

>> GOOD EVENING.

WE ARE HERE TONIGHT FOR THE  
NEW STEPS FOR FAMILIES WITH  
CHILDREN NEWLY IDENTIFIED  
WITH HEARING LOSS:

INTERACTIVE DISCUSSION WITH  
MEDICAL PROFESSIONALS.

>> ALL RIGHT.

GOOD EVENING.

WELCOME TO NEXT STEPS FOR  
FAMILIES WITH CHILDREN NEWLY  
IDENTIFIED WITH HEARING LOSS.

MY NAME IS CANDACE LINDOW  
DAVIES, AND I'M THE

COORDINATOR OF THE PARENT-TO-PARENT ORGANIZATION MINNESOTA HANDS & VOICES.

ON BEHALF OF THE LIONS CHILDREN'S CENTER AND AMPLATZ CHILDREN'S HOSPITAL AND MINNESOTA HANDS & VOICES AND LIFETRACK RESOURCES, WE'RE PLEASED YOU COULD JOIN US. PROVIDING YOU, THE NEWEST MEMBERS OF OUR COMMUNITY, WITH INFORMATION, RESOURCES AND CONNECTIONS TO EACH OTHER IS OUR TOP PRIORITY.

THIS WORKSHOP WEBINAR IS WHAT  
MINNESOTA HANDS & VOICES  
HOPES WILL BE THE FIRST IN A  
THREE-PART SERIES WITH  
FAMILIES WITH CHILDREN  
RECENTLY IDENTIFIED AS DEAF  
OR HARD OF HEARING.

TONIGHT'S WORKSHOP IS AN  
OPPORTUNITY FOR NEW FAMILIES  
TO LEARN ABOUT WHAT ARE  
TYPICALLY NEXT STEPS TO TAKE  
WITH MEDICAL PROFESSIONALS.

WE'RE HONORED TO HAVE  
PRESENTERS REPRESENTING

AUDI OLOGY, OTOLARYNGOLOGY,  
ENT, GENETICS AND INFECTIOUS  
DI SEASE.

THEY' LL BE DRAWING ON THEIR  
TRAINING WORKING WITH  
CHI LDREN WHO ARE DEAF AND  
HARD OF HEARING ABOUT THEIR  
PARTICULAR FIELD OF STUDY.

IT IS NOT THE INTENT OF THIS  
WORKSHOP TO GET INTO  
EDUCATI ON.

I BELIEVE THE PANEL WOULD  
AGREE THIS IS NOT THEIR AREA  
OF EXPERTISE.

WE ARE OFFERING TWO  
ADDITIONAL WORKSHOPS  
ADDRESSING THESE IN THE FALL.  
STAY TUNED TO OUR NEWSLETTER  
FOR MORE INFORMATION.

MINNESOTA HANDS &  
VOICESFULLY RECOGNIZES THE  
DIVERSITY OF THE FAMILIES IN  
OUR COMMUNITY.

WE ACKNOWLEDGE EVERY FAMILY  
MAY NOT CHOOSE TO FOLLOW  
THESE NEXT STEPS.

AS PARENTS OURSELVES, WE ALSO  
WALK THIS JOURNEY AND WE  
RESPECT EACH FAMILY'S CHOICE.

JUST SOME TECHNICAL  
DETAILS.

THOSE ATTENDING THE WEBINAR,  
YOU CAN FIND THAT ON THE  
WEBSITE,

[WWW.MNHANDSANDVOICES.ORG](http://WWW.MNHANDSANDVOICES.ORG),

LOOK UNDER CURRENT EVENTS AND  
NEXT STEPS.

IF YOU WISH TO ASK A  
QUESTION, THE E-MAIL ADDRESS  
IS ON THE SCREEN.

SO PLEASE JOIN ME IN  
WELCOMING BARB FRIEDMAN, OUR  
FIRST PRESENTER.  
SHE'S A STAFF AUDIOLOGIST.  
SHE RECEIVED HER  
UNDERGRADUATE DEGREE AT THE  
UNIVERSITY OF MINNESOTA,  
MASTER'S DEGREE AT UNIVERSITY  
OF WISCONSIN-MADISON, AND HER  
CLINICAL DOCTORATE IN  
AUDIOLOGY AT THE UNIVERSITY  
OF FLORIDA.  
HER AREA OF EXPERTISE  
INCLUDES ASSESSMENT, SHE

WORKS WITH CHILDREN, HEARING  
AIDS AND COCHLEAR IMPLANTS.

THANK YOU, BARB, FOR JOINING  
US.

>> DR. FRIEDMAN:

TECHNOLOGY.

HOW DO I GET TO MY SLIDES?

SO WHILE I'M WAITING FOR

THAT, I'M GOING TO START WITH  
A STORY.

SO I'M GOING TO SPEND SOME

TIME TALKING ABOUT HEARING

LOSS AND AUDIOLOGY, BUT I'VE

DONE THIS FOR A LONG TIME,

AND I'VE MET LOTS OF FAMILIES  
AND LOTS OF CHILDREN.

ONE OF MY FAVORITE STORIES IS  
A CALL I GOT FROM A PARENT.

SHE HAD JUST HEARD A

DIAGNOSIS, THE CHILD WAS

UNDER ONE YEAR OF AGE, THIS

IS 25 YEARS AGO, AND SHE

CALLED THE UNIVERSITY AND SHE

SAID, MY CHILD HAS A

SIGNIFICANT HEARING LOSS, I'M

SO WORRIED, I WANT HIM TO BE

A BIOLOGIST.

AND I REMEMBER SITTING IN MY  
OFFICE THINKING TO MYSELF,  
I'M THINKING ABOUT MY  
CHILDREN IN TOILET TRAINING,  
NOT THINKING THAT FAR AHEAD.

AND I SAY TO THEM, AS I  
SAY TO LOTS OF FAMILIES,  
HEARING LOSS ALONE IS NOT THE  
ONLY REASON YOUR CHILD WILL  
BE A BIOLOGIST.

I STILL SEE THIS CHILD.

MY CHILDREN DID GET TOILET  
TRAINED, BUT THIS CHILD IS

NOW I N A PH. D. PROGRAM, NOT  
I N BI OLOGY, BUT BOTANY.

I OFTEN TELL THE PARENT SHE' S  
FAI LED, AND JUST TO THINK  
ABOUT THE FUTURE.

THE LAST APPOINTMENT TH I S  
YOUNG MAN CAME I N AND HE  
BROUGHT HI S GI RLFRI END I N.  
SHE, TOO, WAS I N THE PH. D.  
PROGRAM I N BOTANY.

I WANT TO SAY TO EVERYBODY,  
WHEN YOU LEARN ABOUT ALL THE  
STUFF, EXPECT GREATNESS FROM  
YOUR CHI LDREN.

HEARING LOSS ALONE IS NOT THE  
REASON.

BUT IT'S A LOT OF WORK FROM  
EDUCATORS, FROM PARENTS, AND  
FROM THE CHILD.

BUT EXPECT GREATNESS.

I AM GOING TO TALK ABOUT  
THE EAR AND HEARING, WHO  
AUDILOGISTS ARE -- YOU'LL BE  
MEETING LOTS OF THEM ALONG  
THE WAY --  
HOW THE EAR WORKS, HOW WE  
TEST HEARING IN CHILDREN,

HEARING LOSS AND ITS EFFECTS  
ON COMMUNICATION, TECHNOLOGY  
AND EDUCATION.

WHEN WE SEE CHILDREN,  
THOUGH, THERE ARE PARTS NOT  
SO TECHNICAL.

WE LOOK THROUGH A LENS OF  
FAMILIES, SOME ARE  
EXPECTATIONS, HOW THEY REACT  
TO THE NEWS, CULTURAL NEEDS  
AND GOALS.

SO YOU NEED TO GO TO EVERY  
APPOINTMENT WHEN YOU SEE ALL

OF US, WHAT YOU WANT TO KNOW  
AND WHAT WE CAN PROVIDE FOR  
YOU.

AUDI OLOGI STS ARE  
SPECI ALI STS WHO WORK WI TH  
CHI LDREN WHO ARE HARD OF  
HEARI NG, WE TEST HEARI NG, WE  
WORK WI TH TECHNOLOGY.  
MANY OF US WORK AT CLINI CS,  
HOSPI TALS, BUT THERE' S  
ANOTHER AUDI OLOGI ST WHO WORKS  
I N THE SCHOOLS, THEY' RE THE

LI AI SON BETWEEN THE HOSPI TALS  
AND THE SCHOOLS.

THEY MAKE SURE THE ROOM  
ACOUSTI CS ARE WORKI NG WELL,  
I N THE CLASSROOM THE HEARI NG  
AIDS ARE WORKI NG WELL AND  
GOOD SUPPORT FOR FAMI LIES,  
LETTI NG THE TEACHER KNOW WHAT  
SOME OF THE PROBLEMS THAT MAY  
HAPPEN WI TH THEIR CHI LDREN.

DOCTOR RI MELL LOOKS AT  
DI SEASES OF THE EAR. HOW THE  
EAR WORKS AND SOUND I S PICKED

UP BY THE OUTER EAR, THE  
PINNA, THAT'S THE OUTSIDE.  
WE HAVE A LARGE ONE HERE.  
AND IT GETS DIRECTED THROUGH  
THE EAR CANAL, AND THAT  
VIBRATION GETS SET UP, THE  
EARDRUM IS THEN MOVING, AND  
THEN WE HAVE THREE SMALL  
MIDDLE EAR BONES, YOU  
PROBABLY LEARNED THEM AS THE  
INCUS, MALLEUS OR STAPES.  
FROM THERE, THAT VIBRATION  
GOES IN THE INNER EAR.

THERE' S A SPIRAL SNAIL-LIKE  
CONFIGURATION.

THAT' S THE COCHLEA.

THAT' S THE ORGAN OF HEARING.

IF I WAS TO OPEN THAT UP,

YOU' D HAVE ABOUT 30,000 HAIR

CELLS THAT VIBRATE TO SOUND.

FROM THAT VIBRATION, THERE

WILL BE AN ELECTRICAL

CHEMICAL MOVEMENT TO THE

HEARING NERVE THAT COMES OFF

FROM THE COCHLEA.

SEE IT UP HERE.

RIGHT NEXT TO IT ARE THE  
SEMI CIRCULAR CANALS, AND  
SOMETIMES THOSE ARE RELATED.  
FOR MOST PERMANENT HEARING  
LOSS, IT'S A PROBLEM OF THE  
HAIR CELLS IN THAT COCHLEA.  
SOME DON'T DEVELOP, SOME HAVE  
DIED OFF.  
SO WE'RE TRYING TO PICK UP  
HEARING THROUGH LESS OF THOSE  
HAIR CELLS TO STIMULATE  
SOUND.

GOING BACK TO THE DIAGRAM OF THE EAR, THE OUTER EAR AND THE MIDDLE EAR CAN HAVE DISEASES OR PROBLEMS IN THAT AREA.

THE OUTER EAR YOU CAN HAVE WAX THAT PLUG THE CANAL.

THOSE PROBLEMS ARE CALLED CONDUCTIVE HEARING LOSS PROBLEMS.

PROBLEM OF CONDUCTING THE SOUNDS TO THE ORGAN OF HEARING.

IF THE PROBLEM'S IN THE INNER  
EAR, WE CALL IT  
SENSORI NEURAL.

YOU' LL BE HEARING THOSE TERMS  
FLOATING AROUND.

CONDUCTIVE, SENSORI NEURAL,  
AND SOMETIMES WE CALL THAT  
TOGETHER A MIXED HEARING  
LOSS.

HOW WE TEST HEARING, WE  
USE A BATTERY OF TESTS.  
NO ONE TEST GIVES US ALL THE  
ANSWERS TO TESTING HEARING.

SOME OF THE INPUT IS FROM PARENTS, AS WELL.

WE APPRECIATE WHEN PARENTS GIVE US THEIR IDEAS.

THIS CHILD THERE WITH THE EARPHONES ON, ONE WAY OF TESTING IS GOING THROUGH THE WHOLE SYSTEM, WE'LL HAVE EARPHONES OVER THE HEAD.

IT GOES THROUGH THE WHOLE SYSTEM, GIVES US INFORMATION ON HEARING.

WE CAN BYPASS THAT CONDUCTIVE PORTION, THE OUTER AND MIDDLE

EAR, BY SOMETHING CALLED A  
BONE VIBRATOR.

IT'S A PIECE PLACED ON THE  
BONY PROTRUSION BEHIND YOUR  
EAR.

AND THAT GIVES US INFORMATION  
ON THAT COCHLEA, IT GIVES US  
HEARING.

WE REALLY ARE OFTEN TRYING TO  
LOOK AT BOTH PIECES.

THERE ARE A NUMBER OF WAYS  
TO GET THAT INFORMATION.

WHEN A CHILD IS AN INFANT, IN  
THE NEWBORN HEARING  
SCREENING, WE'LL OFTEN DO  
OTOACOUSTIC EMISSIONS.

SOUND RADIATED BACK.

SO THOSE HAIR CELLS THAT YOU  
SAW RADIATED BACK AND THEY  
COULD MEASURE IT.

IT DOESN'T GIVE US ANY  
INFORMATION ON HOW MUCH  
HEARING LOSS, WHAT TYPE OF  
HEARING LOSS, BUT IT DOES  
GIVE US A QUICK WAY TO KNOW

IF THERE'S HEARING LOSS OR  
NOT.

IT'S A QUICK WAY TO GET  
INFORMATION IN THE NEWBORN  
HEARING SCREENING.

IT'S NOT WITHOUT FAULTS.

AS I MENTIONED, WE NEED TO  
LOOK AT A BATTERY OF TESTS.

BUT IT GIVES US A QUICK WAY  
OF GETTING SOME INFORMATION.

ANOTHER TEST IS AUDITORY  
BRAIN STEM RESPONSE TESTING.

IT LOOKS LIKE AN EEG  
PROCEDURE AND WE LOOK AT THE  
BRAIN WAVES OF HEARING.

THAT OFTEN TAKES A LITTLE  
LONGER.

IF THEY'RE OVER A FEW MONTHS  
OF AGE, WE'LL NEED TO SEDATE  
THE CHILD.

BUT IT DOES GIVE US A LOT OF  
INFORMATION ON HOW MUCH  
HEARING LOSS, WHAT TYPE OF  
HEARING LOSS.

ANOTHER TERM I'LL TALK ABOUT  
IS CONFIGURATION.

WHAT FREQUENCY THE HEARING  
LOSS IS GREATEST AT.

BOTH OF THOSE TESTS ARE  
ELECTRICAL.

THE CHILD DOESN'T HAVE TO  
COOPERATE FOR THEM, AND IT  
GIVES US INFORMATION, BUT NOT  
ALWAYS THE WHOLE PICTURE.

AS THE CHILDREN GET OLDER, WE  
WANT TO DO WHAT'S CALLED A  
BEHAVIORAL TEST, AND THE  
CHILD HAS TO COOPERATE.

AND THEN FOR ME, THAT'S WHERE  
IT GETS TO BE FUN.

AT ABOUT SIX MONTHS OF  
AGE, WE CAN GET A PRETTY GOOD  
HEARING TEST ON MANY  
CHILDREN.

WHAT WE DO, WE PRESENT THE  
SOUND AND WE ASK THAT 6-  
MONTH-OLD AND RAISE THEIR  
HAND, WHICH THEY DON'T, THEY  
LOOK TO THE SOUND AND WE  
REINFORCE THEM WITH A  
FLASHING BEAR OR DANCING  
ANIMAL BEHIND PLEXIGLASS.

THAT KEEPS THEM GOING QUITE A LONG TIME.

AND SOMETIMES WE CAN GET EARPHONE INFORMATION ON THEM, AND OFTENTIMES QUITE A LOT OF INFORMATION.

AS THEY GET OLDER, THE CHILDREN WILL LOOK AT A PROCEDURE CALLED CONDITIONED PLAY AUDIOMETRY.

THAT WILL KEEP THEM LONGER.

SOMETIMES WE HAVE TO PROVIDE M&MS TO KEEP THEM GOING LONGER STILL.

4 OR 5 YEARS OF AGE THEY  
RAISE THEIR HAND TO SOUND.  
WE TRY TO DO TESTING WITH  
EARPHONES AND THEN WITH BONE  
CONDUCTION TO FIND OUT WHERE  
THE HEARING LOSS IS.

ANOTHER PART OF THE  
MEASUREMENT IS CALLED  
TYMPANOGRAMS AND IT MEASURES  
THE EARDRUM MOVEMENT.  
IT NEEDS TO BE DONE REGULARLY  
TO MONITOR THE HEALTH OF THE  
EAR.

IT GIVES US INFORMATION.

IF THE EARDRUM IS NOT MOVING  
NORMALLY, WE'LL SEE A FLAT  
LINE, THE ONE ON THE LEFT  
BECAUSE MAYBE THERE'S FLUID  
ON THE EARDRUM.

IF WE SEE NORMAL MOVEMENT,  
YOU'LL SEE A PEAK LIKE ON THE  
RIGHT.

MAYBE THERE'S WAX IN THE EAR  
OR HOLE IN THE EARDRUM OR PE  
TUBE IN PLACE.

WE HAVE BEEN STRIVING AS  
AUDI OLOGI STS -- NOT TO GIVE  
YOU TOO MUCH I NFORMATI ON, BUT  
I N AUDI OLOGY SCHOOL, WE  
ALWAYS WANT TO TALK ABOUT THE  
AUDI OGRAM.

OVER TIME YOU, TOO, WILL BE  
TALKI NG ABOUT YOUR CHI LD' S  
AUDI OGRAM.

THAT' S THE GRAPH OF HEARI NG  
AND THAT' S HOW WE DEFINE  
HEARI NG AND HEARI NG LOSS.

AND IF WE LOOK AT ACROSS THE  
TOP, THAT'S FREQUENCY OR  
PITCH.

FOR PEOPLE WHO ARE MUSICALLY  
INCLINED, 250 IS MIDDLE C.

THIS IS ONE, TWO, THREE, FOUR  
OCTAVES ABOVE.

THE HUMAN EAR CAN USUALLY  
HEAR 20,000.

AS WE AGE, OUR HEARING STARTS  
TO DROP.

SO JUST BE AWARE THAT YOUR  
CHILDREN PROBABLY WILL HEAR  
BETTER THAN YOU – CHILDREN

WITH NORMAL HEARING WILL HEAR  
BETTER THAN THEIR PARENTS.

BUT THE MOST IMPORTANT  
FREQUENCIES THAT WE'RE  
LOOKING AT RIGHT NOW IS THE  
FREQUENCY FOR SPEECH.

ALONG THE OTHER AXIS IS  
LANGUAGE.

IF WE'RE LOOKING AT AN  
AUDIOGRAM, IF YOU LOOK AT A  
GLASS OF MILK, IT'S FILLED TO  
THE TOP.

ACROSS HERE, THAT WOULD BE  
NORMAL HEARING.

WE'VE DEFINE THEM BY MILD,  
MODERATE, SEVERE, PROFOUND  
DEGREES OF HEARING LOSS.

WHAT'S IMPORTANT ABOUT IT,  
AS I MENTIONED, IS SPEECH AND  
SPEECH SOUNDS.

I PUT IN THE SPEECH BANANA.

ONE PARENT ONCE SAID THEY

REMEMBER THE AUDIOLOGIST

TALKED ABOUT FRUIT, SO I'M

ASSUMING IT'S THE BANANA THEY

WERE TALKING ABOUT, SO I'M  
GOING TO INTRODUCE YOU TO  
THIS FAMOUS BANANA.

