THE SMITH FAMILY CURSE

AN EIGHTEEN-GENERATIONAL STUDY OF HEREDITARY HEARING LOSS

By Mary R. Simms

CHAPTER 1---OPENING

UNIT 1

AN EIGHTEEN-GENERATIONAL STUDY OF HEREDITARY HEARING LOSS

[aka THE SMITH FAMILY CURSE]

521 YEARS OF DFNA/DFNA1 HEREDITARY HEARING IMPAIRMENT TRACED TO TWO COMMON ANCESTORS!

By Mary R. Simms
marysimms@sc.rr.com
803-996-3567
hildenbrandt.com/simms

UNIT 2---DEDICATION

In memory of

My beloved Great-Grandfather

WILLIAM FRANKLIN SMITH 1865---1921

TEXAS RANGER---HOMESTEADER---PIONEER
HEARING IMPAIRED FROM BIRTH

Rest in Peace, Grandpa Frank

UNIT 3—CONTRIBUTORS

Mary R. Simms ---Author, Research, Communications

Pamela Russell---Art Graphics, Genealogy

Clark Jones---Buchanan Line Genealogy

Cynthia R. Hildenbrandt---Web Site

Wes Brosman---Guest Columnist

David R. Simms---Archives

Barron Simms---Editor

Many thanks to all of the family members who furnished the information contained herein. Without their cooperation, this book could not have been written.

UNIT 4---FOREWORD

521-year chain of hereditary hearing loss.—Although the Smith Family Curse has been traced back to 1490, this report documents mainly the 147-year period between 1864 and 2011. Twelve earlier generations are noted but not documented with fact sheets as little is known about these early ancestors except for their hereditary hearing loss. See Units 35, 60, and 108 for further details regarding the period from 1490 to 1864.

Truth in advertising.—The mortal remains of William Franklin Smith lie buried in a lonely mountain grave, high up in the Colorado Rockies near Craig, Colorado. He was ambushed and murdered during a feud over a few bags of potatoes. After such a violent death, it is unlikely that he rests in peace. His tombstone reads: "WILLIAM FRANKLIN SMITH---1865—1921". Perhaps they should have added: "WARNING!!! BEING A DESCENDANT OF THIS MAN COULD PROVE HAZARDOUS TO YOUR HEARING!" This addendum would certainly comply with the rules and regulations of today's truth in advertising guidelines.

Root of the problem.--After four years of research, our family hereditary hearing loss was traced back 521 years—showing a chain of eighteen consecutive generations with hereditary hearing loss. The DFNA/DFNA1 hereditary hearing loss was previously traced back to only William Franklin Smith [1865] who was nearly deaf at the age of 56 when he came out on the short end of a deadly mountain feud. The chain of hereditary hearing loss has recently been traced back much farther. A most important fact which was discovered recently was that Frank's wife, Sarah, shared the same family lineage of hereditary hearing loss.

Information.--Detailed information from the past 147 years covering six generations and including nearly 300 of Frank and Sarah's genetic descendants has been documented. These acquired facts were compiled, organized, analyzed, and condensed to create this report.

Tracing it back.--A consecutive pedigree chain of inherited hearing loss goes all the way back to 1490 when Frank's great-great-great-great- great-great-great-great-great-great grandfather George Buchanan, was born in Scotland. This same

George Buchanan was also Sarah's great-great-great-great-great-great-great-great-great-great-grandfather.

1490-1864--Very little is known about the period between 1490 and 1864, therefore this report has been keyed to great grandpa Frank, his wife, and their descendants.

Guilty party?--Until recently, it was unknown whether Frank inherited his poor hearing from his mother or from his father. We knew that Frank and Sarah were first cousins once removed but nothing more. Now it becomes evident that the hereditary hearing loss originated within Frank and Sarah's common ancestors—the Buchanan line. Therefore, Sarah was as much responsible for the hereditary hearing loss gene being passed down to future generations as Grandpa Frank.

Inbreeding.-- Between 1490 and 1900 there was a lot of inbreeding. During that period many cousins as well as siblings married and reproduced. This seemed to intensify the hereditary hearing loss gene. The last known marriage of relatives was Frank and Sarah in 1897. By this time the hearing loss gene was strong enough to survive another six generations and still keep going strong! [See Unit 108]

Collecting information.--Family interviews and old memories make up the bulk of information contained in this report. The only concrete evidence of the hereditary hearing loss is contained in thirty audiogram charts which were collected. A condensed fact sheet was prepared on every family member who had any reported history of hearing loss. Some members attributed their hearing loss to heredity while others felt it was old age, disease, and/or injury that caused their hearing loss. All types of reported hearing loss within the family were documented.

Hearing loss statistics.-- Within the first three generations 73% are believed to be afflicted with hearing loss. Within the first four generations, the percentage drops to 45% believed to have a hearing loss. It is suspected that several cases of hearing loss have gone unreported and/or undiagnosed.

Original goal.--Before this information was lost forever, the goal was to obtain facts about as many as possible of Frank and Sarah's descendants who had a

history of hearing loss. Questionnaires were mailed out; audiograms were sought; telephone interviews were conducted; fact sheets were compiled. In the end, all of this collected information was organized and condensed into this research report.

Current goal.--With the completion of this research report, the new goal is to reach as many of the descendants of George and Margaret Edmonstone Buchanan as possible and share this information with them. The hereditary hearing loss is a serious matter and needs to be dealt with intelligently by all of George and Margaret's descendants. Bless them; they had no idea why they could not hear. 521 years later, we should be able to understand the cause of our hereditary hearing loss and deal with it accordingly.

Support group.--A Smith Clan reunion was held in Oklahoma City in September 2008. This reunion brought together over sixty family members, many of whom were afflicted with hereditary hearing loss. Information was shared among participants. Another Smith Clan reunion occurred in August 2010 with the same goal in mind. It was a blessing for those with the affliction to meet and discuss their problems with others of a like nature. Thusly, a hereditary hearing loss support group has been formed.

Future OKC Smith Clan Reunions.--OKC Smith Clan Reunions are to be held on a bi-annual schedule. For more information on attending these reunions, please contact the author at marysimms@sc.rr.com or phone 803-996-3567.

Toni Morrison once said: "If there is a book you really want to read, but it hasn't been written yet, then you must write it." I took Toni's advice and wrote this book.

It has taken four years, countless hours of research and extensive personal interviews, as well as considerable monetary outlay, to create this report. In order to make the text easier to understand, hundreds of pages of technical jargon have been translated into simple, everyday language. Facts from family members were obtained, sorted, and documented. Here then are the results of my efforts.

UNIT 5---TABLE OF CONTENTS

CHAPTER 2---ADVICE

UNIT 6---COMMUNICATING

Time for a change.--The terms "hard of hearing", "deaf", and "hearing impaired" are getting somewhat travel weary. Don't you think it is about time that we became a bit more politically correct and called our less than normal ability to hear by a more proper name? Recently, many other titles have been adjusted to reflect this more respectful attitude. [Fire-fighter, police officer, flight attendant, server, etc.]

Audio challenged.--The time is right for a new classification for those of us who are saddled with less than perfect hearing. One proposed title was "Audio Challenged". Think of it; doesn't it sound much more dignified to be categorized as Audio Challenged than to be labeled "deaf as a post"?

Please ponder this.—Do you think that it was our choice to be deaf? Do you think that we enjoy being deaf? Do you think that we could hear your voice better if we only tried harder? Do you think that wearing hearing aids gives perfect hearing? If you answered "yes" to any of these questions, it is definitely time for you to re-think the situation!

Want to know what it feels like to be hearing impaired?--Try stuffing your ears full of Silly Putty for a day or two and you will learn quite quickly what it is like to walk in our shoes! Not much fun, was it?

Being empathetic.--In communicating with a person who is "audio challenged", there are several things that you should bear in mind. Most of these are just plain

common sense, good manners, consideration, courtesy, respect, kindness, empathy, and patience.

a) Remember that we are doing our best to hear and understand you. We truly want to comprehend what you are saying.

b) Always get our attention prior to speaking to us. We need to know that you are addressing us before you begin to speak. Then we can pay special attention to what you say.

c) Never try to talk to the back of our heads. We do not have ears on the back of our heads; in fact, we don't even have very good ears on the sides of our heads either. ☺

d) Be sure that we can see your face and watch your expression at all times. It gives us clues as to what you are saying and often prevents requests for repeats. You might be surprised to learn that over half of our comprehension of what you are saying does not come from hearing your spoken words. It comes from reading your lips, watching your body language, picking up on your tone, seeing your expression, as well as some just plain guessing on our part.

e) If we do not hear you the first time you say something; do not use an exasperated, acidic tone of voice when repeating it. We really tried to hear what you said the first time around; it was fate that intervened and prevented it.

f) Don't become disgusted with us when we fail to hear even on the second or third repeat. Honest, we are trying our best. Do you think that we asked to be deaf?

g) Be patient and keep your sense of humor. Laugh with us but not at us.

h) When we do not understand what you are saying, please don't smirk and say: "What's the matter with you? Are you deaf or something?" Or perhaps, "I think maybe you need to get your ears cleaned out!" These words cut to the bone.

i) Please don't discuss us with another person in our presence as if we were not there. We may not be able to hear every word you say about us, but we can comprehend enough to know that we are being denigrated. This really hurts.

j) Please remember that we are fellow human beings. Just because we cannot hear perfectly does not make us dull, dim-witted, ignorant, or stupid.

k) Remember before you dismiss us as unfit to live in the same world with you because we do not hear so well, that it could just as easily have been you with the problem instead of us. Consider these two wise old sayings: a) "There but for the Grace of God walk I", and b) "Please do not judge me until you have walked a moon in my moccasins."

l) Remember that we neither requested nor deserved to be audio challenged. This handicap was dumped on us by accident. Do your honestly think that we chose to be deaf? Would you choose to be deaf?

m) Please do not ridicule or make fun of our handicap. That is just plain old rude.

n) Please avoid trying to converse with us when there is a lot of background noise, especially if we are wearing hearing aids. The background noise is magnified, and your voice is obliterated. Give us a break.

o) Be considerate when we misunderstand what you are saying and make a gaffe. Don't laugh at our boo-boo and repeat it for others to laugh with you. Please don't rub our noses in our mistakes as you would a dog that soiled your carpet. It is humiliating to both the dog and us.

p) For us to understand you, your voice control is very important. In order to successfully communicate with us, please consider the following suggestions:

1. Speak slowly. Never talk 90 miles per hour.

2. Don't run your words together or slur them.

3. Enunciate each word clearly. (Listen to Queen Elizabeth sometime, and notice how she speaks. She is a role model for clarity.)

4. Raise your overall volume, but don't yell at us.

5. Monotones are difficult to understand, so add a little enthusiasm and expression to your voice. It helps us to understand you better.

6. Speak in a clear, pleasant manner with facial expressions included. Animation helps greatly!

7. Please don't speak in a tiny, soft, little baby voice as though you are afraid that someone might hear you. We are already having enough trouble as it is!

8. Your voice should come from the diaphragm in the manner of great opera singers. This would help ever so much.

9. Please don't mumble. It is next to impossible to decipher words when they dribble like mush from the speaker's mouth.

The Ball is in your court.--Do remember when communicating with an "audio-challenged" person, that we are already doing our best to understand you. Isn't it about time for you to do your part by following the suggestions outlined above?

Thank you and God bless

UNIT 7---"DEAR WES" ADVICE COLUMN

By Cousin Wes Brosman
[FRANK---TOM---OLA---WES]

"For starters, let me assume the role of Hearing Aid Guru and give free

advice that will be worth every penny you paid for it!'

Behind-the -Ear or In-the-Ear?--I cannot recall how many times I have heard that refrain: "Just as good as or as strong as a BTE aid." [Short answer here? Bull!]

I have to laugh.--Every time I have heard this claim I grin and look at the utter shock on the audiologist's face when he reads my audiogram while shaking his head. No, Virginia, there ain't no Sandy Claus and there ain't no canal aid "as good as a "Behind the Ear Aid."

Telecoils in aids.--98% of the aids sold in Europe have Telecoils, and only half of those sold in the USA have this feature. And of those sold with T coils in the USA, most have T coils with less than half the power of those in Europe. You should ask, or better yet, DEMAND, that the aids you buy have double or triple power T coils.

T coils.--And while I am on this rant, let me point out that to my knowledge, no "in the ear" instrument has a full-strength T-coil and any person who buys any aid without a full strength T coil is doing all of us a disservice.

Rip off.--We don't get Telecoils in more than half of the aids sold in America because the audiologists want to sell you an aid that does not have a separate mold to fit in the ear, so that instead of refitting to accommodate subtle changes in ear canal shape brought on by weight loss/gain or aging, we have to buy brand new aids. Who benefits from this practice? Your friendly neighborhood audiologist/hearing aid salesman!

In-the-Ear Type Aids.-- My view is that these in-the-ear-type aids do not deliver the best quality of sound because that is not their purpose. They are vanity aids that give a little better hearing than no aid at all, but do so while tucked out of sight so the owner will not be embarrassed by being identified as a hearing aid user. At some point every hearing impaired person will come to understand that egos suffer more damage from not hearing than from the openly displayed "behind the ear" aid.

ITE and CIC aids.-- Notice, I don't give much advice on "all in the ear" or "all in the canal" aids? There are not many things the user can do to clean or fix these aids, except for using gadgets to clean out the channel as best you can.

Bottom line wisdom.--The bottom line is this: Behind the Ear aids work better, are easier to maintain, and more comfortable to wear all day long. The BTE also has several adjustments that the user can do for himself. The all-in-the-ear aid has virtually nothing that can be adjusted at home.

Slick salesmen.--Most hearing aid users get an aid the audiologist is eager to sell. This falls under "Turn on the blue light, the man wants a blue suit." Slick salesmanship does not make a happy customer, and I would bet the "unhappy user" category fits many of us on the list.

Lied to.--My current aids are better than the ones before. I make certain of that or they go back. However, they do not fill the bill as advertised "will virtually eliminate microphone feedback." Horse hockey!

Service on aids.--Remember, there is a tremendous markup on the price of hearing aids. You are paying dearly for service after the sale, so keep going back until you are satisfied with the fit and adjustment of your new aids. You have paid for this service---now use it!

Headphones.-- I never use headphones for several reasons. First is the fact that they distort the sound for my reverse-slope hearing loss. Then there is the difficulty in getting the sound loud enough for my 85 dB loss when headsets only give a 45 dB boost. With insufficient boost I am forced to try to put the headset on over my aids.

Oops.--Since the microphones on my aids are up at the top, I have to position the headset so high that it does not adjust to touch my head, and I either hold it in place or let it drop forward and serve a blindfold. Most headsets are low in quality and some are truly stupid. And truly stupid is how I look in a headset.

Don't use headsets and save yourself a lot of frustration. Each of the systems I will cover will come to you through your own hearing aid. The Smith Family Curse sufferer should not try to use any system that does not utilize your own aids to

transmit the final sound because our hearing is different, and our personal aids are set up to take into account all that weirdness that is reflected on our hearing chart.

Those "T" settings again.--This means you MUST have a T setting on your aid. If you do not have a T or Telecoil setting on your aid, then chuck it out the window and claim you lost it in an accident so you get something off on your income taxes. An aid without a T setting is next to worthless for Smith Family Curse folks---trust me on this issue, this is the voice of experience speaking.

$10 Sound System.--Now for the icing on the cake: the best and cheapest Assistive Listening Device we can get is the oldest system and the lowest in technology. I speak of the Magnetic Induction Loop. Here's how you can try out this idea. Go to Radio Shack and ask for a 20-foot piece of speaker wire with a male plug on both ends.

How to do it.--Look at your radio or TV set and find a spot for two plug-ins with the words "audio" or "speakers" or "headset". Plug one end of the wire into one of these outlets [they come in pairs]. Run the wire under a throw rug or along the wall baseboard to go near your favorite chair. Next loop it back around to the second plug-in location which should be right next to where you plugged in the first end of the wire. With both ends of the loop plugged in, sit down and turn on the TV.

Sit back and enjoy.--Switch on your aids, turn to T setting, and listen to the clearest sound you will ever hear. And all for less than ten bucks!!!

Oops.--Unfortunately, this procedure cuts off the normal TV sound and the TV audio can be heard only by the person wearing hearing aids!

Radio Shack to the rescue.-- Radio Shack can remedy this problem easily. They will sell you an inexpensive gismo to attach to the TV that will provide simultaneous audio for both regular listeners as well as the magic loop.

About Cousin Wes.--This is just a small sampling of the wit and wisdom of Wes Brosman. When it comes to advice on the care and feeding of hearing devices, Wes is tops! Be sure to read his latest book *No Place Else* which can be ordered

from Wes Brosman. If you have a personal question to ask Wes about hearing devices, he can be reached at wesbro@techline.com. He would love to hear from you!

Cousin Wes is the most amazing person! He has an 85 dB hearing loss and is an excellent role model for anyone who is hearing impaired. He is happier and more successful than most people with normal hearing. Hats off to Wes!

See Wes's Fact Sheet---Unit 67--- to learn more about this truly remarkable person.

UNIT 8---GUIDE FOR USING THIS BOOK

a) Documented research commences with William Franklin and Sarah Buchanan Smith serving as our most common ancestors.

b) In this report, William Franklin Smith will be referred to as "Frank" and his wife as Sarah.

c) In the beginning, Frank and Sarah were our oldest known ancestors with a hereditary hearing loss. When the Buchanan information was uncovered, it indicated that the hereditary hearing loss dates back to 1490 and even earlier.

d) "Smith Family Curse" and "SFC" are used interchangeably throughout this book to describe the suspected DFNA/DFNA1 hereditary hearing loss found in the 18 generations that began in 1490 with George and Margaret Edmonstone Buchanan.

e) Nearly 300 genetic descendants of Frank and Sarah have thus far been investigated.

f) The following order of generations is used:

Generation 1---Frank & Sarah
Generation 2---Frank & Sarah's 10 children
Generation 3---Frank & Sarah's 18 grandchildren

Generation 4---Frank & Sarah's 58 great grandchildren
Generation 5---Frank & Sarah's 108 great-great grandchildren
Generation 6---Frank & Sarah's 88 great-great-great grandchildren
Generation 7---Frank & Sarah's 2 great-great-great-great grandchildren

g) A "G" following a person's name is used as an abbreviation for "Generation".

h) A number following the "G" denotes which generation that particular person represents in relationship to Frank and Sarah. [See chart "f" above.]

i) On pedigree charts, bold uppercase lettering indicates reported hearing loss.

j) Shaded ovals and/or blocks on a pedigree chart indicate reported hearing loss.

k) On pedigree charts, male names are shown in blocks. Female names are shown in ovals.

l) There are 13 Pedigree Charts consisting of: Chart # 1--Generations 1, 2, and 3, and then Charts # 2 through #13---showing twelve of Frank's 18 grandchildren with reported hearing loss and their genetic descendants.

m) Pedigree charts are limited to those twelve grandchildren with reported hearing loss within their branch of family tree. No reported hearing loss? No pedigree chart in this report.

n) A glossary is provided.

o) A Table of contents as well as a general index are provided.

p) Audiogram, Fact Sheet, and Pedigree Chart indexes are provided.

q) Unit 108 explains how the original 147-year study suddenly expanded to 521 years of continuous hereditary hearing loss.

r) To ensure privacy, initials are used instead of last names.

s) All information contained herein was believed to be true at the time of publication. Sincere apologies for any errors.

CHAPTER 3---HEARING LOSS 101

UNIT 9---CAUSES & TYPES OF HEARING LOSS

There are four basic causes of hearing loss in humans:

a) Injury
b) Disease
c) Age
d) Heredity

The Smith Family Curse is a hereditary hearing loss.--A hereditary hearing loss is in the making prior to the child's conception. One or both of the parents involved will have a defective gene that is passed down to their offspring at conception. Those offspring then have the potential of passing the hereditary hearing loss on down to their offspring.

There are four basic types of hereditary hearing loss:

a) X-Linked---Comprises less than 2% of hereditary hearing loss

b) Mitochondrial---Comprises less than 2% of hereditary hearing loss

c) Autosomal Recessive---Comprises almost 65% of hereditary hearing loss

d) Autosomal Dominant---Comprises almost 30% of hereditary hearing loss

The Smith Family Curse is Autosomal Dominant.--It is an equal opportunity employer and is not sexually discriminating. It affects male and female alike. Either the mother or the father can hand it down to either male or female offspring. It can be conspicuous at birth or not be noticeable before age 50---or it can make its presence known at any time in between. It can vary from a mild loss all the way to a profound loss or any gradient in between. A basic rule of thumb

is that a child with Autosomal Dominant Hearing Loss will have at least one afflicted parent. Approximately 50% of the offspring of an afflicted parent will also be afflicted. The other 50% will be born with normal hearing. It is just the luck of the draw.

Spontaneous Mutation.--A rather frightening fact about Autosomal dominant hereditary hearing loss is that it can occur even though both parents and all 4 grandparents have normal hearing. In this case, it begins at conception and is called "Spontaneous Mutation". This is what is sometimes called a "throwback".

In conclusion.—Research has determined that the Smith Family curse can be classified as Autosomal Dominant Sensorineural Nonsyndromic Hereditary Hearing Loss. This is good news as well as bad news.

a) The good news is that we have solved the mystery of the Smith Family Curse.

b) The bad news is that the only permanent way to eliminate the problem would be for every last one of Frank's genetic descendants to halt all procreation.

[Not very likely!] ☺

UNIT 10---DEGREES OF HEARING LOSS

1. Normal hearing:

a) Scores between 0—25 dB shown on audiogram
b) No hearing loss noticeable by self or others
c) Audiogram confirms normal hearing
d) All "Banana Points" intact on audiogram

2. Mild hearing loss:

a) Scores between 26—40 dB shown on audiogram
b) Hearing loss may go unnoticed
c) Audiogram confirms mild hearing loss
d) Most "Banana Points" are still intact
e) Aid seldom required

3. Moderate hearing loss:

a) Scores between 41—55 dB shown on audiogram
b) Hearing loss noticeable by self and/or family
c) Some "Banana Points" will be missing
d) Talking on regular phone becomes difficult at times
e) Hearing Aid optional

4. Serious hearing loss:

a) Scores between 56—70 dB shown on audiogram
b) Hearing loss obvious to self and family
c) Understanding conversations very difficult
d) A few "Banana Points" may or may not remain
e) Talking on regular phone becomes extremely difficult
f) Aid highly recommended

5. Severe hearing loss:

a) Scores between 71—90 dB shown on audiogram
b) Hearing loss obvious to everyone
c) Understanding conversation next to impossible
d) All "Banana Points" are missing
e) Amplification necessary to talk on phone
f) Strong BTE hearing aid mandatory

6. Profound hearing loss:

a) Scores higher than 90 dB shown on audiogram
b) Hearing loss obvious to all
c) Person nearly deaf
d) Understanding normal conversation impossible
e) All "Banana Points' are missing
f) Very strong BTE hearing aid mandatory
g) May also require other high tech hearing devices

UNIT 11---AUDIOGRAMS EXPLAINED

Welcome to the wonderful world of audiograms! In case audiograms are all Greek to you, the following should help you to understand them a bit better. If

you are already cognizant with the workings of the audiogram, please feel free to skip this explanation.

a) Take a look at the audiograms in this report, and you will see that audiograms are a square with vertical and horizontal lines marking it into grids---graphs.

b) Notice that across the top it measures frequency in Hz. These correspond with the vertical lines on the chart. There may be numbers on the top line of the chart from a "–1 Hz" to "8,000 Hz." Most audiograms only measure the Hz from 250 to 8,000.

c) Let's think of these vertical lines as being the strings on a guitar. The one to the far left [250 Hz] will be the bass sting while the one to the far right [8,000 Hz] will be the soprano string. Those strings in between graduate in tone as they move to the right. Thusly, the 250 Hz will be a low note and the 8,000 Hz will be a high note. [Are you with me so far?]

d) Next we come to those horizontal lines. These are the decibel measurements—or dB for short. Some audiograms range from "–10 dB" at the top of the chart to a "120 dB" at the bottom. These numbers measure how loud a noise has to be in order to get your attention. [Similar to the volume control on your TV set.]

e) Next, think of the dB numbers on the horizontal lines as golf scores. As you know, in golf, the lower your score, the better the game! The same is true in evaluating the dB scale. Therefore, a 20-point golf score or 20-point dB score would be better than an 80 point golf score or an 80 point dB loss on the audiogram.

f) Look at the pencil markings that are written on an audiogram. You will see what looks like strings of beads. There will be a string of "X" beads and another sting of "O" beads.

g) In audiogram language, the "O" beads indicate the right ear while the "X" beads indicate the left ear.

h) Some “necklaces” will go almost straight across while other “necklaces” will go uphill and others will go downhill. Some even have humps and dips in the necklace.

i) An uphill “necklace” would indicate better hearing in the higher tones. A downhill “necklace” would show better hearing in the lower tones. A “necklace” that goes straight across the chart would show that the person’s hearing was nearly equal at all tones.

