Synopsis of Original Research Paper

Establishment of animal models for piebaldism by the regulation of melanocyte stem cells

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Piebaldism is an autosomal dominant genetic pigmentary disorder, characterized by congenital white hair and patches located on the forehead, anterior trunk, and extremities. Most piebald patients have a mutation of the *Kit* gene, which encodes a tyrosine kinase receptor involved in pigment cell development. The white hair and patches of such patients are already completely formed at birth and do not usually expand thereafter. This stability of pigmented spots also applies to Kit^{W} and $Kitl^{Sl}$ mutant mice. However, 2 novel cases of piebaldism were reported in 2001, in which both mother and daughter having a novel Val620Ala mutation in their *Kit* gene showed progressive depigmentation. To prepare an animal model of this mutation to explore undefined functions of KIT signaling for maintaining pigmented melanocytes in the skin or more specifically the integrity of the melanocyte stem cell system in the postnatal skin, we produced transgenic mice expressing Val620Ala Kit. These mice well mimicked the white spotting pattern of patients; however, no change in this pattern was observed after birth, even after increasing the transgene expression by various means. Here, we report the unexpectedly extremely stable maintenance of the melanocyte stem cell system under stringent conditions for KIT signaling.