

## Supplementary table 1: Comprehensive list of included studies and subsequent list of references

Clinical studies of movement disorder aetiologies associated with SIB included in the literature review:

Disease	Study	Study type; patient characteristics and neuropsychiatric comorbidities: [n=SIB patients/total patients suffering from disease]	Details on SIB: - Onset (years): [reported n (SIB)/total patients] - Form [n] - Topography (body part) [n]	Movement disorder [n]	Treatment for SIB [n]
<b>Neurodevelopmental disorders</b>					
Tourette syndrome	[1]	Case-Control study n=26/111 (23.4%) Age: 10y (mean; SD: 2.8y) Sex: male/female (96/15) Comorbidities: • OCD [16/111] • ADHD [52/111]	Onset: not reported  <i>Forms (topography):</i> • Picking/scratching (skin, lips, nasal mucosa) [16/111] • Rubbing, licking, wiping (skin, lips, anus) [7/111] • Biting (finger, nail, lips) [7/111] • Hitting (head) [2/111] • Self-cutting (throat) [1/111] • Self-strangulation [1/111] • Picking (self-pinching; NOS) [1/111]	• Tics	not reported
Tourette syndrome	[2]	Case-Control study n=11/31 (35.5%) Age: 31.4y (mean; SD: 7.6) Sex: male/female (28/3) Comorbidities: • OCD [12/31] • Aggression [11/31] • ADHD [5/31] • Anxiety [4/31]	Onset: not reported  <i>Forms and topography:</i> • Not in detail reported • Examples: self-slapping (face), bumping against walls (NOS)	• Tics (in history and current) [27/31]	not reported

Tourette syndrome	[3]	Case-control study n=23/57 (40%) Age: 11.3y (mean, SD: 2.4) Sex: male/female (45/12) Comorbidities: <ul style="list-style-type: none"> <li>History of hyperactivity [36/57]</li> <li>History of impulsivity [35/57]</li> <li>ADHD [34/57]</li> <li>Compulsive rituals [23/57]</li> <li>History of aggressive behaviour [17/57]</li> <li>Obsessional thoughts [14/57]</li> </ul>	<i>Onset:</i> not reported  <i>Forms and topography:</i> <ul style="list-style-type: none"> <li>Only most common form reported: head banging [12/57]</li> </ul>	<ul style="list-style-type: none"> <li>Tics</li> </ul>	not reported
Tourette syndrome	[4]	Prospective cohort study n=32/148 (21.6%) (total number of patients with available data in study: n=171) Age: 29.1y (mean, SD: 10.8) Sex: male/female (134/37) Comorbidities: <ul style="list-style-type: none"> <li>OCD [97/151]</li> <li>Depression [70/148]</li> <li>Anxiety [53/148]</li> <li>ADHD [43/152]</li> </ul>	<i>Onset:</i> not reported  <i>Form and Topography:</i> not reported	<ul style="list-style-type: none"> <li>Tics</li> </ul>	not reported
Tourette syndrome	[5]	Cross-sectional study n=86/297 (29%) Age: 19.5y (mean, SD: 14.1) Sex: male/female (230/67) Comorbidities: <ul style="list-style-type: none"> <li>OCD [119/297]</li> <li>ADHD [118/297]</li> <li>ADHD+OCD [77/297]</li> <li>Rages, risk taking behaviour, anxiety</li> </ul>	<ul style="list-style-type: none"> <li>Severe SIB in n=12/297</li> </ul> <i>Onset:</i> not reported  <i>Forms:</i> [n not reported] <ul style="list-style-type: none"> <li>Picking/scratching</li> <li>Hitting</li> <li>Banging</li> <li>(Head banging counted as tic form)</li> </ul> <i>Topography:</i> not reported	<ul style="list-style-type: none"> <li>Tics</li> </ul>	not reported

		disorders, mood disorders, conduct disorders, eating disorders [n not determinable]			
Tourette syndrome	[6]	<p>Cross-sectional study</p> <p>n=490/3500 (14%)</p> <p>Age (at diagnosis): 13.2y (mean; SD: 9.6y)</p> <p>Sex: male/female (2846/654)</p> <p>Comorbidities:</p> <ul style="list-style-type: none"> <li>• ADHD [2100/3500]</li> <li>• OCD [945/3500]</li> <li>• OCB [1120/3500]</li> <li>• Learning disability [805/3500]</li> <li>• Conduct disorder/Oppositional defiant disorder [525/3500]</li> <li>• Mood disorder [700/3500]</li> <li>• Anxiety [630/3500]</li> <li>• PDD [158/3500]</li> <li>• Mental retardation/ID [137/3500]</li> <li>• History of anger problems [1295/3500]</li> <li>• Social skill problems [700/3500]</li> <li>• Sexual inappropriate behaviour [210/3500]</li> </ul>	<ul style="list-style-type: none"> <li>• SIB reported without trichotillomania (n=95)</li> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	not reported

Tourette syndrome	[7]	<p>Cross-sectional study</p> <p>n=8/12 (66%)  Age: 35.2y (mean)  Sex: male/female (12/0)  Comorbidities:</p> <ul style="list-style-type: none"> <li>• OCB [12]</li> <li>• Lack of impulse control [8]</li> <li>• Depression [7]</li> <li>• ADHD [4]</li> <li>• Addiction [3]</li> </ul>	<ul style="list-style-type: none"> <li>• Form, onset and topography not reported in detail</li> <li>• Examples named: head banging, hitting, self-biting, more severe SIB</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	not reported
Stereotypies/ stereotypic movement disorder (associated with ID and/or ASD)	[8]	<p>Cross-sectional study</p> <p>n=22/22 (subgroup of study population)  Age: 22.9y (mean, SD: not calculable)  Sex: male/female (10/12)  Comorbidities:</p> <ul style="list-style-type: none"> <li>• ID [22/22]</li> </ul>	<p><i>Onset</i>: not reported  <i>Forms and topography</i>: [n not calculable]</p> <ul style="list-style-type: none"> <li>• Hitting (self-slapping of head)</li> <li>• Others (voluntary falling) (NOS)</li> <li>• Scratching (NOS)</li> <li>• Banging</li> <li>• Biting</li> <li>• Picking</li> </ul> <p>Hitting (head)</p>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported
	[9]	<p>Cross-sectional study</p> <p>n=121/121 (subgroup of study population)  Age: 37.1y (Mean; SD: 12.2y)  Sex: male/female (68/53)  Comorbidities:</p> <ul style="list-style-type: none"> <li>• Mental retardation/ID [121/121]</li> <li>• PDD/ASD [74/121]</li> <li>• Eating disorder [73/121]</li> <li>• Impulse control disorder [58/121]</li> <li>• Anxiety [54/121]</li> <li>• Sexual disorder [53/121]</li> <li>• Mood disorder [50/121]</li> <li>• Schizophrenia [41/121]</li> <li>• Mania [40/121]</li> </ul>	<p><i>Onset</i>: not reported  <i>Form (topography)</i>:</p> <ul style="list-style-type: none"> <li>• Biting (NOS) [56/121]</li> <li>• Hitting (NOS) [53/121]</li> <li>• Banging (against hard objects) (NOS) [n=38/121]</li> <li>• Picking (sores, NOS) [33/121]</li> <li>• Pulling (hair, NOS) [15/121]</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported

	[10]	Open-label clinical trial n=11 (total study population) n=5 (overlap between SIB and stereotypy) Age: 8,7y (mean; SD: 2.2y) Sex: male/female (7/4) Comorbidities: • ID (11/11) • Anxiety (7/11)	<i>Onset:</i> not reported  <i>Form:</i> • Biting [6] • Hitting [5]  <i>Topography:</i> • Head (head, nose) [5] • Hands [5] • Mouth (lips) [1]	• Stereotypies [5]	<i>No effect:</i> • Risperidone [2/11]  <i>Reduction in SIB frequency (intensity, duration unchanged):</i> • Risperidone [9/11]
Angelman syndrome	[11]	Longitudinal observational study n=1/12 Age: 2.4y Sex: not reported Comorbidities: • DD	<i>Onset:</i> not reported <i>Forms (topography):</i> • Swallowing (hand mouthing)	• Stereotypies	not reported
Smith-Magenis syndrome	[11]	Longitudinal observational study n=1/12 Age: 3.1y Sex: not reported Comorbidities: • DD	<i>Onset:</i> not reported <i>Forms and topography:</i> • Swallowing (hand mouthing)	• Stereotypies	not reported

Cornelia de Lange syndrome	[12]	<p>Case-control study</p> <p>n=30/54</p> <p>Age: 13.9y (mean; SD: 9y) Sex: male/female (25/29)</p> <p>Comorbidities:</p> <ul style="list-style-type: none"> <li>• Aggression [17/54]</li> <li>• ID [exact n not reported]</li> <li>• Hyperactivity [exact n not reported]</li> <li>• Compulsions [exact n not reported]</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Picking [40/53]</li> <li>• Biting [30/53]</li> <li>• Banging [15/53]</li> <li>• Hitting [15/53]</li> <li>• Poking [11/53]</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Hand [40/53]</li> <li>• Face [36/53]</li> <li>• Body [34/53]</li> <li>• Head [16/53]</li> <li>• Mouth [6/53]</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies [at least 52/53]</li> </ul>	not reported
Lowe-syndrome	[13]	<p>Cross-sectional study</p> <p>n=36/47 (in 12-15y age group)</p> <p>Age: 16.8y (mean; Range: 6-41y)</p> <p>Sex: male/female (47/0)</p> <p>Comorbidities:</p> <ul style="list-style-type: none"> <li>• Aggressive-irritable behaviour [up to 45/47]</li> <li>• Poor eye contact [36/47]</li> <li>• Poor concentration [33/47]</li> <li>• Mental retardation/ID [24/32]</li> </ul>	<ul style="list-style-type: none"> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Infantile hypotonia [47/47]</li> <li>• Stereotypies/ stereotyped behaviour [40/47]</li> <li>• Repetitive hand, body, head movements [36/47]</li> <li>• Oral stereotypies [33/47]</li> <li>• Waving/shaking body parts [32/47]</li> <li>• Waving /shaking extremities: [31/47]</li> </ul>	not reported

Cri du chat syndrome	[14]	<p>Cross sectional study</p> <p>n=61/66 Age: 14.8y (mean; Range: 6-37y) Sex: male/female (27/39) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Aggressive behaviour [58/66]</li> <li>• ID (feature of syndrome, introduction)</li> <li>• Developmental delay (feature of syndrome, introduction)</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• One topography [4/66], ≥ 2 topographies [n=57/66]</li> </ul> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Others (vomiting/ruminating) [38/66]</li> <li>• Hitting (head or other body parts) [36/66]</li> <li>• Biting (NOS) [31/66]</li> <li>• Banging (head against objects) [31/66]</li> <li>• Rubbing (teeth grinding) [28/66]</li> <li>• Scratching (NOS) [27/66]</li> <li>• Pulling (hair) [23/66]</li> <li>• Penetrative (finger in cavities) [22/66]</li> <li>• Body to body (NOS) [21/66]</li> <li>• Pica [18/33]</li> <li>• Picking (pinching)[17/66]</li> <li>• Banging (body to object) [14/66]</li> <li>• Penetrative (objects in cavities) [13/66]</li> <li>• Swallowing (extreme drinking) [13/66]</li> <li>• Swallowing (air) [3/66]</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies [54/66]</li> </ul>	not reported
Cri du chat syndrome	[11]	<p>Longitudinal observational study</p> <p>n=1/12 Age: 3.8y Sex: male/female (not reported) Comorbidities:</p> <ul style="list-style-type: none"> <li>• DD</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms (topography):</i></p> <ul style="list-style-type: none"> <li>• Swallowing (hand mouthing)</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported

