



D4.9 partner [UNIPV]

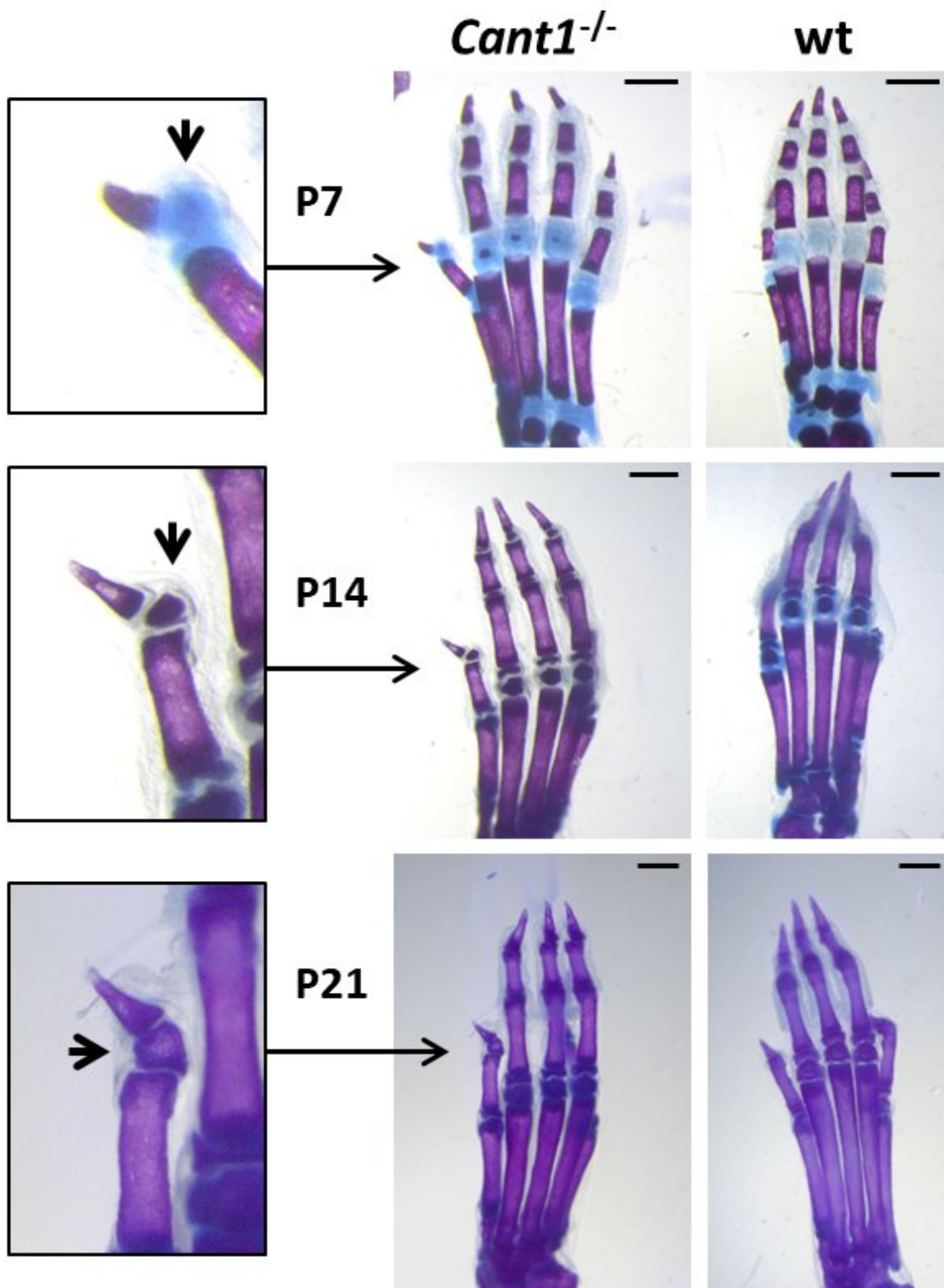
30/08/2018 Prof Antonio Rossi

Growth defect and kyphosis in Cant1^{-/-} mice. X-ray analysis of 60 days old Cant1^{-/-} and wt mice showed reduced skeletal growth in Cant1^{-/-} compared with



wild type animals. In *Cant1*^{-/-} mice moderate thoracic kyphosis was present (arrow). Scale bar 1 cm.