Banana shadows.--The banana shadow derives its name from a banana-shaped area marked off on some audiograms that shows the area where human voices can be heard. [See Unit 97, Ken R, G4 audiogram for a good example of the banana shadow.] A perfect score on banana points would be 12. The greater the number of banana points---the better able a person is to hear normal conversation. The fewer the points, the more difficult it is to understand words. No points at all? Better get some hearing aids!

And there you have it---audiograms in a nutshell!

UNIT 12---GLOSSARY

Banana points---Banana-shaped shadow imposed onto audiogram graph to mark the dB score levels necessary for hearing human speech

Bat Squeak Syndrome---Ability to hear tones at a much higher frequency than those heard by people with normal hearing

Cookie Bite---Small dip or a hump in audiogram “necklace” graph pattern

dB, Hz on audiograms---dB, Decibel, measures volume of sound—Hz, Hertz, measures high/low tone frequencies.

DFNA---Scientific acronym denoting Autosomal Dominant Nonsyndromic Hereditary Hearing Loss [Such as SFC]

DFNA1---Sub group of DFNA---scientific acronym denoting greater loss in lower

tones; less loss in high tones [Frequent in SFC]

Generation 1 [G1]---William Franklin (Frank) Smith & his wife Sarah
Generation 2 [G2]---Offspring of Frank & Sarah
Generation 3 [G3]---Grandchildren of Frank & Sarah
Generation 4 [G4]---Great grandchildren of Frank & Sarah
Generation 5 [G5]---Great-great grandchildren of Frank & Sarah
Generation 6 [G6]---Great-great-great grandchildren of Frank & Sarah
Generation 7 [G7]---Great-great-great-great grandchildren of Frank & Sarah

Hearing Loss Syndromes---Various hearing-related symptoms that often accompany our familial hereditary hearing loss such as hearing impairment, undersize ear canals, bat-squeaks, greater loss in lower tones, tinnitus, etc

Most common ancestors---George and Margaret Buchanan, (born 1490)

Most common documented ancestors---William Franklin (Frank, born 1865) and Sarah Buchanan Smith (born 1864

Nonsyndromic hearing loss---Hereditary hearing loss that is not evidenced by seemingly unrelated physical characteristics, such as white streak in forelock, eyes of two different colors, blindness, etc

Pedigree chart---Shows family tree structure

Smith Family Curse [SFC]---Autosomal Dominant Nonsyndromic Hereditary Hearing Loss---DFNA/ DFNA1---hereditary hearing loss passed down from George and Margaret Edmonstone Buchanan through at least seventeen more consecutive generations.

Spontaneous Mutation---DFNA hereditary hearing loss that occurs even though both parents have normal hearing—quite rare. Any descendant of Frank & Sarah is at risk.

Tinnitus---Annoying noises that originate inside of head and are heard only by affected person. Includes ringing, ocean sounds, car horns, clicking, & other noises.

CHAPTER 4---STATISTICS

UNIT 13---48 CASES OF REPORTED HEARING LOSS IN FRANK & SARAH'S DESCENDANTS

NAME	GEN	AGE	LOSS
Aeron L,	G6	10	Severe
Allison M,	G6	14	Moderate**
Angie R,	G5	28*	Moderate
Belle R,	G3	75	Mild
Betty G,	G3	82*	Severe
Bob R,	G4	65	Severe**
Brad B,	G5	46	Moderate
Cheryl M,	G4	61	Serious**
Clement S,	G6	23	Severe
Cory M,	G5	49	Severe**
David R,	G5	44	Moderate
Ernest S,	G2	78*	Moderate
Floyd L,	G5	35	Serious
Frances R,	G3	71*	Serious**
Frank S,	G1	56*	Severe
Gladys R,	G4	62	Severe
Grant R,	G6	7	Moderate
Irene G,	G4	68	Serious
James S,	G2	28*	Severe
Jay M,	G5	35	Mild
Jim B,	G4	75	Severe**
John S,	G3	70	Moderate
Judy B,	G4	48	Mild**
Julia B,	G2	75*	Serious
Ken R,	G4	72	Severe**
Lillian M,	G2	74*	Moderate
Luke A,	G6	6	Serious**
Marcus W,	G4	55	Moderate
Marion M,	G3	76*	Serious

Mary S,	G4	70	Severe
Nancy M,	G4	54	Moderate**
Nora S,	G5	52	Moderate
Ola M,	G3	86*	Serious
Randy R,	G5	43	Mild
Robert A	G5	42	Moderate
Rosa F,	G3	83	Serious
Rose H,	G3	47*	Moderate
Rosie C,	G2	78*	Serious
Ruth S,	G6	27	Other
Sarah S,	G1	73*	Moderate/Serious
Shirley B,	G4	49	Mild
Stan M,	G4	59	Severe
Steve M,	G4	56	Moderate
Sue P,	G4	51	Severe
Til B,	G3	70	Moderate
Tom S,	G2	71*	Serious
Wes B,	G4	70	Severe**
William S,	G3	65*	Serious

KEY--- * Age at death
** Degree of hearing loss supported by audiogram
All other degrees of loss are estimated

Note.—My apologies for any errors incurred herein. If you feel that any of these facts are incorrect, please feel free to contact me for discussion. If there are actual mistakes, then changes and apologies will be in order.

CHAPTER 5---RESEARCH RESULTS

UNIT 14---MORE HEARING LOSS SYNDROMES?

Additional DFNA/DFNA1 hearing loss syndromes are possible. Along with the loss of hearing attributed to the Smith Family Curse, there seem to be more ear-

related syndromes popping up. Several such syndromes have recently come to my attention.

A. LOSS OF BALANCE SYNDROME.—Our center of balance is located in the middle ear. Nearly all of the descendants of Grandpa Frank that were tested had a defective sense of balance. The challenge was to try to stand on one foot for 30 seconds with eyes open and note how many seconds before toppling. Then close both eyes and repeat the procedure. If a person topples at 30 seconds with eyes open, it stands to reason that the same person should be able to do the same thing with eyes closed.

B. VERTIGO SYNDROME.—Two descendants of Grandpa Frank have reported having unusual vertigo-like symptoms. One was hospitalized and has serious recurring symptoms which are most unpleasant. The second person seems to have her symptoms under control.

Dizziness, inability to walk, vomiting, depression, panic, and nausea are some of the symptoms of this malady. If you are a descendant of Grandpa Frank, please let us know if you have had any similar experiences.

C. **UNDERSIZE EAR CANAL SYNDROME**.—Many of those affected by the Smith Family Curse were reported to have undersize ear canals. This phenomenon even exists in the descendants of John Alvin Smith who was William Franklin Smith's brother. Further research is definitely needed on this syndrome.

D. BAT SQUEAK SYNDROME.—Several cases of the bat squeak syndrome were encountered. Perhaps this syndrome needs further study also.

E. **FRAGILE HEARING GENE SYNDROME**:-- When exposed to loud noises and/or ENT infections, non-Clan members with normal hearing seldom incur any permanent damage to their hearing. On the other hand, when Clan members with "normal" hearing are exposed to similar loud noises and/or ENT infections, it often causes permanent damage to their hearing. Could this indicate a common fragile hearing gene within our family? Does any Clan member over the age of fifty still have normal hearing? Are all Clan members born with fragile hearing?

UNIT 15---FURTHER RESEARCH SUGGESTED

While collecting and analyzing information, several questions became very intriguing. Puzzles and theories abound, but no definite answers were uncovered. Here are a few of the riddles.

I. Does Right hand + left ear = poor handwriting?--In tallying up the Fact Sheets, it was found that nearly all of us with right hand + left ear combination had poor handwriting! With only one exception, those with same hand + same ear or right hand + equal-ear hearing had nice handwriting. From these figures, it is assumed that the majority of the SFC victims suffer an imbalance of body orientation. It is enough to make you wonder what other unknown problems that we suffer because of this particular imbalance. Are other fine motor skills involved? Unfortunately, few other fine motor skills were studied in this research.

II. What other fine motor skills are involved besides poor handwriting?--- Most people have a dominant right or left side of their bodies. For most persons with a dominant right side, the right half of the body is better developed, more agile, better coordinated, stronger, quicker, often larger, more acute, and usually favored over the left side of their body. Vice versa for the majority of people with a dominant left side.

The person with a dominant right side of his body normally has better vision in the right eye, better hearing in the right ear, will be right-handed, lead with the right foot, and have more strength in the right arm, right hand, and right leg. The right hand, arm, leg, and foot are usually slightly larger and better developed than those on the left side of the body. If you don't believe this, just try switching your wedding ring over to your right hand! Doesn't fit, does it!

Vice Versa for the person with a dominant left side of body.

Other fine motor skills: Is there a correlation between right hand/left ear imbalance and fine motor skills, or is it just a strange coincidence? If you believe it to be a coincidence, you would have trouble trying to prove it with the facts that were uncovered in this Smith Family Curse Research Report!

III. Are all Clan members born with fragile hearing genes ?— According the experts, 50% of offspring born to a DFNA hearing impaired parent will have genetic hearing disorders and the other 50% will have normal hearing. Then why is it next to impossible to find a single Clan member by the age of 50 who still has normal hearing? Some blame injury—some blame disease---others blame old age for their hearing problems. Some even deny that a hearing loss exists! But nearly every descendent of Grandpa Frank seems to need multiple repeats before age 60!

Fragile and/or defective hearing genes: After studying all of the information provided by various family members, it becomes obvious that we have too many members who attribute their poor hearing to injury—mostly to loud noise. They state that it was after being exposed to loud noises that they first noticed their hearing was going downhill.

Major permanent hearing damage: People with normal hearing may incur little or no hearing loss when exposed to the same level of loud noise that will cause major permanent hearing loss in a Clan member. While very little or no harm comes to the ordinary person's hearing, the Clan member may suffer major, permanent damage. In other words, the same amount of noise that can put a Clan member half deaf is taken in stride by the person with normal hearing. Could it be that ordinary people are born with much sturdier hearing genes, and Clan members are all born with extremely fragile hearing genes? Are we all predisposed to hearing loss?

IV. Are some of us deaf Geniuses?—Did you ever notice how many of the Clan members who are noticeably hearing impaired have a very high IQ? One member asked me if there is a correlation between hearing loss and higher intellect. Is our higher intellect inherited along with the deafness? Do deafness and genius go hand in glove? Are the deaf genes somehow interconnected with the higher intelligence genes? Are SFC victims actually born with greater intelligence or do they acquire their greater intellect while trying to cope in a hearing world? So, is our higher intelligence hereditary or survival of the fittest? Answer? I have no earthly idea!

V. Do poor balance and poor hearing go hand in glove?-- Are the victims of the SFC doomed to poor balance? Your center of balance is located in your middle

ear. Try this test: a) Stand unaided on one foot for 30 seconds with your eyes open. b) Close both eyes and stand unaided on one foot for 30 seconds. Ideally, you should be able to stand the same length of time whether your eyes are opened or closed. A perfect score would be "30/30". How did you do?

Did you topple over when your eyes were closed? Or did you manage to stand 30 seconds with your eyes closed just as you did with your eyes opened? If you toppled sooner with your eyes closed, you have a problem with your center of balance. Could this be another syndrome of the Smith Family Curse? Sounds likely to me.

VI. Are a few of us victims of spontaneous mutation?—The experts currently believe that spontaneous mutation hereditary hearing loss occurs after several generations of supposedly normal hearing---a throw-back, so to speak. When checking back a few more generations, great and/or great-great-grandparents are usually found who had a hereditary hearing loss. The family has virtually forgotten that many years ago their ancestors had a hearing loss. Then out of the blue, a child is born with poor hearing although both parents and all four grandparents were believed to have normal hearing. Could it be that one of the child's parents and one of the child's grandparents might have had a mild hereditary hearing loss that had gone unnoticed? Even with an unnoticed mild hearing loss, the parent and grandparent could still pass down the hereditary hearing loss gene. Perhaps further study is needed on this question.

Another possible explanation: A Smith parent with a mild hereditary hearing loss plus a non-Smith parent with a mild hereditary hearing loss could produce an offspring with an obvious hearing loss. Many cases of mild hearing loss go unnoticed. Therefore, what was diagnosed as "spontaneous mutation" was actually hereditary hearing loss after all. During my research, this situation was suspected in three family members.

VII. Do non-Smith spouses contribute to our hereditary hearing loss gene pool?
While researching the hereditary hearing loss within the Clan, it was noted that several of the non-Smith parents admitted to having a hearing loss as well as their Smith Clan spouse. Could these hearing impaired non-Smith parents be the ones responsible for the hereditary hearing loss in some instances? Could these hearing impaired non-Smith parents have contributed to the already existing

problem and made it worse? Are we to blame Grandpa George and his wife for all of the Smith Clan's hereditary hearing loss problems—or did they get some help from the outside a few times?

VIII. Are early audiograms needed?—If every member of the Clan were to be given an audiogram at age 10 and every ten years thereafter, it is likely that there would be very few of us who did not register at least a mild hearing loss by age 20. Many people suffer a mild loss and are never aware of it. When a Clan member with the mild hearing loss gets exposed to the loud noises and loses half of his hearing, he insists that it was injury that made him deaf---not the Smith Family Curse. He feels that his hearing was perfect prior to the noise injury.

IX. Is DNA testing suggested?—Along with those audiograms every 10 years, would it be advisable to have a DNA test made on all Clan members? Experts are able to differentiate a normal hearing gene from a defective gene. We just might learn that every Clan member is born with at least some degree o potential hearing loss! Perhaps we all have at least a few warped hearing genes!

How long will these questions go unanswered? Sorry, folks, but the answers to these questions were not discovered during the research. Perhaps someday the experts will solve these puzzles---maybe DNA and audiograms will help to clear the air. **Let's hope so**.--

UNIT 16---SMITH FAMILY CURSE IN A NUTSHELL

a) It is a form of hereditary nerve deafness

b) It is called: "Autosomal Dominant Nonsyndromic Sensorineural Hereditary Hearing Loss"

c) Its acronym is "DFNA" and/or "DFNA1".

d) It is passed down from parent to offspring.

e) It retains its potency indefinitely.

f) Only one parent is needed to pass the gene down to his/her offspring.

g) Approximately 50% of these offspring will demonstrate a hearing loss.

h) Approximately 50% of these offspring will show no symptoms.

i) Rule of thumb: An afflicted offspring will have at least one affected parent. [See "m" below for exception to this rule.]

j) DFNA victims may demonstrate any degree of hearing loss.

k) DFNA symptoms may be evident at birth or may not become noticeable until several years later.

l) Children afflicted with DFNA are seldom born totally deaf.

m) DFNA spontaneous mutation can occur after several generations with no demonstrated hearing loss.

UNIT 17---PURPOSE OF THIS REPORT---FROM A TO Z

This information was documented for the following reasons:

a) To assemble important information before it is totally forgotten;

b) To share information among the victims of the Smith Family Curse;

c) To provide a foundation for professional researchers who may wish to do further study into the intricacies of the Smith Family Curse type of hereditary hearing loss as well as other types of hereditary anomalies plaguing the Smith Clan;

d) To benefit future research in the field of hereditary hearing loss;

e) To help to alleviate unnecessary suffering by those already afflicted with hereditary hearing loss;

f) To end the unhealthy shame felt by many hearing impaired relatives, help them to become whole and get on with their lives;

g) To encourage self-confidence and assertiveness in those with hearing loss;

h) To encourage clan members to consult with genetic advisors prior to procreating;

i) To educate the entire Smith Clan about this type of hearing loss;

j) To encourage more and better communication among afflicted family members;

k) To form support groups for the hearing impaired within the family;

l) To help those purchasing hearing aids to select the aids best suited for their needs;

m) To help those purchasing hearing aids to get the most for their money;

n) To assist family members in their choice of career opportunities that are available, advisable, and suitable for the hearing impaired;

o) To make family members aware of the ADA, which protects the rights of all, handicapped citizens;

p) To provide genealogical information for those who are interested in learning more about the structure of the Smith Clan;

q) To help clan members become more keenly aware of their roots, which have been hidden from view for many years;

r) To aid Audiologists when fitting hearing aids for our DFNA-afflicted clansmen;

s) To benefit ENT doctors when diagnosing our DFNA hearing loss. [Be sure to take a copy of this report with you when you visit your audiologist and/or ENT doctor;

t) To help Genetic counselors who are trying to determine the advisability of propagation by descendants of William Franklin Smith.

u) To aid victims of the Smith Family Curse by helping them to understand their difficult situation and better cope with their handicap;

v) To help family members with normal hearing understand the problems facing the hearing impaired members of the family;

w) To encourage more communication between family members with normal hearing and those persons with hearing problems;

x) To assist future generations of Clan members who become afflicted with hereditary hearing loss;

y) To help educate those with normal hearing to improve their communications skills with the hearing impaired members of the Clan; and

z) To assure those afflicted with the Smith Family Curse that they are not alone---to let them know that they have many other relatives similarly afflicted---and to inform them that the rest of us care and understand how they feel---to let them know that they can turn to us for help.

UNIT 18---SUMMATION OF FINDINGS

CONCLUSION: A high percentage of family members who have descended from two common ancestors demonstrate varying degrees of hearing loss.

The following facts were uncovered during my research:

1. The hereditary hearing loss within the Smith Clan has been diagnosed as "Autosomal Dominant Nonsyndromic Sensorineural Hereditary Hearing Loss".

2. In the Smith Clan, the Autosomal Dominant Nonsyndromic Sensorineural Hereditary Hearing Loss [DFNA/DFNA1] is usually referred to as "The Smith Family Curse" or "SFC" for short.

3. This research report covers eighteen generations beginning with two common ancestors (George and Margaret Buchanan). It spans 521 years---from 1490 to 2011; however, most of this report deals only with the descendants of Frank and Sarah Smith. [1864—2011]

4. Evidence of the SFC gene was originally traced back to William Franklin [Grandpa Frank] Smith, born 1865---died 1921. It was later determined that Frank's wife had the same hearing loss gene. This fact allowed the research to go back another twelve generations—to 1490.

5. Grandpa Frank and his wife, Sarah, serve as our most common documented ancestors for the majority of this research report. They were first cousins once removed, and it is firmly believed that inbreeding strengthens the hearing loss gene.

6. It is now known that Frank inherited his hearing loss from his mother, Julia McConnell. Sarah inherited her hearing loss gene from her father, John Montgomery Buchanan. Both Julia and John are descendents of George and Margaret Buchanan, both born in 1490.

7. It has been documented that Frank and Sarah passed their hereditary hearing loss down to their offspring, grandchildren, great-grandchildren, great-great-grandchildren, and great-great-great grandchildren.

8. It is not yet known whether any of their great-great-great-great grandchildren are also affected. Most likely some will be. Give it a little time.

9. Frank had a severe hearing loss. {Generation 1} Sarah's hearing loss was not as noticeable as Frank's loss. Frank's loss is believed to have been between severe and profound. Sarah's loss is thought to be between moderate and serious.

10. All six of Frank and Sarah's adult children demonstrated a definite hearing loss ranging from moderate to severe. {Generation 2}

11. Ten of Frank and Sarah's eighteen grandchildren are reported to have hearing losses ranging from mild to severe. {Generation 3}

12. Fifteen of Frank and Sarah's great-grandchildren have been found to have hearing losses ranging from moderate to severe. {Generation 4}

13. Nine of Frank and Sarah's great-great-grandchildren have been found to have a hearing loss ranging from mild to severe. {Generation 5}

14. Six of Frank and Sarah's great-great-great-grandchildren have been found to have a hearing loss ranging from moderate to severe. {Generation 6}

15. It is believed that many of Frank and Sarah's fifth and sixth generational descendants are either unaware of their hearing loss or are in denial.

16. The descendants of Frank and Sarah's oldest son, Tom, appear to be the most seriously affected.

17. Four of Tom's six children reported a hearing loss ranging from moderate to severe.

18. Eleven of Tom's twenty grandchildren reported hearing losses ranging from mild to severe.

19. Of the 48 cases of hearing loss investigated, 6 were found to have a mild hearing loss; 16 were found to have a moderate hearing loss; 11 were found to suffer a serious hearing loss; 13 were found to have a severe hearing loss; however, none was found with profound hearing loss.

20. One example of a six-generational study of consecutive hearing loss was documented as follows. Frank/Sarah---their son (serious loss)--- their granddaughter (serious loss)---their great-granddaughter (moderate loss)---their great-great-granddaughter (moderate loss)--- their great-great-great-grandson

(severe loss). This finding indicates that the hereditary hearing loss gene remains just as virulent even after being passed down through six generations.

21. Another example of a six -generational study of consecutive hearing loss was documented as follows: Frank/Sarah--- son (serious loss)--- granddaughter (serious loss)---great-grandson (severe loss)---great-great-granddaughter (severe loss)---great-great-great-granddaughter (moderate loss).

22. Four cases of possible spontaneous mutation were discovered.

23. Many SFC victims have a rather unique audiogram pattern, known as DFNA1 or the reverse ski slope. [DFNA1 family members have greater loss in the lower tones and hear somewhat better in the higher tones.]

24. The "cookie bite pattern" is evident in all of our audiograms.

25. Audiograms show overall average hearing losses to range from less than 1% to 76%.

26. Audiograms show hearing losses to range from 40 dB to 90 dB.

27. Many affected family members suffer from tinnitus.

28. Most affected family members are right handed but hear better with their left ear, making for a lopsided sensory balance.

29. Most family members affected in the above manner have poor handwriting skills.

30. Most affected family members have an impaired sense of balance which is located in the middle ear.

31. Nearly all affected family members have mates with normal hearing.

32. Several occurrences were noted in which a hearing impaired Clan member chose a mate who was also hearing impaired. Most instances resulted in a hearing loss by 100% of their offspring instead of the expected 50% with normal

hearing and 50% with hereditary hearing loss. [See Units 34, 35, 50, 60, 70, 75, and 108.] It appears that when a DFNA/DFNA1 candidate mated with a person with normal hearing that the 50% rule is followed. When a DFNA/DFNA1 candidate mated with a person with similar hearing loss then the 50% rule changed into a 100% rule, and all offspring were afflicted with DFNA/DFNA1. When two related DFNA/DFNA1 mates produced children, the situation became even more serious.

33. The "bat squeak syndrome" has been noticed in several family members who are able to hear ultra high-pitched sounds that should be inaudible to human ears. This trait usually lessens with age.

34. Many family members afflicted with SFC have gained some relief by using hearing aids.

35. Hearing aids have failed to help several family members. Perhaps the audiologists have been fitting them with standard aids when they actually need DFNA1 aids.

