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Quantification of phenylalanine
hydroxylase activity by ...

1 Quantification of Phenylalanine
Hydroxylase Activity by Isotope-Dilution
Liquid Chromatography-Electrospray
Ionization Tandem Mass Spectrometry
Caroline Heintz¹, Heinz Troxler¹, Aurora
Martinez², Beat Thöny^{1,3,4}, Nenad

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Blau^{1,3,4,5}* 1Division of Clinical
Chemistry and Biochemistry, University
Children's Hospital, Zürich, Switzerland;
2Department of Biomedicine, University
of Bergen ...

Quantification of phenylalanine
hydroxylase activity by ...

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With heart failure a leading cause of death, a better understanding of metabolic function in the heart is a welcome advance. Murashige et al. measured more than 270 metabolites using liquid chromatography-mass spectrometry in human blood samples taken from an artery entering the heart and from a vein leaving

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it. Differences thus reflected the metabolic processes at work in the heart.

QUANTIFICATION OF PHENYLALANINE HYDROXYLASE ACTIVITY BY LC ...

Activity of in vitro expressed mutant PAH may predict the patient's phenotype and

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response to tetrahydrobiopterin (BH(4)),
the cofactor of PAH. METHODS:
A robust LC-ESI-MSMS PAH assay for
the quantification of phenylalanine and
tyrosine was developed.

Describe the biochemical relationships
between these ...

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The range of phenylalanine hydroxylase activity was determined by measuring the conversion of radioactive phenylalanine to tyrosine in liver and kidney of various vertebrates. Rodents (rats, mouse, gerbil, hamster and guinea pig) were found to have the highest liver phenylalanine hydroxylase activity among all animals

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Phenylalanine hydroxylase - Wikipedia
quantification of phenylalanine
hydroxylase activity by lc-ms/ms Article
in Journal of Inherited Metabolic Disease
34:S94-S94 · January 2011 with 15 Reads
How we measure 'reads'

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Phenylalanine Hydroxylase Activity in
Liver Biopsies from ...

The data obtained indicate the presence of phenylalanine hydroxylase activity in human leucocytes and fibroblasts. The following methods were used: estimation of accumulation of the oxidized form of

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the pteridine cofactor after Ayling and coworkers and radiochemical method. Probably this activity is \square

Phenylalanine, Tyrosine & Tryptophan
HPLC Assay | Eagle ...

The activity of rat liver phenylalanine hydroxylase has been measured in systems

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which, in addition to the components necessary for hydroxylation, contained ascorbic acid and catalase.

6-Methyltetrahydropterine or the corresponding 6,7-dimethyl compound was used as tetrahydropterine cofactor.

The quantitative determination of

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Extract: Liver biopsy samples from the patients with hyperphenylalaninemia have an average of 5% of the normal hydroxylase activity. The parents of the patients have between 7.3% (excluding the ...

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Distribution of phenylalanine hydroxylase (EC 1.14.3.1) in ...

Phenylketonuria (PKU), caused by phenylalanine hydroxylase (PAH) gene variants, is a common autosomal inherited metabolic disease. So far, 1111 PAH variants have been revealed. The residual activity of the PAH variants is the key

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determinant of the metabolic phenotype
and BH₄responsiveness in PKU patients.

Quantification of phenylalanine
hydroxylase activity by ...

Residual phenylalanine hydroxylase
(PAH) activity is the key determinant for
the phenotype severity in phenylketonuria

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(PKU) patients and correlates with the patient's genotype. Activity of in vitro expressed mutant PAH may predict the patient's phenotype and response to tetrahydrobiopterin (BH 4), the cofactor of PAH.

Quantification of phenylalanine

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hydroxylase activity by ...

On the 20th day of gestation, the liver (and the kidney) is devoid of phenylalanine hydroxylase and at birth contains 20% of the adult activity. During the second postnatal week of development, when the phenylalanine hydroxylase activity was about 40% of the adult value, an injection

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of cortisol doubled this value.

Phenylalanine hydroxylase | enzyme |
Britannica

Phenylalanine deficiency stimulates the
activation of phenylalanine metabolites in
the brain tissues thus resulting in the
impairment of neurotransmitter synthesis

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(Schuck et al., 2015). The inactivity of enzyme phenylalanine hydroxylase lowers phenylalanine metabolism levels thus resulting in the development of an autosomal recessive disorder known as phenylketonuria (PKU).

Quantification of phenylalanine

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hydroxylase activity by ...

The predicted level of phenylalanine hydroxylase activity correlated strongly with the pretreatment serum level of phenylalanine ($r = 0.91$, P less than 0.001 in the Danish patients and $r = 0.74$, P ...

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Quantification Of Phenylalanine
Hydroxylase Activity

Quantification of phenylalanine
hydroxylase activity by isotope-dilution
liquid chromatography-electrospray
ionization tandem mass spectrometry.

Heintz C(1), Troxler H, Martinez A,
Thöny B, Blau N. Author information:

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Biochemistry, University Children's
Hospital, Zürich, Switzerland.

Comprehensive quantification of fuel use
by the failing ...

□ caused by decreased activity of
phenylalanine hydroxylase (PAH), an

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enzyme that converts the amino acid phenylalanine to tyrosine, a precursor of several important hormones and skin, hair, and eye pigments. Decreased PAH activity results in accumulation of phenylalanine and a decreased amount of tyrosine and other metabolites.

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[Detection of phenylalanine hydroxylase activity in human ...

Phenylalanine hydroxylase (PAH) (EC 1.14.16.1) is an enzyme that catalyzes the hydroxylation of the aromatic side-chain of phenylalanine to generate tyrosine. PAH is one of three members of the bipterin-dependent aromatic amino acid

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hydroxylases, a class of monooxygenase that uses tetrahydrobiopterin (BH₄, a pteridine cofactor) and a non-heme iron for catalysis.

Phenylalanine Hydroxylase Deficiency |
AACC.org

Phenylketonuria is the most abundant

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genetic disorder of the amino acid metabolism. It is characterized by a lack of phenylalanine hydroxylase which causes an increase of phenylalanine in cells and body fluids. Due to the lack of enzyme activity, phenylalanine, normally metabolised to tyrosine, is converted to phenylpyruvic acid.

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In vitro residual activities in 20 variants of
...

Thank you for joining me on this Pearl of
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Quantification of phenylalanine
hydroxylase activity by ...

Residual phenylalanine hydroxylase
(PAH) activity is the key determinant for
the phenotype severity in phenylketonuria
(PKU) patients and correlates with the

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patient's genotype. Activity of in vitro expressed mutant PAH may predict the patient's phenotype and response to tetrahydrobiopterin (BH 4), the cofactor of PAH.

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