Attentional Lapse and Inhibition Control in Adults with Williams Syndrome

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Abstract

Research exploring cognitive processing associated with Williams Syndrome (WS) has suggested that executive functioning deficits exist across the developmental spectrum. Such executive functions include problem solving, planning, dividing attention and inhibiting responses. Within a framework of executive functions, the aim of the current study was to explore attentional lapse and inhibition skills in older adults with WS (n=20; aged 36 – 61 years) and consider the implications of deficits within this group. Participants with WS were compared to typical adults of the same chronological age and typical older adults (aged 65+ years) to consider attentional changes seen in the ageing process. The study employed a sustained attention to response task known to assess inhibition and attentional lapse but which had not previously been used with this population. Compared to both groups of typical matches, the results indicated atypicalities of attention and inhibition in adults with WS. Specifically, compared to the typical matches, adults with WS failed to withhold a response (showing inhibition deficits), had problems re-engaging attentional control processes after making an error and showed a generalized deficit of concentration and task engagement. We conclude that further attention should be paid to the cognitive capacity of older individuals with WS in order to consider the everyday challenges faced by this group and to provide adequate intervention and support for daily living.

1. Introduction

Williams Syndrome (WS) is a neurodevelopmental disorder with a prevalence of 1:20,000 (Wang et al., 1997; but see also 1:7,500 Strømme, Bjørnstad, & Ramstad, 2000) that is caused by a micro-deletion of approximately 28 genes on chromosome 7 (Osborne, & Mervis, 2007). Individuals with the disorder tend to function within the mild-moderate range of intellectual difficulty (Searcy et al., 2004) and exhibit a cognitive profile of relative proficiency within the verbal compared to the non-verbal domain (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999). The cognitive profile of the disorder has attracted the attention of cognitive scientists for the last two decades due to this juxtaposition of relatively better verbal than non-verbal skill, but it is critical to emphasise that heterogeneity of cognitive function occurs (Porter, & Coltheart, 2005) and the relative difference between verbal and spatial skill co-exists against a background of mild-moderate intellectual difficulty. Within the cognitive profile, research has recently highlighted the importance of exploring the area of executive functioning (e.g. Rhodes, Riby, Park, Fraser, & Campbell, 2010) since the successful engagement of such processing mechanisms is closely related to everyday cognitive ability. Executive function (EF) is an umbrella term that encompasses a range of higher order cognitive processes that control and regulate functions such as working memory, problem solving, planning, divided attention and inhibition and which are predominantly controlled by frontal brain regions (Alvarez, & Emory, 2006). Here, we focus on response inhibition and lapses of attention as these are executive skills with clear implications for understanding wider deficits related to facets of the WS phenotype (e.g. the inability to inhibit inappropriate social approach behaviour, Little et al., 2013).

In research exploring executive functioning in WS, there is no consensus regarding the precise components of executive ability that are more or less impaired. However, in a recent paper in the area Costanzo et al. (2013) examined a variety of executive function tasks in children, and younger and older adults with WS (range 11-35 year olds) compared to Down Syndrome (DS) and mental-age

matched typical controls. Planning ability was particularly compromised in the WS group, with mixed finding found in categorization and inhibition, particularly with regards the modality of the tests employed (i.e. visual vs. auditory tasks yielding inconsistent results; see Osório et al., 2012 who also employed a battery of executive function tasks and again report inconclusive findings).

Somewhat more informative, research has suggested that some individuals with WS share executive function characteristics with individuals who have Attention Deficit Hyperactivity Disorder (ADHD; Rhodes et al., 2010). Comorbid ADHD is relatively more common in WS (64%; Lefeyer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006) than it is in other disorders such as DS (6-8%; Dykens, 2007). Important here is the fact that ADHD is a neurodevelopmental disorder characterised by impaired attention, hyperactivity, impulsivity and disinhibition (Nigg, 2001; Rhodes, Riby, Matthews, & Coghill, 2011) and which is linked to executive-frontal lobe deficits within the brain (Willcutt, Doyle, Nigg, Faraone, & Pennington, 2005). Focussing specifically on inhibition, possible primary, and at least secondary, causes of the behavioural deficits observed in ADHD can be explained by disinhibitory deficits (Nigg, 2001). Recent fMRI work concurs that the executive impairment observed in WS mirrors the patterns seen in ADHD. In that study, Mobbs et al. (2006) employed fMRI while participants with WS (aged 15-48 years) performed a *Go/No Go* measure of sustained attention and inhibition. The authors concluded that observed dis-engagement of the frontal-striatal networks of the brain contributed to the complex pattern of social and behavioural deficits associated with WS (see Hocking et al., 2013 who examined dual tasking and inhibition in the motor domain). In summary, work that has administered batteries of executive function tasks have been inconclusive while those that have specifically examined inhibition are promising in pinpointing the precise executive cognitive processes impaired in WS.

