

Quarterly Advocate

Spring 2001

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Member Retreat:

Informative, inspirational and loads of fun seem to be the consensus of the attendees, judging by the feedback we have received to date. With over 60 attendees our first annual Member Retreat has been deemed a huge success.

Those in attendance were lavished in comfort at the Bertramm Inn & Conference Center who's employees paid great attention to every detail. The food also received high rankings from our well fed members.

Our member/attendees were presented with information on BCCNS by some of the top Scientists, Researchers, Clinicians and Health Care professionals in the Country.

Chemoprevention:

Our sessions began on Friday with Dr. Ervin Epstein Jr. MD of UCSF General Hospital who is conducting research on mice that they have been able to make grow Basal Cell Carcinomas. He is partnered with Dr. David Bickers MD Professor of Dermatology at the College of Physicians and Surgeons of Columbia University.

DR. Epstein was thorough in his explanation of the genetic mechanics of our disorder. This was a great overview to enable us to grasp the whole picture of his research. DR. Epstein has been successful in growing Basal Cell Carcinomas on genetically engineered mice. A feature that was not possible previously. Having these mice has allowed his team to do extensive research on methods of preventing the growth of Basal Cell Carcinomas in BCCNS Patients.

Dr. Bickers explained that they have developed a proven principal. This is not a theory. They have tested and retested on mice and have sufficient documentation to know that this will work on human subjects. The drug currently being considered has been approved by the FDA and is widely prescribed for other applications. Side effects on human patients have been sorted out as well. This is very exciting news! I can think of nothing that would please me more than to know that our Children and Grandchildren may never have to have surgery for Basal Cell Carcinomas! DR. Bickers did explain quite thoroughly, that this may not be a cure all. It may not prevent the growth of Basal Cell Carcinomas forever but, as long as it works for the duration of our lifetime it has done it's job. Hence the term Chemoprevention.

They need our help! DR. Epstein & Bickers need 60 patients to commit to a study in order to gain FDA approval. The problem is that we are a small group of patients. It is quite difficult to find people with BCCNS / Gorlin Syndrome. When their team is able to locate patients they have to meet a specific criteria and agree to comply with all of the protocol for the study. One of the problems is that the drug being considered is quite popular and the patients can't have been taking it for a period of time prior to the study.

Again, we are a small group of patients. If we do not help with this important study it may never gain approval. For all of us old timers imagine if you had the opportunity to take a pill instead of all of the shots, scraping, burning, Mohs and plastic surgery we have endured all of our lives. I know for sure my Doctor would much rather golf or fish than do what he must do to me. I would prefer to join him! If we don't get involved for ourselves, for our children and for our grandchildren nothing will ever change. We can't leave it for the other guy. In our case there is no other guy.

If you are interested in learning more about the Chemoprevention Study call 1-800-386-1001

DR. Bickers was also generous enough to conduct the breakout sessions where members had an opportunity to get more detailed information on his work. DR. Bickers also fielded questions regarding specific needs of each member and served as a skilled Moderator at our General Session.

Thank You DR. Epstein and DR. Bickers! We are indebted to you for all you've done and are trying to get done.

We will be establishing a website with all the details of the study and we will get you that link as soon as it is available.

Reconstructive Plastic Surgery:

Next was a phenomenal presentation by DR. Brian Windle a reconstructive Plastic Surgeon. We were all impressed by DR. Windle's informative presentation and even more so by his humanitarian work in the Philippines. DR. Windle shared a slide presentation of many of the people he has helped in this endeavor. The dramatic difference in appearance he was able to achieve in these patients was incredible and without a doubt improved their quality of life 100%. Many professionals in DR. Windle's position may vacation in some tropical climate or spend a several weeks touring exotic golf courses. He chooses to travel to a foreign country, do a couple hundred surgeries for people he doesn't even know, all this for free. You are to be saluted DR. Brian Windle, it is an honor to have him as part of our Medical Advisory Board. DR. Windle stressed the importance of saving our spare parts for future need. Some of our members were hearing terms like this and Harvest Sites for the first time. We are pleased that he addressed this important issue. DR. Windle had us all on the edge of our seats and led the attendees through an emotional session that ranged from laughter, tears to roaring cheers! Thank you DR. Windle.

