



All About Us

All About Us is the newsletter of the Foundation for Nager and Miller Syndromes. We took our name from a story submitted by Monica Quinn in our Autumn 1991 issue. Monica captured our entire theme and purpose in three little words.

"Us" is all persons who live with our syndromes, the members of our families, our friends, our neighborhoods, the medical community and everyone we can reach. We offer information and communication for everyone interested in Nager and Miller syndromes.

As you read our newsletter, as you think about getting involved, as you contribute your features, reports, photographs, or your own experiences and poetry, think to yourself — "This is all about us."

FNMS is an international support group dedicated to helping those affected by Nager and Miller syndromes. We serve as a clearinghouse of information and link families seeking support, hope, and advice. FNMS empowers families and guides them through the process of surviving to thriving.

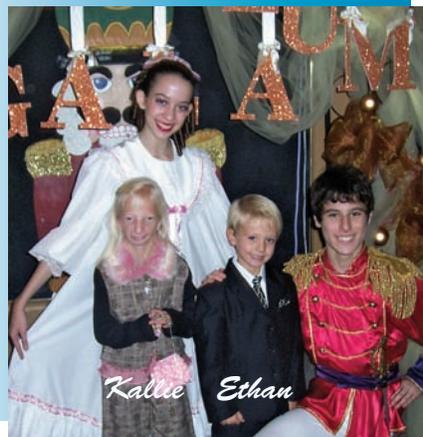
FNMS is a parent-run volunteer organization with a 501 (c) (3) not-for-profit status.

All About Me

Hey ya'll! My name is Kallie Dahl. I live in League City, TX. I am 9 years old and am in the 3rd grade. I live with my mom, dad and little brother Ethan who is 7 years old. We have a dog, Lucy, who I like to play with, and a cat named Chick.

I am special. I was born with four fingers and no thumbs. When I was very young I had to have surgery in order to make one finger on each hand into a thumb. Sometimes I forget to use my thumbs, but my dad always reminds me to. I am still able to do most everything that my friends do like ski, dance, play the Nintendo game Wii, hang on the monkey bars, ride my bike, and all of those fun things. Things like buttoning and zipping my pants and tying my shoes may seem really easy, but are very hard for me to do by myself.

My favorite food is ice cream, I eat it every night. I love to dance and have been in many performances. For the past 2 years I have been in the Bay Area Houston Ballet and Theater Company's production of The Nutcracker. It is so much fun to be on stage in front of all of those people! I also like to sing karaoke, play with my Webkinz, listen to my iPod, and go shopping with mommy. My favorite singer is Hannah Montana.



I am sooo lucky because I got to see her in concert in March!

I also enjoy hanging out with my family. Ethan and I fight sometimes, but we get along most of the time. We like to play school, store and Wii. My family and I went on a ski trip to Ruidoso, New Mexico, this year. We had so much fun skiing, snow tubing and playing in the snow. It was too cold for me, I almost froze to death!

I kind of like school, except for math because it is so hard. My favorite subject is reading.

All About Us

Founder and Fundraising Mgr.
Margaret Ieronimo Hogan
fnms4u@ameritech.net

Director and Treasurer
DeDe Van Quill
ddfms@aol.com

Editor
Leslie Gaffney
leslie.gaffney@gmail.com

Contributing Writers
Lara Beranek Greg Kehler
Castillo Family Harlena Morton
Kallie Dahl Perry Family
Cristy González Charles Randall
Margaret Hogan DeDe Van Quill

Printing
Graphics Plus, Inc., Seattle, WA

Webmaster
Rose Memije, Chicago, Illinois

Table of Contents

All About Kallie Dahl	1
Editor's note	2
New members	3
The Shopping Benefit.....	4
FNMS donor recognition	4
Beranek family fundraiser	5
Triathlon donors	6
Ojeda fundraiser ad	6
A diary of a mandibular distraction	7
Peyton's jaw distraction	10
CCA/FNMS 2008 retreat	12
Our shining stars	15
In memoriam donors for Jim Troka	16

This foundation does not endorse hospitals, teams, products, or treatments. The information in this newsletter is provided to keep you informed of activities and progress internationally regarding Nager and Miller syndromes. Views expressed in this newsletter are not necessarily those of the Foundation for Nager and Miller Syndromes.

Editor's Note: The Foundation for Nager and Miller Syndromes is in its 18th year of service to our member families. It struck me at this year's conference how FNMS is changing. We now have a large number of affected adult and young adult members who have grown up with the benefit of the foundation in their lives, giving them a link to other members who face similar issues. There have been a number of new families finding us via the Internet every year, and we now have more international member families than ever before. Each of these changes make it imperative that FNMS continue to evolve.

We now have a new web mistress, Rose Memije, whom we cannot thank enough for so generously volunteering her time and skills to our cause. Stay tuned for the launch of a new FNMS website later this year that promises to be more dynamic and up-to-date. Leslie Gaffney also deserves a huge helping of gratitude for stepping in as Editor-In-Chief of the *All About Us* newsletter and saving my sanity at the same time. We'd also like to thank Kelly Russell of Chicago, who recently volunteered to help with a large mailing.

We have several inspirational stories this issue, including two mothers' experience having their young child go through jaw distraction. We also get to meet Kallie Dahl of Texas, who tells us about all of her favorite things. Kallie also underwent a recent jaw distraction, which we hope to learn more about next issue. We have a few more inspiring updates from members in the "Our Shining Stars" feature, and we can't forget the reporting from families who attended, as well as pictures of, the always-uplifting FNMS Conference. This summer we gathered in Myrtle Beach, South Carolina, and met many other unforgettable families who are members of the Children's Craniofacial Association.

The growth of FNMS and its families is amazing and inspiring, but it brings with it the need for additional volunteers as well as increased financial contributions to keep the organization running. Since the foundation has always been a parent-run, volunteer organization, we have always been able to direct 100% of our funds directly to helping the families we serve. This makes FNMS fairly unique in the non-profit world and it is the one characteristic that we hope to never change. *Everyone can make a difference!* FNMS member families, such as the Beraneks, who are spotlighted in this edition, have committed to giving back to FNMS through fundraising. They have turned their interests and passions into fun, rewarding community fundraising events. As you read their stories, please take a moment to think about your interests, hobbies and passions. Is there a great fundraising idea to be mined there? Donating even a small amount of your time and talent can make a significant difference in the lives of others. FNMS currently needs volunteers to perform web searches, write donation Thank You notes, and assist with library updates, web link verification, conference committee tasks, mailings and to volunteers to work at The Heart of Glenview fundraiser on February 2, 2009. If you have time, please consider making a difference in the lives of our current and future FNMS families.

