Large Vestibular Aqueduct Syndrome
Is My Child At Risk?

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Large Vestibular Aqueduct---

What is that?

The Vestibular Aqueduct is a narrow, bony canal that carries the endolymphatic duct from the inner ear (cochlea) to a fluid-filled sac between the inside of the skull and the membranes that cover the brain.
In some people, this canal and duct are too large and they allow fluid to back up into the cochlea after a change in head position, or a bump on the head. This can cause a change in the sensorineural hearing that can be temporary or permanent. It can also cause a conductive hearing loss too.
Approximately 40% of the time, there can also be a deformation of the inner ear and/or a fistula or leak in the inner ear.

This can be diagnosed through a CT scan, or measured on an MRI.
At any given time since I started looking for it in 2000, about 30% of the kids that I consult with who have any degree or configuration of hearing loss have LVAS.
SIGNS YOUR CHILD MIGHT HAVE LVAS/EVAS

- Hearing loss of any degree and any configuration, and any loss that has been progressive

- Conductive hearing loss, with no fluid present when physician checks and there is no ear infection

- Hearing seems to fluctuate

- Understanding seems to fluctuate
SIGNING YOUR CHILD MIGHT HAVE LVAS/EVAS cont’d

- CHANGE IN HEARING AFTER THEY BUMP THEIR HEAD, MAY BE A MINOR OR A MAJOR BUMP

- Significant balance problems, especially if they come with vomiting and/or eye movements (nystagmus)

- Family history of Pendred Syndrome

- Child may report that they hear “sirens” when none are present (tinnitus)
SIGNS YOUR CHILD MIGHT HAVE LVAS/EVAS cont’d

- Hearing may decrease after they have a fever

- Hearing may decrease after they play on a trampoline/bouncy house etc.

- Change in hearing after you fly, especially in a small non-pressurized plane

- Change in hearing after they dive into deep water
Consider this...

- This can also be unilateral!

- Remember, this is about 30% of my caseload, not 7% possibly because I look for it.

- THE HEARING LOSS PROGRESSION MAY BE PREVENTED IF THE FAMILY KNOW AND IS TAKING PRECAUTIONS.
Also Consider....

- In some areas, all babies who are diagnosed with a hearing loss get a scan! They may need to be at least one to one and a half years old before the duct can be measured accurately.
- There has been some research about changing measurements, but this has been viewed with skepticism at the present time. One thing experts agree on is that the size of the Vestibular Aqueduct does not predict how or when the child’s hearing levels change.
- The thresholds can go back to previous levels up to 9 – 12 months later.
Case Study: WT  Age 3: September 1999 and November 1999

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Audiological Evaluation

Audiogram

1999 Calibration
FREQUENCY IN HERTZ (Hz)
250 500 1000 2000 4000 8000

Intensity in Decibels (dB)

Audiologist: C.F.T.

Masking Noise Used: White Noise

Aid Notes:
Right
Left

Tested at home with a microphone.

No speech available.

Kathleen G. Johnson
Audiology
WT age 6

AUDILOGICAL EVALUATION

Name: 
Address: 
Parent: 
Refer: 

WT

AUDILOGRAM

ASHR 1969 Calibration
Frequency in Hertz (Hz)

200 500 1000 2000 4000 8000

40 dB HL

COMFORT LEVELS

Mask

SPEECH RECEPTION THRESHOLD

dB HL

R

L

Bone

SF

Aided

Aided

Aided

Aided

Comfort levels: UNCL

SPEECH DISCRIMINATION

% dH.L List Masking

R

L

Bone

Aided

Aided

Aided

Aided

Masking Noise Used:

KEY

MLV + Recorded:

NOTES:

No hearing aids worn.

Significant drop in right thresholds.

Complaint of difficulty in hearing following infection.

New hearing in right ear amplified to 0 dB on this side.

Through tests, hearing was good.

Reported hearing in right.

