



第 54 回 『Aberrant activation of IL-6/JAK/STAT3/FOSL1 signaling induces renal abnormalities in a *Xenopus* model of Joubert syndrome-related disorders』

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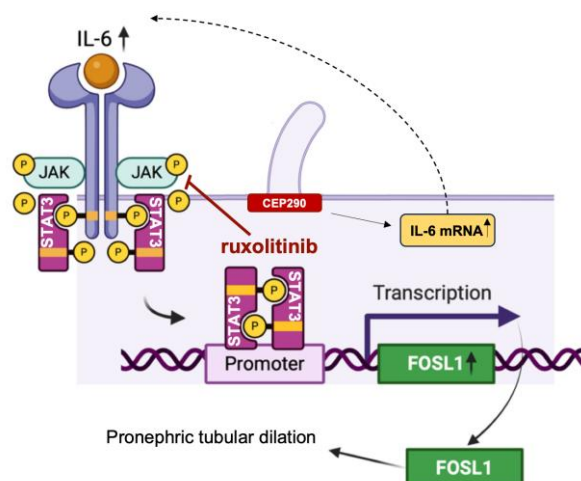
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脳神経科学研究所 5階会議室



Joubert syndrome-related disorders (JSRD) are a group of ciliopathies characterized by the "molar tooth sign" on MRI. *CEP290* is one of the causative genes, whose mutations can lead to brain, retinal and kidney defects. To investigate kidney abnormalities, *CEP290* was depleted in *Xenopus* embryos, resulting in edema and dilated pronephric tubules. Tolvaptan reduced tubular dilation but did not restore normal morphology. RNA-seq revealed activation of the IL-6/JAK/STAT3 pathway in *CEP290* morphants, which was suppressed by tolvaptan. Inhibition of this pathway with ruxolitinib rescued tubular dilation. Moreover, overexpression of FOSL1, a downstream IL-6 target, was sufficient to induce dilation. These results suggest that aberrant IL-6/JAK/STAT3/FOSL signaling drives renal tubule dilation in *CEP290*-associated disease, linking JSRD mutations to cystic kidney pathology.



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