

FSH Watch

A publication of the Facioscapulohumeral Muscular Dystrophy Society
Connecting the community of patients, families, clinicians and investigators

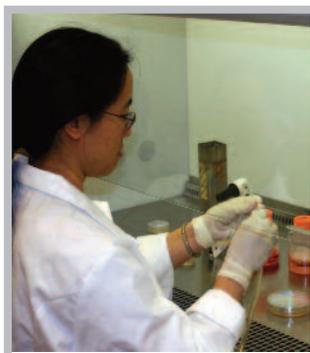
Thank you!

Festive Evening of Music & Song raises over \$100,000 for FSHD research

See page 8

First muscle biopsies arrive at the NIH BBRI Senator Paul Wellstone FSHD Center!

Jennifer Chen, Ph.D., processes muscle stem cells from muscle tissue, obtained from the first facioscapulohumeral (FSHD) patient and family member who volunteered to provide muscle samples. Dr. Chen is a Wellstone investigator at Boston Biomedical Research Institute (BBRI).



Jennifer Chen, Ph.D.,
 Wellstone investigator

The tissue bank at the NIH BBRI Senator Paul Wellstone Muscular Dystrophy Cooperative Research Center will store muscle cells from pairs of individuals, called a cohort. Each cohort contains biopsies from two different places (shoulder and upper arm) from a person affected with FSHD, and two different biopsies from the same matched places from a close genetically related relative (usually a sibling or parent). Two sets of matched pairs are acquired, four biopsies in total.

Muscle cells isolated from each biopsy can be reproduced many times over, providing material for experiments performed by multiple research

labs. The tissue cells remain paired throughout the studies, the FSHD-affected individual with the relative, so that familial and genetic differences can be observed.

Muscle tissue cells are needed by investigators around the world in their search for treatments and cures for FSHD. For example, a researcher takes a first step in experimenting with potential treatments by

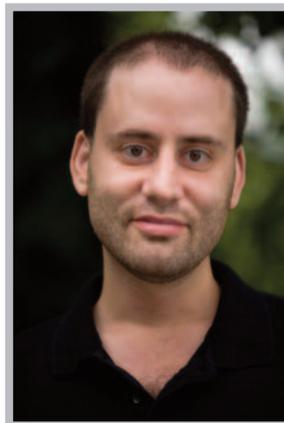
Living with FSHD:

My road to developing a therapy service for people with physical illnesses and their partners and family members

By David Younger, Ph.D.

Coming to terms with having a degenerative physical illness is a lifelong work in progress. I have known that I have FSHD since I was a little boy. I was able to do everything other kids my age could do. I acted like it was not a part of my life. In my teenage years, I saw my mother's health slowly decline. I remember telling my friends that the stair-assist chair installed in our house was to help my parents bring their books up and down the stairs as they are renowned rare book dealers. I was in denial. I did not want to talk about it, see it, or hear about it. Because my symptoms were quite benign through high school, I was able

to ignore, at least on the surface, what was happening to me and to my mother.



David Younger, Ph.D.

The first non-family member that I told about the FSHD was my best friend, Eric. We were in college and FSHD was becoming more and more a part of my consciousness. I could not ignore it any longer. I felt alone in the world, like people did not know the real me because they did not know about this huge secret that I was keeping. I was hyper conscious about hiding FSHD, and I never put myself in situations that would reveal it. Little by little, I started

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testing them on FSHD muscle cells growing in a dish. Promising treatments and drug compounds are then investigated in animal models. If the experiments support further development, the drug compound might go to a clinical

trial including persons affected with FSHD.

The tissue bank hopes to receive muscle biopsy material from fifteen cohorts — fifteen FSHD-affected individuals and fifteen relatives each year.



Muscle tissue cells



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Individuals with FSHD share their work life and their family life



Nancy Van Zant

Dear Friends,

I know you will find the articles by Alan, David, and Lin in this issue of the FSH Watch both inspiring and helpful. We often hear from people with FSHD, and their parents, who have concerns and despair about raising a family, building a career and coping with a progressive disease. These are constant challenges for people in our FSHD community.

The stories contributed within are heartfelt and poignant. Please share these experiences with a young person who faces these questions. He or she just might become a nurse, a producer, or a clinical psychologist!

Sincerely,

Nancy Van Zant
 Executive Director

What's behind the smile?

By Lin McGee McClay, R.N.

Lin is a nurse at St. Mary Medical Center in Galesburg, Illinois; she is also a trauma nurse specialist (TLS), and certified in advanced cardiac life support (ACLS) and pediatric advanced life support (PALS).

When I was a little girl, I dreamed of becoming a mom and a nurse. My childhood was active and happy with school, bike riding, Girl Scouts, baseball, tap dancing, baton twirling and many friends. My dolls always got the best loving care! I occasionally got teased about my "crooked smile" or my "big lips" and my parents would console me with "You just have the McGee mouth." How prophetic those words would become.

After high school graduation my hard working parents and my part time jobs helped me to achieve my dream of becoming a nurse. After I earned a nursing degree in the early 1970s, I headed for the Emergency Department and never looked back. As of June 2009, I will have worked in the ED Level II Trauma Center for 38 years. I love the variety and learning something new each day. I thrive on taking care

of people at their most vulnerable times— all ages and all conditions.

