

Distinct Expression Patterns Of Causative Genes Responsible For Hereditary Progressive Hearing Loss In Non-Human Primate Cochlea

Makoto Hosoya¹, Masato Fujioka¹, Kaoru Ogawa¹, and Hideyuki Okano^{2,*}

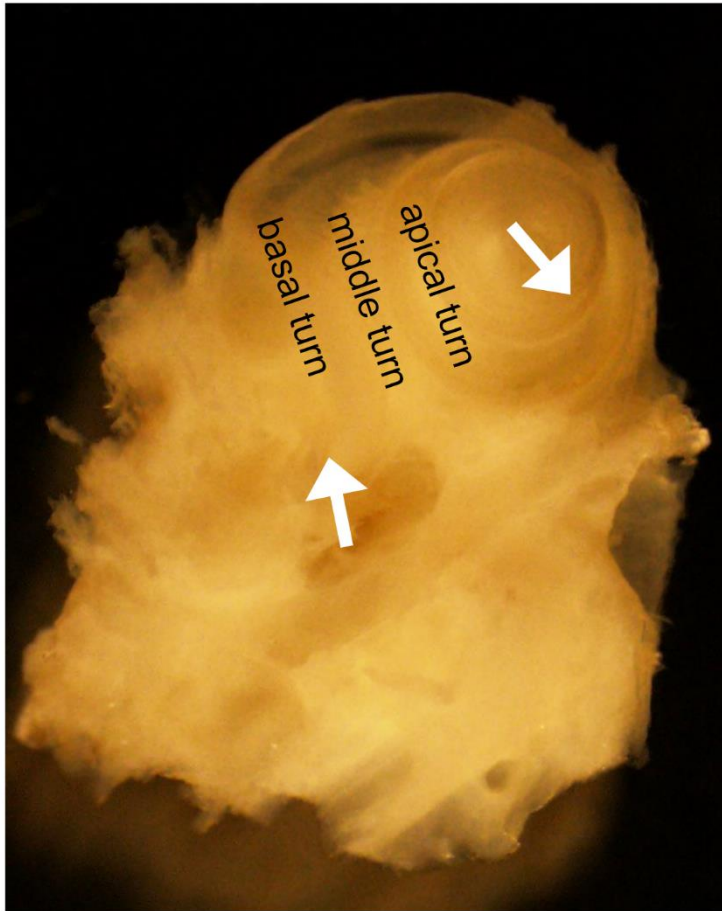
1 Keio University School of Medicine, Department of Otorhinolaryngology, Head and Neck Surgery, 35

Shinanomachi Shinjyuku-ku Tokyo, 160-8582, Japan

2 Keio University School of Medicine, Department of Physiology, 35 Shinanomachi Shinjyuku-ku

Tokyo, 160-8582, Japan

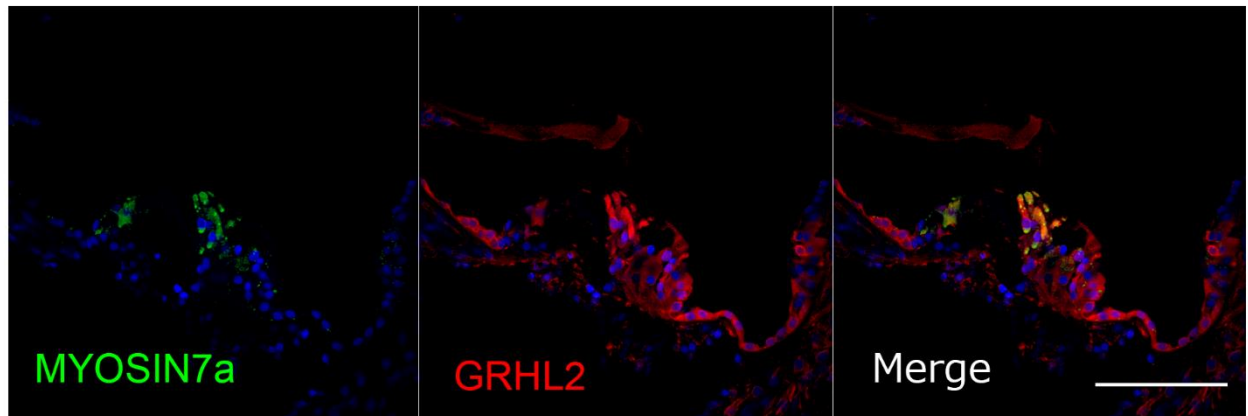
[*hidokano@a2.keio.jp](mailto:hidokano@a2.keio.jp)