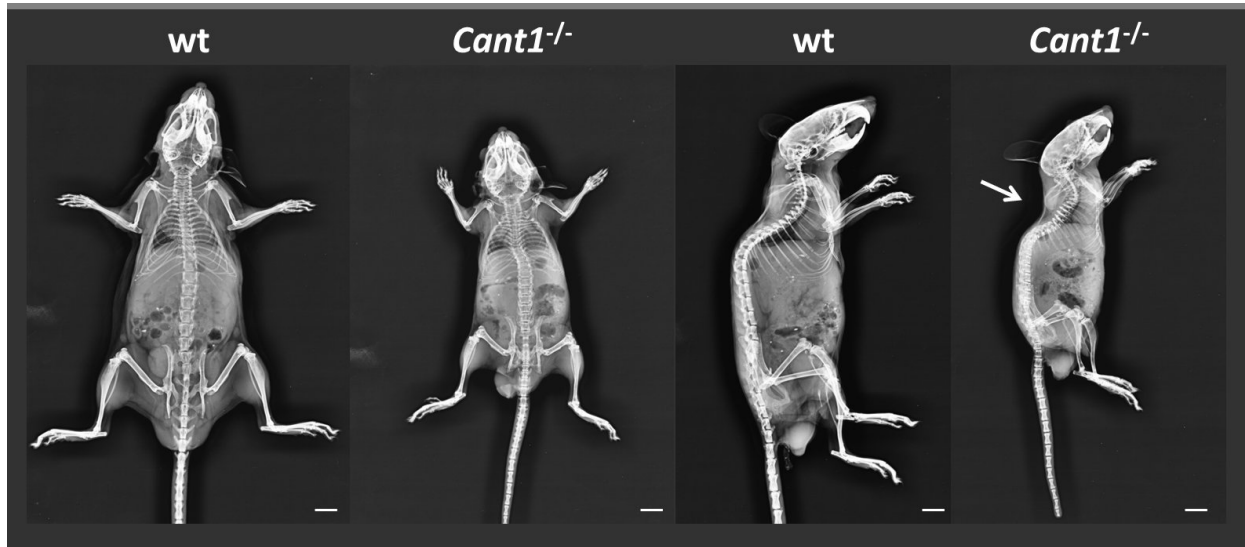
The hind limbs of *Cant1*^{-/-} and wt mice were stained with alcian blue and alizarin red; an additional phalanx (arrow) causing medial deviation of the first digit is evident in *Cant1*^{-/-} mice. This phalanx, named delta phalanx, is observed in patients with Desbuquois dysplasia type 1. Scale bar 1 mm.



delta phalanx.png

SYBIL Project 602300. The SYBIL Project is funded under the European Commission Seventh Framework Programme, grant number 602300, Theme HEALTH.2013.2.1.1-1