SOUNDS GO THROUGH AN ARRAY OF  
FREQUENCIES.

THERE'S SOME THAT ARE REALLY  
IMPORTANT.

THE S SOUND IS A HIGH  
FREQUENCY SOUND, IT'S QUITE  
SOFT, NOT ONLY DO YOU HAVE IT  
IN A WORD LIKE SNOW, BUT IT  
HAS GOT A LOT OF GRAMMATICAL  
MARKERS.

IF YOU WANT TO LEARN PLURALS,  
SHOE, SHOES, YOU WANT TO KNOW  
ABOUT POSSESSION, YOU NEED  
THAT.

SO SOME OF THOSE SOUNDS FOR  
ORAL LANGUAGE ARE GOING TO BE  
REALLY IMPORTANT, SO WE  
REALLY WANT TO MAKE SURE  
CHILDREN ARE HEARING ALL  
THOSE SOUNDS FOR LEARNING  
ORAL LANGUAGE.

ALSO ON THE AUDIOGRAM, WE  
GRAPH OUT RIGHT AND LEFT EAR.

AND THE RED, RIGHT, ROUND  
CIRCLES ARE FOR THE RIGHT  
EAR.

THE BLUE, LEFT, XS ARE FOR  
THE LEFT EAR.

WHEN YOU LOOK AT THAT GRAPH,  
IF THOSE LINES ARE BELOW THAT  
SPEECH BANANA, WE SEE THAT  
WITH THE RIGHT EAR, THAT  
MEANS THOSE SOUNDS AREN'T  
BEING HEARD.

IF THEY'RE ABOVE, SO IF YOU  
LOOK AT THE LEFT EAR, WE SEE  
SOME OF THE SOUNDS ARE BEING

HEARD, BUT SOME OF THE HIGH  
FREQUENCY SOUNDS ARE NOT.

IT'S IMPORTANT TO KNOW  
THAT WE SPEAK THE WAY WE  
HEAR.

SO IF WE'RE NOT HEARING SOME  
OF THOSE SOUNDS, IT WILL BE  
REFLECTED IN THE SPEECH.

IT'S IMPORTANT TO GET ALL  
THAT INFORMATION IN FOR CLEAR  
SPEECH.

TYPES OF HEARING LOSS, AND  
I TALKED ABOUT THAT.

LOTS OF CAUSES, AND I THINK  
PROBABLY DOCTOR RIMELL WILL  
TALK MORE ABOUT THOSE.

CONFIGURATION I MENTIONED.

THAT'S IMPORTANT, BECAUSE IN  
TERMS OF FITTING HEARING  
AIDS, SO THE ONE ON THE  
RIGHT, WE WOULD CALL THAT A  
HIGH PITCH, HIGH FREQUENCY  
HEARING LOSS.

THE ONE ON THE LEFT IS A FLAT  
CONFIGURATION, BUT IT ALSO

GIVES US SOME OTHER  
INFORMATION WHEN WE GRAPH OUT  
THAT AUDIOGRAM THAT YOU'LL BE  
SEEING OVER AND OVER AGAIN.

THIS IS GOING THROUGH THE  
WHOLE SYSTEM, THE RIGHT AND  
LEFT EAR.

THESE HATCH MARKS ARE FOR  
BONE CONDUCTION, THAT GOES  
RIGHT TO THE ORGAN OF  
HEARING, THE COCHLEA.

SO WHAT IT TELLS US IS THE  
POTENTIAL HEARING IS NORMAL,

SOMETHING IS BLOCKING THE WAY  
AND THAT'S CONDUCTIVE HEARING  
LOSS.

ANOTHER PART OF WHEN WE  
TEST HEARING IS WE LOOK AT  
CLARITY.

AND WHEN CHILDREN GET OLDER  
AS THEY AGE, WE WANT TO KNOW  
IF THERE'S DISTORTION IN THE  
HEARING SYSTEM.

SOMEWHAT LIKE AN UNTUNED  
RADIO, YOU CAN TURN IT UP  
LOUDER, BUT IT'S NOT CLEAR.

THAT' S PART OF SOME HEARING  
LOSS, AS WELL.

OFTENTIMES IF THERE' S NO  
CLARITY, YOU CAN GET LINES,  
THEY HEAR WHAT THEY WANT TO  
SEE, WE SEE MORE BEHAVIORAL  
PROBLEMS WITH CHILDREN,  
BECAUSE THEY' RE MISSING  
HEARING AND PEOPLE ASSUME  
THEY' RE HEARING.

ONE PERSON TOLD ME THE CHILD  
ONCE CAME UP AND SAID, "WHY  
DO YOU START EVERY STORY WITH

ONE ON A DIME?" ONCE UPON A  
TIME.

SO THAT'S A CHILD THAT'S  
MISSING THOSE HIGH FREQUENCY  
SOUNDS, THINGS AREN'T CLEAR  
ENOUGH.

IN DOING THIS  
PRESENTATION, I ASKED PARENTS  
OF KIDS WHO HAVE HEARING  
LOSS, WHAT WOULD YOU HAVE  
LIKED TO KNOW AT THE  
BEGINNING?

THEY SAID, I WOULD LIKE TO  
KNOW AS MUCH ABOUT HEARING  
AIDS AS POSSIBLE.

THAT'S ANOTHER LECTURE, BUT I  
WANT TO TALK ABOUT HEARING  
AIDS.

THEY DO AMPLIFY SOUND.

AND IF SPOKEN LANGUAGE IS  
IMPORTANT, WE NEED HEARING  
AIDS TO COUNTERACT THE  
HEARING LOSS.

IT'S NOT ONE-SIZE-FITS-ALL  
WITH HEARING AIDS.

THERE ARE PROGRAMS FOR EACH  
CHILD'S HEARING LOSS AND  
THEY'RE ADJUSTED ACCORDINGLY.  
ODDLY ENOUGH, AS THE EAR  
GROWS, WE NEED TO ADJUST  
THAT.

THINGS THAT HAVE BEEN  
DEVELOPED ALONG THE WAY,  
WE'RE VERY FORTUNATE IN THIS  
STATE TO HAVE THE HEALTH  
LIONS LOANER PROGRAM, SO WHEN  
A CHILD GETS DIAGNOSED,  
THERE'S SIX MONTHS THAT  
HEARING AIDS CAN BE LOANED

UNTIL WE KNOW MORE ABOUT  
HEARING LOSS OR INSURANCE  
THAT WILL COVER THE HEARING  
AIDS.

ALSO SOME VERY WONDERFUL  
PARENTS WENT TO THE  
LEGISLATURE ABOUT FIVE YEARS  
AGO AND GOT HEARING INSURANCE  
COVERAGE FOR HEARING AIDS, SO  
THAT'S SOMETHING THEY DON'T  
HAVE TO WORRY ABOUT.

IF SPOKEN LANGUAGE IS  
IMPORTANT, YOU NEED TO HAVE

HEARING AIDS WORN ALL THE  
TIME, EXCEPT SLEEPING AND  
BATHING.

YOU WANT IT TO BE A POSITIVE  
EXPERIENCE. WE'RE HAVING  
BRIGHTLY COLORED HEARING AIDS  
ON CHILDREN, SOME WITH  
GLITTER.

ODDLY ENOUGH, I SEE A LOT OF  
ADULTS, MY ADULTS WHO HAVE  
HAD HEARING LOSS FOR A LONG  
TIME SAY I DON'T WANT TO EVER  
SEE A BEIGE HEARING AID.

THEY' RE GETTING PINK AND BLUE  
AND THEY HAVE ACCEPTED IT AND  
THEY WANT TO MAKE IT FUN.

THERE' S LOT OF TECHNOLOGY  
FOR HEARING AIDS.

FM TECHNOLOGY WHERE A CHILD  
WEARS A MICROPHONE.

THAT' S GOOD IN LECTURE  
SITUATIONS, IT' S GOOD IN  
SPORT SITUATIONS, SO THERE' S  
LOT OF ADJUSTMENTS MADE FOR  
HEARING AIDS.

STREAMERS ARE A NEW TERM IN  
CONNECTION.

SOME ARE WIRELESS.

NOT THINGS WE PUT ON OUR  
INFANTS, BUT THE FUTURE OF  
CHILDREN IS EXCITING AND IT  
GETS MORE AND MORE EXCITING  
EVERY YEAR.

COCHLEAR IMPLANTS ARE FOR  
CHILDREN WHO HAVE VERY FEW OF  
THOSE HAIR CELLS.

IT'S WHERE HEARING AIDS  
AREN'T PROVIDING ENOUGH

AUDI TORY I NFORMATI ON AFTER A  
HEARI NG AI D TRI AL.

I T BYPASSES THE MI DDLE EAR  
AND ELECTRODES PLACED I N THE  
COCHLEA.

I ' M MI S-SAYI NG THI S, WHEN WE  
LOOK AT THE SMALL BONES I N  
THE BODY, THEY ARE THE  
SMALLEST BONES I N OUR BODY.

I T I S THE SI ZE OF A PEA.

WHEN DOCTOR RI MELL TALKS,  
HE' S DOI NG A VERY GOOD JOB ON  
SURGERY.

TO ME, IT'S HARD TO BE AN  
AUDI OLOGI ST WI THOUT  
I NTRODUCI NG EDUCATI ON.

THERE'S MANDATES, THERE'S  
SERVI CES THAT ARE PROVI DED,  
AND THERE'S A WEBSI TE TO FI ND  
OUT WHAT SERVI CES ARE  
AVAI LABLE THROUGH OUR SCHOOL  
DI STRI CT.

ANY FAMI LY COULD BE A  
CONSUMER AND OBSERVE  
DI FFERENT CLASSROOMS, TALK TO  
I NDI VI DUALS WI TH HEARI NG  
LOSS.

YOU WANT TO KNOW WHAT'S BEST  
FOR YOUR CHILD AND YOUR  
FAMILY.

COMMUNICATION.

LOTS OF THINGS ARE EXCITING.

I'VE DONE THIS FOR A LONG

TIME, WE REALLY HAVE LOTS OF

OPTIONS FOR FAMILIES.

MINNESOTA HANDS & VOICES, WHO

HELPED SPONSOR THIS, IS A

GOOD RESOURCE TO TALK ABOUT

ALL THE DIFFERENT MODES OF

COMMUNICATION.

AGAIN, YOU HAVE TO LOOK AT  
WHAT'S RIGHT FOR YOUR FAMILY  
AND CHILD.

THERE'S NO ANSWER THAT YOU  
MAKE A SELECTION WITH THAT'S  
RIGHT THAT YOU CAN'T CHANGE  
YOUR MIND.

WHAT WE LIKE TO LOOK AT  
WITH CHILDREN IS NOT THE  
PERFECT CHILD, BUT THE CHILD  
WHO HAS GOOD WELL BEING.  
WE DON'T WANT PARENTS TO COME  
IN AND HAVE US FIX THEIR

CHILDREN, WE WANT TO DO  
TECHNOLOGY IF THEY WANT  
SPOKEN LANGUAGE SO THEY CAN  
HEAR, BUT YOU WANT YOUR CHILD  
TO BE HAPPY, YOU WANT YOUR  
CHILD TO LEARN, AND YOU WANT  
EDUCATION.

EARLY INTERVENTION DOES  
WORK.

AGAIN, I'M IN THIS WITH A  
STORY.

I HAD THREE FAMILIES OF FIRST  
GRADERS COME IN, AND ALL

FAMILIES SAID TO ME, THEY  
SAID THEIR CHILD WAS THE TOP  
OF THEIR CLASS AND THEY ALL  
HAD SIGNIFICANT HEARING LOSS.  
OF COURSE, AGAIN I WANTED TO  
GO HOME AND THINK, OH, GOSH,  
WHAT ARE MY CHILDREN DOING  
THROWING SPIT BALLS?  
EARLY INTERVENTION DOES MAKE  
A DIFFERENCE, LOTS OF WORK.  
THERE'S LOTS OF GOOD PUBLIC  
AND PRIVATE EDUCATION,  
OBSERVE.  
FAMILY SUPPORT IS ESSENTIAL.

YOU CAN' T JUST SEND THE CHI LD  
OFF TO SCHOOL AND ASSUME THAT  
GREATNESS WI LL HAPPEN.

BUT GREATNESS CAN HAPPEN.

EXPECT I T, BUT DON' T  
EXPECT PERFECTI ON.

WE SHOULDN' T WI TH ANY CHI LD.

EXPECT GOOD BEHAVI OR, ENJOY  
YOUR CHI LD.

AND LASTLY, HERE ARE SOME  
RESOURCES THAT WI LL BE I N THE  
HANDOUT.

MINNESOTA DEPARTMENT OF  
HEALTH HAS A GREAT WEBSITE.  
IT'S A REALLY GOOD RESOURCE.

[APPLAUSE]

>> ARE THERE ANY  
QUESTIONS FOR BARBARA?

>> AUDIENCE MEMBER:  
SHOULD TESTS BE HAPPENING  
NOW?  
SHOULD WE BE ASKING QUESTIONS  
RIGHT NOW OR SHOULD WE ASK  
THEM AT THE END?  
CAN WE ASK QUESTIONS NOW?

MY QUESTION IS ABOUT THIS:  
SOMETIMES HEARING AIDS WORK  
WELL FOR CHILDREN AND  
SOMETIMES COCHLEAR IMPLANTS.  
HOW DO YOU – HOW DO YOU KNOW  
THAT HEARING AIDS MAY BE NOT  
SUCH A BAD THING FOR A CHILD  
AT SUCH A YOUNG AGE WITHOUT  
THE CHILD TELLING YOU IT'S  
NOT WORKING?  
HOW DO YOU MEASURE THAT TO  
FIGURE OUT IF IT'S BENEFICIAL  
OR IF YOU NEED TO – OR IF

YOU' D LI KE TO DO A COCHLEAR  
I MPLANT?

>> DR. FRI EDMAN: WHAT  
WE DO WI TH YOUNG, YOUNG  
CHI LDREN -- WI TH OLDER  
CHI LDREN, OBVI OUSLY IT' S A  
LI TTLE BI T SI MPLER.

WI TH YOUNGER CHI LDREN, WE DO  
HA VE A TRI AL PE RI OD OF  
HEARI NG AI DS.

IT' S MANDATORY.

AND WHAT WE DO IS GET THE  
SCHOO L I NVOLVED, WE GET THE

PARENTS INVOLVED AND WATCHING  
DIFFERENCES IN BEHAVIOR.

WE DO LOTS OF BEHAVIORAL  
TESTS TO SEE WHAT TYPE OF  
BENEFIT.

WE WANT SPEECH TO BE AUDIBLE,  
AND IF ALL THE TESTS AND  
FAMILY INPUT SUGGESTS THAT  
SPEECH IS INAUDIBLE, THAT'S  
WHEN WE LOOK AT A COCHLEAR  
IMPLANT.

>> AUDIENCE MEMBER:  
SO WOULD YOU SAY MAYBE LIKE  
18-MONTH-OLD PICKING UP

LANGUAGE, IF WE START  
SPEAKING DIFFERENTLY – HOW DO  
YOU MEASURE WITH THE LANGUAGE  
ACQUISITION IF IT'S WORKING?  
' CAUSE MAYBE THEY'RE BEHIND  
FOUR AND THEY'RE STARTING TO  
CATCH UP.

HOW DO YOU MEASURE IF IT'S  
BEING SUCCESSFUL IN THAT WAY?  
IS IT BY BEHAVIOR ALONE, LIKE  
HOW THEY RESPOND TO SPEECH OR  
IS IT – OR WHAT – HOW DO YOU  
MEASURE THAT?

>> DR. FRIEDMAN: WE' RE  
NOT MEASURING LANGUAGE AT  
THAT TIME.

WE' RE LOOKING TO SEE IF  
THEY' RE SHOWING ANY  
BEHAVIORAL RESPONSE TO SOUND,  
TO THEIR NAME.

WE ALSO DO -- AT SIX MONTHS  
OF AGE, WE CAN DO BEHAVIOR  
AUDIOLOGIC TESTING.

WE DO THAT BRAIN STEM  
RESPONSE TEST TO SEE THE  
SEVERITY OF THEIR HEARING  
LOSS.

SO WE LOOK AT A COMBINATION  
OF TESTS TO SEE.

BUT WE'RE NOT LOOKING AT  
LANGUAGE AT THAT TIME, WE'RE  
REALLY LOOKING AT ARE THEY  
SHOWING ANY AWARENESS TO  
SOUND.

AND IF THEY'RE NOT AND WE'VE  
DONE ALL THE BEHAVIORAL  
DIAGNOSTIC TESTS, THEN WE'LL  
LOOK AT COCHLEAR IMPLANT.

>> AUDIENCE MEMBER:

OK.

THANK YOU.

I HAD SOME EXPERIENCE WITH MY OLDEST SON.

WE WERE TESTING TO SEE ABOUT HIS HEARING.

HE WAS BORN DEAF AND SO AT ABOUT 2 YEARS HE – HE WAS PUT INTO, YOU KNOW, EARLY INTERVENTION PROGRAM.

LATER I NOTICED, YOU KNOW, HE – THEY TESTED HIM AND HE WASN' T DEAF.

SO AT FIRST WE THOUGHT THERE WAS HEARING LOSS AND THEN THERE WASN' T LIKE WE THOUGHT

THAT THERE WAS, BUT THERE WAS NEVER A RESPONSE WHEN HE WAS YOUNGER.

THERE WAS NO RESPONSE IF HE WAS -- LIKE YOU WOULD THINK THERE WOULD BE.

SO WE HAD PUT HEARING AIDS ON AND THE CHILD HAD IGNORED THESE THINGS, SO APPARENTLY -- IT TURNED OUT LATER THAT THE CHILD HAD BEEN JUST IGNORING WHAT WAS HAPPENING.

SO WE HAD THESE RESPONSES, WE HAD THESE NOISES AND SHE

RESPONDED AS IF SHE WASN' T  
HEARING ANYTHING, EVEN THOUGH  
SHE ACTUALLY WAS.

SO IT SEEMS LIKE SOME OF THEM  
CAN BE KIND OF INACCURATE OR  
NOT QUITE – NOT WHAT WE WERE  
– IT SEEMS – SO WHAT HAPPENED  
WAS THAT ALTHOUGH – ALTHOUGH  
THE CHILD WAS HEARING, THE  
RESPONSES APPEARED THAT THERE  
WAS HEARING LOSS EVEN THOUGH  
THERE WASN' T.

>> DR. FRIEDMAN: WE  
DON'T LOOK AT ANY ONE TEST  
FOR CHECKING CHILDREN.  
SO BEHAVIORAL TESTS GIVE US  
INFORMATION, AUDITORY BRAIN  
STEM RESPONSE TESTING GIVES  
US INFORMATION, INPUT FROM  
THE SCHOOLS.  
AND THE TEACHES DO COME OUT  
AT INFANCY TO THE HOME AND  
TEST.  
SO ANY ONE TEST ISN'T GOING  
TO MAKE OUR DECISION TO DO A  
COCHLEAR IMPLANT, WE DO A

BATTERY OF TESTS, ESPECIALLY  
WITH YOUNG CHILDREN.

>> AUDIENCE MEMBER:

THANK YOU.

>> DR. RIMMEL: I'M

DOCTOR FRANK RIMMEL, ONE OF  
THE OTOLARYNGOLOGISTS.

I'M WAITING FOR MY SLIDES TO  
GET LOADED HERE.

ALL RIGHT.

