

## Good Vibrations Research Study Update

By Ellen Fung, PhD, RD, Associate Research Scientist,  
Department of Hematology

Patients with thalassemia have many risk factors for bone fracture. Currently there are no non-drug therapies available to patients to help build strong bones. Therefore, the “Good Vibrations” research study was conducted at the Children’s Hospital & Research Center Oakland, between March, 2010 and December, 2011. The intervention consisted of subjects standing on a vibrating platform (the size of a large bathroom scale), for 20 minutes a day, 6 days per week for 6 months. Eighteen patients completed the study, and though it was a rather small group, we found that the vibration platform did improve bone health. Whole body bone mineral content increased by 2.6% and bone mineral density by 1.3% in the adult patients after just 6 months. Though these appear to be small changes, they are quite

exciting for such a small group of subjects in a short period of time. The results are not as clear for the adolescent subjects in the study. This is the first report using this technology (low magnitude mechanical stimulation) to improve bone density in patients with thalassemia. The results suggest promise for this noninvasive intervention in a group of patients with significant risk for bone pain and fracture. The results were recently published in the American Journal of Hematology (Fung EB et al. The effect of whole body vibration therapy on bone density in patients with thalassemia: a pilot study. AJH 2012;87:10:E76-9).

Many of you are frequently asked to participate in research studies here at the hospital, and we are so grateful that you are willing to do so. It is because of you and your



efforts that research such as this can be done and new types of therapies can be tested. “Vibration platform therapy” is still quite expensive (currently \$1,650 per platform), and it is not covered by insurance. However, now that we have shown it can be successful in thalassemia, perhaps we can encourage insurers to invest in these types of therapies. Thank you to all who help with research at Children’s. We look forward to working with you on future research projects.

## Experts on Thalassemia Needed

The California Department of Public Health Genetic Disease Screening Program has received a grant from the Centers for Disease Control and Prevention (CDC) to conduct health promotion in thalassemia, and we’re looking for people to advise us as we get started. We need Californians knowledgeable about the day-to-day impact of thalassemia on the lives of those with the disorder and their families to discuss their experiences with us and help guide us as we determine what kind of information we promote and to whom. Conversations can take place via phone or email or during clinic visits to Children’s Hospital Oakland, and travel to/from any meetings attended will be reimbursed.

### CONTACT/MORE INFORMATION:

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## Thalassemia Research and Care—An Update Third Edition, 2012

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# Italian Catholic Federation (ICF) Convention Blood Drive a Huge WIN!

By Laurice Levine

One of Jane Dianda's first goals as the Italian Catholic Federation's (ICF) Grand President was to revive the blood drive and organ donor awareness committee that was first created by Al Teglia during his presidency. Thus, the "Live to Give" Committee was named. This year, it was obvious that Jane's decision was a wise one as the "Live to Give" Committee put on its first blood drive at the ICF Convention and it turned out to be a tremendous success.

At the helm of the "Live to Give" Committee is Nina Malone, Life Member of the Central Council. Working with Nina are Al Teglia, Life Member Emeritus and Past Grand President of the Central Council, Lisa Crudo (recently nominated to the Central Council), and Laurice Levine from the thalassemia outreach program at Children's Hospital Oakland. The crew was excited to get the blood drive going and hopeful that people would respond.

The ICF always steps up to the plate, and such was the case again with the blood drive. United Blood Services (UBS) was able to collect 26 viable units of blood from 21 generous donors. Blood is always in higher demand around holiday weekends such as Labor Day, so UBS was very grateful for the lifesavers who donated blood. Because each

unit can be broken down into platelets, red blood cells, and plasma, three patients can benefit from one single donation.

Red blood cells, which carry oxygen throughout the body and give us energy, are often used for patients who require surgery, have an organ transplant, have kidney disease, or have a blood disorder such as Cooley's anemia/thalassemia or sickle cell disease.

Platelets are the part of the blood that helps with clotting. Patients with leukemia or who are undergoing a bone marrow transplant (BMT—the only known cure for Cooley's anemia/thalassemia) need platelets, as their bodies do not make enough to control bleeding.

Plasma and its derivatives are used for the treatment of hemophilia, shock, or diseases such as hepatitis.

One person; one unit of blood; three lives saved! Thank you to all of the ICF members who saved a life at convention!

Welcome Jim Acitelli, Central Council Member, to the "Live to Give" Committee. We are honored that you are joining us!

For more information or to schedule an appointment to donate blood, contact:

- Blood Centers of the Pacific, [www.bloodcenters.org](http://www.bloodcenters.org)

- American Red Cross, [www.redcrossblood.org](http://www.redcrossblood.org)

For more information on the "Live to Give" Committee, please contact Nina Malone at [ninamalone0436@comcast.net](mailto:ninamalone0436@comcast.net).



