Behaviour problems and cognitive skills in adolescents with Down syndrome

by

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Submitted in partial fulfilment of the requirements for the degree of

Doctor of Psychology (Health)

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November, 2014
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Date: 24/11/2014
For my cousin Ryan,

who shared my love of psychology
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ABSTRACT

In this thesis there were two primary aims. The first aim was to examine the type and severity of behaviour problems in individuals with Down syndrome (DS) during late childhood and adolescence. The second aim was to investigate the role of cognitive, neuropsychological and maturational variables in explaining externalising and internalising behaviour problems in persons with DS. In examining the literature it was identified that the number of participants with behaviour problems varies between 17% and 38%, and that the age of the sample and assessment techniques might contribute to this variability. Studies undertaken with control groups also suggests that the causes of behaviour problems might be unique to DS. Cognitive skill deficits and Alzheimer’s neuropathology were proposed to be related to behaviour problems. Two studies were conducted to address the aims of this thesis. The rationale for conducting Study 1 was to examine whether maturation and different measures of behaviour problems were associated with the type and severity of behaviour problems. Twenty-four individuals with DS and 24 typically developing (TD) children and adolescents between 10- and 16-years old participated in this study. A parent completed the Child Behavior Checklist (CBCL) and Social Skills Information System Rating Scales (SSIS-RS) for all participants. The results showed that children with DS had significantly more externalising and total behaviour problems than the control group. Measurement differences and maturation were not significantly related to behaviour problems. Study 2 extended Study 1 by examining whether IQ, emotion recognition, theory of mind, language, social skills and neuropsychological markers of Alzheimer’s disease were correlated with behaviour problems. Participants in Study 2 were the same as those who completed Study 1. All participants were presented with standardised tests to assess the aforementioned cognitive and neuropsychological variables. Parents also completed
the pragmatic language items from the Children’s Communication Checklist. It was shown that pragmatic language was significantly correlated with externalising problems in both the DS and control groups, and internalising problems in the DS group. The results showed that better social skills were associated with more behaviour problems in both study groups. Additionally, better theory of mind and poorer performance on a neuropsychological marker of early dementia was positively correlated with internalising problems in the DS group only. Overall the results from this thesis provide the first evidence that certain cognitive skills and neuropathology might be linked to behavioural problems in individuals with DS.
CHAPTER 1: DOWN SYNDROME

1.1 Introduction

Down syndrome (DS) is a genetic disorder identifiable by a distinct set of physical characteristics. Common physical abnormalities associated with DS include brachycephaly (disproportionately wide head), up-slanting palpebral fissures, epicanthal folds, Brushfield spots (white spots on the iris), flat nasal bridge, protruding tongue, small low-set ears, short stature and small broad hands (Roberts, Price, & Malkin, 2007; Visootsak, Sherman, Visootsak, & Sherman, 2007; Zigman, Silverman, & Wisniewski, 1996). In addition, affected individuals have an elevated risk for congenital heart problems, gastrointestinal disorders, hearing impairments, eye disorders (e.g., congenital cataracts), endocrine abnormalities (e.g., hypothyroidism), orthopaedic problems and a high risk of Alzheimer’s disease in adulthood (Batshaw, 2002; Korenberg et al., 1994; Pueschel, 1990; Roizen & Patterson, 2003).

DS is also associated with a number of intellectual and cognitive problems (Määttä, Tervo-Määttä, Taanila, Kaski, & Iivanainen, 2006; Pennington, Moon, Edgin, Stedron, & Nadel, 2003). The intellectual problems seen in people with DS can be severe. The IQ of affected individuals typically ranges between 30 and 70 with the average for the population estimated to be 50 (Chapman & Hesketh, 2000; Mégarbané et al., 2013; Walker, Dosen, Buitelaar, & Janzing, 2011). This means that on average, people with DS have a mild to moderate intellectual disability (American Psychiatric Association, 2000). Additionally, individuals with DS have been repeatedly found to have a range of cognitive impairments including working memory, attention and executive functioning as well as language impairments.
While much is known about the physical and cognitive correlates of DS, the extent and severity of behavioural problems of people with DS is not so well understood. In the DS literature evidence has been presented showing the presence (e.g., Cuskelly & Dadds, 1992; Glenn & Cunningham, 2007; van Gameren-Oosterom et al., 2011) and absence of behavioural problems (e.g., Eisenhower, Baker, & Blacher, 2005; Gau, Chiu, Soong, & Lee, 2008). This thesis examines the extent to which behavioural problems in individuals with DS are related to one or more cognitive problems. Specifically the first question addressed in this thesis is do children and adolescents with DS present with externalising and internalising behaviour problems? The second research question addressed is whether age, cognitive and neuropsychological skills have a role in explaining externalising and internalising behaviour problems in children and adolescents with DS? The cognitive and neuropsychological variables examined in this thesis are IQ, emotion recognition, theory of mind, language, social skills and a neuropsychological marker of Alzheimer’s disease.

Understanding whether cognitive or neuropsychological problems are related to the behaviour problems in individuals with DS is important. One positive outcome is that children with DS who are likely to have behaviour problems may be identified earlier. Second, understanding the cognitive or neuropsychological correlates may further increase our knowledge about why the behaviour problems are present. Such knowledge might be important in order to improve the effectiveness of intervention and support for children with DS and their families.
1.2 Outline of Thesis

Chapters 2 – 5 provide an overview of the state of the literature with regard to behavioural problems in children and adolescents with DS.

The aim of Chapter 2 is to define the key concepts used in the subsequent sections on childhood behavioural problems. This chapter focuses largely on the differentiation of externalising and internalising problems.

Chapters 3 – 5 review literature investigating the extent and nature of behavioural problems in children and adolescents with DS. Chapter 3 examines studies investigating the number of children and/or adolescents with DS who have behaviour problems. It will be shown that the presence of behaviour problems in this clinical group is high. Differences in externalising and internalising problems are discussed.

Chapter 4 reviews research that has compared behavioural problems in DS groups with those in a control group. Control groups include a typically developing group of similar chronological or mental age, or a group of individuals with an intellectual disability or neurodevelopmental disorder. Overall, these studies suggest that IQ is not a strong predictor of behaviour problems in people with DS.

In Chapter 5 it will be shown that individuals with DS have a range of cognitive and neuropsychological impairments. It is proposed that impairments in IQ, emotion recognition, theory of mind, language, social skills and/or Alzheimer’s disease neuropathology might influence the behaviour problems of children and adolescents with DS. The role of maturation is also considered.
On the whole this review of the literature indicates that many children and adolescents with DS experience behaviour problems; however, relatively little is known about the cause of these problems.

Chapter 6 presents a summary of the child variables that are proposed to be related to the behavioural problems in children and adolescents with DS. This chapter also outlines the research hypotheses for the cognitive, social and neuropsychological variables, and externalising and internalising behaviour problems.

Chapter 7 describes Study 1, which examined the percentage of children and adolescents with behaviour problems, and its association with maturation. A comparison of participants with and without DS is undertaken and the results discussed.

Study 2 is presented in Chapter 8. This study investigated the relationship between cognitive, social and neuropsychological skills, and behavioural problems in the DS and TD groups. Correlational analyses are presented, followed by a discussion of these results.

Chapter 9 presents a general discussion of the results of this thesis. The clinical implications of the research are considered. Limitations of the studies presented throughout this thesis and the implications of the results for future research are also discussed.
CHAPTER 2: DEFINING BEHAVIOUR PROBLEMS IN CHILDHOOD

The purpose of this chapter is to present an overview of behaviour problems in childhood. A variety of terms exist to describe the construct of ‘behaviour problems’. These include ‘maladaptive behaviour’, ‘problem behaviours’, ‘challenging behaviours’, ‘aberrant behaviour’ and ‘behaviour disorders’ (Shattuck et al., 2007). In general, behaviour problems refer to behaviours that are dysfunctional, interfere with daily activities, are socially inappropriate or, are detrimental to an individual’s ability to cope with stressors (Bhatia, 2009; Shattuck et al., 2007). A widely adopted taxonomy of behavioural problems distinguishes between externalising and internalising problems (e.g., Achenbach & Edelbrock, 1978; Forns, Abad, & Kirchner, 2011; Mesman, Bongers, & Koot, 2001). An introduction to these different types of behaviour problems is now presented.

2.1 Externalising & Internalising Behaviour Problems

Externalising problems describe behaviours that are harmful, disrupting or damaging to oneself, others or to property (Eisenberg et al., 2001; Keil & Price, 2006; Liu, 2004; Phares, 2008; Zahn-Waxler, Klimes-Dougan, & Slattery, 2000). Table 1 presents a summary of different types of externalising behaviour problems. Common types of externalising behavioural problems include aggressive, delinquent, hyperactive, inattentive, disruptive and oppositional behaviours (Cicchetti & Toth, 1991; Liu, 2004; Phares, 2008; Rapport, Denney, Chung, & Hustace, 2001; Wilmshurst, 2009). Some externalising problems involve acts that verbally or physically harm an individual or non-human animals. Others may involve negative interactions with property such as vandalism or theft. However, not all externalising problems necessarily entail outward behaviour that ultimately harms other individual/s or leads to the destruction of property (Liu, 2004). For example,
hyperactive behaviour describes excessive motor activity that may result in difficulties staying on task. The individual may be constantly fidgeting, talking or yelling inappropriately, have difficulties remaining seated when expected (e.g., in the classroom) or be continually moving, which causes considerable disruption to others (American Psychiatric Association, 2000; Liu, 2004; Selikowitz, 2009).

Table 1. Different Types of Externalising Behaviour Problems

<table>
<thead>
<tr>
<th>Behaviour Problem</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aggression</td>
<td>Behaviours that cause or threaten to cause harm to other people or animals. Aggression may be physical, verbal, proactive or reactive (American Psychiatric Association, 2000; Liu, 2004; Ramírez &amp; Andreu, 2006).</td>
</tr>
<tr>
<td>Delinquent behaviour</td>
<td>A range of criminal and non-criminal anti-social behaviours. These behaviours may include, but are not limited to violence, truancy, vandalism, running away from home, promiscuity, drug use, disobedience, and shoplifting (Liu, 2004; Segen, 2006; Shoemaker, 2008).</td>
</tr>
<tr>
<td>Attention problems / Inattention</td>
<td>Difficulties with sustaining and regulating attention. A child with attention problems may have difficulties following instructions, be easily distracted, appear to not listen when being spoken to, or have difficulty completing tasks (American Psychiatric Association, 2000; Liu, 2004).</td>
</tr>
<tr>
<td>Hyperactivity</td>
<td>Describes over-active motor behaviours or restlessness. For example, a child may have difficulty sitting still, run or climb excessively at inappropriate times, or appear constantly ‘on-the-go’ (American Psychiatric Association, 2000; Liu, 2004; Selikowitz, 2009).</td>
</tr>
</tbody>
</table>

Internalising problems are behavioural difficulties characterised by disordered mood or emotional distress. Common to internalising problems is the absence of overt behaviour that negatively impacts on another individual or property (Forns et al., 2011; Kovacs & Devlin, 1998). Table 2 summarises different types of
internalising behavioural problems. Common internalising problems include depression, anxiety, social withdrawal and somatic complaints. Due to their covert nature, these behaviours may not be immediately visible to others (Forns et al., 2011). For example, a child may choose to avoid certain stimuli (e.g., dogs) or situations (e.g., social gatherings) in an effort to minimise feelings of anxiety. This avoidant behaviour may not be particularly obvious to other people.

Table 2. *Different Types of Internalising Behaviour Problems*

<table>
<thead>
<tr>
<th>Behaviour Problem</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anxiety</td>
<td>A psychological state that is characterised by excessive worry and fear (Blumberg &amp; Izard, 1986). Symptoms include being easily fatigued, concentration difficulties, irritability, muscle tension and sleep difficulties (American Psychiatric Association, 2000).</td>
</tr>
<tr>
<td>Depression</td>
<td>Describes low mood and/or loss of interest and pleasure in doing things. Other symptoms include irritability (particularly in children), significant and unplanned changes in weight, insomnia or excessive sleep, lacking energy, concentration difficulties, feelings of worthlessness or excessive guilt and suicidal ideation (American Psychiatric Association, 2000; Blumberg &amp; Izard, 1986).</td>
</tr>
<tr>
<td>Withdrawal</td>
<td>Consistently exhibiting solitary behaviour when meeting with familiar and/or unfamiliar peers (Rubin, Burgess, &amp; Coplan, 2002).</td>
</tr>
<tr>
<td>Somatic complaints</td>
<td>Physical symptoms of ill-health that do not have an identifiable biological cause, and instead appear to be linked to depression (Chakraborty, Avasthi, Grover, &amp; Kumar, 2010).</td>
</tr>
</tbody>
</table>
2.2 Conclusion

Common to all behaviour problems is that they are maladaptive responses. At the same time, behaviour problems are a multifaceted construct that broadly comprises externalising and internalising problems. These behaviour problems are not specific to individuals with DS. Therefore, the next two chapters review literature that has specifically examined behavioural problems in samples with DS, as well as studies that have compared DS groups with other children that do/do not have a developmental disorder or genetic syndrome.
3.1 Introduction

This chapter reviews research examining behaviour problems in people with DS during the childhood and adolescent years. In the first section of this chapter, criteria for determining whether or not a child or adolescent presents with a behavioural problem is considered. The second part of this chapter investigates how common behaviour problems are in individuals with DS. The third part of the chapter examines the literature to identify whether DS is associated with a specific type of behaviour problem. From this review it will be demonstrated that behaviour problems are common in children and adolescents with DS. With respect to specific types of behaviour problems, there is evidence suggesting a possible maturational or developmental trend. Specifically, preliminary evidence suggests externalising problems appear in childhood and internalising problems become apparent in adolescence. However, there does not appear to be one specific type of internalising and externalising problem that is prevalent amongst children and adolescents with DS.

3.2 Determining the Presence of Behaviour Problems

Behavioural research undertaken with people with DS can be broadly described as assessing problems according to either a behavioural checklist/s, manualised diagnostic criteria or the presence of an existing psychiatric diagnosis. In this section, the advantages and disadvantages of these assessment methods are discussed, with particular consideration of how these apply to the literature reviewed in the following chapters.
Behaviour Problems in Down Syndrome

One way of measuring behaviour problems in developmental research is by behavioural checklists. Depending on the specific measure, a behavioural checklist is completed by a parent or guardian, or teacher. These questionnaires usually include a list of behavioural problems and the respondent is asked to rate how often or severe each problem is for the child on a likert scale. The aggregate of individual items is then used to calculate individual subscales (e.g., externalising problems scale) and/or an overall behavioural problem score (e.g., total problems score). One benefit of behavioural checklists is that they are often standardised measures that provide a researcher with clinical and subclinical cut-off markers, so that the clinical significance of the reported problems can be determined. This allows clinicians and researchers to determine the type and severity of problems experienced by the individual, as well as estimate how many individuals display significant behavioural disturbances in a sub-population.

One difficulty that is encountered when examining behavioural literature for people with DS, is that some studies fail to report the proportion of participants with clinical or subclinical range scores. This makes it difficult to determine how severe the behavioural problems are in each sample. Instead, some studies report the proportion of participants with behavioural problems at an individual item level (e.g., Dykens & Kasari, 1997; Dykens, Shah, Sagun, Beck, & King, 2002). It is problematic to estimate the overall presence of behavioural problems in people with DS using studies that have only reported on individual items, these are less reliable than results obtained using composite scores. However, given that this thesis aims to examine how behavioural problems do/do not present in people with a low prevalence clinical condition it is also problematic to exclude these studies from review. Consequently, in the following sections, where composite scales are available these are reported. Studies that have included only single item results are
also included in the review are also presented, however this is noted in the tables and results are treated with more caution, given the lower reliability of the results.

Another, less frequently used method for measuring behavioural problems is by investigating the proportion of participants that have an existing psychiatric diagnosis or meet the diagnostic criteria for a behavioural disorder. This approach is useful as it provides information about the presence of severe and clinically significant behavioural difficulties in a sub-population. However, it might also provide an under-estimation for the number of children or adolescents that present with behavioural disturbances. This is because only children with the most severe behaviour problems would be considered to display behavioural problems in these studies. In this review any participant with a behavioural diagnosis or that meets diagnostic criteria for a disorder primarily characterised by externalising (e.g., aggression, delinquent behaviours) or internalising symptoms (e.g., depression, anxiety) is considered to have behaviour problems. It is also proposed that this approach does not take into account sub-clinical behaviour problems and therefore might be an under-estimate of the true proportion of children and adolescents that exhibit behavioural difficulties.

3.3 How Common are Behaviour Problems in Down Syndrome?

In this section studies are reviewed that have examined the number of children or adolescents with DS who present with one or more types of behaviour problem (e.g., Nicham et al., 2003; Povee, Roberts, Bourke, & Leonard, 2012; van Gameren-Oosterom et al., 2011). A summary of studies examining the number of individuals with DS in a sample who present with behaviour problems is presented in Table 3.
### Table 3. Background Study Characteristics and Proportion of Total Behaviour Problems in Individuals with Down Syndrome

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% of DS Sample with Behaviour Problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Burke, Fisher, and Hodapp (2012)</td>
<td>42</td>
<td>15.1</td>
<td>Child Behavior Checklist</td>
<td>15.4</td>
</tr>
<tr>
<td>Dykens and Kasari (1997)</td>
<td>43</td>
<td>11.0</td>
<td>Child Behavior Checklist</td>
<td>23.0</td>
</tr>
<tr>
<td>Eisenhower et al. (2005)</td>
<td>12</td>
<td>2.9</td>
<td>Child Behavior Checklist</td>
<td>8.3b</td>
</tr>
<tr>
<td>Fidler, Hodapp, and Dykens (2000)</td>
<td>20</td>
<td>5.8</td>
<td>Child Behavior Checklist</td>
<td>40.0</td>
</tr>
<tr>
<td>Gath and Gumley (1984)</td>
<td>23</td>
<td>8.3</td>
<td>A2 Rutter Behavior Problem Checklist - Parent Reportc</td>
<td>30.0 (A2)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>B2 Rutter Behavior Problem Checklist - Teacher Reportd</td>
<td>23.0 (B2)</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported

- Results are for the Strengths and Difficulties Questionnaire for subsample of participants 5-10 ($n = 14$) and 10-16 ($n = 10$) years old. $^b$Proportion of children with T-scores in the borderline clinical and clinical range. $^c$Rutter, Tizard, and Whitmore (1970). $^d$Rutter (1967). $^e$Scored in deviant range on at least one Rutter scale and had a score of 10 or more on the Additional Behaviour Checklist.
### Table 3. Background Study Characteristics and Proportion of Total Behaviour Problems in Individuals with Down Syndrome (continued)

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% of DS Sample with Behaviour Problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gath and Gumley (1986)</td>
<td>193</td>
<td>51</td>
<td>A2 Rutter Behavior Problem Checklist - Parent Report</td>
<td>34.0 (A2)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>B2 Rutter Behavior Problem Checklist - Teacher Report</td>
<td>31.0 (B2)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Additional Behaviour Checklist</td>
<td>38.3e</td>
</tr>
<tr>
<td>Myers and Pueschel (1991)</td>
<td>261</td>
<td>43</td>
<td>DSM-III-TR Diagnostic Criteria</td>
<td>17.6</td>
</tr>
<tr>
<td>Povee et al. (2012)</td>
<td>224</td>
<td>43</td>
<td>Developmental Behaviour Checklist</td>
<td>26.3</td>
</tr>
<tr>
<td>van Gameren-Oosterom et al. (2011)</td>
<td>320</td>
<td>47</td>
<td>Child Behavior Checklist</td>
<td>26.9</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported

* Results are for the Strengths and Difficulties Questionnaire for subsample of participants 5-10 (n = 14) and 10-16 (n = 10) years old. *Proportion of children with T-scores in the borderline clinical and clinical range. *Rutter et al. (1970). *Rutter (1967). *Scored in deviant range on at least one Rutter scale and had a score of 10 or more on the Additional Behaviour Checklist.
Overall, the number of children and adolescents identified with behaviour problems is typically high. For instance, in the study by van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) 21% of the sample were found to have a ‘Total Problems Score’ in the clinical range on the Dutch translation of the Child Behaviour Checklist (CBCL; Verhulst, Ende van der, & Koot, 1996). This composite index measures the presence of externalising and internalising problems, as well as other types of behaviour problems that fall outside these domains such as attention, thought and social problems. Higher scores on the CBCL indicate the presence of one or more behavioural problems. In a study by Fidler et al. (2000) 40% of children with DS were found to have clinically significant behaviour problems when measured using the Total Problems Score from the CBCL (Achenbach, 1991).

Even though studies have typically reported that a high number of children with DS have behaviour problems, there is variation in these findings. From the studies presented in Table 3, it can be seen that the percentage of children or adolescents with DS presenting with one or more behaviour problems ranges from 15% (Burke et al., 2012) to 40% (Fidler et al., 2000). It is likely that sampling error may account for some of the variation in results here. This is especially likely in those studies that have small sample sizes. The studies summarised in Table 3 that have the smallest sample size often have the greatest variability in study findings. For example, the study by Fidler et al. (2000) found that 40% of children with DS had behaviour problems. The sample size of this study was 20. Dykens and Kasari (1997) found that 23% of their sample had behaviour problems. However, there were 44 children with DS who participated in the study. In a smaller study, Eisenhower et al. (2005) investigated the behaviour problems of 12 young children with DS. The results showed very few participants had behaviour problems, with
only 8% of the sample reported to have a total problem score in the borderline clinical and clinical range on the CBCL. This percentage is considerably lower than those reported in other studies. However, these children were also substantially younger than those assessed elsewhere, with a mean age of three years. Taken together, it can be seen that there is a large disparity in the results reported amongst studies with small samples. Therefore in examining how common behaviour problems are in people with DS, this section continues by reviewing only those studies undertaken with large sample sizes.

There are at least six studies with sample sizes of around 200 or more that have examined behaviour problems in individuals with DS. It is interesting to note that one of these studies found a relatively smaller number of children and adolescents with DS had behaviour problems. Myers and Pueschel (1991) examined behavioural problems in 261 participants with DS. The mean age of the sample was 9.5 years ($SD = 5.4$ years). In this study a participant was considered to have a behaviour problem if s/he met the DSM-III-R criteria (American Psychiatric Association, 1987) for Disruptive, Anxiety, Gastrointestinal, Repetitive, Affective or ‘Other’ disorders, such as Autism. Using this approach a participant could be considered to have an externalising problem if diagnosed with Attention Deficit Disorder, Conduct/Oppositional Disorder or Aggressive behaviour. A child was said to have an internalising problems if s/he were diagnosed with Phobias, Conversion Disorder or Major Depressive Disorder. Using this approach, behaviour problems were diagnosed in 17.6% of the participants with DS. However, it is possible that the method used to identify children with behavioural problems may contribute to the low number. The approach used by Myers and Pueschel required children to have a psychiatric disorder in order to be considered to have a behavioural problem. This approach may underestimate the number of children with DS with behaviour
problems. This is because children who have behaviour problems but do not meet the criteria for a psychiatric disorder would be missed using Myers and Pueschel’s method.

The low number of behaviour problems found by Myers and Pueschel (1991) has not been replicated in other studies. Gath and Gumley (1986) investigated behaviour problems in a sample of 193 children and adolescents with DS. Participants in the study were aged between 6- and 17-years. This study evaluated behaviour problems using the Rutter Rating Scales (Rutter, 1967; Rutter et al., 1970). The Rutter Rating Scales comprises items that ask about whether a child exhibits a specific behaviour (e.g., “Often appears miserable, unhappy, tearful or distressed”). The rater, who may be a parent or teacher, indicates how frequently they observe the behaviour. In the study by Gath and Gumley, a child was considered to have a behaviour problem if s/he scored in the ‘deviant’ range on the Rutter Rating Scales. In this sample 38.3% of the participants were identified to have one or more types of behavioural problems.

In other studies the number of children and adolescents with behaviour problems has been found to be less than 38.3% (as observed by Gath & Gumley, 1986), but greater than 17.6% (as observed by Myers & Pueschel, 1991). Van Gameren-Oosterom et al. (2011) studied behavioural problems in 320 children with DS. The mean age of the sample was of 8.1 years ($SD = 0.2$ years). In this study behaviour problems were assessed using the Total Problems Score on the Dutch version of the CBCL (Verhulst et al., 1996). The CBCL comprises statements describing different types of behaviour (e.g., ‘cruel to animals’). The informant, who may be a parent or someone who spends a lot of time with the child, rates the extent the statement applies to their child. The Total Problem Score measures the severity
of externalising and internalising problems as well as social, thought and attention problems. In the study by Van Gameren-Oosterom et al. 26.9% of children with DS were identified to have clinically significant behaviour problems. Similar results were found by Dykens et al. (2002) who also examined behaviour problems using the CBCL (Achenbach, 1991). In this study, 26.5% of 211 individuals with DS had one or more different types of behaviour problems. The mean age of the sample was 9.7 years.

There is some evidence to suggest that the method used to assess behaviour problems is not entirely related to the instrument used. Povee et al. (2012) investigated behaviour problems in 224 individuals with DS. The mean age of the sample was 13.7 years ($SD = 5.9$ years). Unlike previous studies, behaviour problems were identified using an adapted version of the Developmental Behavior Checklist (Einfeld & Tonge, 1995) designed for use with children/adolescents with an intellectual disability. Using this instrument, the results showed that 26.3% of the individuals with DS had behavioural problems.

### 3.3.1 Summary of Findings for Overall Behaviour Problems

The evidence reviewed thus far has highlighted two main findings. Firstly, behavioural problems are common in individuals with DS. Focusing only on those studies with large sample sizes, it seems that around one quarter of children and/or adolescents with DS have clinically significant behaviour problems. Second, based on the findings of Myers and Pueschel (1991) it seems that for many individuals with DS, behaviour problems may not severe enough to warrant a diagnosis of a psychiatric disorder. The next section examines how frequent specific types of behaviour problems are in children and adolescents with DS.
3.4 Specific Types of Behavioural Problems in Down Syndrome

As noted in Chapter 2, in broad terms, behaviour problems can be categorised as being external or internal in nature. In this section, studies are reviewed that have examined the number of children with DS who present with clinically significant externalising or internalising behaviour problems. A commonly used method in the DS literature to measure the presence of different types of behaviour problems involves using composite scores from the CBCL (Achenbach, 1991). The CBCL permits the summing of related items to create a composite score measuring the presence of externalising or internalising problems (e.g., Fidler et al., 2000; van Gameren-Oosterom, Fekkes, van Wouwe, et al., 2013; van Gameren-Oosterom et al., 2011). Another approach used to quantify the presence of internalising or externalising problems in children with DS is to examine whether a specific type of externalising or internalising behaviour is present. Examples of specific types of externalising problems are aggression and delinquency. Examples of specific internalising problems consist of anxiety, depression, withdrawal behaviours and somatic complaints.

