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| **Study** | **HD****Agea** | **Disease Duration (yrs) or HD stage** | **Drug** | **Study Design** | **Outcome Measure(s)** | **Summary of Primary Findings** |
| Agid et al., 1983 [1] | 38 (2) | 6 (1) | FK 33-824 | crossover, placebo controlled | study specific (involuntary movements per minute) | involuntary movements did not improve after administration of drug |
| Albanese et al., 1995 [2] | range (21-61) | 4.4 | Apomorphine | randomized, placebo controlled | Study specific (motor performance, eye movements, chorea, gait, & posture) | subcutaneous injection improved motor performance |
| Aminoff & Marshall, 1974 [3] | range (35-64) | range (5-20) | Lithium Carbonate (Priadel) | placebo controlled | WAIS FSIQ, Study specific (mood, behavior, mobility, dressing, bathing, feeding) | no improvements over placebo |
| Asher & Aminoff, 1981 [4] | range (26-65) | --- | Tetrabenazine | crossover placebo | study specific (movement severity ratings) | Of 26 patients, 15 improved, 10 were unchanged, and 1 worsened |
| Bamford et al., 1995 [5] | 40 (10.3)39.2 (10.4) | ---- | Baclofen | placebo controlled | TFC, Stroop, Trailmaking Test, PPVT, WAIS Digit Symbol, WAIS Block Design, Verbal Fluency  | some improvements in chorea; less improvement for dystonic/athetoctic states |
| Bassi et al., 1986 [6] | 44.1 (14.2) | 5 | Transdihydrolisuride (TDHL) | placebo controlled | Raven’s Progressive Matrices Test, Kohs Cubes Syndrom Kurz test, Study specific—manual dexterity, gait, speech, chorea severity | improvements in disease severity (as determined by a composite score) in 7 of 10 patients |
| Beglinger et al., 2009 [7] | F&S Stage 1,2 | ---- | Atomoxetine | randomized, placebo controlled | Trailmaking Test, WAIS Digit Symbol, WAIS Letter Number, Stroop, Verbal Fluency, CAARS, UHDRS-m, SCL-90-R | no improvements over placebo |
| Beister et al., 2004 [8] | 39.7 | 2.5 | Memantine | open label | UHDRSb, AIMS, CGI, HD-ADL | no significant increases in chorea (i.e., neuroprotective effect) |
| Bender et al., 2005 [9] | 46 (7.3) | 4 (2.1) | Creatine | ---- | UHDRS-m, TFC, MMSE, H-MRS as metabolite level marker | no effect on clinical ratings |
| Blackwell et al., 2008 [10] | 57 (4) | ----- | Modafinal  | randomized, placebo-controlled crossover | CANTAB, physiology | single dose improved altertness; no significant improvements in cognitive function or mood |
| Bonelli et al., 2004 [11] | 40 (10.3) | 4.5 (2.2) | Minocycline | open label | UHDRS, MMSE | improvement in motor and cognition at 6 months; stabilization in motor and cognition at 3 years |
| Bonelli et al., 2003 [12] | 41.4 (9.1);33.2 (3.4) | 4.8 (2.6)5 (2.9) | Minocycline | age/gender matched controls | UHDRS, MMSE | improvements in motor function and cognition after 6 months |
| Bonelli et al., 2002 [13] | 39.2 (10.1) | 6.3 (2.6) | Olanzapine  | open label | UHDRS | significant improvements on most subscales of the UHDRS |
| Bonelli et al., 2002 [14] | 30 | 6 | high-dose Olanzapine | case study | UHDRS | improvement in grave choreatic attacks |
| Bonuccelli et al., 1994 [15] | 51.8 (8.6) | 7.2 (3.5) | Clozapine | open label | AIMS, M&Q scale, HAM, BPRS | improvements in motor functioning; no side effects during long-term treatment |
| Braun et al., 1989 [16] | range (24-77) | range (2-20) | SKF39393  | placebo controlled | AIMSb | no significant motor improvements |
| Brusa et al., 2009 [17] | 56.3 (12.4) | ---- | Aripiprazole (AP) & tetrabenazine (TBZ) | crossover | UHDRS (motor exam), CGI, HAM, ESS, MMSE | both AP & TBZ improved chorea & motor performance; TBZ associated with increased sleepiness  |
| Caine et al., 1979 [18] | 51.3 | ---- | Clozapine | crossover, placebo controlled | study specific (abnormal movements) | 2 of 3 patients had improvements in abnormal movements |