*Danny Centurioni gives peace and blood; Lifesavers among us.*



*Queen Chelsea Spencer and her Court; Chelsea Spencer*



# Italian Catholic Federation (ICF) 2012: In Giving We Receive

Dr. Vichinsky, Laurice and Matt Levine, and Laurice's mother, Beverly Compagno, were welcomed with open arms at the ICF's 81st Annual Convention in Reno, Nevada. As always, it was an event to remember! The ICF donated \$65,000 for the thalassemia fund at Children's Hospital Oakland, and then ICF Past Grand President and Life Member Nettie Descalso DelNero presented Dr. Vichinsky and Laurice with an additional \$10,000 from the Robert Kemp Estate.

The total contribution of \$75,000 is what keeps the thalassemia outreach program running, especially during these times of ever-increasing economic challenge. ICF funds support medical

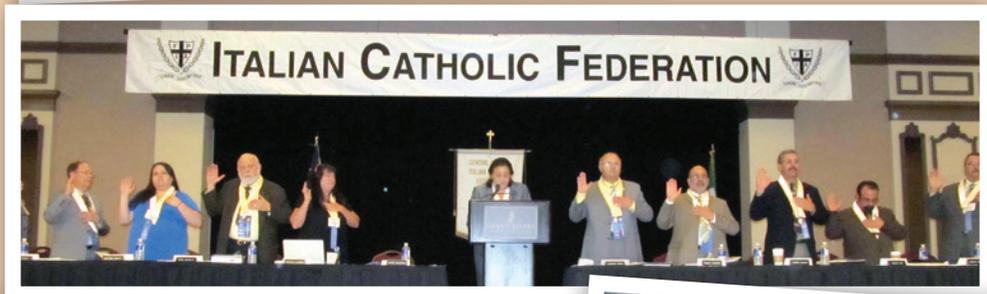
research, which improves the quality of patients' lives, as well as their longevity; community education and outreach; comprehensive care for patients whose parents cannot otherwise afford their care; patient sponsorships and assistance to travel to our center for consultation; and psychosocial programs and events for patients and families, such as our holiday party. The ICF's tireless hard work, immense generosity, and endless support make these endeavors possible. There are not enough words to express our gratitude to the ICF.

This was an exceptional year for Laurice as she received the Pope John Paul XXIII Award, the ICF's greatest honor. "I

will cherish this award always and hold the memories deep in my soul. Thank you to everyone who made this award possible and thank you to the ICF from the bottom of my heart," Laurice stated at the convention. We also feel a deep sense of gratitude for once again being adopted us as the ICF's national charity; we hope that the ICF will continue to utilize our services by inviting us to their branches or districts for meetings and events so that we may provide education and updates on thalassemia and our program. Once again, we thank the ICF for their generosity, kindness, friendship, love, and support.



*ICF members enjoy the banquet; Central Council Members hard at work; Rosemary Cook winner of the ICF Young Adult Leadership Award with her family; Grand President Jane Dianda honors Laurice with the Pope John Award.*



**Many thanks to our donors for their generosity, and support!**

Italian Catholic Federation

Chelsea Spencer and the Italian American Federation

Ligure Club

Charles Abella

Anna and Louis Alberti

Marlene Farber

The Head Family

Naveana Nail Salon on behalf of Bryan Tran

Rosa Radicchi

Mary and Louis Salib

Mary Sperrazzo

Michael Tom

Joanna Van Blaricom

Christine M. Wagstaff - Trustee of the Estate of Robert Kemp

# Interview with Olivia Stahl

**Q: How did you hear about the Painted Turtle Camp and what made you decide to go?**

A: I heard about the Painted Turtle Camp from my doctors down in Oakland. I think what really made me decide to go was that I really wanted to meet people who had similar diseases and who could relate to medical things I was going through. Also, it was a way for me to kind of step out of my comfort zone a little bit. It also happened to be in sunny Southern California, and I didn't want to miss a traveling opportunity like that.

**Q: How did you feel about going to camp for the first time?**

A: Honestly, I was scared to death. I was scared that I wasn't going to meet anyone, or that no one would like me. It was like the first day of freshman year all over again... gahh! But at the same time, I was excited to meet new people, try new things, and take risks.

**Q: What did you do at camp (activities, etc.)?**

A: I went swimming, horseback riding, and fishing; made various arts and crafts; and bonded with my cabin friends.

**Q: How was the food?**

A: The food was really good. I don't think there was a meal I didn't like there. My favorite dish was probably spaghetti, because we tried to eat it with no hands. That was interesting.

**Q: How many girls were in your cabin?**

A: There were seven girls in my cabin, plus the six counselors.

