Catatonic Regression in ASD & other Neurodevelopmental Disorders

Autism Conference
Cork, Ireland
4-14-18

Judith H. Miles, M.D., Ph.D.
Division of Medical Genetics & Thompson Center for Autism & Neurodevelopmental Disorders
Department of Child Health

NO DISCLOSURES
Autism Spectrum Disorders

- Described & Named in 1940s
  - Kanner, 1943
  - Asperger, 1944

- Autism’s Black Past – Bruno Bettlheim → “Refrigerator Mother”
  - 1967 – The Empty Fortress – Autism caused by parent’s wish their child did not exist
  - Children should be taken away from their families
  - Claimed he could cure them at the Orthogenic School
  - Later proved to be a total fraud

- Rediscovery of Autism – Psychology
  - Lorna Wing, England – 1980s, 1990s

- Medicine & Genetics – 1990s
Autism = Behavioral Diagnosis

  - **Autism** = Persistent deficits: Social + Communication + Repetitive
  - Subtypes: Autistic Disorder, Asperger’s, Autism NOS
  - 70% with IQ < 70
  - Increase in Diagnoses (1/2500 to 1/68 kids)

- DSM-5 - 2013
  - **Autism Spectrum Disorders**
  - Social Communication + Restricted, repetitive behaviors…
  - Subtypes – replaced by modifiers for IQ, language, associated diagnoses, severity

**Autism is a Syndrome Diagnosis**

Hillman et al, 2000
300% increase in 5-9yo Missouri 1988 - 1995
Why are the children so different?

Variable expression? Heterogeneity?
ASD Phenotypic Subgroups

- Dysmorphology ~ 15% - 20%
- Microcephaly ~ 5%
- Brain malformations ~ 20%
- Macrocephaly ~ 20%
- Regressive Onset / Language Loss ~ 30%
- Family History of related disorders ~ 40%
  - ASD features, OCD, alcoholism
  - + others based on lab tests, Rx responses, etc
- Catatonia in 13 – 18% - not surprising

Search for Biomarkers
Autism Genetics

- Chromosomal & single gene disorders — Downs, Turners, Fragile X

- Known Autism Genes > **800 autism associated genes**
  - PTEN, Sotos, Neurofibromatosis, Fragile X (macrocephaly)
  - EN2, PTCHD1, HOXA1, GRIK2 (Neurodevelopment)
  - Joubert, Moebius, Chiari 1, EN2 (Cerebellar malformations)
  - Timothy, Cong. Stationary Night Blindness (Calcium channel)
  - Retts (methylation pathways)
  - NRXN1,3, GABRB3, MET, SHANK (connectivity)
  - SLC25A12 (mitochondrial)
  - SLC6A4 (neurotransmitters)

- Big Data — Genotype Content Mapping, 30,000 SSC patients & family
  - Compared groups defined by ASD symptoms
  - 286 genes associate with at least one subgroup
    - 1/3 are known “autism genes” vs. 2/3 are novel
    - **193 new ASD genes found!**

(Spencer et al, 2017)
Catatonia’s Evolution

- 1848 Karl Kahlbaum – described Catatonia (motor, speech & behavior)
- Psychiatry “Catatonia is a complication of Schizophrenia”
  - 1980 & 90s (DSM 3 & 4) – Catatonia = specifier for mental or medical dx
- Since ~ 2000 ➔ catatonia in neurologic & medical disorders
  - Psychiatric – especially affective disorders (bipolar) > Schizophrenia
  - Medication effects – (atypical antipsychotics, amoxicillin, azithromycin, etc)
  - Hydrocephalus, strokes, head trauma injury, seizures, SIADH, Tourettes
  - Infections – encephalitis, hepatitis encephalopathy, meningitis, neuro-syphilis
  - Endocrine disorders (hyper & hypothyroidism, diabetes)
  - Autoimmune diseases (Autoimmune encephalitis, Graves disease, Lupus, celiac)
  - Metabolic encephalopathy – Homocystinuria, carnitine disorder, Wilson’s Disease
  - Folate receptor Alpha Defect ➔ cerebral folate deficiency
  - Stress, bullying
Neurodevelopmental disorders?