Smith-Magenis syndrome	[15]	Case-control study (part I), cross-sectional study (part II)  n=32/35 Age: 9y (mean; SD not calculable) Sex: male/female (15/20) Comorbidities: <ul style="list-style-type: none"> <li>• ID [35/35]</li> <li>• Lability [31/35]</li> <li>• Impulsivity [30/35]</li> <li>• Nervousness [23/35]</li> <li>• Aggression [20/35]</li> <li>• Hyperactivity, disobedience, tantrums, attention-seeking, sleep disturbances [33-35/35]</li> </ul>	Onset: not reported  <i>Topography</i> : not reported  <i>Form</i> : mean number of SIB forms/patient: 3,7 (SD: 2,1) <ul style="list-style-type: none"> <li>• Biting [27/35]</li> <li>• Hitting [23/32]</li> <li>• Pulling (nail) [9/32]</li> <li>• Penetrative (insertion of objects into body-openings) [8/32]</li> </ul> <ul style="list-style-type: none"> <li>• Overlap between SIB and stereotypies</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported
<i>Chromosomal aberrations:</i>					
Trisomy 13	[11]	Longitudinal observational study  n=1/12 Age: 2.2y Sex: not reported Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> </ul>	Onset: not reported <i>Forms (topography)</i> : <ul style="list-style-type: none"> <li>• Swallowing (hand mouthing)</li> <li>• Biting (thumb)</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported
Fragile X syndrome	[16]	Cross-sectional study  n=60/85 Age: 14y (mean; SD: 2,3y) Sex: male/female (85/0) Comorbidities: <ul style="list-style-type: none"> <li>• ID (part of diagnosis)</li> <li>• Aggression [70/85]</li> <li>• ASD [15/85]</li> <li>• ADHD [4/85]</li> <li>• PDD-NOS [1/85]</li> </ul>	Form, onset and topography not reported	<ul style="list-style-type: none"> <li>• Stereotypies [77/85]</li> </ul>	not reported



	[17]	Cross-sectional study n=40/50 Age: 15.6y (mean; SD: 4.3y) Sex: male/female (50/0) Comorbidities: • ID [50/50] • Aggressive behaviour [38/50]	<i>Onset:</i> not reported <i>Topography:</i> not reported  <i>Forms:</i> Most common forms reported: • Hitting (NOS) [25/50] • Biting (NOS) [15/50]	• Stereotypies [49/50]	not reported
	[18]	Cross-sectional study n=38/47 Age: 7.8y (mean; SD: 4.2y) Sex: male/female (35/12) Comorbidities: • ID [42/47] • Aggression [40/47] • Features of ADHD [39/40] • Eating disturbances [23/47] • ASD [20/47] • Avoidant behaviour [16/47] • Tantrum behaviour [11/47] • Conduct • Anxiety [9/47] • Repetitive behaviour [8/47] • Depressive features [8/47]	• Form, onset and topography not reported	• Stereotypies [47/47]	not reported
<b>Neurometabolic</b>					
Lesch-Nyhan syndrome	[19]	Cross-sectional study n=40/43 [n=3 with inconclusive diagnosis; all reports for n=40] Age: 13.9 (mean; SD: 7.7y; Range: 2-32y) Sex: male/female (40/0) Comorbidities: • Aggression [35/40]	<i>Onset:</i> 3y (mean, SD: 1.7y) • Finger biting appeared first [23/40]  <i>Forms (topography):</i> 26 SIB forms in total (biting forms not summarized) • Biting [37/40] (lips [33], fingers [29], arms [16], tongue [14], shoulder [8]) • Banging (throwing arm, leg, head	• Motor involvement characteristic of the disease [40/43]	<i>Not reported:</i> • Use of restraints [32/37] • Preventive teeth extraction [34/40]  <i>No effect on SIB:</i> • Protective mouthguards [11/15]

			<p>out at doorway) [35/40]</p> <ul style="list-style-type: none"> <li>• Banging (arching and head snapping) [34/40]</li> <li>• Banging (head) [30/40]</li> <li>• Others (putting feet under wheelchair) [23/40]</li> <li>• Others (putting fingers in spokes) [14/40]</li> <li>• Poking (eye) [13/40]</li> <li>• Biting (cheeks) [2/40]</li> <li>• Not otherwise specified (putting fingers in dangerous places) [2/40]</li> <li>• Nose banging [2/40]</li> <li>• Arm banging [2/40]</li> <li>• Banging (hitting knee to face) [2/40]</li> <li>• Others (flinging body out of bed) [2/40]</li> <li>• Swallowing (insert fingers down to throat) [2/40]</li> <li>• Others (self-drowning) [2/40]</li> <li>• Not otherwise specified (upsetting wheelchair) [1/40]</li> <li>• Others (flinging out of wheelchair) [1/40]</li> <li>• Others (digging leg on wheelchair) [1/40]</li> <li>• Others (putting arms and legs into closing car door) [1/40]</li> <li>• Burning (touching hot objects) [1/40]</li> <li>• Biting (toe) [1/40]</li> </ul>		<p><i>“Very helpful”:</i></p> <ul style="list-style-type: none"> <li>• Protective mouthguards [3/15]</li> <li>• Parental intervention techniques: “being aware of needs/stress reduction [n not reported]</li> <li>• Parental intervention techniques: “attending to physical comfort” [26/40]; “adjusting restraints” [23/40], “talk to patient [22/40], distraction of attention [19/40], supplying needs [13/40], positive reinforcement techniques [18/40]</li> <li>• Anticonvulsive [5/5], benzodiazepines [6/17], neuroleptics [1/8], antidepressants [1/4], chloralhydrate [1/2]</li> <li>• Psychosocial therapy [4/20]</li> </ul> <p><i>“Least helpful”:</i></p> <ul style="list-style-type: none"> <li>• Punitive parental intervention techniques or time-out or loss of reinforcement [in total 14/40 rated as helpful]</li> <li>• Psychosocial therapy [10/20]</li> </ul> <p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Sinemet [1/1], antidepressants [3/4], opiate antagonists [1/4],</li> </ul>
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					benzodiazepines [1/17] <ul style="list-style-type: none"> <li>• Psychosocial therapy [6/20]</li> <li>• Others: Talking therapy [5/40], hypnosis [1/40], acupuncture [1/40], religion [1/40], cognitive therapy [1/40]</li> </ul>
Lesch-Nyhan syndrome	[20]	Cross sectional study n=21/23 Age: 16.6y (Range 1-28y) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay 21/23</li> </ul>	<ul style="list-style-type: none"> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Spasticity</li> <li>• Hypotonia</li> <li>• Motor developmental delay</li> </ul>	not reported
Lesch-Nyhan syndrome	[21]	Open label trial n=10/14 Age: 28,1 (mean; Range 11-49y) Comorbidities: Anxiety [14]	Form, onset and topography not reported	<ul style="list-style-type: none"> <li>• Movement disorder (not otherwise specified)</li> </ul>	<i>Effect not in detail reported:</i> <ul style="list-style-type: none"> <li>• Physical restraints</li> <li>• Current therapy: allopurinol, gabapentin or pregabalin (9/14), carbamazepine or valproate (4/14)</li> </ul> SAM-Therapy: 4/14 tolerated dose escalation  <i>Reduction:</i> <ul style="list-style-type: none"> <li>• SAM: 3/14 SIB</li> </ul> <i>Worsening:</i> SAM 10/14 with AE (e.g. SIB worsening, increase in anxiety, Sleep disturbances, MD worsening)
Lesch-Nyhan syndrome	[22]	Randomized-controlled trial n=9 Age: 10.3y (mean; SD: 4.6y) Sex: male/female (9/0)	<i>Onset:</i> not reported  <i>Forms</i> <ul style="list-style-type: none"> <li>• Not otherwise specified</li> </ul>	<ul style="list-style-type: none"> <li>• Only stereotyped behaviour reported [9]</li> <li>• Dystonia [9]</li> </ul>	<i>Reduction: (not clear if significant)</i> <ul style="list-style-type: none"> <li>• Ecopipam [4]</li> </ul> <i>SIB as AE:</i>

		Comorbidities: <ul style="list-style-type: none"> <li>• ID [9]</li> <li>• Aggressive behaviour [9]</li> </ul>	<i>Topography:</i> <ul style="list-style-type: none"> <li>• Not reported</li> </ul>		<ul style="list-style-type: none"> <li>• Ecopipam [5]</li> </ul>
Lesch-Nyhan syndrome	[23]	Prospective open-label study n=5 Age: 11y (mean; SD:8.8y) Sex: male/female (5/0) Comorbidities: not reported	<i>Onset:</i> not reported <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting [5]</li> <li>• Banging [2]</li> <li>• Hitting [2]</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia [5]</li> <li>• Hypotonia [4]</li> <li>• Opisthotonos [2]</li> <li>• Spasticity [1]</li> <li>• Chorea [1]</li> <li>• Rigidity [1]</li> <li>• Spasticity [1]</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>• L-dopa [5]</li> </ul>
Lesch-Nyhan syndrome	[24]	Longitudinal observational study n=10 Age:14 y (mean; SD:4.2y) Sex: male/female (10/0) Comorbidities: <ul style="list-style-type: none"> <li>• DD [10]</li> <li>• ID [10]</li> </ul>	<i>Onset:</i> 7y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lip, buccal mucosa, tongue) [10]</li> <li>• Fingers [10]</li> </ul>	<ul style="list-style-type: none"> <li>• Spasticity</li> <li>• Choreoathetosis</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>• Tranquillizers, psychomimetics, l-5-hydroxytryptophan [10]</li> </ul>
<b>Others</b>					
<i>Perinatal brain damage</i>					
Hypoxia at birth	[11]	Longitudinal observational study n=1/12 Age: 2.5y Sex: not reported Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> </ul>	<i>Onset:</i> not reported <i>Form (topography):</i> <ul style="list-style-type: none"> <li>• Banging (head)</li> <li>• Hitting (head)</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported
Cerebral palsy	[11]	Longitudinal observational study n=1/12 Age: 1.9y Sex: not reported	<i>Onset:</i> not reported <i>Form (topography):</i> <ul style="list-style-type: none"> <li>• Banging (head)</li> <li>• Hitting (head)</li> <li>• Biting (thumb, arm)</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> </ul>	not reported

		Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> </ul>			
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Case series and case reports included in the literature review:

Disease	Study	Patient characteristics and neuropsychiatric comorbidities: [n = SIB patients/total patients suffering from disease]	Details on SIB: - Onset (years): [reported n(SIB)/total patients] - Form [n] - Topography (body part) [n]	• Movement disorder [n]	Treatment for SIB [n]
<b>Neurodevelopmental disorders</b>					
<i>Primary tic disorders:</i>					
Tourette syndrome	[25]	n=9 Age: 25y (mean; SD: 10y) Sex: male/female (8/1) Comorbidities: <ul style="list-style-type: none"> <li>• OCD [9]</li> <li>• ADHD [6]</li> <li>• Oppositional behaviour [2]</li> <li>• Depression [1]</li> <li>• Anxiety [4]</li> <li>• Lack of impulse control [1]</li> <li>• Mood disorder [4]</li> </ul>	Onset: not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Hitting [2]</li> <li>• Biting [2]</li> <li>• Swallowing [2]</li> <li>• Amputational [1]</li> <li>• Penetrative [1]</li> <li>• Banging [1]</li> <li>• Poking [1]</li> <li>• Picking [1]</li> <li>• Pulling [1]</li> <li>• Others [2]</li> <li>• NOS [4]</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth [3]</li> </ul>	• Tics	<i>No effect:</i> <ul style="list-style-type: none"> <li>• Clonidine, clonazepam, pimozide, risperidone, quetiapine, paroxetine, fluoxetine, haloperidol [1]</li> <li>• Tetrabenazine, sertraline, alprazolam, risperidone, fluvoxamine, haloperidol, valproic acid, thiothixene, olanzapine, clonidine, carbamazepine, fluoxetine bupropion, citalopram, imipramine, and a variety of CNS stimulants [1]</li> <li>• Haloperidol, ziprasidone, imipramine, clomipramine, bupropion, sertraline,</li> </ul>

