Schizophrenia Spectrum Disorders 10

Colin Hemmings

Introduction

Historical Context

It has long been thought that the relationship between intellectual impairment and psychosis may provide important clues to the aetiology of psychosis in general (Offord & Cross, 1971). Kraepelin (1919) described *pfropfschizophrenie* as being psychosis literally 'engrafted' onto *idiocy* and accounting for up to 7 % of cases of *dementia praecox*. He believed that *pfropfschizophrenie* was an early onset and severe form of psychosis in pre-existing ID. Kraepelin also believed that a schizophrenia-type psychosis beginning in the first decade of life could itself cause 'mental defect' (Heaton-Ward, 1977). For a long time after however most researchers believed that the joint occurrence of both conditions in the same individual was merely a chance combination (Heaton-Ward, 1977). There was accordingly a lack of research interest in the study of co-morbid ID and schizophrenia (Turner, 1989). More recently there has been much renewed interest in the context of much better recognition and understanding of mental disorders in people with ID.

Nomenclature

The term *schizophrenia spectrum disorder* (SSD) is widely accepted to include the schizophrenias as well as schizoaffective, persistent delusional and schizoptypal disorders. The evidence base regarding the epidemiology, risk factors, neuropathology, psychopathology, management and prognosis of SSDs in people with ID will be reviewed. This chapter will focus (whenever possible) on SSDs in people with ID as opposed to the less specific category of (non-affective) psychosis, which is perhaps more diagnosed in clinical practice and thus has often been used instead of SSDs in the research literature.

Epidemiology

Diagnosis

SSDs in people with ID are generally more difficult to detect and diagnose than in the general population. Sometimes a specific diagnosis of an SSD in people with ID is impossible. 'First rank' (Schneider, 1959) positive symptoms of schizophrenia are conceptually complex and thus often difficult to reliably assess in people with ID

C. Hemmings (✉)
Mental Health in Learning Disabilities Service, South London and Maudsley NHS Foundation Trust, London, UK

Institute of Psychiatry, King's College, London, UK
e-mail: Colin.Hemmings@slam.nhs.uk

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(Hucker, Day, George, & Roth, 1979). There may be difficulties distinguishing true delusional beliefs and hallucinations from self-talk, fantasies and talking to imaginary friends, which can be developmentally appropriate for a person with ID (Heaton-Ward, 1977; Hurley, 1996). People with ID often 'remember' conversations and think out loud about them. They might not realise that this is 'socially inappropriate'. Develop-mentally appropriate features may become more prominent and exaggerated when under stress (Hellendoorn & Hoekman, 1992). Formal thought disorder may also be difficult to elicit in people with ID for it can often be difficult anyway to follow the thread of their speech. People with ID are also often hypersensitive to how they are perceived by others and this can often be grounded in real-life experiences of being mocked and rejected (Hurley, 1996). This hypersensitivity may be mistaken for delusional persecutory thinking. It has also been suggested that people with ID are more likely to be able to be persuaded of the falseness of the delusional beliefs. It may be more important to observe for their readiness to restate their beliefs when not so subject to suggestibility (Hurley, 1996). Apparent 'negative' symptoms can also be misattributed to a psychotic disorder when they may be due to many other factors such as institutionalisation, over-sedation, lack of environmental stimulation and severe cognitive impairment (James & Mukherjee, 1996). It may be difficult to discriminate psychotic symptoms from baseline features of ID, a diagnostic problem which Sovner and Hurley (1986) termed 'baseline exaggeration'. However, the difficulty of differentiating psychotic symptoms and features related to a person's ID might also lead to the problem of 'diagnostic overshadowing' (Reiss, Levitan, & Zyszko, 1982) and psychosis therefore being actually under-diagnosed in this service user group.

One of the particular problems with diagnosing schizophrenia in people with ID is the difficulty of differentiating it from autism (Bakken, Friis, Lovoll, Smeby, & Martinsen, 2007; Bradley, Lunsky, Palucka, & Homitidis, 2011; Helevershou, Bakken, & Martinsen, 2008; Palucka, Bradley, & Lunsky, 2008). Sometimes it may be virtually impossible to be definitive as to whether a person has autism instead of, or as well as, schizophrenia unless an adequate developmental history is available (James & Mukherjee, 1996). Unusual preoccupations held rigidly by people with autism may be particularly difficult to differentiate from delusions. Flattened and incongruent affect, poor non-verbal communication and poverty of speech are commonly seen in autism as well as schizophrenia and so also lead to diagnostic difficulties. The term 'autism' itself was actually coined by Bleuler (1911) to describe social withdrawal in schizophrenia. He included autism as one of the four core characteristics of schizophrenia. Kolvin (1971) though clearly later showed that the pervasive developmental disorder of childhood autism was distinct from schizophrenia and other psychoses. There has been growing research activity into the associations between ID, autism and psychopathology including symptoms of SSDs (Matson & Shoemaker, 2009). It still remains unclear as to whether the risk of SSDs in people with ID and autism is greater than in non-autistic individuals with ID.

