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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Data S1. Supplementary Methods

Table S1. Primer pairs used to amplify the *NOTCH3*, *HES1*, and *HES5* coding sequences.

Figure S1. Sequence analysis of the *NOTCH3* mutation in the family of proband B.

Figure S2. Mean ER chaperone/β-actin ratios from densitometry analysis of data presented in Figure 3. The levels of all three chaperones were significantly decreased in cells expressing T900P-NOTCH3 compared with those expressing wild-type NOTCH3 (WT). $^*P < 0.05$.

Figure S3. Western blotting of *NOTCH3* degradation. Stable cells were treated with (Tet+) or without (Tet−) tetracycline (2 μg/mL) for 24 h and then incubated in medium without tetracycline. Cells were harvested on days 0, 1, and 2 and were subjected to SDS-PAGE and western blot analysis. The data are representative of experiments performed for three stable cell lines (WT-17, G840E-36, and T900P-33). Experiments were performed twice for each stable cell line.