

HANDS & VOICES

April 30, 2009

6:30-8:30pm

>> WELCOME.

I'M BARBARA FREEDMAN, AN AUDIOLOGIST FROM THE UNIVERSITY OF MINNESOTA MEDICAL CENTER-FAIRVIEW.

THIS IS THE THIRD YEAR OF THE JOINT VENTURE WITH MINNESOTA HANDS AND VOICES AND LYONS CHILDREN HEARING CENTER.

TODAY WE'RE FORTUNATE TO HAVE SPEAKERS WHO HAVE WORKED WITH CHILDREN WITH AUDITORY NEUROPATHY.

WHEN I STARTED IN AUDIOLOGY MANY YEARS AGO WE DIDN'T HAVE A REASON WHY MANY OF THE KIDS PASSED HEARING TESTS BUT ACTED LIKE THEY WERE DEAF.

NOW WE KNOW A LOT MORE AND WE HOPE BY THE END OF THE EVENING SO WILL YOU.

I'M GOING TO INTRODUCE THE FIRST THREE SPEAKERS TO SAVE TIME.

DR. TINA HUANG IS A NEUROTOLOGIST AND DID HER TRAINING AT NEW YORK UNIVERSITY.

IN ADDITION TO SPECIALIZING IN EAR DISORDERS SHE DOES EVEN MORE INTRICATE SURGERY OF THE INNER EAR.

SHE'S DONE MANY OF THE COCHLEAR IMPLANT SURGERIES FOR ADULTS AND CHILDREN.

DR. FERRELLO DID HER INTERNSHIP WITH US AND DID A LOT TO DO WITH AUDIOLOGY AND WE'RE FORTUNATE SHE'S DEVELOPED A SPECIAL INTEREST IN WORK WITH AUDIOLOGY AND INTEROPERATIVE MONITORING AND PEDIATRIC ASSESSMENT MANAGEMENT AND COCHLEAR IMPLANTS.

LAURA NYHUS IS AT GILLETT CHILDREN'S SPECIALTY HEALTHCARE.

**PRIOR TO THAT WORKING WITH US AS A SENIOR ORAL
REHABILITATION SPECIALIST.**

**SHE'S WELL VERSED IN WORKING WITH HARD OF HEARING AND DEAF
CHILDREN.**

WE'LL START WITH DR. HUANG.

>> SO THIS IS QUITE EXCITING.

I SEE THAT WE HAVE 15 PEOPLE ONLINE RIGHT NOW.

**SO HOPEFULLY ALL THE PEOPLE THAT ARE ONLINE ARE ACTUALLY
ABLE TO HEAR US AS WELL AS SEE THE – AS WELL AS SEE THE
SCREEN.**

**SO I'M JUST GOING TO GIVE AN OVERVIEW ESSENTIALLY OF WHAT
AUDITORY NEUROPATHY OR SOME PEOPLE WILL CALL IT AUDITORY
DYSYNCHRONY IS.**

A LOT OF MY SLIDES ARE PICTURES.

**WITHOUT HAVING ANY IDEA OF WHAT THE AUDITORY PATHWAY IS IT
DOESN'T MAKE SENSE TO PROBABLY EVEN TALK ABOUT THIS.**

SO WE'RE GOING TO TALK ABOUT ANATOMY FIRST.

WE'LL MOVE THINGS AROUND A LITTLE BIT.

I HOPE I'M NOT GIVING YOU MOTION SICKNESS HERE.

**AND SO THIS IS JUST THE MOST BASIC DIAGRAM YOU CAN HAVE OF
AUDITORY PATHWAY WHERE THE SOUND COMES IN THROUGH THE
EAR CANAL, HITS THE EARDRUM THAT'S TRANSMITTED TO THE FLUID
OF THE COCHLEA AND THAT GETS TRANSPORTED TO THE AUDITORY
NERVE AND THAT GOES TO THE BRAIN CENTERS AND UP TO GO INTO
YOUR MORE REFINED PROCESSING TO TURN THAT SOUND INTO
SOMETHING UNDERSTANDABLE AND CERTAINLY TO TURN IT INTO
SPEECH AND MUSIC.**

**AND SO THIS IS JUST A LITTLE HIGHER POWER VIEW AND SO HERE IS
THE EARDRUM RIGHT HERE.**

**AND HERE IS THE CHAIN THAT CONNECTS TO THE COCHLEA ITSELF
AND THIS IS A CROSS SECTION OF THE COCHLEA AND THE COCHLEA**

HAS 2 ½ TURNS IN IT SO IT LOOKS LIKE A LITTLE BIT OF A SPIRAL STAIRCASE AND IN THE MIDDLE OF THAT SPIRAL STAIRCASE IS THE COCHLEAR NERVE.

AND THE NERVE IS RIGHT HERE AND IT IS THE AUDITORY NERVE AND YOU'LL SEE ON THIS DIAGRAM THERE ARE THESE LITTLE TENDRILS THAT GO OUT TO ALL DIFFERENT PARTS OF THE COCHLEA.

THAT'S REALLY THE MAIN NERVE THAT GOES IN.

AGAIN, HERE IS ANOTHER HIGHER POWERED VIEW OF IT.

SO HERE IS THE NERVE THAT GOES IN RIGHT INTO THE CENTER OF THAT SPIRAL AND IT SENDS – IT HAS ALL THESE CONNECTIONS THAT GO INTO ALL THE DIFFERENT PARTS OF THE COCHLEA AND ON THIS PICTURE HERE, THIS IS ACTUALLY A – CALLED A CONFOCAL MICROGRAPH AND THE NERVE CELLS ARE STAINED YELLOW AND THE BONE OF THE EAR IS STAINED RED AND YOU CAN SEE THE CLOVER LEAF CONFIGURATION OF THE COCHLEA.

IN THE MIDDLE OF THE CLOVER LEAF ARE ALL THE YELLOW FIBERS OF THE NERVE.

AND THIS IS THE REST OF THE AUDITORY PATHWAY.

I REALIZE IT'S REALLY SMALL HERE BUT WHAT IS REALLY IMPORTANT TO KNOW IS YOUR COCHLEA IS OUT HERE AND HERE IS YOUR AUDITORY NERVE AND HERE THEY CALL IT THE COCHLEA VESTIBULAR BECAUSE THE NERVES ARE BUNDLED TOGETHER.

THEY GO UP TO THE BRAINSTEM AND GOES INTO THE COCHLEAR NUCLEUS HERE.

AND YOU GET SOME PROCESSING OF INFORMATION RIGHT HERE. SO YOU GET SIGNAL INTENSITY, FREQUENCY, DURATION, A LOT OF THAT IS DONE IN THE COCHLEA ITSELF BUT SOME OF IT IS ALSO PROCESSED IN THE NUCLEUS.

FROM HERE IT ACTUALLY GOES TO THE SUPERIOR OLIVARY COMPLEX.

THIS IS ALSO IN THE BRAINSTEM SO THESE ARE ALL A LITTLE BIT ON THE LOWER CENTER PROCESSING PATHWAYS.

AND FROM HERE IT ACTUALLY GOES UP FURTHER INTO A PORTION OF THE BRAINSTEM HERE AND THERE IS MORE PROCESSING GOING ON AT THIS AREA.

THIS HELPS WITH VOCALIZATION AND THEN FROM HERE IT GOES INTO THE CORTEX OF THE BRAIN ITSELF.

SO HERE IS WHERE YOU HAVE YOUR HIGHER PROCESSING CENTERS WHERE YOU'RE TURNING SOUND AGAIN INTO SOMETHING RECOGNIZABLE.

YOU ARE TURNING IT INTO THAT SYMPHONY.

YOU'RE TURNING IT INTO AN UNDERSTANDABLE CONVERSATION.

SO A LOT OF THAT – SOME OF THAT HAPPENS ALL THROUGH THESE LOWER PROCESSES AND PATHWAYS BUT A LOT OF THAT ACTUALLY HAPPENS UP HERE IN THE CORTEX.

AND SO ONE OF THE PROBLEMS WHICH AUDITORY NEUROPATHY OR DYSYNCHRONY IS WE DON'T KNOW WHERE IN THIS LONG PROCESS THAT THINGS ARE GOING AWRY.

IF THE NERVE ITSELF IS NOT WORKING, THEN NOTHING ABOVE THAT IS GOING TO WORK, EITHER.

AND SO WE CAN'T REALLY TELL, AND IF THE NERVE IS ONLY PARTIALLY WORKING AGAIN WE HAVE NO WAY OF TESTING WHAT IS GOING ON ABOVE THAT, REALLY, TO BE ABLE TO TELL WHAT PART OF THAT PATHWAY IS ACTUALLY ABNORMAL.

THIS IS JUST A PICTURE TO SHOW YOU – THIS IS ACTUALLY A LEFT EAR AND THE RIGHT EAR IS THIS LITTLE SHADED AREA HERE.

BUT WHAT IT'S REALLY SHOWING YOU IS THAT ALL THESE PATHWAYS ACTUALLY CROSS.

THE COCHLEAR NERVE ITSELF ONLY GOES TO ONE COCHLEA, ONLY GOES TO ONE SIDE.

WHEN IT COMES TO THE BRAINSTEM IN THE NUCLEUS THAT IS ALSO ONE SIDED.

ONCE IT PASSES THE BRAINSTEM AND GOES INTO THE OTHER CENTERS OF THE BRAINSTEM THOSE FIBERS GO UP EACH SIDE AND THEY ALSO CROSS OVER.

THERE IS A LOT OF CROSSING OF THE PATHWAYS AS WELL WHICH AGAIN MAKES IT EVEN MORE DIFFICULT TO TRY TO LOCALIZE WHERE THE PROBLEM IS.

THE GOOD PART ABOUT THE CROSSOVER IS THAT, AGAIN, THE BODY IS VERY GOOD, THERE IS A LOT OF REPETITIVE AND KIND OF YOU MIGHT ALMOST SAY USELESS THINGS BUT THAT'S WHY WE HAVE TWO OF A LOT OF OUR THINGS.

THE SAME WITH YOUR VISION.

THERE IS A LOT OF CROSS OVER IN VISION AS WELL.

EVEN IF YOU HAVE ONLY ONE WORKING EAR OR ONE WORKING EYE, YOU CAN STILL GET A LITTLE BIT OF SPATIAL SENSATION WITH ONLY ONE EYE.

YOUR BRAIN FIGURES IT OUT.

WITH ONE EAR PEOPLE HAVE MUCH HARDER TIME VOCALIZING BUT THERE ARE PEOPLE WHO CAN ACTUALLY LEARN TO LOCALIZE ESPECIALLY THE YOUNGER YOU ARE.

KIDS ARE VERY, VERY WHAT WE TERM – ALL THE NEURONS IN THEIR BRAIN ARE MAKING NEW CONNECTIONS ALL THE TIME.

IF YOU STIMULATE THOSE CONNECTIONS THEY'LL MAKE THEM.

AND SO A NEUROPATHY IS ACTUALLY A PROBLEM.

A NERVE HAS DIFFERENT PARTS.

THERE IS ACTUALLY A CELL BODY TO THE NERVE AND THAT HAS ALL THE VITAL FUNCTIONS OF THE NERVE, BUT THEN THERE ARE FIBERS THAT COME OUT TO THE NERVE CALLED AXONS.

THEY GO AWAY FROM THE NERVE AND THOSE ARE WHAT ACTUALLY CARRY THE ELECTRICAL SIGNAL OUT TO WHATEVER THAT AXON IS TOUCHING.

THAT'S HOW YOU MAKE THAT CONNECTION FROM ONE TO THE NEXT. IT IS ALMOST LIKE PLAYING TELEPHONE THROUGH ALL THE DIFFERENT NERVES.

THEY ALL KIND OF TALK TO EACH OTHER AND THE AXON IS WHAT HELPS TAKE THAT SIGNAL DOWN AND WHAT GIVES THAT ABILITY TO TALK TO ONE ANOTHER.

