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**Article:**

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## Highlights

- A mutation at Leu74Pro of OGR1 found in amelogenesis imperfecta was characterized.
- Either a wild-type or a mutant OGR1 receptor was transfected in HEK-293 cells.
- Both receptors are distributed in plasma membranes.
- The wild-type receptor, but not the mutant receptor, was internalized by protons and  $\text{Ni}^{2+}$ .
- $\text{Ca}^{2+}$  response to ligands was lost in the mutant receptor-expressing cells.