

Table S1. Loss-of-function phenotypes for signaling pathway genes

Gene	Organ	Species	Phenotype	References
<b>Fgf</b>				
<i>Fgf7</i>	Kidney	Mouse	-/- Hypoplasia	(Qiao et al., 1999)
<i>Fgf8</i>	Kidney	Mouse	-/- Hypoplasia, dysplasia	(Perantoni et al., 2005)
	SMG <sup>a</sup>	Mouse	-/- Aplasia	(Jaskoll et al., 2004b)
<i>Fgf9</i>	Kidney	Mouse	-/- No phenotype	(Colvin et al., 2001a)
	Lung	Mouse	-/- Hypoplasia	(Colvin et al., 2001b)
<i>Fgf10</i>	Kidney	Mouse	-/- Dysplasia	(Ohuchi et al., 2000)
	Lacrimal	Human	+/- Aplasia, hypoplasia <sup>b</sup>	(Entesarian et al., 2005)
	Lacrimal	Mouse	+/- Agenesis	(Entesarian et al., 2005)
	Lung	Mouse	-/- Aplasia	(Min et al., 1998)
	Mammary	Mouse	-/- Dysplasia	(Mailleux et al., 2002)
	Pancreas	Mouse	-/- Hypoplasia	(Bhushan et al., 2001)
	Salivary <sup>b</sup>	Human	+/- Aplasia, hypoplasia <sup>c</sup>	(Entesarian et al., 2005)
	SMG	Mouse	+/- Aplasia, hypoplasia	(Entesarian et al., 2005)
	SMG	Mouse	-/- Agenesis	(Ohuchi et al., 2000)
	Trachea	Mouse	-/- Aplasia	(Min et al., 1998)
<i>Fgf20</i>	Kidney	Human	-/- Agenesis	(Barak et al., 2012)
<i>Fgf9; Fgf20</i>	Kidney	Mouse	<i>Fgf9</i> <sup>-/-</sup> ; <i>Fgf20</i> <sup>+/-</sup> No phenotype	(Barak et al., 2012)
		Mouse	<i>Fgf9</i> <sup>-/-</sup> ; <i>Fgf20</i> <sup>+/-</sup> No phenotype	(Barak et al., 2012)
		Mouse	<i>Fgf9</i> <sup>+/-</sup> ; <i>Fgf20</i> <sup>-/-</sup> Hypoplasia	(Barak et al., 2012)
		Mouse	<i>Fgf9</i> <sup>-/-</sup> ; <i>Fgf20</i> <sup>-/-</sup> Agenesis	(Barak et al., 2012)
<i>Fgfr1; Fgfr2</i>	Kidney	Mouse	<i>Fgfr1</i> <sup>-/-</sup> ; <i>Fgfr2</i> <sup>-/-</sup> Aplasia	(Poladia et al., 2006)
<i>Fgfr1; Fgfr2llc</i>	Kidney	Mouse	<i>Fgfr1</i> <sup>-/-</sup> ; <i>Fgfr2llc</i> <sup>-/-</sup> Dysplasia, hypoplasia	(Sims-Lucas et al., 2011)
<i>Fgfr2</i>	Kidney	Human	-/- Dysplasia <sup>d</sup>	(Reardon et al., 1994)
	Lung	Mouse	-/- Agenesis	(De Moerlooze et al., 2000)
	Mammary	Mouse	-/- Dysplasia	(Mailleux et al., 2002)
	SMG	Mouse	-/- Agenesis	(De Moerlooze et al., 2000)
<i>Fgfr3; Fgfr4</i>	Lung	Mouse	<i>Fgfr3</i> <sup>-/-</sup> ; <i>Fgfr4</i> <sup>-/-</sup> Dysplasia	(Weinstein et al., 1998)
