

## Loss-of-Function Mutations in the *PRPS1* Gene

### Cause a Type of Nonsyndromic X-linked

### Sensorineural Deafness, DFN2

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Table S1. LOD Scores Reflecting Linkage between Chromosome X Markers and Locus of Inherited Hearing Loss in Family GZ-Z052

Marker	Two-point LOD score at $\theta =$						
	0.00	0.01	0.05	0.1	0.2	0.3	0.4
DXS8020	-2.42	-0.40	0.29	0.55	0.68	0.58	0.35
DXS8096	<b>4.25</b>	<b>4.19</b>	<b>3.93</b>	<b>3.58</b>	<b>2.81</b>	<b>1.96</b>	<b>1.02</b>
DXS1106	2.45	2.41	2.24	2.02	1.56	1.07	0.55
DXS1210	1.50	1.47	1.38	1.25	0.96	0.64	0.31
DXS1059	1.37	1.34	1.24	1.11	0.84	0.55	0.27
DXS8088	0.46	0.45	0.41	0.35	0.23	0.12	0.03
DXS8055	-5.29	-1.25	-0.44	-0.07	0.20	0.24	0.16

Table S2. Expression Profiling of PRPS1 Identified by Quantitative Real-Time RT-PCR

<b>Individual</b>	<b>Sex</b>	<b>Mutation Carrier</b>	<b><math>\Delta C(t) = C(t)PRPS1 - C(t)\beta-ACTIN</math></b>
Affected males:			
IV-1	M	Y	6.53
IV-34	M	Y	6.28
IV-36	M	Y	6.06
IV-9	M	Y	6.45
<b>Average</b>			<b>6.33</b>
Female carriers:			
III-27	F	Y	6.54
III-29	F	Y	6.32
<b>Average</b>			<b>6.43</b>
Unaffected male and normal controls:			
IV-5	M	N	5.71
IV-7	M	N	6.61
C-1	M	N	6.57
C-2	M	N	6.38
<b>Average</b>			<b>6.32</b>