			<ul style="list-style-type: none"> <li>• Extremities [2]</li> <li>• Trunk [2]</li> <li>• Head [2]</li> <li>• Internal [2]</li> <li>• Eye [1]</li> <li>• NOS [6]</li> </ul>		<p>fluoxetine, venlafaxine, clonidine, alprazolam, lithium [1]</p> <p><i>SIB changed under treatment:</i></p> <ul style="list-style-type: none"> <li>• Neck brace (SIB changed to harder punches to anterior neck) [1]</li> </ul> <p><i>Inconclusive effect:</i></p> <ul style="list-style-type: none"> <li>• Successive therapy with fluphenazine, behavioural therapy, cingulotomy, limbic leucotomy [1]</li> </ul> <p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>• DBS: GPi</li> </ul> <p><i>Not reported:</i> n=5</p>
Tourette syndrome	[26]	<p>n=1</p> <p>Age: 12y</p> <p>Sex: male/female (1/0)</p> <p>Comorbidities:</p> <ul style="list-style-type: none"> <li>• ADHD</li> <li>• OCD</li> <li>• Aggressive behaviour</li> </ul>	<p><i>Onset:</i> 8y</p> <p><i>Form:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> <li>• Hitting</li> <li>• Rubbing</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Head</li> <li>• Mouth</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>Partly effective:</i></p> <ul style="list-style-type: none"> <li>• Pharmacological medication (alone or a combination up to 3 drugs): risperidone, fluvoxamine, methylphenidate, aripiprazole</li> </ul> <p><i>SIB changed under treatment:</i></p> <ul style="list-style-type: none"> <li>• Mouth guard (self-hitting instead of self-biting)</li> </ul>
Tourette syndrome	[27]	<p>n=1</p> <p>Age: 54y</p> <p>Sex: male/female (1/0)</p> <p>Comorbidity:</p> <ul style="list-style-type: none"> <li>• OCD</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Form:</i></p> <ul style="list-style-type: none"> <li>• Penetrative</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Single and combination of: SSRI (including Fluoxetine), TCA (including clomipramine), haloperidol; cognitive behavioural therapy</li> </ul>

					<i>Remission:</i> <ul style="list-style-type: none"> <li>• Pregabalin</li> </ul>
Tourette syndrome	[28]	n=1 Age: 27y Sex: male/female (1/0) Comorbidity: <ul style="list-style-type: none"> <li>• OCD</li> </ul>	<i>Onset:</i> not reported  <i>Form:</i> not reported  <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth</li> <li>• Head</li> <li>• Fingers</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<i>Cessation:</i> <ul style="list-style-type: none"> <li>• DBS: Ncl. accumbens</li> </ul>
Tourette syndrome	[29]	n=1 Age: 38y Sex: male/female (1/0) Comorbidity: <ul style="list-style-type: none"> <li>• Anxiety</li> <li>• Irritability</li> <li>• Depressive symptoms</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Hitting</li> <li>• Biting</li> <li>• Poking</li> <li>• Banging</li> <li>• Picking</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth</li> <li>• Head</li> <li>• Eye</li> <li>• NOS</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>• Isolated and combined: clonazepam, haloperidol, risperidone, pimoziide, quetiapine, levomepromazine, sulpiride, olanzapine, clonidine, topiramate, sertraline, citalopram, nicotine patches, supportive and behavioural psychotherapy</li> </ul> <i>Inconclusive effect:</i> <ul style="list-style-type: none"> <li>• 23 Sessions ECT and maintenance therapy with olanzapine, sulpiride, citalopram, clonidine</li> </ul>
Tourette syndrome	[30]	n=1 Age: 28y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Anxiety</li> <li>• Psychotic symptoms</li> <li>• Compulsive features</li> </ul>	<i>Onset:</i> 12y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Hitting</li> <li>• NOS</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Head</li> <li>• NOS</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>• Successive combination of haloperidol, diazepam, biperiden</li> </ul> <i>Inconclusive effect:</i> <ul style="list-style-type: none"> <li>• 7 Sessions ECT and post-ECT pimoziide and carbamazepine (behaviour during next 2</li> </ul>

					years improved, SIB not explicit mentioned)
Tourette syndrome	[31]	n=1 Age: 4.1y Sex: male/female (0/1) Comorbidities: • OCD • Hyperactivity	<i>Onset:</i> About 4y  <i>Forms:</i> • Rubbing  <i>Topography:</i> • Mouth (teeth)	• Tics	<i>Reduction:</i> • Occlusal splint for stabilization/prevention, and behavioural therapy
Tourette syndrome	[32]	n=1 Age: 36y Sex: male/female (0/1) Comorbidities: • Signs of BPD • Aggressive behaviour • Anxiety • Depression • Impulsivity	<i>Onset:</i> 14y  <i>Forms:</i> • Biting • Pulling • Poking • Burning	• Tics	<i>Cessation:</i> • DBS with 4 leads in centromedian-parafascicular complex of thalamus and limbic part of GPi: ○ Bilateral thalamic stimulation: SIB ceased ○ Bilateral pallidal stimulation: SIB ceased ○ Combination of thalamic and pallidal stimulation: SIB ceased
Tourette syndrome	[33]	n=1 Age: 14y Sex: male/female (1/0) Comorbidities: • ADHD • OCD	<i>Onset:</i> 4y  <i>Forms:</i> • Pulling • Swallowing • Picking • Penetrative  <i>Topography:</i> • Mouth • Internal • Extremities • NOS	• Tics	<i>No effect:</i> • Medical history of: methylphenidate, imipramine, clonidine bupropion, valproate, haloperidol, sertraline, pimozone, clomipramine, paroxetine, fluphenazine, fluoxetine, venlafaxine, lithium, fluvoxamine, ziprasidone, topiramate, olanzapine, nicotine patch, clonazepam, lorazepam,



					<p>naltrexone, molindone, gabapentin, (often in combination with family therapy, behavioural and individual therapy)</p> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Stereotactic cingulotomy (area above the corpus callosum): reappearance of SIB after 1 month</li> <li>• Re-surgery with limbic leucotomy (under caudate nucleus): SIB reduced</li> </ul>
Tourette syndrome	[34]	<p>n=1 Age: 16y Sex: male/female (0/1) Comorbidity:</p> <ul style="list-style-type: none"> <li>• OCD</li> </ul>	<p><i>Onset: 16y</i></p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> <li>• Poking</li> <li>• Pulling</li> <li>• Scratching</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth</li> <li>• Eye</li> <li>• Head</li> <li>• Not otherwise specified</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>Reduction:</i></p> <p>Combination out of multiple approaches:</p> <ul style="list-style-type: none"> <li>○ ICU sedation, botulinum toxine for bite blocking (break of severe SIB episodes)</li> <li>○ Behavioural therapy (SIB replaced to less destructive forms)</li> <li>○ Prevention: winter and boxing gloves (SIB frequency reduced)</li> <li>○ Drug regimen at discharge out of: clonidine, docusate sodium, citalopram, clonazepam, clomipramine, aripiprazole, risperidone,</li> </ul>

					diphenhydramine, chlorhexidine, saltwater rinse
Tourette syndrome	[35]	n=1 Age: 25y Sex: male/female (1/0) Comorbidities: • OCD • Depression	<i>Onset:</i> not reported <i>Forms:</i> Poking <i>Topography:</i> Eye	• Tics	<i>Inconclusive reported:</i> • Clomipramine, buspirone hydrochloride
Tourette syndrome	[36]	n=1 Age: 13y Sex: male/female (1/0) Comorbidity: • OCD	<i>Onset:</i> not reported  <i>Forms:</i> • Biting • Picking • Hitting  <i>Topography:</i> • Lip • Head • (Leg; inconclusive reported, probably self-hitting on bed frame)	• Tics	<i>No effect:</i> • Haloperidol, bromazepam, flunitrazepam  <i>Reduction:</i> • Injection of diazepam  <i>Cessation of biting behaviour:</i> • Acrylic splint on mandibular teeth
Tourette syndrome	[37]	n=2 Age: 25.5y (mean) Sex: male/female (1/1) Comorbidity: • OCD [2]	<i>Onset:</i> not reported  <i>Forms:</i> • Biting • Scratching • Rubbing  <i>Topography:</i> • Mouth • Extremities • Hands	• Tics	<i>not reported</i>
Tourette syndrome	[38]	n=1 Age: 30y	<i>Onset:</i> 10y	• Tics	<i>No effect:</i> • Pimozide, fluphenazine,

		Sex: male/female (0/1) Comorbidity: • Not reported	<i>Forms:</i> • Hitting • Biting  <i>Topography:</i> not otherwise specified		chlorpromazine, thioridazine, benzodiazepines, barbiturates, buspirone, clonazepam, carbamazepine, phenytoin, TCA, calcium channel blockers, anticholinergics and others (not in detail reported)  <i>Reduction:</i> • Fluoxetine, oxycodone [1]
Tourette syndrome	[39]	n=1 Age: 19y Sex: male/female (1/0) Comorbidity: • Obsessive thoughts and compulsions • Suicidal thoughts (after SIB, therefore included)	<i>Onset:</i> 7y  <i>Forms:</i> • Banging • Biting • Burning • Poking • Not otherwise specified (finger-mutilative behaviour) • Others (forced Valsalva manoeuvre)  <i>Topography</i> • Head • Mouth • Hands • Eye • Internal	• Tics	<i>No effect:</i> • Sulpiride, chlorprothixene, clomipramine, l-tryptophan [1]  <i>Cessation:</i> • Bilateral limbic leucotomy, behavioural therapy, sulpiride
Tourette syndrome	[40]	n=1 Age: 13y Sex: male/female (1/0) Comorbidity: • Not reported	<i>Onset:</i> 9y  <i>Forms:</i> • Pulling • Not otherwise specified (self-hurting)  <i>Topography:</i> • Eyebrows/eye	• Tics	<i>No effect:</i> • Haloperidol, thioridazine, pimozide

Tourette syndrome	[41]	n=2 Age: 25y (mean) Sex: male/female (2/0) Comorbidities: • Anxiety	<i>Onset:</i> 10.5y (n=1 not reported)  <i>Forms:</i> • Pulling [2] • Poking [2] • Banging [1] • Rubbing [1]  <i>Topography:</i> • Eye [2] • Head [1] • Teeth [1] • Arms (shoulder) [1] • Trunk (neck) [1]	• Tics	<i>No effect:</i> • Pimozide [1] • Thioridazine, haloperidol, chlorpromazine, phenobarbital, phenytoin, narcotic analgesics, benzodiazepines, TCA, anticonvulsants [1]  <i>Inconclusive reported</i> • Haloperidol, individual, family and group therapy [1]
Tourette syndrome	[42]	n=1 Age: 17y Sex: male/female (1/0) Comorbidity: • Not reported	<i>Onset:</i> not reported  <i>Forms:</i> • Not otherwise specified  <i>Topography:</i> • Extremities • Head	• Tics	<i>No effect:</i> • Haloperidol, clonidine  <i>Reduction:</i> • Naloxone i.m.[1]
Tourette syndrome	[43]	n=1 Age: 29y Sex: male/female (1/0) Comorbidity: • OCD • Depression	<i>Onset:</i> not reported  <i>Forms:</i> • Banging • Pulling  <i>Topography:</i> • Head	• Tics	<i>No effect:</i> • Antipsychotics, SSRI and behavioural therapy
Tourette syndrome	[44]	n=1 Age: 27y Sex: male/female (0/1) Comorbidities: • Ultrarapid cycling Bipolar II Disorder • Anxiety • Phobias	<i>Onset:</i> 11y  <i>Forms:</i> • Pulling • Poking • Others (Self-strangulations attempts, self-squeezing of throat)	• Tics	<i>No effect:</i> • Haloperidol, pimozide, clonidine, benzodiazepines, alprazolam, TCA, fluoxetine, fluvoxamine, lithium carbonate, carbamazepine, valproic