Supplementary Figure S.1

The common marmoset cochlea

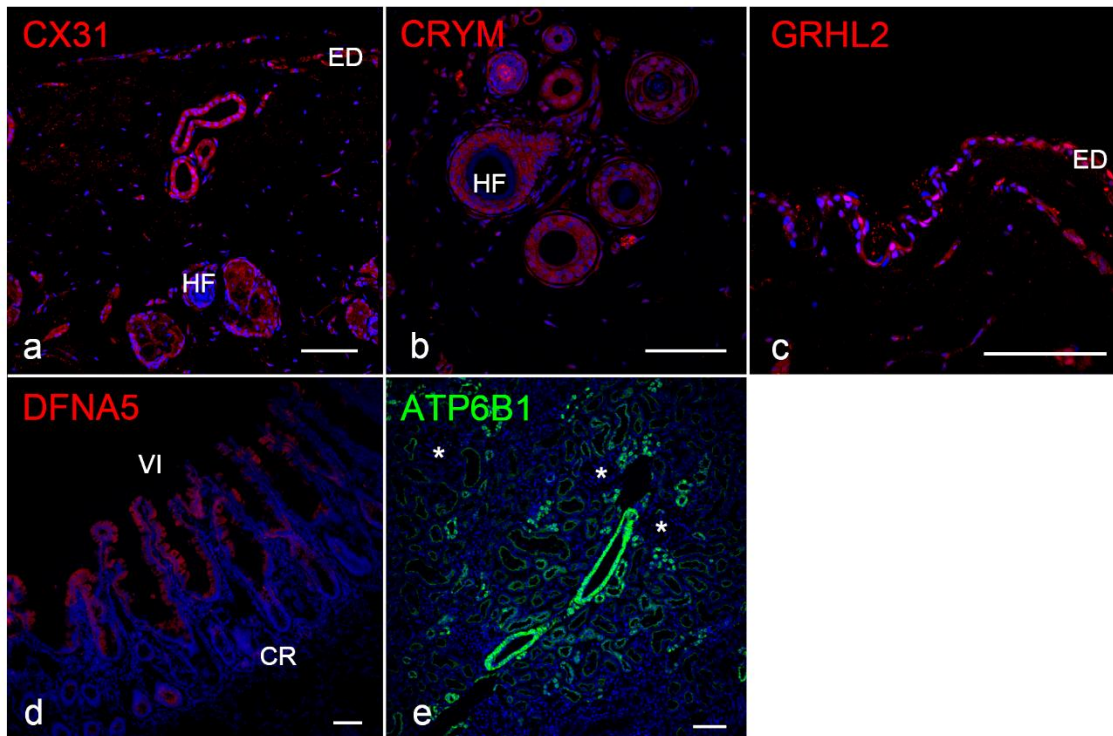
The common marmoset cochlea has 2 and 3/4 turns (between allows) with apical, middle and basal turns as similar to that of human. Note that the cochlear bone capsule was removed.



Supplementary Figure. S2

Cytoplasmic expression of GRHL2

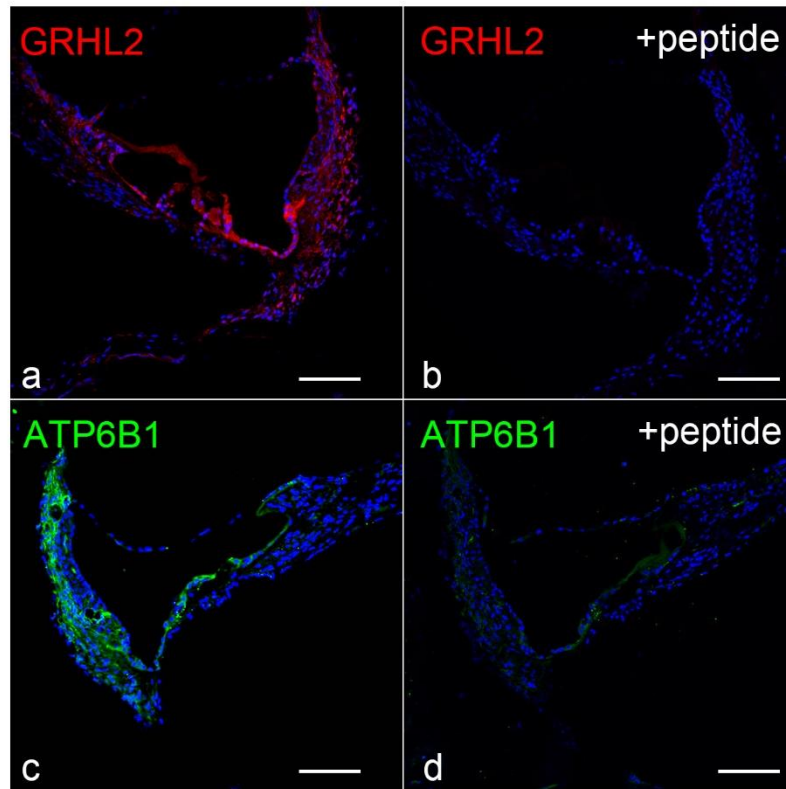
Immunostaining for GRHL2 was observed in the nuclear and cytoplasm. We confirmed the localization by using two different antibodies. Like as shown in Fig.4 (using antibody from Sigma-Aldrich), the immunoreactivity was also detected in cytoplasm with another anti GRHL2 antibody from LSBio. The nuclei were counterstained with Hoechst (blue). Scale bar: 100 μm .



Supplementary Figure. S3

Expressions of CX31, CRYM and GRHL2 in the skin, DFNA5 in the small intestine and ATP6B1 in the kidney

