SCIENTIFIC ARTICLES/ CLINICAL STUDIES

Sodium Phenylbutyrate
(4-Phenylbutyric Acid, Sodium Salt)

CANCER


De-shum, Pan, Wei-qiang, Chen. The effects of sodium phenylbutyrate (SPB) on the expression of p21 and survivine genes on human tongue squamous cancer cell line Tca8113. *Chinese Pharmacological Bulletin,* February 2010


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Cunha De Santis , G; de barros Tammarozzi, M; Sousa, RB; Moreno, Se; Secco, D; Garcia, AB; Lima, ASG; Faccioli, LH; Falcao, RP; Cunha, FQ; Rego, EM. Adhesion molecules and Differentiation Syndrome: phenotypic and functional analysis of the effect of ATRA, As203, phenylbutyrate, and G-CSF in acute promyelocytic leukemia. Haematologica 2007; 92(12):1615-1622.


Lubbert, M; Claus, R. Epigenetic inactivation of gene expression. New therapeutic targets in hematology. Onkologe 2007;13(1):46-55. lindsay@customvite.com or service@nutrilab.com.


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BLOOD


SICKLE CELL ANEMIA, THALASSEMA & COOLEY’S ANEMIA


Witt, Olaf, Mönkemeyer, Sven, Röndahl, Gabi, Erdlenbruch, Bernhard, Reinhardt, Dirk, Kanbach, Katrin and Pekrun, Arnulf. Induction of fetal hemoglobin expression by the histone deacetylase inhibitor apicidin. *Blood*, March 1, 2003, Volume 101, Number 5


**UREA CYCLE DISORDER/ ORNITHINE TRANSCARBAMYLASE DEFICIENCY**


Lee B, Garovoy MR, Gargosky SE, Berry SA. Preliminary data on adult patients with urea cycle disorder (UCD) in an open-label, switch-over, does-escalation study comparing a new ammonia scavenger glyceryl tri (4-phenylbutyrate) (HPN-100), to buphenyl (sodium phenylbutyrate – PBA). *Journal of Inherited Metabolic Disease* 2008; 31 (Suppl. 1): 91.


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LEUCINE MATABOLISM


LIPOPROTEIN RECEPTOR


Tveten K, Holla OL, Ranheim T, Berge KE, Leren TP, Kulseth MA. 4-Phenylbutyrate restores the functionality of a misfolded mutant low-density lipoprotein receptor. FEBS J 2007; 274 (8): 1881-1893.
CYSTIC FIBROSIS


Roque, T., Boncoeur, E., Saint-Criq, V., Bonvin, E., Clement, A., Tabary, O., Jacquot, J. Pro-inflammatory effect of sodium 4-phenylbutyrate in ΔF508-CFTR lung epithelial cells: Involvement of ERK1/2 and JNK signaling. JPET, June 23, 2008, DOI:10.1124/jpet.107.135186


**FAMILIAL HYPERCHOLESTEROLEMIA**


**PHENYLKETONURIA**


**LUNG/ABCA3 DEFECTS**


**ADRENOLEUCODYSTROPHY (ALD)**


**KIDNEY**


Liu Xiao Li, Nephrin cellular trafficking and intracellular interactions, Division of Matrix Biology Dept. of Medical Biochemistry and Biophysics, Karolinska Institutet, Stockholm, Sweden, May 2004.


**BLADDER DYSFUNCTION**


**AMYOTROPHIC LATERAL SCLEROSIS (ALS)**


CEREBRAL ISCHEMIC INJURY


EPILEPSY


CARDIAC INJURY

Okajima, M., Takamura, M., Usui, S., Kaneko, S. Sodium 4-phenylbutyrate protects against myocardial ischemia-reperfusion injury by reducing unfolded protein response-mediated apoptosis in mice. 30th International Symposium on Intensive Care and Emergency Medicine, Brussels, Belgium March 9-12, 2010


HUNTINGTON’S DISEASE


Tolchin, E. Biomarkers of Huntington’s Disease Found in Blood 7/26/05. *Bioscience Technology*, http://www.biomedicalproducts.com


**MULTIPLE SCLEROSIS (MS)**


**PARKINSON’S DISEASE**


Kubota, Kyoko, Niinuma, Yoshifumi, Kaneko, Masayuki, Okuma, Yasunobu, Sugai, Mami, Omura, Tomohiro, Uesugi, Mai, Uehara, Takashi, Hosoi, Toru, Nomura, Yasuyuki. Suppressive effects of 4-Phenylbutyrate on the aggregation of Pael receptors and endoplasmic reticulum stress. *Journal of Neurochemistry*, 2006, 97, 1259-1268

SPINAL MUSCULAR ATROPHY


Davies S.L., Moral M.A. Spinal muscular atrophies – distinctions and therapeutic progress. *Drugs of the*

Wirth, B, Brichta, L, Schrank, B, Lochmouller, H, Blick, S, Baasner, A, Heller, R. Mildly affected patients with spinal muscular atrophy are partially protected by an increased SMN2 copy number. *Hum Genet* 2006; 119 (4): 422-428


**ALZHEIMER’S DISEASE**


**AGING**


**EYE**


Christopher Kai-shun Leung, Chinese University of Hong Kong, Sodium 4-Phenylbutyrate has the ability to prevent steroid-induced glaucoma. *Press Release* August 17, 2010

Gong Bo, Zhang Li-Yun, Lam Dennis Shun-Chiu, Pang Chi-Pui, Yam Gary Hin-Fai. Sodium 4-phenylbutyrate ameliorates the effects of cataract-causing mutant gammaD-crystallin in cultured cells. *Molecular Vision* 2010 June 4, 2010


**RHEUMATOID ARTHRITIS**


**DIABETES**

Srinivasan K, Sharma SS. Sodium Phenylbutyrate ameliorates focal cerebral ischemic/reperfusion injury associated with comorbid type 2 diabetes by reducing endoplasmic reticulum stress and DNA fragmentation. *Behav Brain Res* Nov 20, 2011; 225(1); 110-6


**LIVER**

Van der Velden, L.M., Stapelbroek, J.M., Krieger, E., et al. Folding defects in P-type ATP 8B1 associated with hereditary cholestasis are ameliorated by 4-phenylbutyrate. *Hepatology* 2010;51(1);286-96


**ADDICTION**


**ISCHEMIC SPINAL CORD DAMAGE**


**MAPLE SYRUP URINE DISEASE**


**ENDOPLASMIC RETICULUM STRESS**


SCHIZOPHRENIA

Sandner, Guy, Host, Lionel, Angst, Marie-Josée, Guiberteau, Thierry, Guignard, Blandine, Zwiller, Jean.

OBESITY

Basseri Sana, Lhotak Sarka, Sharma Arya M, Austin Richard C. The chemical chaperone 4-phenylbutyrate inhibits adipogenesis by modulating the unfolded protein response. *Journal of Lipid Research* May 2009

MISCELLANEOUS RESEARCH & ARTICLES


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