**Q: What were your counselors like?**

A: Oh, man, our counselors were super sweet and caring, and they really took the time to get to know each one of us. I got pretty close to a couple of the counselors, which made it super hard to say goodbye. In all honesty, by the last day and a half, the counselors and the campers were sort of butting heads a bit. Let's just say everyone

in the cabin was a bit "sassy." But at the end of the day, we still loved them to death. I thought of some of the counselors like the older sisters I never had.

**Q: What was the highlight of camp? Were there any challenges or hard times at camp?**

A: The highlight of camp was getting to know everyone from the green cabins. For the first time ever, I finally got to meet people with similar blood disorders who know and understand what it's like to get poked all the time, or to get lots of blood transfusions, or to take a ton of medications—to not feel judged or get bombarded with questions is just the best feeling in the world. To know you're not alone and that there are other people going through the same thing is truly amazing. I feel somewhat blessed to have a blood disorder and to have met people who understand what it's like and who can share a common bond. Another cool highlight was the day our cabin went boating/fishing. It was our cabin and one of the other green cabins, which happened to be one of the older guy cabins. I wanted to go fishing, yet all the other ladies wanted to go boating. So I was the only girl from my cabin who went fishing, and I got stuck with all the guys. But I also happened to catch more fish than any of the guys, which was pretty exciting. I caught four fish with a little help from my wonderful counselor, Kailyn, who I dragged along with me. There weren't really any challenges or hard times at camp. I mean, if I had to pinpoint one thing, I think for me it would have to be not having contact with my family. I was a bit homesick at first, but that goes away.

**Q: If a child or teen was not sure about going to camp (or a parent was not sure about sending their child), what advice would you give them?**

A: Go! Go! Go! Seriously, you won't regret going. It definitely is nerve-wracking going to a new place, not knowing anyone, but when you get there, you get so



*Thuy and Olivia at Camp*

caught up in the activities and meeting people that you forget about nerves. The first day, you're getting to know your cabin mates and the next thing you know, you're leaving your new "cabin family." It goes by so quickly, so enjoy every day of it.

**Q: Are you planning to return to the Painted Turtle Camp?**

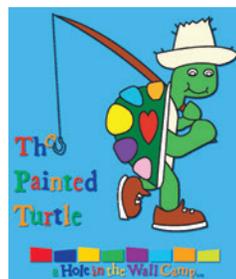
A: Most definitely! I would love to go back as a LIT (Jr. Counselor in Training) or maybe a counselor later. The experience there was really eye-opening and somewhat life-changing for me, so I definitely would love to get back there as soon as possible.

**Q: Any other comments?**

A: I really recommend going to this amazing camp. I guarantee that you'll walk out with a smile on your face—how could you not? I met some of the coolest people there that I hope I can stay in contact with for a really long time.

**Q: Can you tell me a little bit about yourself—age, hobbies, year in school, thalassemia type (and if you want, blood transfusion and chelation regiment)?**

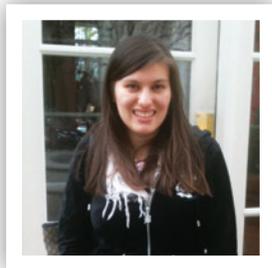
A: I'm 16 years old, I am adopted from Vietnam, and I'm a junior at Lakeridge High School in Lake Oswego, Oregon. My family, friends, health, school, dance, Harry Potter, The Hunger Games, Starbucks, and the NFL are the most important things in my life. On Mondays, Thursdays, and of course, Sundays, I love sitting at home, screaming at my TV, watching pro football. Go, Packers! I have e-beta thalassemia, and I am currently off blood transfusions, but I'm taking Exjade, which is oral chelation, and hydroxyurea.



# Thalassemia Awareness – A Volunteer Experience That Will Last a Lifetime

By Ivanna Pincilotti

Thalassemia is a genetic blood disorder in which the body does not produce enough hemoglobin. Many people have thalassemia trait, but the majority of people don't know what thalassemia is. How could this be possible?



A year ago this June, I graduated from high school. During senior year, my English teacher assigned my class a project. The project consisted of reading a book about a specific topic and presenting it to the class. Most of my classmates had a difficult time finding a topic, whereas I knew exactly what I wanted to talk about: thalassemia. My sister has thalassemia trait, and I wanted to learn more about this disorder. During my presentation, I asked the class if they had donated blood before, and a handful of students raised their hands. Then when I asked the students if they knew what thalassemia was, nobody had a clue. However, by the end of my presentation, my classmates were asking various questions and now had some background knowledge about thalassemia. It became clear to me that not enough people knew what thalassemia is because it is not talked about

in classes, school clubs, or many other places.