<i>Fgfr4</i>	Lung	Mouse	-/- No phenotype	(Weinstein et al., 1998)
<i>Bnl</i>	Trachea	<i>Drosophila</i>	-/- Dysplasia (branching)	(Sutherland et al., 1996)
<i>Btl</i>	Trachea	<i>Drosophila</i>	-/- Impaired cell migration and branching	(Klämbt et al., 1992)
<i>Spry1</i>	Kidney	Mouse	-/- Cystic hyperplasia	(Basson et al., 2006)
<i>Spry1; Spry2</i>	Lung	Mouse	<i>Spry1</i> <sup>-/-</sup> ; <i>Spry2</i> <sup>-/-</sup> Dysplasia (airway)	(Tang et al., 2011)
	SMG	Mouse	<i>Spry1</i> <sup>-/-</sup> ; <i>Spry2</i> <sup>-/-</sup> Dysplasia, hypoplasia	(Knosp et al., 2015)
<b>Egf</b>				
<i>Egfr</i>	Lung	Mouse	-/- Dysplasia	(Miettinen et al., 1997)
	Mammary	Mouse	-/- Dysplasia	(Wiesen et al., 1999)
	Pancreas	Mouse	-/- Dysplasia	(Miettinen et al., 2000)
	SMG	Mouse	-/- Dysplasia	(Jaskoll and Melnick, 1999)
<i>Hb-egf</i>	Lung	Mouse	-/- Dysplasia, hypoplasia	(Jackson et al., 2003)
<b>Tgf</b>				
<i>Tgf-β2</i>	SMG	Mouse	-/- No phenotype	(Jaskoll and Melnick, 1999)
<i>Tgf-β3</i>	SMG	Mouse	-/- No phenotype	(Jaskoll and Melnick, 1999)
<i>TβRII</i>	Lung	Mouse	-/- Dysplasia	(Li et al., 2008)
<i>Lefty1</i>	Lung	Mouse	-/- Left pulmonary isomerism	(Meno et al., 1998)
<b>Gdnf</b>				
<i>Gdnf</i>	Kidney	Mouse	-/- Agenesis	(Pichel et al., 1996)
<i>GFRα1</i>	Kidney	Mouse	-/- Agenesis	(Enomoto et al., 1998)
<i>Ret</i>	Kidney	Mouse	-/- Agenesis, dysplasia	(Schuchardt et al., 1994)
<b>Wnt</b>				
<i>Wnt4</i>	Kidney	Mouse	-/- Dysplasia, hypoplasia	(Stark et al., 1994)
	Lung	Mouse	-/- Hypoplasia	(Caprioli et al., 2015)
	Trachea	Mouse	-/- Dysplasia	(Caprioli et al., 2015)
<i>Wnt5a</i>	Kidney	Mouse	-/- Dysplasia	(Pietilä et al., 2016)
<i>Wnt7b</i>	Kidney	Mouse	-/- Deficient medulla	(Yu et al., 2009)
<i>Wnt9b</i>	Kidney	Mouse	-/- Dysplasia (branching), hypoplasia	(Carroll et al., 2005)
<i>Wnt11</i>	Kidney	Mouse	-/- Mild hypoplasia, dysplasia	(Majumdar et al., 2003)
<i>Wnt11; Ret</i>	Kidney	Mouse	<i>Wnt11</i> <sup>+/-</sup> ; <i>Ret</i> <sup>-/-</sup> Moderate hypoplasia	(Majumdar et al., 2003)
		Mouse	<i>Wnt11</i> <sup>+/-</sup> ; <i>Ret</i> <sup>-/-</sup> Severe hypoplasia	(Majumdar et al., 2003)

