THE CLINICAL PRESENTATION OF CHILDHOOD-ONSET SCHIZOPHRENIA: A LITERATURE REVIEW

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STATEMENT

I, the undersigned, hereby declare that the work contained in this assignment is my own original work, and that I have not previously in its entirety or in part submitted it at any university for a degree.
Abstract

This literature review explores the research on the clinical presentation of childhood onset schizophrenia (COS) that has been conducted over the past ten years. A literature search was done using internet search engines and psychological databases to collect English language journals from 1994 onwards. Research indicates that COS is a stable diagnosis. Generally, there is a clear history of premorbid abnormalities, an insidious onset and a deteriorating course. For the majority of cases there seems to be a poor outcome. In conclusion, despite the limitations in the research conducted thus far, findings provide important insights regarding COS and several possibilities for future research.
Opsomming

Hierdie literere oorsig fokus op navorsing wat die afgelope tien jaar gedoen is oor die kliniese aanbieding van skisofrenie wat in die kinderjare begin (COS). Daar is gebruik gemaak van Internet “soek enjins” en sielkundige databasisse ten einde Engelstalige joernale op te spoor wat vanaf 1994 tot nou oor dié onderwerp verskyn het. Navorsing dui daarop dat COS ‘n stabiele diagnose is. Oor die algemeen toon dit ‘n duidelijke geskiedenis van premorbiede abnormaliteite, ‘n ongemerkte aanvang en verloop en agteruitgang oor tyd. In die meeste gevalle blyk daar ‘n swak uitkoms te wees.

Laastens bied die bevindinge belangrike insigte ten opsigte van COS en heelwat moontlikhede vir toekomstige navorsing, ten spyte van die beperkinge in die navorsing wat tot dusver gedoen is.
Introduction

My interest in childhood-onset schizophrenia (COS) was sparked during a year I spent as a volunteer at a child psychiatric inpatient ward. It seemed that the diagnosis and treatment of this mental illness were fraught with pitfalls and inexplicable features. This led me to explore the literature on this area. The literature seemed sparse, leaving me with more questions than answers. My research, which began as an attempt to come to grips with this form of childhood psychopathology, thus, proceeded to become a literature review which aims to understand what the literature has found about the clinical presentation of childhood-onset schizophrenia.

A literature search was done using internet search engines (google.com and www.sciedirect.com) and psychological databases (PsycINFO, Pubmed and Sabinet) to collect English language journal articles from 1994 onwards which in some way were attempting to explore this issue (in other words, over the past ten years). A few articles prior to 1994 were included in areas where information was particularly scant, and where there was a need for a greater perspective on developments in the given area of research. As my chosen area of focus was the phenomenology of childhood onset schizophrenia several areas of research where not included, namely, neuroimaging and magnetic resonance studies (Ballmaier et al., 2004; Eliez & Reiss, 2000; Hendren, Backer & Pandina, 2000), smooth pursuit eye movements and blink rate studies (Jacobsen et al., 1996; Karatekin & Asarnow, 1999; Ross, 2003), neurobiological and brain development studies (Eggers, 1999; Giedd et al., 1999; Jacobsen & Rapoport, 1998; Keller et al, 2003; McKenna, Gordon & Rapoort, 1994), obstetrical complications and developmental abnormality studies (Matsumoto, Takei, Saito, Kachi & Mori, 1999; Nicolson & Rapoport, 1999), and genetic and hereditary studies (Asarnow et al., 2001; Jacobsen & Rapoport, 1998; Kumra et al., 2001; Nicolson & Rapoport, 1999; Nicolson et al., 2000). The areas of prevention (Faraone, Brown, Glatt & Tsuang, 2002; Tsuang, Stone & Faraone, 2000; Tsuang, Stone & Faraone, 2002) and treatment (American Academy of Child and Adolescent Psychiatry, 1997; American Academy of Child and Adolescent Psychiatry, 2000; Lauriello, Bustillo & Keith, 1999; Malla & Norman, 2001; Volkmar, 1996) were not included either.
The aim of this review is to determine what, according to the literature, the clinical presentation or phenomenology of childhood onset schizophrenia (COS) is. The focus will be specifically on childhood-onset schizophrenia defined loosely as onset of psychosis before 12 years of age. This is to be distinguished from early-onset schizophrenia (EOS), where onset is after 12 years of age and adult-onset schizophrenia (AOS) where age of onset is after 18 years of age (Kumra et al., 2001; Nicolson & Rapoport, 1999; Nicolson et al., 2000).
Clinical Features

Historically, the diagnosis of childhood schizophrenia was given to a broad spectrum of children showing signs and symptoms of psychopathology (Asarnow & Asarnow, 1994). During and after the 1970’s a process of differentiation took place, whereby autism and other childhood mental illnesses were separated from childhood schizophrenia and put into other categories of childhood psychopathology (for further information regarding the historical development of COS as a diagnosis refer to Asarnow, 1994; Asarnow & Asarnow, 1994; Jacobser & Rapoport, 1998; Remschimdt, Schulz, Martin, Warnke & Trott, 1994).

Yet, no separate diagnostic category for COS exists. Rather, it is the case that the adult diagnostic criteria (see Box 1 and Box 2 on pp. 5-6) are used to diagnose children, adolescents and adults in both the DSM system and in the ICD system (American Psychiatric Association, 1994; World Health Organisation, 1992). There are only a few studies which have explored specifically the clinical features of COS. These were four American studies (Alaghband-Rad, Hamburger, Giedd, Frazier & Rapoport, 1997; Green, Padron-Gayol, Hardesty & Bassiri, 1992; Russell, Bott & Sammons, 1989; Spencer & Campbell, 1994).

Auditory Hallucinations

Three studies found that auditory hallucinations predominated in the presentation of COS. In the Green et al. (1992) study, 32 of the 38 children suffering from COS had auditory hallucinations. Russell et al. (1989) found that 80% of the 35 children in their sample had auditory hallucinations, making this the most common positive symptom in their study. All of the children in the Spencer and Campbell study (1994), suffered from auditory hallucinations (sample size was 16).