Because he couldn't hear.
WT age 8: September 9, 2004 and September 27, 2004

WT age 9 May 2005
# Case Study: PH

- **Name:** PH
- **Age:** 6
- **Birthday:** 4/20/92
- **Date of Birth:** 10/27/03
- **School:**
- **Phone:**
- **Doctor:**
- **Audimeter:** G57-1C

## Test Results

### Type of Test

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<th>Type</th>
<th>Score</th>
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<tr>
<td>SSA</td>
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### Test Reliability

- **Air:** Good
- **Bone:** Fair

### Pure Tone Average

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### Speech Reception Threshold

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### Comfort Levels

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<th>UCL</th>
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### Speech Discrimination

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Note: MLV Recorded

## Impression

- **Suspect possible progressive loss present screening last year bilaterally.**
- **Right ear mild to moderate sensorineural hearing loss, left ear normal.**
- **Need ENT referral.**
- **History:**
  - Possibility of implantation
  - Familial history of similar
Case Study HE
SUGGESTIONS FOR FAMILIES WITH CHILDREN WITH LARGE VESTIBULAR AQUEDUCT SYNDROME
Prevention of further hearing loss means taking a lot of precautions, safety measures, restriction of activities, a large measure of luck, and educating everyone in the child’s life. This can be very stressful to both the child and the parents.
Be strict about helmets for ALL of your children and yourselves. This is a idea good for everyone; remember that the possibility exists that other members of the family might have LVAS too.
Remove all benches, bar stools, backless stools, and anything that might tip over easily in your home and yard. The child might have balance problems in addition to the hearing loss. Add handrails around porches. Eliminate things they might want to climb up and stand on.
• Keep bicycles, tricycles, swing sets, etc. in good working order. Teach your child how to use them safely. Use only under adult supervision until you are sure they know what to do.
Carpet all hard wood floors, and remove throw rugs that the child might trip over. Keep floors clear and picked up.
Be sure that all family members, babysitters, child care givers, etc. are aware of the problem, and that they need to report even minor head bumps immediately to you.
If your Ear, Nose, and Throat specialist has suggested the use of steroids after a head bump, ask to have standing orders for steroids already on file with your pediatrician and at your pharmacy. Consider keeping a supply if you go camping or on vacation as it is important to get the child on steroids as quickly as possible after they hit their head.
Make sure that the school is aware of the situation, and that all of the playground aides, gym teacher, classroom teachers, etc. know what restrictions are placed on activities.
Remind the school that the child might have “good hearing days and bad hearing days” and try to come up with ideas to help. The child can have no hearing on some days and then be better the next. This is frustrating for everyone. The child can also have days where their discrimination (understanding) is affected. In other words, they may be able to hear the speech but it isn’t clear enough for them to understand what is being said.
• Be sure to let the school know if the child is on steroids---this can make them pretty busy!

• It is a good idea to have a WRITTEN sheet of instructions available to be given to substitutes, especially in gym class as the child is often too shy to tell a sub or other adult that they shouldn’t be doing something.
Look for alternatives to sports that will still give the child something to do---swimming is OK, as long as they aren’t diving to the bottom of the pool and they are trained to be sure to have their arms in front of them when turning (if swimming laps). Backstroke isn’t a good idea because of the chance they might hit their head. Dance classes can help with balance and coordination, and may help prevent falls and injuries. Be sure the dance teacher knows about LVAS.
Let the child have a say in what they want to give up---for some kids, sports are more important than hearing and they need to have input in these decisions. One child told me his life wasn’t worth it if he couldn’t play football because this was one of the few areas he really excelled in after school. I believed him. His parents let him play because he was going into depression. His hearing stayed stable, luckily.
Some of these kids seem to crave sensory input, and this, unfortunately, makes them WANT to spin upside down on the monkey bars. They seem to like to swing very hard, and very high on the swingset a lot, which the ENT said is OK as long as they don’t let go or jump off!
• If possible, teach phonics at as young an age as possible while they still have hearing. Back it up with visual phonics. Encourage reading, even at a preschool age while they have more hearing.

• Real Time Captioning is offering some promising results in the classroom for good readers. Otherwise, they may need sign language to supplement what they are getting auditorily and visually in the classroom.
• Encourage the family to have “silent evenings” with just sign or lip reading for EVERYONE in the family, so the child can get used to no hearing days, and the family can get used to no hearing days.

Even if the family has chosen to be oral, several “survival signs” such as “no, stop, come here NOW, don’t move (this one saved my son from falling out of a window), and I love you” can be very beneficial even at bath time or at the swimming pool when their hearing aids are out.
Be sure that there is a family plan for fire or other “disasters”, and purchase a vibrating smoke alarm for the child. If they ask, explain what you would do if the electricity went out, or someone was trying to break into the house (a 6 year-old’s question after a scary movie)! Teach them how to use the telephone to call for help. This can alleviate fears---for some reason, my patients with progressive losses are more likely to want to sleep with their hearing aids on because they worry about hearing the smoke alarm---and then end up with otitis externa, broken hearing aids, etc.
• Let them have a flashlight in bed with them at night; use night lights to help prevent falls in the dark.

