

Patient	Age at DBS	Male (M) or Female (F)	Dystonia Group	Age at onset (years)	Distribution of dystonia	Phenotype: Fixed or Phasic	Inherited, Acquired, Idiopathic	Aetiology-	Imaging	Medication at time of surgery	Duration of Dystonia (years)	Proportion of Life Lived with dystonia	Baseline BFMDRS motor score	1 year BFMDRS motor score	% Improvement at 1 year
1	13.9	M	Primary	10.9	Generalised. Neck, trunk and all 4 limbs	Phasic	Inherited	DYT1 positive	Normal	None (previous either ineffective or side effects)	3	0.22	57	23	59.6
2	11.8	M	Primary	8	Generalised, particularly both legs and left arm	Fixed	Inherited	DYT1 positive	Normal	Co-Careldopa	3.8	0.32	59.5	10	83.2
3	7.3	F	Primary	6.3	Generalised, particularly both legs and right arm	Fixed	Inherited	DYT1 positive	Normal	Trihexyphenidyl	1	0.14	50	6	88.0
4	10.8	M	Primary	3.5	Generalised	Fixed	Idiopathic	Idiopathic (DYT1, DYT5 and PANK2 negative)	Normal	None	7.3	0.68	75	26.5	64.7
5	13.9	M	Primary	11.5	Generalised	Fixed	Inherited (positive family history)	Idiopathic	Normal	Diazepam	2.4	0.17	48	15.5	67.7
6	12.9	M	Primary (plus)	2.9	Generalised – particularly face and upper limbs	Phasic	Inherited	DYT11	Normal	None	10	0.78	30	9.5	68.3
7	17.1	F	Primary (plus)	14.1	Predominantly right upper limb	Phasic	Inherited	Idiopathic (DYT11 and TITF1 negative)	Normal	None	3	0.18	8	1	87.5
8	12.2	F	Primary (plus)	0.2	Generalised	Phasic	Idiopathic	Idiopathic	Normal	None	12	0.98	34	35	-2.9
9	8.9	M	Primary (plus)	0.5	Generalised	Fixed	Idiopathic	Unknown	Normal	Baclofen Trihexyphenidyl	8.4	0.94	64.5	55.5	14.0
10	15.9	F	Primary (plus)	0.6	Generalised	Phasic	Idiopathic	Unknown	Normal	None	15.3	0.96	59	N/A *	N/A *
11	13.1	F	Primary (plus)	9.1	Generalised	Phasic	Idiopathic	Unknown	Normal	Intrathecal baclofen Trihexiphenidyl Diazepam	4	0.31	89.5	78	12.8
12	17.4	F	Primary (plus)	4.4	Generalised but predominantly upper limbs (R>L) and R lower limb.	Phasic	Idiopathic	Unknown	Normal basal ganglia. Non-specific left Hippocampus abnormality	None	13	0.75	38	16.5	56.6
13	4.5	M	Primary (plus)	0.8	Generalised	Phasic	Idiopathic	Unknown	Normal basal ganglia. Non-specific left frontal arachnoid cyst	Lorazepam Tizanidine Trihexyphenidyl	3.7	0.82	88.5	96.5	-9.0
14	18.1	M	Primary (plus)	9	Generalised	Fixed	Idiopathic	Unknown	Normal	Sinemet plus	9.1	0.50	24	31	-29.2
15	12.6	F	Secondary static	0.6	Generalised	Fixed	Acquired	Ex-premature 26/40 (Perinatal brain injury)	Thalamus, PVWM	Baclofen Nitrazepam	12	0.95	103	86	16.5
16	7.9	M	Secondary static	0.9	Generalised	Fixed	Acquired	Ex premature 27/40 (Perinatal brain injury)	PVWM	Baclofen Clonidine Trihexiphenidyl Dantrolene	7	0.89	106	109.5	-3.3
17	6.1	F	Secondary static	0.1	Generalised	Phasic	Acquired	Ex-premature 24/40 (Perinatal brain injury)	PVWM	Trihexiphenidyl	6	0.98	106.5	97	8.9
18	10.6	M	Secondary static	0	Generalised	Fixed	Acquired	Ex-premature 25 /40 (Perinatal brain injury)	Normal	Trihexiphenidyl	10.6	1.00	87.5	84	4.0
19	9.2	M	Secondary static	0.2	Generalised	Phasic	Acquired	Ex-premature 32/40 twin, twin-twin transfusion donor, neonatal sepsis (Perinatal brain injury)	Signal change and volume loss in Globi pallidi and PVWM	Clonazepam	9	0.98	109.5	101.5	7.3
20	5.5	F	Secondary static	0	Generalised	Phasic	Acquired	Ex premature 25/40 twin (Perinatal brain injury)	White matter volume loss + Focal right cerebellar cortical dysplasia	None	5.5	1.00	110	111	-0.9
21	12.1	M	Secondary static	0.1	Generalised	Fixed	Acquired	(Perinatal brain injury) Ex-premature 28/40 G6PD deficiency leading to hyperbilirubinaemia	Basal ganglia normal Minimal white matter volume loss,	Baclofen Trihexiphenidyl Nitrazepam, Haloperidol	12	0.99	114.5	106	7.4
22	16.8	F	Secondary static	0.8	Generalised	Phasic	Acquired	Term HIE (Perinatal brain injury)	Gliosis of thalami, lentiform nuclei and periorolandi cortex.	Trihexiphenidyl	16	0.95	107	110	-2.8

