

# **Acute therapy of open ion channel blockers to treat the Slow-Channel Congenital Myasthenic Syndrome.**

Isabelle M. Gonzalez-Montalvo<sup>1</sup>, Vivianette Alicea-Vázquez<sup>1</sup>, José A. Lasalde-Dominicci<sup>1</sup>  
Orestes Quesada<sup>2</sup>

<sup>1</sup>*Department of Biology, University of Puerto Rico, San Juan, Puerto Rico, USA,* <sup>2</sup>*Department of Physical Sciences, University of Puerto Rico, San Juan, Puerto Rico, USA*

The Slow-Channel Congenital Myasthenic Syndrome (SCCMS) is a neuromuscular inherited disorder caused by an abnormality in the muscular acetylcholine receptors (AChR). The mutation has been linked to the prolonged opening in the AChR, thus, causing an overflow of ions to the muscles. The voltage dependent properties of the AChR allows the study of the ion-channel's openings with the usage of the two-electrode voltage clamp electrophysiology technique (TEVC) for *in vitro* studies of SCCMS. Therefore, the aim of the present study is to test the effectivity of drugs with open ion-channel blockers abilities, such as fluoxetine hydrochloride, quinidine sulfate, memantine hydrochloride, ketanserine and amitriptyline, in a transgenic mice line bearing the  $\delta$ S268F mutation. Previous studies shows that fluoxetine hydrochloride and quinidine sulfate have proven to diminish the openings of AChR and currently is the treatment used in patients that have this syndrome.