AND SO THE PROBLEM ISN'T ACTUALLY WITH THE MAIN CELL BODY, IT'S ACTUALLY WITH THOSE LITTLE AXONS.

SO IF YOU THINK ABOUT THE LITTLE TELEPHONE THAT YOU USED TO MAKE AS A KID WITH THE CUP AND THE STRING, IT IS NOT THE CUP, IT'S THE STRING.

THE STRING IS NOT WORKING.

SO THE CUP IS WORKING FINE BUT SOMETHING IS GOING AWRY WITH THE STRING.

CAN'T MAKE THAT CONNECTION FROM ONE TO THE NEXT.

WE DON'T KNOW WHAT'S WRONG WITH THE STRING.

THERE ARE LOTS OF DIFFERENT PARTS TO THAT AXON, THAT STRING. THERE IS A NICE LAYER OF CELLS THAT GO AROUND IT AND HELPS TO INSULATE IT.

THAT INSULATION ACTUALLY HELPS MAKE THAT SIGNAL MUCH FASTER.

IF YOU DON'T HAVE THE CELLS AROUND THE AXON TO HELP INSULATE IT, THE SIGNAL'S EXTREMELY SLOW.

JUST LIKE INSULATION IN YOUR HOUSE, THAT INSULATION KEEPS THE ELECTRICAL SIGNAL WITHIN THAT STRING, OTHERWISE ALL THE SIGNAL JUST LEAKS OUT EVERYWHERE.

SO IT GOES VERY, VERY SLOWLY AND A LOT OF THAT JUST KIND OF LEAKS OUT.

THAT MIGHT BE ONE PROBLEM.

AND THAT'S WHAT DEMYELINATION IS.

SO THE MYELIN IS MADE BY THE NERVES – IT'S MADE BY THE CELLS THAT GO AROUND THE AXON AND THAT IS THE INSULATION.

THAT'S ONE POTENTIAL PROBLEM IS THAT YOU DON'T HAVE THE INSULATION AND SO THEREFORE YOU'RE LOSING SIGNAL, IT IS GOING REALLY SLOW, IT IS NOT ABLE TO TRAVEL WELL.

THERE COULD ACTUALLY BE A PROBLEM WITH THE STRING ITSELF. SO WITH THAT AXON, WITH THAT FIBER, SOMETHING COULD BE WRONG.

THE FIBERS DON'T ACTUALLY CONNECT TO EACH OTHER.

THERE IS ACTUALLY – IT'S – THOSE FIBERS AREN'T SO MUCH LIKE HAIR IN ONE CONTINUES BUT THERE ARE ACTUALLY LOTS OF LITTLE ELEMENTS THAT MAKE UP THAT AXON THAT MAKE UP THAT FIBER. SO THAT COULD BE THE PROBLEM.

AND THEN FINALLY IT COULD BE BOTH.

YOU COULD HAVE SOMETHING WRONG BOTH WITH THE INSULATION AND WITH THE FIBER ITSELF.

AND AGAIN, THE PROBLEM WITH ALL OF THIS IS THAT WE HAVE NO WAY OF DETECTING WITHOUT HAVING THE PERSON BE DEAD AND HAVE DONATED THEIR BRAIN AND TEMPORAL BONE TO FIND OUT. THERE IS NO WAY TO TEST THIS IN A LIVE PERSON.

AND DYSYNCHRONY, THE OTHER NAME FOR THIS, IS THE FACT THAT WHEN THOSE NERVES FIRE, THEY HAVE TO FIRE IN A VERY SPECIFIC PATTERN.

AGAIN, IT'S VERY MUCH LIKE PLAYING TELEPHONE.

YOU KNOW HOW IF YOU START IN A BIG GROUP ONE PERSON SAYS SOMETHING AND BY THE TIME THE 30th PERSON GETS IT, IT DOESN'T HAVE ANYTHING TO DO WITH WHAT WAS ORIGINALLY SAID.

THAT'S BECAUSE THOSE SIGNALS GOT ALL MIXED UP SOMEWHERE IN THERE.

THE FIRING WAS ALL WRONG AND IT WASN'T A NICE, CONTINUOUS SIGNAL IN THE CORRECT PATTERN SO THAT YOUR BRAIN AND ALL THE OTHER CELLS THAT ARE CONNECTED TO IT CAN RECOGNIZE THAT AS WHAT THE CORRECT INPUT IS SUPPOSED TO BE.

AND SO THE GENERAL CRITERIA FOR DIAGNOSING AUDITORY NEUROPATHY IS ONE, YOU NEED TO HAVE AN ESSENTIALLY NORMAL OTOACOUSTIC EMISSION.

AS MANY OF YOU KNOW THAT'S THE NORMAL SOUND AND OUTPUT THAT THE COCHLEAR HAIR CELLS PUT OUT.

THAT'S CRITICAL BECAUSE REMEMBER THIS IS A NERVE PROBLEM, IT IS NOT A COCHLEAR PROBLEM.

THIS TELLS YOU, ONE, THAT THE COCHLEA IS NORMAL.

IT IS WORKING NORMALLY.

THE HAIR CELLS IN THE COCHLEA ARE PUTTING OUT THE SOUNDS, THEY'RE HEALTHY.

SO THAT'S THE SECOND CRITERIA, THEN, IS THAT YOU DON'T HAVE AN AUDITORY BRAINSTEM RESPONSE, WHICH IS THE ACTUAL SIGNAL OF THE NERVE AND THEN ALL THE WAY UP THROUGH THOSE CENTRAL PROCESSING PATHWAYS.

AND THAT'S HOW WE KNOW THAT IT IS NOT A COCHLEAR PROBLEM, IT IS A NERVE OR PROCESSING OR PATHWAY PROBLEM.

AND THEN THE THIRD CRITERIA, AND THIS IS LESS

ELECTROPHYSIOLOGY AND WHAT MORE PEOPLE WILL TELL YOU.

YOU MAY TEST THEIR HEARING AND THEIR HEARING, PER SE, MAY

NOT BE THAT BAD HEARING TONES AND YOU WOULD EXPECT

SOMEONE WHO MAYBE JUST HAS A MILD HEARING LOSS TO BE ABLE

TO UNDERSTAND SPEECH GREAT AS LONG AS IT'S PRESENTED LOUD

ENOUGH TO THEM.

BUT IN PEOPLE WHO HAVE NEUROPATHY OR DYSYNCHRONY WHAT

HAPPENS IS THAT IT DOESN'T MATTER HOW LOUD YOU PRESENT THAT

SOUND TO THEM, ALL THOSE SIGNALS ARE STILL ALL JUMBLED UP AND GARBLED WELL AND NOT UNDERSTAND SPEECH WELL. THEY'LL SCORE A LOT LOWER ON THE SPEECH TESTING. IF IT'S AN ADULT THEY'LL ACTUALLY TELL YOU I CAN HEAR OKAY. I CAN HEAR THE DOOR SLAM AND I CAN HEAR CARS GO BY AND I CAN HEAR SOUND, BUT I DON'T KNOW WHAT PEOPLE ARE SAYING. I HAVE TO ASK THEM TO SAY WHAT – I HAVE TO SAY WHAT ALL THE TIME, THEY HAVE TO REPEAT THINGS AND IT MIGHT NOT EVEN MATTER HOW LOUD THEY SAY IT TO THEM. IT JUST IS REALLY HARD FOR THEM TO UNDERSTAND. AND THEN THE FOURTH THING IS NOT REALLY A CRITERIA BUT IT IS REALLY JUST SOMETHING TO KNOW THAT THERE IS NO CLASSIC PATTERN FOR HEARING LOSS FOR THESE PATIENTS. THEY CAN HAVE A MILD HEARING CONFIGURATION AND MUCH WORSE SPEECH UNDERSTANDING OR THEY CAN HAVE A PROFOUND HEARING LOSS WITH ESSENTIALLY NO SPEECH UNDERSTANDING. BUT YOU CAN'T TELL AND IT IS ACTUALLY NOT ALL THAT PREDICTIVE, NECESSARILY, EITHER. OBVIOUSLY THE MORE HEARING THEY HAVE THE LIKELY THE BETTER THEY'RE GOING TO DO BUT IT ISN'T ALWAYS THAT PREDICTIVE. AND I DON'T EXPECT YOU TO READ THIS LINE AT ALL. THIS IS A SLIDE AND HERE IT GOES THROUGH BASICALLY THAT WHOLE PATHWAY THAT WE WENT THROUGH EARLIER, ALL THE WAY FROM THE HAIR CELLS IN THE COCHLEA ALL THE WAY UP TO THE CORTEX, TO THAT PRIMARY VERY HIGH-PROCESSING CENTER AND THIS OVER HERE ARE ALL POTENTIALLY THE TESTS THAT WE COULD DO TO TRY AND FIGURE IT OUT. BUT AGAIN, MELISSA WILL TALK ABOUT SOME OF THESE TESTS BUT MOST OF THESE TESTS ARE NOT DONE.

THESE ONES THAT ARE UP HERE, YOU KNOW, THIS IS THE AUDITORY BRAINSTEM RESPONSE WAVE FIVE.

WELL, IF YOU DON'T HAVE A GOOD WAVE ONE, YOU AREN'T GOING TO HAVE A WAVE FIVE.

THE PROBLEM IS, IF YOU DON'T HAVE THE FIRST THING THAT'S WORKING, YOU AREN'T EVEN GOING TO KNOW IF THE OTHER FOUR THINGS DOWN THE LINE ARE WORKING OR NOT.

AND THAT'S REALLY STILL THE MAIN PROBLEM.

AND SO AS FAR AS TRYING TO DIAGNOSE PEOPLE, ON PHYSICAL EXAM THEY'LL LOOK TOTALLY NORMAL.

THEY'LL HAVE NORMAL-LOOKING EARS.

THEY DON'T HAVE ANYTHING ABNORMAL WITH THEIR EARS.

THEIR EARDRUMS WILL LOOK NORMAL.

THESE AREN'T THE KIDS WITH LOTS AND LOTS OF EAR INFECTIONS.

IT DOESN'T TELL YOU ONE WAY OR ANOTHER.

AND THEY REALLY HAVE TO MEET THOSE CRITERIA THAT I TALKED ABOUT EARLIER IN ORDER TO REALLY BE ABLE TO SAY YES, YOU'VE GOT A NEUROPATHY.

AND WHAT WE'RE FINDING IS THAT WHEN NEWBORNS ARE SCREENED NOW, MOST NEWBORNS ARE SCREENED ONLY WITH OTOACOUSTIC EMISSIONS ONLY TO SEE IF THE COCHLEA WORKS.

AS WE TALKED ABOUT IN THE CRITERIA THAT CAN BE A PROBLEM BECAUSE IF THEY HAVE NEUROPATHY, YES, THE COCHLEA WORKS GREAT.

THEY'LL PASS THAT OTOACOUSTIC EMISSION AND THOSE WILL BE THE CHILDREN THAT MIGHT COME IN WITH A LATER DIAGNOSIS BECAUSE IT ISN'T UNTIL, GEE, THEY'RE NOT TALKING, BUT MOM AND DAD SAY THEY CAN HEAR ME AND THAT'S TRUE.

THEY CAN HEAR BUT THEY DON'T UNDERSTAND.

THEY'RE NOT GOING TO BE ABLE TO FASHION GOOD SPEECH AND LANGUAGE BECAUSE, AGAIN, THEY CAN'T PROCESS THAT INFORMATION INTO SOMETHING COHERENT.

AND SO THERE HAS BEEN SOME TALK ABOUT DOING NEWBORN HEARING SCREENING WITH BOTH OTOACOUSTIC EMISSIONS AS WELL AS AUDITORY BRAINSTEM RESPONSE.

THAT'S MORE EXPENSIVE AND IT TAKES EXPERTISE.

A LOT OF THE OAEs ARE ALL AUTOMATED AND ANYBODY CAN DO IT, ESSENTIALLY.

THAT'S A PROBLEM.

MOST OF THE CHILDREN THAT ARE IN THE NEONATAL ISUs DO GET SCREENED WITH AUDITORY BRAINSTEM RESPONSE AND THEY'LL BE PICKED UP.