36. Many other affected family members would probably benefit by wearing hearing aids.

37. Only one family member reported using ASL and/or reading lips as a substitute for hearing aids.

38. No affected family member was found to have profound hearing loss. [Over 90dB loss]

39. Several family members attributed their hearing loss to disease, age and/or injury. Heredity is suspected in all instances.

40. Very few of Grandpa Frank's descendants reach the age of 50 without some form of hearing loss.

41. It has recently been discovered that John Alvin Smith (Grandpa Frank's only full sibling) has numerous descendants with DFNA/DFNA1 hearing impairment. Further research on this subject is underway.

42. One family member had cochlear implant surgery which failed.

43. Many affected family members found an excellent support group at the 2008, 2010, and 2011 Clan Reunions.

44. Hereditary hearing loss was a key topic at all three Clan Reunions.

45. A factual book has been written by our cousin, Wes Brosman, about his early life with a severe hereditary hearing loss. It is a must read for all descendants of Grandpa Frank. For information on obtaining this book, contact: wesbro@techline.com and request *No Place Else.*

46. Anyone needing excellent personal advice about the purchase and/or use of hearing aids should contact Wes Brosman at: wesbro@techline.com. He would love to hear from you.

47. Cousin Wes is also very knowledgeable about the Americans with Disabilities Act. Feel free to contact him for information and advice on this subject. wesbro@techline.com

48. Professional researchers are encouraged to contact the author if they are interested in using these findings for further hearing loss study and/or research.

49. To obtain a free CD, or inquire about purchasing a hard copy of the SFC, to ask questions, or make comments about this research, contact the author at mrsimms@peoplepc.com. I would love to hear from you!

NOTE TO FAMILY MEMBERS:

When seeking medical services of an ENT doctor or Audiologist be sure to carry a copy of this report with you, and do not fail to share it with your health care professional.

UNIT 19---REFERENCES

[No authors listed]. ***CIGNA HealthCare Coverage Position. Genetic Testing for Congenital, Profound Deafness***, Coverage Position Number 0254, revised 12/15/2007.

[No authors listed]. ***The Essential Guide to Hearing Loss, Part 2: Understanding Audiograms, Hearing Loss, and Speech Intelligibility***, Hound Dog Hearing, 2002. www.hdhearing.com/Learning/Part2.htm.

[No authors listed]. ***Your Genes and Hearing Loss***, Colorado Adult and Children's Ear, Nose, and Throat, P.O., 2004 AAO-HNS/AAO-HNSF. www.drtomaski.com/Pages/PatientEducation/1028.html.

Berke J. ***Connexin 26---Genetic Cause of Hearing Loss***, ***Your Guide to Deafness,*** The New York Times Company, 2007. http://deafness.about.com/od/diseasesandsyndromes/a/connexin…

Boys Town National Research Hospital. Information on Hearing loss – **Genetics and Deafness – Autosomal Dominant Inheritance**. http//www.boystownhospital.org/Hearing/info/genetics/concepts/auto_dom.asp

Cordes, S., Genetic Hearing Loss**,** Grand Rounds Presentation**, UTMB, Dept. of Otolaryngology, April 5, 2000. www.otohns.net/default.asp?id=14091**

Halpin, Khetarpal, McKenna, ***Autosomal-Dominant Progressive Sensorineural Hearing Loss in a Large North American Family***, *American Journal of Audiology*, V 5, No. 1, 1996

Keats, B. (2005, September 6). ***Genetics and Hearing Loss***, The ASHA Leader, pp. 6-7, 16-18. www.asha.org/about/publications/leader-online/archives/…

Mehr A. ***Understanding Your Audiogram,*** Pacific Audiology, Salem, Oregon. www.pacificaudiology.com/audiogram/uya.html.-
Harvard University, www.hearing.harvard.edu/info/genetics.pdf

Smith J, Camp G. ***Deafness and Hereditary Hearing Loss Overview***. ***Gene Review.*** Funded by the NIH. Developed at the University of Washington, Seattle.

Initial posting 1999 Feb 14. Last Update: 30 January 2007. Accessed October 19, 2007.
www.geneclinics.org/profiles/deafness-overview/details.html

Smith J, Camp G. ***Deafness and Hereditary Hearing Loss Overview***. ***Gene Review.*** Funded by the NIH. Developed at the University of Washington, Seattle. Initial posting 1999 Feb 14. Last Update: 30 January 2007. Accessed October 19, 2007. http://www.ncbi.nlm.nih.gov/bookshelf/br.fcgi?book=gene&part=deafness-overview

Turner A, Smolenyak M. ***Trace your Roots with DNA***. Rodale Press. October 18, 2004. DNACousins@aol.com

World Book Encyclopedia, World Book Inc., Chicago IL. 1994.

CHAPTER 6---INTERVIEWS

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A Fact Sheet was prepared on each of the 48 Clan members reported to have a hearing loss syndrome. Most of the information was obtained from the

person involved; however, some Fact Sheets had to be completed by proxy.

Many thanks to those family members with a hearing impairment who shared private and personal information to benefit this hearing loss research. We are grateful to those who willingly granted these interviews.

My sincere apologies for any errors in these Fact Sheets. If any serious mistakes are found, please notify me and forward a current audiogram for the person involved.

UNIT 21---AERON L, G6, AGE 10

[FRANK/SARAH---TOM--- MARION---SUE---FLOYD---AERON]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Sixth consecutive generation with hearing loss
   c) Siblings—0 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---No
   f) Hearing aids---No
   g) Audiogram--- None available
   h) Other---90% loss on sense of balance test---

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM--- MARION---SUE---FLOYD---AERON]
   b) See Frank & Sarah S, G1--- Tom S, G2---Marion M, G3---Sue P, G4--- Floyd L, G5

3. Other reported causes: Disease

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Disease-related hearing loss

5. Personal comment: FEELS LIKE BUBBLE IN MY EAR AND I CAN'T POP IT

UNIT 22---ALLISON M, G6, AGE 14

[FRANK/SARAH---TOM---OLA---WES---CORY---ALLISON]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Sixth consecutive generation with hearing loss
   c) Siblings— 0 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Sometimes
   f) Hearing aids---No
   g) Audiogram---Shows 3% loss, DFNA1 slope, cookie bite pattern, and 55 dB loss

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---WES---CORY---ALLISON]
   b) See Frank & Sarah S, G1--- Tom S, G2---Ola M, G3---Wes B, G4---Cory M, G5

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 23---ANGIE R, G5, AGE 28 AT DEATH

[FRANK/SARAH---TOM---FRANCES---GLADYS---ANGIE] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings—0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring--None

e) Tinnitus---Not reported
f) Hearing aids---None
g) Audiogram---None available
h) Other---DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---FRANCES---GLADYS---ANGIE]
   b) See Frank & Sarah S, G1--- Tom S G2---Frances R, G3---Gladys R, G4

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 24---BELLE R, G3, AGE 75

[FRANK/SARAH---ERNEST---BELLE] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Moderate
   b) Family history---Third consecutive generation with hearing loss
   c) Siblings—1 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 3 with no reported hearing loss
   e) Tinnitus---None reported
   f) Hearing aids---No
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST---BELLE]
   b) See Frank & Sarah S, G1---ERNEST S, G2

3. Other reported causes: Age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss

UNIT 25---BETTY G, G3, AGE 82 AT DEATH

[FRANK/SARAH---JULIA---BETTY] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Third consecutive generation with hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Some
   f) Hearing aids---2, BTE
   g) Audiogram---None available
   h) Other---DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---JULIA---BETTY]
   b) See Frank & Sarah S, G1---Julia B, G2

3. Other reported causes: Loud noise, age

4. Possible diagnosis:
   a) Hereditary hearing los
   b) Injury-related hearing loss
   c) Age-related hearing loss

UNIT 26---BOB R, G4, AGE 65

[FRANK/SARAH---TOM---FRANCES---BOB]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss—Severe
   b) Family history---Fourth consecutive generation with hearing loss
   c) Siblings—3 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 0 with no reported hearing loss
   e) Tinnitus---Severe
   f) Hearing aids---1 BTE

g) Audiogram---Shows 57% loss, DFNA1 slope, cookie bite pattern, and 85 dB hearing loss
h) Other---DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---FRANCES---BOB]
   b) See Frank & Sarah S, G1 ---Tom S, G2---Frances R, G3

3. Other reported causes: Loud noises, underwater work as Navy Seal, age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Age-related hearing loss

UNIT 27---BRAD B, G5, AGE 46

[FRANK/SARAH---TOM---OLA---WES---BRAD]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus--- Yes
   f) Hearing aids---None
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---WES---BRAD]
   b) See Frank & Sarah S, G1 --- Tom S, G2---Ola M, G3---Wes B, G4

3. Other reported causes: Loud noises

4. Possible diagnosis:
   a) Injury-related hearing loss

b) Hereditary hearing loss

5. Personal comments? YES-- I TRUTHFULLY THINK THAT MY HEARING LOSS IS DUE TO LOUD TOOLS AND MUSIC MORE THAN ANYTHING ELSE. YOU WOULD NOT BELIEVE THE HOURS I'VE SPENT WITH A 90-POUND JACKHAMMER, HEAVY EQUIPMENT, & NAIL GUNS. DAD WILL CONFIRM THAT I DID HEAVY GNARLY WORK IN THE TRADES. I FEEL THAT I WAS TAKEN ADVANTAGE OF DUE TO MY SIZE. I AM WELL INTO MY 40S AND FEEL BLESSED TO HAVE LASTED THAT LONG.

UNIT 28---CHERYL M, G4, AGE 61

[FRANK/SARAH---JULIA---BETTY---CHERYL]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Serious
   b) Family history---Fourth consecutive generation with hearing loss
   c) Siblings—0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Some
   f) Hearing aids---Yes, 2 BTE
   g) Audiogram---Shows 19% loss, DFNA1 slope, cookie bite pattern, and 70 dB loss
   h) Other---Bat squeaks

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---JULIA---BETTY---CHERYL]
   b) See Frank & Sarah S, G1 ---Julia B, G2---Betty G, G3

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 29---CLEMENT S, G6, AGE 23

[FRANK/SARAH---TOM---OLA---IRENE---NORA---CLEMENT]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Sixth consecutive generation with hearing loss
   c) Siblings—0 with reported hearing loss, 1 with undersize ear canals, but no reported hearing loss
   d) Offspring--None
   e) Tinnitus---No
   f) Hearing aids---Yes, 2 ITC
   g) Audiogram---None available
   h) Other---DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---IRENE---NORA---CLEMENT
   b) See Frank & Sarah S, G1---Tom S, G2---Ola M, G3---Irene G, G4---Nora S,G5

3. Other reported causes: Severe ear infections

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Disease-related hearing loss

5. Personal comments? YES---I ALWAYS HAVE HAD THE HEARING PROBLEM SO I AM RESIGNED TO IT.MY PARENTS FIRST NOTICED MY HEARING PROBLEM WHEN I BEGAN TO SPEAK. I MADE UP WORDS FOR WATER, FOOD, ETC. THIS CAUSED THEM TO DOUBT MYINTELLIGENCE, SO THEY GOT A DOCTOR TO CHECK ME OUT. HE TOLD THEM THAT I HAD SEVERE HEARING PROBLEMS.

I ALSO HAD HORRIBLE EAR INFECTIONS AS A CHILD; SOME WERE SO BAD THAT MY EARS WOULD ACTUALLY BLEED.

I DID EVENTUALLY PICK UP THE ENGLISH LANGUAGE BUT HAD A SPEECH IMPEDIMENT TILL MY 14TH YEAR. I STILL HAVE A HARD TIME HEARING PEOPLE

AND IT AFFECTS MY ABILITY TO COMMUNICATE ON THE PHONE OR IN CROWDED SITUATIONS.

I AM IN A BAND, AND I DO QUITE WELL; HOWEVER, SINCE I CAN'T HEAR I HAVE TO WATCH THE HANDS OF THE OTHER MUSICIANS CONSTANTLY IN ORDER TO KNOW WHERE WE ARE IN A SONG.

UNIT 30---CORY M, G5, AGE 49

[FRANK/SARAH---TOM---OLA---WES---CORY]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 2 with no reported hearing loss
   e) Tinnitus---Moderate, both ears, continuous
   f) Hearing aids---2 BTE
   g) Audiogram---Shows 59% loss, DFNA1 slope, cookie bite pattern and 75 dB loss
   h) Other---DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---WES---CORY]
   b) See Frank & Sarah S, G1 ---Tom S, G2---Ola M, G3---Wes B, G4

3. Other reported causes: Injury, disease, age progressive

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Disease-related hearing loss
   d) Age-progressive hearing loss

5. Personal comments? YES---MY FRIENDSHIPS ARE LIMITED--MY SCHOOLING WAS DIFFICULT---I AM SOMEWHAT RECLUSIVE---MY SOCIAL LIFE IS AWKWARD---MY JOB CHOICES ARE EXTREMELY LIMITED.

UNIT 31---DAVID R, G5, AGE 44

[FRANK/SARAH---TOM---FRANCES---KEN---DAVID] [by proxy]

1. Reported hereditary hearing loss syndromes
   a) Hearing loss--- Moderate
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings—0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---FRANCES---KEN---DAVID]
   b) See Frank & Sarah S, G1 --- Tom S, G2---Frances R, G3---Ken R, G4

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 32---ERNEST S, G2, AGE 78 AT DEATH

[FRANK/SARAH---ERNEST] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Moderate
   b) Family history---Second generation with hearing loss
   c) Siblings—5 with reported hearing loss, 0 with no reported hearing loss

d) Offspring—2 with reported hearing loss, 2 with no reported hearing loss
e) Tinnitus--- Unknown
f) Hearing aids---None
g) Audiogram---None available
h) Other---Both parents had reported hearing loss---[first cousins once removed]

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST]
   b) See Frank & Sarah S, G1

3. Other reported causes: Loud noise, age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

UNIT 33---FLOYD L, G5, AGE 35

[FRANK/SARAH---TOM---MARION---SUE---FLOYD] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings— 0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Yes---very bad
   f) Hearing aids---None
   g) Audiogram---None available
   h) Other--- 90% loss on sense of balance test--- DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---MARION---SUE---FLOYD]
   b) See Frank & Sarah S, G1---Tom S, G2---Marion M, G3---Sue P, G4

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 34---FRANCES R, G3, AGE 71 AT DEATH

[FRANK/SARAH---TOM---FRANCES] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring—4 with reported hearing loss, 0 with no reported hearing loss
   e) Tinnitus---Unreported
   f) Hearing aids---2 ITE
   g) Audiogram---Shows 35% loss, DFNA1 slope, cookie bite pattern, and 65 dB loss
   h) Other---Both Frances and her husband had a reported hearing loss---DFNA1 pattern indicated

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM---FRANCES]
   b) See Frank &Sarah S, G1---Tom S, G2

3. Other reported causes: Age, loud noise

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

UNIT 35---FRANK S, G1, AGE 56 AT DEATH

[MOST COMMON ANCESTOR] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Severe
   b) Family history---6 children, 9 grandchildren, 15 great grandchildren, 8 great-great grandchildren, 6 great-great-great grandchildren known to have hearing loss
   c) Siblings— 1 full sibling and 3 half siblings with unknown diagnosis
   d) Offspring—6 with reported hearing loss---0 with no reported hearing loss
   e) Tinnitus--- Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Both Frank and his wife had a reported hearing loss---they were cousins

2. Placed number 13 in 18 generations of consecutive hearing loss: [GEORGE 1490---THOMAS 1510---JOHN 1545---GEORGE 1576---JOHN 1629---ALEXANDER 1670---SAMUEL 1690---MATTHEW 1725---ELIZABETH 1750---JOHN M 1778---ELIZABETH 1816---JULIA 1847---FRANK 1865]

3. Other reported causes: Injury from firearms possible, ear infections, possibly age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Disease-related hearing loss
   d) Age-related hearing loss

UNIT 36---GLADYS R, G4, AGE 62

[FRANK/SARAH---TOM---FRANCES---GLADYS] [by proxy]

1. Reported hereditary hearing loss syndromes:

a) Hearing loss---Severe
b) Family history---Fourth consecutive generation of hearing loss
c) Siblings—3 with reported hearing loss, 0 with no reported hearing loss
d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
e) Tinnitus---Severe---especially after cochlear implant
f) Hearing aids---2 aids
g) Audiogram---None available
h) DFNA1 hearing slope indicated
i) Undersize ear canals
j) Other---Bat squeaks—Both parents had reported hearing loss

2. Consecutive chain of hearing loss:
   a. [FRANK/SARAH---TOM---FANCES—GLADYS]
   b. See Frank & Sarah S, G1 --- Tom S, G2, Frances R, G3

3. Other reported causes: Age, failed cochlear implant

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

UNIT 37---GRANT R, G6, AGE 7

[FRANK/SARAH---ERNEST---ADA---BRAD---CHELSEA---GRANT]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--Moderate-
   b) Family history---Great-great grandfather & great-great-great grandfather with hearing loss
   c) Siblings— 0 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST---(skip 3 generations)---GRANT]
   b) See Frank & Sarah S, G1 --- Ernest S, G2

3. Other reported causes: Tubes in ears, ear infections

4. Possible diagnosis:
   a) Disease-related hearing loss
   b) Injury-related hearing loss
   c) Spontaneous mutation hearing loss
   d) Hereditary hearing loss from non-Smith side of family

UNIT 38---IRENE G, G4, AGE 68

[FRANK/SARAH---TOM---OLA---IRENE]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---Used for work only
   g) Audiogram---None available
   h) Other---Bad tonsils/adenoids---One undersize ear canal---
      DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH ---TOM---OLA---IRENE]
   b) See Frank & Sarah S, G1 --- Tom S, G2---Ola M, G3

3. Other reported causes: Ear infections during childhood, age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Disease-related hearing loss
   c) Age-related hearing loss

UNIT 39---JAMES S, G2, AGE 28 AT DEATH

[FRANK/SARAH---JAMES] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Second generation with hearing loss
   c) Siblings—5 with reported hearing loss---0 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Both parents had reported hearing loss---[first cousins once removed]

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---JAMES]
   b) See Frank & Sarah S, G1

3. Other reported causes: Perhaps injury &/or disease

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Disease-related hearing loss

UNIT 40---JAY M, G5, AGE 35

[FRANK/SARAH---JULIA---BETTY---CHERYL---JAY] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Mild
   b) Family history---Fifth consecutive generation of hearing loss
   c) Siblings—0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 2 with no reported hearing loss

e) Tinnitus---Unknown
f) Hearing aids---No
g) Audiogram---Hearing loss not noticed until detected by audiogram
h) Other--- DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---JULIA---BETTY---CHERYL---JAY]
   b) See Frank & Sarah S, G1 ---Julia, G2---Betty, G3---Cheryl, G4

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

5. Personal comment? YES---SO FAR NO BIG PROBLEM. ONLY SHOWED UP ON AUDIOGRAM AT TIME OF LEAVING AIR FORCE. UNITL THEN HEARING LOSS HAD NOT YET BEEN NOTICED.

UNIT 41---JIM B, G4, AGE 75

[FRANK/SARAH---TOM---OLA---JIM]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—2 with reported hearing loss---1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 3 with no reported hearing loss
   e) Tinnitus---No
   f) Hearing aids---2---ITC
   g) Audiogram---Shows 80% loss, cookie bite pattern, and 80 dB loss
   h) Other---100% loss on sense of balance test

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---JIM]
   b) See Frank & Sarah S, G1 --- Tom S, G2---Ola M, G3

3. Other reported causes: 4 years of exposure to military artillery noise, age

4. Possible diagnosis:
   a) Injury-related hearing loss
   b) Hereditary hearing loss
   c) Age-related hearing loss

UNIT 42---JOHN S, G3, AGE 70

[FRANK/SARAH---ERNEST---JOHN]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Moderate
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—1 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 4 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST ---JOHN]
   b) See Frank & Sarah S, G1--- Ernest S, G2

3. Other reported causes: Loud noises from farm machinery

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss

UNIT 43---JUDY B, G4, AGE 48

[FRANK/SARAH---ROSA---AMY---JUDY]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--Mild
   b) Family history---Grandmother & great grandfather had hearing loss
   c) Siblings—1 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Yes, left ear
   f) Hearing aids---No
   g) Audiogram---Shows <1% hearing loss, cookie bite pattern, and 40 dB hearing loss

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ROSIE---skip 1 generation---JUDY]
   b) See Frank & Sarah S, G1---Rosie C, G3

3. Other reported causes: Father has serious loss. Did mother have unnoticed mild hearing loss?

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Spontaneous mutation
   c) Disease-related hearing loss

5. Personal comments? I HOPE TO SEE A SPECIALIST IN LARGE CITY THIS SUMMER CONCERNING DIZZINESS DUE TO INNER EAR PROBLEM, POSSIBLE SEIZURES.

UNIT 44---JULIA B, G2, AGE 75 AT DEATH

[FRANK/SARAH---JULIA] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Serious

b) Family history---Second generation of hearing loss
c) Siblings— 5 with reported hearing loss---0 with no reported hearing loss
d) Offspring—3 with reported hearing loss---1 with no reported hearing loss
e) Tinnitus---Unknown
f) Hearing aids---No
g) Audiogram---None available
h) Other---Both parents had reported hearing loss---[first cousins once removed]

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---JULIA]
   b) See Frank & Sarah S, G1

3. Other reported causes: Age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss

UNIT 45---KEN R, G4, AGE 72

[FRANK/SARAH---TOM---FRANCES---KEN]

1. Reported hereditary hearing loss syndromes:
   a. Hearing loss---Severe
   b. Family history---Fourth consecutive generation of hearing loss
   c. Siblings—4 with reported hearing loss, 0 with no reported hearing loss
   d. Offspring—1 with reported hearing loss, 1 with no reported hearing loss
   e. Tinnitus---Yes---intermittent
   f. Hearing aids---Yes—difficulty in adjusting to them
   g. Audiogram---Shows 24% loss, cookie bite pattern, and 75 dB loss
   h. Other---DFNA1 pattern indicated in #5 above

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH—TOM---FRANCES---KEN]
   b) See Frank & Sarah S, G1---Tom S, G2---Frances R, G3

3. Other reported causes: Injury from gunfire earlier in life

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss

UNIT 46---LILLIAN M, G2, AGE 74 AT DEATH

[FRANK/SARAH---LILLIAN] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Second generation with hearing loss
   c) Siblings— 5 with reported hearing loss---0 with no reported hearing loss
   d) Offspring--None
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Both parents had reported hearing loss---[first cousins once removed]

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---LILLIAN]
   b) See Frank & Sarah S, G1

3. Other reported causes: Age

4. Possible diagnosis:

   a) Hereditary hearing loss
   b) Age-related hearing loss

UNIT 47---LUKE A, G6, AGE 6

[FRANK/SARAH---ERNEST---ADA---CAROLYN---RODNEY---LUKE]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Great-great grandfather & great-great-great grandfather had hearing loss
   c) Siblings—Only child
   d) Offspring--None
   e) Tinnitus--- Yes, moderate, intermittent
   f) Hearing aids---Yes, 2 BTE
   g) Audiogram---Shows 29% loss, cookie bite pattern, and 65 dB loss

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST---skip 3 generations---LUKE]
   b) See Frank & Sarah S, G1---Ernest S, G2

3. Other reported causes: None

4. Possible diagnosis:
   a) Spontaneous mutation
   b) Hereditary hearing loss from non-Smith parent

5. Personal comments? YES---LUKE GETS AROUND GREAT AND PLAYS LIKE ANY OTHER KID. IF HE DOESN'T HAVE HIS HEARING AIDS ON MOST KIDS DON'T KNOW THAT HE HAS A HEARING LOSS. [mother's comment]

UNIT 48---MARCUS W, G4, AGE 55

[FRANK/SARAH---ERNEST---BELLE---MARCUS]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Fourth consecutive generation of hearing loss

c) Siblings—0 with reported hearing loss, 3 with no reported hearing loss
d) Offspring—0 with reported hearing loss, 3 with no reported hearing loss,
e) Tinnitus---No
f) Hearing aids---No
g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ERNEST---BELLE---MARCUS]
   b) See Frank & Sarah S, G1---Ernest S, G2---Belle R, G3

3. Other reported causes: Injury from shooting guns

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss

UNIT 49---MARION M, G3, AGE 76 AT DEATH

[FRANK/SARAH---TOM---MARION] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Three consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss---2 with no reported hearing loss
   d) Offspring—4 with reported hearing loss symptoms, 0 with no reported hearing loss syndromes
   e) Tinnitus---Unknown
   f) Hearing aids---Yes, BTE
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---MARION]
   b) See Frank & Sarah S, G1---Tom, G2

3. Other reported causes: Age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss

UNIT 50---MARY S, G4, AGE 70

[FRANK/SARAH---TOM---FRANCES---MARY]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus--- Yes, continuous & moderate---mainly in right ear
   f) Hearing aids---Yes, 2 CIC
   g) Audiogram---Shows 53% loss, DFNA1 slope, cookie bite pattern, and 80 dB loss
   h) Other--- Bat squeaks--- 90% loss on sense of balance test--- Both parents had hearing loss---#5 above indicates DFNA1 pattern

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH—TOM---FRANCES---MARY]
   b) See Frank & Sarah S, G1---Tom, G2---Frances, G3

3. Other reported causes: Age, loud noises, Head trauma--- Undersize ear canals

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

5. Personal comments? YES---SINCE BOTH MY MOTHER AND FATHER HAD A HEARING LOSS---IS IT ANY WONDER THAT ALL 4 OF THEIR OFFSPRING HAVE A SEVERE HEARING LOSS?

