

**A child's hearing loss can be genetic even if no one else in the family has a hearing loss.**

## Genetic testing can identify cause of hearing loss

By Lisa A. Schimmenti, M.D.,  
Assistant Professor, Pediatrics,  
University of Minnesota

Since the start of newborn hearing screening many families have brought their children to a genetics clinic at the advice of their doctor. Both the American Academy of Pediatrics and the American College of Medical Genetics recommend a genetics evaluation and ongoing follow-up by a doctor that specializes in clinical genetics for all children who are deaf and hard of hearing.

Part of my job at the University of Minnesota is counseling families about genetic testing. One family I worked with recently included

Emma, a healthy and happy 1-year-old girl who has bilateral severe sensorineural hearing loss. Bedside hearing screening administered the day after her birth identified a potential problem. Audiology tests at

two months of age confirmed her hearing loss. She had been wearing hearing aids since she was six months of age.

Emma and her parents came to the Genetics Clinic at the University of Minnesota where I practice. When I first met Emma, she was busy exploring the brightly lit room and playing with colored building blocks. Her parents appeared to be unsure what to expect from a genetics evaluation. Emma's mother told me that they brought Emma to the Genetics Clinic on the strength of her doctor's recom-

mendation, but they had their doubts about a genetic cause for their daughter's deafness.

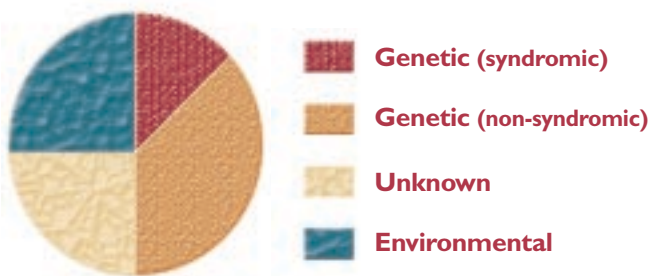
"Neither Emma's father nor I have problems with hearing," the mother said. "We asked our family and went back three generations and not even a grandparent wore a hearing aid. How could Emma's hearing loss be genetic?"

This was a great question, and one that I hear often. To illustrate why genetic testing could help them, I drew a pie chart to show a breakdown of the causes of hearing loss. (See chart at left.) I explained that environmental causes may include infections like cytomegalovirus (CMV). I also told Emma's parents that for a quarter of children with hearing loss the cause will never be known. However, I pointed out that the largest area, the yellow and orange regions, shows that for one half of all children hearing loss is caused by genetic changes. I further divided the genetic category to show that there are "syndromic" or "non-syndromic" forms.

"What is the difference between syndromic and non-syndromic?" Emma's father asked.

I explained that syndrome means "runs with." In about one third of children with hearing loss, a syndrome can be identified. For this group of children, hearing loss "runs with" other medical problems. In a number of common syndromes associated with hearing loss, the medical problems can be hidden. The primary job of the geneticist is to be reasonably certain that the child playing with blocks in the clinic does not have a hidden syndrome.

**Causes of Hearing Loss**



**Half of all hearing loss is genetic. Of that group, one third is associated with a syndrome, meaning other medical issues can accompany the hearing loss.**

## Here's What's Happening

**Thursdays,  
June 9 and 23**

**Bowl from 5 to 7 p.m.** during Monticello Community Education's activity night for students who are deaf or hard of hearing. The \$5 fee includes 2 games of bowling, shoes, pizza, and pop. The location is River City Lanes, 101 Chelsea Road in Monticello. Call 763-295-0320 to register.

**Saturday,  
June 18**

**CSD hosts Deaf Day at Valleyfair Amusement Park** from 10 a.m. to 4 p.m. Tickets are \$23 for adults and \$13 for children less than 48" tall, but older than age 3. Tickets can be purchased at Valleyfair's group sales ticket window on Deaf Day or in advance at CSD of Minnesota, 2055 Rice St., St. Paul between 8 a.m. and 5 p.m. weekdays.