• Be sure to carry night lights with you when you are traveling

• LET THE PEDIATRICIAN KNOW THAT THIS PREDISPOSES THE CHILD TO MENINGITIS. ANY EAR INFECTION SHOULD BE AGGRESSIVELY TREATED AND MONITORED.
Know the symptoms of fistula—vomiting, nystagmus, balance and coordination problems, etc. Seek immediate medical attention if this occurs. Children with Mondini dysplasia in addition to the LVAS are more likely to have fistulas (a leak in the cochlea).

Get out illustrations or an ear model and explain what isn’t working to your child. Teach them to explain it to other people who wonder why they can’t jump on a trampoline.
• Check out activities before your child arrives at the birthday party or a friend’s house. It is really hard to arrive at the house and see all of the other children jumping in the “play pit” and your child can’t do it.

• Practice what they will do and what they should say if an adult (friend’s parent, substitute gym teacher, playground supervisor, etc.) orders them to do something they shouldn’t be doing for safety reasons. Role play how to be polite but firm.
• Let the child ask questions and be prepared for grief reactions when hearing fluctuates.

• DO NOT BECOME ANGRY, RESENTFUL, GUILTY, OR BLAME ANYONE IN FRONT OF THE CHILD WHEN THEY HIT THEIR HEAD. SOMETIMES CHILDREN WANT TO PROTECT THEIR OTHER PARENT OR FRIEND IF THEY ARE AFRAID OF YOUR REACTION.
• **DO NOT BLAME THE CHILD WHEN THEY HIT THEIR HEAD.** STAY CALM, AND EXPLAIN WHAT WILL HAPPEN NEXT.

• **REMEMBER, YOUR REACTION COULD RESULT IN THE CHILD NOT TELLING YOU THE NEXT TIME SOMETHING HAPPENS AND RESULT IN THEM NOT GETTING ON STEROIDS SOON ENOUGH TO HELP.**

• **EVEN VERY YOUNG CHILDREN KNOW THAT YOU FEEL SAD WHEN THEY LOSE MORE HEARING, AND IT AFFECTS HOW THEY FEEL ABOUT THEMSELVES.** LET THEM KNOW THAT THEY ARE OK AND YOU ARE OK NO MATTER WHAT HAPPENS.
• Get professional help for you and your child if you need it to deal with feelings about hearing loss and deafness. Even very young children can have difficulties when they are going through this.

• Be aware that clingy times, night time fears, complaints of feeling pressure in the ears, screaming or using a louder voice, etc. may signal that they may be losing hearing again.

• Sometimes teen “rebellion” takes the form of risky behaviors---no helmet, touch football, etc. Keep your sense of humor, and encourage dialogue with your child.
Remember that the siblings of the child, each parent, grandparents and family dynamics also suffer when the child loses more hearing. This can be an emotional rollercoaster for everyone involved. It is worth mentioning that everyone handles this kind of stress in different ways. These may include extreme sadness, withdrawal, clinical depression, anger, alcohol or drug abuse, becoming a “superparent”, etc. Divorce rates for parents with a special needs child are close to 90%. Get professional help for yourself and your marriage if you need it.
Sometimes the child’s hearing can change even without them hitting their head. Try to keep track of what else might have precipitated the shift including changes in barometric pressure, activity, high fevers, etc. Keep your physician and audiologist informed so they can help manage medical, amplification and classroom issues.
Remember to use precautions when being exposed to loud noise. Even though your child may already have a hearing loss, noise exposure can increase this loss as well. Be sure that everyone in the family wears ear plugs or ear muffs when mowing the lawn, using power tools, attending rock concerts, etc.
• It is a good idea to have a genetics evaluation, and also have your child’s thyroid function tested. Many (but not all) people with LVAS have Pendred’s Syndrome and may also have thyroid issues.

• Remember you are not alone! An excellent online support group is: LVAS@yahoogroups.com

• You can also access this group and get more information about LVAS at: www.hearinglosshelp.com/articles/lvas.htm

• Please feel free to contact me if you have questions.