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**The Relationship between Executive
Function and Challenging Behaviour
in Students with Down Syndrome** □

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العلاقة بين الوظائف العقلية التنفيذية والمشكلات السلوكية لدى التلاميذ ذوي متلازمة داون في الكويت

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ملخص

هدفت هذه الدراسة إلى كشف العلاقة بين الوظائف العقلية التنفيذية وبين المشكلات السلوكية وأيضاً بحث العلاقة بين الوظائف العقلية التنفيذية وبين السلوك الاجتماعي المقبول للتلاميذ ذوي متلازمة داون. كانت عينة الدراسة ٣١ تلميذ من تلاميذ المرحلة الابتدائية في اجنحة تلاميذ متلازمة داون الملحقة في المدارس العادية في الكويت. اشتملت العينة على ١٥ تلميذ و ١٦ تلميذه متوسط أعمارهم بين (٦,٥٨ - ١٥,٤٢). تم تقييمهم بواسطة مقياس الوظائف العقلية التنفيذية، كما تم تقييم سلوكهم من خلال استبيان مواطن القوة والضعف (المشكلات السلوكية والسلوك الاجتماعي المقبول).

كشفت النتائج عن: هناك علاقة موجبه ذات دلالة بين المشكلات السلوكية والقصور في الوظائف العقلية التنفيذية متمثلة في (الكبح- المرونة العقلية - الضبط العاطفي - الذاكرة العاملة- التخطيط - الدرجة الكلية للوظائف التنفيذية) أي كلما زاد القصور في هذه الوظائف زادت المشكلات السلوكية، كما ان هناك سالبه بين الوظائف العقلية وبين السلوك الاجتماعي المقبول أي انه كلما زاد القصور في الوظائف العقلية التنفيذية قل السلوك الاجتماعي المقبول لدى هؤلاء التلاميذ من ذوي متلازمة داون.

الكلمات المفتاحية: صعوبات معرفيه ومتلازمة داون، المشكلات السلوكية، الوظائف العقلية التنفيذية.

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Introduction

Down syndrome (DS) or trisomy 21 is deemed to be the most common genetic disorder (Jones, 2006; Kruszka et al., 2017). DS is one of the most common causes of intellectual disabilities (Canfield et al., 2006; Kinnear et al., 2018). Research has explored the relationship between the cognitive abilities of individuals with DS and other phenotypes that may affect their quality of life (e.g., Asim et al., 2015; Kazemi et al., 2016). Highlighting this relationship is critical if the strengths and weaknesses of those with DS are to be better understood.

Research Problem

Cognitive abilities are pivotal to people's quality of life as they are inextricably linked to learning, behaviour, social relationships and the ability to adapt to changing situations and environments. Executive functions are cognitive abilities responsible for controlling and regulating a range of behavioural and other functions and are essential for successful adaptation (Benavides-Nieto et al., 2017; Costanzo et al., 2013). Executive functions (EF) are critical for decision-making, planning initiating and stopping actions, and adapting and controlling behaviour. Any impairments or delays in executive functions can be demonstrated by reduced adaptation, including challenges with cognitive shifting, inhibition and memory (Costanzo et al., 2013; Karr et al., 2018). Certain aspects of executive functioning has been found to be delayed in adults and children with Down syndrome (e.g., Daunhauer et al., 2017; Fidler, 2005; Thompson, 2003). Research indicates, for instance, that both adults and adolescents with Down syndrome have difficulties with set-shifting, conceptual shifting, sustained attention, planning, inhibition

and working memory (e.g., Godfrey & Lee, 2020; Rowe, Lavender & Turk, 2006; Sabat et al., 2020; Zagaria et al., 2021).

There has also been recent research into the executive functions of children displaying challenging behaviour. Challenging behaviour can include behaviour that is aggressive, impulsive, inappropriate, harmful or self-injurious, or non-compliant or characterised by withdrawal (Adams et al., 2018; Emerson et al., 2001).

