

雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
Kashiwayama Y., Asahina K., Morita M., and Imanaka T.	Hydrophobic regions adjacent to transmembrane domain in 1 and 5 are important for the targeting of the 70-kDa peroxisomal membrane protein.	J. Biol. Chem.	Vol. 282, No. 46	33831-33844	2007
Takahashi N., Morita M., Maeda T., Harayama Y., Shimozawa N., Suzuki Y., Furuya H., Sato R., Kashiwayama Y., and Imanaka T.	Adrenoleukodystrophy: subcellular localization and degradation of adrenoleukodystrophy protein (ALDP/ABCD1) with naturally occurring missense mutations	J. Neurochem.	Vol. 101, No. 6	1632-1643	2007
Takahashi N., Morita M., and Imanaka T.	Adrenoleukodystrophy: Structure and function of ALDP, and intracellular behavior of mutant ALDP with naturally occurring missense mutations.	Yakugaku Zasshi	Vol. 127, No. 1	163-172	2007
Morita M.	Adrenoleukodystrophy: Molecular pathogenesis and development of therapeutic agents.	Yakugaku Zasshi	Vol. 127, No. 7	1059-1064	2007
今中常雄, 柏山恭範, 守田雅志	ペルオキシソームABCタンパク質と副腎白質ジストロフィー.	最新医学	Vol. 62, No. 11	68-75	2007
Toro A., Arredondo C., Cordova G., Araya C., Palacios J. L., Venegas A., Morita, M., Imanaka T., and Santos M. J.	Evaluation of the role of the endoplasmic reticulum-Golgi transit in the biogenesis of peroxisomal membrane proteins in wild type and peroxisomal biogenesis mutant CHO cells.	Biol. Res.	Vol. 40, No. 2	231-249	2007

## 辻 省次

1. Hara K, Momose Y, Tokiguchi S, Shimohata M, Terajima K, Onodera O, Kakita A, Yamada M, Takahashi H, Hirasawa M, Mizuno Y, Ogata K, Goto J, Kanazawa I, Nishizawa M, Tsuji S. Multiplex families with multiple system atrophy. Arch Neurol. 64:545-551, 2007
2. Tsuji S, Onodera O, Goto J, Nishizawa M; Sporadic ataxias in Japan - a population-based epidemiological study. Cerebellum. 2007 Aug 7;:1-9 [Epub ahead of print]

## 加我 牧子

### 雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
白根聖子, 稲垣真澄, 堀口寿広, 中村雅子, 佐々木匡子, <u>加我牧子</u>	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	36	311-317	2004
Inagaki M, Kaga Y, <u>Kaga M</u> , Nihei K	Multimodal evoked potentials in patients with pediatric leukodystrophy.	Clinical Neurophysiology	59 suppl	251-263	2006
古島わかな, 稲垣真澄, 軍司敦子, <u>加我牧子</u> , 山崎広子, 堀口寿広	小児大脳型副腎白質ジストロフィーの超早期発症診断に関する研究: 視覚系心理検査および視覚誘発電位の有用性。	脳と発達	(in press)		2008

加藤 俊一

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
Kato K, Kanda Y, Eto T, Muta T, Gondo H, Taniguchi S, Shibuya T, Utsunomiya A, Kawase T, Kato S, Morishima Y, Kodera Y, Harada M	Allogeneic bone marrow transplantation from unrelated human T-cell leukemia virus-I-negative donors for adult T-cell leukemia/lymphoma: retrospective analysis of data from the Japan Marrow Donor Program.	Biol Blood Marrow Transplant.	13巻	90-9	2007年1月
Morishima Y, Yabe T, Matsuo K, Kashiwase K, Inoko H, Saji H, Yamamoto K, Maruya E, Akatsuka Y, Onizuka M, Sakamaki H, Sao H, Ogawa S, Kato S, Juji T, Sasazuki T, Kodera Y; Japan Marrow Donor Program.	Effects of HLA allele and killer immunoglobulin-like receptor ligand matching on clinical outcome in leukemia patients undergoing transplantation with T-cell-replete marrow from an unrelated donor.	Biol Blood Marrow Transplant.	13(3)	315-28	2007年3月
加藤 俊一	副腎皮質ジストロフィーにおける造血幹細胞移植	BRAIN and NERVE (医学書院)	第59巻4号	339-346	2007年4月
Ishiguro H, Yasuda Y, Tomita Y, Shinagawa T, Shimizu T, Morimoto T, Hattori K, Matsumoto M, Inoue H, Yabe H, Yabe M, Shinohara O, Kato S.	Gonadal shielding to irradiation is effective in protecting testicular growth and function in long-term survivors of bone marrow transplantation during childhood or adolescence.	Bone Marrow Transplant.	39(8)	483-90	2007年4月
Ozawa S, Nakaseko C, Nishimura M, Maruta A, Cho R, Ohwada C, Sakamaki H, Sao H, Mori S, Okamoto S, Miyamura K, Kato S, Kawase T, Morishima Y, Kodera Y;	Chronic graft-versus-host disease after allogeneic bone marrow transplantation from an unrelated donor: incidence, risk factors and association with relapse. A report from the Japan Marrow Donor Program.	British Journal of Hematology	137巻	142-51	2007年4月
加藤俊一	小児に対する造血幹細胞移植の歩みと将来	血液・腫瘍科 (科学評論社)	第55巻特別増刊号	672-678	2007年8月
Y.Nakamura, T.Yahata, Y.Muguruma, T.Uno, T.Sato, H.Matsuzawa, S.Kato, Y.Shirasugi, T.Hotta, K.Ando	Angiopoietin-1 supports induction of hematopoietic activity in human CD34(-) bone marrow cells	Experimental Hematology	35	872-188	2007年8月
Masuda H, Kalka C, Takahashi T, Yoshida M, Wada M, Kobori M, Itoh R, Iwaguro H, Eguchi M, Iwami Y, Tanaka R, Nakagawa Y, Sugimoto A, Ninomiya S,	Estrogen-mediated endothelial progenitor cell biology and kinetics for physiological postnatal vasculogenesis.	Circulation Research	101巻	598-606	2007年9月
Takakazu Kawase, Yasuo Morishima, Keitaro Matsuo, Koichi Kashiwase, Hidetoshi Inoko, Hiroh Saji, Shunichi Kato, Takeo Juji, Yoshihisa Kodera,	High-risk HLA allele mismatch combinations responsible for severe acute graft-versus-host disease and implication for its molecular mechanism	Blood	Vol.110 Number7	235-224	2007年10月

