



D4.9 partner [UNEW]

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All UNEW protocols

Gold standard images of key findings in phenotyping mouse models of skeletal dysplasias (RSD)

This document contains all UNEW protocols together with representative images of the protocol outcomes which have been published on the [Sybil Website](#)

Wild type

V194D matn3



xray matn3.jpg

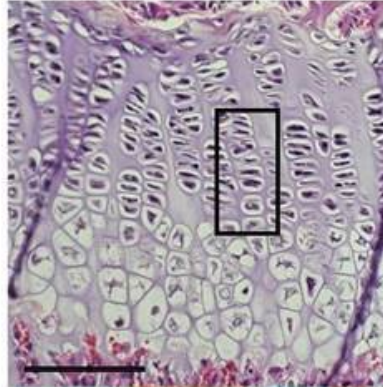
SYBIL Project 602300. The SYBIL Project is funded under the European Commission Seventh Framework Programme, grant number 602300, Theme HEALTH.2013.2.1.1-1



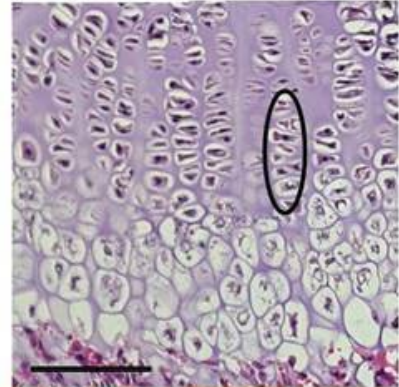
V194D MATN3 Xrays

H&E staining showing disorganised chondrocyte columns at 3 weeks (oval highlight)

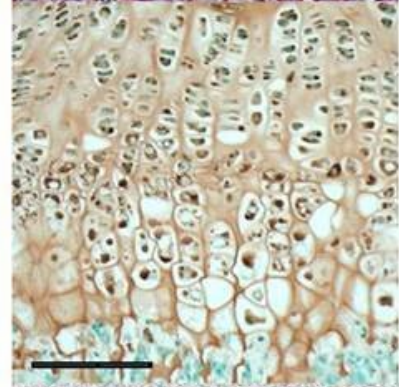
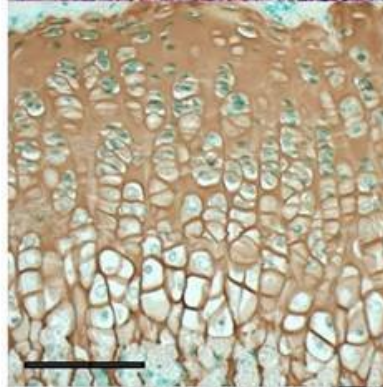
Wild type



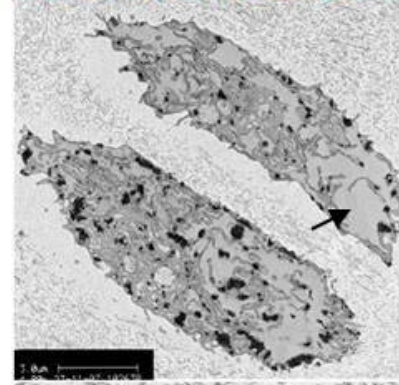
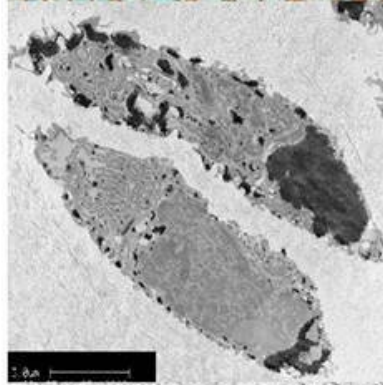
V194D Matn3



Immunohistochemistry for matn3 showing intracellular retention of mutant protein at 3 weeks (brown staining)



Transmission electron microscopy showing retention of mutant protein inside the cell (arrow) at 1 week



Transmission electron microscopy showing abnormal ultrastructure of the mutant extracellular matrix at 1 week