We have noted that EF has been linked to other facets of the WS phenotype. Cognitive aspects of inhibition can be linked to a social phenotype characterised by a tendency to indiscriminately approach both familiar and unfamiliar people (Jones et al., 2000). Using Cluster Analysis to explore heterogeneity of social approach within WS, Little et al. (2013) noted that the participants who showed most indiscriminate and atypically heightened approach ratings to unfamiliar faces were also those individuals who struggled with the Sun-Moon inhibition task (as opposed to relating to emotion processing ability or intellectual capability; Little et al., 2013). The authors proposed that the finding provided preliminary support for a frontal lobe hypothesis of atypical social behaviour within the disorder. The study emphasised the necessity to explore inhibition abilities in individuals with WS due to their link to other facets of the disorder. For example, identifying the role of inhibition in abnormal social approach may mean that intervention can target this skill within a broad intervention approach that considers the cognitive and behavioural needs of the individual.

The first aim of the current study was to investigate inhibitory processing in adults with WS (aged 35+ years), as to date there is limited research that focusses on these behaviours in an older adult cohort. It is not unreasonable to predict particular inhibition deficits in an older WS sample given 1) typically developing older adults suffer from executive deficits (see frontal ageing hypothesis; Greenwood, 2000; inhibition deficit hypothesis; Hasher, & Zacks, 1988) and 2) older adults with WS have been argued to suffer from "mild accelerated ageing" (Krinsky-McHale, Kittler, Brown, Jenkins, & Devenny, 2005; p. 483). For these reasons we also incorporated an elderly typically developing comparison group to help in the data interpretation. The second aim was to employ a task that would enable a comprehensive examination of lapses of attention and inhibition which had previously been demonstrated to be related to real world activities in other populations, including individuals with a neurodevelopmental disorder (e.g. ADHD as well as traumatic brain injury; see Smilek, Carriere, & Cheyne, 2010 for discussion). The paradigm used was the Sustained Attention

to Response Task (SART; Robertson, Manly, Andrade, Baddeley, & Yiend, 1997), a vigilance task which required the participant to respond to a frequent non-target stimulus and withhold a response to an infrequent target stimulus. There were three main metrics derived from the task. First, FA commission errors where participants failed to inhibit a response to non-target infrequent stimuli were used as a measure automaticity and inhibition. Secondly, and arguably the most sensitive measure, pre- and post-error reaction times after a commission error to reflect error monitoring was utilised. Finally, as a general measure of task engagement, differences in the variability of reaction time during the task were gathered as a further measure of attentional lapse (see Dockree et al., 2004; Smallwood, Riby, Heim, & Davies, 2006). Here we aim to elucidate how inhibitory deficits observed in older adults with WS during the SART compare with typically developing individuals matched for chronological age (CA) and gender, and with a group of typically developing adults aged 65 years and over (65yrs). It was hypothesised that 1) the WS group would report greater deficits in failing to withhold a response compared with both the CA and more similar to the over 65yrs groups with known difficulties in inhibitory control (Greenwood, 2000), 2) there would be no difference in the WS group's RT before and after a failure to withhold a response, similar to other populations with known deficits in error monitoring and executive control (e.g. traumatic brain injury, Robertson et al., 1997), whereas both the CA and 65yrs (a wealth of research suggests executive controlled deficits in ageing, however error monitoring in the context of a sustained attention tasks appears spared, e.g. McVay, Meier, Touron, & Kane, 2013) groups would show an increase in RT post-error reflecting an ability to learn from the commission errors, and 3) there would be more variability in reaction times overall during the task reflecting a deficit in task engagement and attentional lapse in the WS group compared to the CA and over 65yrs groups.

