Foundation by completing the Honolulu marathon in December as they provide educational materials and research funding for both EDS and Crohn's disease. I also think it is so very important to exercise to the best of ones ability even if you do have EDS. Although I will primarily be walking, not running, the Honolulu Marathon with other members from the Arthritis Foundations Joints in Motion Team, I consider that they are doing nearly as much for me as I am doing for them. I am getting stronger and am now physically able to do things that I could not do before. I started out approximately one year ago with a mild exercise program at my local gym. At first, I was very careful to just do very low weights and high repetitions on the weight machines along with some stationary rowing. I also began a program of walking using both the track and treadmill. When I first started, I was relatively slow, completing only about 1 mile in twenty minutes and while on the treadmill, my ankles were also likely to give out. I was determined to keep at it however as one of my good friends with EDS who is just sixty and is in a wheelchair said I was much worse physically at age forty than she was. So, gradually, over the course of the year I have been improving. Through both diet and exercise I have lost over twenty pounds, and have also gained muscle. I also began taking glucosamine as there is good evidence that this supplement may prevent the progression of osteoarthritis. Now, instead of walking a mile in twenty minutes, I can walk a mile in just a little over 13 minutes, and

my ankles no longer give out! My daily struggle now is to balance the demands of research, family and marathon training as I need to exercise and walk/run for at least an hour and a half a day in order to log in my needed 50-60 miles a week. There is also some indication that this type of exercise may eventually help build strength in some connective tissues. So far, I have found this to be true as the chronic tendonitis that I was plagued with for over 15 years has now almost gone away! I consider that my Crohn's disease is also significantly better than it was a year ago. If you would like to consider giving a gift to the Arthritis Foundation in order to help me complete my goal, please contact me at the address below. I also would like to hear of any successes that you may also have had, and what has worked for you.

With best personal regards,

Pam Popken-Harris, Ph.D.
David F. Hickok Memorial
Cancer Research Laboratory
Abbott Northwestern Hospital
Minneapolis, MN 55407
Telephone (612) 863-4439
Fax (612) 863-4936

Joyce Norris

By Barbara Uggen-Davis

Despite earning a degree in nursing, Joyce Norris had never learned about Ehlers Danlos Syndrome. At the age of twenty-nine, Joyce first learned of EDS from her physiotherapist who had been treating her for ongoing joint and soft tissue problems. The physiotherapist had recently learned about EDS at a conference. This spark of information led Joyce to the medical library, a bevy of specialists and to the Internet where she not only found more EDS information but the opportunity to interact with others. She finally was diagnosed with Classical, Hypermobility and Vascular EDS at age 32.

After her diagnosis, Joyce needed a hobby to keep her active, yet avoid joint stress. She started making beaded jewelry. Over time, she had made more jewelry than she could wear and needed to find a way to support her hobby. What started with beads has evolved to include crystals, gem stones like amethyst, and sterling silver. Joyce came up with the idea of selling her jewelry online and donating the profit to the Canadian Ehlers Danlos Association. Her website, http://www3.ns.sympatico.ca/whuzzy/, contains information about EDS, Fibromyalgia, and Chronic Fatigue Syndrome, and a catalog of her jewelry.

Most recently, Joyce suggested to the CEDA email list that she could make sterling silver awareness ribbon necklaces for EDS. That idea sparked a discussion of increasing EDS awareness, which evolved into a full-scale EDS Awareness Campaign, to be held in February 2002.

Joyce is selling the necklaces (shown below) for $15 Canadian, plus shipping. A donation of $5 from each ribbon necklace sold will be donated to the EDS organization of your choice, including:

- Canadian Ehlers Danlos Association
- EDS Today
- Ehlers Danlos National Foundation
- Ehlers Danlos Foundation of New Zealand.

Information about the campaign can be found online at http://members.home.com/uggenb/ or by calling (253) 835-1735. If you would like to volunteer for the campaign, please call or email info@edstoday.org. The Awareness Campaign is a collaborative effort of the Canadian Ehlers Danlos Association, EDS Today, the Ehlers Danlos National Foundation, The Ehlers Danlos Foundation of New Zealand, and the Hypermobility and Fibromyalgia Mailing List.

Grete Mikalsen
4/22/64 to 9/9/98

Grete Mikalsen

4-22-64 ~ 9-9-98

By Cathy Bowen

It was EDS that brought us together, but it was the instant bond of compassion, love, friendship and laughter that we shared like sisters, that forever joined us as family. Along with her sister Gunn and nephew Christer (whom she adored), we finally met at the 1998 EDNF Conference, in Tampa. I was in awe of Grete. She radiated such strength, inner peace, and joy for life. She was such a bright light, not only for me, but the numerous others she had touched along the way. I marveled at the empathy she had for other EDSers, while enduring her own pain and uncertainty. She always had time to listen, and offer her help in anyway. Grete lived life to the fullest. She will always live in the heart's and memories of those she came in contact with. She was a loving, courageous, strong, happy (her brilliant smile), compassionate woman, whose life had purpose. She made a difference and left her mark on everyone she met. Grete was a fighter. She never gave up. I told Grete, "I'm very grateful for the week we shared, but I will always remember you this way; dancing to the "Chicken Dance" with the biggest, brightest smile on your face, laughing, having fun, living life."

