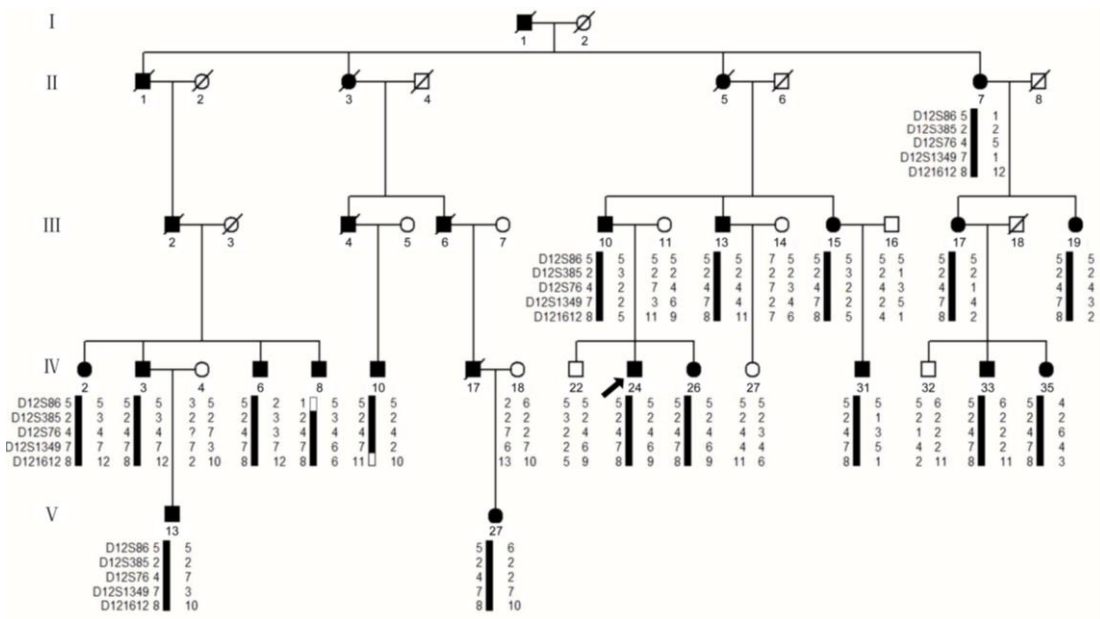


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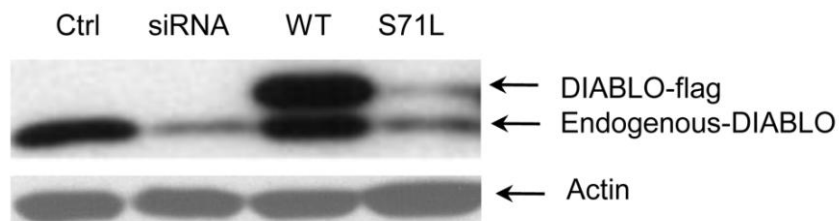
**Supplemental Data**

**Functional Mutation of *SMAC/DIABLO*,  
Encoding a Mitochondrial Proapoptotic Protein,  
Causes Human Progressive Hearing Loss DFNA64**

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**Figure S1. Haplotype analysis of family HN-W078.** The segregating haplotype associated with hearing loss is indicated by a black bar, delimited by markers D12S86 and D12S1612 on chromosome 12.



**Figure S2. Endogenous DIABLO was significantly reduced by siRNA transfection.**

**Table S1. Primers for sequencing of *DIABLO***

Exon		Primer
1	F	5'- GTGGCGACACAGGCAGAG-3'
	R	5'- CAGTAAGAACCCACACCAG-3'
2	F	5'- GGGCGTGGCTATGGGGCGAGGC-3'
	R	5'- GGGGCGGAGGCGGGGCTGTC-3'
3	F	5'- GCCCCTGCAAGTAAATCAA -3'
	R	5'- GCCGGTACTGTGGGGGAAGG-3'
4	F	5'- CGTTTTTGTCTGCCTATTG -3'
	R	5'- AGGCCTGGCTATGTTTCAAT -3'
5+6	F	5'- GAGGCAGCAGGATGGAAGAG -3'
	R	5'- GAGCCACTTAGACCATTACT-3'
7	F	5'- CCCCAGGCCAACGTCCACATG-3'
	R	5'- GCCAAAGTGTAGCAAATG-3'

**Table S2. Variants found in Candidate Genes in Family HN-W078**

Gene	Variant	Amino Acid	Previously Reported	Control Chromosomes
<i>DIABLO</i>	c.377 C>T (NM_019887.4)	p. Ser126Leu	No	0/1000
<i>DIABLO</i>	c.108 A>G (NM_019887.4)	No change	rs12870	
<i>PSMD9</i>	c.50 T>C (NM_002813.4)	p. Val 17 Ala	rs2230681	
<i>SBNO1</i>	c.1698 T>C (NM_001167856.1)	No change	rs61388686	
<i>SBNO1</i>	c.3099 G>A (NM_001167856.1)	No change	rs61751327	
<i>SBNO1</i>	c.3466 C>A (NM_001167856.1)	No change	rs3825141	