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**Cochrane Systematic Reviews**

**Updated Reviews – January 2014**  
Corticosteroids for chronic inflammatory demyelinating polyradiculoneuropathy

**New Reviews – December 2014**  
Interventions for fatigue in peripheral neuropathy

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**New from Up To Date**

What’s new in neurology

Neurology related topics

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**Journal Articles**

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**Corticobasal Degeneration Table of Contents**
1. Apraxia in anti-glutamic acid decarboxylase-associated stiff person syndrome: Link to corticobasal degeneration?

**Corticobasal Degeneration Journal Articles**
1. Apraxia in anti-glutamic acid decarboxylase-associated stiff person syndrome: Link to corticobasal degeneration?.
Citation: Annals of Neurology, January 2015, vol./is. 77(1(173-6)), 0364-5134;1531-8249 (2015 Jan)
Author(s): Bowen LN, Subramony SH, Heilman KM
Language: English
Abstract: Corticobasal syndrome (CBS) is associated with asymmetrical rigidity as well as asymmetrical limb-kinetic and ideomotor apraxia. Stiff person syndrome (SPS) is characterized by muscle stiffness and gait difficulties. Whereas patients with CBS have several forms of pathology, many patients with SPS have glutamic acid decarboxylase antibodies (GAD-ab), but these 2 disorders have not been reported to coexist. We report 2 patients with GAD-ab-positive SPS who also had signs suggestive of CBS, including asymmetrical limb rigidity associated with both asymmetrical limb-kinetic and ideomotor apraxia. Future studies should evaluate patients with CBS for GAD-ab and people with SPS for signs of CBS. ANN NEUROL 2015;77:173-176. Copyright 2014 American Neurological Association.
Publication type: Journal Article
Source: MEDLINE
Full text: Available Annals of neurology at No link? Ask Salisbury Healthcare Library - please click here to request article.

Epilepsy Table of Contents

1. A review of the quality of care following prolonged seizures in 1-18 year olds with epilepsies

2. Association of prone position with sudden unexpected death in epilepsy

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7. Intractable occipital lobe epilepsy: clinical characteristics, surgical treatment, and a systematic review of the literature

8. Memory in children with epilepsy: A systematic review

9. Old versus New: Why Do We Need New Antiepileptic Drugs?.


12. When should clinicians search for GLUT1 deficiency syndrome in childhood generalized epilepsies?

Epilepsy Journal Articles

Citation: Seizure, January 2015, vol./is. 24((88-92), 1059-1311;1532-2688 (2015 Jan)
Author(s): Hunter L, Sidebotham P, Appleton R, Dunkley C
Language: English
Abstract: PURPOSE: To review the quality of care of children and young people with epilepsies who, following a prolonged seizure, received high-dependency or intensive care. To identify and learn from clinical, organisational, management or personal issues that contributed to these admissions, in order to inform practice and improve clinical
services for children across the UK.METHOD: Notifications were collected from consultant paediatricians in England, Wales, Scotland and Northern Ireland over a 10-month period. For all eligible cases a clinical questionnaire was sent to the notifying clinician. A sample of these cases were selected for a detailed case note review. Case notes were reviewed by paediatrician-nurse pairs using a purpose-built assessment tool derived from national guidelines.RESULTS: Data were collected from 135 clinical questionnaires, and 36 sets of case notes were reviewed. Findings were compared to national standards of care and emerging themes identified. There was evidence of good epilepsy management in many cases. In some cases there was evidence of a lack of clear emergency care plans, of delays in administration of emergency medication, and of deviation from established national guidelines.CONCLUSION: The findings of this review suggest there have been improvements in the care of children and young people with epilepsies presenting with prolonged seizures compared to previous studies. Nevertheless, further improvements are needed, particularly in communication with families and prompt administration of emergency medication.Copyright 2014 British Epilepsy Association. Published by Elsevier Ltd. All rights reserved.

Publication type: Journal Article
Source: MEDLINE
Full text: Available Seizure : the journal of the British Epilepsy Association at No link? Ask Salisbury Healthcare Library - please click here to request article.

2. Association of prone position with sudden unexpected death in epilepsy.
Citation: Neurology, February 2015, vol./is. 84/7(703-9), 0028-3878;1526-632X (2015 Feb 17)
Author(s): Liebenthal JA, Wu S, Rose S, Ebersole JS, Tao JX
Language: English
Abstract: OBJECTIVE: To examine the association between prone position and sudden unexpected death in epilepsy (SUDEP).METHODS: We conducted a systematic review and meta-analysis based on a literature search from databases PubMed, Web of Science, and Scopus, using keywords "SUDEP" or "sudden unexpected death in epilepsy" or "sudden unexplained death syndromes in epilepsy." Twenty-five publications met the inclusion and exclusion criteria and were enrolled in this study.RESULTS: Body positions were documented in 253 cases of SUDEP. Of these patients, 73.3% (95% confidence interval [CI] = 65.7%, 80.9%) died in the prone position, whereas 26.7% (95% CI = 16.3%, 37.1%) died in nonprone positions. Binary random-effects analysis showed that prone position is significantly associated with SUDEP, as compared with nonprone position (p < 0.001). In addition, the prone position was reported in all 11 cases of video-EEG-monitored SUDEP. Moreover, in a subgroup of 88 cases of SUDEP in which demographics and circumstances of death were documented, the prone position was observed in 85.7% (95% CI = 74.6%, 93.3%) of patients aged 40 years or younger, but in only 60% (95% CI = 38.7%, 78.9%) of patients older than 40 years. Statistical analysis confirmed that the prone position was significantly more prevalent in the younger patient group, as compared with the older patient group (odds ratio 3.9; 95% CI = 1.4%, 11.4%; p = 0.009).CONCLUSION: There is a significant association between prone position and SUDEP, which suggests that prone position is a major risk factor for SUDEP, particularly in patients aged 40 years and younger. As such, SUDEP may share mechanisms similar to sudden infant death syndrome.Copyright 2015 American Academy of Neurology.
Publication type: Journal Article
Source: MEDLINE
Full text: Available Ovid at Neurology

3. Clinical outcomes and quality of life following surgical treatment for refractory epilepsy: a systematic review and meta-analysis.
Citation: Medicine, February 2015, vol./is. 94/6(e500), 0025-7974;1536-5964 (2015 Feb)
Author(s): Liu SY, Yang XL, Chen B, Hou Z, An N, Yang MH, Yang H
Language: English
Abstract: Surgery for refractory epilepsy is widely used but the efficacy of this treatment for providing a seizure-free outcome and better quality of life remains unclear. This study aimed to update current evidence and to evaluate the effects of surgery on quality of life in patients with refractory epilepsy. A systematic review and meta-analysis of the literature were conducted and selected studies included 2 groups of refractory epilepsy patients, surgical and nonsurgical. The studies were assessed using the Newcastle-Ottawa Scale. The primary outcome was the seizure-free rate. The secondary outcome was quality of life. Adverse events were also reviewed. After screening, a total of 20 studies were selected: 8 were interventional, including 2 randomized controlled trials, and 12 were observational. All of the studies comprised 1959 patients with refractory epilepsy. The seizure-free rates were significantly higher for patients who received surgery compared with the patients who did not; the combined odds ratio was 19.35 (95% CI =
12.10-30.95, P < 0.001). After adjusting for publication bias the combined odds ratio was 10.25 (95% CI = 5.84-18.00). In both the interventional and observational studies, patients treated surgically had a significantly better quality of life compared with the patients not treated surgically. Complications were listed in 3 studies and the rates were similar in surgical and nonsurgical patients. Our meta-analysis found that for patients with refractory epilepsy, surgical treatment appears to provide a much greater likelihood of seizure-free outcome than nonsurgical treatment, although there is a need for more studies, particularly randomized studies, to confirm this conclusion. Based on more limited data, surgical treatment also appeared to provide a better quality of life and did not seem to increase complications.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available *Medicine* at [Medicine](http://www.medicine.com)

### 4. Efficacy of and patient compliance with a ketogenic diet in adults with intractable epilepsy: a meta-analysis.

**Citation:** Journal of Clinical Neurology, January 2015, vol./is. 11/1(26-31), 1738-6586;1738-6586 (2015 Jan)  
**Author(s):** Ye F, Li XJ, Jiang WL, Sun HB, Liu J  
**Language:** English  
**Abstract:** BACKGROUND AND PURPOSE: Despite the successful use of a ketogenic diet in pediatric epilepsy, its application in adults has been limited. The aim of this meta-analysis was to summarize the findings of relevant published studies in order to identify the efficacy of and compliance with a ketogenic diet and its main subtypes (i.e., classic ketogenic diet and modified Atkins diet) in adults with intractable epilepsy, and to provide useful information for clinical practice. METHODS: Electronic searches of PubMed, EMBASE, Google Scholar, and the ISI Web of Science were conducted to identify studies of the efficacy of and patient compliance with a ketogenic diet in adults with intractable epilepsy; the included studies were reviewed. Meta-analyses were performed using STATA to determine combined efficacy rates and combined rates of compliance with the ketogenic diet and its main subtypes. RESULTS: In total, 12 studies qualified for inclusion, and data from 270 patients were evaluated. The results of the meta-analysis revealed combined efficacy rates of all types of ketogenic diet, a classical ketogenic diet, and a modified Atkins diet were 42%, 52%, and 34%, respectively; the corresponding combined compliance rates were 45%, 38%, and 56%. CONCLUSIONS: The results indicate that a ketogenic diet is a promising complementary therapy in adult intractable epilepsy, and that while a classical ketogenic diet may be more effective, adult patients are likely to be less compliant with it than with a modified Atkins diet.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available *Journal of clinical neurology (Seoul, Korea)* at [Journal of Clinical Neurology](http://www.jclinneurokorea.org)