Here's an excerpt from a letter Grete wrote for the United Kingdom's EDS Support Group Newsletter "Fragile Links."

Hi there all of you! I'm Grete, a Norwegian girl diagnosed with EDS, the Vascular Type (IV). I'd like to share some thoughts with you about how it is living with this form of EDS.

I was diagnosed in 1979, without being told much at all about EDS. After my father died in 1983 at the age of 47 (he had EDS too) I received some information, but no one really suggested this could be lethal. In the early 90's I felt the need to know more, and I started digging deeper for information. After reading the papers I received from EDNF, I soon realized that I had to be a Vascular Type. I couldn't relate to the other types when it came to loose joints, stretchy skin etc. My only problems at the time were veins rupturing causing large bruises, and loose finger joints. Also, I had the typical faces of the Vascular Type (IV) what is described as acrogenic type.

In 1994 I had a skin biopsy taken, which confirmed my suspicion. Of course it was hard to accept! That same year I also attended the conference in San Diego, and for the first time I met others with EDS (besides my father). All of a sudden I wasn't alone in this, and it felt great! Coming home I was scared to death, but also very happy to have met a lot of great people.

In 1995, we founded a support group in Norway. I was the board secretary from the beginning. It helps me being able to help others, it's kind of therapy for me. Does that make sense to you guys? I continued to educated myself on EDS, while getting involved with other's suffering from EDS. I attend the 1997 EDS Nosology in France, along with other EDS representatives from other countries.

What I really want to say, is that we all need to take time accepting our EDS, no matter what subtype we have. Other subtypes are lucky they're not a Vascular Type (IV); in a way I feel lucky I don't have to go through all of the problems related to the other types. Compared to most EDSers, my problems are few and far between. The way I see it, at the end it boils down to the same fear for all of us; "when will it end?" (our bodies slowly deteriorating). It's been important for me to find ways to fight the prognosis of the Vascular Type. I've learned to live with the fact that I might get very sick without a moments notice and of course, I'm still scared. It's not fair that I might die young, there are still so many things to do and see. I've learned to live one day at a time, and appreciate the little things in life. One of them being able to have contact with people like you; thanks for being there!

Shaun Michael Bell
6/4/82 to 1/5/96

Shaun Michael Bell

6/4/82 to 1/5/96

By Steve and Chris Bell

On June 4th, 1982 our precious baby boy was born, weighing 8 lbs. 15oz. We proudly named him Shaun Michael Bell. We couldn't possibly imagine that 13 years later our young son's death we would mourn, leaving only memories to cherish and fond stories to tell.

Shaun was always very active throughout his life. He started walking before he was one. He really loved skating and playing hockey and riding bikes. On his 3rd birthday he was so excited to get a bike with training wheels and when he was 12 he was even more excited with his new dirt bike.

Shaun was always busy with his hands. As a young boy he spent many hours playing in the sandbox with his dinky cars and Tonkas. A few years later he was taking apart his remote control cars to clean or repair them. Shaun preferred to keep his hands occupied with Nintendo games, computers and handling his hockey stick rather than turning the pages of a book or holding a pen to do his homework.

Shaun was always trying to make his life as enjoyable as he could and by doing that he made the lives of his family and friends so much more enjoyable. He has left us with so many treasured memories to make us smile.

Shaun died from a spontaneous rupture of his aorta while doing what he loved, playing hockey. Had we known that Shaun had EDS, his lifestyle would have been different, but how happy would he have been?

I was only diagnosed with the Vascular Type (IV) after Shaun's death. As a young girl, I had various tests done, but nobody clued in that I had a connective tissue disorder. My parents were told that I just had thin skin and would bruise easier than other people. So when Shaun bruised easily like me, he just took after his mother. When Shaun was a toddler we had him to the hospital for stitches enough times that they were suspicious of abuse rather than a disorder. Other than the bruising and skin tears Shaun was always healthy. We just didn't imagine that our vessels and organs were weak and a ticking time bomb.

Steve and I got quite a shock when I became pregnant last year. We had tried for 18 years to give Shaun a sibling, but were unable to conceive. We didn't plan this pregnancy, as we knew the risks were just too high. It just happened and we tried not to fear it, but to take it as a true blessing. Shaun's baby brother Everett Steven Bell was born on January 5th, 2001, weighing 7 lbs. 1 oz., on the 5th anniversary of his brother's death. Everett will grow up knowing how very special his older brother was and how much he was loved.

Words on Courage

"You gain strength, courage, and confidence by every experience in which you really stop to look fear in the face. You are able to say to yourself, ' I lived through this horror. I can take the next thing that comes along.'....You must do the thing you think you cannot do."

