Psychosomatics 2017:58:209–212

Case Reports

Refractory Self-Injurious Behavior in Severe Intellectual Disability Responsive to Topiramate: A Case Report

Brandon Hamm, M.D., M.S., Naveed Khokhar, M.D., Xavier F. Jimenez, M.D., M.A.

Introduction

Self-injurious behavior (SIB) is dangerous and difficult-to-manage; however, it is seen in various psychiatric illnesses and is defined as "a class of behaviors, often highly repetitive and rhythmic, that result in physical harm to the individual …[and] the deliberate alteration or destruction of body tissue without conscious suicidal intent.[1]" Such behavior can result in blindness, subdural hematoma, infection/sepsis, or even death.[2] SIB is seen in patients with personality disorders, intellectual disability (ID), autism-spectrum disorders, Lesch-Nyhan syndrome, Prader-Willi syndrome, Cri du Chat syndrome, and Cornella de Lange syndrome among other disorders.[2–5]

Behavioral techniques and psychopharmacological agents are interventions used to decrease SIB, though evidence of efficacy is largely based on isolated case reports and limited randomized controlled trials.[6,7] This article describes a case of refractory SIB in a patient with severe ID that ultimately responded to topiramate augmentation of a complex pharmacological regimen. Though a single case series of patients with Prader-Willi syndrome reports success of topiramate in decreasing skin picking,[8] our case report is the first to describe improvement of protracted and high-intensity SIB (head-banging, self-punching, and finger-biting) in a patient with severe ID after administration of topiramate.

Clinical Presentation

Mr. A, a 27-year-old man had rare chromosomal abnormalities (4.2 mb terminal deletion on chromosome 4 and 3.9 mb duplication on chromosome 19p13.3) complicated by primordial dwarfism (25.4 kg, 137.2 cm, body mass index = 13.5), severe ID (very limited vocabulary with 1–2 word communications at baseline, with an inability to be taught reading or writing), and recurrent aspiration pneumonias, was admitted for an iliac fracture and aspiration pneumonia (in the setting of having a chronic percutaneous endoscopic gastronomy [PEG] tube) after a fall at home. He immediately developed agitation and SIB (beating his face and body with his hands and hitting his head against a wall).

At baseline, Mr. A was verbally limited, but able to participate in a daytime workshop. At home, he exhibited some infrequent SIB (prototypical behaviors include biting and punching himself), predominantly when upset or in pain, but intermittently at his workshop as well. Management of SIB before admission consisted of use of risperidone 2 mg by PEG 3 times a day, fluoxetine 60 mg by PEG daily, gabapentin 500 mg by PEG 4 times a day, buspirone 20 mg by PEG at 10 AM and 30 mg by PEG 3 times a day, and lorazepam (2 mg/mL solution) 1 mL every 6 hours by PEG as needed for agitation. Per the patient's outpatient psychiatrist, none of these medications was prescribed for an anxiety or mood disorder. His care at

Received October 4, 2016; revised November 1, 2016; accepted November 1, 2016. From the Department of Psychiatry (BH, NK, XJ), A. Cleveland Clinic, Cleveland, OH. Send correspondence and reprint requests to Brandon Hamm, M.D., M.S., Department of Psychiatry, A. Cleveland Clinic, 9500 Euclid Ave, P57 Desk, Cleveland, OH; e-mail: hammb@ccf.org

baseline did not include routine mechanical restraints devices, such as a helmet or splints.

On admission, his vital signs were remarkable for an oxygen saturation of 91%. Complete blood count and basic metabolic panel demonstrated mild leukocytosis (13.0 k/μL) and a blood urea nitrogen of 30 mg/dL. Chest X-ray and computed tomography of the head without contrast were unremarkable. Computed tomography of the pelvis demonstrated a left iliac crest fracture. During his hospitalization, medical management included opioid analgesia (oxycodone liquid 2.5 mg by PEG every 6 hours *pro re nata* and occasional 1-time doses of fentanyl 25 mcg IV) for his iliac fracture, as well as acyclovir for the herpetic dermatitis and keratosis. Aspiration pneumonia was treated with vancomycin and piperacillin/tazobactam. An open procedure for percutaneous endoscopic jejunostomy (PEJ) tube placement was performed to prevent future aspiration, which had continued, despite 2 Nissen fundoplications and PEG placement. On presentation to the hospital, SIB was severe, requiring frequent mechanical restraints and complex pharmacological strategies to address agitation.