DeDe Van Quill

FNMS Advisory Board

John W. Polley, MD

Professor and Chairman
Dept. of Plastic and Reconstructive Surgery
Co-Director RUSH Craniofacial Center
RUSH-Presbyterian-St. Luke's Medical Center
1725 W. Harrison
Professional Building, 4th Floor
Chicago, Illinois 60612
toll free phone 888-828-7232
e-mail: jpolley@rush.edu
<http://www.rush.edu/craniofacial>

Jeffrey R. Sawyer, MD

Orthopedic Surgeon
Campbell Clinic
1400 S. Germantown Road
Germantown, TN 38138-2205
phone: 901-759-3100

G.W. Stevenson, MD

Attending Anesthesiologist
Associate Professor of Clinical Anesthesia
Northwestern University School of Medicine
Children's Memorial Hospital
2300 Children's Plaza
Chicago, Illinois 60614
phone: 773-880-4414
<http://www.childmhc.edu>

Eric Wulfsberg, MD

Professor of Pediatrics
Division of Pediatric Genetics
University of Maryland School of Medicine
22 South Greene St., A661
Baltimore, MD 21201
phone: 410-328-3335
email: ewulfsberg@som.umaryland.edu

Call for Updates!



FNMS is in the process of updating the Family Directory and needs your help! Please send an e-mail to Leslie with your family's current mailing address, phone and e-mail address. We have a lot of incomplete information, as well as missing members, that we would like to correct so we can generate an up-to-date directory.

Please send your information to Leslie at leslie.gaffney@gmail.com.

Do you know someone who should be receiving the FNMS newsletter and isn't? Send that person's complete mailing information to DeDe and they will be added to our master mailing list. Our mission is to increase awareness of Nager and Miller syndromes among medical personnel as well as the general population so we can all learn together from sharing our own stories and experiences.

Please send addresses to DeDe at DDFNMS@aol.com. Or you can mail info regarding either update to FNMS, 13210 SE 42nd Street, Auburn, WA 98092, USA. Thanks!

FNMS welcomes new members the Poirier and Wentworth families.



Genève Poirier
Quebec, Canada



Raisa Wentworth
Winslow, Maine, USA

HEART OF GLENVIEW

Glenview's Tastiest Night

Monday, February 2, 2009

Wyndam Glenview Suites, Glenview, Illinois

WE NEED YOUR SPONSORSHIP — PLEASE!

FNMS needs a sponsor for this event

A sponsor of \$1000 is required to participate in the fundraiser.

It could be one sponsor at \$1000 or 2 at \$500 or 10 at \$100.



**Plan ahead and
SAVE THIS DATE!**



DO YOU HAVE AN AUCTION ITEM TO DONATE?

Last year, FNMS raised \$5000 at our auction table and received a \$10,000 donation from the Glenview Chamber of Commerce.

We also need 5 volunteers to help out that night!

Contact Margaret Hogan

fnms4u@ameritech.net

for tickets and information on volunteering!

This is too good to miss!

FNMS Benefits from Bloomingdale's "The Shopping Benefit"

A delightful night was planned just for FNMS supporters. The Bloomingdale's March 27, 2008, event raised over \$3000 for FNMS. Browsers and shoppers gathered for an Ellen Tracy fashion preview while sipping wine, tasting yummy appetizers from Blue Plate catering, and listening to the music served up by the DJ. This event surely put the fun in fundraising! Pictured below are longtime supporters and volunteers of FNMS.



(L-R): Shoppers Cathy Cerone, Mary Carlsen, Beth Atkins, Lizzy Cerone, and Nan Glynn.



Joan Troka is flanked by her loving daughters Margaret and Beth.



(L-R): Beth Grossman, Joan Kitchie, and Cindy Giangreco.

FNMS Donor Recognition (October 2007 through July 2008)

GENERAL DONATIONS

Bloomingdales
Susan B. Cummings
Mr. and Mrs. Irwin Eisen
Marilyn V. Everett
Glenview Chamber of Commerce
Suzanne E. Kach
Ferrell Maynard
Mr. and Mrs. Daniel McGrath
Morton Grove Foundation
Sauganash Woman's Club
Mr. and Mrs. Hans Schaefer
Mr. and Mrs. Robert Winter

DONATIONS HONORING MARGARET AND MIKE HOGAN'S WEDDING

Mary Aamodt
Kathy Brown
Leslie and Scott Cordes
Meg Gagliardo
Kary and Kymn Harp
Peter Maloney
Nina and Bill McNulty
Mary and Hugh McLoone
The Riegert-Dudley Families

DONATIONS IN HONOR OF

Dan and Patty Wolf donated in honor of their son, Derrick Wolf
Marilyn and Richard Gaffney donated in honor of their granddaughter, Tinka Gaffney
JoAnn Eggert donated in honor of Rachel Eggert's 30th birthday
Beth and Patrick Smith donated in honor of Judge Mike Hogan

MATCHING CORPORATE DONATIONS

Merrill Lynch
Scotttrade
US Bankcorp



IN MEMORIAM

Andrea and Don Maynard donated in memory of Lori Maynard

Margaret and Mike Hogan donated in memory of Thomas F. Waldron

The Perry Family donated in memory of Aaron Leffel and Charlotte Holden

David and Brenda Hodson, Kelly Kimberly and Timothy Hodson, Kristen and Ryan Youles, Carl and Delores Hodson, and Maurice, Carla, Janelle, Erica and Sherman Perry all donated to FNMS in memory of George Perry



Bridget and Saul keep the sign up!



Saul and his grammy share a joke



Beranek Family Fundraiser

On June 7th we held our third annual fundraiser for FNMS in honor of our son, Saul, who has Nager syndrome. This summer's event was expanded to include a sprint-length triathlon along with the "ultra-mini" length we had last year. Nearly 100 people registered for the two events. We were so excited to have Margaret Hogan, along with her husband, Mike, daughter, Bridget, and sister, Beth, travel from Illinois to join us. They really put together an impressive team: with Margaret swimming, Beth biking and Mike running. Bridget was there with her faithful camera capturing the day's highlights. We felt like movie stars were coming to town when Margaret said they'd be coming to Marathon, Wisconsin (population 2000)! We are honored that they made the trip to be a part of the big day.

