Technology Offer

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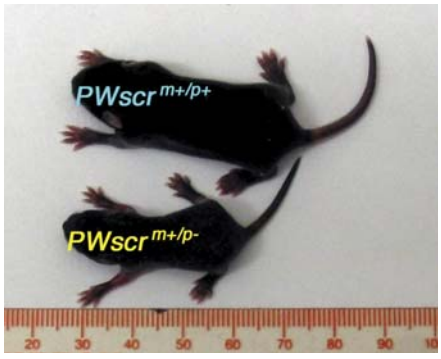
Mouse model for the Prader-Willi syndrome (PWS) with a deletion of MBII-85del+/-

Introduction

Prader-Willi syndrome, or PWS, is a complex neurogenetic disorder and the most common genetic cause of life-threatening childhood obesity. Newborns have poor muscle tone, making suckling difficult, which leads to poor weight gain. After infancy, they experience extreme hunger, leading to obesity. Other symptoms include short stature, mental retardation, and often infertility. In PWS patients, a complex set of genes on the paternal chromosome 15 (in the PWS region) is missing or unexpressed.

The Mouse Model

In an attempt to understand this disorder, various protein-coding genes in this region have been deleted in mice, but none of the resulting phenotypes consistently correlated with the human disease. This region also contains a cluster of genes that encode functional non-protein-coding RNAs. We deleted specifically the MBII-85 small nucleolar RNA (snoRNA) gene cluster on the parental mouse chromosome, which did not affect expression of any of the other snoRNA or protein-coding genes in the PWS region. These mice consistently displayed postnatal growth retardation starting from day 5 to 6, low postnatal lethality only in certain genetic backgrounds (<15%), and no adolescent obesity. Thus, this mouse model, with the deletion of a small, brain-specific non-protein-coding RNA, should prove useful for teasing out the various molecular pathologies of PWS.



Growth Differences among PWScr$^{m+/p-}$ and PWScr$^{m+/p+}$ Siblings

Further Reading

Skryabin et al., PLoS Genet., 2007, 3(12): e235: 2529-2539

Für weitere Informationen wenden Sie sich bitte an:

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