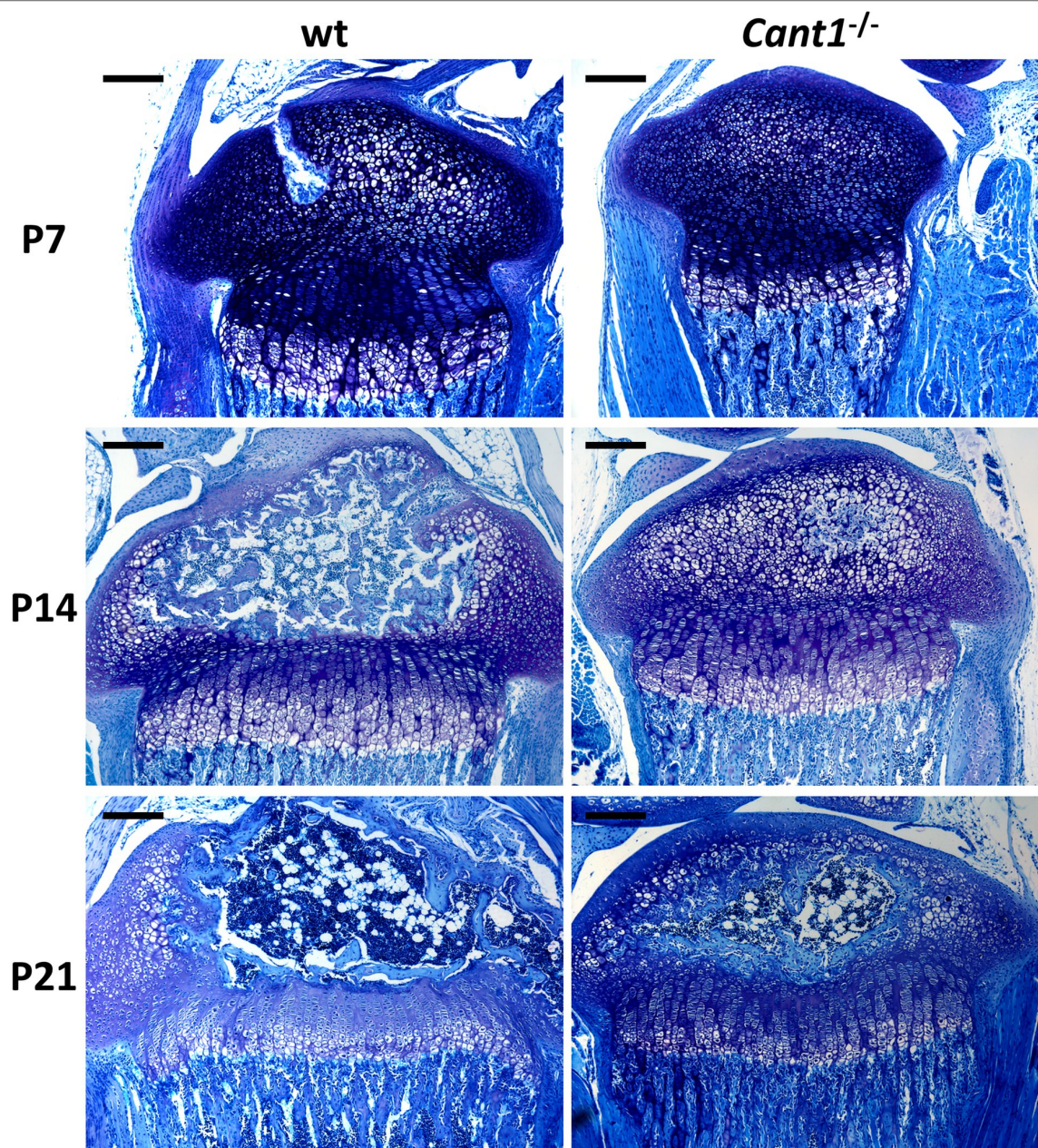
Growth defect and kyphosis in *Cant1*^{-/-} mice. X-ray analysis of 60 days old *Cant1*^{-/-} and wt mice showed reduced skeletal growth in *Cant1*^{-/-} compared with wild type animals. In *Cant1*^{-/-} mice moderate thoracic kyphosis was present (arrow). Scale bar 1 cm.