Delusions

Delusions, though less dominant than auditory hallucinations, were nonetheless a common feature in the clinical presentation of COS. Half the children in Green et al. (1992), 63% of the Russell et al. (1989) study and all of the children of the Spencer and Campbell (1994) study had delusions.
Factors relating to hallucinations and delusions

The Spencer and Campbell (1994) study found that the delusions were related to the hallucinatory experiences of the children in some way. The complexity and elaboration of hallucinations and delusions seemed to be related to the age of the child (Russell et al., 1989; Spencer & Campbell, 1994). Particularly, the greater the age of the child the more complex and elaborate hallucinations and delusions tended to be.

Ego-syntonicity

Russell et al. (1989) make the observation that in their study the children’s psychotic symptoms seemed to be ‘ego-syntonic’ rather than ‘ego-dystonic’ (as found in studies of adults suffering from schizophrenia). These children had difficulty distinguishing their psychotic symptoms from normal experience because they do not have a comparative framework of what normal experience is. Thus, symptoms are not experienced as frightening or disorganising to the extent that this is reported in adults. This may also explain why these children do not report their symptoms to their parents (Russell et al., 1989).

General Comments

These four studies were limited in terms of sample size and thus in generalisability to other populations. Researchers used the DSM-IV or DSM-III-R criteria for schizophrenia to select the participants for these studies. Thus, they were bound to find the symptoms for COS as this had been defined through the selection process, in other words, when judging whether or not to include participants in the study. This limits the ability of these studies to explore variants in clinical presentation of COS and to evaluate the suitableness of the adult criteria for a childhood mental disorder. None of these studies compared their samples to a normal control and only three of the studies (Green et al., 1992; Russell et al., 1989; Spencer & Campbell, 1994) were focusing specifically on the clinical presentation of COS. Thus, any statements made or themes across the studies are tentative and need further corroboration.

Thought disorder is dealt with next (on pp.7) as a sub-section of Clinical Features, the reason being that specific research exists on this area, apart from other studies of the clinical presentation of COS.
Box 1

DSM-IV Diagnostic Criteria for Schizophrenia

A. Characteristic symptoms: Two (or more) of the following, each present for a significant portion of time during a 1-month period (or less if successfully treated):
   (1) delusions
   (2) hallucinations
   (3) disorganized speech (e.g., frequent derailment or incoherence)
   (4) grossly disorganized or catatonic behavior
   (5) negative symptoms, i.e., affective flattening, alogia, or avolition

Note: Only one criterion A symptom is required if delusions are bizarre or hallucinations consist of a voice keeping up a running commentary on the person’s behavior or thoughts, or two or more voices conversing with each other.

B. Social/occupational dysfunction: For a significant portion of the time since the onset of the disturbance, one or more major areas of functioning, such as work, interpersonal relations, or self-care, are markedly below the level achieved prior to the onset (or when the onset is in childhood or adolescence, failure to achieve expected level of interpersonal, academic or occupational achievement).

C. Duration: Continuous signs of the disturbance persist of at least 6 months. This 6-month period must include at least 1 month of symptoms (or less if successfully treated) that meet criterion A (i.e., active-phase symptoms) and may include periods of prodromal or residual symptoms. During these prodromal or residual periods, the signs of the disturbance may be manifested by only negative symptoms or two or more symptoms listed in the criterion A present in an attenuated form (e.g., odd beliefs, unusual perceptual experiences).

D. Schizoaffective and mood disorder exclusion: Schizoaffective disorder and mood disorder with psychotic features have been ruled out because either: (1) no major depressive, manic, or mixed episodes have occurred concurrently with the active-phase symptoms; or (2) if mood episodes have occurred during active phase symptoms, their total duration has been brief relative to the duration of the active and residual periods.

E. Substance/general medical condition exclusion: The disturbance is not due to the direct psychological effects of a substance (e.g., a drug of abuse, a medication) or a general medical condition.

F. Relationship to a pervasive developmental disorder: If there is a history of autistic disorder or another pervasive developmental disorder, the additional diagnosis of schizophrenia is made only if prominent delusions or hallucinations are also present for at least a month (or less if successfully treated).
ICD-10 Diagnostic Criteria for Schizophrenia

This overall category includes the common varieties of schizophrenia together with some less common varieties and closely related disorders.

General criteria for paranoid, hebephrenic, catatonic, and undifferentiated schizophrenia

G1. Either at least one of the syndromes, symptoms, and signs listed under (1) below, or at least two of the symptoms and signs listed under (2) should be present for most of the time during an episode of psychotic illness lasting for at least 1 month (or at some time during most of the days).

(1) At least one of the following must be present:
   (a) Thought echo, thought insertion or withdrawal, or thought broadcasting;
   (b) Delusions of control, influence, or passivity, clearly referred to body or limb movements or specific thoughts, actions, or sensations: delusional perception;
   (c) Hallucinatory voices giving a running commentary on the patient’s behavior, or discussing the patient among themselves, or other types of hallucinatory voices coming from some part of the body;
   (d) Persistent delusions of other kinds that are culturally inappropriate and completely impossible (e.g. being able to control the weather, or being in communication with aliens from another world).

(2) Or at least two of the following:
   (a) persistent hallucinations in any modality, when occurring every day for at least 1 month, when accompanied by delusions (which may be fleeting or half-formed) without clear affective content, or when accompanied by persistent overvalued ideas;
   (b) neologisms, breaks, or interpolations in the train of thought, resulting in incoherence or irrelevant speech;
   (c) catatonic behaviour, such as excitement, posturing or waxy flexibility, negativism, mutism, and stupor;
   (d) “negative” symptoms, such as marked apathy, paucity of speech, and blunting or incongruity of emotional responses (it must be clear that these are not due to depression or neuroleptic medication).