3.4.1 Externalising Problems in Down Syndrome

A summary of studies that have reported the number of children with DS who have externalising problems is presented in Table 4. Summarised in this table is the number of participants in the study and method used to measure the presence of externalising problems. The literature examining externalising problems in children and adolescents with DS has produced mixed findings. Some studies have reported that quite a large number of individuals with DS have externalising problems (e.g., Dykens et al., 2002). In other studies, the number of DS participants with externalising problems has been small (e.g., van Gameren-Oosterom, Fekkes, van...
Wouwe, et al., 2013). In this section the number of children and adolescents with externalising problems is reviewed and an attempt is made to understand some of the inconsistencies in this literature.
### Table 4. Background Study Characteristics and Proportion of Types of Externalising Problems in Individuals with Down Syndrome

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age M</th>
<th>Age SD</th>
<th>Age Range</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Externalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Burke et al. (2012)</td>
<td>42</td>
<td>15.1</td>
<td>2.8</td>
<td>11-21</td>
<td>Child Behavior Checklist</td>
<td>8 Externalising Problems composite</td>
</tr>
<tr>
<td>Cuskelly and Dadds (1992)</td>
<td>21</td>
<td>NR</td>
<td>NR</td>
<td>4-15</td>
<td>Revised Behaviour Problem Checklist</td>
<td>14 Conduct disorder</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>10 Socialised aggression</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>33 Attention problem – immaturity</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>10 Motoric Excess</td>
</tr>
<tr>
<td>Dykens et al. (2002)</td>
<td>211</td>
<td>9.7</td>
<td>3.9</td>
<td>4-19</td>
<td>Child Behavior Checklist</td>
<td>50-76 ‘Argues a lot’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>6-30 ‘Swears’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>27-63 ‘Demands attention’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>38-79 ‘Cannot concentrate’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dykens and Kasari (1997)</td>
<td>43</td>
<td>11.0</td>
<td>NR</td>
<td>4-19</td>
<td>Child Behavior Checklist</td>
<td>65 ‘Argues a lot’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>47 ‘Tantrums’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>72 ‘Disobeys’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>23 ‘Lies, cheats’</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>6 ‘Steals at home’</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; DSM-IV-TR = Diagnostic and Statistical Manual – Fourth edition, text revision (American Psychiatric Association, 2000); ICD-9 = International Classifications for Disease – Ninth edition (World Health Organization, 1978); CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology (Döpfner, Berner, Flechtner, Lehmkuhl, & Steinhausen, 1999); SDQ = Strengths and Difficulties Questionnaire (Goodman, 1997).

* Results refer to individual items on the CBCL (scales not reported). * Data presented for 5-10 year olds (n =22) and 11-30 year olds (n = 18) respectively. * Data presented for 5-10 year olds (n =14) and 10-16 year olds (n = 10) respectively.
### Table 4. Background Study Characteristics and Proportion of Types of Externalising Problems in Individuals with Down Syndrome (continued)

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Externalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ekstein, Glick, Weill, Kay, and Berger (2011)</td>
<td>41</td>
<td>Female: 41</td>
<td>8.0  NR 5 - 16</td>
<td>DSM-IV-TR Diagnostic Criteria</td>
</tr>
<tr>
<td>Fidler et al. (2000)</td>
<td>20</td>
<td></td>
<td>5.8  1.9 3 - 10</td>
<td>Child Behavior Checklist</td>
</tr>
<tr>
<td>Gath and Gumley (1986)</td>
<td>193</td>
<td>51</td>
<td>NR 6 - 17</td>
<td>ICD-9 Diagnostic Criteria</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>5 Aggressive/anti-social/defiant behaviour with depression/</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>anxiety/emotional distress</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>7 Hyperactive/inattention/impulsive with aggressive/</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>anti-social/defiant behaviour</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2 Hyperactive/inattention/impulse with speech delay/</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>reading difficulties/other delays in specific skills</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; DSM-IV-TR = Diagnostic and Statistical Manual – Fourth edition, text revision; ICD-9 = International Classifications for Disease – Ninth edition; CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology; SDQ = Strengths and Difficulties Questionnaire.

a Results refer to individual items on the CBCL (scales not reported). b Data presented for 5-10 year olds (n =22) and 11-30 year olds (n = 18) respectively. c Data presented for 5-10 year olds (n =14) and 10-16 year olds (n = 10) respectively.
Table 4. Background Study Characteristics and Proportion of Types of Externalising Problems in Individuals with Down Syndrome (continued)

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Externalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myers and Pueschel (1991)</td>
<td>261</td>
<td>9.5</td>
<td>DSM-III-TR Diagnostic Criteria</td>
<td>5 Conduct/oppositional disorder</td>
</tr>
<tr>
<td></td>
<td>43</td>
<td>43</td>
<td></td>
<td>7 Aggressive behaviour</td>
</tr>
<tr>
<td></td>
<td>261</td>
<td>9.5</td>
<td></td>
<td>6 Attention deficit disorder</td>
</tr>
<tr>
<td>Nicham et al. (2003)</td>
<td>43</td>
<td>13.1</td>
<td>CASCAP</td>
<td>59-33 Dominant</td>
</tr>
<tr>
<td></td>
<td>49</td>
<td>7.3</td>
<td>SDQ</td>
<td>63-39 Opposing/refusing</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>18-17 Aggression verbal/physical</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>96-67 Inattention/distractible</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>73-39 Impulsive</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>41-11 Increased motor activity</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>29-40 Conduct problems</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>29-10 Hyperactivity/inattention</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; DSM-IV-TR = Diagnostic and Statistical Manual – Fourth edition, text revision; ICD-9 = International Classifications for Disease – Ninth edition; CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology; SDQ = Strengths and Difficulties Questionnaire.

* Results refer to individual items on the CBCL (scales not reported). * CASCAP data available for 40 of the 43 participants. Data presented for 5-10 year olds (n = 22) and 11-30 year olds (n = 18) respectively. * Data presented for 5-10 year olds (n = 14) and 10-16 year olds (n = 10) respectively. * Byrne, Cunningham, and Sloper (1988); Sloper, Knussen, Turner, and Cunningham (1991); Sloper and Turner (1993).
### Table 4. Background Study Characteristics and Proportion of Types of Externalising Problems in Individuals with Down Syndrome (continued)

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Externalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>M, SD, Range</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>40% Female</td>
<td></td>
<td></td>
<td>17 Disruptive</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>94 Disturbed behaviour</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>24 Poor concentration</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>41 Over-activity</td>
</tr>
<tr>
<td>van Gameren-Oosterom et al. (2011)</td>
<td>320</td>
<td>8.1, 0.2, 7.8-9.1</td>
<td>Child Behavior Checklist</td>
<td>15 Externalising Problems composite</td>
</tr>
<tr>
<td></td>
<td>47% Female</td>
<td></td>
<td></td>
<td>3 Delinquent behaviour</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>4 Aggressive behaviour</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>12 Attention problems</td>
</tr>
<tr>
<td>van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013)</td>
<td>322</td>
<td>18.3, 0.8, 16.8-19.9</td>
<td>Child Behavior Checklist</td>
<td>7 Externalising Problems composite</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; DSM-IV-TR = Diagnostic and Statistical Manual – Fourth edition, text revision; ICD-9 = International Classifications for Disease – Ninth edition; CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology; SDQ = Strengths and Difficulties Questionnaire.

a Results refer to individual items on the CBCL (scales not reported). b CASCAP data available for 40 of the 43 participants. Data presented for 5-10 year olds (n = 22) and 11-30 year olds (n = 18) respectively. c Data presented for 5-10 year olds (n = 14) and 10-16 year olds (n = 10) respectively. Byrne et al. (1988); Sloper et al. (1991); Sloper and Turner (1993).
Studies by Burke et al. (2012), Fidler et al. (2000) and van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) observed that the number of children and adolescents with DS who have externalising problems ranged from 7% to 10%. In these studies externalising problems were measured using a composite index. This approach combines different types of externalising problems. The exception to these results was the study by van Gameren-Oosterom et al. (2011) who examined externalising problems in 320 children with DS with a mean age of 8.1 years ($SD = 0.2$ years). The number of behaviour problems was measured by the percentage of children who scored in the clinical range on the Externalising Problems Scale from the CBCL. Results showed that 15% of the DS sample presented with an externalising problem.

In explaining the variability between study findings issues with measurement can be discounted. That is, the number of children identified with a behaviour problem is related to the instrument used to identify the problem. This is because differences between studies have been observed even when behaviour problems have been measured using the same instrument. For example van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) found the percentage of individuals with externalising problems to be 15% and van Gameren-Oosterom et al. (2011) to be 7%. Both studies used the CBCL to measure behaviour problems. Sampling error does not appear to be able explain differences since both studies by van Gameren-Oosterom and colleagues had a sample size larger than 300.

Closer inspection of the literature indicates that age may be related to the presence of externalising behaviour problems in individuals with DS. Specifically, a small number of studies suggest that as children with DS become older, externalising problems may decrease. Studies that have measured externalising problems using a
composite generally appear to follow this trend. Participants in the studies by van Gameren-Oosterom et al. (2011) and Fidler et al. (2000) were children around 6- to 8-years. These studies observed the number of children with externalising problems to be 15% and 10% respectively. In contrast, studies by van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) and Burke et al. (2012) which examined externalising problems in adolescents and young adults with DS reported that 7% to 8% of individuals with DS presented with clinically significant externalising problems.

The next issue addressed in this section is whether one or more specific types of externalising problems are found in children and adolescents with DS. A summary of results from individual studies is presented in Table 4.

Overall, there is some evidence indicating that during the childhood years, there are specific types of externalising problems that are more prevalent than others in individuals with DS. Dykens et al. (2002) investigated externalising behaviours in 211 individuals with DS aged between 4- and 19-years. Behaviour problems were measured using the CBCL. Across all participants, parents of 68% of the sample endorsed the items ‘Argues a Lot’, 56% ‘Demands Attention’ and 68% ‘Cannot Concentrate’. Only 18% of the sample was found to swear. Externalising problems associated with being argumentative/defiant and poor attention have been found in other studies. Nicham et al. (2003) examined specific behaviour problems in 40 participants with DS. The age of the sample varied considerably ranging from 5- to 31-years. Behaviour problems were measured using the Clinical Assessment Scale of Child and Adolescent Psychopathology (CASCAP; Döpfner et al., 1999) interview schedule. The main types of externalising problems found in the sample were problems with attention, which presented in 76% of the sample and impulsive
behaviour found in 57% of the sample. Finally, Turner and Sloper (1996) found that 94% of their sample had ‘Disturbed Behaviour’ such as ‘runs away’, ‘shouting and screaming’ and ‘deliberately tells lies’, and 41% were identified to have over-activity as measured using the Behaviour Problems Questionnaire (Byrne et al., 1988; Sloper et al., 1991; Sloper & Turner, 1993; Turner & Sloper, 1996).

Not all externalising problems appear to be common in people with DS. One common finding across studies is that the number of children and adolescents with aggressive or anti-social behaviour is low. Van Gameren-Oosterom et al.’s (2011) examined specific types of externalising problems in 320 children with DS who were aged between 8- and 9-years. Less than 5% of children were identified with delinquent or aggressive behaviour. Similar results were observed by Gath and Gumley (1986) who examined behaviour problems in 193 children and adolescents with DS. In this study aggressive behaviour was observed in 11% of the sample. Also, Dykens and Kasari (1997) found that 6% of children with DS stole from home.

There is evidence to suggest that some externalising problems may decline as children with DS become older. Age related changes in behaviour problems were studied by Dykens et al. (2002) and Nicham et al. (2003). In the study by Dykens et al. (2002) behaviour problems were studied in groups of participants aged from mid-childhood (4- to 6-years) to late adolescence to early adulthood (14-19 years). In the oldest participants, significantly lower levels of argumentative and demanding behaviour were found. Similarly, Nicham et al. (2003) examined behaviour problems in one group of children with DS aged between 5- and 10-years and another group aged older than 10-years. In the older participants, fewer instances of dominant, opposing/refusing and inattentive
behaviour were observed. There is some evidence that externalising problems may decrease as children with DS become older.

3.4.2 Summary of Findings for Externalising Behaviour Problems

The research reviewed in this section indicates that individuals with DS have fewer externalising problems than is reported for behaviour problems in general. Studies undertaken with larger samples (n > 193) report that up to 15% of participants have externalising difficulties when examining scale or composite scores, or the number of individuals with externalising disorders. Closer inspection of study results reveal that individuals with DS appear to be more likely to present with defiant/argumentative and related externalising problems. Externalising problems that can be considered anti-social behaviour such as aggression have been reported to be low. Since DS only appears to be related to a subset of externalising problems, this might be one reason why there are fewer problems when this composite score is compared with scores for general behavioural problems. Specifically, individuals with DS do not score high on all externalising items. Another trend in the literature is that as children become older, the number of individuals exhibiting externalising behaviour problems appears to decrease.

3.4.3 Internalising Problems in Down Syndrome

The next section examines internalising problems in children and adolescents with DS. Table 5 presents a summary of studies and their results, of literature that has examined this aspect of behaviour problems in individuals with DS. This section continues by reviewing the literature that has examined internalising problems using a composite measure. Following this, studies that have examined specific types of internalising problems in persons with DS are presented.
Studies investigating internalising problems in children and adolescents with DS using composite measures have produced mixed findings. Studies have reported that 11% to 14% of children or adolescents have internalising problems. Studies by Burke et al. (2012) and van Gameren-Oosterom and colleagues (2013; 2011) have found that at least 1 in 10 children or adolescents with DS have an internalising behaviour problem. The studies by van Gameren-Oosterom, et al., (2011) and van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) examined internalising behaviour problems in 320 and 322 children and adolescents with DS. The composite measure of internalising problem was computed using the CBCL. In the study by van Gameren-Oosterom, et al., (2011) the mean age of the participants was 8.1 years. The percentage of children with internalising problems was 11%. In the study by van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) participants were in their late adolescence and early adulthood. Specifically, the mean age of the sample was 18.3 years. The percentage of participants with internalising problems was found to be 14%.

One study has found that only 5% of children with DS have internalising problems. In the study by Fidler et al. (2000) the presence of internalising problems were investigated in 20 children with DS. The mean age of the sample was 5.8 years ($SD = 1.9$ years). The Internalising Problems Scale from the CBCL was used to determine how many children in the sample presented with this type of behaviour problem. Results showed 5% of the sample had internalising problems.

The age of the participants may also account for inconsistent findings with respect to internalising problems in individuals with DS. Closer inspection of the literature indicates that studies undertaken with older participants are more likely to find more participants with internalising problems. The participants in studies that
have found at least 10% of participants have internalising problems have been conducted with either young adults (van Gameren-Oosterom, Fekkes, van Wouwe, et al., 2013) or adolescents (Burke et al., 2012). In contrast, the study which found 5% of their sample had internalising problems was undertaken with children where the mean age was 5.8 years. It might be that children are able to better communicate symptoms of anxiety or low mood to their caregivers as they become older and their language skills improve. Thus, when parents or guardians are completing behavioural checklists, they are more aware of the internalising symptoms of older children and adolescents with DS. However, it is also possible that internalising problems begin to increase somewhere between 8- and 9-years of age. The study by van Gameren-Oosterom et al. (2011) observed that 11% of their sample who were aged between 8- and 9-years have clinically significant internalising problems. Thus, unlike externalising problems, the limited evidence available suggests that internalising problems may increase with age.
Behaviour Problems in Down Syndrome

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Internalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Burke et al. (2012)</td>
<td>42</td>
<td>38</td>
<td>15.1 2.8 11 - 21</td>
<td>Child Behavior Checklist 13 Internalising composite</td>
</tr>
<tr>
<td>Cuskeley and Dadds (1992)</td>
<td>21</td>
<td>43</td>
<td>NR NR 4 - 15</td>
<td>Revised Behaviour Problem Checklist 19 Anxiety/withdrawal</td>
</tr>
<tr>
<td>Dykens et al. (2002)</td>
<td>211</td>
<td>44</td>
<td>9.7 3.9 4 - 19</td>
<td>Child Behavior Checklist* 15-59 'Underactive' 28-66 'Prefers to be alone' 0-35 'Secretive'</td>
</tr>
<tr>
<td>Dykens and Kasari (1997)</td>
<td>43</td>
<td>65</td>
<td>11.0 NR 4 - 19</td>
<td>Child Behavior Checklist* 32 'Compulsions' 39 'Mood changes' 11 'Excessive sleep'</td>
</tr>
<tr>
<td>Fidler et al. (2000)</td>
<td>20</td>
<td>40</td>
<td>5.8 1.9 3 - 10</td>
<td>Child Behavior Checklist 5 Internalising composite</td>
</tr>
<tr>
<td>Gath and Gumley (1986)</td>
<td>193</td>
<td>51</td>
<td>NR NR 6 - 17</td>
<td>ICD-9 Psychiatric Diagnoses 5 Mixed disturbance of conduct and emotions 3 Disturbance of emotions specific to childhood and adolescence - other or mixed</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology; SDQ = Strengths and Difficulties Questionnaire.
*% refer to individual items on the CBCL (scales not reported). *CASCAP data available for 40 of the 43 participants. Data available for 5-10 year olds (n = 22) and 11-30 year olds (n = 18). *Data available for 5-10 year olds (n = 14) and 10-16 year olds (n = 10). *Data only available for children with behaviour problems at Time 1.
Table 5. Background Study Characteristics and Proportion of Types of Internalising Problems in Individuals with Down Syndrome (continued)

<table>
<thead>
<tr>
<th>Study</th>
<th>Sample Size</th>
<th>Age</th>
<th>Measure(s) of behaviour problems</th>
<th>% DS sample with Internalising Problems and Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myers and Pueschel (1991)</td>
<td>261</td>
<td>43</td>
<td>9.5</td>
<td>5.4</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.5 Phobias</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0 Conversion/Affective disorder</td>
</tr>
<tr>
<td>Nicham et al. (2003)</td>
<td>43</td>
<td>49</td>
<td>13.1</td>
<td>7.3</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>59-50 Anxious</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>9-17 Depression/sadness</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0-39 Reduced self-confidence</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0-44 Decreased motor activity</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>5-44 Shy/insecure</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>7-20 Emotional symptoms</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Turner and Sloper (1996)</td>
<td>91</td>
<td>40</td>
<td>9.2</td>
<td>NR</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>88 Fears and worries(^{d})</td>
</tr>
<tr>
<td>van Gameren-Oosterom et al. (2011)</td>
<td>320</td>
<td>47</td>
<td>8.1</td>
<td>0.2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>11 Internalising composite</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0.3 Anxious/depressed</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>6 Withdrawn</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>7 Somatic complaints</td>
</tr>
<tr>
<td>van Gameren-Oosterom, Fekkes, van</td>
<td>322</td>
<td>47</td>
<td>18.3</td>
<td>0.8</td>
</tr>
<tr>
<td>Wouwe, et al. (2013)</td>
<td></td>
<td></td>
<td></td>
<td>14 Internalising composite</td>
</tr>
</tbody>
</table>

Abbreviations. NR = Not reported; CASCAP = Clinical Assessment Scale for Child and Adolescent Psychopathology; SDQ = Strengths and Difficulties Questionnaire.

\(^{a}\) % refer to individual items on the CBCL (scales not reported). \(^{b}\) CASCAP data available for 40 of the 43 participants. Data available for 5-10 year olds (n = 22) and 11-30 year olds (n = 18). \(^{c}\) Data available for 5-10 year olds (n = 14) and 10-16 year olds (n = 10). \(^{d}\) Data only available for children with behaviour problems at Time 1.
Research investigating specific types of internalising behavioural problems in persons with DS suggests a tendency for these individuals to become withdrawn. This is evident in the studies by Dykens and Kasari (1997) and Dykens et al. (2002). In these studies 49% to 60% of the individuals with DS were identified as preferring to be alone. Additionally, studies have found lower levels of motor activity and increased levels of shyness and insecurity (Nicham et al., 2003). The tendency to have ‘fears and worries’ may be present in an overwhelming majority of individuals with DS. Turner and Sloper (1996) examined behavioural problems in 91 children and adolescents with DS who were aged between 7- and 14-years. The mean age of the sample was 9.2 years. In this study the Behaviour Problems Questionnaire (Byrne et al., 1988; Sloper et al., 1991; Sloper & Turner, 1993; Turner & Sloper, 1996) was used to measure specific internalising problems. Using this instrument 88% of the sample were found to have fears and worries, such as ‘fears situations, animals or objects’.

There is evidence that specific internalising problems may increase as children with DS become older. In the study by Dykens et al. (2002) significantly more adolescents and young adults with DS were found to exhibit withdrawn behaviour compared with children. Nicham et al. (2003) found that more children with DS over the age of 10-years presented with low self-confidence and decreased motor activity compared with those under 10-years. Finally, the lowest levels of specific internalising problems were observed by van Gameren-Oosterom et al. (2011). Participants in this study were aged around 8-years and less than 10% of the sample were found to be withdrawn.
3.4.4 Summary of Findings for Internalising Behaviour Problems

Taken together, evidence suggests internalising problems are present in up to 10% of children with DS, but become more prevalent over time. The increase in internalising problems over time appears to reflect an increase with withdrawn behaviour and a reduction in motor activity.

3.5 Conclusions

This chapter reviewed research that had examined how common behaviour problems are in individuals with DS. One clear finding to emerge from this literature is behaviour problems are common in this group. Depending on the study and methodology, large-scale studies have reported between 17% and 38% of individuals with DS present with one or more types of behaviour problems. There is also evidence to suggest that individuals with DS present with externalising and/or internalising symptoms depending on age. Children are more likely to present with externalising problems, and adolescents and young adults with internalising problems.
CHAPTER 4: THE UNDERLYING NATURE OF BEHAVIOUR PROBLEMS

The previous chapter indicates that behaviour problems are common in children and adolescents with DS. This chapter examines the underlying nature of the behavioural problems. Specifically, whether the behaviour problems in individuals with DS are also commonly found in typically developing (TD) children or others with a developmental disorder. This has been achieved by comparing the behaviour problems of children and adolescents with DS to groups comprising TD children of similar chronological age (e.g., Coe et al., 1999; Guralnick, Connor, & Johnson, 2009; van Gameren-Oosterom et al., 2011). Another approach involves comparing children with DS with a group of TD children who are of comparable mental age (MA) or general cognitive ability. Using this method children with DS are usually matched to a TD child based on raw scores from an IQ test. In these studies, children with DS are typically compared with children who are 2.4- to 4.8-years younger (e.g., Glenn & Cunningham, 2007; Guralnick et al., 2009). Another approach has been to compare children with DS to another group of children with developmental disorders characterised by low IQ (e.g., Dykens & Kasari, 1997; Fidler et al., 2000; Griffith, Hastings, Nash, & Hill, 2010). These comparisons are informative because they provide information about the potential cause of the behaviour problems. That is, whether the problems are related to low IQ or the presence of a developmental disorder.

4.1 Comparisons between Children with Down Syndrome and their Typically Developing Peers

This section reviews studies that have compared behaviour problems of children with DS with TD children. Firstly, research is reviewed that has compared DS and TD groups of comparable chronological age (CA). The second section
Behaviour Problems in Down Syndrome

examines studies that have used matched DS and TD groups on the basis of IQ or MA.

4.1.1 Behavioural Problems in Down Syndrome: Comparisons with Typically Developing Children of Similar Chronological Age

A well replicated finding in the literature is that children with DS have more behavioural problems when compared to TD children who are of the same age (e.g., Guralnick et al., 2009; Pueschel, Bernier, & Pezzullo, 1991; van Gameren-Oosterom et al., 2011). Van Gameren-Oosterom et al. (2011) compared 320 children with DS to 661 TD children. The average age of the participants was approximately 8-years. One analysis examined differences between the groups on a measure of general behavioural problems. The measure used was the Total Problems Score from the CBCL. This score provides an overall measure of problem behaviours and includes externalising and internalising difficulties, as well as social, thought, and attention problems. On this measure, the average Total Problem Score for the DS group was found to be significantly higher than the TD group.

Comparable results have also been reported in studies with smaller samples. Guralnick et al. (2009) compared the behavioural problems of 27 children with DS and 27 TD children of comparable age. The mean age of the participants was around 5.5 years. The Total Problems Score from the CBCL was used to measure any type of problem behaviour. Analyses showed that children with DS scored significantly higher on CBCL Total Problems Score compared with the TD children. Similarly, Pueschel et al. (1991) examined behaviour problems in 40 children and adolescents with DS and 34 TD controls of comparable age. Participants were between the ages of 4- and 16-years old. Again, the CBCL was
Behaviour Problems in Down Syndrome

used as a measure of general problem behaviour. The analyses showed the DS group scored significantly higher on the CBCL Total Problems Score.

Interestingly, research investigating differences between children with DS and age-matched controls on measures of externalising behaviour problems report fewer or smaller differences (e.g., Coe et al., 1999; Pueschel et al., 1991; van Gameren-Oosterom et al., 2011). In the study by van Gameren-Oosterom et al. (2011) externalising problems were compared for DS and TD groups of comparable age. Participants were 320 children with DS and 661 TD-CA controls (the mean age of the sample was around 8-years). The Externalising Problems Scale from the CBCL was used as the dependent variable. On this measure, even though the DS group had significantly more externalising problems than the TD age-matched controls, the effect size for the comparison was small. In another study by van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) no significant differences between individuals with DS and age-matched controls were found on a measure of externalising behaviour problems. In this study externalising behaviour problems of 322 individuals with DS were compared with 2076 TD individuals of comparable age. The mean age of the sample was around 18-years. The CBCL Externalising Problems Scale was also used as the measure of externalising behavioural problems in this study.

It is possible that DS and TD children only differ on specific types of externalising behaviours. Consistent with this suggestion several studies have found individuals with DS have more aggressive, delinquent, attention, hyperactive and conduct behavioural problems than TD-CA controls (e.g., Laws, Taylor, Bennie, & Buckley, 1996; van Gameren-Oosterom et al., 2011). Also, other studies have not found significant differences in aggression and delinquency between TD and DS
groups (e.g., Pueschel et al., 1991; van Gameren-Oosterom, Fekkes, van Wouwe, et al., 2013). For example, Coe et al. (1999) compared a group of 44 children with DS with 44 TD children closely matched on CA, gender and socioeconomic status. Participants in both groups had mean ages of 9.7 years. Externalising behaviour problems were measured using the Revised Behavior Problem Checklist (Quay & Peterson, 1983; Quay & Peterson, 1987), parent and teacher report forms. Four scales on this standardised instrument measure types of externalising behaviour problems. These were conduct problems, aggression, hyperactive behaviour and attention deficits. The results showed that children with DS were rated as having significantly more attention and conduct behaviour problems. However, no significant differences were found between groups on the aggression subscale.