**Saturdays,  
June 11,  
July 9,  
August 13**

**Cool off at Cascade Bay Water Park Social Day for the Deaf and Hard of Hearing Community** from 1 to 8:30 p.m. at Cascade Bay, 3830 Pilot Knob Road in Eagan. Tickets are \$8, \$6 for guests under 42" and are available at the water park. Directions to the park are at [www.cascadebay.com](http://www.cascadebay.com). The event is sponsored by CDS and St. Paul Parks and Recreation.

**Saturday,  
July 23**

**"Cue at the Zoo" at Como Park** in St. Paul. Meet at 10 a.m. at the giraffe statue near the African-Hoofed animals exhibit. A transliterator will cue the 11:30 a.m. "Sparky the Sea Lion" show. Bring a picnic lunch. If you have questions contact Katherine Burns-Christenson at [KBC29@aol.com](mailto:KBC29@aol.com) or 952-929-3965.

**Summer dates**

**The Cued Speech Association of Minnesota (CSAM) sponsors "Play and Cue" events** from 10 a.m. to noon for kids who use Cued Speech and their parents. June 17 meet at Rosland Park, 4300 W. 66th St., Edina. July 8 meet at Pearl Park, 414 E. Diamond Lake Rd., Minneapolis. July 22 at Jensen Lake Park in Eagan. Aug. 5 at Cliff Sen Park in Burnsville. Aug. 20 at Staring Lake Park, 14800 Pioneer Trail in Eden Prairie. If it's raining, events will be moved. Contact Katherine Burns-Christenson at [KBC29@aol.com](mailto:KBC29@aol.com) or 952-929-3965 for more information.

**Thursday,  
August 4**

**Make friends at the Family Support Connection's Family Picnic** from 5 to 8:30 p.m. at Como Park in St. Paul. Reservations are needed by Aug. 1. See the attached flyer for details.



## ASL-Interpreted Performances

### **Flint Hills International Children's Festival**

Saturday, June 4, 11:30 a.m. - 3:30 p.m.  
Sunday, June 5, 12:30 - 2:30 p.m.  
Ordway Center for the Performing Arts,  
345 Washington, St. Paul; 651-224-4222,  
651-282-3100(tty); [www.ordway.org](http://www.ordway.org).  
Includes mainstage plus free events in  
Rice Park with circus stunts, mimes,  
clowns, worldwide foods, and art.

### **And Then There Were None**

Friday, June 24, 7:30 p.m.  
Tickets: \$12; \$10 seniors/students  
Bloomington Center for the Arts, 1800  
W. Old Shakopee Road; 952-563-8587;  
[www.bloomingtoncivictheatre.org](http://www.bloomingtoncivictheatre.org).

### **Fiddler on the Roof**

Sunday, June 26, 2 p.m.  
Tickets: Reduced to \$3-5  
Cross Community Players, Maple Grove  
Senior High School, 9800 Fernbrook  
Lane N., 763-391-ARTS (2787);  
[www.crossplayers.org](http://www.crossplayers.org).

### **Peter Pan**

Friday, July 8, 8 p.m.  
Ordway Center for the Performing Arts,  
345 Washington, St. Paul; 651-224-4222,  
651-282-3100(tty); [www.ordway.org](http://www.ordway.org).

### **Little Shop of Horrors**

Sunday, July 24, 1 p.m.  
Request ASL: [accessible@orpheum.com](mailto:accessible@orpheum.com).  
Orpheum Theatre, 910 Henne-pin  
Ave.S., Mpls.; 612-373-5637, -5655(tty);  
[www.hennepintheatredistrict.com](http://www.hennepintheatredistrict.com).

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[herwell@wwt.net](mailto:herwell@wwt.net)

Please note that information about events, services, or other organizations does not imply endorsement by the Family Support Connection.

The Family Support Connection's mission is to build better lives for children who are deaf and hard of hearing by providing parent-to-parent support to families. Please visit our website at [www.familysupportconnection.org](http://www.familysupportconnection.org).