		<ul style="list-style-type: none"> <li>• Body dysmorphic disorder</li> <li>• Eating disorder</li> <li>• Personality disorders: avoidant, dependent, BPD, histrionic, narcissic</li> <li>• Obsessive and compulsive symptoms</li> </ul>	<i>Topography:</i> <ul style="list-style-type: none"> <li>• Trunk (throat)</li> <li>• Eye</li> <li>• Mouth (tongue)</li> </ul>		<p>acid</p> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Behavioural therapy (temporary relief)</li> </ul>
Tourette syndrome	[45]	<p>n=1</p> <p>Age: 31y</p> <p>Sex: male/female (1/0)</p> <p>Comorbidity:</p> <ul style="list-style-type: none"> <li>• ADHD</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms and Topography:</i> not otherwise specified</p>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Flupentixol</li> </ul>
Tourette syndrome	[46]	<p>n=1</p> <p>Age: 12y</p> <p>Sex: male/female (1/0)</p> <p>Comorbidity: Not reported</p>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Pulling (Teeth extraction)</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Psychotherapy, haloperidol, benzodiazepines, TCA, pimozide</li> </ul>
Tourette syndrome	[47]	<p>n=1</p> <p>Age: 20y</p> <p>Sex: male/female (1/0)</p> <p>Comorbidity:</p> <ul style="list-style-type: none"> <li>• ADHD</li> <li>• OCD</li> </ul>	<p><i>Onset:</i> 18y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Hitting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Head</li> <li>• Eye</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>Preventive:</i></p> <ul style="list-style-type: none"> <li>• Helmet, gloves</li> </ul> <p><i>Cessation or remarkable reduction:</i></p> <ul style="list-style-type: none"> <li>• Posteroventral GPi-DBS in combination with bilateral anterior capsulotomy</li> </ul>
Tourette syndrome	[48]	<p>n=5/8</p> <p>Age:</p> <ul style="list-style-type: none"> <li>• SIB patients: 30.4y (mean; SD: 13y)</li> </ul> <p>Sex: male/female (5/0)</p> <p>Comorbidities:</p> <ul style="list-style-type: none"> <li>• OCD [5]</li> <li>• Depression [2]</li> <li>• Symptoms of ADHD [1]</li> <li>• Symptoms of generalized anxiety disorder [1]</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Hitting [3]</li> <li>• Poking [1]</li> <li>• Pulling [1]</li> <li>• Biting [1]</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Head [2]</li> <li>• Eye/eyelid [1]</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Haloperidol, pimozide, risperidone, clonidine, fluoxetine, clonazepam, pergolide [1]</li> <li>• Pimozide, risperidone, olanzapine, quetiapine, fluoxetine, fluvoxamine, sertraline, clomipramine, clonazepam [1]</li> <li>• Haloperidol, pimozide,</li> </ul>

			<ul style="list-style-type: none"> <li>• Mouth (cheek) [1]</li> <li>• Trunk (chest) [1]</li> <li>• Not otherwise specified [2]</li> </ul>		<p>risperidone, fluphenazine, clonidine, fluvoxamine, imipramine, nortriptyline, clonazepam, pergolide [1]</p> <ul style="list-style-type: none"> <li>• Haloperidol, risperidone, aripiprazole, fluphenazine, sertraline, clonazepam, tetrabenazine, guanfacine, topiramate, etanercept, N-acetyl cysteine [1]</li> <li>• Pimozide, risperidone, ziprasidone, aripiprazole, fluphenazine, clonidine, guanfacine, fluoxetine, clonazepam, topiramate [1]</li> </ul> <p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• DBS: Thalamus [3]</li> <li>• DBS: GPi [2]</li> </ul>
Tourette syndrome	[49]	<p>n=4/14</p> <p>Age:</p> <ul style="list-style-type: none"> <li>• Total TS cohort: 11.3y (mean; SD: 2.7y)</li> <li>• SIB patients: 13.3y (mean; SD: 2.5)</li> </ul> <p>Sex:</p> <ul style="list-style-type: none"> <li>• Total TS cohort: male/female (10/4)</li> <li>• SIB patients: male/female (3/1)</li> </ul> <p>Comorbidities: not reported</p>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Hitting [3]</li> <li>• Biting [1]</li> <li>• Rubbing [1]</li> <li>• Picking [1]</li> <li>• Banging [1]</li> <li>• not otherwise specified [2]</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth [4]</li> <li>• Extremities [2]</li> <li>• Trunk (chest) [2]</li> <li>• Not otherwise specified (skin) [1]</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p>not reported</p>
<i>Syndromes due to chromosomal aberrations</i>					
Fragile X	[50]	n=5	<i>Onset:</i>	<ul style="list-style-type: none"> <li>• Stereotypies [5]</li> </ul>	not reported

syndrome		Age: 15.2y (mean, SD: 4.3y) Sex: male/female (5/0) Comorbidities: <ul style="list-style-type: none"> <li>• ID [5]</li> <li>• Mood instability [5]</li> <li>• ASD (CARS rating) [3]</li> <li>• Anxiety [1]</li> <li>• Hyperactivity [1]</li> <li>• Tantrums/Aggression [1]</li> <li>• Risk behaviour [1]</li> </ul>	<ul style="list-style-type: none"> <li>• 7y [mean; n=2]</li> <li>• Not reported [3]</li> </ul> <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>• Biting (hands) [2]</li> <li>• Biting (NOS) [5]</li> </ul>	<ul style="list-style-type: none"> <li>• Scissor-like hand movements [1]</li> <li>• Hyperkinesia [1]</li> </ul>	
17q23.2 duplication	[51]	n=1 Age: 7.5y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• ID</li> <li>• ADHD</li> </ul>	<i>Onset:</i> not reported  <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>• Scratching (NOS)</li> <li>• Pulling (attempt to peel nails)</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotyped, repetitive hand movements</li> <li>• Fine motor skill impairment (writing, finger movements)</li> </ul>	not reported
Tourettism associated with Xq25 micro-duplication	[52]	n=1 Age: 16y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Anxiety</li> <li>• Aggressive behaviour</li> <li>• Developmental delay</li> </ul>	<ul style="list-style-type: none"> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> <li>• Mild tremor</li> </ul>	not reported
1q24-q25 genetic micro-deletion related phenotype	[53]	n=1 Age: 3.8y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay</li> <li>• Hyperactivity</li> <li>• Inattention</li> </ul>	<i>Onset:</i> not reported <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>• Banging (head):</li> <li>• Hitting (NOS)</li> </ul>	<ul style="list-style-type: none"> <li>• Diffuse hypotonia</li> </ul>	not reported
Cri du chat syndrome	[54]	n=5/7 (2 without reported movement disorder) Age: 35.4y (mean, SD: 12.5y) Sex: male/female (3/2) Comorbidities: <ul style="list-style-type: none"> <li>• ID [5]</li> <li>• DD [5]</li> <li>• Hyperactivity [2]</li> </ul>	<i>Onset:</i> not reported <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>• Not otherwise specified (automutilation) [4]</li> <li>• Banging (head) [3]</li> <li>• Scratching (NOS) [2]</li> <li>• Biting (hands, NOS) [2]</li> <li>• Biting (nails, NOS) [1]</li> </ul>	<ul style="list-style-type: none"> <li>• Spasticity [2]</li> <li>• Gait abnormalities [3]</li> <li>• Ataxia [2]</li> </ul>	not reported

		<ul style="list-style-type: none"> <li>Aggressive behaviour [1]</li> </ul>	<ul style="list-style-type: none"> <li>Picking (NOS) [1]</li> <li>Pulling (trichotillomania, NOS) [1]</li> <li>Others (self-luxation: patella, shoulder) [1]</li> </ul>		
Prader-Willi syndrome	[55]	<p>n=1 Age: 9y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>Compulsions/Obsession</li> <li>Eating disorder</li> <li>Temper tantrums</li> <li>ID</li> <li>Aggressive behaviour</li> </ul>	<p>Onset: 3y Forms (topography):</p> <ul style="list-style-type: none"> <li>Picking (wounds, mainly shoulder, nose)</li> <li>Picking (legs)</li> <li>Others (self-eating)</li> <li>Biting (lip)</li> <li>Others (sucking on wounds)</li> </ul>	<ul style="list-style-type: none"> <li>Muscular hypotonia</li> </ul>	<p>No effect on SIB:</p> <ul style="list-style-type: none"> <li>Arm casts</li> <li>Fluoxetine</li> </ul> <p>SIB controlled:</p> <ul style="list-style-type: none"> <li>Combination of: fluoxetine, naltrexone, restraints (NOS) and behavioural program</li> </ul>
18q genetic deletion syndrome	[56]	<p>n=1 Age: 32y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>ID</li> <li>ASD</li> <li>Aggressive behaviour</li> </ul>	<p>Onset: 1.3y Forms:</p> <ul style="list-style-type: none"> <li>Biting</li> </ul> <p>Topography:</p> <ul style="list-style-type: none"> <li>Fingers</li> </ul>	<ul style="list-style-type: none"> <li>Ataxia</li> </ul>	<p>Inconclusive:</p> <ul style="list-style-type: none"> <li>Primidone, carbamazepine</li> </ul>
<i>Others</i>					
GNAO1 encephalopathy	[57]	<p>n=2 Age: 8.1y (mean; SD: 2.7y) Sex: male/female (2/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>Severe ID/DD [2]</li> </ul>	<p>Onset: 5.6y (mean; SD: 3.4y) Forms (topography):</p> <ul style="list-style-type: none"> <li>Biting (lips) [2]</li> <li>Pulling (hair) [1]</li> </ul>	<ul style="list-style-type: none"> <li>Stereotypic hand movements [1]</li> <li>Dystonia [2]</li> <li>Paroxysmal choreoathetosis [2]</li> <li>Facial dyskinesia [2]</li> <li>Hypotonia [2]</li> </ul>	<p>No effect:</p> <ul style="list-style-type: none"> <li>Calcium folinate substitution [2]</li> </ul>
Mitochondriopathy (G8363A transfer RNA <sup>Lys</sup> genetic mutation)	[58]	<p>n=1 Age: 4.3y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>Hyperactivity</li> <li>Autism</li> <li>DD</li> </ul>	<ul style="list-style-type: none"> <li>Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>Motor dyspraxia</li> <li>Repetitive oral behaviours</li> </ul>	not reported



		<ul style="list-style-type: none"> <li>• ID</li> </ul>			
Salt-and-pepper syndrome ( <i>ST3GAL</i> -mutation)	[59]	<p>n=2 Age: not reported Sex: not in detail reported Comorbidities:</p> <ul style="list-style-type: none"> <li>• ID [2]</li> <li>• DD [2]</li> </ul>	<ul style="list-style-type: none"> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Choreoathetosis [2]</li> </ul>	not reported
Gaucher's disease (II)	[60]	<p>n=1 Age: 1.8y Sex: male/female (1/0)</p>	<ul style="list-style-type: none"> <li>• Rubbing (tongue thrusting; wounds by teeth)</li> </ul>	<ul style="list-style-type: none"> <li>• Involuntary movements</li> <li>• Myoclonus</li> </ul>	<p><i>Preventive effect, effect on SIB inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Self-restraint (tongue protector)</li> </ul>
Stereotypic movement disorder associated with ID and/or ASD	[61]	<p>n=1 Age: 11y Sex: male/female (1/0) Comorbidity:</p> <ul style="list-style-type: none"> <li>• ID</li> <li>• Aggressive behaviour</li> <li>• Hyperactivity</li> <li>• Social isolation</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lip)</li> <li>• Fingers</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypies</li> <li>• Athetoid movements of arms, head</li> </ul>	<p><i>Cessation: (effect lasting for about 30min)</i></p> <ul style="list-style-type: none"> <li>• Naloxone i.m.</li> </ul>
Primrose syndrome	[62]	<p>n=1 Age: 43y Sex: male/female (0/1) Comorbidities:</p> <ul style="list-style-type: none"> <li>• DD</li> <li>• Loose of vocalization</li> </ul> <p>Mental retardation (part of syndrome)</p>	<p><i>Onset:</i> not exactly reported, first named with 42y</p> <p><i>Forms (topography):</i></p> <ul style="list-style-type: none"> <li>• Hitting (Head)</li> </ul> <p>Banging ("hand-to-object" punching)</p>	<ul style="list-style-type: none"> <li>• Hand stereotypies</li> <li>• Tics</li> <li>• Spasticity</li> </ul>	not reported
Rett syndrome	[63]	<p>n=2 Age: 7y (mean) Sex: male/female (0/2) Comorbidity:</p> <ul style="list-style-type: none"> <li>• ID [2]</li> </ul> <p>Developmental delay [1]</p>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting [2]</li> </ul> <p><i>Topography:</i></p> <p>Hands</p>	<ul style="list-style-type: none"> <li>• Stereotypic movements [2]</li> <li>• Extrapyramidal cerebral palsy [1]</li> <li>• Axial hypotonia [1]</li> <li>•</li> </ul>	<p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Behavioural therapy (operant learning)</li> </ul>
	[64]	<p>n=1 Age: 3.5y Sex: male/female (0/1) Comorbidity:</p>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul>	<ul style="list-style-type: none"> <li>• Stereotypic hand mouthing</li> <li>• Hand stereotypies</li> <li>• Gait apraxia</li> </ul>	<p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Behavioural therapy</li> </ul>