Similarly, Gau et al. (2008) examined the behaviour problems of 45 individuals with DS between 2- and 14-years old ($M = 7.8; SD = 3.1$ years) using the CBCL. A comparison group of 50 TD children was included and had a mean age of 8.4 years. Participants with DS had significantly higher scores than the control group on the Attention Problems scale, but had a slightly lower mean T-score on Aggressive Behaviour, albeit a non-significant difference.

The magnitude of the difference between DS and TD children on internalising measures may be related to the participant’s age. In the study by van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) adolescents and young adults with DS were found to have significantly more internalising behaviour problems compared with an age-matched control group. The measure of internalising problems was a composite index from the CBCL. However, van Gameren-Oosterom et al. (2011) did not find significant differences when using the same composite measure of internalising behaviour problems in 8 year-old children with and without DS. It is also possible that DS is associated with some but not all types of
internalising behaviour problems. Van Gameren-Oosterom et al. (2011) found children with DS had higher levels of withdrawal behaviours and somatic complaints compared to TD children of comparable age. Interestingly, the DS group had significantly lower levels of depression and anxiety. A similar result was also reported in the smaller study by Gau et al. (2008). Results on the CBCL showed that participants with DS ($M = 7.8; SD = 3.1$ years old) had significantly higher scores on the Somatic Complaints and Withdrawn scales compared with a TD control group ($M = 8.4; SD = 2.8$ years old). However, there was a non-significant group difference on the Anxious/Depressed scale, with the DS group having a lower mean score on this scale than the TD control group.

4.1.2 Behavioural Problems in Down Syndrome: Comparisons with Typically Developing Children of Similar Mental Age

The next section reviews research examining behaviour problems in individuals with DS and TD children of comparable intellectual functioning or MA. As noted earlier, this match is typically achieved by pairing a child with DS to a TD child on the basis of raw score from an IQ test. This comparison is potentially informative because it provides information about the role of IQ in the behaviour problems.

Few studies have been undertaken that have compared behavioural problems in persons with DS and MA controls. However, the results from research undertaken so far suggest that individuals with DS have more behaviour problems compared to TD children of comparable mental ability. In one study, Guralnick et al. (2009) compared the behaviour problems of children with DS and TD participants matched on MA. The Total Problems Score from the CBCL was used to measure overall problem behaviour. In this study, 27 children with DS were compared with
27 MA controls. The mean chronological ages of participants were 5.6 ($SD = 0.6$) and 3.2 ($SD = 0.4$) years for the DS and MA controls respectively. Children in the control group were individually matched to a child with DS based on their MA, ± 3 months using the Stanford-Binet Intelligence Scales. In this study, children in the DS group were found to have significantly more problems than TD-MA participants. Similar results were observed by Glenn and Cunningham (2007). In this study, behaviour problems of children and adolescents with DS were compared to a MA comparison group. Groups were matched using the British Picture Vocabulary Scales, a version of the Peabody Picture Vocabulary Test (Dunn & Dunn, 1981) which correlates with IQ (Childers & Durham, 1994; Hodapp & Gerken, 1999; Smith, Smith, & Dobbs, 1991). The DS and TD-MA control groups had mean ages of 13.3 ($SD = 4.9$) and 4.3 ($SD = 1.2$) years respectively. The CBCL Total Problems Score was used to measure problem behaviour. In this study the results indicated that the DS group had significantly more behaviour problems in comparison with MA controls.

Overall, it seems that the nature of behaviour problems in people with DS are different to those seen in typical development. In the first instance children and adolescents with DS appear to have generally more behavioural problems than is expected for their age. Also, older children and adolescents with DS generally present with more internalising problems for their age.

4.2 Comparisons between Down Syndrome and Other Clinical Groups

Research has also been undertaken investigating whether the behavioural problems in children and adolescents with DS are found in other types of developmental disorders. In this area of research, one approach has been to compare behaviour problems of individuals with DS to groups comprising individuals with an
intellectual disability (but not DS). Another approach has been to compare
behaviour problems in individuals with DS to a group of individuals with other types
of neurodevelopmental disorders. Comparisons between children with DS and other
types of developmental disorders provide useful information about the underlying
nature of behaviour problems. Specifically, whether the behaviour problems
observed in individuals with DS are typical (or atypical) for a developmental
disorder. This section reviews these studies.

4.2.1 Behavioural Problems in Down Syndrome and Intellectual Disability

Research has been conducted investigating the behavioural problems of
people with DS to those individuals with an intellectual disability (ID) of unknown
origin. In this literature, it is typically observed that children and adolescents have
significantly less behaviour problems than those of a comparable CA with an ID. A
study by Stores, Stores, Fellows, and Buckley (1998) examined the total behaviour
problems of 91 individuals with DS and 71 individuals with an ID from other causes.
The mean age of participants in the study was around 10.5 years. Behaviour
problems were assessed with the Aberrant Behavior Checklist (Aman & Singh,
1986). The total score on this instrument includes the sum of items measuring
irritability, lethargy and withdrawal, hyperactivity, repetitive behaviour and
inappropriate or repetitive speech. In this study, the DS group had significantly
fewer behaviour problems compared with the ID group.

Interestingly, differences in behavioural problems between individuals
with DS and an ID have not been observed after statistically controlling for
individual differences in intellectual functioning. Dykens and Kasari (1997)
examined the behavioural problems in 43 individuals DS and an equal number of
individuals with non-specific ID on the Total Problems Score from the CBCL.
Participants were aged 11-years (range: 4-19 years) and IQ was used as a covariate in all comparisons between groups. The mean FSIQ for the DS group was 48 and 56 for the ID group. In this study there were no significant differences between DS and ID groups on the measure of behaviour problems.

Finally, research has found that children with DS have fewer or, comparable levels of externalising problems as children with an ID (e.g., Dykens & Kasari, 1997; Grizenko, Cvejic, Vida, & Sayegh, 1991; Hodapp, Ricci, Ly, & Fidler, 2003). In the study by Dykens and Kasari (1997) 43 individuals with DS and non-specific ID were compared on the Externalising Problems Scale from the CBCL. Participants in both groups had a mean age of 11-years (range: 4-19 years). No significant differences were found between the two groups on externalising behavioural problems.

In the domain of internalising problems, individuals with DS have been reported to have similar or lower levels of internalising problems when compared to groups comprising children with an ID. Hodapp et al. (2003) compared the behavioural problems of 27 children with DS and 15 children with an ID from unknown or other causes. Participants in both groups had mean ages of around 8-years (range: 3-12 years). The CBCL Internalising Problems Scale was used to measure overall problems with depression, anxiety, withdrawal and somatic complaints. The analyses revealed that children with DS had significantly fewer and/or less severe internalising problems than children in the ID group.
4.2.2 Behavioural Problems in Down Syndrome: Comparisons with Children with Neurodevelopmental Disorders

Another series of investigations has compared behaviour problems of children and adolescents with DS to those with other types of developmental disorders. To date, behavioural problems in individuals with DS have been compared with Williams and Smith-Magenis syndromes (Fidler et al., 2000), Prader-Willi syndrome (Dykens & Kasari, 1997) and Autism (Griffith et al., 2010; Grizenko et al., 1991). In these comparisons the children with DS were found to have significantly fewer behavioural problems than the comparison groups.

The study by Fidler et al. (2000) compared behaviour problems of 20 children with DS and 20 with Williams and Smith-Magenis syndromes. Participants in the study were aged around just under 6-years. Children with Williams syndrome show physical characteristics such as full lips and cheeks, puffiness around eyes and short stature. Smith-Magenis syndrome is characterised by brachycephaly (i.e., disproportionately wide head), cleft palate, prominent chin, mid-facial hypoplasia and short stature. Common to Williams and Smith-Magenis syndromes is that they are genetic disorders characterised by ID (Batshaw, 2002). Behavioural problems were assessed using the Total Problems Score from the CBCL. Results showed that the DS group had significantly fewer behaviour problems compared with the children with Williams and Smith-Magenis syndromes. Additional analyses showed there were significantly fewer children with DS who had clinically significant behaviour problems compared with the other developmental disorders studied. Specifically, 80% and 75% of the children with Smith-Magenis syndrome and Williams syndrome had clinically significant levels of behaviour problems respectively. In comparison only 40% of the children with DS had clinically significant behaviour problems.
Dykens and Kasari (1997) compared behavioural problems in individuals with DS and Prader-Willi syndrome. Characteristics of Prader-Willi syndrome include short stature, decreased muscle tone, small hands and feet, hypogonadism and ID (Batshaw, 2002). Participants had an average age of 11-years. However the age range of the participants was wide, varying from 4- to 19-years. After controlling for IQ, participants with DS had significantly fewer behaviour problems compared to the other study group. Furthermore, fewer of the participants with DS had clinically significant behaviour problems compared to the other group. In this study 23% of the participants with DS were found to have clinically significant problems compared to 72% with Prader-Willi syndrome.

With respect to specific types of behaviour problems, evidence suggests DS is associated with both fewer externalising and internalising problems compared with other developmental disorders (e.g., Dykens & Kasari, 1997; Griffith et al., 2010; Grizenko et al., 1991). For instance, in the study by Fidler et al. (2000) children with DS were compared to those diagnosed with Williams and Smith-Magenis syndromes. Externalising behaviour was measured using a composite score from the CBCL. On the externalising composite measure, the DS group were found to have significantly less/fewer externalising problems compared with participants with Williams and Smith-Magenis syndromes. This same pattern of results was observed on the internalising composite measure as well.

Grizenko et al. (1991) examined externalising and internalising behaviour problems in 40 individuals with DS and 136 participants with Autism Spectrum Disorder (ASD). The sample comprised children, adolescents and adults with the age of participants ranging from 11- to 58-years. The Revised Child Behavior Profile (Achenbach & Edelbrock, 1983) was used to measure externalising
and internalising problems. Overall, individuals with DS demonstrated significantly lower levels of externalising behaviour compared to the ASD group. On the internalising measure, overall there were no differences between the groups.

4.3 Summary

This chapter reviewed studies that compared behaviour problems in children and adolescents with DS with TD individuals and also those with different types of developmental disorders. Comparisons between individuals with DS and TD children and adolescents revealed a consistent trend. This was that individuals with DS have more behaviour problems compared with TD children of comparable chronological or mental age. Thus it seems that the behaviour problems observed in people with DS do not seem to form part of the typical developmental trajectory. At the same time, it does not appear to be the case that behaviour problems in children and adolescents with DS mirror those observed in other developmental disorders. The behaviour problems in people with DS appear to be less severe or less frequent compared with other developmental disorders such as Prader-Willi syndrome and ASD. This might suggest that aspects of the behaviour problems are unique to individuals with DS. This suggestion is forwarded on the basis that children with DS display greater problems than TD children matched on mental age, yet have fewer behaviour problems than other children with developmental disorders.
CHAPTER 5: CORRELATES OF BEHAVIOUR PROBLEMS IN DOWN SYNDROME

The review of studies investigating behaviour problems so far indicates children and adolescents with DS have severe and/or frequent problems for their chronological or mental age (e.g., Glenn & Cunningham, 2007; Guralnick et al., 2009; van Gameren-Oosterom et al., 2011). However, it is also noted that DS is characterised by fewer behaviour problems compared with other types of developmental disorders and impairments (e.g., Dykens & Kasari, 1997; Fidler et al., 2000; Griffith et al., 2010). The pattern of results indicates there may be something unique to DS influencing or contributing to their behaviour problems. This chapter examines whether cognitive and neuropsychological problems, and/or maturation in people with DS may be related to the behaviour problems in this group. In particular, this chapter will explore the ways in which the behaviour problems of children and adolescents with DS might be affected by intellectual ability or IQ, emotion recognition, theory of mind, language and pragmatic language skills, social skills, neurodegeneration associated with Alzheimer’s disease or dementia and general maturation.

This chapter focuses on whether ‘within-child’ variables might be related to some of the variation of behaviour problems observed in children and adolescents with DS. There are many other variables external to the child that might also impact on their behavioural patterns. It was not possible to examine all of these potential variables within the one study. Therefore, the decision was made to focus on whether specific cognitive, social or neuropsychological problems might relate to behavioural difficulties for people with DS.
5.1 IQ and Behaviour Problems

Intellectual impairment co-occurs with DS (Sherman, Allen, Bean, & Freeman, 2007; Visootsak & Sherman, 2007). However, it has been found that the severity of the intellectual impairment in people with DS varies considerably between individuals (e.g., Carr, 1994; van Gameren-Oosterom, Fekkes, van Wouwe, et al., 2013; Visootsak & Sherman, 2007). For example, a recent study by Costanzo et al. (2013) measured the IQs of 15 individuals with DS between 8- and 21-years old ($M = 14.5$, $SD = 3.7$ years). Large variability was found between IQ scores on two standardised intelligence measures, with the mean IQ equalling 53 and a standard deviation of 13.5 points (range: 36-83). The variability in IQ may be related to variability in behavioural problems in individuals with DS. In this section a model implicating low IQ in behavioural problems is presented with respect to DS.

5.1.1 How Does Low IQ Underlie Behaviour Problems?

It has been proposed that intellectual impairments underlie or contribute to behavioural problems in childhood (e.g., Einfeld & Tonge, 1996; Goodman, Simonoff, & Stevenson, 1995; Lynam, Moffitt, & Stouthamer-Loeber, 1993). Moffitt (1993) proposed that low IQ might directly lead to the presence of behavioural problems. According to the theory, lower levels of intellectual functioning limit the extent to which an individual can moderate his/her own behaviour. This argument is forwarded on the basis that intellectual impairment reflects the presence of executive dysfunction, poor verbal reasoning and poor problem solving skills. One outcome of this collection of cognitive deficits are difficulties negotiating social interactions. For instance, if a child has poor executive functioning, the child might have difficulty controlling his/her own behaviour when upset or do not have his/her needs met. Moffitt noted that executive functioning is associated with a range of skills such as
maintaining concentration, abstract reasoning, self-awareness and the ability to plan
behaviour. According to this model, children with DS who have a lower IQ are more
likely to have more behaviour problems.

Intellectual impairment may indirectly impact on behavioural problems
as well (e.g., Goodman et al., 1995; Hirschi, 1969; Lynam et al., 1993). According
to Lynam et al. (1993), lower IQ is viewed as an index for the likelihood a child will
experience academic success. Presumably a child with lower IQ may experience
more difficulties dealing with and solving academic and social challenges. It is
proposed that repeatedly failing in social and academic settings may lead to
increased levels of frustration (Goodman et al., 1995). As a consequence of this
frustration the child might be more likely to engage in rule-breaking or aggressive
behaviour that oppose the values advocated by their school (Hirschi, 1969; Lynam et
al., 1993). According to this perspective children with DS who have lower IQ scores
may be more likely to have externalising behavioural problems.

It is also possible that low IQ may lead to internalising problems as well.
Goodman et al. (1995) suggest that individual’s with lower IQ will experience more
difficulties completing academic and non-academic activities in school. The
continual struggle and failure in these tasks may lead a child to be in an ongoing state
of hopelessness and experience lower levels of self-esteem. The demands placed on
children at school might also cause considerably more anxiety or emotional distress
for individuals with lower IQs, where completing academic tasks is substantially
more difficult.
5.1.2 The Relationship between IQ and Behaviour Problems in Down Syndrome

Even though intellectual impairment is a common feature in people with DS, relatively few studies have examined whether this impairment is related to behaviour problems. Evidence supporting the proposal that IQ is related to behaviour problems is found from the studies reviewed earlier comparing children with DS and those with an ID (Dykens & Kasari, 1997; Griffith et al., 2010). The study by Dykens and Kasari (1997) found no difference in behaviour problems between these groups after statistically controlling for differences in intellectual ability. That is, once group differences related to intellectual ability were removed, differences in behavioural problems were no longer observed.

However, there is also evidence that questions whether IQ is associated with the behavioural problems of people with DS. Dykens et al. (2002) measured IQ using the Kaufman Brief Intelligence Test (Kaufman & Kaufman, 1990) or Stanford–Binet Intelligence Test (Thorndike, Hagen, & Sattler, 1986) for 211 individuals with DS between 4- and 19-years old. No significant correlations were found between overall IQ and externalising, internalising or total problems on the CBCL. Dykens and Kasari (1997) also found no significant correlation between IQ and behaviour problems in 43 individuals aged 4- to 19-years old.

Overall, it seems the evidence implicating variability in IQ to behavioural problems in individuals with DS is weak. When IQ has been measured using a well validated individual assessment instrument no association has been found (Dykens & Kasari, 1997; Dykens et al., 2002). However, in these studies reporting no association, the age range of the participants was wide. This might be a problem if the association between age and IQ decreases as children with DS become older.
Thus it could be that an association between IQ and behaviour problems in people with DS exists, but it is only present in children.

5.2 Emotion Recognition and Behaviour Problems

Emotion recognition refers to being able to accurately detect and label emotional cues (Izard et al., 2001). An example of emotion recognition is the ability to recognise or interpret an emotional state, such as happiness from facial expression (Carr & Lutjemeier, 2005). Deficits in emotion recognition have been proposed to contribute to externalising and internalising problems (e.g., Larkin, Jahoda, & MacMahon, 2013; Rojahn, Rabold, & Schneider, 1995; Walker, 1981). There is also evidence that individuals with DS have difficulties with recognising facial expressions (Kasari, Freeman, & Hughes, 2001; Williams, Wishart, Pitcairn, & Willis, 2005). This section reviews theoretical and empirical evidence that poor emotion recognition might be one contributing factor to behavioural problems for persons with DS.

5.2.1 How Does Poor Emotion Recognition Underlie Behaviour Problems?

Deficits in emotion recognition have been proposed to lead to the development of externalising and internalising problems (e.g., Walker, 1981; Walz & Benson, 1996). According to Walker (1981) difficulties recognising another individual’s emotions makes interpersonal interactions less predictable. This may lead to problems identifying social cues necessary for effective communication.

Another theory implicating emotion recognition deficits in behaviour problems was presented by Rojahn et al. (1995). According to the emotion specificity hypothesis, people with an ID have an impairment in the ability to recognise emotion based on facial expressions. This impairment can lead to
behavioural problems. Difficulties with recognising emotion from facial expression may result in problems understanding others’ intentions. Larkin et al. (2013) suggested that emotion recognition difficulties might reduce sensitivity to emotional cues or lead to more difficulties empathising with another person’s point of view. This may predispose an individual to misinterpretation of others intentions, thereby increasing the chance of encountering social conflict. More exposure to social conflict is thought to also increase the likelihood of aggressive behavioural responses.

5.2.2 Evidence Linking Emotion Recognition and Behaviour Problems in DS

No study known to the author has examined whether an association exists between emotion recognition impairments and behaviour problems in people with DS. However, there is evidence suggesting emotion recognition deficits are present in individuals with DS. Studies by Williams et al. (2005) and Wishart, Cebula, Willis, and Pitcairn (2007) investigated the ability of children and adolescents with DS to recognise six basic emotions (i.e., happiness, sadness, anger, fear, disgust and surprise). Emotion recognition was assessed using a pictorial emotion-matching task. Participants were shown a photograph of a facial expression and asked to identify which other photo showed the same emotion (e.g., happy) from three response options. The DS group had poorer performance recognising all six emotions compared to a control group comprising TD children of comparable MA.

Further evidence of an emotion recognition deficit in children with DS was presented by Kasari et al. (2001). In this study 6-year old children with DS were compared to TD children on an emotion recognition task. Emotion recognition was assessed using labelling, recognition and identification tasks. In the labelling task, children were shown puppets with different facial expression. Children were asked
to describe how a puppet was feeling (e.g., angry) based on the facial expression. On the recognition task children were shown four photographs of different facial expressions. The children were asked to identify a specific emotion following the prompt “where is the … (e.g., happy) face?” Finally, on the identification task children were asked to identify how a puppet was feeling in different scenarios (e.g., when a puppet was given ice-cream). Results showed that children with DS performed significantly worse on the labelling, recognition and identification tasks compared to TD children of comparable age. At present it appears that DS is associated with poor emotion recognition abilities. Whether variability in this skill might be contributing to the presentation of behavioural problems in this clinical group has yet to be investigated.

5.3 Theory of Mind and Behaviour Problems

Theory of mind (ToM) is the ability of an individual to infer others’ intentions, feelings, and perspectives, enabling him/her to predict behaviour (Baron-Cohen, Leslie, & Frith, 1985; Buitelaar, Van der Wees, Swaab-Barneveld, & Van Der Gaag, 1999; Stone, Baron-Cohen, & Knight, 1998). This section considers whether ToM deficits in people with DS is related to their behavioural problems. In this section a model implicating ToM deficits in behavioural problems is presented. Then, evidence suggesting individuals with DS have ToM deficits is presented.

5.3.1 How Do Theory of Mind Impairments Underlie Behaviour Problems?

ToM deficits have been proposed to cause externalising and internalising problems (e.g., Blair, 1995; Inoue, Yamada, & Kanba, 2006; Sharp, 2008). Hughes and Leekam (2004) proposed that problems with ToM may lead to externalising and internalising behavioural problems. They suggested that during social interactions ToM is necessary to avoid interpersonal conflict that arises through
misunderstanding others’ intentions and/or emotions. For instance, an individual might observe another child crying, but have difficulty understanding why they are upset and therefore not respond in an appropriate way, eliciting an aggressive response.

ToM problems may also be related to internalising behaviour problems. Hughes and Leekam (2004) suggested that ToM skills might help to increase social harmony amongst peers because it helps to prevent social misunderstandings. This was proposed to decrease the likelihood of the child encountering embarrassing or emotionally distressing social situations. As an example, a child that has difficulties understanding another person’s expectations might be more likely to either withdraw from others, or experience anxiety in social situations due to his/her reduced ability to know the appropriate way to respond. These children might also be at higher risk for being reprimanded or embarrassed by others if they misinterpret other individuals’ intentions or beliefs and thus interact with them in an undesirable manner. This implies that more accurately predicting others’ behaviours might better equip a child to prevent negative social experiences that may increase internalising problems.

5.3.2 Evidence Linking Theory of Mind and Behaviour Problems in Down Syndrome

It is not known whether ToM functioning is correlated with behaviour problems in children and adolescents with DS. However, research has been conducted examining ToM skills in people with DS (e.g., Abbeduto et al., 2001; Baron-Cohen et al., 1985; Giaouri, Alevriadou, & Tsakiridou, 2010; Yirmiya, Solomonica-Levi, Shulman, & Pilowsky, 1996). Studies examining ToM in persons with DS typically report a deficit in this aspect of cognitive functioning (e.g.,
Abbeduto et al., 2001; Giaouri et al., 2010; Yirmiya et al., 1996). Abbeduto et al. (2001) examined the ToM skills of a sample of adolescents and young adults with DS. In this study 25 individuals with DS who were aged between 11- and 23-years and MA controls were presented with a false belief task. False beliefs refer to understanding that others can hold beliefs that are different from reality. To pass a false belief task the participant is required to take on the perspective of a protagonist in a short story. Results showed that the DS group perform poorer on the task compared with MA controls. These findings suggest that the difficulty individuals with DS have with ToM extend beyond limited mental reasoning capacity.

However, not all studies report ToM impairments in individuals with DS. Baron-Cohen et al. (1985) examined ToM in 14 children with DS and TD controls ($n = 27$). The mean age of the children with DS was 10.9 years ($SD = 4.1$ years), and 4.4 years ($SD = 0.6$ years) for the TD participants. The nonverbal and verbal MA of participants with DS was measured by Leiter International Performance Scale (Leiter, 1980) and British Picture Vocabulary Test (Dunn, Dunn, & Whetton, 1982) respectively. ToM was measured using a false belief task similar to the one used by Abbeduto et al. (2001). In this study both groups passed the test with equal levels of proficiency. Thus it could be that ToM impairments are not present in all individuals with DS.

Collectively, past research suggests there might be variability in ToM in people with DS. Some individuals may have ToM impairments, while others may not show deficits. Research has yet to be undertaken to investigate whether variability in ToM functioning might be correlated with behavioural problems in children and adolescents with DS.
5.4 Language and Behaviour Problems

Language skills might also be related to the behaviour problems of individuals with DS. Substantial research exists demonstrating an association between language difficulties and the presence of behaviour problems (e.g., Botting & Conti-Ramsden, 2000; Cantwell & Baker, 1987; Cohen et al., 2000; Lindsay & Dockrell, 2000; Van Daal, Verhoeven, & Van Balkom, 2007). It is well documented that children with DS experience language difficulties, however the extent and severity of problems has been found to vary between individuals (e.g., Chapman, 1997; Chapman, Schwartz, & Kay-Raining Bird, 1991; Chapman et al., 1998; Roberts et al., 2007). This section examines the extent to which language problems might contribute to the behavioural problems in children and adolescents with DS.

For the purpose of this thesis, language problems refer to problems in receptive, expressive and pragmatic language. Receptive language is the ability to understand words and sentences spoken by others. Expressive language refers to the ability to produce grammatically correct sentences that can be understood by others (Bishop, 1997; Rice, 1997). Pragmatic language describes the social, emotional and communicative aspects of social interactions (Adams, Baxendale, Lloyd, & Aldred, 2005; Martin & McDonald, 2003) and includes skills necessary to use and interpret language correctly in relation to its context (Bignell & Cain, 2007; Roberts et al., 2007).

5.4.1 How Do Language Impairments Underlie Behaviour Problems?

Difficulties being able to produce and understand language has been proposed to contribute to externalising and/or internalising problems in children (e.g., Benner, Nelson, & Epstein, 2002; Gallagher, 1999; Moffitt, 1993). According to Benner et al. (2002) poor language might contribute to externalising problems by
limiting a child’s ability to understand and respond appropriately to verbal instruction. When a child fails to comprehend what is being asked of him/her they may appear to be non-compliant with instructions or repeated warnings about their behaviour. This may result in ongoing miscommunication, thereby potentially leading to frustration. Frustration is thought to increase the chance of aggressive behavioural responses.