THOSE ARE THE KIDS AT MUCH HIGHER INCIDENCE.

AND THIS IS WHY.

SO PEOPLE ARE – WHEN THIS WAS FIRST – THIS NEW ENTITY AND NOBODY REALLY KNEW MUCH ABOUT IT OR WHAT THE CRITERIA WERE, PEOPLE USED TO THINK IT WAS EXTREMELY UNCOMMON.

GOSH, HOW CAN IT BE YOUR COCHLEA WORKS BUT THE NERVE DOESN'T WORK.

THAT CAN'T BE RIGHT.

IT'S ALWAYS A COCHLEAR PROBLEM.

AS PEOPLE HAVE STARTED BECOMING MORE AWARE OF THIS AND TESTING KIDS WITH BOTH TESTS, THEY FIND THAT THE PREVALENCE MAY ACTUALLY BE UP TO 8% OF CHILDREN WHO HAVE HEARING LOSS WHO ARE SCREENED FOR HEARING LOSS AND ACTUALLY FOR THE KIDS THAT ARE ACTUALLY CONFIRMED TO HAVE A PERMANENT HEARING LOSS IT COULD BE AS HIGH AS 10% 11% WHICH IS ACTUALLY A LOT OF KIDS, IF YOU THINK ABOUT IT.

AND AS I SAID, THOSE KIDS IN THE NEONATAL ICU WITH THE HIGH RISK FACTORS THEIR RATE ACTUALLY REALLY GOES UP.

AND SO THOSE – THAT’S PARTLY WHY MOST OF THE BABIES THAT NEED TO GO TO THE INTENSIVE CARE UNIT ARE SCREENED WITH AUDITOR BRAINSTEM RESPONSE AND NOT JUST WITH OAE BECAUSE HYPOXIA OR LACK OF OXYGEN AND TOO MUCH BILIRUBIN, THE YELLOW BABIES WHERE YOU HAVE TO GO UNDER THE LIGHT THOSE ARE TWO RISK FACTORS ACTUALLY FOR NEUROPATHY.

SO TREATMENT, OBVIOUSLY CLOSE MONITORING.

WE ALWAYS RECOMMEND THAT FOR ANY HEARING LOSS, THOUGH. ESPECIALLY FOR NEUROPATHY BECAUSE IT MAY CHANGE OVER TIME AND THAT’S REALLY UNKNOWN.

WE DON’T KNOW HOW THESE KIDS ARE GOING TO DO OVER TIME. ESPECIALLY IF THEY’RE DIAGNOSED AS A NEWBORN, THERE HAVE BEEN SOME REPORTS, BECAUSE NEWBORNS ESPECIALLY IF THEY WERE IN THE INTENSIVE CARE UNIT OR IF THEY’RE PREMATURE IS THAT THEIR AUDITORY PATHWAY, THEIR AUDITORY CORTEX IS STILL NOT DEVELOPED.

THEY CAME OUT TOO EARLY.

WHEN YOU FIRST SCREEN THEM THEY MAY LOOK LIKE A NEUROPATHY BABY BECAUSE EVERYTHING IS TOO IMMATURE.

IT ISN’T ALL THERE YET.

SOME PEOPLE WILL SAY THAT THESE ARE KIDS THAT YOU MAYBE DON’T WANT TO IMPLANT ASAP ESPECIALLY IF THEY’RE PREMATURE AND HAD OTHER BRAIN DEVELOPMENTAL ISSUES.

YOU MAYBE WANT TO GIVE THEM A LITTLE BIT OF A CHANCE TO MATURE TO SEE IF SOME OF THAT CORTEX AND SOME OF THOSE PATHWAYS WILL CONTINUE TO MATURE.

SO THAT GOES A LITTLE BIT MAYBE AGAINST THE GRAIN OF WHAT WE USUALLY SAY ABOUT COCHLEAR IMPLANTS SAYING WE WANT TO GET IT IN AS SOON AS POSSIBLE.

BUT AGAIN THERE MAY BE SOME CIRCUMSTANCES IN WHICH YOU'D SAY WELL, LET'S SEE AND LET'S MAKE SURE ESPECIALLY IF THIS CHILD IS DELAYED IN OTHER WAYS.

YOU KNOW, MOTOR, THINGS LIKE THAT, YOU MIGHT WANT TO SAY WELL, LET'S GIVE IT ANOTHER FEW MONTHS AND SEE IF THEY CATCH UP OR THEY CAN DEVELOP A LITTLE BIT MORE.

AND JUST CONTINUE TO TEST THEM TO SEE IF IT GETS BETTER.

SPEECH AND LANGUAGE THERAPY OBVIOUSLY IS REALLY CRUCIAL.

AND THEN FINALLY FOR THE CHILDREN WHO DO HAVE THE

OBVIOUSLY IF THEY HAVE A PROFOUND HEARING LOSS AND A

TOTALLY ABSENT AUDITORY BRAINSTEM RESPONSE, THOSE ARE THE

KIDS YOU'LL START THINKING ABOUT COCHLEAR IMPLANTATION

FOR.

I ALREADY GAVE YOU ONE CAVEAT.

YOU WANT TO LONG DISCUSSION WITH MOM AND DAD ABOUT THERE

ARE SOME STUDIES THAT SAY THAT CHILDREN WITH AUDITORY

NEUROPATHY DO PRETTY MUCH THE SAME AS THE OTHER – AS ANY

OTHER CHILD BUT THERE ARE PLENTY OF OTHER STUDIES THAT SAY

THAT CHILDREN WITH AUDITORY NEUROPATHY HAVE A MUCH

HARDER TIME, EVEN WITH AN IMPLANT.

AND SO – IT'S VARIABLE AND THERE IS NO WAY TO KNOW.

WE DON'T HAVE ANY PREDICTIVE FACTORS.

IT ISN'T LIKE GOSH, IF YOUR ABR LOOKS LIKE THIS YOU'LL DO WELL

AND IF IT LOOKS LIKE THAT YOU'RE NOT GOING TO DO WELL.

THERE IS REALLY NO WAY TO PREDICT THAT.

AND SO I OFTEN WILL TELL MY PARENTS WHO HAVE NEUROPATHY,

YOUR CHILD MAY HAVE TO WORK A LOT HARDER.

IT MAY TAKE THEM LONGER.

AND YOU JUST REALLY HAVE TO CAUTION THEM THAT THEY MIGHT

NOT HAVE THE KIDS WHERE THREE MONTHS LATER THEY'RE DOWN

THE STREET YELLING AND TALKING AND, YOU KNOW, YOU'RE ABLE TO READ BEDTIME STORIES TO.

THERE IS DEFINITELY SOME PRECAUTIONARY THINGS THAT YOU HAVE TO TALK TO MOM AND DAD ABOUT.

FOR THESE CHILDREN, THESE ARE THE KIDS THAT WE LIKELY WOULD GET AN MRI SCAN ON AND NOT A CT SCAN.

BECAUSE AGAIN, WE DON'T THINK THERE IS ANYTHING WRONG WITH THE COCHLEA SO WE DON'T NECESSARILY EXPECT THAT THERE IS GOING TO BE ANY PROBLEMS WITH THE BONE.

THE PROBLEM IS ALL UP HIGHER AND SO WE'RE GOING TO GET THE MRI SCAN TO MAKE SURE THAT THE BRAIN LOOKS NORMAL.

THAT, YOU KNOW, THERE ISN'T SOMETHING ELSE GOING ON.

SO THAT'S ANOTHER DIFFERENCE WITH THE DIAGNOSIS IN THESE CHILDREN AS WELL.

AND I THINK THAT IS PRETTY MUCH ALL I HAVE FOR MY PART.

BARB, ARE WE GOING TO ASK QUESTIONS WITH EACH OR WAIT UNTIL THE END?

>> IF THAT'S OKAY WHY DON'T WE ASK QUESTIONS.

>> ANY QUESTIONS FOR THIS QUICK LITTLE INTRODUCTORY SEGMENT?

>> YOU MENTIONED GENETICS.

ARE THERE SOME TYPE OF GENETIC DISORDERS THAT YOU SEE?

>> NOT THAT WE KNOW OF.

THIS IS SO NEW THAT WE REALLY DON'T KNOW THAT THERE IS ANY CLEAR GENETIC LOW SIDE THAT WE CAN SAY OH, YEAH, IT'S ASSOCIATED WITH NEUROPATHY.

>> ARE THERE ANY DISEASE PROCESSES THAT WITH ADULTS YOU MIGHT GET A NEUROPATHY?

>> ANY DISEASE PROCESS THAT IS GOING TO AFFECT YOUR NERVE BASICALLY, SO ANY DEMYELINATING DISEASES.

THERE ARE SOME AUTOIMMUNE DISEASES THAT CAN DO THAT.

THERE ARE OTHER SYNDROMES THAT CAN BE ASSOCIATED WITH JUST KIND OF GENERAL NERVOUS SYSTEM PROBLEMS AND SO THEY OFTEN WILL HAVE MOTOR ISSUES AND SENSORY ISSUES AND OTHER THINGS THAT ARE ASSOCIATED.

SO IF YOU HAVE ESPECIALLY AN ADULT WHO COMES IN SAYING YOU NEED TO – YOU NEED TO REALLY ASK, YOU KNOW, ANY NUMBNESS, TINGLING IN YOUR FINGERS, ANY WEAKNESS, ARE YOU FEELING KIND OF DISCOORDINATED, ANY TROUBLE SPEAKING?

BECAUSE AGAIN, THAT MIGHT ACTUALLY WARRANT A REFERRAL TO NEUROLOGY TO LOOK FOR SOMETHING THAT IS FULL BODY GOING ON WITH ALL THEIR NERVES AND NOT JUST WITH THE HEARING NERVE.

>> WE HAVE A QUESTION ON THE SCREEN THERE.

THEY ASKED IF YOU COULD REPEAT THE QUESTIONS SO THEY KNOW WHAT IT IS.

>> OKAY.

SO SOMEBODY IS ASKING CAN YOU DETERMINE AUDITORY NEUROPATHY FROM GENETIC TESTING OR DO YOU HAVE TO DO THE MRI?

ACTUALLY, YOU AREN'T GOING TO BE ABLE TO DETERMINE IT FROM EITHER GENETIC TESTING OR MRI.

THE VAST MAJORITY OF THE CHILDREN WHO HAVE AUDITORY NEUROPATHY HAVE NOT A SINGLE THING WRONG ON THE MRI SCAN. IT WILL LOOK COMPLETELY NORMAL.

AND AGAIN, THERE IS NO GENETIC TESTING AS OF YET FOR THIS AND SO YOU REALLY HAVE TO GO BY JUST THE AUDIO METRIC CRITERIA. THERE IS NO OTHER WAY TO DIAGNOSE NEUROPATHY.

>> YOU MENTIONED WAITING A LITTLE BIT OF THE TIME WITH THOSE CHILDREN WHO ARE VERY, VERY YOUNG AND SHOWING SOME POSSIBLE – IS THERE ANY TRENDS THAT HOW LONG BEFORE YOU SEE

SOME OF THAT DEVELOPMENT OR COMING BACK OF THAT EARLY AUDITORY FUNCTION?

>> REPEAT THE QUESTION.

>> THE QUESTION WAS HOW LONG, IF YOU YOU WERE GOING TO WAIT ON A CHILD LET'S SAY WHO WAS PREMATURE AND IN THE ICU AND HAD OTHER DEVELOPMENTAL DELAY ISSUES, I HAVE SEEN SOME PAPERS THAT ADVOCATE FOR 12 MONTHS.

I'VE EVEN SEEN SOMEBODY ADVOCATE FOR 18 MONTHS, WHICH SEEMS LIKE A PRETTY LONG TIME.

I WOULD THINK BY 18 MONTHS POSTNATAL, EVEN IF YOU WERE TWO OR THREE MONTHS PREMATURE, THAT WOULD STILL BE A YEAR AND I WOULD THINK THAT BY THEN FOR SURE ALL OF YOUR CORTEX AND EVERYTHING SHOULD BE PRETTY WELL DEVELOPED.

