

## CONT...

## Human Subjects

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- ONL thickness measured at each location by the mean formal ONL thickness at the conceponding retinal location
- Senal data from study eyes (both treated and untreated retinal locations) of RPE65-LCA were similarly processed to derive ONL fraction.
- Visual sensitivity of patients was determined with a modified computerized perimeter (Humphrey Field Analyzer; Zeiss)
- In treated eyes, multiple tests were performed with overlapping dynamic ranges; floor or ceiling values were discarded, and best sensitivities were used.

## <u>Dogs</u>

- Dogs were dark-adapted overnight, premedicated, and anesthetized
- In some case, high-energy flashes were used to evoke photo responses
- Using the en face retinal images and ONL topography maps as guides, eyes were trimmed to section through the treated and untreated regions.
- As control, we used retinal sections processed in the same manner from dogs that were WT or homozygous mutants at the RPE65 locus.



Photoreceptor (ONL) thickness topography in a normal dog compared with two Rpe65-mutant dogs at different ages. ONL thickness topography is mapped to a pseudo color scale. I, inferior; N, nasal; S, superior retina; T, temporal. Reconstituted OCT scans along a superior–inferior meridian (line) are shown in Lower with the ONL layer highlighted in blue. Retinal distances are specified relative to the point closest to the optic nerve. Note that the OCTs shown are reconstituted from ultra-wide angle maps covering more than  $80^\circ \times 80^\circ$  (~ $24 \times 24$  mm) sampled at 1°. (D–F) ONL thickness quantified as a function of age at five retinal locations (shown as squares in A-C): two inferior loci