Photodynamic Therapy (PDT):

DR. Allan Oseroff MD. of Roswell Park Cancer Institute is a pioneer in the field of Photodynamic Therapy (PDT). He explained these procedures in a very detailed yet understandable manner. Many of our members expressed interest in having PDT as part of their course of treatments. I have been in contact with several members for whom this treatment seems to be a Godsent. The staff at Roswell has been diligent in communicating with their medical teams and the families. We are also very fortunate that DR. Oseroff has agreed to join us as part of our Medical Advisory Board. He brings a great deal of experience in treating patients with BCCNS. DR. Oseroff also hosted several break out sessions where members could have answered, more detailed and personal questions, as to the best treatment for their needs. For more information on Photodynamic Therapy at Roswell Park Cancer Institute visit their website at:

<http://www.roswellpark.org/search/home.asp?keyword=pdt>

Feel free to contact DR. Allan Oseroff MD at 1-800-ROSWELL or 716-845-5813 e-mail allan.oseroff@roswellpark.org

Mohs Surgery:

DR. William Lynch MD. the Director of our Medical Advisory Board was kind enough to rearrange his schedule in order to spend a great deal of time with us. DR. Lynch presented us with information on Melanoma and Non Melanoma Skin Cancers. Included in DR Lynch's presentation was a description of the dynamics of the Basal Cell Carcinomas in BCCNS Patients and how their care may differ from the

general population. We were also impressed by the slide presentation that accompanied DR. Lynch's presentation. He was able to illustrate for us that not all surgical wounds should be immediately closed and reconstructed. In BCCNS patients it may be more important to insure there is no recurrence prior to covering over a tumor removal site. He also described in great detail the benefits of Mohs Micrographic Surgery for BCCNS Patients. The Mohs method is more likely to remove all of the BCC while conserving as much good tissue as possible. Curved disc shaped sections of the tumor are mapped and analyzed. If there are any remaining cells they are detected. Mapping the region allows the Surgeon to know where to go back for additional removal while preserving the good cells. DR. Lynch has worked diligently in organizing our Medical Advisory Board and is establishing a network of Health Care Providers Nationwide who have experience in dealing with BCCNS.

Thank You DR. Lynch!

Odontogenic Keratocysts:

DR. James Hanna MD is also a huge contributor to our Medical Advisory Board. He has extensive experience with Odontogenic Keratocysts in Gorlin Syndrome. Dr. Hanna presented slides on some of his extensive experience and even a complete history of a BCCNS Patient he has followed for some years. His overview of how these cysts develop, the damage they can do, how difficult they can sometime be to diagnose and the importance of regular panorex exam to prevent extensive damage.

DR. Hanna also covered the different treatments for removing these cysts. I am aware that some of our members are just having the cysts drained & later pulled out of a drain hole. DR. Hanna cautioned us against this method, especially having BCCNS. DR. Hanna reviewed some of our member's cases and has communicated with their local Surgeons to insure the best possible care.

Thank You DR. Hanna!

Gorlin Group UK Meeting:

Our International Liaison Jim Costello of the Gorlin Group UK was also an honored guest and presenter at the retreat. Jim and his family traveled from England to be at the event. He and his wife Margaret gave us a multi media presentation on the syndrome, their family experience with BCCNS and their work around the world spreading the word about the full impact of the syndrome and supporting patients.

Margaret was also a crucial component of our support persons session with Social Worker Sheryl Cohen. It was emphasized that the patient is by no means the only member of the family effected by the disorder. Attention should be taken to insure that all members of the family are cared for accordingly.