SOME OF WHAT BARB COVERED I  
COVERED IN MY SLIDES, FOR THE  
SAKE OF TIME, I MAY GO

QUICKER THROUGH SOME SLIDES  
HERE.

THE PREVALENCE OF HEARING  
LOSS IS QUITE FREQUENT IN THE  
UNITED STATES.

IT'S THE MOST COMMON ISSUE  
THAT WE FIND AS PHYSICIANS.

I'M SUPPOSED TO STAND ON THE  
BLACK PAD, AM I FINE?

IT'S PROBABLY BETTER THEY  
DON'T SEE MY FACE.

[LAUGHTER]

>> DR. RIMMELL: ALL  
RIGHT.  
SO IT IS THE MOST COMMON  
ISSUE WE FIND WITH CHILDREN  
WHEN THEY'RE BORN COMPARED TO  
ANY OTHER ISSUES WE FIND WHEN  
A CHILD IS BORN.  
3 OUT OF 1,000 CHILDREN ARE  
BORN WITH HEARING LOSS.  
THAT CAN GIVE YOU THE NUMBER  
HERE.  
AND THAT INCREASES BY SCHOOL  
AGE TO 6 PER 1,000, AS

OPPOSED TO 3 PER 1,000 AT  
BIRTH.

THE ISSUE, OBVIOUSLY, IS  
ONCE THEY'RE IDENTIFIED EARLY  
NOW DAYS IS HOW TO HANDLE  
THEM AND TREATMENT OPTIONS.  
CHILDREN – AND IT'S NOT JUST  
BILATERAL HEARING LOSS, BUT  
IT'S UNILATERAL HEARING LOSS.  
IF THEY'RE IN A HEARING  
ENVIRONMENT, DON'T DO WELL IN  
SCHOOL, AND ABOUT A THIRD OF  
CHILDREN CAN FAIL A GRADE IF

THEY' RE IN A STRAIGHT HEARING ENVIRONMENT IF THEY HAVE A ONE-SIDED HEARING LOSS.

IT' S VERY IMPORTANT THAT THESE ISSUES GET IDENTIFIED AND TREATED PROPERLY.

WE' LL SKIP THAT ONE, GO TO THE NEXT SLIDE.

AS BARB ELUDED, THIS IS REALLY – TREATMENT OF CHILDREN WITH HEARING ISSUES IS DONE BY A TEAM AND YOU' RE MEETING SOME OF THAT TEAM.

THERE ARE OBVIOUSLY NUMEROUS  
ISSUES THAT ARE USED FOR  
TREATING HEARING LOSS.

THERE'S DEAF COMMUNITY,  
INSURANCE ISSUES, TEACHERS,  
GENETICS, INFECTIOUS DISEASE,  
ALL THESE PEOPLE ARE  
IMPORTANT IN TRYING TO COME  
UP WITH A PLAN FOR THE FAMILY  
IN TREATING THAT CHILD TO  
EVERYONE'S EXPECTATIONS,  
ESPECIALLY PARENTS'  
EXPECTATIONS FOR WHAT THEY  
DESIRE FOR THEIR CHILD.

AS BARB DISCUSSED, THE  
HEARING LOSS IS DISCUSSED BY  
THE DEGREE AND THE TYPE.

AS NOTED, ONCE YOU GET INTO  
THE DECIBELS OF 50 AND  
HIGHER, THEN I USUALLY START  
GETTING INVOLVED.

I'M GOING TO TALK ABOUT THE  
DIFFERENT TYPES OF HEARING  
LOSS.

THIS IS THE SAME PICTURE  
BARBARA SHOWED YOU.

WHERE DID THE LASER POINTER  
GO?

BARB STOLE THE LASER POINTER.  
THAT'S A FREE GIFT, WE ALL  
GET ONE WHEN WE'RE DONE.

SO AS BARBARA DISCUSSED,  
THIS IS THE OUTER EAR TO THE  
EARDRUM.

BETWEEN THE EARDRUM AND THE  
COCHLEA IS THE INNER EAR  
SPACE AND THEN THE MIDDLE  
EAR.

IT'S INTERESTING THAT NEXT TO THE HEARING NERVE RUNS A NERVE CALLED THE FACIAL NERVE, WHICH IS IMPORTANT FOR US AS A SURGEON AND BRINGS RISK TO ANY CHILD THAT HAS EAR SURGERY AT ALL.

IT'S OBVIOUSLY – IF THAT NERVE IS DAMAGED, THAT'S THE NERVE THAT HELPS CONTROL EYE CLOSURE, MOUTH MOVEMENT.

IF THAT NERVE IS INJURED AND THAT RESULTS IN INJURIES TO THE FACE.

THE FIRST KIND OF LOSS IS  
CONDUCTIVE HEARING LOSS.  
AND CONDUCTIVE HEARING LOSS  
HAS TO DO WHEN THERE'S A  
PROBLEM IN THAT MIDDLE EAR.  
SO IT WOULD BE DISEASES OF  
THE EAR CANAL ITSELF,  
DISEASES OF THE TYMPANIC  
MEMBRANE, DISEASES OF THAT  
SPACE WITH THE LITTLE HEARING  
BONES AT THE MIDDLE EAR  
SPACE, DISEASES OF THE  
OSSICLES.

THE EUSTACHIAN TUBE IS THAT  
TUBE WHEN YOU GO IN THE  
AIRPLANE AND THE AIRPLANE  
LANDS AND YOU FEEL THAT  
PRESSURE BEHIND YOUR EARDRUM.

VARIOUS DISEASES CAUSE BACK  
PRESSURE IN THAT EAR SPACE  
CAN CAUSE PROBLEMS.

YOU CAN HAVE DISEASES OF THE  
FACIAL NERVE OR OF YOUR  
JUGULAR VEIN OR CAROTID  
ARTERY.

THE JUGULAR VEIN OR CAROTID  
ARTERY CAN BE INJURED,  
THEY' RE IN A TIGHT SPACE AND  
CORNER.

IN THEORY, ANY TYPE OF  
CONDUCTIVE LOSS IS TREATED  
EITHER MEDICALLY OR  
SURGICALLY OR CAN BE  
ATTEMPTED IF ONE DESIRES.  
GETTING BACK TO THAT  
CONFIGURATION AGAIN, JUST TO,  
AGAIN, EMPHASIZE, THIS IS THE  
CONDUCTIVE LOSS.

IT USUALLY HAPPENS BETWEEN  
THE EARDRUM, THE THREE  
HEARING BONES, THE EUSTACHIAN  
TUBE, IN THAT AREA.

THE JUGULAR VEIN AND CAROTID  
ARTERY RUN THROUGH THERE.

SENSORY HEARING LOSS IS  
DAMAGE TO THE HAIR CELLS.

I'LL GO BACK TO THAT. THE  
HAIR CELLS LINE INSIDE THE  
COCHLEA.

IT AFFECTS, OBVIOUSLY, SPEECH PERCEPTION AND HAS MULTIPLE CONFIGURATIONS ON AUDIOGRAM.

IN THEORY FOR US, ALL FORMS OF SENSORY HEARING LOSS CAN BE ADDRESSED SURGICALLY, ASSUMING THERE'S RELATIVELY NORMAL NEUROLOGIC STATUS AND IF THE FAMILY CHOOSES TO HAVE THAT TREATED.

FOR US AS SURGEONS, WE VIEW THE TIMING OF THAT AS SOMEWHAT CRITICAL.

NO SENSORI NEURAL HEARING LOSS  
CAN BE TREATED MEDICALLY TO  
DATE.

BUT IT'S A CHANGE IN OUR  
FIELD.

20 YEARS AGO WE DIDN'T VIEW  
SENSORI NEURAL HEARING LOSS AS  
BEING ABLE TO BE TREATED  
SURGICALLY.

SURGICAL TECHNIQUES ARE  
VOWING TO TRY TO TREAT  
SENSORI NEURAL HEARING LOSS  
WHERE HEARING AIDS CAN'T BE  
USED OR DON'T FUNCTION.

THERE' S AN APPROACH TO  
INSERTING HEARING AIDS, AND  
WE' LL SEE HOW THAT EVOLVES  
OVER THE NEXT DECADE.

THERE' S MIXED LOSS WHICH  
INVOLVES SENSORY AND  
CONDUCTIVE COMPONENTS.  
FOR US AS SURGEONS, AND  
OBTAININGLY FOR AUDIOLOGISTS,  
IT' S VERY IMPORTANT TO  
CHARACTERIZE THEM AS  
BILATERAL AND UNILATERAL FOR  
THE CHILD.

HEARING LOSS – I TEND TO VIEW PROBLEMS AS LIFE-THREATENING OR NOT LIFE-THREATENING PROBLEM.

HEARING LOSS IS NOT A LIFE-THREATENING PROBLEM.

SO THERE'S TIME TO SIT DOWN, DISCUSS, TALK ABOUT IT, SEE WHAT THE OPTIONS ARE, HAVE TIME TO MAKE DECISIONS.

ALL RIGHT.

I'M GOING TO SKIP THESE FOR THE SAKE OF TIME HERE.

SO BEAR WITH ME, I APOLOGIZE  
IT'S A LITTLE MISORGANIZED.

SOME OF THE COMMON RISK  
FACTORS FOR HEARING LOSS,  
PREMATURE BIRTH, VARIOUS  
INFECTIONS THAT DOCTOR  
SCHLEISS IS GOING TO TALK  
ABOUT LATER.

THESE ARE VERY IMPORTANT TO  
TALK ABOUT AND FIGURE OUT.

BRAIN INFECTIONS, THERE ARE  
VARIOUS DRUGS THAT CAN DAMAGE  
THE EARS THAT ARE GIVEN

PARTI CULARLY SMALL BABI ES AND  
PREMATURE BABI ES EARLY ON.

THERE' S CHI LDREN THAT HAVE  
LATE ONSET.

WE HAVE EARLY SCREENI NG.

SOMETI MES, AS PEOP LE KNOW,

OUR TESTS AREN' T PERFECT AND

THEY MAKE A MI STAKE AND

THI NGS GET MI SSED, SO IT' S

VERY I MPORTANT TO PAY

ATTENTI ON TO PARENTS WHO FEEL

LI KE THEIR CHI LD HAS A

HEARI NG LOSS AND WE DI SCOVER

THESE LATER JUST BY REPEATING TESTS.

THERE' S VARIOUS SYNDROMES ASSOCIATED WITH HEARING LOSS, SO IT MAY NOT BE THE HEARING, IT MAY BE KIDNEY PROBLEMS, VISUAL PROBLEMS. FOR US, AGAIN THE EARLIEST WAY TO IDENTIFY THE HEARING LOSS, THIS IS WHERE THE OTHER PHYSICIANS GET INVOLVED, NOT JUST THE EAR, NOSE AND THROAT DOCTOR, THE INFECTIOUS

DI SEASE PHYSI CI ANS, THE  
PEDI ATRI CI AN TO FIND THOSE  
OTHER ISSUES SO THEY CAN BE  
DI SCOVERED AND TREATMENT  
OPTI ONS EXPLAINED TO THE  
PARENTS.

AND THERE ARE MANY OF THEM.

IT WOULD BE A WHOLE OTHER  
HOUR OF LECTURE TO HEAR ABOUT  
ALL THE DI FFERENT KI NDS THERE  
ARE.

AGAIN, IT'S VERY IMPORTANT  
TO PAY ATTENTION TO THE  
PARENTS' CONCERNS AND WHAT  
THEY'RE NOTICING.

I JUST WANT TO MAKE A COMMENT  
HERE.

IT'S VERY COMMON FOR PARENTS  
TO SAY, WELL, YOU KNOW, MY  
CHILD STARTLES WHEN THEY HEAR  
SOMETHING DROP OR THEY HEAR A  
LOUD NOISE.

AGAIN, OUR OBJECT WITH  
HEARING IS THAT IT'S NOT  
NOISE, IT'S SPEECH.

WE TRY TO DIAGNOSE THE  
CHILDREN EARLY AND HELP THEM.

50% ARE ENVIRONMENTAL AND  
50% ARE GENETIC, THAT'S  
GROSSLY HOW WE CHARACTERIZE  
THEM.

THERE ARE VARIOUS GENETIC  
CAUSES, AND WE'LL LEAVE THAT  
TO DOCTOR SCHIMMENTI TO  
DISCUSS.

WE FEEL IT'S IMPORTANT TO  
GIVE THE PARENTS THE OPTION  
TO DECIDE WHICH ROUTE THEY

WANT TO GO AND IF THEY' RE  
GOING TO BE ORAL TO WORK ON  
THOSE SERVICES.

THE MEDICAL WORKUP, AGAIN,  
IS A COMPLETE HISTORY, BOTH  
FAMILY HISTORY, THE BIRTH  
HISTORY, LOOKING AT THE EXAM  
OF THE EARS AND THE REST OF  
THE BODY.

WE OFTEN GET X-RAYS, CT SCAN  
AND MRI .

THE ISSUES WITH THOSE THE  
CHILD HAS TO HOLD STILL, MOST  
CHILDREN DON'T HOLD STILL.

THE CT SCAN CAN BE DONE  
MAKING THEM HUNGRY FOR FOUR  
OR FIVE HOURS, THEY'LL EAT  
AND THEY'LL FALL ASLEEP AND  
THEY'RE STILL.

THE CT SCAN LOOKS AT BONE,  
THE MRI LOOKS AT THE TISSUE  
INSIDE THE BONE, THAT'S WHY  
WE DO BOTH.

THE MRI SHOWS US A HEARING NERVE, THE CT SCAN DOES NOT. THERE CAN BE PROBLEMS WITH THE BONE THAT FORM THE COCHLEA, THAT'S WHY WE GET BOTH, THEY SHOW US TOTALLY DIFFERENT THINGS.

OBVIOUSLY WE GET THE HISTORY AND THE COMPLICATIONS DURING BIRTH.

FAMILY HISTORY, WHICH I DISCUSSED, SEEING THE EYE DOCTOR, WHICH IS IMPORTANT AT

SOME POINT, AGAIN BECAUSE  
HEARING LOSS CAN BE  
ASSOCIATED WITH VISUAL LOSS.  
AND IT'S IMPORTANT TO PICK  
THOSE UP AHEAD OF TIME.

NOW I'M GOING TO GO ON TO  
COCHLEAR IMPLANTS A LITTLE  
BIT HERE.

AGAIN, THE HEARING AID  
BASICALLY JUST MAKES THE  
SOUND LOUDER.

AND I THINK BARB SHOWED YOU  
THE BANANA HERE, AND IT'S

USUALLY FOR THOSE CHILDREN  
THAT ARE IN THAT LOWER END ON  
CONSISTENT MULTIPLE TESTING  
HERE WHERE WE KNOW THAT  
DEVELOPING SPEECH IS MORE  
DIFFICULT OR IMPOSSIBLE WITH  
THE STANDARD HEARING AIDS.

COCHLEAR IMPLANT, THE  
SIGNAL'S PICK UP BY  
MICROPHONE LOCATED IN THE  
HEADSET WHICH IS WORN AT EAR  
LEVEL.

IT LOOKS A LITTLE LIKE A  
HEARING AID.

IT'S CARRIED TO A SPEECH  
PROCESSOR.

THE PROCESSOR DIGITIZES THE  
SOUND AND THE SIGNALS.

THE COIL SENDS THE SIGNALS TO  
THE IMPLANT IN THE SKIN.

THE INTERNAL PART IS PUT IN  
SURGICALLY UNDER THE SKIN,

AND THREE WEEKS LATER THE

EXTERNAL PART AFTER THE

INTERNAL PART IS HEALED.

AS I PREVIOUSLY SAID,  
THERE'S A TEAM INVOLVED.  
PEOPLE FEAR THE SURGERY THE  
MOST.

I'LL DO THE SURGERY, I'LL SEE  
YOU ONCE POST OPERATIVELY AND  
THEN THAT'S IT.

IT'S REALLY SEEING THE  
AUDI OLOGI STS LIKE BARB AND  
THE SPEECH PEOPLE AND THERE'S  
LOTS OF VISITS OVER MANY  
YEARS.

IT'S NOT MAGIC, IT DOESN'T  
WORK WITHOUT THE REST OF THE  
TEAM INVOLVED.

THE BENEFITS OF COCHLEAR  
IMPLANTATION, THE EARLIER  
THEY GET IN, THE BETTER.

THIS IS WHERE SPEECH COMES  
IN, IT LOOKS LIKE HEARING  
AID, HEARING AID LEVEL.  
THERE'S A MAGNET OF OPPOSITE  
POLARITY UNDER THE SKIN.

THIS ALLOWS IT TO STAY ON AND TRANSMIT SIGNALS THROUGH A WIRE THAT WE PUT INSIDE THE COCHLEA WHICH STIMULATES THE HEARING ORGAN AND BYPASS THE HAIR CELLS.

BEING FASCINATED WITH SPORTS AND I MARRIED A JOCK, I LIKE TO SEE HOW THEY STAY ON WHEN THEY DO SPORTS, IT'S OBVIOUSLY AN ISSUE.

PARENTS COME UP WITH UNIQUE WAYS.

THERE' S K I D S W H O P L A Y S P O R T S  
T H A T C A N ' T W E A R H E L M E T S A N D  
T H E R E A R E S P O R T S W H E R E K I D S  
A R E W E A R I N G T H E M .

I T H I N K W E N E E D T O I M P R O V E  
U P O N T H E M F O R K I D S W H O A R E  
W E A R I N G T H E M .

T H E Y ' R E G E T T I N G S M A L L E R A N D  
W I D E R , T H E D E V I C E S .

S O A S T H E S U R G E R Y G O E S , M Y  
J O B I S T O D R I L L A H O L E R I G H T  
H E R E I N T H E C O C H L E A A N D F E E D  
T H A T W I R E I N T H E R E .

AND IT'S ABOUT A THREE-HOUR SURGERY.

MY OBJECT IS NOT TO DAMAGE THE FACIAL NERVE.

EVEN THOUGH THE FACIAL NERVE IS THE MOST FEARED INJURY, IT'S THE LEAST COMMON INJURY. THAT IS, IT RARELY OCCURS AND THAT'S DUE TO MODERN TECHNOLOGY.

IN OUR CENTER, THERE'S BEEN ONE INJURY 20 YEARS AGO OUT OF HOW MANY IMPLANTS OUT OF 800?

WE HAVEN' T HAD ONE SINCE  
THEN.

I THINK MOST MAJOR CENTERS IN  
THE UNITED STATES DON' T HAVE  
THESE, BUT IT IS A  
DEVASTATING INJURY WHEN IT  
HAPPENS.

BUT IT' S PRETTY UNCOMMON NOW  
DAYS.

THIS IS THE DEVICE THAT' S  
PUT UNDER THE SKIN.

THIS IS THE MAGNET OF  
OPPOSITE POLARITY THAT THE

OUTER PART STICKS TO AND THIS IS ALL THE ELECTRONIC PACKAGE.

THIS IS THE WIRE WE INSERT WITH THE VARIOUS ELECTRODES ON IT.

AGAIN, JUST ANOTHER PICTURE OF DRILLING THE HOLE AND INSERTING THE WIRE INSIDE OF IT.

THE OUTER PIECES THAT GO BEHIND THE EAR WITH THE MAGNET.

AGAIN, AS I STATED, THIS IS  
REALLY DONE BY A TEAM  
APPROACH.

THE SURGEON I CONSIDER IS  
PROBABLY ONE OF THE LEAST  
IMPORTANT PARTS OF IT.