~Eleanor Roosevelt ~ (1884 - 1962)

" There is courage that springs from battle fever, or from a desperate emergency. And there is a courage that is rooted in the acceptance of a dreadful circumstance, and all that it entails -- a courage that brings sanity and cheerfulness and hope to live that could be utterly consumed by sorrow. This is the courage that endures. This is the greatest bravery. "

~ Pam Brown ~ 1928

(Continued from page 5)

dibular joint (TMJ) symptoms defined at least as pain and clicking (53.5%) (Table 4). In regards to the prevalence of various arthritic signs and symptoms, our analyses indicated that in general, these were not EDS type specific, with the exception that there may be an increased tendency for those individuals of the type III (hypermobile type) to report osteoarthritis (Table 4). This was particularly true for those type III individuals over age 40 where osteoarthritis as indicated by an X-ray change was found in 5/5 or 100% of the respondents. Despite the presence of osteoarthritis as indicated by a x-ray change across a broad range of types, the only individuals under age 40 to report osteoarthritis were those who had experienced one or more dislocations in this same joint (Table 4). Three of the oldest type I (classical) respondents, (ages 55, 64 and 73), did not indicate the presence of osteoarthritis. Together, this data suggests that at least in the Ehlers-Danlos syndrome, that the presence of increased hypermobility and or dislocation seemed to be more significant than age as a risk factor for the subsequent development of osteoarthritis. Of relevance to this, we found the rate of dislocation to be quite high as 29/43 or 67.4% of respondents indicated at least one prior dislocation (Table 4). Overall, in decreasing order of frequency, joints which were reported to be affected by osteoarthritis were the knee (n=9), temporomandibular joint (TMJ) (n=3), hip (n=2), spine (n=1), shoulder (n=1) and hand (n=1). Of those who reported osteoarthritis, 7/17 or 41.2% (with a mean age of 56.4 years) also reported requiring a total joint replacement. Altogether, there were 3 reports from individuals with a single knee joint prosthesis, 3 individuals with a Christenson TMJ prosthesis (including one individual with bilateral implants), and 1 individual with a distal phalangeal index finger prosthesis. There was also one individual who in addition to her index finger prosthesis, had required both an anterior cruciate ligament replacement and a spinal fusion.

In addition to actual osteoarthritis as indicated by an X-ray change, there were also several other types of non-specific musculoskeletal arthritic signs and symptoms that were commonly reported (Table 4). Unlike osteoarthritis, which was associated with hypermobility and or dislocation, no characteristic patterns emerged as these signs and symptoms seemed to be reported by individuals of every type. Somewhat surprisingly however, several individuals (16.2% of the total) did report occasional inflammatory type changes in association with exacerbations of their arthritis including mild elevations of their erythrocyte sedimentation rate, white blood cell count or C-reactive protein, indicating that the various arthritic signs and symptoms associated with the connective tissue defect may have at times, a mild inflammatory component.

Genitourinary type Complications

As mentioned above, a large number of our female respondents over 21 years of age (35.3%) indicated the need for hormone replacement therapy, thus verifying a high rate of hysterectomy in adult women with this syndrome. Problems that may be associated with a need for hysterectomy included the common report of endometriosis (20.6%) or adenomyosis (uterine fibroids) in (32.3%) of the adult women within in our study population.

With special attention to the urinary tract, we also found that there were 6 reports from adults of actual pyelonephritis (Table 5), and one of a "bladder reflux" from the mother of a seven-year-old type I (classical type) female which was associated with cystitis, but not pyelonephritis. Although we requested further information from all the adults who reported pyelonephritis to determine whether there was a common predisposing factor, we did not find one. Reports which we received included three associated with stone formation, one of nephritis associated with lupus erythrematosus, one of a nephritis and recurrent infection associated with type II diabetes, and one with no other association.

Immune Abnormalities

We received no report from any individual with any

known immunodeficiency; however, we did receive one report from a type IV individual of a mildly decreased gamma globulin level, specifically the IgG_2 and IgG_4 fractions. Furthermore, even though our data set was relatively small, our inspection of the data by type of EDS for the presence of certain fungal, bacterial or viral infections at the rate that would be expected if a specific immunodeficiency was present, indicated that none of these types of infections were prevalent enough to raise this suspicion (data not shown, and see below).

General and Post-operative Infections

In Table 5 both the prevalence of general and surgical post-operative infections as analyzed by the presence or absence of a cardiac and or thoracic abnormality are presented. Analyses were carried out on the group of individuals reporting one or more of these types of abnormalities (cardiac, thoracic or both) (n=22) using the no associated defect group as an internal control (n=21) (Table 3). In general, we found a modest trend for the presence of streptococcal associated infections in those who also reported a cardiac or thoracic defect. The only statistically significant association was with those who also reported at least one post-operative type infection (p= 0.0268). However, these types of infections had occurred only in a minority of their procedures.

Allergy, Asthma and Recurrent Respiratory Problems

In regard to the incidence of allergy, asthma, and respiratory disorders, we observed a high incidence of these conditions (Table 6). The most striking group includes those with at least one severe allergic reaction to medication, reported by 20.9% of the total respondents. Only 1-3% of the general population would normally experience this type of reaction (67). Quite unexpectedly, when we carried out the same type of analyses as we performed above on those reporting certain types of infections, we also detected significant differences for those conditions associated with atopy and whether or not there was a co-existing cardiac or developmental defect. We found the most significant differences between the two groups for those reporting at least one severe or life threatening allergic reaction to medication (p= 0.0212), and those reporting allergic induced asthma (p=0.0089) (Table 5). However, no significant differences between these two groups for those reporting other chronic respiratory conditions including intrinsic asthma due to stress, cold or exercise, or recurrent problems with sinusitis, bronchitis, or pneumonia was found (Table 6).

