

CHARGE Accounts



Summer 2006

A Quarterly Newsletter for Families and Friends

Vol. 16 No. 2

CHARGE Syndrome:
CHARGE has four
major features -
Coloboma, Choanal atresia,
Cranial nerve abnormalities, and
Characteristic ears
More information on website

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Congratulations to Dr. Sandra Davenport and Meg Hefner on their Nomination for the Genetic Alliance 2006 Art of Listening Award!

Here are some quotes from letters submitted in support of their nomination:

I have seen the dedication that both Sandy and Meg have for helping children with CHARGE and their families. . . . They both take the time to listen and understand families' frustrations, joys and concerns, as opposed to just getting enough facts to determine a medical diagnosis.

Over the past dozen years, I've watched the energy, devotion, and compassion of this dynamic duo who assisted [the CHARGE Syndrome Foundation] grow from its infancy to its current flourishing status These are women of warmth, accessibility, and humor. They are available emotionally, at meetings, by email and for frequent hugs.

The information that Meg and Sandy bring to new parents, doctors, and other professionals can have a dramatic effect on the development of the children in their lives Meg and Sandy are dedicated to our kids and our families and make themselves accessible to us for information and support.

We, in the CHARGE Syndrome Foundation, could not have accomplished all that we have without these two women. They listen, they respond, they support our families and the professionals who work with them in so many ways.

Again - congratulations to Meg and Sandy from all of us! Although they were not selected as the final Award winners, they are winners here at the CHARGE Syndrome Foundation!

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YOUR HELP IS NEEDED!

Please share your stories, parent tips, questions, book reviews and suggestions for other features you would like to see included.

Remember this is your newsletter!

!!! Deadline for the Fall Issue - August 15 !!!

Adele's Cochlear Implant Journey

by Julie Brandrup

PERSONAL ACCOUNT

The road leading up to a cochlear implant for my daughter, Adele, seemed long and sometimes steep. Now that she has an implant, we know the journey to hearing is just beginning. We hope telling you some of our story will be helpful.

When Adele was first diagnosed with CHARGE syndrome, her lack of hearing was not a top concern for anyone; we were just trying to keep her alive. However, her three-month hospitalization provided audiologists ample opportunity to test her level of hearing. After several ABR's, they were confident that she showed profound loss in both ears.

From what we were learning about CHARGE, we knew there was a chance Adele had no auditory nerves at all. [Ed. Note: the auditory nerve takes the message from the cochlea in the inner ear to the brain. The auditory nerve is independent of the cochlea—one being normal or not does not tell you about the other.] If she did not have working nerves, we wanted to move forward with sign language. But if she did have at least one viable nerve, we wanted to give her a chance to hear, and expected that chance to depend on a cochlear implant. Determining the condition of her nerves proved to be very challenging for everyone, but it was worth the wait.

The first step was a CT scan when she was 2 ½ months old. It provided our surgeon, Dr. Audie Woolley, with valuable information, such as her lack of semi-circular canals and malformed cochleas, but it was inconclusive for the presence (or absence) of her auditory nerves. An MRI was recommended but we had to wait for her medical condition to become stable

enough to receive general anesthesia for that lengthy, movement-sensitive imaging.

While we waited for the MRI, Adele was fitted with a hearing aid on her right side. (Her left ear was too malformed to keep the ear piece in place.) We were also introduced to our speech therapist, who would later become part of the cochlear implant assessment team. After a couple of months of therapy, Adele began to turn her head to noisemakers. That was our first bit of encouragement because we knew she had at least one auditory nerve.

With high hopes, she was fitted with an aid on her left side. To keep the aid on her tiny, malformed ear, her dad came up with a good trick. We placed a large piece of Tegaderm over her entire ear and aid so the outer edges of the tape stuck to her scalp (she was bald at the time). We used a hole-punch to create a small opening for the microphone. It was strange to look at, but it worked well and gave her a chance to respond with her left ear.

Dr. Woolley had always expressed his preference for implanting the left side. Not only because he was right-handed, making the left side more accessible, but also because Adele has facial palsy on her left side. We knew damage to the facial nerve is a risk with the cochlear implant surgery and we all liked the idea of not risking her “good” side.

