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Zebrafish model for cardiac arrhythmia

POPDC1S191F zebrafish mutant

Proposed use

This fish line has been produced by gene editing and exhibits reduced cAMP binding associated with severe consequences. The fish mutant displays cardiac arrhythmia, muscular dystrophy and myofiber rupture and models a mutation found in patients with limb-girdle muscular dystrophy and AV-block.

Technology overview

The Popeye domain containing (Popdc) genes (Popdc1, Popdc2 and Popdc3) encode a family of transmembrane proteins with an evolutionary conserved Popeye domain, which acts as a high affinity cAMP binding domain [1,2]. These genes are abundantly expressed in cardiac myocytes and striated muscle tissue. It has been demonstrated that Popdc1 and Popdc2 in mutant mice [3] and popdc2 in zebrafish are important for striated muscle differentiation and function [4]. In addition these genes are particularly required in the cardiac conduction system.

Benefits

- Displays cardiac arrhythmia, muscular dystrophy and myofiber rupture
- Models mutation found in patients with limb-girdle muscular dystrophy and AV-block
- Exhibits reduced cAMP binding associated with severe consequences

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Technology reference: **7585**

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Intellectual property information

Reagent/material

Link to published paper(s)

Andrée B, Hillemann T, Kessler-Icekson G, Schmitt-John T, Jockusch H, Arnold HH, Brand T. Isolation and characterization of the novel popeye gene family expressed in skeletal muscle and heart. Dev Biol. Jul 15;223(2):371-82. http://www.ncbi.nlm.nih.gov/pubmed/22290329

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