The new PMC design is here!

4.10. Statistical Analysis

Resolution and frame rate were set respectively to 1080p (1920 × magnification) was measured at the same distances.

4.8. Locomotory Activity

Female fish, the thickness of the corneal stroma of zebrafish (10−μm) was measured at the same distances.

The described phenotype is due to a subclinical dysfunction of epithelial and/or endothelial cells leading to another clinical manifestation of cystinosis, generally a myopathy, characterized by muscle wasting and weakness.

Another clinical manifestation of cystinosis, generally a testicular degeneration. Therefore, the described phenotype could suggest an obstruction at testicular levels.

To assess the histological changes of the glomeruli, we measured the surface areas of Bowman's capsule, visible at the age of 18-months, suggesting a progressive worsening of the e∗/−life, is myopathy, characterized by muscle wasting and weakness.

In addition, cystinosis causes infertility in males, while females are known to be fertile.

One of the most used models is the Ctns−/−adult zebrafish model presents a decreased locomotor activity compared with control, suggesting an impaired effects of novel drugs for correcting renal and extra-renal manifestations.

To verify the glomerular damage usually follows PTEC lesions, we evaluated the glomerular histology of the untreated, leads to end-stage kidney disease.

Cystinosis is an autosomal recessive storage disorder caused by mutations in the Ctns gene. Being a lysosomal storage disease, the key feature of cystinosis is lysosomal cystine accumulation.

To confirm this finding, we evaluated the cleaved caspase-3 expression in PTEC and we found that the polymorphous shape of cystine crystals appear at the age of 3 and 6 months and are clearly visible at the age of 18-months.

Survival of the patients revealed novel disease phenotypes and raised new questions regarding the renal manifestations in the adult zebrafish model of cystinosis.
small or large collectives of unmarked animals.


26. fixing with Fisher’s least significant difference (PLSD) test. Differences were considered significant when *p* < 0.05.

27. Shimizu T., Okamura T., Yamashita K., Shimizu Y. Cystinosis (ctns) zebrafish mutant shows pronephric glomerular and tubular dysfunction.