Autoimmunity

There were only three reports of autoimmunity (Table 2). These included one with a systemic lupus erythrematosus (SLE) or mixed connective tissue disease (MCTD) like syndrome in a 38 year old type III female which presented initially as a red cell aplasia and T cell deficiency, one of a Sjogren's syndrome in a 36 year old type II female which presented initially as numerous dental caries, and one of Hashimoto's thyroiditis in a ten year old type I female.

Interestingly, 18.6% of the adult population also reported the association of a flare of their arthritis with an ophthalmologic symptom such as redness or photophobia. However, as the majority of those individuals reporting eye symptoms did not likely have an actual autoimmune disease or inflammatory bowel disease, a more satisfactory explanation for these types of symptoms may be the common use of certain medications (Table 2) or possibly the consequence of a common physiologic mechanism related to the connective tissue defect.

Cancer

No verifiable reports of malignancy were found in our study population. In fact, the only report of tumor formation which we were able to verify was that of a probable benign pituitary micro-adenoma detected by MRI in an adult female with type unknown EDS.

Discussion

Collagen Biosynthesis

Defects in the composition or processing of collagen characterize the collection of connective tissue defects known as the Ehlers-Danlos syndrome. To date, 19 different types of collagen, each with varying compositions, have been described (27, 44, 48, 64) and these can conveniently be divided into the fibrillar versus the nonfibrillar collagens (48). However, in the Ehlers-Danlos syndrome, only mutations in the genes that code for, or the enzymes responsible for the processing of the fibrillar collagens I, III, and V have been definitively linked (20, 30, 34, 42). Theoretically however, as a mutation in any gene that encodes a protein or enzyme responsible for the production, modification, or regulation of a fully functional collagen gene product could also be responsible, we wish to briefly review the common synthetic pathway that all fibrillar type collagens must undergo prior to their incorporation into the various tissue types.

After translation as a prepro precursor molecule in the endoplasmic reticulum, the first step that a precursor fibrillar collagen chain undergoes is an enzymatic hydroxylation resulting in the cleavage of the signal pre-leader sequence to produce a procollagen precursor. The procollagen precursor then exits the endoplasmic reticulum and enters the Golgi, where additional post-translational modifications occur, including the addition of carbohydrates and the linkage of glycine residues. After exiting the Golgi, the procollagen chain is then secreted into the extracellular matrix (ECM) with its secretory pro-leader sequence intact. Once in the ECM, the procollagen is then processed by both amino and carboxy-terminal propeptidases with the resultant cleavage of the pro-leader sequence where the final steps of collagen fibril aggregation, lysyloxidation, and crosslinking occur (25). An analysis on the collagen content of various tissues has also been carried out (24). Type I collagen, constituting approximately 90% of total body collagen, was found in the skin, tendons, bones and internal organs. Type II collagen was a major component of cartilage, and a minor component of intervertebral discs, notochords and the vitreous body of the eye, while type III collagens were found in the skin, blood vessels and internal organs. Types V and XI collagens while minor, only 1-2% of the whole in most tissues, were recently found to be co-expressed in various heterotrimeric forms and broadly distributed (23).

Molecular Genetic Analyses and the New Nosology

As mentioned above, and summarized in Table 1, mutations resulting in either the aberrant expression or processing of fibrillar collagens type I, III, and V have been described in the Ehlers-Danlos syndrome. However, almost every collagen mutation identified has been unique for that one individual or one family (20, 30, 34, 42). For instance, mutations in the a1 and a 2 chains of the COLA5 chain (collagen type V) have now been linked to several pedigrees with the phenotypic characteristics of Ehlers-Danlos types I and II (classical type of EDS) (12, 13, 20, 37, 41, 45, 53, 63, 66) and recent experiments in fact suggest that type V collagen plays a central role in both fibrillogenesis and wound healing (23). However, demonstrating the heterogeneity of this syndrome, one study has excluded COL5A1 as the candidate gene in one pedigree with Ehlers-Danlos type II (28). While the collagen defect responsible for type III EDS (hypermobile type) still remains undefined, mutations associated with EDS type IV (vascular type) have all been associated with defects in the synthesis or secretion of collagen type III (14, 57, 59). The mutation responsible for EDS type V, described in only one family, remains undefined (8, 38).

Other rare types of EDS are also currently recognized; these include the autosomal recessive kyphoscoliosis type, formally known as EDS type VI, associated with defects in lysyl hydroxylase (PLOD), an enzyme which forms stable hydroxylysine crosslinks between collagen molecules (31, 49), the autosomal recessive dermatosporaxis type, formally known as EDS type VIIC, associated with a defect

in the ability to remove the amino-terminal propeptide of type I procollagen (26, 36, 52), and the autosomal dominant arthrochalasia type, formally known as EDS types VII A and B, associated with base substitutions that result in exon-skipping and deletion of the N-terminal cleavage site in the procollagen alpha chains of type I collagen (COL1A1 and COL1A2)(15, 18, 36). However, according to current classification, the recognition of either type VIII (the periodontal type), or type X (the fibronectin type) as distinct entities remains uncertain (5). Additionally, EDS type IX, now known to be caused by a defect in the copper transporting ATPase, ATP7A, has been completely eliminated from the current classification (5, 51).