Poor language might also lead to behaviour problems by disrupting the child’s ability to participate effectively in social interactions. A number of researchers have highlighted that children with poor language and communication skills experience lower levels of peer acceptance (e.g., Durkin & Conti-Ramsden, 2010; Durkin & Conti-Ramsden, 2007; Laws, Bates, Feuerstein, Mason-Apps, & White, 2012; Menting, van Lier, & Koot, 2011). Pragmatic difficulties might also contribute to misinterpretation and inappropriate responses during social discourse. Difficulty meeting others’ communication expectations may put these children at risk of being socially rejected or neglected. Peer rejection is proposed to lead to social isolation, which then may contribute to the development of internalising behaviours. Specifically, social isolation might contribute to internalising problems by lowering self-worth and increasing feelings of loneliness.

5.4.2 Evidence Linking Language and Behaviour Problems in Down Syndrome

The role of poor language skills in behaviour problems has been observed in children with language impairments (e.g., Gallagher, 1999; Lindsay, Dockrell, & Strand, 2007; Toppelberg & Shapiro, 2000; Van Daal et al., 2007). Research has yet to investigate whether there is a relationship between language functioning and behaviour problems in persons with DS. However, substantial evidence has been obtained indicating expressive and receptive language problems
are present in individuals with DS. The presence of language problems in persons with DS was demonstrated by a meta-analysis undertaken by Næss, Lyster, Hulme, and Melby-Lervåg (2011). In this review a total fifteen studies were included that summarised the results from 379 participants with DS and 428 controls. The meta-analysis showed that overall, individuals with DS performed significantly worse than controls on expressive and receptive language measures compared to chorological and mental age comparison groups. Evidence has also been presented showing individuals with DS present with pragmatic language problems. In one study 50% of participants with DS between 10- and 22-years old were rated as having severe pragmatic difficulties (Laws & Bishop, 2004a). Only 17% of participants with DS were found to have language scores that were comparable with a mental age control group. Whether there is a relationship between behaviour problems and language functioning in individuals with DS has yet to be examined.

5.5 Social Skills and Behaviour Problems

Social skills difficulties may also be a contributing factor to the behavioural problems in people with DS. Social skills refer to learned behaviours that are socially acceptable and permit an individual to engage with others in ways that encourage positive responses, while deterring negative responses (Elliott, Racine, & Busse, 1995). Some examples of social skills are the ability to make compromises, listen to others, allow others to join an activity and be kind, polite and respectful toward others.

5.5.1 How Do Social Skill Deficits Underlie Behaviour Problems?

Engels et al. (2002) proposed that social skill deficits may lead to externalising and internalising problems by causing poor peer relationships. Children who have insufficient social skills are suggested to have more difficulties
making friends, and have lower levels of intimacy and involvement in established relationships. It is proposed that children with poor peer experiences may eventually realise that they are disliked. One outcome is reduced self-confidence and self-esteem, feelings of loneliness and depression, and increased feelings of hostility and perceived failure (Hymel, Rubin, Rowden, & LeMare, 1990; Masten, 2005). Along similar lines, Lewinsohn (1975) proposed that individuals with poor social skills were at higher risk for depression because of a lack of positive reinforcement. Specifically, an individual with poorer social skills is likely to elicit lower rates of positive reinforcement from others, which may lead to low mood and depressive symptoms (Lewinsohn, 1975; Segrin, 2000).

There is some evidence to support a role for social skills in behaviour problems. A meta-analysis has shown that improving social skills also reduces internalising and externalising problems. Specifically, Cook and colleagues (2008) reviewed previous meta-analyses of social skills training programs in secondary school students. In total, data from 77 original studies, and approximately 5000 adolescents between 11- and 19-years old were included. The results showed that social skills training led to an improvement/reduction in behaviour problems. Overall, around 65% of adolescents who attended social skills training evidenced a reduction in behaviour problems compared to 35% of participants who received no intervention.

5.5.2 The Relationship between Social Skills and Behaviour Problems in Down syndrome

At present little is known about the association between social skills and externalising and/or internalising problems in individuals with DS. Additionally, there appears to be few studies that have examined social skills in children with DS.
However, a recent investigation by van Gameren-Oosterom, Fekkes, Reijneveld, et al. (2013) indicated that adolescents with DS have impaired social skills. In this study, 322 adolescents with DS between 16- and 20-years old were compared with normative data from 400 adolescents of similar age (supplied in the test manual) on social skills according to the Children’s Social Behaviour Questionnaire (Hartman, Luteijn, Moorlag, De Bildt, & Minderaa, 2007; Hartman, Luteijn, Serra, & Minderaa, 2006; Luteijn, Luteijn, Jackson, Volkmar, & Minderaa, 2000). Results showed that adolescents with DS had significantly more problems with social functioning compared to the control group, and a large effect size was found. Whether a relationship exists between social skills and behaviour problems in children and adolescents with DS has yet to be tested.

5.6 Neurodegeneration and Behaviour Problems

It has been proposed that the behaviour problems of individuals with DS are related to neurodegeneration. According to Dykens and colleagues (1997; 2002) the emergence of internalising problems during adolescence may occur as a consequence of early neuropathological change associated with Alzheimer’s disease (AD).

Neurodegeneration is common in people with DS (Oliver & Holland, 1986; Visser, Aldenkamp, Van Huffelen, & Kuilman, 1997; Wilcock & Griffin, 2013; Zigman, Schupf, Zigman, & Silverman, 1993). It is well-established within the literature that DS is associated with a high risk for developing AD (Ball et al., 2006; Menéndez, 2005; Zigman & Lott, 2007). There is evidence that almost all adults with DS over 40 years old present with neuropathological signs of AD (Iwatsubo, Mann, Odaka, Suzuki, & Ihara, 1995; Mann & Esiri, 1989), however only approximately 50% develop observable symptoms (Zigman et al., 1993). Cognitive
impairment that follows the neurological problems typically includes memory dysfunction, disorientation, diminished daily living skills, reduced speech and difficulties with language comprehension (Haxby, 1989). Behavioural and personality changes are often the earliest signs of AD in individuals with DS, preceding cognitive change (Alyward, Burt, Thorpe, & Dalton, 1997; Ball et al., 2006; Dykens et al., 2002; Holland, Hon, Huppert, & Stevens, 2000). Early symptoms include depression (Burt, Loveland, & Lewis, 1992; Nelson, Orme, Osann, & Lott, 2001), thus Dykens and colleagues (1997; 2002) forwarded that internalising behaviours during adolescence may be caused by neurodegeneration.

The neurodegenerative hypothesis attempts to account for behaviour problems in persons with DS with respect to neurological dysfunction. According to this hypothesis, behaviour problems occur in individuals with DS because of neuropathological changes associated with AD (e.g., accumulation of neuritic plaques and neurofibrillary tangles) which results in functional brain changes (Visootsak & Sherman, 2007). There is some evidence for neuropathological signs of AD existing in children and adolescents with DS (Mehta, Capone, Jewell, & Freedland, 2007); however, it remains unclear whether these neurological factors have a causal role in behaviour problems.

### 5.6.1 Empirical Evidence Linking Neurodegeneration and Behaviour Problems

At present, no known study has investigated whether internalising problems in adolescents with DS is associated with neurodegeneration. Despite this, there is some evidence that children with DS have signs of brain changes associated with AD. AD is caused by the formation of neuritic plaques and neurofibrillary tangles that is linked with accumulating abnormally high levels of amyloid-β protein (A-β; Englund et al., 2007). According to the amyloid cascade hypothesis, greater
accumulation of A-β is due to an overproduction of the protein (Hardy & Higgins, 1992; Teller et al., 1996) which is especially likely in people with DS because A-β is produced by the amyloid precursor protein gene located on chromosome 21.

Children with DS have higher levels of A-β than TD children (e.g., Leverenz & Raskind, 1998; Mehta et al., 2007). Mehta et al. found that 35 children and adolescents with DS ($M=7.2$, $SD=3.8$ years) had significantly higher levels of soluble forms of A-β in their blood than did 34 TD siblings ($M=10.7$, $SD=4.5$ years). It was concluded that the amyloid precursor protein gene does lead to greater production of A-β prior to the development of neuritic plaques. Englund et al. (2007) observed that A-β levels increased in the cerebrospinal fluid of children with DS from 8- to 20-40- and 54-months old. It was suggested that this was indicative of an overproduction of A-β and that A-β levels increase with age.

Leverenz and Raskind (1998) investigated A-β levels in the brains of individuals with DS between 4-days and 38-years old with no history of AD compared with TD controls between 1-day and 41-years old. Neuritic plaques were found in children with DS from 8-years old, but not in some 20 year-olds. No control participant was found to have any neuritic plaques. Thus neuritic plaques are certainly present in some individuals with DS.

5.7 Maturation and Behaviour Problems

It is also possible that the behaviour problems in individuals with DS do not occur as a consequence of cognitive or neurological impairment. Instead, the problems appear as individuals with DS become older and experience the world. That is the behaviour problems in people with DS are related to age. There is some epidemiological evidence that indicates that behavioural problems change over the course of child and adolescent development (e.g., Crijnen, Achenbach, & Verhulst,
1997). Broadly consistent with this view is that evidence shows that as children become older externalising behavioural problems decrease, while internalising problems increase during adolescence (e.g., Dykens & Kasari, 1997; Dykens et al., 2002).

Korhonen et al. (2014) proposed that cognitive maturation might account for changes in behavioural functioning in all groups of children. It was suggested that young children are less able to verbally express their emotions and regulate their own behaviour in comparison with adolescents and adults. Limitations in these skills are thought to contribute to more externalising problems being present in younger children. Thus, as individuals develop these skills the number and/or severity of externalising problems decreases during adolescence. Presumably, internalising problems become more common with maturation as individuals become more practiced at managing their behavioural and emotional responses internally or in ways that do not cause disruption to others. If this proposal is accurate it would be expected that age correlates with behaviour problems rather than a specific cognitive ability or measure or neurological functioning.

5.7.1 Evidence Supporting a Relationship between Maturation and Behaviour Problems.

To the author’s knowledge only two peer-reviewed study has investigated the influence of maturation factors on behaviour problems in children and adolescents with DS. In the study by Turner and Sloper (1996) the behaviour problems of 91 children and adolescents with DS were measured using a longitudinal method. At Time 1 the mean age of the participants was 9.2 years (range: 7-14 years). Behavioural problems were measured using the Behaviour Problems Questionnaire. This provides an overall measure of problems relating to
management of emotional states and difficulties relating to and interacting appropriately with other people or objects. Participants’ behavioural problems were measured again 3- to 4-years later, when the mean age of the sample was 13.8 years. In this study a significant decrease in the number and frequency of total behaviour problems was observed from Time 1 to Time 2.

A small study by Eisenhower et al. (2005) showed that externalising problems significantly increased for children with DS between 3- and 5-years of age. According to the results on the CBCL the total problems T-scores also increased over this developmental period, however no statistically significant change was reported. This might suggest that externalising behaviour problems increase during early childhood, however only 12 participants with DS took part in this study. Therefore these results need to be replicated with larger study groups before any firm conclusions can be drawn with regard to these findings.

There is also cross-sectional evidence suggesting that age might moderate the type of behaviour problems present in an individual with DS. In the study by Dykens et al. (2002) the behavioural problems of 211 children, adolescents and young adults with DS were assessed using the CBCL. Problem behaviours were compared across four groups of individuals varying in age from 4- to 19-years. The results showed that children aged between 10- and 13-years had significantly higher externalising problem compared with 4-6 year-olds. Older children and adolescents were found to have lower levels of externalising problems. With regard to internalising problems, individuals between 10- and 19-years old had significantly more internalising problems than 4-6 year-old children.

Nicham et al. (2003) also investigated behaviour problems in 40 children, adolescents and adults with DS. Participants were aged between 5- and 31-years old
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(M = 13.1, SD = 7.3 years). Behaviour problems were measured with the CASCAP interview schedule (Döpfner et al., 1999) and Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997). Only the SDQ provides a Total Problems score, which is a composite derived from the Emotional Symptoms, Conduct Problems, Hyperactivity/ Inattention and Peer Relationship Problems subscales.

Analyses were conducted on a subgroup of participants aged 5-10 (n = 14) and 10-16 (n = 10) years old. Results showed that the 5-10 year-olds presented with more externalising behaviour problems than the older participants. In particular, 5-10 year-olds were described as having significantly more ‘opposing/refusing’ and ‘dominant’ than 11-31 year-olds. With regard to internalising symptoms, significantly more participants in the 11-31 year-old group were described as shy or insecure than in the younger participant group. All items that related to symptoms of depression, except for the ‘depression/sadness’ symptoms were more common in the older group. These behaviours were ‘reduced self-confidence’, ‘decreased motor activity’, ‘tics, stereotypes, abnormal habits’ and ‘increased food intake’. Thus, this study provides evidence that internalising symptoms may increase in adolescence and adulthood, while some externalising symptoms might be less common with greater maturity.

The idea that age or general maturational factors are related to behaviour problems in people with DS has not always been replicated (e.g., Fidler et al., 2000; Povee et al., 2012; Stores et al., 1998). A study by Fidler et al. (2000) investigated whether a relationship existed between age and Total, Externalising and Internalising Problems on the CBCL. Twenty children with DS with a mean age of 5.8 years participated (range: 3-10 years). The results showed that there was no significant correlation between age and the Total, Externalising or Internalising Problem scores for the children with DS. The reason for inconsistent findings is unclear. One possibility is that to observe a relationship between age and behaviour problems in
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persons with DS, a sample requires adolescents. This is because it is during adolescence past research shows a decline in externalising problems and an increase in internalising problems.
CHAPTER 6: SUMMARY AND HYPOTHESES

At present there are number of factors that may be related to behaviour problems in children and adolescents with DS. In this review potential correlates considered were IQ, emotion recognition, ToM, language, social skills, early neurodegeneration and maturation. Research is required to test whether a relationship exists between each of the aforementioned variables and behaviour problems in individuals with DS. It is also possible that there are multiple causes underlying maladaptive behaviour in this clinical group, and that these may interact with one another to affect the type or severity of the problems.

On the basis of the theoretical and empirical evidence presented thus far, seven hypotheses are proposed relating to each of the cognitive variables.

1. It is predicted that there will be a negative correlation between IQ and behaviour problems.
2. Theory of mind will be negatively correlated with problem behaviours.
3. Poorer emotion recognition will be associated with more behavioural problems.
4. Participants with poorer language and pragmatic skills will have more behaviour problems.
5. Poorer social skills will be associated with more behavioural problems.
6. Poorer performance on neurodegeneration screening tasks will be associated with more internalising behavioural problems in the DS group only.
7. Measures of total and externalising behaviour problems will be negatively correlated with age, while a positive association will be found for internalising problems.
CHAPTER 7: STUDY 1. BEHAVIOUR PROBLEMS IN DOWN SYNDROME

7.1 Introduction

The aim of Study 1 was to examine the relationship between maturational factors and behaviour problems in children and adolescents with DS. An additional aim was to replicate past research to examine the number of children with DS presenting with behaviour problems. The literature review identified that around 25% of children and adolescents with DS present with behaviour problems (e.g., Dykens et al., 2002; Povee et al., 2012; van Gameren-Oosterom et al., 2011). Also identified in the review was that between late childhood and adolescence/early adulthood there appears to be a change in the nature of problems. Specifically, evidence from several sources suggested that from adolescence to adulthood, externalising behaviour problems decrease and internalising problems increase (Dykens & Kasari, 1997; Dykens et al., 2002; Nicham et al., 2003). One possibility investigated in this study was that maturational factors might account for this change. Study 1 examined this issue further by investigating behavioural problems in children and adolescents with DS who were aged between 10- and 15-years of age. This age range was selected because previous research (Dykens et al., 2002) indicated that during this time period internalising behaviour problems begin to appear. This study adds to the existing literature by assessing behavioural problems specifically in the transitional period from childhood to adolescence. A group of TD children of comparable age were also recruited to participate in the study to serve as a comparison group.

In this study behavioural problems in the participants were assessed. Based on previous research it was hypothesised that overall, the group comprising children and adolescents with DS would present with more behavioural problems than the control group. It was also hypothesised that the DS group would also
present with more externalising and internalising problems when compared with the TD participants. Finally, assuming general maturational factors influenced behaviour problems in individuals with DS, a significant correlation between participants’ age and the presence of externalising and internalising problems was expected.

7.2 Method

7.2.1 Participants

Two groups of participants were recruited to take part in the study. One group comprised 24 children and adolescents with DS. The second group, which served as a comparison group, comprised 24 TD children and adolescents. Participants in the TD group were individually matched to children and adolescents in the DS group on the basis of age, gender and parental skill level. It was beyond the scope of this study to include two comparison groups. As one key variable of interest was the impact of age on behavioural problems, it was important that participants had a similar level of life experience. Consequently no MA comparison group was included in this study.

7.2.1.1 Recruitment of children and adolescents.

7.2.1.1.1 Recruitment of participants with Down syndrome.

Participants with DS were recruited via Down Syndrome Victoria and Down Syndrome Tasmania. The recruitment of participants via Down Syndrome Victoria involved sending families of children or adolescents with DS aged between 10- and 15-years an information pack about the study. This consisted of a plain language statement and letter of informed consent (see Appendix B). To ensure the members confidentiality, the information packs were provided to Down Syndrome Victoria.
These were addressed onsite, and then mailed out directly by the staff. Parents who consented to their child participating in the study were given the option to have testing conducted at school or at Deakin University. Six parents indicated they would prefer these sessions to be conducted at home. Sixteen children and adolescents were tested at their respective schools. For these participants written consent from the school principal was obtained prior to assessment sessions at schools. In the case of testing at home or at Deakin University, appropriate arrangements for testing were made directly with parents. A total of 22 children and adolescents with DS were recruited via this process.

The children with DS were also contacted via Down Syndrome Tasmania by sending the plain language statement and informed consent form to parents of children and adolescents. Parents of individuals between 10- and 15-years old that were taking part in an annual family camp were contacted. This information was provided to Down Syndrome Tasmania, which was then passed on to the eligible families. The participants located in Tasmania were assessed during this camp that ran over three consecutive days. Four participants were recruited for the study using the process.

7.2.1.1.2 Inclusionary criteria for Down syndrome sample. The participants with DS were included in the study if they met the following criteria. First, they were reported by parents to have been diagnosed with DS. Second, they were aged between 10- and 15-years old. Third, the child came from a home where English was spoken as the first language. A child or adolescent with DS was excluded from the study if s/he had been diagnosed with a co-morbid developmental disorder. The criterion aimed to exclude children with a dual-diagnoses that may also account for variability in behaviour problems. Two participants with DS had
received a diagnosis of Autism Spectrum Disorder and were excluded from the sample, resulting in a final sample of 24 participants.

7.2.1.1.3 Identification of specific types of Down syndrome. For children in the DS group, parents were asked to indicate the specific type of DS that their child had been diagnosed with; Trisomy 21, Mosaic Trisomy 21 or Translocation DS. The reason for this was that individuals with Mosaic DS may present with cognitive skills in the average range, while Trisomy 21 is characterised by intellectual disability (de A. Moreira, San Juan, Pereira, & de Souza, 2000; Fishler & Koch, 1991; Fishler, Koch, & Donnell, 1976). In the final DS sample 23 children were identified as having Trisomy 21 and one child had a diagnosis of Mosaic DS.

To assess if the child with Mosaic DS differed significantly on cognitive skills from those with Trisomy 21, a modified $t$-test developed by Crawford and Howell (Crawford & Garthwaite, 2006; Crawford, Garthwaite, Azzalini, Howell, & Laws, 2006; Crawford & Howell, 1998) was used. This test evaluates whether an individual’s score is significantly different from the mean score of a control group, and is specifically designed to be used with small control groups (i.e., $n < 50$). Analyses were conducted with two core measures of cognitive skills; IQ and language. Specifically, the Full Scale IQ score from the Wechsler Abbreviated Scale of Intelligence (Wechsler, 1999) and Core Language standard score from the Clinical Evaluation of Language Fundamentals, 4th Edition (Semel, Wiig, & Secord, 2003) were used for these analyses. No significant differences in the scores were observed for the child with Mosaic DS compared with participants with Trisomy 21 on IQ ($t = 0.55, p = .591$) or language skills ($t = 0.38, p = .706$). Consequently, all further analyses were conducted with all 24 participants.
7.2.1.4 Recruitment of typically developing participants. The TD children and adolescents were recruited from schools in metropolitan areas and also referred to the study from other families whose children had participated in the research. The recruitment procedure at schools included first sending a letter of invitation to school principals (see Appendix C) and then a plain language statement and consent form to children in Grades 4 to 9 (see Appendix D). Letters were sent to children in these grades in order to obtain a comparison group that was of comparable age to the DS group. Participants who were referred to the study were directly supplied with a copy of the plain language statement and informed consent forms. Arrangements were made with parents of these children to run assessment sessions at home.

7.2.1.5 Inclusionary and exclusionary criteria for typically developing sample. To participate in the study children and adolescents in the TD group were required to meet the following criteria: (i) present with no known developmental or diagnosed psychiatric disorder, (ii) a chronological age between 10- and 16-years, (iii) come from a home where English was spoken as the first language (iv) being able to be matched to the gender and chronological age (where possible, ± 6 months) of one of the participants in the DS sample. A discrepancy exists between the age ranges for the DS group (10-15 years) and TD group (10-16 years). Due to difficulties recruiting older TD adolescents, it was necessary to broaden the age range to include 16 year-olds in this group. As will be shown below, no significant differences were found between the two study groups with respect to age.

Table 6 presents the demographic and clinical characteristics of the DS and TD samples. Also reported in Table 6 are the results for independent samples $t$-tests and chi-squared analyses testing whether there were significant differences
between the groups on demographic variables. This Table shows that there were no significant between group differences with regard to gender, age and parental skill level. Initially there was no intention to match the participant groups on parental skill level, however when parents complete the CBCL they are asked to indicate their occupation. As this data was available to the researchers, it was possible to assess whether there were any differences on parental skill levels that might confound the results. Parental skill level was determined by converting the reported occupation of parents to a rating between 1 (highest) and 5 (lowest) according to the Australian and New Zealand Standard Classification of Occupations (Trewin & Pink, 2006). No mothers of participants with DS reported an occupation on the lowest skill level. Thus there was a disparity on the range for mother’s skill level between the DS (1-4) and TD group (1-5). The reason for this difference is not clear from the available data. Significantly more children with DS were reported to have chronic illnesses. This was to be expected, as ongoing health problems are common in children with DS (Chapman et al., 1998; Laws, 2004; Laws & Bishop, 2004b; Roberts et al., 2007).
Table 6. *Sample Characteristics for Down Syndrome and Typically Developing groups (n = 48) and Group Comparisons using t or $\chi^2$ tests*

<table>
<thead>
<tr>
<th></th>
<th>DS (n = 24)</th>
<th>TD (n = 24)</th>
<th>t or $\chi^2$</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gender – female</strong></td>
<td>10 (41.7)</td>
<td>10 (41.7)</td>
<td>0.00</td>
</tr>
<tr>
<td><strong>Age in years</strong></td>
<td>12.6 ± 1.5</td>
<td>12.9 ± 1.8</td>
<td>0.66</td>
</tr>
<tr>
<td><strong>Type of DS - Trisomy 21</strong></td>
<td>23 (95.8)</td>
<td>23 (95.8)</td>
<td>0.00</td>
</tr>
<tr>
<td><strong>Number of chronic illnesses</strong></td>
<td>0.5 ± 0.7</td>
<td>0.0 ± 0.2</td>
<td>2.96**</td>
</tr>
<tr>
<td><strong>Mother’s skill levela</strong></td>
<td>2.3 ± 1.4</td>
<td>2.4 ± 1.6</td>
<td>-0.18</td>
</tr>
<tr>
<td><strong>Father’s skill levela</strong></td>
<td>2.3 ± 1.2</td>
<td>2.5 ± 1.6</td>
<td>-0.43</td>
</tr>
</tbody>
</table>

Note. Total n reported is not always consistent with total sample size due to missing data on some items.

*aParental occupation was classified with the Australian and New Zealand Standard Classification of Occupations (Trewin & Pink, 2006) and converted to skill levels between 1 (highest skill) and 5 (lowest skill level). DS group n = 18 and 19; Control group n = 19 and 22 for mothers and fathers respectively. *p<.05, **p<.01

7.2.2 Materials

7.2.2.1 Measures of Behaviour Problems. Behaviour problems of participants were measured using the CBCL (Achenbach & Rescorla, 2001) and the Social Skills Improvement System - Rating Scales (Gresham & Elliott, 2008).

Parental checklists were selected for the present study as they are a well-validated method for assessing childhood behavioural problems. Other methods of assessment, such as collecting observational data or undertaking clinical assessments using diagnostic criteria would also provide valuable insight into the behavioural profiles of children and adolescents with DS. However, as many of the participants in this study were located in rural areas, it was impractical to select such a time-intensive method of assessment. The CBCL and SSIS-RS are now described in turn.

The CBCL consists of 113 items. Each item presents a statement about the child’s behaviour. For example, ‘disobedient at home’, ‘too fearful or anxious’, ‘impulsive or acts without thinking’. The respondent, who may be a parent or guardian is asked to rate how often their child has exhibited the behaviour within the
past 6 months using a 3-point likert scale where 0 = ‘not true (as far as you know)’, 1 = ‘somewhat or sometimes true’ and 2 = ‘very true or often true’. Two items relating to suicidal ideation and self-harm were omitted due to concerns with research ethics.