2 **Method**

2.1 Participants

Twenty adults with WS (Table 1 displays participant characteristics) were recruited via the UK Williams Syndrome Foundation. Fifteen individuals had previously had their clinical diagnosis confirmed with fluorescent in situ hybridization testing to detect the deletion of one copy of the Elastin gene on chromosome 7. The remaining 5 individuals had a clinical diagnosis but this took place prior to the implementation of routine genetic testing. Three of the WS adults lived independently and seventeen lived with their parents / carers or in sheltered accommodation. Five of the WS adults were in employment while the rest attended daycare centres or received state-provided care assistance. Seventeen adults with WS completed the Wechsler Abbreviated Scale of Intelligence (WASI; mean Full Scale IQ = 60.82 indicative of mild intellectual impairment). WASI data were unavailable for 3 adults with WS due to difficulties complying with the demands of testing.

An age and gender matched typically developing group were recruited for the CA matches. Twenty healthy typically developing older adults were recruited from an existing database of older adults held at Northumbria University and through local older adult groups within the Newcastle-upon-Tyne area. The additional comparison group was tested to help in the interpretation of the data since the WS population tested here were an older adult sample, older adults with WS have been reported to suffer from accelerated ageing (include cognitive; Krinsky-McHale et al., 2005) and inhibition deficits observed in normal ageing may mirror the difficulties observed in an older WS population (Hasher, & Zachs, 1988). The participants in the two comparison groups received £9.00 for their participation. This study received ethics approval from the local ethics committee prior to commencement.

Insert Table 1 about here

2.2 Materials

The Sustained Attention to Response Task (SART; Robertson et al., 1997) is a vigilance task which has been used extensively in neuropsychological research to examine the nature of inhibition deficits (e.g. traumatic brain injury; Robertson et al., 1997; ageing; Carriere, Cheyne, Solman, & Smilek, 2010; ADHD; Johnson et al., 2007) and importantly has validity in terms of everyday attention and inhibition (e.g. Smilek et al., 2010). Participants have to respond to a non-target (the letter 'X') and withhold a response to a target (the letter 'Y'). Stimuli were presented on-screen in Courier New font size 28. Stimulus duration was 300ms interspersed by an inter-stimulus fixation cross presented for 900ms. There were 6 blocks of 20 stimuli, with 120 stimuli in total. The 'Y' stimulus frequency was 20%, with targets and non-targets presented in fully randomised order. The task was programmed using Eprime v2.00 (Psychology Software Tools, Inc.) and stimuli were presented on a Toshiba laptop with a twelve inch screen. A4 laminated examples of the stimuli were used as visual aids for all participants during explanation of the task.

2.3 Procedure

The testing sessions with the WS group took place in their homes, with a parent / carer present at the session or nearby. The comparison groups' testing sessions took place in the Psychology Department at Northumbria University. To commence the session, the participants were greeted by the experimenter and seated in a comfortable chair in front of the computer. The experimenter outlined the experimental procedure and invited each participant to read and sign an informed consent form. Written informed consent was provided by the WS group where possible and by all parents / carers. Before beginning the SART the participants were presented with the following instructions:

> "In this task you will see the letters X and Y appear on the screen. Your task will be to push the space bar whenever you see the letter X. Do nothing when the letter Y appears on the

screen. We would like you to give equal weight to responding to the stimulus and also to minimising errors"

These instructions were reiterated verbally by the experimenter and the participants shown the laminated examples of the stimuli. All participants performed a practice block of 10 stimuli (9 'X's / 1 'Y') prior to performing the main session. Task duration was approximately four minutes.

3. **Results**

3.1 False Alarm Commission Errors (Frequency of failures to withhold on the SART)

Summary data are presented in Table 2. The mean probability of making a commission false alarm (FA) error was considered in a univariate analysis of variance (ANOVA), with Group as the between subjects factor. There was a main effect of group on FA rates [$F(2,59) = 7.832$, $p=.001$]. Tukey post hoc analyses revealed the WS group made significantly more FA than the 65yrs group ($p=.001$) but not the CA group ($p=.207$). The difference between the CA and 65yrs group approached significance ($p=.08$) in that the over 65yrs made fewer FA. The analysis was repeated on the response times when making a FA. ANOVA identified a main effect of group on RT [$F(2,59) = 10.035$, $p<.001$]. Tukey post hoc analyses found the WS group's RT when making a FA was significantly slower than the CA group ($p=.009$) but not the 65yrs group ($p=.418$). There was a significant difference between the CA and 65yrs comparison groups ($p<0.001$).