G2. Most commonly used exclusion clauses

(1) If the patient also meets criteria for manic episode or depressive episode, the criteria listed under G1(1) and G1(2) above must have been met before the disturbance of mood developed.

(2) The disorder is not attributable to organic brain disease or to alcohol- or drug-related intoxication, dependence, or withdrawal.
Thought Disorder

Thought disorder and communication deficits have been studied as specific features of the clinical presentation of COS. Some studies that explored the phenomenology of COS have included findings related to thought disorder. Loose associations, illogical thinking, poverty of speech and incoherence were found in all the children of the Green et al. (1992) study, while 60% of the Russell et al. (1990) sample were labelled as showing likely or definite thought disorder. Thirteen of the sixteen COS participants in the Spencer and Campbell (1994) study were diagnosed with thought disorder. However, the three studies defined thought disorder differently, using the DSM-IV criteria as a basis but differing in the stringency with which the criteria were met.

Thus, to gain a clearer picture of thought disorder as it occurs in COS, studies that focused specifically on thought disorder were reviewed. Six research studies, all from the University of California (UCLA) studied the presentation of thought disorder and communication deficits in COS children (Abu-Akel, Caplan, Guthrie & Komo, 2000; Caplan, 1994a; Caplan, 1994b; Caplan, Guthrie & Foy, 1992; Caplan, Guthrie & Komo, 1996; Caplan, Guthrie, Tang, Komo & Asarnow, 2000). Caplan et al. (1992) found that children with COS differed from adult schizophrenia in three areas. They used fewer conjunctions, used less repetition of words and word roots and were more likely to omit a part of a clause, making the assumption that the listener retained enough information from the clause. The COS participants were similar to adults with schizophrenia in that they spoke less than normal controls and they did not provide links to previous utterances. They often broke the flow of speech to make
reference to something or someone in their immediate environment. Reference to people was often unclear and ambiguous (Caplan et al., 1992).

Caplan (1994a; 1994b) suggests that there are three components to thought disorder:

1. discourse measures - these are related to conversation competence
2. distractibility - a verbal factor and loose associations, which Caplan (1994a) argues may be positive signs of COS
3. illogical thinking and exophora - these may be related to the negative signs of COS

A principal component analysis of a sample of children with COS provided evidence for these three factors (Caplan, 1994a). Based on this it is argued that COS children may have specific communication deficits rather than a generalised impairment of communication ability.

Studies comparing COS to normal children found that COS participants used significantly less referential revision, false starts and fillers (Caplan et al., 2000) and that their use of speech functions was significantly less than that of normal children (Abu-Akel et al., 2000). Specifically, COS children showed an impaired ability to organise their thoughts, did not prepare the listener for a change in topic and showed a lack of adequate reasoning (Caplan et al., 1992; Caplan et al., 1996; Caplan et al., 2000). Interestingly, it was found that those children suffering from COS who were medicated performed more poorly than those who were not medicated, whereas in adults the reverse was true (Caplan et al., 1992; Caplan et al., 1996). This indicates that medication may have been influencing the ability of COS participants to perform on the measures of thought disorder and communication deficits.
In summary, there is evidence for the suggestion that children suffering from COS present with specific communication deficits rather than a generalised impairment in communication (Abu-Akel et al., 2000; Caplan, 1994a, Caplan, 1994b, Caplan et al., 1992; Caplan et al., 1996; Caplan et al., 2000). However, the influence of medication needs to be explored further. In addition, only a few studies have explored thought disorder and communication deficits. Sample sizes were small. There seems to be a group of researchers exploring this area and though this means that the same measures are used, it does limit the generalisability of the findings.
Neuropsychological studies of COS

There is a paucity of research studies that explore the neuropsychological abilities of children suffering from COS. Only two studies (Bedwell et al., 1999; Hendren et al., 1995) and one review (Asarnow et al., 1994) were found.

The Hendren et al. (1995) study compared the performance of children with symptoms of COS and schizotypal personality disorder to a group of normal children. They found that the schizo-spectrum disorder group performed significantly worse in all the areas assessed. This group had the most deficits in frontal lobe ability, verbal memory and story memory. It seems that the schizo-spectrum disorder group suffered from a broad range of neuropsychological deficits. The Bedwell et al. (1999) study compared the pre- and postpsychotic intelligence of a sample of children diagnosed with COS. They found a significant decline in the postpsychotic full scale intelligence (IQ) of their COS participants. Three areas - picture arrangement, information and block design - were particularly impaired.

The lack of research in this area is surprising as one could assume that neuropsychological testing may be useful as part of the process of diagnosis. Both studies found that COS participants had a markedly poor performance on neuropsychological assessments. However, given the small sample size of the studies and the lack of research in this area, these findings are explorative at best. In addition, both samples had severely ill, medicated participants and this must have impacted on the ability to perform. Extraneous variables such as concentration,

1 The DSM requires that a person must be over the age of 18 for a clinician to make a diagnosis of a personality disorder (American Psychiatric Association, 1994). However, the researchers made this diagnosis in children.
attention and anxiety, which, may also have affected the ability to perform were not explored by these studies (Bedwell et al., 1999; Hendren et al., 1995).

All seven studies that explored the potential causes of CDS showed evidence of a history of behaviour or temper problems. These are similar to what Bedwell et al. (1995) reported for a sample of children from a clinic for emotionally disturbed children. However, the studies did not report any significant differences in the incidence of CDS between children who were referred due to behaviour problems and those who were referred due to other reasons. This may indicate that children who are referred for emotional problems are more likely to suffer from anxiety and depression, which may affect their ability to perform at school. These studies also highlighted the importance of assessing the child's personal and family history in order to identify any potential risk factors.
Personal and Family History

Premorbid and Personal History

A careful examination of a child's premorbid history is an important step towards making a decision about diagnosis. This seems to be particularly true for COS as the literature suggests that there is a marked deterioration in functioning before the onset of psychotic symptoms, more severe than that of EOS and AOS (Alaghband-Rad et al. 1995; Russell, 1994).

