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This week in techniques

Approach	Summary	Licensing status	Publication and contact information
Disease models			
Fibrillin 1 (Fbn1)-mutant mouse models of systemic scleroderma	Mice harboring mutations in the integrin-binding domain of Fbn1 could help model systemic scleroderma. The mice exhibited multiple disease features seen in patients with systemic scleroderma or patient fibroblasts, including high levels of collagen, activated integrin β_3 (GPIIIa; CD61) in the skin and high levels of antinuclear antibodies in circulation. Compared with control IgG, an integrin β_1 (CD29)-activating antibody decreased CD61 activity and collagen expression in patient fibroblasts, skin fibrosis and levels of circulating antinuclear antibodies in the mice. Future studies could include using the models to compare the efficacy and safety of CD29-activating and CD61-inhibiting antibodies. Mitsubishi Tanabe Pharma Corp.'s Venoglobulin IH (GB-0998), a liquid human IgG preparation derived from donated plasma, is in Phase III testing to treat systemic scleroderma. arGentis Pharmaceuticals LLC's ARG201, a solubilized type I native bovine collagen, is in Phase II testing to treat systemic scleroderma.	Patent and licensing status unavailable	Gerber, E.E. *et al. Nature*; published online Oct. 9, 2013; doi:10.1038/nature12614 **Contact:** Harry C. Dietz, The Johns Hopkins University School of Medicine, Baltimore, Md. e-mail: hdietz@jhmi.edu

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