7．Matsuzaki M., Hasegawa T., Takeda A., Kikuchi A., Furukawa K., Kato Y., Itoyama Y., Histochemical features of stress-induced aggregates in α-synuclein overexpressing cells. Brain Research (in press), 2004.

中島　孝

1．中島隆：難病の基礎知識I,II, 難病ホームヘルパーテキスト第6版2004（印刷中）
2．川上英孝、中島孝：神経・筋ー変性疾患、認定医・専門医のための内科学レビューー2004、総合医学社（印刷中）
3．中島孝：ALS早期診断のための新しい展開口脳SPECT画像での検討、神経内科2004（印刷中）
4．中島孝：緩和ケアとはなにか、難病と在宅ケア、9:7-11,2003
5．中島孝：筋萎縮性側索硬化症患者に対する生活の質（QoL）向上への取り組み、神経治療学、20:139-147,2003
6．Nemoto H, Toda H, Nakajima T, Hosokawa S, Okada Y, Ymamaoto K, Horicuchi R, Endo K, Masahiko M, Goto, F : Fluvoxamine modulates pain sensation and affective processing of pain in human brain, NeuroReport、14:791-797,2003

永井義隆

1．NagaiY., Fujikake,N., Ohno,K., Higashiyama ,., Popiel,H.A., Rahadian,J., Yamaguchi,M., Strittmatter,W.J., Burke, JR., Toda,T. : Prevention of polyglutamine oligomerization and neurodegeneration by the peptide inhibitor QBP 1 in Drosophia. Hum.Mol.Genet. 12:1253-1259,2003
2．永井義隆、戸田達史：神経変性疾患ポリグルタミン病に対する治療ペプチドQBP1.バイオインダストリー、20:52-63,2003
3．永井義隆:ポリグルタミン鎖結合ペプチドQBP1によるポリグルタミン病に対する分子治療法の確立。神経化学、42:443-456,2003

吉田邦広

1．Hashimoto,T., Sasaki,O., Yoshida,K., Takei,Y., Ikeda,S. : Periodic alternating nystagmus and rebound nystagmus in spinocerebellar ataxia 6. Mov Disord,18:1201-1204,2003.

業績

鈴木康之

1．鈴木康之：ペルオキシソーム病、無カタラーゼ血症。南山堂医学大事典（南山堂、東京）、2001

2．鈴木康之：ペルオキシソームでの代謝異常症。小児科学第２版（医学書院、東京）、2001

3．Takemoto.K., Suzuki,Y., Horibe.R., Shimozawa,N., Wanders,RJA., & Kondo,N. : Gas chromatography/mass spectrometry analysis of very long chain fatty acids, docosahexaenoic acid, phytanic acid and plasmalogen for the screening of peroxisomal disorders. Brain & Development,27:481-487,2003

4．Jia,Y., Qi,C., Zhang,Z., Hashimoto,T., Rao,MS., Huyghe,S., Suzuki,Y., Verdhoven,Van,PP., Baes,M., Reddy,JK. : Overexpression of PPAR α regulated genes in liver in the absence of peroxisome proliferation in mice deficient in both L- and D- forms of enoyl-CoA hydratase/dehydrogenaase enzymes of peroxisomal β -oxidation system. J Biol Chem,., 278:47232-47239,2003

5．Une,M., Iguchi,Y., Sakamoto,T., Tomita,T., Suzuki,Y., Morita,M., Imanaka,T. : ATP-dependent transport of bile acid intermediates across rat liver peroxisomal membranes. J Biochem, 134:225-230,2003

6．Shimozawa,M., Nagase,T., Takemoto,Y., Ohura,T., Suzuki,Y. Kondo,N. : Genetic heterogeneity of peroxisome biogenesis disorders among Japanese patients: evidence for a founder haplotype for the most common PEX10 gene mutation. Am J Med Genet, 120A:40-43,2003

7．Matsumoto,N., Tamuta,S., Furuki,S., Miyata,N., Moser,A., Shimozawa,N., Moser,W., Suzuki,Y., Kondo,N., Fujiki,Y. : Mutations in novel peroxin gene PEX26 that cause peroxisome-biogenesis disorders of complementation group 8 provide a genotype-phenotype correlation. Am J Hum Genet,73:233-246,2003

8．Raas-Rothschild,A., Wanders, RJ., Mooijer,PA., Gooties,J., Waterham,HR., Gutman,A., Suzuki,Y., Shimozawa,N., Kondo,N., Eshel,G., Espeel,M., Roels,F., Korman,SH. : A PEX6-defective peroxisomal biogenesis disorder with severe phenotype in an infant, versus mild phenotype resembling Usher syndrome in the parents. Am J Hum Genet, 70:1062-1068,2002

9．Shimozawa,N., Nagase,T., Takemoto,Y., Suzuk,Y., Fujiki,Y., Wanders,RJ., Kondo,N. : A novel aberrant splicing mutation of the PEX16 gene in two patients with Zellweger syndrome. Biochem Biophys Res Commun.,292:109-112,2002

10．Susuki,Y., Iai,M., Kamei,A., Tanabe,Y., Chiba,s., Yamaguchi,S., Zhang,Z., Takemoto,Y., Shimozawa,N., Kondo,N. : Peroxisomal acyl Coa oxidase deficiency. J Pediatr.,140:128-130,2002

11．Takemoto,Y., Suzuki,Y., Shimozawa,N., Tamakoshi,A., Onodera,O., Tsuji,S., Kondo,N. : Epidemiology of X-linked adrenoleukodystrophy in Japan. J Hum Genet.,47:590-593,2002

12．Suzuki,Y., Shimozawa,N., Imamuea,A., Fukudaa,S., Zhang,Z., Orri T., Kondo,N. : Clinical, biological, genetic aspects and neuronal migration in peroxisome biogenesis disorders. J InheritMetabDis.,24:151-165,2001