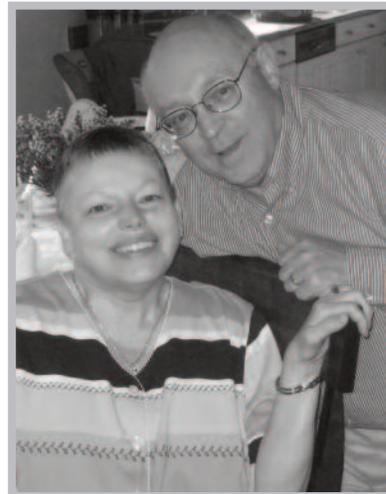
I married in 1974, and my other dream of becoming a mom was not so easily

obtained—at least not by the conventional method. Unable to have children, my husband and I became foster parents to over 200 children in a 12-year span. As a bonus, we adopted eight of these children, raising seven boys and one girl. I always had that desire to care for and to love kids especially in their time of greatest need.

I was very busy with work and a house full of kids, and had little time to care for myself. It seemed

I was always tired, tripped a lot, had to drag myself up the stairs, and everything seemed too heavy to lift or carry. I compensated as best I could and went on.

Soon I noticed that my legs felt like lead walking from the parking lot into work each day. I was on my feet a lot and being



Lin and John McClay, April 2009

What's behind the smile?

the “self diagnostician” that nurses are so proficient at, I decided I must have heart failure. I was convinced but too busy to go to the doctor.

In 1988, my husband of 14 years announced one morning that he was moving to New York and was leaving the next day with four of the older kids who were in high school. Papers were quickly signed, and he and the boys left early the next morning with a few worldly goods packed in an old van and a fishing boat. I was numb as I stood in front of a big old house with my two youngest (ages 8 and 9) who were asking when their dad would be home. The answer to that would be NEVER. Time to pick ourselves up, dust ourselves off, and move forward!

I put my growing list of symptoms on hold for the next six years while we moved, worked, established ourselves in a new church, found new great friends, and I organized my life to head a single-parent family. My own parents were both still alive and were unwavering in their support, as well as my older adopted son, Kevir, who was always available for the kids or me.

In the early 1990s, I met a wonderful Christian man who was first an acquaintance, then a good friend. In 1997, we married and my husband John and I were able to blend our families. For the first time in my adult life, I was able to experience the joy of a loving marriage.

John was concerned about my worsen-

ing symptoms, but I was unwilling to face any news that might not be good. Finally, I accepted my husband's urging that I go to the doctor. My internist discovered my bilateral foot drop and an elevated CPK. He referred me to a tertiary care facility. Many tests later there were still no answers except that “something was wrong.” As I awaited a referral to Iowa City, I looked up every conceivable diagnosis—or so I thought.



*Lin McGee, age 5
Lin found this picture recently and was surprised to see her strong facial symptoms at an early age*

Upon arriving at the University of Iowa Hospitals and Clinics, we had a diagnosis ten minutes after the attending physician and his intern entered the room. “Can you whistle?” “Can you drink through a straw?” There was a quick exam of my scapular regions and the muscle wasting on my arms, hands and legs (not to mention my protruding little belly). The doctor looked at my husband and at me and announced that he

would do a genetic test, but that he was almost sure I had FSHD.

RELIEF—I did have something! I was not a hypochondriac! FEAR! What the heck is FSH muscular dystrophy? And the all important WHY ME? The genetic results arrived in December 2004, and I began my ongoing adjustment and learning curve. My life did change but not necessarily for the worse.

Over these past years I have learned a lot about my disease and some of what to expect. I have taught numerous family, friends, and coworkers about my disease

and the need and reasons for some of my accommodations. I continue to work and have learned the hardest lesson for me—to be able to accept help when I need it instead of always taking care of everyone else. I have learned that my father's “McGee” mouth and the difficulties in ambulation and strength that he experienced for years before his death at 80 were the result of that secret gene that none of us even knew existed. I have reviewed old family pictures and can see evidence of FSHD in many family members on my dad's side.

Attending the FSH Society International Patient and Researcher Network Meeting in Iowa last summer with my sister was a wonderful opportunity for me to actually see and talk to others with FSHD. Until that day, I had never seen another person with FSHD except in pictures. Although I could see that some people appeared less afflicted than me, there were many others who appeared to have more problems than I did. And this experience presented an opportunity again for me to count my blessings and to emulate the strength and the hope that I saw there.

I have been blessed with a wonderful husband who exhibits unconditional love as he sacrifices for me everyday—we work—we travel—we plan—we move forward. We even dance a little—not too pretty on my part, but still wonderful! I have a devoted sister, children, grandchildren, family and friends who support me yet understand my need to be a mom and a nurse. I am fortunate to be able to have a job that I love and still be able to continue to work. (Ankle-foot orthotics and stubborn determination work their wonders on most days.) My prayer is to touch at least one life each day and to hopefully let God use me to encourage others to keep going when they want to give up.

My husband and I have a strong faith life. We begin each day with devotions and a prayer. We believe our relationship with Christ will bring us through each day. We pray for more research and knowledge that will treat and cure FSHD for all of us now and into the future.

I can do all things through Christ who strengthens me. Philippians 4:13

It is the editorial policy to report on developments regarding FacioScapuloHumeral Muscular Dystrophy (FSHD), but not to endorse any of the drugs or treatments discussed. We urge you to consult with your own physician about the procedures mentioned.

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David Younger: Living with FSHD, *continued from front page*

telling more people that I had FSHD and what it entailed.

When I was 22, a college grad, working in marketing for a family business, and living in New York City, I decided to go into therapy. My college years were an emotional roller coaster and even though I had a job, a girlfriend who loved me and a great place in the city, I was battling depression. The physical symptoms were slowly and steadily progressing, and I had to deal with the fact that this illness was a part of my life no matter how hard I tried to deny it.