After I presented my English project, I knew I wanted to raise awareness and volunteer. Laurice Levine invited me to Children's Hospital Oakland's annual Hematology and Oncology

conference to learn more about thalassemia and to meet other hospital staff members. During the conference, I had a chance to sit down with doctors and staff to further my knowledge on thalassemia. Last August, I started volunteering for the Thalassemia Outreach Department. My first project was to help with the department's annual holiday party. I helped Laurice prepare and execute the party, while also writing many letters to solicit donations.

Around the same time, I was asked to speak about thalassemia at the Student California Teachers Association's monthly meeting at Sonoma State University. During the meeting, the students were engaged with the presentation. Afterward, they were determined to help collect toys for the holiday party. Together, we gathered about 200 toys for the party! Once December rolled around, we were ready to kick off the Rock 'n' Roll Holiday Party. All of the

patients, families, and staff members had a wonderful time. When I saw each child at the party smiling, laughing, and having fun, I knew my goal had been accomplished.

I also had the opportunity to help write donation letters for this year's annual blood drive. Various generous donors gave to the blood drive's raffle. This year, the blood drive collected enough pints of blood to help 60 Bay Area patients. Having a chance to work alongside Laurice is an amazing opportunity; she is a warm-hearted person that wants the best for the patients around her. Laurice isn't only my colleague and best friend, but she is also someone I deeply admire. I am looking forward to planning and executing this year's holiday party! My experience volunteering for the CHO Thalassemia Outreach Department has not only given me the opportunity to receive work experience but has also allowed me to learn more about thalassemia.

Ivanna is currently attending Sacramento Community College, but her goal is to transfer out of state to the University of Washington. When she isn't volunteering, she enjoys spending time with her family and friends, taking day trips to San Francisco, and vacationing—especially to Disneyland.

## Thalassemia Program Donation Guidelines

Due to hospital policy, limited storage space, fire hazard, and the prevention of infectious disease, the Thalassemia Outreach Program at Children's Hospital Oakland has a set of donation guidelines. If you would like to make a donation, please adhere to the following guidelines:

- All toys must be new and unused
- Stuffed animals must be new, clean and not off the store shelf for more than a month
- Books must be new or in very good condition
- Used DVD's are acceptable; used VHS video tapes are not.

### GIFT CARDS

Gift cards are great for ages 10 and older, especially our teens and adults. We also are able to purchase the many supplies, arts, crafts and other wares needed to put on a party, You can designate if you would like your gift card to be used as a gift, as party expenses, or wherever needed.

**Best options for gift cards:** Target, Best Buy, Game Stop, Safeway (for party supplies), Albertsons (for party supplies), Gap, Old Navy, Banana Republic, Claires, See's Candy, Barnes and Noble, Big 5, Sports Authority, Regal and AMC Movie Theaters, Costco, Starbucks, Jamba Juice. Denominations can be \$5 to higher. Usually \$10 and \$20 are preferable.

If you would like to make a donation please contact Laurice Levine at 510-428-3885, ext. 5427 or [LLevine@mail.cho.org](mailto:LLevine@mail.cho.org)  
Thank you in advance for your generosity and support!

## PERSPECTIVE

## Italian Youth Giving Back to the Community—That's Amore!

By Chelsea Spencer

The Italian American Federation (IAF) has been a part of the local Italian-American community since 1935. At that time there were several Italian-American clubs in the East Bay. Although they progressed well individually, the leaders of these clubs felt that an association should be formed so that their shared heritage could be celebrated and gain recognition. Thus, the Italian American Federation was formed.

Since 1935, a Columbus Day queen has been appointed to represent these clubs. In the early years of the IAF's existence, the Columbus Day queen was chosen by the number of raffle tickets that were sold in her name.

The queen's coronation was held in October and was viewed as a prestigious event. On the day following coronation, parades and ceremonies were observed throughout the East Bay, thus concluding a joyous occasion, as well as a celebration of the original founder of the American and Italian Heritage. In the present day, the queen is selected by a panel of nonaffiliated judges based on multiple criteria. All prospective queens have to respond to a number of selected questions related to how they would conduct themselves were they to be appointed Columbus Day queen.

In October 2011, Chelsea Spencer was honored to be chosen as 2012 queen for the IAF. Her main responsibilities have been to represent the 14 Italian clubs in the East Bay and to attend their functions. In the past, this has been the extent of a queen's participation. However, as queen this year, Spencer chose to do more. She aspired to bring the Columbus Day Court—which included the remaining girls who ran for Columbus Day queen and herself—closer together under a common goal. The intended result was to encourage young adults to involve themselves more in the clubs and most importantly give back to the Oakland community where their Italian-American ancestors originally established new lives for themselves.

