# Acute presentation of an acquired neurosensory syndrome

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#### Disclosures

- There are no conflicts of interest or financial interests to report
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## Overview: "This is the State Department – we have a problem"

- Individuals began experiencing symptoms late 2016
  - Ear pain
  - Tinnitus
  - Dizziness
  - Cognitive Issues
- Profile
  - All experienced a loud noise or pressure phenomenon before and during the symptoms
  - All were members or family members of individuals stationed with the US Diplomatic Mission in Havana, Cuba
  - The sound was localized and "followed" the individual and would shut off if the door to the outside was opened



#### Miami Experience

- 35 individuals who were symptomatic or at risk evaluated in Miami
  - 25 who had exposure and symptoms
  - 10 who had no exposure, no symptoms but where co-inhabitants of symptomatic individuals and in the dwelling at the same time that the exposure occurred
- 105 unaffected embassy members evaluated in Cuba
  - Largely selected by Embassy
  - US Marines assigned to Embassy Detail were also seen at our request

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#### Intervention

- All individuals
  - Standard history and physical with an additional targeted neurotologic history and physical
  - Eye movement tests
    - Nystagmus
    - Smooth pursuit
    - Saccades and anti-saccades
    - Optokinetic Responses
    - Vergence Response
  - Audiometry
- Subset of individuals
  - Additional vestibular testing
  - Neuropsychological testing

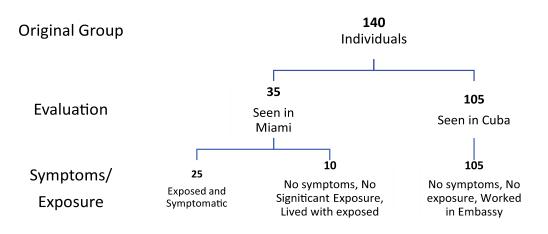


#### Methods

- Only clinically relevant data was collected (all testing was justified and required approval)
- Individuals were acute and unaffected by the influences of time, variable pre-treatment modalities, compensation (workers compensation issues), and media attention



#### **Population Studied**





#### Presenting Symptoms

SYMPTOM	Affected Group (N=25)	Unaffected group (N=10)	Difference	99% Confidence Interval	Fisher Exact P (2 tail)
Dizziness	23 (92%)*	0(0%)	92%	66- >99%	<0.001
Cognitive	14 (56%)*	0 (0%)	56%	32-78%	0.002
Hearing Loss	8 (32%)*	0 (0%)	32%	14-58%	0.073
Tinnitus	8 (32%) *	0 (0%)	32%	14-58%	0.073
Ear Pain	7 (28%) *	0 (0%)	28%	11-54%	0.084
Headache (HA)	6 (24%)	2 (20%)	4%		1.000
At least 2 Symptoms					
Including HA	24 (96%)*	0	96%	71->99%	<0.001
Excluding HA	24 (96%)*	0	96%	71->99%	<0.001
At least 3 Symptoms					
Including HA	16 (64%)*	0	64%	39-83%	<0.001
Excluding HA	14 (56%)*	0	56%	32-78%	0.002

#### Group data

CLINICAL FINDING (Affected Patients)	Number Tested (N)	Abnormal (Percentage)	Prevalence 99% Confidence Interval
Subjective Visual Vertical (SVV)	25	22 (88%)	65-98%
Antisaccade test (abnormal error rate)	25	13 (52%)	31-73%
Standard Audiometry	25	2 (8%)	0-31%
Central Vestibular Findings	25	9 (36%)	18-59%
Chair Impulse Test (HVOR)	12	10 <b>(83%)</b>	48-98%
Cervical Vestibular Evoked Myogenic Potential (VEMP)	12	8 <b>(67%)</b>	34-89%
Ocular VEMP	12	8 (67%)	34-89%
At least one VEMP	12	11 <b>(92%)</b>	56- >99%

#### What was abnormal

- Subjective Visual Vertical/Vestibular evoked myogenic potential (100% one or both)
  - Indicative of abnormal function of the utricle and saccule resulting in abnormal gravity sense and misrepresentation of the body's gravitational inertial vector (GIV)
  - This results in effort dedicated to staying upright with less energy available or cognitive tasks and increased fatigue
  - Can be seen in those without vestibular pathology but not with the definitions of abnormal applied here
- Head rotation tests (83%)
  - Corresponding dysfunction in the semi-circular canals
  - Can result in dizziness



#### **Abnormal Definitions**

- SVV -greater than or equal to 3.2 degrees deviation (lower 5<sup>th</sup> percentile of normative data from 300 subjects)
- Antisaccade -error rate (moving in the wrong direction) greater than or equal to 43% (lower 5<sup>th</sup> percentile of normative data from 300 subjects)
- Standard Audiometry Battery– audiogram, word identification, speech recognition test, tympanometry, reflexes
- Central Vestibular Findings Abnormality on any central vestibular test
- Chair Impulse Test -HVOR gain less than 0.80 at 100 degrees/sec impulse
- Cervical VEMP- Abnormal if amplitude less than 100 microvolts and/or greater than 35% amplitude asymmetry between sides
- Ocular VEMP Abnormal if amplitude less than 3 microvolts and/or greater than 35% amplitude asymmetry between sides abnormal if amplitude typically less than 5 microvolts
- This level of abnormality is not seen without vestibular pathology

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### Cognitive/Neuropsychological

- Complaints
  - Cognitive fog
  - Inattention
  - Problems retrieving information
  - Increased irritability
- Neuropsychological Testing
  - Below expected level
    - Verbal fluency
    - Working memory
    - Sustained attention
  - Difficulty with auditory processing
  - Difficulty with increasing levels of cognitive load



#### Exposure

- Unknown to date
  - Ultrasonic energy
  - Microwave energy
- Directed energy can produce cavitation bubbles
  - Bubble formation and bubble bursting can produce damage
  - Candidate spaces exist in the area of vestibular end organs



#### Is this mTBI?

- Definitions do not match
- Findings do not match
  - anti and predictive saccades and head thrust in mTBI vs. otolith findings in this syndrome
  - High incidence of headaches in mTBI vs. low incidence in this case
  - Differences in neuropsychological test outcome



#### What is this

- An acquired neurosensory dysfunction
  - Essentially universal otolithic disorders
  - Some additional vestibular disorders
  - Unique pattern of cognitive findings
- Site of injury
  - Could be limited to inner ear with secondary cognitive dysfunction
  - Could be in multiple areas
- This is real physical injury in those truly symptomatic



#### **Diagnostic Screen**

- These findings suggest the ability to screen potential cases
  - Otolithic tests are easy to transport and quick to perform
  - A quick cognitive screen could be easily designed
- Screening techniques are critically important to distinguish worried well (many documented cases already) from truly affected
- Any current or future evaluation of cases should include this short battery and include individuals with expertise in this area



#### Thank you

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