- **1970 Rutter** – 12% adolescents with autism regress in language, inertia, intellectual decline

- **1998 Dhossche** – catatonia with auditory hallucinations, waxy flexibility, posturing, aggression, responded to neuroleptics & benzodiazepines

- **2000 & 2006 Wing & Shaw** –
  - Catatonia – like deterioration; 17% of autism (17-50 yo)
  - Obvious, marked deterioration – gradual
  - Catatonic stupor is rare
  - Waxing & waning of symptoms
  - Parents suggest precipitating stress (leaving school, job loss)
  - Suggests stress reduction therapy

- **Autism** – multiple reports but little consensus on etiology or treatment

- **Down syndrome** – 2013 & 2015

- **Other** - Kleefstra syndrome (9p-), Fragile X, cerebellar dysgenesis, congenital hydrocephalus, Prader Willi syndrome
Javon’s Story

ASD dx – 3 yr 4mo, IN; regressed at 18mo
Described as happy, interactive, loved cars

Two year hx of regression
Respiratory infection in Jan, Feb, Mar, 2013
Regression in motor, speech, behavior
- stopped going to store, using iPad, trampoline, swimming,
- motor slowing, freezing, aggression & skin picking,
pulls pubic hair, weight loss, neck tics, sleep, constipation,
sits, stares, head down, hand over eyes, grimacing, no eye contact
- removed from school

Dx with depression

Lisa Bailey (BCFS) – Catatonia dx Sept 2014
Javon – 1st Clinic Visit

→ 2 mg Lorazepam
Javon on Lorazepam

- Walks quickly to the bus → now grabs book-bag & runs to bus
- Back in School
- Walks up stairs without stopping
- Goes to store with Dad, puts stuff in cart
- Plays with his cars
- Stopped skin picking until family fights
- Less impulsive
- Plays with neighborhood boys
Success due to:

1. Catatonia dx & appropriate treatment
2. Apraxia dx & communication device
3. Trichotillomania dx & NAC rx
4. Transfer to appropriate school
1st – Clear & obvious regression

- **Motor activity** - slowing, getting stuck, hyper outbursts
- **Speech** - decreased, mute, slow, whispering
- **Withdrawal** - ↓engagement (people/environment) ↓noticing
- **Mood** - flat, ↓enjoyment, depression, aggression
- **Negativism** – refusing to participate, follow instructions
- **Stereotypic movements** - tics, posturing, grimace
- **Abilities** - ↓skills, self care/daily living skills, bathroom
- **Eating, sleeping** – slow, refusal

**Bush-Francis Catatonia Rating Scale** *(handout)*
Screening Score = # of items 1-14 that are present.
Diagnosis = 2 or more items
• **Onset in adolescents & young adults** (may be earlier)
  - Often gradual, sometimes sudden
  - Always emergence of new symptoms

• **Additional symptoms noted**
  - Sleep disturbance
  - Incontinence & loss of independent toileting
  - Scared
  - Need prompting

• **ASD & Catatonia symptoms overlap**
  - Mutism, echolalia, repetitive behaviors, withdrawal,
  - Misdiagnoses: Depression, OCD, puberty, psychosis etc.
Bush-Francis Catatonia Rating Scale

Screening Score (Presence or absence of items/symptoms 1 - 14) ______
Severity Score (Number of points for items/symptoms 1 - 23) ______

1. **Immobility/stupor**: Extreme hypoactivity, immobile, minimally responsive to stimuli.

2. **Mutism**: Verbally unresponsive or minimally responsive.

3. **Staring**: Fixed gaze, little or no visual scanning of environment, decreased blinking.

4. **Posturing/catalepsy**: Spontaneous maintenance of postures, (sitting, standing for long periods)

5. **Grimacing**: Maintenance of odd facial expressions.

6. **Echopraxia/echolalia**: Mimicking examiner's movements (echopraxia) or speech (echolalia).

7. **Stereotypy**: Repetitive, non-goal-directed motor activity (e.g. finger-play, touching, patting)

8. **Mannerisms**: Odd, purposeful movements (hopping or walking tiptoe, saluting passers-by)

9. **Stereotyped & meaningless repetition of words & phrases**: Repetition of phrases/sentences

10. **Rigidity**: Maintenance of a rigid position despite efforts to be moved

11. **Negativism**: Apparently motiveless resistance to instructions or attempts to move/examine patients. Contrary behavior, does exact opposite of instruction.