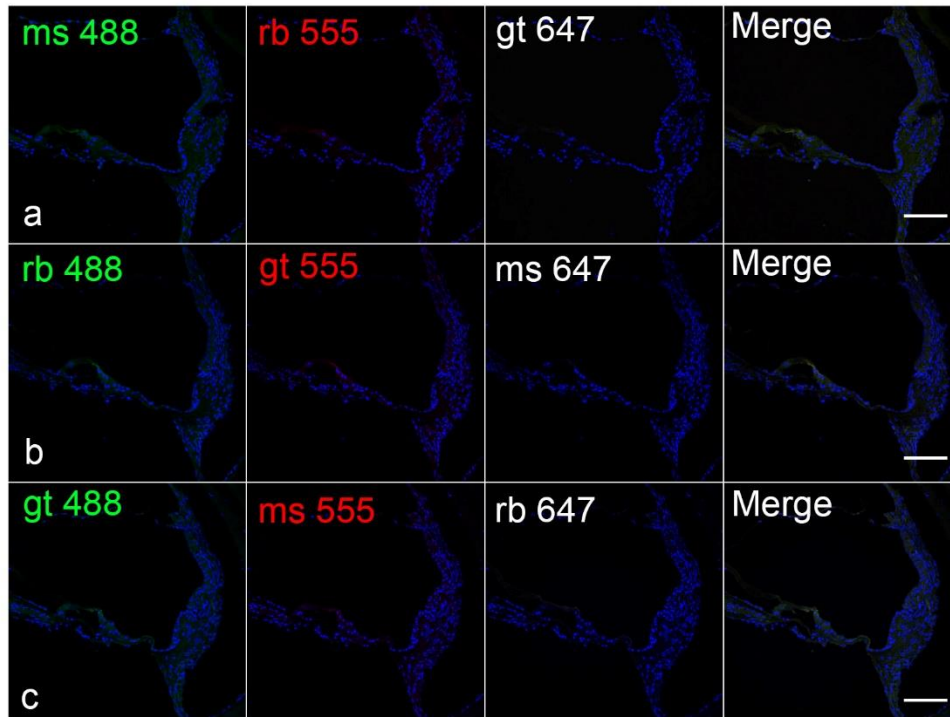
For validating antibodies specificities, we performed immunostaining of the skin, the small intestine and the kidney of common marmoset. (a-c) CX31, CRYM and GRHL2 immunoreactivities were detected in hair follicles (HF) or epidermal layers (ED) and not detected in surrounding connective tissues. (d) DFNA5 immunoreactivities were detected in villus (VI) and not detected in crypt (CR) of the small intestine. (e) ATP6B1 immunoreactivities were detected in collecting ducts and tubules and no immunoreactivities was detected in glomeruli (*). The nuclei were counterstained with Hoechst (blue). Scale bar: 100 μ m.



Supplementary Figure. S4

Antibody absorption test by antigen peptide

For validating antibodies specificities, we performed antibody absorption test by antigen peptide. Immunoreactivities (a and c) were abolished by incubations with antigen peptide (b and d). The nuclei were counterstained with Hoechst (blue). Scale bar: 100 μ m.



Supplementary Figure. S5

Validations of secondary antibodies.

For validating secondary antibodies' specificities, we performed immunostaining without primary antibody incubations. No immunoreactivities were observed. The nuclei were counterstained with Hoechst (blue). Scale bar: 100 μ m. ms: anti-mouse IgG rb: anti-rabbit Ig G gt: anti-goat IgG, 488: Alexa 488, 555: Alexa 555, 647: Alexa 647

Supplementary Figure. S6

a. CX31

Homo sapiens MDWKTLLQALLSGV NKYSTAFGR I WLSVVFVFWVLVYVVA AERVWGDEQKD 50
Callithrix jacchus MDWKTLLQALLSGV NKYSTAFGR I WLSVVFVFRVLVYVVA AERVWGDEQKD 50
Mus musculus MDWKRLQDLLSGV NQYSTAFGR I WLSVVFVFRVLVYVVA AERVWGDEQKD 50

Homo sapiens FDCNTKQPGCTNVCYDNYFPI SNI RLWALQLI FVTCPSLLVI LHVAYREE 100
Callithrix jacchus FDCNTKQPGCTNVCYDNFFPI SNI RLWALQLI FVTCPSLLVI LHVAYREE 100
Mus musculus FDCNTKQPGCTNVCYDNFFPI SNI RLWALQLI FVTCPSMLVI LHVAYREE 100

Homo sapiens RERRHRQKHGDQCAKLYDNAGKKHGGWLWWTYLFSLI FKLII EFLFLYLLH 150
Callithrix jacchus RERRHRQKHGDQCSKLYDNAGKKHGGWLWWTYLFSLI FKLII EFLFLYLLH 150
Mus musculus RERKHRQKHGEQCAKLYSHPGK HGGWLWWTYLFSLI FKLII EFLFLYLLH 150

Homo sapiens TLWHGFNMPRLVQC ANVAPCPNI VDCYI ARPTEKKI FTYF MVGASAVCI V 200
Callithrix jacchus TLWHGFNMPRLVQC ANVAPCPNI VDCYI ARPTEKKI FTYF MVGASAVCI V 200
Mus musculus TLWHGFTMPRLVQCA SIVPCPNFVDCYI ARPTEKKVFTYF MVGASAVCI I 200

Homo sapiens LTI CELCYLI CHRVLRLG LHKDKPRGGCSPSSS ASRASTCRCHHKLVEAGE 250
Callithrix jacchus LTI CELCYLI CHRVLRLALR KDKPQGGHTPSSFASRASTCRCHHKLVEAGE 250
Mus musculus LTI CEICYLI FHRMRGISK GKSTKSLSPKSSASRASTCRCHHKLLESGD 250

Homo sapiens VDPDPGNNKLGASAPNLTPI 270
Callithrix jacchus LDPHPGNNKLGASAPNMTPI 270
Mus musculus PEADPA SEKLGASAPSLTPI 270

b. CRYM

Homo sapiens MSRVPAFLSAAEVEEHLRSS SLLI PPLETALANFSSGPEGGV MQPVRTVV 50
Callithrix jacchus MSRVPAFLSAAEVEEHLRSS SLLI PPLETALANFSSGPEGGV MQPVRTVV 50
Mus musculus MKRAPAFLSAE EVDHLRSS SLLI PPLEAALANFSSGPDGGVMQPVRTVV 50

