

Trisomy 21 Update

A Newsletter from the Trisomy 21 Program at The Children's Hospital of Philadelphia

Fall 2009



Interview with:

Erin Cipriano, Au.D., CCC-A, Division of Audiology

Why is it important to test hearing?

Almost all children with Down syndrome have problems with their ears at some point, including ear infections, middle ear fluid, narrow ear canals, wax build up, and hearing loss. All hearing loss is significant and even mild hearing loss can affect speech development. The earlier a hearing loss is found and treated, the less likely it is to affect speech.

Are there different types of hearing loss?

There are three different types of hearing loss, conductive, sensorineural, and mixed.

Conductive hearing loss may be caused by a narrow ear canal or wax. It may also be caused by fluid or infection behind the ear drum. Conductive hearing loss can be prevented by keeping the ear canals free of wax and keeping middle ear fluid from building up in the middle ear space. Monitoring and treatment by an Ear, Nose and Throat physician is important to remove wax from ear canals and diagnose and treat ear infections. Conductive hearing loss is the most common type of hearing loss in children with Down syndrome. Sensorineural hearing loss may be caused by damage to the cochlea, or nerve of hearing. Sensorineural hearing loss is permanent hearing loss. Some children with Down syndrome are born with sensorineural hearing loss. Sometimes serious infections, such as meningitis or certain medications can cause damage to the cochlea or nerve. Some children with Down syn-

drome have both a conductive and sensorineural hearing loss called mixed hearing loss.

How often should children with Down syndrome have their hearing tested?

Up to 80 percent of children with Down syndrome have hearing loss. The American Academy of Pediatrics (AAP) recommends testing hearing at birth and every 6 months up to age 3 years. After age 3 years, a hearing test should be completed at least annually. Hearing loss can appear at any time so even if a child has normal hearing on several occasions, he or she still needs regular hearing tests.

How do you screen hearing in a baby?

Newborn hearing screenings use either otoacoustic emissions (OAE) testing, or automated Auditory Brainstem Response (ABR) testing. These measures do not require the baby to respond to sounds, but rather measure the electrical responses in the brain.

What does a failed newborn hearing screening mean?

A failed hearing screen means the baby may have hearing loss. He/she needs another test to confirm this because sometimes fluid in the middle ear, noise in the testing room, or excessive movement from the baby can also cause an abnormal test. Thus, babies who

We've Moved

The Trisomy 21 Program is now located at 3550 Market St., 3rd Floor. There is free parking in the Sheraton Hotel located on 36th St. between Market St. and Chestnut St.

Resources

We are trying to expand our Trisomy 21 resource list and we would love your input! Please email us the names and contact information of any doctors, specialists, resources, support groups, camps, etc. that you have found helpful; email resources to trisomy21@email.chop.edu.

DUFFERFEST

The rain of June couldn't stop music lovers of all ages from attending DufferFest on June 20, 2009. Hosted by Duffer's Tavern and Team Kara Marie Kenney, this music festival featured local bands, a disc jockey, children's crafts, games for the big kids, food and drinks, dancing and good times. The event raised over \$15,000 for the Trisomy 21 (Down syndrome) Program at CHOP. The three year cumulative giving for the Trisomy 21 Program is over \$70,000. Thanks Team Kara Marie and Duffer's Tavern!!

Trisomy 21 Team

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Ask the Doctor

We welcome all questions! Please send to trisomy21@email.chop.edu.

Questions will be presented anonymously to our T21 team, and answers may appear in upcoming newsletters. Please submit your questions about caring for children with Down syndrome. Subjects can include child development, neurology or other issues.

SPOTLIGHT

It's never too soon to plan for the future! In anticipation of the inevitable aging of our current pediatric population we have recently taken steps to formalize our adult Trisomy 21 program. Patients over the age of 18 are evaluated through the division of neurology and assessed from a neurological and social/vocational perspective. A full medical history and physical examination are performed to ensure that all recommended testing for individuals with Trisomy 21 have been addressed. Social issues are managed as they arise and may include guidance on transitions to adulthood, employment, creative living options, sexuality and issues surrounding aging adults with Trisomy 21. Individuals with Trisomy 21 are living longer, healthier, and more fulfilling lives; our goal is to follow individuals over time so that we can learn more about this uniquely aging population. Help us spread the word!

Buddy Walk 2009

The Children's Hospital of Philadelphia Buddy Walk® and Family Fun Day is being held on Sunday, Oct. 4, 2009, at 10 a.m. at Villanova University Stadium on the campus of Villanova University. The walk raises funds for patient care, research and education for the Trisomy 21 Program at Children's Hospital. Trisomy 21, or Down syndrome, affects more than 350,000 people in the United States. This event is great fun for the whole family and includes a walk, food, music and activities for all. This year, there will also be a special appearance by singer and Down syndrome activist Chris Burke with Joe and John DeMasi. For more information, visit www.tri21center.org, or contact Rebecca J. Elias at 267-426-5600 or by e-mail at events@email.chop.edu.

7th Annual T21 Symposium

The seventh annual Trisomy 21 symposium will be held on Saturday, March 20th, 2010 at The Children's Hospital of Philadelphia. We would love to hear any suggestions you may have; together let's make this year's event the best we've had yet! Please email your ideas to trisomy21@email.chop.edu. Suggestions may include potential topics, speakers, or exhibitors you'd like to see.

fail their newborn hearing screening need another hearing screen to confirm abnormal results, and they may need more specific testing if the second test is also abnormal.