		<ul style="list-style-type: none"> <li>ID</li> <li>DD</li> </ul>	<i>Topography:</i> Hands	<ul style="list-style-type: none"> <li>Hypotonia</li> </ul>	
Smith-Magenis syndrome	[65]	n=4/10 Age: 27y (mean; SD: 14y) Sex: male/female (2/8) Comorbidities: <ul style="list-style-type: none"> <li>ID [10/10]</li> <li>Impulsivity [8/10]</li> <li>Attention-seeking [8/10]</li> <li>Temper tantrums [7/10]</li> <li>Anxiety [7/10]</li> <li>Aggression [7/10]</li> <li>Hyperactive [5/10]</li> </ul> DD [exact n not reported]	<i>Onset:</i> not reported <i>Topography:</i> not reported <i>Forms:</i> <ul style="list-style-type: none"> <li>Examples named (picking [nail picking], penetrative [inserting objects into body orifices])</li> </ul>	<ul style="list-style-type: none"> <li>“Peculiar” movements [9/10]</li> <li>Unusual motor movements [7/10]</li> </ul>	not reported
				<ul style="list-style-type: none"> <li></li> </ul>	
<b>Neurometabolic disorders</b>					
Lesch-Nyhan syndrome	[66]	n=3 Age: 26y (mean; Range: 19-39y) Sex: male/female (3/0) Comorbidities: <ul style="list-style-type: none"> <li>DD [3]</li> <li>Aggression [1]</li> </ul>	<i>Onset:</i> not reported <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting [3]</li> <li>Hitting [3]</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth [3]</li> <li>Finger [2]</li> <li>Not otherwise specified [3]</li> </ul>	<ul style="list-style-type: none"> <li>Dystonia [3]</li> <li>Ballism [3]</li> <li>Strong retropulsive reaction [1]</li> </ul>	<i>Reduction:</i> <ul style="list-style-type: none"> <li>Intrathecal baclofen [2]</li> </ul> <i>Cessation:</i> <ul style="list-style-type: none"> <li>Intrathecal baclofen [1]</li> </ul>
Lesch-Nyhan syndrome	[67]	n=1 Age: 15y Sex: male/female (1/0) Comorbidities: not reported	<i>Onset:</i> 3y <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting</li> <li>Not otherwise specified</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Finger</li> <li>Eye</li> </ul>	<ul style="list-style-type: none"> <li>Dystonia</li> </ul>	<i>Preventive:</i> <ul style="list-style-type: none"> <li>Physical restraints (wrist restraints, mitts), teeth extraction</li> </ul> <i>Reduction:</i> <ul style="list-style-type: none"> <li>DBS: bilateral GPi electrodes (hint for SIB lateralisation after unilateral)</li> </ul>

			<ul style="list-style-type: none"> <li>• Mouth</li> <li>• Head (nose)</li> </ul>		electrode dysfunctionality)
Lesch-Nyhan syndrome	[68]	n=1 Age: 12y Sex: male/female (1/0) Comorbidities: not reported	<p><i>Onset:</i> 1y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> <li>• Not otherwise specified</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Finger</li> <li>• Mouth (lips, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Axial spasms</li> <li>• Hypotonia</li> </ul>	<p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• DBS: bilateral electrodes in GPi (anterior and posterior)</li> </ul>
Lesch-Nyhan syndrome	[69]	n=1 Age: 10y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay</li> </ul>	<p><i>Onset:</i> 8y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Banging</li> <li>• Biting</li> <li>• Scratching</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Head (ear)</li> <li>• Mouth (lip)</li> <li>• Hand</li> </ul>	<ul style="list-style-type: none"> <li>• Athetosis</li> </ul>	not reported
Lesch-Nyhan syndrome	[70]	n=2 Age: 12y (mean; Range10-14y) Sex: male/female (2/0) Comorbidities: <ul style="list-style-type: none"> <li>• ID [2]</li> <li>• Anxiety</li> </ul>	<p><i>Onset:</i> 3y (mean)</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (cheeks)</li> </ul>	<ul style="list-style-type: none"> <li>• Athetosis [2]</li> <li>• Spasticity [2]</li> </ul>	not reported
Lesch-Nyhan syndrome	[71]	n=1 Age: 4.3y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay</li> <li>• ID</li> <li>• Irritability</li> </ul>	<p><i>Onset:</i> 4y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (cheeks)</li> <li>• Fingers</li> </ul>	<ul style="list-style-type: none"> <li>• Athetoid movements</li> <li>• Opisthotonos</li> </ul>	<p><i>Preventive:</i></p> <ul style="list-style-type: none"> <li>• Gloves</li> </ul> <p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Phenobarbital, dilantin, forced hydration, allopurinol, low purine diet, anticonvulsants</li> </ul>
Lesch-	[72]	n=1	<i>Onset:</i> not reported	<ul style="list-style-type: none"> <li>• Spastic tetraplegia</li> </ul>	<i>Inconclusive:</i>

Nyhan syndrome		Age: 6y Sex: male/female (1/0) Comorbidities: • Developmental delay • ID • Emotional disturbance	<i>Forms:</i> • Biting  <i>Topography:</i> • Mouth (lip)		• Diazepam, restraints (hands, feet), phenobarbital, probenecid, sodium bicarbonate [1]
Lesch-Nyhan syndrome	[73]	n=1 Age: 8y Sex: male/female (1/0) Comorbidities • ID	<i>Onset:</i> not reported  <i>Forms:</i> • Biting  <i>Topography:</i> • Fingers • Mouth (lip)	• Hypertonicity • Choreoathetosis	<i>Preventive:</i> • Boxing gloves [1]
Lesch-Nyhan syndrome	[74]	n=1 Age: 4.5y Sex: male/female (1/0) Comorbidities • DD	<i>Onset:</i> not reported  <i>Forms:</i> • Biting • Rubbing (grinding)  <i>Topography:</i> • Fingers • Mouth (teeth)	• Athetosis • Spastic diplegia	<i>Preventive:</i> • Hand wrapping [1]  <i>Inconclusive:</i> • Alkali treatment, low purine diet [1]
Lesch-Nyhan syndrome	[75]	n=6 Age: 8.3y (mean; n=3 not reported) Sex: male/female (6/0) Comorbidities: • DD [6] • Aggressive behaviour [2] • ID [1]	<i>Onset:</i> 2y (mean; n=1 not reported)  <i>Forms:</i> • Biting [4] • Banging (against hard objects) [1] • Not otherwise specified [1] • Not reported [1]  <i>Topography:</i> • Mouth (lips) [5] • Finger [4] • Head (nose, head) [3]	• Opisthotonic spasms [3] • Opisthotonos [2] • Choreoathetosis [2] • Spasticity [2] • Dystonia • Spasms [2] • Involuntary movements (limbs) [1]	<i>Preventive:</i> • Restraints (elbow splints) [1] • Teeth extraction [2]  • Not reported [3]
Lesch-Nyhan syndrome	[76]	n=1 Age: 29y Sex: male/female (1/0)	<i>Onset:</i> 16y  <i>Forms:</i>	• Dystonia	<i>No effect:</i> • Allopurinol, baclofen, clonazepam, l-carnitine,

		Comorbidities • DD	<ul style="list-style-type: none"> <li>Scratching</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Eye, eyelid</li> </ul>		<p>clozapine, carbamazepine, paroxetine, gabapentin, amitriptyline, risperidone, olanzapine, thioridazine</p> <p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>DBS: Bilateral GPi (posteroventral) and post-DBS treatment with allopurinol, baclofen, clonazepam, l-carnitine</li> </ul>
Lesch-Nyhan syndrome	[77]	n=1 Age: 14y Sex: male/female (1/0) Comorbidities • DD	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>Mouth (lips, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>Ataxia</li> <li>Spasticity</li> <li>Hyperkinetic</li> <li>Choreoathetoid movements</li> </ul>	<p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>Acrylic dental bite plate</li> </ul>
Lesch-Nyhan syndrome	[78]	n=1 Age: 30y Sex: male/female (1/0) Comorbidities • ID	<p><i>Onset:</i> 0.5y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>Mouth (lips)</li> <li>Not otherwise specified</li> </ul>	<ul style="list-style-type: none"> <li>Dystonia</li> <li>Hypertonicity</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>Allopurinol, baclofen, clonazepam, quetiapine, l-carnitine, teeth-extraction</li> </ul> <p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>Bilateral BTX-A-injection in zygomatic muscles</li> </ul>
Lesch-Nyhan syndrome	[79]	n=1/4 (n=3 without SIB) Age: 32y Sex: male/female (1/0) Comorbidities • Developmental delay	<p><i>Onset:</i> 20y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Biting</li> <li>Burning</li> <li>Not otherwise specified (producing wounds)</li> <li>Self-dropping on the ground (throwing self forward)</li> </ul> <p><i>Topography:</i></p>	<ul style="list-style-type: none"> <li>Dystonia</li> <li>Spasticity</li> <li>Hypertonicity</li> </ul>	not reported

			<ul style="list-style-type: none"> <li>• Hands</li> <li>• Head (chin)</li> <li>• Not otherwise specified</li> </ul>		
Lesch-Nyhan syndrome	[80]	n=1 Age: 5y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lips)</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> </ul>	<i>Reduction:</i> <ul style="list-style-type: none"> <li>• Mouth guard</li> <li>• Lip bumper</li> </ul>
Lesch-Nyhan syndrome	[81]	n=1 Age: 10y Sex: male/female (1/0) Comorbidities: not reported	<i>Onset:</i> 8y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Hand</li> <li>• Mouth (lips, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Hyperkinetic dyskinesia</li> <li>• Spasticity</li> </ul>	<i>Reduction:</i> <ul style="list-style-type: none"> <li>• Repeated injection of BTX-A in combination with physical restraint</li> </ul>
Lesch-Nyhan syndrome	[82]	n=1 (only in one patient movement disorder reported) Age: 2.1y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• ID</li> </ul>	<i>Onset:</i> 2,1y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Hand</li> <li>• Mouth (lips, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>• Non-voluntary movements</li> </ul>	<i>Reduction:</i> <ul style="list-style-type: none"> <li>• Moderate effect: pulpotomy and crown amputation on causal teeth</li> </ul>
Lesch-Nyhan syndrome	[83]	n=2 Age: 15y (mean; Range: 8-22y) Sex: male/female (1/1) Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> </ul>	<i>Onset:</i> 5.7y (mean)  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lips, cheek)</li> <li>• Finger</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia [2]</li> <li>• Athetosis</li> <li>• Involuntary movements</li> </ul>	not reported
Lesch-Nyhan syndrome	[84]	n=2 Age: 5y (mean; Range: 4-6y) Sex: male/female (2/0) Comorbidities:	<i>Onset:</i> 1.6y (mean)  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul>	<ul style="list-style-type: none"> <li>• Athetoid quadriplegic palsy [2]</li> </ul>	<i>Cessation:</i> <ul style="list-style-type: none"> <li>• Physical restraints (arm restraint, gloves, mouth guard, self-controlled)</li> </ul>