I HAVE SEEN PEOPLE ADVOCATE FOR UP TO 12 MONTHS.

INSTEAD OF IMPLANTING THEM AT SIX MONTHS, WHICH IS KIND OF THE TREND IS TO TRY AND GET TOWARDS SIX MONTHS OR EARLIER AND EARLIER, TO MAYBE WAIT UNTIL YOU GET TO THAT ONE YEAR MARK AND JUST MAKE SURE THAT THAT TESTING STAYS EXACTLY THE SAME.

NOW, THE PROBLEM IS, IS THAT LET'S SAY THEY GET A LITTLE BETTER.

LET'S SAY THEIR ABR LOOKS A LITTLE BETTER.

THE REAL QUESTION IS, THEY ARE STILL ONE YEAR OLD.

THEY ARE NOT TALKING.

THEY AREN'T DEVELOPING LANGUAGE PROBABLY YET ANYWAY AND YOU STILL THINK THEY HAVE A NEUROPATHY, IS IT – IS IT SO MUCH BETTER THAT THEY ARE GOING TO BE ABLE TO OVERCOME THE FACT THAT THEY CAN HEAR BUT THEY CAN'T PROCESS?

SO I THINK THAT STILL GETS TO BE SOMETHING THAT NEEDS TO BE A JUDGMENT CALL, REALLY.

BECAUSE IF YOU SEE OH, GREAT, I HAVE A LITTLE WAVE FIVE, I HAVE A LITTLE WAVE THREE NOW.

IT'S A JUDGMENT CALL TO SAY IS THAT ENOUGH TO PUT THE HEARING AIDS ON AND KEEP THEM ON FOR ANOTHER YEAR AND, YOU KNOW, AT AGE 2, GEE, THEY AREN'T TALKING.

MAYBE WE SHOULD HAVE DONE THIS A YEAR AGO.

AGAIN, THAT'S VERY MUCH A JUDGMENT CALL AND SOMETHING THAT REALLY, REALLY NEEDS TO BE DISCUSSED WITH MOM AND DAD. AND IN A SENSE KIND OF LET THEM DECIDE AND REALLY FOLLOW THEM CLOSELY OVER THAT TIME PERIOD TO SEE ARE THEY GOING TO DEVELOP SOME LANGUAGE.

CLEARLY, IF THEY'RE NOT, THEN YOU PROBABLY WANT TO GO AHEAD AND START TALKING ABOUT AN IMPLANT OR SOME OTHER TREATMENT.

ANYTHING ELSE?

YEAH.

>> HOW ABOUT IF LATER ON YOU DO NOT SEE THE COCHLEAR – THAT GOES AWAY.

>> THE QUESTION IS WHAT HAPPENS IF YOUR TESTING CHANGES SO YOU DON'T HAVE AN ABR AND YOU HAVE AN OMISSIONS AND COCHLEAR MICROPHONICS AND IT DISAPPEARS.

THEN YOU TREAT THEM AS A PROFOUND LOSS AND NOW YOU'RE NOT GOING TO KNOW DO THEY JUST HAVE THE NEUROPATHY, DO THEY HAVE A COCHLEAR PROBLEM OR DO THEY HAVE BOTH?

AND AT THAT POINT I WOULD SAY OKAY, THEY'VE GOT A PRETTY PROFOUND HEARING LOSS AND THERE IS PROBABLY NO POINT IN WAITING AT THAT POINT.

BECAUSE EVEN IF THEIR ABR STARTED TO IMPROVE, IF NOW THEIR COCHLEA ISN'T WORKING NO END ORGANS ARE GOING TO HELP ANYWAY.

WE'LL GO TO MELISSA NOW.

>> ALL RIGHT.

SO AGAIN MY NAME IS MELISSA, I'M WITH THE AUDIOLOGISTS HERE AT THE UNIVERSITY OF MINNESOTA AND I'M GOING TO GO THROUGH SOME OF THE AUDIOLOGICAL TESTS WE GO THROUGH TO DIAGNOSE SOMEONE WITH DYSYNCHRONY.

AUDITORY NEUROPATHY.

AD OR A MORE RECENT TERM IS AUDITORY NEUROPATHY SPECTRUM DISORDER.

AUDITORY NEUROPATHY WAS FIRST SUGGESTED IN 1986.

THERE ARE 10 INDIVIDUALS WITH DIFFERENT TYPES OF NEUROPATHY. FOR BETTER OR WORSE THAT NAME HAS STUCK THROUGH THE YEARS.

LATER ON AS WE'LL TALK ABOUT THERE WERE SOME OTHER THINGS DR. BERLIN IN 2001 SUGGESTED AUDITORY DYSYNCHRONY BECAUSE THEY FELT LIKE DR. HUANG MENTIONED ABOUT DYSSYNCHRONOUS NERVE FIRINGS OCCURRING.

WHAT IS AUDITORY NEUROPATHY?

PATIENT DISPLAYS AUDITOR – AN ABNORMAL NEURAL FUNCTION AT THE LEVEL OF VIIIth NERVE.

AGAIN, IN 96 WAS THE FIRST TIME WE HEARD THIS TERM.

IT IS NOT A NEW ENTITY AT ALL.

JUST NEW BECAUSE OF THE TESTING THAT WE HAVE TO DIAGNOSE IT. INITIALLY PATIENTS WERE MODERATE PROGRESSIVE HEARING LOSSES AND THEY WERE ABLE TO ATTRIBUTE IT SPECIFICALLY TO THE VIIIth NERVE WHICH IS WHY WE'VE BROADENED THE TERM TO BE DYSYNCHRONY.

FOUND IN CHILDREN AND ADULTS BUT TODAY WE'RE FOCUSING ON CHILDREN.

SO WHY ARE WE SEEING IT SO MUCH MORE?

WHY ARE WE EVEN TALKING ABOUT IT TODAY?

I THINK A BIG REASON IS THE UNIVERSAL NEWBORN HEARING SCREENING, ESPECIALLY THOSE BABIES BORN IN THE TWO STAGE SCREENINGS THEY AREN'T GETTING THE OTOEMISSION TEST THEY'RE GETTING THE OTHER TEST THAT ARE SHOWING ABNORMALITIES.

AUDIOLOGISTS ARE BECOMING MORE FAMILIAR WITH IT AND MANAGING IT IN A LITTLE DIFFERENT WAY.

THAT'S JUST A QUOTE THAT I LIKED IN ONE OF THE PAPERS I READ. IT'S NOT A NEW DISEASE BUT AN ENTITY WHICH IS RECOGNIZED BECAUSE OF IMPROVEMENT IN HEARING ASSESSMENT.

IT'S BEEN AROUND ALL THESE YEARS.

BOOKS AS RECENT AS FIVE OR TEN YEARS AGO DIDN'T MENTION IT IN THERE AND NOW WE HAVE A WHOLE EVENING ON IT.

BRIEFLY AGAIN SOME OF THE SUSPECTED CAUSES OF NEUROPATHY DYSYNCHRONY INCLUDE INNER HAIR CELL LOSS OR DAMAGE.

AUDITORY NERVE DISORDER OR DAMAGE TO THE CONNECTION BETWEEN THE INNER HAIR CELLS AND THE VIIIth NERVE.

SOME RISK FACTORS AND A LOT YOU'LL SEE IN THE BABIES THAT HAD TO SPEND TIME IN THE NICU ARE LACK OF OXYGEN IN THE NEONATAL PERIOD.

HYPOXIA, PREMATUREITY, INFECTIOUS PROCESSES, LOW BIRTH WEIGHT AND THERE ARE SOME GENETIC DISORDERS I WAS READING ABOUT THAT COULD INVOLVE DYSYNCHRONY AS WELL.

BUT THEN AGAIN SOME HAVE NO RISK FACTORS AT ALL.

LIST ALL THE RISK FACTORS AND YOU CAN HAVE AN OTHERWISE NORMAL, HEALTHY-APPEARING CHILD THAT HAS DYSYNCHRONY.

THE INCIDENCE IS REALLY UNKNOWN BECAUSE WE'RE JUST IN THE LAST 20 YEARS STARTING TO DIAGNOSE THIS MORE AND MORE.

THE AVERAGE IS ANYWHERE FROM 7 TO 10 OF THOSE DIAGNOSIS FROM THE HEARING LOSS BUT THERE IS A RANGE.

BUT SOME OF THE STATISTICS THAT I THOUGHT WERE REALLY STRIKING UP TO 40% OF THE HEARING LOSS DIAGNOSIS IN NICU BABY COULD BE DYSYNCHRONY.

AS I'LL TALK ABOUT LATER, ONE OF THE REALLY IMPORTANT REASONS WHY WE NEED A TWO-STAGE SCREENING ON THOSE NICU BABIES.

FOR MANY OF YOU THIS WILL BE REVIEW BUT THESE ARE THE DIAGNOSTIC TESTS WE USE TO DIAGNOSE THE PROBLEM.

THE AUDITORY BRAINSTEM RESPONSE.

I'LL SHOW YOU A GRAPH VERSUS NORMAL VERSUS ABNORMAL.

USE THE TRANSIENT STIMULUS THAT LOOKS AT THE FREQUENCIES BETWEEN 1 AND 4,000 HERTZ AND CHANGING THE POLARITY.

OTOACOUSTIC EMISSIONS.

AS DR. HUANG MENTIONED IT IS MORE OF A COCHLEA TEST.

IT DOESN'T GO BEYOND.

MIDDLE EAR MUSCLE REFLEXES, PURE TONE THRESHOLDS ARE VARY FROM NORMAL, MILD ALL THE WAY TO PROFOUND.

SPEECH RECOGNITION AND NOISE.

A LOT OF THESE INDIVIDUALS INITIALLY WERE DIAGNOSED AS HAVING CENTRAL AUDITORY PROCESSING DISORDER BECAUSE THERE WAS SUCH A DISPARITY IN WHAT YOU WOULD SEE IN THEIR THRESHOLDS VERSUS WHAT YOU WERE SEEING ON THE AUDIOGRAM. SO THE ABR TEST WILL BE ABNORMAL WITH ONLY THE PRESENCE OF A COCHLEAR MICROPHONIC.

I KNOW THOSE OF YOU AT HOME TELL ME YOU CAN'T SEE ME MOVING THE CURSOR.

IF YOU LOOK AT THE BEGINNING OF THE AXIS SEE THE SQUIGGLY LINES?

IT WILL REVERSE WHEN YOU CHANGE THE POLARITY FROM NEGATIVE TO POSITIVE.

THIS IS HARD TO SEE BUT IT ACTUALLY FLIPS AROUND THERE.

OTOACOUSTIC EMISSIONS WOULD BE PRESENT SO AGAIN WHY SOME BABIES ARE MISSED INITIALLY IS THE TEST THAT IS USED MORE SO WITH THE NEWBORN HEARING SCREENING.

MIDDLE EAR MUSCLE REFLEXES CAN BE ABSENT IN THE OTHERWISE NORMAL MIDDLE EAR FUNCTION AND I'VE SEEN NORMAL, MILD, SLOPING THAT LOOKS JUST LIKE A STANDARD YOUR SENSORY NEURAL HEARING LOSS.

SPEECH RECOGNITION AND NOISE IS QUITE POOR.

SO THESE LAST FEW ARE HARD WITH KIDS, I THINK, THOUGH BECAUSE IT'S HARD TO TEST KIDS SPEECH AND NOISE.

KNOWING ALL THAT IT REMINDS YOU WHY IT'S IMPORTANT IF THERE IS A QUESTION TO DO THAT ABR TEST.

THIS IS JUST A NORMAL – LET'S GO BACK HERE.

WE SEE THE COCHLEAR MICROPHONICS AND NOTHING REPEATABLE AFTER THAT.

WHAT YOU LIKE TO SEE IS THIS BEAUTIFUL WAVE THAT REPEAT NO MATTER WHAT THE POLARITY LOOKS LIKE.