| Caine et al., 1978 [19] | 51.9 | range (1-8) | Chlorpromazine, Carbidopa followed by Levodopa, Bromocriptine | randomized, placebo | plasma prolactin (PRL) levels | all treatments showed diminished PRL response (i.e., support for dopamine mediated HPA function) |
| Cankurtaran et al., 2006 [20] | 32 | 8 | Risperidone | ---- | AIMS, BPRS, MMSE | psychotic symptoms disappeared and improvements in motor symptoms |
| Caraceni et al., 1980 [21] | 42.6 | 5.9 | Apomorphine, Bromocryptine, Lysuride, Diazepam, Cyproheptadine | ---- | study specific (abnormal involuntary movements, physiology) | Apomorphine, diazepam, and cypro improved movements; bromocryptine and lysuride had no effect or worsened movements |
| Caraceni et al., 1978 [22] | 49 | 4.9 | Deanol | crossover, randomized | Wechsler Memory Scale, study specific (hyperkinesia) | no improvements in chorea |
| Como et al., 1997 [23] | 43.7a (11.4) | 6.6 (4.5) | Fluoxetine | randomized, placebo controlled | TFC, HAM, MMSE, CBRS, Buschke Selective Reminding Test, Benton Visual Retention Test, Verbal Fluency, Ruff Figural Fuency Test, Trailmaking Test, Stroop, Digit Symbol Modalities Test | no improvements |
| Consroe et al., 1991 [24] | 47.8 (15.3) | 5 (2.8) | Cannabidiol (CBD) | randomized, crossover, placebo controlled | M&Q scale, TFC, Cannabis side effect inventory, plasma CBD levels | no group differences between placebo and CBD on chorea severity |
| Consroe et al., 1991 [25] | median 52.2, range (17-66) | ---- | oral cannabidiol | randomized, placebo controlled crossover | Plasma CBD levels | elimination half-life of CBD estimated at 2-5 days |
| Constantinescu et al., 2011 [26] | 47 (14) | 7 (4.0) | OPC-14117 (free-radical scavenger) | randomized, placebo controlled | UHDRS, CGI, BDI, Trailmaking Test, Hopkins Verbal Learning Test, WAIS Digit Span, Digit Ordering Task, Brief test of Attention, Luria Nebraska Mental Rotation Item, CSF [tau] | CSF total tau may be reliable biomarker but is not appropriate for use in isolation for diagnosis or clinical assessment |
| Corsini et al., 1978 [27] | 47.5 | 4.3 | Apomorphine HCl | placebo controlled | EEG, polygraph (for abnormal movements) | improvements in abnormal movements |
| Cubo et al., 2006 [28] | 46.2 (9.2) | 4.6 (3.4) | Donepezil | randomized, placebo controlled | UHDRS, MMSE, ADAS-c, SIPb | no improvements (chorea, cognition, nor quality of life) |
| Cudkowicz, 2004 [29] | 46.7 (8.9)47.4 (9.3) | 5.3 (3.5)6.9 (3.9) | Minocycline | randomized, placebo controlled | UHDRS, tolerability | drug well tolerated |
| Cudkowicz, 2010 [30] | 47.1 (10.3) | 6.4 (5.4) | Minocycline | randomized, controlled | TFC, UHDRS-m, UHDRS-f, UHDRS-I | Safe and tolerable;  |
| Curtis et al., 2009 [31] | 50.6 (9.5)54.3 (9.4) | ----- | Nabilone | crossover, placebo controlled | UHDRS-m, UHDRS-c, UHDRS-b, NPI | no improvement in motor or cognitive functioning. |
| Dallocchio et al., 1999 [32] | 51 (7) | 6 (3) | Risperidone | ---- | M&Q scale | improvement in motor disability; no side effects |
| Danivas et al., 2013 [33] | 585148 | 2343 | Lithium | Case studies | Study specific (patient reported abnormal movements) | abnormal movements did not worsen |
| Davis & Berger, 1978 [34] | Range (47-59) | ≥10 | Physotigmine, choline chloride | ---- | study specific (abnormal movements) | improvement in involuntary movements in 3 of 6 patients |
| Davis et al., 1978 [35] | ---- | -- | Physotigmine | placebo controlled | study specific (abnormal movements) | improvements in abnormal movements for 50% of patients |
| De Tommaso et al., 2004 [36] | 53.0 (8.7) | 8.9 (4) | Rivastigmine | randomized, controlled | TFC, M&Q scale, MMSE, AIMS | trend for improvements in cognition and movements |