At the outset of her reign, Spencer organized a group meeting with the

princesses, holding a brainstorming session to decide what this common goal would be and how to raise funds in its support. The group chose to support thalassemia (also called Cooley's anemia) research at Children's Hospital Oakland (CHO), and an initial goal was set. Thalassemia is a genetic blood disorder that primarily affects individuals, including Italians, of Mediterranean descent.

The members of the 2012 Columbus Day Court, along with their family and friends, have more than doubled their goal and have raised \$5,900.00 through hard work and determination (\$300.00 will be given to the new incoming court, and \$5,600.00 will be donated). The court accomplished this by setting up and cleaning up for club events and waiting tables. Spencer also organized a small sponsored walk at a local trail.

All 14 Italian club presidents, officers, boards of directors and their members have all supported and embraced the goals

and directives set forth by this year's court. Spencer's reign drew to a close on October

14, 2012, at the Colombo Club in Oakland. This day also marked the coronation of 2012–2013 IAF Columbus Day Queen Michelle West, who will carry on the Italian tradition, as well as establish the goals of her court for the coming year.

CHO Thalassemia Outreach Coordinator Laurice Levine was honored to attend this special event and was present to accept the generous donation from Spencer and the

individuals who helped raise funds for thalassemia. States Spencer, "I am humbled by the love and support of so many throughout the last year. The goals we set and partnerships we developed with the 14 Italian clubs, along with the unconditional support of our families and friends, has truly been inspiring. When I presented the check on October 14, 2012, to Laurice, I did so with the same love and support we have received." Now that's *amore!*



## Volunteers are needed!

### We are looking for:

- **Volunteers for outreach office administration or events.**
- **Writers for Perspectives and thalassemia.com (Have a story to share? Healthcare tips? We want to listen!)**
- **Fundraisers: The economy is a challenge. We need to keep our program going. Please let us know if you want to help.**

If you are interested, please contact Laurice Levine at [LLevine@mail.cho.org](mailto:LLevine@mail.cho.org) or call 510-428-3885, ext. 5427.



## Welcome to Maggie and Clara

Volunteers from Cardinal Newman High School in Santa Rosa



My name is Maggie Leinen, and I am currently a senior at Cardinal Newman High School. My friend, Clara Knapp, and I are working on a senior service project called Project Thal. My goal is to raise awareness of thalassemia and educate our community. I also hope to raise money for research on thalassemia. I

came across thalassemia while looking for a subject for my project and found it fascinating. There is a lot for me to learn about thalassemia, and I love finding new information about it. Apart from doing schoolwork and my thalassemia project, I spend most of my time swimming for the Santa Rosa Neptunes or playing water polo for my school team.



My name is Clara Knapp and, like my partner, Maggie Leinen, I am a senior at Cardinal Newman High School. In my free time, I work as a hostess and sing for my church youth group, in addition to currently participating in the school play. For my senior service project, I will be working as a volunteer with Laurice Levine and Children's Hospital Oakland to raise awareness of thalassemia within my Santa Rosa community.

I have a lot to learn about both the medical side of thalassemia and the psychological side, as well. I hope to learn more about how patients with chronic illness feel it affects them and observe how treating their illnesses ultimately contributes to who they are as people. I enjoy serving the community and am excited to volunteer and get to know the patients I will be working with personally.

## UPCOMING EVENTS

### DECEMBER

**Dec. 13, 2012:** Asian Outreach Advisory Meeting, Alta Bates Summit Medical Center, Oakland, CA.

### UPCOMING EVENTS IN 2013

**Jan. 13, 2013:** ICF Branch #425 Green Valley and ICF Branch #436 Glendale Meeting/Thalassemia Presentation, Tucson, AZ.

**Jan. 30, 2013:** Blood Drive in Honor of Thalassemia at Cream, Berkeley, CA.

**Feb. 12, 2013:** ICF Branch #343 Thalassemia Presentation/Dinner Meeting, Sebastopol, CA.

**March 2, 2013:** ICF Branch #217 Thalassemia Presentation/Lunch, Redlands, CA.

**April 20, 2013:** Second Thalassemia Wellness Conference, Children's Hospital Los Angeles, Los Angeles, CA

**May 8, 2013:** 12th Annual Blood Drive in Honor of World Thalassemia Day, in collaboration with Blood Centers of the Pacific, Oakland, CA.