13．Suzuki,Y., Shimozawa,N., Orii,T., Tsukamoto,T., Osumi,T., Fujiki,Y., Kondo,N.. : Genetic and molecular bases of *peroxisome biogenesis disorders. Genetics in Medicine,3*:372-376,2001

14．Suzuki,Y., Imamura,A., Shimozawa,N., Kondo,N. : The clinical course of childhood and adolescent adrenoleukodystrophy before and Lorenzo's oil. Brain & Development,23:30-33,2001

15．Tamura,S., matsumoto,N., Imamura,S., Shimozawa,N., Suzuki,Y., Kondo,N., Fujiki,Y. : Phenotype-genocyte relationships in peroxisome biogenesis disorders of PEX1-defective complementation group 1 are defined by *Pex1p-Pex6p interaction. Biochem J.*,357:417-426,2001

16．Natsumoto,N., Tamura,S., Moser,A., Moser,H., Braverman,N., Shimozawa,N., Suzuki,Y., Kondo,N., Fujiki,Y. : The peroxin pex6p gene is impaired in peroxisome biogenesis disordes of complementation group 6. J Hum Genet,46:273-277,2001

17．Yamaguchi,S., Iga,M., Kimura,M., Suzuki,Y., Shimozawa,N., Gukao,T., Kondo,N., Tazawa,Y., Orii,T. : Urinary

organic acids in peroxisomal biogenesis disorders: a simple screening method. J Chromat B.,758:81-86,2001
18. Ito,R., Hunag,Y., Yao,C., Shimozawa,N., Suzuki,Y., Kondo,N., Imanaka,T., Usuda,N., Ito,M. : Temperature-sensitive phenotype of Chinese hamster ovary cells defective in PEX5 gene. Biochem Biophys Res Commun.,288:321-327,2001
19. Nakano,K., Suzuki,Y., Zhang,N.,Shimozawa,N., Ishii,N., Funatsuka,M., Shiraakawa,S., Itoh,M., Takashima,S., Une,M., Kana^aki,RR., Mukai,K., Osawa,M., Kondo,N. : D-bifunctional protein deficiency with fetal ascites,polyhydramnios, and contractures of hands and toes. J Pediatr,139:865-867,2001
20. Iwamura,A., Shimozawa,N., Suzuki,Y., Zhang,Z., Tsukamoto,T., Orii,T., Osumi,T., Kondo,N. : Temperature sensitive acyl-CoA oxidase import in group A peroxisome biogenesis disorders. J Med Genet., 38:871-874,2001

今中常雄

1. Kurisu, M., Morita, M., Kashiwayama, Y., Yokota, S., Hayashi, H., Sakai, Y., Ohkuma, S., Nishimura, M., and Imanaka, T.: Existence of catalase-less peroxisomes in Sf21 insect cells. Biochem. Biophys Res Commun. 306, 169-176, 2003.
2. Une,M., Iguchi, Y., Sakamoto, T., Tomita, T., Suzuki, Y., Morita, M., and Imanaka, T.: ATP-dependent transport of bile acid intermediates across rat liver peroxisomal membranes. J. Biochem. 134, 225-230, 2003.
3. Kikuchi, M., Hatano, N., Yokota, S., Shimozawa, N., Imanaka, T., and Taniguchi, H.: Proteomic analysis of rat liver peroxisome: Presence of peroxisome-specific isozyme of lon protease. J. Biol. Chem. 279, 421-428, 2004.
4. Fujino, T., Une, M.; Imanaka, T., Inoue, K., and Nishimaki-Mogami, T.: Structure-activity relationship of bile acids and bile acid analogs in regard to FXR activation. J. Lipid Res. 45, 132-138, 2004.
5. 柏山恭範、今中常雄： ペルオキシソーム ABC タンパク質と脂肪酸代謝. 膜 28, 263-270, 2003.

加藤俊一

1. Yahata.T., Kato,S., et al: : A highly sensitive strategy for SCID-repopulating cell assay by derect injection of primitive human hematopoietic cells into NOD/SCID mice bone marrow. Blood,.2003;101:2905-13.
2. Hagihara,M., Kato.S., et al. : The in vitro generation of Ph1+ ALL-specific HLA-A24- restricted cytotoxic T lymphocytes using a synthetic 16 mer minor bcr-abl peptide. Leuk Res. 2003;27:253-7.
3. Hagihara,M., Kato.S., et al. : Increased frequency of CD3/8/56-positive umbilical cord blood T lymphocytes after allo-priming in vitro. Ann Hematol. 2003.82:166-7.
4. Gansuvd,B., Kato,S. et al. : Umbilical cord blood dendritic cells are a rich source of soluble HLA-DR: synergistic effect of exosomes and dendritic cells on auto logous or allogeneic T-Cell prolife ration. Hum Immunol. 2003;64:427-39.
5. Inoue,H., Kato,S., et al : The kinetics of immune reconstitution after cord blood transplantation and selected CD34+ stem cell transplantation in children: comparison with bone marrow transplantation . Int J Hematol. 2003;77:399-407.
6. Ueda, Y., Kato,S., et al. : The effects of alphaGalCer-induced TCR Valpha24 Vbeta 11(+) natural killer T cells on NK cell cytotoxicity in umbilical cord blood. Cancer Immunol Immunother. 2003;52:625-31.
7. Yoshida,F., Kato,S., et al. : Complete resolution of severe chronic active Epstein-Barr virus infection by cultured, activated donor T lymphocyte infusion after nonmyeloablative stem cells allografting. Bone Marrow Transplant. 2003;32:107-10.
8. Hagihara.M., Kato,S., et al. : Clinical effects of infusing anti-Epstein-Barr virus (EBV)- specific cyto toxic T-lymphocytes into patients with severe chronic active EBV infection. Int J Hematol.2003;78:62-8.
9. Tsuboi,K., Kato,S., et al. : Multivariate analysis of risk factors for hemorrhagic cystitis after hematopoietic stem cell transplantation. Bone Marrow Transplant. 2003;32:903-7.
10. Ueda,Y., Kato,S., et al : Frequencies of dendritic cells (myeloid DC and plasmacytoid DC) and their ratio

reduced in pregnant women: comparison with umbilical cord blood and normal healthy adults. Hum Immunol.2003;64:1141-51.

