

Partial Reversal of Respiratory Dysfunction Shown with Experimental Drugs

One of the more serious consequences of Rett syndrome is the intermittent episodes of breath holding, which can put individuals at risk for brain damage due to oxygen deprivation. In a study published last month in the Journal, PNAS, an international team of researchers based in the U.S. and UK revealed that they were able to halt the potentially lethal, breath holding episodes associated with Rett syndrome.

The team led by Professor John Bissonnette, M.D., of Oregon Health and Science University, Portland OR and Professor Julian FR Paton, PhD, at the University of Bristol in the UK discovered a way to prevent the frequent episodes of breath holding in a mouse model of Rett syndrome using a unique combination of drugs. Earlier work found that an area of the brain that allows breathing to persist throughout life, without interruption, has reduced levels of a vital transmitter substance called gamma-aminobutyric acid (GABA). The researchers took a two-pronged approach, using one set of drugs to increase the amount of GABA, and others to target a specific type of serotonin receptor to reduce activity in brain cells that normally depress inhalation. Both of these approaches halted the life threatening episodes of breathing arrests in Rett syndrome mice and confirmed the investigators' initial theory.

Dr. Bissonnette, co-principal investigator on the IRSF funded study commented "When the phrenic nerve going to the diaphragm is silent, nerves going to muscles for expiration are excessively active. Building on our earlier studies that showed a defect in inhibition within the brain's respiratory areas, we reasoned that expiratory neurons were not receiving enough inhibition. When we boosted inhibition, or separately used a drug known to silence expiratory neurons, the pattern of breath holding was markedly improved." "While the specific drugs used in this mouse study are not available for human use, drugs with similar modes of action have been used in other conditions". Dr. Bissonnette added.

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