Research indicates that children with DS are more likely to possess the behavioural phenotype of challenging behaviour compared to typically developing children, given the higher rates of this kind of behaviour in this population (Feeley & Jones, 2006; Grieco et al., 2015). In particular, DS is linked to problems such as non-compliance, social isolation, attention-hyperactivity, compulsions and self-talk, all of which may increase with age (Feeley & Jones, 2006). Other behavioural issues of those with DS include distraction, avoidance, attention-seeking and escape behaviours, that are thought to underly challenges with on-task learning, non-compliance and misguided social skills (Feeley & Jones, 2008; Grieco et al., 2015).

Despite findings indicating that executive functioning may be somewhat improved in children with DS (e.g., Diamond & Lee, 2011), it remains uncertain if there is a link between EF and challenging behaviour in DS and specifically, if interventions to improve EF may also help to improve challenging behaviour in children with Downs syndrome. There is some research indicating that challenging behaviour (CB) may be associated with delays in EF in those with DS (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998; Wilding et al., 2002); factors that

may play a specific role in contributing to CB in those with DS include a lack of effective decision-making (Cuskelly, Gilmore, Glenn & Jobling, 2016), and delays or difficulties with cognitive flexibility (Zelazo, Burack, Benedetto & Frye, 1996). Individuals with DS may face challenges in attempting to understand things from another's point of view (Theory of Mind) (Tavakoli, Demehri & Azizi, 2019). Therefore, given the range of EF-related developmental issues faced by those with DS, it is clear that this genetic condition impacts individuals on at least three different levels: biological, cognitive and behavioural level.

This research aims to investigate whether any particular weaknesses in EF correlate with CB in children with DS. Moreover, this research also aims to investigate whether the pattern of strengths in some EFs in individuals with DS correlates with social behaviour.

Literature Review

Down Syndrome: Epidemiology, Genetic Basis and Health Issues

Worldwide, 1 in 1000 live births every year are diagnosed with DS, although there may be variation in this rate in different geographical regions (WHO, 2016). DS is the most common genetic disorder, and accounts for 8% of the total congenital disorders diagnosed each year (Weijerman & de Winter, 2010). Individual with DS have long-term health, social and educational support requirements that can adversely affect their quality of life if they remain unmet (Buckley, 2012). However, the overall life expectancy of individuals with DS has increased in recent years, with many people with DS living for 50-60 years as a result of advances in both health and social care (Kliegma, 2011; Startin et al., 2020).

DS is also the most common chromosomal cause of intellectual disability (Daunhauer & Fidler, 2013). The cause of DS is generally trisomy 21, although sometimes may be caused by mosaicism or translocation, and there is a higher incidence of DS births amongst older mothers (Doria-Rose et al., 2003). In a small minority of cases (usually 1-2%), mosaicism may occur, which is when some (but not all) cells are affected by trisomy 21 (Fisher, 2013); in these individuals, intelligence is generally higher (Fisher, 2013). This compares with translocation, which occurs in approximately 3-4% of individuals with DS; this is where a section of genetic material (an extra copy of chromosome 21) may be attached to another chromosome (Fisher, 2013).

Individuals with DS often present with comorbid health disorders, including mood disorders, sleep difficulties, cardiovascular disorders, imbalances in the central nervous system, neurological structural abnormalities and hormonal disturbances (Mazurek & Wyka, 2015). Other disorders associated with DS include early onset Alzheimer's disease and reduced muscle tone (hypotonia) (Smith, 2001). Therefore, continual care and treatment is needed by those with DS (Dimopoulos & Kempny, 2016). Some research reveals that individuals with DS can often present with a range of visual and hearing difficulties (Dennis & Brian, 2006) and some degree of hearing impairment (Kreicher et al., 2018). It is possible that many of the associated health challenges experienced by children with DS, including hearing and vision impairment, could contribute to both cognitive impairment and CB (Dennis & Brian, 2006).