Y. Atsuta, R. Suzuki, A. Yoshimi, H. Gondo, J. Tanaka, A. Hiraoka, K. Kato, K. Tabuchi, M. Tsuchida, Y. Morishima, M. Mitamura, K. Kawa, S. Kato, T. Nagamura,	Unification of Hematopoietic Stem Cell Transplantation Registries in Japan and Establishment of the TRUMP System	International Journal of Hematology (社)日本血	86号	269-274	2007年 10月
Yabe M, Ishiguro H, Yasuda Y, Takakura I, Matsuda S, Shimamura K, Kato S, Yabe	Fatal giant cell myocarditis after allogeneic bone marrow transplantation.	Bone Marrow Transplant.	4 1 卷	93-4	2008年 1月

## 加藤 剛二

Kobayashi R, Yabe H, Hara J, Morimoto A, Tsuchida M, Mugishima H, Ohara A, Tsukimoto I, Kato K, Kigasawa H, Tabuchi K, Nakahata T, Ohga S, Kojima S; Japan Childhood Aplastic Anemia Study Group.

Preceding immunosuppressive therapy with antithymocyte globulin and ciclosporin increases the incidence of graft rejection in children with aplastic anaemia who underwent allogeneic bone marrow transplantation from HLA-identical siblings.

Br J Haematol. 2006 Dec;135(5):693-6.

Yoshihara T, Okada K, Kobayashi M, Kikuta A, Kato K, Adachi N, Kikuchi A, Ishida H, Hirota Y, Kuroda H, Nagatoshi Y, Inukai T, Koike K, Kigasawa H, Yagasaki H, Tokuda K, Kishimoto T, Nakano T, Fujita N, Goto H, Nakazawa Y, Kanegane H, Matsuzaki A, Osugi Y, Hasegawa D, Uoshima N, Nakamura K, Tsuchida M, Tanaka R, Watanabe A, Yabe H.

Outcome of non-T-cell-depleted HLA-haploidentical hematopoietic stem cell transplantation from family donors in children and adolescents.

Int J Hematol. 2007 Apr;85(3):246-55.

## 鈴木康之

### 書籍

鈴木康之 : adrenoleukodystrophy (ALD). 小児科学第3版 医学書院 東京 2008 (in press)

### 雑誌

Takahashi N, Morita M, Maeda T, Harayama Y, Shimozawa N, Suzuki Y, Furuya H, Sato R, Kashiwayama Y, Imanaka T: Adrenoleukodystrophy: subcellular localization and degradation of adrenoleukodystrophy protein (ALDP/ABCD1) with naturally occurring missense mutations. J Neurochem 2007;101:1632-43

Izumi Kuratsubo, Yasuyuki Suzuki, Nobuyuki Shimozawa, Naomi Kondo. Parents of Childhood X-linked Adrenoleukodystrophy: High Risk for Depression and Neurosis. Brain & Development (2008 in press)

