

We claim:

1. A genetically modified mouse whose genome comprises a modified mouse Activin A receptor type 1 (Acvr1) gene comprising:

- i) a human exon 5 of the Acvr1 gene encoding glutamic acid at the first codon instead of aspartic acid in sense orientation flanked upstream and downstream by first pair of recombination recognition sites; and
- ii) a mutant exon 5 of a mouse Acvr1 gene comprising an R206H mutation in antisense orientation flanked upstream and downstream by second pair of recombination recognition sites that are different than the first pair of recombination recognition sites;

wherein the first and second recombination recognition sites are oriented so that a recombinase can invert the mutant exon 5 into sense orientation, delete the human exon 5, and allow a mutant Acvr1 comprising the mutant exon 5 to be expressed resulting in ectopic bone formation.

2. A transgenic mouse derived from the mouse of claim 1, wherein said transgenic mouse has a genome comprising a nucleic acid sequence encoding the mutant Acvr1 comprising the mutant exon 5, and wherein the mouse expresses the mutant Acvr1 comprising the mutant exon 5 resulting in ectopic bone formation.

3. The genetically modified mouse of claim 2, wherein the first and second pairs of recombination recognition sites are Lox2372 and LoxP or vice versa.

4. The genetically modified mouse of claim 2, wherein the recombinase is a tamoxifen-inducible CreER^{T2}.