Homo sapiens PVTKHRGYLGVM PAYSAAEDALTTKLVTFYEDRGI TSVVPSHQATVLLFE 100
Callithrix jacchus PVTKHRGYLGVM PAYSAAEDALTTKLVTFYEDRGI TSVVPSHQATVLLFE 100
Mus musculus PVAKHRGF LGVM PAYSAAEDALTTKLVTFYE GHSNTAVPSHQASVLLFD 99

Homo sapiens PSNGTLLAVMDGNVI TAKRTAAVSAI ATKFLKPPSSEVL CI LGAGVQAYS 150
Callithrix jacchus PSNGTLLAVMDG SVI TAKRTAAVSAI ATKFLKPPSSEVL CI LGAGVQAYS 150
Mus musculus PSNGSLLAVMDGNVI TAKRTAAVSAI ATKLLKPPGSDVLCI LGAGVQAYS 149

Homo sapiens HYEI FTEQFSFKEVRI WNRTKENAEKFADTVQGEVRVCSSVQEAVAGADV 200
Callithrix jacchus HYEI FTEQFSFKEVRI WNRTKENAEKFADTVQGEVRVCSSVQEAVT GADV 200
Mus musculus HYEI FTEQFSFKEVRMWNRTRENAEKFASTVQGDVVRVCSSVQEAVT GADV 199

Homo sapiens IITVTLATEPI LFG EWVKPGAHI NAVGASRPDWRELDDEL MKEAVLYVDS 250
Callithrix jacchus IITVTLATEPI LFG EWVKPGAHI NAVGASRPDWRELDDEL MKEAVLYVDS 250
Mus musculus IITVTMATEPI LFG EWVKPGAHI NAVGASRPDWRELDDEL MRGAVLYVDS 249

Homo sapiens QEAALKESGDVLLS GAEIFAELGEVI KGVKPAHCEKTTVFKSLGMAVEDT 300
Callithrix jacchus QEAALKESGDVLLS GAEIFAELGEVI KGVKPAHCEKTTVFKSLGMAVEDT 300
Mus musculus REAALKESGDVLLS GAEIFAELGEVI S GAKPAHCEKTTVFKSLGMAVEDL 299

Homo sapiens VAAKLIYDSWSSGK 314
Callithrix jacchus VAAKLIYDSWSSGK 314
Mus musculus VAAKLYYDSWSSGK 313

c. GRHL2

<i>Homo sapiens</i>	T L E A T K S L R Q K Q G E G P M T Y L N K G Q F Y A I T L S E T G D N K C F R H P I S K V R S V V	300
<i>Callithrix jacchus</i>	T L E A T K S L R Q K Q G E G P M T Y L N K G Q F Y A I T L S E T G D N K C F R H P I S K V R S V V	300
<i>Mus musculus</i>	T L E A T K S L R Q K Q G E G P M T Y L N K G Q F Y A I T L S E T G D N K C F R H P I S K V R S V V	300
<hr/>		
<i>Homo sapiens</i>	M V V F S E D K N R D E Q L K Y W K Y W H S R Q H T A K Q R V L D I A D Y K E S F N T I G N I E E I	350
<i>Callithrix jacchus</i>	M V V F S E D K N R D E Q L K Y W K Y W H S R Q H T A K Q R V L D I A D Y K E S F N T I G N I E E I	350
<i>Mus musculus</i>	M V V F S E D K N R D E Q L K Y W K Y W H S R Q H T A K Q R V L D I A D Y K E S F N T I G N I E E I	350
<hr/>		
<i>Homo sapiens</i>	A Y N A V S F T W D V N E E A K I F I T V N C L S T D F S S Q K G V K G L P L M I Q I D T Y S Y N N	400
<i>Callithrix jacchus</i>	A Y N A V S F T W D V N E E A K I F I T V N C L S T D F S S Q K G V K G L P L M I Q I D T Y S Y N N	400
<i>Mus musculus</i>	A Y N A V S F T W D V N E E A K I F I T V N C L S T D F S S Q K G V K G L P L M I Q I D T Y S Y N N	400
<hr/>		
<i>Homo sapiens</i>	R S N K P I H R A Y C Q I K V F C D K G A E R K I R D E E R K Q N R K K G K G Q A S Q T Q C N S S S	450
<i>Callithrix jacchus</i>	R S N K P I H R A Y C Q I K V F C D K G A E R K I R D E E R K Q N R K K G K G Q T S Q T Q C N S S S	450
<i>Mus musculus</i>	R S N K P I H R A Y C Q I K V F C D K G A E R K I R D E E R K Q N R K K G K G Q A S Q A Q C N S S	450
<hr/>		
<i>Homo sapiens</i>	D G K L A A I P L Q K K S D I T Y F K T M P D L H S Q P V L F I P D V H F A N L Q R T G Q V Y Y N T	500
<i>Callithrix jacchus</i>	D G K L A A I P L Q K K S D I T Y F K T M P D L H S Q P V L F I P D V H F A N L Q R T G Q V Y Y N T	500
<i>Mus musculus</i>	D G K M A A I P L Q K K S D I T Y F K T M P D L H S Q P V L F I P D V H F A N L Q R T G Q V Y Y N T	500
<hr/>		
<i>Homo sapiens</i>	D D E R E G G S V L V K R M F R P M E E E F G P V P S K Q M K E E G T K R V L L Y V R K E T D D V F	550
<i>Callithrix jacchus</i>	D D E R E G S V L V K R M F R P M E E E F G P V P S K Q M K E E G T K R V L L Y V R K E T D D V F	549
<i>Mus musculus</i>	D D E R E G S S V L V K R M F R P M E E E F G P T P S K Q T K E E N V K R V L L Y V R K E N D D V F	550
<hr/>		
<i>Homo sapiens</i>	D A L M L K S P T V K G L M E A I S E K Y G L P V E K I A K L Y K K S K K G I L V N M D D N I I E H	600
<i>Callithrix jacchus</i>	D A L M L K S P T V K G L M E A I S E K Y G L P V E K I A K L Y K K S K K G I L V N M D D N I I E H	599
<i>Mus musculus</i>	D A L M L K S P T V K G L M E A T S E K Y G L P V E K I T K L Y K K S K K G I L V N M D D N I I E H	600
<hr/>		
<i>Homo sapiens</i>	Y S N E D T F I L N M E S M V E G F K V T L M E I	625
<i>Callithrix jacchus</i>	Y S N E D T F I L N M E S M V E G F K V T L M E I	624
<i>Mus musculus</i>	Y S N E D T F I L N M E S M V E G F K T T L M E I	625