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## In Your Corner

By Candace Lindow-Davies,  
Family Support Coordinator

We hope you enjoy this month's *FOCUS*, discussing genetics and hearing loss. We are very pleased to have had Dr. Lisa Schimmenti write our cover article for us. On several occasions, I have been lucky enough to hear Dr. Schimmenti present and find her talks very fascinating.

We know so much more about genetics than even four years ago when I first came to work at Lifetrack. I can only imagine what we will know in the next four years.

When I mentioned we were planning to cover this topic to some parent advocates in other states, two of them reacted strongly. They both had more than one child who is deaf or hard of hearing, a clear sign that the hearing loss is genetic. They both wanted to know if our program regularly recommends genetic testing to parents. What I tried to explain and want our readers to know is that the Family Support Connection is all about choices. (I wrote about this last month in my article about bilateral implants.) We really want to give parents all the information we can so parents can make informed decisions.

So, if a parent reads this newsletter, does some research, and decides to pursue genetic testing, I will feel we have done our job. If another parent reads this newsletter, does research, and decides to decline testing, I will also feel like we have done our job. We just hope parents will be given enough resources to come to those decisions.

With the start of June, summer is truly right around the corner. When this newsletter is released, my children will be counting down the very last days of school, I will have finally figured out a summer schedule for speech, day camps and work. And maybe, just maybe, we will have some drier, warmer weather. I do have to say I love summer with the longer days, nights without jackets on kids, and more time with family. I hope your family is looking forward to some good times, as well.

We surely hope that many of you will join us for our second annual Family Picnic on Thursday, August 4th at Como Park. Please come and vote whether you would like to move the picnic to Minneapolis next year.

Have a lovely summer—hope to see you out and about at local parks and beaches! Our next issue of *FOCUS* comes out mid-July.

## Legislature passes bills of interest to deaf, hard of hearing

Two bills affecting people who have hearing loss passed through the Minnesota Legislature last month.

The Hearing Aid Consumer Protection Bill has been signed by the Governor. This extends the money-back-guarantee period by 15 days and allows the consumer to obtain the audiogram used to fit a hearing aid. It also offers protections when hearing aids are returned, setting a limit on fees for fitting to \$250 and requiring dispensers to return hearing aids that were traded in for a discount on new aids.

The captioning bill received its final passage May 18. The bill allocates \$300,000 a year to caption live news broadcasts one hour daily for stations serving Alexandria, Duluth, Rochester, Austin and Mankato, plus real-time captioning of public television's "Almanac."

Five other bills are still pending in the Legislature. For details contact Mary Hartnett, Minnesota Commission Serving Deaf and Hard of Hearing People, at 651-297-7305 or e-mail [mary.hartnett@state.mn.us](mailto:mary.hartnett@state.mn.us).

## Parent seeks support for changing rules about bus aides

A local parent who is concerned about getting an aide who signs to ride the school bus with her child wants to connect with other parents in a similar situation. She is interested in investigating whether there is potential to make changes to current laws. E-mail [needs2005@comcast.net](mailto:needs2005@comcast.net).

## Group needs signing volunteers to tutor deaf Jamaicans

Global Deaf Connection (GDC) is looking for volunteers to tutor deaf Jamaican young adults in English skills to help them pass the government's college entrance exam and enter teacher training college.

A trip to Jamaica for volunteers who sign runs from July 2 to 30. The trip costs \$1,990 excluding airfare. \$200 scholarships are available. Contact Jason Sem at [jsem@deafconnection.org](mailto:jsem@deafconnection.org) or see the GDC website at [deafconnection.org](http://deafconnection.org).