My first therapist, Linda, was incredibly warm and nurturing and supportive. She shared with me at some point in the process her own struggle with an illness. She created a safe place for me to begin to explore and connect with what I was experiencing. I learned fairly quickly that the business world was not for me. What fascinated me the most about my marketing job was the interpersonal dynamics. I decided to join a research team at New York University (NYU) to work on a psychology research project. I enrolled in a master's program in psychology a few months later.

After completing the master's degree at NYU, I moved to London, England, to further my studies. I started a one-year full-time intensive master's degree in child psychology at the Anna Freud Centre in the picturesque streets of north London. I remember never sitting by the chairs that were stacked in the corner of the room because I knew that it was often necessary to pass them down if students needed more seats. In large part, I was still living in avoidance of myself.

I began psychoanalysis with another amazing woman. Trudy was more like my London mother to me, definitely not a traditional analyst. Trudy helped connect me to the place that I did my clinical training and to my Ph.D. thesis advisor. When I fell in love with my wife, Debbie, Trudy told me where I needed to go to buy an engagement ring. More importantly, she helped me process the fear and anxiety and anticipation that I had regarding telling Debbie about FSHD. I did a tremendous amount of work with her, integrating FSHD with my

self representation in a way that the FSHD became less and less a parasite that was outside me, and more a part of who I was and who I had become.

Debbie and I were classmates in the master's program at the Anna Freud Centre. We started dating a few months into the program. When I eventually told her about FSHD, she clearly could not have had any idea about all that it entailed. I was relieved and elated when she told me she loved me no matter what, and I did not wait long to propose. We were engaged before the program ended. It made studying for finals a near impossible task.

We were married a year after we got engaged. A year later, Debbie was pregnant with our son Eric, who is now 4 ½. When I knew I was going to become a father, I became acutely aware of the things I might

not be able to do with him. I saw dads in the park playing soccer with their kids and immediately questioned my capacities and limitations. I knew that there would inevitably be activities I could not do with Eric that other dads could, but I also knew that I would bring much to our relationship that is unique and special.

Debbie, Eric, and I moved to New York in December of 2005, with a brief stint in Lima, Peru, while I was completing my doctoral dissertation. A year after we settled in New York, I became very depressed. I had had a rough few years physically and it was becoming increasingly difficult to climb stairs. Once again, FSHD simply refused to be a compliant partner in my life. It was breaking me down and I was sinking fast. A friend and colleague of mine referred me to my current therapist, Gerry.

Group Therapy Services of New York provides customized treatment

At Group Therapy Services of New York (GTS-NY), our priority is to customize the treatment for the individual and not the other way around. We provide individual, couple and group therapy; and if appropriate, we will at times offer a combination of these modalities. We emphasize group treatment because we passionately believe in the effectiveness of group to heal and bring about lasting change. We specialize in a number of different concerns; you can read more about these specialties and several of our specialty groups on our website at www.gtsnewyork.com. Above all, we value the creative and growth potential of every human being. We believe that change is possible no matter how entrenched one may be in a given situation. Please take a few minutes to look through our site. We encourage you to give us a call to find out more.

About the directors...

David Younger, Ph.D., P.C., is a New York state licensed psychologist. In addition to having completed a master's degree in psychology from New York University, a master's degree in child psychology from the Anna Freud Centre in London, and a doctorate in psychology from the University of London, Dr. Younger is a trained couple (Tavistock Centre for Couple Relationships in London) and group psychotherapist (Eastern Group Psychotherapy Society in New York). Dr. Younger also works as a couple therapist at Advanced Cognitive Therapy of New York and as a clinical psychologist at Dr. Robins and Associates doing trauma work in English and Spanish.

Debbie Radzinsky, M.Sc., is a Peruvian born therapist and trained group therapist. Ms. Radzinsky also has a master's degree in child psychology from the Anna Freud Centre in London, a postgraduate diploma in working with adolescents from the Tavistock Institute in London and a postgraduate certificate in group psychotherapy from the Eastern Group Psychotherapy Society. Ms. Radzinsky is an expert in parenting, infertility, and adolescence.

David Younger: Living with FSHD

FSHD not only impacted my body, but it also impacted my mind and spirit as well. In an effort to find a balance, I have explored many different options in addition to psychotherapy. Individuals who have influenced me and FSHD the most, in addition to my therapists, have been Meir Schneider, a physical therapist in San Francisco, California, who specializes in working with people with FSHD, and Dr. Manik Hiranandani, a doctor in the South of India who works holistically with people with chronic and incurable illnesses.

I completed group therapy training shortly after moving to New York. It was by far the most excited I had been about my work. I was in a therapy group for two years as part of my training in London, and I was in another during my group training in New York. I have been running a therapy group for men with long-term HIV diagnoses for over two years. It has been one of the most enriching aspects of my career as a psychologist and has inspired Debbie, who is also a therapist, and me, to create a group therapy service specializing in working with people with physical ill-

nesses as well as with partners of people with physical illnesses and their family members. Integrating FSHD in our lives in a way that is meaningful and inspiring is an accomplishment that has come on the back of really hard work.

Group therapy is so effective because it is not only the therapist that assists in the change process, rather, every member of the group. The environment of group therapy affords the unique opportunity to explore and understand who we are in the context of others. It is natural and inevitable that one person's struggle will serve as another person's valuable lesson. In group, we learn to see, experience and work through love, pain, grief and shame alongside others and even through the eyes of others.

If you are shy or it is hard for you to open up to others, as long as you are willing to engage, group therapy is the ideal place to work on these and other issues because it is a safe and contained environment designed to promote exploration and growth.