d. DFNA5

<i>Homo sapiens</i>	MFAKATRNFLREVDADGDLI AVSNLNDSDKLQLLSLVTKKKRFWCWQRPK	50
<i>Callithrix jacchus</i>	MFAKATK NFLREVDADGDLI AVSNLNDSDKLQLLSLVTKKKRFWCWQRPK	50
<i>Mus musculus</i>	MFAKATRNFLKEVDA GGDLI S VSHLNDSDKLQLLSLVTKKKR YWCWQRPK	50
<i>Homo sapiens</i>	YQFLSLTLGDVLI EDQFPSPVVVESDFVKYEGKFANHVS GTLETALGKVK	100
<i>Callithrix jacchus</i>	YQFLS I TLGDVL K EDQFPSPVVVESDFVKY ESKFANHVS GTL D TALGKVK	100
<i>Mus musculus</i>	Y Q T L S A T L E D V L T E G H C L S P V V E S D F V K Y E S K C E N H K S G A I G T V V G K V K	100
<i>Homo sapiens</i>	LNLGGSSRVESQSSF GTLRKQEV DLQQLI RDSAERTI NLRNPVLQQVLE G	150
<i>Callithrix jacchus</i>	L N V G G S S R V E S Q S S F G T L R K Q E V D L Q Q L I R D S A E R T I N L R N P V L Q Q V L E E	150
<i>Mus musculus</i>	L N V G G K G V V E S H S S F G T L R K Q E V D V Q Q L I Q D A V K R T V N M D N L V L Q Q V L E S	150
<i>Homo sapiens</i>	RNEVLCVLTQKI ITTMQKCVI SEHMQVVEEKCGGIVG IQTKTVQVSATEDGN	200
<i>Callithrix jacchus</i>	RNEVLCVLTQKI ITTMQKCVI SEH T QVVEEKCGG M VGIQTKTVQVSATEDGN	200
<i>Mus musculus</i>	RNEVLCVLTQKI M T QKCVI SEH V Q S E E T C G G M V G I Q T K T I Q V S A T E D G T	200
<i>Homo sapiens</i>	VTKDSNVVLEIPAATTIAYGVIELYVKLDGLF EFCLLRGKQG GFENKKRI	250
<i>Callithrix jacchus</i>	V M K D S N V V L E I P A A T T I A Y G V I E L Y V K L D G H F E F C L L Q G K E S G F E N K R R I	250
<i>Mus musculus</i>	V T T D T N V V L E I P A A T T I A Y G T M E L F V K G D G Q F E F C L L Q G K H G G F E H E R K L	250
<i>Homo sapiens</i>	DSVYLDHLVFRFAFIDMPDA AHGISSQDGPLSVLKQATLL LERNFHPFA	300
<i>Callithrix jacchus</i>	DSVYLD P L A F R E F A F I D M P D A G H G I S S Q D G P L S V L K Q A T P L L E R N F H P F V	300
<i>Mus musculus</i>	D S V Y L D P L A Y R E F A F I D M P D G G G I S S Q D G P L R V V K Q A T L H L E R S F H P F A	300
<i>Homo sapiens</i>	ELPEPQQTALSDFQAVVFDDELLMVLEPV CDDLVSGLSPTVA VLG--EL	348
<i>Callithrix jacchus</i>	ELPEPQQTALSDFQAVLFDN ELLMVLEPV CDDV V S G L S P P L V V L G -- E L	348
<i>Mus musculus</i>	V L P A Q Q R A L F C V I Q K I L F D E E L L R A L E Q V C D D V A G G L W S S Q A V L A M E E L	350
<i>Homo sapiens</i>	KPRQQQDLVAFQLVGC SLQGCGPPE DAGSKQ-LFMTAYFLVSALAEMP	397
<i>Callithrix jacchus</i>	M P R Q Q Q D V M A F L Q L V G C S L Q G E R P G P E D A G S K R - L F T T A Y F L V S A L A E M P	397
<i>Mus musculus</i>	T D S Q Q Q D L T A F L Q L V G Y R I Q G E H P G P G D E V S N Q K L F A T A Y F L V S A L A E M P	400
<i>Homo sapiens</i>	DSAAALLGTCKLQI IPTLCHLLRALSDDGVSDLEDPTLT PLK DTERFGI	447
<i>Callithrix jacchus</i>	D N A A A L L G T C Y K L Q I I P T L C H L L R A L S D D G V S D L E D P T L A P L K D T E R F G I	447
<i>Mus musculus</i>	D N A T V F L G T C C K L H V I S S L C G L L H A L S D D S V C D F H N P T L A P L R D T E R F G I	450
<i>Homo sapiens</i>	VQRLFASADISLERLKSSVKAVILKDSKVFP LLLCITLNGLCALGREHS-	496
<i>Callithrix jacchus</i>	VQRLFASADI T L E R L K S S V K A V I L K D S E I F P L L L C I T L N G L C A L G R E H S -	496
<i>Mus musculus</i>	VQRLFASADI A L E R M Q F S A K A T I L K D S C I F P L I L H I T L S G L S T L S K E H E E	500
<i>Homo sapiens</i>	-----	
<i>Callithrix jacchus</i>	-----	
<i>Mus musculus</i>	E L C Q S G H A T G Q D	512