SIB on presentation to the hospital was worse in frequency and intensity than at baseline, and it included near-constant head-banging on nearby structures, biting fingers (resulting in open wounds and cellulitis), punching his own face (causing extensive bruising), and unrestrained kicking of hard objects. Mr. A was often restrainted for most waking hours. Opioid analgesia was increased (up to a total of oxycodone 27 mg liquid by PEJ on 1 day) without alleviation of agitation, and naloxone was required on 3 separate days (total doses were 1, 2.3, and 1.4 mg) for unresponsiveness with desaturation. Alternative neuroleptics to risperidone were trialed, but haloperidol (received up to 30 mg IV in 1 day) administration was complicated by a prolonged QTc with bradycardia, chlorpromazine (up to 350 mg total IV in 1 day) was limited by hypotension and acute transaminitis, and olanzapine (up to 40 mg by PEJ in 1 day) had little benefit with increased blunt sedation. Lorazepam 4 mg IV every 4 hours as needed was often used for severe agitation, though we initially avoided this owing to concern for disinhibition in the setting of possible hyperactive delirium. Other psychotropic trials included buspirone 20 mg by PEJ 4 times a day (negligible benefit), gabapentin 800 mg by PEJ 3 times a day (negligible benefit and increased sedation), clonidine 0.1 mg by PEJ 3 times a day (negligible benefit and possibly contributed to mild hypotension), and valproic acid (250 mg/5 mL solution) 5 mL by PEJ 3 times a day (with transaminitis and hyperammonemia, negligible benefit). Mr. A was briefly transferred out of the intensive care unit; however, excessive agitation prompted his return to the intensive care unit. Behavioral interventions included frequent distraction with pictures of enjoyed things/people and being wheeled around the unit by nursing staff. Given continued agitation, topiramate 50 mg by PEJ twice a day was initiated, resulting in a rapid and remarkable decrease in the severity and frequency of SIB. Mr. A's affect and vocalizations became calmer and friendlier for the 7 days of topiramate administration before discharge, without any adverse effects. Owing to the desire to continue his improved safety, an on-off-on trial to confirm topiramate's effect in SIB control was not attempted. At the time of discharge, Mr. A's regimen included topiramate 50 mg by PEJ twice daily, buspirone 20 mg by PEJ 4 times daily, fluoxetine 60 mg by PEJ daily, gabapentin 800 mg by PEJ 3 times a day, and clonidine 0.1 mg by PEJ 3 times a day.

Discussion

Complex neurocircuitry involving frontal and limbic cortices (as well as their connections to the basal ganglia) are thought to be involved in aggressive and perseverative behaviors.[9,10] Excitatory glutamatergic activity in cortico-striato-thalamocortical neurons may be involved, as cortico-striato-thalamocortical circuitry is implicated in obsessive-compulsive disorder psychopathology.[9] Such complex circuitry suggests the role for psychopharmacological agents with several mechanisms of action. Pertinent to this case, individuals with ID often have decreased global white matter and heterogeneous regional anomalies in neuroanatomy,[11] impairing neural connectivity. Topiramate inhibits voltage-gated sodium channels, high voltage–activated calcium channels, and α-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid (AMPA) receptors, but agonizes gamma-aminobutyric acid (GABA) isoreceptor A.[12] Inhibitory effects on aforementioned cortico-striato-thalamocortical neurons could decrease compulsive and repetitive behaviors. A small double-blinded placebo-controlled trial of