FOR A COCHLEAR IMPLANT, THE  
PATIENT UNDERGOES A PRETTY  
INTENSE MEDICAL EVALUATION.  
BOTH SCANS ARE BEING DONE,  
THE MORE INFORMATION I HAVE  
ABOUT ANATOMY, THE MORE

DISCUSSION I CAN HAVE ABOUT  
THE RISKS WITH THE FAMILY.

IT CAN ALSO TELL US IT MAY  
NOT WORK THAT WELL.

THERE ARE CERTAIN ISSUES WE  
CAN FIND, IT ALLOWS US TO  
COUNSEL THE FAMILY COCHLEAR  
IMPLANT MAY NOT GIVE US THE  
BEST RESULT.

IF THE COCHLEA IS VERY  
UNDERDEVELOPED, IT GIVES US  
THE OPPORTUNITY TO TALK ABOUT  
THAT.

WE' RE ABLE TO PUT P I E C E S  
TOGETHER AND FOLLOW DATA AND  
SEE WHO DOES WELL AND  
DOESN' T.

AS A SURGEON, WE DON' T WANT  
TO PUT THESE I N U N L E S S  
THEY' RE GOING TO WORK WELL  
AND SUBJECTING ANYBODY TO  
THOSE K I N D O F R I S K S O F  
SURGERY.

THE I S S U E N O W I S B I L A T E R A L  
I M P L A N T S , S I M U L T A N E O U S O R  
S E Q U E N T I A L .

THAT MEANS THEY' RE GOING IN  
TOGETHER AT THE SAME TIME,  
AND SEQUENTIAL MEANS I' M  
DOING ANOTHER ONE TWO OR  
THREE YEARS LATER.

WE JUST DID ONE YESTERDAY,  
IT' S ABOUT 6 HOURS OF SURGERY  
FOR A 1-YEAR-OLD.

THERE ARE RISKS THAT ARE  
INVOLVED.

MOST CENTERS ARE FINDING OUT  
IT' S NOT VERY RISKY,  
SIMULTANEOUS.

IT SAVES THEM ANOTHER SURGERY  
DOWN THE ROAD.

THE DISADVANTAGES TO IT ARE  
JUST THE LENGTH OF SURGERY.

AND WE OBVIOUSLY WON'T DO IT  
IF THERE'S QUESTIONABLE  
HEARING IN ONE EAR.

IF ONE EAR IS PROFOUND AND  
THE OTHER EAR IS 80 OR 90, WE  
WOULD PROBABLY JUST DO THE  
BAD EAR.

THOSE ARE THE KIND OF  
DISCUSSIONS WE HAVE WITH THE  
PARENTS.

SEQUENTIAL, THE PROBLEM IS  
GOING THROUGH A SECOND  
SURGERY.

HAVING TWO IMPLANTS, IF ONE  
WANTS TO BE ORAL IS AN  
ADVANTAGE OVER ONE,  
PARTICULARLY AS THE CHILD  
BECOMES SOCIALIZED IN SCHOOL.  
AND MOST OF THE DATA SHOWS  
MOST PATIENTS WHO HAVE  
IMPLANTS PREFER 2 OVER 1.

AGAIN, JUST TO GO OVER THE  
RISKS OF IMPLANT SURGERY.

FACIAL NERVE INJURY RARELY HAPPENS, BUT IS THE MOST CONCERNING.

WE'VE PUBLISHED OUR DATA IN MAJOR MEDICAL JOURNALS AT THIS INSTITUTE ABOUT DEVICE INFECTIONS AND DEVICE FAILURES.

IT GETS INFECTED, IT HAS TO BE PULLED WITH SURGERY AND HAS TO BE REPLACED 3 MONTHS LATER WITH ANOTHER SURGERY.

THE SURGERY IS EASIER ONCE  
THE DEVICE IS DONE, BUT IT'S  
STILL A SURGERY.

DEVICE FAILURES DON'T  
HAPPEN FREQUENTLY.

THEY HAPPEN A LITTLE MORE  
FREQUENTLY IN CHILDREN THAN  
THEY DO IN ADULTS.

STATISTICALLY, THESE ISSUES  
OCCUR ABOUT 2 TO 3% OF ADULTS  
AND 5% OF CHILDREN, DEPENDING  
ON WHICH INSTITUTION YOU'RE  
AT.

MENINGITIS IS A RISK JUST BECAUSE IF THE HEARING LOSS IS CAUSED BY ANATOMY ISSUES, THEY'RE GENERALLY AT AN INCREASED RISK FOR MENINGITIS.

COCHLEAR IMPLANTS HAVE AN INCREASED RISK FOR MENINGITIS.

IT'S SMALL, THERE ARE CERTAIN VACCINATIONS THEY NEED TO HAVE TO TRY TO PREVENT MENINGITIS.

THERE CAN BE PROBLEMS WITH  
GETTING A WIRE INSERTED  
PROPERLY.

SO WE GO TO THE ORGAN AND I  
CAN'T GET IT IN ALL THE WAY,  
WHEN IT DOES, THAT MEANS  
THEY'RE NOT GOING TO PERFORM  
WELL.

SO IT'S FRUSTRATING FOR BOTH  
THE FAMILY AND MYSELF.

BUT THAT RARELY HAPPENS.

IF WE DO AN IMPLANT AND  
THAT DOESN' T WORK, CAN WE GO  
BACK TO HEARING AIDS?

THE ANSWER IS YES.

IF THE SURGERY DOESN' T WORK,  
THEY TECHNICALLY COULD USE  
THEIR HEARING AIDS.

I CAN' T GUARANTEE IT, WE  
CERTAINLY STRIVE TO DO IT,  
BUT IT DEPENDS ON HOW  
DIFFICULT THE SURGERY IS.

SURGERY TAKES 3 TO 4 HOURS  
PER SIDE.

THERE'S AN INCISION BEHIND  
THE EAR, IT'S 2 INCHES BEHIND  
THE EAR AND ABOUT 3 INCHES  
LONG.

IT SITS IN THIS AREA RIGHT  
HERE IS THE INCISION LINE,  
IT'S A C INCISION IN THAT  
AREA.

IN A BOY, THEY WERE REALLY  
NOTICEABLE AND UGLY FROM A  
FEW YEARS AGO, BUT WE'VE  
CORRECTED THAT AND THEY'RE  
MUCH BETTER NOW.

I THINK THAT'S – WE'LL  
SKIP THAT.

I TALKED ABOUT QUALITIES  
ALREADY.

I WANT TO TALK BRIEFLY ABOUT  
BAHAS, WHICH STANDS FOR BONE  
ANCHORED HEARING AIDS AND  
WE'RE FINDING USEFULNESS FOR  
THIS.

THIS USED TO BE FOR PEOPLE  
WHO COULDN'T HAVE HEARING  
AIDS BECAUSE OF INFECTION.

WE'RE FINDING OUT BAHA AIDS  
CAN BE HELPFUL.

WHERE THIS IS COMING IN PLAY  
IS WITH CHILDREN WHO ARE IN  
SCHOOL WHO HAVE A ONE-SIDED  
DEAFNESS OR ONE-SIDED HEARING  
LOSS AND WE KNOW THAT THOSE  
CHILDREN, A LARGE PERCENT OF  
THEM DO POORLY IN SCHOOL.  
THERE AREN'T GREAT WAYS TO  
HELP THOSE KIDS OUT.

FM SYSTEMS, WHERE THERE'S A  
SPEAKER AT THE DESK, IT'S  
HELPFUL, BUT ONCE THEY GET  
INTO JUNIOR HIGH AND HIGH

SCHOOL, THEY TEND NOT TO HAVE FM SYSTEMS.

THEY HAVE PROBLEMS GETTING FM SYSTEMS.

BY PUTTING THESE SURGICAL IMPLANTS IN, WE'RE ABLE TO GET THEM HEARING FOR ONE-SIDED LOSS.

AND WE'RE FINDING IT CAN BE QUITE HELPFUL FOR THEM.

AGAIN, IT'S A SURGERY, I CALL IS THE FRANKENSTEIN SURGERY.

WE DRILL A HOLE IN THE SKULL  
RIGHT HERE AND THE INCISIONS  
CAN BE PRETTY UGLY.

OFTEN IT RESULTS IN A BALD  
SPOT.

WE' RE GETTING BETTER.

WE' RE DOING BETTER.

IT' S NOT NOTI CEABLE IN A  
WOMAN WI TH LONG HAI R, BUT  
IT' S NOTI CEABLE WI TH A MAN  
WI TH SHORT HAI R.

THERE' S A LOT OF MAI NTENANCE  
AND CARE.

YOU HAVE TO KEEP THEM CLEAN,  
THEY CAN EASILY GET INFECTED.  
THAT'S KIND OF WHAT IT LOOKS  
LIKE.

THERE'S A SCREW THAT ALWAYS  
HANGS OUT UNDER THE SKIN.

AND WHEN YOU'RE WEARING THE  
DEVICE, IT'S EVEN MORE  
NOTICEABLE.

THE DEVICE IS A BLACK BOX  
THAT SNAPS IN.

YOU HAVE THIS POST PUT IN.

ONCE THE POST IS HEALED SO

THE BONE GROWS INTO THE SCREW

AND HOLDS IT IN, YOU COME  
BACK WITH A SECOND SURGERY TO  
PUT IN THE DEVICE.

IT'S A TWO-STAGE SURGERY FOR  
THIS.

AND I THINK THAT'S PRETTY  
MUCH THE END OF MY TALK.

SHOULD I TAKE QUESTIONS AT  
THIS POINT?

>> WE HAVE A FEW E-  
MAILED QUESTIONS.

THE FIRST IS WHAT ABOUT  
CHILDREN THAT HAVE PROFOUND  
LOSS THAT GET A SIGNIFICANT

GAIN WITH HEARING AIDS, WOULD THEY STILL BE A CANDIDATE?

>> DR. RIMMELL: NO, NOT IF THEY'RE GETTING A GOOD RESULT.

OUR VIEW AS A TEAM, AND WE DISCUSS THESE PATIENTS AS A TEAM.

IF THE CHILD – I'LL ASSUME GAIN IS IMPROVEMENT OF VOCABULARY, SPEECH OR SIGNS THAT THE CHILD IS RESPONDING TO HEARING AIDS, WE WOULD

NEVER IMPLANT A PATIENT  
THAT'S RESPONDING TO HEARING.  
WE WOULD MONITOR THE PATIENT  
PRETTY CLOSELY IF THEY HAVE A  
SEVERE LOSS.

BUT IF THEY'RE – HEARING AIDS  
IS ALWAYS THE FIRST CHOICE  
FOR US IF THEY'RE RESPONDING  
TO IT.

>> AUDIENCE MEMBER:

THAT'S MY QUESTION, HOW DO  
YOU KNOW WITH – HOW CAN YOU  
FIGURE OUT AND KNOW WHEN  
YOU'RE LOOKING AT CHILDREN,

LET' S SAY THEY' RE AGE 0 TO 2,  
I KNOW THE EARLIER THE BETTER  
WITH COCHLEAR IMPLANTS, HOW  
DO YOU KNOW THAT HEARING AIDS  
ARE SUCCESSFUL IN THE WAY YOU  
WANT THEM TO BE SO YOU MAKE  
SURE THEY HAVE THE PROPER  
THING FOR THEIR HEARING LOSS?  
' CAUSE YOU NEED TO WORK WITH  
ALL THESE DIFFERENT PEOPLE,  
THE PSYCHOLOGIST, THE SPEECH  
TEACHERS, EVERYONE, AND I' M  
WONDERING HOW YOU MAKE SURE

TO HAVE THE PROPER FIT FOR  
THAT CHILD?

' CAUSE THEY' RE SO LITTLE WHEN  
THEY' RE UNDER 2 AND IT' S HARD  
TO GET ALL THE – MAYBE ALL  
THE CORRECT SIGNS TO KNOW  
EXACTLY WHAT THEY' RE GETTING  
OR NOT GETTING.

>> DR. RIMELL: RIGHT.

IT' S A SERIES OF MULTIPLE  
TESTS.

IF A CHILD HAS MULTIPLE TESTS  
AND SHOWING RESPONSE AT 90 TO  
100 DECIBELS IN BOTH EARS,

WE' RE CONFIDENT THEY' RE NOT  
GOING TO DEVELOP SPEECH AND  
LANGUAGE ORALLY WITHOUT  
COCHLEAR IMPLANTS.

BEHAVIORALLY, THEY' RE NOT  
RESPONDING TO SOUNDS AND  
THEY' RE SHOWING NO RESPONSE  
TILL 90 OR 100 DECIBELS IN  
BOTH EARS, WE CAN BE  
CONFIDENT THAT THAT CHILD IS  
NOT GOING TO BENEFIT FROM A  
HEARING AID.

IF WE WAIT UNTIL 2 YEARS WITH COCHLEAR IMPLANT AS IF THEY GOT IT AT AGE 1.

THAT'S HOW WE ASSESS IT.

OBVIOUSLY IF A CHILD IS COMING IN AT 70 TO 80, WE'RE GOING TO WAIT.

WE'RE GOING TO WAIT AND SEE THAT CHILD.

SOME KIDS ARE VERY TOUGH TO TELL, AND WE ERR ON THE SIDE OF WAITING AND SEE WHAT HAPPENS OVER TIME.

>> AUDIENCE MEMBER: I  
READ AN ARTICLE A FEW YEARS  
AGATHA WAS ENCOURAGING – FROM  
THE FDA THAT WAS TALKING  
ABOUT – IT WAS ENCOURAGING TO  
GET THE COCHLEAR IMPLANTS  
ABOUT – SO A FEW YEARS AGO I  
WAS READING AN ARTICLE, I'M  
NOT SURE WHICH ONE THAT WAS  
FROM – AND THE FDA WAS TRYING  
TO CHANGE – I'M NOT SURE IF  
IT WAS 90 AND ABOVE – FOR  
TRYING TO PRESSURE THE FDA  
TRYING TO PUT IT DOWN TO 60,

AND I ASKED ONE OF MY FRIENDS  
WHO WAS HARD OF HEARING AND  
THEY SAID THEY WERE ABOUT 70  
OR 80 AND THEY SAID THEY  
COULD HEAR SOUND WITH THE  
HEARING AID AND THEY COULD  
TALK ON THE PHONE AND  
UNDERSTAND, SO I'M WONDERING  
WHY THE – WHY THEY WERE  
TRYING TO PRESSURE THEM TO  
PUT THAT TO 60, DO YOU KNOW  
WHY THAT IS?

>> DR. RIMMELL: THAT  
WOULD BE FOR ADULTS ONLY AND  
NOT CHILDREN.

AT LEAST NOT INFANTS AND  
TODDLERS.

>> AUDIENCE MEMBER:  
THERE WASN'T A SPECIFIC AGE  
GROUP AND JUST IN GENERAL.

>> DR. RIMMELL: JUST  
FOR ADULTS WHEN ADULTS ARE  
NOT ABLE TO GET INFORMATION  
FROM HEARING AIDS.  
AGAIN, IT'S ABOUT SPEECH AND  
NOT ABOUT JUST THE AUDIOGRAM.

SO THERE ARE ADULTS WHO MAY  
HAVE AUDIOGRAMS AT 60, 70  
DECIBELS WHO GET VERY POOR  
SPEECH UNDERSTANDING, SO  
THERE MAY BE ADULTS THAT HAVE  
60 AND 70 AND THEY HAVE A  
WORD LIST OF ONLY ABOUT 10 OR  
20% OF WORDS.

THERE ALREADY ADULTS WHO GET  
80 OR 90% OF WORDS, THEY' RE  
NOT GOT TO GET AN IMPLANT.

A 1 OR 2-YEAR-OLD WILL NEVER  
GET IMPLANTED WITH A 60 OR 70  
DECIBEL RANGE NEVER.

>> AUDIENCE MEMBER:  
NEVER?

>> DR. RIMELL: AS THEY  
GET OLDER AND WE SEE A STOP  
IN PROGRESSION AND THEY'RE  
ABLE TO GIVE US WORD  
RECOGNITION SCORES, THAT WILL  
CHANGE POSSIBLY, BUT IT  
DEPENDS ON THE INDIVIDUAL  
PATIENT.

>> AUDIENCE MEMBER:  
OK.  
I UNDERSTAND THAT.  
THANK YOU.

>> WE HAVE A COUPLE  
OTHER QUESTIONS.  
ONE, HAVE THERE BEEN ALLERGIC  
REACTIONS TO THE DEVICE?

>> DR. RIMMEL: THERE  
HAVE BEEN NO ALLERGIC  
REACTIONS TO THE DEVICE, BUT  
REACTIONS FROM SURGERY.  
I'VE NEVER HEARD OF THAT  
EVER.

>> THE LAST QUESTION  
IS FOR YOU.  
ARE YOU ABLE TO GENERALIZE  
WHAT PORTION OF PATIENTS WHO

MI GHT BE AUDI OLOGI C  
CANDI DATES FOR COCHLEAR  
I MPLANTS MI GHT NOT GET THEM  
DUE TO ANATOMI CAL OR OTHER  
PHYSI CAL REASONS?

>> DR. RI MELL: NOT  
ONLY THAT, BUT NEUROLOGI C  
REASONS WOULD BE THE THI RD  
ONE.

LET' S TALK ABOUT EACH ONE  
REAL QUI CKLY.

THE ANATOMI C CONDI TI ONS ARE  
PRETTY RARE.

EVERY ONCE IN A WHILE YOU GET  
A SEVERE ONE WHERE THERE' S NO  
COCHLEA, THAT' S VERY RARE.  
THEY CAN' T HAVE AN IMPLANT.  
HOWEVER, WE HAVE NOW WHAT' S  
CALLED DIRECT BRAIN STEM  
IMPLANTS AND LAY AN IMPLANT  
IN THERE FOR THOSE CHILDREN.  
SO THAT' S – NOW DAYS AS TIME  
GOES ON, THERE' S SOME OPTION  
TO DO WITH ANY ANATOMI C  
PROBLEM.

WHAT WAS THE OTHER ONE?

>> PHYSICAL,  
ANATOMICAL.

>> DR. RIMMEL:  
NEUROLOGIC IS AN ISSUE THAT I  
ACTUALLY SAW A PATIENT TODAY  
WHO WAS SEVERELY  
NEUROLOGICALLY DEVASTATED, HE  
DOESN'T WEAR AN IMPLANT,  
THOSE ARE THE KIND OF  
PATIENTS WE WANT TO AVOID.  
BUT I CAN'T PREDICT THAT.  
SOME CHILDREN HAVE SEVERE  
CEREBRAL PALSY BUT IT'S  
MUSIC.

THEY MAY NOT SPEAK AND MAY NOT DO MUCH, BUT THEY LOVE HEARING THE SOUNDS THEY GET OUT OF IT.

WHEN WE HAVE A CHILD WHO'S NEUROLOGICALLY DEVASTATED BECAUSE OF A BRAIN INJURY OR WHATEVER ELSE IS GOING ON, WE HAVE A HARD DISCUSSION WITH THE PARENTS SAYING THIS MAY NOT DO ANYTHING AT ALL, OR IT MAY BE HELPFUL, EVEN THOUGH THEY DON'T SPEAK, THAT THEY ENJOY WEARING IT, BECAUSE

WHATEVER INFORMATION OR  
STIMULUS IT GRANTS.

FOR THOSE PEOPLE WHERE  
THEY'RE GEARED FOR HEARING,  
HUMAN VOICE AND MUSIC CAN BE  
A POSITIVE STIMULUS MUCH LIKE  
FOOD CAN BE.

SO THERE ARE – AGAIN, WHEN  
IT'S A NEUROLOGIC ISSUE, IT'S  
A HARDER ISSUE.