What happens at an audiologic evaluation?

Even when a child passes his or her newborn hearing screening, a pediatric audiologic evaluation should occur when the child is 6 months of age. During this evaluation the audiologist examines the ear, measures how well the eardrum is moving (tympanometry), and completes a hearing test. The hearing test provides information on how a child behaviorally responds to different levels of pitch and loudness. Children might be taught to look at puppets, play a game, or simply raise their hand when they hear a sound.

Sometimes it's difficult to get the child to cooperate with the hearing test. What can parents do to make the test successful?

Behavioral testing does require a child's cooperation to obtain accurate test results. If a child is tired, hungry, or not feeling well, it may be difficult to obtain reliable results regarding a child's hearing sensitivity. The hearing test should be scheduled during a time of day that is best for the child. Additionally, prepare the child ahead of time by explaining what will happen. Children can also practice wearing headphones at home. Avoid pairing the hearing test with other stressful office visits. If a child is sick that day, you may want to reschedule.

What happens if the hearing test is abnormal or the child just can't do it?

Regardless of a child's age or developmental level, sometimes behavioral hearing tests just aren't successful, or the results are inconclusive. When behavioral results are inconclusive or suggest hearing loss, diagnostic Auditory Brainstem Response (ABR) testing should be considered. Diagnostic ABR testing does not require a child's cooperation and results from ABR testing provide information regarding the type, degree, and configuration of hearing loss. ABR testing can also confirm normal hearing. To complete ABR testing, the child must be sleeping and in older children (usually above 6 months of age), this may require sedation.

What are the best ways to make sure a child will sleep during ABR testing?

Children with Down syndrome require a higher level of medical support during sedation so when possible, a natural sleep ABR should be attempted. Children younger than 6 months are often able to sleep naturally through the duration of testing. Parents should do the best they can to make sure their baby is tired on the day of the evaluation. Waking the child early and either shortening or skipping the child's nap may help. In addition, it is important that the baby remain awake during the ride to the office. We also suggest that parents come prepared to feed their child prior to testing as many infants sleep best when they have a full tummy. If a child is not a good sleeper or attempts to complete a natural sleep ABR test are unsuccessful, sedation should be considered. Here at CHOP, children with Down syndrome are sedated for testing at the main hospital only.

What is the best way to make sure a child receives the most beneficial audiologic follow-up?

It helps to find a managing audiologist for all audiologic follow-up. By establishing a relationship with one audiologist, parents and children develop a comfort level that may allow for more successful testing. In addition, if testing does not go as expected, parents can work with the audiologist to develop a plan for the next office visit. Whether it is scheduling the appointment at another time of day, practicing wearing headphones at home, or working on a listening game, everyone will know what to expect at the next evaluation.

MY GREAT STORY CAMPAIGN

Join the National Down Syndrome Society's (NDSS) My Great Story campaign to honor and celebrate the amazing stories of the 400,000 Americans with Down syndrome. If you have Down syndrome, tell us about your achievements, dreams, aspirations, and successes. If you know someone who has Down syndrome tell us how they've inspired you. Visit www.ndss.org/stories, click the 'submit' bookmark, and enter your great story.

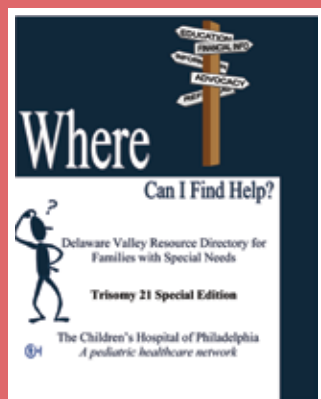
CHALLENGER DAY

Challenger Day is for youth interested in learning about or trying out sports. Activities will include baseball, basketball, cheerleading, football, lacrosse, swimming, face painting, arts and crafts and lots more. This year's Challenger Day is being held on Saturday, October 3, 2009 from 2 to 4PM at the Upper Merion Middle School in King of Prussia, PA. This event is free and open to families wherever they live. For more information, to register or to volunteer, please e-mail Emily Miller at emiller@umtowsnhip.org or call the Park & Recreation department at (610) 265-1071

Resource directory

The resource directory, updated each year, is on the Trisomy 21 Program Web site.

Go to www.chop.edu and type “Trisomy 21” in the search box and choose the first option. Then choose “Resources



for Families” from the list on the left side of the page. The resource directory is under “Downloadable Material.”

Other Resources Available Through the T21 Program

Care Binder

This is an organizational tool to keep all medical and educational materials in one place, making it easy to retrieve the right piece of information for the doctor, the school, the babysitter and the rest of the family. Call or e-mail to sign up for a one-on-one or group session, at which you will learn how to use the notebook, including which records to keep in the notebook and how to maintain records on a continuing basis. Contact Symme Trachtenberg at 215-590-7444 or by emailing Trachtenberg@email.chop.edu

Symposium

Mark your calendars! The 6th Annual T21 Symposium will be held on Saturday, March 20th, 2010. Look for details on our program website.

 The Children's Hospital of Philadelphia®

Hope lives here.

34th Street and Civic Center Boulevard

Philadelphia, PA 19104-4399

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