All seven articles that explored the premorbid course of COS showed evidence of a history of behavioural and psychiatric disturbances (Alaghband-Rad et al., 1995; Eggers, Bunk & Krause, 2000; Eggers, Bunk, Volberg & Röpcke, 1999; Mezaide, Gringras, Rodrique et al., 1996; Russell, 1994; Russell, 1995; Schaeffer & Ross, 2002). One male subject in the study by Eggers et al. (2000), however, was an exception. Three studies found developmental delays, particularly in language (Alaghband-Rad et al., 1995; Mezaide, Gringras, Rodrique et al., 1996; Schaeffer & Ross, 2002). In contrast, the study by Eggers et al. (2000) found no signs of developmental delay in their sample. This could be due to the fact that their sample size was significantly smaller than the other studies (only 11 participants), but also raises the question of generalisability, as theirs was the only study outside of the United States of America.

Those studies that did consider premorbid intelligence, found that generally these children were of low intellectual functioning (Alaghband-Rad, et al., 1995; Mezaide, Gringras, Rodrique et al., 1996). Four of the studies indicated a lack of social
development, using words such as withdrawn, avoidant and reserved to describe the premorbid social functioning of their participants (Alaghband-Rad et al., 1995; Eggers et al., 2000; Mezaide, Gringras, Rodrique et al., 1996; Schaeffer & Ross, 2002). In two of the studies a small percentage of the sample had shown some pervasive developmental disorder symptoms, such as odd behaviour; signs of psychotic symptoms such as delusions and auditory hallucinations and/or obsessive-compulsive traits (Alaghband-Rad et al., 1995; Schaeffer & Ross, 2002). Behaviour problems and disruption at school were characteristic of premorbid history in several of the studies. Specifically, 30% of the children in the Alaghband-Rad et al. (1995) study were diagnosed with Attention Deficit Hyperactivity Disorder (ADHD) and 9% of the sample fulfilled the criteria for Conduct disorder. Thirty-two percent of the Mezaide, Gringras, Rodrique et al. (1996) study showed externalised symptoms (defined as partially meeting the diagnostic criteria for ADHD, Conduct Disorder or Oppositional Defiant Disorder). Five of the eleven participants in the Eggers et al. (2000) study showed bizarre or aggressive behaviour and twenty-nine percent of the Schaeffer & Ross (2002) study showed ADHD type symptoms.

Alaghband-Rad et al. (1995) argue that the premorbid history of COS has significantly more abnormalities than that of adults. The Eggers et al. (1999) study comparing premorbid abnormalities of COS with EOS (in other words onset before twelve years of age and after twelve years of age, respectively) found that those children with COS had significantly more premorbid abnormalities than the EOS group did. This provides evidence for a distinction between COS and EOS. There was evidence that the tendency for withdrawal and disturbances in adaptive social
behaviour could be a sign of social impairment in the later course of COS as well as a probable risk factor COS (Eggers et al., 1999; Eggers et al., 2000).

Russell (1994) provides a concise description of the premorbid history and prodromal course of COS. He reports that COS begins with a broad spectrum of behavioural disturbances at a young age. There is a gradual emergence of psychotic symptoms. With the combined effect of the behavioural disturbance and psychotic symptoms there is a marked deterioration in functioning, leading to hospitalisation and finally to a full diagnostic assessment. However, others suggest that this course is not as clear-cut, since often these children receive numerous differential diagnoses before a final diagnosis is decided upon (Schaeffer & Ross, 2002).

**Gender, family and social factors**

Two studies found that there were significantly more males than females presenting with COS (Green et al., 1992; Spencer & Campbell, 1994). The Alaghband-Rad et al. (1997) study found that males were more likely to have an insidious onset of COS than females. A family history of psychiatric illness or hospitalisation was found in the Green et al. (1992) and the Spencer and Campbell (1994) studies. Socio-economic status did not seem to have an influence on the presentation of COS, according to the Green et al. (1992) study.

In summary, findings suggest that there is a marked deterioration in several areas of functioning in the premorbid stages of COS. This includes developmental delays, poor social functioning, pervasive developmental disorder symptoms and behavioural symptoms (Alaghband-Rad et al, 1995; Mezaide, Gringras, Rodrique et al, 1996;
Russell, 1994; Schaeffer & Ross, 2002). It has also been found that COS has significantly more premorbid abnormalities than EOS and AOS (Eggers et al, 1999; Eggers et al, 2000). Further research is required, specifically to deal with the contradictions between the American (Alaghband-Rad et al, 1995; Mezaide, Gringras, Rodrique et al, 1996; Russell, 1994; Russell, 1995; Schaeffer & Ross, 2002) and the German (Eggers et al, 1999; Eggers et al, 2000) studies of the premorbid history of COS.
Onset

Age and type of onset differentiates COS from EOS and AOS. Thus, onset is an important consideration for diagnosing COS. There were eight articles that explored the onset of COS (Alaghband-Rad, et al., 1995, Bunk, Eggers & Klapal, 1999; Eggers & Bunk, 1997; Eggers et al., 1999; Eggers et al., 2000; Frazier et al., 1997; Russell, 1995; Schaeffer & Ross, 2002). Four were from the Essen study in Germany, (Bunk, Eggers & Klapal, 1999; Eggers & Bunk, 1997; Eggers et al., 1999; Eggers et al., 2000), and of these four, three included early-onset schizophrenia in their sample group. Thus, conclusions made about the onset of COS are tentative at best, as the actual sample of COS subjects is small. In addition, any statistical analysis (including means and ranges) included the EOS group which interferes with findings regarding only COS (see Table 1 below).

