**Supplementary Table 1.** Short-term and long-term behavioural outcomes of deep brain stimulation in Huntington’s disease

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  |  | Behavioural symptoms | | | | | | | |
| Authors, publication year | n | overall | apathy | irritability & aggression | depression | anxiety | perseveration | psychotic symptoms | pseudobulbar affect |
| Gonzalez et al., 2014 | 7 | progression of unspecified behaviour disorder in one case | NR | NR | NR | NR | NR | NR | NR |
| Wojtecki et al., 2015 | 6 | no statistically important change in BPRS at 6 months | NR | NR | significantly better mood at 6-months as assessed with HADS-D but not with BDI or MADRS | NR | NR | NR | NR |
| Zittel et al., 2018 | 6 | no psychiatric side-effects | NR | NR | NR | NR | NR | NR | NR |
| Sanrey et al., 2021 | 13 | constant psychiatric comorbidities but neuroleptics were added in two patients during follow-up due to behaviour disorder | NR | NR | NR | NR | NR | NR | NR |

\* BDI, the Beck Depression Inventory; BPRS, the Brief Psychiatric Rating Scale; HADS, the Hospital Anxiety and Depression Scale; MADRS, the Montgomery-Åsberg Depression Rating Scale;

**Supplementary Table 2.** Short-term and long-term cognitive and language outcomes of deep brain stimulation in Huntington’s disease

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
|  |  | Cognitive function and speech | | | | | | |
| Authors, publication year | n | screening | speech & language function | visuospatial functions | episodic memory and learning | information processing & working memory | executive functions | social cognition |
| Gonzalez et al., 2014 | 7 | no deterioration in MDRS at 12-month follow-up | NR | NR | NR | NR | NR | NR |
| Wojtecki et al., 2015 | 6 | no change in MDRS total score at 6-month follow -up (atypical score reporting: percent of performable points) | NR | no change in MDRS construction | NR | deterioration in MDRS concentration (possibly initiation/perseveration scale) at 6-months | NR | NR |
| Zittel et al., 2018 | 6 | no definite deterioration of cognitive function | NR | NR | NR | NR | NR | NR |
| Sanrey et al., 2021 | 131 | deterioration of MDRS score at 3rd and 4th follow-up  two youngest cases (< 60 years old at surgery) demonstrated a dramatic drop of their cognitive abilities 1 year before they died (severe dementia — MDRS < 10) | NR2 | NR | deterioration in MDRS memory & orientation at 4th follow-up; only 1 patient assessed at 4th follow up with verbal learning (RL/  RI16 verbal memory test) | deterioration on Digit Span forward and MDRS attention at 4th follow-up | no change in fluency2 and TMT at4th follow-up | NR |

MDRS, Mattis Dementia Rating Scale; TMT, Trail Making Test

1 full cognitive data available for 1-10 patients depending on test and time-point

2 the authors reported fluency as language, here we grouped it under executive function

**Supplementary Table 3.** Adverse events of deep brain stimulation in Huntington’s disease

|  |  |
| --- | --- |
| Authors, publication year | Adverse events |
|
| Gonzalez et al., 2014 | hardware dysfunction leading to extension replacement (n = 1);  repositioning of one of the leads after 1 year due to suboptimal control of chorea (n = 1);  suicide attempt (n = 1) |
| Wojtecki et al., 2015 | gait impairment and hyperkinesia after reprogramming (n = 1) – serious adverse event leading to hospital admission and requiring reprogramming;  postoperative malignant hyperthermia (n = 1) – serious adverse event life-threatening and leading to prolonged hospital stay;  hyperthermia (n = 1);  bradykinesia (n = 1);  gait impairment and fall (n = 1),  increased chorea after reprograming due to bradykinesia (n = 1);  deactivation of impulse generator (n = 1);  hospitalization-related (n=3): thrombophlebitis, MRSA nose infection, superficial nose abrasion |
| Zittel et al., 2018 | frequency-dependent increase of chorea (n = 2);  stupor (n = 1);  dysphagia (n = 1);  small haemorrhage (n = 1);  limbs spasticity (n = 3);  postural instability (n = 2);  gait impairment (n = 3) |
| Sanrey et al., 2021 | not reported |