1 1．Muguruma,Y., Kato,S., et al. : In vivo and in vitro differentiation of myocytes from human bone marrow-derived multipotent pregenitor cells. Exp Hematol. 2003;31:1323-30.

1 2．Yasuda,Y., Yabe,H., Kato,S., et al. : Comparison of PCR-amplified JC virus control region sequences from multiple brain regions in PML. Neurology. 2003,Dec,9;61(11):1617-9.

1 3．Tazume,K., Hagihara,M., Kato,S., et al. : Induction of cytomegalovirus-specific CD4+ cytotoxic T lymphocytes from seropositive or negative healthy subjects or stem cell transplant recipients. Exp Hematol. 2004,Jan;32(1):95-103.

加我牧子

1．Kaga,K., Kaga,M., Tamai,F., Shindo,M. : Auditory agnosia in children after herpes encephalitis. Acta Otolaryngol,123:232-235,2003

2．Hatori,T., Inagaki,M., Shirane,S., Kaga,M. : Developmental changes of auditory P300; Difference between two stimuli conditions, non-verbal sound and verbal sound. Seisin Hoken Kenkyu,49:159-167,2003

3．田中恭子、稲垣真澄、加我牧子：発達障害のスクリーニングと早期発見 知的障害の子ども。小児看護、26：1637-1641,2003.

4．稲垣真澄、白根聖子、加我牧子：ＡＤ/ＨＤ児の高次脳機能評価：視覚性弁別課題による検討。臨床脳波、45:767-772,2003

5．白根聖子、稲垣真澄、堀口寿広、中村雅子、佐々木匡子、加我牧子：副腎白質ジストロフィー症における両耳分離聴能検査（Dichotic Listening Test）異常。脳と発達、2004、印刷中

佐々木秀直

1．佐々木秀直，田代邦雄：協調運動の診かた．Clinical Neuroscience 21 (3)：288-289, 2003

2．佐々木秀直，矢部一郎，田代邦雄：脊髄小脳変性症の臨書像と鑑別診断．日本医事新報 4119：16-24, 2003

3．矢部一郎，佐々木秀直，田代邦雄：Spinocerebellar ataxia type 6 (SCA6) の臨床像についての再考．脳神経 55：299-306, 2003

4．矢部一郎，佐々木秀直：フリードライヒ病． 総合リハビリテーション 31:445-450, 2003

5．Sasaki H, Yabe I, Tashiro K : The hereditary spinocerebellar ataxias in Japan. Cytogenetic and Genome Research100:198-205, 2003

6．Yabe I, Sasaki H, Takeichi N, Takei A, Hamada T, Fukushima K, Tashiro K : Positional Vertigo and Macroscopic DownbeatPositioning Nystagmus in Spinocerebellar Ataxia Type 6(SCA6). J Neurol 250：440-443, 2003

7．Li M, Ishikawa K, Toru S, Tomimitsu H, Takashima M, Goto J, Takiyama Y, Sasaki H, Imoto I, Inazawa J, Toda T, Kanazawa I, Mizusawa H : Physical map and haplotype analysis of 16q-linked autosomal dominant cerebellarataxia (ADCA) type III in Japan. J Hum Genet , 48:111-118, 2003

8．Yabe I, Sasaki H, Chen H-H, Raskind WH, Bird TD, Yamashita I, Tsuji S, Tashiro K : Spinocerebellar ataxiatype 14 caused by a mutation in protein kinase C gamma. Arch Neurol 60:1749-1751, 2003

水澤英洋

1．Miura,Y., Misawa,N., Inagaki,Y., Yamamoto,N., Ito,M., Yagita,H., Okumura,K., Mizusawa,H., Koyanagi,Y. : TNF-related apoptososis-inducing ligand on HIV-1-infected macrophages induces neuronal apoptosis in murine model of HIV encephalopathy.