**Deaf and hard of hearing students can take drivers' education classes with ASL interpreters this summer through St. Paul Community Education. Classes run from 1 to 4 p.m. July 11-26 at Central High School, 275 N. Lexington Pkwy. Classes are open to adults and students who are least 15 years old. Cost is \$290 (\$10 discount for St. Paul residents/public school students). Applications are available at the Community Education offices in St. Paul public high schools. For more information call 651-632-6020 or see [www.commed.spps.org](http://www.commed.spps.org).**

**Genetics...continued from Page 1**

Most of the syndromes associated with hearing loss may be recognized by obvious health issues, however there are a handful that have hidden medical issues. It is better to catch these problems before symptoms occur. For example, in Jervell & Lange-Nielsen syndrome, hearing loss runs with life-threatening heart rhythm abnormalities that would need to be treated by a heart specialist. In Usher syndrome, children can develop vision loss and need an eye exam every year to watch for development of eye disease that sometimes can be treated with special vitamins. For other syndromes, we need to test for proper thyroid or kidney function or perform CT scans to check for inner ear malformations.

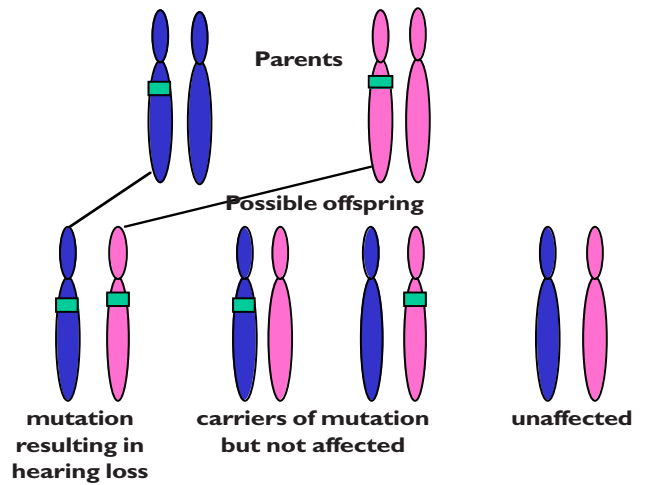
For many children, I cannot tell immediately if they will fall into the non-syndromic or syndromic group and I will recommend further testing.

In Emma’s case, the cause of her hearing loss was not clear. As a first step, I suggested conducting genetic testing for mutations in the genes that code for Connexin 26 and Connexin 30, the most common genetic cause of hearing loss. It recently was discovered that about half of children with non-syndromic hearing loss have a mutation (change) in the genes that encode Connexin 26 and Connexin 30. Mutations in Connexin 26 and Connexin 30 are the most common cause of hearing loss worldwide. Connexins are proteins important for recycling of potassium in the cochlea. A person has two copies of each gene. If a person has a mutation in both copies of either gene or in both genes, then that person will have hearing loss.

We can perform genetic testing from DNA that is taken from a blood sample to learn if a child’s hearing loss is caused by mutations in Connexin 26 or Connexin 30. Extensive research has shown that children with mutations in Connexin 26 and 30 have non-syndromic sensorineural hearing loss. The hearing loss will not get much worse over the years (it’s not a progressive loss). And, there are no other hidden medical issues to worry about.

Emma’s mother reminded me, “No one in our family

**Inheriting hearing loss from genetic mutation**



has hearing loss. Could Emma’s hearing loss still be caused by Connexin 26 and Connexin 30?”

“Yes,” I told her. This is because hearing loss caused by mutations in Connexin 26 or Connexin 30 is inherited in a recessive manner. Recessive means that both parents are carriers of the mutation, but they are not affected by it.

Each cell in our bodies contains the entirety of our DNA, which is packaged into chromosomes. We have two of each chromosome, one from our mother and one from our father for a total of 46 chromosomes. The genes for Connexin 26 and Connexin 30 are on chromosome pair 13. Having one copy of an altered gene does not cause hearing loss. A parent can be a carrier and not even know until they have a child with hearing loss.

In the figure above, the father’s chromosomes are blue and the mother’s are pink. A child receives one chromosome from each parent. If one of the chromosomes from each parent contains a gene that has a mutation (represented by the green bar), there would be a 1 in 4 chance with each pregnancy of having a child with hearing loss.

If genetic testing for hearing loss shows that Emma has two copies of Connexin 26 or Connexin 30 with a mutation, then we know why she has hearing loss. This would also mean that her hearing loss will not be progressive and that she is not at risk for other health issues. If the testing for Connexin 26 and 30 does not find two mutations, then it does not mean that the hearing loss is not genetic, but only that the most common cause has been ruled out.

