



Dietary Pyrophosphate Modulates Calcification in a Mouse Model of Pseudoxanthoma Elasticum: Implication for Treatment of Patients

Submitted by Beatrice Guillaumat on Tue, 01/08/2019 - 15:27

Titre	Dietary Pyrophosphate Modulates Calcification in a Mouse Model of Pseudoxanthoma Elasticum: Implication for Treatment of Patients
Type de publication	Article de revue
Auteur	Pomozi, Viola [1], Julian, Charnelle B [2], Zoll, Janna [3], Pham, Kevin [4], Kuo, Sheree [5], Tókési, Natália [6], Martin, Ludovic [7], Váradi, András [8], Le Saux, Olivier [9]
Editeur	Elsevier
Type	Article scientifique dans une revue à comité de lecture
Année	2018
Langue	Anglais
Date	20 Nov. 2018
Titre de la revue	The journal of investigative dermatology
ISSN	1523-1747
Résumé en anglais	<p>Pseudoxanthoma elasticum (PXE) is a heritable disease caused by ABCC6 deficiency. Patients develop ectopic calcification in skin, eyes and vascular tissues. ABCC6, primarily found in liver and kidneys, mediates the cellular efflux of ATP, which is rapidly converted into pyrophosphate (PPi), a potent inhibitor of calcification. PXE patients and Abcc6 mice display reduced PPi levels in plasma and peripheral tissues. PXE is currently incurable, although some palliative treatments exist. In recent years, we have successfully developed therapeutic methodologies to compensate the PPi deficit in animal models and humans. Here, we inadvertently discovered that modulating dietary PPi can also be an effective approach to reducing calcification in Abcc6 mice. Our findings were prompted by a change in institutional rodent diet. The new chow was enriched in PPi, which increased plasma PPi, and significantly reduced mineralization in Abcc6 mice. We also found that dietary PPi is readily absorbed in humans. Our results suggest that the consumption of food naturally or artificially enriched in PPi represents a possible intervention to mitigate calcification progression in PXE, that dietary preferences of patients may explain PXE heterogeneous manifestations and that animal chow has the potential to influence data reproducibility.</p>
URL de la notice	http://okina.univ-angers.fr/publications/ua18549 [10]
DOI	10.1016/j.jid.2018.10.040 [11]
Lien vers le document	https://www.jidonline.org/article/S0022-202X(18)32826-4/pdf [12]
Titre abrégé	J. Invest. Dermatol.
Identifiant (ID) PubMed	30468740 [13]

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- [1] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=29176>
- [2] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32431>
- [3] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=31472>
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- [12] <https://www.jidonline.org/article/S0022-202X>
- [13] <http://www.ncbi.nlm.nih.gov/pubmed/30468740?dopt=Abstract>

Publié sur *Okina* (<http://okina.univ-angers.fr>)