IT'S HARD TO PREDICT THOSE  
ISSUES SOMETIMES.

>> AUDIENCE MEMBER: I WAS CURIOUS, I'VE HEARD MIXED AGES LIKE WITH CERTAIN THINGS LIKE CRANIAL ATRESIA, ARE THERE GUIDELINES WHEN YOU DO SURGERY, EVEN FOR THE BAHAS OR THE COCHLEAR IMPLANT?

>> COULD YOU BRIEFLY REPEAT WHAT SHE SAID?

>> DR. RIMMEL: THE QUESTION WAS ARE THERE GUIDELINES PARTICULARLY WITH REGARDS TO BAHAS AND COCHLEAR IMPLANTS.

YOU HAVE TO HAVE A THICK  
ENOUGH SKULL WITH BONE  
ANCHORED HEARING AIDS.

MOST OF US LIKE TO WAIT UNTIL  
4 OR 5 YEARS OF AGE WHERE THE  
SKULL IS THICK ENOUGH FOR THE  
SCREW.

THE COCHLEAR IMPLANT, THE FDA  
HAS A LIMIT OF 12 MONTHS OF  
AGE.

MOST CENTERS ARE DOING IT  
YOUNGER, FOR INSTANCE IF BOTH  
EARS ARE 100 DECIBELS.

TECHNICALLY IT CAN BE DONE AT SIX MONTHS OF AGE QUITE EASILY WITH NO RISKS.

MOST CENTERS ARE USED TO DOING THEM.

THE PARENTS ARE SURE THERE'S 100 DECIBEL ON BOTH SIDE OR SEVERE-PROFOUND HEARING LOSS ON BOTH SIDES WITH TESTS,

WE'LL DISCUSS FDA APPROVAL AT 12 MONTHS AND LEAVE THE DECISION WITH THE FAMILY.

>> AUDIENCE MEMBER:

WHAT ABOUT WITH OTHER SORTS

OF ANATOMICAL THINGS LIKE EAR  
FLAPS, ATRESIA?

I'VE HEARD MIXED THINGS.

IF YOU'RE WONDERING IF THE  
EAR CANAL IS PRESENT TO HAVE  
THAT FAMILY.

THEY WERE GOING TO GO IN AT  
LIKE SIX MONTHS BUT THEN I  
HEARD SOME SURGEONS WAIT  
UNTIL 2.

I DIDN'T KNOW IF THERE WAS  
THINGS --

>> DR. RIMMELL: THE  
QUESTION WAS DEALING WITH

ATRESIA, WHERE THERE' S NO  
EXTERNAL EAR CANAL OR IF  
THERE' S QUESTION IF IT  
EXISTS.

THE EXTERNAL EAR CANAL  
PROBLEMS ARE SENSORINEURAL  
HEARING LOSS AND ONE-SIDED,  
NOT BOTH SIDED.

WHEN YOU' RE BORN WITHOUT AN  
OUTER EAR CANAL.

NOTHING SHOULD BE DONE AS  
LONG AS THE OTHER EAR IS  
HEARING NORMALLY.

BUT IF BOTH EARS ARE HAVING PROBLEMS HEARING, WE CAN DO A BONE-ANCHORED HEARING AID.

WE WOULD NEVER OPERATE TO MAKE AN EAR CANAL BEFORE THE AGE OF 5 OR 6.

I KNOW THERE'S A CHILD IN SOUTH DAKOTA THAT HAD A SURGERY ATTEMPT, I JUST SAW HIM IN THE OFFICE AT 2 YEARS OF AGE, NO MAJOR CENTER WOULD DO THAT.

>> I DO HAVE ONE MORE QUESTION.

DO YOU HAVE STATISTICS SHOWING THAT CHILDREN WHO GET IMPLANTS AND THEIR CARING AFTER THE IMPLANT WHAT ARE THE PERCENTAGE OF RANGES?

>> DR. RIMELL: THE QUESTION WAS DO WE HAVE STATISTICS ON – I GUESS THEY WANT TO KNOW GETTING UP TO WHERE WE WOULD CONSIDER NORMAL HEARING RANGE OF 20 DECIBELS, I'M ASSUMING THAT'S

– AGAIN, THERE'S A LOT OF ISSUES THAT GO INTO IT.

IF THE CHILD HAS NO OTHER MEDICAL PROBLEMS, IF THERE'S A GOOD FAMILY THAT SHOWS UP FOR APPOINTMENTS AND GOES TO PROGRAMMING AND SPEECH AND ALL THE OTHER ISSUES, THE CHANCE IS VERY HIGH.

IF I DO A PERFECT SURGERY AND THE CHILD IS NORMAL AND THE PARENTS DON'T SHOW UP FOR APPOINTMENTS, THEY'RE NOT GOING TO DO WELL.

IF THERE'S NO OTHER MEDICAL PROBLEMS, IF THE PARENTS ARE GOOD PARENTS AND SPEND TIME IN WORKING WITH THE CHILD, THEY GO TO THEIR APPOINTMENTS, THE CHANCE IS 90%, 85% THAT THEY'RE GOING TO HAVE RELATIVELY NORMAL HEARING.

>> DR. FRIEDMAN:  
SPEECH WILL BE AUDIBLE.  
THAT'S THE QUESTION.

>> DR. RIMMELL: THAT'S  
WHAT MOST CENTERS ARE SHOWING

NOW DAYS WITH MODERN  
IMPLANTS.

>> AUDIENCE MEMBER:  
WHAT KIND OF NUMBERS, WHAT  
KIND OF STATISTIC IS THAT  
FROM?

>> DR. RIMELL: THAT'S  
FROM MULTIPLE CENTERS AND  
CLINICAL TRIALS.  
THESE DEVICES HAVE BEEN  
AROUND FOR 20 YEARS.

>> AUDIENCE MEMBER:  
800 SURGERIES HAVE BEEN DONE  
HERE.

WHAT ARE THE STATISTICS?

DO YOU HAVE STATISTICS FOR  
YOUR ACTUAL SURGERIES YOU'VE  
DONE HERE, OR IS THAT THE  
GENERAL?

>> DR. RIMMELL: IF HE  
WANTS TO E-MAIL ME, I'LL  
SUPPLY HIM WITH SOME  
ARTICLES.

>> AUDIENCE MEMBER:  
OK.

>> DR. RIMMELL: BUT  
THIS DATA IS PUBLISHED.

>> AUDIENCE MEMBER: IS THAT PUBLIC KNOWLEDGE? IT SEEMS LIKE SOMETHING THAT SHOULD BE MORE PUBLIC KNOWLEDGE.

' CAUSE THAT'S A NUMBER THAT SHOULD BE – THAT SEEMS LIKE YOU COULD BE PROUD OF.

>> DR. RIMELL: IT IS. AND THERE ARE MULTIPLE LARGE CENTERS IN THE UNITED STATES THAT PUBLISH DATA ON THEIR OUTCOMES AND FOLLOW-UP FOR CHILDREN.

LIKE I SAID, IF HE WANTS TO  
E-MAIL ME, I WILL E-MAIL HIM  
SOME ARTICLES.

>> AUDIENCE MEMBER:

OK.

THANK YOU.

[APPLAUSE]

>> THANK YOU VERY

MUCH, DOCTOR RIMMEL.

NEXT WE HAVE DOCTOR SCHLEISS

FROM INFECTIOUS DISEASE.

>> DR. SCHLEISS:

THANKS, MELISSA.

IT'S A PLEASURE TO BE ABLE TO  
SPEAK TO EVERYONE THIS  
EVENING.

I WANT TO THANK THE MINNESOTA  
HANDS & VOICES AND ALSO THE  
LIONS CLUBS OF MINNESOTA FOR  
THE OPPORTUNITY TO SHARE SOME  
INFORMATION WITH YOU.

#### MY TAKE-HOME MESSAGE

TONIGHT IS GOING TO BE THAT  
THE MAJORITY OF IDENTIFIABLE  
CAUSES OF HEARING LOSS,  
OUTSIDE OF GENETIC CAUSES

THAT YOU' LL HEAR ABOUT IN A FEW MINUTES, ARE ACTUALLY DO TO ONE INFECTION, CMV.

THAT' S THE COMMON INFECTION THAT NO ONE HAS EVER HEARD OF.

ONE THING I' D LIKE TO SEE HAPPEN, IT' S BEEN HAPPENING OVER THE LAST SEVERAL YEARS AS WE' VE ALL BEEN WORKING TOGETHER IN THIS IS TO GET THE WORD OUT, TO MAKE WOMEN AWARE OF THIS, TO MAKE HEALTH

CARE PROVIDERS AWARE OF IT,  
AND ALSO TO EDUCATE  
INDIVIDUALS WHO ARE INVOLVED  
IN THE CARE OF THESE CHILDREN  
THAT WE HAVE TREATMENT  
AVAILABLE AND HOPEFULLY WE'LL  
SEE A VACCINE ON THE HORIZON  
THAT WILL SOLVE THIS PROBLEM.

THIS IS A SLIDE SHOWING A  
CYTOMEGLAOVIRUS CELL.  
THESE ARE EYE INCLUSIONS,  
BECAUSE IT LOOKS LIKE A BARN-  
YARD OWL.

THESE WERE FOUND UNDER A  
MICROSCOPE 100 YEARS AGO, BUT  
IT WASN'T UNTIL THE 1940S  
WHEN THE CELL WAS DISCOVERED.  
IT WAS NAMED CMV,  
CYTOMEGLAVIRUS, MEANS IT'S  
THE CELL THAT MAKES IT LARGE.  
YOU CAN APPRECIATE THIS IS A  
VERY LARGE, SWOLLEN CELL.  
THIS IS A CARTOON SHOWING  
WHAT THE SLIDE IS ITSELF.  
THE INTERESTING THING ABOUT  
CMV, IT IS THE LARGEST VIRUS  
THAT INFECTS HUMAN BEINGS.

THE LARGEST VIRUS NOT ONLY IN THE SIZE OF THE PARTICLE ITSELF, BUT ALSO THE SIZE OF THE G NOME, THE GENETIC CODE THAT THE VIRUS USES TO REPLICATE ITSELF AND CAN ACTUALLY DAMAGE THE DEVELOPING BABY IN THE WOMB.

IF A PREGNANT WOMAN ENCOUNTERS THIS VIRUS, THEN THERE'S A SUBSTANTIAL RISK THAT THAT BABY WILL BE INFECTED IN THE WOMB AND ONCE

BORN MAYBE HAVE PERMANENT  
INJURY, THE MOST COMMON OF  
WHICH IS HEARING LOSS.

IN OTHER ERA, THE MOST  
COMMON INFECTIOUS DISEASE  
THAT CAUSED HEARING LOSS IN  
BABIES WAS ANOTHER VIRUS  
CAUSED RUBELLA.

IT CAUSED THE GERMAN MEASLES.  
THE GERMAN MEASLES WAS A VERY  
BENIGN TRIVIAL ILLNESS IN  
CHILDREN, THEY WOULD GET THE  
GERMAN MEASLES, IN CONTRAST

TO THE HARD MEASLES, IT WAS  
VERY MILD, RUNNY NOSE AND  
VERY MILD RASH.

MOST KIDS WEREN' T EVEN VERY  
ILL.

THE ONLY REASON WAS THAT  
RUBELLA WAS A PROBLEM, THEY  
WERE AROUND WOMEN WHO WERE  
GETTING PREGNANT.

IF THE WOMAN GOT THE GERMAN  
MEASLES, THAT BABY WOULD BE  
AT HIGH RISK FOR BRAIN  
INJURY, CATARACTS AND HEARING  
LOSS.

WE SOLVED THIS INFECTION BY  
VACCINATION AND THE GOAL WILL  
BE TO DEVELOP A VACCINE FOR  
CMV AND ERADICATE THE  
INFECTION TO BABIES FROM  
THAT.

AFTER RUBELLA WAS  
VACCINATED, THE FIRST DEAF  
MISS AMERICA ACTUALLY HAD  
MENINGITIS WHEN SHE WAS A  
LITTLE GIRL FROM THAT  
BACTERIA AND THAT IS WHY SHE  
HAD HEARING LOSS.

AND WE NOW HAVE A VACCINE TO PREVENT THAT INFECTION, AS WELL.

CMV CAN CAUSE MONO, SO ABOUT THREE-QUARTERS OF TEENAGERS GET IT FROM KISSING EACH OTHER IS DUE TO A VIRUS, BUT OTHERS IS FROM THE CYTOMEGLAOVIRUS.

WE SEE THIS IN TRANSPLANT PATIENTS, CANCER PATIENTS AND HIV-INFECTED PATIENTS.

IT'S CONGENITAL CMV THAT'S  
THE BIG CONCERN.

CONGENITAL CMV, MEANING CMV  
THAT'S TRANSMITTED TO THE  
BABY WHILE THE BABY IS STILL  
DEVELOPING IN THE WOMB.

UP TO 2% OF ALL NEWBORNS HAVE  
THIS.

ALL OF THE THINGS THAT WE  
SCREEN FOR, CMV IS A VIRUS  
THAT IS INCREDIBLY COMMON.

CMV CAN INFECT THESE BABIES  
AND THEY LOOK ASYMPTOMATIC,

AND THEY LOOK NORMAL IN THE NURSERY.

30 TO 40% OF ALL NONSYNDROMIC DEAFNESS IN NEWBORNS IS DUE TO CMV.

CMV IS GENERALLY BILATERAL, IN OTHER WORDS, BILATERAL DEAFNESS IS THE RULE, NOT THE EXCEPTION, AND IT CAN BE PROGRESSIVE IN NATURE.

THEY MAY HAVE MILD LOSS AND PROGRESS IN THE YOUNGER YEARS OF LIFE TO MAJOR DEAFNESS.

AND IT BECOMES VERY IMPORTANT  
ISSUE TO MONITOR FOR IN THE  
CLINIC.

THERE ARE 4 MILLION BABIES  
BORN IN THE UNITED STATES  
EVERY YEAR, IF WE ESTIMATE  
CONSERVATIVELY THAT 1 OUT OF  
100 BABIES THERE ARE ABOUT  
40,000 BABIES INFECTED WITH  
CMV.

AND ALTHOUGH MOST OF THESE  
BABIES LOOK NORMAL AT BIRTH  
AND ARE ASYMPTOMATIC, UP TO

10 OR 15% OF THESE BABIES MAY GO ONTO DEVELOP HEARING LOSS. THAT'S SOMEWHERE AROUND 4 TO 6,000 BABIES A YEAR WHO HAVE DISABILITIES RELATED TO CMV.

CONGENITAL CMV CAN OCCUR IN YOUNG WOMEN, IT'S VERY COMMON ALSO IN WOMEN WHO HAVE INCREASED EXPOSURE OPPORTUNITIES, WOMEN IN GROUP DAYCARE CENTERS OR MOTHERS WHO HAVE TODDLER ATTENDING

GROUP DAYCARE, PARTICULARLY  
CHILDREN UNDER THE AGE OF 3.  
AT THE AGE OF 3, MOST  
CHILDREN BECOME TOILET  
TRAINED AND THAT'S THE BREAK  
POINT.

PRIOR TO BEING TOILET  
TRAINED, THEY CAN TRANSMIT  
THAT TO MOTHERS THROUGH  
DIAPER CHANGES.

THERE AREN'T ANY STATE  
REGULATIONS FOR  
CYTOMEGLAOVIRUS.

ONE OF THE THINGS I'VE BEEN WORKING ON WITH MY COLLEAGUES AT THE LIONS CLINIC AND THE DEPARTMENT OF HEALTH IS TO PUSH FORWARD SCREENING ALL NEWBORNS FOR CMV.

THESE ARE SOME DRAMATIC PICTURES WITH BABIES WITH SEVERE CMV.

DOCTOR RIMMELL AND SCHIMMENTI AND I TALK ABOUT IN THE CLINIC, WHEN BABIES SHOULD HAVE THEIR BRAIN CAT SCANNED AND THEIR EAR AND MIDDLE EAR.

THESE ARE A COUPLE OF OTHER  
INFANTS WITH CMV INFECTION AT  
BIRTH.

YOU MIGHT APPRECIATE THIS  
BABY IS COVERED WITH A RASH.  
THESE ARE SMALL DISCREET DARK  
SPOTS.

YOU CAN ACTUALLY FEEL THESE  
BULGING OUT OF THE SKIN.

IT LOOKS LIKE A BLUEBERRY  
MUFFIN RASH.

THESE BABY CAN BE SICK ALL  
OVER, THEY CAN HAVE HEPATITIS  
AND PNEUMONIA.

THE REST OF THESE ARE SELF-  
RESOLVING OR RAPIDLY CLEARED  
WITH ANTI VIRAL DRUGS.

THIS IS A CAT SCAN OF THE  
BRAIN.

WHAT YOU CAN APPRECIATE HERE  
IS THE DARKNESS.

THAT'S FLUID, THAT'S NOT  
NORMAL.

THIS IS A BABY WHO HAS VERY,  
VERY LARGE VENTRICLES.

THIS IS A DETAILED SLIDE  
AND THE REFERENCE IS ON THERE

FOR THOSE INTERESTED IN  
LEARNING MORE ABOUT IT.

THIS IS A COHORT OF ABOUT 50  
BABIES THAT WERE FOLLOWED  
OVER TIME TO ASK WHEN DID  
THEIR HEARING LOSS BECOME A  
PROBLEM, WHEN COULD IT BE  
DEMONSTRATED BY THE  
AUDI OLOGI ST.

THESE WERE ALL TESTED TO HAVE  
CMV.

THE REST OF THESE BABIES  
DEVELOPED HEARI NG LOSS

GRADUALLY OVER THE FIRST FEW YEARS OF LIFE.

AND SO WHEN WE SCREEN

NEWBORNS WITH THE NEWBORN

HEARING SCREENING TOOLS THAT

WE HAVE TODAY, WE NEED TO

CONTINUE TO DO THAT, IT'S

ESSENTIAL, WE HAVE TO

RECOGNIZE THAT WON'T CAPTURE

ALL THE BABIES THAT HAVE CMV

HEARING LOSS.

MOST BABIES DESTINED TO HAVE

HEARING LOSS DUE TO CMV WILL

ACTUALLY HAVE A NORMAL  
NEWBORN SCREENING.

THE ANSWER WOULD BE TO SCREEN  
NEWBORN BABIES NOT ONLY FOR  
HEARING, BUT ALSO FOR THE  
VIRAL INFECTION.

I'LL SAY MORE ABOUT THAT IN A  
MOMENT.

THERE'S A TREATMENT FOR CMV,  
AND THIS IS AN IMPORTANT  
MESSAGE TO GET OUT THERE.

I THINK A LOT OF AUDIOLOGISTS  
IN THE COMMUNITY, EVEN A LOT  
OF PEDIATRICIANS AND FAMILY

PRACTITIONERS ARE UNAWARE OF THE STUDIES THAT SHOW THE ANTIBIOTIC THAT ACTUALLY ATTACKS THE CMV VIRUS CAN BE USEFUL IN PREVENTING HEARING LOSS IN BABIES WITH CMV.

THIS WAS A STUDY PUBLISHED A FEW YEARS AGO, IT DID TESTING OF BABIES WITH CMV INFECTION.

IT SHOWED BABIES THAT GOT THE DRUG GANCICLOVIR AND WERE

LESS LIKELY TO DEVELOP  
HEARING LOSS.

THIS IS NOT A INTEREST DRUG,  
AND MOST INFANTS THAT HAVE  
HEARING LOSS DUE TO CMV ARE  
GOING TO CONTINUE TO HAVE  
PROBLEMS, EVEN THEY ARE  
TREATED WITH MEDICATION.