After a couple more months of therapy and testing her left ear, there was no response. About this time, Adele was approved for general anesthesia for the elective MRI. Unfortunately, the results were still inconclusive. Our surgeon did not feel confident about the implant's chance of success on her left but we were also weighing the risk of her facial nerve on the right. To help with the decision, he ordered a fairly uncommon procedure called promontory stimulation. This test would allow direct stimulation of the auditory nerve by sticking an electrode-tipped needle through her ear drum and cochlea.

We were referred to the University of Michigan at Ann Arbor, where we were seen by audiologist, Dr. Paul Kileny and implant surgeon, Dr. Steven Telian. During the “prom stim” procedure, a medical doctor places the needle and the audiologist administers the hearing test (EABR). While she was under anesthesia, they also repeated an MRI to get another look at the nerves.

For Adele, the promontory stimulation had definitive results: her left side had poor auditory response and her right side response was almost normal. The words “almost normal” still bring a smile to our faces. The repeat MRI generated better images and validated the recommendation to implant the right side. We all agreed that the risk to her “good” facial nerve was worth the tremendous opportunity of hearing with a cochlear implant. The facial nerve is monitored closely during surgery. [Ed note:



Adele Brandrup

this is VERY important, as many children with CHARGE have a facial nerve which is ectopic, that is, not running exactly where it is expected to be.]

Back home with the test and MRI results, we only needed a surgery date and one final CT scan. After waiting for insurance approval and for Adele to get over a sickness, she went into cochlear implant surgery in September 2005 (she was 16-months old). We expected a 4-5 hour procedure. After 2 hours, the surgeon entered the room and told us bad news: during drilling, they had encountered an unexpected “tissue” in what should have been the mastoid cavity. (This cavity is the usual passageway to place the implant’s electrodes in the cochlea.) A neurosurgeon was consulted and they determined the tissue was actually a large vein carrying blood from the brain. This was obviously a serious complication that needed further assessment. The surgery was stopped for now.

Over the next few weeks, our surgeon discussed options with the neurosurgeon as well as his colleague, Dr. Telian, in Ann Arbor. Dr. Telian, a specialist in otology neurotology and skull base surgery, said he had seen similar vascular anomalies in his adult tumor patients. He felt he would be able to implant Adele successfully and safely, either by working around the vein or by approaching the cochlea from the ear canal (her ear drum would be removed.) We had renewed hope.

The second surgery was two months later, in December 2006. Drs. Telian and Woolley performed the surgery together and it was a success. They created a passageway for the electrodes by drilling away bone above the vein and by cauterizing the top of the vein. It was the best Christmas gift ever, but not without complications. A few days later, Adele’s incision site was extremely swollen. She was readmitted for probable hematoma and the site was drained of excess blood. Everything has been fine with the site since that time – she even has grown some hair!

In January 2006, the implant was turned on and the external pieces were programmed. As exciting as “turn-on” day was, we were prepared not to see an immediate reaction. It would be about a month before we would see a dramatic response – to the deep grinding noise of the garbage disposal. It was music to all of our ears.

Over four months of weekly therapy later, we are seeing more measurable progress. Adele turns her head to sound and mimics simple sounds for specific toys, such as “ah ah” for airplane and “ba ba ba” for bus. We and her therapists are very encouraged. We know she has a long way to go but we are so excited and grateful that she has the opportunity to hear and eventually speak. When we look back to those first months Adele was in the hospital, when hearing seemed like a luxury we couldn’t afford to consider, it makes us especially grateful how far she has come.

We continue to expect great things.

Jay and Julie Brandrup
Birmingham, Alabama



Adele Brandrup

Note: Adele was also included in recent additions of *ADVANCE for Speech-Language Pathologists and Audiologists* featuring CHARGE syndrome:

Web link to *ADVANCE* Magazine article:

<http://speech-language-pathology-audiology.advanceweb.com/Common/editorial/editorial.aspx?CC=71206>

Web link to *ADVANCE* E-newsletter article:

<http://speech-language-pathology-audiology.advanceweb.com/Common/editorial/editorial.aspx?CC=71199>

“CHARGE Syndrome Miranda Statement”

I am the parent of a child with very complex problems.

I need information about my child that may be time-consuming and difficult to obtain.

My child needs coordinated services from health care professionals and educators who know or are willing to learn about CHARGE syndrome.