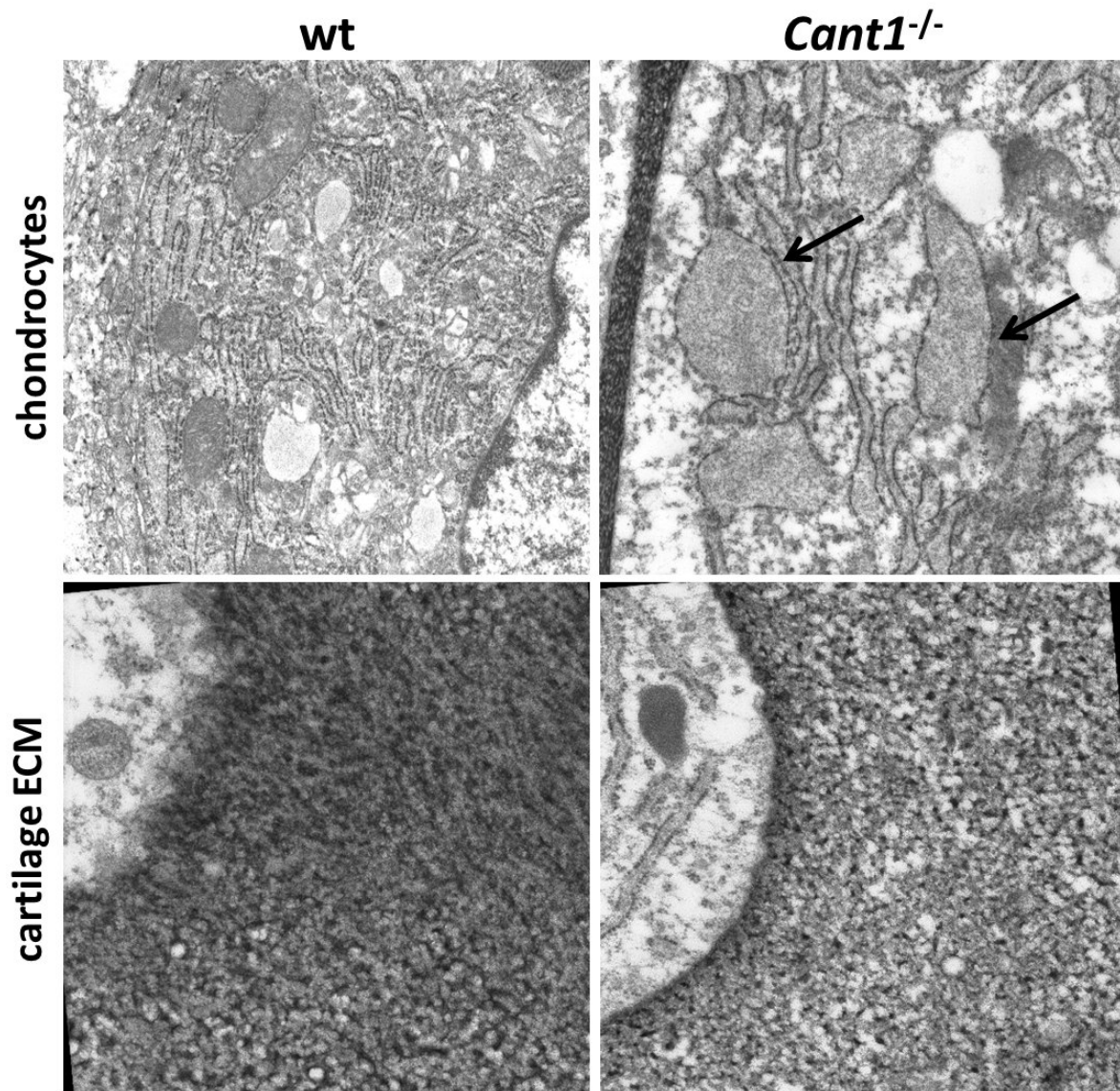
X ray.png

***Cant1*^{-/-} mice showed a delay in the formation of the secondary ossification centre. Proximal tibia sections of P7, P14 and P21 *Cant1*^{-/-} and wild type mice were**