## BLOOD DRIVE IN HONOR OF THALASEMIA JAN. 20, 2013

Please come out and support us!



www.creamnation.com



www.bloodcenters.org



**Children's Hospital Oakland  
C.R.E.A.M  
Blood Centers of the Pacific**

**are collaborating to hold a blood drive in honor of thalassemia**

**EVENT LOCATION:**  
2399 Telegraph Ave., Berkeley  
Near UC Berkeley campus

**DATE & TIME :**  
Jan. 30, 2013, 1-6 pm

**For more information contact  
Children's Thalassemia Department:**

- Email LLevine@mail.cho.org
- Call 510-428-3885, ext. 5427

**Please sign-up for an appointment:**

1. Go to [www.bloodheroes.com](http://www.bloodheroes.com)
2. Click on "Donate Blood"
3. Enter Sponsor Code: THAL

# Welcome Thalassemia Interns

By Sushrita Neogi

The Thalassemia Outreach Program at Children's Hospital Oakland is always doing its best to increase thalassemia awareness across the country for various and varied individuals and groups. Recently, we have had the opportunity to welcome—all current students at or recent graduates of UC Berkeley—to our team to create an extremely talented, diligent, and innovative taskforce! These individuals have taken on projects ranging from conducting outreach at health fairs and blood drives to collaborating with various foundations such as the American Red Cross, Blood Centers of the Pacific, and the Alta Bates Asian Outreach group. These interns have also taken the initiative to start a DeCal about thalassemia at UC Berkeley. A DeCal is a student-led undergraduate class. Right now, the DeCal is in the planning stages as the interns create a syllabus, gather lecture materials, and recruit guest speakers. The DeCal on thalassemia is set to start in Spring 2013.

The thal interns are some of the most hardworking volunteers! The following biographies and images will give you a chance to get to know them a little bit better!

My name is **Sushrita Neogi**. I recently graduated from UC Berkeley, majoring in Molecular and Cell Biology, Cell and Developmental. I am currently a volunteer researcher at Children's Hospital Oakland. In my free time, I enjoy reading books, baking, and hanging out with my friends. I have been working with Laurice Levine on thalassemia outreach for over a year, and I am delighted to have this new group of motivated and hardworking interns on board! With their help and support, the thal outreach group has been able to expand and raise awareness on a larger scale. I look forward to a wonderful year of spreading the word about thalassemia and achieving new goals.



My name is **Wendi Gu** and I am currently finishing up my last year at UC Berkeley, pursuing a degree in Molecular Cell Biology, with an emphasis on Neurobiology, as well as a minor in Italian Studies. Not only do I love doing outreach and spreading the word about Thalassemia with my team members at the Children's Hospital Oakland, I also love



volunteering with the Suitcase Clinic, an organization on the UC Berkeley campus dedicated to offering free health and social services to underserved and homeless populations in the Bay Area. In addition, I am doing research in a vision science lab that studies cataract formation in the eye and potential treatment options for this leading cause of blindness. When I'm not knee-deep in schoolwork or extracurricular activities, I like to practice yoga, root for the Los Angeles Lakers & Oklahoma City Thunder, learn new languages, be a foodie, spend time with friends and travel!

My name is **Amina Khimani** and I am currently a second year undergraduate student at the University of California: Berkeley. I absolutely love to dance, run, and play/watch basketball. Aside from dancing and running, I am currently an intern for the thalassemia outreach team at the Children's Hospital: Oakland. When I'm not spreading awareness about thalassemia, I am also involved in Public Health research on pediatric malnutrition. Working on the thalassemia outreach team has allowed me to gain more knowledge than ever before on thalassemia, and in a sense helped me learn more about myself, as I am a carrier of the trait. I hope to expand our outreach as much as possible, and to educate people on what thalassemia really means!



My name is **Jeff Nathan**, and I am a senior at UC Berkeley. I study Molecular and Cell Biology, with an emphasis in Immunology and Pathogenesis, and I have aspirations for pharmacy school upon graduation. My work for the thalassemia outreach team under the guidance of Laurice Levine began in the summer of 2012 and has continued into the fall. Throughout this term, I have enjoyed attending various health fairs and blood drives, as well as an outreach meeting at Alta Bates Hospital. Currently, I am cooperating with the other team members on creating a syllabus for a DeCal class on thalassemia, which will be offered in the spring of 2013. The overarching goal of this course will be to spread awareness and education



about thalassemia, while also providing a forum for student discussion about many of the social, psychological, and health effects of the disease. In addition to my work for the outreach team, I enjoy playing basketball and singing with my a cappella groups. I also work in an environmental toxicology lab on campus, for which I perform various genomic experiments to determine the LD50 for certain local environmental toxicants.

My name is **Andrew Shieh** and I graduated from UC Berkeley in May 2012 with a degree in Molecular & Cell Biology, Neurobiology. In addition to being an active intern for the Thalassemia Outreach Team, I currently work in a lab at San Francisco and also teach and tutor chemistry at UC Berkeley and other tutoring companies in the Bay Area. Aside from spreading awareness, I love to play basketball, ride my longboard, and learn new recipes. My goal as an intern at outreach events is to provide knowledge for my audience so they leave my table having learned something new. My future goals for this program include extending outreach to middle and high schools in the Bay Area, engrossing myself in more literature on thalassemia, and teaching our thalassemia decal as a guest speaker at UC Berkeley. Go Team!