BUT WHAT WE'RE TRYING TO DO  
WITH THIS IS GET THE BABY TO  
THE POINT WHERE THEY'RE BIG  
ENOUGH FOR DOCTOR RIMMEL TO  
PUT IN COCHLEAR IMPLANT, BUY  
SOME TIME TO WE CAN CORRECT

THAT BABY' S HEARING WITH  
HEARING AIDS, PREVENT IT FROM  
DETERIORATING TO COMPLETE  
DEAFNESS AND THAT' S THE POINT  
OF ANTI VIRAL MEDICATION.

IT' S NOT GOING TO CURE IT,  
BUT IT' S GOING TO DELAY AND  
IN SOME BABIES LEAD TO SOME  
IMPROVEMENT.

I' VE ARGUED AND PUBLISHED  
AND TESTIFIED IN ST. PAUL  
THAT WE NEED TO DEVELOP  
SCREENING PROGRAMS FOR CMV,

AND I'D LOVE TO SEE MINNESOTA  
TO BE THE FIRST TO DO THIS.

WE'RE WAY BEHIND OUR EUROPEAN  
COLLEAGUES, FRANCE, NORTHERN  
ITALY WHERE CMV SCREENING HAS  
BECOME ROUTINE.

I BELIEVE WE SHOULD INTEGRATE  
NEWBORN SCREENING WITH  
FUNCTIONAL HEARING SCREENING  
AND CHILDREN WITH UNEXPLAINED  
HEARING LOSS WHO HAD THEIR  
GENETIC EVALUATION, THEIR CAT  
SCAN, THEY SHOULD BE  
EVALUATED FOR THE POSSIBILITY

OF CMV INFECTION AND OFFERED  
THIS MEDICATION TO BABIES WHO  
HAVE HEARING LOSS.

THE PROBLEM IS WHO TO  
SCREEN.

THERE WAS A PERSON WHOSE WIFE  
GAVE BIRTH TO A BABY WHO  
FAILED THE SCREENING.

THE FAMILY WAS TOLD THIS  
MIGHT BE A FALSE FAILURE,  
LET'S JUST COME BACK IN A FEW  
WEEKS AND REASSESS IT.

IT WAS STILL ABNORMAL.

WELL, LET'S KEEP AN EYE ON  
IT.

AFTER THEIR 6-WEEK POST-  
PARTUM VISIT, THE  
PEDIATRICIAN THOUGHT IT COULD  
BE CMV.

THEY GOT A TEST ON THE WRONG  
PATIENT AT THE WRONG TIME.

WE CAN'T DIAGNOSE CMV BY  
TESTING WOMAN'S BLOOD.

WE CAN'T GET A VERY GOOD  
HANDLE ON WHETHER A BABY HAS  
CMV INFECTION OR NOT BY  
TESTING THE BABY'S BLOOD WITH

ANTIBODIES, BECAUSE THOSE MAY  
HAVE BEEN FROM THE MOTHER.

WE NEED TO LOOK FOR THE VIRUS  
ITSELF BY A SPECIAL TEST  
CALLED PCR TO LOOK FOR VIRAL  
DNA.

WHAT BETTER SOURCE TO GET  
THAT BLOOD THAN THE NEWBORN  
METABOLIC SCREEN.

ALL NEWBORN INFANTS HAVE  
THEIR HEEL PRICKED AND TESTED  
FOR DISORDERS.

MY LABORATORY HAS BEEN  
WORKING ON PCA ASSAYS.

WE' RE WORKING WITH THE  
COLLABORATION OF THE  
DEPARTMENT OF HEALTH.

ABOUT 65 TO 70,000 BABIES ARE  
BORN IN MINNESOTA EVERY YEAR.

1 TO 2% OF ALL BABIES IN  
MINNESOTA HAVE CMV, WE' RE  
TALKING ABOUT CHILDREN WITH  
CMV-ASSOCIATED HEARING LOSS.

WE ALREADY TALKED ABOUT  
BRINGING DOWN THAT MEDIAN  
AGE.

PROGRAMS NEED TO CONTINUE TO  
PUSH VERY AGGRESSIVELY.

BUT WE'VE BEEN DOING SOME  
PILOT PROJECTS TO ASK CAN WE  
FIND THESE DNA BLOOD SPOTS.  
WE COLLECTED BLOOD SPOTS FROM  
BABIES THAT FAILED THE TEST.  
WE THEN ASKED WHETHER OR NOT  
WE COULD EXTRACT THE VIRAL  
DNA FROM THESE BLOOD SPOTS,  
THE ANSWER WE COULD.  
THESE ARE EXAMPLES OF  
POSITIVE CONTROLS USING THIS  
PCR ASSAY.  
THESE ARE 3 REAL BABIES, BUT  
WE DO KNOW THEY FAILED THEIR

HEARING SCREEN AND ALL OF  
THESE BABIES HAD CMV IN THEIR  
NEWBORN BLOOD SPOTS.

WE RECENTLY PUBLISHED THIS  
AND OTHER COLLEAGUES HERE AT  
THE UNIVERSITY, AS WELL AS  
THE MINNESOTA DEPARTMENT OF  
HEALTH, AND WE FOUND ABOUT 3%  
OF BABIES THAT FAIL THAT  
NEWBORN SCREENING HAVE CMV IN  
THEIR BLOOD SPOT.

WHAT I'D LIKE TO SEE HAPPEN  
IS THESE STUDIES MOVE FORWARD  
AND DO A TRIAL IN WHICH WE

GET THE NAMES OF THESE BABIES  
AND FOLLOW THESE BABIES.

I'LL JUST TAKE A COUPLE OF  
MINUTES OF YOUR TIME AND JUST  
SAY A FEW WORDS ABOUT CMV  
VACCINE.

AGAIN, WHEN WE TALK ABOUT  
RUBELLA, INFLUENZA TYPE B,  
THE ULTIMATE WAY TO SOLVE  
THOSE PROBLEMS WAS NOT TO  
DEVELOP MORE TREATMENT, BUT  
TO PREVENT THEM ALTOGETHER  
WITH VACCINATION.

I WORKED IN MY LABORATORY  
WITH ANIMALS.

IT TURNS OUT GUINEA PIGS  
TRANSMIT THE VIRUS TO THE  
FETUS.

THE INFECTION OF THE PUP CAN  
BE PREVENTED BY VACCINATION.

WE'RE VERY INTERESTED IN  
LOOKING AT GENE PRODUCTS.

THESE ARE SOME NEWBORN GUINEA  
PIG PUPS WHO HAD CMV AND DIED  
QUICKLY AFTER BIRTH, THEY HAVE  
STUNTING OF GROWTH, HIGH

LEVELS OF VIRUS IN THEIR  
BLOOD.

YOU CAN PREVENT THIS BY  
VACCINATING THE FEMALE GUINEA  
PIG.

THIS IS A CONTROL ANIMAL ON  
THE LEFT SHOWING NORMAL  
HEALTHY PLACENTA AND THE  
OTHER SHOWING THE VIRUS.

IT GETS TO THE BABY THROUGH  
THE MATERNAL BLOOD, ACROSS  
THE PLACENTA TO THE BABY  
BLOOD.

WE ARE VERY INTERESTED IN  
PURSUING THE QUESTION OF  
WHETHER OR NOT WE CAN PREVENT  
INFECTION OF THE COCHLEA IN  
GUINEA PIGS, AND IF WE CAN,  
TO THEN APPLY THAT KNOWLEDGE  
TO THE HEALTH OF THE INFANTS.

THERE ARE OTHER NEW DRUGS  
ON THE HORIZON.

IN A NUMBER OF ANIMALS, WE  
EITHER INFECTED THEM WITH  
JUST ASSAULT WATER, OR  
INFECTED THEM WITH VIRUS.

AND THE ANIMALS THAT WEREN' T  
TREATED WITH ANTI VIRAL DRUGS,  
HAD A HIGH DEGREE OF HEARING  
LOSS OF AT LEAST 30 DECIBELS.  
THERE WAS NO HEARING LOSS  
WHATSOEVER IN THE ANIMALS  
TREATED WITH THE ANTI VIRAL  
DRUGS.

THESE ARE NORMAL AVR.

PANEL B IS AN ANIMAL INFECTED  
WITH CMV, THEN TREATED WITH  
THE ANTI VIRAL DRUG.

WE HOPE IF WE CAN IDENTIFY  
INFANTS WITH CMV EARLY AND

TREAT THEM AGGRESSIVELY WITH  
ANTI VIRAL DRUGS AND TAKE THIS  
ALONG TO HUMANS.

WE DON' T KNOW HOW CMV  
INJURES THE COCHLEA.  
WE THINK IT' S IN THIS  
COMPARTMENT OF THE COCHLEA.  
THIS IS A CROSS-SECTION OF  
THE COCHLEA, THE SCALA  
VESTIBULAR AND SCALA TYMPANI .  
BUT THE MAJOR WAY IN WHICH IT  
CAUSES HEARING LOSS IS  
INFLAMMATION THAT LEADS TO

SCLEROSIS AND SCARRING AND  
THAT'S THE MAJOR WAY IN WHICH  
CMV DOES THIS.

I'M OFTEN ASKED BY PARENTS IN  
THE CLINIC, IF MY BABY HAD  
CMV, WOULD THEY BE A  
CANDIDATE FOR COCHLEAR  
IMPLANT?

AND THE ANSWER IS YES.

THESE SHOW SOME OF THE  
INFLAMMATORY RESPONSES THAT  
YOU SEE IN THE COCHLEA.

AND THESE WHITE BLOOD CELLS  
COME IN AND ELABORATE THAT

THEN LEAD TO THIS DEGREE OF SCARRING AND INJURY.

AND THE GOAL IS TO PREVENT THIS BY EITHER VACCINATING THE MOTHER PRIOR TO PREGNANCY TO ELIMINATE THE INFECTION ALTOGETHER OR HAVE VACCINES.

THIS IS A LATE-STAGE FINDING, THERE'S A LOT OF VERY FIBROTIC TISSUE.

NORMALLY WE LIKE TO SEE NICE, PINK TISSUE, THIS IS JUST ALL SCARRED, BASICALLY.

AND THE SCAR LEADS TO A  
COMPONENT OF THE HEARING LOSS  
WE SEE WITH CMV.

SO IN ADDITION TO CLINICAL  
PROGRAMS AND NEWBORN  
SCREENING, WE'RE INTERESTED  
IN LOOKING AT MATERNAL  
VACCINATIONS, ANTI-VIRAL  
DRUGS.

THESE ARE SOME OF THE AVENUES  
OF EXPERIMENTATION IN THE  
LABORATORY.

I WANT TO LEAVE YOU WITH THE TAKE-HOME MESSAGE, THE MOST COMMON CAUSE OF HEARING LOSS IS DUE TO ONE INFECTION, CMV. WE DO HAVE TREATMENTS FOR CMV, WE NEED TO BE ABLE TO SCREEN BABIES AND IDENTIFY THIS EARLY TO OFFER INTERVENTION AND IT'S A VERY, VERY HIGH PRIORITY AREA FOR VACCINE DEVELOPMENT IN THE UNITED STATES TODAY TO DEVELOP VACCINES TO PREVENT THIS INFECTION.

ALTHOUGH, I SHOWED YOU SOME  
EXAMPLES OF VERY SEVERE BRAIN  
INJURY AND OTHER DAMAGE DUE  
TO CMV, MOSTLY THE MAJOR ONE  
IS HEARING LOSS.

WITH THAT, I'D BE GLAD TO  
TAKE QUESTIONS.

>> I WAS GOING TO ASK  
ABOUT THE BRAIN INJURY THAT  
YOU WERE SPEAKING IN THE  
LARGE VENTRICLES.

>> DR. SCHLEISS: 10 TO  
15%.

USUALLY THE CLUE IS THE  
BABY' S HEAD IS TOO SMALL,  
WHEN YOU MEASURE THE HEAD,  
THERE' S A DI SCONNECT.

BABY MAY HAVE A REASONABLE  
WEI GHT AND HEI GHT OR LENGTH,  
BUT THE HEAD IS TOO SMALL,  
BECAUSE THE BRAIN HAD BEEN  
I N F E C T E D I N T H E W O M B .

S O I T ' S U N C O M M O N , B U T M A Y B E  
10 OR 15% OF THE TIME WE  
W O R R Y A B O U T T H A T A N D I D O C T  
S C A N S O N T H O S E B A B Y ' S B R A I N S .

A LOT OF PARENTS COME IN AND  
THEY'RE REALLY, REALLY  
UNDERSTANDABLY CONCERNED  
BECAUSE THEY THINK THE BABY  
IS FINE, THE BABY HAS BEEN  
FOUND TO HAVE CMV FOR  
WHATEVER REASON, AND THEN  
THEY LEARN A LITTLE BIT MORE  
ABOUT IT AND FIND THAT CMV IS  
ASSOCIATED WITH MENTAL  
RETARDATION.

IT'S COMMONLY ASSOCIATED WITH  
CMV, SO IT CAN BE VERY, VERY  
SCARY NEWS.

MOST OF THE PROBLEMS BABIES  
HAVE ARE HEARING LOSS.

>> AUDIENCE MEMBER:

I'M CURIOUS, WITH CMV, WHEN A  
CHILD GETS TO BE ABOUT 5 OR  
6, ARE THERE ANY OTHER  
PROBLEMS WITH MAYBE PICKING  
UP LANGUAGE OR ARE THERE  
OTHER PROBLEMS THAT HAPPEN  
WITH THAT?

OR IS THAT WITH EITHER  
SIGNING OR SPEAKING, DO THOSE  
TEND TO DEVELOP IN A

DIFFERENT WAY IF A CHILD HAS  
HAD CMV?

>> DR. SCHLEISS: IS HE  
ASKING ABOUT THOSE CHILDREN  
WITH CMV THAT HAVE HEARING  
LOSS SPECIFICALLY?

>> AUDIENCE MEMBER:  
NO, JUST GENERALLY, WITH  
HEARING LOSS OR JUST ANY  
CHILD THAT HAS CMV, ARE THERE  
BABIES THAT HAVE MENTAL  
DELAYS OR LANGUAGE DELAYS OR  
IS THAT – HOW DOES THAT  
INFLUENCE THAT, WITH EITHER

HEARING LOSS OR NO HEARING  
LOSS?

>> DR. SCHLEISS:

THAT'S A GREAT QUESTION.

BABIES THAT ARE BORN WITH CMV  
THAT HAVE HEARING LOSS,

OBVIOUSLY IF THERE'S NOT A

CORRECTION OF THAT OR EARLY

INTERVENTION, THEN THERE WILL

BE DELAYS TO SPEECH AND

LANGUAGE.

SOME BABIES ARE BORN WITH CMV

AND ARE NORMAL AND THEY

REMAIN NORMAL.

THAT FIRST-GRADE CHILD WHO  
HAS A MILD LEARNING  
DISABILITY OR ATTENTION  
DEFICIT DISORDER OR AUTISM,  
THERE ARE SUBTLE NEUROLOGIC  
INJURIES THAT ARE COMPLETELY  
DISTINCT FROM HEARING LOSS.  
THAT'S SOMETHING WE NEED TO  
LEARN MORE ABOUT.

>> CAN THE HEARING  
FLUCTUATE WITH CMV, OR DOES  
IT ALWAYS GET WORSE?

>> DR. SCHLEISS: SO  
THE QUESTION IS CAN THE

HEARING FLUCTUATE IF THE BABY IS BORN WITH CMV OR DOES IT ALWAYS GET WORSE?

YOU KNOW, AGAIN, THAT'S A GREAT QUESTION.

THE ANSWER IS THAT IT CAN FLUCTUATE QUITE A BIT.

AND SO BABY MAY COME IN AT 2 MONTHS OF AGE AND HAVE AN ABNORMAL OAE AND THEN AT THE 6-MONTH VISIT MIGHT LOOK A LITTLE BETTER.

FOR IT TO FLUCTUATE OVER TIME  
IS SEEN AND WE DON'T KNOW WHY  
THAT HAPPENS.

THE OVERALL TREND IS TO  
WORSEN, SORT OF LOOKS LIKE  
THE STOCK MARKET IN THE LAST  
2 OR 3 YEARS.

MAYBE THAT'S NOT A GOOD  
ANALOGY.

IT MAKES STUDIES OF ANTI VIRAL  
DRUGS HARD TO INTERPRET.

THAT'S WHY YOU NEED BLINDED  
STUDIES, NEITHER THE  
INVESTIGATOR NOR THE PARENT

KNOWS IF THE CHILD IS GETTING  
THE ANTI VIRAL MEDI CATION.

WE ARE A PARTI CIPATI NG SI TE

HERE AT AMPLATZ CHI LDREN' S

HOSPI TAL FOR A NATIONAL

STUDY, SO WE' RE VERY

I NTERESTED I N HEARI NG ABOUT

BABI ES, YOU KNOW, I N

MI NNESOTA WHO HAVE CMV OR MAY

HAVE CMV AND WE' RE DELI GHTED

TO TALK TO THE FAMI LIES ABOUT

THE OPTI ON OF ENROLLI NG I N

THE STUDI ES.

>> DID YOU SAY IT  
IMPROVES OR STOPS IT?

>> DR. SCHLEISS: BOTH.  
SOMETIMES YOU GET IMPROVEMENT  
BUT STABILIZATION, IN OTHERS  
IT DOESN'T MAKE ANY  
DIFFERENCE AT ALL.

WHAT I TELL PARENTS, I CAN  
TELL YOU WHAT WILL HAPPEN TO  
100 BABIES, BUT I CAN'T  
PREDICT WHAT'S GOING TO  
HAPPEN TO YOUR BABY.

WE BELIEVE IT'S STANDARD –  
IT'S A STANDARD OF CARE TO AT

LEAST OFFER THIS ANTI VIRAL  
MEDI CATION FOR 6 WEEKS IN A  
BABY THAT' S BORN WITH CMV AND  
HAS SOME DEGREE OF NEUROLOGIC  
I NJURY I NCLUDING HEARI NG  
LOSS.

I BELIEVE THE ANTI VIRAL  
MEDI CATION SHOULD BE OFFERED  
TO ALL OF THESE I NFANTS.

WHAT WE DON' T KNOW I S WHETHER  
TREATING BEYOND 6 WEEKS  
I MPROVES THOSE OUTCOMES EVEN  
MORE.

IT'S NOT GOING TO CURE IT,  
IT'S NOT GOING TO MAKE THE  
HEARING LOSS GO AWAY  
COMPLETELY IN ALMOST ALL  
CASES, BUT WHAT WE HOPE WILL  
BE, IT ALLOWS THE HEARING TO  
BE STABLE FOR A LONG ENOUGH  
PERIOD OF TIME THAT BABY CAN  
GET BIG ENOUGH TO TOLERATE A  
COCHLEAR IMPLANT SURGERY, IF  
THAT'S WHAT'S EVENTUALLY  
NEEDED.

>> IS THE ONLY WAY  
ONE CAN GET CMV IS TO BE BORN  
WITH IT?

>> DR. SCHLEISS: IS  
THE ONLY WAY THAT ONE CAN GET  
CMV TO BE BORN WITH IT?  
NO.

PEOPLE TRANSMIT CMV BY CLOSE  
PERSON CONTACT.

SO THE MOST COMMON WAYS IN  
WHICH CMV IS TRANSMITTED ARE  
THROUGH EXPOSURE TO URINE,  
SALIVA, SEMEN.