This year we raised donations in a few ways. First, we sent letters to local businesses and asked them to sponsor the triathlon. For a \$100 donation, their name would be printed on the event t-shirts. Second, we collected \$20 per entry in the triathlon (\$50 for a team, \$40 for a family). Third, we had some events at local schools in the weeks before the race, including a family movie night and a dunk tank. Finally, we held a raffle, giving out a hand-made knife, money, some Avon products, DVDs, and two gorgeous pieces of pottery that Margaret made. All in all, we raised just over \$6000! Getting donations was harder this year. If we're not in a recession, people still think we are. We just kept trying — and ended up raising more than last year!

Organizing "our" triathlon has become something we enjoy so much. The community comes together through volunteers, donors and athletes. We were touched by so many things this year, from the wonderful people who were there that day, to the amazing abilities of the athletes we saw, to the unexpected \$500 donation from a friend, to the birth of our niece just as the last athletes were finishing the race. It was an amazing day. We can't wait 'til next year!

Lara Beranek
Marathon, Wisconsin



Mike, Bridget and Beth



Sorry Margaret, Coppertone commercial try-outs were last week.



Lara with Saul and big sister Mira



Beranek Ultra-Mini Triathlon Donors 2008

Individuals and participants

Jody Apfelbeck
Curtis Axness
Nicholas Barden
Lisa Beck
Joyce Beranek
Gary and Joyce Binning
Carol Boote
Tony and Sandy Braun
Linda Brickheimer
Kelly and Jim Christensen
John and Ann Colby
Gina and Kevin Cornell
Meghann Crowe
Timothy Deets
Jane Detert
Jennifer Doering
Liza Doering
Bob and Connie Furger
LaVerne and Mary Jane Furger
Kory Gertshen
Ed Gibbons
Gifts From Heaven
Lisa Giordano
Lori Haight
Tina Halverson
Michael and Margaret Hogan
Linda Hornung
Beverlee Huset

Annaluna Karkar and James Collison
Christine Kepner
Lenny Kersten
Ann Kumm
Tony Lenard
Jean Lubbe
Jessica and James Lynott
Marathon Elementary
Marathon High School
Marathon High School French Club
Dick and Sharon Masek
Mary Montana
Jodi Neumann
Janet Nichols
Marty Olson
Tony and Kay Pickar
Gary Raether
Willie and Kathy Reed
Patricia Riske
Marty and Traci Robbins
Tille and Mary Rosensprung
Taylor Saracoff
Robb and Sarah Sigler
St. Mary's School
Dennis Wendt
Barb Wilichowski
Wendy Zimmer

Businesses

Airrunner Networks LLC
Alois Dreikosen Post 469, American Legion
Aspirus Marathon Clinic
Berens Insurance Agency
Bilco, Inc. (McDonald's)
Brickner Motors, Inc.
Color Creations
County Center Animal Hospital
County Market
County Materials Corporation
Fischer Transportation
Gourmet Kitchen Essentials.com
Hurtis Companies
Kipper Konstruktion
Kohl's
Marathon Cheese Corporation
Marathon Family Dentistry
Marks Chiropractic
Masek Custom Knives
Mega Pick 'n Save
Menzner Lumber and Supply Company
Morgan Sand and Gravel
Peterson/Kraemer Funeral Homes
Podiatry Associates of Wausau, SC
Quality Foods IGA
Quirt Family Dentistry
Rib Mountain Cycles
Star Environmental, Inc.
Sun Country Tees
Wal-Mart
Walter Jansen Builders

The Bellamy Brothers
Journey
Stevie Nicks
The Smothers Brothers
Chubby Checker
B.B. King
Mel Tillis
Krokus
Cheap Trick
Air Supply
Lee Greenwood
Pat Benatar
Peter, Paul & Mary
Eddie Money
Anne Murray

Donations for FNMS Fundraiser Needed

We are looking for donations of new or used guitars, acoustic or electric, in any condition (nonworking guitars accepted)! We are also looking for new or used drumheads of any size.

These items will be signed by famous musicians and then auctioned in an effort to help raise money for FNMS.

Please contact Jesse Ojeda at 928-770-4467 or jroojeda2001@yahoo.com for more information or to make a donation. Items can be sent to Jesse at 5532 Wishing Well Way, Fort Mohave, Arizona 86426, USA.

THANKS!

The start of the journey — a diary of a mandibular distraction

The date was set. On Monday, March 3, 2008, my son Theo went for his first mandibular distraction, 3 days short turning 9 months old. As our ENT surgeon told us, this is what we have been working toward since before Theo was born. We were lucky to know that Theo had a small jaw ante-nataly, so were able to plan an EXIT procedure birth that would mean that Theo was not without oxygen at any stage, cancelling out the risk of brain damage. After his safe arrival, he was taken immediately for a tracheotomy, then 7 days in PICU. It was during this time that we met with Glenn Bartlett, the leading craniofacial surgeon in New Zealand. His news was that, although the jaw was very small, he would be able to perform a mandibular distraction before Theo's first birthday and hopefully get the tracheotomy removed by then also.

In the whirlwind that was the first few weeks of Theo's life, this was a lifeline. Although I had no idea what would be involved, how it would be done and what equipment would be used, it was a little ray of hope to cling to during the next few months of intense learning of how to care for Theo. Knowing there was someone out there who could help Theo's breathing issues was a comforting thought.

Over the next few months I started my research and had a long list of questions regarding the procedure when we had our consultation with Dr. Bartlett 4 months later. We knew he was aiming to use internal distractors that are reabsorbed by the body over a period of time, and that the process involved breaking Theo's jaw and slowly turning pins to bring the jaw forward. The Internet and my FNMS friends were a blessing at this time, giving me things to think about and more questions to ask. Dr. Bartlett reassured us that internal distraction would be the best for Theo, and that if we chose not to do this, then he couldn't distract until Theo was 2–3 years old because the external devices he had would be too big for him. He explained the procedure and devices thoroughly and we left with a rough timeframe for when Theo was about 8 months old.