### 5. Epilepsy surgery in patients with bilateral temporal lobe seizures: A systematic review.

**Citation:** Epilepsia, December 2014, vol./is. 55/12(1892-901), 0013-9580;1528-1167 (2014 Dec)  
**Author(s):** Aghakhani Y, Liu X, Jette N, Wiebe S  
**Language:** English  
**Abstract:** We explored the association between magnetic resonance imaging (MRI) lesion, degree of seizure laterality on intracranial electroencephalography (iEEG), and seizure outcome in patients with ambiguous or presumed bilateral temporal lobe epilepsy (BiTLE) on scalp EEG. We systematically reviewed the literature using Embase and MEDLINE up to May 31, 2012. Patients with bilateral iEEG, temporal lobe surgery, and follow-up >1 year were included. We undertook three separate analyses on patients whose scalp EEG showed ambiguous onset or BiTLE (1) group data of those whose iEEG demonstrated unilateral TLE, (2) group data of those whose iEEG demonstrated BiTLE, (3) individual patient analysis in those with BiTLE for whom iEEG seizure laterality data were provided. Of 1,403 patients with ambiguous or presumed BiTLE on scalp EEG, 1,027 (73%) proved to have unilateral TLE on iEEG and contributed to the first analysis. Of these, 58% had Engel class I and 9% Engel class II outcomes. Of 132 patients in the second analysis (true BiTLE), Engel class I and II outcomes were achieved in 23% and 14%, respectively. Of 41 patients in the third analysis, 66% and 2% had Engel class I and II outcomes, respectively. The median proportion of seizures ipsilateral to the resection on iEEG did not differ between BiTLE patients with Engel class I-III (76%) and Engel III-IV (78%) outcomes ($p = 0.87$). Patients with ambiguous or independent bitemporal seizure onset on scalp EEG achieved good surgical outcomes. Overall, a significantly higher proportion of patients achieved good outcomes when iEEG showed unilateral TLE (67%) than when it showed true BiTLE (45%). However, the degree of seizure lateralization in those with BiTLE was not associated with seizure outcome, and it has a limited role in selecting the side of surgery. Copyright Wiley Periodicals, Inc. 2014 International League Against Epilepsy.

**Citation:** Journal of Neurosurgery. Pediatrics., January 2015, vol./is. 15/1(34-44), 1933-0707;1933-0715 (2015 Jan)

**Author(s):** Griessenauer CJ, Salam S, Hendrix P, Patel DM, Tubbs RS, Blount JP, Winkler PA

**Language:** English

**Abstract:** OBJECT Evidence in support of hemispherectomy stems from a multitude of retrospective studies illustrating individual institutions' experience. A systematic review of this topic, however, is lacking in the literature. METHODS A systematic review of hemispherectomy for the treatment of refractory epilepsy available up to October 2013 was performed using the following inclusion criteria: reports of a total of 10 or more patients in the pediatric age group (<20 years) undergoing hemispherectomy, seizure outcome reported after a minimum follow-up of 1 year after the initial procedure, and description of the type of hemispherectomy. Only the most recent paper from institutions that published multiple papers with overlapping study periods was included. Two reviewers independently applied the inclusion criteria and extracted all the data. RESULTS Twenty-nine studies with a total of 1161 patients met the inclusion criteria. Seizure outcome was available for 1102 patients, and the overall rate of seizure freedom at the last follow-up was 73.4%. Sixteen studies (55.2%) exclusively reported seizure outcomes of a single type of hemispherectomy. There was no statistically significant difference in seizure outcome and type of hemispherectomy (p = 0.737). Underlying etiology was reported for 85.4% of patients with documented seizure outcome, and the overall distribution of acquired, developmental, and progressive etiologies was 30.5%, 40.7%, and 28.8%, respectively. Acquired and progressive etiologies were associated with significantly higher seizure-free rates than developmental etiologies (p < 0.001). Twenty of the 29 studies (69%) reported complications. The overall rate of hydrocephalus requiring CSF diversion was 14%. Mortality within 30 days was 2.2% and was not statistically different between types of hemispherectomy (p = 0.787). CONCLUSIONS Hemispherectomy is highly effective for treating refractory epilepsy in the pediatric age group, particularly for acquired and progressive etiologies. While the type of hemispherectomy does not have any influence on seizure outcome, hemispherotomy procedures are associated with a more favorable complication profile.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available *Journal of neurosurgery. Pediatrics* at [No link? Ask Salisbury Healthcare Library - please click here to request article.](#)

7. Intractable occipital lobe epilepsy: clinical characteristics, surgical treatment, and a systematic review of the literature.

**Citation:** Acta Neurochirurgica, January 2015, vol./is. 157/1(63-75), 0001-6268;0942-0940 (2015 Jan)

**Author(s):** Yang PF, Jia YZ, Lin Q, Mei Z, Chen ZQ, Zheng ZY, Zhang HJ, Pei JS, Tian J, Zhong ZH

**Language:** English

**Abstract:** PURPOSE: We reported our experience in the surgical treatment of a relatively large cohort of patients with occipital lobe epilepsy (OLE). We also carried out a systematic review of the literature on OLE.METHODS: Thirty-five consecutive patients who underwent occipital resection for epilepsy were included. Diagnoses were made following presurgical evaluations, including magnetic resonance imaging (MRI), fluorodeoxyglucose-positron emission tomography (FDG-PET), scalp video-electroencephalogram (EEG) monitoring, and intracranial EEG monitoring. At last follow-up, seizure outcome was classified using the Engel classification scheme.RESULTS: Twenty-five of 35 patients experienced/had experienced >1 type of aura before the seizure. Invasive recordings were used to define the epileptogenic area in 30 of 35 patients (85.7%). All patients underwent occipital lesionectomies or topectomies. Histopathology revealed: cortical dysplasias, gliosis, dysembryoplastic neuroepithelial tumor, ganglioglioma, and tuberous sclerosis. After a mean follow-up of 44 months, 25 patients (71.4 %) were seizure free (Engel class I), 3 (8.6 %) rarely had seizures (Engel class II), 5 (14.3 %) improved more than 75 % (Engel class III), and 2 (5.7 %) had no significant improvement (Engel class IV). Preoperatively, 12 of 33 patients (36.4 %) had visual field deficits. Postoperatively, 25 patients (75.8 %) had new or aggravated visual field deficits.CONCLUSIONS: The management of OLE has been aided greatly by the availability of high-resolution diagnosis. Postoperative visual field deficits occur in a significant proportion of patients. Comprehensive intracranial EEG coverage of all occipital surfaces helps to define the epileptogenic area and preserve visual function, especially in cases of focal cortical dysplasia undetectable by MRI.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available *Journal Article* at [No link? Ask Salisbury Healthcare Library - please click here to request article.](#)
8. Memory in children with epilepsy: A systematic review.
Citation: Seizure, February 2015, vol./is. 25/(126-35), 1059-1311;1532-2688 (2015 Feb)
Author(s): Menlove L, Reilly C
Language: English
Abstract: PURPOSE: Research suggests an increased risk for cognitive impairment in childhood epilepsy with memory being one area of cognition most likely to be affected. Understanding the prevalence and predictors of memory difficulties may help improve awareness of the difficulties and allow efficacious supports to be put in place.
METHOD: A systematic review was carried out using the search terms 'memory', 'children' and 'epilepsy' in the database PUBMED. Eighty-eight studies met inclusion criteria. The review focuses on comparisons of memory scores of children with epilepsy and controls, and comparison of memory scores of children with epilepsy to normative scores. Predictors of memory impairment and the effect of surgery on memory functioning are also reviewed.
RESULTS: The majority (78%) of studies reviewed revealed that children with epilepsy scored lower than controls and normative scores on measures of memory. Post-surgery, memory scores were reported to improve in 50% of studies. Predictors of memory impairment included a greater number of AEDs used, younger age of onset, increased seizure frequency and longer duration of epilepsy.
CONCLUSION: Children with epilepsy have a high frequency of memory impairments. However, the exact prevalence of difficulties is not clear due to the lack of population-based data. Most studies have not controlled for IQ and thus it is unclear if difficulties are always related to global cognitive difficulties. There is need for future population-based studies and studies focussing on the neurobiology of memory problems in children with epilepsy.
Publication type: Journal Article
Source: MEDLINE
Full text: Available Seizure : the journal of the British Epilepsy Association at No link? Ask Salisbury Healthcare Library - please click here to request article.

9. Old versus New: Why Do We Need New Antiepileptic Drugs?.
Citation: Journal of Epilepsy Research, December 2014, vol./is. 4/2(39-44), 2233-6249;2233-6249 (2014 Dec)
Author(s): Lee SK
Language: English
Abstract: Achieving complete seizure remission without adverse events is the goal of epilepsy treatment. Recently, many new antiepileptic drugs (AEDs) have been developed. Even though the efficacy of new AEDs is not stronger than that of old AEDs, there are advantages in using new AEDs. They have unique or different mechanisms of action that enable the creation of possible synergistic combinations. They usually exhibit fewer or no pharmacokinetic drug interactions. Furthermore, the response to AEDs varies individually. A similar efficacy does not imply a similar response from all patients. Many new AEDs have fewer adverse events, including induction of congenital malformations. Other concerns about the long-term effects of established AEDs, such as bone health and development of atherosclerosis, may be alleviated by the use of new AEDs. New AEDs are needed to achieve better care of patients with epilepsy.
Publication type: Journal Article, Review
Source: MEDLINE