Table S1. Loss-of-function phenotypes for signaling pathway genes (Continued)

Gene	Organ	Species	Phenotype	References
<b>Bmp</b>				
<i>Bmp4</i>	Kidney	Mouse	+/- Dysplasia, hypoplasia	(Miyazaki et al., 2000)
	Lung	Mouse	-/- Dysplasia	(Eblaghie et al., 2006)
<i>Bmp7</i>	Kidney	Mouse	-/- Dysplasia	(Dudley et al., 1995)
<i>Bmpr1a</i>	Lung	Mouse	-/- Dysplasia	(Eblaghie et al., 2006)
<i>Dpp</i>	Trachea	<i>Drosophila</i>	-/- Impaired cell migration and branching	(Vincent et al., 1997)
<b>Pdgf</b>				
<i>Pdgf-a</i>	Lung	Mouse	-/- Dysplasia (alveogenesis)	(Boström et al., 2002)
<i>Pdgfar</i>	Lung	Mouse	-/- Hypoplasia	(Boström et al., 2002)
<b>Eda</b>				
<i>Eda</i>	-	Human	+/- or -/- Ectodermal dysplasia <sup>a</sup>	(Kere et al., 1996)
<i>Edar<sup>dl</sup></i>	-	Human	+/- or -/- Ectodermal dysplasia	(Monreal et al., 1999)
	SMG	Mouse	-/- Dysplasia	(Jaskoll et al., 2003)
<i>Eda<sup>ta</sup></i>	SMG	Mouse	-/- Hypoplasia	(Jaskoll et al., 2003)
<b>Others</b>				
<i>Adam17 (Tace)</i>	Lung	Mouse	-/- Hypoplasia	(Zhao et al., 2001)
	Mammary	Mouse	-/- Dysplasia (branching)	(Sternlicht et al., 2005)
<i>C-met (Hgf)</i>	Lung	Mouse	-/- Dysplasia	(Yamamoto et al., 2007)
<i>Dll</i>	Vascular	Mouse	+/- Aplasia (sprouting and branching)	(Napp et al., 2012)
<i>Dnm2</i>	Vascular	Mouse	-/- Dysplasia	(Lee et al., 2014)
<i>Igf-Ir</i>	Mammary	Mouse	-/- Hypoplasia	(Bonnette and Hadsell, 2001)
<i>Irs-1/2</i>	Mammary	Mouse	-/- Hypoplasia	(Heckman et al., 2007)
<i>P120</i>	SMG	Mouse	-/- Dysplasia	(Davis and Reynolds, 2006)
<i>P190-B</i>	Mammary	Mouse	-/- Hypoplasia	(Heckman et al., 2007)
<i>Shh</i>	Lung	Mouse	-/- Aplasia, dysplasia	(Pepicelli et al., 1998)
	Pancreas	Mouse	-/- Hyperplasia	(Hebrok et al., 2000)
	SMG	Mouse	-/- Aplasia, dysplasia	(Jaskoll et al., 2004a)
<i>Vegf-A</i>	Lung	Mouse	-/- Dysplasia	(Yamamoto et al., 2007)

The list of signaling pathway genes may not be complete. +/+, wild type; +/-, heterozygous; -/-, homozygous. Agenesis is defined here based on the published study as a failure of organ formation, aplasia as incomplete development, dysplasia as abnormal development, hyperplasia as increased size, and hypoplasia as underdevelopment. *Bmp*, bone morphogenetic protein; *Bnl*, branchless; *Btl*, breathless; *Dll*, delta-like ligand; *Dnm*, dynamin; *Dpp*, decapentaplegic; *Eda*, ectodysplasin A; *Edar<sup>dl</sup>*, ectodysplasin-A receptor, downless; *Fgf*, fibroblast growth factor; *Gdnf*, glial cell line-derived neurotrophic factor; *Hb-egf*, heparin-binding epidermal growth factor; *Hgf*, hepatocyte growth factor; *Hgfr*, hepatocyte growth factor receptor; *Igf*, insulin-like growth factor; *Irs*, insulin receptor substrate; *Pdgf*, platelet-derived growth factor; *Pdgfr*, platelet-derived growth factor receptor; *Shh*, sonic hedgehog; *Spry*, sprouty; *Tace*, tumor necrosis factor- $\alpha$ -converting enzyme; *Tgf*, transforming growth factor; *Vegf*, vascular endothelial growth factor.