IMPORTANT WHY YOU NEED A BATTERY OF TESTS, NOT JUST ONE TEST.

NOT JUST THE ABR OR THE OEAs.

SO WHAT DO WE DO WHEN WE DECIDE IT'S NOT SENSORINEURAL HEARING LOSS AND IT'S DYSYNCHRONY.

SOME OF THE RECOMMENDATIONS HAVE CHANGED BUT SOME IN THE 90s ARE APPLICABLE TODAY AND IT'S CASE BY CASE.

THE MAIN RECOMMENDATIONS INCLUDED AMERICAN SIGN LANGUAGE AND CUEING BECAUSE THEY REALLY THOUGHT IT WAS DUE TO IMPAIRED TEMPORAL PROCESSING AND THAT IT WAS A NERVE DISORDER SO THAT SOME OTHER INTERVENTION SUCH AS A COCHLEAR IMPLANT WOULDN'T HELP.

AUDIOLOGISTS BEGAN EXPERIMENTING WITH LOW GAIN HEARING AIDS IN ONE EAR.

SOME DID NO AMPLIFICATION AT ALL, SINCE THERE WAS PRESENT OUTER HAIR CELL FUNCTION YOU DIDN'T WANT TO DESTROY THAT RESIDUAL HEARING THAT PATIENTS HAD.

BUT MOST ALL THE SUGGESTIONS I THINK TODAY STILL APPLIES WITH AN FM SYSTEM BECAUSE OF THE POOR PERFORMANCE IN NOISE. THE PROBLEM – NOT NECESSARILY PROBLEM BUT ONE OF THE ISSUES WITH TRADITIONAL AMPLIFICATION IT WASN'T AM MRI FILING THE SPEECH SOUNDS.

WE WEREN'T SURE WHAT WAS GETTING THROUGH THE AUDITORY PATHWAY SO THERE WAS A FEW CENTERS OUT THERE IN THE LATE 90s WHO BEGAN EXPERIMENTING WITH COCHLEAR IMPLANTS.

SOME OF THE STUDY INITIALLY, THOUGH, SUGGESTED THAT IT WASN'T SOMETHING THAT WAS BENEFICIAL.

THEY WENT BACK AND LOOKED AGAIN AND FOUND THAT THE ELECTRICAL ABR THEY DID AFTER THE COCHLEAR IMPLANT SURGERY LOOKED QUITE SYNCHRONOUS AND STUDIES STARTED SHOWING THAT MAYBE IT WAS PROVIDING SOME BENEFIT.

PUT THIS IN AGAIN BUT COMMUNICATION MODALITIES, SPOKEN LANGUAGE, ASL, CUEING.

IT REALLY IS A CASE BY CASE BASIS.

I THINK THAT YOU WANT TO TRY TRADITIONAL AMPLIFICATION AND SEE WHAT HAPPENS WITH THEIR PROGRESS.

I THINK THAT'S REALLY IMPORTANT.

I THINK IT'S REALLY IMPORTANT TO WORK AS A TEAM.

TO WORK WITH YOUR EAR, NOSE AND THROAT POSITION AND WORK WITH THE SCHOOL SYSTEM TO MAKE SURE THEY'RE PROGRESSING IN THEIR SPEECH/LANGUAGE MILESTONES AND DECIDE IF YOU ARE BEGINNING WITH SPOKEN LANGUAGE, IF THAT'S SOMETHING THAT NEEDS TO CONTINUE, IF THAT'S NOT PROGRESSING, IF YOU NEED TO TRY A DIFFERENT COMMUNICATION MODALITY.

FM SYSTEM FOR NOISIER SITUATIONS.

THEN A COCHLEAR IMPLANT ONLY IF THERE IS LACK OF PROGRESS WITH TRADITIONAL APPLICATION.

WE HAVE SEEN KIDS WHO HAVE DONE WELL WITH HEARING AIDS. IT IS NOT NECESSARILY A GIVEN THAT THEY WON'T PERFORM OR WON'T DEVELOP SPEECH AND LANGUAGE.

ONE OF THE NAMES IS A SPECTRUM.

THERE IS SUCH A WIDE RANGE OF VARIABLE PERFORMANCES WITH IT.

I WOULD SAY IF YOU START WITH HEARING AIDS IN GENERAL, A LOT OF THE KIDS ARE TINY BUT YOU WANT TO FIT IT BASED ON THE AUDIOGRAM.

YOU DON'T WANT A PROGRAM THAT BASED OFF THE ABR MOST OF THE TIME YOU'LL GET NO RESPONSE AND YOU DON'T WANT TO GIVE THEM THAT PROFOUND LOSS AND HEARING AIDS.

HOW DO WE DECIDE WHAT INTERVENTION IS BEST?

AGAIN, BY A TEAM APPROACH.

SO DECIDE BASED ON THE DEVELOPMENTAL LEVEL OF THE CHILD, CONSISTENCY OF AMPLIFICATION USE AND YOUR CLINICAL JUDGMENT.

I JUST PUT IN A SLIDE HERE JUST TALKING ABOUT WHY COCHLEAR IMPLANT MIGHT BE AN OPTION.

IF THE SITE OF LESION, AN IMPASSE CAN DIRECTLY STIMULATE THE AUDITORY NERVE.

LIKE I SAID BEFORE, SOME OF THE ELECTRIC ABRs DONE IN THE OPERATING ROOM DURING THE COCHLEAR IMPLANT SURGERY HAVE SUGGESTED THAT THAT'S HAPPENING.

SO IF THE SITE OF LESION IS THE AUDITORY NERVE.

AUDITORY SYNCHRONIZATION IS – EXPECTATIONS SHOULD BE SET MORE REALISTICALLY BECAUSE THERE IS SUCH A WIDE RANGE OF PERFORMANCE WITH THESE KIDS.

SO WHAT ARE THE OUTCOMES?

I KEEP USING THE WORD VARIABLE AND SPECTRUM BUT IT'S VARIABLE.

THERE IS A SMALL CHANCE THAT AUDITORY FUNCTION CAN IMPROVE IN THE FIRST ONE TO TWO YEARS AND IT IS ANOTHER REASON WE WATCH THE SPEECH/LANGUAGE MILESTONES SO CLOSELY.

SOME STUDIES SUGGEST THAT LESS THAN 10% CAN DEVELOP NORMAL SPEECH AND LANGUAGE.

THAT MIGHT SEEM DIRE BUT SOME STUDIES CONSIDERING THE DELAYS IN DIAGNOSIS AND NEEDING TO CATCH UP AND REALLY SOME OF THOSE STUDIES ARE SIMILAR TO THOSE WITH SENSORINEURAL HEARING LOSS.

RECEPTIVE LANGUAGE OBVIOUSLY IS PROBABLY GOING TO BE DELAYED COMPARED TO NORMAL HEARING BUT SIMILAR TO MATCH GROUPS WITH SENSORINEURAL HEARING LOSS.

MORE AND MORE WE'RE SEEING MORE STUDIES IN THE LAST DECADE ON COCHLEAR IMPLANTS AND AUDIO DYSYNCHRONY SO WE'LL BE ABLE TO GAUGE THAT BETTER AS MORE STUDIES BECOME AVAILABLE.

THE SPEECH PRODUCTION IS VARIABLE THOUGH IT'S INTELLIGIBLE.

I THINK A LOT OF TIMES PEOPLE SAY WHAT ARE THEY HEARING?

WHAT ARE WE AMPLIFYING WITH THE HEARING AIDS?

IT IS SOUND OR NOISE AND SPEECH FOR THE MOST PART BECOMES INTELLIGIBLE.

THESE ARE MY TAKE HOME MESSAGES FOR YOU.

THE IMPORTANCE OF NEWBORN HEARING SCREENING, MULTIPLE TESTING IN NICU BABIES.

THE STATISTIC I READ WAS UP TO 40% OF THOSE DIAGNOSED WITH HEARING LOSS HAVE DYSYNCHRONY.

THE IMPORTANCE OF A TEAM EVALUATION.

THE SPEECH PATHOLOGIST, ENT, GENETICIST AND PEDIATRICIAN.

DON'T BE AFRAID TO TRY AMPLIFICATION OR BASED ON THE PARENTS' PREFERENCE.

IF THEY WANT TO HELP THE CHILD DEVELOP SPOKEN LANGUAGE THAT'S SOMETHING YOU CAN START WITH.

GET BEHAVIORAL INFORMATION AND ORDER THE HEARING AIDS IF YOU CAN AND MONITOR SPEECH AND LANGUAGE CLOSELY.

IF THERE IS A LACK OF BENEFIT COCHLEAR IMPLANTS ARE A POSSIBILITY.

THEY'RE NOT ALWAYS A GIVEN, THOUGH, SO THAT'S WHY YOU HAVE YOUR TEAM APPROACH.

THESE ARE ALL MY REFERENCES THAT ARE VERY TINY AND YOU CAN'T READ.

I CAN EMAIL THEM OUT TO YOU.

ANY QUESTIONS?

OKAY.

THANK YOU.

>> YOU MENTIONED THAT INTEROPERATIVELY THEY'VE BEEN FINDING THE ELECTRICAL STIMULATION OR ELECTRICAL ABR DOES IMPROVE IN SOME CASES.

DO THEY HAVE STUDIES THAT THOSE ARE THE ONES THAT DO BETTER WITH THE IMPLANTS?

DO WE HAVE A SOMEWHAT EARLY PREDICTOR OF SUCCESS?

>> -- IS THE ONLY ONE SO FAR FOR IMPLANT.

>> MY QUESTION IS, AT THE OPERATIVE STAGE YOU CAN PREDICT THIS PERSON MIGHT REALLY SUCCEED WITH THEIR IMPLANT AS OPPOSED TO ITS NOT WORKING WELL.

>> AS A GENERAL I WOULD SAY IT'S A PREDICTOR OF GOOD PERFORMANCE WHO GETS GOOD SPEECH RECOGNITION ALL THE TIME BUT IT IS SUGGESTIVE OF SOME BENEFIT.

>> OKAY.

>> CAN YOU REPEAT A LITTLE BIT OF WHAT WAS JUST SAID OVER HERE FOR THOSE THAT ARE LISTENING?

>> HE WAS ASKING IF THE ELECTRIC ABR DONE IN THE OPERATING ROOM IS A PREDICTOR OF PERFORMANCE LATER ON.

>> THERE IS A QUESTION.

>> IS THERE ONE ABOVE THAT?

>> JUST THAT ONE.

>> SOMETIMES THE EMISSIONS WHICH ARE A TEST OF OUTER HAIR CELL FUNCTION SOMETIMES THOSE GO AWAY NATURALLY WITH THOSE IN AUDITORY DYSYNCHRONY.

THEY END UP BEING ABSENT ANYWAY AND THAT'S WHY DR. HUANG WAS SAYING IT LOOKS MORE LIKE A SENSORY HEARING NEURAL LOSS AND IT MIGHT BE EASIER TO MAKE A DECISION ON WHETHER A COCHLEAR IMPLANT.

>> SOME PEOPLE DON'T KNOW OTOACOUSTIC EMISSIONS.

EXPLAIN THAT PROCESS.

>> IN OUR COCHLEA WE HAVE HAIR CELLS.

OUTER HAIR CELLS ARE SOMETHING THAT BOOST THE SOUND AND TO HELP THE NERVE FIRE OR TO HELP THOSE OTHER HAIR CELLS FIRE THE NERVE AND SO WHAT THE OTOACOUSTIC EMISSION DOES IT SENDS A SOFT LEVEL SOUND INTO THE COCHLEA AND THE OUTER HAIR CELLS HAVE AN ECHO THAT RESPONDS BACK.

IN AUDITORY DYSYNCHRONY SINCE THE SITE OF LESION IN BEYOND THAT THE HAIR CELLS ARE STILL SENDING AN ECHO BACK.