12. **Waxy flexibility**: During repositioning of patient, patient offers initial resistance before allowing him/herself to be repositioned

13. **Withdrawal**: Refusal to eat, drink and/or make eye contact.

14. **Excitement**: Extreme hyperactivity, constant motor unrest which is apparently non-purposeful.

Dx – 2 or more symptoms
15. **Impulsivity:** Patient suddenly engages in inappropriate behavior (e.g. runs down hallway, starts screaming or takes off clothes) without provocation.

16. **Automatic obedience:** Exaggerated cooperation with examiner's request or spontaneous continuation of movement requested.

17. **Passive Obedience:** Patient raises arm in response to light pressure of finger, despite instructions to the contrary.

18. **Muscle Resistance:** Involuntary resistance to passive movement of a limb to a new position.

19. **Motorically Stuck:** Patient appears stuck in indecisive, hesitant motor movements.

20. **Grasp reflex:** Striking the patient’s open palm with two extended fingers of the examiner’s hand results in automatic closure of patient’s hand.

21. **Perseveration:** Repeatedly returns to same topic or persists with the same movements.

22. **Combativeness:** Belligerence or aggression, Usually undirected, without explanation.

23. **Autonomic abnormality:** Abnormality of body temperature (fever), blood pressure, pulse, respiratory rate, inappropriate sweating, flushing.
Andy’s Story

till 2013…
Andy trying to close a door

note:
Motor slowing
Freezing
Repetitive attempts
Withdrawn facial expression
Andy trying to eat

note:
Arm & shoulder movements
Motor slowing
Freezing
Repetitive attempts
Facial grimaces, shoulder shrugs, & body tics
note:
Speed
Looking
Noticing
Smiling
Andy following lorazepam & ECT - 6-20-15

note:

Fast
Interactive
Smiling
**Clinical Protocol**

1\(^{st}\) Diagnosis

- **History** – change from baseline, timeline
- Bush Francis Catatonia Rating Scale

2\(^{nd}\) Assess causes of motor & cognitive regression, known causes of catatonia & autoimmune dysfunction

- **Physical exam** - observation, neurologic
- Lorazepam 2mg IV test dose (E’s mother’s report)

- **Neurologic** – MRI, EEG, LP
- **Immune dysfunction**:
  - ASO (Streptolysin O Ab), DNase B Ab, Thyroglobulin Ab, Thyroid Peroxidase Ab, FANA, Lupus Anti StaClot, Celiac serology, IgG NMDAR Ab, GAD, Cunningham Immune Panel (Moleculara lab)

- **Brain Metabolism**:
  - Dopamine metabolism disorder (low HVA & 5HIAA), CSF Neopterin, Cerebral folate deficiency (www.mnnglab.com)

