This week in techniques

Approach	Summary	Licensing status	Publication and contact information
Disease models			
Mouse model of methylmalonic acidemia-associated renal pathology	<p>A mouse model of methylmalonic acidemia-associated renal pathology could help identify new treatments for the condition, which is a metabolic disorder caused primarily by mutations in methylmalonyl CoA mutase (MUT). In this model, <i>Mut</i> knockout mice were engineered to express a liver-specific <i>Mut</i> transgene. The mice showed severe proximal tubular mitochondrial changes that recapitulate the renal pathology of methylmalonic acidemia seen in patient biopsy samples. In the mouse model, dietary supplementation with the antioxidants ubiquinone and vitamin E decreased weight loss compared with no supplementation and improved markers of renal function. Next steps include evaluating additional antioxidant compounds in the mice and then conducting a clinical trial in patients who have methylmalonic acidemia to evaluate the most effective compound.</p> <p>SciBX 6(33); doi:10.1038/scibx.2013.907 Published online Aug. 29, 2013</p>	<p>Model unpatented; available for licensing from the NIH Contact: Tara Kirby, National Institutes of Health, Bethesda, Md. e-mail: kirbyt@mail.nih.gov</p>	<p>Manoli, I. <i>et al. Proc. Natl. Acad. Sci. USA</i>; published online July 29, 2013; doi:10.1073/pnas.1302764110 Contact: Charles P. Venditti, National Institutes of Health, Bethesda, Md. e-mail: venditti@mail.nih.gov</p>