Executive Functions in DS

Executive functioning describes a range of cognitive functions that facilitate a range of adaptive, goal-directed actions, including planning, working memory, inhibition and shifting (Lanfranchi, Jerman, Dal Pont, Alberti & Vianello, 2010). EFs start to take shape during infancy (Miller & Marcovitch, 2015) and continue to develop throughout adolescence until early adulthood (Anderson, 2001; Henry & Bettenay, 2010). The EF of working memory involves the ability to retain and manipulate information; shifting involves the ability to flexibly adapt to different rules, such as thinking about things from a different viewpoint or thinking creatively; planning involves generating appropriate steps to solve problems and achieve goals; and inhibition involves the ability to regulate one's behaviour and respond appropriately to situations. This includes controlling one's impulses and the ability reduce distractions and resist temptation (Diamond, 2013).

Studies have attempted to identify specific patterns of cognitive delay in DS, which often include, for example, a lower IQ (Daunhauer et al., 2014), challenges in maintaining attention and controlling impulses (Dieleman et al., 2018), yet strengths when it comes to empathy and social skills (Buckley, 2012). In general, therefore, individuals with DS appear to demonstrate a low level of cognitive abilities (Startin et al., 2020) with significant EF challenges and delays (Will et al., 2017; Tomaszewski et al., 2018). For example, Rowe et al. (2006) investigated EFs in adults with DS and compared them to an age-matched control with a learning disability (without DS) and revealed that those with DS performed at a significantly lower level on various EF tests measuring working memory, planning,

inhibition, shifting and problem solving. Another study by Costanzo et al. (2013) revealed that those with DS exhibited various challenges with EF that depended on the type of EF and the task modality (whether it was verbal or visuo-spatial) compared to 16 mental-age-matched typically developing children and 16 adults and children with Williams syndrome. In particular, individuals with DS struggled with working memory (WM), inhibition and shifting, especially in the verbal modality. This is supported by other research, such as from Carney et al. (2012), which compared children with DS to children with Williams syndrome and typically developing children, revealing (similarly to Costanzo et al., 2013) that individuals with DS displayed relative difficulty in the verbal modality of set-shifting. Daunhauer et al. (2014) also revealed, from questionnaires completed by teachers and parents of children with DS, that there are particular delays and challenges experienced by individuals with DS in the domains of inhibition, planning and WM.

The patterns of strengths and weaknesses in the cognitive profile of DS has led some researchers to conceptualise EFs into either ‘hot’ or ‘cool’ categories (Lee et al., 2015). “Cool” EFs are those functions that do not involve an emotional element, including planning and WM (Zelazo & Müller, 2011). By comparison, “hot” EFs describe those functions that are associated with reward, reinforcement and motivation, describing the cognitive abilities required to make or set emotionally-salient decisions and goals (Zelazo & Müller, 2011). Various researchers have utilised standardised EF measures and revealed that children with DS demonstrate greater difficulties in ‘cool’ EFs, such as WM and monitoring, compared to ‘hot’ EFs, such as emotional control (e.g., Lee et al., 2015). It must be

noted however that individuals with DS often demonstrate atypical development scores compared to typically developing individuals in both hot and cold test scores (Lee et al., 2015), indicating that individual differences and different developmental trajectories must be taken into account. The results from a range of studies however indicate that overall, both hot and cool EFs may be delayed in individuals with DS, beyond any impairments in general intellectual functioning.

Therefore, in spite of varied findings in the research a lack of consensus amongst researchers, it appears that, in general, an overall cognitive profile has emerged that maps the general EF abilities of individuals with DS in specific domains (Daunhauer & Fidler, 2012). Challenges in particular appear to be experienced in the domains of working memory (Godfrey & Lee, 2018; Lanfranchi, Jerman & Vianello, 2009), planning (Fidler, Hepburn, Mankin & Rogers, 2005; Schworer et al., 2020), shifting (Daunhauer et al., 2017; Rowe, Lavender & Turk, 2006) and inhibition (Edgin, 2003; Fontana et al., 2021; Rowe et al., 2006).