The British Broadcasting System BBC has been following Jim and his family for the last six months filming for part of a four part series on Genetic Disorders And Their Impact On Families. They followed to our meeting in Ohio as well. Gorlin Syndrome has been chosen (Due to Jim and Margaret's efforts) as one of the four disorders. The broadcasting of this program will certainly be one of the biggest events to give exposure to BCCNS / Gorlin Syndrome that has ever occurred. Thanks to Jim and Margaret for allowing this intrusion in their lives to benefit us all.

Beginning to regret that you were unable to attend the Member Retreat in Ohio? Well here is your second chance. Jim has just set the date for the Gorlin Group UK 2001 Convention in England. Mark your calendars it is October 13th, 2001. What a great excuse to go to Europe! There seems to be enough interest from our group already that I have contacted my Travel Agent to see about group rates. Of course,

I will shop them out and solicit discounts from the Airlines directly. If you are interested in traveling with the group just let me know. Some of us attended their meeting last year and had only one problem. Nobody wanted to leave!

For more details on the Gorlin Group UK 2001 Convention watch their website:

<http://www.gorlin-group.pwp.blueyonder.co.uk/>

Thanks Jim & Margaret!

Living With BCCNS:

Social worker Sheryl Cohen MSW, LISW conducted a very informative session on how to live with a long term illness. She was also wonderful at discussing how to help your children deal with the various aspects of the disease. Several important issues were handled such as how to function as a family when one or some of the family members are affected with a chronic illness. It was stressed that the members of the family not afflicted be included in discussions, decisions and therapy. Sheryl also conducted a session for our care givers being spouses or significant others. This was the highest rated session on our feedback forms. Even the people from the BBC could not hold back their emotion during this session. Sheryl is a graduate of Case Western Reserve University, Mandel school for Nonprofit Leadership Masters of Nonprofit Organization Candidate Spring 2003. University of Pennsylvania School of Social Work. Sheryl is a camp director for the American Cancer Society Camp Centers of Excellence Program, and Camp Friendship Director (Camp for children with cancer sponsored by the American Cancer Society. Sheryl Cohen is also the Senior Pediatric Oncology Social Worker for University Hospitals of Cleveland. Sheryl has also agreed to offer her services to our organization by being part of our Medical Advisory Board.

Thanks ever so much Sheryl and our group sends a big hug!

Professor Gorlin:

We were honored by the attendance and presentation of DR. Robert J. Gorlin DDS. Regent's Professor of Oral Pathology and Genetics Department of Pathology, Dermatology, Pediatrics, Obstetrics-Gynecology and Otolaryngology School of Dentistry and Medicine Professor Gorlin described every known feature of the syndrome and had awesome slides to document them all. He gave us the details of how he happened to come upon the syndrome and even had a slide of Muriel the first patient ever to be diagnosed with what would become Gorlin Syndrome a.k.a., Gorlin - Goltz Syndrome a.k.a., Nevoid Basal Cell Carcinoma Syndrome a.k.a., Basal Cell Nevus Syndrome a.k.a., Basal Cell Carcinoma Nevus Syndrome, etc., etc., etc. We were also enlightened on the story behind a family of Mummies, now in Torino, Italy that have the features of Gorlin Syndrome. Care to guess where I hope to stop on the way to England?

The genetics of the syndrome were also laid out for us in a very understandable manner and I believe our members are better able to understand why we have the individual features we possess. Professor has a new publication coming out in September and he has assured us that we will get a copy as soon as it is available.

Professor Gorlin is a great guy and a rare personality. We are extremely grateful to him for all of his years of research and attending our meetings and giving such an informative presentation. What he may not be aware of is that I am most thankful for how wonderful he was with all of the children. He mixed with

many of our members, but made a special effort to interact with as many of the kids as he could. After meeting him I think he still has a little of that kid inside of him.

Thank You Professor Gorlin.