Table 1: Summary of mean age, range of onset and type of onset of COS

<table>
<thead>
<tr>
<th>Study</th>
<th>N</th>
<th>Mean age of onset</th>
<th>Range of onset</th>
<th>Type of onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Russell (1994)</td>
<td>35</td>
<td>6.9 years</td>
<td>3 to 11 years</td>
<td>Predominantly insidious</td>
</tr>
<tr>
<td>Alaghband-Rad et al (1995)</td>
<td>23</td>
<td>**</td>
<td>**</td>
<td>15 participants = insidious</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>8 participants = subacute</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 participant = episodic</td>
</tr>
<tr>
<td>Eggers &amp; Bunk (1997)</td>
<td>71</td>
<td>13 years</td>
<td>6 to 14 years</td>
<td>11 participants = insidious</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>33 participants = acute</td>
</tr>
<tr>
<td>Frazier et al (1997)</td>
<td>28</td>
<td>10 years</td>
<td>**</td>
<td>**</td>
</tr>
<tr>
<td>Bunk et al (1999)</td>
<td>44</td>
<td>**</td>
<td>**</td>
<td>**</td>
</tr>
<tr>
<td>Eggers et al (1999)</td>
<td>44</td>
<td>**</td>
<td>7 to 14 years</td>
<td>11 participants = insidious</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>33 participants = acute</td>
</tr>
<tr>
<td>Eggers et al (2000)</td>
<td>11</td>
<td>11 years</td>
<td>7 to 16 years</td>
<td>7 participants = insidious</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>4 participants = acute</td>
</tr>
<tr>
<td>Schaeffer &amp; Ross (2002)</td>
<td>17</td>
<td>8.6 years (s = 2.9 years)</td>
<td>**</td>
<td>**</td>
</tr>
</tbody>
</table>

*included EOS participants in their study
**not stated
The average age of onset ranged from 6.9 years (Russell, 1994) to 13 years (EOS subjects were included in the Eggers et al., 1999 study). The study by Schaeffer and Ross (2002) reported one child who had onset of psychosis at two years of age. There seems to be no general trend, except that researchers for the last ten years have a tacit agreement that onset before 12 years should be considered to be COS, whereas onset after 12 years and before approximately 18 years is considered to be EOS (Eggers et al., 1999; Rosenbaum Asarnow, Tompson and Goldstein, 1994). Though this distinction between EOS and COS is not applied by all who do research in this area, (for example, Eggers and Bunk (1997) classified COS as being onset before 14 years of age), findings, particularly in regards to type of onset, have provided evidence that this distinction may be justified. Research that included both EOS and COS samples reveal a possible difference in the type of onset. Insidious onset was more frequent for children with onset before 12 years of age, whereas acute onset was more often found in children with onset after 12 years of age (Eggers et al., 1997; Eggers et al. 1999). Those studies who only included a sample which had been classified as having onset before 12 years found that insidious onset predominated (Alaghband-Rad, McKenna, Gordon et al., 1995; Russell, 1994).

In looking for an explanation for this difference in type of onset, a study by Frazier, Alaghband-Rad, Jacobsen et al. (1997) explored the influence of pubertal development on onset of psychosis. No clear relationship could be found, though it was stated tentatively that onset for females may in some way be related to pubertal development. Bunk et al. (1999) found that at onset of psychosis there was no clear division of positive and negative symptoms into different dimensions. Based on their
findings, Alaghband-Rad, McKenna, Gordon et al. (1995) suggest that girls may have a less insidious onset than boys.

Inclusion of EOS in several studies that explored onset makes estimating the average age of onset of psychotic symptoms problematic. However, findings in regards to type of onset have quite possibly provided some evidence for the distinction between EOS and COS, in that it has been found that type of onset is usually insidious before 12 years of age, whereas type of onset is acute after 12 years of age (Bunk, Eggers & Klapal, 1999; Eggers & Bunk, 1997; Eggers et al., 1999; Eggers et al., 2000). However, it should be noted that sample sizes were small and COS as being onset before 12 years of age was not applied systematically in the research. Thus, conclusions based on these findings are made with caution.
Course

Another important aspect of the clinical presentation of COS is the course of this disorder. Six studies monitored the course of COS by two or more follow-up procedures (Bunk et al., 1999; Eggers et al., 2000; Mezaide, Bouchard, Gringras et al., 1996; Mezaide, Gringras, Rodrique et al., 1996; Asarnow, Tompson & Goldstein, 1994; Asarnow & Thompson, 1999). For all of the studies the majority diagnoses remained stable. This suggests that there is a continuity of COS overtime. In addition, it was found that for the most part there was a deteriorating course (Mezaide, Gringras, Rodrique et al., 1996; Asarnow & Tompson, 1999), though the Eggers et al. (2000) study reports that courses differed with regards to number of episodes, expression of subtypes and intervals between episodes.

Three of the studies looked specifically at the course of the positive and negative symptoms of COS (Bunk et al., 1999; Eggers et al., 2000; Mezaide, Bouchard, Gringras et al., 1996). The Mezaide, Bouchard, Gringras et al. (1996) study found poor continuity for both positive and negative symptoms. However, their study included EOS and thus may not be entirely applicable to COS. The study by Bunk et al. (1999) saw a significant decrease in the average frequency of positive and global symptoms while the average frequency of negative symptoms remained stable. They argue that this does not indicate an independent course for positive symptoms as opposed to negative symptoms because for both the positive and negative symptoms their disabling impact increased through the course of COS. The Eggers et al. (2000) study found that the course of COS did not set in with negative symptoms. Rather, it was the positive symptoms that occurred very early on in the course.
Little research has been done on the course of COS. The research that has been conducted has found evidence for a deteriorating course and a stable diagnosis (Bunk et al., 1999; Eggers et al., 2000; Mezaide, Bouchard, Gringras et al., 1996; Mezaide, Gringras, Rodrigue et al., 1996; Asarnow, Tompson & Goldstein, 1994; Asarnow & Thompson, 1999). However, small sample sizes and the inclusion of EOS participants in two studies (Mezaide, Bouchard, Gringras et al., 1996; Mezaide, Gringras, Rodrigue et al., 1996) make any conclusions tentative.
Outcome

There were two measures of outcome for COS. The DAS-M (Disability Assessment Schedule-Mannheim) was used by the German studies (Eggers & Bunk, 1997; Eggers et al., 1999; Eggers et al., 2000). The American studies (Mezaide, Bouchard, Gringras et al., 1996; Mezaide, Gringras, Rodrigue et al., 1996; Asarnow et al., 1994; Asarnow & Tompson, 1999) used the C-GAS (Children’s Global Assessment Scale).