Insert Table 2

3.2 Hit rates for the frequent non-target stimuli

ANOVA were also applied to hit rates (correctly responding to the non-target). A significant main effect of group was observed [$F(2,59) = 30.677$, $p<.001$]. The WS group reported a significantly lower hit rate when responding to the non-target than both the CA and 65yrs groups (both $p<.001$), while the CA group reported a significantly greater hit rate than the 65yrs group ($p=.05$). The ANOVA also revealed a significant main effect of RT to hit rates [$F(2,59) = 15.913$, $p<.001$]. Tukey post hoc analyses reported no difference in RT between the WS and CA groups ($p=.943$), but significantly longer latency by the 65yrs group when responding to the non-target than both the WS and CA groups ($p<.001$).

3.3 Reaction Time before and after a failure to withhold a response

In order to identify the effect of a failure to withhold a response on RT and error monitoring by the participants, the mean RT was calculated on the two stimuli presented immediately before and immediately after each FA. Smallwood et al., (2006) use this analytical approach and argue that after a FA error attention tends to be re-directed back to the task after a period of task disengagement resulting in slower reaction times. Data were only included into the mean if a participant correctly responded to four non-target stimuli (i.e. two responses before and two responses after an error), resulting in RT data from 8 of the WS group, 17 from the CA group and 10 from the 65yrs group being included in this analysis. Separate t-tests for each group (WS, CA and 65yrs) were employed to compare their RT before and after a FA commission error. The WS group reported no difference in RT before and after a FA [$t(7) = 0.196$, $p=.85$]. In contrast the CA group reported significantly slower RT post FA [$t(16) = 3.329$, $p=.004$], whilst the latency in the 65yrs group approached significance [$t(9) = 2.251$, $p=.051$]. These data are presented in Figure 1.

Insert Figure 1

3.4 Mean variability in RT during performance of the SART task

ANOVA were also applied to task variability measure (SDs of response time throughout the whole task for each participant). A significant main effect of group was observed [$F(2,57) = 26.48$, $p<.001$]. Tukey post hoc analyses revealed greater variability in the WS group compared to both the CA and over 65yrs (both $p<.001$). There was no difference in variability between the CA and over 65 groups ($p=.77$). These data are displayed in Figure 2.

Insert Figure 2 about here

4. Discussion

The findings of the current study demonstrate that the SART task is sensitive for examining different aspects of attentional lapse and inhibition in WS. It has previously been proposed that older adults with WS may suffer accelerated ageing and work on children and adolescents points to executive functioning deficits accompanying the disorder (Rhodes et al., 2010). While inhibition has been studied with regards cognitive (e.g. Costanzo et al., 2013) and social functioning (Little et al., 2013), neither of these research endeavors have provided a comprehensive comparison of different metrics of attentional lapse and inhibition, in an older WS group, and when completing a task known to be related to everyday cognitive failures (Smilek et al., 2010).

Consider first the effects observed using FA errors of commission to the infrequent target stimuli and the response times pre- and post-error as dependent variables. Robertson et al. (1997) argue that as well as errors being an indicator of poor inhibition, quicker responses prior to, and increase in

response time following, an error "…are the result of drift of controlled processing into automatic responding consequent on impaired sustained attention to task" (p.747). However, in the present study the WS participants did not follow this pattern and their performance was in line with other populations with known frontal lobe and associated executively controlled processing deficits (e.g. traumatic brain injury, TBI; Robertson et al., 1997). The comparison of younger and older control participants demonstrated that FA commission errors were greater in the CA group but this difference was accompanied by slower responses for the typical adults over 65 years of age. Although this finding failed to reach significance for the FA data it seems plausible that the elderly participants were sacrificing their speed to maintain task performance. Speed-accuracy trade-offs of this nature and individual differences in strategies employed during cognitive task are typical in ageing research where adults attempt to compensate and minimize errors during task completion (Starns, & Ratcliffe, 2010). Interestingly, if we were to predict (due to the proposal of accelerated ageing in WS) a similar pattern of results this did not occur. Numerically (and significant for the 65yrs vs. WS comparison), WS participants produced the highest FA commission errors. This alone suggests an inhibition deficit, especially when considering the response times were equivalent to the over 65yrsand slower than the CA match. The increased response time for the WS group did not lead to reduced FAs as a speed-accuracy trade-off would have predicted. This finding is consistent with inhibition deficits found on more traditional neuropsychological measures (e.g. West, Schwarb, & Johnson, 2010) and work suggesting ADHD characteristics associated with WS in children and younger adults (Rhodes et al., 2011)