PNAS, 100:2777-2782, 2003
2. Kubodera, T., Yokota, T., Ohwada, K., Ishikawa, K., Miura, H., Matsuoka, T., Mizusawa, H. : Proteolytic cleavage and cellular toxicity of the human α1A calcium channel in spinocerebellar ataxia type 6 (SCA6). Neuroscience Letters, 341:74-78, 2003.
3. Furukawa, Y., Hashimoto, N., Yamakuni, T., Ishida, Y., Kato, C., Ogashiwa, M., Kobayashi, M., Kobayashi, T., Nonaka, I., Mizusawa, H., Song, S. :Down-regulation of an ankyrin repeat-containinig protein, V-1, during skeletal muscle differentiation and its re-expression in the regenerative process of muscular dystrophy. Neuromuscular Disorders, 13:32-41, 2003.
4. Kanda, T., Yamawaki ,M., Mizusawa, H., : Sera from Guillain-Barre patients enhance leakage in blood-nerve barrier model. Neurology, 60:301-306, 2003
5. Li, M., Ishikawa, K., Toru, S., Tomimitsu, H., Takashima, M., Goto, J., Takiyama, Y., Sasaki, H., Imoto, I., Imazawa, J., Toda, T., Kanazawa, I., Mizusawa, H. : Physical map and haplotype analysis of 16q-linked autosomal dominant cerebellar ataxia(ACDA)type III in Japan. J Hum Genet, 48:111-113, 2003
6. Mitoma, H., Ishida, K., Ikeda, M., Mizusawa, H. : Dual impariment of GABAA- and GABAB-receptor-mediated synaptic responses by autoantibodies to glutamic acidic decarboxylase. J Neurol Sci, 208:51-56, 2003
7. Owada, K., Uchihara, T., Ishida, K., Mizusawa, H., Watabiki, S., Tsuchiya, K. : Motor weakness andd cerebellar ataxa in Sjogren syndrome-indentification of antineuronal antibody: a case report. J Neurol Sci, 197:79-84, 2002.
8. Miura, Y., Kobayashi, Y., Mizusawa, H. : TNF-related apoptosis-inducing ligand(TRAIL) induces neuronal apoptosis in HIV-encephalopathy. J Med Dent Sci, 50:17-25, 2003.
9. Ishibashi, S., Kuroiwa, T., Endo, S., Okeda, R., Mizusawa, H. : Neurological dysfunction versus regional infarction volume after focal ischemia in mongolian gerbils. Stroke, 34:1501-1506, 2003.
10. Ishibashi, S., Yokota, T., Shiojiri, T., Matunaga, T., Tanaka, H., Nishina, K., Hirota, H., Inaba, A., Yamada, M., Kanda, T., Mizusawa, H. : Reversible acute axonal polyneuropathy associated with Wernicke-Korsakoff syndrome: impaired physiological nerve conduction due to thiamine deficiency? J Neurol Neurosurg Psychiatry, 74(5):674-676, 2003
11. Yamada, M., Sodeyama, N., Itoh, Y., Takahashi, A., Matsushita, M., Mizusawa, H : Association of neprilysin polymorphism with cerebral amyloid angiopathy. J Neurol Neurosurg Psychiatry, 74(6):749-751, 2003
12. Moinul Hossain AKM, Murata, T., Zhang, L., Taura, S., Saitoh, Y., Mizusawa, H., Oda, K., Matsushita, E., Okubo, Y., Shibuya, H. : Brain perfusion SPECT in patients with corticobasal degeneration: analysis using statistical parametric mapping. Movement Disorders, 18(6):697-703, 2003
13. Yokota, T., Sakamoto, N., Enomoto, N., Tanabe, Y., Miyagishi, M., Maekawa, S., Yi, L., Kurosaki, M., Taira, K., Watanabe, M., Mizusawa, H : Inhibition of intracellular hepatitis C virus replication by synthetic and vector-derived small interfering RNAs. EMBO reports, 4(6):602-608, 2003
14. Mizusawa, H., Clark, HB., Koeppen, AF. : Spinocerebellar ataxias. (Olsson, Y., Dickson, D., Berberon, C., Hardy, JA., Hauw, JJ., Jellinger, K., Lantos, P., Mizuno, Y., Mizusawa, H., eds), Neurodegeneration: The Molecular Pathology of Dementia and Movement Disorders, International Society of Neuropathhology Press, Woerthsenee p242-256, 2003

金澤一郎

1. Yazawa,I., Hazeki,N., Nakase, H., Kanazawa,I., Tanaka,M. : Histone H3 is aberrantry phosphorylated in

glutamine-repeat diseases. Biochemical Biophisical Research Communications,302:144-149,2003

2. Okabe,S., Ugawa,Y., Kanazawa,I. : 0.2-Hz retitive transcranial magnetic stimulation has no add-on effects as compared to a realistio sham stimulation in Parkinson's disease. Movement Disorders,18(4):382-388,2003
3. Kawahara,Y., Kwak,S., Sun,H., Ito,K., Hashida,H., Aizawa,H., Jeong,SY., Kanazawa,I. : Human spinal motoneurons express low relative sbundance of GluR2 mRNA: an implication for excitotoxity in ALS. Journal of Neurochemistry,85:680-689,2003
4. Takahashi,Y., Jeong,SY., Ogata,K., Goto,J., Hashida,H., Ishihara,K., Uchiyama,Y., Kanazawa,I. : Human skeletal muscle calcium channel α1S is expressed in the basal ganglia: distinctive expression pattern among L-type Ca2+ channels. Neuroscience research,45:129-137,2003
5. Ugawa,Y., Hanajima,R., Terao,Y., Kanazawa,I. : Exaggerated 16-20 Hz motor cortical oscillation In patients with positive or negative myoclonus. Clinical Neurophysiology, 114:1278-1284,2003
6. Kawahara,Y., Ito,K., Sun,H., Kanazawa,I., Kwak,S. : Low editiing efficiency of GluR2 mRNA is associated with a low relative abundance of ADAR2 mRNA in white matter of normal human brain. European J Neurosci.,18:23-33,2003
7. Hanajima,R., Furubayashi,T., Kobayasshi,N., Shiio,Y., Olabe,S., Kanazawa,I., Ugawa,Y. : Furthea evidence to support different mechanisms underlying intracortical inhibitation of the motor cortex. Exp Brain Res., 151:427-434,2003
8. Kaida,K., Kusunoki,S., Kamakura,K., Motoyoshi,K., Kanazawa,I. : GalNAc-GD1a in human peripheral nerve. 81:465-470.2003
9. Liu,W.,Goto,j.,Wang,YL.,Murata,M.,Wada,K., : Specific inhibition of Huntington's disease gene expression by siRNAs in cultured cells. Proc Japan Aacd SerB.,79:293-298,2003
10. Iwata,A., Maruyama,M., Akagi,T., Hashikawa,T., Kanazawa,I., Tsuji,S., Nukina,N. : Alpha-synuclein degradation by sarine protease neurosin: implication for pathogenesis of synuclainopathias. Human Molecular Genetics,12(20):2625-2635,2004