UNIT 51---NANCY M, G4, AGE 54

[FRANK/SARAH---ROSIE---AMY---NANCY]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Mild
   b) Family history---Grandmother & great grandfather had hearing loss
   c) Siblings—1, with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 2 with no reported hearing loss
   e) Tinnitus---No
   f) Hearing aids---No
   g) Audiogram---Shows <1% hearing loss, cookie bite pattern, with 45 dB loss
   h) Other---67% loss on sense of balance test---DFNA1 pattern indicated.

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ROSIE---skip one generation---NANCY]
   b) See Frank & Sarah S, G1---Rosie C, G3

3. Other reported causes: Father has serious loss. Did mother have unnoticed mild hearing loss?

4. Possible diagnosis:
   a) Hereditary hearing loss from non-Smith parent
   b) Hereditary hearing loss from Smith parent
   c) Spontaneous mutation

UNIT 52---NORA S, G5, AGE 52

[FRANK/SARAH---TOM---OLA---IRENE---NORA] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Fifth consecutive generation of hearing loss
   c) Siblings 0with reported hearing loss , 1 with no reported hearing loss

d) Offspring—1 with reported hearing loss, 1 with reported undersize ear canals & no reported hearing loss
e) Tinnitus--- Unknown
f) Hearing aids---No
g) Audiogram---None available
h) Other---Bad tonsils/adenoids problem like mother had--- DFNA1 pattern indicated

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---IRENE---NORA]
   b) See Frank & Sarah S, G1---Tom S, G2---Ola M, G3---Irene G, G4

3. Other reported causes: Ear infections as a child, tonsillitis

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Disease-related hearing loss

UNIT 53---OLA M, G3, AGE 86 AT DEATH

[FRANK/SARAH---TOM---OLA] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring—3 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---Yes---unused
   g) Audiogram---None available

2. Consecutive chain of hearing loss:

   a) [FRANK/SARAH—TOM---OLA]
   b) See Frank & Sarah S, G1---Tom, G2

3. Other reported causes: Child abuse, age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related loss
   c) Injury-related loss

UNIT 54---RANDY R, G5, AGE 43

[FRANK/SARAH--- TOM---FRANCES---BOB---RANDY]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Mild
   b) Family history---Fifth consecutive generation with hearing loss
   c) Siblings—Only child
   d) Offspring—0 with reported hearing loss, 3 with no reported hearing loss
   e) Tinnitus---Severe
   f) Hearing aids---No
   g) Audiogram---Shows <1% hearing loss, cookie bite pattern, with 40 dB loss
   h) Other---Vertigo---Nausea---90% loss on sense of balance test

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH—TOM---FRANCES---BOB---RANDY]-
   b) See Frank & Sarah S, G1---Tom S, G2---Frances R, G3---Bob R, G4

3. Other reported causes: Loud noises during military duty in Afghanistan

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss

UNIT 55---ROBERT A, G5, AGE 42

[FRANK/SARAH---ERNEST---ADA---CAROLYN---ROBERT]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss-- -Moderate
   b) Family history--- Great grandfather & great-great grandfather had hearing loss
   c) Siblings—0 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 2 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids--- none
   g) Audiogram---None available

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH--- ERNEST---{ADA NORMAL}---{CAROLYN NORMAL}--- ROBERT]
   b) See Frank & Sarah S, G1---Ernest, G2

3. Other reported causes: Loud noise

4. Possible diagnosis:
   a) Injury-related hearing loss
   b) Hereditary hearing los/ Spontaneous mutation

UNIT 56---ROSA F, G3, AGE 83

[FRANK/SARAH---ROSIE---ROSA]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Serious
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—0 with reported hearing loss, 3 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 3 with no reported hearing loss
   e) Tinnitus---Unknown

f) Hearing aids---Yes, both ears BTE
g) Audiogram---None available
h) Other---39% loss on sense of balance test

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH---ROSIE---ROSA]
   b) See Frank & Sarah S, G1---Rosie C, G2

3. Other reported causes: Age

4. Possible diagnosis:
   a) Age-related hearing loss
   b) Hereditary hearing loss

UNIT 57---ROSE H, G3, AGE 47 AT DEATH

[FRANK/SARAH---JULIA---ROSE] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Moderate
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 3 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available
2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH—JULIE---ROSE]
   b) See Frank & Sarah S, G1---Julia B, G2

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss

UNIT 58---ROSIE C, G2, AGE 78 AT DEATH

[FRANK/SARAH---ROSIE] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Second generation of hearing loss
   c) Siblings—5 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—1 with hearing loss, 3 with no reported hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Both parents reported to have hearing loss---[first cousins once removed]

2. Consecutive chain of hearing loss:
   a) [FRANK/SARAH—ROSIE]
   b) See Frank & Sarah S, G1

3. Other reported causes: Age

4. Possible diagnosis:
   a) Age-related hearing loss
   b) Hereditary hearing loss

UNIT 59---RUTH S, G6, AGE 27

[FRANK/SARAH---TOM---OLA---IRENE---NORA---RUTH] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Hearing reported as being normal
   b) Family history--- Five prior generations with hearing loss--- some with undersized canals

c) Siblings—1 with reported hearing loss, 0 with no reported hearing loss
d) Offspring--None
e) Tinnitus--- No
f) Hearing aids---No
g) Audiogram---None available
h) Other---Undersize ear canals similar to mother and grandmother

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---IRENE---NORA]
   b) See Frank & Sarah S, G1---Tom, G2---Ola, G3---Irene, G4---Nora, G5

3. Other reported causes: None

4. Possible diagnosis: Hereditary undersize ear canals

UNIT 60---SARAH S, G1, AGE 73 AT DEATH

[MOST COMMON ANCESTOR] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate to serious
   b) Family history---6 offspring---9 grandchildren---15 great grandchildren---8 great-great grandchildren---6 great-great-great grandchildren with reported hearing loss. Previous 11 generations of continuous hearing loss dating back to 1490.
   c) Siblings--Unknown hearing status
   d) Offspring—6 with reported hearing loss--- 0 with normal hearing
   e) Tinnitus---Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Both Sarah and her husband had a reported hearing loss

2. Number 12 in continuous chain of hearing loss:
   a) [GEORGE 1490---THOMAS 1510---JOHN 1545---GEORGE 1576---JOHN 1629---ALEXANDER 1670---SAMUEL 1690---MATTHEW 1725---ELIZABETH

1750---JOHN M 1778---JOHN L 1834---SARAH 1868]
b) See Unit 108.

3. Other reported causes: Age

4. Possible diagnosis:
   a) Hereditary hearing loss from her non-Smith parents
   b) Age-related hearing loss

UNIT 61---SHIRLEY B, G4, AGE 49

[FRANK/SARAH---TOM---MARION---SHIRLEY] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss—Reported as having some mild hearing loss as a child
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss--- 0 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 2 with no reported hearing loss
   e) Tinnitus--- Unknown
   f) Hearing aids---No
   g) Audiogram---None available
   h) Other---Bat squeaks as child

2. Continuous chain of hearing loss syndromes:
   a) [FRANK/SARAH---TOM---MARION---SHIRLEY]
   b) See Frank & Sarah S, G1---Tom, G2---Marion, G3

3. Other reported causes: None

4. Possible diagnosis: Hereditary hearing loss syndromes

UNIT 62---STAN M, G4, AGE 59

[FRANK/SARAH---TOM---MARION---STAN]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss--- Severe
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Often severe
   f) Hearing aids---2---1 BTE, 1 ITE
   g) Audiogram---None available

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM---MARION---STAN]
   b) See Frank & Sarah S, G1---Tom, G2---Marion, G3

3. Other reported causes: Age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age related hearing loss

5. Personal comments? WOULD LIKE TO SEE RESULTS OF RESEARCH WHEN FINISHED

UNIT 63---STEVE M, G4, AGE 56

[FRANK/SARAH---TOM---MARION---STEVE]

1. Reported hereditary hearing loss syndromes:
   a. Hearing loss---Moderate
   b. Family history---Fourth consecutive generation of hearing loss
   c. Siblings—3 with reported hearing loss, 0 with no reported hearing loss
   d. Offspring—0 with reported hearing loss, 2 with no reported hearing loss

e. Tinnitus---No
f. Hearing aids---No
g. Audiogram---None available

2. Continuous chain of hearing loss:

a) [FRANK/SARAH---TOM---MARION---STEVE]
b) See Frank & Sarah S, G1---Tom, G2---Marion, G3

3. Other reported causes: Loud noises during military duty

4. Possible diagnosis:
a) Injury-related hearing loss
b) Hereditary hearing loss

5. Personal comments? WHEN PEOPLE TALK BEHIND MY BACK I SOMETIMES MISS WHAT IS SAID.

UNIT 64---SUE P, G4, AGE 51

[FRANK/SARAH---TOM---MARION---SUE]

1. Reported hereditary hearing loss syndromes:
a) Hearing loss---Severe
b) Family history---Fourth consecutive generation of hearing loss
c) Siblings—3 with reported hearing loss, 0 with no reported hearing loss
d) Offspring—1 with reported hearing loss, 1 with no reported hearing loss
e) Tinnitus---Severe, progressively worse with age
f) Hearing aids---Tried twice---could not tolerate them
g) Audiogram---Shows 44% loss, DFNA1 slope, cookie bite pattern, and 90 dB loss
h) Other---Bat squeaks
i) Other---90% loss on sense of balance test---DFNA1 pattern indicated

2. Continuous chain of hearing loss:
a) [FRANK/SARAH---TOM---MARION---SUE]

b) See Frank & Sarah S, G1---Tom, G2---Marion, G3

3. Other reported causes: Loud noises from explosives, age

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Age-related hearing loss

5. Personal comments? YES---I JUST LIVE WITH IT AS BEST I CAN. WHEN FIRST DIAGNOSED, ONE OF THE THINGS THEY DID TO ME WAS ELECTRO-SHOCK THERAPY. I WAS GIVEN A PILL TO CAUSE AMNESIA, SO I REALLY DON'T REMEMBER THE SHOCKS, BUT I HAVE AN INTENSE NEED TO MAKE PEOPLE THINK I CAN HEAR THEM. IT'S BEEN BETTER SINCE I FOUND OUT ABOUT THE SHOCKS. I TRY TO LET THEM KNOW I DIDN'T HEAR, BUT I WAS 32 BEFORE I LEARNED OF THIS FROM MOM. I DO REMEMBER BEGGING DAD NOT TO MAKE ME KEEP GOING TO THAT AUDIOLOGIST AND HE FINALLY PUT A STOP TO IT AS IT WASN'T HELPING. WAS TOLD BY MY AUDIOLOGIST YEARS AGO THAT SINCE I HAD NEVER BEEN ABLE TO HEAR, I WOULD LIKELY NOT DO WELL WITH HEARING AIDS. THEY DID MAKE ME TERRIBLY NERVOUS, SO I GAVE UP ON THEM.

UNIT 65---TIL B, G3, AGE 70

[FRANK/SARAH---JULIA---TIL]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Moderate
   b) Family history---Third consecutive generation with hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---No
   f) Hearing aids---No
   g) Audiogram---None available

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---JULIA---TIL]

b) See Frank & Sarah S, G1---Julia B, G2

3. Other reported causes: Age, loud noises

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Injury-related hearing loss
   c) Age-related hearing loss

UNIT 66---TOM S, G2, AGE 71 AT DEATH

[FRANK/SARAH---TOM] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Second generation of hearing loss
   c) Siblings— 5 with reported hearing loss, 0 with no reported hearing loss
   d) Offspring—4 with reported hearing loss, 2 with no reported l hearing loss
   e) Tinnitus---Unknown
   f) Hearing aids---Yes---1 BTE
   g) Audiogram---None available
   h) Other---Both parents reported to have hearing loss---[first cousins once removed]

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM]
   b) See Frank & Sarah S, G1
3. Other reported causes: Age, severely beaten during police work
4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

UNIT 67---WES B, G4, AGE 70

[FRANK/SARAH---TOM---OLA---WES]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Severe
   b) Family history---Fourth consecutive generation of hearing loss
   c) Siblings—2 with reported hearing loss, 1 with no reported hearing loss
   d) Offspring—3 with reported hearing loss, 1 with no reported hearing loss
   e) Tinnitus---Mild---occasionally
   f) Hearing aids---Two BTE plus other hearing devices
   g) Audiogram---Shows DFNA1 slope, 76% hearing loss, cookie bite pattern, and 85 dB loss,
   h) Other---Bat squeaks---0% loss on sense of balance test---DFNA1 pattern indicated

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM---OLA---WES]
   b) See Frank & Sarah S, G1---Tom, G2---Ola, G3

3. Other reported causes: Physical abuse as child, age, ear infections, sinus infections

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss
   d) Disease-related hearing loss

5. Personal comments? YES
   a) I HAVE BEEN CALLED “THE HIGHEST FUNCTIONING PERSON EVER FOUND TO HAVE SUCH A SEVERE IMPAIRMENT”.
   b) I HID MY 80+ DB LOSS FROM ALL BUT MY CLOSEST FRIENDS. I LEARNED TO READ LIPS ON MY OWN TO ACCOMPLISH THIS FEAT. I TRIED TO LEARN ASL. BUT COULD NOT MASTER IT.

c) MY RIGHT EAR WAS A LAZY EAR—GOOD FOR NOISE AND NO WORD RECOGNITION UNTIL 1995 WHEN I TAUGHT MYSELF TO DISTINGUISH SPEECH IN MY USELESS RIGHT EAR BY USING THE LINDAMOOD SYSTEM.
d) MY HEARING LOSS HAS MADE ME A BETTER LEARNER BUT SOMETIMES LED TO DISAGREEMENTS AND EVEN FIGHTS BEFORE BEING FITTED FOR HEARING AIDS IN 1973.
e) I HAVE BEEN VERY SOCIAL ALL OF MY LIFE. I HAVE BEEN A LEADER IN MANY ORGANIZATIONS.

5. Author's comment: Wes is truly an amazing person. He was born nearly deaf and has accomplished much more during his lifetime than most people who are born with normal hearing. He has also achieved a great measure of happiness and fulfillment during his first 70 years.

Wes is a born leader.wes has a very positive attitude toward living that should be an inspiration to other hearing-impaired persons. Wes has a wonderful sense of humor and genuinely enjoys being around other people.

Wes seems to consider his severe hearing loss as more of a challenge than a handicap. He meets challenges head on and sorts them out.

You could say that when life handed Wes lemons, instead of becoming bitter about it, he has made lemonade!

Wes has also written a book [*no place else*] that describes his life as a deaf child who is struggling to survive in a hearing world. The book describes what it is like for him to grow up extremely poor during the 1940s with a severe hearing impairment. It is intended to bring awareness to those with normal hearing and consolation to those who are similarly afflicted--- "been there---done that." This book could be best described as being a sequel to John Steinbeck's *The Grapes of Wrath*.

Wes is a well-educated, highly intelligent human being who has written an advice column for this research report. Hopefully the general advice that Wes hands out in this report will answer many questions that baffle other victims of the SFC. Wes will also answer questions and give one-on-one advice about hearing aids, as well as how to deal with a hearing loss, online via his web site. You can contact

him personally at: wesbro@techline.com. Wes makes an excellent role model for other persons with hearing impairment.

Hearing-impaired or not, this guy has a marvelous zest for life, and is filled with a lot of pure determination.

UNIT 68---WILLIAM S, G3, AGE 65 AT DEATH [FRANK/SARAH---TOM---WILLIAM] [by proxy]

1. Reported hereditary hearing loss syndromes:
   a) Hearing loss---Serious
   b) Family history---Third consecutive generation of hearing loss
   c) Siblings—3 with reported hearing loss, 2 with no reported hearing loss
   d) Offspring—0 with reported hearing loss, 5 with no reported hearing loss
   e) Tinnitus--- Unknown
   f) Hearing aids---Yes for work
   g) Audiogram---None available

2. Continuous chain of hearing loss:
   a) [FRANK/SARAH---TOM---WILLIAM]
   b) See Frank & Sarah S, G1---Tom, G2

3. Other reported causes: Age, loud noises

4. Possible diagnosis:
   a) Hereditary hearing loss
   b) Age-related hearing loss
   c) Injury-related hearing loss

CHAPTER 7---PEDIGREE CHARTS

UNIT 69---PEDIGREE CHART INDEX

NOTE:

Males are shown in blocks

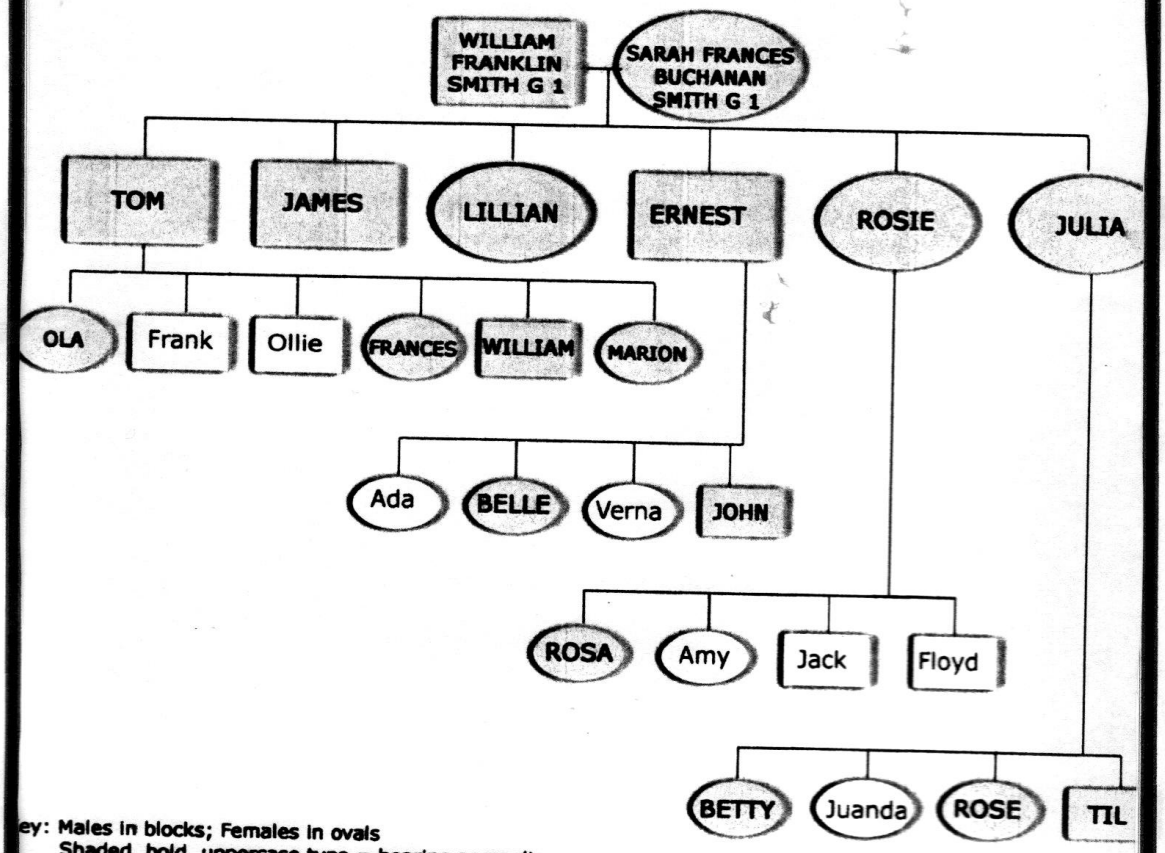
Females shown in ovals

Shaded areas with uppercase bold type indicate hearing loss syndromes

No hearing loss was reported in six branches: Frank G3, Ollie G3, Verna G3, Jack G3, Floyd G3, and Juanda G3, hence no pedigree charts are included on these six branches.