Some percentage of all the studies showed complete remission as classified by these two measures. Twenty-eight percent of 21 participants in Asarnow et al. (1994) showed good outcome and four of the five participants with good outcome showed remission of their schizophrenic symptoms. Twenty-five percent of 44 participants in the study by Eggers and Bunk (1997) had full remission from symptoms. In the Mezaide, Gringras, Rodrigue et al. (1996) study, twenty-six percent of the 41 participants in the study had good to moderately good long-term outcome. Two of these (in other words, five percent of the sample) had a total recovery. Eleven of the 44 participants had a very good remission in the study by Eggers et al. (1999).

Twenty-eight percent of 18 participants in the Asarnow and Tompson (1999) study had a good outcome. Lastly, two of the eleven participants in the study by Eggers, Bunk and Krause (2000) were classified as having a good outcome. One of the participants was living with her family, without psychiatric treatment and the other was being treated in an outpatient clinic. These findings counterbalance the overriding sense that COS is a chronic and more severe form of schizophrenia, with a poor prognosis (Asarnow et al., 1994; Asarnow & Tompson, 1999).
However, the majority of participants in studies had a poor outcome. Forty-four percent of the participants in the study by Asarnow et al. (1994) had severe impairment and a further seventeen percent of the sample had persistent impairment with a deteriorating course. Seventy-four percent of the Mezaide, Gringras, Rodrigue et al. (1996) study were found to have poor or very poor outcomes. Eleven of the forty-one participants in the sample were institutionalised in a psychiatric hospital. Fifty percent of the Eggers et al. (1997) sample showed poor outcome and severe residual symptoms. Eggers et al. (1999) saw that fifty percent of their sample showed no episodes of remission and that no subject with insidious onset completely remitted from social disability. Eight of the eleven participants in the Eggers et al. (2000) study showed poor to very poor social adjustment.

Contradictory findings included the study by Asarnow and Tompson (1999) which found that only seventeen percent of the 18 participants in their sample had a deteriorating course. They argue that there was no significant picture of poor outcome in their study. Eggers et al. (2000) could not find a systematic relationship between diagnosis of COS and outcome, though those participants with predominantly catatonic symptoms were the ones to show a poor outcome. And though Mezaide, Gringras, Rodrigue et al. (1996) concluded that COS has a very poor outcome, they could not find a relationship between age of onset and outcome.

Tentatively, the findings with regards to outcome show a generally negative picture, however there is evidence that some COS patients will recover and function well (Eggers et al., 2000; Asarnow et al. 1994.; Asarnow and Tompson, 1999). However, small sample sizes and the use of different measures of outcome by the German
studies as opposed to the American studies limit the extent that conclusions can be made based on these findings.

The DSM-IV criteria for the diagnosis of a pervasive developmental disorder do not specify the duration of symptomatology. There is insufficient evidence for the clinical validity of a pervasive developmental disorder on the condition that the child's and later in childhood are present for at least a month. American studies have shown that this is not true.

Several studies have analyzed the validity of diagnosis. Shellenberg et al. (1994) suggested that the criteria were better defined and more clearly related to childhood autism in the United States. They also suggested that the criteria were more clearly defined and more clearly related to childhood autism in the United States. Only a few studies have been conducted to determine if childhood autism is a definite clinical entity. However, there are some indications that this may be the case. The DSM-IV criteria for a pervasive developmental disorder do not specify the duration of symptomatology. There is insufficient evidence for the clinical validity of a pervasive developmental disorder on the condition that the child's and later in childhood are present for at least a month.
Differential Diagnoses

The DSM-IV states that the differential diagnoses, schizoaffective disorder and mood disorder, need to be excluded to make a diagnosis of schizophrenia. There is provision made for a co-morbid diagnosis of a pervasive developmental disorder on the condition that delusions and hallucinations are present for at least a month (American Psychiatric Association, 1994).

Several studies have explored differential diagnosis. McKenna et al. (1994) screened seventy-one of the two-hundred and sixty patients who had been referred to the National Institute for Mental Health between 1990 and 1993 with onset of psychosis before age 12. They found that the COS diagnosis was used overinclusively for children who were severely disturbed and had some psychotic symptoms. Only nineteen of the seventy-one participants in their study were diagnosed as definitely suffering from COS. Fourteen received a diagnosis of some kind of affective disorder, four were diagnosed with Asperger’s disorder and two with Pervasive Developmental Disorder not otherwise specified. The largest group of participants (twenty-one) did not meet the criteria of the DSM-IV for hallucinations and delusions. They had a few symptoms consistent with borderline personality disorder, conduct disorder and schizotypal disorder. However, none of this group met the criteria for any of these disorders. Eighty-five percent had co-morbid ADHD, however this diagnosis did not explain their clinical presentation. They displayed excessive age-inappropriate fantasy, significant neuropsychological deficits and perceptual disturbances, behaviour problems and difficulties with language. Yet, there seemed to be no deterioration of functioning over time. McKenna et al. (1994: 638) labelled this
group as “Multidimensionally Impaired” (MDI), suggesting these children may
present a “particular diagnostic challenge because so many of their symptoms are seen
in other disorders” (McKenna et al., 1994: 642). This diagnosis does not appear in the
DSM system.