Challenging Behaviour

Challenging behaviour (CB) is defined by the Royal College of Psychiatrists et al. (2007) as behaviour that “is of such an intensity, frequency or duration as to threaten the quality of life and/or the physical safety of the individual or others and is likely to lead to responses that are restrictive, aversive or result in exclusion”. This definition, whilst acceptable, may not encompass all behaviour that is perceived to present a challenge, given that ‘challenging’ behaviour may not be entirely objective. CB is generally perceived to include behaviour that is

aggressive, impulsive, self-injurious or socially inappropriate, and can also involve withdrawal and/or non-compliance (Emerson et al., 2001). CB can be disruptive to an individual's educational development across the lifespan as well as being detrimental to the wellbeing and quality of life of parents, carers and teachers (Feeley & Jones, 2006). Given the profound impact that CB can have on the health, wellbeing, opportunities and safety of individuals (Dennis & Brian, 2006), it is essential to devise and implement effective interventions aiming to reduce such behaviour from an early age.

The causes and contributors to CB can be wide and varied, including medical, cognitive and environmental factors, necessitating a 'biopsychosocial model' of CB in order to understand it more holistically (Koritsas & Iacono, 2012). Similarly, Macleod (2010) notes that a more coherent understanding of CB is prevented by the varied approaches from different disciplines, such as sociology and neuropsychology; as a result, Macleod (2010) calls for an interdisciplinary approach that brings in an educational perspective as a matter of urgency.

Relationship between Executive Functions and Challenging Behaviour in Down Syndrome.

The causes of CB in individuals with DS may be varied, yet research indicates it may be attributed—in part, at least—to challenges encountered in daily living skills (Daunhauer, 2011) and in different EF domains (Daunhauer & Fidler, 2013; Lee et al., 2011; Rowe, Lavender & Turk, 2006). Difficulties with EFs could prompt disruptive or challenging behaviour in those with DS (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998), due to ineffective decision-making

(Cuskelly, Einam, & Jobling, 2001), and delays in cognitive flexibility in relation to mental age (Sabat et al., 2020).

Children with DS may encounter specific difficulties in school functioning, which must be addressed if they are to take advantages of the learning opportunities presented to them. One study of children with DS in elementary school asked teachers and parents to rate the difficulties exhibited by the children and revealed that participants perceived them to be most competent in planning and WM, and least competent when it came to skills involving behavioural regulation or understanding/following social conventions, and interacting positively with their peers (Daunhauer, Fidler & Will, 2014). Thus, this study highlights that challenges with self-regulation and behavioural management negatively impacted the children's educational experience within the classroom. Clearly, support services must work harder to implement inclusive education and provide a tailored, needs-based assessment for each individual to establish their unique strengths and weaknesses (Buckley, 2012).

Research Questions

- 1- **Is there relationship between EF abilities and challenging behaviour in students with DS?**
- 2- **Is there relationship between EF abilities and social behaviour?**

Research Aims

- To investigate the correlation between executive functions and challenging behaviour.
- To investigate the correlation of executive functions and social behaviour.

Method**Research Design**

This study adopted a descriptive, quantitative design.

Ethical Considerations

Consent forms were presented to parents of students with DS and the procedure of the research was fully explained. All parents agreed to participate in this research by signing the consent form.

Sample

The pupils' sample was comprised of 31 pupils with DS (15 male and 16 female, aged between 6.58 years to 15.42 years), who attended one of four special units in Kuwait for pupils with DS. All pupils were Kuwaiti and of Arabic ethnicity.

Data Collection**Questionnaires**

Questionnaires were utilized as a cost-effective, practical and standardised tool (Basit, 2010). This study utilised two questionnaire tools, to be completed by parents of students with DS.

1- Behaviour Rating Inventory of Executive Function – Preschool version (BRIEF-P) Questionnaire.