### パンフレット

副腎白質ジストロフィー早期診断のために

## 服部 孝道

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版
原著					
Shirai W, Ito S, Hattori T	Linear T2 Hyperintensity along the Medial Margin of the Globus Pallidus in Patients with Machado Joseph Disease and Parkinson Disease, and in Evakuating posterolateral linearization of the putaminal margin with magnetic resonance imaging to diagnose the Parkinson variant of multiple system atrophy	AJNR Am J Neuroradiol	28巻10号	1993-5	2007
Ito S, Shirai W, Hattori T	Skin vasodilator response to local heating in multiple system atrophy	Mov Disord	22巻4号	578-81	2007
Yamanaka Y, Asahina M, Mathias CJ, Akaogi Y, Koyama Y, Hattori T	The raphae magnus/pallidus regulates sweat secretion and skin vasodilation of the cat forepaw pad: a preliminary electrical stimulation study	Mov Disord	22巻14号	2405-8	2007
Asahina M, Sakakibara R, Liu Z, Ito T, Yamanaka Y, Nakazawa K, Shimizu E, Hattori T	Genetically confirmed Huntington's disease masquerading as motor neuron disease	Neurosci Lett	415巻3号	283-7	2007
短報					
Kanai K, Kuwabara S, Sawai S, Nakata M, Misawa S, Iose S, Hirano S, Kawaguchi N, Katayama K, Ning Y, Kanai K, Tomiyama H, Li Y, Funayama M, Yoshino H, Sato S, Asahina M, Kuwabara S, Takeda A, Hattori T, Mizuno Y, Hattori N	PARK9-linked parkinsonism in Eastern Asia: Mutation detection in ATP13A2 and clinical phenotype	Mov Disord	Epub ahead of print		2008
Sakakibara R, Yamaguchi T, Uchiyama T, Yamamoto T, Ito T, Liu Z, Odaka T, Yamaguchi C, Hattori T	Calcium polycarbophil improves constipation in primary autonomic failure and multiple system atrophy	Neurology	In press		2008
Kanai K, Sakakibara R, Uchiyama T, Liu Z, Yamamoto T, Ito T, Hirano S, Asahina M, Kuwabara S, Hattori T, Fukami G, Arai K, Yamaguchi C, Nomura F	Sporadic case of spinocerebellar ataxia type 17: treatment observation for managing urinary and psychotic symptoms	Mov Disord	22巻11号	1673-2	2007
Sakakibara S, Murakami E, Katagiri A, Hayakawa S, Uchiyama T, Yamamoto T, Hattori T	Moxibustion, an alternative therapy, ameliorated disturbed circadian rhythm of plasma arginine vasopressin and urine output in multiple system atrophy	Mov Disord	22巻3号	441-3	2007
Sakakibara S, Murakami E, Katagiri A, Hayakawa S, Uchiyama T, Yamamoto T, Hattori T	Moxibustion, an alternative therapy, ameliorated disturbed circadian rhythm of plasma arginine vasopressin and urine output in multiple system atrophy	Intern Med	46巻13号	1015-8	2007
総説					
榊原隆次、内山智之、服部孝道	中枢性排尿障害	Clinical Neuroscience	25巻4号	435-7	2007
山本達也、榊原隆次、服部孝道	パーキンソン病および変性性パーキンソン症候群における排尿障害	プラクティス	14巻4号	331-6	2007

## 中島 健二

### 論文発表

- Sakuma K, Adachi Y, Fukuda H, Kai T, Nakashima K. Triple stimulation technique in patients with spinocerebellar ataxia type 6. *Clin Neurophysiol.* 2005;116(11):2586-91.
- Fukuda H, Kusumi M, Nakashima K. Epidemiology of primary focal dystonias in the western area of Tottori prefecture in Japan: Comparison with prevalence evaluated in 1993. *Mov Disord.* 2006;21(9):1503-6.
- Hayashi M, Adachi Y, Mori M, Nakano T, Nakashima K. Clinical and genetic epidemiological study of 16q22.1-linked autosomal dominant cerebellar ataxia in western Japan. *Acta Neurol Scand.* 2007;116(2):123-7.
- 佐久間研司ほか、Triple stimulation techniqueを用いた脊髄小脳変性症、パーキンソン類縁疾患での皮質脊髄路機能の検討。臨床脳波50(1):27-31, 2008.

## 糸山 泰人

### 雑誌

- 1) Hasegawa T., Sugeno N., Takeda A., Matsuzaki-Kobayashi M., Kikuchi A., Furukawa K., Miyagi T., Itoyama Y., Role of Neu4L sialidase and its substrate ganglioside GD3 in neuronal apoptosis induced by catechol metabolites. *FEBS Lett.* 581: 406-412, 2007.
- 2) Satake W, Mizuta I, Suzuki S, Nakabayashi Y, Ito C, Watanabe M, Takeda A, Hasegawa K, Sakoda S, Yamamoto M, Hattori N, Murata M, Toda T., Fibroblast growth factor 20 gene and Parkinson's disease in the Japanese population. *Neuroreport* 18:937-940, 2007
- 3) Takeda A., Kikuchi A., Itoyama Y., Olfactory dysfunction in Parkinson's disease. *J. Neurol.* IV/2-7, 2007.