		<ul style="list-style-type: none"> <li>• DD [1]</li> <li>• Not reported [1]</li> </ul>	<i>Topography:</i> <ul style="list-style-type: none"> <li>• Finger</li> <li>• Mouth (lips)</li> </ul>		helmet-mask [1]  <i>No effect:</i> <ul style="list-style-type: none"> <li>• Physical restraints (arm restraint, gloves, mouth guard, self-controlled helmet-mask [1])</li> </ul>
Lesch-Nyhan syndrome	[85]	n=1/2 Age: <ul style="list-style-type: none"> <li>• Both patients: 4.5y (mean; Range: 2.1-7y)</li> <li>• SIB patient:7y</li> </ul> Sex: male/female (2/0) Comorbidities: <ul style="list-style-type: none"> <li>• DD [2]</li> </ul>	<i>Onset:</i> 2.3y (mean; SD:0.7y)  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Finger</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Choreoathetosis</li> </ul>	not reported
Lesch-Nyhan syndrome	[86]	n=1 Age: 10y Sex: male/female (1/0) Comorbidities: not reported	<i>Onset:</i> 4y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lip)</li> <li>• Fingers</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Athetosis</li> </ul>	<i>Preventive:</i> <ul style="list-style-type: none"> <li>• Teeth extraction, elbow restraints</li> </ul> <i>Cessation:</i> <ul style="list-style-type: none"> <li>• DBS: Bilateral pallidal stimulation</li> </ul>
Lesch-Nyhan syndrome	[87]	n=1 Age: 16y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> <li>• Rubbing (grinding)</li> <li>• Poking</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lip, tongue, teeth)</li> <li>• Finger/hands</li> <li>• Eye</li> </ul>	<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Athetosis</li> <li>• Stereotypical behaviour</li> </ul>	<i>Preventive.</i> <ul style="list-style-type: none"> <li>• Ties for hands</li> </ul> <i>No effect:</i> <ul style="list-style-type: none"> <li>• Baclofen, diazepam, clomipramine, cyamemazine</li> </ul> <i>Cessation:</i> <ul style="list-style-type: none"> <li>• DBS: Double stimulation of limbic (anterior-ventral) and motor (posterior) GPi bilaterally (SIB disappeared with anterior-ventral stimulation)</li> </ul>

Lesch-Nyhan syndrome	[88]	n=1 Age: 1.4y Sex: male/female (1/0) Comorbidities: • Developmental delay • Aggressive behaviour	<i>Onset:</i> not reported  <i>Forms:</i> • Biting  <i>Topography:</i> • Mouth (lip, oral mucosa, cheeks)	• Indirectly reported: no head control	<i>No effect:</i> • Soft resin bite plate  <i>Inconclusive:</i> • Teeth extraction
Lesch-Nyhan syndrome	[89]	n=1 Age: 3y Sex: male/female (1/0) Comorbidities: not reported	<i>Onset:</i> 1.8y  <i>Forms:</i> • Biting  <i>Topography:</i> • Mouth (cheeks, lip)	• Spasticity • Hypertonicity	<i>No effect:</i> • Diazepam  <i>Cessation:</i> • Gabapentin
Lesch-Nyhan syndrome	[90]	n=1 (one case also reported in [73]) Age: 5y Sex: male/female (1/0) Comorbidity: • Developmental delay • ID	<i>Onset:</i> not reported  <i>Forms:</i> • Biting  <i>Topography:</i> • Mouth (lip) • Fingers	• Dystonia • Choreoathetosis • Spasticity • Hypertonicity • Double hemiparesis	<i>Inconclusive:</i> • Teeth extraction
Lesch-Nyhan syndrome	[91]	n=1 Age: 19y Sex: male/female (1/0) Comorbidity: • ID	<i>Onset:</i> 4y  <i>Forms:</i> • Biting  <i>Topography:</i> • Mouth (lip) • Fingers • Extremities (forearm)	• Dystonia • Opisthotonos	<i>Preventive:</i> • Arm protectors, gloves  <i>Cessation:</i> • DBS: Quadripolar DBS bilateral in GPi
Lesch-Nyhan syndrome	[92]	n=2/6 Age: • Total LNS cohort: 4.4y (mean, SD: 2.5y) • SIB patients: 7y (mean) Sex: male/female (2/0) Comorbidities:	<i>Onset:</i> 4y (n=1: not reported)  <i>Forms:</i> • Biting • Rubbing (grinding)  <i>Topography:</i>	• Athetoid movements [2] • Spasticity [2] • Mild generalized spasms [1]	<i>Preventive:</i> • Mouth guard, teeth extraction [1]  <i>Cessation:</i> • Probenecid [1]



		<ul style="list-style-type: none"> <li>• Developmental delay [2]</li> <li>• ID [2]</li> <li>• Irritability [1]</li> </ul>	<ul style="list-style-type: none"> <li>• Fingers [2]</li> <li>• Mouth (teeth, lip, oral mucosa)</li> </ul>		<p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Allopurinol [1]</li> <li>• Probenecid [1]</li> </ul>
Lesch-Nyhan syndrome	[93]	<p>n=4/5 Age: Total LNS cohort: 8.1y (mean; Range: 1.9-15y SIB patients: 9.6y (mean; 3.5-15y) Sex: male/female (3/1) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Aggressive behaviour [4]</li> <li>• DD [4]</li> <li>• Compulsions [1]</li> </ul>	<p><i>Onset:</i> 3.4y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting [4]</li> <li>• Banging [1]</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Finger</li> <li>• Mouth (lips, tongue)</li> <li>• Head</li> </ul>	<ul style="list-style-type: none"> <li>• Extrapyrmidal symptoms [4]</li> <li>• Dystonia [4]</li> <li>• Spasticity [2]</li> <li>• Hypotonia [2]</li> <li>• Hypertonicity [1]</li> </ul>	<p><i>Onset of SIB associated with:</i></p> <ul style="list-style-type: none"> <li>• Allopurinol therapy [3]</li> </ul> <p><i>Preventive:</i></p> <ul style="list-style-type: none"> <li>• Dental plates, physical restraints [1]</li> </ul> <p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Physical restraints (dental plates, others), allopurinol, purine restricted diet, diazepam, carbamazepine [1]</li> </ul> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• S-adenosylmethionine [3]</li> </ul> <p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>• S-adenosylmethionine [1]</li> </ul>
Lesch-Nyhan syndrome	[94]	<p>n=1 Age: 28y Comorbidities:</p> <ul style="list-style-type: none"> <li>• Developmental delay</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> <li>• Hitting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lips)</li> <li>• Finger</li> <li>• Extremities (legs, elbow)</li> </ul>	<ul style="list-style-type: none"> <li>• Spasticity</li> <li>• Dystonia</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Allopurinol, folic acid, thioridazine</li> </ul> <p><i>Poor effect:</i></p> <ul style="list-style-type: none"> <li>• Physical restraints</li> </ul> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Risperidone</li> </ul>
Lesch-Nyhan syndrome	[95]	<p>n=1 Age: 6.5y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Developmental delay</li> </ul> <p>Absence of alertness and curiosity</p>	<p><i>Onset:</i> 3y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <p>Mouth (lips)</p>	<ul style="list-style-type: none"> <li>• Choreoathetosis</li> <li>• Spasticity</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Allopurinol, 5-hydroxytryptophan, carbidopa [1]</li> <li>• Oral surgery, parent education, program for behaviour modification</li> </ul>

					<i>Inconclusive:</i> • Limp restraints not reported
Lesch-Nyhan syndrome	[96]	n=4 Age: 10y (mean; SD: 3.1y) Sex: male/female (4/0) Comorbidities: • ID [3] • Antisocial behaviour [1]	<i>Onset:</i> 8y [1] (n=3 not reported)  <i>Forms:</i> • not otherwise specified [2] • Biting [1] • Pulling (hair pulling) [1]  <i>Topography:</i> • Hands [3] • Mouth (lips) [3] • Head [1]	• Spasticity [3] • Rigidity [3] • Double hemiparesis [2] • Choreiform movements [1] • Choreoathetosis [1]	
Lesch-Nyhan syndrome	[97]	n=5 Age: 6y (mean, SD: 2.9y) Sex: male/female (5/0) Comorbidities: • Developmental delay [5] ID [5]	<i>Onset:</i> not reported  <i>Forms:</i> • Biting [5] • Scratching [5]  <i>Topography:</i> • Head (nose) [5] • Extremities (arms, forearms) [5] • Fingers [5] • Mouth [5]	• Chorea [5] • Spasticity [5] • Ataxia [5] • Dystonia [4]	<i>Preventive:</i> • Gloves, other forms of restraints (not otherwise specified) [5]  <i>Worsening:</i> • Carbidopa/levodopa [2] • Bromocriptine [1] • Tetrabenazine [1] • Fluphenazine [1]  <i>No effect:</i> • Fluphenazine [2] • Tetrabenazine [2]  <i>Reduction:</i> • Tetrabenazine [2] • Carbidopa/levodopa [1] • Bromocriptine [1]
Lesch-Nyhan syndrome	[98]	n=5 Age: 7.3y (mean, SD: 2.3y) Sex: male/female (5/0) Comorbidities: • ID [5]	<i>Onset:</i> 2.7y [5]  <i>Forms:</i> • Biting [5]	• Choreoathetosis [2] • Dystonia [2] • Opisthotonos [2] • Athetoid	<i>No effect:</i> • Low purine diet, benzbromarone [1] • Probenecid, sulfinpyrazone,

		Developmental delay [5]	<i>Topography:</i> <ul style="list-style-type: none"> <li>Fingers [5]</li> <li>Mouth (lips, oral mucosa) [5]</li> </ul>	<ul style="list-style-type: none"> <li>movements [2]</li> <li>Spastic paraplegia [1]</li> <li>Spasticity [1]</li> </ul>	benzbromarone, allopurinol [1] <ul style="list-style-type: none"> <li>Azathioprine [1]</li> </ul> <i>Reduction:</i> <ul style="list-style-type: none"> <li>Benzbromarone (SIB reduced only for 2 month) [1]</li> </ul> <i>Inconclusive:</i> <ul style="list-style-type: none"> <li>Probenecid, allopurinol, azathioprine, phenobarbital, mephobarbital, diazepam [1]</li> <li>Allopurinol [3]</li> </ul>
Lesch-Nyhan syndrome	[99]	n=9/22 Age: <ul style="list-style-type: none"> <li>Total cohort: 12.2y (mean; Range: 0.4y-35y)</li> <li>SIB patients: 14.5y (mean; SD: 10.9y)</li> </ul> Sex: male/female (22/0) Comorbidities: <ul style="list-style-type: none"> <li>ID [8]</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>Not otherwise specified</li> </ul> <i>Topography:</i> not reported	<ul style="list-style-type: none"> <li>Choreoathetosis [9]</li> <li>Spasticity [9]</li> </ul>	not reported
PTPS deficiency	[100]	n=1 Age: 21y Sex: male/female (0/1) Comorbidities: <ul style="list-style-type: none"> <li>Developmental delay</li> <li>ID</li> <li>Aggressiveness</li> </ul>	<i>Onset:</i> 9y  <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>Biting (fingers, hands, lips)</li> <li>Banging (head)</li> <li>Banging (arms against walls)</li> <li>Others (neck-squeezing)</li> </ul>	<ul style="list-style-type: none"> <li>Limb dystonia</li> <li>Stereotypies</li> <li>Choreic movements</li> </ul>	not reported
Hepato-lenticular de-generation	[101]	n=1 Age: 16y Sex: male/female (0/1) Comorbidity: <ul style="list-style-type: none"> <li>Anxiety</li> <li>Irritability</li> </ul>	<i>Onset:</i> 3 months before examination (about 16y)  <i>Forms (topography):</i> <ul style="list-style-type: none"> <li>Skin scratching (chest)</li> </ul>	<ul style="list-style-type: none"> <li>Hand tremor</li> </ul>	<i>SIB reduced:</i> <ul style="list-style-type: none"> <li>Low-dose haloperidol and escitalopram</li> </ul>