My name is **Pang Vang**, and I am a fourth-year undergraduate student from UC Berkeley, majoring in Integrative Biology with a minor in South and Southeast Asian Studies. My interest in thalassemia started when I took a public health seminar at UC Berkeley and did a research project on the disorder. From there, I started interning for the thalassemia department through the volunteer program at Children's Hospital Oakland, where I worked with Gabe Wong, MSW, for almost a year. I just recently started interning with the thal outreach team. In addition to being a thal intern, I work as a student assistant at the UC Berkeley Early Childhood Program, and I am currently an officer for the Hmong Student Association on campus. Aside from academics, I really enjoy cooking and working out at the gym. My goal as an intern is to learn as much as I can about thal and continue to spread awareness in the Bay Area. My future goals are to help the team create a DeCal on campus and possibly extend outreach to Sacramento and the Central Valley.



*The UCB Thal Outreach Team enjoy dinner at Skates with Laurice Levine; Pang tabling at a community blood drive; Andrew and Wendy at a Blood Centers of the Pacific Blood drive at the Studio Art Center in Oakland.*



# Hemoglobin H Disease

By Ash Lal, MD

Hemoglobin H disease (HbH) is a form of alpha thalassemia in which moderately severe anemia develops due to reduced formation of alpha globin chains. In this condition, as in the other forms of thalassemia, there is an imbalance of globin chains needed to form hemoglobin. Normally, there are four genes to produce alpha globin chains. When three out of four of these genes become inactive, there are too few alpha globin chains to combine with beta chains and give rise to normal hemoglobin (hemoglobin A). The excess beta globin chains then combine with each other to form hemoglobin H, which is the origin of the name “hemoglobin H disease.”

While most individuals with HbH do not require transfusions, there is heterogeneity in the clinical course. This is an important component of the counseling for the family at the first clinic visit. HbH caused by deletion of three genes (deletional HbH) is less severe than cases in which two genes are deleted and the third gene has a point mutation (non-deletional HbH). HbH Constant Spring (HCS) is the most common form of non-deletional HbH in the United States.

In California, all newborns with HbH are identified through newborn screening, followed by identification of the alpha globin gene deletions or mutations by the Hemoglobin Reference Laboratory located at Children’s Hospital & Research Center Oakland. Older patients who are seen for the first time should have DNA testing to identify alpha globin gene deletions and the presence of the Constant Spring (CS) mutation. If only two alpha genes are deleted and the CS mutation is absent, further testing for uncommon mutations should be done. These patients should not be categorized as deletional HbH. Patients with HbH should also be screened for beta globin gene mutations with multiplex PCR. The complete genotype is used as the basis for discussion of a future clinical course and genetic counseling.

## Routine Care

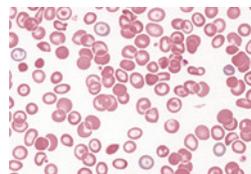
Patients should be seen frequently in the first year after diagnosis to establish hemoglobin level and monitor growth. Communication with the primary care provider is also important so that care can be coordinated. Later visits to a thalassemia center should occur once or twice a year, and routine health maintenance should be provided by the primary care provider. Patients with HCS should be followed closely by the thalassemia center because of the potential for severe anemia, growth delay, iron overload, and the need for splenectomy. All routine childhood vaccines should be completed and seasonal influenza vaccine given every year. All patients should receive folic acid, 0.5 to 1.0 mg per day.

## Management of Fever

Owing to the risk of severe anemia during infections in HCS, such patients should be seen on the same day in the clinic or emergency room. Patients with deletional HbH can usually be seen in the clinic on the next day, unless an ER visit is warranted by the symptoms. A blood count with reticulocyte count and bilirubin level should be obtained. An admission for observation or transfusion may be needed if the hemoglobin has fallen below baseline. Antibiotic treatment is determined by assessing the source of infection. All splenectomized patients with fever should be seen on the same day and started on antibiotics (ceftriaxone is preferred). An admission is recommended until sepsis can be excluded. Oxidant drugs, which cause hemolysis in G6PD deficiency, should be avoided.

## Splenectomy

Splenectomy is not required for deletional HbH disease. It may be required for patients with HCS when there are multiple episodes of sudden fall in hemoglobin level requiring transfusion, or if anemia is severe and affecting growth.



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## Transfusion Therapy

Common infectious diseases, such as common cold or viral fever, can lead to a rapid fall in hemoglobin level in HCS patients. The fall in hemoglobin level in HbH patients is much smaller, and the development of severe anemia needing transfusion is unlikely. Transfusions are given when hemoglobin falls below 6 g/dL. As mentioned above, splenectomy is recommended if there is a need for frequent transfusions. There is little role for chronic transfusion therapy such as that given to individuals with beta thalassemia major.