Cardiac and Thoracic Defects

As shown in Table 3, we found that skeletal thoracic abnormalities such as scoliosis, a straight back, lordosis, kyphosis and pectus deformities were common as 22/43 or 51.2% of our respondents reported this. Three of our respondents also indicated the presence of a combination of these abnormalities. Even though we relied on self-reported data, we also found that our overall cumulative incidence for these types of abnormalities is in substantial agreement with that of 55.6% previously reported in 18 patients with types I-IV EDS (4). Further, our inspection of the data presented in Table 3 did not indicate a higher frequency of cardiac or thoracic defects in any one type of EDS; however, we also note that many of these types of abnormalities are characteristic of connective tissue diseases in general, and therefore are not specific to the Ehlers-Danlos syndrome (58).

In regards to cardiac abnormalities, we found a cumulative incidence of 11/43 or 25.6%. with 9 mitral and 2 tricuspid valve abnormalities reported. Of these, there were two reports of mitral valve repair, one in a 64 year old type I male who also reported a recurrent endocarditis, and a endocarditis related CVA, while the other report of a mitral valve repair was from a 48 year old type II female who indicated no other complications. There was also one report of a tricuspid valve repair in a 53 year old type I female with subsequent bacterial endocarditis who experienced a cerebral embolization directly related to the endocarditis. Therefore, 3/11 of our respondents with a cardiac valve defect eventually experienced a substantial complication(s). We did find however, that of our 11 respondents indicating a cardiac valve defect that only 5 of these had indicated the need for a daily cardiac type medication, and that 3/5 of these were older than 50 years of age. Together, this data suggests that many of the cardiac associated abnormalities that were reported may in fact be relatively mild. In fact, many of our respondents who indicated a MVP were likely diagnosed by the older less strict single M-mode echocardiographic criteria and if re-evaluated under the current stricter criteria might not be found to have MVP (21). It has been suggested however, that patients with EDS should be evaluated by serial echocardiography as a significant enlargement of the aortic root was recently detected in five patients with EDS types I, II and III (62).

Musculoskeletal Complaints and Osteoarthritis

In this report, we have found evidence validating the assumption that there may be an increased risk for "wear and tear arthritis" in this population(35). We also found evidence that non-specific arthritic signs and symptoms were common, and that in general these were not EDS type specific, although type III or hypermobile individuals may be more susceptible to actual osteoarthritis as determined by an X-ray change (Table 4). We also suspect from the majority of reports that we received that pain management and possibly associated problems with depression might be problematic in this population. In fact, pain relievers and tranquilizers/anti-depressants/anti-anxiety type medications were one of the two most frequent types of daily medications reported as being used in this population (Table 2). Although this data must be interpreted with caution as tranquilizers/anti-depressants/anti-anxiety type medications are also commonly prescribed as sleep

aids, a significant association with depression, anxiety disorders and the presence of mitral valve prolapse or joint hypermobility syndrome have recently been reported (10, 16, 29).

Various soft tissue complaints that may be associated with micro-trauma such as tendinitis and bursitis were also common, as well as what seems like a very high rate of temporo-mandibular joint symptoms as 23 of the 43 or 53.5% of the respondents reported TMJ associated pain and/or clicking. This in fact may not be too surprising, as a high rate of TMJ symptoms have previously been reported in hypermobile individuals (9), and TMJ complications have been reported in the Ehlers-Danlos syndrome (39, 43, 46). However, to our knowledge, this is the first report to actually document the actual incidence of TMJ symptoms in this population, as well as the actual frequency of severe TMJ associated osteoarthritis in that 3/43 or 7.0 % of the respondents indicated actually requiring a TMJ joint prosthesis.

We found that in general, that the presence of increased hypermobility and/or repeated dislocation seemed to be the most significant risk factor for the subsequent development of osteoarthritis as type III (hypermobile) individuals reported the highest incidence of osteoarthritis (Table 3). We also found that type II (classical type) (n=8) individuals who were thought to be mild in their EDS symptoms, as they do not show skin hyperextensibility, did not appear to be spared in the incidence of any of the major arthritic signs and symptoms, and except for type III (hypermobile type) (n=9) respondents reported the highest rate of arthritic signs and symptoms in every single category (Table 3). Paradoxically, the type I respondents (n=11), who all also reported having hyperextensible skin, seemed to have the fewest arthritic signs and symptoms overall. Although the cause of fibromyalgia is as yet unknown, it is suspected to be associated with disturbed sleep patterns (1), and the relatively high incidence of fibromyalgia found in the EDS population would be consistent with this as disruptions in sleep patterns has previously been reported as being a common problem in EDS (55).

