

Title: Prolidase Deficiency *GeneReview* - Laboratory techniques used in the diagnosis of prolidase deficiency

Authors: Ferreira C, Wang H

Date: June 2015

Note: The following information is provided by the authors listed above and has not been reviewed by *GeneReviews* staff.

Measurement of prolidase enzyme activity in erythrocytes, leukocytes, or cultured fibroblasts. Assays are performed with high concentrations (10-50 mM) of imidodipeptides as substrates [Hechtman 2014]. The preferred substrate is Gly-Pro [Royce & Steinmann 2002]. Following incubation at 37°C, the reaction is terminated by heating or by adding trichloroacetic acid [Hechtman 2014]. The released proline is quantitated by spectrophotometry, using either Chinard's original method [Chinard 1952] or its modification by Myara et al [1982]. Other techniques used for measuring prolidase activity include reverse-phase high-performance liquid chromatography [Harada et al 1990], capillary electrophoresis [Zanaboni et al 1997], and more recently MALDI-TOF mass spectrometry [Kurien et al 2004].

A review of the different laboratory techniques used in the diagnosis of prolidase deficiency can be found elsewhere [Kurien et al 2006, Viglio et al 2006].

References

- Chinard, F.P., 1952. Photometric estimation of proline and ornithine. *J. Biol. Chem.* 199:91–95.
- Harada, M., Fukasawa, K.M., Hiraoka, B.Y., Fukasawa, K., Mogi, M., 1990. High-performance liquid chromatographic procedure for the determination of serum prolidase activity. *J. Chromatogr.* 530:116–121.
- Hechtman, P., 2014. Prolidase Deficiency, in: Beaudet, A.L., Vogelstein, B., Kinzler, K.W., Antonarakis, S.E., Ballabio, A., Gibson, K.M., Mitchell, G. (Eds.), *The Online Metabolic and Molecular Bases of Inherited Disease*. The McGraw-Hill Companies, Inc., New York, NY.
- Kurien, B.T., Patel, N.C., Porter, A.C., D'Souza, A., Miller, D., Matsumoto, H., Wang, H., Scofield, R.H., 2006. Prolidase deficiency and the biochemical assays used in its diagnosis. *Anal. Biochem.* 349:165–175. doi:10.1016/j.ab.2005.10.018
- Kurien, B.T., Patel, N.C., Porter, A.C., Kurono, S., Matsumoto, H., Wang, H., Scofield, R.H., 2004. Determination of prolidase activity using matrix-assisted laser desorption/ionization time-of-flight mass spectrometry. *Anal. Biochem.* 331:224–229. doi:10.1016/j.ab.2004.04.043
- Myara, I., Charpentier, C., Lemonnier, A., 1982. Optimal conditions for prolidase assay by proline colorimetric determination: application to iminodipeptiduria. *Clin. Chim. Acta* 125:193–205.
- Royce, P., Steinmann, B., 2002. Prolidase deficiency, in: Royce, P., Steinmann, B. (Eds.), *Connective Tissue and Its Heritable Disorders*. Wiley-Liss, New York, pp. 727–738.
- Viglio, S., Annovazzi, L., Conti, B., Genta, I., Perugini, P., Zanone, C., Casado, B., Cetta, G., Iadarola, P., 2006. The role of emerging techniques in the investigation of prolidase deficiency: from diagnosis to the development of a possible therapeutical approach. *J. Chromatogr. B Analyt. Technol. Biomed. Life Sci.* 832:1–8. doi:10.1016/j.jchromb.2005.12.049
- Zanaboni, G., Viglio, S., Dyne, K.M., Grimm, R., Valli, M., Cetta, G., Iadarola, P., 1997. Direct monitoring of prolidase activity in cultured skin fibroblasts using capillary electrophoresis. *J. Chromatogr. B Biomed. Sci. Appl.* 695:77–84.