- **Intermediary metabolism**:
  - Homocystinuria, carnitine disorders, Wilson’s disease, vitamin B12, folate
<table>
<thead>
<tr>
<th>Medical conditions associated with development of catatonia</th>
<th>Medical conditions that may have presentations similar to catatonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infections</td>
<td>Arteriovenous malformations</td>
</tr>
<tr>
<td>Illicit drug use</td>
<td>Cerebrovascular accident</td>
</tr>
<tr>
<td>Cerebrovascular dx</td>
<td>Encephalitis</td>
</tr>
<tr>
<td>Electrolyte imbalance</td>
<td>Fibromuscular dysplasia</td>
</tr>
<tr>
<td>Vitamin B12 def.</td>
<td>Huntington’s disease</td>
</tr>
<tr>
<td>Seizures</td>
<td>Meningitis</td>
</tr>
<tr>
<td>Hepatic transplant</td>
<td>Neurosyphilis</td>
</tr>
<tr>
<td>Thyroid disease</td>
<td>Parkinson’s disease</td>
</tr>
<tr>
<td>Diabetic - ketoacidosis</td>
<td>Progressive multifocal Leukoencephalopathy (PML)</td>
</tr>
<tr>
<td>Lupus</td>
<td>Seizure disorder</td>
</tr>
<tr>
<td>Sheehan syndrome</td>
<td>Central pontine myelinolysis</td>
</tr>
<tr>
<td>Encephalitis/opathy due to:</td>
<td>Progressive - supranuclear palsy</td>
</tr>
<tr>
<td>Herpes simplex</td>
<td>Strychnine poisoning</td>
</tr>
<tr>
<td>Liver disease</td>
<td>Cortical basal – ganglionic degenerate.</td>
</tr>
<tr>
<td>Drug withdrawal</td>
<td>Cortical basal – ganglionic degenerate.</td>
</tr>
<tr>
<td>Lethargica</td>
<td></td>
</tr>
<tr>
<td>NMDA receptor</td>
<td></td>
</tr>
<tr>
<td>Poor nutrition</td>
<td></td>
</tr>
<tr>
<td>Homocystinuria</td>
<td></td>
</tr>
<tr>
<td>Renal transplant</td>
<td></td>
</tr>
<tr>
<td>Wilson’s disease</td>
<td></td>
</tr>
<tr>
<td>Head trauma</td>
<td></td>
</tr>
<tr>
<td>Metabolic abn</td>
<td></td>
</tr>
<tr>
<td>Severe weight loss</td>
<td></td>
</tr>
<tr>
<td>Porphyria</td>
<td></td>
</tr>
<tr>
<td>Iatrogenic illness</td>
<td></td>
</tr>
<tr>
<td>Med side effects</td>
<td></td>
</tr>
<tr>
<td>Cerebral folate def.</td>
<td></td>
</tr>
<tr>
<td>Post Partum</td>
<td></td>
</tr>
<tr>
<td>SIADH</td>
<td></td>
</tr>
<tr>
<td>Lesions of the CNS</td>
<td></td>
</tr>
<tr>
<td>Fabry disease</td>
<td></td>
</tr>
<tr>
<td>Critical Illness</td>
<td></td>
</tr>
<tr>
<td>Arteriovenous malformations</td>
<td></td>
</tr>
<tr>
<td>Cerebrovascular accident</td>
<td></td>
</tr>
<tr>
<td>Encephalitis</td>
<td></td>
</tr>
<tr>
<td>Fibromuscular dysplasia</td>
<td></td>
</tr>
<tr>
<td>Huntington’s disease</td>
<td></td>
</tr>
<tr>
<td>Meningitis</td>
<td></td>
</tr>
<tr>
<td>Neurosyphilis</td>
<td></td>
</tr>
<tr>
<td>Parkinson’s disease</td>
<td></td>
</tr>
<tr>
<td>Progressive multifocal Leukoencephalopathy (PML)</td>
<td></td>
</tr>
<tr>
<td>Seizure disorder</td>
<td></td>
</tr>
<tr>
<td>Central pontine myelinolysis</td>
<td></td>
</tr>
<tr>
<td>Progressive - supranuclear palsy</td>
<td></td>
</tr>
<tr>
<td>Strychnine poisoning</td>
<td></td>
</tr>
<tr>
<td>Hallervorden-Spatz</td>
<td></td>
</tr>
<tr>
<td>Lewy body dementia</td>
<td></td>
</tr>
<tr>
<td>Neurosarcoidosis</td>
<td></td>
</tr>
<tr>
<td>Other white matter dx</td>
<td></td>
</tr>
<tr>
<td>Parkinsonism</td>
<td></td>
</tr>
<tr>
<td>Progressive - supranuclear palsy</td>
<td></td>
</tr>
<tr>
<td>Strychnine poisoning</td>
<td></td>
</tr>
<tr>
<td>Cortical basal – ganglionic degenerate.</td>
<td></td>
</tr>
<tr>
<td>Cortical basal – ganglionic degenerate.</td>
<td></td>
</tr>
</tbody>
</table>
1. **GABA decreased** → **(hypoactivity)**
   - GABA/Glutamate neurotransmitter system disruption
     - GABA – inhibitory neural transmission
     - Glutamate – excitatory neural transmission
   - Goal = increase GABA
     - Benzodiazepines, ECT, Barbiturates (GABA agonists)

2. **Glutamate increased** → **(hyperactivity)**
   - NMDA receptor dysfunction
   - Goal = decrease glutamate
     - Memantine, Amantadine, & Nuedexta (NMDAR antagonists)