“This is what we have been working toward since before Theo was born”

Upon returning home, doing more research and talking to more parents who had to make the decision about whether to distract or not, I started to get very nervous. What if it didn't work? What if Theo's TMJs were to fuse? Would it be better to wait until he was older and use the external distractors? He would only be the second baby in New Zealand to have these internal distractors fitted, so what if something went wrong with the devices? If it wasn't successful, would it cause more problems with regard to distracting in the future? Were there other doctors who we could get a second opinion from? What if the internal distractors didn't have a high success rate for Nager babies? What was the expected

Written by
Sarah de Heer,
Wellington, NZ,
shown here with
son Theo the
day after his
surgery



damage to muscles, tissues and cells around where the distractors were placed? What about risk of infection from the device? These were all questions that I e-mailed to Dr. Bartlett, hoping for some straight answers to help make the decision. I was very pleased with the responses that came within a day or two. He answered the questions without talking down to us or thinking that we wouldn't understand, and told us that he was confident that this process would work for Theo. In turn, the confidence that I had in Dr. Bartlett went through the roof after this and there was not much more I could do. He was the expert and I had to trust his judgement. It's not like we had any other choice of surgeons — New Zealand being such a small country, there was no one else of his expertise and experience that could perform the surgery. So, the decision was made.

It amazes me how quickly 5 months can pass. We had planned Theo's gastrostomy surgery for February 15th, and after a 4-day stay in hospital we arrived home to a letter saying that we were needed in Auckland in 2 weeks for the distraction. We wanted to avoid having back-to-back surgeries, but we had waited for this one for some time so there was no question in our minds — it was full steam ahead! Theo bounced back well after having his Mic-Key feeding button placed, however, 3 days before leaving for Auckland he caught a stomach bug. This was the last thing he needed before a big procedure, but once again he bounced back in time for the 1-hour flight from Wellington on Sunday.

During the 2 weeks when I knew the surgery was finally happening, I tried hard to put it out of my mind. I realized that it was going ahead anyway and that worrying about it wasn't going to change anything. I had every confidence in the team that was performing the procedure, and they were the experts, not me. It was hard to forget, though, when friends and family were asking questions and projecting their curiosity about the procedure and fears for Theo to me. I had made peace with it, but had forgotten that others were not armed with the same information as me. I always seemed to end these conversations with people by saying that Theo has overcome so much already in his short life, that I was confident that he would overcome this challenge too, and that really, he was in the best hands possible with a surgeon who had been doing this for many years. In hindsight, I didn't really have

time to get worried myself, because I was spending much of my time reassuring other people! In my mind, it was out of my control — I wasn't going to be in the operating room (OR) performing the procedure and so there was nothing I could do about it. I found that these thoughts were strangely comforting.

The hardest part was the morning of the surgery. We were called for an x-ray at 10 am, so off we went to the hospital. At 12:30 pm the doctors descended for the usual pre-op discussions and for us to sign our lives away. We had ear, nose and throat (ENT) doctors, speech and language pathologists, anesthetists and craniofacial surgeons all explaining the part they would be playing. It was overwhelming, although good to ask any last-minute questions. I asked to see some photos of the device and Dr. Bartlett was able to show me the previous child he had just completed 3 months prior. I was amazed at how little 'hardware' was on show and how small the scars and pins were. It did a lot to settle me, and seeing him before he performed the surgery also gave me a chance to see his confidence (which was oozing!). I had a few tears at one point when they were all there — it was a lot of last-minute information to take in, but I knew that I needed to be strong for my boy. It was important to me that he didn't see us worried, but confident and happy. We were making the best decision for him and everything was going to be fine.

At 1:30 pm we were called into the OR and I asked to go in with Theo until he was put to sleep. Once again, it was important to me that he knew we hadn't abandoned him and that we were going through this with him. Although he is only young, he knows when I leave him and doesn't like it. So, with my emotions tightly under control, I held his hands while he was slowly put to sleep. When his eyes closed, the tears started as I knew they would. It was the hardest thing walking out of the OR and leaving him in there, knowing what was about to happen. My husband was also worried, as I found by the very tight hug he gave me. But, it was not in our control now; we just had to wait until it was all over. We had decided the day before that we would hold off on doing the grocery



Theo 2 days before surgery.

shopping until when Theo was in the OR, so that it gave us something to do other than loiter around the hospital nervously, waiting for it to be over. The surgeon thought that it would take about 2–3 hours, so we had some unusual time to ourselves without any responsibility. We found that although it was a good idea, it was hard to put into practice. We didn't spend too long away and were back at the hospital by 4 pm, ready and waiting for news.

“I had made peace with it, but had forgotten that others were not armed with the same information as me”

At 5 pm the surgery was completed and Theo was in recovery. We went to PICU at 5:15 pm, hoping that they would have some news about when we could see him. They found out for us that he was still coming out of the anesthetic and that he had come through so well that he wouldn't need to spend any time in PICU, but was to be put straight up on the ward. This was great news — not only had he made it in one piece, but he was so strong that he didn't need special nursing. At 6:10 pm, Theo came up from recovery with a bandage around his head and on the usual pulse-ox monitors. He was put on a morphine infusion and slept most of the night. I was up suctioning his trach every 5–10 minutes throughout the night because his airway was irritated by the poking around and anesthetic. The surgeon phoned telling us that he was very pleased with the way things went and that he would be back to see us on Wednesday to do the first turning of the pins. Round One was over and Round Two was about to start.

The first thing that hit me on Tuesday morning was how swollen Theo's face was. I had prepared myself so well for the surgery, getting the details straight in my head, that I hadn't even given a thought to how he would look after the surgery. Of course there would be swelling, but somehow my brain had failed to think of that eventuality. He looked so different. After 9 months of seeing a little face, all of a sudden it was huge — he just looked like a well-fed baby! But as the day went on, the swelling only got worse. It was so bad by the afternoon that we had to change his trachey ties, as the others were getting too tight and looked as if they were strangling him. Although he was on morphine, they also gave him regular paracetamol to help reduce pain and swelling and antibiotics as a preventative measure. With all the drugs in his system, I was surprised that Theo was able to sit up in his bed and play, even if it was only for 10 minutes at a time before he needed another sleep. We could also see his personality emerging again — cheeky, cheeky, cheeky! We were called down to x-ray again at 6 pm to check the position of the pins and that everything was in place before the distraction began in the morning.