UNIT 70

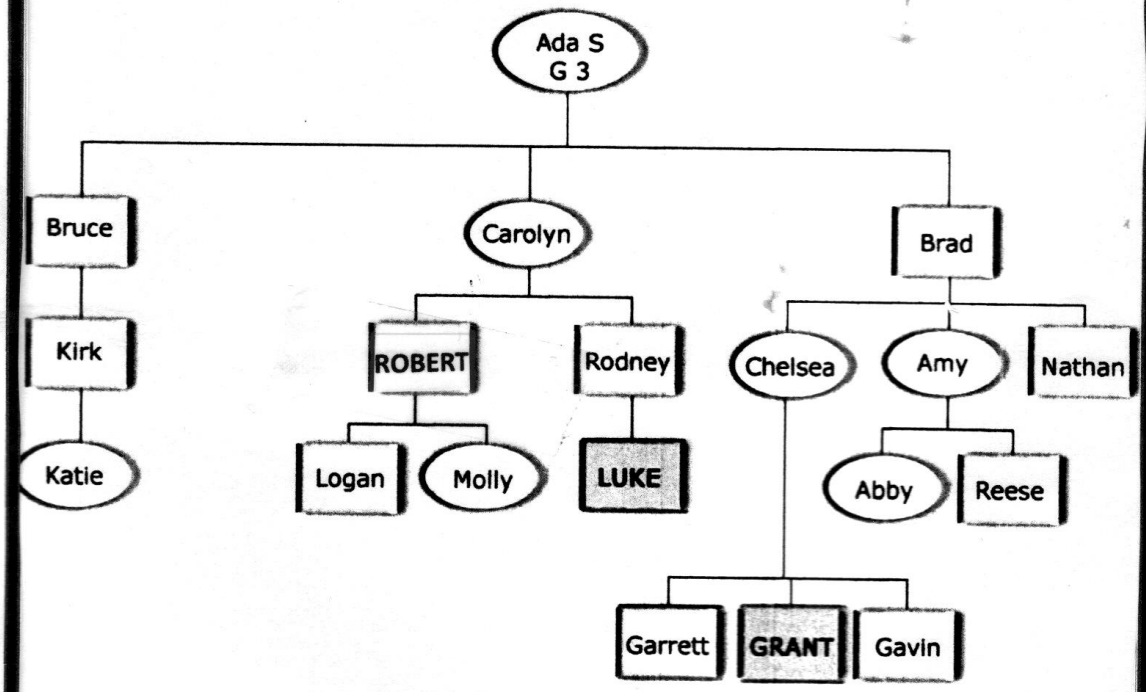
HEARING LOSS SYNDROMES---CHART #1
FRANK, WIFE, CHILDREN, & GRANDCHILDREN



ey: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 71

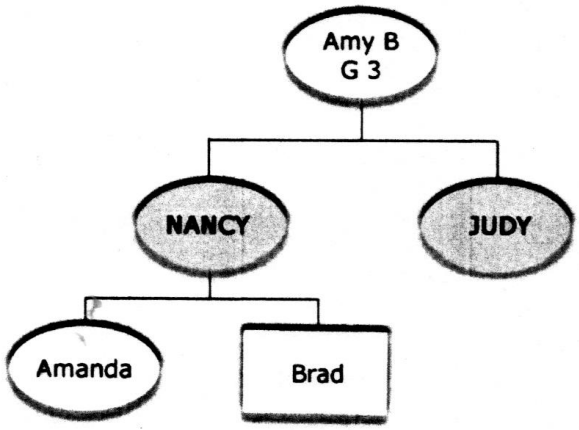
HEARING LOSS SYNDROMES---CHART # 2
ADA S, G3 & GENETIC DESCENDANTS



Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 72

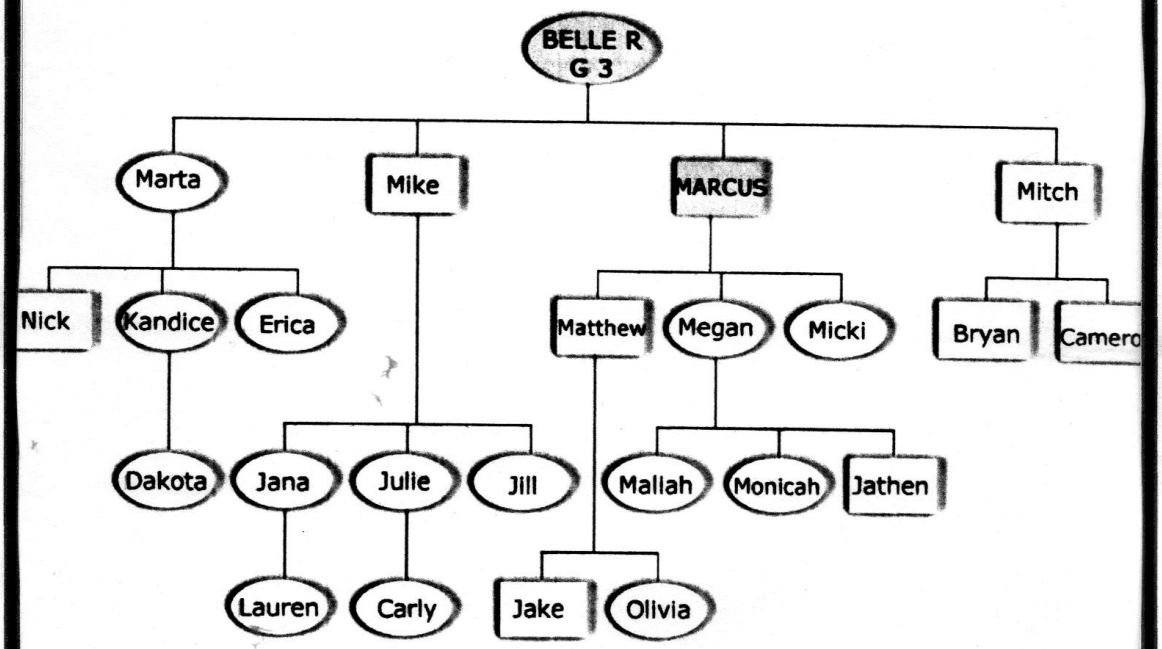
HEARING LOSS SYNDROMES---CHART # 3
AMY B, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 73

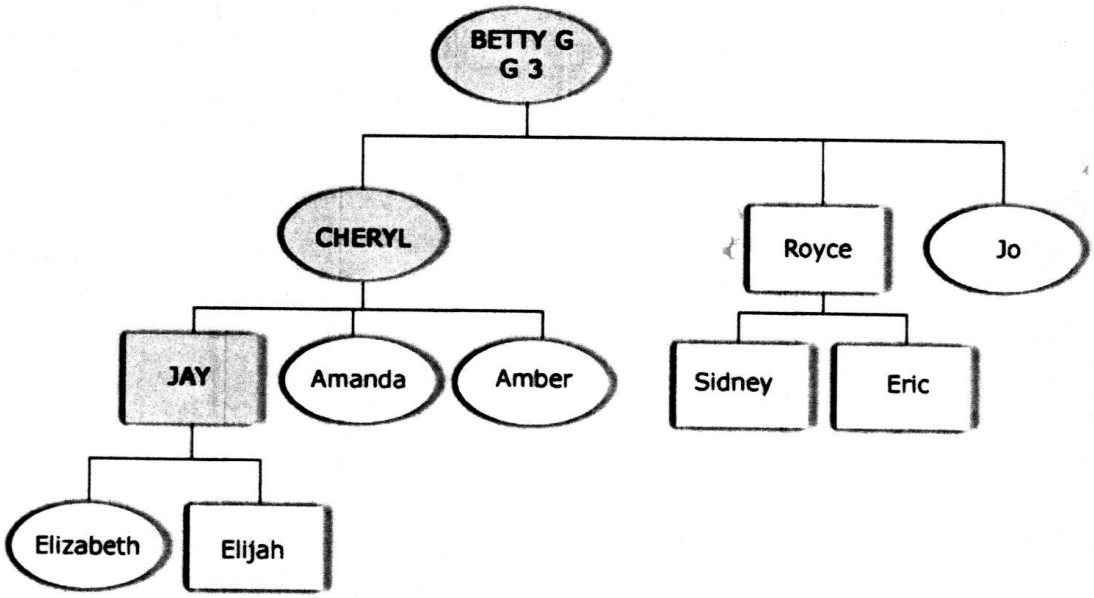
HEARING LOSS SYNDROMES---CHART # 4
BELLE R, G3 & GENETIC DESCENDANTS



ey: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 74

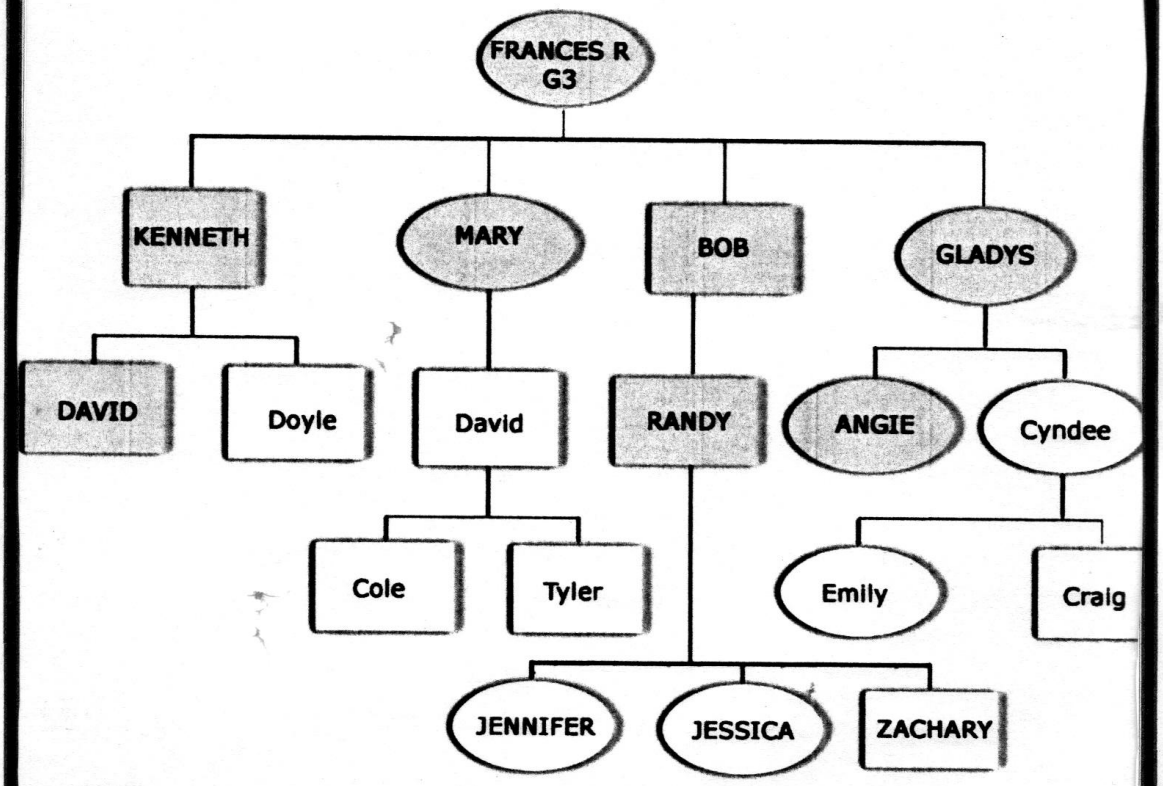
HEARING LOSS SYNDROMES---CHART # 5
BETTY G, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 75

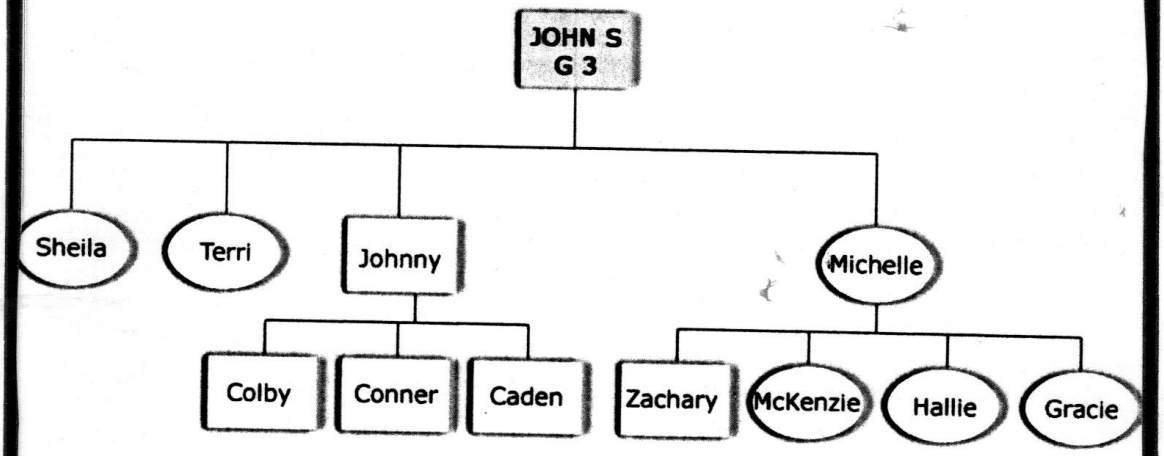
HEARING LOSS SYNDROMES CHART # 6
FRANCES R, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 76

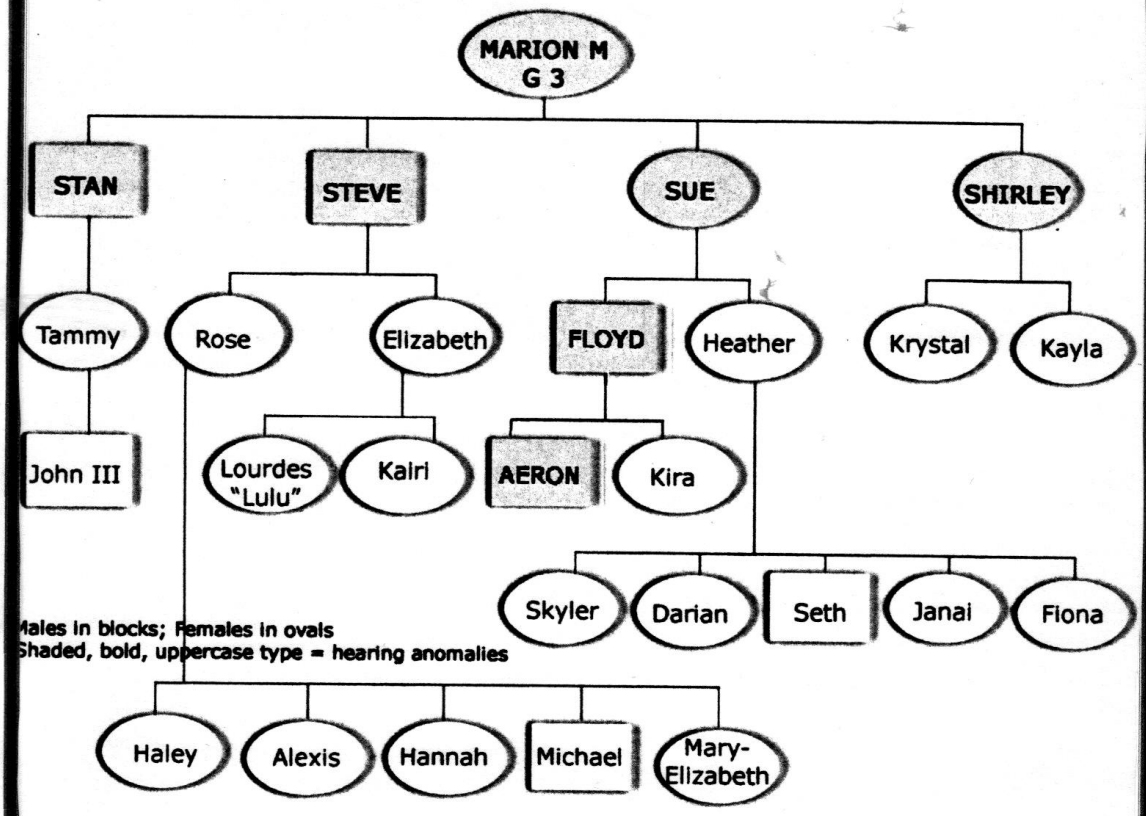
HEARING LOSS SYNDROMES---CHART # 7
JOHN S, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 77

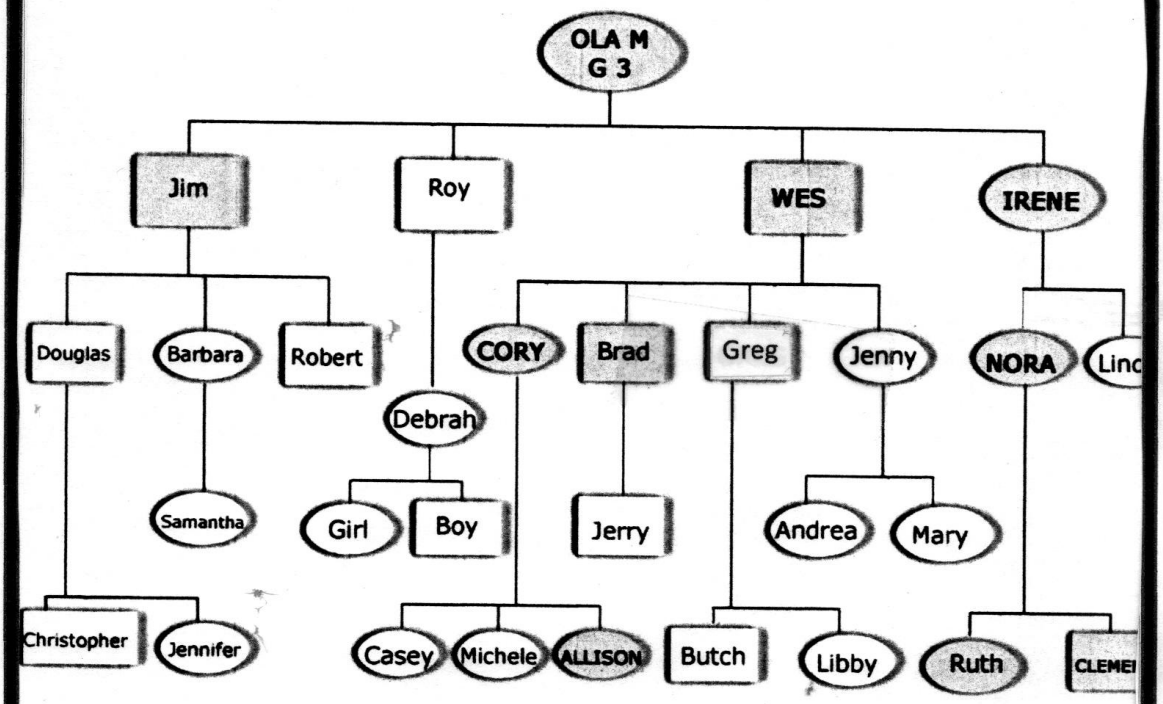
HEARING LOSS SYNDROMES---CHART # 8
MARION M, G3 & GENETIC DESCENDANTS



Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

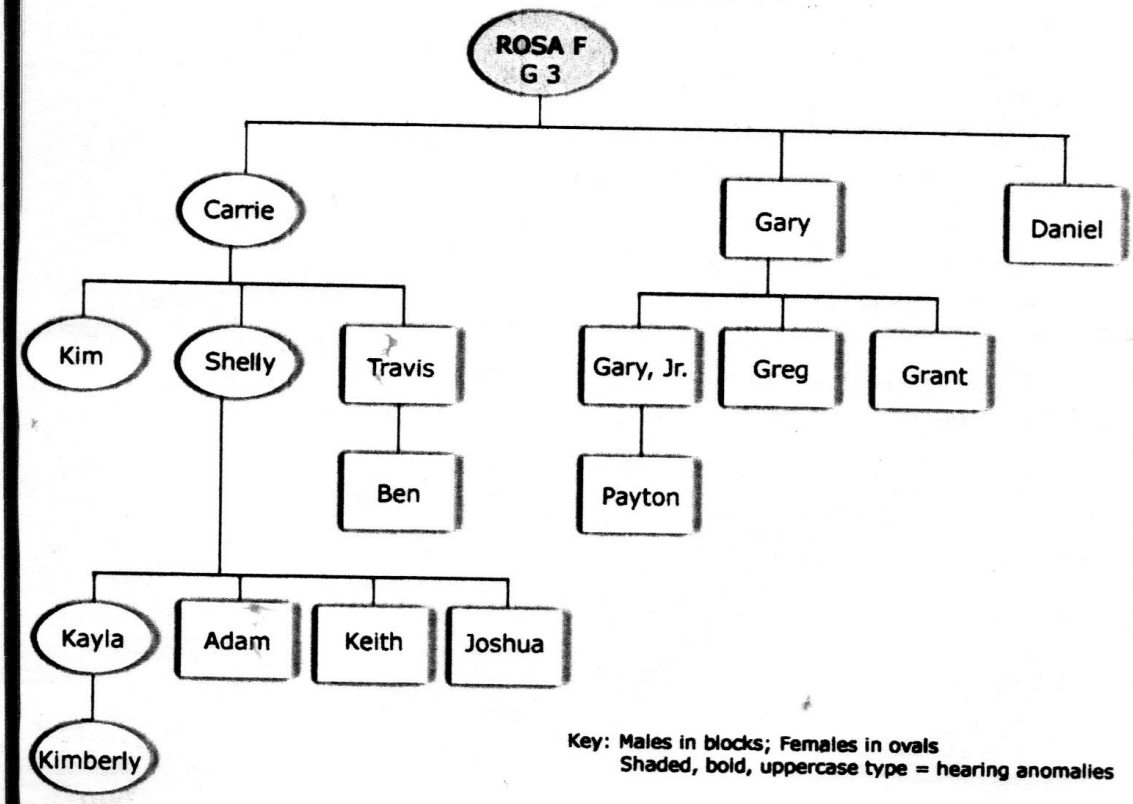
UNIT 78

HEARING LOSS SYNDROMES---CHART # 9
OLA M, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

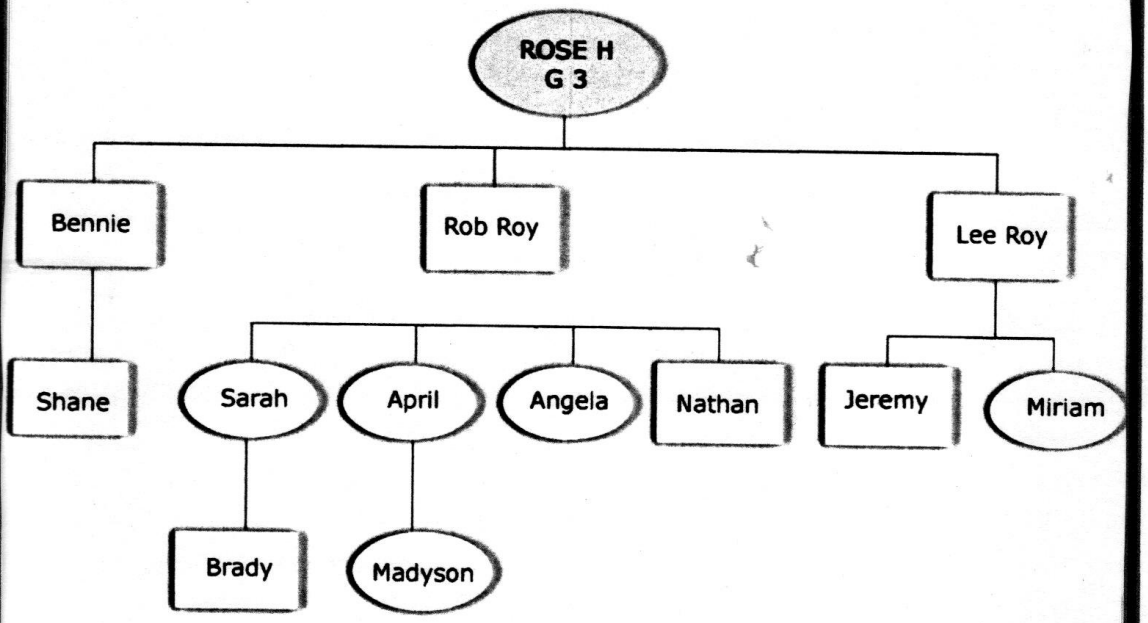
HEARING LOSS SYNDROMES---CHART # 10
ROSA F, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 80

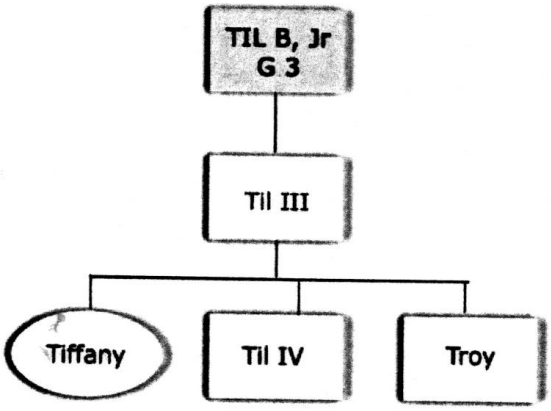
HEARING LOSS SYNDROMES---CHART # 11
ROSE H, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 81

