

POSTER PRESENTATION



Autoimmunity and cystatin SA 1 deficiency behind chronic mucocutaneous candidiasis in autoimmune polyendocrine syndrome

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From 7th European Workshop on Immune-Mediated Inflammatory Diseases Noordwijk aan Zee, the Netherlands. 28-30 November 2012

Patients with the monogenic disease autoimmune polyendocrine syndrome type I (APS I) develop autoimmunity against multiple endocrine organs and suffer from chronic mucocutaneous candidiasis (CMC), a paradoxical complication with an unknown mechanism. We report that saliva from APS I patients with CMC was defective in inhibiting growth of C. albicans in vitro and had reduced levels of a salivary protein identified as cystatin SA1. In contrast, APS I patients with no CMC expressed salivary cystatin SA1 and could inhibit C. albicans to the same extent as healthy controls. We evaluated the anti-fungal activity of cystatin SA1 and found that synthesized full length cystatin SA1 efficiently inhibited growth of C. albicans in vitro. Moreover, APS I patients exhibited salivary IgA autoantibodies recognizing myosin-9, a protein expressed in the salivary glands that also produce cystatin SA1, thus linking autoimmunity to cystatin SA1 deficiency and CMC. This data suggests an autoimmune mechanism behind CMC in APS I and provide rationale for evaluating cystatin SA1 in antifungal therapy.

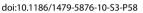
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Published: 28 November 2012

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Cite this article as: Lindh *et al.*: **Autoimmunity and cystatin SA 1** deficiency behind chronic mucocutaneous candidiasis in autoimmune polyendocrine syndrome. *Journal of Translational Medicine* 2012 **10**(Suppl 3):P58.

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