Wednesday morning started early — 5 am to be exact — when Theo's intravenous line ruptured in his foot. After the house surgeon tried twice to replace it and couldn't find a vein, the registrar was called and also took two goes to get it in. By this time Theo was in some pain after being used as a pin-cushion and also because he had been an hour without any medication. At 7 am he was given a bolus of morphine in preparation for the first turning of the pins. Dr. Bartlett arrived, along with the ENT team who were going to turn the pins when Dr. Bartlett couldn't come in to do it himself. Theo just gazed up from the bed and didn't flinch once. The whole process took about 2 minutes and that was it for the day. The swelling had gotten worse again overnight, but Dr. Bartlett told us that it was nothing to be concerned about — in fact his words were, “Swelling? What swelling? That is nothing! It could be heaps worse!” Theo regained his will to play and create mischief as the day wore on and the morphine drip was stopped. He also had the bandages around his head taken off. We

had gone and got some hats and headbands to use instead, just so that he couldn't pull at the pins or get them caught on anything and hurt him.

Thursday arrived and the pins were turned at 8 am. After such a good reaction from Theo the previous day, I wasn't prepared to see him in so much pain this time. It was heartbreaking. Even though he had had some morphine 30 minutes before, he cried and tried his best to get away. He took about 10 minutes to settle afterward, then had a good sleep — the morphine had done its job. As the day went on, Theo's swelling got visibly less and he started to enjoy his surroundings again. He found that he could poke his tongue out and was really enjoying our reaction when he did it. At this stage, there was some bruising becoming evident at the incision sites. Although not nearly as much as I thought there would be — just the yellow tinge you get when bruises fade. He was amazing me with how he was handling all of this and how his body was reacting to the procedure and healing so quickly.

The next 6 days went by much the same. We were discharged from hospital and spent time at the Ronald McDonald House, which was only a 3-minute walk away. Every morning we went to the ward at 7 am, gave Theo his morphine elixir, and had his pins turned at 7:30 am. He would always cry and be in pain at the turning, taking about 10 minutes to settle again. As the jaw came forward, he seemed to feel it more each time and would take a bit longer to settle. The blessing was that we would take him back to the room and he would sleep for about an hour, waking up happy, smiley and ready for a fun-filled day again. I am sure he came to dread the mornings, as did I, but there was no stopping now. It had to be completed. There was no point coming this far and not continuing. During these 6 days Theo had two more x-rays, just to see what was happening inside his mandible.

Dr. Bartlett was happy with everything so far and wanted to see if we could squeeze another millimeter or two out of the devices. On Thursday, Theo had already been distracted 16 mm. The devices, on paper, were only meant to be able to go 15 mm. But as Dr. Bartlett tried to turn

“...the bigger picture is becoming much clearer now”

the pins carefully to get a bit more out of them, he found that he could actually go another 2 mm on each side. To say Theo hated it was an understatement. This was by far the worst morning, as things were obviously really hurting in Theo's jaw. But in the end we managed 18 mm in all — a huge difference in only 9 days.



Theo, 8 mm distracted.



Halfway there!



Pin sites — the bits they turned (one above each ear).



Our cheeky boy and his dad, 18 mm.

And boy could we see it. Theo finally had a chin in the usual place you would find a chin. His top and bottom lips could meet. We could see his bottom teeth when he opened his mouth, rather than having to peer into his mouth while holding his bottom lip out of the way! He had what I refer to as ‘chops’ — the part of your jaw under your ears, which often gets chubby on babies. His whole face looked fuller and more round. It was a transformation. The x-ray confirmed that his airway was open and his tongue was sitting in the correct place. It couldn't have gone any better. My boy was a star. He was still here, he was smiling and he was happy. That's all I wanted.

Yes it was tough to see him in pain, but the bigger picture is becoming much clearer now. We know that this is only the first of several mandibular distractions for Theo, but he came through it with flying colors. Now all we have to do is wait for 6 weeks for the new bone to grow and harden, then the pins can come out. What a day that will be. His little body is creating a miracle while we wait for that day — growing new bone at a great rate, fixing the gap that was created. Will Theo be able to get his tracheostomy removed after all this? We don't know the answer to that question yet, but we do know that we are a huge step closer to that result than we were 2 weeks ago.

This has been an amazing process to go through — a difficult one, but life-changing also. Theo has taught me so much in these 2 weeks. If I were in his shoes I would have been sore, felt sorry for myself and generally wallowed in self-pity for as long as anyone would give me the attention. Not my brave little boy. He cried when it hurt, wanted cuddles and love to help him feel better, then just got on with his day. He had a life to live and nothing was going to stop him from doing that. What an amazing character that shows. That at 9 months of age he just knew that it would stop hurting and he could carry on learning, growing and living. I knew there was a reason this special boy was in my life and he has taught me so much in such a short time. I can't wait to see the other lessons he has for me on this journey that we are taking together.

Another family's view — Peyton's jaw distraction

by Tracy Smith
Elk Grove, California

Choosing a surgeon and method

Six weeks prior to Peyton's birth my husband and I discovered through an ultrasound she had a small jaw, but the doctors weren't able to give us a diagnosis at that time. So when she was born via emergency c-section 4 days after her due date, we were not completely surprised that there were medical issues. She was transferred from our local hospital to the NICU at UC Davis Medical Center within hours of her birth.

Due to a difficult intubation that first day, we were given a recommendation to have her trached. The doctors were concerned that should the breathing tube ever fall out or be pulled out, she may not survive the time it would take to reinsert it. Knowing very little about micrognathia, my husband and I trusted in her physicians and agreed to the trach. A few weeks later, we also agreed to a g-tube when she failed several feeding tests. Although a bottle was attempted in the first few months of life, beyond a milliliter or two, Peyton was 100% dependent on the g-tube.



Ami and Adam meet Tracy and Peyton at the hospital.

Peyton was discharged from the NICU at 6 weeks old. We were told to keep her healthy and fatten her up and around 6 months of age she should be a candidate for an external jaw distraction. At 6 weeks old the surgeon felt her jaw was too small for an external distraction and her jaw was one of the most severe he had ever seen. As she neared 6 months old, UC Davis took another CT scan and gave us their evaluation. The surgeon planned on an external distraction as soon as they could work her into the schedule. While still inpatient and within 7–10 days of placing the distractors, he would attempt to decannulate (remove the trach). He would repair her cleft palate at 15–18 months of age. Although the “d” word was music to our ears, my husband and I both had an overwhelming feeling that this surgeon was not as experienced as we felt she deserved and we sought opinions from two other surgeons.

A surgeon at Primary Children's Medical Center in Salt Lake City, Utah, also recommended an external jaw distraction as soon as it could be scheduled. Since we live in Northern California, we would need to stay in Salt Lake for several weeks until she was discharged and then return every few weeks for the next several months. He felt the cleft palate should be repaired before attempting the decannulation and that would be many months into the future. The surgeon came highly recommended but the travel involved seemed overwhelming.