Debbie is my partner in this therapy

service, and this further provides a unique combination of perspectives. Just as I am inevitably in tune with the physical, psychological and emotional aspects of living with FSHD, Debbie is in tune with all that is involved with being a partner of someone with FSHD. I have seen and personally learned that my psychological and emotional well-being is definitely not inversely proportional to my physical well-being.

Even though I have many more physical challenges today than I did ten years ago, I am much more at peace today. I am more optimistic, open, and full of life. Living with FSHD brings challenges with every day. It is crucial to be attentive to one's needs and to be proactive in creating a support network. Our therapy service, Group Therapy Services of New York (GTS-NY), was created as an effort to help others help themselves. If we can do it, others can too. Our goal is to help as many people as we can who struggle with living with physical illnesses lead more fulfilling lives.

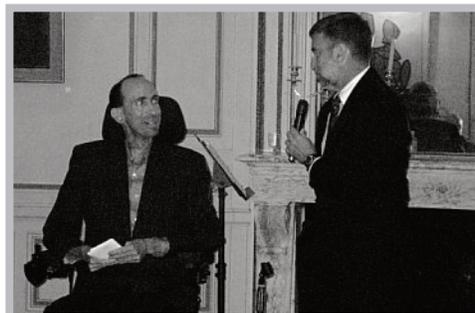
Tango and Comedy Night



Tango dancers

Members of the FSH Society and aficionados of comedy and tango gathered at the Embassy of Argentina, Washington, D.C., on April 17, to support FSHD research in Argentina and around the world. Manuel Gomez chaired the evening which included the comedy of Brett Leake, tango singing by Claudia Gargiulo, and

music and live tango by Tango Reo. The audience also enjoyed tapas and wines from Argentina. Thanks to everyone who contributed to the evening, including Martin Cerda, Amber Rasmussen and Sarah Lamb.



Brett Leake and Manuel Gomez



Manuel Gomez and daughter, Alina

Photos courtesy of *Beth Johnston*

How the FSH Society Makes Discoveries and Progress that Help You!

By Daniel Paul Perez

The FSH Society is without peer in initiating new research ideas in FSHD, in making scientific discoveries about FSHD, in bringing new scientists into the field of FSHD, and in generating knowledge about and data for FSHD. We work quietly behind the scenes and get the job done on a shoestring budget. The FSH Society board, scientific advisory board, staff and volunteers are unparalleled in getting the job done and making continual

progress thanks to your dollars and support. I can think of no other organization that has such a remarkable track record of pioneering the advances in the field of FSHD and the growing knowledge of our own disease. There are many other organizations that seek to emulate the FSH Society on a local level, with whom we are happy to work and always extend our wishes to do so.

About ten years ago, with the help of Dr. Stekly, we brought non-invasive technologies to clinical research of FSHD. At that time, using MRI/MRS for dystrophy research was not widely entertained. Thanks to the generosity of the **Sam E. and Mary F. Roberts Foundation** we have initiated multiple projects using MRI/MRS and oral creatine supplementation to study biomarkers and further develop clinical trial endpoint markers in FSHD.

Thanks to Dr. Stekly's meeting with Dr. Heerschap, of Nijmegen, the Netherlands, and the Sam E. and Mary F. Roberts Foundation, the Society initiated nutrition research grant FSHS-SMRF-003, a research project that has resulted in a paper that describes a new MRI/MRS imaging method to provide biomarkers for clinical trials and to evaluate muscle damage in FSHD.

Researchers in Nijmegen, the Netherlands, theorized that quantitative assessment of MR imaging observations found in previously funded FSH Society

Sam E. and Mary F. Roberts Nutrition Research projects on MRI by Dr. Graham Kemp at Liverpool, England, could provide an objective non-invasive biomarker for individual muscle involvement in FSHD. Drs. Hermien Kan and Arend Heerschap developed a new MRI method to "quantitatively separate muscular and fat content in

different muscles of the lower leg in healthy volunteers and patients with FSHD, using differences in T2 relaxation times of fat and muscle tissue, to provide an objective biomarker for individual muscle involvement in FSHD."

The researchers acknowledged the Society's role in developing this much needed non-invasive tool and, thanks to initial funding by the FSH Society, they have been able to perform the scientific work described in this publication. It describes a new imaging method to evaluate muscle damage in FSHD. They have recently acquired substantially larger funding from the Dutch Prinses Beatrix Fund to continue with this work. This research helps us to understand the difficulties in FSHD research and how major differences can occur within the same muscle. Findings showed that muscle involvement was non-uniform even within one muscle, indicating that MRI can be used as a valuable tool to study pathophysiology and therapy effects in FSHD. Researchers also found considerable differences in the degree of involvement within one muscle and that the degree of involvement was larger at the distal end. This observation of non-uniformity over the muscle provides important clues for the patho-physiology of the disease.

The authors make a note that helps us understand perhaps why so many different research labs have different research results on FSHD – "when evaluating therapies or assessing the disease progression, great care

should be taken to assess a muscle at the same location every time, or to assess a muscle as a whole. Finally, when taking a muscle biopsy, MRI might be helpful to guide the optimal location as 20% of the muscle biopsies in FSHD are reported to be normal."

Reference

Neuromuscul Disord. 2009 May;19(5):357-62. Epub 2009 Mar 28

Quantitative MR imaging of individual muscle involvement in facioscapulohumeral muscular dystrophy.

Kan HE, Scheenen TW, Wohlgenuth M, Klomp DW, van Loosbroek-Wagenmans I, Padberg GW, Heerschap A.