## 吉良 潤一

発表者氏名: Iwaki A, Kawano Y, Miura S, Shibata H, Matsuse D, Li W, Furuya H, Ohyagi Y, Taniwaki T, Kira J, Fukumaki Y

論文タイトル名: Heterozygous deletion of ITPR1, but not SUMF1 in spinocerebellar ataxia type 16

発表誌名: J Med Genet

巻号: 45

ページ: 32-35

出版年: 2008

佐々木 秀直

雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
Yabe I, Matsushima M, Soma H, Basri R, Sasaki H.	Usefulness of the Scale for Assessment and Rating of Ataxia (SARA)	J Neurol Sci			2007 (in press)
Basri R, Yabe I, Soma H, Sasaki H.	Spectrum and prevalence of autosomal dominant spino- cerebellar ataxia in Hokkaido, the northern island of Japan: a study of 113 Japanese families	J Hum Genet	52(10)	848-855	2007
Soma H, Yabe I, Takei A, Fujiki N, Yanagihara T, Sasaki H.	Heredity in multiple system atrophy	J Neurol Sci	240	107-110	2006
Basri R, Yabe I, Soma H, Takei A, Nishimura H, Machino Y, Kokubo Y, Kosugi M, Okada R, Yukitake M, Tachibana H, Kuroda Y, Kuzuhara S, Sasaki H.	Four mutations of the <i>spastin</i> gene in Japanese families with spastic paraplegia	J Hum Genet	51	711-715.	2006
Yabe I, Soma H, Takei A, Fujiki N, Yanagihara T, Sasaki H.	MSA-C is the predominant clinical phenotype of MSA in Japan: Analysis of 142 patients with probable MSA	J Neurol Sci	249	115-121	2006
Chen DH, Cimino PJ, Ranum LPW, Zoghbi HY, Yabe I, Schut L, Margolis RL, Lipe HP, Feleke A, Matsushita M, Wolff J, Morgan C, Lau D, Fernandez M, Sasaki H, Raskind WH, Bird TD.	The clinical and genetic spectrum of spino- cerebellar ataxia 14	Neurology	64	1258-1260	2005
Takei A, Hamada T, Yabe I, Sasaki H.	Treatment of cerebellar ataxia with 5-HT1A agonist	Cerebellum	4	211-215	2005



黒岩 義之

原著論文

著者	論文タイトル	発表誌名	巻	出版年
			頁	
Omoto S	The effect of attended color on the P1/N1 component of visual event-related potentials	Neuroscience Letters	429:22-27	2007
Johmura Y	Acute mountain Sickness with reversible Vasospasm.	Journal of the Neurological Sciences	263:174-176	2007
Rino Y	Vitamin E malabsorption and Neurological Consequences after Gastrectomy for Gastric Cancer	Hepato-Gastroenterology	54:1858-1861	2007
西山毅彦	てんかん重積	臨床神経生理学	35 (1) : 63-70	2007

吉田 邦広

原著論文

著者	論文タイトル	掲載誌名	巻	出版年
			頁	
Yoshida K, Okano T, Hoshi K, Yahikozawa H, Suzuki K, Banno H, Tamura T, Sobue G, Ikeda S	Congenital fibrosis of the extraocular muscles (CFEOM) syndrome associated with progressive cerebellar ataxia	Am J Med Genet	143(A): 1494-1501	2007
Yoshida K, Yanagawa S, Tsuchiya A, Nakajima T, Fukushima Y, Ikeda S	Huntington's disease with onset ages greater than 60 years	Geriatr Gerontol Int	7: 80-82	2007
Yoshida K, Wada T, Sakurai A, Wakui K, Ikeda S, Fukushima Y	Nationwide survey on predictive genetic testing for late-onset, incurable neurological diseases in Japan	J Hum Genet	52: 675-679	2007

## 祖父江 元

### 日本語論文

1. 祖父江元 神経変性疾患の病態抑止療法への展望 治療研究のパラダイムシフト 神経治療学 24 巻 4 号 Page403-405 (2007.07)

### 英語論文

1. Suenaga M, Kawai Y, Watanabe H, Atsuta N, Ito M, Tanaka F, Katsuno M, Fukatsu H, Naganawa S, Sobue G: Cognitive impairment in spinocerebellar ataxia type 6. *J Neurol Neurosurg Psychiatry*. 2007; [Epub ahead of print]
2. Ito M, Watanabe H, Kawai Y, Atsuta N, Tanaka F, Naganawa S, Fukatsu H, Sobue G: Usefulness of combined fractional anisotropy and apparent diffusion coefficient values for detection of involvement in multiple system atrophy. *J Neurol Neurosurg Psychiatry*. 78: 722-8, 2007.
3. Kawai Y, Suenaga M, Takeda A, Ito M, Watanabe H, Tanaka F, Kato K, Fukatsu H, Naganawa S, Kato T, Ito K, Sobue G: Cognitive impairments in multiple system atrophy: MSA-C vs. MSA-P. *Neurology*. (in press).
4. Suzuiki K, Katsuno M, Banno H, Takeuchi Y, Atsuta N, Ito M, Watanabe H, Yamashita H, Hori N, Nakamura T, Hirayama M, Tanaka F, Sobue G: CAG repeat size correlates to electrophysiological motor and sensory phenotypes in SBMA. *Brain*. 131: 51-61, 2008.
5. Oki Y, Koike H, Iijima M, Mori K, Hattori N, Katsuno M, Nakamura T, Hirayama M, Tanaka F, Shiraishi M, Yazaki S, Nokura K, Yamamoto H, Sobue G: Ataxic vs painful form of paraneoplastic neuropathy. *Neurology*. 69: 564-72, 2007
6. Niwa J, Yamada S, Ishigaki S, Sone J, Takahashi M, Katsuno M, Tanaka F, Doyu M, Sobue G: Disulfide bond mediates aggregation, toxicity, and ubiquitylation of familial amyotrophic lateral sclerosis-linked mutant SOD1. *J Biol Chem*. 282: 28087-28095, 2007
7. Tanaka KI, Namba T, Arai Y, Fujimoto M, Adachi H, Sobue G, Takeuchi K, Nakai A, Mizushima T: Genetic evidence for a protective role for heat shock factor 1 and heat shock protein 70 against colitis. *J Biol Chem*. 282: 23240-23252, 2007
8. Adachi H, Waza M, Tokui K, Katsuno M, Minamiyama M, Tanaka F, Doyu M, Sobue G: CHIP overexpression reduces mutant androgen receptor protein and ameliorates phenotypes of the spinal and bulbar muscular atrophy transgenic mouse model. *J Neurosci*. 27(19): 5115-26, 2007
9. Jiang YM, Yamamoto M, Tanaka F, Ishigaki S, Katsuno M, Adachi H, Niwa JI, Doyu M, Yoshida M, Hashizume Y, Sobue G: Gene expressions specifically detected in motor neurons (Dynactin 1, Early Growth Response 3, Acetyl-CoA Transporter, Death Receptor 5, and Cyclin C) differentially correlate to pathologic markers in sporadic amyotrophic lateral sclerosis. *J Neuropathol Exp Neurol*. 66: 617-627, 2007
10. Yang Z, Chang YJ, Yu IC, Yeh S, Wu CC, Miyamoto H, Merry DE, Sobue G, Chen LM, Chang SS, Chang C: ASC-J9 ameliorates spinal and bulbar muscular atrophy phenotype via degradation of androgen receptor. *Nature Med*. 13: 348-353, 2007

