

FOPの解析から明らかとなった新しいBMP受容体の活性化機構

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1

Fibrodysplasia ossificans progressiva; FOP

- Heterotopic ossification (HO) in skeletal muscle

- Mutations in the *ALK2 (ACVR1)* gene

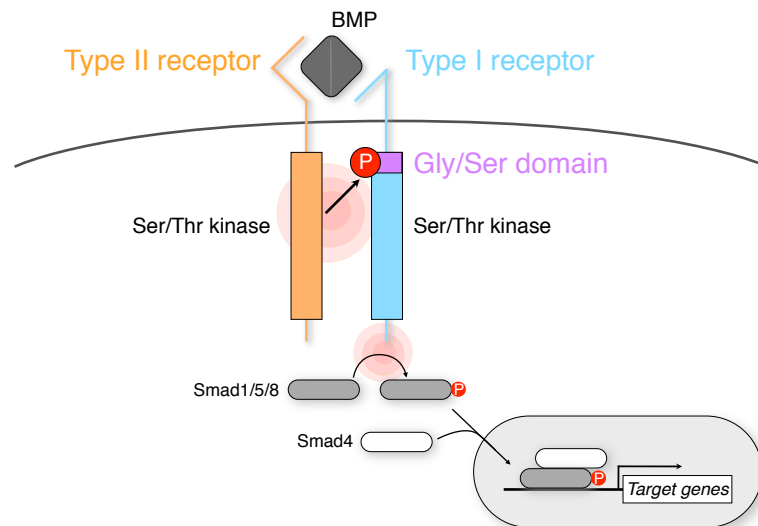
Shore EM, et al., Nat. Genet. (2006)

- Gain-of-function mutants

Fukuda T, et al., J. Biol. Chem. (2009)

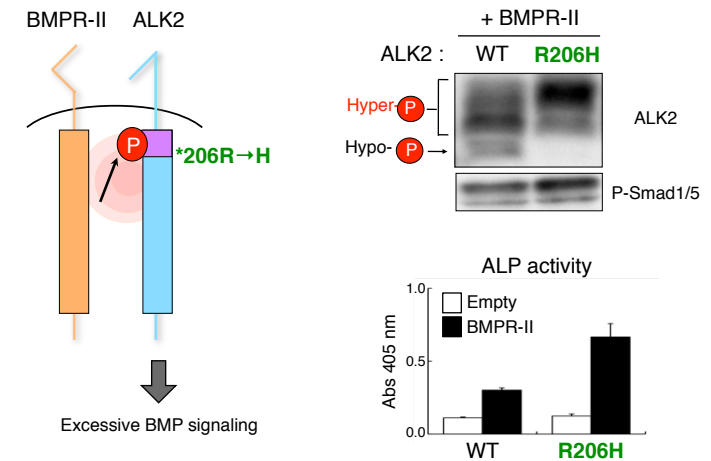
2

Scheme of signal transduction of BMPs



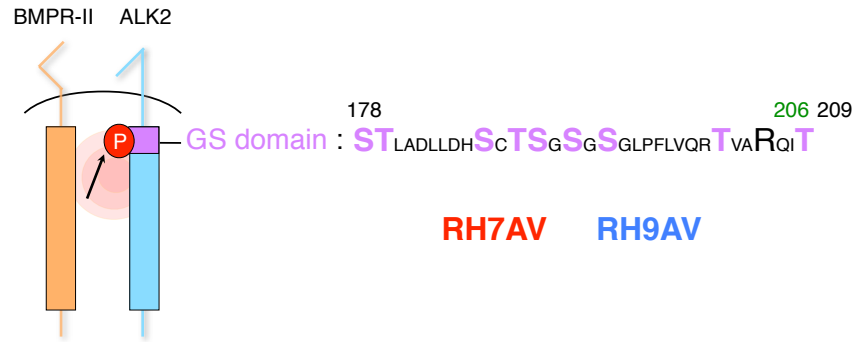
3

ALK2(R206H) is further activated by BMP type II receptor



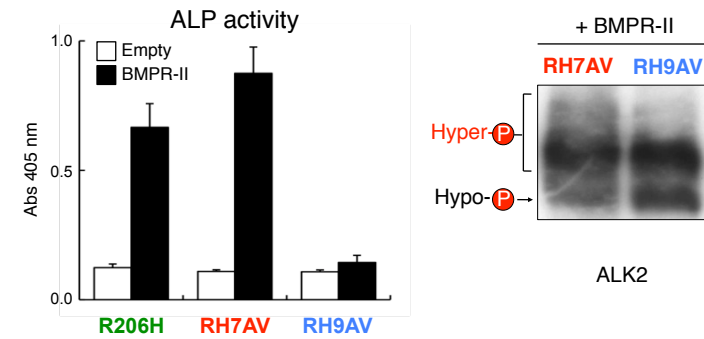
4

Construction of RH7AV and RH9AV mutants



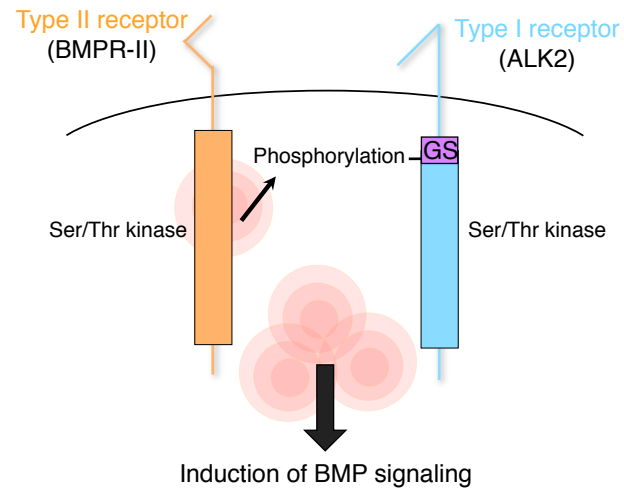
5

RH7AV is activated by BMPR-II



6

Conclusion



7