| De Tommaso et al., 2005 [37] | 53.2 (7.8) | 7.4 (3.2) | Levetiracetam (LEV)  | age/gender matched controls | UHDRS-m, UHDRS-b, TFC, MMSE | small improvement in involuntary movements and functional ability |
| De Tommaso et al., 2007 [38] | 55.7 (13) | 8.9 | Rivastigmine | HD controls  | UHDRS-m, UHDRS-f, TFC, M&Q scale, AIMS, MMSE | improvements in motor functioning; trend for improvements in cognition and function |
| De Yebenes et al., 2011 [39] | 50.6 (10.5) | 4.8 (3.5) | Pridopidine | randomized, placebo controlled | UHDRS-mb(modified) | no improvements in motor functioning |
| Deroover et al., 1984 [40] | ---- | -- | Tiapride | randomized, placebo controlled crossover | Study specific (choreatic movements, motor skills) | improvements in choreatic movements and motor skills |
| Destee et al., 1984 [41] | 52 | 5.8 | Piracetam | placebo controlled | Study specific (involuntary movements) | increased choreic movements |
| Dorsey, 2008 [42] | 52.3 (9.8) | 6.8 | ethyl-EPA (omega3 fatty acid) | randomized, placebo controlled | UHDRS-m, UHDRS-c, MMSE, TFC | no improvements |
| Dorsey et al., 2011 [43] | ---- | ----- | Tetrabenazine | Randomized substudy, active drug group only | depressed mood item from UHDRS | no improvement in depressed mood  |
| “randomized, double-blind, placebo-controlled…” JAMA Neurol, 2013 [44] | 53.3 (9.7) | 4.4 (3.6) | Latrepirdine | randomized, placebo controlled | UHDRS-m, MMSE, NPI, CIBIC-plus, AD-ADL | no improvements |
| Dubinsky & Gray, 2006 [45] | 49.2 | 6.6 | Cysteamine (cystagon) | open label | UHDRS-m, UHDRS-b, UHDRS-I, TFC, max tolerated dose | no improvements; tolerated at 20 mg/kg per day with several limiting side effects |
| Dupont et al., 1978 [46] | 66 | 12 | Somatostatin | crossover, saline controlled case study | Study specific (neurological scores, involuntary movements, motor function) | no improvements |
| Esmaeilzadeh et al., 2011 [47] | range (39-75) | ----- | Pridopidine | ----- | PET, MRI | increased metabolic activity in several brain regions after treatment |
| Fahn, 1972 [48] | 51.3 | ---- | Perphenazine | placebo controlled crossover | study specific (chorea severity) | improvements in chorea for 70% of patients |
| Fekete et al., 2012 [49] | 58 (11) | 8.4 (3.5) | Tetrabenazine | ----- | UHDRS-m, study specific (computerized dynamic posturography system) |  moderately improvement in balance  |
| Fernandez et al., 2000 [50] | 58 | 7.5 | Donepezil HCL | open label | UHDRS, MMSE, supplemental neuropysch battery | improvements in motor function; no improvement in other domains |
| Foster et al., 1983 [51] | range (25-59) | ---- | THIP | placebo controlled | AIMS, BPRS, Columbia Rating scaleb | no improvement in motor or behavioral functioning |
| Frank, 2009 [52] | 50.9 (11.5) | 8.5 (4.5) | Tetrabenazine | placebo controlled | UHDRS | improvements in chorea for up to 80 weeks |
| Frank et al., 2008 [53] | 56.1 (9.7)55.9 (8.5)59.8 (14.2) | 10.1 (4.5)8.9 (5.6)11.4 (4.8) | Tetrabenazine withdrawal | randomized, placebo controlled | UHDRS, CGI | trend for reemergence of chorea in patients who were withdrawn from study drug |
| Frattola et al., 1977 [54] | 49 (3.1) | 7 | Bromocriptine | crossover, placebo controlled | study specific (chorea severity, finger dexterity, gait, speech) | improvements in involuntary movements |
| Frattola et al., 1983 [55] | 48.4 (3.3) | range (3-15) | Lisuride | placebo controlled | study specific (abnormal involuntary movements, chorea) | temporary improvement of abnormal involuntary movements |
| Gessa et al., 1991 [56] | 62 | ----- | Sch 23390  | placebo controlled case study | BPRS, study specific (involuntary movements) | 1mg dose improved involuntary movements |
| Gilligan et al., 1972 [57] | 38 (15.2) | ----- | Tetrabenazine | placebo controlled | study specific (motor tasks) | slight to moderate improvement in involuntary movements |