服部孝道

1. Ito,S., Kawahara,S., Sakakibara,R., Oki,T., Arai,H., Oda,S., Hattori,T. : Combined treatment with LDL-apheresis,chenodeoxycholic acid and HMG-CoA reductase inhibitor for cerebrotendinous xantomatosis. J Neurol Sci.,216:179-182,2003
2. Kanai,K., Kuwabara,S., Arai,K., Sung,JY., Ogawara,K., Hattori,Y. : Muscle cramp in Machado-Joseph disease.:Altered motor axonal excitability properties and mexiletine treatment. Brain, 126:965-973,2003
3. Uchihara,T., Tanaka,J., Funata,N., Arai,K., Hattori,T. : Influences of intanuclear inclusion on nuclear sizw-morphometric study on pontine neurons of neuronal intranuclear inclusion disease cases. Acta Neuropathol., 105:103-108.2003
4. Yamaguchi,M., Arai,K., Asahina,M., Hattori,T. : Larygeal stridor in maltiple system atrophy.Eur Neurol,49:154-159,2003
5. Asahina,M., Kikkawa,Y., Uchiyama,T., Yoshiyama,M., Yamanishi,T., Hattori,T. : Cataneous sympathetic function in patients with multiple system atrophy,. Clin Auton Res.,13(2):91-93,2003
6. Sakakibara,R., Matsuda,S., Uchiyama,T., Yoshiyama,M., Yamanishi,T., Hattori,T. : The effect on intranasal desmopressionon noctumal waking in urination in multiple system atrophy patients with noctumal polyuria. Clin Auton Res.,1382):106-108,2003
7. 内山智之、榊原隆次、服部孝道：各種神経疾患における排尿障害。臨床と研究、80(8):1442-1446,2003
8. 内山智之、榊原隆次、服部孝道：中枢疾患に伴う尿失禁の。Urology View,1(5):41-47,2003
9. 服部孝道：脊髄小脳変性症。山口徹、北原光夫総編集、「今日の治療指針」2004年度版 p.638,（医学書院、東京）2004

祖父江　元

1．Katsuno,M., Adachi,H., Sobue,G : Sweat relief for Huntington's disease. Nat Med., in press,2004

2．Katsuno,M., Adachi,H., Tanaka,F., Sobue,G : Spinal and Bulbar musculea atrophy(SBMA): Ligand-degendent pathogenesis and therapeutic perspective. J Mel Med., in press,2004

3．Koike,H., Misu,K., Sugiura,M., Iijima,M., Mori,K., Yamamoto,M., Hattori,M., Mukai,E., Ando,Y., Ikeda,S., Sobue,G : Pathologic differences between early-and late-onset type 1(TTR Met30) familial amyloid polyneuropathy. Neurology, in press,2004

4．Takeuchi,H.,Niwa,j.,Hishikawa,N.,Ishigaki,S.,Tanaka,F.,Doyu,M., : Dorfin prevents cell death by reducing mitchondrial localizing mutant superoxide dismutase 1 in a neuronal cell model of familial amyotrophic lateral sclerosis.J Neurochem., in press,2004

5．Watanabe,H., Fukatsu,H., Katsuno,M., Sugiura,M., Hamada,K., Okada,Y., Hirayama,M., Ishigaaki,T., Sobue,G : Multiple regional 1H-MR spectoroscopy in multiple system arophy:NNA/Cr reduction in pontine base as a valuable diagnotic marker. J Neurol Neurosurg Psychiatry,75:103-109,2004

6．Nodera,H., Bostock,H., Kuwabara,S., Sakamoto,T., Asanuma,K., Jia-Ying,S., Ogawara,K., Hattori,N., Hirayama,M., Sobue,G, Kaji,R. : Narve excitabilitypropertion in Charcot-Marie-Tooth disease type 1A. Brain,127:203-211,2004

7．Ito,T., Niwa,J., Hishikawa,N., Ishigaki,S., Doyu,M., Sobue,G : Dorfin localizes to Lawy bodies and ubiquitilates synphilin-1. J boil Chem.,278:29106-29114,2003

8．Ishihara,K., Yamagishi,N., Saito,Y., Adachi,H., kobayashi,Y., Sobue,G, Otsuka,K., Hatayama,T. : Hsp105a suppresses the aggregation of truncated androgen receptor with expanded CAG repeats and celltoxicity. J Biol Chem.,278:25143-25150,2003

9．Hishikawa, n., Niwa,J., Doyu,M., Ito,T., Ishigaaki,S., Hashizume,Y., Sobue,G : Dorfin localizes to the ubiquitylated inclusions in Parkinson's disease, dementia with Lewy bodies, multiple system atrophy,and amyotrophic lateral sclerosis. Am. J Pathol.,163:609-619,2003

10．Hattori,N., Yamamoto,M., Yoshihara,T., Koike,M., Nakagawa,N., Yoshikaawa,H., Ohnishi,A., Hayasaka,K., Onodera,O., Baba,M., Yasuda,H., Saito,T., Nakashima,K., Kira,J., Kaji,R., Oka,N., Sobue,G and the Study Group for Hereditary Neuropathy in Japan : Demyelinating and axonal features of Charcot-Marie-Tooth disease with mutations of myelin-related proteins (PMP22,MPZ, and Cx32): a clinicopathological study of 205 Japanese patients. Brain,126:134-151m2003

11．Hishikawa,N., Hashizume,Y., Yoshida,M., Sobue,G : Clinical and neuropathological cortelates of Lewy body disease. Acta Neuropathol.,105:341-350,2003

12．Hamada,K., Hirayama,M., Watanabe,H., Kobayashi,R., Ito,H., Ioda,T., Koike,Y., Sobue,G : Onset age and severity impairment are associated with reduction of myocardial 1231-MIBG uptake in Parkinson's disease. J Neurol Neurosurg Psychiatry, 74:423-425,2003

13．Adachi,H., Katsuno,M., Minamiyama,M., Sang, C., Pagoulatous,G, Angelidis,C., Kusakabe,M., Yoshida,A., Kobayashi,Y., Doyu,M., Sobue,G : Heat shock protein 70 chaperone overexpression ameliorates phenotypes of the spinal and bular muscular atrophy transgenic mouse model by reducing nuclear-localized mutant androgen receptor protein. J Neurosci.,23:2208-2211,2003