Department of Radiology, Radboud University Nijmegen Medical Center, Nijmegen, The Netherlands. h.e.kan@lumc.nl

The purpose of this study was to implement a quantitative MR imaging method for the determination of muscular and fat content in individual skeletal muscles of patients with facioscapulohumeral muscular dystrophy (FSHD). Turbo Inversion Recovery Magnitude (TIRM) and multiecho MR images were acquired from seven FSHD patients and healthy volunteers. Signal decay in the multiecho MR images was fitted to a biexponential function with fixed relaxation rates for muscle and fat tissue and used to calculate the degree of fatty infiltration in eight muscles in the lower leg. Considerable differences in fatty infiltration between different muscles were observed in FSHD patients, suggesting that this could be used as a biomarker for disease progression. TIRM imaging indicated an inflammatory component of the disease previously only observed in muscle biopsies. Typically, muscle involvement was non-uniform even within one muscle, indicating that MRI can be used as a valuable tool to study pathophysiology and therapy evaluation in FSHD.

PMID: 19329315 [PubMed - in process]

{Excerpt From Paper}

Acknowledgements

The authors gratefully acknowledge Frank Vergeldt (Wageningen University, Wageningen, The Netherlands) for his contribution to the fitting algorithm and the FSH Society Sam E. and Mary F. Roberts Nutrition Research Fellowship Grant FSHS-SMRF-003 and Prinses Beatrix fonds WAR06-0217 for funding.

In memoriam -- Zdenek J. J. "John" Stekly

By Daniel Paul Perez

With deep sadness and sorrow we extend our condolences to the family on the loss of an FSH Society Board member and dear friend – Zdenek J. J. "John" Stekly.

In life, there are individuals who have a remarkable impact on so many others without their even knowing. Z. John Stekly was one of those people. John was a brilliant scientist and retired CEO. He had remarkable connections in Washington, D.C., and he was able to walk through walls with credentials like none I had ever seen. He was actively involved as a member of the National Academy of Engineering and the New England Council. He often made visits to the White House and had an undying love of magnets, magnetic phenomenon and superconductivity. He was a Nobel-quality scientist and pioneer in magnetic resonance imaging and magnetic resonance spectroscopy (MRI/MRS). Dr. Stekly traveled to the International Society for Magnetic Resonance in Medicine (ISMRM)

meetings all over the world and engaged leading MRI/MRS scientists and centers in working on FSHD. His efforts recently led to a landmark paper on MRI and FSHD.

John was the most kind, thoughtful and humorous fellow. When he travelled, he would also send a little souvenir to us. He called and offered support to my wife and to myself in our darker hours. He was not afraid of any illness, nor did he let his own health issues deter him from being out and active in the world. He wanted to see solutions for FSHD for his wife and son and was always sincere, direct, sure, with fine

intellect, sharp thinking and good feeling. In our comings and goings at the science meetings conferences, at Board meetings, out and about for a bite to eat, and in his home alongside the magnificent Sudbury river basin in Wayland, Massachusetts, and charming vista, he was always gentle, loving, creative, resourceful and grandfatherly.

And now looking back, I see clearly a reflection of John's remarkable capacity as a scientist and engineer to bring ideas into real form and to make the world a better place for all.

Save the date

Gatherings of FSH Society members and friends!

- ▶ **Highland Park, New Jersey**, Luncheon, Sunday, June 28
- ▶ **Denver, Colorado**, Gathering of FSH Society members and friends, Sunday, June 28
- ▶ **Alexandria, Minnesota**, Picnic, Saturday, July 18
- ▶ **Twin Cities (Coon Rapids), Minnesota**, Picnic, Sunday, July 19
- ▶ **Cape Cod, Massachusetts**, Walk & Roll, Saturday, October 10
- ▶ **FSH Society International Patient and Researcher Network Meeting**, Summer 2010

For more information, contact
nancy.vanzant@fshsociety.org
or (617) 658-7878

Zdenek J. J. "John" Stekly 1933 – 2009

Dr. Zdenek J. J. "John" Stekly, 75, succumbed on April 3, 2009 after a long battle with coronary heart disease.

He was born on October 11, 1933 in Prague, Czechoslovakia, the son of the late Karel A. Stekly and Jindriska (Wolfstahl) Stekly.

Dr. Stekly was the beloved husband of Suzanne Gibbs Stekly of Wayland, Massachusetts. He was the loving father of Susan Stekly Williams and her husband Stephen W. Williams of Framingham, Paul F. Stekly and his wife Ashby Free of Cave Creek, AZ and of the late J. Steven Stekly. He leaves five grandchildren, a niece and two nephews.

After escaping Nazi occupied Czechoslovakia, Dr. Stekly relocated temporarily to England before moving to Rio De Janeiro, Brazil where he spent the majority of his youth. Accepted into MIT at the age of sixteen, Dr. Stekly completed his studies, the first in his class, receiving a BS in Mechanical Engineering, and a Masters in Mechanical and Electrical Engineering in 1955. In 1959 he received his Doctorate in Mechanical Engineering.

After working for AVCO Everett Research Lab, Dr. Stekly worked as chairman of Magnetic Corporation of America, specializing in the production of superconducting magnets for use in MRI Scanners, Maglev research, Dept of Defense and the Dept of Energy.

A pioneer in superconductivity applications, Dr. Stekly developed the 'Stekly Stability Criterion' that defines the maximum efficient operating capacity of superconducting wire.

Inducted into the National Academy of Engineering in 1981, Dr. Stekly was also a member of the American Physical Society and the New England Council. He was elected to the Board of Directors of the FSH Society, Inc. (Muscular Dystrophy). He belonged to the Phi Kappa Sigma Fraternity.

At the request of the family, there will be no services at this time. Private services will be held for the family at a later date. For those who desire, gifts in his memory may be sent to the FSH Society Inc., 64 Grove St, Watertown, MA 02472.