| Gimenez-Roldan & Mateo, 1989 [58] | 43.5 | 5.5 (3.6) | Tetrabenazine; Haloperidol | crossover | Kartzinel scale for involuntary movements | TBZ and haloperidol both improved chorea significant |
| Giuffra et al., 1992 [59] | Range (26-52) | 6 | Milacemide | randomized, placebo controlled | AIMSb, Buschke Selective Reminding Test, Verbal Fluency, ADAS-c | 1200 mg/day did not improve chorea or cognition |
| Goety et al., 1990 [60] | 51.6 (16.7) | 6.3 (3.6) | L-Acetyl-Carnitine | crossover, placebo controlled | TFC, AIMS, HAM, MMSE, Verbal fluency | drug had neither efficacy nor toxicity |
| Growdon, 1978 [61] | ---- | -- | Choline | healthy controls | biomarkers, study specific (speech, balance, gait) | transient improvement in balance and gait |
| Haslam, 1967 [62] | 2735 (n=2) | 79 | Penicillamine | age/gender matched controls | biomarkers, study specific (dexterity, conversation, gait)  | mild improvement in one patient and no improvement in the other |
| Hersch et al., 2006 [63] | 44.7 | 7.9 (3.5) | Creatine  | randomized, placebo controlled | UHDRS, tolerability | study drug well tolerated; no improvements on clinical measures  |
| Holl et al., 2010 [64] | 48.6 (10.5) | 5.9 (4.3) | Venlafaxine XR | ---- | HAM, BDI | significant improvement of depression |
| Hyson et al., 2010 [65] | 50.4 (8.4) | ---- | high dosage Coenzyme Q10 | healthy controls | Tolerability, AE frequency, blood levels | well tolerated; 82% achieved target dosage |
| Jankovic, 1982 [66] | 34 | ---- | Tetrabenazine | crossover, placebo controlled case study | study specific (hyperkinesia) | improvement of chorea |
| Jankovic & Beach, 1997 [67] | 54.8 | 7.27 (3.9) | Tetrabenazine | ---- | study specific (abnormal movements) | improvement in abnormal movements in 83% of patients |
| Kartzinel et al., 1976 [68] | range (41-54) | ---- | Bromocriptine | crossover, placebo controlled | study specific (motor disability) | high doses increased involuntary movements |
| Kenney et al., 2007 [69] | 56.3 (11.6) | 10.4 (5.8) | Tetrabenazine | ---- | UHDRS-m, BDI | single dose improves chorea for an average of 5 hours |
| Kieburtz, 2001 [70] | 47.5 (10.1)47.4 (11)48.2 (10.7) | 3.3 (2.3)2.6 (2.2)2.3 (2.0) | CoQ10, Remacemide | randomized, placebo controlled | UHDRS | no improvements with either drug |
| Kieburtz et al., 1996 [71] | 44.3 (14.8)48.8 (11.6) | 9.58.1 | Remacemide | randomized, placebo controlled | TFC, HDMRS, MMSE, Trailmaking Test, Symbol Digit Modalities Test, Stroop, HVLT, BDI, tolerability | no differences between treatment arms; drug generally well tolerated |
| Kieburtz et al., 2010 [72] | 53.7 (10.9) | 4.4 | Latrepirdine | randomized, placebo controlled | UHDRS, MMSE, ADAS-cog, tolerability | improved cognition (MMSE), but no improvements on other measures |
| “randomized, double-blind, placebo-controlled…” Mov Disord, 2013 [73] | 54.3 (11)50.5 (10.5)50.9 (9.7) | 4.6 (3.5)4.4 (3.6)4.1 (2.8) | Pridopidine | randomized, placebo controlled | UHDRS, CGI, HADS, Trailmaking A, Tolerability, lab tests | drug generally well tolerated; no improvements |
| Kotzailias et al., 2003 [74] | 60 (4) | ---- | Tiapride | age/gender matched controls | platelet counts | increased platelet counts |
| Kremer et al., 1999 [75] | 46.1 | 2.9 | Lamotrigine | placebo controlled | TFC | no improvements |
| Laks et al., 2004 [76] | 49(n=1) | ---- | Olanzapine | Case reports | UHDRS | long term tolerability; improvement in motor and behavioral functioning |
| Lal et al., 1973 [77] | 5446 (n=2) | 126 | Apomorphine, Pimozide, L-dopa | placebo crossover | Probenecid test, CSF levels | HVA and 5HIAA levels increased (i.e., unimpaired turnover of DA and 5HT) |
| Landwehrmeyer et al., 2007 [78] | 44.9a (9.6) | 5.0 (3.6) | Riluzole | randomized, placebo controlled | UHDRS, BDI, CGI | no improvements |