## 宮井 一郎

### 書籍

1. 宮井一郎. 神経疾患のリハビリテーション. 小林祥泰, 水澤英洋編. 運動療法. 神経疾患再診の治療 2006-2008. p. 295-299, 南江堂, 2006.
2. 宮井一郎. 光脳機能イメージング. 鳥羽研二編. 日常診療に生かす老年病ガイドブック 7. 高齢者への包括的アプローチとリハビリテーション. p.252-253, メジカルビュー社, 2006.
3. 関有香子, 畠中めぐみ, 三原雅史, 矢倉一, 宮井一郎. 日本リハビリテーション医学会診療ガイドライン委員会、リハビリテーション連携パス策定委員会編. 脳卒中リハビリテーション連携パス 基本と実践のポイント, p.139-144, 医学書院, 2007.
4. 宮井一郎. 脳に障害がある場合の器用さの学習のメカニズム-リハビリテーション医療への応用. 久保田競, 宮井一郎, 虫明 元. ライブラリ脳の世紀-ー心のメカニズムを探る⑥『学習と脳』, p.107-157, サイエンス社, 2007.
5. 宮井一郎. 神経リハビリテーションと損傷脳の機能的再構成. 「脳を活かす研究会編. ブレイン・マシン・インターフェース. 脳と機械をつなぐ. p.140-155, オーム社, 2007.

### 雑誌

1. Miyai I, Suzuki M, Hatakenaka M, Kubota K. Effect of body weight support on cortical activation during gait in patients with stroke. *Exp Brain Res* 2006; 169(1):85-91.
2. Yagura H, Hatakenaka M, Miyai I. Does therapeutic facilitation add to locomotor outcome of BWSTT in nonambulatory patients with stroke? A randomized controlled trial, *Arch Phys Med Rehab* 2006;87(4):529-535.
3. Hatakenaka M, Miyai I, Mihara M, Sakoda S, Kubota K. Frontal regions involved in learning of motor skill -A functional NIRS study-. *NeuroImage* 2007;34(1):109-116.
4. Hatakenaka M, Miyai I, Sakoda S, Yanagihara T. Proximal paresis of the upper extremity in patients with stroke. *Neurology* 2007;69:348-355.
5. Yagura H, Miyai I, Hatakenaka M, Yanagihara T. Inferior olivary hypertrophy is associated with lower functional state after pontine hemorrhage. *Cerebrovasc Dis* 2007;24:369-374.
6. Mihara M, Miyai I, Hatakenaka M, Sakoda S, Kubota K. Sustained prefrontal activation during ataxic gait: A compensatory mechanism for ataxic stroke? *Neuroimage* 2007;37:1338-45.
7. Kohno S, Miyai I, Seiyama A, Oda I, Ishikawa A, Tsuneishi S, Amita T, Shimizu K. Removal of the skin blood flow artifact in functional near-infrared spectroscopic imaging data through independent component analysis. *Biomedical Optics* 2007;12: 062111-1-9.
8. Suzuki M, Miyai I, Ono T, Kubota K. Activities in the frontal cortex and gait performance are modulated by preparation. An fNIRS study. *NeuroImage* 2008;39:600-607.
9. 宮井一郎. 脳卒中患者の歩行障害への対応. *リハ医学* 2006;43(1):33-39.
10. 三原雅史, 畠中めぐみ, 宮井一郎. 運動時の大脳皮質活動. *体育の科学* 2006;56(1):13-17.
11. 畠中めぐみ, 三原雅史, 矢倉一, 宮井一郎. 脳卒中後の歩行の再獲得と転倒. *バイオメカニズム学会誌* 2006;30(3):128-131.
12. 宮井一郎. 回復期リハビリテーション病棟の転帰と転機. *総合リハ* 2006;34(4):309.
13. 宮井一郎. 神経科学的知見に立脚した脳卒中リハビリテーションの方法論. *日本臨牀* 2006;64(増刊号 7):778-782.
14. 宮井一郎. 神経内科の医療・介護 -現状と課題-. *神経疾患のリハビリテーション. 神経内科* 2006;65(6):560-565
15. 宮井一郎. リハビリテーション医学の神経科学. *BRAIN and NERVE* 59(4):347-355, 2007
16. 宮井一郎. 専門医レポート. 研修施設紹介. 森之宮病院. *臨床リハ* 2007;16(10):994-999.
17. 服部憲明, 宮井一郎. 画像診断. 神経画像法の進歩. *総合リハ* 2007;35(10):1007-1013.
18. 畠中めぐみ, 宮井一郎. 脳可塑性を促進するリハビリテーションプログラム *MB Med Reha* No.85:17-24, 2007.
19. 三原雅史, 畠中めぐみ, 矢倉一, 宮井一郎. 歩行運動時の大脳皮質活動. *神経内科* 2007;67(5):447-452.
20. 三原雅史, 矢倉一, 畠中めぐみ, 宮井一郎. 歩行のリハビリテーション. *Brain Medical* 2007;19(4):368-372.