Emma’s parents chose to have the genetic testing for Connexin 26 and 30 performed. They returned to clinic two months later to learn the result.

In Emma’s case, both copies of the gene for Connexin 26 have a mutation. Thanks to genetic testing, Emma’s parents now understand why she has a hearing loss and do not need to worry that she could have other medical issues. No further tests are necessary. Her hearing loss should remain stable.

**More information on genetics & hearing loss:**

Boys Town National Research Hospital,  
[www.boystownhospital.org/parents/info](http://www.boystownhospital.org/parents/info) (scroll down and click on “Genetics and Deafness”)

Deafness and Hereditary Hearing Loss Overview,  
[www.geneclinics.org/profiles/deafness-overview/details.html](http://www.geneclinics.org/profiles/deafness-overview/details.html)

Genetic services are available at the University of Minnesota, Mayo Clinic and Minneapolis and St. Paul Children’s Hospitals.

## Study seeks information from cochlear implant users

A graduate student at Tennessee Tech University who also has a child with a cochlear implant is conducting research on the success of cochlear implants.

Deanna Meador is studying which factors most significantly predict cochlear implant success as measured by speech and hearing improvement post-implantation. Participants just need to fill out a 5-page survey and send in audiograms; there's no testing involved. She's hoping to develop a formula that will predict success for individuals who are considering the procedure.

To be eligible for this study, participants must have been deafened prior to age three and have had a cochlear implant at least one year. Contact Deanna at 615-633-1917 or e-mail [dnmeador21@tntech.edu](mailto:dnmeador21@tntech.edu).

## Summer worship services accessible to deaf signers/cuers

ASL-interpreted services will be offered this summer at 9:30 on Sundays at Macalester Plymouth United Church, 1658 Lincoln Ave., in St. Paul. The church is a blended Presbyterian and United Church of Christ faith community. For more information see [www.macalester-plymouth.org](http://www.macalester-plymouth.org).

Cued Speech-transliterated services will be offered once a month this summer at Diamond Lake Lutheran Church, 5760 Portland Ave. S., in Minneapolis. The 9:30 worship service on June 12, July 10, and Aug. 14 will be transliterated.

Diamond Lake Lutheran Church also will have a transliterator for Vacation Bible School June 27-July 1 from 9:30 a.m. to 12:15 p.m. Cost is \$10 per child (\$20 per family). For more information call 612-866-2579 or see [www.dllc.org](http://www.dllc.org).

**An ASL interpreter will be on hand for the free Summer Reading Program at 2 p.m. on the following Tuesdays at Merriam Park Library, 1831 Marshall Ave. in St. Paul. For details call 651-642-0385 (v) or 651-298-4184 (tty).**  
**July 12—Dakota Wild Animals**  
**July 19—Steve Abrams**  
**Puppeteer**  
**July 26—Professor Bear**  
**August 2—Airplane Annie**  
**August 9—Magician Matt Dunn**



## A Little Grin

By Linda Werness-Jaroszewski  
 Contributed by Denise Pederson

My poor son, Noah! He is still trying to figure out who has hearing loss and who doesn't. You see, hearing loss is genetic in my husband's family. I have typical hearing and my husband Mark has a hearing loss and uses hearing aids. Our 2-year-old son, Noah, also has a hearing loss and wears aids. Noah goes to school with many children who use a cochlear implant, which he calls "buttons."

One day, he saw his father's hearing aid case and handed it to his dad, saying "hearing aid." Then he climbed on Mark's lap to see if the aids were in. Only one was. He was not happy about it. Then, he climbed on my lap and ran his fingers through my hair and said "NO button" really mad. "Where's your button?"

He probably thinks everyone wears hearing aids or a button!

Share your 'Grins' by emailing [fsc@lifetrackresources.org](mailto:fsc@lifetrackresources.org).

