
International Journal for Vitamin and Nutrition Research
Internationale Zeitschrift
für Vitamin- und Ernährungsforschung
Journal international de Vitaminologie et de Nutrition

EDITOR-IN-CHIEF

G. RITZEL, ST. ALBANVORSTADT 19, CH-4052 BASEL

CURATORES

K. Bernhard, Basel	R.S. Harris, Cambridge, Mass.	R.E. Stone, Chicago
H.D. Cremer, Giessen	C. den Hartog, Wageningen	A.V. Szent-Györgyi, Woods Hole
K. Folkers, Austin	T. Reichstein, Basel	

EDITORES

J.T. Abrams, Cambridge, GB	G. Brubacher, Basel	J. Ganguly, Bangalore
H. Berger, Innsbruck	R. Buzina, Zagreb	J. Mauron, La Tour-de-Peilz
H. Bickel, Zürich	D.B. Coursin, Lancaster	H.E. Sauberlich, Birmingham, USA
A. Bondi, Rehovoth	P.M. Dreyfus, Sacramento	M.L. Scott, Ithaca
M. Brin, Nutley	F. Fidanza, Perugia	

The "International Journal for Vitamin and Nutrition Research" is published at irregular intervals (about 4 fasc. per annum) 4 fasc. = 1 volume, prize per volume = Sw. Fr. 134.— / Die «Internationale Zeitschrift für Vitamin- und Ernährungsforschung» erscheint in zwangloser Reihenfolge (ca. 4 Hefte pro Jahr). 4 Hefte ergeben einen Band. Bezugspreis pro Band = sFr. 134.—, DM 170.— / Le «Journal international de Vitaminologie et de Nutrition» paraît irrégulièrement en fascicules (env. 4 fasc. par année). 4 fasc. = 1 volume, prix par volume = sfr. 134.—

Hans Huber Publishers Berne Stuttgart Toronto

Western Hemisphere: Jack K. Burgess Inc., 2175 Lemoine Avenue, Fort Lee, N.J. 07024

This journal is regularly listed in Current Contents and Index Medicus

The journal serves as the official organ of the Swiss Society for Nutrition Research

Inhalt / Contents

ARAFA A.M. and HUSSEIN L.: The Assessment of The Vitamin B ₆ Status among Egyptian School Children by Measuring the Urinary Cystathionine Excretion	321
ARIMANANA L. and LEATHWOOD P.D.: Effects of Prior Carbohydrate Intake on Protein/Carbohydrate Selection by the Rat	283
ARNAUD M.J., WELSCH C., GARREL D. and TOURNAIRE J.: Reproducibility of the Measurement of Whole Body Protein Turnover Using the End Product Method	282
BASU T.K., WEISER, T. and DEMPSTER J.F.: An <i>in vitro</i> Effect of Ascorbate on the Spontaneous Reduction of Sodium Nitrite Concentration in a Reaction Mixture	233
BAYLISS R.M., BROOKES R., McCULLOCH J., KUYL J.M. and METZ J.: Urinary Thiamine Excretion after Oral Physiological Doses of the Vitamin	161
BICKEL R., REINHARDT C.A. and LÜTHY J.: <i>In vitro</i> Growth Inhibition Assay for the Detection of Fusarium Toxins in Feedstuffs	287
BIESALSKI H.K.: Retinol and Retinyl Ester in Separated Structures of the Guinea Pig Inner Ear . . .	113
BLUM M., PRABUCKI A.L. and SCHÜRCH A.: The Influence of Different Dietary Fats on the Life Span of Rats	276
BLUM M., PRABUCKI A.L. and SCHÜRCH A.: The Influence of Various Dietary Fats on the Fatty Acid Composition of Organ Lipids in the Rat	277
BONJOUR J.-P., BAUSCH J., SUORMALA T. and BAUMGARTNER E.R.: Detection of Biocytin in Urine of Children with Congenital Biotinidase Deficiency	223
BOUCHER-EHRENSPERGER M. and PETTER C.: Prevention of Thrombocytic Defects in the br/br Rabbit with Folic acid and Vitamin B ₁₂ : Analogy with the T.A.R. Syndrome in Humans	199
BRANCA D., SCUTARI G. and SILIPRANDI N.: Pantethine and Pantothenate Effect on the CoA Content of Rat Liver	211
CASTER W.O., DRUMMOND S.K. and TANNER M.A.: Maternal Diet and Mental Retardation in Southern Georgia, USA	371
CHANGBUMRUNG S., POSHAKRISHANA P., VUDHIVAI N., HONGTONG K., PONGPAEW P., MIGASENA P. <i>et al</i> : Measurements of B ₁ , B ₂ , B ₆ Status in Children and their Mothers Attending a Well-baby Clinic in Bangkok	149
CULLUM M.E., OLSON J.A. and VEYSEY S.: Analysis of Deuterated Analogs of Vitamin A by Electron Impact and Chemical Ionization Modes in Gas Chromatography Coupled to Mass Spectrometry	3
CULLUM M.E., JOHNSON C. and ZILE M.H.: Comparison of Fetal and Adult Retinol and Retinoic Acid Binding Proteins in Bovine Serum and Pigment Epithelium	297
CULLUM M.E., ZILE M.H. and VEYSEY S.W.: Analysis of Retinol and Dideuterated Retinol in Rat Plasma by Gas Chromatography Combined Mass Spectrometry	11
DÉCOMBAZ J., SARTORI D., ARNAUD A.L., THELIN A.L., SCHÜRCH P. and HOWALD H.: Oxidation and Metabolic Effects of Fructose or Glucose Ingested before Exercise in Man	281
DOREA J.G., SOUZA J.A., GALVÃO M.O. and IUNES M.A.F.: Concentration of Vitamin A in the Liver of Foetuses and Infants dying of Various Causes in Brasilia, Brazil	119
FIDANZA A.A.: Nutritional Status of the Elderly, IV	361
FIDANZA F., BRUBACHER G., SIMONETTI M.S. and CUCCHIA L.M.: Nutritional Status of the Elderly, III.	355
FIDANZA F., FIDANZA A.A., COLI R. and MENCARINI C.A.: Food and Nutrient Consumption of Two Rural Italian Population Groups Followed for Twenty Years	91
FIDANZA F., SIMONETTI M.S., CUCCHIA L.M., BALUCCA G.G. and LOSITO G.: Nutritional Status of the Elderly, II.	75
FISHBAINE B. and BUTTERFIELD G.: Ascorbic Acid Status of Running and Sedentary Men	273
FLATT J.P., ACHESON K.J., RAVUSSIN E. and JÉQUIER E.: Dietary Fat Substrate Utilization and Short Term Body Composition Changes in Man	279
FLORIDI A., PUPITA M., PALMERINI C.A., FINI C. and FIDANZA A.A.: Thiamin Pyrophosphate Determination in Whole Blood and Erythrocytes by High Performance Liquid Chromatography .	165
FRIGG M. and BROZ J.: Relationships between Vitamin A and Vitamin E in the Chick	125