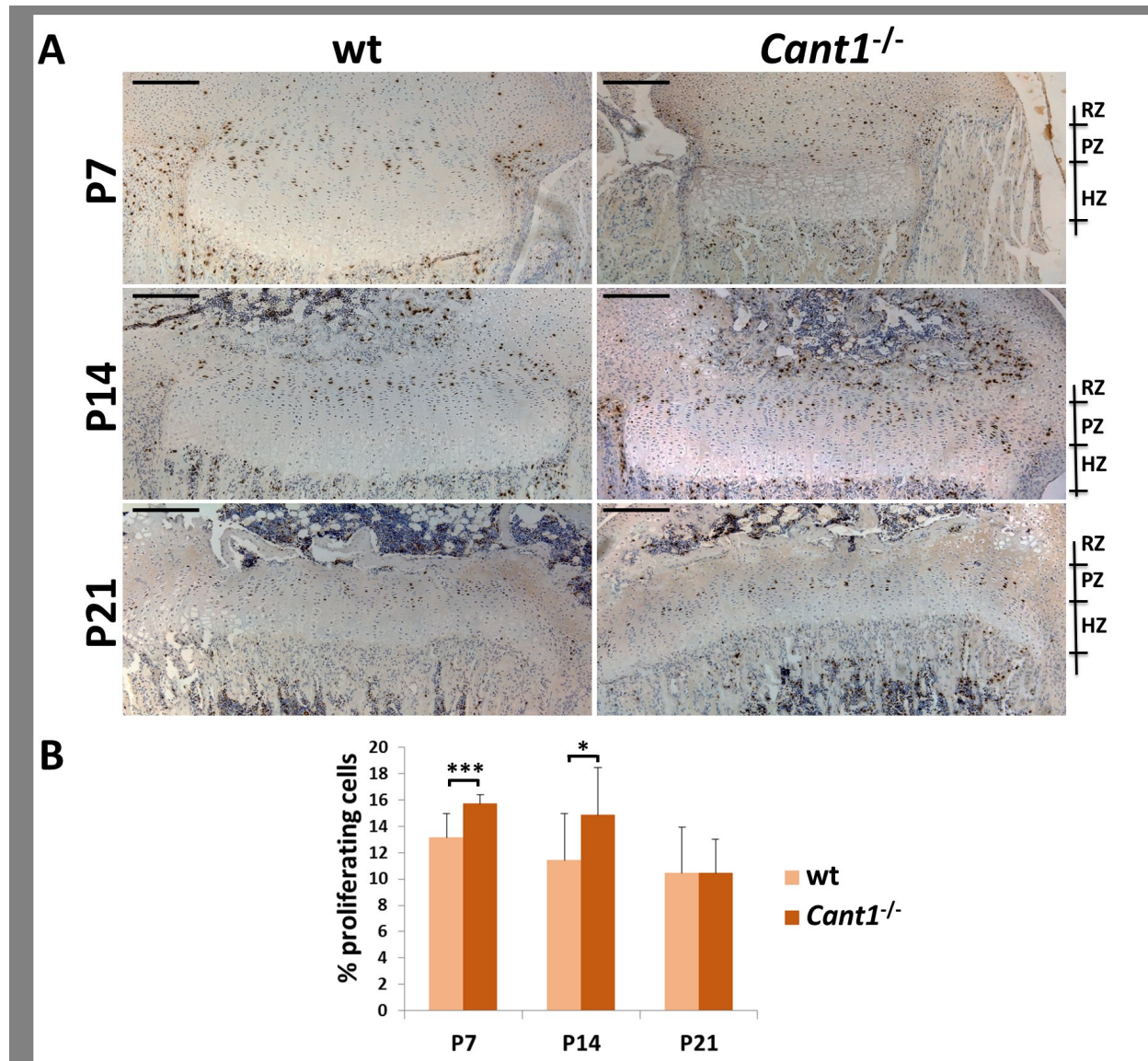
stained with toluidine blue. The overall architecture of *Cant1*^{-/-} growth plate was preserved w