For the purposes of this study three composite scores were obtained from the CBCL that measured externalising problems and internalising problems as well as overall behaviour problems. A summary of participants’ externalising problems was obtained using the ‘Externalising Problems Scale’. The Externalising Problems Scale is computed by summing scores on the Rule Breaking and Aggressive Behaviour subscales. Participants’ internalising problems was measured using the Internalising Problems Scale. This scale is obtained by combining scores from the Anxious/Depressed, Withdrawn/Depressed and Somatic Complaints subscales. Finally, the scores from all problem items on the CBCL are combined to create a ‘Total Behaviour Problems Scale’. This scale assesses the severity of a child’s overall problem behaviour. For all scales and subscales, higher values indicate the presence of a behaviour problem. Table 7 presents a description of the constructs measured by the Externalising, Internalising and Total Behaviour Problem Scales, as well as component subscales.
<table>
<thead>
<tr>
<th>CBCL scale / subscale</th>
<th>Description and sample items</th>
<th>Min. – Max. raw score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Behaviour Problems</td>
<td>Composite measure of all subscales and scores from an additional 17 ‘other’ items that do not contribute to subscale scores (e.g., ‘talks too much’, ‘showing off or clowning’, ‘wets the bed’). Subscales that contribute to composite are: Anxious/Depressed, Withdrawn/Depressed, Somatic Complaints, Rule Breaking Behaviour, Aggressive Behaviour, Social Problems, Thought Problems and Attention Problems.</td>
<td>0 – 240</td>
</tr>
<tr>
<td>Externalising Problems</td>
<td>Composite measure of the Rule Breaking and Aggressive subscales.</td>
<td>0 – 70</td>
</tr>
<tr>
<td>Internalising Problems</td>
<td>Composite measure of the Anxious/Depressed, Withdrawn/Depressed and Somatic Complaints subscales.</td>
<td>0 – 64</td>
</tr>
<tr>
<td>Anxious/Depressed</td>
<td>Symptoms associated with low mood (e.g., sadness, guilt) and anxiety (e.g., worry, fear). Sample items: ‘feels he/she has to be perfect’, ‘feels worthless or inferior’</td>
<td>0 – 26</td>
</tr>
<tr>
<td>Withdrawn/Depresseda</td>
<td>Signs of social withdrawal that are related to low mood. Sample items: ‘would rather be alone than with others’, ‘unhappy, sad or depressed’</td>
<td>0 – 16</td>
</tr>
<tr>
<td>Somatic Complaints</td>
<td>Physical complaints that have no known biological cause. Sample items: ‘overtired without good reason’, ‘Physical problems without known medical cause: headaches’</td>
<td>0 – 22</td>
</tr>
<tr>
<td>Rule Breaking Behaviourb</td>
<td>Behaviours that violate social norms or expectations. Sample items: ‘breaks rules at home, school, or elsewhere’, ‘lying or cheating’</td>
<td>0 – 34</td>
</tr>
<tr>
<td>Aggressive Behaviour</td>
<td>Behaviours that can cause harm to the self, others, or property, as well as difficulties with managing negative emotion (e.g., anger). Sample items: ‘cruelty, bullying, or meanness to others’, ‘destroys his/her own things’</td>
<td>0 – 36</td>
</tr>
</tbody>
</table>

Abbreviations. CBCL= Child Behavior Checklist

*a This scale is the counterpart to the ‘withdrawn’ scale on the CBCL (1991) and thus was referred to as this in the previous chapters. *b This scale was referred to as ‘delinquent behaviour’ on the CBCL (1991) and therefore this term was used throughout the previous chapters. (Achenbach & Rescorla, 2001).
The CBCL subscales have good reliability. The internal consistency for the eight syndrome subscales range from .82 to .92 (Achenbach & Rescorla, 2001). Raw scores for each of these scales have been converted to T-scores using separate norms for boys and girls, and children between 6-11 and 12-18 years. T-scores have a distribution which has a mean of 50 and standard deviation of 10. These standard scores provide information about whether the precise behavioural syndrome score should be interpreted as non-clinical ($T < 65$), borderline-clinical ($T = 65-69$) or within the clinical range ($T \geq 70$). The Internalising, Externalising and Total Behaviour Problems scores have an internal consistency coefficients of .91, .92 and .94 respectively (Achenbach & Rescorla, 2001). Raw scores on the CBCL range between 0-70 (Externalising scale), 0-64 (Internalising scale) and 0-240 (Total Behaviour Problem scale). On Externalising, Internalising and Total Behaviour Problems scales a T-Score that is greater than 64 indicates the presence of a clinically significant behaviour problem. T-scores between 60-63 indicate behaviour problems that are borderline or subclinical. That is, a behaviour problem is present, but may not be clinically significant as determined by the CBCL. A T-Score that is below 60 indicates the behaviour is in the normal range. That is, no behaviour problems are present as determined by the CBCL.

An additional measure of behaviour problem was also used in this study. By using two measures the reliability with which behaviour problems were measured should be improved. The second measure of behaviour problems used was the Social Skills Improvement System - Rating Scales (SSIS-RS; Gresham & Elliott, 2008). The SSIS-RS is a standardised, norm-referenced instrument that measures problem behaviours as well as social skills in children, adolescents and young adults aged between 3- and 18-years. In total there are 79 items on the SSIS-RS. However, only data from the 33 items that measure internalising, externalising and total
behaviour problems were analysed for this study. The SSIS-RS comprises statements about the child’s behaviour. Example items include ‘acts anxious with others’ and ‘is aggressive toward people or objects’. The informant who may be a parent or guardian is asked to rate each statement on a four-point likert scale, indicating how often the behaviour has occurred during the past 2 months. Response options are 0 = ‘never’, 1 = ‘seldom’, 2 = ‘often’ and 3 = ‘almost always’. In this study the participant’s parent completed the SSIS-RS.

Table 8 presents a description of the constructs measured by the Externalising, Internalising and Total Behaviour Problems scales, sample items and the minimum/maximum raw scores for each index. In the analyses, only raw scores could be used for the measures of Externalising and Internalising Behaviour Problems as no standard scores are available. The Total Problem Behaviours Scale from the SISS-RS is obtained by summing items from the Externalising and Internalising Behaviour scales, as well as Bullying, Hyperactive/Inattention and Autism Spectrum (problem items only). The Total Problem Scale has raw scores that can range between 0 and 99 and is standardised to a mean of 100 and standard deviation of 15.
Table 8. Description of Problem Behaviour Indexes on the Social Skills Improvement System- Rating Scales

<table>
<thead>
<tr>
<th>SSIS-RS index</th>
<th>Description and sample items</th>
<th>Min. – Max. raw score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Problem Behaviours</td>
<td>Composite measure of general behaviour problems. This includes externalising and internalising behaviour problems, as well as items assessing bullying, hyperactivity/inattention and behaviour problems typically associated with autism spectrum disorders.</td>
<td>1 – 99</td>
</tr>
<tr>
<td>Externalising Problems</td>
<td>Verbal or physical aggression, difficulty controlling temper or being argumentative. Sample items: ‘disobeys rules or requests’; ‘fights with others’.</td>
<td>0 – 36</td>
</tr>
<tr>
<td>Internalising Problems</td>
<td>Feelings of anxiety, sadness and loneliness or having poor self-esteem. Sample items: ‘withdraws from others’; ‘has low energy or is lethargic’.</td>
<td>0 – 30</td>
</tr>
</tbody>
</table>

(Gresham & Elliott, 2008)

The SSIS-RS has been shown to have good reliability, with the internal consistency coefficients ranging between .73 and .92 for the individual subscales (Gresham & Elliott, 2008; Gresham, Elliott, Vance, & Cook, 2011). The scales of particular interest in this study show excellent internal consistency. The Externalising scale alpha coefficient ranges between .88 and .92 while the Internalising domain and Total Problems composite have alpha coefficients ranging .82-.90, and .93-.96 respectively (Gresham & Elliott, 2008; Gresham et al., 2011).

7.2.3 Procedure

Prior to testing, ethics approval for this study was granted from Deakin University Research Ethics Committee (see Appendix E), the Department of Education and Early Childhood Development (see Appendix F) and Catholic Education Office (see Appendix G). Parents of all children in the DS and control groups were supplied with a copy of the CBCL and SSIS-RS, and a reply-paid
envelope. One parent or guardian was asked to complete the questionnaires and return them directly to the researchers using the reply-paid envelope.

7.3 Results

7.3.1 Behaviour Problems in Children and Adolescents with Down Syndrome using the CBCL

The first set of analyses investigated the number of children and adolescents with behaviour problems in the DS and TD groups as determined using the CBCL. The percentage of adolescents with behaviour problems in the non-clinical, borderline-clinical and clinical range on the CBCL are presented by group in Table 9. Classifications were made according to T-Scores using the recommended clinical cut-off scores provided in the test manual (Achenbach & Rescorla, 2001). That is, a T-Score on the subscales (e.g., Rule-breaking) below 65 is considered non-clinical, between 65 and 69 is borderline-clinical and 70 or above is considered within the clinical range. For the Externalising, Internalising and Total Problems composites, behaviour problems in the non-clinical, borderline-clinical and clinical ranges are indicated by T-scores below 60, 60-63 and above 64 respectively.
# Behaviour Problems in Down Syndrome

## Table 9. Frequency of CBCL Behaviour Problems, According to Clinical Range and Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>DS group (n = 24)</th>
<th>TD group (n = 24)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Non-clinical</td>
<td>Borderline</td>
</tr>
<tr>
<td></td>
<td>n</td>
<td>%</td>
</tr>
<tr>
<td><strong>Subscales</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anxious / Depressed</td>
<td>23</td>
<td>95.8</td>
</tr>
<tr>
<td>Withdrawn / Depressed</td>
<td>22</td>
<td>91.7</td>
</tr>
<tr>
<td>Somatic Complaints</td>
<td>19</td>
<td>79.2</td>
</tr>
<tr>
<td>Rule Breaking</td>
<td>23</td>
<td>95.8</td>
</tr>
<tr>
<td>Aggressive Problems</td>
<td>18</td>
<td>75.0</td>
</tr>
<tr>
<td><strong>Composites</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Internalising Problems</td>
<td>19</td>
<td>79.2</td>
</tr>
<tr>
<td>Externalising Problems</td>
<td>17</td>
<td>70.8</td>
</tr>
<tr>
<td>Total Problems</td>
<td>16</td>
<td>66.7</td>
</tr>
</tbody>
</table>

Abbreviations. CBCL = Child Behavior Checklist (Achenbach & Rescorla, 2001)
Table 9 shows that on the Total, Externalising and Internalising Problems Scales, there are generally more participants with DS who scored within the borderline or clinical range compared with the control group. On the Total Problems Scale 33.3% of the DS sample obtained a score that was either in the borderline or clinical range. In comparison only 4.2% or one participant in the TD group obtained an equivalent score. Chi-square analysis revealed that whether a participant had a score that was in the borderline or clinical range was dependent upon whether s/he had DS ($\chi^2 (2) = 7.056, p = .029$).

Externalising behaviour problems were found to be more prevalent in the DS group compared with the comparison group. As seen in Table 9 there were more children/adolescents with DS who scored in the borderline and clinical range of the CBCL compared to the control group. In total, 29.1% of the DS sample obtained a score in the borderline or clinical range on the Externalising Problems Scale. None of the participants in the TD group obtained a score on this scale that was in the borderline or clinical range. Chi-square analysis indicated that the presence of externalising behaviour problems in the borderline or clinical range was related to whether the participant was in the DS group ($\chi^2 (2) = 8.195, p = .017$). Interestingly, it is not the case that all externalising behaviours are elevated in the DS group compared with the TD group. Table 9 shows no differences between the groups with on the Rule Breaking subscale ($p > .999$). However, borderline or clinically significant behaviour problems for the Aggressive Behaviour subscale was significantly related to the DS group ($\chi^2 (2) = 6.857, p = .032$).

On the Internalising Problems Scale, there were generally more children in the DS within the borderline or clinical range compared to the TD group. In the DS group 20% of the sample obtained a score that was at least in the borderline
range compared to 12.5% in the TD group. However, no significant relationship between groups and scores on the Internalising Problems Scale was found ($\chi^2 (2) = 2.233, p = .327$). Non-significant relationships were also observed on each of the internalising behaviour problems subscales (Anxious/Depressed: $\chi^2 (2) < 0.001, p = .999$; Withdrawn/Depressed: $\chi^2 (2) = 3.022, p = .221$; Somatic Complaints: $\chi^2 (2) = 0.226, p = .893$). However, it is interesting to note that the largest difference between groups was found on the Withdrawn/Depressed subscale. On this index two children in the DS group obtained a score in the clinical range and one child in the TD group obtained a score in the borderline range. Thus there is a non-significant trend of elevated levels of Withdrawn/Depressed behaviour in participants with DS.

**7.3.2 Behaviour Problems in Children and Adolescents with Down Syndrome using the SSIS-RS**

In the second set of analyses, differences in behaviour problems in the DS and TD groups were compared using the SSIS-RS. This instrument provides composite measures for Externalising, Internalising and Total behaviour problems. Summary statistics for each of these groups are presented in Table 10. As noted earlier, for this instrument no standard scores are available for the measures of internalising and externalising behaviour. Thus, for these composites, only raw scores are presented. Scores from the Total Problem Behaviours Scale are standardised to a mean of 100 and standard deviation of 15. For all measures, higher scores indicate more problem behaviours.
To investigate the differences between the DS and TD group on the behavioural measures independent samples $t$-tests and Cohen’s $d$ effect sizes were computed for the externalising, internalising and total problem scales on the CBCL and SSIS-RS. According to Cohen’s (1988) taxonomy effect sizes are classified as small ($d = 0.2$), medium ($d = 0.5$) or large ($d = 0.8$).
Table 10. Summary Statistics for Behaviour Problems on CBCL and SSIS-RS, Reported by Groups

<table>
<thead>
<tr>
<th>Variable</th>
<th>DS group (n = 24)</th>
<th>TD group (n = 24)</th>
<th>Comparison of Means</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>Min.</td>
</tr>
<tr>
<td>CBCL</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Composites</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Externalising</td>
<td>54.04</td>
<td>9.26</td>
<td>40</td>
</tr>
<tr>
<td>Internalising</td>
<td>51.04</td>
<td>10.22</td>
<td>34</td>
</tr>
<tr>
<td>Total Behaviour Problems</td>
<td>56.67</td>
<td>8.29</td>
<td>36</td>
</tr>
<tr>
<td>SSIS-RS</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Externalising</td>
<td>9.67</td>
<td>5.79</td>
<td>1</td>
</tr>
<tr>
<td>Internalising</td>
<td>4.25</td>
<td>3.77</td>
<td>0</td>
</tr>
<tr>
<td>Composite</td>
<td>Total Problem Behaviours</td>
<td>110.38</td>
<td>15.96</td>
</tr>
</tbody>
</table>

Abbreviations. CBCL = Child Behavior Checklist; SSIS-RS = Social Skills Improvement System – Rating Scale

Note. CBCL scores are standardised to a mean of 50 and standard deviation of 10. SSIS-RS externalising and internalising scores are raw scores, as no standard scores are available. SSIS-RS Total Problem Behaviours scores are standardised to a mean of 100 and standard deviation of 15.

<sup>a</sup> Positive values indicate DS children were reported to have higher scores on the measure.
On the SSIS-RS the pattern of results largely mirror those observed in the first set of analyses undertaken with the CBCL. On the Total and Externalising Problem Behaviours scales the DS group obtained a significantly higher score than the comparison group. This result indicates that on these measures the DS group had significantly more Total Problem Behaviours and Externalising Problem Behaviours. Also on these comparisons, the magnitude of the effect size was found to be large. The difference between the groups on the Internalising Problem Behaviours composite was not found to be statistically significant. However, it is interesting to note that the magnitude of the effect size between these groups on this index was found to be medium. That is, the value of Cohen’s $d$ was found to be 0.49 indicating the difference between the groups was around half a standard deviation. Thus, low statistical power rather than an absence of an effect might account for the non-significant result in this instance.

### 7.3.3 Relationship between Age and Behaviour Problems in Down Syndrome

The final set of analyses examined the relationship between participants’ age and the presence of behaviour problems. In the following analyses Pearson’s $r$ was used to compute the relationship between the participants’ age and measures of internalising, externalising and total behaviour problems from the CBCL and SSIS-RS. Correlations between age and behaviour problems for the CBCL and SSIS-RS are presented in Table 11 for the DS and TD control group.
Table 11. *Pearson’s Correlations for Externalising, Internalising and Total Behaviour Problems on the CBCL and SSIS-RS, Reported by Group*

<table>
<thead>
<tr>
<th>Variables</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>DS group (n = 24)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Age in Years</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. CBCL Externalising Problems</td>
<td>.077</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. CBCL Internalising Problems</td>
<td>.217</td>
<td>.313</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. CBCL Total Problems</td>
<td>.144</td>
<td>.812**</td>
<td>.718**</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. SSIS-RS Externalising Problems</td>
<td>-.027</td>
<td>.796**</td>
<td>.291</td>
<td>.755**</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. SSIS-RS Internalising Problems</td>
<td>.144</td>
<td>.396</td>
<td>.832**</td>
<td>.696**</td>
<td>.474’</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>7. SSIS-RS Total Problems</td>
<td>-.011</td>
<td>.743**</td>
<td>.551**</td>
<td>.835**</td>
<td>.915**</td>
<td>.739**</td>
<td>—</td>
</tr>
<tr>
<td><strong>TD group (n = 24)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Age in Years</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. CBCL Externalising Problems</td>
<td>.018</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. CBCL Internalising Problems</td>
<td>-.043</td>
<td>.626**</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. CBCL Total Problems</td>
<td>-.056</td>
<td>.931**</td>
<td>.810**</td>
<td>—</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. SSIS-RS Externalising Problems</td>
<td>-.054</td>
<td>.739**</td>
<td>.432’</td>
<td>.734**</td>
<td>—</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. SSIS-RS Internalising Problems</td>
<td>-.205</td>
<td>.561**</td>
<td>.466’</td>
<td>.578**</td>
<td>.483’</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>7. SSIS-RS Total Problems</td>
<td>-.183</td>
<td>.715**</td>
<td>.529**</td>
<td>.745**</td>
<td>.843**</td>
<td>.843**</td>
<td>—</td>
</tr>
</tbody>
</table>

*p < .05, **p < .01, ***p < .001
Table 11 reports no significant correlations between age and any of the specific behavioural scales on CBCL or SSIS-RS. Age was also not correlated with the Total Problem Behaviours scores in participants with DS. Low reliability might explain the non-significant associations between age and behaviour problems in the DS group. To increase the reliability of the results new composite scores were created using the item scores from both behavioural measures. These scores were created by averaging the z-scores obtained from raw scores on the CBCL and SSIS-RS.

Spearman’s rho bivariate correlations were calculated between chronological age and the Externalising, Internalising and Total Behaviour Problems Composite scores for the DS group and are presented in Table 12. The results showed that age was not significantly related to any of the behaviour problem scores in adolescents with DS.

Table 12. Spearman Rho Correlations between Age and Behaviour Composite Scores in the Down Syndrome Group (n = 24)

<table>
<thead>
<tr>
<th>Variables</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Age in years</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>2. Externalising Problems Composite</td>
<td>-.091</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>3. Internalising Problems Composite</td>
<td>.223</td>
<td>.334</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>4. Total Behaviour Problems Composite</td>
<td>-.022</td>
<td>.867***</td>
<td>.688***</td>
<td>—</td>
</tr>
</tbody>
</table>

*p < .05, **p < .01, ***p <.001

7.3.4 Exploratory Analysis of Non-Linear Relationships in Behaviour Problems

One possible reason for finding no correlation between behavioural problems and age on the CBCL might be that there is a non-linear trend between the two variables. To investigate this possibility, some exploratory analysis were conducted by producing scatter plots for the Externalising, Internalising and Total Problem Composites. The scatter plots are presented in Figure 1 (a) Total (b)
Behaviour Problems in Down Syndrome

Externalising and (c) Internalising Problems. Inspection of these plots shows no non-linear associations between age and any type of behavioural problem in adolescents with DS.

Figure 1. Scatter plots of relationship between age and (a) Total, (b) Externalising and (c) Internalising Composite Scores in DS group
Figure 1. Scatter plots of relationship between age and (a) Total, (b) Externalising and (c) Internalising Composite Scores in DS group (continued)
7.4 Discussion

Study 1 investigated behavioural problems in children and adolescents with and without DS. The results from Study 1 supported the hypothesis that overall, more behaviour problems would be present in the DS group compared to the TD age-matched controls. On the Total Behaviour Problems Scale from the CBCL the DS group had significantly more participants considered to be in the borderline or clinical range. Around 33% of the participants with DS obtained scores that fell within the criteria. This result is comparable to a number of past studies that also show the number of children and adolescents with behaviour problems is high (Dykens et al., 2002; Povee et al., 2012; van Gameren-Oosterom et al., 2011). This study provides further evidence that behaviour problems are prevalent in children and adolescents with DS.

However, the results of this study did not show that all types of behaviour problems are present in individuals with DS in the age range studied. Analyses showed the DS group had significantly more externalising behaviour problems compared with the TD group. This finding appears robust since the result was observed using the CBCL and SSIS-RS. Also, data from the CBCL revealed that just under 17% of the sample with DS had externalising behaviour problems in the clinical range. This finding is consistent with results from other studies (Fidler et al., 2000; van Gameren-Oosterom et al., 2011).

The difference between the groups on the internalising measures was not found to be statistically significant. This result is inconsistent with van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) who reported significantly higher levels of internalising problems in adolescents and young adults with DS compared with an age-matched control group. The age of the participants in the current study
Behaviour Problems in Down Syndrome

cOMPARED TO THOSE IN VAN GAMEREN-OOSTEROM, FEKES, VAN WOUWE, ET AL. (2013) MAY ACCOUNT FOR THIS DISCREPANCY. ONE POSSIBILITY IS THAT INTERNALISING PROBLEMS BECOME MORE PREVALENT IN OLDER INDIVIDUALS WITH DS. AS NOTED EARLIER, VAN GAMEREN-OOSTEROM ET AL. (2011) DID NOT FIND DIFFERENCES ON BEHAVIOUR PROBLEMS IN 8-YEAR OLD CHILDREN WITH DS AND AGE-MATCHED CONTROLS.

IN THE CURRENT STUDY IT WAS CERTAINLY NOT THE CASE THAT THE DS AND THE COMPARISON GROUP WERE UNIFORM WITH RESPECT TO INTERNALISING PROBLEMS. ANALYSES SHOWED THAT ON THE CBCL, THE PERCENTAGE OF PARTICIPANTS WITH DS SCORING WITHIN THE BORDERLINE AND CLINICAL RANGE FOR INTERNALISING PROBLEMS WAS 20.8% AND 12.5% FOR TD PARTICIPANTS. ON THE SSIS-RS, THE DS GROUP SCORED AROUND HALF A STANDARD DEVIATION HIGHER ON THE INTERNALISING PROBLEMS COMPOSITE COMPARED WITH CONTROLS. thus, observed in this study were effect sizes that were medium in magnitude suggesting the presence of internalising problems between the ages of 10-15 years. This pattern of result is different to another study that found small effect sizes in internalising problems in young children with DS compared to controls (van Gameren-Oosterom et al., 2011). Based on this result it is tentatively suggested that internalising problems may emerge during late childhood for people with DS.

FInally, this study also examined whether general maturational factors, as measured by a participant’s age, was related to behaviour problems. Specifically, the hypothesis tested in this study was that age would be positively correlated with internalising behaviour problems, and negatively associated with externalising problems. The results did not support this hypothesis. No significant correlation was found between age and a measure of internalising or externalising behaviour problems. This result was found using correlations were computed using age and measures of behaviour problems from the CBCL and SSIS-RS.
Possible explanations for the non-significant relationship between age and behavioural problems are that (i) behaviour problems are relatively stable for individuals with DS aged 10-15 years old (ii) the age range of participants may have contributed to the non-significant findings or (iii) there is a non-linear relationship between behavioural problems and age. These explanations will now be considered.

This is the first known study to specifically examine the relationship between age and behavioural problems during the transition from late childhood to early adolescence. Thus this study aimed to examine changes in behavioural problems that were hypothesised on the basis of empirical work undertaken with participants aged 4-19 years old (e.g., Dykens & Kasari, 1997; Dykens et al., 2002). Therefore, it is possible that there is not a significant change in externalising and/or internalising problems for individuals with DS between 10 and 15 years old. Alternatively, there is some support for explanations (ii) and (iii). Specifically, Dykens and Kasari (1997) reported a significant positive relationship between internalising problems and age that was not replicated in the present study. As noted above, the Dykens and Kasari (1997) study was undertaken with participants 4-19 years old, while the current sample included individuals between 10 and 15 years old. Therefore, it may be that the age differences in internalising problems only emerge if taken over a wider developmental period. However, no known study has reported a significant correlation between externalising problems and age, suggesting that externalising problems may be influenced by factors other than age. Additionally, studies by Dykens et al. (2002) and Povee et al. (2012) showed that total and/or externalising problems increased until children reached approximately 13 to 14 years of age, and then decreased in older adolescents and young adults with DS (e.g., Dykens et al., 2002; Povee et al., 2012). Thus, this supports the possibility of a non-linear trend to behavioural problems in this population. Due to sample size
limitations it was not possible to formally test this possibility in the current study. However, inspection of the composite score scatterplots indicated no obvious non-linear trend between age and externalising, internalising or total behaviour problems.

Overall, the results of this study show that behaviour problems are more prevalent in individuals with DS aged between 10-15 years compared with TD children and adolescents of comparable age. However, during this period of time, the results suggest that while externalising problems are common, internalising problems appear to be emerging. This change in the nature of behaviour problems does not appear to be related to general maturational factors as indexed by the age of participants.
CHAPTER 8: STUDY 2. BEHAVIOUR PROBLEMS, COGNITIVE AND NEUROPSYCHOLOGICAL SKILLS IN DOWN SYNDROME

8.1 Introduction

In Chapter 5 a range of cognitive skills and a neurological problem were presented which might account for behaviour problems in children and adolescents with DS. The aim of Study 2 was to examine the relationship between these variables and behaviour problems in individuals DS. Specifically, in Study 2 participants with DS and an age matched control group were presented with a test battery that measured IQ, emotion recognition, ToM, language and social skills and a neuropsychological measure associated with dementia. The first set of analyses examined differences between the DS and control group on each of these measures. The second set of analyses examined the relationship between the aforementioned variables and behaviour problems. Based on the theories presented in Chapter 5, a significant association was expected between the behaviour problems and cognitive/neuropsychological measures. The hypotheses are summarised in Table 13.
Table 13. *Hypothesised Relationships between Behaviour Problems and Cognitive / Neuropsychological Variables*

<table>
<thead>
<tr>
<th>Variable</th>
<th>Hypothesis</th>
</tr>
</thead>
<tbody>
<tr>
<td>IQ</td>
<td>Children and adolescents with lower IQ will have more externalising, internalising and total behavioural problems.</td>
</tr>
<tr>
<td>Emotion recognition</td>
<td>Participants with poorer emotion recognition skills will show more externalising, internalising and total behaviour problems.</td>
</tr>
<tr>
<td>Theory of mind</td>
<td>Poorer theory of mind will be associated with more externalising, internalising and total behavioural problems.</td>
</tr>
<tr>
<td>Language</td>
<td>Participants with more language and pragmatic language difficulties will have more externalising, internalising and total problems.</td>
</tr>
<tr>
<td>Social skills</td>
<td>Children and adolescents with poorer social skills are predicted to have more externalising, internalising and total behaviour problems.</td>
</tr>
<tr>
<td>Neurodegeneration</td>
<td>Poorer performance on measures of dementia will be associated with more internalising problems in individuals with DS. No significant association is expected between the measure of dementia and externalising or total behaviour problems.</td>
</tr>
</tbody>
</table>

8.2 Method

8.2.1 Participants

Participants in this study were the same children and adolescents who participated in Study 1. A summary of the participants’ age and background characteristics are re-presented in Table 14. As noted in Study 1, it was possible to have only one TD comparison group. As the participant groups were the same for both studies, there was also no MA comparison group in Study 2.
Table 14. Summary of Sample Characteristics for Down Syndrome and Typically Developing groups (n = 48) and Group Comparisons Using t or χ² Tests

<table>
<thead>
<tr>
<th></th>
<th>DS (n = 24)</th>
<th>TD (n = 24)</th>
<th>t or χ²</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n* (%) or</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Mean ± SD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gender – female</td>
<td>10 (41.7)</td>
<td>10 (41.7)</td>
<td>0</td>
</tr>
<tr>
<td>Age in years</td>
<td>12.6 ± 1.5</td>
<td>12.9 ± 1.8</td>
<td>0.66</td>
</tr>
<tr>
<td>Type of DS - Trisomy 21</td>
<td>23 (95.8)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number of chronic illnesses</td>
<td>0.5 ± 0.7</td>
<td>0.0 ± 0.2</td>
<td>2.96**</td>
</tr>
<tr>
<td>Mother’s skill levelᵃ</td>
<td>2.3 ± 1.4</td>
<td>2.4 ± 1.6</td>
<td>-0.18</td>
</tr>
<tr>
<td>Father’s skill levelᵃ</td>
<td>2.3 ± 1.2</td>
<td>2.5 ± 1.6</td>
<td>-0.43</td>
</tr>
</tbody>
</table>

Note. Total n reported are not always consistent with total sample size due to missing data on some items.