GIBSON J. and WESTHUYZEN J. VAN DER: Effect of L-Dihydroxyphenylalanine (L-dopa) and Methionine on Tissue S-Adenosylmethionine Concentrations in Cobalamin-inactivated Fruit Bats .	329
GLENN B.P. and ELY D.G.: Effect of Lipid-coated Lysine on Digestion and Nitrogen Metabolism by Wethers	377
GRECO A.M., BOSCHI G., STICCHI R., VETRANI A. and SALVATORE G.: Effects of a Purified Diet Enriched with Animal Protein on Female Rats. Preliminary Report.	263
GUIDOUX R.: Ca ²⁺ Regulation by Isolated Rat Liver Mitochondria Influence of the Intramitochondrial Citrate Content on the Extramitochondrial Ca ²⁺ Level	288
GUIGOZ Y. and JULLERAT M.: Induction of Tyrosine Aminotransferase (TAT) by a Physiological Stress as Function of Dietary Fat	289
GULLAND J.C., BEREKSI-REGUIG B., LEQUEU B., MOREAU D. and KLEPPING J.: Evaluation of Pyridoxine Intake and Pyridoxine Status among Aged Institutionalised People	185
HIDIROGLOU M. and WILLIAMS C.J.: Vitamin D ₃ Levels in Certain Sheep Tissues at Various Times after the Intramuscular Administration of Vitamin D ₃	17
HOLLOWAY D.E., GUIRY V.C., HOLLOWAY B.A. and RIVERS J.M.: Influence of Dietary Ascorbic Acid on Cholesterol 7- α -Hydroxylase Activity in the Rat	333
HOLM J., HANSEN I. and LYNGBYE J.: Quantitative and Qualitative Effects of N ¹⁰ -Methylfolate on High-affinity Folate Binding in Human Leukocytes	195
HORIUCHI S. and ONO S.: Effects of Riboflavin Administration on the Phospholipid Metabolism of Rat Liver Impaired with Carbon Tetrachloride	173
HUNT C., CHAKRAVORTY N.K. and ANNAN G.: The Clinical and Biochemical Effects of Vitamin C Supplementation in Short-stay Hospitalized Geriatric Patients	65
KIRCHGESSNER M. and MÜLLER H.L.: Thermogenesis with a Ketogenic Diet in Sows	99
KORPELA H., LOUENIVA R., YRÄNHEIKKI E. and KAUPPIA A.: Selenium Concentration in Maternal and Umbilical Cord Blood, Placenta and Amniotic Membranes	257
KRATZING C.C., KELLY J.D. and OELRICHS B.A.: Ascorbic Acid Changes in Brain	349
KRAUS K.-H., BONJOUR, J.-P. and BERLIT P.: Biotin Levels in Plasma of Patients with Friedreich Ataxia and Other Spinocerebellar Degenerations	272
KRAUSE K.-H., KOCHEN W., BERLIT P. and BONJOUR J.-P.: Excretion of Organic Acids Associated with Biotin Deficiency in Chronic Anticonvulsant Therapy	217
KUMPULAINEN J., VUORI E. and SIIMES M.A.: Effect of Maternal Dietary Selenium Intake on Selenium Levels in Breast Milk	251
KUNZ C., NIESEN M., LILIENFELD-TOAL L. VON, and BURMEISTER W.: Vitamin D, 25-Hydroxy-Vitamin D and 1,25-Dihydroxy-Vitamin D in Cow's Milk, Infant Formulas and Breast Milk during Different Stages of Lactation.	141
LABADARIOS D., SHEPHARD G.S., MINEUR L.G., VAN BUUREN A.J., HUTCHISON M.E. and OOSTHUIZEN O.J.: Biochemical Vitamin B ₆ Deficiency in Adults with Chronic Glomerulonephridies with and without the Nephrotic Syndrome	313
LANGHANS W. and SCHARRE E.: Feeding Rats a High Fat Diet Changes the Effect of Various Metabolic Challenges on Food Intake	280
MOSER U. and WEBER F.: Uptake of Ascorbic Acid by Human Granulocytes.	47
MOËNNOZ D. and ASHLEY D.V.: Comparison of Plasma Free Tryptophan and Plasma Tryptophan to Large Neutral Amino Acid Ratios as Physiological Predictors of Brain Tryptophan.	285
MOËNNOZ D., EXELL N. and ASHLEY D.V.: Lack of Influence of Free Fatty Acids on Tryptophan-albumin Binding in Plasma.	286
NADIGER H.A., RAD A.S. and SADASIVUDU B.: Effect of Simultaneous Administration of Vitamin E and Pyridoxin on Erythrocyte Membrane Na ⁺ K ⁺ ATPase Activity.	307
OELRICHS B.A., KRATZING C.C., KELLY J.D. and WINZOR D.J.: The Binding of Ascorbate to Bovine Serum Albumin.	61
PIIRONEN V., VARO P., SYVÄOJA E.-L., SALMINEN K., KOIVISTOINEN P. and ARVILOMMI H.: High Performance Liquid Chromatographic Determination of Tocopherols and Tocotrienols and its Application to Diets and Plasma of Finnish Men, II.	41
PIIRONEN V., VARO P., SYVÄOJA E.-L., SALMINEN K. and KOIVISTOINEN P.: High Performance Liquid Chromatographic Determination of Tocopherols and Tocotrienols and its Application to Diets and Plasma of Finnish Men, I.	35

