

## **SIGNIFICANCE STATEMENT**

Pseudoxanthoma elasticum (PXE) is a genetic disease caused by *ABCC6* gene mutations that results in low systemic pyrophosphate levels and soft tissue calcifications. This report describes the presence of massive papillary calcifications (Randall plaque) and an increased prevalence of kidney stones in patients with PXE. *Abcc6* knockout mice develop Randall plaque, showing the major role of pyrophosphate in the prevention of kidney calcifications.