e. ATP6B1

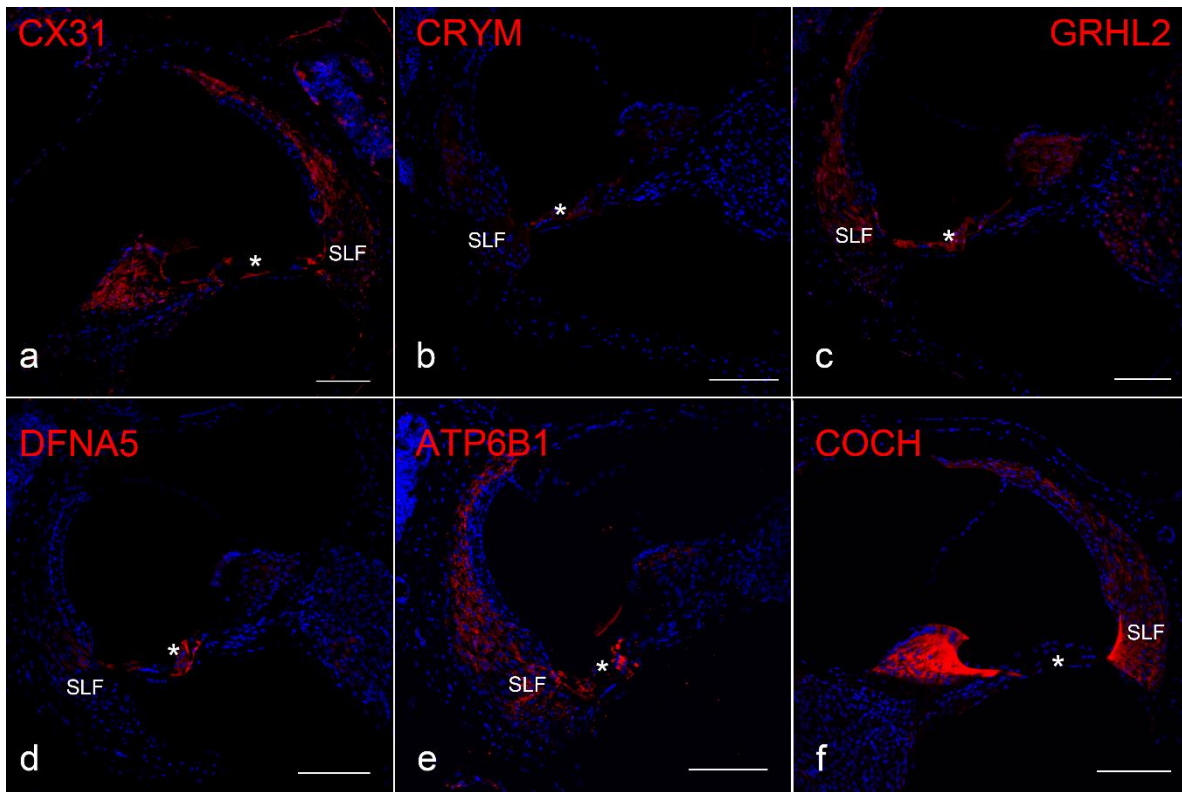
<i>Homo sapiens</i>	M A M E I D S R P G G L P G S S C N L G A A R E H M Q A V T R N Y I T H P R V T Y R T V C S V N G P	50
<i>Callithrix jacchus</i>	M A M E I D S R P G G L P G S G F K L G A A R E H M Q A V T R N Y I T H P R V T Y R T V C S V N G P	50
<i>Mus musculus</i>	M A T T V D S R S S G F T G N S C D P G T A G E H V Q A V T R N Y I T H P R V T Y R T V C S V N G P	50
<i>Homo sapiens</i>	L V V L D R V K F A Q Y A E I V H F T L P D G T Q R S G Q V L E V A G T K A I V Q V F E G T S G I D	100
<i>Callithrix jacchus</i>	L V V L D Q V K F A Q Y A E I V H F T L P D G T Q R S G Q V L E V A G T K A V V Q V F E G T S G I D	100
<i>Mus musculus</i>	L V V L D G V K F A Q Y A E I V N F T L P D G T Q R S G Q V L E V A G T K A I V Q V F E G T S G I D	100
<i>Homo sapiens</i>	A R K T T C E F T G D I L R T P V S E D M L G R V F N G S G K P I D K G P V V M A E D F L D I N G Q	150
<i>Callithrix jacchus</i>	A R K T T C E F T G D I L R T P V S E D M L G R V F N G S G K P I D K G P V V M A E D F L D I N G Q	150
<i>Mus musculus</i>	S Q K T T C E F T G D I L R T P V S E D M L G R I F N G S G K P I D K G P A V M A E E F L D I N G Q	150
<i>Homo sapiens</i>	P I N P H S R I Y P E E M I Q T G I S P I D V M N S I A R G Q K I P I F S A A G L P H N E I A A Q I	200
<i>Callithrix jacchus</i>	P I N P H S R I Y P E E M I Q T G I S P I D V M N S I A R G Q K I P I F S A A G L P H N E I A A Q I	200
<i>Mus musculus</i>	P I N P H G R I Y P E E M I Q T G I S P I D V M N S I A R G Q K I P I F S A A G L P H N E I A A Q I	200
<i>Homo sapiens</i>	C R Q A G L V K K S K A V L D Y H D D N F A I V F A A M G V N M E T A R F F K S D F E Q N G T M G N	250
<i>Callithrix jacchus</i>	C R Q A G L V K K S K A V L D Y H D D N F A I V F A A M G V N M E T A R F F K S D F E Q N G T M G N	250
<i>Mus musculus</i>	C R Q A G L V K K S K A V L D Y H E D N F A I V F A A M G V N M E T A R F F K S D F E Q N G T M G N	250