A Festive Evening of Music and Song raises over \$100,000 for research in FSHD

Nearly 250 concert goers enjoyed a splendid concert to benefit the FSH Society on March 25th. The performance was held at Merkin Concert Hall, at the Kaufman Center, Goodman House, 129 West 67th Street, New York.

Hanna Lachert, violinist, and member of the New York Philharmonic, accompanied by pianist H el ene Jeanney; and Steven Blier, co-founder & artistic director, New York Festival of Song, with Paul Appleby, tenor, and Jesse Blumberg, baritone, played and sang a wonderful concert!

Hanna and H el ene played *Sonata for violin and piano in three movements: Allegro Vivo, Intermezzo: Fantasque et L eger, and Finale: Tr es Anim e* by Claude Debussy. Hanna recalled that she first played in Merkin Hall at least 30 years ago, and she and H el ene have played together for 10 years.

Steven, Paul and Jesse played and sang a wide range of music—German art songs, Latin American pieces, French popular songs from the 1930s and 40s, and American music from all walks of life—the concert stage, the musical theater, the opera house, and the big bands. Steven delighted the concert goers with humor, good spirit and commentary in his introductions.

The evening concluded with a champagne encore reception, an occasion to meet the performers and to continue the festive occasion.

Special thanks from all the FSH Society to these wonderful musicians, to Judy Seslowe, chair of the concert; Rosalind Devon for her donation of the concert hall; and the concert committee for making this the most successful evening to date for the Society in New York.

Music, FSH and Me A Message from Steven Blier

I play many concerts all year, and each one takes on a tremendous amount of meaning for me. But participating in this one, a benefit to raise money for research on FSH Muscular Dystrophy, has a special place in my heart. I cope with this disability on a daily basis, but I rarely get a chance to do anything about it. What a joy to take up my own cause by playing some of my favorite songs with one of my favorite performers—tenor Paul Appleby, a natural balladeer and my own personal Orpheus. I think of March 25 as the “Send Steve to Dancing School” concert, and I remain hopeful that one day I’ll do the meringue at my own wedding. I have learned a lot from having a disability, and I am not afraid of it. My father always wanted me to be tougher, and FSH Dystrophy has certainly brought out my grit. (It has also made me far more sensitive to the people around me.) Still, I’d welcome a bit of a letup. I think I’ll be just as tough and sensitive after they find a cure for FSH. In the meantime, music continues to give me vitality; I can’t dance with my feet, but I can still dance at the keyboard. Tonight, Strauss, Gershwin, Hoagy Carmichael, and Villa-Lobos will lift me into the air, and Paul Appleby’s voice will keep me soaring. It doesn’t get any better than that.

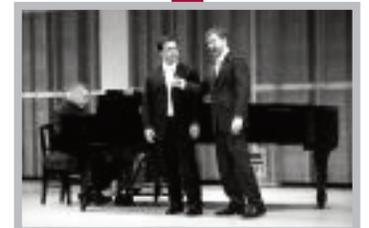
Steven Blier



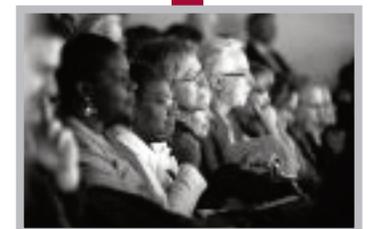
Judy Seslowe, chair of the concert,
and Steven Blier



Hanna Lachert, violin,
and H el ene Jeanney, piano



Steven Blier, piano; Paul Appleby,
tenor; and Jesse Blumberg, baritone



Attentive audience

Charity Navigator rates FSH Society ★★★★★!

America's premier independent charity evaluator has awarded the FSH Society its coveted 4-star rating for sound fiscal management. In the letter announcing this achievement, Charity Navigator cited the Society for "its ability to efficiently manage and grow its finances."

Charity Navigator applies data-driven analysis to the charitable sector. It works to advance a more efficient and responsive philanthropic marketplace by evaluating the financial health of over 5,400 of America's largest charities.

This "exceptional" designation differentiates the Society from its peers and demonstrates to the public it is worthy of their trust. To read the analysis, go to www.charitynavigator.org and search on FSH Society.

End of Tax Season Celebration--Spring Fling for FSH Muscular Dystrophy raises over \$20,000 for Society



Christopher and Ellen Stenmon (center) hosted the 11th annual End of Tax Season Celebration, a very successful fundraiser, on May 1.

Guests enjoyed good food and drink, music, dancing, a door prize, a silent auction and a raffle. They had great fun and supported a great cause. Chris is a member of the Society's board of directors.

As Society's Facebook page succeeds ... a new FSH Society online forum to be launched

Since its launch in early 2008, the Society's Facebook page has attracted hundreds of viewers and many fans. Go to www.facebook.com and sign up. It is free and easy. Search on FSH Society and become a fan. Bookmark it and come back often.

A number of Society members have also asked how we might provide other venues for their connecting. In the next few weeks, we will be launching a forum community where you will have access to specific topics and be able to communicate privately with other members, respond to polls and participate in other special features.

Watch for this new feature.



Young and older members meet for lunch in LA

Southern California members and friends of the FSH Society gathered in the Brentwood area on January 31. Guests, including the three above, enjoyed meeting each other and sharing experiences and great Chinese food. Many thanks to Barbara Rosenblum, Mimi Garcia, and Sixto Garcia for organizing the lunch meeting for the Society.