## Iron Overload

Iron overload occurs in adults with HbH. In HCS, there is early iron overload that may need treatment. Patients’ ferritin and liver iron concentration should be monitored and measured via MRI or ferritometer.

## Outreach

Patients should be provided with a card that shows the diagnosis and emergency contact number for their hematology service. Clinical summaries should be sent to the primary care provider with treatment recommendations. Families should be given a letter for school to explain the need for clinic visits. Children are allowed to determine their own limits of activity during physical education with no routine restrictions.

## Adults with HbH

Genetic counseling is extremely important for adults. Ideally, a full testing of an adult patient's partner for alpha and beta thalassemia mutations should be performed. At a minimum, testing for alpha thalassemia trait should be done to determine any risk for alpha thalassemia major which can be fatal to a fetus.

Patients with HbH who become pregnant should be monitored for further drop in hemoglobin level. Most of these patients will not need transfusions. Pregnant patients with HCS need close observation and should start regular transfusions if hemoglobin drops below 7 g/dL. Transfusions are performed every three to four weeks with the aim of maintaining pre- and post-transfusion hemoglobin levels at 9 and 12 g/dL, respectively.

All adults should have echocardiograms to screen for pulmonary hypertension, more frequently in those who are splenectomized. Older patients, particularly with HCS, should be evaluated for fatigue, difficulty in coping at work, and family stress.

## Conclusions

Deletional HbH is asymptomatic during infancy and childhood, although deficits in growth may appear among older children. These individuals should receive all routine care through a primary care physician, with periodic evaluation by a hematology center. The key points are counseling the family and adopting strategies to avoid blood transfusion. In contrast, HCS is a potentially serious disease that needs close follow-up by a thalassemia specialty center to plan for emergency and elective transfusions, measure iron overload, monitor growth failure, and evaluate the need for splenectomy.

## www.thalassemia.com

CHECK OUT OUR NEW AND IMPROVED WEBSITE:

- New look
- Updated information
- Improved psychosocial and resource section
- Patient stories
- Event calendar and more.

Special thank you to Shanda Robertson for her work on the website.

The image displays two screenshots of the updated website. The top screenshot is the 'Genetics of Thalassemia' page, which includes a navigation menu (Home, About us, News & Events, For Physicians, How you can help, Site Map) and a sub-menu (What is Thalassemia?, Genetics, Diagnosis, Treatment, Research, Links with Thalassemia). The main content area is titled 'The Inheritance of Thalassemia' and features a diagram showing a mother with alpha thalassemia trait (one normal alpha gene, one deleted) and a father with beta thalassemia trait (one normal beta gene, one deleted) having children with various genotypes: alpha thalassemia trait, normal, and beta thalassemia trait. Below the diagram are two karyotypes (46,XX and 46,XY) highlighting the alpha and beta globin gene clusters on chromosomes 16 and 11, respectively. The bottom screenshot is the 'Welcome' page, featuring a navigation menu, a 'Welcome' message, a photo of a doctor, and a search bar.

## Second Thalassemia Wellness Conference

PLEASE SAVE THE DATE:  
APRIL 20, 2013

The Second Thalassemia Wellness Conference will be held on April 20, 2013. The conference is sponsored by the Thalassemia Support Foundation in conjunction with Children's Hospital Los Angeles (CHLA) and Children's Hospital Research Center Oakland.

**Location:** Children's Hospital Los Angeles

**Time/Details:** TBA

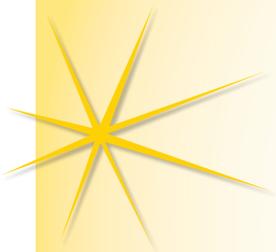
**Information:** Laurice Levine at 510-428-3885, ext. 5427.



## P E R S P E C T I V E S

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Children's Hospital & Research Center Oakland  
747 52nd Street, Oakland, CA 94609

Address change requested



**Dr. Vichinsky, Laurice Levine,  
and the Thalassemia staff  
would like to wish everyone  
a joyous holiday season and  
a New Year filled with  
good health, peace and happiness.**



*Perspectives* is produced by Thalassemia Outreach Coordinator Laurice Levine, MA, CCLS, at Children's Hospital & Research Center Oakland. For questions regarding the newsletter or for more information on thalassemia, call 510-428-3885, ext. 5427, or visit [www.thalassemia.com](http://www.thalassemia.com). If you no longer wish to receive *Perspectives*, please email Laurice at [llevine@mail.cho.org](mailto:llevine@mail.cho.org).