

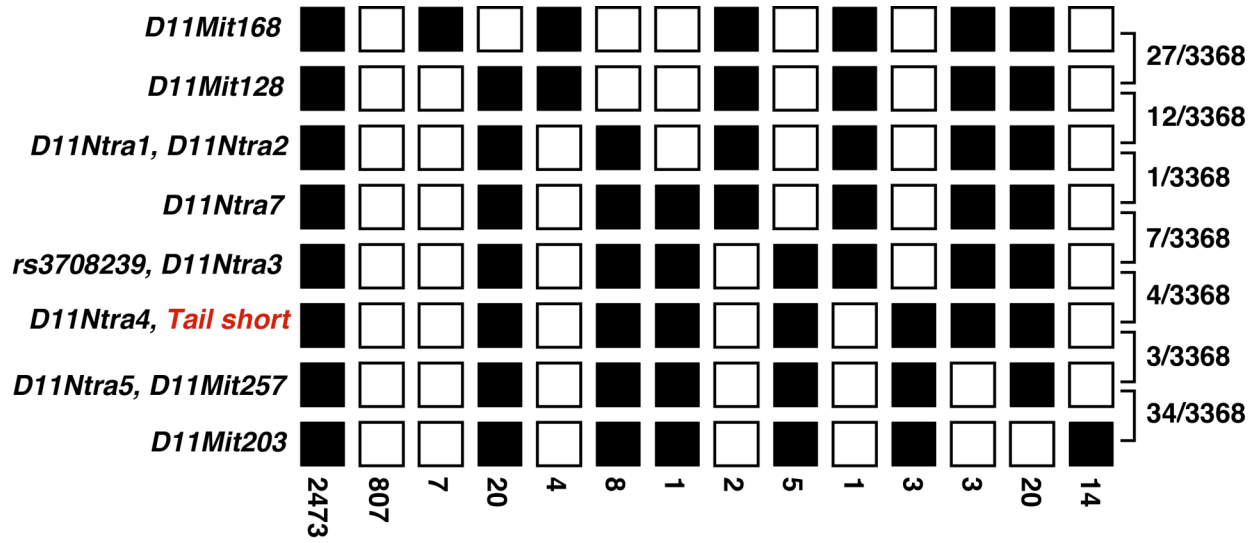
## Supplemental Information

**Supplemental Figure S1. Haplotypes of *Ts* F2 progeny.** Haplotypes of 1684 F2 intercross progeny at the marker loci given on the left. Black box represents the wildtype Black Swiss allele and the white box refers to the *Ts* chromosome. Number at the bottom of each column represents the number of chromosomes encountered in the cross and the numbers on the right indicate the number of recombinants per meioses. The skewed number of wildtype chromosomes in the F2 cross is due to the embryonic and perinatal lethality of the *Ts* mutation.

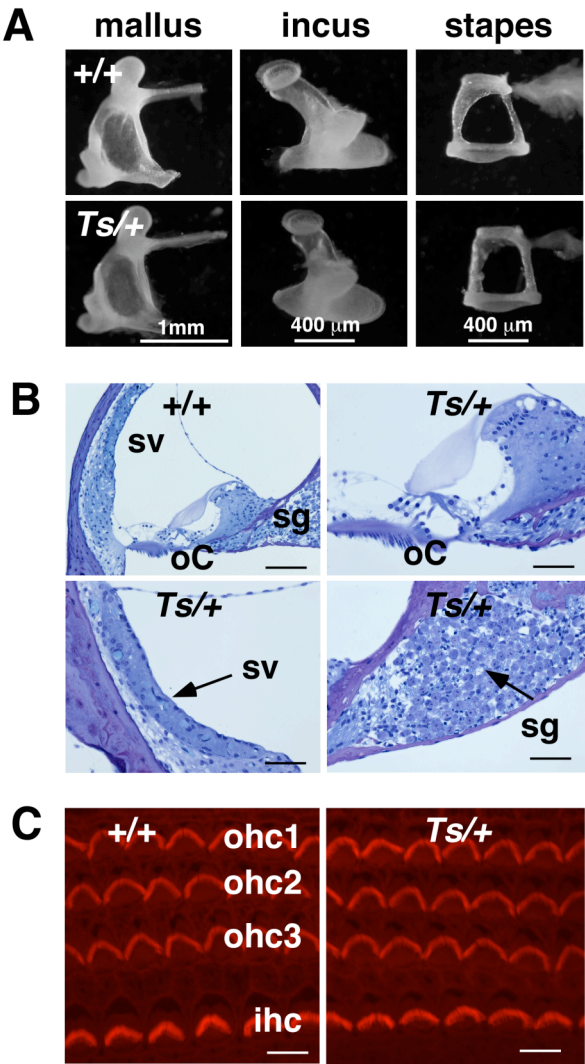
**Supplemental Figure S2. Inner ear morphology in *Ts* mice.** (A) Photographs of middle ear bones in +/+ and *Ts*/+ mice. Note the cellular debris around the stapes in *Ts*. (B) Images of plastic-embedded toluidin-stained cross sections through the cochlear coil. Sv, stria vascularis; oC, organ of Corti; sg, spiral ganglion. Scale bar = 50  $\mu$ m. (C) Confocal surface images of organs of Corti whole mounts stained with phalloidin revealing the stereociliary hair bundle structure. ohc, outer hair cell; ihc, inner hair cell. Scale bar = 5  $\mu$ m.