<sup>a</sup>SMG, submandibular salivary gland.

<sup>b</sup>Salivary, major salivary glands.

<sup>c</sup>In aplasia of lacrimal and salivary glands (ALSG).

<sup>d</sup>In some cases, renal dysplasia is observed.

<sup>e</sup>Hypohidrotic ectodermal dysplasia is characterized by dysplasia of hair, teeth, and several exocrine glands.

Table S2. Loss-of-function phenotypes for transcription factors

Gene	Organ	Species	Phenotype	References
<b>Homeobox</b>				
<i>Hoxa11 Hoxd11</i>	Kidney	Mouse	-/- No phenotype	(Patterson et al., 2001)
	Kidney	Mouse	-/- No phenotype	(Patterson et al., 2001)
<i>Hoxa11; Hoxd11</i>	Kidney	Mouse	<i>Hoxa11</i> <sup>-/-</sup> ; <i>Hoxd11</i> <sup>-/-</sup> Agenesis, aplasia	(Patterson et al., 2001)
<i>Pax2</i>	Kidney	Human	+/- Hypoplasia, renal impairment	(Sanyanusin et al., 1995)
	Kidney	Mouse	-/- Aplasia (no ureteric bud outgrowth)	(Brophy et al., 2001)
<i>Pdx1</i>	Pancreas	Mouse	-/- Aplasia (rudimentary bud), dysplasia	(Offield et al., 1996)
<i>Nkx2.1</i>	Lung	Mouse	-/- Hypoplasia, dysplasia	(Minoo et al., 1999)
<i>Nkx2.3</i>	Sublingual	Mouse	-/- Dysplasia	(Biben et al., 2002)
<i>Nkx3.1</i>	Prostate	Mouse	+/- Mild hypoplasia and dysplasia	(Bhatia-Gaur et al., 1999)
	Prostate	Mouse	-/- Hypoplasia, dysplasia	(Bhatia-Gaur et al., 1999)
	Prostate	Mouse	-/- Hypoplasia, dysplasia (duct)	(Schneider et al., 2000)
	Minor SG <sup>a</sup>	Mouse	-/- Hypoplasia, dysplasia (duct)	(Schneider et al., 2000)
<b>Ets</b>				
<i>Etv4</i>	Kidney	Mouse	-/- Occasional agenesis	(Lu et al., 2009)
<i>Etv4; Etv5</i>	Kidney	Mouse	<i>Etv4</i> <sup>+/-</sup> ; <i>Etv5</i> <sup>+/-</sup> Occasional agenesis	(Lu et al., 2009)
		Mouse	<i>Etv4</i> <sup>-/-</sup> ; <i>Etv5</i> <sup>-/-</sup> Dysplasia	(Lu et al., 2009)
		Mouse	<i>Etv4</i> <sup>-/-</sup> ; <i>Etv5</i> <sup>-/-</sup> Agenesis, severe hypoplasia and dysplasia	(Lu et al., 2009)
<b>Fox</b>				
<i>Foxa1</i>	Lung	Mouse	-/- Delayed development, no postnatal phenotype	(Besnard et al., 2005)
<i>Foxa2</i>	Lung	Mouse	-/- Dysplasia, goblet cell hyperplasia	(Wan et al., 2004)
<i>Foxa1; Foxa2</i>	Lung	Mouse	<i>Foxa1</i> <sup>-/-</sup> ; <i>Foxa2</i> <sup>-/-</sup> Dysplasia, hypoplasia	(Wan et al., 2005)
<b>Gata</b>				
<i>Gata3</i>	Kidney	Human	+/- Hypoplasia, dysplasia, aplasia <sup>b</sup>	(Ali et al., 2007)
		Mouse	-/- Dysplasia	(Grote et al., 2008)
<i>Gata4</i>	Pancreas	Human	+/- Agenesis <sup>c</sup>	(Shaw-Smith et al., 2014)
	Pancreas	Mouse	-/- Agenesis (ventral pancreas)	(Watt et al., 2007)