“Three different surgeons and three different opinions”

Our final evaluation was with Dr. Schendel at Lucile Packard Children's Hospital at Stanford — a 2-hour drive from our home. While waiting to get into the exam room, we had the opportunity by sheer chance to meet Ami and her son, Adam, who both have Nager Syndrome. What an amazing experience! Adam had just completed his first jaw distraction and we could actually see what the end result would look like. After a quick photo of the mothers and children together, we were off to the exam room. We immediately felt comfortable with Dr. Schendel. His nurse had told us previously that he had treated 11 other children with Nager's over the years. We knew he was knowledgeable but he was also warm and friendly. His recommendation was to use an internal distractor. He felt it may take two attempts to get the length we needed. After completing the distraction he would allow 3–4 months to heal and then we would return to have the distractors removed and the cleft palate repaired. Due to ankylosis in one of Peyton's TM joints, Dr. Schendel would attempt to “loosen up” the joint at the time he removes the distractors. After another 6–8 weeks of healing, we would then begin the process of scoping Peyton's airway in preparation for decannulation.

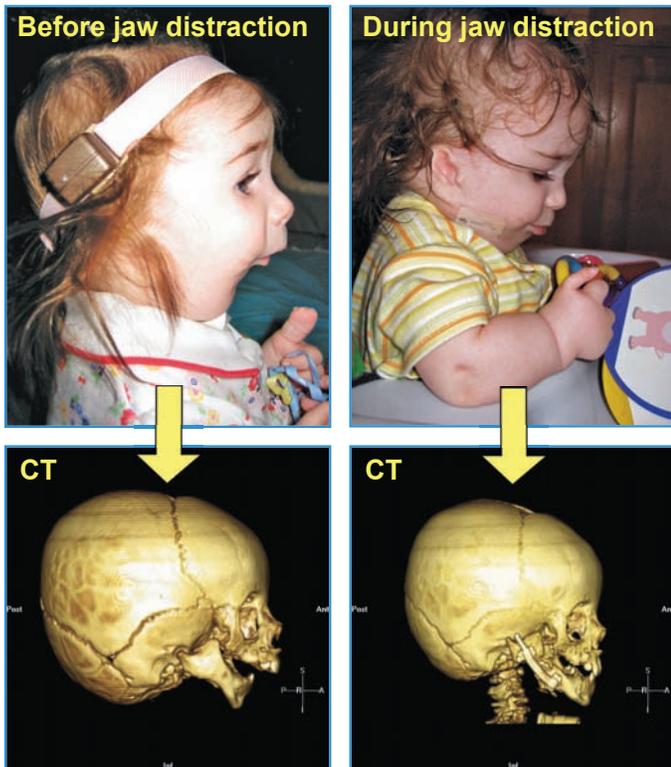
Three different surgeons and three different opinions. After some discussion, we felt confident that Lucile Packard was where we were supposed to be. We made an appointment for surgery and the next 2 months flew by as we tried to ensure Peyton remained healthy.

Surgery

Monday, June 2, 2008, was the big day. Peyton was in surgery for several hours. The Osteomed Logic Jr. distractors were implanted on each side of her jaw. Pins were inserted right above her ears and would be turned by a hex screwdriver. By Tuesday morning, Dr. Schendel was already turning the pins. There was a lot of swelling but it seemed to peak at 2–3 days and then started to reduce a little more each day. One side was much more swollen than the other, which we were told is normal for this procedure. Peyton was back up to full g-tube feedings within 24 hours of the surgery and tolerated them well. Peyton was admitted on a Monday morning and discharged on Thursday morning — only three nights in the hospital. The rest of the distraction process would be done at home.

Going home

The care at home was very simple. We would clean the pin sites several times a day with a hydrogen peroxide/water mix and then



Peyton before distraction and her related CT scan (left top and bottom pictures), and during distraction (right pictures) showing the device in place and bottom jaw lengthened.

apply ointment. We were given several options of how often to turn the screws: 3 turns 1x a day, approximately 24 days to complete; 3 turns 2x a day, approximately 12 days to complete; 6 turns 1x a day, approximately 12 days to complete. We chose the second option to only turn a little at a time, but still resulting in a shorter process all together by doing it twice a day.

Followup

Peyton was seen one week following discharge to monitor the growth. At that time we were told that she had enough growth already that a second jaw distraction should not be needed for several years.

We returned a week later to have the pins and tape removed. Although the removal of the pins was painful, it only took a few minutes and within moments of picking her up, Peyton was smiling and back to her pre-surgery self. The nurse explained that older patients have indicated that there is a tremendous sense of relief when the pins are removed after weeks of the pressure it puts on the skull.

The change in Peyton's demeanor since the pins were no longer being turned and then eventually removed was remarkable. She was back to the happy baby we had thought we might never see again.

Lessons learned

There were several lessons learned. We would not have discharged until Peyton's system was back to normal. She had gone almost a week without a bowel movement and within a few days of being home we had to perform an enema to avoid an emergency room visit. We have since been told that most hospitals will not discharge a child if there has not been a bowel movement and unfortunately, we didn't realize constipation was very common following surgery — and could become such a serious problem very quickly.

Although the professionals could prepare us for turning the pins, how to clean the site and how to administer the medications, there was no way they could adequately prepare us for the emotional anguish we would feel inflicting pain on our child several times a day. Peyton has always had sponge baths at night on her changing table. After several days of turning the pins on the changing table she would start sobbing as soon as we would lay her down — even if we were just changing a diaper. Despite Motrin every 6 hours and Tylenol with Codeine 30 minutes prior to turning of the pins, Peyton was miserable. We discovered that once the Tylenol kicked in she seemed the most comfortable, but as soon as we knew it had kicked in we had to turn the screws so it was short-lived. We know that she was in significant amounts of pain because she was normally a very easy going child, but during the pin turning process at home, she would cling to me and start crying as soon as I tried to put her down. While she had been in the hospital she was getting Morphine in place of the Motrin and that did a much better job of controlling the pain. She would cry for 15 minutes or so after the pins were turned and then fall asleep. At home, the Motrin just wasn't enough. In addition, one of her top teeth erupted just as we were finishing up that only added to her discomfort. When we need to do this process again in the future, we will insist on stronger pain medications or will keep her inpatient so that she can get the stronger pain medications she needs to stay comfortable.