**Supplemental Figure S3. qPCR of *Rp138* expression in the cochlea.** Poly-A<sup>+</sup> RNA extracted from *Ts*/+ and +/+ cochlea (three cochleae pooled for each genotype) at postnatal day 14, was reversed transcribed and used as template in qPCR using *Rp138*- and *GAPDH*-specific Taqman probes (Applied Biosystems) and TaqMan Universal PCR Master Mix (Applied Biosystems) following manufacturer's instruction. Plotted is the mean  $\Delta$ Ct from three experiments performed in triplicates. n.s., not significant ( $p > 0.05$ ; t-test).

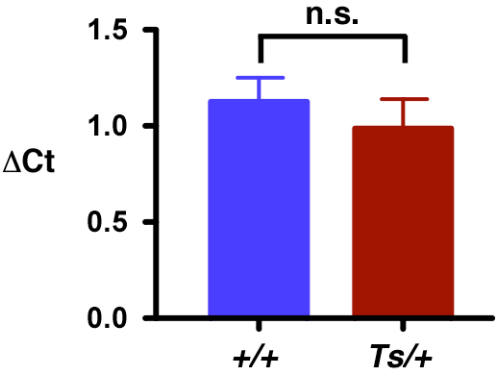
Supplemental Fig. S1



Supplemental Fig. S2



Supplemental Fig. S3



**Supplemental Table S1 • Serum chemistry and hematology in *Ts* mice**

	<b>+/+</b>		<b><i>Ts</i>/+</b>		<b><i>p</i></b>	<b>Unit</b>
	<b>mean</b>	<b>SD</b>	<b>mean</b>	<b>SD</b>		
<b>albumin</b>	2.55	0.19	2.57	0.12	0.858	g/dL
<b>alkaline phosphatase</b>	160.83	21.54	172.83	24.79	0.392	U/L
<b>ALT</b>	35.67	2.73	46.33	24.09	0.392	U/L
<b>AST</b>	92.83	43.79	88.33	47.64	0.868	U/L
<b>blood urea nitrogen</b>	20.67	5.57	25.67	7.20	0.208	mg/dL
<b>calcium</b>	10.07	0.72	10.75	0.53	0.091	mg/dL
<b>cholesterol</b>	98.67	16.80	91.67	16.07	0.478	mg/dL
<b>creatine kinase</b>	379.33	373.85	187.17	71.06	0.244	U/L
<b>creatinine</b>	0.22	0.08	0.23	0.12	0.780	mg/dL
<b>glucose</b>	175.67	33.10	215.17	58.14	0.179	mg/dL
<b>LDH</b>	325.83	183.54	265.33	74.76	0.179	U/L
<b>organic phosphate</b>	6.63	0.85	8.20	1.23	0.028	mg/dL
<b>total bilirubin</b>	0.38	0.04	0.37	0.08	0.664	mg/dL
<b>total protein</b>	4.62	0.27	4.67	0.26	0.750	g/dL
<b>triglyceride</b>	178.50	61.94	140.67	57.52	0.299	mg/dL
<b>white blood cells</b>	5.81	2.22	5.06	2.35	0.581	K/ $\mu$ L
<b>red blood cells</b>	8.55	0.70	8.25	0.39	0.383	M/ $\mu$ L
<b>hemoglobin</b>	13.75	0.99	13.28	0.89	0.410	g/dL
<b>hematocrit</b>	53.60	3.85	51.42	2.23	0.257	%
<b>MCV</b>	62.67	2.97	62.35	1.65	0.824	fL
<b>platelets</b>	833.83	41.12	823.50	85.35	0.795	K/ $\mu$ L
<b>PMNs</b>	0.67	0.46	0.85	0.32	0.451	K/ $\mu$ L
<b>lymphocytes</b>	4.69	1.83	3.75	2.10	0.429	K/ $\mu$ L
<b>monocytes</b>	0.41	0.13	0.42	0.23	0.939	K/ $\mu$ L
<b>eosinophils</b>	0.04	0.03	0.04	0.05	1.000	K/ $\mu$ L
<b>basophils</b>	0.01	0.01	0.01	0.01	1.000	K/ $\mu$ L
<b>MCH</b>	16.11	0.65	16.11	0.83	0.995	fL
<b>MCHC</b>	25.68	1.15	25.84	1.34	0.826	g/dL
<b>body weight</b>	25.08	12.32	18.70	18.83	0.019	g

ALT, alanine aminotransferase; AST, aspartate aminotransferase; LDH, lactate dehydrogenase; MCV, mean corpuscular volume; PMN, polymorph nuclear cells; MCH, mean corpuscular hemoglobin; MCHC, mean corpuscular hemoglobin concentration; means are pooled data from three males and three females of the indicated genotype at 12 weeks of age.