Background

- Pseudobulbar affect (PBA) occurs secondary to a variety of otherwise unrelated neurologic diseases or brain injury and is characterized by uncontrollable, inappropriate outbursts of laughing and/or crying.

- The total US prevalence of PBA was conservatively estimated at 1.5-2 million persons in a large, online survey of patients/caregivers of patients with neurologic conditions predisposing to PBA.

- Moreover, for each underlying neurologic diagnosis, such as multiple sclerosis (MS), published prevalence estimates vary dramatically (Figure 1).

- PBA remains under-recognized. Clinicians seldom inquire about PBA symptoms and patients and caregivers may not recognize PBA as a distinct treatable condition.

- Disease registries are powerful tools for obtaining prospective epidemiologic data. A disease registry was established to estimate PBA prevalence in a clinic sample using a validated rating instrument.

Objective

- To estimate PBA prevalence and quality of the QoL impact across disease states commonly associated with PBA, utilizing a registry.

Methods

- Recruitment goal: The Pseudobulbar Affect Registry Series (PRISMA) intends to recruit ~500 sites nationwide, which in turn will enroll up to 10,000 patients at risk for PBA due to underlying MS, Alzheimer’s disease, amyotrophic lateral sclerosis, Parkinson’s disease, disease, stroke, or traumatic brain injury.

- Recruitment procedures: Sites enroll in PRISMA via a centralized web portal and register with a central institutional review Board (IRB). Upon IRB approval, each active site begins screening consecutive patients with the above primary neurologic conditions.

- Data acquisition:
  - Enrolled patients complete the Center for Neurologic Study—Lability Scale (CNS-LS), a validated PBA screening tool, and a QoL measure asking patients to rate on a scale of 0-”10” how strongly affected, “How has your neurologic condition affected your quality of life?”
  - Demographic/disease characteristics are also collected.
  - Investigators enter study data via a Web portal.

- Initial investigator recruitment efforts uncovered several obstacles, chiefly:
  - Unfamiliarity with IRB approval processes
  - Requirement (at some sites) to use a local versus the central IRB
  - Lack of recognition of time commitment and associated costs necessary to complete investigative site start-up
  - Implementation of strategies to overcome identified barriers increased enrollment fivefold, to approximately 500 per month (Figure 2. These include:)
    - Step-by-step and on-site assistance navigating IRB application procedures
    - Reimbursement for startup costs, including limited costs for local IRB
    - Investigator newsletter providing summary of study progress and addressing key issues
    - Medical Affairs assistance to recruit and train investigators on research procedures
    - Distribution of brochures describing the registry
  - As of April 30, 2013:
    - 560 sites have registered
    - 216 sites activated

Figure 1: Published PBA Prevalence Estimates by Primary Neurologic Condition

<table>
<thead>
<tr>
<th>Primary Diagnosis</th>
<th>AD</th>
<th>ALZ</th>
<th>MS</th>
<th>PD</th>
<th>Stroke</th>
<th>TBI</th>
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<tr>
<td>ALS</td>
<td>60 (10)</td>
<td>70 (14)</td>
<td>65 (13)</td>
<td>65 (15)</td>
<td>58 (10)</td>
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Table 1: Characteristics of Current PRISMA Registry Participants

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<tr>
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<th>Total</th>
<th>AD</th>
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<th>MS</th>
<th>PD</th>
<th>Stroke</th>
<th>TBI</th>
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<tbody>
<tr>
<td>Age (years)</td>
<td>Mean (SD)</td>
<td>60 (10)</td>
<td>70 (14)</td>
<td>65 (13)</td>
<td>65 (15)</td>
<td>58 (10)</td>
<td>47 (15)</td>
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<tr>
<td>Gender</td>
<td>Relative %</td>
<td>63% (37)</td>
<td>63% (36)</td>
<td>63% (40)</td>
<td>60% (81)</td>
<td>63% (13)</td>
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Conclusions

- The PRISMA Registry is a novel tool for estimating national PBA prevalence, giving practitioners unprecedented insight into local and national PBA epidemiologic patterns.

- Successful implementation of disease registries requires training and support of clinicians unfamiliar with clinical research procedures and familiarity to accommodate local IRB use.

- The high prevalence of positive PBA screening scores relative to published prevalence estimates re-emphasizes a need for greater education regarding PBA and proactive screening for PBA in at-risk populations.

References


Disclosures

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