23	13.7	M	Secondary static	1.7	Generalised	Fixed	Acquired	Term HIE (Perinatal brain injury)	Gliosis of thalami, lentiform nuclei and periorolandi cortex	Trihexiphenidyl Baclofen	12	0.88	102.5	93	9.3
24	17.8	F	Secondary static	0.8	Generalised	Phasic	Acquired	Term HIE (Perinatal brain injury)	Gliosis of thalami, lentiform nuclei and periorolandi cortex	Baclofen	17	0.96	72	73	-1.4
25	7.2	M	Secondary static	0	Generalised	Phasic	Acquired	Term kernictuerus (Perinatal brain injury)	Gliotic change in Globi pallidi consistent with previous elevated bilirubin	None	7.2	1.00	66	77.5	-17.4
26	3.5	M	Secondary static	0	Generalised	Fixed	Acquired	Term kernictuerus (Crigler-Najjar) (Perinatal brain injury)	Gliotic change in Globi pallidi consistent with previous elevated bilirubin	Trihemiphenidyl Nitrazepam	3.5	1.00	98	106.5	-8.7
27	12.3	M	Secondary static	0	Generalised	Fixed	Acquired	Term kernictuerus (Perinatal brain injury)	Signal change in Globus pallidi	Baclofen, Trihexiphenidyl Co-careldopa	12.3	0.98	111.5	107	4.0
28	10.6	F	Secondary static	0.6	Generalised	Fixed	Inherited – inborn error of metabolism	Glutaric aciduria type 1 (Toxic/metabolic)	Gliotic change and volume loss in basal ganglia and white matter	Baclofen Chloral hydrate Diazepam Clonazepam Tetrabenazine	10	0.94	112.5	95.5	15.1
29	15.7	M	Secondary static	1.7	Generalised	Fixed	Inherited – inborn error of metabolism	Glutaric aciduria type 1 (Toxic/metabolic)	Gliotic change and volume loss in basal ganglia and white matter	Diazepam Baclofen Trihexyphenidyl	14	0.89	114.5	105.5	7.9
30	10.4	F	Secondary static	3	Left hemidystonia	Phasic	Acquired	Focal infarct (Vascular)	Infarct right caudate and pallidum	None	7.4	0.71	30.5	16.5	45.9
31	3.3	M	Secondary static	1.3	Generalised	Fixed	Inherited – Acquired	Bilateral nigrostriatal necrosis, Aicardi Goutieres type 6 ADAR1 positive	Bilateral nigrostriatal necrosis	Diazepam Baclofen Trihexiphenidyl	2	0.61	102.5	74	27.8
32	6.7	M	Secondary static	1.7	Generalised	Fixed	Inherited - Acquired	Bilateral nigro-striatal necrosis, Aicardi Goutieres type 6. ADAR1 positive	Bilateral nigrostriatal necrosis	Baclofen Carbamazepine	5	0.75	83	62	25.3
33	13.1	F	Secondary static	0.1	Generalised	Phasic	Uncertain	Unknown. Bilateral perisylvian polymicrogyria with epilepsy and dystonia	Malformation: Perisylvian polymicrogyria and cerebellar atrophy	Tetrabenazine Trihexiphenidyl	13	0.99	104	97	6.7
34	15.7	M	Secondary static	3.7	Generalised but predominantly right sided	Fixed	Probably Acquired	Unknown	Left putaminal reduced volume		12	0.76	80	65.5	18.1
35	7.1	M	Secondary static	0.1	Generalised	Fixed	Probably inherited: affected sister	Unknown	PVWM	Tetrabenazine Co-careldopa	7	0.99	97	101	-4.1
36	8.9	F	Secondary static	6	Generalised	Phasic	Probably inherited	Unknown. Congenital neuromuscular disorder: LMNA mutation	Normal	Trihexiphenidyl	2.9	0.33	52.5	64	-21.9
37	17.2	F	Secondary progressive	8.2	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	None	9	0.52	59.5	78	-31.1
38	7.9	F	Secondary progressive	5.9	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	Baclofen Diazepam Trihexiphenidyl	2	0.25	64	102	-59.4
39	10.5	M	Secondary progressive	5.5	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	Baclofen Diazepam	5	0.48	98	95.5	2.6
40	4.3	M	Secondary Progressive	1.5	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	Trihexiphenidyl Haloperidol Clonidine	2.8	0.65	97.5	N/A **	N/A**
41	13.6	F	Secondary progressive	3.6	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	Intrathecal baclofen	10	0.74	120	18	85.0
42	9.6	F	Secondary progressive	3.1	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-striatal tract.	Baclofen Trihexiphenidyl Gabapentin	6.5	0.68	82.5	85	-3.0
43	16.4	M	Secondary	4.4	Generalised	Fixed	Inherited – inborn	NBIA- PANK2 deficiency	Iron deposition in GP and nigro-	Diazepam	12	0.73	114	98.5	13.6

			progressive				error of metabolism		striatal tract.	Baclofen Tetrabenazine Clonazepam					
44	6	M	Secondary progressive	2	Generalised	Fixed	Inherited – inborn error of metabolism	NBIA- PANK2 deficiency	Iron deposition in GP and nigro- striatal tract.	Co-Careldopa, Trihexiphenidyl Sodium valproate	4	0.67	86.5	107	-23.7

Table S1 – Clinical Data for individual patients

PVWM = Periventricular White Matter signal change, GP = Globi pallidi

* patient had DBS electrodes removed 3 months post procedure due to infection

** patient had DBS electrodes removed 1 month post procedure to due brain abscess associated with coincident acute hepatitis A infection which had provoked status dystonicus prior to DBS surgery and followed by nephrotic syndrome after DBS surgery.