Research Note

Biotin Levels in Plasma of Patients with Friedreich Ataxia and Other Spinocerebellar Degenerations KLAUS-HENNING KRAUSE¹, JEAN-PIERRE BONJOUR² and PETER BERLIT¹

In patients with Friedreich ataxia low pyruvate carboxylase (PC) activity in liver and fibroblasts has been described recently [1]. PC activity can be lowered due to reduced circulating biotin levels caused by disturbances of the metabolism, absorption or transport of biotin. We determined plasma biotin levels using *Lactobacillus plantarum* as test organism [2], in 12 patients with spinocerebellar disorders, and compared the concentrations with those of normal controls, matched for age (± 3 ys) and sex. 5 non-related patients (cases 1-5) exhibited the typical characteristics of Friedreich ataxia with recessive familial involvement, 2 patients (cases 6 and 7) showed a sporadic olivopontocerebellar atrophy, 3 siblings (cases 8-10) a combination of cerebellar ataxia with degeneration of anterior column cells, and 2 siblings (cases 11 and 12) cerebellar atrophy of Nonne-Pierre-Marie type. As shown in the table, the patients with Friedreich ataxia did not have low biotin plasma levels; this finding indicates, that the reduced PC activity observed in Friedreich ataxia is not due to a decrease in biotin level. In contrast to all other patients, the two patients with sporadic olivopontocerebellar atrophy showed low biotin levels of 188 and 185 ng/l. The clinical implication of this finding is not clear. Oral medication with biotin in a dose of 10 mg per day over a period of three months showed no certain improvement of the disorder in patients 6 and 7.

Table I: Biotin levels in plasma (ng/l) in patients with spinocerebellar disorders and in controls

No of patient	biotin in plasma of patients (ng/l)	biotin in plasma of controls (ng/l)
1	348	373
2	322	367
3	347	263
4	317	435
5	588	348
6	188	382
7	185	357
8	332	243
9	317	343
10	247	338
11	300	375
12	312	345

References: 1. DIJKSTRA, U.J., WILLANS, J.L., JOOSTEN, E.M.G., GABRIEELS, F.J.M.: *Ann. Neurol.* 13, 325 (1983). - 2. FRIGG, M., BRUBACHER, G.: *Internat. J. Vit. Nutr. Res.* 46, 314 (1976).

¹ Neurologische Universitätsklinik, Voss-Str. 2, D-6900 Heidelberg, F.R.G.

² Department of Vitamin and Nutrition Research, F. Hoffmann-La Roche & Co., Basel, Switzerland.