Post error slowing after a FA commission error is an important indicator of the executive functions of error monitoring and the re-establishment of controlled processing during sustained attention. In ageing this aspect of executive function is relatively well preserved during continuous performance tasks like the SART described here (e.g. McVay, Meier, Touron, & Kane, 2013). However, with

more severe frontal lobe deficit the pattern is somewhat different. For example, individuals who have suffered from traumatic brain injury, characterised by frontal lobe and white matter damage, fail to decelerate RTs after an error on the SART (Robertson et al., 1997, see also Dockree et al., 2004). It is important to exercise caution in the interpretation of these data in the current study due to the reduced sample size in this analysis brought on by insufficient trials to create a mean in some participants. However, WS participants in the current study showed this precise pattern, implying that under conditions of automaticity brought on by the presentation of long streams of non-target stimuli, these individuals are unable to re-establish executive control of behaviour to maintain sustained attention performance. As an example of this sort of behaviour in other domains of cognition, it is worthwhile emphasizing error monitoring in spatial cognition where inefficient visual search performance is characterized by a lack of monitoring of previously visited spatial locations (Smith et al., 2009). Therefore rather than showing parallels to a 'normal' ageing profile, WS older adults display inhibitory processing deficits consistent with those who have received traumatic brain injury (Robertson et al., 1997). Elsewhere, in the working memory domain lower hit rates accompanied by higher FAs were observed in a TBI population which supports our study suggesting similarities WS and TBI profiles (Slovarp, Azuma, & LaPoine, 2012). The profile of older adults with WS being comparable with TBI is not surprising given our abovementioned discussion of executive deficits in ADHD and WS (section 1.1). Rhodes and colleagues (e.g. Rhodes et al., 2011) have been influential not only highlighting the relationships between the executive deficits and dis-inhibition observed in WS and ADHD but also stressing the importance of this avenue of research given how such cognitive measures predict everyday behavioural difficulties (e.g. reported via parents and carers; Rhodes et al., 2010).

As a broad measure of attentional lapse and task engagement it is desirable to consider the mean hit rates to non-target frequent stimuli. Perhaps not surprising given predictable large individual differences within neurodevelopmental disorders, indeed considering the cognitive heterogeneity we

know to be associated with WS (Porter, & Coltheart, 2005) the hit rate was low (48% WS vs 93% CA vs 79% over 65yrs) and the standard deviation was high (28%). Regarding our analysis of variability in response times during the duration of the task it is evident that WS participants were unable to exert controlled processes to maintain focus during the task. Both the CA and over 65yrs were comparable, but for the WS group a lapse of attention in general was evident as well as an inability in learning from a commission error. Sustained attention metrics including RT variability have been used in previous research when assessing the key cognitive markers of ADHD and indeed proved to be strong predictors of impairment further highlighting the similarities of the cognitive difficulties observed between WS and ADHD (Williams et al., 2010). Much like other disorders of development, it has been argued that sensitive cognitive measures of inhibition may serve as a phenotype marker (Crosbie, & Schachar, 2001).

The aforementioned results show the benefit of including an older typical sample of matched individuals in that the results seen in the WS group cannot be linked directly to an ageing hypothesis or interpretation. Exploring any possible association with ageing in the WS group was a key aim of the current study. However, it would have also been useful to include one further group of typical individuals of comparable mental age to ensure that the pattern of findings for the WS sample was not associated with mental capacity. This additional comparison may be useful in future research of this nature.

Regarding underlying neuro-cognitive mechanism sub-serving inhibition impairment a future avenue of research would be to extend Mobbs et al. (2006) finding of deficit in frontal-striatal systems using fMRI and investigate how these inhibition networks differ from more social aspects of self-regulation and control seen in orbitofrontal-amygdala interactions (Meyer-Lindenberg et al., 2005). Furthermore, converging evidence from event-related potential studies with the aim to pinpoint the temporal dynamics of inhibition deficits (see N200 work; e.g. Schmajuk, Liotti, Busse, & Woldorff, 2006)

would be advantageous. Indeed, in other domains such as face processing ERPs have been successful at pinpoint the processing mechanisms impaired and spared with early markers related to attention being spared (Mills et al., 2000).