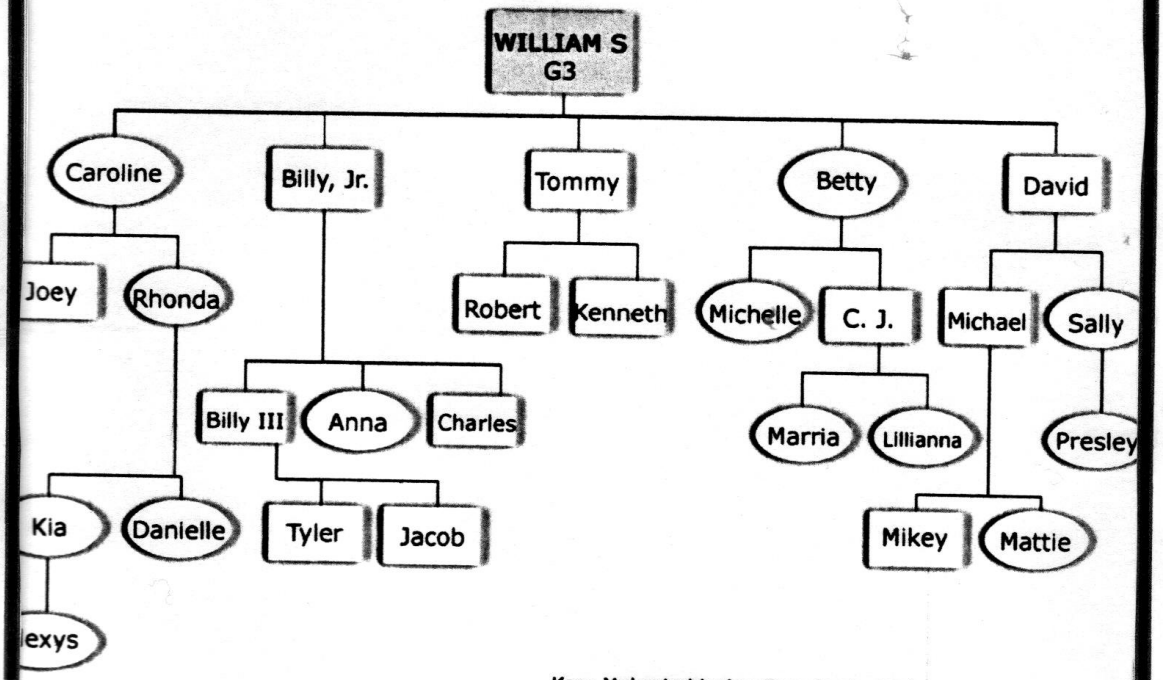
HEARING LOSS SYNDROMES---CHART # 12
TIL B, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

UNIT 82

HEARING LOSS SYNDROMES---CHART # 13
WILLIAM S, G3 & GENETIC DESCENDANTS



Key: Males in blocks; Females in ovals
Shaded, bold, uppercase type = hearing anomalies

CHAPTER 8---AUDIOGRAMS

UNIT 83---AUDIOGRAM COMPARISON CHART

[For actual audiograms, see Units 85-107]

NAME	% LOSS	BANANA POINTS	dB LOSS	SLOPE	PATTERN	AIDS
Judy B	<1%	12	40	DDNA1	C-bite	No
Nancy M	<1%	11	45	DFNA1	C-bite	No
Randy R	<1%	10	40	Downward	C-bite	No
Allison M	3%	8	55	Center dip	C-bite	No
Cheryl M	19%	8	55	Center dip	C-bite	No
Ken R	24%	2	75	DFNA1	C-bite	No
Luke A	29%	7	65	Ski slope	C-bite	2 BTE
Frances R	35%	3	65	Ski slope	C-bite	2 ITE
Jim B	39%	4	80	Ski slope	C-bite	2 ITC
Sue P	44%	0	90	DFNA1	C-bite	No
Mary S	53%	0	80	DFNA1	C-bite	2 CIC
Bob R	57%	0	85	Center dip	C-bite	1 BTE
Cory M	59%	0	75	DFNA1	C-bite	2 BTE
Wes B	76%	0	85	DFNA1	C-bite	2 BTE

How we stacked up.--Audiogram results are listed from lowest percentage of hearing loss to greatest percentage of hearing loss. You will notice that ratings of banana points and dB loss do not follow in exactly the same pattern. It all depends on which figures that a person is looking at.

Summary of audiograms

a) Percentages of hearing loss ranged between <1% to a high of 76%.

b) Banana point scores ranged from one perfect score of 12 to five scores with zero points.
c) Decibel loss ranged from 40 dB to 90 dB.
d) About half of the audiograms showed a definite DFNA1 slope. Three showed downward slopes. Two showed center peaks while two showed center dips.
e) All audiograms showed Cookie Bite patterns. [Notches]
f) Without hearing aids, those who have no "banana" points whatsoever experience extreme difficulty in comprehending normal conversation.
g) Randy, Judy, Allison, and Nancy's overall scores indicated the least hearing loss.
h) With a 76% overall loss, Wes must have excellent hearing aids to function as well as he does.
i) If an audiogram were to be made on every member of the Smith Clan, you might be surprised at how many of us would show a measureable hearing loss!

Summary of hearing aids

a) With less than a 5% hearing loss, Judy, Randy, Nancy, and Allison do not wear hearing aids.

b) With a 75 and 90 dB loss, and very few banana points, Ken and Sue would benefit with the correct hearing aids. Hopefully they could find some that will help instead of hinder. [See Fact Sheets on Ken R, Unit 45, and Sue P, Unit 64 }

c) The remaining eight charted clan members, with overall hearing loss ranging from 19% to 76% with 65 to 85 dB losses, wear one or two hearing aids.

UNIT 84---AUDIOGRAM INDEX

NOTE.—

a) All submitted audiograms have been included

b) Audiograms on Randy R, G5, were submitted as narratives and converted into graph format

UNIT 85

ALLISON M, G6

4/18/2000

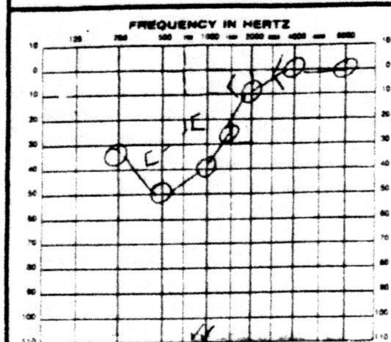
AGE 6

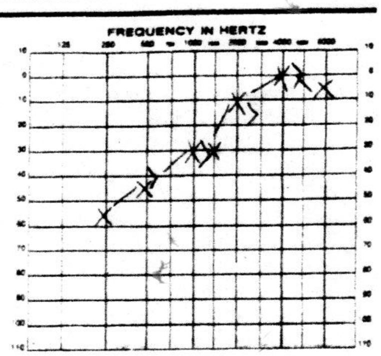
Vancouver Ear Nose & Throat
Head & Neck Surgery Clinic
1405 S.E. 164th Ave.
Suite 102
Vancouver, WA [illegible]

Audiologic Examination Report

AUDIOLOGICAL ASSESSMENT for Allison

Address ______ Referred by AW





RIGHT EAR: (Red) Air, Bone <, No response, With masking △ [

LEFT EAR: (Blue) Air x, Bone >, No response, With masking □]

SPEECH AUDIOMETRY

	Speech Reception Threshold 1	2	Most Comfortable Level 1	2	Discrimination Score (PB Max) 1	2	Threshold Discomfort
Right Ear	10						
Left Ear	15						
Sound Field							
Aided							

SHORT INCREMENT SENSITIVITY INDEX (SiSi Test)

Freq	500	1K	2K	4K	
Right					
Left					

THRESHOLD TONE DECAY

Right					
Left					

PURE TONES 500 - 1000 - 2000 CPS AVERAGE SPEECH RANGE

Ear	3 Freqs	2 Freqs
Right		
Left		

Examiner J. S[illegible] MA CCC A

Audiometer Used GSI-16

Hearing Constant ______

Hearing Today Same ___ Better ___ Worse ___

Tinnitus Right ______ Left ______

HEARING AID RECOMMENDED

Ear ______ Receiver ______ Gain ______

Remarks

PEDIGREE CHAIN [FRANK---TOM---OLA---WES---CORY---ALLISON]

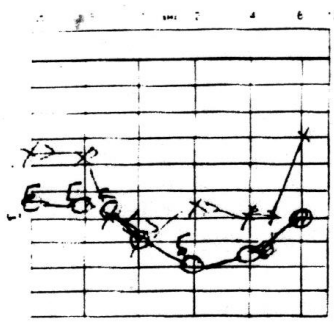
UNIT 86

BOB R, G4

8/21/02---AGE 60

Serial Audiograms

8/22/06---AGE 64

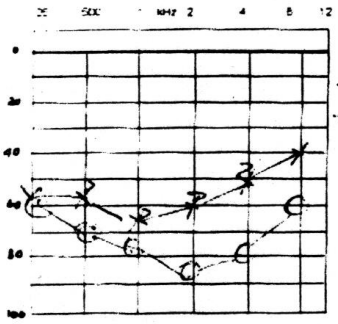


Name:

Date 8/21/02

Right Ear
SRT 5 dBHL
MCL 15 dBHL
SDS 100 %

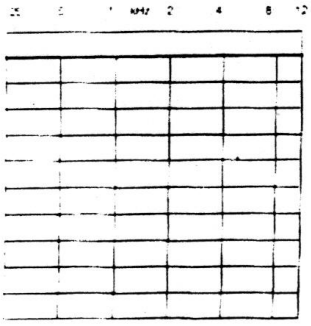
Left Ear
SRT 35 dBHL
MCL 70 dBHL
SDS 60 %



Date 2/22/06

Right Ear
SRT 20 dBHL
MCL 50 dBHL
SDS 70 %

Left Ear
SRT 60 dBHL
MCL 75 dBHL
SDS 70 %



Date _____

Right Ear
SRT _____ dBHL
MCL _____ dBHL
SDS _____ %

Left Ear
SRT _____ dBHL
MCL _____ dBHL
SDS _____ %

William E. Lentz, Ph.D. & Associates

PEDIGREE CHAIN [FRANK---TOM---FRANCES---BOB]

UNIT 87

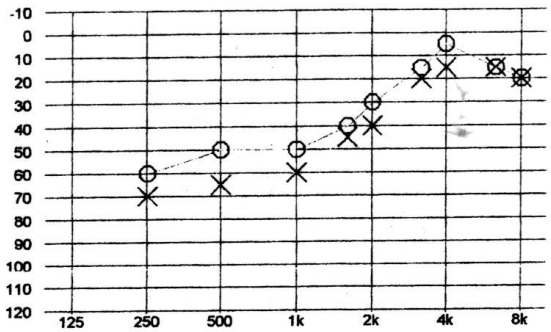
CHERYL M, G4

3/29/09

AGE 63

Aurical Tone Audiometer

Left Ear/Right Ear - HL



AC:

BC:

SF:

Masking Levels

	Right	Masking					Left	Masking	
SRT:	40 dB					SRT:	45 dB		
	Right	Masking					Left	Masking	
MCL:									
	Right	Masking					Left	Masking	
UCL:									
	Right						Left		
PTA (A):									

Unaided Discrimination:	Right	Level	Masking	Binaural	Level	Masking	Left	Level	Masking
	92 %	75 dB					92 %	70 dB	
Aided Discrimination:	Right	Level	Masking	Binaural	Level	Masking	Left	Level	Masking
Unaided Discrimination, noise:	Right	Level	S/N	Binaural	Level	S/N	Left	Level	S/N

PEDIGREE CHAIN [FRANK---JULIA---BETTY---CHERYL]

UNIT 88

CORY M, G5

3/31/08

KAISER PERMANENTE.

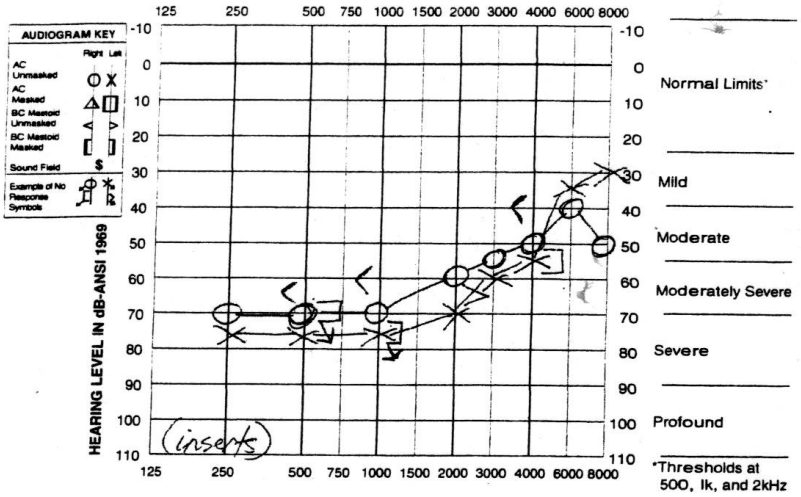
AGE 49

Audiological Record

Pure Tone Audiometry

Referred By:

FREQUENCY IN HERTZ



THRESHOLD TONE DECAY

Right						
Left						

SPEECH AUDIOMETRY

	Speech Reception Threshold 1	2	Most Comfortable Level 1	2	% Discrimination Score 1	LIST	2	LIST	Threshold Discomfort
Right	60		75		96	4A			DNT
Left	60		77*		84	4B			
Sound Field	* could not tolerate 80 dB								
Aided	Level in (L) ear								

Comments: Otosc-WNL, Au

Test Reliability: (Good) Fair Poor Examiner: L. Gladstone Audiometer Used: GSI-61

PEDIGREE CHAIN [FRANK---TOM---OLA---WES---CORY]

UNIT 89

CORY M, G5

10/15/03

AGE 45

ersity of Washington
artment of Speech and Hearing Sciences
ech and Hearing Clinic

ECORD OF AUDIOMETRIC EVALUATION

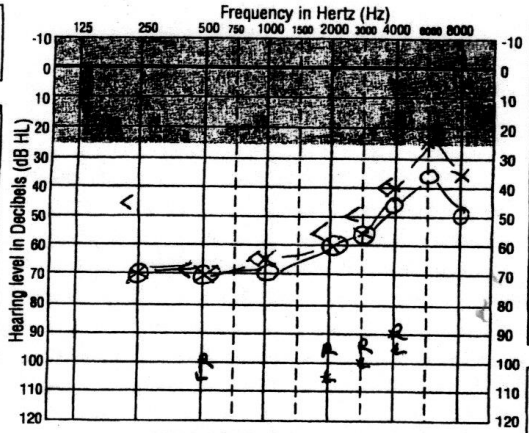
IBRATION
ent ANSI reference
s for pure tones and
ch

REVIATIONS
Air conduction
Bone conduction
Could not establish
Did not test
Hearing level
Most comfortable level
Monitored live noise
Narrow band noise
No response
Sound pressure level
Speech noise
Threshold of discomfort
White noise
Within normal limits

ONSE RELIABILITY
od) Fair Poor

IMULUS
dy (Pulse) FM

= ucls



AUDIOGRAM KEY
Right Left
AC unmasked
AC masked
BC mastoid unmasked
BC mastoid masked
BC forehead masked
BC forehead unmasked
Sound field
Aided
Example of no response symbols

AUDIOMETER
No.: 1169568
Make: GSI
Model: 61

TRANSDUCER
EE-3A

Effective Masking levels to non-test ear

		250 Hz	500 Hz	1K Hz	2K Hz	4K Hz	8K Hz
AC	L						
	R						
BC	L						
	R						

H AUDIOMETRY

	PTA	SRT	Speech discrimination		MCL	UCL
	66	65	32 %	80 %	70	90
in left			dB	LIST 24 dB LIST		
	65	70	38 %	80 %	70	90
in right			dB	LIST 11 dB LIST		
ld			%	%		

ACOUSTIC REFLEX

PROBE	STIM	Frequency	Decay
Right	IPSI	CNE	
	Contra		
Left	IPSI	CNE	
	Contra		

IMMITTANCE

Y-226 Hz	Right	Left
Press (da Pa)	-25	10
Vol (cc)	2.3	2.1
Admit (ml)	.5	1.1

PEDIGREE CHAIN [FRANK---TOM---OLA---WES---CORY]

UNIT 90

CORY M, G5

3/06/98

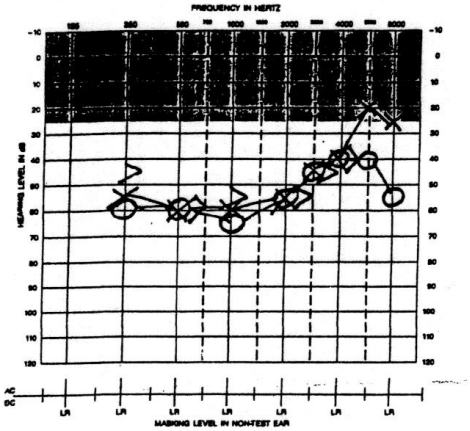
AGE 39

Cascade Medical-Dental Center
411 N.E. 87th Avenue, Suite B
Vancouver, Washington 98664
(360) 892-9367 FAX (360) 253-3801

Audiology Clinic

Birthdate

Date: 3-6-98 Audiometer: GSI-16 Referral Source: RETURN / Dr. John Kass



Test Reliability: (Good) Fair Poor
Calibration Standard ANSI: 1989
Calibration Date: 1-26-98
Time of Test: 2:30 PM
Last Noise Exposure: —

AUDIOGRAM KEY

RIGHT LEFT
AC UNMASKED
AC MASKED
BC MASTOID UNMASKED
BC MASTOID MASKED
BC FOREHEAD UNMASKED
BOTH
BC FOREHEAD UNMASKED SOUND FIELD
EXAMPLE OF NO RESPONSE SYMBOLS

RESPONSES
☒ CONSISTENT ☐ INCONSISTENT
☐ RAPID ☐ DELAYED

TONE DECAY RESULTS

	500Hz	1000Hz	2000
RIGHT	NT	<	1C
LEFT	NT		1C

THRESHOLD ☒ SUPRATHRESHOLD

SPEECH HEARING LEVELS

	APT	SPEECH RECEPTION THRESHOLD 1	2	MOST COMFORTABLE LEVEL 1	2	THRESHOLD OF DISCOMFORT
RIGHT		50		70	80	95
LEFT		50		80		95
MASKING						

SPEECH DISCRIMINATION SCORES

	TEST 1		TEST 2		TEST 3		TEST 4
RIGHT	60%	HL 80	80%	HL 80	%	HL	%
MASKING	dB	LIST M	dB	LIST O	dB	LIST	d
LEFT	76%	HL N	%	HL	%	HL	%
MASKING	dB	LIST N	dB	LIST	dB	LIST	d

Speech Source: TAPE ☐ LIVE VOICE ☒ NU-6 ☐ CAMPBELL ☐ W-22 ☐ WIPI ☐

PATIENT REPORTED HISTORY OF:

- ☒ Difficulty understanding speech: ☒ in noise ☐ in quiet
- ☐ Hearing poorer in: ☐ right ☐ left
- ☒ Gradual loss of hearing
- ☐ Recent change in hearing
- ☒ Familial hearing loss
- ☐ Tinnitus: ☐ right ☐ left
- ☐ Middle ear pathology/surgery: ☐ right ☐ left
- ☐ High blood pressure
- ☐ Upper respiratory problems
- ☐ Dizziness/vertigo
- ☐ Head/neck trauma
- ☐ Diabetes
- ☐ Other

TEST RESULTS

Right Ear: Otoscopic CLEAR
☐ Pure tone and speech thresholds within or near normal limits
☒ A significant hearing loss is present, which is MODERATE TO MILD
☐ mixed (conductive and sensorineural)
☒ sensorineural ☐ conductive ☐ unspecified

Left Ear: Otoscopic CLEAR
☐ Pure tone and speech thresholds within or near normal limits
☒ A significant hearing loss is present, which is MODERATE TO MILD
☐ mixed (conductive and sensorineural)
☒ sensorineural ☐ conductive ☐ unspecified

COMMENTS:

PEDIGREE CHAIN [FRANK---TOM---OLA---WES---CORY]

UNIT 91

CORY M, G5

7/30/91

AGE 33

Al Hicks, Ph.D
Audiologist, CCC-A

Nancy M. Bowen, M.S.
Audiologist, CCC-A

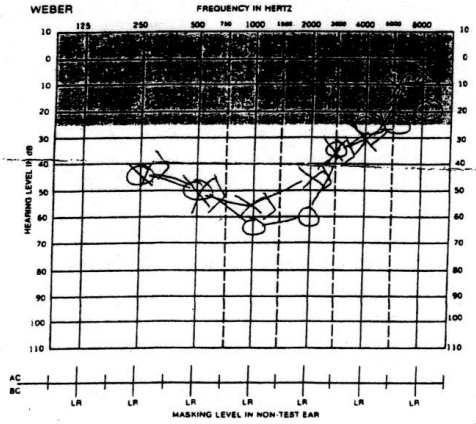
Audiology Clinic

Date: 7/30/91 Audiometer: 11H24-M Referral Source:

Test Reliability: (Good) Fair Poor

Calibration Standard ANSI: 1969

Calibration Date: 9/11/90



AUDIOGRAM KEY

	RIGHT	LEFT
AC UNMASKED		
AC MASKED		
BC MASTOID UNMASKED		
BC MASTOID MASKED		
BC FOREHEAD MASKED		
	BOTH	
BC FOREHEAD UNMASKED SOUND FIELD	$	
EXAMPLE OF NO RESPONSE SYMBOLS		

RESPONSES

CONSISTENT X INCONSISTENT ☐
RAPID ☐ SLOW ☐
DELAYED ☐

TONE DECAY RESULTS

	500Hz	1000Hz	2000Hz
RIGHT			
LEFT			

THRESHOLD ☐ SUPRA-THRESHOLD ☐

SPEECH HEARING LEVELS

	APT	SPEECH RECEPTION THRESHOLD 1	2	MOST COMFORTABLE LEVEL 1	2	THRESHOLD OF DISCOMFORT
RIGHT		40		60		80
LEFT		35		60		85
MASKING						

SPEECH DISCRIMINATION SCORES

	TEST 1		TEST 2		TEST 3		TEST 4	
RIGHT	72 %	HL 60	92 %	HL 75	88 %	HL 65	%	HL
MASKING	dB 40	LIST M	dB 50	LIST D	dB 45	LIST M	dB	LIST
LEFT	92 %	HL 60	92 %	HL 75	%	HL	%	HL
MASKING	dB 40	LIST M	dB 55	LIST D	dB	LIST	dB	LIST
Speech Source:	TAPE ☐ LIVE VOICE ☐		NU-6 ☐ CAMPBELL ☐		W-22 ☐			

Impressions & Recommendations:

PEDIGREE CHAIN [FRANK---TOM---OLA---WES---CORY]

UNIT 92

FRANCES R, G3

4/16/86

AGE 68

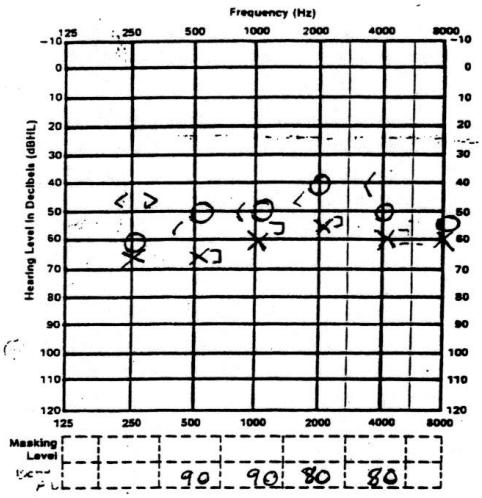
COLORADO STATE UNIVERSITY
Hearing Clinic
HEARING EVALUATION

Patient's Name Cindy Bloyer . Age 68 Sex F

Test Reliability: Good

ASHA Certified Clinical Audiologist

Clinician Gregory F Moore, B.S.