ᵃParental occupation was classified with the Australian and New Zealand Standard Classification of Occupations (Trewin & Pink, 2006) and converted to skill levels between 1 (highest skill) and 5 (lowest skill level). DS group n = 18 and 19; Control group n = 19 and 22 for mothers and fathers respectively.

8.2.2 Materials

Children and adolescents participating in the study were presented with a test battery that measured behaviour problems, cognitive skills, and a neuropsychological measure previously found to be an indicator of dementia. Each measure is now described.

8.2.2.1 Measure of behaviour problems. Study 2 measured behaviour problems with the same instruments that were used in Study 1, these were the CBCL and SSIS-RS. As a reminder, these are standardised checklists of behaviour problems where a parent or guardian rates the extent they have observed specific types of behaviour problems in their child using a likert scale. The results from Study 1 showed that the composite scores created from the CBCL and SSIS-RS had better reliability coefficients that the CBCL and SSIS-RS alone. Given this, for Study 2 a
Composite scores of Total, Externalising and Internalising Problems were created by summing each participant’s respective score from the CBCL and SSIS-RS. To ensure that the CBCL and SSIS-RS made an equal contribution to the composite, prior to summing scores it was necessary to transform raw scores from each instrument to a $z$-score. The $z$-score was referenced to the median and standard deviation of the entire sample. For the final analyses each participant’s behaviour problems were measured by a composite measure of Total Behaviour Problems, Externalising Problems and Internalising Problems. Each of these composites was the sum of two $z$-transformed raw scores from the CBCL and SSIS-RS.

Reliability analyses were conducted to assess the internal consistencies for the new behavioural composite scales. The new composite variables were found to have excellent reliability. Cronbach’s alpha was found to be .91, .87 and .96 for Externalising (37 items), Internalising (38 items) and Total Behaviour Problem composites (135 items) respectively. These reliability coefficients exceed the internal consistencies reported for the CBCL and SSIS-RS (Achenbach & Rescorla, 2001; Gresham & Elliott, 2008). Items that did not have demonstrate any participant variability (i.e., all participants scored zero on the item) were removed from the composite scores.

**8.2.2.2 Measure of IQ.** To assess the IQ of participants in this study the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999) was administered. The WASI is a brief, standardised measure of IQ for people aged between 6- and 89-years old (Wechsler, 1999). The WASI was selected as it is
widely used measure of intelligence that has previously been used in research with children and adolescents with DS (e.g., Edgin, Pennington, & Mervis, 2010; Groen, Laws, Nation, & Bishop, 2006). Furthermore, the WASI subtests are similar in format and item selection to those in other Wechsler intelligence scales, for example the Wechsler Intelligence Scale for Children – 3rd edition (Wechsler, 1991).

Wechsler intelligence scales have also been used extensively in research with participants with DS (e.g., Devenny, Krinsky McHale, Sersen, & Silverman, 2000; Dressler, Perelli, Feucht, & Bargagna, 2010; Kittler, Krinsky McHale, & Devenny, 2004; Pennington et al., 2003; Yirmiya et al., 1996; Youn & Youn, 1991). Multiple studies have shown the WASI to be a valid screening tool for general intelligence (e.g., Canivez, Konold, Collins, & Wilson, 2009; Hays, Reas, & Shaw, 2002; Saklofske, Caravan, & Schwartz, 2000; Wechsler, 1999).

Participants in the study were presented with the two-subtest version of the WASI, which consists of the Vocabulary and Matrix Reasoning tasks. The Vocabulary subtest involves the researcher giving the child a word and asking him/her what this is, or what the word means, for example, ‘what is an alligator’.

On the Matrix Reasoning subtest participants are presented with an incomplete matrix (i.e., picture) with five or six response options below. Children are asked ‘which one of these [researcher points to response options] goes here [points to missing piece]’. Performance from both subtests are combined to estimate a Full Scale IQ (FSIQ). The FSIQ is standardised to a mean of 100 and SD of 15.

The WASI demonstrates good reliability with its internal consistency coefficients for children between 10- and 16-years old ranging between .86 and .93 for Vocabulary, .86 and .92 for Matrix Reasoning and between .92 and .95 for the FSIQ score. The WASI also shows good test-retest reliability for Vocabulary (.85),
Matrix Reasoning (.77) and FSIQ (.85) for children between 6- and 16-years old (Wechsler, 1999).

**8.2.2.3 Measure of emotion recognition.** The Affect Recognition subtest from the Neuropsychological Assessment, 2nd Edition (NEPSY-II; Korkman, Kirk, & Kemp, 2007) was used to measure participants’ ability to accurately identify facial expression (i.e., affect). The Affect Recognition subtest involves first presenting the examinee a picture of a child’s face. The examinee is then asked to identify from an array of four other faces, which one “feels the same way” (e.g., happy, sad, angry). Raw scores from this subtest were used in the analyses. The maximum score that could be obtained is 35. The Affect Recognition subtest shows excellent internal consistency with coefficients ranging between .84 and .88 for children between 10- and 16-years old Edition (NEPSY-II; Korkman et al., 2007). This is the first known study to assess children with DS using the Affect Recognition subtest from the NEPSY-II.

**8.2.2.4 Measure of theory of mind.** ToM was measured using the ‘Theory of Mind’ subtest from the NEPSY-II (Korkman et al., 2007). This subtest measures a child’s ability to comprehend others’ perspectives, intentions and beliefs (Korkman et al., 2007). According to the NEPSY-II test manual the Theory of Mind subtest is appropriate for children aged 3- to 16-years old. Unfortunately, at present normative scores have only been provided for children between 3- and 6-years old on this task. The decision was made to include the subtest in spite of this limitation, because it includes a non-verbal assessment task for ToM. This was an important consideration, as language difficulties are commonly experienced by people with DS. Thus, this task was selected to minimise the expressive language demands placed
upon the participant and consequently reduce the impact that language difficulties might have on the results for this subtest.

The Theory of Mind subtest comprises two different tasks: verbal and contextual. On the verbal task the examinee is shown pictures of a person engaging in an activity. The examinee is then asked to describe or infer the mental state of the person in the picture. For example, one item shows a man with his finger on his lip and gazing into the distance. The examinee is asked what the man is doing in the picture. In this example the correct answer is ‘thinking’. In other items participants are asked to explain the meaning of abstract phrases such as “two peas in a pod”. On the contextual task the examinee is shown a picture of ‘Julia’ in a particular situation. As an example, in one scene Julia can be seen having an argument with a boy. Julia’s face is not visible in any of the scenes. The examinee is asked identify how Julia feels by pointing to one of the four pictures which shows different facial affect. In the above example the response options include neutral, angry, sad and surprised facial expressions (see Figure 2). Participants’ performance on the Theory of Mind subtest is quantified by adding raw scores from the verbal and contextual subtests. This was necessary since standard scores for this subtest are not available for children older than 6-years. The maximum score that could be obtained on the Theory of Mind subtest is 28 (NB: the maximum raw score on the verbal test is 22 and the maximum score on the contextual task is 6).
The reliability for the Theory of Mind subtest is good. Data reported in the test manual (Korkman et al., 2007) shows Cronbach’s Alpha to be between .76 and .84 for children aged between 3- to 6-years of age. For the current sample reliability was found to be substantially better. The analyses showed that Cronbach’s alpha was .92 on the verbal task (15 items) and .76 for the contextual task (6 items). The reliability for the total Theory of Mind score (21 items) was .93. To the authors knowledge, no study has assessed children or adolescents with DS with this Theory of Mind subtest.

8.2.2.5 Measure of language. Children’s language skills were measured using the Core Language Score (CLS) from the Clinical Evaluation of Language Fundamentals 4th Edition – Australian (CELF-4; Semel et al., 2003). The CLS provides an overall measure of a child’s ability to produce and understand language. The CELF-4 was selected for this study as it provides a comprehensive assessment of children’s ability to produce and understand language and has been standardised with an Australian sample of individuals aged between 5- and 21-years. Prior research has investigated the language skills of children and adults with DS using a variety of
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CELF editions (e.g., Heller et al., 2003; Laws, 2004; Laws & Bishop, 2003; Pennington et al., 2003).

The CLS is calculated by summing standard scores from four subtests. These are Concepts and Following Directions, Recalling Sentences, Formulating Sentences and Word Classes subtests. For children aged 13-years or older, the CLS is calculated using scores from the Word Definition subtest instead of Concepts and Following Directions. Each of these subtests are briefly described.

The Concepts and Following Directions subtest measures receptive language. On this subtest the examinee is first shown a picture of horizontally placed objects (e.g., ball, fish, house, shoe, car, apple). S/He is then asked to point to the pictures in an order specified by the examiner. For example on one item the examinee is asked to ‘Point to the big shoe, then point to the little apple’. This subtest measures how well children can understand language.

Recalling Sentences and Formulating Sentences measure expressive language. On the Recalling Sentences subtest the examinee is verbally presented with a sentence such as ‘the tractor was followed by the bus’ and asked to repeat it back verbatim. On the Formulating Sentences subtest children are shown a picture of an everyday event. Pictures include children playing a game or catching a bus. After a picture is shown children are provided with a word (e.g., playing). The child’s task is to make a sentence about the picture using the target word. Both the Recalling Sentences and Formulating Sentence subtests measure productive language skills.

On the Word Classes subtest children are verbally presented with four words. The examinee is first asked which two words are semantically related and
then asked to explain how they are related. For example, on one item the examinee is presented with the words ‘pillow’, ‘door’, ‘blanket’ and ‘lamp’. The two semantically related words are ‘pillow’ and ‘blanket’. Acceptable responses to explain how the two are related on this item are ‘they go on a bed’ or ‘you sleep on/with them’. The Word Classes measures both expressive and receptive language skills.

Finally, on the Word Definition subtest children are asked to define or provide a definition of a presented word. For example, on one item children are presented with the word ‘giraffe’ and asked to explain what this word means.

The Core Language Score (CLS) is standardised to a mean of 100 and standard deviation of 15. The CLS has been shown to have excellent reliability, with the CLS reliability coefficients ranging between .94 and .97 for children between 10- and 16-years old (Semel et al., 2003).

8.2.2.6 Measure of pragmatic language. The pragmatic scales from the Children’s Communication Checklist, 2nd Edition (CCC-2; Bishop, 2003) was used to measure pragmatic language skills. This instrument was selected for the current study because it provides a standardised measure of pragmatic language skills which are not assessed by other traditional language assessments (Bishop, Maybery, Wong, Maley, & Hallmayer, 2006). The first edition of the Children’s Communication Checklist has been used in previous research with children with DS (Laws & Bishop, 2004a).

The CCC-2 comprises 70-items, however of this total, 28-items measure pragmatic language skills. Only these items were used in the study. The CCC-2 is a checklist that is completed by a parent or guardian. Each item consists of a statement
describing an aspect of the child’s pragmatic language for example ‘Does not look at
the person s/he is talking to’. The respondent is asked to rate how often this
behaviour occurs on a 4-point likert scale. The response options are 0 = ‘less than
once a week (or never)’, 1 = ‘at least once a week, but not every day’, 2 = ‘once or
twice a day’ and 3 = ‘several times (more than twice) a day (or always)’. For the
analyses a total pragmatic language composite variable was created by summing
responses. Higher raw scores indicate poorer pragmatic language skills. However,
to aid in the accurate interpretation of the relationships between all cognitive
variables and behaviour problems, the CCC-2 scores were reversed prior to
undertaking correlational analyses in Study 2. Therefore higher scores on the CCC-2
are now representative of better pragmatic language skills. The pragmatic composite
had excellent internal consistency with Cronbach’s alpha equalling .94.

8.2.2.7 Measure of social skills. Social skills were measured using the
SSIS-RS parent report form (Gresham & Elliott, 2008). Although not widely used in
previous research with children with DS, at least one previous study has included this
instrument (Dinnebeil et al., 2013). The SSIS-RS consists of 46 items that measure
social skills. It should be noted that the items that measure social skills on the SSIS-
RS do not contribute to the composites that measure behaviour problems. The SSIS-
RS is a checklist that is completed by a parent or guardian about a child’s social
skills. Each item describes a prosocial behaviour such as ‘takes care when using
other people’s things’, ‘follows your directions’ and ‘shows concern for others’. A
parent or guardian rates each item on how frequently his/her child has displayed the
behaviour during the previous 2 months using a four-point likert scale. The response
options are 0 = ‘never’, 1 = ‘seldom’, 2 = ‘often’ and 3 = ‘almost always’.
There are seven subscales that measure social skills. These include Communication, Cooperation, Assertion, Responsibility, Empathy, Engagement and Self-Control. The Total Social Skills score is computed by summing all of the social skill items on the SSIS-RS and is an overall measure of learned behaviours that promote positive interactions with others (Gresham & Elliott, 2008). The Total Social Skills score was used in the analyses. Total raw scores range between 0 and 138, and the standardised score has a mean of 100 and SD of 15.

The social skills scales from the SSIS-RS has good reliability. The internal consistencies of the social skill subscales range from .74 to .87, and .95 to .96 on the total score for individuals 5-18 years old (Gresham & Elliott, 2008; Gresham et al., 2011). Total standard scores were used in the preliminary analyses. Raw scores were used for all dependent variables for the main analyses in Study 2.

**8.2.2.8 Behavioural measure of neurodegeneration.** Two subtests from the Cambridge Neuropsychological Test Automated Battery (CANTAB; Cambridge Cognition, 2006) were used to measure early signs of dementia related neurodegeneration. These were the Paired Associates Learning (PAL) and Graded Naming Test (GNT). The PAL and GNT were selected because both tests have been shown to be a sensitive measure of early cognitive changes related to dementia and Alzheimer’s disease (e.g., Blackwell et al., 2004; De Jager, Blackwell, Budge, & Sahakian, 2005; Égerházi, Berecz, Bartók, & Degrell, 2007; Swainson et al., 2001). The PAL test has been used in other research undertaken with children and adults with DS (e.g., Boada et al., 2012; Breslin et al., 2014; Edgin, Spano, Kawa, & Nadel, 2014; Visu-Petra, Benga, incaș, & Miclea, 2007) This GNT was developed by McKenna and Warrington (1983), which has previously been used studies undertaken with children and adolescents (e.g., Bishop, 2001; Bishop, North, &
Donlan, 1996; Goulandris, Snowling, & Walker, 2000). No study known to the author has used this task specifically with children with DS.

The PAL test is a measure of learning and memory for visuo-spatial information. In the PAL task, participants are seated in front of a touchscreen computer display. The computer display initially shows six white boxes placed around the screen (see Figure 3). During the first phase of testing each of the boxes ‘open’ revealing a non-verbalisable pattern. Once all boxes have been opened a target pattern appears in the middle of the screen. The examinee’s task is to touch the box where they saw this pattern before. If the examinee selects the wrong box, that is makes an error, all boxes are opened again. The examinee is then asked again to find the correct box. At the start of the subtest the examinee is required to find the location of one target pattern. However, as s/he continues through the test the difficulty increases by having the examinee identify two, three, then six and eight target patterns. Testing on the subtest is discontinued if the examinee makes one or more errors after ten attempts at a trial. The dependent variables obtained from the PAL subtest are the highest number of targets that could be identified and the total number of errors made.

Figure 3. Paired Associates Learning (PAL) task
The PAL task has good reliability, with test-retest reliability coefficients of .87 and .68 for the number of stimuli and error scores respectively (Cambridge Cognition, 2008). In the current study, the error scores have been reversed scored, so that a higher score indicates better performance on the PAL. From this point onwards this will be referred to as the ‘PAL performance’ score. Raw scores have been used in all statistical analyses. The preliminary analyses show that ceiling effects were found for the PAL number of stimuli measure in the TD group. Consequently, only the performance score is used in the correlational analyses.

The GNT is a measure of semantic memory. Semantic memory is a repository of everyday general knowledge and facts about the world. This includes knowing the concept of a ‘dog’ and ‘table’. On the GNT task children are shown a series of black and white pictures on a computer screen and are asked to identify the picture. For example, children are shown a picture of a kangaroo and asked “what is this?” Children are able to attain a maximum raw score of 30 on this subtest. The test-retest reliability of this task is excellent, with a reliability coefficient of .92 (Bird, Papadopoulou, Ricciardelli, Rossor, & Cipolotti, 2004). Raw scores were used in the analyses and indicate the number of pictures correctly identified.

Throughout this thesis the PAL performance and GNT scores are used as an indicator for a greater risk of developing dementia. It is acknowledged that dementia or AD is characterised by a decline in functioning during adulthood. For this reason, it is not possible to identify any child or adolescent with AD, as baseline cognitive functioning is still being established. However, as noted in the literature review, there is biological evidence that children with DS may begin to develop the neuropathology associated with AD from childhood (e.g., Leverenz & Raskind, 1998; Mehta et al., 2007). It was beyond the expertise of the author to examine these
biological markers and therefore it was necessary to select measures that have been shown to be sensitive to early cognitive decline. It is not the intention of the author to suggest that poor performance on the CANTAB tasks indicates that a participant has dementia. Rather it is cautiously suggested that poorer performance on these tasks might indicate a higher risk of developing AD in the future. Thus, this study seeks only to investigate whether a behavioural marker of early neuropathological change might be related to behavioural problems for children and adolescents with DS.

8.2.3 Procedure

Prior to the assessment sessions written informed consent was provided by a parent or legal guardian for all participants. Each participant was tested individually in a quiet room in his/her home, school or at the camp. The tasks were presented over three sessions with each lasting no more than 60 minutes. When participants had sufficient verbal skills, assent was also obtained from the child. During the sessions, no participant verbally indicated that they wanted to stop the session. For individuals that had little or no verbal communication skills, the researcher monitored the level of cooperation as an indicator for wanting to withdraw from the study. All participants were able to stay engaged in the activities with minimal need for reorienting them to the tasks, thus testing was not discontinued with any individual.

In the first session, the participants were presented with the WASI and the CANTAB tasks. In the second session the CELF-4 subtests were administered. In the final session, children completed the tasks from the NEPSY-II. Presentation of the assessments was counterbalanced within each session to average any carryover practice effects. Additionally, one parent or guardian was asked to complete the
CBCL parent report form, SSIS-RS parent report form and the Pragmatic Language Scale from the CCC-2. Parents were provided with a reply-paid envelope so that they could return the questionnaire directly to the researchers.

8.3 Results

8.3.1 Differences between the DS and Control Group on the Cognitive and Neuropsychological Measures

The first set of analyses compared the DS and age-matched control group on each of the cognitive and neuropsychological measures. Summary statistics for each of these tests are reported by group in Table 15. First, differences between the groups were examined using independent samples $t$-tests and the magnitude of the difference or effect size was summarised using Cohen’s $d$ (Cohen, 1988). To control for multiple comparisons $p$-values were adjusted using the Holm’s Step Down Procedure (Holm, 1979).

On all measures significant differences were found between the two groups. Specifically, the DS group obtained significantly lower scores on all subtests compared to the age-matched control group. Inspection of effect sizes reveals differences between the groups were largest for the measure of intelligence and language. On the measure of intelligence the difference between the groups was 7.07 SD units and for language 9.50 SD units. However, on these two measures the standard deviation of the control group is around five times larger compared to the DS group. This lack of variability in the scores in the DS group indicates potential floor effects for these measures. To address this problem composite scores for the IQ and language measures were recalculated by summing $z$-transformed raw scores from each subtest. For example, on the measure of IQ, raw scores from the Matrices
and Vocabulary subtests were converted to a $z$-score. The $z$-score from both subtests were then added together. For each subtest $z$-scores were calculated using the mean and standard deviation of the entire group.
### Table 15. Summary Statistics for all Cognitive and Neuropsychological Tasks, According to Group

<table>
<thead>
<tr>
<th>Variable</th>
<th>DS group (n = 24)</th>
<th>TD group (n = 24)</th>
<th>Comparison of Means</th>
</tr>
</thead>
<tbody>
<tr>
<td>WASI (Intelligence)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Full Scale IQ</td>
<td>56.13 ± 2.07</td>
<td>106.63 ± 9.89</td>
<td>t-value 24.48, p &lt; .001, Cohen’s d -7.07</td>
</tr>
<tr>
<td>NEPSY-II (Emotion Recognition &amp; ToM)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Affect Recognition</td>
<td>2.04 ± 1.55</td>
<td>9.63 ± 2.90</td>
<td>t-value 11.31, p &lt; .001, Cohen’s d -3.27</td>
</tr>
<tr>
<td>Theory of Mind</td>
<td>9.79 ± 4.48</td>
<td>25.33 ± 2.32</td>
<td>t-value 15.09, p &lt; .001, Cohen’s d -4.36</td>
</tr>
<tr>
<td>CELF-4 (Expressive &amp; Receptive Language)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Core Language Score</td>
<td>40.75 ± 1.96</td>
<td>104.71 ± 10.22</td>
<td>t-value 30.12, p &lt; .001, Cohen’s d -9.50</td>
</tr>
<tr>
<td>CCC-2 (Pragmatic Language)</td>
<td>51.29 ± 9.55</td>
<td>78.04 ± 6.17</td>
<td>t-value 11.52, p &lt; .001, Cohen’s d -3.33</td>
</tr>
<tr>
<td>Pragmatic Language Composite</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SSIS-RS (Social Skills)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total Social Skills Composite</td>
<td>83.25 ± 11.24</td>
<td>100.67 ± 11.34</td>
<td>t-value 5.35, p &lt; .001, Cohen’s d -1.54</td>
</tr>
<tr>
<td>CANTAB (Dementia)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PAL number of stimuli</td>
<td>5.96 ± 2.99</td>
<td>8.00 ± 0.00</td>
<td>t-value 3.35, p &lt; .003, Cohen’s d -0.97</td>
</tr>
<tr>
<td>PAL performance</td>
<td>13.92 ± 6.81</td>
<td>26.71 ± 1.08</td>
<td>t-value 9.08, p &lt; .001, Cohen’s d -2.62</td>
</tr>
<tr>
<td>Graded Naming Test</td>
<td>4.71 ± 2.48</td>
<td>13.96 ± 3.51</td>
<td>t-value 10.56, p &lt; .001, Cohen’s d -3.05</td>
</tr>
</tbody>
</table>


*All variables have been scored so that higher scores indicate better performance. Positive values indicate DS children were reported to have higher scores on the measure.
The recalculated scores for intelligence and language are presented in Table 16. Differences between the groups on the transformed IQ and language scores were examined using independent samples $t$-tests. Transformed scores obtained for the DS group were found to be significantly lower than the comparison group ($t(46) = 23.63, p < .001$ and $t(46) = 27.96, p < .001$ for intelligence and language respectively). However, no significant differences in standard deviations were observed between the groups on measures of IQ ($F = 0.51, p = .480$) or language ($F = 0.13, p = .132$).
Table 16. Re-calculated z-scores for IQ and Language, According to Group

<table>
<thead>
<tr>
<th>Variablea</th>
<th>DS group (n = 24)</th>
<th>TD group (n = 24)</th>
<th>Comparison of means</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>Min.</td>
</tr>
<tr>
<td><strong>WASI</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Full Scale IQ</td>
<td>-0.83</td>
<td>0.24</td>
<td>-1.12</td>
</tr>
<tr>
<td><strong>CELF-4</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Core Language Score</td>
<td>-1.12</td>
<td>0.24</td>
<td>-1.29</td>
</tr>
</tbody>
</table>

a All variables have been scored so that higher scores indicate better performance
8.3.2 Association between Cognitive Skills and Behaviour Problems

The next set of analyses investigated whether one or more of the cognitive/neuropsychological variables were related to different types of behavioural problems in the DS and TD groups. Spearman’s Rho correlations were used to investigate whether one or more of the cognitive/neuropsychological variables correlated with a measure of Externalising, Internalising or Total Behaviour problems (as noted earlier behavioural composite scores were derived by averaging z-scores (using raw scores) on the CBCL and SSIS-RS broadband behaviour scales). Spearman’s Rho analyses were selected because the correlations were being computed for a set of ordinal variables. These correlation analyses were undertaken separately for each group and are presented in Table 17 and Table 18 for the DS and control group respectively. This approach was used to examine whether the cognitive and neuropsychological measures were related to the behaviour problems in each group.

