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Craniofacial deformity and malocclusion in a mouse model of Neurofibromatosis type 1 (NF1)

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Neurofibromatosis type I (NF 1) is an autosomal dominant disorder affecting approximately 1 in 3500 individuals. NF1 exhibits multiple manifestations such as the presence of café-au-late spots, learning disabilities and bone deformities. A large proportion of NF 1 patients display skeletal deformities including alteration in bone size and shape, the presence of scoliosis, and tendency to develop pseudoarthroses. Although the skeletal manifestations of NF1 have long been recognized and studied but only recently recognized as skeletal dysplasia by bone researchers. Craniofacial abnormality occurs in about 7% of NF1 patients and characterized by hypoplasia or absence of greater wing of sphenoid bone. This dysplasia is progressive and always unilateral, results in bulging of one eye and mid-facial bone associated with malocclusion, and is termed Sphenoid Wing Dysplasia (SWD). We have established a breeding colony of neurofibromin (NF1 gene) osteoblast conditional knockout mice. Preliminary result indicate that the NF1ob-/- mice present with cranial asymmetry with eye bulging and malocclusion as the animal age. Micro-CT of these animals shows a progressive (12-24 weeks) loss of craniofacial symmetry at the sphenoid bone and other cranial bones. This phenotype is strikingly similar to SWD seen in NF1 patients. We propose to eliminate the possibility of tumor as a cause of facial deformity, by histological examination of numerous specimens and there by establish an osteoblast origin for the phenotype, to characterize the time course of the progression of craniofacial defect and to identify metrics to follow the progression of the phenotype, and to test the effect of treatment with diet, PTH and Ras antagonist on the development and progression of the phenotype. There are currently no treatment known that block or slow the progression of sphenoid wing dysplasia in NF1. Our ultimate goal is to identify the original cause of sphenoid wing dysplasia in NF1 and try to block or prevent such deformity, which will help the NF1 patients not to develop malocclusion and facial asymmetry.

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