山田 光則

雑誌

発表者氏名	論文タイトル	発表雑誌	巻号	ページ	出版年
Hara K, Momose Y, Tokiguchi S, Shimohata M, Terajima K, Onodera O, Kakita A, Yamada M, Takahashi H, Hirasawa M, Mizuno Y, Ogata K, Goto J, Kanazawa I, Nishizawa M, Tsuji S.	Multiplex families with multiple system atrophy	Arch Neurol	64	545-551	2007
山田光則	DRPLA	Clinical Neuroscience	25	850-851	2007
Yamada M, Sato T, Tsuji S, Takahashi H	CAG repeat disorder models and human neuropathology: similarities and differences	Acta Neuropathol	115	71-86	2008

西澤 正豊

小野寺 理

雑誌

1. Takahashi T, Kikuchi S, Katada S, Nagai Y, Nishizawa M, Onodera O. Soluble polyglutamine oligomers formed prior to inclusion body formation are cytotoxic. *Hum Mol Genet* 2008;17(3):345-56.
2. Takado Y, Hara K, Shimohata T, Tokiguchi S, Onodera O, Nishizawa M. New mutation in the non-gigantic exon of SACS in Japanese siblings. *Mov Disord* 2007;22(5):748-9.
3. Tada M, Onodera O, Tada M, et al. Early development of autonomic dysfunction may predict poor prognosis in patients with multiple system atrophy. *Arch Neurol* 2007;64(2):256-60.
4. Nozaki H, Ikeuchi T, Kawakami A, et al. Clinical and genetic characterizations of 16q-linked autosomal dominant spinocerebellar ataxia (AD-SCA) and frequency analysis of AD-SCA in the Japanese population. *Mov Disord* 2007;22(6):857-62.
5. Shimohata T, Hara K, Sanpei K, et al. Novel locus for benign hereditary chorea with adult onset maps to chromosome 8q21.3-q23.3. *Brain* 2007;130(Pt 9):2302-9.
6. Hara K, Momose Y, Tokiguchi S, et al. Multiplex families with multiple system atrophy. *Arch Neurol* 2007;64(4):545-51.
7. Hara K, Shimbo J, Nozaki H, Kikugawa K, Onodera O, Nishizawa M. Sacsin-related ataxia with neither retinal hypermyelination nor spasticity. *Mov Disord* 2007;22(9):1362-3.
8. Takahashi T, Tada M, Igarashi S, et al. Aprataxin, causative gene product for EAOH/AO A1, repairs DNA single-strand breaks with damaged 3'-phosphate and 3'-phosphoglycolate ends. *Nucleic Acids Res* 2007;35(11):3797-809.
9. Shimohata T, Shinoda H, Nakayama H, et al. Daytime hypoxemia, sleep-disordered breathing, and laryngopharyngeal findings in multiple system atrophy. *Arch Neurol* 2007;64(6):856-61.
10. Tsuji S, Onodera O, Goto J, Nishizawa M. Sporadic ataxias in Japan - a population-based epidemiological study. *Cerebellum* 2007:1-9.

