

MOLECULAR NETWORKS UNDERLYING DENTAL DEFECTS IN THE DIGEORGE SYNDROME

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INTRODUCTION & DISCUSSION:

TBX1 is a key player in the aetiology of the DiGeorge/Velo-cardio-facial syndrome (DGS/VCFS), which is a complex developmental disorder associated with a variety of abnormalities, including facial dysmorphism, submucous cleft palate and incisor hypoplasia or aplasia. Targeted disruption of *Tbx1* in mice results in facial abnormalities that are very similar to those presented in the human DGS. We investigate the role of *Tbx1* in normal tooth development and analyse the consequences of *Tbx1* deletion on the formation of teeth. *Tbx1* expression is restricted to the epithelial component of tooth primordia and appears to mark the epithelial cells destined to give rise to the enamel matrix producing ameloblasts. Using *Tbx1*^{-/-} mice we have identify genes whose expression is altered in the absence of *Tbx1* in dental structures, as compared to the wild type mice. In addition, some of the *Tbx1* activities may be mediated through the control of FGF expression in the developing teeth. Regulators and targets of *Tbx1* should unveil genetic networks that are affected in tooth developmental disorders.

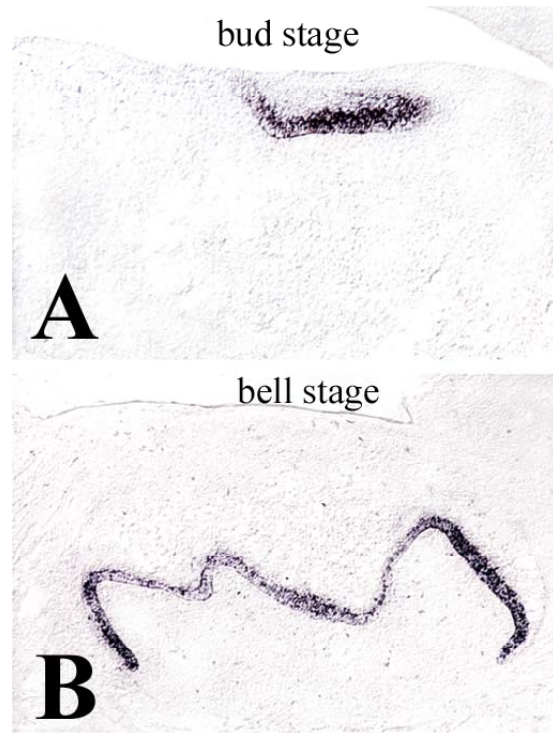


Figure: Expression of Tbx1 in the epithelium of the developing tooth during the bud (A) and bell (B) stages.

REFERENCE: M. Zoupa, M. Seppala, T. Mitsiadis, M. Cobourne (2006). *Int. J. Dev. Biol.* 50: 504-510.