14．Abe,Y., Kachi,T., Arahata,Y., Yamada,T., Washimi,Y., Iwai,K., Ito,K.,Yanagisawa,N., Sobue,G : Occipital hypoperfusion in Parkinsom's disease without dementia correlation to impaired cortical visual processing. J Neurol Neurosurg Psychiatry, 74:419-422,2003

15．Koike,H., Iijima,M., Sugiura,M., Mori,K., Hattori,N., Ito,H., Hirayama,M., Sobue,G : Alcoholic neuropathy is

clinicopathologically distant from thisminc-deficiency neuropathy. AnnNeurol.,54:19-29,2003
16. Katsuno,M., Adachi,H., Doyu,M., Minamiyama,M., Sang,C., Kobayashi,Y., Inukai,A., Sobue,G : Leuprotein rescue polyglutamine-dependent phenotypes ina transgenic mouse model of spinal and bulbar ,uscular atrophy. Nature Medicine,9:768-773,2003
17. Wada,M., Kimura,M., Daimon,M., Kurita,K., Kato,T., Johmura,Y., Johkura,K., Kuroiwa,Y., Sobue,G : As usual phenotype of McLeod syndrome with late onset axonal neuropathy. J Neurol Neurosurg Psychiatry, 74:1697-1698,2003
18. Katsuno,M., Adachi,H., Inukai,A., Sobue,G : Trangenic mouse models of spinal bulbar meucular strophy(SBMA). Cytogenet Genome Res.,100:243-251,2003
19. Mori,K., Iijima,M., Sugiura,M., Koike,H., Hattori,N., Ito,H., Hirayama,M., Sobue,G : Sjogren's syndrome associated painful sensory neuropathy without sensory atxia. J Neurol Neurosurg Psychiatry, 74:1230-1322,2003

小野寺　理

1. Sano, Y., Date, H., Igarashi, S., Onodera, O., Oyake, M., Takahashi, T.,Hayashi, S., Morimatsu, M., Takahashi, H., Makifuchi, T., et al. (2004).Aprataxin, the causative protein for EAOH is a nuclear protein with a potential role as a DNA repair protein. Ann Neurol 55, 241-249.
2. Sekijima, Y., Hashimoto, T., Onodera, O., Date, H., Okano, T., Naito, K.,Tsuji, S., and Ikeda, S. (2003). Severe generalized dystonia as apresentation of a patient with apraxin gene mutation. Mov Disord 18,1198-1200.
3. Toyoshima, Y., Yamada, M., Onodera, O., Shimohata, M., Inenaga, C., Fujita,N., Morita, M., Tsuji, S., and Takahashi, H. (2004). SCA17 homozygoteshowing Huntington's disease-like phenotype. Ann Neurol 55, 281-286.

西澤正豊

1. Nishizawa,M., et.al : Identification of a SACS gene missense mutation in ARSACS. Neurology,52(19) : 107-109,2004
2. 西澤正豊：最近の研究情報、特集「脊髄小脳変性症の最新情報」。「難病と在宅ケア」9(2)：7-9,2003
3. 西澤正豊他：遺伝性けい性対麻痺の遺伝子的研究。「脳神経」55(9)：757-763,2003
4. 西澤正豊他：脳梁ひ薄化を伴う遺伝性けい性対麻痺。「脳神経」55(9)：765-770,2003
5. 西澤正豊他：脊髄小脳変性症の治療の試み。「BIO Clinica」18(12)：24(1064)-27(1067),2003

黒岩義之

1. Wang L, Kuroiwa Y, Kamitani T, Li M, wang J ; Do P1 and N1 evoked by the ERP task reflect primary visual processing in Parkinson's disease? Documenta Ophthalmologica,2001 102:83-93.
2. Omoto S, Kuroiwa Y, Li M, Doi H, Shimamura M, Koyana S, Segawa F, Suzuki Y: Modulation of event-related potentials in normal human subjects by visual divided attention to spatial and color factors. Neurosci Lett 2001 311:198-202.
3. Yamazaki T, Kamijo K, Kinuya T, Takaki Y, Kuroiwa Y: Multiple dipole analysis of visual event-related potential during oddball paradigm with silent counting. Brain Topography,2001 13:161-168.
4. Yamazaki T, Kamijo K, Kinuya T, Takaki Y, Kuroiwa Y, Ochi A, Otsubo H: PC-based multiple equivalent current dipole source localization system and its applications. Res Adv in Biomedical Eng 2001 2:97-109.
5. Johkura K, Komiyama A, Kuroiwa Y: Sustained downgaze in coma after cardiac arrest. J Neurol Neurosurg Psychiatry 2001 71:278-279.