Citation: JAMA, January 2015, vol./is. 313/3(285-93), 0098-7484;1538-3598 (2015 Jan 20)
Author(s): Jobst BC, Cascino GD
Language: English
Abstract: IMPORTANCE: Epilepsy surgery is indicated for patients with focal seizures who do not respond to appropriate antiepileptic drug therapy consisting of 2 or more medications. OBJECTIVES: To review resective surgery outcomes for focal epilepsy, to identify which patients benefit the most, and to discuss why epilepsy surgery may not be universally accepted. EVIDENCE REVIEW: Medline and Cochrane databases were searched between January 1993 and June 2014 for randomized clinical trials, meta-analyses, systematic reviews, and large retrospective case series (>300 patients) using Medical Subject Headings and indexed text terms. Fifty-five articles were included. Subpopulations and prognostic factors were identified. Systematic reviews for cognitive, psychiatric, quality-of-life, and psychosocial outcomes were included. FINDINGS: Two randomized clinical trials enrolling 118 patients with temporal
lobe epilepsy found greater freedom from seizures with surgery when compared with continued medical treatment (58% vs 8% [n=80] and 73% vs 0% [n=38], P<.001). Nine systematic reviews and 2 large case series of medically refractory patients treated with surgery reported seizure-free outcomes in 34% to 74% of patients (median, 62.4%). The remainder of systematic reviews and meta-analyses examined subpopulations. Epilepsy surgery was less effective when there were extratemporal lesions, the epilepsy was not associated with a structural lesion, or both. Seizure-free outcomes were similar between children and adults. Hippocampal sclerosis and benign tumors were associated with better outcomes relative to other pathologies. Similar procedures such as selective amygdalohippocampectomy and temporal lobectomy for temporal lobe epilepsy were associated with subtle differences in seizure and neuropsychological outcome. There is low perioperative mortality (0.1%-0.5%) from epilepsy surgery. The most frequent neurologic complication is visual field defect occurring from temporal lobe resection. Quality of life improved after surgery but improved the most in patients who were seizure-free after surgery.CONCLUSIONS AND RELEVANCE: Epilepsy surgery reduced seizure activity in randomized clinical trials when compared with continued medical therapy. Long-term cognitive, psychiatric, psychosocial, and quality-of-life outcomes were less well defined. Despite good outcomes from high-quality clinical trials, referrals of patients with seizures refractory to medical treatment remain infrequent.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available American Medical Association at JAMA


**Citation:** Journal of Advanced Nursing, March 2015, vol./is. 71/3(478-97), 0309-2402;1365-2648 (2015 Mar)  
**Author(s):** Lewis SA, Noyes J, Hastings RP  
**Language:** English  
**Abstract:** AIM: To determine the effectiveness of epilepsy self-management interventions and explore the views and experiences of medication and seizures by children and young people.BACKGROUND: Experiencing seizures and side-effects from anti-epileptic medicines have negative impacts on children and young people managing their epilepsy. Children commonly experiment with not taking epilepsy medication as prescribed and engage in unhealthy lifestyles. DESIGN/REVIEW METHODS: Mixed-method systematic review with theory development. Cochrane quantitative methods and thematic synthesis of qualitative and survey evidence.DATA SOURCES: Eight databases were searched from earliest dates to July 2013.RESULTS: Nineteen studies were included. Meta-analysis was not possible. Zero of nine intervention studies showed improvement in anti-epilepsy medication adherence. Skill-based behavioural techniques with activities such as role play and goal setting with young people increased epilepsy knowledge and seizure self-management (small effects). Intervention studies were methodologically weak and no studies reported if improvement in self-management was sustained over time. Synthesis of nine qualitative and one mixed-method studies generated six themes encapsulating anti-epilepsy medication and epilepsy effects. There was a lack of fidelity between intervention programme theories and what children and young people found difficult with medication self-management and managing the effects of epilepsy.CONCLUSION: Children and young people knowingly and/or unknowingly take risks with their epilepsy and give reasoned explanations for doing so. There are no effective interventions to change epilepsy medication adherence behaviours. There is an urgent need for more innovative and individually tailored interventions to address specific challenges to epilepsy self-management as identified by children and young people themselves.Copyright 2014 John Wiley & Sons Ltd.  
**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available EBSCOhost EJS at Journal of Advanced Nursing

### 12. When should clinicians search for GLUT1 deficiency syndrome in childhood generalized epilepsies?.

**Citation:** European Journal of Paediatric Neurology, March 2015, vol./is. 19/2(170-5), 1090-3798;1532-2130 (2015 Mar)  
**Author(s):** Lebon S, Suarez P, Alija S, Korff CM, Fluss J, Mercati D, Datta AN, Poloni C, Marcoz JP, Campos-Xavier AB, Bonafe L, Roulet-Perez E  
**Language:** English  
**Abstract:** UNLABELLED: GLUT1 deficiency (GLUT1D) has recently been identified as an important cause of generalized epilepsies in childhood. As it is a treatable condition, it is crucial to determine which patients should be investigated.METHODS: We analyzed SLC2A1 for mutations in a group of 93 unrelated children with generalized epilepsies. Fasting lumbar puncture was performed following the identification of a mutation. We compared our results
with a systematic review of 7 publications of series of patients with generalized epilepsies screened for SLC2A1 mutations. RESULTS: We found 2/93 (2.1%) patients with a SLC2A1 mutation. One, carrying a novel de novo deletion had epilepsy with myoclonic-atonic seizures (MAE), mild slowing of head growth, choreiform movements and developmental delay. The other, with a paternally inherited missense mutation, had childhood absence epilepsy with atypical EEG features and paroxysmal exercise-induced dyskinesia (PED) initially misdiagnosed as myoclonic seizures. Out of a total of 1110 screened patients with generalized epilepsies from 7 studies, 2.4% (29/1110) had GLUT1D. This rate was higher (5.6%) among 303 patients with early onset absence epilepsy (EOAE) from 4 studies. About 50% of GLUT1D patients had abnormal movements and 41% a family history of seizures, abnormal movements or both. CONCLUSION: GLUT1D is most likely to be found in MAE and in EOAE. The probability of finding GLUT1D in the classical idiopathic generalized epilepsies is very low. Pointers to GLUT1D include an increase in seizures before meals, cognitive impairment, or PED which can easily be overlooked. Copyright 2014 European Paediatric Neurology Society. Published by Elsevier Ltd. All rights reserved.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available [European journal of paediatric neurology : EJPN : official journal of the European Paediatric Neurology Society](http://group.bmj.com/group/rights/licensing/permissions).

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**Motor Neurone Disease Table of Contents**


2. Dignity therapy for people with motor neuron disease and their family caregivers: a feasibility study


**Motor Neurone Disease Journal Articles**

1. **Cortical thickness in ALS: towards a marker for upper motor neuron involvement.**  
   **Citation:** Journal of Neurology, Neurosurgery & Psychiatry, March 2015, vol./is. 86/3(288-94), 0022-3050;1468-330X (2015 Mar)  
   **Author(s):** Walhout R, Westeneng HJ, Verstraete E, Hendrikse J, Veldink JH, van den Heuvel MP, van den Berg LH  
   **Language:** English  
   **Abstract:** OBJECTIVE: Examine whether cortical thinning is a disease-specific phenomenon across the spectrum of motor neuron diseases in relation to upper motor neuron (UMN) involvement. METHODS: 153 patients (112 amyotrophic lateral sclerosis (ALS), 19 patients with a clinical UMN phenotype, 22 with a lower motor neuron (LMN) phenotype), 60 healthy controls and 43 patients with an ALS mimic disorder were included for a cross-sectional cortical thickness analysis. Thirty-nine patients with ALS underwent a follow-up scan. T1-weighted images of the brain were acquired using a 3 T scanner. The relation between cortical thickness and clinical measures, and the longitudinal changes were examined. RESULTS: Cortical thickness of the precentral gyrus (PCG) was significantly reduced in ALS (p=1.71x10(-13)) but not in mimic disorders (p=0.37) or patients with an LMN phenotype (p=0.37), as compared to the group of healthy controls. Compared to patients with ALS, patients with a UMN phenotype showed an even lower PCG cortical thickness (p=1.97x10(-3)). Bulbar scores and arm functional scores showed a significant association with cortical thickness of corresponding body regions of the motor homunculus. Longitudinal analysis revealed a decrease of cortical thickness in the left temporal lobe of patients with ALS (parahippocampal region p=0.007 and fusiform cortex p=0.001). CONCLUSIONS: PCG cortical thinning was found to be specific for motor neuron disease with clinical UMN involvement. Normal levels of cortical thickness in mimic disorders or LMN phenotypes suggest that cortical thinning reflects pathological changes related to UMN involvement. Progressive cortical thinning in the temporal lobe suggests recruitment of non-motor areas, over time. Copyright Published by the BMJ Publishing Group Limited. For permission to use (where not already granted under a licence) please go to [http://group.bmj.com/group/rights-licensing/permissions].

**Publication type:** Journal Article  
**Source:** MEDLINE

Citation: Journal of Palliative Medicine, January 2015, vol./is. 18/1(31-7), 1557-7740;1557-7740 (2015 Jan)

Author(s): Aoun SM, Chochinov HM, Kristjanson LJ

Language: English

Abstract: UNLABELLED: Abstract Background: There are calls to explore psychological interventions to reduce distress in patients with motor neuron disease (MND) and their family caregivers. Dignity therapy is a short-term psychotherapy intervention shown to alleviate distress for people with life-limiting illnesses.OBJECTIVES: To assess the acceptability, feasibility, and effectiveness of dignity therapy to reduce distress in people with MND and their family caregivers.METHODS: The study used a repeated-measures design pre- and post-intervention. Acceptability and feasibility were assessed using participants’ ratings of the helpfulness of the intervention across several domains and time and resources required. Effectiveness measures for patients included: dignity-related distress, hopefulness, and spiritual well-being; and those for family caregivers included burden, hopefulness, anxiety, and depression.RESULTS: Twenty-seven patients and 18 family caregivers completed the intervention. Dignity therapy was well accepted, including those patients who required assisted communication devices. The feasibility may be limited in small or not well-resourced services. There were no significant differences in all outcome measures for both groups. However, the high satisfaction and endorsement of dignity therapy by patients suggests it has influenced various important aspects of end-of-life experience. Family caregivers overwhelmingly agreed that the dignity therapy document is and will continue to be a source of comfort to them and they would recommend dignity therapy to others in the same situation.CONCLUSIONS: This is the first dignity therapy study to focus on MND and on home-based caregiving. RESULTS established the importance of narrative and generativity for patients with MND and may open the door for other neurodegenerative conditions.