## 高嶋 博

### 書籍

著者氏名	論文タイトル名	書籍全体の編集者名	書籍名	出版社名	出版地	出版年	ページ
高嶋 博	常染色体劣性遺伝形式の末梢神経障害および小脳失調症を示す疾患 —spinocerebellar ataxia with axonal neuropathy (SCAN1)	柳澤信夫 篠原幸人 岩田 誠 清水輝夫 寺本 明	Annual Review (神経 2007)	中外医学社	東京	2007	208-213
荒田 仁 高嶋 博	Gerstmann-Straussler-Scheinker syndrome 遺伝子異常と臨床的特徴	柳澤信夫 篠原幸人 岩田 誠 清水輝夫 寺本 明	Annual Review (神経 2008)	中外医学社	東京	2008	In press
高嶋 博 有村公良	末梢神経-基礎から臨床まで 末梢神経疾患の発生機序・臨床所見 遺伝性ニューロパチーの進歩	なし	Clinical Neuroscience	中外医学社	東京	2007	752-754

### 雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
市來征仁, 有銘 工, 高嶋博, 有村 公良ら	精神症状を主徴とした 高齢発症のミトコンド リア脳筋症の1例	日本内科学会 雑誌	96巻11号	2536-2538	2007
高嶋 博	末梢神経疾患の所見の とり方と臨床検査 遺 伝性ニューロパチーの 診断と病態	末梢神経	18巻2号	145-151	2007
高嶋 博 有村公良	Spinocerebellar ataxia with axonal neuropathy (SCAN1)	神経進歩	50(3)	379-386	2006
荒田 仁, 高嶋 博	遺伝性プリオン病(GSS, 家族性CJD, FFI)	日本臨床	65(8)	1433-1437,	2007

Hirano R, Takas hima H et al.	Spinocerebellar ataxia with axonal neuropathy: consequence of a Tdpl recessive neomorphic mutation?	EMBO J	26(22)	4732-4743	2007
----------------------------------	---	--------	--------	-----------	------

※掲載論文の別刷りを一部添付のこと。

## 水澤英洋

### [原著]

1. Amino T, Ishikawa K, Toru S, Ishiguro T, Sato N, Tsunemi T, Murata M, Kobayashi K, Inazawa J, Toda T, Mizusawa H. Redefining the disease locus of 16q22.1-linked autosomal dominant cerebellar ataxia. J Hum Genet. 2007; 52(8):643-9. Epub 2007 Jul 5.

### [著書]

1. Ishikawa K., Flanigan K., Mizusawa H. The chromosome 16q22,1-linked autosomal dominant cerebellar ataxia and SCA4. 編集中

### [総説]

1. 石川欽也, 水澤英洋. 第 16 番染色体長腕連鎖型脊髄小脳変性症. 臨床医のための神経病理 Clinical Neuroscience 2007; 25(7): 738-739.
2. 石川欽也, 融 衆太, 水澤英洋. 脊髄小脳変性症の新しい遺伝子: *puratrophin-1*. Current Insights In Neurological Science. 15(3); 8-9, 2007.



矢澤 生

脳バンク日本における現状と問題点. 医学のあゆみ. 220 巻 820-823 頁、2007 年

貫名 信行

雑誌

1. Khan, L. A., Yamanaka, T., & Nukina, N. Genetic impairment of autophagy intensifies expanded polyglutamine toxicity in *Caenorhabditis elegans*. *Biochem Biophys Res Commun* (2007) in press.
2. Doi, H., Okamura, K., Bauer, P.O., Furukawa, Y., Shimizu, H., Kurosawa, M., Machida, Y., Miyazaki, H., Mitsui, K., Kuroiwa, Y. & Nukina, N. RNA-binding protein TLS is a major nuclear aggregate-interacting protein in Huntingtin exon 1 with expanded polyglutamine-expressing cells. *J Biol Chem* (2007) in press.

瀧山 嘉久

書籍

1. Takiyama, Y.: Machado-Joseph disease/Spinocerebellar ataxia type 3. *Advances in spinocerebellar degeneration and spastic paraplegia*, eds by Takiyama, Y. and Nishizawa, M., Research Signpost, Kerala, India (in press)
2. Takiyama, Y.: Sacsinopathies. *Advances in spinocerebellar degeneration and spastic paraplegia*, eds by Takiyama, Y. and Nishizawa, M., Research Signpost, Kerala, India, 2007 (in press)
3. Shimazaki, H. and Takiyama, Y.: Spinocerebellar ataxia type 6 (SCA6). *Advances in spinocerebellar degeneration and spastic paraplegia*, eds by Takiyama, Y. and Nishizawa, M., Research Signpost, Kerala, India (in press)
4. Shimazaki, H. and Takiyama, Y.: Ataxia oculomotor apraxia type 1 (AOA1). *Advances in spinocerebellar degeneration and spastic paraplegia*, eds by Takiyama, Y. and Nishizawa, M., Research Signpost, Kerala, India (in press)

5. Sakoe, K., Namekawa, M., and Takiyama, Y.: Spastic paraplegia type 4 (SPG4). *Advances in spinocerebellar degeneration and spastic paraplegia*, eds by Takiyama, Y. and Nishizawa, M., Research Signpost, Kerala, India (in press)
6. Sakoe, K. and Takiyama, Y.: Heat shock proteins and neurodegenerative diseases. *Heat-Shock Proteins: International Research*, ed by Columbus, F., Nova Science Publishers, Inc., NY, USA (in press)
7. 瀧山嘉久: 脊髄小脳変性症, 看護のための疾患・症候事典, 永井良三監修, メディカルフレンド社, 東京 (印刷中)
8. 瀧山嘉久: 神経難病 多発性硬化症・脊髄小脳変性症, 運動リハビリ実践マニュアル, 伊藤博元・岩本幸英・岩谷 力・越智光夫・立花新太郎・星野雄一編 (日本整形外科学会・日本リハビリテーション学会監修), 全日本病院出版会, 東京 (印刷中)
9. 瀧山嘉久: 遺伝性痙性対麻痺, Annual Review 神経 2008, 柳澤信夫・篠原幸人・岩田誠・清水輝夫・寺本 明編, 中外医学社, 東京 (印刷中) .