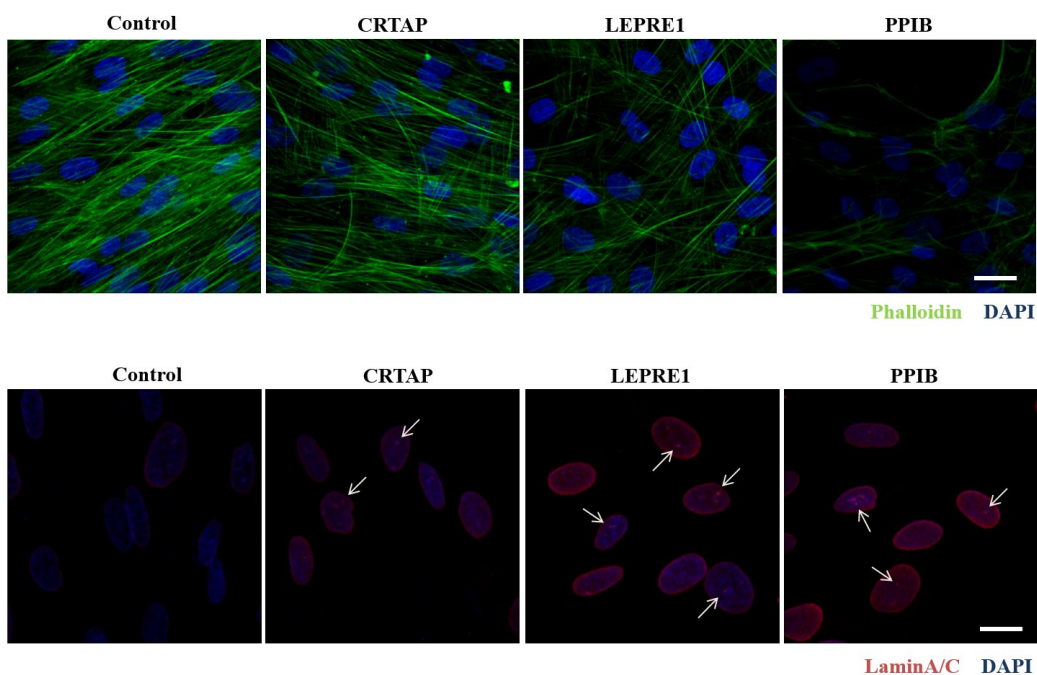
toluidine blue.png



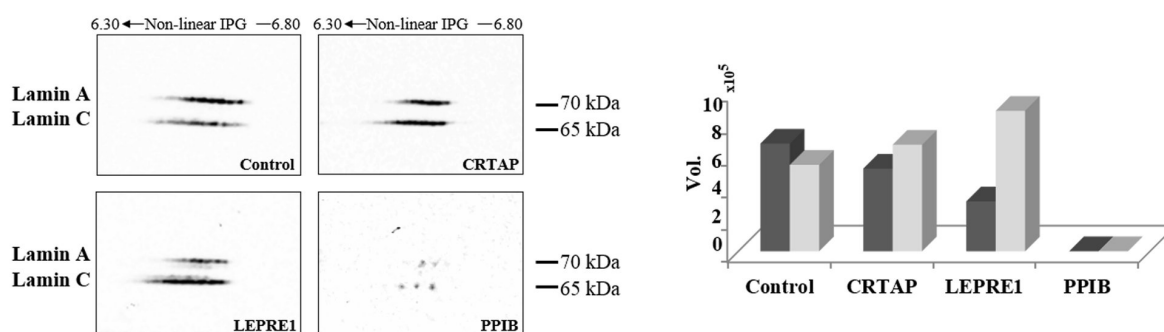
TEM.png



proliferation.png



Recessive 1.png



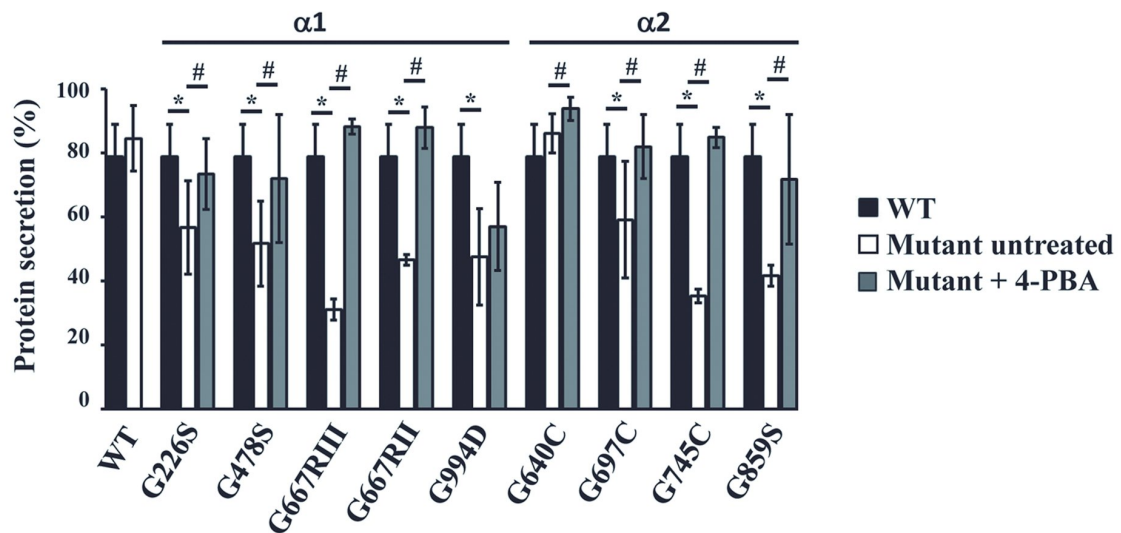
Recessive 2.tif

4-PBA ameliorates OI fibroblasts homeostasis by stimulating protein secretion. After 4-PBA treatment OI patients' fibroblasts were labeled with [35S] EXPRESS35S Protein Labeling Mix and total