Publication type: Journal Article

Source: MEDLINE

Full text: Available Journal of palliative medicine at No link? Ask Salisbury Healthcare Library - please click here to request article.


Citation: Neuropathology & Applied Neurobiology, February 2015, vol./is. 41/2(201-26), 0305-1846;1365-2990 (2015 Feb)

Author(s): Raman R, Allen SP, Goodall EF, Kramer S, Ponger LL, Heath PR, Milo M, Hollinger HC, Walsh T, Highley JR, Olpin S, McDermott CJ, Shaw PJ, Kirby J

Language: English

Abstract: AIMS: Amyotrophic lateral sclerosis (ALS) and primary lateral sclerosis (PLS) are two syndromic variants within the motor neurone disease spectrum. As PLS and most ALS cases are sporadic (SALS), this limits the availability of cellular models for investigating pathogenic mechanisms and therapeutic targets. The aim of this study was to use gene expression profiling to evaluate fibroblasts as cellular models for SALS and PLS, to establish whether dysregulated biological processes recapitulate those seen in the central nervous system and to elucidate pathways that distinguish the clinically defined variants of SALS and PLS.METHODS: Microarray analysis was performed on fibroblast RNA and differentially expressed genes identified. Genes in enriched biological pathways were validated by quantitative PCR and functional assays performed to establish the effect of altered RNA levels on the cellular processes.RESULTS: Gene expression profiling demonstrated that whilst there were many differentially expressed genes in common between SALS and PLS fibroblasts, there were many more expressed specifically in the SALS fibroblasts, including those involved in RNA processing and the stress response. Functional analysis of the fibroblasts confirmed a significant decrease in miRNA production and a reduced response to hypoxia in SALS fibroblasts. Furthermore, metabolic gene changes seen in SALS, many of which were also evident in PLS fibroblasts, resulted in dysfunctional cellular respiration.CONCLUSIONS: The data demonstrate that fibroblasts can act as cellular models for ALS and PLS, by establishing the transcriptional changes in known pathogenic pathways that confer subsequent functional effects and potentially highlight targets for therapeutic intervention.Copyright 2014 The Authors. Neuropathology and Applied Neurobiology published by John Wiley & Sons Ltd. on behalf of British Neuropathological Society.

Publication type: Journal Article

Source: MEDLINE

Full text: Available Neuropathology and applied neurobiology at No link? Ask Salisbury Healthcare Library - please click here to request article.
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2. A systematic review and meta-analysis of strength training in individuals with multiple sclerosis or Parkinson disease.


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7. Towards a better understanding of MS pain: a systematic review of potentially modifiable psychosocial factors.

Multiple Sclerosis Journal Articles

   **Citation:** Therapeutic Advances in Neurological Disorders, January 2015, vol./is. 8/1(31-45), 1756-2856;1756-2856 (2015 Jan)
   **Author(s):** Havrdova E, Horakova D, Kovarova I
   **Language:** English
   **Abstract:** Alemtuzumab is a humanized monoclonal antibody therapy that has recently been approved in over 30 countries for patients with active relapsing-remitting multiple sclerosis. It acts by targeting CD52, an antigen primarily expressed on T and B lymphocytes, resulting in their depletion and subsequent repopulation. The alemtuzumab clinical development program used an active comparator, subcutaneous interferon beta-1a, to show that alemtuzumab is a highly efficacious disease-modifying therapy, with benefits on relapses, disability outcomes, and freedom from clinical disease and magnetic resonance imaging activity. The safety profile was consistent across studies and no new safety signals have emerged during follow-up in the extension study. Infusion-associated reactions are common with alemtuzumab, but rarely serious. Infection incidence was elevated with alemtuzumab in clinical studies; most infections were mild or moderate in severity. Autoimmune adverse events occurred in approximately a third of patients, manifesting mainly as thyroid disorders, and less frequently as immune thrombocytopenia or nephropathy. A comprehensive monitoring program lasting at least 4 years after the last alemtuzumab dose allows early detection and effective management of autoimmune adverse events. Further experience with alemtuzumab in the clinic will provide needed long-term data.
   **Publication type:** Journal Article, Review
   **Source:** MEDLINE
   **Full text:** Available National Library of Medicine at [Therapeutic Advances in Neurological Disorders](https://www.therapeuticsadvances.com)

2. A systematic review and meta-analysis of strength training in individuals with multiple sclerosis or Parkinson disease.
   **Citation:** Medicine, January 2015, vol./is. 94/4(e411), 0025-7974;1536-5964 (2015 Jan)
   **Author(s):** Cruickshank TM, Reyes AR, Ziman MR
   **Language:** English
   **Abstract:** Strength training has, in recent years, been shown to be beneficial for people with Parkinson disease and multiple sclerosis. Consensus regarding its utility for these disorders nevertheless remains contentious among healthcare professionals. Greater clarity is required, especially in regards to the type and magnitude of effects as well as the response differences to strength training between individuals with Parkinson disease or multiple sclerosis.
study examines the effects, magnitude of those effects, and response differences to strength training between patients with Parkinson disease or multiple sclerosis. A comprehensive search of electronic databases including Physiotherapy Evidence Database scale, PubMed, EMBASE, Cochrane Central Register of Controlled Trials, and CINAHL was conducted from inception to July 2014. English articles investigating the effect of strength training for individuals with neurodegenerative disorders were selected. Strength training trials that met the inclusion criteria were found for individuals with Parkinson disease or multiple sclerosis. Individuals with Parkinson disease or multiple sclerosis were included in the study. Strength training interventions included traditional (free weights/machine exercises) and nontraditional programs (eccentric cycling). Included articles were critically appraised using the Physiotherapy Evidence Database scale. Of the 507 articles retrieved, only 20 articles met the inclusion criteria. Of these, 14 were randomized and 6 were nonrandomized controlled articles in Parkinson disease or multiple sclerosis. Six randomized and 2 nonrandomized controlled articles originated from 3 trials and were subsequently pooled for systematic analysis. Strength training was found to significantly improve muscle strength in people with Parkinson disease (15%-83.2%) and multiple sclerosis (4.5%-36%). Significant improvements in mobility (11.4%) and disease progression were also reported in people with Parkinson disease after strength training. Furthermore, significant improvements in fatigue (8.2%), functional capacity (21.5%), quality of life (8.3%), power (17.6%), and electromyography activity (24.4%) were found in individuals with multiple sclerosis after strength training. The limitations of the study were the heterogeneity of interventions and study outcomes in Parkinson disease and multiple sclerosis trials. Strength training is useful for increasing muscle strength in Parkinson disease and to a lesser extent multiple sclerosis.

**Publication type:** Journal Article

**Source:** MEDLINE

**Full text:** Available Medicine at Medicine


**Citation:** Therapeutic Advances in Neurological Disorders, January 2015, vol./is. 8/1(20-30), 1756-2856;1756-2856 (2015 Jan)

**Author(s):** Bomprezzi R

**Language:** English

**Abstract:** Multiple sclerosis (MS) shares an immune-mediated origin with psoriasis. Long-term safety and efficacy data generated in Europe from usage of fumaric acid formulations in the latter disease constituted grounds to investigate their effects in MS patients. Dimethyl fumarate (DMF) was found to be the active principle in those formulations and in vitro studies have demonstrated that DMF has immune-modulatory properties exerted through abilities to divert cytokine production toward a Th2 profile, both on lymphocytes and microglial cells. More importantly, DMF was discovered to impact the anti-oxidative stress cell machinery promoting the transcription of genes downstream to the activation of the nuclear factor (erythroid derived 2)-like2 (NRF2). DMF exposure increases the cytosol concentrations of NRF2, which besides immune regulatory effects, has the potential for cytoprotection on glial cells, oligodendrocytes and neurons. Extensive and rigorous clinical trials have assessed the efficacy and safety of DMF at the dose of 240 mg twice and three times a day in relapsing-remitting MS patients during one phase IIb and two phase III trials. Robust, positive results were obtained across a number of clinical and paraclinical parameters. In one study (DEFINE), the relative reductions of the adjusted annualized relapse rate of the low and high dose regimens in comparison with placebo were 53% and 48%, respectively (p<0.001 for both comparisons). In the other trial (CONFIRM), DMF decreased the annualized relapse rate in comparison with placebo by 44% in the lower and by 51% in higher dosage group (also p < 0.001). The number and size of lesions as detected by magnetic resonance imaging were also significantly decreased in comparison with the patients receiving DMF at every dosage. Multiple post hoc and subgroup analyses corroborated the clinical data, rendering DMF an appealing medication whose potential for impacting the degenerative aspects of MS remains to be explored.

**Publication type:** Journal Article, Review

**Source:** MEDLINE

**Full text:** Available National Library of Medicine at Therapeutic Advances in Neurological Disorders

4. Falls in people with MS-an individual data meta-analysis from studies from Australia, Sweden, United Kingdom and the United States.

