

The Kir2.1 potassium channel is required for palate closure, jaw development, and craniofacial bone development

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Mutations that disrupt an inwardly rectifying K⁺ channel, Kir2.1, cause dominantly inherited Andersen-Tawil Syndrome (ATS). Symptoms of ATS include cardiac arrhythmia, periodic paralysis, cognitive deficits, and morphological abnormalities in craniofacial and limb development. Although the known role of Kir2.1 in muscle and neurons may explain the phenotypes in muscle and brain, it is less clear how loss of this ion channel could affect morphogenesis. We show that deletion of Kir2.1 causes cleft palate, reduction of the size of the jaw, loss of craniofacial bone, and digit defects. Deletion of Kir2.1 drastically reduces proliferation of the cells in the palate shelf. Conditional deletion of Kir2.1 reveals that Kir2.1 is required in the cranial neural crest cells that make up the mesenchyme of the palate shelves, and not in the overlying ectoderm for palate closure.