| Leonard et al., 1975 [79] | ---- | ---- | Lithium Carbonate, Haloperidol | randomized, crossover, placebo controlled | study specific (ultrasonic measurement, psychological variables) | no improvements |
| Lieberman et al., 1975 [80] | ---- | ---- | Piribedil | case study (n=2) | study specific (abnormal involuntary movements) | increase in abnormal involuntary movements |
| Lucetti et al., 2002 [81] | 60.4 (8.4) | 5.0 (1.9) | oral Amantadine | Open label | UHDRS-m, AIMS | improvement in dyskinesias |
| Lucetti et al., 2003 [82] | 60.7 (7.9) | 5.1 (1.8) | IV and oral amantadine | randomized, crossover, placebo controlled | AIMS, UHDRS-m, HAM, BDI, BPRS | improvements in choreic dyskinesias; no adverse effect on cognition |
| Lundin et al., 2010 [83] | 50.2 (7.0)a | 6.8 (5.0)a | Pridopidine (ACR16) | randomized, placebo controlled | UHDRS-m, HADS, CGI, Symbol Digit Modalities Test, Verbal Fluency, Stroop, Trailmaking Test A | no improvements in cognition |
| Manyam et al., 1990 [84] | 46 (15)a | Range (3-10)--- | Isoniazid (plus pyridoxine) | randomized, crossover, placebo controlled | CSF levels of Choline and AChE | reduced level of choline in CSF on patients |
| Manyam et al., 1981 [85] | 41 | Range (3-20) | Isoniazid (plus pyridoxine) | randomized, crossover, placebo controlled | biomarkers, study specific (abnormal involuntary movements) | no significant improvements; serious side effects |
| Manyam et al., 1987 [86] | 46 (15) | ---- | Isoniazid (plus pyridoxine) | crossover, placebo controlled | amino acid levels in CSF and plasma | significant elevation in average CSF levels of several amino acids |
| Manyam et al., 1980 [87] | 35 (10) | ---- | Isoniazid (plus pyridoxine) | placebo controlled | GABA levels in CSF and plasma | elevation of brain GABA |
| Marshall, 2006 [88] | ---- | ---- | Tertrabenazine | randomized, placebo controlled | CGI, UHDRS | improvements in chorea and disease severity |
| Marshall, 1998 [89] | 47.7 (14.8)47.1 (13.7)45.3 (13.1) | 7.8 (5.6)6.6 (3.6)6.6 (4.6) | OPC-14117 (lipophilic free-radical scavenger) | randomized, placebo controlled | CGI, UHDRS, tolerability, free radical levels in CSF and plasma | no significant differences between treatment arms; drug well tolerated |
| Marshall, 2003 [90] | 46.6 (9.1)47.9 (8.7) | 5.0 (3.7)6.3 (3.8) | Riluzole | randomized, placebo controlled | UHDRS | improvements in chorea, but not in other domains |
| Mateo & Gimenez-Roldan, 1996 [91] | ---- | ---- | Piracetam | placebo controlled | study specific (involuntary movements) | increased chorea |
| McLean, 1982 [92] | range (35-59) | ---- | Isoniazid | randomized, placebo controlled, crossover | study specific (movement frequency) | no improvements in chorea |
| McLellan et al., 1974 [93] | 51 | ---- | Tetrabenazine, thiopropazate | placebo controlled | study specific (chorea) | both study drugs improved chorea |
| Metman et al., 2002 [94] | Median 52 | Median 6 | Amantadine | randomized, placebo controlled crossover | UHDRS-m, UHDRS-c, RBANS, blood samples | chorea improved in all but one patient |
| Muller-Vahl et al., 1999 [95] | 58 | ---- | Nabilone | case study | Folstein chorea and motor impairment scale, WAIS Digit Span, CVLT, Benton Visual Retention Test, Raven’s Progressive Matrices Test | increased chorea |
| Murman et al., 1997 [96] | 48.4 (12.8) | 4.7 (3.9) | NMDA-receptor antagonist ketamine | randomized crossover (placebo administered as part of crossover) | UHDRS-m, BPRS, SRS, Buschke Selective Reminding Test, Washington Square Picture Memory Test, Verbal Fluency, WAIS Digit Span | declines in cognitive functioning |
| Newman et al., 1985 [97] | Range (30-50) | ---- | EMD 23,448 | crossover, placebo controlled | AIMS, hormone levels in blood | increased involuntary movements; plasma prolactin levels fell  |