## 雑誌

1. Ouyang Y and Takiyama Y.: 16q-linked autosomal dominant cerebellar ataxia in Japan. In: R.M. Mohan, ed. Research Advances in Neurology 4. Kerala: Global Research Network, 2007; 1-7.
2. Takiyama Y.: Sacsinopathies: saccin-related ataxia. *Cerebellum* 6: 353-359, 2007.
3. Shimazaki H, Sakoe K, Nijima K, Nakano I and Takiyama Y.: An unusual case of a spasticity-lacking phenotype with a novel *SACS* mutation. *J Neurol Sci* 255: 87-89, 2007.
4. Ouyang Y, Segers K, Bouquiaux O, Wang FC, Janin N, Andris C, Shimazaki H, Sakoe K, Nakano I, and Takiyama Y.: Novel *SACS* mutation in a Belgian family with saccin-related ataxia. *J Neurol Sci* 264: 73-76, 2008.
5. Craig K, Takiyama Y. Soong BW, Jardim LB, Saraiva-Pereira ML, Lythgow K, Morino H, Maruyama H, Kawakami H and Chinnery PF: Pathogenic expansions of the SCA6 locus are associated with a common CACNA1A haplotype across the globe: founder effect or predisposing chromosome? *Eur J Hum Genet* (in press)

## 松浦 徹

### 雑誌

1. Kurosaki T, Matsuura T, Ohno K, Ueda S Long-range PCR for the diagnosis of spinocerebellar ataxia type 10. **Neurogenetics** 2008, in press.
2. #Gao R, #Matsuura T, #Coolbaugh M (#equally contributed), Zühlke C, Nakamura K, Rasmussen A, Siciliano MJ, Ashizawa T, Lin X. Instability of expanded CAG/CAA repeats in spinocerebellar ataxia type 17. **Eur J Hum Genet** 2008; 16: 215-222.
3. Saito T, Amakusa, Y, Kimura T, Yahara O, Aizawa H, Ikeda Y, Day JW, Ranum LPW, Ohno K, Matsuura T. Myotonic dystrophy type 2 in Japan: ancestral origin distinct from Caucasian families. **Neurogenetics** 2008, in press.
4. Sahashi K, Masuda A, Matsuura T, Shinmi J, Zhang Z, Takeshima Y, Matsuo M, Sobue G, Ohno K. In vitro and in silico analysis reveals an efficient algorithm to predict the splicing consequences of mutations at the 5' splice sites. **Nucleic Acids Res** 2007; 35: 5995-6003.
5. Ichihara M, Murakumo Y, Masuda A, Matsuura T, Asai N, Jijiwa M, Ishida M, Shinmi J, Yatsuya H, Qiao S, Takahashi M, Ohno K. Thermodynamic instability of siRNA duplex is a prerequisite for dependable prediction of siRNA activities. **Nucleic Acids Res** 2007; 35: e123.
6. 松浦 徹. 脊髄小脳失調症 10 型の分子遺伝学的解析. **臨床神経学** 2008; 48:1-10.

## 永井 義隆

### 書籍

著者氏名	論文タイトル名	書籍全体の編集者名	書籍名	出版社名	出版地	出版年	ページ
永井義隆	神経変性疾患治療戦略としての低分子による蛋白質凝集阻害	高橋良輔	神経変性疾患のサイエンス	南山堂	東京	2007	181-197

### 雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
Takahashi T, Kikuchi S, Katada S, <u>Nagai Y</u> , Nishizawa M, Onodera O.	Soluble polyglutamine oligomers formed prior to inclusion body formation are cytotoxic.	Hum Mol Genet	17 (3)	345-356	2008
Takahashi Y, Okamoto Y, Popiel HA, Fujikake N, Toda T, Kinjo M, * <u>Nagai Y</u> .	Detection of polyglutamine protein oligomers in cells by fluorescence correlation spectroscopy.	J Biol Chem	282 (33)	24039-24048	2007
* <u>Nagai Y</u> , Inui T, Popiel HA, Fujikake N, Hasegawa K, Urade Y, Goto Y, Naiki H, Toda T.	A toxic monomeric conformer of the polyglutamine protein.	Nat Struct Mol Biol	14 (4)	332-340	2007
永井義隆	ポリグルタミンタンパク質の毒性構造体の発見	蛋白質・核酸・酵素			印刷中
永井義隆、ポピエル明子、藤掛伸宏、戸田達史	ポリグルタミン病に対する治療戦略	BRAIN and NERVE	59 (2)	393-404	2007

※掲載論文の別刷りを一部添付のこと。