The BRIEF-P is widely used within the field of intellectual disabilities research. It consists of a 63-item rating form that is designed to measure children's EF in both home and school contexts. It is able to be completed by both parents and teachers (Isquith, Gioia & Espy, 2004). It assess five functions using five different subscales: inhibition, shift,

emotional control, working memory (WM) and planning/organising (PO), providing statements that can be answered ‘never, ‘sometimes’ or ‘often’ (rated 1, 2 and 3, respectively). The scores of each sub-section can then be totalled to provide a global EF score. The reliability of this questionnaire is between acceptable to high, as shown:

Variable	Cronbach's Alpha
Inhibition	.834
Shift	.865
Emotional control	.785
Working memory	.937
Plan organize	.873
Global executive function	.956

2- *Strengths and Difficulties Questionnaire (SDQ).*

The SDQ is a 25-item measure including five subscales (containing five statements for each subscale). The total score from four of the subscales (emotional symptoms, conduct problems, hyperactivity, peer problems) indicates total behavioural difficulties, and prosocial behaviour is measured on the remaining subscale. The statements within each subscale require a response of either, ‘not true’, ‘somewhat true’ or ‘definitely true’ (rated 0, 1 and 2 respectively). The SDQ also contains five statements that are phrased in the opposite direction to assess social behaviour, which must be reversed prior to scoring (SDQ, 2018).

All 20 items delineating total behavioural difficulties were assessed for internal consistency reliability; values of the Cronbach’s alpha scale showed acceptable internal consistency (.71). The remaining prosocial behaviour scale demonstrated high levels of internal consistency (.88).

Results

To ascertain whether there was a correlation between EF scores on the BRIEF-P assessment and challenging or prosocial behaviour (SDQ) scores in students with DS, data were analysed using Pearson correlation coefficient (Table 1).

	SDQ PROSOCIAL items	SDQ PAR Total Difficulties items
BRIEF PARENT INHIBIT SCALE	-.588**	.768**
BRIEF PARENT SHIFT SCALE	-.555**	.575**
BRIEF PARENT EMOTIONAL CONTROL SCALE	-.537**	.718**
BRIEF PARENT WORKING MEMORY SCALE	-.489**	.511**
BRIEF PARENT PLAN_ORGANISE SCALE	-.493**	.572**
BRIEF_PARENT__GLOBAL_EXECUTIVE COMPOSITE	-.650**	.761**

** Correlation is significant at the 0.01 level.

* Correlation is significant at the 0.05 level.

Table 1 show that there is a significant positive correlation between the score of total difficulties items that assess CB and all BRIEF-P subscales (inhibition, shifting, emotional control, WM, PO and the total score of global EF) in students with DS. This means that difficulty in EF was significantly correlated with CB. In addition, the results revealed a significant negative correlation between prosocial behaviour and all BRIEF-P subscales (inhibition, shifting, emotional control, WM, PO and the total score of global EF) in students with DS. This means that greater

difficulties in EF were significantly negatively correlated with prosocial behaviour (those rated as possessing greater difficulties on the BRIEF-P (significant low level of EF) were rated as more prosocial and vice versa).

Discussion

The findings of this study correspond with previous research that demonstrated that CB is associated with delays in EF in individuals with DS (Memisevic & Sinanovic, 2014; Pennington & Bennetto, 1998; Wilding et al., 2002). Challenges to EF can heavily impact cognitive functioning, such as effective decision-making (Cuskelly et al., 2016), to retain and work with important information or rules (working memory) (Lee et al., 2015), and the ability to view things from another's perspective (cognitive flexibility) (Tavakoli, Demehri & Azizi, 2019). This can lead individuals, such as those with DS, to face everyday challenges in following instructions, switching between and completing tasks, maintaining attention, and deciding which course of action to follow. Delays in cognitive flexibility in particular may result in a lack of emotional intelligence (Yazdi et al., 2018), which can negatively impact prosocial behaviour, explaining the finding in the present study that greater EF difficulties was associated with a reduction in prosocial behaviour. Given the behavioural impact of delays to EF, it is perhaps unsurprising that incidences of CB are greater in those experiencing challenges to their executive functioning, such as those with DS. This coincides with the view that the genetic condition of DS impacts those affected on a biological, cognitive and behavioural level.