Results from the correlation analyses revealed few significant associations between cognitive/neuropsychological and behaviour measures for both the DS and TD group. Common to both groups was that the measure of pragmatic language was found to be significantly correlated with externalising behaviour problems. That is, in the TD and DS groups children/adolescents with few pragmatic language problems also experienced fewer externalising behaviour problems. For the DS group, this trend was also observed for internalising problems. Also common to both groups was that the measure of social skills was correlated with the Total Behaviour Problems composite. Specifically, participants with or without DS that had poorer social skills, showed fewer behaviour problems overall.
The results also showed significant associations that were specific to each group. For the TD group, children with higher IQ were found to have more externalising problems. In the DS group, individuals who performed better on the ToM tasks had more internalising problems. The behavioural measure of neurodegeneration was found to be significantly negatively correlated with internalising problems. This association was not observed in the TD group.
Table 17. Spearman’s Rho Correlations between Behaviour Problem Composites and Cognitive/Neuropsychological Variables in Down Syndrome Group (n = 24)

<table>
<thead>
<tr>
<th>Variable</th>
<th>1.</th>
<th>2.</th>
<th>3.</th>
<th>4.</th>
<th>5.</th>
<th>6.</th>
<th>7.</th>
<th>8.</th>
<th>9.</th>
<th>10.</th>
<th>11.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Total Problems Composite</td>
<td></td>
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<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>2. Externalising Problems Composite</td>
<td>.332</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Internalising Problems Composite</td>
<td>.540**</td>
<td>.332</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Full Scale IQ</td>
<td>.247</td>
<td>-.298</td>
<td>.189</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Affect Recognition</td>
<td>-.160</td>
<td>-.249</td>
<td>-.044</td>
<td>.329</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Theory of Mind - Total</td>
<td>.314</td>
<td>-.048</td>
<td>.502&quot;</td>
<td>.518**</td>
<td>.429&quot;</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>7. Core Language Score</td>
<td>.204</td>
<td>-.197</td>
<td>.386</td>
<td>.604**</td>
<td>.203</td>
<td>.449&quot;</td>
<td></td>
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</tr>
<tr>
<td>8. Pragmatic Language</td>
<td>-.097</td>
<td>-.494&quot;</td>
<td>-.440&quot;</td>
<td>-.012</td>
<td>.233</td>
<td>-.188</td>
<td>-.093</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>9. Total Social Skills</td>
<td>.606**</td>
<td>-.350</td>
<td>-.010</td>
<td>.317</td>
<td>.006</td>
<td>.138</td>
<td>.167</td>
<td>.499&quot;</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>10. Paired Associates Learning</td>
<td>-.284</td>
<td>-.342</td>
<td>-.485&quot;</td>
<td>.015</td>
<td>-.079</td>
<td>-.282</td>
<td>-.069</td>
<td>.032</td>
<td>-.103</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11. Graded Naming Test</td>
<td>.055</td>
<td>-.030</td>
<td>.296</td>
<td>.205</td>
<td>.116</td>
<td>.366</td>
<td>.544&quot;</td>
<td>-.179</td>
<td>-.085</td>
<td>-.173</td>
<td></td>
</tr>
</tbody>
</table>

*p < .05, **p < .01, ***p < .001
Table 18. Spearman’s Rho Correlations between Behaviour Problem Composites and Cognitive/Neuropsychological Variables in Typically Developing Group (n = 24)

<table>
<thead>
<tr>
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</thead>
<tbody>
<tr>
<td>1. Total Problems Composite</td>
<td></td>
<td></td>
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<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>2. Externalising Problems Composite</td>
<td>.333</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Internalising Problems Composite</td>
<td>.308</td>
<td>.570**</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Full Scale IQ</td>
<td>.324</td>
<td>.415*</td>
<td>.121</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Affect Recognition</td>
<td>.304</td>
<td>.139</td>
<td>-.138</td>
<td>.412*</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Theory of Mind - Total</td>
<td>.111</td>
<td>.217</td>
<td>-.010</td>
<td>.612**</td>
<td>.448*</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Core Language Score</td>
<td>.408*</td>
<td>.337</td>
<td>.081</td>
<td>.798***</td>
<td>.524**</td>
<td>.646**</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Pragmatic Language</td>
<td>-.022</td>
<td>-.514*</td>
<td>-.403</td>
<td>-.053</td>
<td>.273</td>
<td>.068</td>
<td>.167</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Total Social Skills</td>
<td>.683***</td>
<td>-.300</td>
<td>-.345</td>
<td>.014</td>
<td>.275</td>
<td>-.055</td>
<td>.207</td>
<td>.354</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11. Graded Naming Test</td>
<td>-.026</td>
<td>.373</td>
<td>-.005</td>
<td>.671***</td>
<td>.257</td>
<td>.586**</td>
<td>.642**</td>
<td>.099</td>
<td>-.300</td>
<td>.263</td>
<td></td>
</tr>
</tbody>
</table>

*p < .05, **p < .01, ***p < .001
8.4 Discussion

The aim of this study was to examine the extent to which cognitive skills and a neuropsychological measure of dementia related to behaviour problems in children and adolescents with DS. Results from this study provide evidence that a range of cognitive problems are present in individuals with DS. The initial set of analyses showed that the DS group performed significantly poorer than the control group on all cognitive and neuropsychological measures. The correlation analyses indicated that for the DS group, pragmatic language, ToM, social skills and a neuropsychological measure linked to dementia was significantly correlated with some of the behaviour problems. This study provides evidence that some of the behaviour problems in persons with DS may be related to one or more cognitive impairments. Each hypotheses tested in this study is now addressed.

8.4.1 The Relationship between IQ and Behaviour Problems

It was hypothesised that IQ would negatively correlate with behavioural problems. That is participants with lower IQ’s would have more behaviour problems. For the DS group, no significant correlations were observed between IQ and any measure of behaviour problems. The results from the control group were also inconsistent with this hypothesis, where a positive significant association was observed between IQ and externalising problems.

As noted earlier few studies have investigated whether IQ is related to behaviour problems in children and adolescents with DS. The results observed in this study are consistent with research by Dykens et al. (2002) and Dykens and Kasari (1997) who also observed no significant correlations between IQ and behaviour problems for participants with DS. The current study extends this work by replicating past findings using a different standardised test of intelligence. In the
current study IQ was measured using the WASI (Wechsler, 1999). In the research by Dykens et al., the Kaufman Brief Intelligence Test or Stanford–Binet Intelligence Test was used. Thus the finding that behaviour problems are not significantly correlated with IQ appears to be quite robust. Also, in the research by Dykens et al. participants with DS were aged between 4- and 19-years. One potential criticism of this method is that changes in the type of behaviour problems over the lifespan may mask associations with IQ. The results of this study discount this as an explanation. The participants with DS in the current study were aged between 10- and 15-years, which is relatively narrower compared to previous work.

The results of the current study were inconsistent with the findings of van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013). In their research a significant association between lower intellectual functioning and behavioural problems was reported. One potential reason for this discrepancy might have arisen from the method used to quantify IQ. In the studies by Dykens et al. and also the current study, IQ was measured using an individually administered test of intelligence. In contrast, van Gameren-Oosterom, Fekkes, van Wouwe, et al. (2013) measured IQ with the Dutch social competence rating scale (Kraijer, Kema, & De Bildt, 2004). This is a validated instrument that measures self-help skills and independence in individuals with an intellectual disability between 4- and 18-years old. It provides a broad estimate of degree of intellectual disability and IQ; (i) profound = <20, (ii) severe = 20-34, (iii) moderate = 35-49 and (iv) mild = 50-69.

Collectively, the results of Dykens et al. and those observed in the current study question whether IQ is related to behavioural problems in people with DS. As noted in Chapter 5, it has been proposed that low IQ represents a limit in cognitive resources that individuals have to regulate their own behaviour and
negotiate social interaction (Moffitt, 1993). Another proposal was that individuals with low IQ might become frustrated with having difficulty completing mental tasks such as completing a classroom activity (Goodman et al., 1995). One consequence of this problem is an increase in externalising behaviour problems. The results of this study question whether such proposal can be applied to individuals with DS.

The positive relationship found between IQ and behaviour problems in the control group also cast some doubt on its applicability to TD children and/or adolescents, given that this finding was also inconsistent with the theorised relationship between IQ and behaviour problems.

### 8.4.2 The Relationships between Emotion Recognition and Behaviour Problems

It was also predicted in this study that emotion recognition would be correlated with behaviour problems in children and adolescents with DS. That is, participants with poor emotion recognition skills would have more behaviour problems. The results from the study did not support this hypothesis in the DS or control group. Initial analyses showed that the DS group performed significantly poorer than the control group on the emotion recognition task. However, the correlation between the measure of emotion recognition and behaviour problems was not found to be significant for the DS or control group. The results showed that emotion recognition deficits are present in children and adolescents with DS, and this finding is consistent with past research. Studies by Williams et al. (2005) and Wishart et al. (2007) have both reported that individuals perform more poorly than controls on a task similar to the one used in this study.

While the DS group appeared to have difficulty with emotion recognition, correlation analyses did not show that this was related to their behaviour problems. This same result was also observed in the control group. These findings...
question the proposal by Walker (1981), Rojahn et al. (1995) and also Larkin et al. (2013) who proposed emotion recognition problems may lead to behaviour problems. It is possible that emotion recognition problems might lead to behaviour difficulties in other groups. For example, Aspan, Vida, Gadoros, and Halasz (2013) did find that emotion recognition problems were related to externalising behaviour problems in male children identified with conduct problems. However, there was no evidence found in the current study to suggest that an association exists in individuals with DS.

**8.4.3 The Relationship between ToM and Behaviour Problems**

In this study it was predicted that a measure of ToM would be related to both internalising and externalising behaviour problems. The results partially supported this hypothesis. For the DS group, a significant correlation was found between ToM and internalising behaviour problems. However, unexpectedly, better ToM performance was associated with more internalising problems. As anticipated the children with DS performed significantly worse than the control group on the ToM task overall. Thus, it appears that children with DS that had less severe deficits in ToM showed more internalising problems. The correlation between ToM and externalising problems was not significant. The relationship between ToM and internalising problems appears to be specific to the DS group. There were no significant correlations between ToM and behaviour problems for the control group.

To the author’s knowledge, this study is the first to examine whether ToM is related to behaviour problems in children and adolescents with DS. The significant association observed between the ToM measure and internalising behaviour problems is inconsistent with Hughes and Leekam’s (2004) position. They argued that problems in understanding the mental states of others might lead to
both internalising and externalising problems. It is interesting to note that ToM was only found to be related to internalising problems. An alternative explanation of the relationship between internalising problems and ToM might be that better ToM skills predispose individuals to be more aware of how others’ perceive them. Consequently these individuals might be more attuned to others’ criticisms, putting them at greater risk for developing negative self-perceptions and internalising problems (Cutting & Dunn, 2002). However, there was a verbal component to the ToM task that might also partially account for these results. It is possible that children with DS who had better expressive language were able to perform better on the verbal ToM task, as well as communicate to others their feelings of anxiety or depression. It would be interesting to examine the association between ToM and behavioural problems with a larger sample of children with DS, where it was possible to control for the effects of language.

8.4.4 The Relationship between Language and Behaviour Problems

Another hypothesis forwarded in this study was that behaviour problems would be correlated with language skills. In this study two aspects of children’s language skills were measured. One was their ability to understand and produce language. The other was the extent participants correctly used communicative conventions during social exchanges (otherwise known as pragmatic language skills). Results showed mixed support for this hypothesis. The ability to understand and produce language was not found to be related to any behaviour problem measure for the DS group. In contrast to expectations, children/adolescents with better expressive/receptive language had more total behaviour problems in the control group. However, for both groups, pragmatic language skills were related to externalising problems, as well as internalising behaviour problems in the DS group.
Initial analyses conducted found that the DS group performed significantly poorer than the controls on the Core Language Score that measures comprehension and production skills and also the measure of pragmatic language. These findings are consistent with a number of previous studies showing poor communicative functioning in individuals with DS. As noted earlier, a meta-analysis by Næss et al. (2011) found that DS was associated with poor production and comprehension skills. Also, a study by Laws and Bishop (2004a) found poorer pragmatic skills in persons with DS. Thus the results of this study provide further evidence that individuals with DS experience a range of communicative problems.

The correlation analyses indicated pragmatic language, but not oral language skills, were related to externalising and internalising behaviour problems. This association was observed in the DS and control group for externalising problems, and DS group for internalising problems. This is the first study known by the author to examine the relationship between pragmatic language skills and types of behavioural problems in children and adolescents with DS. Therefore, the current study provides evidence that for individuals with DS, their ability to respond appropriately in social situations may be contributing to their behavioural problems. The relationship between pragmatic language and externalising behaviour problems was also found in the control group. Collectively, this pattern of results supports previous researchers’ claims that communication skills play an important role in behaviour problems (e.g., Durkin & Conti-Ramsden, 2010; Durkin & Conti-Ramsden, 2007; Laws et al., 2012; Menting et al., 2011).

8.4.5 The Relationship between Social Skills and Behaviour Problems

Poorer social skills were predicted to be related to more behaviour problems. The correlation analyses did not support this hypothesis. For both the DS
and control group there was a positive relationship between the measure of social
skills and total behaviour problems. A somewhat confusing finding was that in both
the non-significant negative correlation between social skills and externalising and
internalising behaviour problems was found. This was not reflected in the direction
of the relationship between social skills and total behaviour problems. One reason
for this may be that the total problem scores on the CBCL and SSIS-RS include
items that do not contribute to the externalising or internalising subscales. The
CBCL includes 15 items that assess Thought Problems, 11 items on Social Problems,
10 items on Attention Problems and 11 items assessing ‘Other’ Problems, that do not
contribute to a specific scale. Similarly, the SSIS-RS includes Bullying,
Hyperactivity/Inattention and Autism Spectrum problems scales that consist of 5-, 7-
and 7-items respectively. As the composite scores used in the analyses were
computed from the total scores on both of these measures, these additional items
were included in the total problems composites. Therefore, it is possible that the
strong positive association between social skills and the total behaviour problems
reflects a relationship between other forms of behavioural dysfunction that are not
classified by the instruments as either externalising or internalising. As noted earlier,
the study by van Gameren-Oosterom, Fekkes, Reijneveld, et al. (2013) reported
poorer social skills in 322 adolescents aged between 16.8 and 19.1 years, with DS.
The current study extends this research by showing that poor social skills are also
evident in a younger sample of children and adolescents with DS. The age range of
the participants with DS in the current study was 10- to 15-years. Thus it seems that
poor social skills might be present from childhood onwards. Poor social skills have
been linked to academic and social problems (Malecki & Elliot, 2002; McClelland,
Morrison, & Holmes, 2000). However, the evidence in this study indicated that
better social skills were related to more overall behaviour problems for participants
with and without DS. This finding contrasts with evidence suggesting that social
skills interventions reduce behavioural problems (Cook et al., 2008). As this is the
first study known to the author that has examined the association between
behavioural problems and social skills in children and adolescents with DS, this
unexpected result requires validation in larger samples.

8.4.6 The Relationship between Early Neurodegeneration and Behaviour
Problems

The final hypothesis tested in this study was that a neuropsychological
marker linked to dementia would be correlated with internalising behaviour problems
in children and adolescents with DS. The results did provide support for this
hypothesis. A significant correlation was observed between internalising problems
and a behavioural measure of early onset dementia for the DS group.

In this study, early neurodegeneration was measured using the PAL
subtest from the CANTAB. Previous research has shown this subtest to be sensitive
to the early onset of dementia in Alzheimer’s patients (e.g., Blackwell et al., 2004;
De Jager et al., 2005; Égerházi et al., 2007; Swainson et al., 2001). The first set of
analyses showed that the DS group performed significantly poorer on this task
compared with the control group. This was consistent with expectations, given that
previous research has demonstrated a range of cognitive impairments and high risk
of dementia in people with DS (Korenberg et al., 1994; Määttä et al., 2006;
Pennington et al., 2003; Pueschel, 1990).

The significant correlation observed between internalising behaviour
problems and the PAL subtest provide support to claims by Dykens and colleagues
(Dykens & Kasari, 1997; Dykens et al., 2002). The proposal by these researchers is
that behaviour problems occur in people with DS because of neuropathological changes associated with AD (e.g., accumulation of neuritic plaques and neurofibrillary tangles) which results in functional brain changes (Visootsak & Sherman, 2007). Thus, the significant association observed between the PAL subtest and internalising behaviour problems provide initial support for Dykens et al.’s proposal.

In the DS group, poorer performance on the PAL task was significantly related to more internalising problems. It is interesting that this association was specific to internalising symptoms. Thus, this supports Dykens and colleagues (1997; 2002) neurodegenerative hypothesis. At a theoretical level it is of importance that this association was found to be exclusive to internalising behaviour problems (i.e., PAL performance did not significantly relate to externalising or total behaviour problems), and that it was only found in the DS group.

This is the only known study to empirically investigate a relationship between behaviour problems and behavioural markers of early AD in children and adolescents with DS. Consequently, the results here provide the first evidence that there may be a link between early dementia and internalising problems in children and adolescents with DS. Even though the PAL task was the only subtest to emerge as relating to internalising symptoms in this group, it is possible that the semantic memory problems might be related to internalising symptoms in older individuals with DS, when presumably signs of AD will become more common. Thus, it would be interesting to examine this relationship in late adolescence and early adulthood.

Obvious limitations of the current study design are also apparent, as completing assessment tasks on a single occasion does not allow for testing cognitive decline over time. Thus, there is certainly a need to examine this finding using larger
samples, and measuring performance on neuropsychological tasks using a longitudinal design.

8.4.7 Conclusions

This study examined the correlates of behaviour problems in a sample of children and adolescents with and without DS. The first finding from this study was that the participants with DS performed more poorly on measures of IQ, emotion recognition, ToM, language and pragmatic language, social skills and neuropsychological measures of dementia than individuals in the TD group. The second finding was that for the DS group, only the measures of pragmatic language, ToM, social skills and dementia were related to behaviour problems.
CHAPTER 9: GENERAL DISCUSSION & CONCLUSIONS

This thesis examined behaviour problems in children and adolescents with DS. The research question investigated was whether behavioural problems in children and adolescents with DS were associated with cognitive and neuropsychological deficits, and/or reflect an issue related to maturation. Study 1 examined behaviour problems in detail in persons with DS and a comparison group comprising TD individuals of comparable age. Study 2 investigated cognitive and neuropsychological correlates of the behaviour problems in children and adolescents with and without DS. This chapter discusses the overall findings of the study with respect to the underlying nature of DS that was considered in Chapters 3 – 5. Specifically, discussion focuses on whether the types of behaviour problems found in people with DS are unique to this group. The second section considers the clinical implications of the study findings with respect to identifying and supporting behaviour problems in children and adolescents with DS. In the final section, the limitations of the current study and avenues for future research are considered.

9.1 The Nature of Behaviour Problems in Down Syndrome: The Role of Cognitive and Social Skills

The survey of the literature presented in Chapter 3 revealed that between 17% and 38% of children and adolescents with DS present with clinically significant behaviour problems. Furthermore, between 7% and 15% can be identified with externalising problems and 5% and 14% with internalising problems. These rates were comparable to the number of problems in the sample of individuals with DS who participated in the studies in this thesis. As a reminder, 16.7% of participants in this thesis were identified as having problems in the clinical range on all three composite scores on the CBCL. Thus, the total problem scores for the current sample were consistent with the lower estimates of problems reported in other studies, while
the externalising and internalising problems were slightly above the range reported elsewhere. Externalising and internalising problems in childhood have been linked to a range of negative social, academic and psychosocial outcomes (Loeber, Stouthamer-Loeber, & Raskin White, 1999; Masten et al., 2005; Pine, Cohen, Cohen, & Brook, 1999; Rapport et al., 2001). Given this, it is important to understand the reasons why behaviour problems are present in this group.

Understanding the underlying reasons for the behaviour problems in people with DS can be considered particularly challenging. As noted in Chapter 3, it is certainly not the case that all individuals with DS have the same problems. The results of Study 1 provide further support to this claim. In particular, clinical and sub-clinical aggressive behaviours and somatic complaints were observed in 25% and 21% of the DS sample respectively. Conversely, other externalising and internalising problems affected fewer individuals with DS. Only 4.2% of participants showed symptoms of anxiety or depression, or rule-breaking behaviours within the borderline or clinical ranges on the CBCL. Thus, it can be seen that a small percentage of the sample were reported to have different or additional externalising and internalising problems compared with the remainder of participants.

Another issue to note is that the severity and the type of behaviour problems in individuals with DS are not comparable to the problems seen in TD children or even those seen in other types of developmental disorders characterised by low IQ. In Study 1 the DS group were found to have significantly more behaviour problems compared to an age-matched control group. The studies reviewed in Chapter 4 showed that the behaviour problems in people with DS were not comparable to those seen in children or adolescents with Prader-Willi, Williams
and Smith-Magenis syndromes and Autism Spectrum Disorder. Thus, based on the research findings of this thesis and past research it seems that the type and nature of behaviour problems found in children and adolescents with DS are quite unique to this group.

Chapter 5 considered whether one or more cognitive problems might be contributing to the behaviour problems of people with DS. The research reviewed in Chapter 5 identified studies that had reported poorer communicative, cognitive and social skills in children and/or adolescents with DS (e.g., Laws & Bishop, 2004b; Næss et al., 2011; van Gameren-Oosterom, Fekkes, Reijneveld, et al., 2013; van Gameren-Oosterom, Fekkes, van Wouwe, et al., 2013). A finding to emerge from Study 2 was that it is not the case that individuals with DS have a single cognitive or social impairment. Rather, evidence from this study indicates multiple cognitive and social problems. Specifically, in Study 2, the DS group performed significantly more poorly on the measures language, emotion recognition, ToM, social skills as well as intelligence.

It was interesting to find that only pragmatic language, ToM, social skills and a behavioural marker of neurodegeneration was related to behaviour problems in participants with DS. Pragmatic language was found to be related to behaviour problems in the control group as well. Thus it seems that despite multiple cognitive impairments in people with DS, not all are related to behaviour problems. The significant relationship observed between the measure of early dementia and internalising problems appears to be consistent with a proposal forwarded by Dykens and colleagues (1997; 2002). According to their proposal internalising problems that present in people with DS are related to neurodegeneration. Should this proposal be accurate, it would mean there are aspects of behaviour problems that are unique to
this group. In this case it is possible that early dementia may be contributing to internalising behaviour problems not found in other genetic disorders.

There may be aspects of behaviour problems in individuals with DS that are common to both other developmental disorders and typical development as well. In Study 2 a significant association was observed between pragmatic language and externalising problems in the DS and control groups. This might indicate that there are aspects of behaviour problems in people with DS that are similar to that seen in typical development. That is, children and adolescents with poor pragmatic skills will be more likely to have behaviour problems irrespective of whether they have DS or are typically developing. This might indicate that some cognitive deficits lead to behavioural problems regardless of the overall cognitive ability of the individual.

The research presented in this thesis examined cognitive influences on the behaviour problems of children and adolescents with DS. However, research has shown that there are parental and environmental influences that also play a role in the behaviour problems of children without DS. As an example, aggressive parenting practices such as spanking has been linked with higher levels of child aggression (Stormshak, Bierman, McMahon, & Lengua, 2000). Parental discipline has also been shown to be important for behavioural problems. Authoritarian (i.e., strict and non-responsive), lax or permissive and inconsistent discipline, as well as punitive practices have been linked with childhood externalising and/or internalising problem behaviours (Campbell, 1995; Keown & Woodward, 2002; Rankin Williams et al., 2009; Stormshak et al., 2000).

It is possible that some of the non-significant associations between cognitive and behavioural variables relate to environmental or family factor mediation effects. To use the example of parenting practices again, the way in which
a parent interacts with and disciplines her/his child might not only impact the child’s behaviour, but also their cognitive development. In applying this to some of the cognitive skills examined in this thesis, there is evidence that this might be the case for emotion recognition and language skills. There is empirical support for the early child-caregiver relationship influencing the emotion recognition skills of older children. Steele, Steele, and Croft (2008) conducted a longitudinal study examining the relationship between attachment during infancy and emotion recognition ability at the ages of 6- and 11-years. The results indicated that children who had a stronger attachment to their mothers performed significantly better when asked to identify emotions from facial affect (e.g., happy, sad, angry, disgust) at 6-years of age. At 11-years of age, this association was approaching statistical significance. This suggests that children’s early cognitive development is influenced by the quality of the caregiver-child relationship, however this association might weaken over time.

Similarly, there is research linking parental practices with language development (Karrass & Braungart-Rieker, 2003; Scheffner Hammer, Bruce Tomblin, Zhang, & Weiss, 2001). Scheffner Hammer et al. (2001) found that parents of preschool children with a developmental language disorder engaged in more frequent discipline of their child than those without a language impairment. In this study it was also found that parents of TD children engaged their child in more conversational activities compared with the parents of participants with language disorders. This suggests that exposure to language in the family context might be important for language development. Therefore, it might be that there is a relationship between language skills and behavioural problems in individuals with DS, however this is mediated by the degree to which parents attempt to engage their child in conversation.
9.2 Clinical Implications of the Findings

The results of this thesis have clinical implications with respect to identifying and supporting children with DS. It is clear from the literature and also from Study 1 that behaviour problems are not present in all children or adolescents with DS. A challenge for families and clinicians is to identify those who might be at risk. An ideal scenario might be to provide support before the problems emerge in order to prevent the negative outcomes associated with behaviour problems. Study 2 delineates two potential cognitive markers of behavioural problems in children and adolescents with DS. The first is performance on the Paired Associates Learning (PAL) task from the CANTAB, which in this thesis was used to measure early onset of dementia (Cambridge Cognition, 2006). The PAL task may be useful for screening children with DS who are at risk for developing internalising problems. This has the potential to be a valuable clinical tool because the test does not depend on participants providing a verbal response. This is important following results presented in Study 2 showing very severe language problems in the group.

A test of pragmatic language skills might also be useful to identify children with DS who are at risk of behaviour problems. In Study 2 this measure correlated with both externalising and internalising problems. The instrument used in this task was the Children’s Communication Checklist-2 (CCC-2; Bishop, 2003). This instrument is a parental checklist and therefore, completion of the form is also not dependent on the oral language skills or cooperation of the child or adolescent.

The results of this research offer potential suggestions for remediating behaviour problems in individuals with DS. One promising avenue is that internalising and externalising behaviour problems might be reduced by improving pragmatic language skills. Existing interventions have been successful in
significantly improving children’s pragmatic language skills (e.g., Adams, Lloyd, Aldred, & Baxendale, 2006; Adams et al., 2012; Hyter, Rogers-Adkinson, Self, Simmons, & Jantz, 2001). As an example, Adams et al. (2012) conducted a randomised controlled trial of 88 children between 5.9- and 10.7-years old, with pragmatic language or communication needs. This study examined the effectiveness of a manualised intervention program for improving structural and pragmatic aspects of language. The intervention included 16-20 one-hour sessions with a speech and language therapist or specially trained therapy assistant. Sessions were individualised to the needs of each child but targeted high-level language skills such as semantics, pragmatics, social interaction, and interpretation of social cues.

