The Double D: Assessment tools for the diagnosis of Dementia in adults with Down syndrome
Lior Fishman
SUNY Plattsburgh- Department of Communication Sciences and Disorders

Introduction
Down syndrome (DS), also known as trisomy 21, is the most common genetic disorder resulting from an extra copy of the 21st chromosome. This genetic disorder results in apparent physical and developmental delays.

Dementia is an umbrella term referring to disorders that result in decreased memory, personality changes and impaired reasoning which interfere with an individual’s daily activities. Alzheimer’s disease (AD) is the most common form of dementia resulting from build up of abnormal proteins between the nerve cells, which cause the destruction of brain cells (Moriconi, Schlamb, & Harrison, 2015).

Dementia Scale for Down Syndrome (DSDS)
The DSDS was created to diagnose dementia and measure baseline for individuals with intellectual disabilities who may be at risk for dementia. The DSDS was designed to specifically avoid psychological long impairments that may result in a false positive diagnosis. Questions on the assessment are broken into three categories: early, middle, and late characteristics of dementia (Jozavai, Kartakis, & Gedy, 2009).

Tempel et al. (2001) conducted a study on 35 adults with DS between the ages of 29 and 67 as cited in Jozavai et al., 2009, p. 58. Participants in the study were followed for a minimum of six months and were noted with the DSDS several times throughout the study to ensure accurate results. 20% of participants were diagnosed with early stage dementia and 6% were diagnosed with middle and late stage dementia.

Dementia Questionnaire for people with Intellectual Disabilities (DMR)
The DMR is a tool completed by family and staff members for individuals with intellectual disabilities to screen for possible dementia diagnosis. A single administration of the DMR is highly discouraged as it does not provide accurate results and may not take into consideration secondary behaviors and health concerns. Studies have found that a longitudinal study using the DMR provides the most reliable and accurate diagnosis. The assessment is interpreted in combination with other diagnostic information by the diagnosing physician, psychologist and behavioral therapist. The DMR is rated with three response options based on behaviors that have occurred within the last two months: 0 points-- no deficit, 1 point-- moderate deficit, 2 points -- severe deficit (Evenhuis, Kengen, & van Wieringen, 2009).

Deb and Braganza (1999) compared ratings of several informant based scales based on diagnoses made on 62 adults with DS (as cited in Evenhuis et al., 2009). The study found the use of observer rated scales to be more useful than neurophysiological tests in the diagnosis of dementia.

Dementia Screening Questionnaire for People with Intellectual Disabilities (DSQUID)
The DSQUID is a user friendly questionnaire that is to be completed by a caregiver who has known the individual before and after the onset of symptoms, created to screen for dementia in adults with DS. The sections of the assessment cover information regarding loss of memory, confusion, loss of skills, behavioral changes and speech abnormalities. The first section contains questions regarding the individuals “best ability” they had and currently have (i.e. could speak fluently and understandably, could live independently and needed a lot of help with self-help skills). The second section contains 43 questions associated to behaviors and symptoms typically associated with dementia (i.e. cannot find words, cannot dress without help, obsessive and repetitive behaviors). The third section contains 10 comparative questions to be answered yes or no (i.e. speaks or signs less, walks slower, generally appears more confused) (Deb, Hare, Prior, & Bhaumik, 2007).

A study conducted by Deb et al. (2007) examined 193 adults with DS from the UK. Clinicians diagnosed dementia in 49 participants, found an absence of diagnosis in 68 participants and 76 participants were not examined and therefore received no diagnosis.

Recommendations
• Adults with DS should take part in annual medical examinations to review medications and screen for expected comorbidities. Due to communicative difficulties adults with DS are unable to provide accurate descriptive information (Glasson, Dye, & Bittles, 2014). A failure to provide annual assessments results in delayed intervention and further health complications.
• The DSQUID should be administered at regular intervals to identify change in scores (Deb et al., 2007).
• Test batteries for early identification of individuals with mild to moderate intellectual disabilities should include measures of memory in addition to caregiver observations (Jozavai et al., 2009).
• The DMR should be administered if there is any observed deterioration (Evenhuis et al., 2009).

Conclusion
The normal ageing process in adults with DS is not well enough understood and requires more evidence and research in order to better understand expected cognitive changes (Moriconi et al., 2015). Due to the absence of a "gold standard" in diagnosis of dementia in adults with DS it is challenging to establish a standard score that would allow for definitive diagnosis. Determining accurate validity measures for assessment is challenging due to the absence of a standard cut off score. Clinical decisions are made based on screening tools which are used for identifying persons at risk rather than allowing for a concrete diagnosis (Jozavai et al., 2009). Based on the advantages and disadvantages it may be assumed the DSQUID is a more robust and reliable tool for dementia diagnosis in adults with DS when compared to the DSDS and DMR. The DSQUID has been found to be within good test retest reliability and validity (Deb et al., 2007).