<i>Homo sapiens</i>	V C L F L N L A N D P T I E R I I T P R L A L T T A E F L A Y Q C E K H V L V I L T D M S S Y A E A	300
<i>Callithrix jacchus</i>	V C L F L N L A N D P T I E R I I T P R L A L T T A E F L A Y Q C E K H V L V I L T D M S S Y A E A	300
<i>Mus musculus</i>	V C L F L N L A N D P T I E R I I T P R L A L T T A E F L A Y Q C E K H V L V I L T D M S S Y A E A	300
<i>Homo sapiens</i>	L R E V S A A R E E V P G R R G F P G Y M Y T D L A T I Y E R A G R V E G R G G S I T Q I P I L T M	350
<i>Callithrix jacchus</i>	L R E V S A A R E E V P G R R G F P G Y M Y T D L A T I Y E R A G R V E G R G G S I T Q I P I L T M	350
<i>Mus musculus</i>	L R E V S A A R E E V P G R R G F P G Y M Y T D L A T I Y E R A G R V E G R G G S I T Q I P I L T M	350
<i>Homo sapiens</i>	P N D D I T H P I P D L T G F I T E G Q I Y V D R Q L H N R Q I Y P P I N V L P S L S R L M K S A I	400
<i>Callithrix jacchus</i>	P N D D I T H P I P D L T G F I T E G Q I Y V D R Q L H N R Q I Y P P I N V L P S L S R L M K S A I	400
<i>Mus musculus</i>	P N D D I T H P I P D L T G F I T E G Q I Y V D R Q L H N R Q V Y P P I N V L P S L S R L M K S A I	400
<i>Homo sapiens</i>	G E G M T R K D H G D V S N Q L Y A C Y A I G K D V Q A M K A V V G E E A L T S E D L L Y L E F L Q	450
<i>Callithrix jacchus</i>	G E G M T R K D H G D V S N Q L Y A C Y A I G K D V Q A M K A V V G E E A L T S E D L L Y L E F L Q	450
<i>Mus musculus</i>	G E G M T R K D H G D V S N Q L Y A C Y A I G K D V Q A M K A V V G E E A L T S E D L L Y L E F L Q	450
<i>Homo sapiens</i>	K F E K N F I N Q G P Y E N R S V F E S L D L G W K L L R I F P K E M L K R I P Q A V I D E F Y S R	500
<i>Callithrix jacchus</i>	K F E K N F I N Q G P Y E N R S V F E S L D L G W K L L R I F P K E M L K R I P Q A M I D E F Y S R	500
<i>Mus musculus</i>	K F E K N F I T Q G P Y E N R T V F E S L D L G W K L L R I F P K E M L K R I P Q S M T D E F Y S R	500
<i>Homo sapiens</i>	E G A L Q D L A P D T A L	513
<i>Callithrix jacchus</i>	E G A P Q D P A P D T A L	513
<i>Mus musculus</i>	Q G A Q Q D P A S D T A L	513