Participants in the control group received their treatment as usual with their local speech and language specialist. The CELF-4 (Semel, 2006) and CCC-2 (Bishop, 2003) were used to assess structural and pragmatic language skills respectively. The results indicated that the intervention did not have a significant effect on the structural components of language, however there was a significant improvement in pragmatic language skills. Investigation of whether pragmatic language interventions are able to reduce behavioural problems in children and adolescents with DS might be more useful at present than focusing on the relationship between dementia and internalising problems. One reason for this is that pragmatic language problems appear to relate to both externalising and internalising behavioural problems in this group. Therefore, by targeting these skills clinicians might be able to simultaneously reduce the impact of both types of behavioural difficulties in individuals with DS. Additionally, finding that internalising problems may relate to early dementia does not provide a promising avenue for prevention or reduction of internalising symptoms at present. That is, until medical advances are made to be
able to reduce or reverse the neuropathological effects of AD, there may not be a lot that can be done for dementia-related internalising problems.

This thesis did not investigate the role that external factors may have in determining the behaviour of children and adolescents with DS. However it is likely that there are environmental variables that also influence whether a child may develop externalising or internalising behaviour problems. Research is needed to examine how different environmental variables might serve to increase or decrease specific problems for children with DS. For example, family and school variables might be particularly important for determining how behaviour problems develop and change over time. Therefore, it is proposed that once research identifies other environmental influences on behaviour problems seen in children with DS, pragmatic language interventions might be used together with environmental approaches in order to achieve the greatest reductions in problem behaviour. 9.3 Lessons Learnt Regarding the Sensitivity of Standardised Assessments in Populations with Intellectual Disability

The research undertaken as part of this thesis unexpectedly revealed limitations in the sensitivity of the language and intelligence tests used. In Study 2 it was found that converting raw scores to standard scores for the CELF-4 (which measures language) and the WASI (which measures IQ) removed virtually all the variability in the scores in the DS group. As a consequence it was difficult to examine differences between the children and adolescents with DS. The extent of this insensitivity was unexpected as both the CELF-4 and WASI have previously been used in research with individuals with DS (e.g., Edgin et al., 2010; Groen et al., 2006; Heller et al., 2003; Laws & Bishop, 2003; Pennington et al., 2003). In clinical settings this may have the impact of masking strengths and weakness in language
and intelligence. This is a concern since planning the support required for a child is influenced by scores from standardised tests. Moreover, the unanticipated finding in this study indicates that in the cognitive assessment of children, tests are required that can better discriminate between different ability levels at the lower end of the distribution.

9.3 Limitations and Future Directions

There are several limitations which must also be considered when interpreting the results of this thesis. Firstly, the sample of participants in the current study was small. Findings from small studies are less likely to accurately estimate an effect or association compared with large samples (Christley, 2010; Hackshaw, 2008). Several unexpected findings were observed with regard to the direction of the relationship between some behaviour problems and IQ, language and social skills in the TD and/or DS group. Thus future research conducted with a larger sample size will be required to determine the accuracy of the results presented in this thesis.

A second limitation is that the relationships between behaviour problems and the cognitive/neuropsychological variables were analysed using a correlational method. As a reminder, one of the results from Study 2 suggested that poor pragmatic language is associated with more externalising problems. The results were interpreted to suggest that children or adolescents with poor pragmatic language skills are more likely to develop externalising behavioural problems. However, it is also possible that externalising problems cause pragmatic language difficulties or that there is another variable that is responsible for these two variables co-occurring. It seems unlikely that externalising problems would cause pragmatic language impairment in this population. Indeed, it has been noted elsewhere that there is little evidence for behavioural problems causing communication impairments in children.
(Cross, 2004). However, the possibility of another variable underlying the correlation between behaviour problems and pragmatics cannot be excluded. One such variable might be a diagnosis of Autism. Children with co-occurring DS and ASD might be more prone to behavioural problems. In the current sample, no participant with or without DS was known to have any other clinical disorder. However, participants were not actively assessed for ASD. Impaired communication and pragmatic language skills can be symptoms of Autism (American Psychiatric Association, 2000) and it is estimated that around 10% of individuals with DS also have ASD (Dykens, 2007; Pary & Hurley, 2002). In Chapter 4 it was shown that ASD comparison groups have more behaviour problems than children and adolescents with DS (Griffith et al., 2010; Grizenko et al., 1991). Thus, if some children have undiagnosed ASD then this might be contributing to the behavioural problems in this clinical group. An additional consideration here is that a significant correlation between pragmatic language and externalising problems was also observed in the control group. Tentatively, this might suggest that an association between pragmatics and externalising behavioural problems may exist irrespective of whether a child does or does not have a developmental disorder.

Longitudinal research would build upon the results from this study by helping to clarify the nature of the relationships between variables. The current study found that externalising and internalising behavioural problems varied according to pragmatic language in individuals with DS and to a lesser extent, TD participants. Conversely, only internalising problems in the DS group differed on the basis of ToM and a neuropsychological measure of AD (PAL task). Longitudinal studies would help to clarify whether poor pragmatic language and neurodegeneration cause internalising and/or externalising problems, or if pre-
existing behavioural problems cause pragmatic language difficulties or poor performance on the PAL task.

The results of this thesis showed that the behaviour problems in children and adolescents with DS appear to be largely unique to this group. In particular, behaviour problems were rarely reported in the TD group, and there was little similarity between the DS and control group with regard to how the independent variables related to behaviour problems. Unexpectedly, relatively few cognitive, neuropsychological and/or maturational variables were significantly related to behavioural problems in this study. Future research may incorporate additional variables that might relate to externalising and/or internalising problems. While the focus of this thesis was on the relationship between cognitive and neuropsychological variables and behavioural problems, other variables have been suggested elsewhere. In particular, Dykens (2007) proposed a range of biological and social variables that might influence the risk of psychopathology in individuals with DS. These variables included but are not limited to hypothyroidism, serotonin levels, health and pain, obesity, type of DS, abuse, stigma, personality, life stressors, and family genetics. It is clear that the variables in this study cannot completely account for the behaviour problems in people with DS. Accordingly, consideration of additional influences on maladaptive behaviours may be beneficial in attempting to understand how behaviour problems develop and/or are maintained in this group.

9.4 Concluding Comments

This thesis has added novel information to the pre-existing DS literature in several ways. It has synthesised previous research on the presentation of behavioural problems in this clinical group. Additionally, it has provided evidence that behaviour problems are not intrinsically linked with DS, but rather appear to be
associated with very specific cognitive and neuropsychological skill deficits that may be helpful in identifying if children that are at risk of developing behavioural problems. Furthermore, it is possible that with the right support strategies these cognitive difficulties may be minimised and thus hopefully have a positive impact on a child’s well-being by minimising the risk or impact of externalising and/or internalising behaviour problems during childhood and adolescence.
References


Appendix A. Plain Language Statement and Informed Consent Form for Participants with Down Syndrome

DEAKIN UNIVERSITY
PLAIN LANGUAGE STATEMENT AND CONSENT FORM

TO: Parents & Guardians

Plain Language Statement

Date:

Full Project Title: Understanding the behaviour of children and adolescents

Principal Researcher: Miss Laura Smith

Supervisor: Dr Jarrad Lum

This Plain Language Statement and Consent Form is four pages long. Please make sure you have all the pages.

Dear
My name is Miss Laura Smith and I am a doctoral student in health psychology from Deakin University. I am currently undertaking research examining behaviour in children and adolescents and I am writing to invite your child to participate. This research is being supervised by Dr Jarrad Lum who is a lecturer in psychology from Deakin University.

What I am researching in this study:
The purpose of this research project is to examine the development of behavioural patterns in children and adolescents. The reason that some children and adolescents develop behavioural difficulties is not completely understood. This project examines a range of processes that may be involved in influencing behaviour including intellectual, academic and neurological functioning, language, social skills and friendship.

Why have I received this letter?
In this study we are examining behaviour in children and adolescents with Down syndrome. Earlier this year I asked Down Syndrome Association of Victoria to pass this letter on to parents who might be interested in participating in this research.
What will my child be asked to do?
Children participating in the research will be presented with a number of different interesting task and activities presented. These can be presented at your child’s school or at Deakin University. These activities will be presented over three 30 minute sessions spaced over three weeks. The activities are summarized below:

- **Intellectual Functioning Tasks**: Children are presented with a range of tasks which look at their verbal and non-verbal reasoning skills.

- **Language Tasks**: Children will be presented with a range of tasks which look at their comprehension and production skills.

- **Neurological Tasks**: Children will be seated in front of a computer and watch a series of ‘boxes’ which open individually, and some which contain a pattern. Each pattern will then be displayed and children will then be asked to identify which box contained the pattern.

- **Academic Tasks**: Children are presented with tasks which examine spelling, reading and mathematic skills.

- **Friendship Questionnaire**: Children and are asked to complete a 40-item questionnaire about their friendship with their best friend. Items will be presented in both written and verbal form.

- **Behaviour Questionnaire**: Parents/guardians will be asked to complete a questionnaire about their child’s behaviour.

Does my child have to take part?
Your child does **not** have to take part in this study. Also if you decide to take part and then later change your mind, your child can withdraw from (leave) the study. This can be before your child starts the study, during it or afterwards.

Will my child’s data be confidential?
All the information collected will be confidential (not told to anyone else, including teachers at your child's school). I will **not** write your child’s name or school on your recording sheet or on the computer database. Your child will be given a number instead. **The data collected will be retained in a locked filing cabinet at Deakin University for a period of six years from date of publication.** Your child’s name will not appear in any publication of the results arising from this study.
Will I have access to the results of the study?
When the study has been completed we will produce a handout for teachers and parents outlining the findings of the study. These results can be given to you by mail, e-mail or through a letter that will be given to your child to take home. If you would like a copy of the main findings of the study please complete the form titled “Request for results from study”.

If you would like your child to participate:
If you would like your child to participate in this research please complete the attached consent. I will collect it from you when I visit your child.

Where can I obtain more information?
If you have any questions, comments or require further clarification about this research project please contact Dr Jarrad Lum who is a lecturer in psychology and is the supervisor for this project. His contact details are:

Mail: School of Psychology
Deakin University, Waterfront Campus.
Level 3, 27 Brougham St.
Geelong
3217
VIC

Phone: 03 5227 8474
Fax: 03 5227 8621
E-mail: jarrad.lum@deakin.edu.au

Yours Sincerely,

Miss Laura Smith
Doctoral Health Psychology) Student

IMPORTANT NOTE: If you have any complaints or concerns about any aspect of the project, the way it is being conducted or any questions about your rights as a research participant, then you may contact:

The Executive Officer, Human Research Ethics Committee, Deakin University, 221 Burwood Highway, Burwood Victoria 3125, Telephone: 9251 7123, Facsimile: 9244 6581; research-ethics@deakin.edu.au.
DEAKIN UNIVERSITY
PLAIN LANGUAGE STATEMENT AND CONSENT FORM

TO:

Consent Form

Date:

Full Project Title: Social Understanding in Children

I have read and I understand the attached Plain Language Statement.

I give my permission for ..........................................................(child’s name)

who was born on the ..............................................(day/month/year) to participate in this project

according to the conditions in the Plain Language Statement.

I have been given a copy of Plain Language Statement and Consent Form to keep.

The researcher has agreed not to reveal my identity and personal details, including where

information about this project is published, or presented in any public form.

Name (printed) ..........................................................

Name of Person giving Consent (printed) ..........................................................

Relationship to Participant: ..........................................................

Phone number: ..........................................................

Signature ................................................................. Date.................................

Contact Details for Dr Jarrad Lum

Mail: School of Psychology

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Geelong

3217

VIC

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Fax: 03 5227 8621

E-mail: jarrad.lum@deakin.edu.au
Appendix B. Letter of Invitation to School Principals and Organisational Consent Form

Faculty of Health, Medicine, Nursing and Behavioural Sciences
School of Psychology
Burwood campus
221 Burwood Highway, Burwood
Victoria 3125 Australia
Telephone: 0413 119 231
lsmi@deakin.edu.au

Friday, 25 October 2013

Dear PRINCIPAL,

My name is Miss Laura Smith and I am a Doctoral student studying towards a degree in health psychology. I am currently undertaking research looking at behaviour in children and adolescents with and without Down syndrome. I am writing to you to ask your permission to conduct part of the research at your school. This research project is being supervised by Dr Jarrad Lum who is lecturer in Psychology at Deakin University.

Written parental consent has already been provided for some children with Down syndrome. We are requesting permission to present these children with a series of tasks at your school. Children between 10 and 15 years old will be presented with a number of interesting tasks designed to assess intellectual and academic functioning, language, neurological functioning and friendship quality. Behaviour of children will be examined by a parent-report of activities, social and school behaviour. Tasks presented to children will be presented over three 45 minute sessions. The tasks would be presented to children on school grounds, in an open room which is clearly visible and accessible to staff (e.g., library, staffroom) and carried out at a time which is convenient for teachers and yourself. The tasks are presented in the context of a game and children are given constant positive feedback.

The types of tasks children are presented with include:

- **Intellectual Functioning Tasks**: Children are presented with a range of tasks which look at their verbal and non-verbal reasoning skills. These tasks will be used to estimate a child’s overall intellectual abilities.

- **Language Tasks**: Children will be presented with a range of tasks which look at their comprehension and production skills, and provide a measure of a child’s language skills.

- **Neurological Tasks**: Children will be seated in front of a computer and watch a series of ‘boxes’ which open individually, and some which contain a pattern. Each pattern will then be displayed and children will then be asked to identify which box contained the pattern. These tasks will be used to measure cognitive skills, such as memory, that are related to different areas of the brain.

- **Academic Tasks**: Children are presented with tasks which examine spelling, reading and mathematic skills. These tasks will be used to assess a child’s school-based skills.

- **Friendship Questionnaire**: Children and are asked to complete a 40-item questionnaire about their friendship with their best friend. Items will be presented in both written and verbal form and will be used to measure friendship quality.

- **Behaviour Questionnaire**: Parents/guardians will be asked to complete a questionnaire about their child’s behaviour. This will be used to assess a child’s behavioural strengths and weaknesses.

This study has received ethical approval from the Deakin University Human Ethics Committee, Department of Education and Early Childhood Development and Catholic Education Office.

I very much hope you would be interested in taking part and I would be delighted to discuss the project and tasks with you in more detail. I have enclosed in this correspondence copies of the information letters and consent forms that would be used in this study. I will contact you shortly to discuss the application with you further. Alternatively, feel free to contact me by phone or e-mail on the details listed in the letter head.

Yours Sincerely,

[Signature Redacted by Library]

Miss Laura Smith
Health Psychology Doctoral Student
PLAIN LANGUAGE STATEMENT AND CONSENT FORM

TO: School Principals.

Organisational Consent Form

Date:

Full Project Title: Understanding the behaviour of children and adolescents

I have read and I understand the attached Plain Language Statement.

I give my permission for students of Mornington Special Developmental School to participate in this project according to the conditions in the Plain Language Statement.

I have been given a copy of Plain Language Statement and Consent Form to keep.

The researcher has agreed not to reveal the participants’ identities, personal details or the school’s identity if information about this project is published or presented in any public form.

I agree that

1. The institution/organisation WILL NOT be named in research publications or other publicity without prior agreement.

3. I EXPECT to receive a copy of the research findings or publications.

Name of person giving consent (printed) ..........................................................

Signature ................................................................. Date .............................
Appendix C. Plain Language Statement and Informed Consent Form for Typically Developing Participants

Deakin University
Faculty of Health, Medicine, Nursing and Behavioural Sciences
School of Psychology
Burwood campus
221 Burwood Highway, Burwood
Victoria 3125 Australia
Telephone: 0408 534 866
lsmi@deakin.edu.au

An invitation for your child to participate in research

[DATE]

Dear

My name is Miss Laura Smith and I am a doctoral student in health psychology from Deakin University. I am currently undertaking research examining behaviour in children and adolescents and I am writing to invite your child to participate. This research is being supervised by Dr Jarrad Lum who is a lecturer in psychology from Deakin University.

What I am researching in this study:
The purpose of this research project is to examine the development of behavioural patterns in children and adolescents with and without Down syndrome. The reason that some children and adolescents develop behavioural difficulties is not completely understood. This project examines a range of processes that may be involved in influencing behaviour including intellectual and neurological functioning, language, social skills and friendship.

Why have we come to your school?
Your child’s school has been selected on the grounds that it is located in the Melbourne and surrounding areas. Prior to coming to your child’s school ethics permission was obtained from the Deakin University Ethics Committee and also from the Department of Education and Early Childhood Development to carry out the research. I also have met with and received permission from [school’s principal inserted here] to carry out this research. During this meeting the aims of the study and task were discussed and presented.

Why has my child received this letter?
When we visit a school we hope that a large number of children in each year level participate in the study. Because of this, we have asked teachers to distribute this letter out to all children in Grades 4 through to 6 at [insert child’s school here].
Behaviour Problems in Down Syndrome

What will my child be asked to do?
Children participating in the research will be presented with a number of different interesting task and activities presented at the school. These activities will be presented over three 45 minute sessions spaced over three weeks. The activities are summarized below:

- **Intellectual Functioning Tasks**: Children are presented with a range of tasks which look at their verbal and non-verbal reasoning skills. These tasks will be used to estimate a child’s overall intellectual abilities.

- **Language Tasks**: Children will be presented with a range of tasks which look at their comprehension and production skills, and provide a measure of a child’s language skills.

- **Neurological Tasks**: Children will be seated in front of a computer and be presented with a number of different activities. For example, in one task children will watch a series of ‘boxes’ which open individually, and some which contain a pattern. Each pattern will then be displayed and children will then be asked to identify which box contained the pattern. These tasks will be used to measure cognitive skills, such as memory, that are related to different areas of the brain.

- **Friendship Questionnaire**: Children and are asked to complete a 21-item questionnaire about their friendship with their best friend. Items will be presented in both written and verbal form and will be used to measure friendship quality.

What will I be asked to do?
One parent/guardian will be asked to complete a questionnaire about their child’s behaviour and social skills, based on the child’s activities, social relations and school performance during the past 6 months. This questionnaire will take approximately 40 minutes to complete.

Does my child have to take part?
Your child does not have to take part in this study. Also if you decide to take part and then later change your mind, your child can withdraw from (leave) the study. This can be before your child starts the study, during it or afterwards.

Will my child’s data be confidential?
All the information collected will be confidential (not told to anyone else, including teachers at your child’s school). I will not write your child’s name or school on your recording sheet or on the computer database. Your child will be given a number instead. Your child’s name will not appear in any publication of the results arising from this study.
Behaviour Problems in Down Syndrome

Will I have access to the results of the study?
When the study has been completed we will produce a handout for teachers and parents outlining the findings of the study. These results can be given to you by mail, e-mail or through a letter that will be given to your child to take home. If you would like a copy of the main findings of the study please complete the form titled “Request for Feedback”. Please note that your child does not have to participate in the study in order for you to receive this handout.

If you would like your child to participate:
If you would like your child to participate in this research please complete the attached consent form and return it by mail in the reply-paid envelope provided or alternatively, your child may return it to school.

Where can I obtain more information?
If you have any questions, comments or require further clarification about this research project please contact me at:

Mail: Miss Laura Smith  
School of Psychology  
Deakin University, Burwood Campus.  
221 Burwood Highway (Building W),  
Burwood  
3125  
VIC  

Phone: 0408 534 866  
E-mail: lsmi@deakin.edu.au

Yours Sincerely,

Miss Laura Smith  
Doctoral Health Psychology Student

IMPORTANT NOTE: If you have any complaints about any aspect of the project, the way it is being conducted or any questions about your rights as a research participant, then you may contact:

The Manager, Office of Research Integrity, Deakin University, 221 Burwood Highway, Burwood Victoria 3125, Telephone: 9251 7129, Facsimile: 9244 6581; research-ethics@deakin.edu.au.

Please quote project number 2009-187
Consent Form

Date: Monday, 11 May 2015

Full Project Title: Understanding the behaviour of children and adolescents

I have read and I understand the attached Plain Language Statement.

I give my permission for .................................................................(child’s name)

who was born on the .................................................(day/month/year) to participate in this project according to the conditions in the Plain Language Statement.

I have been given a copy of Plain Language Statement and Consent Form to keep.

The researcher has agreed not to reveal my identity and personal details, including where information about this project is published, or presented in any public form.

Name of Child (printed) .................................................................

Name of Person giving Consent (printed) .................................................................

Relationship to Participant: .................................................................

Phone number: .................................................................

Name of child’s school:

.................................................................

Signature ................................................................. Date .................................................................

Please send to Miss Laura Smith at:

Mail: School of Psychology
     Deakin University, Burwood Campus.
     221 Burwood Highway (Building W).
     Burwood
     3125
     VIC

Phone: 0408 534 866
E-mail: lsmi@deakin.edu.au
Appendix D. Deakin University Ethics Approval

DEAKIN UNIVERSITY
Human Ethics Research
Office of Research Integrity
Research Services Division
70 Elgar Road Burwood Victoria
Postal: 221 Burwood Highway
Burwood Victoria 3125 Australia
Telephone 03 9251 7123 Facsimile 03 9244 6581
research-ethics@deakin.edu.au

Memorandum

To: Dr Jarrad Lum
School of Psychology
F

cc: Miss Laura Smith

From: Deakin University Human Research Ethics Committee (DU-HREC)

Date: 17 February, 2010

Subject: 2009-187

Understanding the behaviour of children and adolescents

Please quote this project number in all future communications

The application for this project was considered at the DU-HREC meeting held on 07/12/2009.

Approval has been given for Miss Laura Smith, under the supervision of Dr. Jarrad Lum, School of Psychology, to undertake this project from 17/02/2010 to 17/02/2013.

The approval given by the Deakin University Human Research Ethics Committee is given only for the project and for the period as stated in the approval. It is your responsibility to contact the Human Research Ethics Unit immediately should any of the following occur:

- Serious or unexpected adverse effects on the participants
- Any proposed changes in the protocol, including extensions of time.
- Any events which might affect the continuing ethical acceptability of the project.
- The project is discontinued before the expected date of completion.
- Modifications are requested by other HREC's.

In addition you will be required to report on the progress of your project at least once every year and at the conclusion of the project. Failure to report as required will result in suspension of your approval to proceed with the project.

DU-HREC may need to audit this project as part of the requirements for monitoring set out in the National Statement on Ethical Conduct in Human Research (2007).

Human Research Ethics Unit
research-ethics@deakin.edu.au
Telephone: 03 9251 7123
Appendix E. Department of Education and Early Childhood Development
Ethics Approval

Department of Education and
Early Childhood Development

Office for Policy, Research and Innovation

2 Treasury Place
East Melbourne, Victoria 3002
Telephone: +61 3 9637 2000
DX 210083
GPO Box 4367
Melbourne, Victoria 3001

2010_000535

Miss Laura Smith
School of Psychology (Building W)
221 Burwood Highway
Deakin University
BURWOOD 3125

Dear Miss Smith

Thank you for your application of 15 March 2010 in which you request permission to conduct a research study in government schools titled: Understanding the behaviour of children and adolescents.

I am pleased to advise that on the basis of the information you have provided your research proposal is approved in principle subject to the conditions detailed below.

1. Should your institution’s ethics committee require changes or you decide to make changes, these changes must be submitted to the Department of Education and Early Childhood Development for its consideration before you proceed.

2. You obtain approval for the research to be conducted in each school directly from the principal. Details of your research, copies of this letter of approval and the letter of approval from the relevant ethics committee are to be provided to the principal. The final decision as to whether or not your research can proceed in a school rests with the principal.

3. No student is to participate in this research study unless they are willing to do so and parental permission is received. Sufficient information must be provided to enable parents to make an informed decision and their consent must be obtained in writing.

4. As a matter of courtesy, you should advise the relevant Regional Director of the schools you intend to approach. An outline of your research and a copy of this letter should be provided to the Regional Director.

5. Any extensions or variations to the research proposal, additional research involving use of the data collected, or publication of the data beyond that normally associated with academic studies will require a further research approval submission.

6. At the conclusion of your study, a copy or summary of the research findings should be forwarded to Education Policy and Research Division, Department of Education and Early Childhood Development, Level 3, 33 St Andrews Place, GPO Box 4367, Melbourne, 3001.

Every child, every opportunity

Victoria
The Place To Be

165
I wish you well with your research study. Should you have further enquiries on this matter, please contact Jonathan Howcroft, Policy and Research Officer, Education Policy and Research, by telephone on (03) 9947 1892 or by email at <howcroft-jonathan.j@edumail.vic.gov.au>.

Yours sincerely

Dr Elizabeth Hartnell-Young
Group Manager
Education Policy and Research

21/04/2010

enc
Appendix F. Catholic Education Office Ethics Approval

In reply please quote:

GE10/0009
1578

10 March 2010

Miss L. Smith
C/- Dr Jarrad Lum
School of Psychology (Building W)
Deakin University
BURWOOD VIC 3125

Dear Miss Smith

I am writing with regard to your research application received on 9 March 2010 concerning your forthcoming project titled Understanding the behaviour of children and adolescents. You have asked approval to approach Catholic schools in the Archdiocese of Melbourne, as you wish to involve Year 5–9 students and their parents.

I am pleased to advise that your research proposal is approved in principle subject to the nine standard conditions outlined below.

1. The decision as to whether or not research can proceed in a school rests with the school's principal, so you will need to obtain approval directly from the principal of each school that you wish to involve.
2. You should provide each principal with an outline of your research proposal and indicate what will be asked of the school. A copy of this letter of approval, and a copy of notification of approval from the university's Ethics Committee, should also be provided.
3. A Working with Children (WWC) check – or registration with the Victorian Institute of Teaching (VIT) – is necessary for all researchers visiting schools. Appropriate documentation must be shown to the principal before starting the research in each school.
4. No student is to participate in the research study unless s/he is willing to do so and informed consent is given in writing by a parent/guardian.
5. You should provide the names of schools which agree to participate in the research project to the Knowledge Management Unit of this Office.
6. Any substantial modifications to the research proposal, or additional research involving use of the data collected, will require a further research approval submission to this Office.

1 of 2
7. Data relating to individuals or schools are to remain confidential.

8. Since participating schools have an interest in research findings, you should consider ways in which the results of the study could be made available for the benefit of the school communities.

9. At the conclusion of the study, a copy or summary of the research findings should be forwarded to this Office. It would be appreciated if you could submit your report in an electronic format using the email address provided below.

I wish you well with your research study. If you have any queries concerning this matter, please contact Mr Mark McCarthy of this Office. The email address is <km@ceomelb.catholic.edu.au>.

Yours sincerely

[Signature Redacted by Library]

Nancy Bicchieri
DEPUTY DIRECTOR