Bleeding Tendencies and Coagulation Abnormalities

In agreement with several other reports (3, 19, 47), we also found that deficiencies of coagulation factors can sometimes occur in EDS, but suspect that the actual incidence of coagulation factor deficiencies in this population may actually be quite low as only 2 of the 43 respondents spontaneously provided this information. Furthermore, these deficiencies, one a factor VII, and one factor XI deficiency were seen to be quite mild, and had not led to any serious bleeding events. Our observations are in agreement with other reports (33, 61) which propose that the observed bruising tendency in many patients with EDS is likely due to the associated connective tissue defect and problems with vascular integrity and or wound healing rather than a primary coagulation factor deficiency. If however, in any given EDS patient, clinical lab studies do find evidence for a coagulation factor deficiency, and a surgical or invasive procedure is being contemplated, then treatment would be empirical based on the type of deficiency found, but prophylactic 1-deamino-8-D-arginine vasopressin (DDAVP) has also been reported as being beneficial in preventing the bleeding complications sometimes seen in EDS (54, 61).

Infections, Allergy and Asthma

In specific regards to the immune system, and in agreement with data on the incidence of post-operative surgical infections in EDS patients recently collected by Weinberg and MacFarland (65), we did find evidence for a slightly higher prevalence of post-operative surgical infections (Table 4). However, as we could detect no specific immunodeficiency, it is possible that the majority of post-operative infections were related either to the presence of an abnormal mechanical factor such as tissue dehiscence, or a physiological factor associated with the connective tissue defect such as an abnor-

mality in wound healing. Furthermore, although a large number of respondents (37.2%) indicated at least one post-operative infection, the majority of respondents also indicated that they had only occurred in a minority of their procedures. Another type of infection, pyelonephritis, was more frequent than expected in this population. It is possible that the root cause of many urinary type of infections is mechanical, not immunologic in nature as bladder distension, diverticulae, and even rupture have (7, 11, 17, 22, 32, 56) previously been reported in EDS. Lastly, since we documented four episodes of bacterial endocarditis, it would seem prudent to advise routine antibiotic prophylaxis in all patients with EDS who have a cardiac valve abnormality prior to their dental or surgical procedures.

Quite unexpectedly, when we also analyzed the prevalence of allergy and or asthma by the presence of a cardiac or skeletal thoracic abnormality we also found an association with the presence of these abnormalities and those conditions previously associated with the presence of atopy, that of allergy in general (p=0.0546), severe drug allergy (p= 0.0212), and extrinsic asthma (p= 0.0089) (Table 5) (50). Although this was in a smaller series, our results are also in agreement with those presented by Ayres et al. (4), who also found an approximately two-fold increase in allergy as revealed by skin testing in those patients with EDS who had a pre-existing skeletal or thoracic abnormality. Explanations for this phenomenon may be associated with the thoracic defects in our respondents (Table 3) which can lead to mechanical and physical factors such as reduced expiratory air volumes, abnormal air flow, or reduced cilary clearance of allergens. However, if this was entirely the case, one would expect the presence of intrinsic asthma due to mechanical or physical factors to be positively associated with the presence of a thoracic defect, yet no difference was found between the two groups for this parameter (p=0.4121)(Table 5). Our results suggest that pulmonary and immune function studies to further confirm the presence of atopy with a cardiac or thoracic skeletal abnormality in a more objective manner is warranted.

Autoimmunity

We found no evidence for an increased prevalence of autoimmune disease, as we received only three confirmed reports, including one of Sjogren's syndrome, one of a systemic lupus erythrematosus (SLE) like or mixed connective tissue disease (MCTD) like syndrome, and one of a Hashimoto's thyroiditis thought to be inherited from the non Ehlers-Danlos parent. Although a relatively high percentage of respondents (18.6 %) also reported the presence of eye symptoms including redness, pain, and photophobia which they thought was associated with their arthritis, a physiological explanation for this observation including the common use of certain medications or possibly a common physiologic condition related to the connective tissue defect may be more likely.

Summary

In conclusion, we found that various non-surgically related complications related to the musculoskeletal, cardiac, gastrointestinal, and genitourinary tract systems were common in the EDS population. Our results also suggest that many of these complications were likely directly related to the connective tissue defect. Further, although we found that most of these complications were relatively benign in nature, and compatible with a long life, recognition of the connective tissue defect and how this may contribute to the presenting sign(s) and or symptom (s) could result in an improved therapeutic outcome for patients with EDS. We also found that although patients with type III or the hypermobile type of EDS had evidence for an increased rate of actual osteoarthritis, that the treatment of pain associated with every type of EDS was often problematic. We also believe that more objective research into the possible causes of the association which we found between EDS and gastrointestinal problems, particularly GERD and IBD, as well as the possible association that we found between the condition of

atopy, particularly atopic asthma and the presence of either a cardiac or skeletal thoracic defect will be necessary to understand the mechanism(s) behind this association.

Primary care physicians as well as specialists who have patients with EDS should be aware of the various types of non-surgical complications that we found in this syndrome, as early recognition of these complications, and how a connective tissue disease could contribute to their development may result in an improved therapeutic outcome for their patients. Individuals with the phenotypic manifestations that can be seen in EDS should also have a genetic evaluation to confirm their diagnosis and determine which type of EDS they have.