	Pancreas	Mouse	-/- Mild dysplasia	(Xuan et al., 2012)
<i>Gata5</i>	Kidney	Mouse	-/- No phenotype	(Molkentin et al., 2000)
	Lung	Mouse	-/- No phenotype	(Molkentin et al., 2000)
<i>Gata6</i>	Pancreas	Human	+/- Agenesis	(Lango Allen et al., 2011)
	Pancreas	Mouse	-/- Mild dysplasia	(Xuan et al., 2012)
<i>Gata4; Gata6</i>	Pancreas	Mouse	<i>Gata4</i> <sup>-/-</sup> ; <i>Gata6</i> <sup>-/-</sup> Agenesis	(Xuan et al., 2012)
<b>Myc</b>				
<i>N-Myc</i>	Lung	Mouse	-/- Hypoplasia	(Moens et al., 1992)
<i>c-Myc</i>	Vascular	Mouse	-/- Aplasia	(Baudino et al., 2002)
<b>Pas</b>				
<i>Npas3</i>	Lung	Mouse	-/- Dysplasia (branching)	(Zhou et al., 2009)
<i>Trh</i>	Trachea	<i>Drosophila</i>	-/- Aplasia (no tubulogenesis)	(Isaac and Andrew, 1996)
	SMG	<i>Drosophila</i>	-/- Aplasia (no tubulogenesis)	(Isaac and Andrew, 1996)
<b>Sox</b>				
<i>Sox4</i>	Pancreas	Mouse	-/- Dysplasia	(Wilson et al., 2005)
<i>Sox8; Sox9</i>	Kidney	Mouse	<i>Sox8</i> <sup>-/-</sup> ; <i>Sox9</i> <sup>+/-</sup> Hypoplasia	(Reginensi et al., 2011)
		Mouse	<i>Sox8</i> <sup>-/-</sup> ; <i>Sox9</i> <sup>-/-</sup> Almost agenesis	(Reginensi et al., 2011)
<i>Sox9</i>	Kidney	Human	+/- Renal malrotation, nephrocalcinosis	(Cost et al., 2009)
	Lung	Mouse	+/- Dysplasia, hypoplasia <sup>d</sup>	(Foster et al., 1994)
	Lung	Mouse	-/- ( <i>Shh</i> <sup>Cre</sup> ) Hypoplasia, dysplasia	(Chang et al., 2013)
<i>Sox11</i>	Pancreas	Mouse	-/- Hypoplasia	(Seymour et al., 2007)
	Lung	Mouse	-/- Hypoplasia	(Sock et al., 2004)
	Pancreas	Mouse	-/- Hypoplasia	(Sock et al., 2004)
<i>Sox17</i>	Lung	Mouse	-/- Microvasculature, hypoperfusion	(Lange et al., 2014)
<b>Ptf1</b>				
<i>Ptf1a</i>	Pancrea	Human	-/- Agenesis <sup>e</sup>	(Sellick et al., 2004)
	Pancreas	Mouse	-/- Aplasia, hypoplasia	(Kawaguchi et al., 2002)
<i>Ptf1-p48</i>	Pancreas	Mouse	-/- Agenesis	(Krapp et al., 1998)
<b>Tcf/Lef</b>				
<i>Lef1</i>	Trachea	Mouse	-/- Agenesis (submucosal gland)	(Duan et al., 1999)
<i>Tcf2</i>	Pancreas	Mouse	-/- Agenesis	(Haumaitre et al., 2005)
<b>T-box</b>				
<i>Tbx2</i>	Mammary	Mouse	-/- No phenotype	(Jerome-Majewska et al., 2005)
<i>Tbx3</i>	Mammary	Human	+/- Ulnar-mammary syndrome	(Bamshad et al., 1997)
		Mouse	-/- Aplasia	(Davenport et al., 2003)
		Mouse	+/- Aplasia, dysplasia (nipple and ductal tree)	(Douglas and Papaioannou, 2013)
<i>Tbx2; Tbx3</i>	Mammary	Mouse	<i>Tbx2</i> <sup>-/-</sup> ; <i>Tbx3</i> <sup>+/-</sup> ↑ aplasia compared to <i>Tbx3</i> <sup>-/-</sup>	(Jerome-Majewska et al., 2005)