**Citation:** Multiple Sclerosis, January 2015, vol./is. 21/1(92-100), 1352-4585;1477-0970 (2015 Jan)

**Author(s):** Nilsagard Y, Gunn H, Freeman J, Hoang P, Lord S, Mazumder R, Cameron M

**Language:** English

**Abstract:** BACKGROUND: Falls are common in people with multiple sclerosis (PwMS). Previous studies have generally
included small samples and had varied methods. OBJECTIVES: The objectives of this paper are to compile fall rates across a broad range of ages and disease severity and to definitively assess the extent to which MS-associated and demographic factors influence fall rates. METHODS: Individual data from studies in four countries that prospectively measured falls for three months were analyzed. We determined fall rates, prevalence of fallers (>1 falls) and frequent fallers (>2 falls), location and timing of falls, and fall-related demographic factors. RESULTS: A total of 537 participants reported 1721 falls: 56% were fallers and 37% frequent fallers. Most falls occurred indoors (65%) between 6 a.m. and 6 p.m. (75%). Primary progressive MS was associated with significantly increased odds of being a faller (odds ratio (OR) 2.02; CI 1.08-3.78). Fall risk peaked at EDSS levels of 4.0 and 6.0 with significant ORs between 5.30 (2.23-12.64) and 5.10 (2.08-12.47). The fall rate was lower in women than men (relative risk (RR) 0.80; CI 0.67-0.94) and decreased with increasing age (RR 0.97 for each year, CI 0.95-0.98). CONCLUSION: PwMS are at high risk of falls and there are important associations between falls and MS-associated disability, gender, and age. Copyright The Author(s), 2015.

Publication type: Journal Article
Source: MEDLINE
Full text: Available ProQuest at Multiple Sclerosis Journal

Citation: Journal of Clinical Neurology, January 2015, vol./is. 11(1)(9-19), 1738-6586;1738-6586 (2015 Jan)
Author(s): Kim W, Zandona ME, Kim SH, Kim HJ
Language: English
Abstract: Classical multiple sclerosis (MS) treatments using first-line injectable drugs, although widely applied, remain a major concern in terms of therapeutic adherence and efficacy. New oral drugs recently approved for MS treatment represent significant advances in therapy. The oral route of administration clearly promotes patient satisfaction and increases therapeutic compliance. However, these drugs may also have safety and tolerability issues, and a thorough analysis of the risks and benefits is required. Three oral drugs have been approved by regulatory agencies for MS treatment: fingolimod, teriflunomide, and dimethyl fumarate. This article reviews the mechanisms of action, safety, and efficacy of these drugs and two other drugs that have yielded positive results in phase III trials: cladribine and laquinimod.

Publication type: Journal Article, Review
Source: MEDLINE
Full text: Available Journal of clinical neurology (Seoul, Korea) at Journal of Clinical Neurology

Citation: European Journal of Neurology, March 2015, vol./is. 22/3(443-e34), 1351-5101;1468-1331 (2015 Mar)
Author(s): Dalgas U, Stenager E, Sloth M, Stenager E
Language: English
Abstract: BACKGROUND AND PURPOSE: The purpose of this study was to perform a systematic review of the literature on the effects of exercise on depressive symptoms in patients with multiple sclerosis (MS), as well as to apply meta-analytical procedures to the results. METHODS: A systematic search covering eight databases was conducted. The included studies were randomized controlled trials applied to people with definite MS who completed a structured exercise intervention which were compared to any comparator, including other forms of exercise. The outcomes included a primary measure of depression/depressive symptoms or an instrument with a clearly defined depression subscale. RESULTS: Fifteen randomized controlled trial studies were identified including a total of 331 exercising subjects and 260 controls. The average Physiotherapy Evidence Database (PEDro) score was 5.6 +/- 1.3 points. Only one study applied depressive symptoms as the primary outcome. Four studies showed positive effects of exercise on depressive symptoms. An in-depth analysis of the studies revealed that the baseline level of depressive symptoms, patient disability level, choice of depression instrument and exercise intensity may influence the results. The meta-analysis included 12 studies reflecting a total of 476 subjects. The standardized mean difference across studies was g = -0.37, 95% confidence interval (-0.56; -0.17), and the null hypothesis of homogeneity within the sample could not be rejected (Q = 12.05, df = 11, P = 0.36). DISCUSSION: Exercise may be a potential treatment to prevent or reduce depressive symptoms in individuals with MS, but existing studies do not allow solid conclusions. Future well-designed studies evaluating the effects of exercise on depressive symptoms and major depression disorder in MS are highly warranted. Copyright 2014 EAN.

Publication type: Journal Article
7. Towards a better understanding of MS pain: a systematic review of potentially modifiable psychosocial factors.

**Citation:** Journal of Psychosomatic Research, January 2015, vol./is. 78/1(12-24), 0022-3999;1879-1360 (2015 Jan)

**Author(s):** Harrison AM, McCracken LM, Bogosian A, Moss-Morris R

**Language:** English

**Abstract:** OBJECTIVE: Pain is a common symptom of Multiple Sclerosis (MS). Biomedical treatments achieve only modest reductions in pain severity suggesting that this approach may be too narrow. The aim of this systematic review was to assess evidence for associations between modifiable psychosocial factors and MS pain severity and pain interference and use this evidence to develop a preliminary biopsychosocial model of MS pain.

METHODS: Empirical studies of pain in MS utilising standardised pain severity and pain interference measures were included. Online databases (Cochrane, PsychInfo, EMBASE, CINAHL, Medline, Web of Science and World Cat) and reference sections of included articles were searched, and corresponding authors contacted to identify unpublished studies. Information about design, sample size, MS type, time since diagnosis, psychosocial and pain measures and key findings were extracted. Thirty-one studies were assessed for quality and a narrative synthesis was conducted.

RESULTS: Similar to primary chronic pain, most studies reported small to medium associations between several psychosocial factors and pain severity and interference. Pain catastrophizing showed consistently strong associations with pain interference. Preliminary findings revealed a strong correlation between pain acceptance and pain interference. However, fear-avoidance appeared less important in MS, and other forms of behavioural avoidance were not explored.

CONCLUSIONS: A preliminary model of MS pain outlining specific psychosocial factors is presented with a conceptual formulation from both traditional, and contextual, cognitive-behavioural perspectives. Pain catastrophizing, acceptance, and endurance, as opposed to fear avoidance, responses are highlighted as potentially important treatment targets in MS, and directions for future research are outlined.

**Publication type:** Journal Article, Research Support, Non-U.S. Gov't

**Source:** MEDLINE

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2. Autonomic predominant multiple system atrophy in the context of Parkinsonian and cerebellar variants

3. Changes in total cell numbers of the basal ganglia in patients with multiple system atrophy - A stereological study.

5. Distinct functional and macrostructural brain changes in Parkinson's disease and multiple system atrophy.

6. Multiple system atrophy is not caused by C9orf72 hexanucleotide repeat expansions.

7. Putaminal hypointensity on T2*-weighted MR imaging is the most practically useful sign in diagnosing multiple system atrophy: A preliminary study

8. The role of transcriptional control in multiple system atrophy

9. The utility of cerebral perfusion SPECT analysis using SPM8, eZIS and vbSEE for the diagnosis of multiple system atrophy-parkinsonism.

**Multiple System Atrophy Journal Articles**

1. Application of diffusion tensor imaging in multiple system atrophy: the involvement of pontine transverse and longitudinal fibers.

**Citation:** International Journal of Neuroscience, January 2015, vol./is. 125/1(18-24), 0020-7454;1563-5279 (2015 Jan)

**Author(s):** Yang H, Wang X, Liao W, Zhou G, Li L, Ouyang L
were only observed in the putamen (p=0.04) and globus pallidus (p=0.01). In the MSA brains the total number of
oligodendrocytes (p<0.001), and, to a lesser extent in the caud
substantia nigra (p=0.001), putamen (p=0.001), and globus pallidus
were estimated in brains from 11 patients with multiple system atrophy (MSA) and 11 age
control subjects with unbiased stereological methods. Compared to the control subjects, the MSA patients had a
substantially lower number of neurons in the substantia nigra (p=0.001), putamen (p=0.001), and globus pallidus
(p<0.001), and, to a lesser extent in the caudate nucleus (p=0.03). A significantly lower number of oligodendrocytes
were only observed in the putamen (p=0.04) and globus pallidus (p=0.01). In the MSA brains the total number of


dysfunction at first examination in MSA patient that may have relevance in its early diagnosis.METHODS: Clinical
features, MSAp (Parkinsonian) and MSAc (cerebellar) variants are known. We studied the severity of auto
symptoms for several months (38.7+/-26.11, median 36 months) before the appearance of cerebellar (1 patient) or Parkinsonian (6 patients) features.CONCLUSION: Autonomic dysfunction may be the only presenting feature in some patients with MSA. Routine autonom
abnormalities were detected in hand grip test (86.8%), deep breathing test (79.2%), cold pressor test (71.7%), Valsalva
maneuver (75.5%), heart rate variability at 30:15 beats after standing (50.9%) and systolic BP on standing (41.5%). Six
(16.2%) MSAp and 1 (6.3%) MSAc patients continued to have autonomic symptoms for several months (38.7+/-26.11,
median 36 months) before the appearance of cerebellar (1 patient) or Parkinsonian (6 patients) features. Cross sign was divided into three grades as follows: 0, no cross sign; 1, vertical line only; 2, clear cross sign. Spearman rank correlation analysis was used
between FA, MD, and the cross grade in patients with MSA-C.RESULTS: FA and MD in the MSA-C group, and each cross grade, showed statistically significant differences compared to control groups. There was a close correlation between all measures. FA decreased and MD increased, and cross grade formed gradually in the patients.CONCLUSION: DTI can identify microstructural abnormalities in pontine transverse and longitudinal fibers even in patients without abnormalities on conventional MRI. Along with pontine transverse tract degeneration, the cross sign develops accompanied by the start of longitudinal tract degeneration, ultimately resulting in the complete formation of a cross sign.

Publication type: Journal Article, Research Support, Non-U.S. Gov't
Source: MEDLINE
Full text: Available The International journal of neuroscience at No link? Ask Salisbury Healthcare Library - please click here to request article.