| Nutt et al., 1978 [98] | 47 | ---- | Arecoline  | ---- | physiology, study specific (involuntary movements) | increased chorea; significant alterations in vital signs |
| Nutt et al., 1978 [99] | 47.3 | ---- | Naltrexone | crossover, placebo controlled | study specific (involuntary movements) | no improvements |
| Ondo et al., 2007 [100] | 53.5 (20.8) | ---- | Memantine | open label | UHDRS | improved motor symptoms, but not other domains |
| Ondo et al., 2002 [101] | 56.3 (12.4) | 8.1 (5.3) | Tetrabenazine | ---- | AIMS | improved motor symptoms |
| O'Suilleabhain & Dewey, 2003 [102] | 51 (13.0) | ---- | Amantadine | randomized, placebo controlled crossover | study specific (chorea, self-report improvement survey) | no objective improvements, although patients “felt better” |
| Paleacu et al., 2002 [103] | 47.6 (11.4) | 11.2 (3.3) | Olanzapine | ---- | UHDRS-b, UHDRS-m, CGI | improvements in behavioral symptoms; no improvements in motor symptoms |
| Perry et al., 1980 [104] | 37.6 | 5.7 | Aminooxyacetic acid (AOAA) | placebo controlled, crossover | study specific (chorea, motor functioning) | Inconclusive |
| Perry et al., 1979 [105] | 43.8 | 6.3 | Isoniazid (INH) | open label | biomarkers, study specific (mental function, movement) | 3 of 6 patients had some improvement. |
| Perry et al., 1982 [106] | 42.4 | 4.6 | Isoniazid (INH) | crossover, placebo controlled | biomarkers, study specific (motor function, neurologic assessment, psychometric testing) | minority of patients had improvements following high dose therapy |
| Peyser et al., 1995 [107] | ---- | Mild-moderate severity | a-tocopherol | randomized, placebo controlled | QNE, MMSE, informant rated HD-ADLS, WAIS Digit Span, Verbal Fluency, Trailmaking Test, CERAD Verbal Learning, Wisconsin Card Sorting Test, Benton Visual Retention Test, Go/No Go Test, Stroop, Design Fluency Test | drug may slow rate of motor decline early in the course of HD |
| Piolti et al., 1995 [108] | 50.5 (15.5) | 6.4 (3.0) | Proglumide | placebo controlled | BPRS, Raven’s Progressive Matrices Test, Kohs Cubes, study specific (abnormal movements, motor function) | no improvements |
| Puri et al., 2005 [109] | 50 (9.3) | -------- | Ethyl-EPA | randomized, placebo controlled | UHDRS | no group differences on motor functioning |
| Puri et al., 2002 [110] | 53.1 (11.1) | -------- | Ethyl-EPA | randomized, placebo controlled, pilot | UHDRS-m, MRI | improvements in cerebral structure and function (relative to placebo) |
| Puri et al., 2008 [111] | 51.3 (2.5) | -------- | Ethyl-EPA | randomized, placebo controlled | MRI  | significant group-level reductions in brain atrophy  |
| Quinn & Marsden, 1984 [112] | 53 | 6.7 | Sulpiride | randomized, crossover | study specific (movement, chorea, functional scale) | improvements in dyskinesia and movements; no functional improvements |
| Ranen et al., 1996 [113] | 43.9 (13.7) | 5.9 | Idebenone  | randomized, placebo controlled | HD-ADL, QNE, MMSE, Buschke Selective Reminding Test, Benton Visual Retention Test, WAIS Arithmetic, Trailmaking Test, Stroop | no group differences |
| Rosas et al., 1999 [114] | 45 (10.2) | 6.1 (4.1) | Riluzole | Open label | UHDRS-chorea, dystonica, TFC, lactate levels | improvements in chorea; no effect on dystonia or functional capacity |
| Rubin et al., 1993 [115] | 42.2 (10.1) | 8.1 (3.4) | Baclofen  | placebo controlled | study specific (saccade latency and velocity) | declines in saccade latency and mean velocity in controls |
| Saft et al., 2006 [116] | 10.25 | ---- | Valproic acid | ----  | UHDRS-m | improvements in motor score for 7 of 8participants |
| Satoh et al., 2009 [117] | 54.8 | 19.3 | Yi-Gan San, chaihu-Jia-Longgu-Muli Tan | crossover | UHDRS-m, MMSE, Barthel Index, Lab tests | both drugs improved chorea |