The finding that those demonstrating greater delays to their EFs exhibit more CB is supported in the literature. Schuiringa et al. (2017) for example assessed the EFs of WM, cognitive flexibility and inhibition, as well as externalising behaviour problems (CB) in children with average and low IQ (mild borderline intellectual disability). They found that those with mild borderline intellectual exhibit more CB *and* greater delay in some EFs – particularly WM, and (to a lesser extent), inhibition. Interestingly, however, their study showed no link between CB, IQ and cognitive flexibility – it was not more impaired in those children with mild borderline intellectual disability with CB than those with mild borderline intellectual disability without CB. This indicates that more research is clearly required into the role of cognitive flexibility in CB, and perhaps more importantly, into the role that WM may play in contributing to CB. This could lead to targeted WM interventions with the aim of reducing CB.

There are potentially other factors that may contribute to or exacerbate CB in individuals found to have delays in their EFs. Those with DS, for example, are well-documented in the literature to be more likely to suffer from sleep difficulties, which has been found to correlate with EF weaknesses. For example, Joyce et al. (2019) found that obstructive sleep apnoea (OSA) contributes to the advancement of EF delay, as it impairs cognitive ability over time. Furthermore, sleep problems such as OSA correlate with CB in individuals with DS; Esbensen et al. (2018) for instance found that individuals with DS that had poor quality sleep demonstrated greater inattention, hyperactivity and impulsivity during the day, all of which can manifest in CB. This indicates that sleep may be one area requiring targeted intervention in those demonstrating high levels of CB, delays in EF, or both.

The finding that fewer difficulties in EFs is associated with greater prosocial behaviour has been documented in the non-DS literature. For instance, studies such as that by Diamantopoulou et al. (2007) revealed that EF challenges and ADHD behaviour led to reduced prosocial behaviour, with peers and teachers rating students with more severe EF and ADHD challenges as exhibiting ‘poor’ social functioning. These students tended to exhibit greater physical aggression and other CB and, moreover, there was a gender discrepancy, where girls with poor EF were less accepted by their peers than their male counterparts. Other studies indicate that typically developing children with higher EF abilities, especially in WM and inhibition, often demonstrate greater prosocial behaviour (Williams et al., 2016).

Prosocial behaviour in the form of social cognition and its link to EF was explored in individuals with DS in a study by Amado et al. (2016). These researchers used a battery of EF and social cognition tasks in children with DS and found that, whilst there was an increase in abilities in social cognition in children with DS as they increased in chronological age, WM explained more than 50% of the variability in social cognition in the individuals with DS. This compared to 31% variability in the chronological age-matched typically developing control group. It appears, therefore, that WM in particular plays a critical role in the development of social cognition, which in the literature is widely evidenced to be pivotal in prosocial behaviour (e.g., Bakopoulou & Dockrell, 2016; Conte et al., 2018).

The link between EF (especially WM) and CB and prosocial behaviour indicates that there is a need to conduct training and interventions with the aim of improving EFs in children with DS, which

could, in turn, improve children's behaviour. These interventions could be conducted to train teachers how to modify the behaviour of students with DS at school, or undertaken by parents with the aim of improving CB in the home. Given the evidence base that specific EFs such as WM are linked to greater CB and reduced prosocial behaviour, it is arguably more cost and time effective to focus on developing and implementing WM interventions, as this may improve functioning in a range of areas, such as academic performance, daily living and quality of life, as well as reducing CB at school and in the home.

Decision makers also have significant responsibilities towards individual with ID. As those people experiencing reduced cognitive abilities face challenges in all aspects of their lives, not only CB, there is a need to establish ongoing guidelines for and reviews of conducting interventions with the aim of improving their cognitive abilities, such as EFs. One of the main factors underpinning challenging behaviour is task avoidance; thus, improving EFs abilities could reduce task difficulties, which in turn could reduce CB associated with task avoidance. It should be considered that task difficulties and task avoidance may not only occur in an academic context; it can occur anytime and in any place, as people require cognitive skills in their daily lives need, such as when shopping or interacting with others. This also highlights the strong need to investigate the daily life skills that correlate with specific EFs and design strategies to improve both.

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