Role of Immunotherapy in Down Syndrome Disintegrative Disorder (DSDDD)
Nidhiben Anadani, Deepti Chrusciel

Objective
To describe case series of patients with DSDDD, successfully treated with immunotherapy including Intravenous Immunoglobulin (IVIG) at a single academic center.

Background
Down syndrome is the most common chromosomal disorder, and in most cases, is due to trisomy of chromosome 21. DSDDD is under-recognized, rapidly progressive neuropsychiatric syndrome with various postulated etiology including psychological stress, primary psychiatric disorder and autoimmune.

Design/Methods
Case-1: A 20-year-old fun loving female with trisomy-21 and infantile spasms started having complex partial seizures, hallucinations, speech regression, tics, abnormal head movement and obsessive-compulsive behavior. Case-2: A 20-year-old female cheerleader with trisomy-21, started having rapid regression in language, cognition, social skills and agitation over one year. Case-3: A 22-year-old female dancer with trisomy-21, started having subacute onset depression, hallucinations, sleep changes, anorexia and speech regression over one year.

Results
Case-1: MRI brain and cerebrospinal fluid (CSF) studies were normal including negative autoimmune encephalitis panel. Serum thyroglobulin and thyroid peroxidase antibody were high. Prolonged oral steroid therapy helped but caused adverse effects. She was able to return to her premorbid baseline with chronic IVIG therapy every 10 weeks. Case-2: A 20-year-old female cheerleader with trisomy-21, started having rapid regression in language, cognition, social skills and agitation over one year. Case-3: A 22-year-old female dancer with trisomy-21, started having subacute onset depression, hallucinations, sleep changes, anorexia and speech regression over one year.

Conclusions
DSDDD should be considered in patients with down syndrome with rapid regression. It is often associated with positive thyroid peroxidase antibody suggesting immune mediated etiology. Various immunotherapy treatments have been reported in literature including steroid, IVIG, mycophenolate and rituximab with significant improvement in selected patient with autoimmunity.

Disclosure: Dr. Anadani has nothing to disclose. Dr. Chrusciel has nothing to disclose.

EEG Characteristics in Hospitalized Patients With Acute COVID-19 Symptoms
Ganesh Murthy, Daniel Fayard, Ryan Chung, Steve Chung

Objective
Our objective was to evaluate the incidence of seizures, pattern of EEG abnormalities, and localization of abnormal discharges in hospitalized patients with COVID-19.

Background
The COVID-19 epidemic has revealed significant neurological manifestations including de novo seizures in patients who do not have a prior history of epilepsy or clear epilepsy risk factors. Our center is located in Arizona, which in the early part of January 2021 had more cases per capita than any other place in the world.

Design/Methods
We performed a retrospective review to observe the electroencephalogram (EEG) patterns of hospitalized adult patients with COVID-19 between March 2020 and February 2021.

Results
We identified 99 patients who were COVID-19 positive and had EEG testing during the same hospitalization. The most common EEG abnormality was diffuse background slowing, which was seen in 63.6% of patients (n = 63/99), compare to 15.1% of focal background slowing. Epileptiform discharges were seen in 11.1% of patients and seizures were found in 5.1% of patients, as newly diagnosed seizures. When combining all focal abnormalities, the most common location for these abnormalities was in the frontal regions 36.4% (n = 8/22). Even though 21 patients had acute focal neuroradiologic findings, only 5 had correlated EEG abnormalities within the same region. When EEG was obtained with suspected seizures (n = 33), 4 cases (12.1%, n = 4/33) indeed showed ictal pattern compared to 1.6% when seizures was not suspected (p = 0.087).

Conclusions
Abnormal EEG findings are most commonly found in the frontal lobe among hospitalized patients with acute COVID-19 symptoms. De novo seizures may be seen with COVID-19 infection. Suspicion of seizures should be raised in patients with COVID-19 encephalopathy. The utility of an EEG may help allow us better insight into how and where the COVID infection affects our central nervous system.

Disclosure: Dr. Murthy has nothing to disclose. Dr. Fayard has nothing to disclose. Dr. Chung has nothing to disclose. The institution of Dr. Chung has received personal compensation in the range of $10,000-$49,999 for serving as a Consultant for ucb pharma. Dr. Chung has received personal compensation in the range of $500-$4,999 for serving as a Consultant for SK Life sciences. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for eisai. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for ucb pharma. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for eisai. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for eisai. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for ucb pharma. Dr. Chung has received personal compensation in the range of $50,000-$99,999 for serving as a Consultant for ucb pharma.

Progressive Multifocal Leukoencephalopathy Associated With Sarcoidosis: A Multi-Center Case Series
Caleb R.S. McEntire, MD, Anita Fletcher, MD, Michel Toledano, MD, Samantha Epstein, MD, Sabrina Tan, MD, Yang Mao-Draayer, MD, PhD, Samantha Banks, MD, Allen Aksamit, MD, Jeffrey M. Gelfand, MD, MAS, Kiran Thakur, MD, Irene Cortese, MD, Shamik Bhattacharyya, MD
Role of Immunotherapy in Down Syndrome Disintegrative Disorder (DSDD)
Nidhiben Anadani and Deepti Chruscieł

Neurology 2022;99;S69
DOI 10.1212/01.wnl.0000903552.74099.b4

This information is current as of December 5, 2022

Updated Information & Services
including high resolution figures, can be found at:
http://n.neurology.org/content/99/23_Supplement_2/S69.1.full

Subspecialty Collections
This article, along with others on similar topics, appears in the following collection(s):
Cerebrospinal Fluid
http://n.neurology.org/cgi/collection/cerebrospinal_fluid
CT
http://n.neurology.org/cgi/collection/ct
Low pressure syndrome
http://n.neurology.org/cgi/collection/low_pressure_syndrome

Permissions & Licensing
Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints
Information about ordering reprints can be found online:
http://n.neurology.org/subscribers/advertise