| Scigliano et al., 1984 [118] | 48.8 | 6.8 | y-vinyl GABA | placebo controlled, crossover | physiology, study specific (chorea, motor function) | no improvements |
| Scott, 2011 [119] | 49.2 | 8.3 | Tetrabenazine | placebo controlled | CGI, UHDRS | improvements in chorea |
| Seppi et al., 2001 [120] | 46.4 (9.3) | 8 (3.3) | Riluzole | open label | UHDRS | transient improvement in chorea; sustained improvements in psychomotor speed and behavior |
| Shoulson & Chase, 1975 [121] | 56 | 6.0 | Caffeine | crossover, placebo controlled | study specific (tremor, rigidity, akinesia) | no improvements |
| Shoulson et al., 1989 [122] | 38.0 (10.2) | 5.2 (3.7) | Baclofen | randomized, placebo controlled | TFC | no improvements |
| Shoulson, et al., 1977 [123] | ---- | --- | Muscimol | crossover, placebo controlled | lab tests, study specific (motor and cognitive performance, EEG) | no improvement in hyerkinesia; improved chorea in the most severely hyperkinetic patient |
| Shoulson et al., 1978 [124] | 43.9 | 7.1 | Muscimol | crossover, placebo controlled | Trailmaking Test, Wechsler Memory Scale, study specific (motor, chorea, dystonia, ADLs) | no improvements in motor or cognition |
| Shoulson et al., 1976 [125] | 47 | 5.0 | DPA & GABA | crossover | study specific (chorea, finger dexterity, gait, speech) | at maximum dose, combined drugs increased turnover of dopamine and serotonin |
| Shults et al., 1986 [126] | range (28-52) | ---- | Cysteamine | placebo controlled, crossover | Road Map test, Recurring figures test, Visual form discrimination, Dichotomous listening test, study specific (chorea, rigidity, akinesia) | no improvements on motor or cognitive functioning |
| Squitieri et al., 2009 [127] | 46.7 (12.0) | -------- | Riluzole | placebo controlled | UHDRS-m, UHDRS-b, TFC, BDNF serum levels | placebo showed significantly greater volume loss of grey matter; no differences between groups on UHDRS scores |
| Squitieri et al., 2013 [128] | 50.6 (10.60 | ----- | Pridopidine | Placebo controlled | UHDRS-m, UHDRS-c, tolerability, lab tests | acceptable safety profile and generally well tolerated |
| Stocchi et al., 1989 [129] | 52.2 | 3.8 | Transdihydrolisuride (TDHL) | crossover, placebo controlled | M&Q scale, AIMS | no improvements on chorea |
| Symington et al., 1978 [130] | 45.7 | ---- | Sodium Valproate | open label | biomarkers, study specific (body movements) | no improvements on movements |
| Tabrizi et al., 2003 [131] | 42.2 (5.6) | ---- | Creatine | age matched controls | UHDRS, MRS | no change in motor functioning, functional capacity or cognition at 12-months; Creatine was significantly elevated in brain and muscle tissue |
| Tabrizi et al., 2005 [132] | 44.2 (5.7) | ---- | high dose Creatine | placebo controlled | UHDRS, MRS | no change in motor functioning, functional capacity or cognition at 24-months; Creatine was significantly elevated in brain and muscle tissue |
| Tan et al., 1976 [133] | 50 (n=1) | ---- | Sodium Valproate | placebo controlled | biomarkers, study specific (chorea) | tolerance to study drug |
| Tarsy & Bralower, 1977 [134] | 55.7 | 9.7 | Deanol Acetamidobenzoate | placebo controlled, crossover | study specific (involuntary movements, motor function) | no improvements in chorea |
| Tell et al., 1981 [135] | 48 | 6.5 | y -acetylenic GABA | placebo controlled | biomarkers, study specific (abnormal movements, motor function) | GABA concentration increased; no motor improvement |
| Terrence, 1976 [136] | 52.2 | 2.7 | Fluphenazine decanoate | placebo controlled | study specific (chorea) | improvements in chorea |
| Thomas et al., 2004 [137] | range (21-66) | 9 | Minocycline | pilot | UHDRS, AIMS, MMSE, lab tests | drug tolerated; no improvements on outcome measures |