McKenna et al. (1994) noted that hallucinations and disorganised behaviour were
common in other disorders such as depression. The differentiation between
developmentally immature thought and deviant thought was difficult to make and
results differed depending on which diagnostic tool was used. Distinguishing flat
affect from psychomotor retardation presented another difficulty. They also found
that incoherence, poverty of speech and derailment were seen in children with severe
language impairments. This made diagnosis of psychotic disorders difficult. In
conclusion, they emphasise that though COS is a valid diagnosis it is used too
frequently. This may be because no diagnostic category exists for children with
intermittent psychotic symptoms, who also have developmental impairments.

A study by Tompson, Asarnow, Burney Hamilton, Newell and Goldstein (1997)
compared the thought disorder and communication problems of children diagnosed
with depressive disorders to children diagnosed with schizophrenia-spectrum
disorders and to a normal control group. Schizophrenia-spectrum disorders showed
the highest levels of thought disorder. Major depression showed less thought disorder
and normal controls had the lowest levels of thought disorder. Children with
schizophrenia and schizotypal personality disorder were similar on both thought
disorder and communication measures. Thus, a clear distinction could be made
between children suffering from depressive disorders and children suffering from
schizophrenia-spectrum disorders based on their performance on measures of thought disorder and communication.

In a comparison of multidimensionally impaired disorder (MDI) to COS and ADHD, it was suggested that MDI may be a variant of COS (Kumra et al., 1998). The Kumra et al. (1998) study found that MDI and COS had a common pattern of premorbid developmental delays. The MDI group and COS group seemed to have difficulty with comprehension of what they read despite showing the necessary word recognition skills. They seemed to share a deterioration in cognitive ability, and there was evidence that they have a greater incidence of schizophrenia-spectrum disorders in their first degree relatives. However, the transient Pervasive Developmental Disorder symptoms were more prevalent in the MDI group and this group also had an earlier onset of both cognitive and behavioural problems, and of psychotic symptoms. The ADHD group and the COS group performed significantly lower than the MDI group on measures of attention and concentration. Thus, no clear-cut evidence could be found for the supposition that MDI may be a variant of COS. In addition the sample size was small, a number of the participants were on drug treatments at the time of the study and the majority of information gathered was retrospective. Conclusions made based on the findings of this study are tentative.

A study by Karatekin and Asarnow (1998) compared the working memory of a COS group, an ADHD group and a normal control group. They found that the ADHD participants and the COS participants did not differ on any of the measures of working memory. No impairments could be found that were specific to either of the disorders. Both groups showed deficits in verbal and spatial memory, performing poorly as
opposed to the normal children. Interestingly, the COS group though deficient on verbal working memory tasks, performed as well as the normal group on immediate recall, but were impaired on delayed recall. ADHD children had more errors than the normal group on both immediate and delayed recall tasks. Karatekin and Asarnow (1998) queried the capacity of the sensory buffers of these two groups and described the “availability and allocation of resources to the central executive” as quite possibly limited.

In a study comparing the neuropsychological deficits of children diagnosed with COS and children diagnosed with Pervasive Developmental Disorder-not otherwise specified (PDD-NOS), it was found that deficits were similar in these groups (Kumra et al., 2000). An eight-hour battery of neuropsychological tests was administered to both groups over two sessions. The majority of subjects in both groups had cognitive difficulties on the same types of tasks, though the COS group were more impaired in terms of verbal learning and less impaired in regards to coding. However, once again the sample size was small. A large proportion of the sample was on medication at the time of testing and the test battery was lengthy and administered over a short period of time. In addition, the PDD-NOS group were younger than the COS group. These factors may have influenced the performance on tasks and make comparison between the groups difficult (Kumra et al., 2000).

Thirty-three participants who had been excluded from a study of COS on the basis of an evaluation and were given a diagnosis of a mood disorder, together with six children who had been diagnosed as definitely suffering from COS, were re-evaluated by three psychiatrists who were blind to the diagnoses given (Calderoni et al., 2001).
It was found that consensus reliability was significantly good between raters. Mood disorders had a specific clinical presentation as opposed to COS. Hallucinations were mood-congruent rather than mood-incongruent. There was no grossly disorganised behaviour or speech noted in the mood disorder group and often this group had a co-morbid disorder typical of mood disorders such as anxiety. In addition, mood disorders had a different prognosis with no deteriorating course and no chronic psychotic symptoms. However, schizoaffective disorder presented as diagnostically problematic. A retrospective evaluation of the period of overlap between the mood disorder and psychotic symptoms was difficult to make. Depressive symptoms resembled the negative symptoms of schizophrenia and it was difficult to distinguish mania from the agitation and disorganisation of schizophrenia. Thus, schizoaffective disorder rather than COS was the most difficult to identify. Based on their findings, Calderoni et al. (2001) emphasised the need to assess, firstly the nature of psychotic symptoms (in other words, whether hallucinations and delusions are mood-congruent or not) and secondly to consider the overall course of the illness and the level of functioning of the child before making a diagnosis. They argue that psychosis is a common feature of childhood disorders and that differentials should be considered carefully before making any diagnosis.