CMV CAN BE A SEXUALLY-  
TRANSMITTED INFECTION.

IT CAN ALSO BE TRANSMITTED  
MOTHER TO BABY THROUGH BREAST  
MILK.

THAT'S WHY IT'S SO IMPORTANT  
TO TEST BABIES RIGHT WHEN  
THEY'RE BORN.

WE MAY HAVE A BABY COME IN  
FOR TESTING THAT'S BEEN  
BREAST FED.

WE DON'T KNOW THAT THAT  
INFECTION WAS PRESENT IN THE  
WOMB, THAT'S WHY WE NEED TO

FIND WAYS TO TEST BABIES IN  
THE FIRST WEEK OF LIFE.

>> IF A CHILD IS  
OLDER, CAN THEY STILL BE  
TESTED FOR CMV?

>> DR. SCHLEISS: YEAH,  
THAT'S ANOTHER GREAT  
QUESTION.

YOU CAN TEST AN OLDER CHILD.  
IF THAT CHILD ATTENDED GROUP  
DAYCARE OR IF THAT CHILD WAS  
BREAST FED.

WE DO HAVE ANOTHER STUDY  
THAT'S BEING CONDUCTED BY ONE

OF MY FORMER FELLOWS WHO WILL  
BE JOINING OUR FACULTY SOON,  
AND WHAT HIS STUDY HAS LOOKED  
AT WAS TO TAKE CHILDREN WHO  
HAVE COME INTO THE LIONS  
CLINIC, THE 2-YEAR-OLD, 3-  
YEAR-OLD, WHO HAVE HEARING  
LOSS AND WE CAN'T FIND A  
REASON FOR IT.

WE CAN GO BACK TO THAT BLOOD  
SPOT THAT WAS OBTAINED AT  
BIRTH.

THE STATE OF MINNESOTA SAVES  
THOSE BLOOD SPOTS.

IN THE STATE OF MINNESOTA WE  
DO.

WITH THE PARENTS' PERMISSION,  
WE CAN PULL THAT BLOOD SPOT  
AND TEST FOR CMV YEARS LATER.  
THE DNA IS VERY STABLE.

30 OR 35% OF THE TIME, WHEN  
THERE'S NO OTHER EXPLANATION  
FOR HEARING LOSS, THAT THAT  
BLOOD SPOT WILL HAVE CMV IN  
IT.

AND PARENTS ARE GLAD TO KNOW,  
THEY'RE GLAD TO HAVE AN  
ANSWER.

IN A 5-YEAR-OLD, THERE MIGHT NOT BE ANY TREATMENT WE CAN OFFER.

BUT SOMETIMES IT'S VERY SATISFYING TO HAVE AN EXPLANATION.

I THINK OF THE 150 FAMILIES THAT WE'VE APPROACHED TO GET CONSENT TO DO THIS, WE HAD 149 SAY YES, GO AHEAD.

WE ONLY HAD ONE FAMILY DECLINE.

THANK YOU.

[APPLAUSE]

>> I HAVEN'T GOTTEN A  
CHANCE TO GIVE YOU THE  
BIOGRAPHY OF THE PRESENTERS,  
BUT WE'LL PUT THEM UP ON THE  
WEBSITE FOR YOU.

THIS IS DOCTOR LISA  
SCHIMMENTI FROM GENETICS.

>> DR. SCHIMMENTI :  
THANK YOU FOR LETTING ME  
BEING HERE.  
THANK YOU FOR HANGING OUT  
THIS LATE AT NIGHT FOR THE  
LAST LECTURE.

I'M GOING TO TALK ABOUT  
HEARING AND GENETICS, BECAUSE  
HEARING AND GENETICS ARE  
IMPORTANT TO THE CARE OF KIDS  
WITH HEARING LOSS.

SO THERE'S 2 MESSAGES THAT I  
WANT YOU TO TAKE HOME.

THE GENETIC – A GENETIC BASIS  
FOR DEAFNESS OR HARD OF  
HEARING CAN BE IDENTIFIED IN  
HALF OF ALL INDIVIDUALS WHO  
ARE DIAGNOSED DEAF OR HARD OF  
HEARING.

TO UNDERSTAND THE GENETIC  
BASIS INFORMS MEDICAL  
MANAGEMENT.

AND IF YOU LEARN THOSE TWO  
THINGS TONIGHT, YOU'VE  
LEARNED A LOT ABOUT WHY  
GENETICS IS IMPORTANT.

SO THIS IS A SLIDE THAT  
SUMMARIZES A LOT OF WHAT WE  
KNOW ABOUT GENETICS AND  
DEAFNESS.

SO FOR THE CAUSES OF  
DEAFNESS, HALF IS GENETIC.

DOCTOR SCHLEISS TALKED ABOUT ENVIRONMENTAL CAUSE AND ONE OF THEM BEING CYTOMEGLAOVIRUS, AND 25% WE NEVER QUITE FIND OUT WHY A CHILD IS DEAF.

IN THE GENETIC GROUP, 30% OF KIDS WHO ARE DEAF AND HARD OF HEARING WILL HAVE A GENETIC SYNDROME.

THERE ARE OVER 400 DIFFERENT GENETIC CONDITIONS THAT CAUSE HEARING LOSS.

I'M GOING TO TALK ABOUT SOME OF THE MORE COMMON ONES THIS EVENING.

ONE IS USHER SYNDROME, THAT IS HEARING LOSS OR DEAFNESS AND VISION LOSS LATER ON IN LIFE.

BRANCHO-OTO-RENAL SYNDROME, WHICH IS A CONDITION WHERE INDIVIDUALS HAVE DIFFERENCES IN THEIR EAR SHAPE, DIFFERENCES IN STRUCTURES THAT ARE FORMED DURING DEVELOPMENT OF THE FETUS AND

SOMETIMES KIDNEY  
ABNORMALITIES THAT CAN LEAD  
TO KIDNEY FAILURE.

JERVELL LANGE-NIELSEN  
SYNDROME, THIS IS HEARING  
LOSS ASSOCIATED WITH HEART  
RHYTHM AND CAN BE LIFE-  
THREATENING.

AND ENLARGED VESTIBULAR  
AQUEDUCT PENDRED SYNDROME.  
THIS CAN BE ASSOCIATED WITH  
THYROID ABNORMALITIES.

BUT THE MAJORITY OF KIDS WHO ARE DEAF OR HARD OF HEARING HAVE WHAT'S CALLED NONSYNDROMIC HEARING LOSS, MEANING THEY HAVE NO OTHER PHYSICAL FINDINGS BUT THEIR HEARING LOSS AND THE MAJORITY OF THESE INDIVIDUALS HAVE HEARING LOSS ON A GENETIC BASIS.

AND A MAJORITY OF THESE KIDS WILL HAVE A CHANGE IN THE GENES IN GJB2 AND GJB6.

THERE ARE OTHER FORMS OF DEAFNESS IN ABOUT 15 TO 24% OF KIDS.

X LINK THAT OCCUR IN 1 TO 2% AND LESS THAN 1% OF KIDS WILL HAVE HEARING LOSS CAUSED BY CHANGES IN THE MITOCHONDRIAL GENE.

THIS IS SORT OF A SUMMARY OF THE GENETIC BASIS OF HEARING LOSS BASED ON WHERE THE DIFFERENT GENES ARE EXPRESSED IN THE COCHLEA.

AND THIS BEAUTIFUL  
ILLUSTRATION COMES FROM A  
PAPER THAT WAS PUBLISHED IN  
THE NEW ENGLAND JOURNAL ABOUT  
4 YEARS AGO.

AND IF YOU DO GET A CHANCE TO  
PULL THIS PAPER, HERE IS THE  
REFERENCE.

I KNOW THIS IS PROBABLY TOO  
SMALL TO SEE RIGHT NOW, BUT  
WHAT THEY SHOW IN THIS  
PICTURE IS WHERE THE  
DIFFERENT GENES ARE EXPRESSED  
IN THE INNER EAR, IN THE

COCHLEA, IN THE HAIR CELLS,  
IN THE DIFFERENT STRUCTURES  
THAT HAVE BEEN ASSOCIATED  
WITH DEAFNESS.

BUT BY FAIR AND AWAY GJB2  
AND GJB6 ARE THE TWO GENES  
THAT ENCODE TWO GENES,  
CONNEXIN 26 AND CONNEXIN 30  
WORLDWIDE.

IN STUDIES THAT WERE DONE IN  
THE MIDWEST ABOUT 10 YEARS  
AGO, THEY LOOKED AT 52  
INDIVIDUALS WHO WERE DEAF.

42% OF THEM HAD 2 GENETIC  
CHANGES IN THE GENE GJB2 AND  
29 OF THE 41 VARIANTS WERE  
35DELG THAT WERE DELETED.

THIS IS A RECESSIVE FORM OF  
HEARING LOSS.

IT'S VERY DIFFERENT FROM WHAT  
YOU LEARNED IN HIGH SCHOOL.

WHAT I MEAN BY RECESSIVE IS  
AN INDIVIDUAL HAS A CHANGE IN  
THE COPY OF THE GENE, A  
CHANGE IN THE GENE INHERITED  
FROM THEIR FATHER AND MOTHER  
TO HAVE THAT CONDITION.

SO I HAVE – IT LOOKS LIKE WE HAD A PC TO MAC CONVERSION ISSUE.

THERE WAS A COLOR BLUE TO SHOW THERE WAS A DIFFERENCE BETWEEN CONNEXIN 26.

WE WOULD SAY THAT INDIVIDUAL MIGHT BE A CARRIER, ALTHOUGH THERE'S SOME DATA TO SUGGEST IT MIGHT BE MORE INVOLVED IN THAT IN AN INDIVIDUAL WHO HAS DEAFNESS.

IF WE DON'T FIND ANY CHANGES IN THE GJB2 GENE, THEN WE

WOULD SAY THIS IS NOT  
CONNEXIN HEARING LOSS.

WHEN WE SEQUENCE THE  
CONNEXIN 26 GENE, THERE ARE  
MANY VARIANTS IN THE GENE.  
THEY FOUND THERE'S TWO  
DIFFERENT KINDS OF CHANGES IN  
THE GENE.

THERE ARE WHAT ARE CALLED  
TRUNCATING BARRIERS THAT  
CAUSE THE DNA TO NOT PRODUCE  
A FULL PROTEIN.

THERE WAS EITHER SMALL OR  
ABSENT PROTEIN.

THERE'S ALSO MIS-SETS.

IT DOESN'T FUNCTION LIKE IT  
SHOULD FUNCTION.

PEOPLE WITH TRUNCATING  
VARIANTS HAVE SLIGHT HEARING  
LOSS.

THIS IS THE MOST COMMON  
FORM OF HEARING LOSS  
WORLDWIDE.

PEOPLE IN THE POPULATION WHO  
ARE NOT DEAF OR HARD OF

HEARING AND ARE JUST WALKING  
AROUND AND NOT – DON'T KNOW  
ANYTHING ABOUT WHAT'S IN  
THEIR GENE, LIKE MOST OF US.  
2 TO 4% OF EUROPEAN HERITAGE  
WILL BE CARRIERS OF THAT  
35DELG CHANGE.

PEOPLE OF THE JEWISH HERITAGE  
HAVE 4 TO 7%.

PEOPLE FROM JAPAN HAVE 1 TO  
2% OF 235DELG.

AND THE OTHER IS THE W371,  
AND THOSE INDIVIDUALS HAVE A

VERY HIGH CARRIER FREQUENCY  
OF 11.6%.

WE KNOW A LOT ABOUT GJB2  
AND GJB6 RELATED DEAFNESS AND  
HARD OF HEARING.

WE KNOW THAT INDIVIDUALS WHO  
HAVE THIS CONDITION HAVE  
SENSORINEURAL DEAFNESS, WE  
KNOW THAT THEY'RE MOST LIKELY  
GOING TO HAVE BILATERAL  
DEAFNESS.

SO DEAFNESS IN BOTH EARS,  
ALTHOUGH THERE ARE RARE CASES

OF PEOPLE WHO HAVE UNILATERAL  
DEAFNESS.

IT'S GENERALLY NOT  
PROGRESSIVE.

THERE ARE REPORTS THAT  
SUGGEST SLOW PROGRESSION OVER  
TIME WITH MODERATE FORMS OF  
HEARING LOSS, BUT IT TENDS TO  
BE NONPROGRESSIVE.

INDIVIDUALS WILL HAVE NORMAL  
TEMPORAL BONE MORPHOLOGY.

AND THAT BECOMES AN IMPORTANT  
POINT WHEN I TALK ABOUT  
DIFFERENCES IN TEMPORAL BONES

THAT LEAD US TO MAKE  
DIFFERENT GENETIC TESTING  
CHOICES.

WITH THE PROTEIN TRUNCATING  
VARIANTS FALLING IN THE  
SEVERE TO PROFOUND RANGE AND  
THE NON-TRUNCATING VARIANTS  
IN THE MODERATE LOSS.

WHEN THEY HAVE THIS HEARING  
LOSS, WE CAN BE ASSURED  
THEY' RE NOT GOING TO  
EXPERIENCE OTHER SYNDROMIC  
ITEMS WITH OTHER HEARING  
LOSS.

THIS IS A PICTURE OF A  
WOMAN WHO HAS A GOITER.

THIS IS A RETINA WITH SOMEONE  
WITH USHER SYNDROME.

SYNDROME HAS SUCH NEGATIVE  
MEDICAL CONNOTATIONS.

SYN MEANS WITH AND DROME  
MEANS RUN TOGETHER.

THE PHRASE WAS COINED, AS  
CLINICIANS WE'RE ALERT TO  
THINGS THAT RUN TOGETHER SO  
WE DON'T MISS SOMETHING.

AN EXAMPLE WOULD BE PENDRED  
SYNDROME, THAT RUNS TOGETHER  
WITH THYROID GOITER.

THIS CREATES A PREVENTATIVE  
MEDICINE MODEL FOR KIDS WHO  
ARE DEAF OR HARD OF HEARING.  
WE CAN CREATE PLANS FOR  
MEDICAL CARE.

SO I WANT TO TALK ABOUT  
THE ENLARGED VESTIBULAR  
AQUEDUCT SYNDROME.

IT CAN BE ISOLATED OR NOT AND  
THAT DEAFNESS IS THE ONLY  
ISSUE SOMEONE HAS.

BUT YOU COULD SEE IT WITH  
OTHER GENETIC CONDITIONS,  
THIS INCLUDE PENDRED  
SYNDROME, BRANCHO-OTO-RENAL  
SYNDROME, WAARDENBERG  
SYNDROME.

SO WHAT IS THE VESTIBULAR  
AQUEDUCT?

IT'S A BONY STRUCTURE THAT  
CONNECTS THE INNER EAR TO THE  
– IN THE SKULL TO THE BRAIN

AREA AND IT CARRIES INNER EAR  
FLUID AND BLOOD VESSELS.

THE REASON THIS MATTERS IS  
THAT IT IS THE MOST COMMON  
BONY INNER EAR MALFORMATION  
IN SENSORINEURAL DEAFNESS, IT  
SEEMS TO BE MORE COMMON IN  
GIRLS, THE MAIN ISSUE IT'S  
IMPORTANT IS CAN BE  
ASSOCIATED WITH PROGRESSION  
OF HEARING LOSS OVER TIME.  
AND SO FOR PARENTS WHO ARE  
ASKING WILL MY CHILD'S

HEARING GET WORSE OVER TIME,  
IN CHILDREN WHERE WE FIND  
ENLARGED VESTIBULAR AQUEDUCT,  
WE CAN SAY YES, THAT'S  
PROBABLY TRUE BECAUSE OF THIS  
FINDING.

THIS IS A PICTURE OF THE  
INNER EAR, HERE'S THE  
ENDOLYMPHATIC SACK AND DUCT  
AND HERE'S THE ENLARGEMENT OF  
THE VESTIBULAR AQUEDUCT.

NOW WE DON'T HAVE BEAUTIFUL  
COLOR DRAWINGS IN THE CLINIC

AND WE HAVE TO RELY ON THINGS  
LIKE CT SCANS.

AND SO HERE'S AN ENLARGED – I  
MEAN, A REGULAR AQUEDUCT IN A  
CT SCAN, HERE'S AN INDIVIDUAL  
WITH AN ENLARGED VESTIBULAR  
AQUEDUCT.

IT ALMOST LOOKS LIKE A THUMB  
PRINT IN CLAY IN THE AREA OF  
THE VESTIBULAR AQUEDUCT.

ONE OF THE FIRST THINGS WE  
DO WHEN WE SEE CHILDREN AT

LIONS CLINIC, WE TAKE A CT  
SCAN.

WE CAN THEN MAKE SOME  
DECISIONS ON GENETIC TESTING  
FOR THAT INDIVIDUAL.

SO THE MOST COMMON  
SYNDROME ASSOCIATED WITH  
ENLARGED VESTIBULAR AQUEDUCT  
IS PENDRED SYNDROME.

THEY HAVE VARIABLE  
SENSORINEURAL DEAFNESS, IT'S  
PROGRESSIVE, I'VE SEEN  
CHILDREN AS YOUNG AS AGE 5

DEVELOP A GOITER AND PEOPLE  
HAVING ENLARGED VESTIBULAR  
AQUEDUCT.

THE INHERITANCE OF THIS IS  
AUTOSOMAL RECESSIVE.

GENE SLC26A4 IS THE MOST  
COMMON GENE INVOLVED AND IT'S  
THE SECOND MOST COMMON CAUSE  
OF HEARING LOSS.

### BRANCHO-OTO-RENAL

SYNDROME IS ANOTHER SYNDROME  
ASSOCIATED WITH ENLARGED  
VESTIBULAR AQUEDUCT, IT'S

FOUND IN 7.5% OF DEAFNESS AT BIRTH.

YOU ONLY NEED A CHANGE OF ONE COPY OF THE GENE TO HAVE THE CONDITION.

THE MOST COMMON GENE ASSOCIATED WITH BRANCHED-OTO-RENAL SYNDROME IS EYA1.

BUT THERE ARE OTHER GENES THAT HAVE BEEN IDENTIFIED AND TESTING IS NOT YET AVAILABLE, BUT WE HOPE TO SEE IT SOON.

SOME OF THE OTHER FINDINGS  
IN BRANCHO-OTO-RENAL  
SYNDROME, SENSORINEURAL,  
CONDUCTIVE OR MIXED DEAFNESS  
OR HARD OF HEARING, CYSTS OR  
FISTULA, INDIVIDUALS CAN HAVE  
ABNORMALITIES OF THE KIDNEYS,  
THERE CAN BE ABNORMAL SHAPES  
OF THE OUTER EAR OF THE PINNA  
OR EAR PITS OR TAGS THAT CAN  
BE PRESENT.

WAARDENBERG SYNDROME IS ANOTHER CONDITION THAT HAS AN ENLARGED VESTIBULAR AQUEDUCT. IT'S ALSO A DOMINANT CONDITION. THERE ARE MANY GENES THAT CAUSE WAARDENBERG SYNDROME, WITH PAX3 AS THE MOST COMMON. SOME OF THE CLINICAL FINDINGS INCLUDE PROFOUND SENSORINEURAL HEARING LOSS, INDIVIDUALS CAN HAVE DIFFERENT COLOR BLUE EYES, A WHITE PATCH OF HAIR, AND

SOMETIMES CHILDREN WILL HAVE SEVERE CONSTIPATION BECAUSE THEY HAVE ABSENT NERVE CELLS IN THEIR INTESTINE AND CAN PRESENT THAT WAY.