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proteins were extracted from medium and cell layer.
Determinati

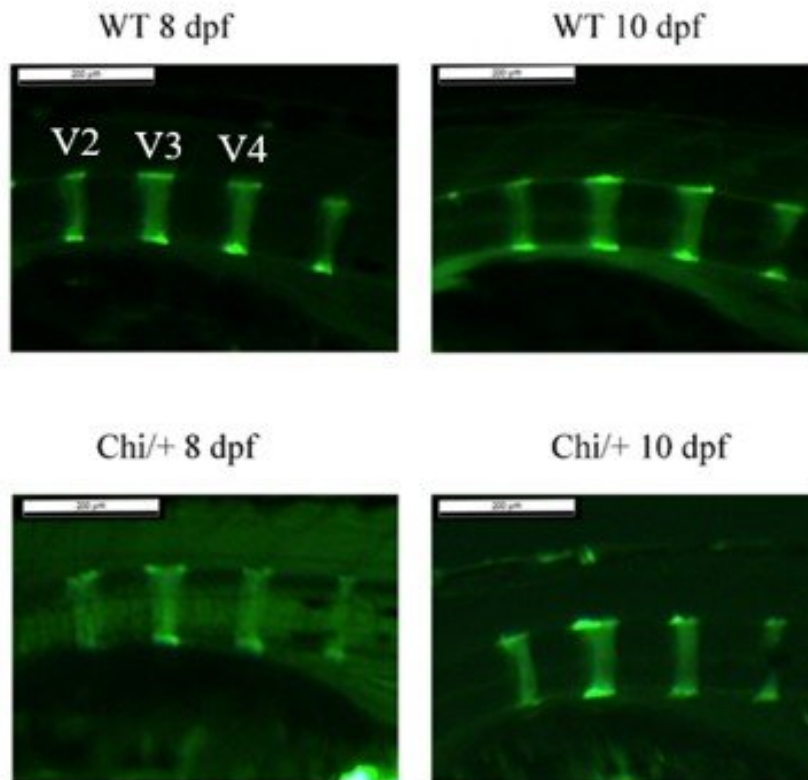


PBA effect 1.tif

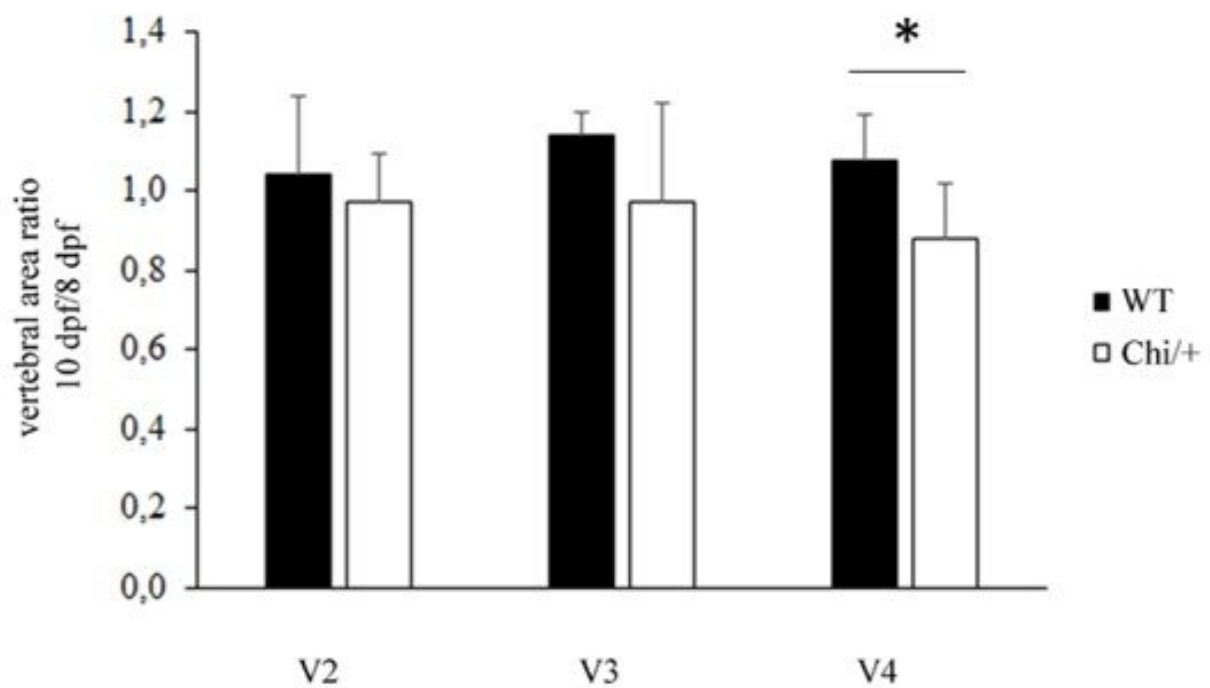
A. Vertebral centra of 8 and 10 dpf WT and Chi/+ larvae stained with calcein. B. Chi/+ show a delay of the bone formation indicated as the ratio between the

area measured at 10 and the area measured at 8 dpf