f. COCH

<i>Homo sapiens</i>	MSAAWIPALGLGVCLLLLPGP--AGSEGAAPIAITCFTTRGLDIRKEKADV	48
<i>Callithrix jacchus</i>	MSAAWIQTFLFLGVCLQLLPGP--MGSEGAAPIAITCFTTRGLDIRKEKADV	48
<i>Mus musculus</i>	MPSSRIPALGLGAWLLLLLLPFRFAEAIVPIPVITCFTTRGLDIRKEKADV	50
<i>Homo sapiens</i>	LCPGGCPLEEFVSYGNIVYASVSSICGAAVHRGVISNSGGPVRVYSLPGR	98
<i>Callithrix jacchus</i>	LCPGGCPLEEFVSYGNIVYASVSSICGAAVHRGVISNSGGPVRVYSLPGR	98
<i>Mus musculus</i>	LCPGGCSLEEFVSYFGNIVYASVSSICGAAVHRGVIGTISGGPVRVYSLPGR	100
<i>Homo sapiens</i>	ENYSSVDANGIQSQMLSRWSASFVTVKGKSSTQEATGQAVSTAHPPTGKR	148
<i>Callithrix jacchus</i>	ENYSSVDANGIQSQMLSRWSASFVTVKGKSSTQEATGQAVSTAHPPTGKR	148
<i>Mus musculus</i>	ENYSSVDANGIQSQMLSRWSASFAVTKGKSSTQEATGRAVSTAHPPTSGKR	150
<i>Homo sapiens</i>	LKKTPEKKTGNKDCKADIAFLIDGGSFNIGQRRFNLQKNFVGKVALMLGIG	198
<i>Callithrix jacchus</i>	LKKTPEKKTGNKDCKADIAFLIDGGSFNIGQRRFNLQKNFVGKVALMLGIG	198
<i>Mus musculus</i>	LKKTPEKKTGNKDCKADIAFLIDGGSFNIGQRRFNLQKNFVGKVALMLGIG	200
<i>Homo sapiens</i>	TEGPHVGLVQASEHPKIEFYLNFTSAKDVLFAIKEVGFRRGNSNTGKAL	248
<i>Callithrix jacchus</i>	TEGPHVGLVQASEHPKIEFYLNFTSAKDVLFAIKEVGFRRGNSNTGKAL	248
<i>Mus musculus</i>	TEGPHVGLVQASEHPKIEFYLNFTSAKDVLFAIKEVGFRRGNSNTGKAL	250
<i>Homo sapiens</i>	KHTAQKFFTVDAGVRKGIKVVVVVIDGWPSDDIEEAGIVAREFGVNVFI	298
<i>Callithrix jacchus</i>	KHTAQKFFTVDAGVRKGIKVVVVVIDGWPSDDIEEAGIVAREFGVNVFI	298
<i>Mus musculus</i>	KHTAQKFFTADTGVVRKGIKVVVVVIDGWPSDDIEEAGIVAREFGVNVFI	300
<i>Homo sapiens</i>	VSVAKPIPEELGMVQDVTFVDKAVCRNNGFFSYHMPNWFQTTKYVKPLVQ	348
<i>Callithrix jacchus</i>	VSVAKPIPEELGMVQDVSFVDKAVCRNNGFFSYHMPNWFQTTKYVKPLVQ	348
<i>Mus musculus</i>	VSVAKPIPEELGMVQDVAFVDKAVCRNNGFFSYHMPNWFQTTKYVKPLVQ	350
<i>Homo sapiens</i>	KLCTHEQMMCSKTCYNSVNI AFLIDGSSSVGDSNFRMLLEFVSNI AKTFE	398
<i>Callithrix jacchus</i>	KLCTHEQMMCSKTCYNSVNI AFLIDGSSSVGDSNFRMLLEFVSNI AKTFE	398
<i>Mus musculus</i>	KLCTHEQMMCSKTCYNSVNI AFLIDGSSSVGDSNFRMLLEFVSNI AKTFE	400
<i>Homo sapiens</i>	ISDIGAKIAAVQFTYDQRTEFSFTDYSTKENVLAVIRNIRYMSGGTATGD	448
<i>Callithrix jacchus</i>	ISDIGAKIAAVQFTYDQRM EFSFTDYNTKENVLAVIRNIRYMSGGTATGD	448
<i>Mus musculus</i>	ISDIGAKIAAVQFTYDQRTEFSFTDYNTKENVLAVLANIRYMSGGTATGD	450
<i>Homo sapiens</i>	AISFTVRNVFGPIRES PNKNFLVIVTDGQSYDDVQGPAAAAHDAGITIFS	498
<i>Callithrix jacchus</i>	AISFTVRNVFGPIRES PNKNFLVIVTDGQSYDDVQGPAAAAHDAGITIFS	498
<i>Mus musculus</i>	AIAFTVRNVFGPIRES PNKNFLVIVTDGQSYDDVRGPAAAAHDAGITIFS	500
<i>Homo sapiens</i>	VGVAWAPLDDLKDMASKPKESHAFFTREFTGLEPIVSDVIRGICRDFLES	548
<i>Callithrix jacchus</i>	VGVAWAPLDDLKDMASKPKESHAFFTREFTGLEPIVSDVIRGICRDFLES	548
<i>Mus musculus</i>	VGVAWAPLDDL RDMASKPKESHAFFTREFTGLEPIVSDVIRGICRDFLES	550
<i>Homo sapiens</i>	Q Q	550
<i>Callithrix jacchus</i>	Q Q	550
<i>Mus musculus</i>	Q Q	552

Comparisons of amino-acid sequences between human (*Homo sapience*), common marmoset (*Callithrix jaccus*) and mouse (*Mus musculus*)

We compared amino-acid sequences of these three animals and we showed in these six deafness genes amino-acid sequences of common marmoset is more similar to human than mouse. (a) Homology of amino-acid sequences of CX31 was shown. Human vs common marmoset: 95.19%, human vs mouse 82.22% (b) Homology of amino-acid sequences of CRYM was shown. Human vs common marmoset: 99.04%, human vs mouse 88.50% (c) Homology of amino-acid sequences of GRHL2 was shown. Human vs common marmoset: 98.88%, human vs mouse 94.72% (d) Homology of amino-acid sequences of DFNA5 was shown. Human vs common marmoset: 92.14%, human vs mouse 71.37% (e) Homology of amino-acid sequences of ATP6B1 was shown. Human vs common marmoset: 98.44%, human vs mouse 94.72% (f) Homology of amino-acid sequences of COCH was shown. Human vs common marmoset: 98.55%, human vs mouse 93.45%



Supplementary Figure. S7

Immunostaining of mouse specimens.

(a) Anti-CX31 antibody recapitulated previous report and strong immunoreactivities were detected in the spiral ligament fibrocytes of mouse and no immunoreactivities was detected in the organ of Corti. (b) Anti-CRYM antibody did not work well against mouse specimens. (c) Anti-GRHL2 antibody used in this report partially recapitulated previous report and immunoreactivities were detected in the organ of Corti. (d) Anti-DFNA5 antibody used in this report partially recapitulated previous report and immunoreactivities were detected in the organ of Corti. (e) Anti-ATP6B1 antibody recapitulated previous report and strong immunoreactivities were detected in the spiral ligament fibrocytes of mouse. (f) Anti-COCH antibody recapitulated previous report and strong immunoreactivities were detected in the spiral ligament fibrocytes of mouse and no immunoreactivities was detected in the organ of Corti. SLF: Spiral Ligament Fibrocytes, *. Organ of Corti. The nuclei were counterstained with Hoechst (blue). Scale bar: 100 μ m.