6．Nishiyama T, Johkura K, Johmura Y, Momoo T, Yamada H, Kuroiwa Y: Encephalitis with MRI abnormality as a manifestation of central nervous system involvement of adult T cell leukemic lymphoma. Eur Neurology 2001,146:218-220.
7．Susuki K, Johkura K, Yuki N, Hasegawa O, Kuroiwa Y: Rapid resolution of nerve conduction blocks after plasmapheresis in Guillain-Barre syndrome associated with anti-GM1b IgG antibody. J Neurol 2001 248:148-150.
7．Li M, Kuroiwa Y, Wang L, Kamitani T, Takahashi T, Suzuki Y, Omoto S: Early sensory information processes are enhanced on visual oddball and S1-S2 tasks in Parkinson's disease. A visual event-related potentials study. Parkinsonism and Related Disorders. 2002, 9:329-340.
8．Yamada H, Momose T, Okada M, Kuroiwa Y: Anticholinergic drugs:response of parkinsonism not responsive to levodopa. J Neurol Neurosurg Psychiatry 2002 72:111-113.
9．Kamitani T, Kuroiwa Y, Wang L, Li M, Suzuki Y, Takahashi T, Ikegami T, Matsubara S: Visual event related potential changes in two subtypes of multiple system atrophy, MSA-C and MSA-P. J Neurol 2002 249: 975—982.
10．Yamazaki T, Kamijo K, Kinuya T, Takaki Y, Kuroiwa Y: Multiple dipole analysis of visual event-related potentials during oddball paradigm International Congress Series(Excerpta Medica). 2002, 1232:863-870.
11．Kamijo K, Yamazaki T, Kinuya T, Takaki Y, Kuroiwa Y :Visual event-related potentials during movement imagery and the dipole analysis. Brain Topography 2002 14:279-292.
12．Li M, Kuroiwa Y, Omoto S, Hotta S, Suzuki Y, Kamitani T, Koyano S, Segawa F:: The effect of stimulus-onset asynchrony on human visual event-related potentials during simple and choice reaction paradigms under constant or randam conditions. Neuroscience Letters, 2003, 345:109-112.
13．Kamitani T, Kuroiwa Y, Li M, Ikegami T, Matsubara S: Relationship between cerebellar size and variation of the reaction time during a visual cognitive task in normal subjects. J Neurol 2003 250: 1001—1003.
14．Kamitani T, Kuroiwa Y, Wang L, Li M, Ikegami T, Matsubara S: Event-related potentials during visual S1-S2 paradigm in multiple system atrophy: relation to morphologic changes on brain MRI measurement.changes Parkinsonism and Related Disorders. 2003, 10:93-100.
15．Yamada H, Dezawa M, Kuroiwa Y: Transfer of the von Hippel-Lindau gene to neuronal progenitor cells in treatment for Parkinson's disease. Ann Neurol, 2003, 54:352-359.
16．Shimamura M, Momose T, Takahashi T, Uchida T, Kuroiwa Y: Measurement using 18F-dopa and 11 C-N-methylspiperone PET of striatal dopaminergic function in corticobasal degeneration: comparison with progressive supranuclear palsy and normal controls. (in submission)

中島健二

1．村上丈伸、中曽一裕、佐久間研司、楠見公義、中島健二：フェニトイン長期内服中に認めた小脳半球外側に限局した萎縮性病変。神経内科 58：338-339, 2003
2．Yasuhiro Watanabe, Yasutaka Shimizu, Katsuya Urakami, Eiji Matsushima, Kenji Nakashima, : Vertical ophthalmoplegia in a demented patient with striatopallidodentate calcification. Psychiat Clin Neurosci 57: 447-450.
3．Yosuke Wakutani, Hisanori Kowa, Masayoshi Kusumi, Kazuhiro Nakaso, Kenji Isoe- Wada, Hidetaka Yano, Katsuya Urakami, Takao Takeshima, Kenji Nakashima, : A haplotype of the methylenetetrahydrofolate reductase gene is protective against late-onset Alzheimer's disease. Neurobiol Aging 25: 291-294,2004

貫名信行

1. Tanaka, M., Machida, Y., Niu, S., Ikeda, T., Jana, N. R., Doi, H., Kurosawa, M., Nekooki,M., and Nukina, N. (2004). Trehalose effectively alleviates polyglutamine· mediated pathology in a transgenic mouse model of Huntington's disease. Nat Med 10, 148-154.
2. Zemskov, E. A., Jana, N. R., Kurosawa, M., Miyazaki, H., Sakamoto, N., Nekooki, M., and Nukina, N. (2003). Pro-apoptotic protein kinase C delta is associated with intranuclear inclusions in a transgenic model of Huntington's disease. J Neurochem 87, 395-406.
3. Zemskov, E. A., and Nukina, N. (2003). Impaired degradation of PKCalpha by proteasome in a cellular model of Huntington's disease. Neuroreport 14, 1435-1438.
4. Iwata, A., Maruyama, M., Akagi, T., Hashikawa, T., Kanazawa, I., Tsuji, S., and Nukina, N. (2003). Alpha-synuclein degradation by serine protease neurosin: implication for pathogenesis of synucleinopathies. Hum Mol Genet 12, 2625-2635.
5. Wen, F. C., Li, Y. H., Tsai, H. F., Lin, C. H., Li, C., Liu, C. S., Lii, C. K., Nukina, N., and Hsieh, M. (2003). Down-regulation of heat shock protein 27 in neuronal cells and non-neuronal cells expressing mutant ataxin-3. FEBS Lett 546, 307-314.
6. Tanaka, M., Machida, Y., Nishikawa, Y., Akagi, T., Hashikawa, T., Fujisawa, T., and Nukina, N. (2003). Expansion of polyglutamine induces the formation of quasi-aggregate in the early stage of protein fibrillization. J Biol Chem 278, 34717-34724.
7. Lee, J. A., Lim, C. S., Lee, S. H., Kim, H., Nukina, N., and Kaang, B. K. (2003). Aggregate formation and the impairment of long-term synaptic facilitation by ectopic expression of mutant huntingtin in Aplysia neurons. J Neurochem 85, 160-169.

山田光則

1. Toyoshima,Y., Yamada,M., Onodera,O.et al. : SCA17 homozygote showingHuntington's disease-like phenotype. Annalz of Neurology,55:281-286,2004
2. 山田光則、高橋均：ポリグルタミン病の神経病理。脳と神経、55:921-9312003
3. 山田光則、高橋均：ポリグルタミン病の病理。大脳皮質における diffuse nuclear staining(1C2)。Cognition and Dementia,2:23-27,2003

垣塚　彰

1. Matsumoto, M., Yada,M., Hatakeyama,H., Ishimoto,H., Tanimura,T., Tsuji, S., Kakizuka, A., Kitagawa, M., Nakayama,K.I.. : Molecular clearance of ataxin-3 is regulated by a mammalian E4. EMBOJ, in press, 2004
2. Kobayashi,T., & Kakizuka,A. : Molecular analyses of Machado-Joseph disease. Cytogenet. Genome Res. 100:261-275,2003
3. Kimura.Y., & Kakizuka,A. : Polyglutamine diseases and molecular chaperones. IUBMB Life, 55:337-345,2003
4. Mizuno,Y., Hori,S., Kakizuka,A., & Okamoto,K. : Vacuole-creating protein in neuro-degenerative diseases. Neurosci.Lett.,343:77-80,2003