3. **Dopamine (D2) hypoactivity**
   - Amantadine – facilitates central dopamine release & delays uptake
First Line Treatments
GABA agonists

- **High dose benzodiazepines** – 1\textsuperscript{st} line therapy
  - Lorazepam – start at 2 mg/day PO, may go up to 25 mg/day (slowly)
  - Side effects – sleepiness, dizziness
  - Risks - withdrawal seizures, overdose

- **Modified ECT** – 2\textsuperscript{nd} line therapy → 80\% - 100\% effective
  - Ambulatory surgery suite (anesthesiologist/psychiatrist/nurse)
  - Sedation – brief with etomidate, methohexital, propofol
  - Muscle blockade – succinylcholine
  - Oxygenation
  - MECTA 5000Q - Brief-pulse (4 sec) bitemporal/bifrontal electrode
  - **Resistance: lack of knowledge, media, legal restrictions**
Electroconvulsive shock – ECT
for Catatonia in adolescents – 2nd line treatment

• Adults: > 75 years; standard of care for catatonia
  • Refractory depression, bipolar, mania, psychosis, neuroleptic malignant syndrome
  • Efficacy - 80-100% for catatonia
  • Safety – no structural, histopathologic or cognitive damage after ECT with prolonged maintenance
    • 4 deaths/100,000 treatments – mainly due to cardiac disease in the elderly

• Children: should be safer than adults
  • 3 controlled studies, 1 analysis of 59 adolescents
  • Am Acad Child Adol Psychiatry best practice parameters (2004) - similar to adults
  • No deaths reported in adolescents or children
  • Risks similar to short term anesthesia

• Resistance: lack of knowledge, media portrayal, legal
  • Side effects: transient memory loss, prolonged seizure, headache, nausea, muscle aches
  • Lack of long term studies:
  • Laws vary by State – California & Texas are most restrictive if < 18.
    • Missouri – court approval for incompetent individuals
Electroconvulsive therapy, procedure--prohibitions--attorney's fees.

630.130. 1. Every patient, whether voluntary or involuntary, in a public or private mental health facility shall have the right to refuse electroconvulsive therapy.

2. Before electroconvulsive therapy may be administered voluntarily to a patient, the patient shall be informed, both orally and in writing, of the risks of the therapy and shall give his express written voluntary consent to receiving the therapy.

3. Involuntary electroconvulsive therapy may be administered under a court order after a full evidentiary hearing where the patient refusing such treatment is represented by counsel who shall advocate his or her position. The therapy may be administered on an involuntary basis only if it is shown, by clear and convincing evidence, that the therapy is necessary under the following criteria:

   (1) There is a strong likelihood that the therapy will significantly improve or cure the patient's mental disorder for a substantial period of time without causing him any serious functional harm; and

   (2) There is no less drastic alternative form of therapy which could lead to substantial improvement in the patient's condition. At the conclusion of such hearing, if the petitioner has sustained his burden of proof, the court may order up to a specified number of involuntary electroconvulsive therapy treatments to be performed over a specified period of time.

4. Parents of minor patients or legal guardians of incompetent patients shall be required to obtain court orders authorizing electroconvulsive therapy under the procedures specified in subsection 3 of this section.

5. Persons who are diagnosed solely as mentally retarded shall not be subject to electroconvulsive therapy.

6. If the judge finds that the respondent is unable to pay attorney's fees for the services rendered in the proceedings the judge shall allow a reasonable attorney's fee for the services, which fee shall be assessed as costs and paid together with all the costs in the proceeding by the state, in accordance with rules and regulations promulgated by the state court administrator, from funds appropriated to the office of administration for such purposes provided that no attorney's fees shall be allowed for services rendered by any attorney who is a salaried employee of a public agency or a private agency which receives public funds.