2. Autonomic predominant multiple system atrophy in the context of Parkinsonian and cerebellar variants.

Citation: Clinical Neurology & Neurosurgery, March 2015, vol./is. 130/(110-3), 0303-8467;1872-6968 (2015 Mar)
Author(s): Tandon R, Pradhan S
Language: English
Abstract: OBJECTIVES: Autonomic dysfunction is often a late feature of multiple system atrophy (MSA). Based on early
features, MSAp (Parkinsonian) and MSAc (cerebellar) variants are known. We studied the severity of autonomic
dysfunction at first examination in MSA patient that may have relevance in its early diagnosis.METHODS: Clinical
(including autonomic), radiological and lab features of 53 MSA patients were analyzed in the context of MSAp (16
patients) and MSAc (37 patients).RESULTS: Most frequent autonomic symptoms were erectile dysfunction in 30 out of
38 males (78.9%), bladder symptoms in 35 (66%), followed by blackouts/fainting attacks in 33 (62.3%). Autonomic
abnormalities were detected in hand grip test (86.8%), deep breathing test (79.2%), cold pressor test (71.7%), Valsalva
maneuver (75.5%), heart rate variability at 30:15 beats after standing (50.9%) and systolic BP on standing (41.5%). Six
(16.2%) MSAp and 1 (6.3%) MSAc patients continued to have autonomic symptoms for several months (38.7+/-26.11,
median 36 months) before the appearance of cerebellar (1 patient) or Parkinsonian (6 patients) features. Cross sign was divided into three grades as follows: 0, no cross sign; 1, vertical line only; 2, clear cross sign. Spearman rank correlation analysis was used
between FA, MD, and the cross grade in patients with MSA-C. RESULTS: FA and MD in the MSA-C group, and each cross grade, showed statistically significant differences compared to control groups. There was a close correlation between all measures. FA decreased and MD increased, and cross grade formed gradually in the patients. CONCLUSION: DTI can identify microstructural abnormalities in pontine transverse and longitudinal fibers even in patients without abnormalities on conventional MRI. Along with pontine transverse tract degeneration, the cross sign develops accompanied by the start of longitudinal tract degeneration, ultimately resulting in the complete formation of a cross sign.

Publication type: Journal Article
Source: MEDLINE
Full text: Available Clinical neurology and neurosurgery at No link? Ask Salisbury Healthcare Library - please click here to request article.

3. Changes in total cell numbers of the basal ganglia in patients with multiple system atrophy - A stereological study.

Citation: Neurobiology of Disease, February 2015, vol./is. 74/(104-13), 0969-9961;1095-953X (2015 Feb)
Author(s): Salvesen L, Ullerup BH, Sunay FB, Brudek T, Lokkegaard A, Agander TK, Winge K, Pakkenberg B
Language: English
Abstract: Total numbers of neurons, oligodendrocytes, astrocytes, and microglia in the basal ganglia and red nucleus
were estimated in brains from 11 patients with multiple system atrophy (MSA) and 11 age- and gender-matched
control subjects with unbiased stereological methods. Compared to the control subjects, the MSA patients had a
substantially lower number of neurons in the substantia nigra (p=0.001), putamen (p=0.001), and globus pallidus
(p<0.001), and, to a lesser extent in the caudate nucleus (p=0.03). A significantly lower number of oligodendrocytes
were only observed in the putamen (p=0.04) and globus pallidus (p=0.01). In the MSA brains the total number of
astrocytes was significantly higher in the putamen (p=0.04) and caudate nucleus (p=0.01). In all examined regions a higher number of microglia were found in the MSA brains with the greatest difference observed in the otherwise unaffected red nucleus (p=0.001). The results from the stereological study were supported by cell marker expression analyses showing increased markers for activated microglia. Our results suggest that microgliosis is a consistent and severe neuropathological feature of MSA, whereas no widespread and substantial loss of oligodendrocytes was observed. We have demonstrated significant neuronal loss in the substantia nigra, striatum, and globus pallidus of patients with MSA, while neurons in other basal ganglia nuclei were spared, supporting the region-specific patterns of neuropathological changes in MSA. Copyright 2014 Elsevier Inc. All rights reserved.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available Neurobiology of disease at [No link? Ask Salisbury Healthcare Library - please click here to request article.](#)
OBJECTIVE: To identify useful MRI abnormalities in the putamen for diagnosing multiple system atrophy.

METHODS: Patients with multiple system atrophy (n=15), Parkinson’s disease (n=16), or progressive supranuclear palsy (n=9) and healthy controls (n=10) were enrolled. Using a visual analog scale, 4 examiners independently rated high-intensity signals along the lateral putamen on T2-weighted and T2*-weighted images, low-intensity signals within the putamen on T2-weighted and T2*-weighted images, and putaminal atrophy. Receiver operating characteristic analyses were performed, and the area under the receiver operating characteristic curve was calculated.

RESULTS: For differentiating multiple system atrophy from progressive supranuclear palsy, Parkinson’s disease, and healthy controls, the mean area under the curve values was the highest for low-intensity signals within the putamen on T2*-weighted images (0.797, 0.867, 0.896, respectively). Variations in the area under the curve values among the 4 examiners were the smallest in low-intensity signals within the putamen on T2*-weighted images. Good inter-rater reliability was achieved for low-intensity signals within the putamen on T2*-weighted images and high-intensity signals along the lateral putamen on T2*-weighted images.

CONCLUSION: Low-intensity signals within the putamen on T2*-weighted images is the most useful MRI abnormality for diagnosing multiple system atrophy.

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Publication type: Journal Article
Source: MEDLINE
Full text: Available Journal of the neurological sciences at No link? Ask Salisbury Healthcare Library - please click here to request article.

8. The role of transcriptional control in multiple system atrophy.

Citation: Neurobiology of Aging, January 2015, vol./is. 36/1(394-400), 0197-4580;1558-1497 (2015 Jan)

Author(s): Chen J, Mills JD, Halliday GM, Janitz M

Language: English

Abstract: Multiple system atrophy (MSA) is an alpha-synucleinopathy that is clinically characterized by varying degrees of parkinsonian, autonomic, and cerebellar features. Unlike other alpha-synucleinopathies such as Parkinson’s disease, MSA is unique in that the principal alpha-synuclein lesions, called glial cytoplasmic inclusions, occur in oligodendroglia rather than neurons, with significantly more alpha-synuclein accumulating in MSA brain compared with Parkinson’s disease. Although well defined clinically, the molecular pathophysiology of MSA has barely been investigated. In particular, there have been no systematic studies of the perturbation of the brain transcriptome during the onset and progression of this disease. Interestingly, measurements of alpha-synuclein gene (SNCA) expression in MSA brain tissue have not revealed overexpression of this gene in oligodendroglia or neurons. It has therefore become clear that other genes and gene networks, both directly as noncoding RNAs or through protein products, contribute to the accumulation of the alpha-synuclein protein in the brain. This review provides a summary of current developments in the investigation of the transcriptional causes of MSA and outlines perspectives for future research toward the elucidation of the molecular pathology of MSA-specific neurodegeneration. Copyright 2015 Elsevier Inc. All rights reserved.

Publication type: Journal Article, Research Support, Non-U.S. Gov't
Source: MEDLINE
Full text: Available Neurobiology of aging at No link? Ask Salisbury Healthcare Library - please click here to request article.

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9. The analysis of C9orf72 repeat expansions in a large series of clinically and pathologically diagnosed cases with atypical parkinsonism

10. Therapeutic strategies to prevent and manage dyskinesias in Parkinson's disease

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**Parkinson's Disease and Atypical Parkinsonism Journal Articles**

1. **A Meta-Analysis on the Efficacy of Tai Chi in Patients with Parkinson's Disease between 2008 and 2014.**
   **Citation:** Evidence-Based Complementary & Alternative Medicine: eCAM, 2015, vol./is. 2015/(593263), 1741-427X;1741-427X (2015)
   **Author(s):** Zhou J, Yin T, Gao Q, Yang XC
   **Language:** English
   **Abstract:** Objective. The purpose of this systematic review is to evaluate the evidence on the effect of Tai Chi for Parkinson's disease (PD). Methods. Six electronic databases up to June 2014 were searched. The methodological quality was assessed with PEDro scale. Standardised mean difference and 95% confidence intervals of random-effects model were calculated. Results. Nine studies were included in our review. The aggregated results are in favor of Tai Chi on improving motor function (P = 0.002) and balance (P < 0.00001) in patients with PD. However, there is no sufficient evidence to support or refute the value of Tai Chi on improving gait velocity (P = 0.11), stride length (P = 0.21), or quality of life (P = 0.40). And there is no valid evidence in follow-up effects of Tai Chi for PD. Conclusion. The current results suggest that Tai Chi can significantly improve the motor function and balance in patients with PD, but there is indeed not enough evidence to conclude that Tai Chi is effective for PD because of the small treatment effect, methodological flaws of eligible studies, and insufficient follow-up. Consequently, high-quality studies with long follow-up are warranted to confirm current beneficial findings.
   **Publication type:** Journal Article, Review
   **Source:** MEDLINE
   **Full text:** Available National Library of Medicine at Evidence-based Complementary and Alternative Medicine : eCAM

2. **A systematic review of treatments for Impulse Control Disorders and related behaviours in Parkinson's disease.**
   **Citation:** Psychiatry Research, February 2015, vol./is. 225/3(402-6), 0165-1781;1872-7123 (2015 Feb 28)
   **Author(s):** Tanwani P, Fernie BA, Nikcevic AV, Spada MM
   **Language:** English
   **Abstract:** Impulse Control Disorders (ICDs) are a set of behaviours characterised by impulsivity despite known harm. Related to ICDs is the dopamine dysregulation syndrome (DDS), which is characterised by an addiction-like consumption of dopaminergic medication and punding. These behaviours all have an increased prevalence in Parkinson's disease (PD). The aim of this review is to identify treatments available for patients suffering from ICDs, DDS and punding in PD. Searches of The Cochrane Controlled Trials Register, Embase, Medline and PsychInfo were conducted, using the entire timescale available. Seven out of the 688 papers retrieved met the inclusion criteria and were considered in this systematic review. One class I study, one class II study, and five class IV studies were identified. All studies demonstrated a positive effect on ICDs in PD. Research in this field is still in its early stages. At present, there is insufficient evidence to recommend any treatment over another. There is a need for more methodologically robust research, using larger, more generalisable samples, randomisation and meaningful follow-up periods. In addition, the use of a validated outcome measures should be implemented in future research efforts. Copyright 2014 Elsevier

Citation: Neurologic Clinics, February 2015, vol./iss. 33/1(39-56), 0733-8619;1557-9875 (2015 Feb)

Author(s): Stamelou M, Bhatia KP

Language: English

Abstract: Atypical parkinsonism comprises typically progressive supranuclear palsy, corticobasal degeneration, and multifluid system atrophy, which are distinct pathologic entities; despite ongoing research, their cause and pathophysiology are still unknown, and there are no biomarkers or effective treatments available. The expanding phenotypic spectrum of these disorders as well as the expanding pathologic spectrum of their classic phenotypes makes the early differential diagnosis challenging for the clinician. Here, clinical features and investigations that may help to diagnose these conditions and the existing limited treatment options are discussed. Copyright 2015 Elsevier Inc. All rights reserved.