**A new technology called "Signlinks" makes it possible for websites to be presented in American Sign Language (ASL).**

**Application developers in Canada created the software to help native signers understand web content that typically is written in English. Using Quicktime 6.5, Signlinks shows a person signing the information on a website. Signlinking currently is used on two websites: [aslpah.com](http://aslpah.com) and the website of the Canadian Hearing Society ([www.chs.ca](http://www.chs.ca)).**

## Survey looks at kids evaluated for cochlear implants

Cochlear America, the manufacturer of Nucleus brand implants, is surveying parents of children who were evaluated for a cochlear implant between Jan. 1, 2002 and March 15, 2005. Participants who complete the survey by June 13 will receive a \$5 Starbucks card. For a survey, contact Donna Sorkin at [dsorkin@cochlear.com](mailto:dsorkin@cochlear.com).

## U of M studies new hearing test

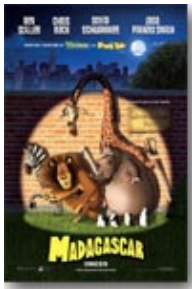
The University of Minnesota is looking for children ages 5 to 12 who have mild to severe hearing loss to participate in a study for developing a better hearing test.

Funded by the National Institutes of Health, the study involves testing a child's hearing twice: once with the traditional method where the child pushes a button or raises his hand when he hears a beep and once with a new computer-based test using a touch screen similar to a computer game.

For more information contact Allison L. Kohtz, M.A., CCC-A, at 612-624-7699 or [kohtz001@umn.edu](mailto:kohtz001@umn.edu).

**Local theaters show captioned movies**

Oakdale Cinemas (Marcus Theatres) offers an open-captioned movie every weekend in June. The theater is at 5677 Hadley Ave. N. in Oakdale. Movie showtimes are recorded at 651-770-4994 or on-line at [www.marcustheatres.com](http://www.marcustheatres.com). “Kicking and Screaming,” rated PG, shows June 3-5. “Madagascar,” rated PG, shows June 10-12. “Sisterhood of the Traveling



Pants,” rated PG-13, shows June 17-19. And, “Cinderella Man,” rated PG-13, shows June 24-26.

Kerasotes Theaters will pick up “Kicking and Screaming” June 6-7 in Coon Rapids and June 8-9 in Inver Grove Heights. The theaters also will show “Cinderella Man” June 13-14 in Coon Rapids and June 15-16 in Inver Grove Heights.

To find out when open-captioned movies are showing at local theaters, see [www.mnocfilms.org](http://www.mnocfilms.org). The site also lists movies showing at theaters that have rear-window captioning systems.

**A couple of engineering students recently won an award for designing a device that uses a laptop computer to alter incoming speech so that it all falls within the frequency range heard by an individual who has a severe to profound sensorineural hearing loss. The device also uses voice-recognition software to produce text of speech. The Pennsylvania State University students hope to perfect the device to help deaf people understand lectures and meetings.**

**Plans move forward for signing town in South Dakota**

A tiny town in South Dakota that doesn’t even exist yet has caught worldwide attention.

Laurent, a planned community near Mitchell where ASL would be the common language, is still in the planning phase. Since town planners first put out notice about the community, 108 reservation forms have arrived with a total of 166 adults and 109 children making a commitment to live in Laurent. Groundbreaking is expected this Fall.

Last March, town planners and interested residents held a weeklong design meeting at Camp Lakodia near Madison, S.D. Reporters from the *Minneapolis Star Tribune*, *People* magazine and the *New York Times* interviewed participants, generating news stories in about 170 newspapers worldwide. The news stories along with drawings of the town plans are on the Laurent Company’s website: [www.LaurentSD.com](http://www.LaurentSD.com).

Talk of creating a signing community first began in December 2002 when Marvin Miller and his mother-in-law, M.E. Barwacz, decided to make their dream become reality. They named the future town Laurent after Laurent Clerc, considered the father of American Sign Language. Clerc, who was deaf, brought sign language to America from France in 1815 and helped establish the first permanent school for the deaf in America.

