

**Supplementary Table 1:** Comparison of Lung Phenotypes in *Tbx4cKO* Mice and Human TBX4 Syndrome

<b>Feature</b>	<b>Mouse Model (<i>Tbx4cKO</i>)</b>	<b>Human TBX4 Syndrome</b>
<b>Hemodynamics</b>		
Pulmonary Hypertension	Present	Present
PH Age of Onset	Adult	Variable (neonatal to geriatric)
<b>Vasculature</b>		
Medial Thickening	Present	Present
Neointimal Lesions	Variable	Variable
Distal Muscularization	Present	Present
Muscularized vessels near pleura	Present	Present
PA Branching Pattern	Elaborated	Not reported
<b>Development</b>		
Lobe Volume	Reduced at E18.5	Hypoplastic
Distal Lung Development	Underdeveloped distal airspaces reported <sup>38</sup>	Maldeveloped acini or abnormal alveolar growth
<b>Airways</b>		
Distal Muscularization	Present	Present
SMC Ring Spacing	Widened	Not specifically characterized
<b>Abnormal Mesenchymal Features</b>		
Ectopic Interstitial "Myofibroblasts"	Present	Present
Subpleural SMC Bands	Present	Present
Heterologous Bone Formation	Not seen	Variable