AUDIOGRAM KEY

Ear	AIR Unmasked	AIR Masked	BONE Masked	BONE Unmasked	No Response
Right	O	Δ	[	<	↓
Left	X	□	]	>	↓

COMMENTS

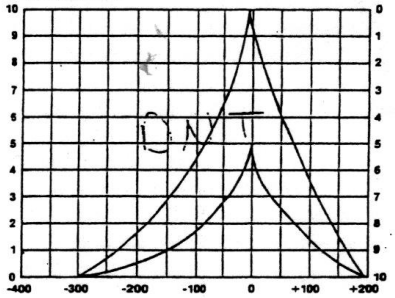
Otoscopy: WNL Bilaterally

RE: LE:

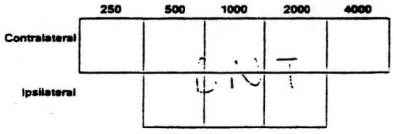
SPEECH HEARING TESTS

Ear	SRT	Speech Discrim Scores	Speech Discrim Scores (Rollover)
Right	55 db	100% 85 db	%/ db
Left	60 db	96 %/ 91 db	%/ db
Sound Field	db	%/ db	%/ db

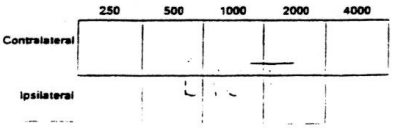
TYMPANOGRAM



RIGHT



LEFT



PEDIGRE CHAIN [FRANK---TOM---FRANCES]

UNIT 93

FRANCES R, G3

10/05/76

AGE 58

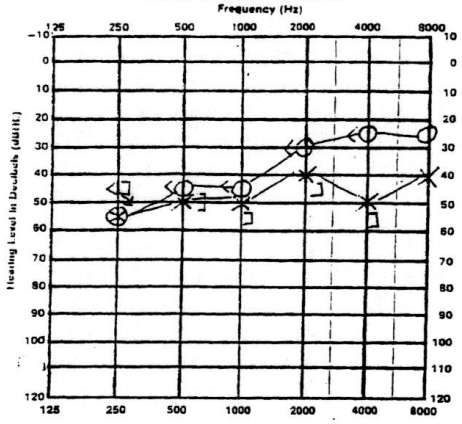
COLORADO STATE UNIVERSITY
Speech and Hearing Clinic
HEARING EVALUATION

Patient's Name _____ Age 57 Sex F

Referred by Dr. Robt. Conlon (re-eval) Examiner Linville

Audiometer Grason-Stadler 1701 Test Reliability: Good X Fair ____ Poor ____

PURE TONE AUDIOGRAM



500-2000 Hz Average
Best 2 Frequency Average X

AC Right 40 dB HL
Left 48 dB HL

SPEECH RECEPTION THRESHOLD
Right 38 dB HL
Left 48 dB HL
Sound Field 38 dB HL

LOUDNESS DISCOMFORT THRESHOLD
Right 106+ dB HL
Left 106+ dB HL
Sound Field DNT dB HL

SPEECH DISCRIMINATION

	Test	List	Hearing Level
Right	98 %	W-22 1-A('½)	70 dB
Left	100 %	2-A('½)	80 dB
Sound Field	88 %	3-A('½)	50 dB
	68 %	4-A	50 dB

S/NFC

MASKING APPLIED TO NONTEST EAR*

		250	500	1000	2000	3000	4000	8000	SRT	Discrim.
AC	Right									55
	Left									
BC	Right	60	60	70	60		70			
	Left									

Type of Masking for:
Speech: Sp.B
Puretones: NBN

*EFFECTIVE MASKING RE: odBHL

AUDIOGRAM KEY

		UNMASKED	MASKED
AC	Right	O	△
	Left	X	□
BC	Right	<	[
	Left	>	]

Sound Field S
Examples of No Response

Hearing is Within Normal Limits ☐

Approximate Loudness of Conversational Speech ☐

COMMENTS

PEDIGRE CHAIN [FRANK---TOM---FRANCES]

UNIT 94

JIM B, G4

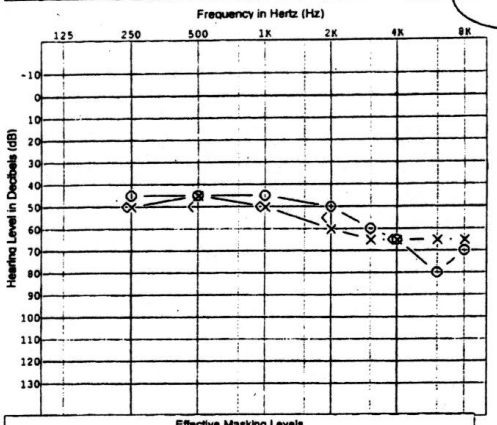
2/13/07

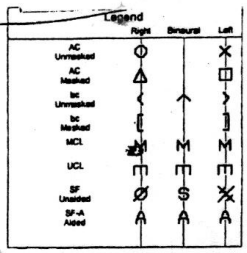
AGE 75

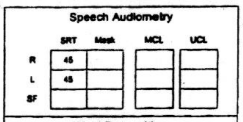
2007

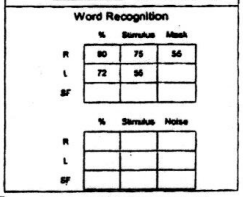
Hearing Evaluation Report

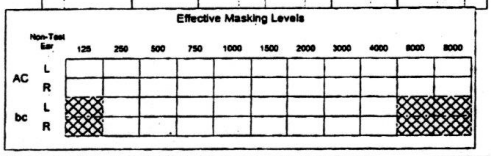
Date of Evaluation

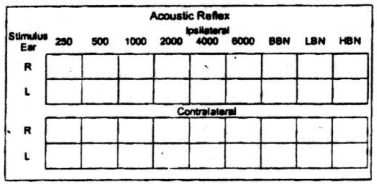


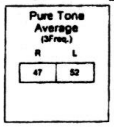


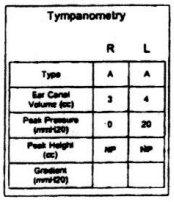












Report Comments

VET TESTED BY GAIL TERNES, M.ED. Date of last audi 1-9-04

Signature: Gail Ternes, MEd Date: 2-13-07

PEDIGRE CHAIN [FRANK---TOM---OLA---JIM]

UNIT 95

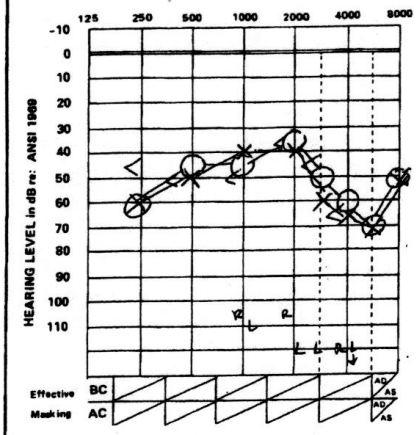
JIM B, G4

1/09/04

AGE 72

Department of Veterans Affairs | 2004 | AUDIOLOGICAL EV/
ADDENDUM TO VA FOI

AUDIOGRAM



(OVER FOR S.O.A.P.)

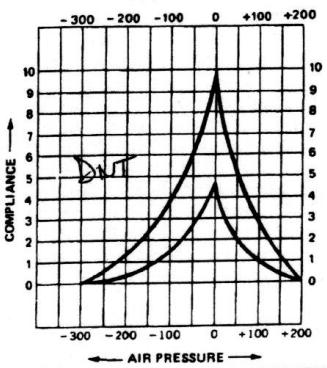
last audio 3-6-98

Air conduction	Ear R	Ear L
Unmasked	○	X
Masked	△	□
Bone conduction		
Unmasked	<	>
Masked	[	]
No Response		
Unmasked		
Masked		

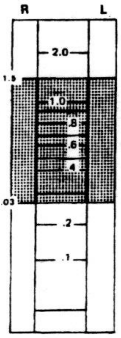
MLV TAPE/CD CNC/-3

MCLdBHL	EAR	SRT	DISCRIMINATION dB	dB	dB	UCLdBHL
75	R	50	84 %	%	%	100
	MASK L		75dB 50dB			
75	L	50	76 %	%	%	105
	MASK R		75dB 50dB			

TYMPANOGRAM



STATIC COMPLIANCE



Stapedius Reflex

FREQ.		500	1K	2K	4K
CON	R				
CON	L	DNT			
IPS	R				
IPS	L				

REFLEX DECAY 500 1k

Static Compliance

R	L
C2	C2
C1	C1
Cx	Cx

LAST NAME - FIRST NAME - MIDDLE INITIAL	AGE	CLAIM NO.
	71	17005081

NAME OF EXAMINING STATION OR CLINIC	SIGNATURE OF EXAMINING AUDIOLOGIST	DATE OF EXAM
VA OPC MARTINEZ - AUDIOLOGY	Diane Fernandez, M.S.	1-9-04

VA FORM 10-2364a EXISTING STOCK OF VA FORM 10-2364a DEC 1983

PEDIGREE CHAIN [FRANK---TOM---OLA---JIM]

UNIT 96

JUDY B, G4

3/27/08

AGE 48

Karr Audiology & Hearing Aids

499 Richland Ave, Athens, OH 45701

Phone: (740) 594-6333

_____

_____

_____

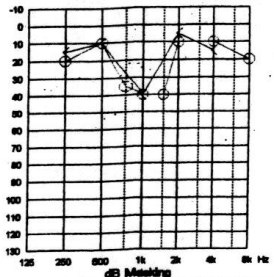
_____

.45701_____

_____

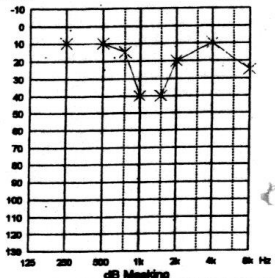
Tone audiogram

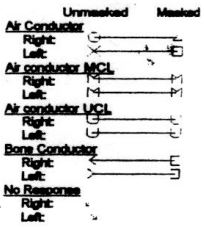
dB HL — Right Ear

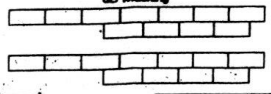


Tone audiogram

dB HL — Left Ear







Speech

	SRT	Mask.	WRS HL	WRS %	WRS Mask.	HL	%	Mask.	MCL	UCL
AC Right	35 dB		60 dB	80 %						
AC Left	35 dB		55 dB	84 %						
Ground										
BC										
BF										
BF Aided										

SRT = Speech Reception Threshold

MCL = Most Comfortable Level

UCL = Uncomfortable Level

WRS = Word Recognition Score

HL = Hearing Level

Mild sensorineural hearing loss @ 1KHz and 1.5KHz. Bilaterally - normal hearing for all other frequencies. Mild loss for speech, also in both ears; good word discrimination

PEDIGREE CHAIN [FRANK---ROSIE---AMY---JUDY]

UNIT 97

KEN R, G4

1/18/05

AGE 69

MASSACHUSETTS INSTITUTE OF TECHNOLOGY
MEDICAL DEPARTMENT

AUDIOLOGY REPORT

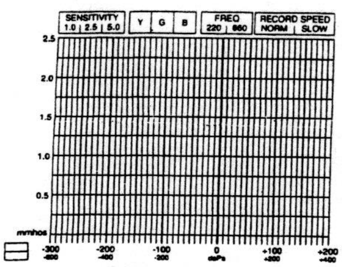
PATIENT NAME ____ 990530 (01/18/2005) ________ MR # ____________

AUDIOl ____ Sex M ________ DATE ____________

RELIABILITY *Good* AUDIOMETER *Madsen 602* *insert phones*

Sound Field	SF
Aided	A

TYMPANOGRAM re-plotted



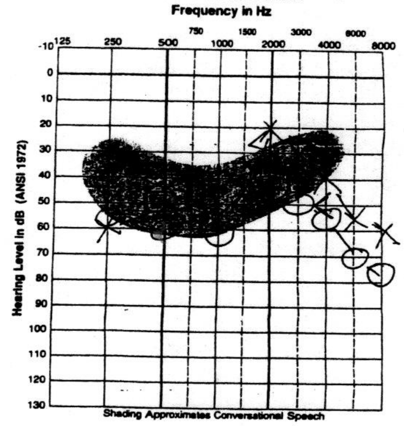
Contralateral Acoustic Reflex

Stim.	Probe	.5K	1K	2K	4K / NOISE	Decay .5K	Decay 1K
R	L						
L	R						

Ipsilateral Acoustic Reflex

Stim.	Probe	.5K	1K	2K	NOISE	Decay .5K	Decay 1K
R	R						
L	L						

PURE TONE AUDIOGRAM
Frequency in Hz



Comments:
Pt saw ENT for ear cleaning
notes RE-meatus narrower than LE-

uses hearing aids sporadically
mostly for meetings —
c/o RE aid of no benefit.
(+) family Hx - maternal side
siblings all have hrg loss
his children (in 40's) ok

SPEECH AUDIOMETRY

	Right	Left	Mask	SF	Aided
Speech Awareness Threshold	dB	dB			
Spondee Threshold	45 dB	30 dB			
Speech Recognition Score	84% / 85 dBHL	88% / 70 dBHL	60		
Speech Recognition Score	75 dBHL	dBHL			
MCL / UCL	dBHL	dBHL			

Discrimination Word List: W22 ☒ NU6 ☐ PBK ☐ WIPI ☐
Live Voice ☐ Tape/CD ☒ *1A*

PEDIGREE CHAIN [FRANK---TOM---FRANCES---KEN]

UNIT 98

LUKE A, G6

9/17/07

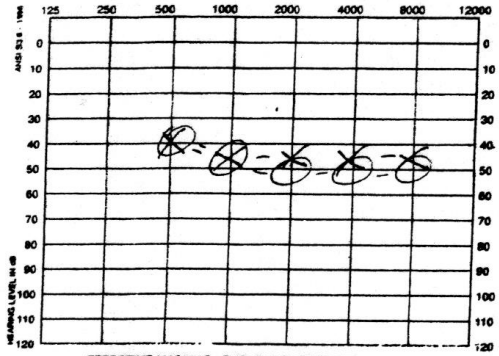
AGE 6

HEARING CLINIC
AUDIOLOGICAL RECORD

NAME _____ D _____ TE 9-17-07

TEST BY METZGER AUDIOMETER AURICAL REFERRED _____

PURE TONE AUDIOGRAM
FREQUENCY IN Hz
EARPHONES ☒ INSERT ☐ SUPRA-AURAL



ACOUSTIC IMMITTANCE
(SEE TRACES ON BACK)

EAR	PHYSICAL VOLUME (ml)	MIDDLE EAR PRESSURE (daPa)	ACOUSTIC ADMITTANCE NORMAL RANGE: (226HZ) Adults: 0.3 - 1.6 Children: 0.3 - 0.85
R			
L			

ACOUSTIC REFLEX THRESHOLDS (dBHL)

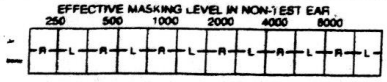
	Stimulus in Right Ear				Stimulus in Left Ear			
	500	1000	2000	4000	500	1000	2000	4000
CONTRA								
IPSI								
CONTRA DECAY								
IPSI DECAY								

Technique: ☒ Conventional ☐ Conditioned Play
☐ Visual Reinforcement ☐ Behavioral Observation
Reliability: ☐ Good ☐ Fair ☐ Poor

SPECIAL TESTS

Frequency	
Right	
Left	

EFFECTIVE MASKING LEVEL IN NON-TEST EAR



AUDIOGRAM KEY

MODALITY	EAR: RIGHT	UNSPECIFIED	LEFT
EARPHONES UNMASKED	O		△
EARPHONES MASKED	△		□
B.C. VIBRATOR UNMASKED	<		>
B.C. VIBRATOR MASKED	[		]
LOUDSPEAKER UNAIDED	S	S	S
LOUDSPEAKER AIDED	[S]	[S]	[S]
NO RESPONSE	↘		↙

SPEECH AUDIOMETRY
☐ EARLY ☐ RECORDED
WORD RECOGNITION

EAR	PHYSICAL	SRT	PB	MCL	UCL
RIGHT	48	45			
LEFT	45	40			
Soundfield		SRT/SAT			

☐ W-22 ☐ BYU ☐ WIPI ☐ SPANISH ☐ OTHER

COMMENTS:

- results: no change in thresholds re: previous audiogram of 7/19/07
- rx: annual HE

PEDIGREE CHAIN [FRANK---ERNEST---ADA---CAROLYN---RODNEY---LUKE]

UNIT 99

LUKE A, G6

8/29/06

AGE 5

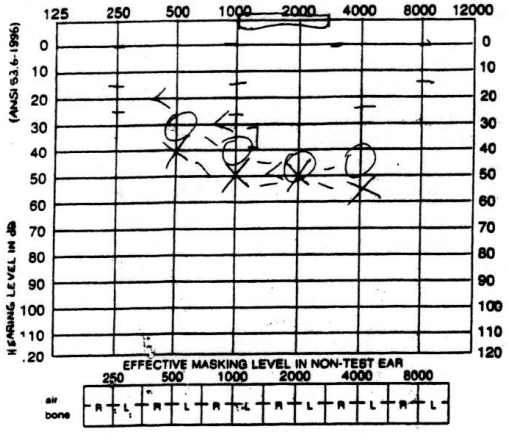
HEARING CLINIC AUDIOLOGICAL RECORD

NAME _____

TEST BY MET2GER AUDIOMETER AURICAL

PURE TONE AUDIOGRAM
FREQUENCY IN Hz

EARPHONES: ☒ INSERT ☐ SUPRA-AURAL



EFFECTIVE MASKING LEVEL IN NON-TEST EAR

ACOUSTIC IMMITTANCE
(SEE TRACES ON BACK)

EAR	PHYSICAL VOLUME (ml)	MIDDLE EAR PRESSURE (daPa)	ACOUSTIC ADMITTANCE NORMAL RANGE (226Hz) Adults: 0.30 - 1.40 Children: 0.20 - 0.80
R	0.82	35	1.04 (A)
L	088	-5	0.86 (A)

ACOUSTIC REFLEX THRESHOLDS (dBHL)

	Stimulus in Right Ear				Stimulus in Left Ear			
	500	1000	2000	4000	500	1000	2000	4000
CONTRA	80	80	80	85	80	80	90	95
IPSI		80	80			80	80	
CONTRA DECAY								
IPSI DECAY								

Technique: ☒ Conventional ☐ Conditioned Play ☐ Visual Reinforcement ☐ Behavioral Observation
Reliability: ☒ Good ☐ Fair ☐ Poor

SPECIAL TESTS	
Frequency	
Right	
Left	

AUDIOGRAM KEY

MODALITY	EAR: RIGHT	UNSPECIFIED	LEFT
EARPHONES UNMASKED	O		X
EARPHONES MASKED	△		□
B.C. VIBRATOR UNMASKED	<		>
B.C. VIBRATOR MASKED	[		]
LOUDSPEAKER UNAIDED	S	S	S
LOUDSPEAKER AIDED	R	A	L
NO RESPONSE	↙		↘

SPEECH AUDIOMETRY

☒ MLV ☐ RECORDED — WORD RECOGNITION

EAR	PVT AV	SRT / SAT	HL for SRT/SAT	WR PB	HL for WR PB	MCL / UCL
RIGHT	38	35	70 dB	96 %		
LEFT	46	35	70 dB	88 %		
Soundfield		SRT/SAT				

☐ W-22 ☐ BYU ☐ WIPI ☐ SPANISH ☐ OTHER

COMMENTS: - Pt. failed HE w/ Lara Ciasulli 8-31-06. Pt was born 5 wks early, and weighed 5 lbs, 2 oz, and had jaundice. ϕ family hx. of HL. No concerns re: speech/language; ϕ hx of OM. Pt ~~dx~~ tinnitus AS recently. No memory of newborn screening. ϕ noise hx.

- results: mild sloping to moderate SNHL, AU
- rx: ① flu w/ Dr. Taylor as soon as possible
② binaural amplification pending [illegible]

AUDIOLOGIST _____

PEDIGREE CHAIN [FRANK---ERNEST---ADA---CAROLYN---RODNEY---LUKE]

UNIT 100

LUKE A, G6

8/21/06

AGE 5

SAN LUIS VALLEY BOARD OF COOPERATIVE SERVICES
P.O. BOX 1198, ALAMOSA, CO 81101, (719) 589-585:

AUDIOMETRIC EVALUATION/SCREENING

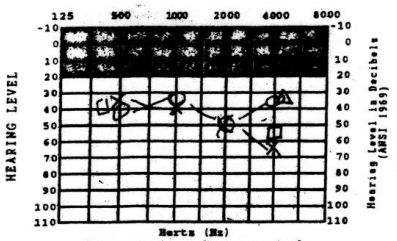
Æ _____ 5 TEST

CHOOL Hooper ___ TEACHER

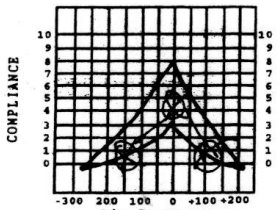
DATE 8-21-06 REFERRAL SOURCE parent / J. Kunugi

	Air	Air Masked	Unmasked Bone	Bone Masked	No Response Air	No Response Bone	VRA
Right	O	△	<	⊏	0↘	↘	(
Left	X	□	>	⊐	↙X	↘	)

AUDIOGRAM



TYMPANOGRAM



Physical Volume: RE 0.7 LE 0.9

Middle Ear Function:

Normal: Right X Left X

Middle Ear Fluid: Right ___ Left ___

Negative Pressure: Right ___ Left ___

Otoacoustic Emissions: Absent for both ears.

Speech

	Speech Reception Threshold	Discrimination	Speech Awareness Level
R.E.	dB HL	% dB HL	dB HL
L.E.	dB HL	% dB HL	dB HL

RECOMMENDATIONS:

X Medical Referral to ENT to evaluate hearing loss

X Audiological Recheck Every 3 months

X Annual Hearing Assessment

( ) Monitor Hearing Through School Screening

( ) Full Assessment in a Sound Suite

X Preferential Seating: _____

X Gain child's attention prior to beginning instruction.

( ) Child will need to be able to see the speaker's face in order to follow communication.

X Encourage the child to ask for clarification of concepts or instructions.

( ) Hearing may vary from day to day. Encourage child to ask questions.

X Use of ear protection when exposed to loud sounds.

( ) Change from last evaluation.

( ) No Audiologic recommendations at this time.

Results: Mild sensorineural hearing loss for the right ear.
Mild-moderate sensorineural hearing loss for the left ear.