If you cannot afford the time or do not have the capability to provide these special services, please refer us to someone who can.

Anonymous 1997

PRESIDENT'S NOTE

Neal Stanger

Hello everyone,

As I have been telling you in past issues of *CHARGE Accounts*, I feel that this and next year will be truly ground breaking years for the Foundation. So far this year we have redone our web site (go take a look at www.chargesyndrome.org), we have made our premier publication, *CHARGE Syndrome: A Management Manual for Parents* available on line to read and/or download at no charge from our web site (or you can go directly to <http://www.chargesyndrome.org/resources-manual.asp>), and the Board of Directors has recently introduced a new family lifetime membership option available for only \$150. In the next few months we will also be announcing a new fundraising campaign that Dennis O'Toole, our fundraising chairperson will be kicking off. We will be launching this initiative sometime in June or July on our website and also in a mailing to all of our members. We hope as many of you as possible will participate.

It isn't too early to start thinking about our 8th International Conference in Costa Mesa, California next summer. The dates are July 27th – July 29th and it will be held at the Hilton Costa Mesa. You will be able to make room reservations through a link on our web site beginning sometime in August. We were fortunate enough to negotiate the low room rate of only \$129 per night. The program committee is starting to put together a list of topics and speakers and will always value your comments. If you would like to make any suggestions for the program in California, please email Jim Thelin at jim@chargesyndrome.org.

Please keep checking our web site at www.chargesyndrome.org for more updates on the Foundation. We will be having even more exciting news to announce to you throughout this year and next year. Lastly, if there is anything that you would like to have

FOUNDATION ACCOUNT

the Foundation do for you, please do not hesitate to let me know.

Neal Stanger

EXECUTIVE DIRECTOR'S NOTE

Marion Norbury

I Need Your Help

I have had the same email address (mnorbury@coin.org) for many years but I have had to make a change this month to mnorbury@centurytel.net. Before we had our website and the chargesyndrome.org email addresses, my email address was listed as the contact information on many other websites. So my request to you is if you find a site that has my coin.org email, please send me a message with the website address so we can get it changed. Also if you have my old email in your address book, be sure to update it also.

By the way, if your email changes, please let us know. Thank you for your help.

In September 2004, we moved into our Vandiver Drive office and out of the back room of my house. We also added another staff member - Kyna Byerly. Kyna is a genetic counselor and very knowledgeable about CHARGE syndrome. It was great to have her in the office and work with her. But good things don't last forever - in April Kyna took a consultant position with the Heartland Regional Genetics Coordinating Center. However we haven't lost her completely. She is continuing to work with Meg Hefner on the update of the Manual.

THANK YOU KYNA!

At the Board of Directors meeting in April, a new membership level was approved. For \$150, you may become a Lifetime Silver Member. A Lifetime Gold Membership is available for a donation of \$1,000.

The brochure that most of you have received was written in 2001. It has been revised and will be available soon.

We are so pleased that the Manual is now available on the web site. However, we know that some of our families do not have access to the Internet so I want to remind everyone that we still have copies of both the English and Spanish editions. You will find them listed on the insert in this issue.

COMMITTEES

For a description of what each committee is responsible for, please go to www.chargesyndrome.org or contact the chairperson or the Foundation Office.

Conference Committee

Chairperson: Marilyn Ogan

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Family Services Committee

Chairperson: Marion Norbury

Adult Services Committee

Chairperson: Bonnie Haggerty

Education Committee

Chairperson: Suzy Morales

Collaboration Committee

Chairperson: Neal Stanger

Public Awareness Committee

Chairperson: John Wynne

All board members have a special email address:

[\(firstname@chargesyndrome.org\)](mailto:(firstname@chargesyndrome.org))



The CHARGE Accounts newsletter is intended for general information only. Medical or treatment information and/or opinions are not necessarily endorsed nor recommended by CHARGE Syndrome Foundation, Inc. or its officers. Readers are reminded that the best source of medical advice is always their physician.

As a result of a meeting of the Board of Directors in September 2005, an Adult Services Committee has been created within the CHARGE Syndrome Foundation. Due to the fact our children are growing, changing and getting older, the Board felt it important that we too should grow and support our children through their ever-changing lives. As this is a new and developing committee there is much work to be done by many individuals around the United States and around the world.