THERE IS A QUESTION IS IT POSSIBLE TO HAVE SENSORINEURAL HEARING LOSS IN ONE EAR AND AUDIO DYSYNCHRONY IN THE OTHER? IT'S POSSIBLE.

WE'VE SEEN IT QUITE RECENTLY.

IT IS RARE BUT, YEAH, IT IS.

IT'S AN INTERESTING PHENOMENON.

ANY OTHER QUESTIONS?

>> DR. HUANG IS THERE ANYTHING IN A PREOPERATIVE MRI – I SEE YOU SHAKING YOUR HEAD NO.

THAT WOULD INDICATE THE SITE OF LESION IS HIGHER OR MIGHT BE CONTAINED TO THE BRAINSTEM OR NOT?

>> THE QUESTION WAS IS THERE ANYTHING THAT SHOWS UP ON THE MRI PREOPERATIVELY?

>> NOT UNLESS THERE IS AN ACTUAL LESION THAT YOU CAN SEE ON THE MRI.

I MEAN, A LOT OF THE CHILDREN, ESPECIALLY THE ONES THAT NEEDED INTENSIVE CARE UNIT STAYS HAVE BRAIN ABNORMALITIES. THEY HAVE UNDERDEVELOPED BRAINS OR THEY HAVE PARTS OF THEIR BRAIN THAT ARE UNDEVELOPED AND AGAIN, THOSE ARE THE KIDS THAT ARE OFTEN GOING TO HAVE MORE GLOBAL PROBLEMS AND NOT JUST THAT SO—

>> CAN YOU REPEAT THAT?

>> YOU'LL HAVE TO REPEAT IT.

>> JUST VERBALLY.

>> DO YOU WANT TO COME BACK TO THE MIKE?

>> I'LL JUST SIT IN THIS CHAIR.

I JUST WANTED TO SAY THAT NO, THERE IS NO PARTICULAR SITE OF LESION.

WE KNOW WHERE THE PATHWAY RUNS AND WE KNOW WHERE THE BRAINSTEM NUCLEI ARE.

AND ROUGHLY – AND WHERE THE AUDITORY CORTEX IS, EVERYTHING.

BUT THE KIDS THAT ARE AGAIN IN THE INTENSIVE CARE UNIT, THEY OFTEN WILL HAVE ABNORMAL MRI SCANS.

THEY'LL HAVE PROBLEMS WITH MYELINATION, THEY'LL HAVE UNDERDEVELOPED PARTS OF THEIR BRAIN AND AGAIN LIKE I SAID, THOSE ARE KIDS THAT OFTEN HAVE MORE PROBLEMS THAN JUST THE HEARING LOSS.

BUT CLEARLY IF THEY HAVE WHAT LOOKS LIKE A DYSYNCHRONY ON TESTING, THAT IT'S SOMETHING THAT YOU'LL KNOW AND ON THOSE CHILDREN ACTUALLY SOMETIMES THOSE ARE THE KIDS THAT WE LOOK VERY, VERY CLOSELY TO MAKE SURE THEY EVEN HAVE A COCHLEAR NERVE.

CHILDREN WHO HAVE A VERY ABNORMAL MRI SCAN OR UNDERDEVELOPED MRI SCAN THERE ARE CHILDREN WHO HAVE NERVE AGENESIS, THEY HAVE NO NERVE WHATSOEVER. SO THOSE KIDS ARE CLEARLY NOT GOING TO BENEFIT FROM A COCHLEAR IMPLANT.

THOSE ARE THE CHILDREN THAT, YOU KNOW, UNFORTUNATELY AT LEAST IN THIS COUNTRY ARE GOING TO NEED TO LEARN SIGN. AND – BECAUSE AUDITORY BRAINSTEM IMPLANT ISN'T AVAILABLE HERE IN THIS COUNTRY.

UNLESS THEY WANT TO GO TO ITALY, THAT'S UNFORTUNATELY THE ONLY OPTION WE HAVE FOR THEM.

>> MY NAME IS LAURA NYHUS.

I'M GOING TO TALK BRIEFLY ABOUT THE SPEECH AND LANGUAGE CONSIDERATIONS THAT WE TAKE WITH CHILDREN WHO HAVE AUDITORY NEUROPATHY.

A FEW OF THE AREAS THAT I'LL TALK ABOUT ARE HOW EARLY INTERVENTION IS IMPORTANT.

THERE ARE DIFFERENT CONSIDERATIONS FOR SPEECH AND LANGUAGE.

HOW TO KIND OF DEVELOP A GREATER AWARENESS OF SOUND AND WHAT YOUR CHILD IS HEARING ON A DAY-TO-DAY BASIS.

AND DIFFERENT COMMUNICATION SYSTEMS AND MODALITIES.

INFANTS AND CHILDREN WHO ARE DIAGNOSED WITH AUDITORY NEUROPATHY WILL OR SHOULD HAVE CONSISTENT EARLY INTERVENTION.

IT'S IMPORTANT TO SEEK OUT THE SPECIALISTS WHO ARE – IF CAN WHO ARE FAMILIAR WITH CHILDREN WITH HEARING LOSS AND FIND SOMEONE FAMILIAR WITH AUDITORY NEUROPATHY THAT'S TYPICALLY BEST.

TEAM MEMBERS MIGHT INCLUDE YOUR AUDIOLOGIST, YOUR SPEECH PATHOLOGIST, PEOPLE AT SCHOOL, YOUR TEACHERS, ENTs, NEUROLOGISTS AND JUST DIFFERENT PARENT ADVISORS OR PARENT ADVOCATES THAT YOU HAVE.

SINCE YOU AS A PARENT OUT THERE, SINCE YOU'RE A MEMBER OF THE TEAM, IT'S VERY IMPORTANT YOU TO SHARE YOUR INSIGHTS AND OBSERVATIONS OF THE MEMBERS OF THE TEAM.

YOU KNOW YOUR CHILD THE BEST.

WE CAN GET A LOT OF INFORMATION FROM TESTS AND THINGS BUT YOU KNOW YOUR CHILD THE BEST AND HOW THEY ACT ON A DAY-TO-DAY.

AND THAT WILL HELP THE TEAM MEMBERS GET A BETTER PICTURE OF YOUR CHILD SINCE WE SEE THEM FOR A QUICK SNAPSHOT OF TIME WHEN THEY'RE IN FOR TESTING OR WHEN THEY'RE IN FOR SPEECH. SO IF YOU – YOUR INSIGHTS AND OBSERVATIONS ARE REALLY IMPORTANT.

SO I JUST WANTED TO HAVE MY OWN SEPARATE SLIDE ON THAT.

I THINK THAT'S SOMETIMES IT'S HARD FOR FAMILIES TO REALIZE THAT YOU ARE A MEMBER OF THE TEAM AND THAT YOU DO KNOW YOUR CHILD THE BEST.

DIFFERENT SPEECH AND LANGUAGE CONSIDERATIONS.

ONE THING THAT IS IMPORTANT IS THAT COMMUNICATION IS MORE THAN JUST TALKING AND WITH THESE LITTLE KIDS THAT ARE DIAGNOSED WITH AUDITORY NEUROPATHY WE WANT TO MAKE SURE THEY'RE MEETING THEIR DEVELOPMENTAL MILESTONES.

NOT JUST SAYING THINGS TO US BUT THEIR GESTURES, HOW THEY INTERACT WITH OTHERS, THEIR ATTACHMENT TO THEIR FAMILY.

THEIR SOCIAL SKILLS WITH FAMILIAR AND UNFAMILIAR PEOPLE. PLAY SKILLS, LANGUAGE COMPREHENSION, WHAT THEY UNDERSTAND AND THEIR LANGUAGE EXPRESSION AND WHAT THEY'RE SAYING TO US.

SO IT IS IMPORTANT TO FOLLOW UP CONSISTENTLY WITH AN SLP TO MAKE SURE THAT THESE MILESTONES ARE BEING MET.

IF YOUR CHILD IS NOT BEING SEEN ON A WEEKLY BASIS, WHEN THEY'RE STILL YOUNG AND KIND OF GETTING THE FINE-TUNING OF THE HEARING AIDS I WOULD RECOMMEND THAT THEY BEING SEEN AT LEAST ONCE A MONTH TO MAKE SURE THAT MONTHLY THEY'RE KEEPING UP WITH THOSE MILESTONES.

DEVELOPING A BETTER AWARENESS OF SOUND AND WAYS TO MONITOR YOUR CHILD'S AWARENESS OF SOUND ON A DAILY BASIS. SOME – IN SOME OF THE LITERATURE IT RECOMMENDED, YOU KNOW, DOES YOUR CHILD CONSISTENTLY RESPOND TO SOUNDS DAY-TO-DAY? ARE THEY RESPONDING TO YOUR FAMILY MEMBERS' VOICE, THE TELEPHONE RINGING, MUSIC PLAYING, YOUR DOG BARKING? ARE THEY CONSISTENTLY SOMETHING THEY'RE DETECTING OR RESPONDING TO?

WHEN THEY HEARD THESE SOUNDS WAS THERE A VISUAL CUE OR WAS IT JUST THE SOUND ON ITS OWN?

ARE THE SOUNDS TYPICALLY LOUDER SOUNDS OR SOFT SOUNDS? THESE ARE ALL THINGS JUST AT HOME YOU CAN DO ON YOUR OWN. WHEN THEY COME IN FOR TESTING YOU'LL GET MORE ACCURATE NUMBERS BUT THIS IS SOMETHING AS A PARENT YOU CAN CALL IN AND SAY TO YOUR AUDIOLOGIST THEY HAVEN'T BEEN HEARING THE DOORBELL RING FOR A COUPLE OF WEEKS.

I FEEL LIKE SOMETHING MIGHT BE CHANGING.

THIS KIND OF THE SAME THING.

IT MIGHT BE USEFUL TO – WITH THIS INFORMATION IT'S USEFUL FOR ALL THE PEOPLE INVOLVED.

SPEECH PATHOLOGIST, AUDIOLOGIST AND TEACHERS SO AS A TEAM WHEN WE'RE WORKING WITH YOUR CHILD WE CAN ALL ADDRESS IT THE SAME WAY.

A LOT OF PARENTS LIKE TO KEEP A NOTEBOOK OF OBSERVATIONS FROM DAY-TO-DAY OR WEEK TO WEEK OF WHAT THEY NOTICED THEIR CHILD DOING.

IF THEY NOTICE SOMETHING NEW THEY HEAR.

A PARENT NOTICED HER SON HEARING THE DOG DRINKING WATER. SOMETHING THEY NEVER NOTICED BEFORE.

THOSE THINGS ARE FUN TO MONITOR PROGRESS OR TO MONITOR REGRESSION IF SOMETHING IS CHANGING.

MANY RESEARCHERS SAY THAT A VISUAL COMMUNICATION SYSTEM SHOULD BE INCLUDED IN EARLY PROGRAMMING FOR CHILDREN DIAGNOSED WITH AUDIO NEUROPATHY.

THIS IS DEBATABLE WITH DIFFERENT GROUPS BECAUSE SOME PEOPLE BELIEVE THE VISUAL COMMUNICATION MIGHT BE A NEGATIVE INFLUENCE TO SPEECH DEVELOPMENT.

THE MAIN THING WITH THIS IS THAT COMMUNICATION SYSTEMS CAN BE CONTROVERSIAL BUT THEY'RE INDIVIDUAL.

IT'S WHAT WORKS BEST FOR YOU, YOUR CHILD AND YOUR FAMILY.

SO THAT'S JUST THE MAIN THING TO KEEP IN MIND.

SO WHEN PICKING A DIFFERENT – IF YOU'RE GOING TO USE A DIFFERENT COMMUNICATION SYSTEM IT'S IMPORTANT TO CHOOSE SOMETHING THAT IS BASED ON THE FAMILY'S ABILITIES AND THEIR GOALS.

YOU KNOW, IF YOU DON'T HAVE THE ABILITY TO LEARN SIGN LANGUAGE, THEN THAT'S NOT THE RIGHT CHOICE FOR YOU AND THAT'S FINE.