At this point, the town has optioned approximately 275 acres of land. Developers expect about 2,500 people to live there. The town will have an integrated ASL public school system with K-12 being served in one facility. Updates about the town are sent out periodically through an e-newsletter, *The Laurent Town Crier*. To subscribe, visit [www.LaurentSD.com](http://www.LaurentSD.com).

**Curtis Pride, the only deaf player in Major League baseball, moved up to the Los Angeles Angels last month to play left field as #19.**

**Position for Cued Speech open**

The National Cued Speech Association is looking for a Director of Programs in a one-year paid position.

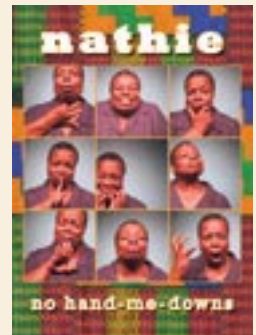
The Director will be responsible for new programs including a pilot program offering Cued Speech workshops and camps nationwide, creating a Virtual Library, and launching a national professional-level conference.

The position starts July 1, 2005 and ends July 31, 2006. For details contact [MinnCuer@aol.com](mailto:MinnCuer@aol.com).

**Documentary features famous Deaf storyteller**

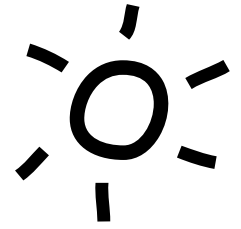
A new documentary called “Nathie: No Hand-Me Downs” profiles the American Sign Language storyteller and poet, Nathie Marbury.

In this 125-minute documentary, Nathie signs about her upbringing in Pittsburgh, where she was the 16th child in a family of 17 children. She also shares her ASL poems. The DVD is voice-interpreted and subtitled. It is available for \$29 online at webstores including [www.thetactilemind.com](http://www.thetactilemind.com) and [www.harriscomm.com](http://www.harriscomm.com).



Make friends at the **Family Support Connection's**

# Family Picnic!

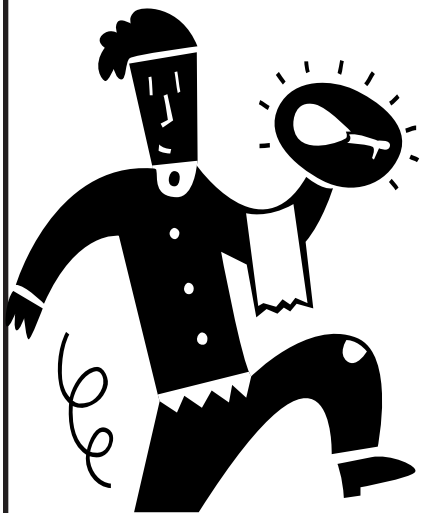


For families with children who are deaf or hard of hearing.

**Thursday, August 4<sup>th</sup>**  
**5-8:30 P.M** (Rain or Shine!)  
**Como Park Midway Pavilion South**  
(Midway Parkway and Horton Avenue in St. Paul)

## Free for the family:

- Dinner at 5:30
- Face Painting & Games
- Temporary Tattoos
- Door Prizes!



## Also at Como Park:

- Zoo and Conservatory (open 10-6)
- Cafesjian Carousel (open 11-4)
- **NEW!** Amusement Rides (open 10-8)
- **Mini Golf** (open 12-6)

For more information about Como Park, Zoo or Conservatory call 651/487-8200 or visit [www.comozooconservatory.org](http://www.comozooconservatory.org)

## Please RSVP!

Call before Monday, August 1 to let us know how many from your family will attend



**Lifetrack**  
**Resources**

709 University Avenue West • St. Paul, MN 55104-4804  
651/265-2435 (voice) • 651/265-2379 (TTY)  
1-866-DHOHKID (toll-free) • 1-866-857-2379 (toll-free TTY)  
[www.familysupportconnection.org](http://www.familysupportconnection.org)

**Family Support Connection's**

# Family Picnic!

For families with children who are deaf or hard of hearing.

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## **Como Park Midway Pavilion South (Midway Parkway and Horton Avenue in St. Paul)**