4. Conformation determines the seeding potencies of native and recombinant tau aggregates.

Citation: Journal of Biological Chemistry, January 2015, vol./iss. 290/2(1049-65), 0021-9258;1083-351X (2015 Jan 9)


Language: English

Abstract: Intracellular Tau inclusions are a pathological hallmark of several neurodegenerative diseases, collectively known as the tauopathies. They include Alzheimer disease, tangle-only dementia, Pick disease, argyrophilic grain disease, chronic traumatic encephalopathy, progressive supranuclear palsy, and corticobasal degeneration. Tau pathology appears to spread through intercellular propagation, requiring the formation of assembled "prion-like" species. Several cell and animal models have been described that recapitulate aspects of this phenomenon. However, the molecular characteristics of seed-competent Tau remain unclear. Here, we have used a cell model to understand the relationships between Tau structure/phosphorylation and seeding by aggregated Tau species from the brains of mice transgenic for human mutant P301S Tau and full-length aggregated recombinant P301S Tau. Deletion of motifs (275)VQIINK(280) and (306)VQIVYK(311) abolished the seeding activity of recombinant full-length Tau, suggesting that its aggregation was necessary for seeding. We describe conformational differences between native and synthetic Tau aggregates that may account for the higher seeding activity of native assembled Tau. When added to aggregated Tau seeds from the brains of mice transgenic for P301S Tau, soluble recombinant Tau aggregated and acquired the molecular properties of aggregated Tau from transgenic mouse brain. We show that seeding is conferred by aggregated Tau that enters cells through macropinocytosis and seeds the assembly of endogenous Tau into filaments. Copyright 2015 by The American Society for Biochemistry and Molecular Biology, Inc.

5. Disorders of the oral cavity in Parkinson's disease and parkinsonian syndromes.

Citation: Parkinsons Disease, 2015, vol./iss. 2015/(379482), 2090-8083;2042-0080 (2015)

Author(s): Zlotnik Y, Balash Y, Korczyn AD, Giladi N, Gurevich T

Language: English

Abstract: Awareness of nonmotor symptoms of Parkinson’s disease is growing during the last decade. Among these, oral cavity disorders are, although prevalent, often neglected by the patients, their caregivers, and physicians. Some of these disorders include increased prevalence of caries and periodontal disease, sialorrhea and drooling, xerostomia, orofacial pain, bruxism, and taste impairment. Though many of these disorders are not fully understood yet and relatively few controlled trials have been published regarding their treatment, physicians should be aware of the body of evidence that does exist on these topics. This paper reviews current knowledge
regarding the epidemiology, pathophysiology, and treatment options of disorders of the oral cavity in Parkinson’s disease patients.

**Publication type:** Journal Article, Review  
**Source:** MEDLINE  
**Full text:** Available National Library of Medicine at Parkinson’s Disease

**Citation:** Neurology Research International, 2015, vol./is. 2015/(345285), 2090-1852;2090-1860 (2015)  
**Author(s):** Chauhan A, Jeans AF  
**Language:** English  
**Abstract:** Parkinson’s disease (PD) is the world’s second most common neurodegenerative disease and most common movement disorder. Characterised by a loss of dopaminergic neurons and the development of intraneuronal inclusions known as Lewy bodies, it has classically been thought of as a cell-autonomous disease. However, in 2008, two groups reported the startling observation of Lewy bodies within embryonic neuronal grafts transplanted into PD patients little more than a decade previously, suggesting that PD pathology can be propagated to neighbouring cells and calling basic assumptions of our understanding of the disease into question. Subsequent research has largely served to confirm this interpretation, pointing towards a prion-like intercellular transfer of misfolded alpha-synuclein, the main component of Lewy bodies, as central to PD. This shift in thinking offers a revolutionary approach to PD treatment, potentially enabling a transition from purely symptomatic therapy to direct targeting of the pathology that drives disease progression. In this short review, we appraise current experimental support for PD as a prion-like disease, whilst highlighting areas of controversy or inconsistency which must be resolved. We also offer a brief discussion of the therapeutic implications of these discoveries.

**Publication type:** Journal Article, Review  
**Source:** MEDLINE  
**Full text:** Available National Library of Medicine at Neurology Research International

**Citation:** Journal of Neurosciences in Rural Practice, January 2015, vol./is. 6/1(65-76), 0976-3147;0976-3155 (2015 Jan)  
**Author(s):** Grover S, Somaiya M, Kumar S, Avasthi A  
**Language:** English  
**Abstract:** Parkinson’s disease (PD) is essentially characterized by the motor symptoms in the form of resting tremor, rigidity and bradykinesia. However, over the years it has been recognized that motor symptoms are just the "tip of the iceberg" of clinical manifestations of PD. Besides motor symptoms, PD characterized by many non-motor symptoms, which include cognitive decline, psychiatric disturbances (depression, psychosis and impulse control), sleep difficulties, autonomic failures (gastrointestinal, cardiovascular, urinary, thermoregulation) and pain syndrome. This review evaluates the various aspects of psychiatric disorders including cognitive decline and sleep disturbances in patients with PD. The prevalence rate of various psychiatric disorders is high in patients with PD. In terms of risk factors, various demographic, clinical and treatment-related variables have been shown to be associated with higher risk of development of psychiatric morbidity. Evidence also suggests that the presence of psychiatric morbidity is associated with poorer outcome. Randomized controlled trials, evaluating the various pharmacological and non-pharmacological treatments for management of psychiatric morbidity in patients with PD are meager. Available evidence suggests that tricyclic antidepressants like desipramine and nortriptyline are efficacious for management of depression. Among the antipsychotics, clozapine is considered to be the best choice for management of psychosis in patients with PD. Among the various cognitive enhancers, evidence suggest efficacy of rivastigmine in management of dementia in patients with PD. To conclude, this review suggests that psychiatric morbidity is highly prevalent in patients with PD. Hence, a multidisciplinary approach must be followed to improve the overall outcome of PD. Further studies are required to evaluate the efficacy of various other measures for management of psychiatric morbidity in patients with PD.

**Publication type:** Journal Article, Review  
**Source:** MEDLINE  
**Full text:** Available Journal of Neurosciences in Rural Practice at Journal of Neurosciences in Rural Practice  
**Full text:** Available Journal of Neurosciences in Rural Practice at No link? Ask Salisbury Healthcare Library - please click here to request article.
8. Safety and efficacy of rasagiline in addition to levodopa for the treatment of idiopathic Parkinson's disease: a meta-analysis of randomised controlled trials.

Citation: European Neurology, 2015, vol./is. 73/1-2(5-12), 0014-3022;1421-9913 (2015)

Author(s): Cai JP, Chen WJ, Lin Y, Cai B, Wang N

Language: English

Abstract: BACKGROUND: To assess the safety and efficacy of rasagiline for the treatment of Parkinson's disease (PD) among individuals currently receiving levodopa.

METHODS: A systematic literature search was conducted to identify randomised controlled trials (RCT) comparing rasagiline with placebo/no treatment in individuals with PD currently receiving levodopa. Outcome measures included improvement in motor functions; symptomatic improvement; improvement in quality of life; adverse effects. Random-effect meta-analytical techniques were conducted for the outcome measure and subgroup analyses.

RESULTS: Three RCTs were included (n = 1002). The results showed significantly greater improvements in daily 'on' time without dyskinesia in levodopa-treated participants with idiopathic PD receiving 1 mg/day rasagiline compared to placebo (n = 712, 2 RCTs, MD 0.80, CI 0.45 to 1.15; p < 0.00001), and significantly greater improvements in Unified Parkinson's Disease Rating Scale motor performance scores during 'on' time in participants receiving 0.5-1 mg/day rasagiline (0.5 mg/day: n = 282, MD -2.91, CI -4.59 to -1.23; p = 0.0007; 1 mg/day: n = 712, 2 RCTs, MD -2.91, CI -4.02 to -1.80; p < 0.00001). There were no significant differences in adverse effects.

CONCLUSION: 0.5 to 1 mg/day rasagiline in addition to levodopa is a safe and well-tolerated combination therapy for individuals with Parkinson's disease. 2014 S. Karger AG, Basel.

Publication type: Journal Article

Source: MEDLINE

Full text: Available European neurology at No link? Ask Salisbury Healthcare Library - please click here to request article.

9. The analysis of C9orf72 repeat expansions in a large series of clinically and pathologically diagnosed cases with atypical parkinsonism.