PEDIGREE CHAIN [FRANK---ERNEST---ADA---CAROLYN---RODNEY---LUKE]

UNIT 101

MARY S, G4

4/25/07

AGE 70

Patient Information

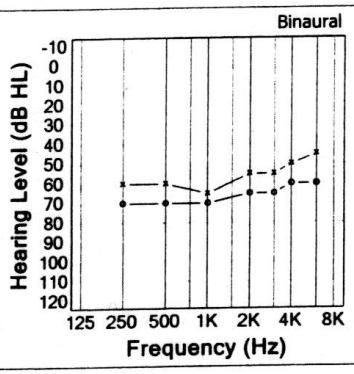
Exam Date:

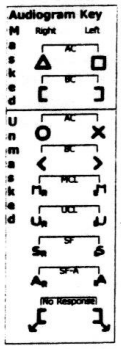
Office Information

Office Name: GREENVILLE OFFICE
Address: 110 Mills Avenue

Phone: (864) 232-3999
Fax:

Examiner Name: P. JEROME WILKERSON
License: 272
Calibration Date: 12/15/2006





Air Conduction

	Right				Left				Binaural		
Unaided	HL (...	EM	(P)		HL (...	EM	(P)		HL-R	HL-L	(P)
SRT											
MCL											
UCL											

Aided	HL (...	EM	(P)	HL (...	EM	(P)	HL-R	HL-L	(P)
SRT									
MCL									
UCL									

(P) = Parameter Key

U = Unaided
A = Aided
MLV = Monitored Live Voice
REC = Recorded
H = Headphone
S = Soundfield
Q = Quiet
N = Noise

PTA - 3 Frequencies
Right - 110 dB HL
Left - 108.3 dB HL

	Right				Left				Binaural				
WR	**%**	HL...	EM	(P)	%	HL...	EM	(P)	%	HL-R	HL-L	(P)	Word List	Count
1														
2														
3														
4														

Comments

PEDIGREE CHAIN [FRANK---TOM---FRANCES---MARY]

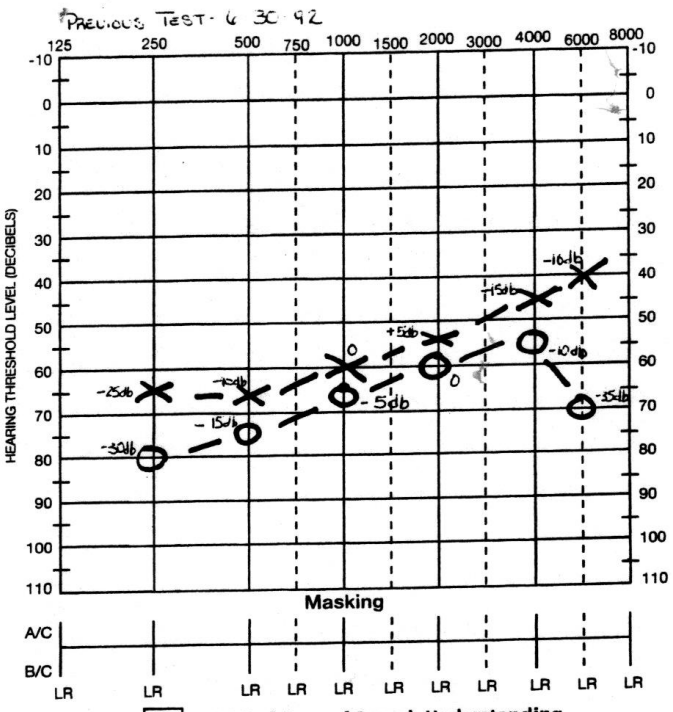
UNIT 102

MARY S, G4

3/24/03

AGE 66

PURE TONE AUDIOGRAM
FREQUENCY, HZ (CPS)



= Critical Area of Speech Understanding

AUDIOGRAM KEY

	LEFT	RIGHT
AC Unmasked	X	O
AC Masked	□	△
BC Mastoid Unmasked	>	<
BC Mastoid Masked	]	[
BC Forehead Unmasked	⌐	¬
MCL		
UCL		

BOTH
BC Forehead Unmasked
Sound Field S

Examples of No Response Symbols

Calibration ☐SPL ☐HTL ☐Tape ☐MLV
Audiometer Make Model
☐Headphones ☐Inserts

SPEECH	Ear Tested	Left	Right	Binaural
	SRT	dB	dB	dB
	MCL	dB	dB	dB
	UCL	dB	dB	dB
	Discrim. %	%	%	

MASTER HEARING AID		Left	Right	Binaural
	Low Cut	dB	dB	
	MCL	dB	dB	
	UCL	dB	dB	
	Discrim. %	%	%	

PURE TONE AVERAGE		Left	Right
	Air Conduction		
	Bone		

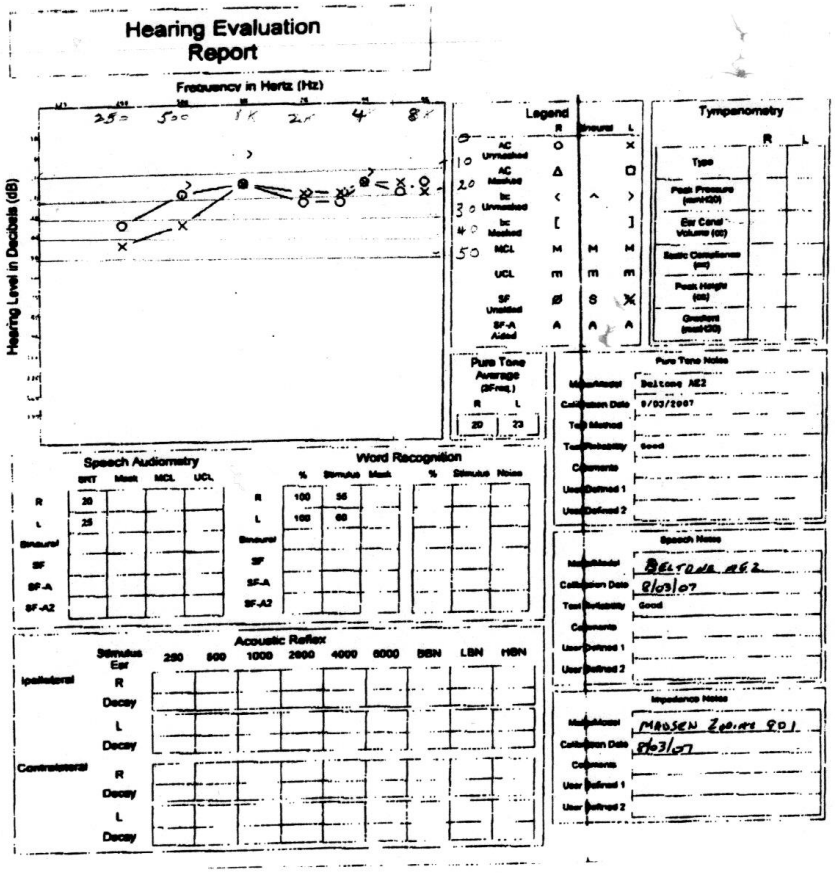
PEDIGREE CHAIN [FRANK---TOM---FRANCES---MARY]

UNIT 103

NANCY M, G4

8/01/08

AGE 54

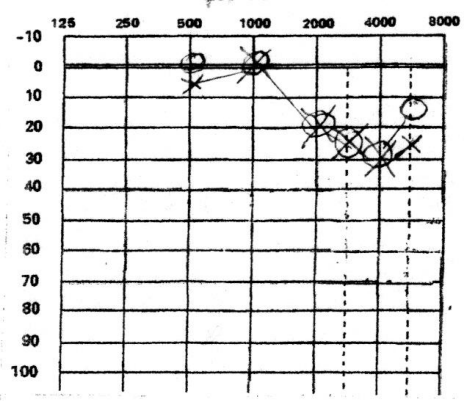


PEDIGREE CHAIN [FRANK---ROSIE---AMY---NANCY]

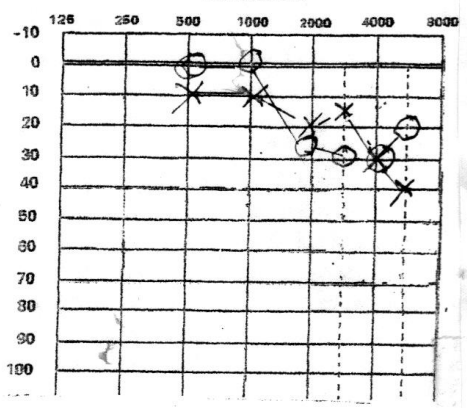
UNIT 104

RANDY R, G5
2004—2007
AGES 39—42

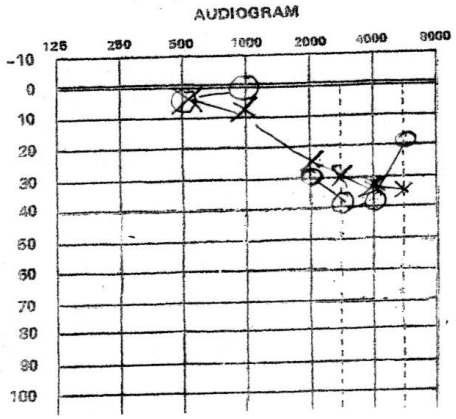
9/21/07---AGE 42



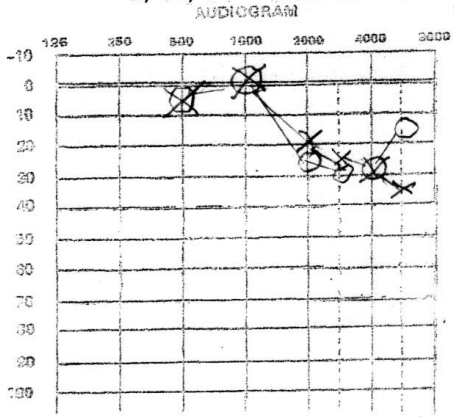
12/02/06---AGE 41



4/17/06---AGE 40



2/02/04---AGE 39

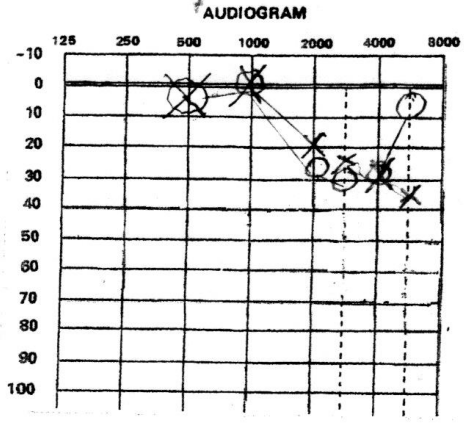


PEDIGREE CHAIN [FRANK---TOM---FRANCES---BOB---RANDY]

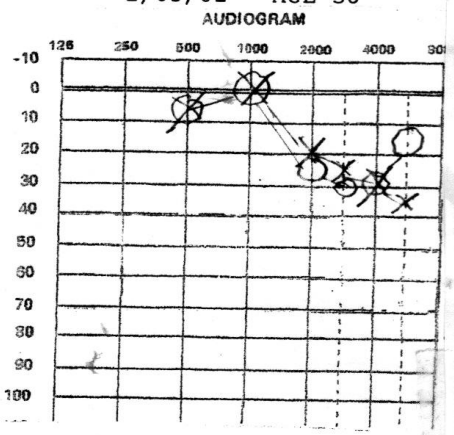
UNIT 105

RANDY R, G5
2001—2002
AGES 35—36

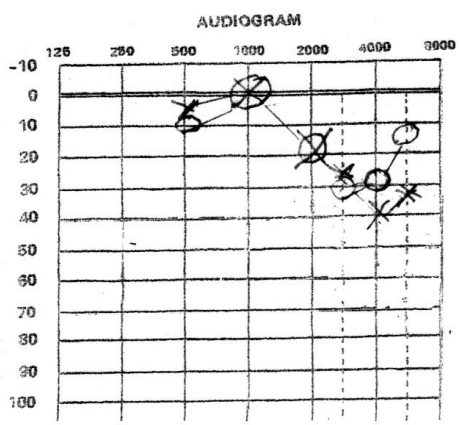
2/09/02---AGE 36

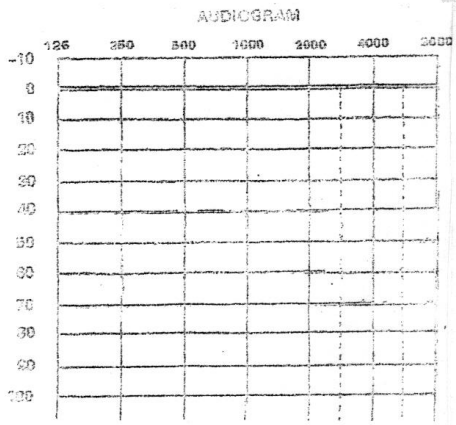


2/05/02---AGE 36



3/11/01---AGE 35





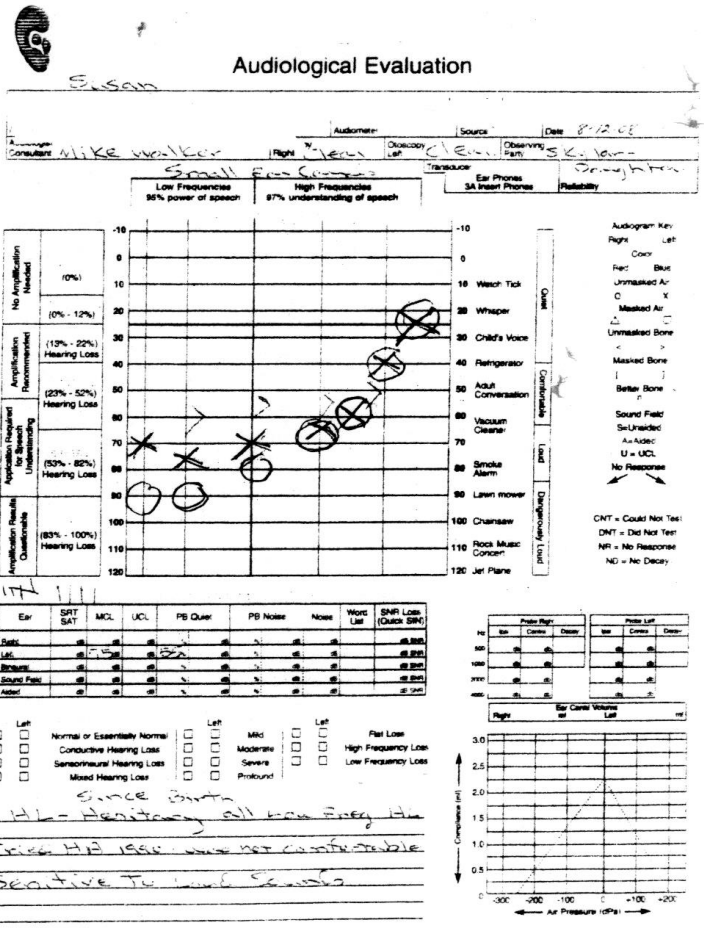
PEDIGREE CHAIN [FRANK---TOM---FRANCES---BOB---RANDY]

UNIT 106

SUE P, G4

8/12/08

AGE 51



PEDIGREE CHAIN [FRANK---TOM---MARION---SUE]

UNIT 107

WES B, G4

4/16/07

AGE 70

UNIVERSITY OF WASHINGTON
Department of Speech and Hearing Sciences
Speech and Hearing Clinic

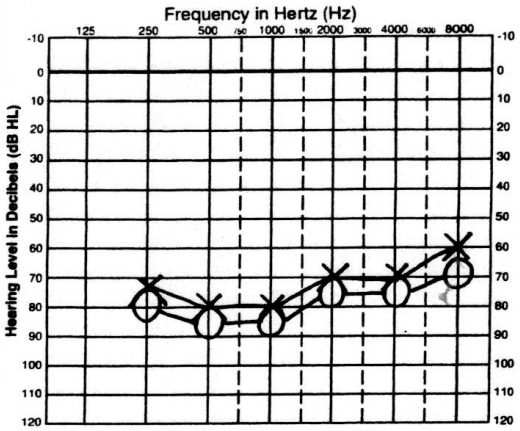
RECORD OF AUDIOMETRIC EVALUATION

CALIBRATION
Current ANSI reference levels for pure tones and speech

ABBREVIATIONS
AC: Air conduction
BC: Bone conduction
CNE: Could not establish
DNT: Did not test
HL: Hearing Level
MCL: Most comfortable level
MLV: Monitored live voice
NBN: Narrow band noise
NR: No response
SPL: Sound Pressure level
SPN: Speech noise
TD: Threshold of discomfort
WN: White noise
WNL: Within normal limits

RESPONSE RELIABILITY
Good / Fair / Poor

PT STIMULUS
Steady / Pulsed / FM



Effective Masking Levels to non-test ear

		250 Hz	500Hz	1K Hz	2K Hz	4K Hz	8K Hz
AC	L						
	R						
BC	L						
	R						

AUDIOGRAM KEY
Right Left
AC unmasked
AC masked
BC mastoid unmasked
BC mastoid masked
BC forehead masked
BC forehead unmasked
Sound field
Aided
Example of no response symbols

AUDIOMETER
No: GSI 61
Make:
Model: 0305

TRANSDUCER
ER-3A

SPEECH AUDIOMETRY

	PTA	SRT	Speech discrimination		MCL	UCL	
Right	82	70	54 % dB List	95 % HL List	60 dB HL	100 dB HL	½ list W-22 PB-50
Masking in left							CNE WNL
Left	76	70	72 % dB List	95 % HL List			WIPI Other:
Masking in right							Tone SPN MLV
Soundfield							

ACOUSTIC REFLEX

PROBE	STIM	Frequency 5	1	2	Decay
Right	IPSI				
	Contra				
Left	IPSI	DNT			
	Contra				

IMMITTANCE

Y-226 Hz	Right	Left
Press (da Pa)		
Vol (cc)	DNT	
Admit (ml)		

COMMENTS
audiologic re-eval. Thresholds stable as compared to previous audio on 11.07.05. DNT BC today.

PEDIGREE CHAIN [FRANK---TOM---OLA---WES]

CHAPTER 9---CLOSING

UNIT 108---POST SCRIPT---THE BUCHANAN FAMILY CURSE?

Recent research uncovered some rather shocking facts. It seems that for hundreds of years ancestral inbreeding, along with hereditary hearing loss has been an old family tradition. This includes marriages between full siblings, half siblings, first cousins, second cousins, etc. Here then is an update on the latest findings.

Shortly before submitting this manuscript for online publication, new evidence was discovered regarding the origins of the Smith Family Curse. The hereditary gene has now been traced all the way back to the fifteenth century--1490. And, of course, it goes back much farther than that. The original contents of the research report have been updated somewhat to reflect this new information.

During the original research, it was never determined whether the hearing loss gene originated with Great-Great Grandpa Alvin Smith or with his wife, Julia Addeline McConnell Smith. It was obvious that one or the other had to have possessed the DFNA/DFNA1 gene—as one or the other had most definitely passed the defective gene down to their descendants.

Remember, it only takes one parent to pass the hereditary hearing loss gene down to his/her offspring. Approximately ½ of his/her offspring will inherit the hereditary hearing loss gene and show symptoms. When two parents both carry the hereditary hearing loss gene, it doubles the chances of their offspring having a hereditary hearing loss. This means that virtually every one of their offspring will be hearing impaired. It is also suspected that when both parents posses the DFNA/DFNA1 gene that the gene becomes even stronger.

Sad to say, but the hereditary hearing loss gene that is shared by all descendants of Alvin Smith, Frank Smith, and John Alvin Smith has been kept active and even magnified by careless inbreeding of family members prior to 1900. There are at least two documented cases of siblings marrying and producing offspring. There

are numerous cases of cousins marrying and producing children. Sorry to report, but every one of us descended from these inbred ancestors.

The DFNA/DFNA1 gene was traced all the way beck to a George Buchanan and his wife—both born in 1490 in Scotland. They were not related to Great-Grandpa Alvin Smith; however, George and Margaret Buchanan were Great-Great Grandma Julia's relatives. In fact, George and Margaret Buchanan were Julia's great- great-great-great-great-great-great-great-great-grandparents!

Therefore, the answer to the riddle of whether it was Alvin or Julia who carried the hereditary hearing loss gene has been solved---it was Julia Addeline McConnell Smith who handed it down to approximately 50% of her descendants. In fact, Julia Addeline's Uncle Rolly, born in 1840, was both deaf and dumb and listed as such in the 1850 census. Rolly Buchanan was also an uncle to Sarah Frances Buchanan Smith.

It has not yet been determined whether it was George or Margaret [1490] that carried the hereditary hearing loss gene. Best guess would be both of them as they were probably cousins or even half siblings!

Frank's 521-year, 18-generational hearing loss chain reads:

1. GEORGE BUCHANAN-1490/1562 & or MARGARET EDMONSTONE-1490/1530
2. THOMAS BUCHANAN-1510/1590 + [JANET BUCHANAN-1520/1595] siblings
3. JOHN BUCHANAN-1545/1577
4. GEORGE BUCHANAN-1576/?
5. JOHN BUCHANAN-1629/1662
6. ALEXANDER BUCHANAN-1670/1728
7. SAMUEL BUCHANAN-1690/1774
8. MATTHEW BUCHANAN-1725/1779 married cousin

9. ELIZABETH BUCHANAN-1750/1792 married ½ brother/first cousin

10. JOHN M BUCHANAN-1778/1847--(Offspring of brother/sister parents--father of deaf and dumb Rolly-- Frank & Sarah's most common ancestor)

11. ELIZABETH JANE BUCHANAN-1826/? (Sister to deaf & dumb Rolly)

12. JULIA ADDELINE MCCONNELL SMITH-1847/1924 (Rolly's niece)

13. WILLIAM FRANKLIN (FRANK) SMITH-1865/1921 (married cousin—Rolly's great nephew)

14. THOMAS SMITH-1889/1960

15. OLA M SMITH-1911/1997

16. WES-living

17. CORY-living

18. ALLISON-living

Sarah's 521-year, 17-generational hearing loss chain reads:

1. GEORGE BUCHANAN-1490/1562 & or MARGARET EDMONSTONE-1490/1530

2. THOMAS BUCHANAN-1510/1590 + JANET BUCHANAN-1520/1595 (siblings)

3. JOHN BUCHANAN-1545/1577

4. GEORGE BUCHANAN-1576/?

5. JOHN BUCHANAN-1629/1672

6. ALEXANDER BUCHANAN-1670/1728

7. SAMUEL BUCHANAN-1690/1774

8. MATTHEW BUCHANAN-1725/1779 (married cousin)

9. ELIZABETH BUCHANAN-1750/1792 (married ½ brother/first cousin)

19. JOHN M BUCHANAN-1778/1847----(Offspring of brother/sister parents--father of deaf and dumb Rolly-- Frank & Sarah's most common ancestor)

10. JOHN L T BUCHANAN-1834/1907 (brother to deaf & dumb Rolly)

11. SARAH FRANCES BUCHANAN SMITH (married cousin—Rolly's niece}

12. THOMAS SMITH-1889/1960

13. OLA M SMITH-1911/1997

14. WES-living

15. CORY-living

16. ALLISON-living

Tracing the hereditary hearing loss chain dead ended with George and Margaret Buchanan (1490) as it is not known which one or both carried the DFNA/DFNA1 gene. Probably both of them!

The majority of our active Smith Clan members are either Frank or John Smith's great-grandchildren. In this case George and Margaret Buchanan—both born in 1490—are our great-great-great-great-great-great-great-great-great-great-great-great-great-grandparents. We are the sixteenth consecutive generation with traceable hereditary hearing loss in this chain, and there are two more generations of documented hereditary hearing loss that come after us!

It is awesome when you stop to realize that our hereditary hearing loss gene goes back 521+ years! This is one potent gene.

UNIT 109---DIRECTORY

Unit 25---- Betty G
Unit 26---- Bob R
Unit 27----- Brad B
Unit 28---- Cheryl M
Unit 29---- Clement S
Unit 30---- Cory M
Unit 31---- David R
Unit 32---- Ernest S
Unit 33---- Floyd L
Unit 34---- Frances R
Unit 35---- Frank S
Unit 36---- Gladys R
Unit 37---- Grant R
Unit 38---- Irene G
Unit 39---- James S
Unit 40---- Jay M
Unit 41---- Jim B
Unit 42---- John S
Unit 43---- Judy B
Unit 44---- Julia B
Unit 45---- Ken R
Unit 46---- Lillian M
Unit 47---- Luke A
Unit 48---- Marcus W
Unit 49---- Marion M
Unit 50--- Mary S
Unit 51---- Nancy M
Unit 52---- Nora S
Unit 53---- Ola M
Unit 54---- Randy R
Unit 55---- Robert A
Unit 56---- Rosa F
Unit 57---- Rose H
Unit 58---- Rosie C
Unit 59---- Ruth S
Unit 60---- Sarah S
Unit 61---- Shirley B

Unit 62---- Stan M
Unit 63---- Steve M
Unit 64--- Sue P
Unit 65---- Til B
Unit 66---- Tom S
Unit 67---- Wes B
Unit 68---- William S

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Unit 99---- Luke A, age 5
Unit 100--- Luke A, age 5
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