		<ul style="list-style-type: none"> <li>• Mood instability</li> <li>• Impulse control disorder</li> </ul>			
PKU (late treated)	[102]	<p>n=1 Age: 73y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>• ID</li> </ul>	<p><i>Onset:</i> not reported <i>Forms and topography:</i></p> <ul style="list-style-type: none"> <li>• Banging (“body-to-object banging”; [head, back, body])</li> </ul>	<ul style="list-style-type: none"> <li>• Ataxia</li> <li>• Spasticity</li> <li>• (Leg tremor and spasms preceded SIB)</li> </ul>	<p><i>SIB controlled:</i></p> <ul style="list-style-type: none"> <li>• Strict dietary control</li> <li>• During SIB episodes: lorazepam and paracetamol</li> </ul>
<b>Neurodegenerative</b>					
Hereditary spastic paraplegia (with <i>B4GALNT1</i> mutation)	[103]	<p>n=2 Age: in the 30s Sex: not reported Comorbidities:</p> <ul style="list-style-type: none"> <li>• ID [2]</li> <li>• Developmental disability [2]</li> <li>• Autistic features [2]</li> <li>• Social phobia [2]</li> <li>• Paranoia [2]</li> </ul>	<ul style="list-style-type: none"> <li>• Occasional SIB</li> <li>• Form, onset and topography not reported</li> </ul>	<ul style="list-style-type: none"> <li>• Spasticity [2]</li> <li>• Gait abnormalities [2]</li> <li>• Hyperextended knees [2]</li> <li>• Manneristic movements [2]</li> </ul>	<ul style="list-style-type: none"> <li>• not reported</li> </ul>
Neuro-acanthocytosis	[104]	<p>n=1 Age: 28y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Agitation</li> <li>• Depressive symptoms</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Picking</li> <li>• Banging</li> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Finger</li> <li>• Mouth (oral mucosa, lips)</li> <li>• Head</li> <li>• Not otherwise specified (skin)</li> </ul>	<ul style="list-style-type: none"> <li>• Shoulder jerks</li> <li>• Involuntary movements</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Biperiden, pimozide, haloperidol</li> </ul> <p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Self-made apparatus for distraction of biting urge</li> </ul> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Quetiapine within medication scheme of paroxetine and biperiden (re-emerging SIB after dosage reduction)</li> </ul>
Neuro-acanthocytosis	[105]	<p>n=1 Age: 32y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Self-neglect</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul>	<ul style="list-style-type: none"> <li>• Tics</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Patient-initiated physical restraint (scarf in mouth) and fluoxetine together with sequential trials of</li> </ul>

		<ul style="list-style-type: none"> <li>Abulia</li> <li>Features of OCD</li> </ul>	<i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (lips)</li> </ul>		quetiapine, clozapine, topiramate, clonazepam  <i>Reduction:</i> <ul style="list-style-type: none"> <li>Soft-mouth guard</li> </ul>
Neuro-acanthocytosis	[106]	n=1 Age: 39y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>Self-neglect</li> <li>Agitation</li> <li>Restlessness</li> <li>Impaired attention, memory, concentraion</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>Scratching</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Head</li> </ul>	<ul style="list-style-type: none"> <li>Choreiform movements</li> <li>Nonpurposeful movements</li> <li>Repetitive movements</li> </ul>	not reported
Neuro-acanthocytosis	[107]	n=1 Age: 46y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>Episode of psychosis</li> <li>Mild cognitive impairment</li> </ul>	<i>Onset:</i> 36y  <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting</li> </ul> <i>Topography:</i> Mouth (lips, tongue)	<ul style="list-style-type: none"> <li>Ataxia</li> <li>Bradyimia</li> <li>Bradykinesia</li> <li>Rigidity</li> <li>Dystonia</li> <li>Tics</li> <li>Choreiform movements</li> </ul>	<i>Inconclusive:</i> <ul style="list-style-type: none"> <li>Pimozide, buspirone</li> </ul>
Neuro-acanthocytosis	[108]	n=1/2 Age: 36y Sex: male/female (0/1) Comorbidities: <ul style="list-style-type: none"> <li>Eating difficulties</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (lip, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>Choreatic movements</li> <li>Dystonia</li> <li>Oral dyskineic movements and uncontrolled tongue protrusions</li> <li>Ataxia/Gait disorder</li> <li>Rigidity</li> </ul>	not reported
Neuro-acanthocytosis	[109]	n=1 Age: 52y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>ID</li> <li>Aggressiveness</li> </ul>	<i>Onset:</i> 40y  <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (lip, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>Chorea</li> <li>Orofacial dyskinesia</li> <li>Hyperkinetic movements (facial, perioral, periorbital, lingual)</li> </ul>	not reported

				<ul style="list-style-type: none"> <li>• Dystonia</li> <li>• Dysarthria"</li> </ul>	
Neuro-acanthocytosis	[110]	<p>n=2 Age: 48y (mean; Range: 42-54y) Sex: male/female (0/2) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Episodes of anxiety, episodes of depression [1]</li> <li>• Not reported [1]</li> </ul>	<p><i>Onset:</i> 38.5y (mean)</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lip, tongue, oral mucosa)</li> </ul>	<ul style="list-style-type: none"> <li>• Involuntary movements</li> <li>• Orofacial dyskinesia</li> <li>• Chorea</li> <li>• Hypotonia</li> <li>• Tics</li> </ul>	<p><i>Limited success:</i></p> <ul style="list-style-type: none"> <li>• Sequential trials with: Benzodiazepine, oral nondeposit form of haloperidol, biperiden hydrochloride, trihexyphenidyl hydrochloride, clonazepam, levodopa, fluoxetine hydrochloride, vitamin B complex therapy [1]</li> <li>• not reported [1]</li> </ul>
Neuro-acanthocytosis	[111]	<p>n=1 Age: 36y Sex: male/female (1/0) Comorbidities: not reported</p>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lip, tongue)</li> </ul>	<ul style="list-style-type: none"> <li>• Orofacial dyskinesia</li> <li>• Chorea</li> <li>• Intermittent fast bending of trunk/neck</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Tetrabenazine, haloperidol, olanzapine, clonazepam</li> </ul> <p><i>Cessation (almost):</i></p> <ul style="list-style-type: none"> <li>• DBS: bilateral implantation of electrodes in GPi</li> </ul>
Neuro-acanthocytosis	[112]	<p>n=1 Age: 36y Sex: male/female (0/1) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Cognitive impairment</li> </ul>	<p><i>Onset:</i> 32y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lip)</li> </ul>	<ul style="list-style-type: none"> <li>• Orofacial dyskinesia</li> <li>• Chorea</li> <li>• Dystonia</li> <li>• Dysarthria</li> <li>• Involuntary vocalisations"</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Haloperidol, clonazepam</li> </ul>
Neuro-acanthocytosis	[113]	<p>n=1/2 Age: 33y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>• Mild cognitive impairment</li> </ul>	<p><i>Onset:</i> 29y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (lip)</li> </ul>	<ul style="list-style-type: none"> <li>• Involuntary movements</li> <li>• Chorea</li> <li>• Orofacial dyskinesias</li> <li>• Dysarthria</li> <li>• Sudden flexion/extension of</li> </ul>	<p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Tetrabenazine</li> </ul> <p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Tiapride (reported as reduced dyskinesias)</li> </ul>

				<ul style="list-style-type: none"> <li>arms</li> <li>Dystonia</li> <li>Dysdiadochokinesia</li> <li>Parkinsonism</li> <li>Involuntary vocalisations</li> </ul>	
NBIA syndromes	[114]	<p>n=1 Age: 10y Sex: male/female (0/1) Comorbidities:</p> <ul style="list-style-type: none"> <li>DD</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>Mouth (lip, tongue)</li> <li>Fingers</li> </ul>	<ul style="list-style-type: none"> <li>Dystonia</li> <li>Oromandibular dyskinesias</li> <li>Choreiform movements</li> <li>Apraxia</li> <li>Opisthotonos</li> <li>Retrocollis</li> </ul>	<p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>Local applications and antibiotics for oral ulcerations, oral trihexyphenidyl and baclofen for dystonia</li> </ul>
NBIA syndromes	[115]	<p>n=1 Age: 11y Sex: male/female (0/1) Comorbidities:</p> <ul style="list-style-type: none"> <li>DD</li> </ul>	<p><i>Onset:</i> 11y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Rubbing (thrusting)</li> <li>Not otherwise specified</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>Mouth (tongue, lip, oral mucosa)</li> </ul>	<ul style="list-style-type: none"> <li>Oral dyskinesia</li> <li>Generalized dystonia</li> <li>Spasticity</li> <li>Opisthotonos</li> </ul>	<p><i>Reduction (for tongue-thrusting movements)</i></p> <ul style="list-style-type: none"> <li>Trihexyphenidyl hydrochloride</li> </ul>
NBIA syndromes	[116]	<p>n=1 Age: 7y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>Learning disability</li> </ul>	<p><i>Onset:</i> not reported</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>Biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>Mouth (tongue, lip, oral mucosa)</li> </ul>	<ul style="list-style-type: none"> <li>Orofacial dystonia</li> <li>Spasms</li> </ul>	<p><i>Cessation:</i></p> <ul style="list-style-type: none"> <li>Bite guard, teeth extraction of primary incisors</li> </ul>
juvenile Parkinsonism with <i>FBOX07</i> mutation	[117]	<p>n=1 Age: 16y Sex: male/female (1/0) Comorbidities:</p> <ul style="list-style-type: none"> <li>ID</li> <li>Behavioural disturbances (aggression, mood instability)</li> </ul>	<p><i>Form, onset and topography not reported</i></p> <ul style="list-style-type: none"> <li>Drug-induced onset under 1mg/d Rasagiline (not reported in detail)</li> </ul>	<ul style="list-style-type: none"> <li>Postural instability</li> <li>Bradykinesia</li> <li>Rigidity</li> <li>Resting tremor</li> <li></li> </ul>	<p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>Clozapine (acceptable level of "psychiatric symptoms")</li> </ul>
Cortico-	[118]	n=1	<ul style="list-style-type: none"> <li>Episodic SIB</li> </ul>	<ul style="list-style-type: none"> <li>Arrhythmic tremor</li> </ul>	not reported

basal syndrome (antiglycine receptor mediated)		Age: 55y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Visual hallucinations</li> <li>• Loss of language</li> <li>• Visual agnosia</li> <li>• Global aphasia</li> <li>• Apathy</li> </ul>	<ul style="list-style-type: none"> <li>• <i>Form, onset and topography not reported</i></li> </ul>	<ul style="list-style-type: none"> <li>• Rigidity</li> <li>• Anterocollis</li> <li>• Hemifacial myoclonus</li> </ul>	
Alzheimer's disease (associated with PS-1 gene mutation)	[119]	n=1 Age: 35y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Depression</li> <li>• Decreasing memory, attention</li> <li>• Mood instability</li> </ul>	<ul style="list-style-type: none"> <li>• SIB not otherwise specified (onset, form, topography)</li> </ul>	<ul style="list-style-type: none"> <li>• Occasional multifocal myoclonus</li> </ul>	Not reported
SCA 17	[120]	n=2 Age: 66y [1; n=1 not reported] Sex: male/female (0/2) Comorbidities: <ul style="list-style-type: none"> <li>• Low mood [2]</li> <li>• Cognitive deterioration [1]</li> <li>• Mild impairment of memory, attention and concentration [1]</li> </ul>	<i>Onset:</i> 64y [1; n=1 not reported]  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Scratching</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Extremities (upper limbs, arms)</li> <li>• Trunk (chest)</li> </ul>	<ul style="list-style-type: none"> <li>• Gait abnormalities [2]</li> <li>• Choreic movements [1]</li> <li>• Generalised chorea [1]</li> </ul>	not reported
<b>Characteristic structural brain changes</b>					
<i>RTTN</i> mutation	[121]	n=1 Age: 5.8y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>• Developmental delay</li> <li>• Agitation</li> </ul>	<i>Onset:</i> about 5y  <i>Forms and topography:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Fingers</li> </ul>	<ul style="list-style-type: none"> <li>• Motor stereotypies</li> <li>• Truncal hyperkinetic movements</li> <li>• Limb spasticity</li> <li>• Axial hypotonia</li> </ul>	not reported
GLHS	[122]	n=1 Age: 3y Sex: male/female (1/0)	<i>Forms, onset and topography not reported</i>	<ul style="list-style-type: none"> <li>• Ataxia</li> </ul>	not reported