| Tolosa, 1976 [138] | 41.5 | ---- | Levodopa, Haloperidol | open label | Study specific (chorea) | Levodopa did not improve chorea; Haloperidol improved chorea |
| Tourian, 1972 [139] | 50.8 | 7 | 5-hydroxy-L- tryptophan | crossover | WAIS Digit Span, WAIS Block Design, WAIS Arithmetic, WAIS Digit Symbol, WAIS FSIQ, study specific (physical and neurologic exam) | no improvements in cognitive, behavioral or motor functioning |
| Vaddadi et al., 2002 [140] | 49.8 | ---- | HUFA (highly unsaturated fatty acids) | randomized, placebo controlled | UHDRS, Rockland-Simpson Dyskinesia Scale (motor) | improvement in motor function; no significant cognitive improvements |
| Van Vugt et al., 1997 [141] | 43.6 (11.4)50.5 (7.9) | 9.3 (7.5)7.3 (3.8) | Clozapine | randomized, placebo controlled | UHDRS-chorea, AIMS | improvement in motor function (as per AIMS, not UHDRS) |
| Verbessem et al., 2003 [142] | 49.6 (1.9) | 8.6 (1.5) | Creatine | randomized, placebo controlled pilot | UHDRS, study specific (strength and coordination tasks) | no improvements  |
| Verhagen Metman et al., 2002 [143] | median 52range (32-68) | median 6range (2-15) | Amantadine | crossover, placebo controlled | UHDRS-chorea, MMSE, RBANS, Verbal Fluency, Symbol Digit Modalities Test, Stroop | improvements in chorea |
| Vestergaard et al., 1977 [144] | 49 | 6.7 | Lithium | crossover, placebo controlled | study specific (motor, hyperkinesia, global health) | no improvements |
| Vitale et al., 2007 [145] | 45.9 | 6.3 | Apomorphine HCL | randomized, crossover trial | UHDRS-m, AIMS, HAM | improvements in chorea for some patients with continuous injection |
| Walker & Hunt, 1989 [146] | 44.4 | ---- | Dextromethorphan | Open label | TFC, QNE, Benton Visual Retention Test, Verbal Fluency | declines in functioning  |
| Wesseling & Lakke, 1980 [147] | range (38-72) | range (8-20) | 4-aminopyridine | placebo controlled | study specific (involuntary movements) | no group differences |
| Zesiewicz et al., 2006 [148] | 51.9 (9.3) | ---- | Levetiracetam (LEV) | open label | UHDRS-m, CGI, ESS | slight improvements in motor functioning; significant improvements in chorea |
| "randomized, placebo controlled trial…" Neurology, 2001 [149] | 47.5c(10.1)47.4 (11.0)48.2 (10.7) | 5.3 (2.7)c4.9 (2.6)4.6 (3.2) | Coenzyme Q10, Remacemide HCL | randomized | TFC | declines in functional capacity |
| "TBZ as anti chorea therapy…." Neurology, 2006 [150] | ---- | ---- | Tetrabenazine | randomized, placebo | UHDRS, CGI, ESS, HAM | improvements in chorea (relative to controls) |

a Reported as mean (standard deviation) unless otherwise specified.

b Scale was modified

c Numbers reported in text and table were discrepant for this publication, we report the number from the table in this appendix

ADAS-cog, Alzheimer Disease Assessment Scale (cognitive subsection); AIMS; Abnormal Involuntary Movement Scale; BDI; Beck Depression Inventory; BPRS, Brief Psychiatric Rating Scale; CAARS, Connors’ Adult ADHD Rating Scale; CBRS, Cognitive Behavior Rating Scale; CGI, Clinical Global Impression scale; ESS, Epworth Sleepiness Scale; HADS, Hospital Anxiety and Depression Scale; HAM, Hamilton Depression scale; HD-ADL, Huntington disease activities of daily living scale; HDMRS, HD motor rating scale; MMSE, Mini-Mental State Exam; M&Q scale, Marsden and Quinn chorea severity scale; NPI, Neuropsychiatric Inventory; QNE, quantitative neurologic examination; RSDRS, Rockland-Simpson Dyskinesia Rating Scale; SIP, sickness impact profile; SRS, National Institute of mental health's self-rating score; UHDRS, Unified Huntington’s Disease Rating Scale (-m: motor; -c: cognitive; -f: functional; -b: behavioral; -I: independence scale; -TFC: total functional capacity)

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