4.1 Conclusion

With the SART tasks used in this study a myriad of controlled processes related to inhibition and attentional lapse were found to be problematic for older adults with WS. Failing to withhold a response, re-engaging attentional control processes after an error and an overall deficit of concentration and task engagement was apparent. To be clear, we believe that under certain conditions a deficit in executive control prevents WS adults effectively monitoring and shifting from automatic to control modes of processing. By examining different aspects of attention and inhibition within the same task we are in agreement with research elsewhere that stresses that we should not consider inhibition as a single construct (e.g. Sinoplolia, & Dennis, 2012). Indeed, although it could be argued that those with WS suffer a global deficit in inhibition, further work is needed to investigate the different aspects to get a fuller understanding of cognitive and social components of inhibition across the lifespan in WS. The inclusion of an older adult control group was informative since the pattern of results was not consistent with the accelerated ageing hypothesis (Krinsky-McHale et al., 2005). Research endeavours should mimic those carried out with TBI where a systematic examination of the types of inhibition impaired will allow interventions and strategies to be employed to minimise difficulties in "…adaptive functioning, poor psychosocial outcomes, and decrements to academic, vocational, and social successes" Sinoplolia & Dennis, 2012; p.213).

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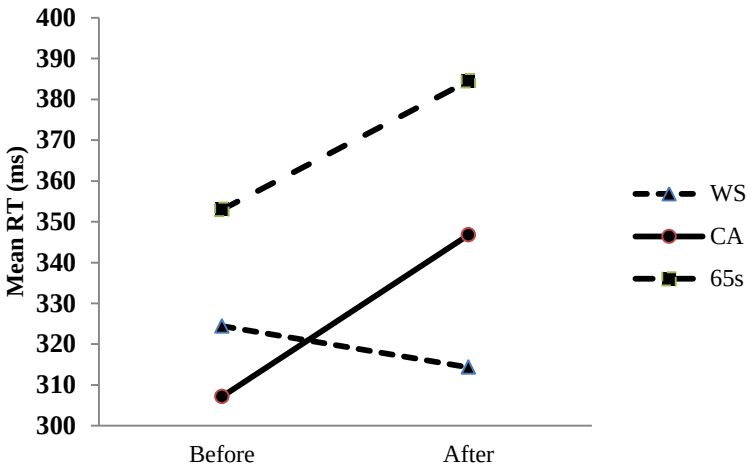

Figure 1: Mean reaction time (RT) in ms of responses before and after a false alarm commission error: WS, CA, over 65yrs(squares)

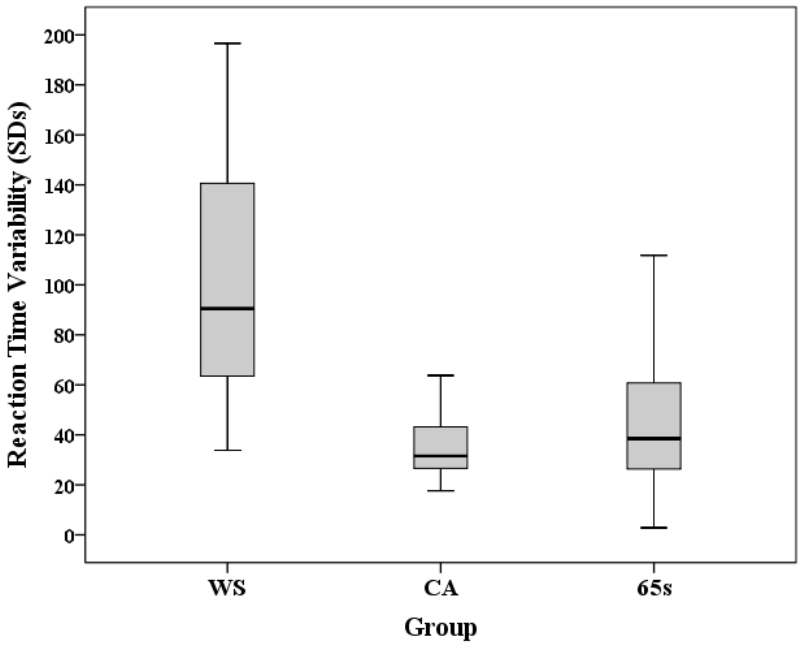

Figure 2: Mean variability in RT during task across WS, CA and over 65yrs groups