		Comorbidities: <ul style="list-style-type: none"> <li>• ASD</li> <li>• Restlessness</li> <li>• Aggressiveness</li> </ul>			
GLHS	[123]	n=1/2 Age: 11y Sex: male/female (0/1) Comorbidities: <ul style="list-style-type: none"> <li>• ID</li> <li>• DD</li> </ul>	<i>Onset:</i> 3y  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Rubbing</li> <li>• not otherwise specified (tearing off)</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Head (nose)</li> <li>• Eye</li> </ul>	<ul style="list-style-type: none"> <li>• Ataxia</li> </ul>	not reported
GLHS	[124]	n=1/3 Age: not reported Sex: male/female (0/1) <ul style="list-style-type: none"> <li>• ID</li> <li>• Developmental delay</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Rubbing</li> <li>• not otherwise specified</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Head (face)</li> <li>• Eye</li> </ul>	<ul style="list-style-type: none"> <li>• Ataxia</li> </ul>	not reported
Hypomyelination with atrophy of the basal ganglia and cerebellum (H-ABC)	[125]	n=1 Age: 13y Sex: male/female (0/1) Comorbidities: <ul style="list-style-type: none"> <li>• DD</li> <li>• ID</li> </ul>	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth</li> </ul>	<ul style="list-style-type: none"> <li>• Oromandibular dystonia</li> <li>• Global hypertonicity</li> <li>• Progressive motor impairment</li> </ul>	<i>Inconclusive:</i> <ul style="list-style-type: none"> <li>• Botulinum toxin, teeth extraction, clonazepam, trihexyphenidyl, levodopa</li> </ul>
Megalencephalic leucoencephalopathy (MLC)	[126]	n=1 Age: 10y Sex: male/female (1/0)	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>• Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>• Mouth (lips)</li> </ul>	<ul style="list-style-type: none"> <li>• Oromandibular dystonia</li> </ul>	<i>Cessation:</i> <ul style="list-style-type: none"> <li>• Botulinum toxin-A injection in orbicularis oris muscle and mentalis muscle</li> </ul>
Rhombencephalo-	[127]	n=1 Age: 22y	<i>Onset:</i> 19y	<ul style="list-style-type: none"> <li>• Ataxia</li> </ul>	<i>Relative ineffective:</i> <ul style="list-style-type: none"> <li>• Serotonergic medications</li> </ul>

synopsis		Sex: male/female (1/0) Comorbidities: • ID • OCD	<i>Forms:</i> • Not otherwise specified (stereotypical oral self-mutilation) • Not otherwise specified (self-touching)  <i>Topography:</i> • Mouth (teeth and oral mucosa)		
Joubert syndrome	[128]	n=2 Age: 5y (mean; SD: 3y) Sex: male/female (1/1) Comorbidities: • ID [2]	• Form, onset and topography not reported	• Abnormal eye movements • Floppiness • Ataxia	not reported
Chiari malformation type II	[129]	n=1 Age: 2.8y Sex: male/female (1/0)	<i>Onset:</i> not reported  <i>Form:</i> • Biting  <i>Topography:</i> • Mouth (lip)	• Quadriplegia	<i>Cessation:</i> • Protective lip bumper
<b>Others</b>					
<i>Autoimmune</i>					
Anti-NMDA receptor encephalitis	[130]	n=1 Age: 34y Sex: male/female (1/0) Comorbidity: • Aggressive behaviour	<i>Form, onset and topography not reported</i>	• Rigidity • Orofacial dyskinesia • Cyclical motions of legs and arms	not reported
<i>Drug induced</i>					
Dopamine agonist (PD)	[131]	n=1 Age: 51y Sex: male/female (1/0) Comorbidities: • Mild depression	<i>Onset:</i> 51y <i>Forms:</i> • Picking (after increased ropinirole dosage to 2-3mg/d)	• Dyskinesia • Tremor • Rigidity	<i>Cessation:</i> • Stop of ropinirole therapy

		<ul style="list-style-type: none"> <li>Mild subcortical dementia</li> <li>Anxiety</li> <li>Hyperactivity</li> </ul>	<i>Topography:</i> <ul style="list-style-type: none"> <li>Head</li> </ul>		
Neuroleptic (main diagnosis not reported)	[132]	n=1 Age: 21y Sex: male/female (1/0) Comorbidities: <ul style="list-style-type: none"> <li>Psychotic symptoms</li> <li>ID</li> </ul>	<i>Onset:</i> 21y  <i>Forms and topography:</i> <ul style="list-style-type: none"> <li>Biting (after injection of Flupentixol 50mg i.m. [+ Diazepam 7.5mg p.o.])</li> <li>Amputational</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (tongue)</li> </ul>	<ul style="list-style-type: none"> <li>Orolingual dyskinesia</li> </ul>	<i>Cessation (for short span of time; afterwards auto-amputation of tongue):</i> <ul style="list-style-type: none"> <li>Biperiden, diazepam, antibiotics (ampicillin, cloxacillin, metronidazole), hyoscine i.m., pethidine i.m.</li> </ul>
<i>Post infectious</i>					
Secondary to meningitis in infancy	[133]	n=1 Age: 14.2y Sex: male/female (0/1) Comorbidities: not in detail reported	<i>Onset:</i> several months before consultation  <i>Form:</i> <ul style="list-style-type: none"> <li>Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (lip, oral mucosa)</li> </ul>	<ul style="list-style-type: none"> <li>Mandibular dystonia</li> </ul>	<i>Successful prevention:</i> <ul style="list-style-type: none"> <li>Physical restraint: orthodontic bands, lip bumper</li> </ul> <i>No effect:</i> <ul style="list-style-type: none"> <li>Diazepam</li> </ul>
<i>Post traumatic</i>					
Secondary to traffic accident	[134]	n=1 Age: 12y Sex: male/female (1/0)	<i>Onset:</i> not reported  <i>Forms:</i> <ul style="list-style-type: none"> <li>Biting</li> </ul> <i>Topography:</i> <ul style="list-style-type: none"> <li>Mouth (tongue)</li> </ul>	<ul style="list-style-type: none"> <li>Spasms (jaws)</li> </ul>	<i>Inconclusive:</i> <ul style="list-style-type: none"> <li>Bite guard</li> </ul>
<i>Not otherwise specified (NOS)</i>					
Dystonia (prenatal diagnosis)	[135]	n=1 Age: 1.4y Sex: male/female (0/1)	<i>Onset:</i> 1.4y  <i>Forms:</i>	<ul style="list-style-type: none"> <li>Oromandibular dystonic movements</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>Paracetamol, morphine, diazepam, fosphenytoin</li> </ul>

of por-encephalic cavity and palsy of III. right cranial nerve)			<ul style="list-style-type: none"> <li>• Severe biting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Mouth (tongue)</li> </ul>	<ul style="list-style-type: none"> <li>• Head stereotypies</li> <li>• Bruxism</li> <li>• Spasms</li> <li>• Trismus</li> <li>• Choreodystonic movements</li> <li>• Upper limb hemiparesis</li> <li>• Upper limb hypertonicity</li> </ul>	<p>sodium, hydroxyzyine, oxcarbazepine, levodopa, trihexyphenidyl hydrochloride</p> <p><i>Inconclusive:</i></p> <ul style="list-style-type: none"> <li>• Tetrabenazine and together with trihexyphenidyl hydrochloride</li> </ul> <p><i>Partial efficacy:</i></p> <ul style="list-style-type: none"> <li>• Clonazepam</li> </ul> <p><i>Effective:</i></p> <ul style="list-style-type: none"> <li>• Preventive sedation and analgesia, preventive bite guard, composite application on sharp teeth, botulinum toxine injection in masseter muscle and orbicularis oris muscle</li> </ul>
Stereotypic movement disorder	[136]	<p>n=1 Age:49y Sex: male/female (1/0) Comorbidity:</p> <ul style="list-style-type: none"> <li>• Episode of Anorexia nervosa</li> </ul>	<p><i>Onset:</i> 22y</p> <p><i>Forms:</i></p> <ul style="list-style-type: none"> <li>• Banging</li> <li>• Hitting</li> </ul> <p><i>Topography:</i></p> <ul style="list-style-type: none"> <li>• Head</li> <li>• Trunk (stomach)</li> </ul>	<ul style="list-style-type: none"> <li>• Occasional tics</li> <li>• Stereotypies</li> <li>• Ataxia</li> </ul>	<p><i>Preventive:</i></p> <ul style="list-style-type: none"> <li>• Helmet</li> </ul> <p><i>No effect:</i></p> <ul style="list-style-type: none"> <li>• Haloperidol, risperidone, thioridazine, chlorpromazine, high-dose fluoxetine, lorazepam, buspirone, trazodone, multiple attempts for physical restraint</li> </ul> <p><i>Reduction:</i></p> <ul style="list-style-type: none"> <li>• Naltrexone, discontinuation of haloperidol, behavioural therapy</li> </ul>

History of hydrocephalus, bilateral thalamic venous angiomas and brain hemorrhage	[137]	n=1 Age: 16y Sex: male/female (1/0) Comorbidity: • DD	<i>Onset:</i> about 15y  <i>Forms:</i> • Biting (cheeks/buccal mucosa)	<ul style="list-style-type: none"> <li>• Spasticity</li> <li>• Dystonia</li> <li>• Involuntary movements</li> </ul>	<i>No effect:</i> <ul style="list-style-type: none"> <li>• Baclofen, olanzapine</li> <li>• Fluvoxamine, bupropion hydrochloride, acrylic occlusal splint, composite bulbs for prevention</li> </ul> <i>Cessation:</i> <ul style="list-style-type: none"> <li>• Modified hawley retainer with buccal shields</li> </ul>
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**Supplementary table 1. Comprehensive list of all included studies and SIB patients, including patient characteristics, details on SIB (onset, form, topography), associated movement disorder and reported treatment.**

ADHD, attention-deficit/hyperactivity disorder, AE, adverse event, ASD, autism spectrum disorder, BPD, borderline personality disorder, BTX, botulinum toxin, CARS, childhood autism rating scale, CNS, central nervous system, DBS, deep brain stimulation, DD, developmental disability, ECT, electroconvulsive therapy, GLHS, Gomez-Lopez-Hernandez syndrome, Gpi, internal globus pallidus, ICU, intensive care unit, ID, intellectual disability, i.m., intramuscular, ncl., nucleus, NBIA, neurodegeneration with brain iron accumulation, NMDA, N-methyl-D-aspartate, NOS, not otherwise specified, OCD, obsessive-compulsive disorder, PDD, pervasive developmental disorder, PD, parkinson's disease, PKU: phenylketonuria, *PS-1*, presenilin 1, PTPS, 6-pyruvoyl-tetrahydropterin synthase, SD, standard deviation, SIB, self-injurious behaviour, SSRI, selective serotonin reuptake inhibitor, TCA, tricyclic antidepressants, TS, Tourette syndrome

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