(2006) Section allows a healthcare provider who has a duty of care to act in the patient's best interest, even if the patient has refused treatment, if the patient is in need of the therapy. In re Dunn, 181 S.W.3d 601 (Mo.App.E.D.).
Other Treatments

- **Glutamate antagonists**: ↓ glutamate
  - Namenda (*Menantine*)
  - N-acetylcysteine (*NAC*)
  - Topiramate/Topamax
  - Ketamine
  - Nuedexta 20/10 (*Dextromethorphan + quinidine*)

- **Anti-inflammatory**: Minocycline – Cerebral inflammation
  - Autoimmune dysfunction → inflammation
  - May also be a NMDA receptor antagonist

- **Behavioral therapy**
  - Mitigate stressful exposures
  - Provide enjoyed activities

- **Good medical care**
  - Stop most other meds
  - Sleep hygiene
  - Dietary
EVALUATION, DIAGNOSIS AND TREATMENT OF CATATONIA IN CHILDREN AND ADOLESCENTS

Possible catatonia

Medical work-up + urine toxicology

Apply catatonia criteria

Assess severity of catatonia (eg, using rating scales)

Search and eliminate culprit substances or medications

Catatonia

LZP challenge test

Improved

3-day LZP Trial

Much improved

LZP continuation

When relapsing, LZP maintenance

Somewhat improved

LZP + Bilateral ECT

When relapsing, maintenance LZP + ECT

Not improved

Bilateral ECT

When relapsing, maintenance ECT

LZP=lorazepam; ECT=electroconvulsive therapy.

Treatment Monitoring Essential

- Principles
  - Team work
  - Ease of access
  - Longitudinal monitoring
    - Catatonia Impact Scale (CIS)
### Amount of Movement

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>During the past 1 week, how often he/she had?</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>1. Is immobile</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Holds a stiff posture</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Slow movements and daily activities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Low activity level</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Gets stuck in movements -</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Kinds of Movements

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>6. Makes odd facial expressions</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Makes repetitive, stereotypic movements</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Has involuntary movements</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Has odd mannerisms -</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10. Has episodes of extreme hyperactivity with constant motion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11. Resists being moved</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12. Will move an arm with minimal pressure or suggestion</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Talking

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>13. Doesn’t talk – quiet</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14. Doesn’t initiate conversations</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15. Mimics other's speech</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16. Repeats words or phrases that don’t mean anything or are not in context</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17. Perseverates</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Withdrawal

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>18. Withdrawn from people around her</td>
<td></td>
<td></td>
</tr>
<tr>
<td>19. Stares into space</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. Doesn’t respond to requests</td>
<td></td>
<td></td>
</tr>
<tr>
<td>21. Doesn’t eat all her food &amp;/or isn’t drinking enough water</td>
<td></td>
<td></td>
</tr>
<tr>
<td>22. Doesn’t focus and engage in school, work or recreational activities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>23. Doesn’t seem to notice the things around him/her</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Total Scores**

<table>
<thead>
<tr>
<th>Amount of Movement</th>
<th>Score</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>58</td>
<td>52</td>
</tr>
</tbody>
</table>

**Kinds of Movements**

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>13. Doesn’t talk – quiet</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14. Doesn’t initiate conversations</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15. Mimics other's speech</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16. Repeats words or phrases that don’t mean anything or are not in context</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17. Perseverates</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Talking**

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>13. Doesn’t talk – quiet</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14. Doesn’t initiate conversations</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15. Mimics other's speech</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16. Repeats words or phrases that don’t mean anything or are not in context</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17. Perseverates</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Withdrawal**

<table>
<thead>
<tr>
<th>Movement Description</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>18. Withdrawn from people around her</td>
<td></td>
<td></td>
</tr>
<tr>
<td>19. Stares into space</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. Doesn’t respond to requests</td>
<td></td>
<td></td>
</tr>
<tr>
<td>21. Doesn’t eat all her food &amp;/or isn’t drinking enough water</td>
<td></td>
<td></td>
</tr>
<tr>
<td>22. Doesn’t focus and engage in school, work or recreational activities</td>
<td></td>
<td></td>
</tr>
<tr>
<td>23. Doesn’t seem to notice the things around him/her</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Withdrawal

<table>
<thead>
<tr>
<th>During the past 1 week, how often he/she had?</th>
<th>Frequency</th>
<th>Impact interfere with everyday functioning?</th>
</tr>
</thead>
<tbody>
<tr>
<td>18. Withdrawn from people around her - doesn’t make eye contact or seem to remember people</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>19. Stares into space – fixed gaze, without scanning the environment, decreased blinking, trance-like</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>20. Doesn’t respond to requests - getting up, taking medicine, getting dressed, needs prompting</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>21. Doesn’t eat all her food &amp;/or isn’t drinking enough water</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>22. Doesn’t focus and engage in school, work or recreational activities</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>23. Doesn’t seem to notice the things around him/her</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
</tbody>
</table>

