



## ABCC6 Deficiency Promotes Development of Randall Plaque

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Type de publication	Article de revue
Auteur	Letavernier, Emmanuel [1], Kauffenstein, Gilles [2], Huguet, Léa [3], Navasiolava, Nastassia [4], Boudierlique, Elise [5], Tang, Ellie [6], Delaitre, Léa [7], Bazin, Dominique [8], de Frutos, Marta [9], Gay, Clément [10], Perez, Joëlle [11], Verpont, Marie-Christine [12], Haymann, Jean-Philippe [13], Pomozi, Viola [14], Zoll, Janna [15], Le Saux, Olivier [16], Daudon, Michel [17], Lefthériotis, Georges [18], Martin, Ludovic [19]
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Mots-clés	Calcium [20], genetic renal diseaseminera [21], l metabolism [22]
Résumé en anglais	<p><b>BACKGROUND:</b> Pseudoxanthoma elasticum (PXE) is a genetic disease caused by mutations in the gene that result in low pyrophosphate levels and subsequent progressive soft tissue calcifications. PXE mainly affects the skin, retina, and arteries. However, many patients with PXE experience kidney stones. We determined the prevalence of this pathology in patients with PXE and examined the possible underlying mechanisms in murine models.</p> <p><b>METHODS:</b> We conducted a retrospective study in a large cohort of patients with PXE and analyzed urine samples and kidneys from mice at various ages. We used Yasue staining, scanning electron microscopy, electron microscopy coupled to electron energy loss spectroscopy, and Fourier transform infrared microspectroscopy to characterize kidney calcifications.</p> <p><b>RESULTS:</b> Among 113 patients with PXE, 45 (40%) had a past medical history of kidney stones. Five of six computed tomography scans performed showed evidence of massive papillary calcifications (Randall plaques). mice spontaneously developed kidney interstitial apatite calcifications with aging. These calcifications appeared specifically at the tip of the papilla and formed Randall plaques similar to those observed in human kidneys. Compared with controls, mice had low urinary excretion of pyrophosphate.</p> <p><b>CONCLUSIONS:</b> The frequency of kidney stones and probably, Randall plaque is extremely high in patients with PXE, and mice provide a new and useful model in which to study Randall plaque formation. Our findings also suggest that pyrophosphate administration should be evaluated for the prevention of Randall plaque and kidney stones.</p>
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## Liens

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- [26] <http://www.ncbi.nlm.nih.gov/pubmed/29991491?dopt=Abstract>

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