Supplementary table 3.

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| No | Sex | TNR | Diagnosis/  Presenting symptom | Cere  -bellar | Parkinson  -ism | Psychiatric | Cognitive impairment | Chorea | Age  of onset |
| Bauer et al, 2004[1](#_ENREF_1) | M | 52/40 | Hyperkinesia | (-) | (-) | (-) | (-) | (-) | 50 |
|  | F | 51/35 | Ataxia | (+) | (-) | (+) | (+) | (-) | 32 |
|  | M | 50/36 | Ataxia | (+) | (-) | (-) | (+) | (-) | 32 |
|  | M | 48/37 | Ataxia | (+) | (-) | (-) | (+) | (-) | 35 |
|  | F | 46/36 | Ataxia | (+) | (-) | (-) | (+) | (+) | 36 |
|  | M | 46/35 | Ataxia | (+) | (-) | (+) | (-) | (+) | 37 |
|  | M | 46/35 | HDL | (+) | (-) | (-) | (+) | (+) | 38 |
| Brockmann et al, 2012[2](#_ENREF_2) | F | 54 | Ataxia | (+) | (-) | (-) | (-) | (-) | 36 |
|  | M | 51 | Ataxia | (+) | (-) | (-) | (-) | (-) | 40 |
|  | M | 45 | Ataxia | (+) | (-) | (-) | (-) | (-) | 46 |
|  | M | 54 | Ataxia | (+) | (-) | (-) | (-) | (-) | 33 |
|  | M | 51 | Ataxia | (+) | (-) | (-) | (-) | (-) | 35 |
| Silveira et al, 2002[3](#_ENREF_3) | M | 43 | Ataxia | (+) | (-) | (-) | (+) | (-) | 52 |
| Fujigasaki et al, 2001[4](#_ENREF_4) | M | 46 | Ataxia | (+) | (-) | (-) | (+) | (-) | 50 |
|  | M | 46 | Ataxia | (+) | (-) | (-) | (+) | (+) | 37 |
|  | F | 46/37 | Ataxia | (+) | (-) | (-) | (-) | (-) | 55 |
|  | F | 46/39 | Ataxia | (+) | (-) | (-) | (-) | (-) | 34 |
| Gunther et al, 2004[5](#_ENREF_5) | M | 50/37 | Ataxia | (+) | (-) | (-) | (-) | (-) | 31 |
| Koide et al,1999[6](#_ENREF_6) | F | 63/36 | Ataxia | (+) | (-) | (-) | (+) | (-) | 6 |
| Lin et al, 2007 | M | 45/39 | Ataxia | (+) | (+) | (-) | (-) | (-) | 43 |
| Nakamura et al, 2001[7](#_ENREF_7) | M | 55/37 | Ataxia | (+) | (+) | (-) | (+) | (-) | 19 |
|  | F | 55/39 | Ataxia | (+) | (+) | (-) | (+) | (-) | 25 |
|  | M | 48/36 | Ataxia | (+) | (+) | (-) | (+) | (-) | 39 |
|  | M | 48/36 | Ataxia | (+) | (+) | (-) | (+) | (+) | 48 |
|  | F | 47/36 | Ataxia | (+) | (+) | (-) | (+) | (+) | 40 |
|  | F | 47/32 | Ataxia | (+) | (+) | (-) | (+) | (-) | 28 |
| Oda et al, 2004[8](#_ENREF_8) | F | 56/36 | Ataxia | (+) | (-) | (-) | (+) | (-) | 30 |
|  | F | 48/36 | Ataxia | (+) | (-) | (-) | (+) | (-) | 27 |
|  | M | 47/44 | Ataxia | (+) | (-) | (-) | (+) | (-) | 25 |
|  | M | 47/36 | Ataxia | (+) | (-) | (-) | (+) | (-) | 42 |
|  | F | 45/37 | Ataxia | (+) | (-) | (-) | (-) | (-) | 52 |
|  | F | 45/36 | Ataxia | (+) | (+) | (-) | (+) | (+) | 60 |
|  | M | 44/36 | Ataxia | (+) | (+) | (-) | (+) | (+) | 29 |
|  | F | 43/33 | Ataxia | (+) | (-) | (-) | (-) | (-) | 47 |
| Rolfs et al, 2003[9](#_ENREF_9) | F | 49/37 | Parkinsonism | (-) | (+) | (+) | (-) | (-) | 38 |