YOU ALSO HAVE TO CONSIDER THE ABILITIES AND PRIORITIES OF YOUR CHILD AND YOUR WHOLE FAMILY.

IF YOUR GOAL IS FOR YOUR CHILD TO BE ABLE TO COMMUNICATE WITH GRANDMA AND GRANDPA AND BROTHER AND SISTER IT IS ALL THOSE KINDS OF THINGS TAKEN INTO ACCOUNT AS WELL AS THE RESOURCES AVAILABLE IN YOUR SCHOOLS AND COMMUNITIES. SOME HERE IN THE CITIES WE HAVE A LOT OF RESOURCES AVAILABLE.

SMALLER AREAS OR OUT FURTHER AWAY IT'S HARD TO FIND ALL THOSE RESOURCES.

AS A PARENT YOU ARE ENTITLED TO THE EDUCATION PLAN YOU FEEL IS MOST APPROPRIATE FOR YOUR CHILD.

I FORGOT TO CHANGE THE SLIDES.

NO, I DIDN'T.

SO MOVING ON TO COMMUNICATION MODALITIES.

THERE ARE FIVE MAIN COMMUNICATION MODALITIES.

THE FIRST TWO AUDITORY ORAL AND AUDITORY VERBAL ARE SOMEWHAT SIMILAR AND MELISSA AND I WERE TALKING THAT THEY'RE KIND OF BECOMING COMBINED TOGETHER AND THEN THERE IS CUE SPEECH, TOTAL COMMUNICATION AND AMERICAN SIGN LANGUAGE.

I'LL JUST BRIEFLY TALK ABOUT EACH ONE.

AUDITORY ORAL FOCUSES ON THE DEVELOPMENT OF SPOKEN LANGUAGE THROUGH LISTENING AND SPEECH READING OR LIP READING.

CHILDREN WEAR HEARING AIDS AND THEY DON'T TYPICALLY USE SIGN LANGUAGE.

THE AUDITORY VERBAL MODALITY EMPHASIZES THE USE OF RESIDUAL HEARING AND PRIMARY MEANS OF DEVELOPING SPOKEN LANGUAGE.

VISUAL CUES ARE TYPICALLY NOT ENCOURAGED IN ORDER TO HELP THIS CHILD DEVELOP THE SPEECH.

AND THIS IS A LOT OF TIMES CLINICALLY BASED.

CUED SPEECH IS A VISUAL CLARIFICATION OF LIP READING THAT IS USED THROUGH A SYSTEM OF EIGHT HAND SHAPES MADE IN FOUR DIFFERENT LOCATIONS NEAR YOUR FACE.

A LOT OF PEOPLE THAT USE THIS SAY IT HELPS WITH READING BETTER, READING SKILLS.

AND TOTAL COMMUNICATION IS JUST KIND OF THAT'S WHAT IT IS, TOTAL COMMUNICATION BASED ON SPOKEN LANGUAGE THAT UTILIZES A WIDE RANGE OF STRATEGIES USING ALL THE STRATEGIES, WHATEVER WE CAN COMMUNICATE, WHICHEVER WAY WE CAN YOU'LL USE ALL OF THOSE.

AND THEN AMERICAN SIGN LANGUAGE IS COMPLETELY VISUAL AND IT IS – THE LANGUAGE OF THE DEAF CULTURE IS CONSIDERED THE NATURAL LANGUAGE OF THE DEAF AND IT – PEOPLE THAT HAVE AMERICAN SIGN LANGUAGE AS THEIR PRIMARY MODE OF COMMUNICATION WILL TEACH THAT FIRST AND ENGLISH IS TAUGHT AS A SECOND LANGUAGE.

AND THEN JUST TO WRAP UP SOME DIFFERENT IDEAS FOR PARENTS FOR WORKING WITH YOUR CHILD ARE AUDITORY NEUROPATHY. IF YOUR CHILD WEARS AMPLIFICATION OR HAS A COCHLEAR IMPLANT NOTE DIFFERENCES IN AUDITORY BEHAVIOR AND WRITE DOWN YOUR OBSERVATIONS AND SHARE IT WITH YOUR AUDIOLOGIST, DOCTOR.

DOES YOUR CHILD RESPOND DIFFERENTLY IN NOISE?

WHAT TYPES OF NOISE DO THEY RESPOND DIFFERENTLY?

JOT ANY OF THE CHANGES DOWN THAT YOU MIGHT NOTICE.

BE SURE TO KEEP COPIES OF ALL YOUR TESTS AND ASSESSMENTS.

THIS IS GOOD BOTH FOR WHEN YOU GO TO THE SCHOOL AND WHEN YOU GO TO SPEECH, A LOT OF TIMES THEY'LL SAY DO YOU HAVE AN IEP.

HAVE YOU HAD A RECENT TEST DONE WITH YOUR AUDIOLOGIST AND IT'S NICE BECAUSE YOU CAN DOCUMENT CHANGES THAT ARE MADE AND LOOK BACK AND SAY THEY'RE MAKING PROGRESS.

SOMETIMES YOU SEEM LIKE OH, THEY'RE KIND OF STUCK LOOKING BACK AND SEEING ALL THIS HAPPENS AT THE BEGINNING AND WHERE YOU ARE NOW.

ANY QUESTIONS?

>> THERE IS ONE ON THE BOARD.

>> THIS IS DR. HUANG, I'M ANSWERING HEATHER'S QUESTION ON THE SCREEN.

MELISSA TALKED ABOUT THIS A LITTLE BIT.

WE GET THIS QUESTION EVEN FROM OUR RESIDENTS BECAUSE IT SEEMS COUNTERINTUITIVE BECAUSE YOU HEAR TIME AND TIME AGAIN THE TRADITIONAL LEARNING ABOUT COCHLEAR IMPLANTS IS THAT YOU'RE BYPASSING THE COCHLEA AND STIMULATING A NERVE AND IF THERE IS SOMETHING WRONG WITH THE NERVE IT WON'T WORK.

IT'S DYSYNCHRONY.

AGAIN YOU'RE GETTING BAD SIGNALS.

I TELL MY RESIDENTS IT IS JUST LIKE GETTING A PACEMAKER.

YOUR HEART DOESN'T GIVE OUT THE RIGHT SIGNALS.

IT IS NOT BEATING CORRECTLY.

THE PACEMAKER DRIVES THAT WORK.

IT PUTS THE RIGHT SIGNALS INTO YOUR HEART BY GIVING IT THE ELECTRICAL SIGNALS IT NEEDS TO WORK.

AND THAT'S EXACTLY WHAT THE IMPLANT DOES.

IT DRIVES THE COCHLEAR NERVE INTO SYNCHRONY AND THAT'S WHY IT WORKS.

IF IT'S A HIGHER PROCESSING PROBLEM THAT IMPLANT PROBABLY ISN'T GOING TO WORK GREAT AND THERE IS A QUESTION EARLIER ABOUT IF YOU TEST THE ELECTRICAL ABR IN SURGERY AFTER YOU

PUT THE IMPLANT IN, THERE HAVE BEEN SOME STUDIES THAT SHOW THOSE CHILDREN THAT HAVE A NORMAL OR NEAR NORMALISH LOOKING ABR THAT THEY'LL BE THE KIDS THAT DO BETTER.

AND AGAIN YOU'RE DRIVING THE COCHLEAR NERVE.

IF YOU DRIVE THE COCHLEAR NERVE IS THE PROBLEM OR THAT SYNAPSE BETWEEN THE COCHLEA AND THE NERVE IS THE PROBLEM AND EVERYTHING HIGHER WORKS YOU'RE GOING TO GET A MORE NORMAL ABR.

IF YOU'RE DRIVING THE NERVE AND STUFF HIGHER ISN'T WORKING YOU'RE GOING TO HAVE AN ABNORMAL ABR AND IT IS CONSTITUENT PROBABLY NOT GOING TO WORK BECAUSE YOU KNOW IT'S A HIGHER ORDER PROCESSING PROBLEM.

IT DOESN'T MATTER WHAT'S GOING ON WITH THE NERVE.

>> SO IF YOU HAD A LATE ONSET CASE WHERE IT LOOKS LIKE NEUROPATHY, ALL THE AUDIO METRIC FINDINGS ARE SIMILAR TO WHAT YOU DESCRIBE SO A PERSON WOULD HAVE A PRETTY NORMAL SPOKEN LANGUAGE BASE, WOULDN'T YOU KNOW EARLY ON IN A SURGICAL PROCESS WHETHER OR NOT THAT PERSON MIGHT REALLY SAIL WITH AN IMPLANT THEN?

>> NOT REALLY.

BECAUSE NO MATTER WHAT – THE QUESTION WAS IF THIS IS A LATE ONSET LIKE AN ADULT WHO DEVELOPS THE NEUROPATHY OR THE DYSYNCHRONY, DO YOU HAVE A BETTER IDEA OF HOW THEY'RE GOING TO DO, AND MY ANSWER IS NOT REALLY BECAUSE YOU STILL DON'T KNOW WHERE THAT PROCESS—

>> I'M SAYING ONCE YOU'RE IN SURGERY.

IF YOU CONFIRM INTEROPERATIVELY THERE IS A GOOD ELECTRICAL.

>> YOU DON'T KNOW IT UNTIL THE IMPLANT IS IN.

THERE IS NO WAY TO CHECK THAT UNTIL THE IMPLANT IS IN AND YOU'RE PRETTY MUCH ALL CLOSED UP AND DONE WITH THE SURGERY.

>> THAT'S STILL FAIRLY EARLY IN THE PROCESS POST SURGERY THAT MIGHT HAVE A STRONG PREDICTOR.

>> TRUE.

AND THAT CAN BE DONE AT YOUR FIRST MAPPING AND PROGRAMMING, TOO.

YOU CAN GET AN ELECTRICAL ABR AT YOUR MAPPING AND PROGRAMMING.

IT DOESN'T NEED TO BE DONE DURING SURGERY.

THAT'S PROBABLY WHY MOST PEOPLE DON'T DO IT DURING SURGERY.

THAT'S A REALLY EXPENSIVE EABR ALL THE SURGICAL ANESTHESIA TIME.

IT'S PROBABLY MORE COST EFFECTIVE TO DO IT AT THE FIRST MAPPING AND PROGRAMMING.

IF YOU GET A BAD RESULTS ON IT YOU WON'T TAKE OUT THE IMPLANT AND SAY TOO BAD.

YOU'RE STILL GOING TO FINISH AND CLOSE AND CROSS YOUR FINGERS AND HOPE THAT THEY ARE GOING TO BEAT THE ODDS AND BE AN OKAY USER.

SO IT DOESN'T NECESSARILY MATTER WHEN YOU DO IT BECAUSE IT ISN'T GOING TO CHANGE WHAT YOU'VE ALREADY DONE.

>> I HAVE A QUESTION FOR LAURA.

MOST OF WHAT YOU'VE TALKED ABOUT IS FOR EARLY CHILDHOOD AND INFANTS.

DO YOU HAVE ANY SUGGESTIONS FOR SCHOOL-AGE CHILDREN?

>> IF THEY'RE DIAGNOSED LATER OR WHEN THEY'RE SCHOOL AGE.

>> THE QUESTION WAS WHAT I TALKED ABOUT WAS MOSTLY FOR YOUNGER CHILDREN AND IF I HAD SUGGESTIONS ON SCHOOL AGE CHILDREN.

I WOULD SAY THE SAME SUGGESTIONS.

THEY'RE MEETING THEIR LANGUAGE MILESTONES.

WE HAVE DIFFERENT TESTING THAT WE CAN DO.

SPEECH AND LANGUAGE TESTING THAT IS NOT NECESSARILY ON KIDS WITH HEARING LOSS BUT A GOOD PREDICTOR OF WHERE THEY SHOULD BE COMPARED TO THE NORMAL HEARING PEERS.

IT IS A GOOD WAY TO SET GOALS FOR THEM TO SEE WHERE THEY'LL BE.

LANGUAGE DEVELOPMENT AT THAT AGE.