NPR Interview:

Our Legal Council and great friend Mr. Bill Ginn Esq. not only was Bill our first financial contributor he was kind enough to arrange an interview National Public Radio which was broadcast on Tuesday April 24th, 2001. You can hear a rebroadcast of the interview on the Internet here:

<http://www.wcpn.org/spotlight/ra/0424Skin.ram>

If you have trouble loading the broadcast you may need to download realplayer from <http://real.com> this is a free download.

Help us spread the word by contacting the local station and emphasize the importance of the need for follow up programming. You can send an e-mail from here:

<http://www.wcpn.org/talk/general.html>

Bill also provided a very informative session on Patient Rights and a breakout session on End of Life Issues.

Thanks Bill!

BCCNS Life Support Network Young Persons Committee:

Our newly formed BCCNS Life Support Network Young Persons Committee will be charged with the tasks of submitting future newsletter articles of their interest. Planning young persons activities for annual meetings. Hosting occasional web chats. Monitoring and managing the BCCNS Young Persons Forum and nominating future chairpeople. Tami Neimeister has agreed to serve as an assistant to this committee.

Thanks Mamapugz3!

Members under the age of 18 can access the Forum here:

<http://www.delphi.com/bccns/start>

Web chats for this group will also be hosted from here. The Committee was named in Ohio. The Chairpeople of this Committee will serve a one year term and have agreed to volunteer their time to do so. Serving as our first official chairpeople are (in alphabetical order)

Gina (New Jersey)

Steve (Pennsylvania)

Congratulations to all involved and Best of Luck!

Patient Forum:

I recently posted some statistics on the increase of communication on our bulletin board. The activity here is up over 225%! If you have not visited this site in the past or have not been there in a while I encourage you to click the link below:

<http://www.delphi.com/Gorlinsyndrome/start>

Donations and Fund-raisers:

Mr. & Mrs. Wilbur & Eleanor Schmitt were recognized at our Member Retreat for donating \$30,000.00 to the cause. Thank you very much Wilbur and Eleanor! It is a great thing your Daughter, Kristie Schmitt Burr our Founder, is doing! It is also a great thing that you have done for all of us!

Another member who is new to our group, from Tennessee sells crafts for a living and she has pledged 15% of her profits to the Organization! How wonderful. Thank you so much for your support. It is a great feeling to know that so many people feel as strongly as we do that this work must be done!

Many of our members and attendees have inquired about donating money to the Basal Cell Carcinoma Nevus Syndrome Life Support Network. If you are interested in helping to support our cause, contact Sharon Burger at 1-866-834-1895 or by e-mail at info@bccns.org Sharon will be glad to assist you however necessary. BCCNS Life Support Network is a Not For Profit Organization.

Snail Mail:

Here we slow! We are moving the format of this newsletter to Snail Mail or Ordinary Postal Service for delivery of future issues. We have several reasons this has become necessary.

We don't want to lose you! Each time we send out a newsletter we discover that there is a significant number of undeliverable messages returned. This occurs for a few reasons. Many people are moving to high speed Internet access and have switched providers losing their old e-mail address. People just change their e-mail address. People lose their ability to pay for Internet service or a computer to access.

Until this time the Internet was the only way you could get this newsletter. Many of our newer contacts have come from soliciting Dermatologist, Mohs Surgeons and Oral Surgeons across the Country. Most of these newer members do not have access to the Internet and would never have found us if we restrict our services here.

We hope to dramatically improve the quality of the look and content of the newsletters. Having them professionally prepared will help insure this goal. This is just one of the many improvements we will be making.

In order to get a current mailing address list for all of our members, we are asking that you send us an e-mail info@bccns.org with your complete address and phone number. Be sure to put newsletter in the subject.

With all of the ongoing research there will be many new developments in treatments and updates on existing and future clinical trials. One of our goals is to get information out as it develops to as many BCCNS Patients as we possibly can.

BCCNS Life Support Network

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