Life has changed — a little

We are only a few days out from the pins being removed. Despite that, we have already noticed some changes. Trach changes and daily trach care are more difficult now that Peyton has a chin. In addition, she has just discovered her trach since her "new" chin is hitting up against it. She has learned that if her trach filter is off, she can cover her trach with her chin and make interesting sounds.

While the pins were still in place, Peyton would start crying each time she would yawn. She would barely open her mouth and we never saw her tongue. She refused to use her pacifier any longer. In the two days since the pins were removed, she is back to opening her mouth as wide as we had seen prior to surgery. She is slowly introducing toys back to her mouth and using her tongue to explore past her teeth. Her first feeding therapy appointment was the day before the pins were removed so we didn't make



Post-distraction (25 mm) and happy to try new foods.

much progress as she would not open her mouth wide enough to take any baby food. Since the pins have been removed, she is tasting the baby food with her tongue and using the LilDippers by Gerber to get it just a little past her teeth.

We are now on hold for the next several months to allow time for the jaw to heal. Our goals are to continue to introduce small bites of baby food to try to get her to stretch her mouth even farther and develop the muscles in her mouth.

Obviously, the most dramatic difference is her appearance. Over the next several months the swelling will continue to reduce and eventually most observers would never guess that she had any surgery or how small her jaw used to be. We are very pleased with the outcome and hopeful this procedure will allow for a decannulation in her future.

CCA/FNMS 2008 Retreat



Tinka Gaffney and her dad, Frank van den Elzen, help decorate a surfboard at the CCA meet-and-greet ice cream social.



Erica, Carla, Sherman and Maurice Perry pose with college-bound Bridget Ieronimo.



Derrick Wolf and his mom, Patty, relax on a comfy hotel couch.



Lucas Bueno waits for the bus to take his family to the aquarium with his mom, Lanessa, and sister Nina.

"We often get asked why we go such a long way to the FNMS conferences, as it is costly in terms of time and money, and our children are adults now. We reply that we are a family and families support one another. Our adult children are there to act as role models and mentors to the younger children coming on, and as an encouragement to the parents, just as families with older children were our support and mentors when we went to our first conference.

We have been to every conference since Margaret Ieronimo-Hogan planned the first one in Chicago in 1996. Each conference has been special in its own way. We enjoy them all, but feel we get the most benefit from the time spent "just us" as the FNMS family. We share joys and sorrows interwoven, and we all know that the smallest milestone is huge for our children. We learn about new medical issues and treatments, and share the joy of new members in our family.

We survive without the contact, but we thrive and grow and do so much better when we share time, experiences, knowledge and information as a family. We are very isolated here in New Zealand, with our family having the only children with Miller Syndrome, so getting together with the other families at conference continues to be important and special for all of us in our nuclear family.

Thank you DeDe and Margaret for all your hard work behind the scenes, that makes each and every one of our conferences so worthwhile and valuable to our family. We had a great time in Myrtle Beach, even if we did 'cook' a bit in the heat, since we had just come from our mid-winter!"

Carla, Maurice, Janelle, Erica and Sherman Perry
New Plymouth, New Zealand

This year the Foundation for Nager and Miller Syndromes (FNMS) joined the Children’s Craniofacial Association’s (CCA) 18th Annual Cher’s Family Retreat, which was held this year in Myrtle Beach, South Carolina, from June 26–29, 2008. Nine FNMS families joined another 90 families from CCA enjoying everything from an ice cream social, Ripley’s Aquarium and Broadway at the Beach, dinner, dancing, a raffle, talent show and lots of trips to the beach. After many of the families went home, FNMS spent another cherished day together playing miniature golf, swimming, and a special FNMS dinner where we spent time working on the next Heart of Glenview submission, sharing information, and celebrating MacKenzie Kehler’s 11th birthday. We were all happy to be together and wished we could be sharing that time with even more of our families. Perhaps we’ll see you all next time!

DeDe Van Quill with Jackson’s sisters Paris (left) and Peyton at Ripley’s Aquarium.



Cristy González, Bridget Ieronimo and Margaret Hogan.



Jackson Van Quill and David González check out the hotel swimming pool.

“My favorite moments are when I see the children playing together as if they were brothers and sisters — they know they look alike and they are free to be who they are. David said about Jackson, ‘He is a little me’, and with those words I realize that we are all part of a big family.”

Cristina González
Guadalajara, Mexico



Riding the waves in the warm Atlantic Ocean: Lucas Bueno (far left), muscleman MacKenzie Kehler (middle) and David González.





GOLF!



HEARTS!



Quinn Stevens, Harlena Morton, Sharon Sawyer and Brittany Stevens celebrate at the FNMS family dinner.



"The Myrtle Beach retreat was wonderful! It was great to be able to meet with families from FNMS and CCA. I truly appreciated having the opportunity to sit down and talk with teens and parents. We were able to learn more about each other and therefore, able to help and share information. The various settings, (such as the beach, the hotel pool, Boardwalk at the Beach, the heart painting and interview activity, the outdoor breakfast as well as the dinner dance) provided unique opportunities to help me and my family remember the purpose of getting together. My children had a fabulous time! Overall, this was one of the best retreats we've attended. The combination of large gatherings as well as intimate meetings was perfect. The only thing that could have made the retreat better would have been to have more FNMS families present. Finally, my family was blessed with help from FNMS and CCA in the form of partial scholarships. We would not have been able to attend this retreat without the generous help of donors from both organizations. We are very thankful for the financial support. Hopefully, the fundraiser I'm hosting in honor of my daughter's 18th birthday in August will assist other families in need in the future."

Harlena Morton
Philadelphia, Pennsylvania, USA



Tinka Gaffney with parents Frank and Leslie.



From left: Frank van den Elzen, Ryan Van Quill, Alex and Lanessa Bueno, Paris and DeDe Van Quill, and Cristy González.

MacKenzie and Greg Kehler are bachelors for the weekend. Happy Birthday Mac!

"The conference was fantastic! Mac and I had a great time 'batching it'. It would have been nice if Joanne and Emma could have made the trip, but maybe next time. Myrtle Beach was a great location — we both had a blast at the beach and had great times on the mini golf courses. The night at the dance was fab. I've never seen Mac dance so much! He had a blast with all the group dancing that was happening. We (I) remembered a few people from Atlanta when both groups were together last. I was sad that more FNMS families were unable to attend, but it was great to see the Kiwi's again, reconnect with the Buenos and the rest of the contingency."