SO THE NEXT GROUP OF CONDITIONS I WANT TO TALK ABOUT VISION AND HEARING. AND THE MOST COMMON OF THIS IS USHER SYNDROME.

AND STUDIES HAVE SHOWN THAT 3 TO 6% OF DEAF CHILDREN HAVE USHER SYNDROME.

THIS IS A RECESSIVE  
CONDITION.

THERE ARE A NUMBER OF  
DIFFERENT FORMS AND A NUMBER  
OF DIFFERENT GENES THAT ARE  
ASSOCIATED WITH USHER  
SYNDROME.

THE ISSUE FOR CHILDREN WITH  
USHER SYNDROME IS THAT  
CHILDREN WILL USUALLY SHOW UP  
FIRST WITH BEING DEAF OR HARD  
OF HEARING, BUT THEN SOMETIME  
IN LATE CHILDHOOD OR EARLY  
TEENS WILL BEGIN TO LOSE

THEIR VISION TO AN EYE  
CONDITION CALLED RETINITIS  
PIGMENTOSA WHERE THE CELLS  
STOP LIVING AND DIE.

THE THREE TYPES THAT WE  
SEE, TYPE I, THEY ARE  
PROFOUND SENSORINEURAL  
DEAFNESS, AND THE CAUSE OF  
THAT WILL HAVE BALANCE ISSUES  
AND HAVE DELAYED WALKING.  
RETINITIS PIGMENTOSA WILL  
START EARLY, SOMETHING LIKE  
NIGHT BLINDNESS.

TYPE II HAS MILD TO SEVERE  
SENSORINEURAL HEARING LOSS AT  
BIRTH AND MAY BE IN THE  
HIGHER FREQUENCIES, THE  
PROGRESSION CAN BE INTENSIVE  
AND RETINITIS PIGMENTOSA CAN  
BEGIN LATER.

AND THE AGE OF ONSET OF  
VISION LOSS IS VARIED.

JERVELL LANGE-NIELSEN  
SYNDROME.

WHEN THEY COME TO THE LIONS  
CLINIC, WE TAKE AN EKG.

THERE ARE GENES THAT ARE  
IMPORTANT FOR TRANSPORT OF  
POTASSIUM BETWEEN CELLS THAT  
ARE INVOLVED IN JERVELL  
LANGE-NIELSEN SYNDROME.

PARENTS WHO ARE CARRIERS CAN  
ALSO HAVE LQT SYNDROME.

IF YOU CAN IMAGINE THIS, WE  
CAN GO TO THE SISTERS AND  
BROTHERS AND AUNTS AND UNCLES  
AND TEST THAT.

CLINICAL FINDINGS, SEVERE  
TO PROFOUND CONGENITAL

SENSORI NEURAL DEAFNESS,  
PATIENTS HAVE PRESENTED WITH  
PASSING OUT.

WE'VE ALL HEARD STORIES ABOUT  
A CHILD WHO WAS DEAF WHO HAD  
TO SPEAK IN FRONT OF AN  
AUDIENCE WHO WOULD PASS OUT  
AND THERE HAVE BEEN REPORTS  
OF SUDDEN DEATH.

SO LET ME TALK TO YOU  
ABOUT SOME CASE STUDIES FROM  
THE LIONS CHILDREN'S HEARING  
CLINIC WHERE THE GENETIC

TESTING HAS BEEN HELPFUL IN PROVIDING CARE FOR KIDS.

SO IN THIS CASE, THIS IS A NEWBORN WHO HAD THE NEWBORN HEARING SCREENING.

THE CHILD WAS FITTED WITH HEARING AIDS, A CT SCAN WAS PERFORMED THAT SHOWED NO NORMAL BONE MORPHOLOGY.

THIS CHILD'S PARENTS WERE OF EUROPEAN HERITAGE AND NO HISTORY OF HEARING LOSS.

GENETIC TESTING WAS PERFORMED FOR GJB2 AND GJB6.

THEY WERE FOUND TO HAVE  
CHANGES IN THE 167DELT AND  
THE 35DELG.

WE DON'T HAVE CONCERNS FOR  
OTHER HEALTH ISSUES RELATED  
TO HEARING LOSS.

AND THIS CHILD'S HEARING HAS  
BEEN STABLE OVER A 2-YEAR  
TIME PERIOD.

THE SECOND CASE IS A CHILD  
WHO WAS 15 MONTHS OLD WHO HAD  
FLUCTUATING BILATERAL  
SENSORINEURAL DEAFNESS.

THE CHILD HAS HEARING AIDS  
PLACED BUT HAD PROBLEMS,  
WOULD CHANGE OVER SHORT  
PERIOD OF TIME WITH  
PROGRESSIONS VARIED.

AND WAS TESTED FOR CHANGES IN  
GJB2 AND GJB6 BUT NO CHANGES  
WERE IDENTIFIED.

THERE'S NO HISTORY OF HEARING  
LOSS AND THEN A CAT SCAN WAS  
PERFORMED WHICH SHOWED  
ENLARGED VESTIBULAR AQUEDUCT.

WE IDENTIFIED TWO CHANGES IN THE GENE AND THIS CONFIRMED THE DIAGNOSIS.

AND WE THEN HAD A CARE PLAN IN PLACE FOR THIS CHILD THAT THIS CHILD SHOULD BE CHECKED BY THE PRIMARY CARE PROVIDER FOR THYROID FUNCTION AND FOLLOWED FOR DEVELOPMENT OF GOITER, THAT CHILD IS ALSO BEING FOLLOWED WITH PROGRESSION FOR THEIR HEARING LOSS AND MONITORED CLOSELY.

IN THIS SITUATION, A  
LITTLE BOY PRESENTED AT 5  
YEARS OF AGE WITH PROFOUND  
SENSORINEURAL HEARING LOSS.  
HE WALKED LATE.  
AND THIS CHILD HAD A COCHLEAR  
IMPLANT AT AGE 6.  
HIS CT SCAN WAS READ AT  
NORMAL.  
TWO VARIANTS IN THE GENE WERE  
IDENTIFIED CONFIRMING THE  
DIAGNOSIS OF USHER SYNDROME.  
THIS CHILD HAS CLOSE  
MONITORING AND IS DEVELOPING

SOME NIGHT BLINDNESS AND  
PROGRESSIVE VISION LOSS.  
THE ONE THING ABOUT USHER  
SYNDROME THAT WE'LL ALL SEE  
IS THAT GENE THERAPY LOOKS  
LIKE IT MIGHT BECOME A  
REALITY WITH KIDS WITH USHER  
SYNDROME.

THERE ARE CLINICAL TRIALS.  
SO THE HOPE IS WITH EARLY  
IDENTIFICATION AND AS GENE  
THERAPY MATURES, THIS IS  
SOMETHING WE'RE GOING TO BE

ABLE TO OFFER KIDS WITH USHER SYNDROME.

THE LAST SITUATION I'M GOING TO TALK ABOUT IS A CHILD WHO IS 5 MONTHS OLD WITH PROFOUND SENSORI NEURAL HEARING LOSS.

THIS CHILD HAS HEARING AIDS PLACED BY 2 MONTHS OF AGE BUT HAD LITTLE RESPONSE.

HIS MOTHER REPORTED HE RESPONDED TO A LOUD ALARM AT CLOSE AGE.

A COCHLEAR IMPLANT IS BEING  
DISCUSSED WITH THIS CHILD AND  
THE CT SCAN WAS NORMAL.  
GENETIC TESTING WAS PERFORMED  
FOR GJB2 AND GJB6 AND SOC264.  
SO THE CAUSE OF HEARING LOSS  
IS UNKNOWN.

AND STATISTICALLY WHAT WE  
FIND IS THAT 14% OF THE TIME  
IN KIDS THAT WE DON'T  
IDENTIFY A GENETIC CHANGE, WE  
WILL SEE A FUTURE BROTHER OR

SISTER HAVE SENSORI NEURAL  
HEARING LOSS.

SO THE CHANCE THAT WE TELL  
FAMILIES FOR HAVING ANOTHER  
CHILD WITH HEARING LOSS IF  
THE GENETIC TESTING IS ABOUT  
14%.

THERE'S ONGOING MEDICAL  
MANAGEMENT FOR THIS CHILD IN  
ANTICIPATION THAT THERE COULD  
BE PROBLEMS WE'VE NOT  
IDENTIFIED.

AND WE FOLLOW THIS CHILD FOR  
DEVELOPMENT OF NORMAL  
MILESTONES.

SO I HOPE THAT I FULFILLED  
MY OBJECTIVES TONIGHT, AND  
THAT IN CHILDREN WHO ARE DEAF  
AND HARD OF HEARING HALF OF  
THEM WILL HAVE A GENETIC  
CAUSE THAT WE CAN IDENTIFY  
AND THEN UNDERSTANDING THE  
GENETIC BASIS OF DEAFNESS  
WILL INFORM MEDICAL  
MANAGEMENT.

SO THANK YOU VERY MUCH FOR  
YOUR ATTENTION AND IN  
STICKING WITH ME.

>> AUDIENCE MEMBER: I  
HAVE A QUESTION.

I HAVE TAKEN A TYMPANIC  
GENETIC TEST A WHILE AGO,  
ABOUT 6 YEARS AGO, IT WAS A  
BLOOD TEST FOR CONNEXIN 26.  
ONE FORM OF THAT IS FROM MY  
DAD'S SIDE.

I'M NOT SURE IF IT WAS FROM  
MY DAD'S SIDE OR MOTHER'S

SIDE, BUT THEY FOUND ONE FROM THAT.

THEY SAID REALLY IT'S RARE, 1 OF ONLY 15 PEOPLE IN THE WORLD FOUND TO HAVE THAT.

>> SIGN INTERPRETER:

I'M SORRY, WE'RE GOING TO HAVE TO SWITCH INTERPRETERS, I'M HAVING A HARD TIME SEEING FROM BACK HERE.

I APOLOGIZE.

>> AUDIENCE MEMBER:

THIS WILL BE BETTER IF I  
STAND UP.

SO I GOT THE CONNEXIN 26 FROM  
ONE OF MY PARENTS, NOT SURE  
EXACTLY WHICH ONE I GOT IT  
FROM, SO WE HAD THE BLOOD  
TEST TO FIGURE OUT WHICH ONE.  
MY FATHER AND I HAD THE SAME  
THING WITH THE CONNEXIN 26,  
SOME OTHER THING WITH A LONG  
NAME, THAT WAS PASSED ON FROM  
MY FATHER – MY GRANDPARENTS  
WERE HEARING, SO WE WERE

TRYING TO FIGURE OUT HOW THAT ALL GOT PASSED DOWN AND WHERE IT CAME FROM.

AND SO MY GRANDPARENTS WERE CARRIERS, BUT – THEY WERE HEARING BUT THEY DIDN'T REALIZE THEY WERE CARRIERS.

AND IN EUROPE, 1 AND 3 ARE CARRIERS FOR THIS AND THE NUMBER STATISTICS WERE HIGHER IN EUROPE THAN AMERICA.

IS THERE AN EXPLANATION WHY THERE'S MORE CARRIERS IN EUROPE THAN IN AMERICA?

>> DR. SCHIMMENTI: THE NUMBER OF CARRIERS IN EUROPE AND AMERICA ARE PRETTY SIMILAR, BUT THERE SEEMS TO BE A VARIANT IN MEDITERRANEAN.

AMERICA IS A MELTING POT. ABOUT 3 TO 4% OF AMERICANS, AT LEAST IN THE MIDWESTERN STUDIES, ARE CARRIERS, THAT'S WHAT WE SEE IN MINNESOTA, TOO.

ONE OF THE THINGS YOU MIGHT HAVE ELUDED TO, AND LET ME

MAKE SURE I UNDERSTAND IT  
CORRECTLY, THEY ONLY FOUND  
ONE GENETIC CHANGE IN YOU  
THAN TWO, IS THAT CORRECT?  
OR DID THEY FIND TWO?

>> AUDIENCE MEMBER: I  
DO HAVE TWO.

>> DR. SCHIMMENTI: OK.

AUDIENCE MEMBER: I  
HAVE THE COMMON KIND, THE  
CONNEXIN 26 AND THE ONLY ONE  
IS THE 1 IN 15 PEOPLE IN THE  
WORLD.

THAT'S WHY I CAME TO BE DEAF.

MY FATHER AND I HAVE THE SAME ONE, BOTH MY FATHER AND I ARE 2 OF THE 15.

ON MY MOM'S SIDE, THEY CARRIED THE MORE COMMON ONE. I WAS WONDERING ABOUT THOSE STATISTICS.

>> DR. SCHIMMENTI: YOU KNOW, THERE ARE SO MANY DIFFERENT VARIANTS IN THIS GENE, OVER 100 VARIANTS. WE JUST IDENTIFY THEM IN FAMILIES.

WE' RE STILL IDENTIFYING NEW  
CHANGES THAT WE HAVEN' T SEEN  
BEFORE.

>> I HAVE A QUESTION  
FROM HOME.

SHOULD A CT AND MRI BE DONE  
AS A ROUTINE PART OF  
DIAGNOSIS?

MY CHILD IS 14 WEEKS, MANY OF  
THOSE TESTS HAVE NOT BEEN  
DONE.

MANY PRESENTED IN DOCTOR  
RIMELL' S PRESENTATION HAVE  
BEEN.

>> DR. SCHIMMENTI: WE  
DO.

I THINK IN OTHER PRACTICES WE  
DO IT AT OTHER TIMES OR HAVE  
A DIFFERENT STYLE.

THE QUESTION IS AT WHAT TIME  
SHOULD A CT OR MRI BE  
PERFORMED IN IDENTIFIED  
TEMPORAL BONES.

AT LEAST FROM MY PRACTICE  
STYLE, IT TENDS TO INFORM THE  
GENETIC TESTING I'M GOING TO  
DO AND ALSO INFORM THE KIND

OF PROGNOSIS WE CAN GIVE  
FAMILIES.

NONE OF THIS IS WRITTEN IN  
STONE YET, THERE ARE  
PRACTICED GUIDELINES THAT ARE  
DONE YET.

AND I THINK EVERYBODY DOES  
THINGS A LITTLE DIFFERENTLY.

>> MY QUESTION IS  
ABOUT THE SYNDROMIC CAUSES OF  
YOUR LOSS.

IS IT SOMETHING THAT'S SOLELY  
DETERMINED BY GENETIC TEST OR  
BLOOD TEST, OR ARE THERE

CHARACTERISTICS THAT YOU TAKE  
INTO CONSIDERATION?

>> DR. SCHIMMENTI :

YEAH, GENETIC TESTING IS SO  
EXPENSIVE.

IT'S \$1,000 FOR TESTING.

GJB2 OR GJB6 ARE LESS  
EXPENSIVE.

SO WE WANT TO TAKE AN  
APPROACH WHERE WE MAKE THE  
BEST DECISION BASED ON  
CLINICAL FINDINGS IN ON  
INDIVIDUAL.

THE REASON I'M KIND OF SAYING IT THIS WAY, THERE ARE NEW METHODS THAT ARE GOING TO BE AVAILABLE IN THE NEXT FEW YEARS WHERE YOU CAN TEST MANY GENES AT ONCE AT A VERY LOW PRICE.

WE JUST DID A VISION – SO THESE ARE DONE ON WHAT'S CALLED A GENE CHIP.

WE DID A RESEARCH STUDY USING A VISION LOSS CHIP AND WE TESTED 112 GENES FOR \$8,000.

WE EXPECT THE PRICES ARE  
GOING TO DROP.

AND THE PRACTICE OF TESTING  
ONE GENE AT A TIME WE DO NOW  
IS GOING TO CHANGE.

AND CHANGE SOON.

>> SO SAY A PARENT  
GETS A GENETIC TEST AND IT  
COMES BACK NEGATIVE, DOES  
THAT MEAN THE SYNDROME IS NOT  
PRESENT?

>> DR. SCHIMMENTI : NO.

I THINK WE HAVE TO GO ON WHAT  
THE CLINICAL FINDINGS ARE  
FIRST FOR MANY CONDITIONS.

IF GENETIC TESTING IS  
NEGATIVE, IT DOESN'T RULE OUT  
A GENETIC COST.

>> THANK YOU.

[APPLAUSE]

>> WELL, I JUST  
WANTED TO THANK THE  
PRESENTERS FOR ALL THEIR TIME  
AND EFFORT PUT INTO THOSE  
PRESENTATIONS.

I'VE SEEN I THINK EACH OF YOU  
AT LEAST ONCE, IF NOT SEVERAL  
TIMES, AND EVERY TIME I LEARN  
SOMETHING NEW.

SO I APPRECIATE YOU TOOK THIS  
EVENING TO SPEND SOME TIME  
WITH OUR NEW FAMILIES.

SO THANK YOU VERY MUCH TO  
EACH ONE OF YOU.

AND WE HAVE A LITTLE GIFT, SO  
DON'T YOU LEAVE BEFORE YOU  
GET YOUR GIFT.

ALSO, I WANTED TO THANK  
ALYSSA ANDERSON WHO HAS BEEN  
A GODSEND TO US.

SO THANK YOU.

THANK YOU TO THE LIONS  
CHILDREN'S HEARING CENTER.

I WANT TO THANK THE ASL  
INTERPRETER AND THE CAPTIONER  
FROM RIVERSIDE FOR ALL YOUR  
WORK.

THANK YOU FOR THE  
PARTICIPANTS FOR COMING OUT.

ALSO, THOSE THAT ARE  
ATTENDING BY WEBINAR.

WE' RE GOING TO BE CUT OFF IN  
A MI NUTE.

YOU CAN FIND THIS ON OUR  
WEBSI TE AT  
WWW. MNHANDSANDVOI CES. ORG.

I F YOU HAVE QUESTI ONS THAT  
YOU HAVE, YOU CAN E-MAI L.  
WE WI LL HAVE THE CAPTIONED  
TRANSCRI PT AND I BELIEVE ALSO  
THE VI DEO SCREAM, WE' RE  
HOPI NG TO ALSO POST UP ON  
THERE.

SO LOOK FORWARD TO THOSE AND  
SHARE THAT WITH PEOPLE WHO  
HAVE NOT BEEN ABLE TO  
PARTICIPATE TONIGHT.

AGAIN, I'M JUST VERY INDEBTED  
TO EACH AND EVERY ONE OF YOU.

>> AUDIENCE MEMBER: I  
HAVE A QUICK QUESTION.

SO THE WAY TO CONTACT WOULD  
BE JUST THAT ONE.

IS THE POWER POINT SOMETHING  
THAT WOULD BE E-MAILED OUT?

>> IT'S ON OUR  
WEBSITE.

RIGHT NOW IT'S THERE ON THE  
WEBSITE.

ANY OTHER LAST-MOMENT  
QUESTIONS?

OTHERWISE THEY'RE CUTTING US  
OFF.

I'LL QUICKLY INTRODUCE OUR  
STAFF.

THESE GUYS ARE METRO FOLKS,  
THEY DO THE MAJORITY OF THE  
WORK, I SIT AROUND AND EAT  
CHOCOLATE.

LAURA GODFREY, JEN.

THESE LADIES ARE A HUGE PART  
OF OUR GROUP.

THANK YOU.

I HAVE CEUS FOR

PROFESSIONALS, I ALSO HAVE

PARENT CEUS, ' CAUSE I FEEL

PARENTS DESERVE RECOGNITION

FOR ALL THEY DO FOR THEIR

CHILDREN.

PEOPLE WILL SAY WHAT HAVE YOU

DONE LATELY, HUH, SEE.

THANKS AGAIN.