in both WT and mutant fish. *: $p < 0,05$. (V2, V3

A

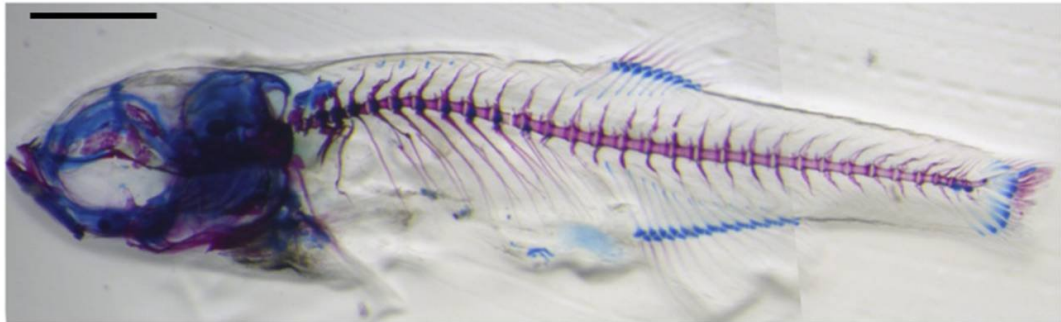


B

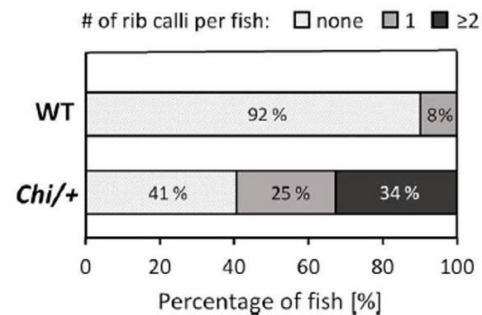
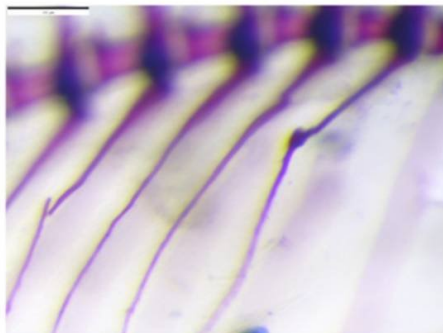


A. Adult mutant *Chi/+* stained with Alcian blue and Alizarin Red. Scale bar 1mm. B. Magnification of a bone callus (red arrowhead) at the level of the ribs. Bone calli in the ribs were observed in 59% of *Chi/+* and in 8% of the WT, suggesting the presence o

A



B



fratture (1).jpg