IF THEY HAVE LANGUAGE AND THEY'RE AT THE HIGHER PROCESSING, IT WOULD BE KIND OF TRADITIONAL SPEECH/LANGUAGE THERAPY WORKING ON A HIGHER LEVEL FUNCTIONING.

THEY MIGHT NEED ONE-ON-ONE, YEAH.

>> GREAT, THANK YOU.

>> I'M WITH MINNESOTA HANDS AND VOICES, I'M CANDACE.

IS THAT BETTER?

I WOULD LIKE TO INTRODUCE TO YOU A PARENT WHO HAS COME TO SHARE THEIR STORY WITH US.

AND HER NAME IS LIZ, SHE HAS A BEAUTIFUL YOUNG DAUGHTER. SHE IS GOING TO TELL YOU ABOUT HER AND I THINK LIZ AND I TALKED EARLIER AND WE BOTH AGREE HER FAMILY STORY IS ONE STORY.

AND WE DID ACTUALLY TRY TO BRING A COUPLE MORE FAMILIES IN BECAUSE THERE IS SUCH DIVERSITY IN THE CHOICES THAT FAMILIES MAKE.

UNFORTUNATELY THOSE FAMILIES WERE NOT AVAILABLE TONIGHT AND WE HAD ONE LAST MINUTE CANCELLATION.

WE'RE GLAD LIZ IS HERE AND TELL US ABOUT HER DAUGHTER, MAVE.

>> I FEEL LIKE I'M FROM THE OLDEN DAYS.

I'M LIZ AND MY DAUGHTER, MAVE, WAS BORN ALMOST 11 YEARS AGO NOW AND SHE HAD SOME OF THE CLASSIC RISK FACTORS FOR HAVING AUDITORY NEUROPATHY.

SHE WAS BORN PREMATURELY AND HYPER BILIRUBIN – THE NURSE PRACTITIONER SAYING WHEN WE WERE TAKING HER HOME YOU

**MIGHT WANT TO HAVE AN ABR DONE IN SIX TO NINE MONTHS
BECAUSE SHE'S AT RISK FOR THE KIND OF HEARING LOSS THAT CAN'T
GO CAUGHT ON AN OAE.**

WE WAITED.

I'M ASSUMING THAT'S NOT THE ADVICE GIVEN NOW.

**WE WAITED AND SHE WAS NOT RESPONSIVE TO US AT HOME AT ALL IN
TERMS OF HEARING.**

**AND SO WE BROUGHT HER IN, HAD THE ABR AND IT WASN'T AN
ABSENT ABR.**

**IT WAS ATYPICAL AND SHE HAD A PROFOUND HEARING LOSS IN
ADDITION TO THAT.**

THIS WAS 1999.

**AND AT THAT TIME WHAT WE WERE TOLD WAS SHE – USE SIGN
LANGUAGE.**

HEARING AIDS WON'T REALLY WORK.

**AT THAT TIME THEY DIDN'T THINK ANYTHING ABOUT COCHLEAR
IMPLANTS.**

AND SO THAT'S WHAT WE DID.

WE CONTACTED THE SCHOOL DISTRICT.

**WE GOT THE SCHOOL DISTRICT INVOLVED AND ABOUT A YEAR LATER
THE AUDIOLOGIST CALLED US AND SAID WE'VE HEARD ABOUT A
COUPLE OF KIDS WHO HAD AUDITORY NEUROPATHY, HAD THE
COCHLEAR IMPACT AND HAVING SOME SUCCESS.**

THAT WAS THE PATH WE TOOK WITH MAVE.

SHE WAS 2 ½ WHEN SHE GOT HER IMPLANT.

**I DO REMEMBER NOW THE AUDIOLOGIST IN THE SURGERY WHO SAID
HER ABR LOOKED GREAT AFTER SURGERY.**

HE WAS SO EXCITED AND I THOUGHT BIG – TIME WILL TELL.

**IT REALLY FOR THE FIRST YEAR AFTER SHE HAD HER IMPLANT WE
JUST WONDERED WAS THIS WORTH IT?**

SHE REALLY WASN'T VERY RESPONSIVE.

SHE WAS RESPONSIVE BUT NOT TALKING.

AND THEN ALL OF A SUDDEN SHE'S TAKEN OFF.

CANDACE HAS SEEN HER.

SHE DOESN'T STOP TALKING NOW.

THAT'S MANY YEARS NOW DOWN THE LINE.

**SHE IS IN A CLASSROOM WHERE THEY USE TOTAL COMMUNICATION,
EXPRESSIVELY SHE TALKS AND SHE HAS GOOD ARTICULATION.**

SHE CERTAINLY DOES FOR US.

SHE USES AN FM SYSTEM.

**WHEN WE'RE OUT AT COMMUNITY EVENTS OR OTHER PLACES, SHE
USES AN INTERPRETER FOR RECEPTIVE COMMUNICATION AND THE
FM SYSTEM.**

**SO REALLY KIND OF A TOTAL COMMUNICATION IS WHAT'S WORKED
FOR HER.**

AND ABOUT TWO YEARS AGO WE HAD A SECOND IMPLANT.

**SHE – I DON'T THINK KNOWS SHE HAS AUDITORY NEUROPATHY.
SHE'S JUST A DEAF KID.**

THAT'S WHAT SHE'S DOING.

THAT'S KIND OF OUR STORY.

ANY QUESTIONS?

I WISH SHE WAS HERE.

IT'S A LITTLE BIT TOO LATE FOR HER TO BE OUT AND ABOUT.

>> WHEN DID SHE GET THE SECOND IMPLANT?

>> TWO YEARS AGO.

THAT WAS QUITE – I MEAN, SHE WAS OLD.

**SHE WAS EIGHT YEARS OLD AND SHE – IT'S NOT HER STRONG SIDE.
WHAT WE SOMETIMES DO IS HER FIRST IMPLANT SOMETHING
HAPPENS.**

IT IS NOT WORKING.

AND WE FORCE HER TO REALLY DEPEND ON HER SECOND SIDE.

OTHERWISE THEY DOESN'T DO THAT WILLINGLY.

BUT IT'S GOOD PRACTICE FOR HER AND SHE REALLY CAN DO WELL WITH IT.

IT IS JUST NOT HER STRONG SUIT SO –

>> THANK YOU.

>> THERE IS ANOTHER QUESTION.

THAT'S PROBABLY NOT FOR YOU.

THAT'S FOR LAURA, I THINK.

>> THERE IS A QUESTION ON THE SCREEN THAT SAYS WHAT DO YOU SUGGEST FOR THERAPY, EDUCATIONAL SERVICES IF A CHILD HAS BILATERAL CIs AND THEIR LANGUAGE DEVELOPMENT ISN'T DEVELOPING AT AN EXPECTED RATE?

FOR EDUCATIONAL SERVICES HOPEFULLY THEY'VE GOT AN IEP IN PLACE IF THEY DO HAVE BILATERAL COCHLEAR IMPLANTS THEY PROBABLY DO.

READDRESSING WITH THE SCHOOL WHAT THE GOALS ARE AND IF THEY'RE GETTING SOME THERAPY THROUGH SCHOOL OR GETTING SOMEONE ON ONE THERAPY IN THE SCHOOL AND THE MODIFICATIONS IN THE CLASSROOM.

IF THEY'RE SCHOOL AGE.

I DON'T KNOW IF THIS IS A YOUNG CHILD.

ARE THEY SITTING IN THE BACK OF THE ROOM BY THE DOOR?

ARE THEY IN THE FRONT BY THE TEACHER?

ARE THEY BY THE NOISY FRIEND THAT'S MAKING NOISE AND MOVING THEIR CHAIR?

AS FAR AS THERAPY I WOULD RECOMMEND IF THEY AREN'T MEETING THEIR MILESTONES TO GET SOME OUTSIDE SPEECH THERAPY DEPENDING ON THEIR AGE, SCHEDULE IS ONE TO TWO TIMES A WEEK TO GET A LITTLE BOOST AND SEE IF THEY CAN'T CATCH UP AND CLOSE THAT GAP A LITTLE BIT.

LOOKS LIKE HEATHER IS TYPING.

THAT'S IT.

**>> I JUST WANT TO THANK EVERYONE FOR COMING TONIGHT AND
THANK GOODNESS FOR THE RAIN STOPPING SO WE COULD GET HERE
DRY.**

I WANTED TO THANK OUR PRESENTERS.

THANK YOU – OKAY, THAT ONE IS NOT FOR ME, EITHER.

WHO WOULD LIKE TO COME UP ON THE HOT SEAT?

>> WE CAN GET HER EMAIL.

**SHE CAN SEND PRIVATELY AND WE COULD ANSWER HER QUESTION A
LITTLE MORE SPECIFIC.**

**>> HEATHER, THE SUGGESTION IS IF YOU WOULD LIKE TO EMAIL
ALYSSA ON THE NOTE PORTION OF THE SCREEN HERE A SPECIFIC
QUESTION, THEN THE PRESENTERS WERE OFFERING THEIR ADVICE
AND COUNSEL PRIVATELY IF THAT WOULD WORK FOR YOU.**

I WOULD LIKE TO THANK OUR PRESENTERS TONIGHT.

**I WOULD LIKE TO THANK LIZ FOR SHARING HER FAMILY STORY AND
HER DAUGHTER IS EXTREMELY INTELLIGENT AND VERY FUN TO BE
AROUND.**

**SO OBVIOUSLY THE AUDITORY NEUROPATHY HAS NO BEARING ON
HOW MUCH INFORMATION SHE KNOWS AND HOW SMART SHE IS.**

**I WOULD LIKE TO THANK DR. HUANG, MELISSA AND LAURA FOR
TAKING TIME AND WORKING ON YOUR POWER POINT AND
PRESENTING TONIGHT FOR US AND I WOULD LIKE TO THANK – THANK
OUR SUPPORT PERSON TO MAKE THIS HAPPEN.**

THANK YOU.

**A SPECIAL THANK YOU TO ALYSSA ANDERSON FOR ALL THE RUNNING
AROUND.**

**YOU HAVE NO IDEA WHAT SHE'S BEEN DOING TO GET THE
TECHNOLOGY AND REFRESHMENTS ARRANGED.**

YOU'VE BEEN A HUGE, HUGE HELP.

**THE WORKSHOP WILL BE AVAILABLE FOR VIEWING AFTER TONIGHT
AND AS ALYSSA HAS NOTED HERE CONTACT HER FOR FURTHER**

INFORMATION ABOUT THAT AND TALK FURTHER ABOUT ANOTHER WAY TO PUT A LINK ON OUR HANDS AND VOICES WEBSITE OR WHATEVER AND THERE WILL ALSO BE A TRANSCRIPT OF TONIGHT'S CAPTIONING, TOO, SO THAT WILL BE HELPFUL FOR THOSE DEAF OR HARD OF HEARING INDIVIDUALS WHO I KNOW WANTED TO PARTICIPATE BUT WE WEREN'T ABLE TO DO THE CAPTIONING OTHER THAN IN THIS ROOM.

WE APOLOGIZE FOR THAT.

THE POWER POINTS ARE UP ON THE HANDS AND VOICES WEBSITE. FEEL FREE TO GET THEM FROM THERE.

IF THE PRINT AND SOME OF THIS IS A LITTLE TOO SMALL OR FAINT, WHICH I REALIZED AS WE GOT HERE, WE'D LOVE TO HAVE FEEDBACK ABOUT THE WORKSHOP SO IF YOU WOULDN'T MIND CONTACTING HANDS AND VOICES YOU CAN REACH US AT MNHV STANDING FOR MINNESOTA HANDS AND VOICES AT LIFETRACK RESOURCES.ORG. LET US KNOW HOW THE WORKSHOP WENT FOR YOU AND WE'D LOVE TO KNOW IDEAS FOR FUTURE TOPICS AS WELL.

AND I THINK THAT IS IT FOR TONIGHT.

SO THANK YOU VERY MUCH AGAIN FOR COMING.

AND HAVE A GOOD EVENING.

[APPLAUSE]