In summary, various research studies have been conducted comparing COS to other diagnoses. It is argued that psychotic symptoms in children may be more common than previously assumed and that COS may be used overinclusively (Calderoni et al., 2001; McKenna et al., 1994). Though, different aspects have been compared such as thought disorder, neuropsychological abilities and working memory (Kumra et al., 1998; Kumra et al., 2000; Tompson et al., 1997), it seems that there is a lack of
extensive, systematic research comparing COS to its differential diagnoses. Though Calderoni et al. (2001) make some attempt to provide for this by exploring the elimination process to find a diagnosis of COS, their findings need to be supported by further research.
Discussion

The purpose of this study was to explore what the last ten years of literature suggests about the clinical presentation of COS. It seems that auditory hallucinations are predominant, and delusions, while not occurring as regularly, are a common feature of COS (Green et al., 1992; Russell et al., 1990; Spencer & Campbell, 1994). With regards to thought disorder, findings suggest that COS sufferers have specific rather than generalised communication deficits (Abu-Akel et al., 2000; Caplan, 1994a; Caplan, 1994b; Caplan et al., 1996). Neuropsychological studies indicate that children with COS may perform poorly on neuropsychological tests (Bedwell et al., 1999; Hendren et al., 1998). In terms of premorbid history, there seem to be marked behavioural and psychological disturbances including, a lack of social development, developmental delays, pervasive developmental disorder symptoms, behaviour problems and ADHD symptoms. This history of premorbid abnormalities is significantly greater for COS than for EOS and AOS (Eggers, 1997; Eggers et al., 1999; Eggers et al., 2000; Frazier et al., 1997; Russell, 1994; Russell, 1998; Schaeffer & Ross, 2002). COS seems to have an insidious onset rather than an acute onset. This distinguishes it from EOS which the findings indicate generally has an acute onset (Alaghband-Rad et al., 1995; Bunk et al., 1999; Eggers & Bunk, 1997; Eggers et al., 1999; Eggers et al., 2000). Though there are exceptions, to a large extent, COS seems to have a deteriorating course and the diagnosis remains stable (Mezaide, Gringras, Rodrigue et al., 1996; Asarnow et al., 1994; Asarnow & Tompson, 1999). There is evidence for a complete remission from COS. However, for the majority of cases there seems to be a poor outcome (Eggers et al., 1999; Eggers et al., 2000; Mezaide, Bouchard, Gringras et al., 1996; Mezaide, Gringras, Rodrigue et al., 1996;
Asarnow et al., 1994; Asarnow & Tompson, 1999). In terms of differential diagnoses, COS seems to be used overinclusively for childhood psychosis. It seems that psychotic symptoms in children are more frequent than commonly believed (Calderoni et al., 2001; Kumra et al., 1998; McKenna et al., 1994).

Limitations

There is a definite paucity of research on the phenomenology of COS. Sample sizes are relatively small and the number of studies focusing on particular aspects (for example, thought disorder, neuropsychological abilities and course) are limited. Participants were often severely ill and medicated. This is of particular relevance as it would impact on the clinical presentation of COS and on the ability of a child diagnosed with COS to perform tests such as communication measures and psychometric assessments. The studies on onset, course and outcome were often retrospective, relying on parent and teacher reports as well as medical records, limiting their reliability.

Different studies used different measures. This limits one's ability to make broad conclusions based on the findings. Related to this, is the fact that the differentiation between COS, EOS and AOS was not applied systematically in the research. This may be because two particular schools of researchers are working in this area: the Essen studies in Germany (Eggers et al., 2000) and the American work (Calderoni et al., 2001; McKenna et al., 1994). Thus, there has not been an exploration of how COS may present on a wider scale, particularly in developing countries and in cross-cultural contexts.
Recommendations

Systematic, extensive research, with larger sample sizes needs to be done in this area. Particularly, an exploration of the comparison between the presentations of COS and its differentials would be fruitful. Research on the clinical presentation of COS in developing countries and cross-cultural settings may add insights that could be particularly relevant to the South African context. Further research on the differences between COS, EOS and AOS may pave the way to a better system of diagnosis and treatment of schizophrenia. This is particularly relevant in the case of young children (COS), where the research seems to indicate several differences in presentation, namely in the premorbid history (COS seems to have a greater degree of premorbid abnormalities as opposed to EOS and AOS), in the type of onset (insidious as opposed to acute) and prognosis (deteriorating course and poor outcome). The possibility that COS may be a more debilitating type of schizophrenia suggests that research should continue in the area of prevention, early diagnosis and treatment.

Due to the lack of research pertaining to COS no definite conclusions can be made about the implications of the findings for clinical practice. At present, the DSM and ICD-10 do not provide detailed descriptions of COS and EOS as opposed to AOS. Though generally it has been argued that COS is identical to AOS in presentation, the research suggests that a distinction between the two may be warranted. Three differences are indicated. Firstly, COS seems to show a more severe premorbid history. Secondly, it seems that COS is more likely to have an insidious onset. Lastly, COS appears to have a poorer prognosis (deteriorating course and poor outcome). Further research is needed to provide more substantive evidence for these three differences.
The research suggests that psychotic symptoms do not necessarily imply schizophrenia and that these symptoms may be more common in children than previously believed. Thus, clinicians are advised to consider the child’s premorbid history, level of functioning and the ‘appropriateness’ of the psychotic symptoms for the context that the child finds herself in. These considerations may provide valuable information for the process of diagnosis and thus, will impact on treatment (how soon it is implemented and how appropriate it is for the symptomatology).

A final consideration with regards to the research on COS, is the use of the adult criteria from the DSM system to select participants for the studies. This creates a situation where researchers find what they are looking for because of the criteria they are using. This presents future research with a dilemma. Using the DSM system criteria means that they will in essence be assuming a similarity between COS and AOS, despite possible differences (as mentioned above). However, if they were to discard the DSM system criteria, the question would be: which selection criteria to use? This suggests a tension between having too narrow a category as opposed to having too broad a category on which to base selection criteria. Research is needed to explore this further.

In conclusion, the scant information available indicates that COS is a more severe form of schizophrenia which in most cases has a clear premorbid history, an insidious onset, a deteriorating course and a poor outcome. Further research needs to have larger sample sizes, be more systematic in its approach (particularly with regards to the classification system and measures used) and extraneous variables (such as the
influence of medication, the impact of the illness on performance and the problematic nature of retrospective material) need to be considered. Despite the lack of research on the clinical presentation of COS, the findings over the last ten years provide important insights regarding the presentation of this disease, and a wide array of possibilities for future research.
References