|  | F | 49/36 | Ataxia | (+) | (-) | (+) | (+) | (-) | 16 |
|  | F | 49 | Ataxia | (+) | (-) | (+) | (+) | (-) | 16 |
|  | M | 54/37 | Ataxia | (+) | (-) | (+) | (+) | (+) | 36 |
|  | M | 54/37 | Ataxia | (+) | (-) | (-) | (+) | (-) | 18 |
|  | F | 54/37 | Ataxia | (+) | (-) | (+) | (-) | (+) | 23 |
|  | M | 54/34 | Ataxia | (+) | (-) | (-) | (-) | (-) | 18 |
|  | F | 45/39 | Ataxia | (+) | (+) | (+) | (-) | (-) | 30 |
|  | M | 45/37 | Ataxia | (+) | (-) | (-) | (-) | (-) | 43 |
|  | F | 45/37 | Ataxia | (+) | (-) | (-) | (-) | (+) | 30 |
|  | M | 45/36 | Ataxia | (+) | (-) | (-) | (-) | (-) | 23 |
|  | F | 54 | Ataxia | (+) | (-) | (-) | (+) | (-) | 33 |
|  | F | 54 | Ataxia | (+) | (-) | (-) | (+) | (-) | 33 |
|  | F | 54 | Ataxia | (+) | (-) | (+) | (-) | (+) | 8 |
|  | M | 54 | Ataxia | (+) | (-) | (-) | (-) | (-) | 18 |
| Salvatore et al, 2006[10](#_ENREF_10) | M | 53 | Ataxia | (+) | (+) | (+) | (+) | (+) | 23 |
|  | F | 51 | Ataxia | (+) | (+) | (+) | (+) | (+) | 40 |
|  | M | 52 | Ataxia | (+) | (+) | (+) | (+) | (+) | 40 |
| Toyoshima et al, 2004[11](#_ENREF_11) | M | 48/48 | Ataxia | (+) | (-) | (-) | (+) | (+) | 39 |
| Wu et al, 2004[12](#_ENREF_12) | F | 46 | PD | (-) | (+) | (-) | (-) | (+) | 75 |
| Zuhlke et al,2005[13](#_ENREF_13) | M | 53 | Ataxia | (+) | (-) | (-) | (-) | (+) | 41 |
|  | F | 52 | Ataxia | (+) | (-) | (-) | (-) | (+) | 50 |
| Zuhlke et al,2003[14](#_ENREF_14) | F | 48/38 | Ataxia | (+) | (-) | (-) | (+) | (+) | 38 |
|  | M | 48/38 | Ataxia | (+) | (-) | (-) | (+) | (-) | 43 |
|  | F | 48/37 | Ataxia | (+) | (-) | (-) | (+) | (-) | 36 |
|  | M | 48/37 | Ataxia | (+) | (-) | (-) | (+) | (+) | 33 |
| Stevanin et al, 2003[15](#_ENREF_15) | F | 46 | HDL | (+) | (+) | (-) | (+) | (+) | 23 |
|  | F | 44 | HDL | (+) | (-) | (-) | (+) | (+) | 29 |
| Maltecca et al, 2003[16](#_ENREF_16) | F | 53 | Ataxia | (+) | (-) | (+) | (+) | (-) | 35 |
|  | M | 53 | Ataxia | (+) | (-) | (+) | (+) | (-) | 34 |
|  | M | 53 | Ataxia | (+) | (-) | (+) | (+) | (-) | 23 |
|  | F | 66 | Ataxia | (+) | (-) | (+) | (+) | (-) | 3 |
| Alibardi et al, 2014[17](#_ENREF_17) | F | 41 | Chorea/  Psychiatric symptom. | (-) | (-) | (+) | (-) | (+) | 23 |
| Nanda et al, 2007[18](#_ENREF_18) | M | 41 | Ataxia | (+) | (-) | (+) | (+) | (-) | 50 |
| Nielsen et al, 2012[19](#_ENREF_19) | M | 41 | Dementia | (+) | (-) | (-) | (-) | (-) | 42 |
| Nolte et al, 2010[20](#_ENREF_20) | F | 42 | Ataxia | (+) | (-) | (-) | (-) | (-) | 41 |
|  | F | 42 | Ataxia | (+) | (-) | (-) | (+) | (-) | 32 |
|  | M | 42 | Ataxia | (+) | (-) | (-) | (+) | (-) | 45 |
|  | M | 42 | Ataxia | (+) | (-) | (-) | (-) | (-) | 44 |

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