Table 1. The New Nomenclature in the Ehlers-Danlos Syndrome:Correlation of Genotype with Phenotype

Former	**New**	**Genotype**	**Phenotype**
Gravis (EDS type I) Mitis (EDS type II)	Classical Type	COL5A1 & COL5A2 (ref: 14, 15, 22, 44, 49, 54, 64, 75, 78)	Variable skin hyperextensibilty, widened atrophic scars (tissue fragility), joint hypermobility, easy bruising, ± mulluscoid pseudo-tumors, and subcutaneous spheroids
Hypermobile (EDS type III)	Hyper-mobility Type	Unknown	Generalized joint hypermobility, skin involvementincluding hyperextensibility and/or smooth, velvety skin, chronic joint pain, and recurrent joint dislocations
Vascular (EDS type IV)	Vascular Type	COL3A1	Thin, translucent skin, arterial/intestinal/uterine fragility or rupture, extensive bruising, characteristic facial appearance, ± acrogeria, hypermobility of small joints, early onset varicose veins, and pneumothorax
Ocular-Scoliotic (EDS type IV)	Kyphoscoliotic Type	PLOD	Generalized joint laxity, severe muscle hypotonia at birth, scoliosis at birth which is progressive, scleral fragility and rupture of the ocular globe, ± tissue fragility, easy bruising, arterial rupture and osteopenia
Arthrochalasis multiplex	Arthrochalasia Type	COL1A1 & COL1A2	Severe generalized joint hypermobility with recurrent congenita (EDStypes COL1A2 subluxations, congenital bilateral hip dislocation, VIIAand VIIB) (ref: 17, 20, 43) skin hyperextensibility, tissue fragility (atrophic scars), easy bruising, muscle hypotonia and osteopenia
Human Dermatosparaxis	Dermatosparaxis Type	deficiency of procollagen 1 N-terminal peptidase	Severe skin fragility, sagging redundant skin, ± soft doughy skin texture, easy bruising, premature rupture of fetal membranes, and large hernias (umbilical and (ref: 31, 43, 63) inguinal hernia
	Other Types		

Current classification of EDS based upon the revised nosology (5). [b]The corresponding genetic mutation if known, with relevant references provided underneath. [c]The major phenotypic manifestations of each type according to the current nosology, with minor diagnostic criteria prefaced by a ± sign. [d]The definate classification of other rare forms of EDS which have previously been described including X-linked EDS (EDS type V), the Periodontitis type (EDS type VIII) and Fibronectin-deficient EDS (EDS type X) await further molecular definition before inclusion. The occipital horn syndrome (EDS type IX), and the benign familial hypermobility syndrome (EDS type XI) have been removed from classification (5)

Table 2. Medication Use by Category and Frequency			
Daily Medications ([a])		**Immune Modulators ([b])**	
NSAID Type Pain Releiver	19/43	Cortisone Type Injection	19/43
Non-NSAID Type Pain Reliever	8/43	Prophylactic Antibiotics	
Narcotic Type Pain Reliever	3/43	- for cardiac valve	11/43
Anti-Reflux Agent	14/43	- for joint prosthesis	5/43
Anti-Depressant/Anxiety/Tranquilizer	14/43	Oral Steroids (prednisone)	
Allergy Medication	6/43	- for allergy	2/43
Asthma Medications (inhalers)	3/43	-for asthma	5/43
Herbal or Vitamin Supplement	5/43	-for joint pain	3/43
Muscle Relaxant	3/43	-for autoimmune disorders	3/43
Thyroid Supplement	5/43	Other Immune Modulators [d]	
Hormone Replacements [c]	12/34	Mesalamine	3/43
Calcium Channel Blocker	2/43	Methotrexate	2/43
Blood Thinning Agent	2/43	Chloroquine	1/43
ß-Blocker	1/43		
Anti-Glaucoma Agent	1/43		
Osteoporosis Modifier	1/43		

a.The number of respondents who reported the use of various medications by category. [b]The non-daily use of immune modulators, both injectable and oral were also occasionally reported. [c]Women respondents over the age of 21 (n=34). [d]The use of other types of daily immune modulators were also occasionally reported: 2 reports of mesalamine for Crohn's disease, 2 reports of methotrexate (including one for Crohn's disease and one for costochondritis), and 1 report of the use of chloroquine for a systemic lupus or mixed connective tissue disease like syndrome.

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Table 3. Demographic Characteristics of Survey Respondents by Type and the Presence or Absence of a Cardiac and or Thoracic Defect

	I	II	III	IV	VI	Unknown	Total
Males [a]	-	3	-	-	-	2	5
Females [b]	8	8	9	2	1	10	38
Mitral Valve Defect	1	3	4	-	-	1	1
Tricuspid Valve Defect	1	-	1	-	-	-	2
Scoliosis	3	4	2	-	1	2	12
Straight Back	-	1	1	-	-	1	1
Kyphosis and Lordosis	1	-	1	-	-	-	1
Pectus Deformity c	2	1	3	-	-	-	6
No Abnormality	7	2	1	2	-	9	21

[a]The age range for male respondents was 28-72, and [b] for female respondents was 7-73. [c]Pectus deformities reported included 4 reports of "funnel chests"and 2 reports of "barrel chests."