Citation: Neurobiology of Aging, February 2015, vol./is. 36/2(1221.e1-6), 0197-4580;1558-1497 (2015 Feb)


Language: English

Abstract: A GGGGCC repeat expansion in the C9orf72 gene was recently identified as a major cause of familial and sporadic amyotrophic lateral sclerosis and frontotemporal dementia. There is suggestion that these expansions may be a rare cause of parkinsonian disorders such as progressive supranuclear palsy (PSP), multiple system atrophy (MSA), and corticobasal degeneration (CBD). Screening the C9orf72 gene in 37 patients with features of corticobasal syndrome (CBS) detected an expansion in 3 patients, confirmed by Southern blotting. In a series of 22 patients with clinically diagnosed PSP, we found 1 patient with an intermediate repeat length. We also screened for the C9orf72 expansion in a large series of neuropathologically confirmed samples with MSA (n = 96), PSP (n = 177), and CBD (n = 18). Patients were found with no more than 22 GGGGCC repeats. Although these results still need to be confirmed in a larger cohort of CBS and/or CBD patients, these data suggest that in the presence of a family history and/or motor neuron disease features, patients with CBS or clinical PSP should be screened for the C9orf72 repeat expansion. In addition, we confirm that the C9orf72 expansions are not associated with pathologically confirmed MSA, PSP, or CBD in a large series of cases. Copyright 2015 The Authors. Published by Elsevier Inc. All rights reserved.

Publication type: Journal Article

Source: MEDLINE

Full text: Available Neurobiology of aging at No link? Ask Salisbury Healthcare Library - please click here to request article.

10. Therapeutic strategies to prevent and manage dyskinesias in Parkinson's disease.

Citation: Expert Opinion on Drug Safety, February 2015, vol./is. 14/2(281-94), 1474-0338;1744-764X (2015 Feb)

Author(s): Pilleri M, Antonini A

Language: English

Abstract: INTRODUCTION: Chronic treatment with levodopa is associated with the development of motor fluctuations and dyskinesias particularly in young Parkinson patients. In some cases, dyskinesias become so severe that they interfere with normal movement and negatively impact quality of life.

AREAS COVERED: In this review,
we discuss benefits and limits of available therapeutic approaches aimed at delaying or managing dyskinesias as well as new strategies that are currently under investigation.

EXPERT OPINION: Among available treatments, monotherapy with dopamine agonists in the early phases of the disease reduces the risk for dyskinesias compared with levodopa. Nevertheless, dopamine agonists are unable to prevent dyskinesias once levodopa is added, which is always required once disease severity progresses. Convincing evidence of dyskinesia improvement has been shown only for deep brain stimulation and to some extent also for duodenal levodopa infusion and subcutaneous apomorphine. These approaches are expensive, have restrictive inclusion criteria and can cause potentially serious side effects. Alternative therapies include drugs targeting nondopaminergic neurotransmitter systems. Amantadine improves dyskinesias but its long-term effect is often unsatisfactory. Glutamatergic and gabaergic compounds have been tested in clinical trials, with promising results. By contrast, adrenergic drugs, fipamezole and idazoxan, did not show antidyskinetic effect.

**Publication type:** Journal Article  
**Source:** MEDLINE  
**Full text:** Available Expert opinion on drug safety at No link? Ask Salisbury Healthcare Library - please click here to request article.


**Citation:** Neurological Sciences, February 2015, vol./is. 36/2(275-9), 1590-1874;1590-3478 (2015 Feb)  
**Author(s):** Gomez-Caravaca MT, Caceres-Redondo MT, Huertas-Fernandez I, Vargas-Gonzalez L, Carrillo F, Carballo M, Mir P  
**Language:** English  
**Abstract:** Drooling is a common symptom in parkinsonian disorders. Our aim was to assess the safety and effect of botulinum toxin when applied to parotid glands without ultrasound guidance for sialorrhea in parkinsonian disorders in a retrospective study with a long-term follow-up. We evaluated 53 patients (64.2 % male and 35.8 % female) with a mean age of 70.18 +/- 9.25 years who were treated in our centre between 2007 and 2013. We analysed the mean dose, latency, effect duration, response and adverse effects of treating sialorrhea by injecting botulinum toxin type A (Botox) into the parotid glands without ultrasound guidance. A total of 41 patients with Parkinson's disease, 6 with progressive supranuclear palsy, 4 with multiple system atrophy and 2 with corticobasal degeneration were included. The mean duration of the disease at onset was 10.51 +/- 6.81 years and the mean sialorrhea duration was 1.99 +/- 1.55 years. The initial dose used for each parotid gland was 14.53 +/- 3.95 units of Botox, with a mean dose of 22.17 +/- 8.76 units. There was an improvement after treatment in 65.22 % of patients with an average score of 6.85 +/- 1.58 points on a scale from 0 to 10. The duration of the treatment effect was 4.38 +/- 2.11 months, with a latency period of 10.06 +/- 9.63 days. Adverse effects were mild and infrequent. Botulinum toxin is a safe and effective therapy for the treatment of sialorrhea in parkinsonian disorders and there is no requirement for ultrasound guidance. It has a rapid onset and lasting effect without requiring a high dosage.  
**Publication type:** Journal Article  
**Source:** MEDLINE
PPN stimulation in progressive supranuclear palsy (PSP) patients with short-term (6 months) and long-term (18 months) follow-ups. Patients with PSP who had gait disturbances, but were able to walk with or without assistance, were selected. The patients' median age was 64 years and the disease duration 3 years. Bilateral PPN deep brain stimulation (DBS) was performed. The pacemaker was programmed using a bipolar mode and lower frequencies (20-45 Hz). The PSP rating scores (PSPRS) and their gait subscores (No. 25, 26, 27 and 28) along with PSP staging scores were used as primary end points. The total Unified Parkinson’s Disease Rating Scale (UPDRS), UPDRS III and the 39-item Parkinson’s Disease Questionnaire were considered as secondary end points. Video recordings of the gait were performed before surgery and at the 6- and 18-month follow-ups. These were retrospectively reviewed by a blinded neurologist for the primary end points. At the 6- and 18-month follow-ups, the median change in PSPRS was from 33 (baseline) to 37.5 and 47, respectively. Similarly, the PSP staging changed from 3 to 2.5 and 3.5, item 25 from 1.5 to 2 and 3.5, item 26 from 2.5 to 2 and 3.5, item 27 from 3.5 to 3 and 3.5 and item 28 from 1.5 to 1.5 and 3. Two patients in the study with the PSP-parkinsonism phenotype experienced improvement in their gait until the last follow-up. Bilateral PPN DBS can be safely performed in PSP patients despite mid-brain atrophy. 2015 S. Karger AG, Basel.

Publication type: Journal Article
Source: MEDLINE
Full text: Available Stereotactic and functional neurosurgery at No link? Ask Salisbury Healthcare Library - please click here to request article.

2. Three sib-pairs of autopsy-confirmed progressive supranuclear palsy.
Citation: Parkinsonism & Related Disorders, February 2015, vol./is. 21/2(101-5), 1353-8020;1873-5126 (2015 Feb)
Author(s): Fujioka S, Sanchez Contreras MY, Strongosky AJ, Ogaki K, Whaley NR, Tacik PM, van Gerpen JA, Uitti RJ, Ross OA, Wszolek ZK, Rademakers R, Dickson DW
Language: English
Abstract: OBJECTIVE: To describe the clinical, pathological, and genetic features of three sib-pairs of pathologically-confirmed progressive supranuclear palsy (PSP).METHODS: We searched the Mayo Clinic neurodegenerative diseases brain bank for cases of PSP in which more than one family member had pathologically-confirmed PSP. Clinical and pathological data were reviewed and all individuals were screened for mutations in MAPT, by sequencing exons 1, 7, and 9-13.RESULTS: We identified three sib-pairs of pathologically-confirmed PSP. Sufficient information was available to suggest an autosomal dominant inheritance in two. The mean age at symptom onset was 41 years in one pair, and 76 years in the other two. The young onset pair had a p.S285R mutation in MAPT, but no mutations were detected in the other two.CONCLUSIONS: All sib-pairs had typical pathological features of PSP; however, the age at onset of the sib-pair with MAPT mutation was significantly younger than sporadic PSP. Future studies are warranted to identify a possible genetic basis for PSP associated with late onset and typical PSP pathology.Copyright 2014 Elsevier Ltd. All rights reserved.
Publication type: Journal Article
Source: MEDLINE
Full text: Available Parkinsonism & related disorders at No link? Ask Salisbury Healthcare Library - please click here to request article.

3. Utility of Frontal Assessment Battery in detection of neuropsychological dysfunction in Richardson variant of progressive supranuclear palsy.
Citation: Neurologia i Neurochirurgia Polska, 2015, vol./is. 49/1(36-40), 0028-3843;0028-3843 (2015)
Author(s): Sitek EJ, Konkel A, Dabrowska M, Slawek J
Language: English
Abstract: Progressive supranuclear palsy is characterized by motor, cognitive and behavioral features. In Richardson's syndrome of PSP (PSP-RS) executive dysfunction is quite prominent. Frontal Assessment Battery (FAB) is one of the most popular screening tests in the differential diagnosis of bradykinetic rigid syndromes. The study aimed at analyzing FAB subscores in relation to neuropsychological assessment results. Twenty patients with PSP-RS (12 with probable and eight with possible diagnosis) participated in the study. Sixteen PSP-RS patients scored below 15 on FAB. Among four patients having scored above cut-off (12 points) on FAB, two demonstrated both executive and language deficits, while the other two presented with only selective executive deficits on comprehensive neuropsychological evaluation. FAB is a useful screening measure in PSP, but it may not detect subtle executive deficits. Moreover, language performance seems to contribute significantly to FAB scores. Thus, FAB should be treated as "frontal" rather than "executive" screening task, in line with its name. Copyright 2014 Polish Neurological Society. Published by Elsevier Urban & Partner Sp. z o.o. All rights reserved.
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