Society members gather across the country

Lunch in Spring Lake New Jersey in November

New friends share their experiences with FSHD—keeping up with family, jobs, friends and all the activities of busy lives—over lunch at The Breakers in Spring Lake, November 2008. They report that they also talked about baseball. Conversation was lively and today they feel less isolated and happy to be new friends. They will meet again on June 28. If you would like to join them, please let the FSH Society office know!



Happy to be in Florida in February

Mimi Brown hosted a group of Treasure Coast members and friends of the FSH Society in her home on February 21, some of whom are pictured here. Guests had a great afternoon meeting each other and sharing experiences.



It's what you can do that matters...

By Alan Brown

Alan is Director of Project Management, NFL Films, and also serves as producer on special projects. He lives in Bucks County, Pennsylvania.

Living with FSHD is like having two lives. There is one life in which the world is boundless; then slowly it changes into another life bound by limitations. Being diagnosed at 25 years of age was a life changing experience for me. But over the intervening years, the diagnosis became a positive force in my life. This may seem odd, but in some ways being diagnosed with FSHD provided me with the impetus to set goals in my career and lifestyle.

From the outset, my career in film and television production needed to be steered, nurtured and cultivated in a way that would allow me use my mind rather than depending on physical strength. I set out on a deliberate path to experience and learn as much as I possibly could to provide me with the best overall view of the production world. My end game has been to one day be in a position to know all the elements and “gears” that need to “mesh” in order to produce a show or commercial.

I am proud to say that in my 21-year career, I have done it all—from clearing a toilet on a recreational vehicle used by a 40-person crew on a hot summer day, to walking the red carpet for the premiere of a film I produced, at the Chinese Theatre on Hollywood Boulevard. Everything in between those two extremes has been filled with adventure, friendship and challenges, but most of all, life.

Filmmaking, like any other art form, is about pushing the envelope of your limitations.

For the musician, it is the note that you do not play that speaks volumes. For the painter, it is the stroke that is not painted that finishes the painting. It is about working smart and knowing your limitations, and working intelligently within these parameters, to maximize the end product while at the same time minimize the physical impact. This describes how I live my life. I maximize what I can do and I know how to outsource what I cannot do. I have to build and or join the right team.

For me professionally, NFL Films was the right team to join. They have been an incredible team to work with. Over the

past 21 years, they have seen my declining physical condition, yet my career has prospered because NFL Films has allowed me to take charge of my career and contribute to the company in a positive way. The wonderful people who have allowed me to do this are Kennie Smith and Steve Sabol. The two of them foster an environment of risk taking and growth that provides me with the perfect environment to achieve my career goals. It is also the hallmark of what makes NFL Films one of the most successful names in the business.

Most corporate mission statements live in a brochure somewhere or get shown in some boring meeting via a Power Point display. NFL Films lives its mission statement everyday and on every project. The mantra is “Finish Like a Pro.” That is how we do our work. We utilize the best talent to do specific tasks, and I am honored to be one of the utilized people.

At NFL Films, we make sure that whatever we start also ends with the same dedication and attention as we had at the beginning. However, as we filmed the *24 Hours of LeMans* in France last summer—we worked for 39 hours straight—it seemed a little less likely that we could “Finish like a Pro.” In the end, we all rose to the occasion and transcended ourselves and our limitations with the end result being the film *Truth in 24*. Producing this film was more physically demanding than other jobs in recent years. FSHD compounded with a heavy travel schedule, overall physical assertion, and fatigue due to the duration of the shoot days made the experience more of a challenge. However, if you ask for help when you need it and do all the little things to make your life a little easier, then it is possible to focus on the task at hand and maximize your productivity regardless of your limitations.

Being a part of the right team starts at home. In my personal life I am blessed to have a supporting wife, Rona, who intuitively knows what I can and cannot do. Having her as a partner allows us to work together as any couple should toward common goals. Tasks as simple as planting flowers require us to communicate and

compensate for what we both do not know or cannot do. The same goes for raising our 7-year-old daughter, Rachel, who knows there is something different about me. I have never lifted her into the air or picked her up when she has fallen. Rona has had to compensate for many of the normal father-daughter activities. Despite this though, it all comes back to maximizing what you can do. Rachel knows that although I have some physical bounds, what I would do for her and how much I love her has no bounds. She will always have every ounce of strength and love I can give.

As I conclude, it has become quite apparent what this letter is about: Being part of the right team and communicating with that team. For me being with NFL Films has afforded me the opportunity to do what I love with people I love. At home, we are a family that is compassionate, loving and generous in our approach to life. We are enriched by our abilities and our disabilities.

Although we all have our disadvantages, sometimes FSHD or something else, the key is to be honest enough with ourselves to know what we can do and what we cannot do. It is also up to us to make the decision as to how we will internalize and utilize our disadvantages or shortcomings. These disadvantages or shortcomings actually bring our strengths more into focus. And that focus provides us with an opportunity to do more with what we have. This is a true blessing. It is what you cannot do that makes what you can do more poignant. Lastly, I refuse to be remembered for what I could not do. I will be remembered for what I did.

Visit the Society's website, www.fshsociety.org, to:

- Volunteer to provide tissue samples
- Read about new research
- Make a gift online
- Learn more about FSHD
- Get involved!

Society post-doctoral fellowship award brings important results & new publications

Gene associated with FSHD vision problems identified?

Over 95% of FSHD patients carry a genetic abnormality proposed to affect expression of the FRG1 (FSHD Region Gene 1) gene; previous studies of FRG1 in frogs demonstrate that it is important for skeletal muscle development. University of Illinois scientists investigated the possibility that the FRG1 gene might also be responsible for the blood vessel abnormalities in FSHD patients' eyes. Their report describes how they examined the FRG1 gene in the frog and found the protein that it encodes, or converts, is highly expressed in blood vessels. Additional experiments show that normal FRG1 protein expression is important for blood vessel growth and organization.