**Withdrawal subscale scores**

<table>
<thead>
<tr>
<th>Behaviors</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>24. Impulsive, engages in sudden inappropriate behaviors.</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>25. Seems fearful or apprehensive or anxious</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>26. Becomes belligerent or aggressive, strikes out</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>27. Resists suggestions or instructions. Refuses to do activities he/she previously enjoyed</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
</tbody>
</table>

**Behaviors subscale score**

<table>
<thead>
<tr>
<th>Activities of Daily Living</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>28. Is more dependent on others for bathing, eating, picking out clothes,</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>29. Refuses to do daily tasks that previously were enjoyed - folding clothes, cleaning the room, going to a job or school</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>30. Toileting – doesn’t go easily and quickly</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
</tbody>
</table>

**Activities of daily living subscale scores**

<table>
<thead>
<tr>
<th>Medical</th>
<th>Frequency</th>
<th>Impact</th>
</tr>
</thead>
<tbody>
<tr>
<td>31. Has episodes of high fevers, inappropriate sweating, facial flushing, fast heart rate or high blood pressure</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
<tr>
<td>32. Change in amount or continuity of sleep - sleeps more or less</td>
<td>0 1 2 3 4</td>
<td>0 1 2 3 4</td>
</tr>
</tbody>
</table>

**Medical subscale scores**

### Total Scores

**scoring directions**

0 = never
Catatonia Impact Scale

Andy's Catatonia Scores - June 2013 - June 2015

https://showmeportal.missouri.edu/redcap/surveys/?s=PNTUbpiHI8
Catatonia Impact Scale

July 2016 – July 2017
Catatonia Impact Scale

5/20/14 to 8/1/17

Screening

- Lorazepam (mg)
- Nac (100mg)
- Nuedexta (mg)
- ECT (date)

Lorazepam

Nuedexta

ECT
Complications of Catatonia
Physical & Medical

- **Malignant Catatonia** –
  - Severe autonomic nervous system impairment
  - Associated with neuroleptic medication
  - Fever, hypertension, incontinence
  - Neuroleptic Malignant syndrome & Serotonin syndrome are probably medication induced Malignant Catatonia

- **Malnutrition, starvation, dehydration**

- **Disorders of immobility**
  - Venous thrombosis
  - Bed sores

- **Unable to live at home**
  - Nursing home or residential care facility
Problems we need to Solve

• Symptoms may look a little different in patients with developmental disorders – “It’s not your grandfather’s catatonia”

• Treatment responses - may take longer than in some psychiatric disorders

• Catatonia in Neurodevelopmental disorders more apt to be chronic

• ECT scares people - parents, pediatricians, psychiatrists, courts - Going to court is an impediment (expensive, time consuming)

• Parental caregiving is 24/7 - help for families

• Increase awareness & treatment facilities

• Awareness, Advocacy, Education, Research
Conclusions
Treatable, Diagnosable & Common

- **Severe neuropsychiatric disorder** → inability to function at home, school, work, & life threatening

- **Pathophysiologically similar to Catatonia** in schizophrenia, depression, lupus, encephalitis, developmental & neurologic disorders
  - All diagnosed by Bush-Francis Scale
  - All respond to the same basic treatments

- ? **ASD or DS specific mechanisms or triggers**
  - Ex. Immune dysfunction

- **Common cause of deterioration** in autism teens
  - Most diagnoses are missed

- **Urgently need research, advocacy and awareness**
  - prevalence, symptom profile, basic neuroscience, program development

**ASD Catatonia ≡ Catatonia in psychiatric dx**
Thank you to the young people & their families who teach us & help every step of the way.

Catatonia Team

Medical Genetics/Autism - Judith Miles
Psychiatry - Muaid Ithman, John Hall
Neurology/Autism - David Beversdorf
Psychology - Kerri Nowell
Research Core - Nicole Takahashi & Julie Muckerman