Table S2. Loss-of-function phenotypes for transcription factors (Continued)

Gene	Organ	Species	Phenotype	References
<b>Rar</b>				
<i>Rara</i> ; <i>Rarb2</i>	Kidney	Mouse	<i>Rar-α<sup>-/-</sup></i> ; <i>Rar-β2<sup>-/-</sup></i> Hypoplasia, Hydronephrosis	(Mendelsohn et al., 1994)
	Lung	Mouse	<i>Rar-α1<sup>-/-</sup></i> ; <i>Rar-β2<sup>-/-</sup></i> Agenesis (left), hypoplasia, dysplasia	(Mendelsohn et al., 1994)
<i>Rara</i> ; <i>Rary</i>	Kidney	Mouse	<i>Rara<sup>-/-</sup></i> ; <i>Rary<sup>-/-</sup></i> Agenesis, aplasia	(Mendelsohn et al., 1994)
	Lung	Mouse	<i>Rara<sup>-/-</sup></i> ; <i>Rary<sup>-/-</sup></i> Dysplasia	(Mendelsohn et al., 1994)
<i>Rarb2</i> ; <i>Rary</i>	Kidney	Mouse	<i>Rarb2<sup>-/-</sup></i> ; <i>Rary<sup>-/-</sup></i> Hydronephrosis	(Mendelsohn et al., 1994)
<b>Others</b>				
<i>Hes1<sup>f</sup></i>	Lung	Mouse	-/- Dysplasia (nodule), hypoplasia	(Ito et al., 2000)
<i>Nfib</i>	Lung	Mouse	-/- Hypoplasia	(Gründer et al., 2002)
<i>Pdx1</i> ; <i>Sox9</i>	Pancreas	Mouse	<i>Pdx1<sup>-/-</sup></i> ; <i>Sox9<sup>-/-</sup></i> Agenesis (ventral), hypoplasia (dorsal)	(Shih et al., 2015)
<i>Smad1</i>	Lung	Mouse	-/- Dysplasia	(Xu et al., 2011)
<i>Smad5</i>	Lung	Mouse	-/- No phenotype	(Xu et al., 2011)
<i>Sna1</i>	Vascular	Mouse	-/- Aplasia	(Lomeli et al., 2009)

List of transcription factors may not be complete. *Pax*, paired box; *Pdx*, pancreatic and duodenal homeobox; *Ets*, E26 transformation-specific; *Fox*, forkhead box; *Trh*, tracheless; *Sox*, SRY-related HMG-box; *Ptf*, pancreas transcription factor; *Tcf*, T cell-factor; *Lef*, lymphoid enhancer binding factor; *Rar*, retinoic acid receptor; *Hes*, hairy and enhancer of split; *Nf*, nuclear factor; *Sna*, snail.

<sup>a</sup>SG, salivary glands.

<sup>b</sup>In hypothyroidism, sensorineural deafness, and renal dysplasia.

<sup>c</sup>Other phenotypes include neonatal and childhood-onset diabetes mellitus.

<sup>d</sup>In some cases, pulmonary hypoplasia or tracheopulmonary defects are observed.

<sup>e</sup>Other phenotypes include cerebellar agenesis and permanent neonatal diabetes mellitus.

<sup>f</sup>Downstream of Notch.

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