Previous work linked aberrant FRG1 expression to the skeletal muscle defects of FSHD, and this study shows that FRG1 expression similarly contributes to abnormal blood vessel growth in the retina. Thus, the FSHD mutation, by affecting expression of the FRG1 gene may contribute to both the skeletal muscle and visual deficits associated with FSHD pathology. This study supports the idea that FRG1 expression is a main cause of this type of muscular dystrophy and might be an important target for therapeutic intervention.

The report, "FSHD region gene 1 (FRG1) is crucial for angiogenesis linking FRG1 to facioscapulohumeral muscular dystrophy-associated vasculopathy" was written by Ryan D. Wuebbles, Meredith L. Hanel, and Peter L. Jones of the Department of Cell and Developmental Biology at the University of Illinois at Urbana-Champaign. The paper was published in the May/June issue of *Disease Models & Mechanisms*. The publication acknowledges support from a number of funders, including the FSH Society Landsman Charitable Trust Fellowship award.

Attention all young artists and parents!

By Aubrie Lee

A few years ago, many of you will recall that Justin Cohen led a greeting card business to raise funds for the FSH Society. The project began as his service project, customary for bar- and bat-mitzvah children at his synagogue. Other young people affected with FSHD contributed artwork, and the project was a very successful fundraiser for the Society. Now, we want to resume this business, using fresh artwork. We hope other young artists (middle school age and above) will join with us.

If you are a parent of someone with FSHD, please encourage your child to participate by submitting artwork. If you are a young artist, please send your original, two-dimensional artwork, with your name and age, to

Dr. and Mrs. Gary Cohen
58 Arnold Drive
Kingston, NY 12401 USA

Or, if it is more convenient, you may also email attachments to me at mystic.4444@gmail.com.

Thank you very much!

Is your email address current at the Society?

We communicate with many of you by e-mail and many of you encourage the Society to use more electronic media. New databases and software will make this easier in the coming months. If we do not have your current e-mail address, and if you want to be sure of receiving up-to-the-minute information from the Society as news breaks, please e-mail your address to us – for the first time or when you have changes.

E-mail jennifer.lazzaro@fshsociety.org. Thank you!



Picnics in Minnesota in July...

Below is an invitation for a picnic near the Twin Cities on July 19. Another will be hosted in Alexandria, Minnesota, on July 18. If you or someone in your family would like to attend, please contact the Society office.



Please Join Us for a Picnic!

Gathering of FSH Society Members and Friends

An occasion to meet each other and celebrate summer

With a brief program about advances in FSHD Research

Sunday, July 19, 2009, 1:00-4:00 p.m.

**Coon Rapids Dam Regional Park
Coon Rapids, Minnesota**

RSVP by July 10 to nancy.vanzant@fshsociety.org or 617-658-7878

Food:

- Please provide your own table service, drinks and anything you must have for a picnic.
- Bring a salad, hot dish, snack or dessert to pass.
- Brats, hamburgers, hotdogs, buns and condiments will be provided.
(A small donation to cover this cost will be collected at the picnic.)

Accessibility: *The pavilion is near parking, accessible restrooms, the river and playground and on paved pathways. There is room to maneuver mobility devices between picnic tables. The park has miles of accessible trails, fishing on a pond or the river. It is clean and safe.*

Help for FSHD patients must continue on. Full speed.

Each membership in the FSH Society costs \$50 and you can increase your membership contribution many times over!

Your optional increased donation to the FSH Society goes straight to the FSH Society programs and initiatives. Every additional \$50 enables the FSH Society to answer another FSHD patient's or family's or friend's call to Society.

Since 1991, the FSH Society, Inc., has been supporting and advocating for people with FSHD. Every day, individuals with FSHD need to talk to the FSH Society about a problem they face. The support and advice they receive calms fears, solves problems - and even saves lives!

We want to ensure that all FSHD individuals get the help they need. However, to do that, we need to answer more calls. The reality is that FSH Society can only answer the hundreds of calls it receives each year

with your continued support. Remember, the FSH Society, Inc., may be an FSHD patient's only hope - and we cannot achieve any of this without you. Please give

generously today!

If you have not made a membership gift in 2009, please send your gift in the enclosed contribution envelope. Or,

make your gift at the new website,

www.fshsociety.org.

Go to **Contribute** and then to **Membership & Other Gifts**, and then to **Membership**.

Your help, support and participation are vitally needed in these challenging economic times!

The funds you donate today will:

- Help FSH Society staff respond to more calls from patients with FSHD each year.
- Help FSHD researchers and clinicians have access to patients they need to help solve and treat FSHD.
- Enable patients with FSHD and those involved with FSHD worldwide to e-mail or post messages directly to FSH Society patient outreach staff.

FSHD Future Fund growing

In early 2008, several members of the board of directors shared with FSH Society members their estate planning—that their wills or other trust documents include a bequest to the Society. By way of this good news, the Society launched the FSHD Future Fund, and declared these individuals charter members.

Since then, many other individuals and families have informed the Society of

its inclusion in their estate plans.

We can all help the Society and its future work by becoming members of the FSHD Future Fund by including a bequest to the Society in our will or other estate planning documents.

If you have already included the FSH Society in your will, we hope you will let us know. If you will allow the Society to recognize your dedication in our Annual

Donor Report, your example might inspire others. If you have questions about your planning and how it can support the work of the Society in the future, or if you would like a copy of the booklet, *Questions and Answers about Wills and Bequests*, let the Society office know by calling (617) 658-7878.

Thank you!