**Why do mutations of *ACTB*, coding for β-actin, cause kidney malformations?**

Being born with abnormal kidneys is an important cause of renal failure in children and young adults. We recently discovered a fault in a gene that normally controls the shape of cells. People with this genetic fault are born with abnormal kidneys and can often also have abnormal hearts and brains. In this study, we will begin to model this devastating human disease to better understand how the gene problem causes the disease. The results of our project will help answer the question “Why was our child born with a devastating disease”. In the longer term, the results may also help design new therapies for poorly formed kidneys.