Greg Kehler
Sylvan Lake, Canada



FUN!

Our daughter, **Tiffany Castillo**, (Long Beach, California) turned 15 years old and had her traditional quinceanera party. It was beautiful and she was very happy. Isn't it amazing that she is 15 years old now after the doctors told us that she would not live?



Our Shining Stars!

In our last newsletter issue we shared stories on some of our talented FNMS stars. Here are several more, though they are only a small fraction of our amazing kids' accomplishments.

My name is Charles Randall and I have a 10-year-old son named **Zachary Randall** who has Nager syndrome. We live in Mesquite, Texas. My son has had to deal with all of the normal problems associated with Nager syndrome: small jaw with multiple surgeries, thumbs and fingers, trached (not anymore as of age 6), and problems eating as well as hearing. He has a BAJA for hearing. I bought him a Little Tykes drum when he was 1 year old and he beat that thing senseless. I then bought him a little drum set when he was 2 — he destroyed that thing in 6 months. When he was 3 years old I bought him a First Act drum set. He played that for 3 years. At 6 he got a CB700 full-size adult drum set. He played that until age 8. He now plays a \$2000 Pearl ELX black burst set. He has always been creative in his drumming and has been taking professional lessons for the past 2 years. Zachary practices and plays everyday. His love and passion for drums and percussion simply blows me away. I have always said I would do anything I could to help my son have a better life, and I believe this is what he was put here to do. Zachary is very popular and well liked at school, which really helps. He has played with other musicians and has a couple of future projects in the works. Not bad for a 10-year-old Nager kid.



Bridget Ann Ieronimo graduated from Glenbrook South High school in Glenview, Illinois, on June 1, 2008. The Science Department awarded Bridget for outstanding achievement on the State Horticulture Team. After summer, Bridget plans to attend Oakton Community College in Des Plaines, Illinois, to study graphic design and pottery.

James (Jim) D. Troka
April 5, 1956 – November 30, 2007



Jim was close to his niece Bridget Ieronimo, who was the inspiration for starting FNMS in 1989. Both Bridget and FNMS were dear to his heart. Through your generosity, Jim's loving spirit is able to continue to help those with Nager and Miller syndromes. He would be so touched and grateful for your generosity. Please know the entire Troka family appreciates your thoughtful prayers, masses and donations in his memory.

Jim enjoyed fishing on Lake Geneva and hosting his family's Father's Day BBQ.



Mary Eileen Aamodt
 Mary Pat Adams
 Mary Adler
 Mr. & Mrs. Terry Athas
 Beth Atkins
 Michael P. Atkins
 Rosemary Beaumont
 Mr. & Mrs. Chuck Beck
 Mr. & Mrs. David Bernauer
 Paul Bernin
 Susan M. Bohm
 Tom Briska
 Cheryl Bronkema
 Regina A. Brown
 Vernon A. Brunner
 Mr. and Mrs. Wallace Butler
 Mr. & Mrs. Richard D. Cagney, Jr.
 Mr. & Mrs. Jack Cerone
 Mr. & Mrs. Patrick Cerone
 Mr. & Mrs. Robert Cerone
 Robert J. Cinq-Mars, Jr.
 Sr. Patricia Coughlin
 E.A. Cushman
 Eugenia M. Dallas
 Richard F. Dousman
 Joan A. Doyle
 Jere A. (Troka) Edel
 John Ehlert, Jr.

Susan Fegan
 Joseph F. Finn
 Meg M. Fiorenza
 Barbara Forst
 Chip Frank
 Mr. & Mrs. Robert Friedman
 Nancy C. Gallivan
 Mr. & Mrs. Michael Georgopoulos
 Susan Mary Gibbs
 Salvatore Giganti
 Mary Beth Ginley
 Mr. & Mrs. Dennis Glynn
 Margaret Green
 Gary L. Griffin
 Mr. & Mrs. Gary Griffin
 Donna Grzegorzczuk
 Mr. & Mrs. John Haben
 Dr. & Mrs. Robert Havey
 Mr. & Mrs. Gerard Hillsman
 Mr. & Mrs. Frank Hogan III
 Mr. & Mrs. James Hogan
 Hon. & Mrs. Michael J. Hogan
 June Traina Holmes
 Judith R. Huff
 Mr. & Mrs. Frank Hussey III
 Maureen Jacobs
 Lois J. Jedd
 Madelyn B. Kaufman

Maureen Keane
 Grace S. Kelly
 Mr. & Mrs. Michael J. Kelly
 Mr. & Mrs. Jeffrey Kruckman
 Andrew J. Kubisco, JR.
 Barbara J. Larson
 Mr. & Mrs. Jerome Latin
 John G. Locallo
 Mr. & Mrs. Lawrence Lucas
 John J. Lynch
 Peter Maloney
 Mr. & Mrs. David Mankowski
 Nina McNulty
 Mr. & Mrs. Kevin Medick
 Barbara Monahan
 Mr. & Mrs. James Murphy II
 Thomas R. Nangle
 John P. O'Malley
 Mary G. Osmanski
 Marjorie Ostrowski
 Claire Owen
 Mr. & Mrs. Robert Perkaus
 Mary H. Perry
 Mr. & Mrs. John Person
 Mr. & Mrs. George Pontarelli
 Jeanne Potokar
 Jan. A. Puoci
 Mr. & Mrs. Joseph Rago

Mr. & Mrs Wayne Rapp
 Ellen A. Russell
 Michael Russell
 Jeannie Sargent
 Mr. & Mrs. Donald Schmidt
 Douglas D. Schultz
 Joel Schwartz
 Sandra L. Simon
 Mr. & Mrs. James Snediker
 Buddy Sokolick
 Angela Scully Steger
 Essie & Rick Streicher
 John J. Strickland
 Suzanne M. Sullivan
 Carole C. Tarpey
 Rebecca C. Thompson
 Mr. & Mrs. William Troka
 Colleen (Bowler) Vandendreissche
 Mr. & Mrs. Larry Van Der Bosch
 Mr. & Mrs. Mark Van Guyse
 Mrs. Richard Vlerick
 M. H. Wade
 Flora Ward
 Mr. & Mrs. Thomas Zanoni



FNMS

13210 SE 342nd Street
 Auburn, WA 98092
 1-800-507-3667
 fax 253-288-7679
 www.fnms.net

PRESORT STANDARD
 U.S. POSTAGE
 PAID
 SEATTLE, WA
 PERMIT NO. 1441