The mission of the Adult Services Committee is to continue the Foundation mission to identify and sustain formal/informal resources to support and sustain the lifetime management and appropriate independence of adult individuals who have CHARGE syndrome.

The committee has had two meetings thus far. Our first step was to identify the term "adult" for our purposes as age 14. This age was chosen because formulating an educational plan specifically for the transition process into adult services for children who receive special educational services generally begins when the child turns age 14. To help provide resources for transition, we need to identify those young adults who are beginning that transition process.

We are also developing a geographic (regional) database of adults who have CHARGE syndrome. This contact information will enable us to compile, develop and expand information about adult resources. We hope this will help others understand, support and discover appropriate medical, educational, independent living services and other providers in the individual's region. It will also enable the committee to become aware of and perhaps evaluate programs and models of outcome-based approaches to transition from school to adulthood.

The first meeting was attended by parents, one adult with CHARGE syndrome, and numerous professionals. At this meeting, we tried to recognize some of the unique and continuing specific needs of our adults, with topics ranging from medical, social, emotional, educational and independent living services. It was apparent that adults who have CHARGE syndrome and their parents/care givers face similar difficulties that are a characteristic of CHARGE across the spectrum and around the world. A pilot program is now in the process of compiling data about positive and negative interactions, needs and struggles. This information will then be used in one state for the empowerment of adults to improve programs and services. If fruitful this may then be expanded to other areas.

One of the most important goals of the Foundation and the Adult Services Committee is to promote greater participation of our young adults who have CHARGE syndrome. To help promote this goal, a subcommittee was formed within the Adult Services Committee, comprised of adults who have CHARGE. We hope these individuals will be active participants in much of the Foundation's growth. Their first task is to create a universal sign for CHARGE syndrome without input from parents or specialists. They will follow through with this task in following specific rules and guidelines of ASL. Once they have jointly decided on the most appropriate and empowering sign for CHARGE syndrome, it will be announced through the Foundation. As you can imagine we are all elated that there is such an enthusiastic group of adults who have CHARGE syndrome who will be deciding, in part, their own identity. We are pleased that these individuals will be in control of their own destiny as so often it is others who make decisions concerning nearly

every aspect of their lives. We have a devoted group of individuals with CHARGE on this subcommittee, but welcome others who are interested. Again, this subcommittee is only open to individuals ages 14 and up who actually have CHARGE syndrome. Anyone wishing to join should contact Bonnie Haggerty at bonnie@chargesyndrome.org.

The CHARGE Syndrome Foundation is also planning to provide time and space for adults who have CHARGE to meet informally (and independent of parents, other family members or specialists) at the next conference in California in 2007. This gathering will be facilitated by one or two Foundation "specialists" with the focus of helping to increase self-determination and empowerment of all our current and future adult children.

Another activity the CHARGE Foundation wishes to create for adults who have CHARGE syndrome is a peer-mentoring program connecting adults with adolescents, children and/or their families within their geographic region and/or through email.

We have only just begun. There is now much work to be done and will take many individuals around the world to fulfill the goals of this committee. Currently there are over 26 members of the Adult Services Committee with 13 being parents, 7 professionals and 6 adults with CHARGE. If you would like to join the Adult Services Committee please contact Bonnie Haggerty, Adult Services Chair, at bonnie@chargesyndrome.org.



*Thank You to Everyone for Your Generous Support of the
CHARGE Syndrome Foundation*

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CALENDAR

2006

- August 15** Submit items for the next issue of CHARGE Accounts
- September** Fall Issue of CHARGE Accounts (Latest Information on 2007 Conference)
- Sept. 29 - Oct. 1** Australasian CHARGE Syndrome Association Limited Conference, Novotel Twin Waters Resort, Sunshine Coast, Queensland. More details: www.austcharge.com.au
- October 7** First TEXAS CHARGERS Retreat at Peaceable Kingdom Retreat in Killeen, Texas

2007

- July 27 - 29** Eighth International CHARGE Syndrome Conference, Costa Mesa, California
More details will be available in future issues of CHARGE Accounts
- September 25-30** Deafblind International Fourteenth World Conference, Burswood International Resort Convention Centre, Perth, Western Australia. More details: www.dbiconference2007.asn.au/