Table 4. Arthritic Signs and Symptoms by Type of Ehlers-Danlos Syndrome

Sign or Symptom	I	II	III	IV	VI	Unknown	Total %
Bone Spur	2/11	5/8	3/9	0/2	1/1	5/12	37.2
Bursitis	6/11	5/8	6/9	1/2	1/1	8/12	62.8
Costochondritis	1/1	4/8	3/9	0/2	0/1	2/12	23.2
Dislocating Joint	6/11	8/8	7/9	1/2	0/1	7/12	67.4
Fibromyalgia	2/11	3/8	3/9	0/2	0/1	1/12	20.9
Neuritis	1/11	5/8	4/9	1/2	1/1	4/12	37.2
Osteoarthritis <40	1/5	0/5	2/4	0/1	0/0	1/6	19.0
Osteoarthritis >40	3/6	2/3	5/5	0/1	0/1	1/6	50.00
Plantar Faciitis	1/11	2/8	3/9	2/2	0/1	3/12	20.9
Spondylitis	1/11	2/8	2/9	0/2	0/1	0/12	11.6
Tendonitis	2/11	3/8	6/9	0/2	1/1	7/12	44.2
TMJ [c]	3/11	7/8	6/9	0/2	0/1	7/12	53.5

[a]Neuritis was defined as the presence of numbness or tingling in any extremity not caused by sitting or lying down. [b]The presence of osteoarthritis in those age 40 and under and those over age 40 was scored as positive only if an x-ray change was present. [c]TMJ= Temporomandibular joint symptoms that were defined at least as the presence of pain and or clicking.

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Table 5. Incidence of General Infections by the Presence or Absence of Cardiac and or Thoracic Defect

Type of Infection	No Defect (n=21)	Defect (n=22)	p-value [a]	Total %
Endocarditis	0/21	2/22	0.4884	2.7
Post Operative	4/21	12/22	0.0268	37.2
Ear Infection(s)	5/21	9/22	0.3319	32.5
Strep Throat	1/21	6/22	0.0946	16.3
Oral Thrush [b]	1/21	1/22	1.0000	4.6
Bladder	5/21	6/22	1.0000	25.6
Kidney	4/21	2/22	0.4121	14.0
Yeast [c]	4/21	10/22	0.1040	32.6
Other Fungus	0/21	4/22	0.1078	9.3
Viral	4/21	7/22	0.4876	25.6

[a]See materials and methods for derivation, no significant difference if p>0.05. [b]Oral thrush not associated with diabetes or the use of steroids. [c] Not including oral thrush, these infections were often associated with previous antibiotic use.

Table 6. Incidence of Allergy, Asthma and Recurrent Respiratory Problems by the Presence or Absence of a Cardiac and or Thoracic Defect

Condition	No Defect (n=21)	Defect (n=22)	P-value	Total %
Allergy [b]	4/21	11/22	0.0546	34.9
Severe Drug Allergy [c]	1/21	8/22	0.0212	20.9
Intrinsic Asthma d	2/21	5/22	0.4121	16.3
Extrinsic Asthma e	0/21	7/22	0.0089	16.3
Sinusitis	8/21	14/22	0.3880	51.2
Bronchitis	5/21	10/22	0.2027	34.9
Pneumonia	1/21	5/22	0.1853	14.0

[a] See materials and methods for derivation, no significant difference if p>0.05. [b]Allergy was defined by a positive skin or blood test to any allergen, including contact, food and respiratory type allergens. [c]A severe allergic reaction was defined as one or more anaphylactic type reactions with angioedema, or severe dermatitis. [d]Intrinsic asthma was defined as asthma induced by stress, cold, or exercise. [e]Extrinsic asthma was defined as allergic asthma.

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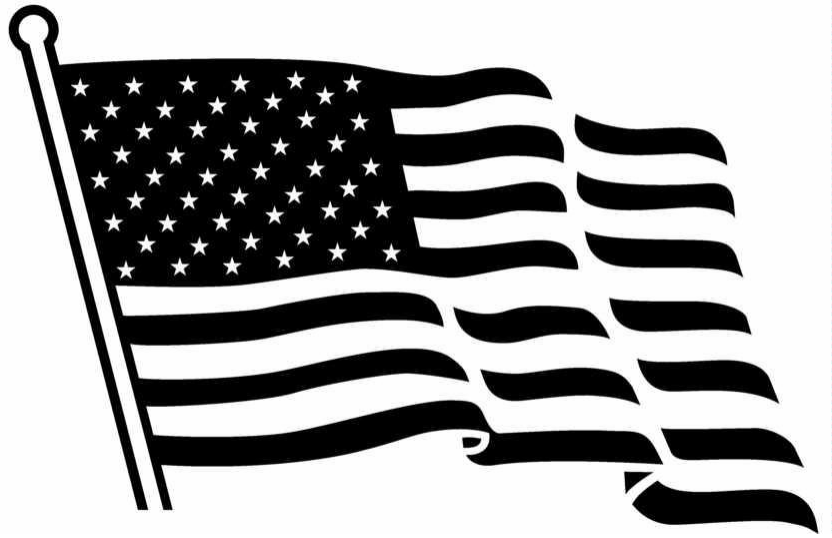
We will never forget.



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