川上秀史

1. Oda,M., Maruyama,H., Komure,O., Morino,H., Terasawa,H., Izumi,Y., Imamura,T., Yasuda,M., Ichikawa,K., Ogawa,M., Mastumoto,M., Kawasaki,H. : Expansion of 44-47 CAG/CAA repeats in the TATA-binding protein gene may not be fully penetrat in spinocerebellar atxia17. Archives of Neurology, in press
2. Terasaawa,H., Oda,M., Morita,H., Miyachi,T., Izumi,Y., Maruyama,H., Matsumoto,M., Kawakami,H. : Molecular basis of prevalence and founder a¥effect for Japanese SCA6 population. Neuroscience Lett., in press.

3．Honji,K., Ohshita,H., Kawakami,H., Naka,H., Imon,Y., Maruyama,H., Mimori,Y., Matsumoto,M. : Quantitative assessment of cerebral blood flow in genetically confirmed spinocerebellar ataxia type6. Archives of Neurology, in press

久野貞子

1．Izumi,Y., Maruyama,H., Oda,M., Morino,H., Okada,T., Ito,H., Sasaki,I., Tanaka,H., Komure,O., Udaka,F., Nakamura,S., Kawakami,H. : SCA8 repeat expansion: large CTA/CTG repeat alleles are more common in ataxic patients, including those with SCA6. Am J Hum Genet,72:704-9,2003

武田　篤

1．Nunomura A., Chiba S, Kosaka K, Takeda A, Castellani RJ, Smith MA., Perry G, Neuronal RNA oxidation is a prominent feature of dementia with Lewy bodies. NeuroReport 13 : 2035-2039, 2002.
2．Tateyama M., Takeda A., Onodera Y., Matsuzaki M., Hasegawa T., Nunomura A., Hirai K., Perry G, Smith MA., Itoyama Y., Oxidative stress and predominant Abeta42(43) deposition in myopathies with rimmed vacuoles. Acta Neuropathologica 105 : 581-585, 2003.
3．Hasegawa T., Matsuzaki M., Takeda A., Kikuchi A., Furukawa K., Shibahara S., Itoyama Y., Increased dopamine and its metabolites in SH-SY5Y neuroblastoma cells that express tyrosinase. J. Neurochem. 87 : 470-475, 2003.
4．Masaki T., Matsushita S., Arai H., Takeda A., Itoyama Y., Mochizuki H., Kamakura K., Ohara S., Higuchi S., Association between a polymorphism of BDNF gene and sporadic Parkinson's disease. Ann. Neurol. 54 : 276-277, 2003.
5．Tateyama M., Saito N., Fujihara K., Shiga Y., Takeda A., Narikawa K., Hasegawa T., Taguchi Y., Sakuma R., *Onodera Y., Ohnuma A., Tobita M., toyama Y., Familial inclusion body myositis: a report on two Japanese sisters.* Internal Med. 42 : 1035-1038, 2003.
6．Kikuchi A., Takeda A., Fujihara K., Kimpara T., Shiga Y., Tanji H., Nagai M., Ichinose H., Urano F., Okamura N., Arai H., Itoyama Y., The Arg (184) His mutant GTP cyclohydrase I, causing recessive hyperphenylalanemia, is responsible for dopa-responsive dystonia: a case. Movement Disorders (in press), 2004.
7．Matsuzaki M., Hasegawa T., Takeda A., Kikuchi A., Furukawa K., Kato Y., Itoyama Y., Histochemical features of stress-induced aggregates in α -synuclein overexpressing cells. Brain Research (in press), 2004.

中島　孝

1．中島　隆：難病の基礎知識I,II, 難病ホームヘルパーテキスト第6版2004（印刷中）
2．川上英孝、中島孝：神経・筋一変性疾患、認定医・専門医のための内科学レビュー2004、総合医学社（印刷中）
3．中島　孝：ALS早期診断のための新しい展開 - 脳SPECT画像での検討、神経内科2004（印刷中）
4．中島　孝：緩和ケアとはなにか、難病と在宅ケア、9:7-11,2003
5．中島　孝：筋萎縮性側索硬化症患者に対する生活の質（QoL）向上への取り組み、神経治療学、20:139-147,2003
6．Nemoto H, Toda H, Nakajima T, Hosokawa S, Okada Y, Ymamaoto K, Horicuchi R, Endo K, Masahiko M, Goto, F : Fluvoxamine modulates pain sensation and affective processing of pain in human brain, NeuroReport、14:791-797,2003

永井義隆

1．NagaiY., Fujikake,N., Ohno,K., Higashiyama ,., Popiel,H.A., Rahadian,J., Yamaguchi,M., Strittmatter,W.J., Burke,

JR., Toda,T. : Prevention of polyglutamine oligomerization and neurodegeneration by the peptide inhibitor QBP 1 in Drosophia. Hum.Mol.Genet. 12:1253-1259,2003
2．永井義隆、戸田達史：神経変性疾患ポリグルタミン病に対する治療ペプチドQBP 1.バイオインダストリー、20:52-63,2003
3．永井義隆：ポリグルタミン鎖結合ペプチドQBP 1によるポリグルタミン病に対する分子治療法の確立。神経化学、42:443-456,2003

吉田邦広

1．Hashimoto,T., Sasaki,O., Yoshida,K., Takei,Y., Ikeda,S. : Periodic alternating nystagmus and rebound nystagmus in spinocerebellar ataxia 6. Mov Disord,18:1201-1204,2003.

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