Diagnostic Criteria

Research has been hampered by inconsistencies in diagnostic criteria and rating instruments used (Sturmey, 1999). A major reason for the inconsistency of approaches has been the continued doubts about the usefulness of standard diagnostic classification systems in diagnosing mental disorders in those with more severe ID. The major difference in the various approaches used has been whether researchers have used standardised criteria or criteria modified for people with ID. For example, Hucker et al. (1979) modified the Research Diagnostic Criteria of Feighner et al. (1972) in their study of schizophrenia in people with ID and thus included features such as 'behaves as if hallucinated' as evidence of the disorder. Nor have definitions of the term 'intellectual disabilities' been consistent. However, recent improvement in consistency of criteria used in studies of mental disorders in people with ID has allowed greater comparison between them. The specific problems of diagnosis of schizophrenia

in people with ID have improved in recent years with the development of standardised assessment instruments for this service user group (e.g. Hatton et al., 2005). In people with mild ID and good verbal skills, it seems that the standard classification systems can be used with reliability and validity (Dosen & Day, 2001). By analogy, ICD and DSM criteria for schizophrenia can be reliable when used with children with typical IQ as young as 7 years old (Green, Padron-Gayol, Hardesty, & Bassiri, 1992). The linguistic competence of a normal 7-year old is similar to an adult with moderate ID (Melville, 2003). The developmental perspective is helpful conceptually, as complex delusions and hallucinations are unusual in young children but become more frequent in older children and children with a higher IQ (Watkins, Asarnow, & Tanguay, 1988).

Most of the research literature on SSDs in ID has focused on people with mild ID (IQ 50–70). Reid (1972) influentially argued that it is impossible to diagnose schizophrenia in people with limited verbal communication. In practice, this means below an IQ level of around 45. A person with more severe ID has limited verbal expression and ability to employ abstract reasoning, which are essential for the manifestation of the complex, subjective experiences of the language-based symptoms of schizophrenia. Sovner (1986) has described this phenomenon as 'intellectual distortion'. Sovner and Hurley (1986) also argued that 'cognitive disintegration', or psychotic-type reactions to stress, occurred in people with ID because of their limited coping abilities. Higher incidences of SSDs have been found in those with mild ID compared with more severe ID (Bouras & Drummond, 1992; Cowley et al., 2004). It remains unclear whether this reflects reality or whether it merely reflects the problems in detection of psychosis in people with more severe ID. Most researchers probably share the view of Heaton-Ward (1977) that a person's degree of insight and touch with reality cannot be fully assessed if they do not have a reasonable level of intelligible communication. However the study of Einfield et al. (2007) showed that at least there was good inter-rater reliability among experienced clinicians about the clinical diagnoses of psychosis in people with ID.

Prevalence and Incidence

There have been long-standing difficulties with establishing true incidence and prevalence rates of SSDs in people with ID. Taking into consideration all of the foregoing problems in accurately diagnosing SSDs in people with ID, epidemiological studies have converged around point prevalence rates of around 3 % and this figure is now widely accepted (Deb, Thomas, & Bright, 2001; Fraser & Nolan, 1994). This prevalence rate is around three times higher than in the general population although still thought to be a possible underestimate of the true figure (Turner, 1989). In a population-based cohort study of people with ID, Cooper et al. (2007) found the 2-year incidence of non-affective psychoses to be 1.4 % and for the first episode incidence to be 0.5 %.

There are three major possibilities for the increased prevalence rates of SSDs in people with ID (Blackwood, Thiagarajah, Malloy, Malloy, & Muir, 2008). First, a lowered IQ could be a premorbid sign of schizophrenia (Gunnell, Harrison, Rasmussen, Fouskakis, & Tynelius, 2002). Second, cerebral abnormalities caused by genetic factors or in utero may cause both ID and SSDs. This has been described as the 'common aetiology' hypothesis (David, Malmberg, Brandt, Allebeck, & Lewis, 1997). Third, low IQ could lead to incorrect social assumptions and thus increased risk of symptoms such as delusions and hallucinations. David et al. (1997) showed that low intellectual ability in itself is a risk factor for schizophrenia and other psychoses, although their study, like the vast majority in schizophrenia research, excluded those with premorbid ID.

Risk Factors

Genetics

Kallman (1938) influentially reported no increase in ID in the relatives of people with schizophrenia and vice versa. This study concluded that the co-morbid state was a chance occurrence of two unrelated conditions and this contributed to a period of subsequent decreased research interest into co-morbid ID and schizophrenia (Turner, 1989).

However Kallman's study did not consider the co-morbid probands and, when Doody, Johnstone, et al. (1998) did so, they found that both the prevalence of ID and schizophrenia were increased in the relatives of the co-morbid probands. This supported the idea that a highly familial form of schizophrenia might occur in the ID population. Doody, Johnstone, et al. (1998) also showed that there was a high rate of chromosomal variants in karyotype testing in the co-morbid group. Cytogenetic anomalies, which are often associated with intellectual impairment, may be associated with an earlier age of onset of schizophrenia (Bassett et al., 1998). Schizophrenia and ID in association have been reported with a wide range of chromosomal abnormalities (Blackwood et al., 2008). Psychotic symptoms are sometimes associated with Prader-Willi syndrome (15q11-13 partial deletion), particularly the maternal disomy form (Boer, Holland, Whittington, Butler, & Webb, 2002). High rates of schizophrenia have been reported in adults with velo-cardio-facial syndrome (VCFS), which is associated with a deletion on chromosome 22 and is often associated with ID (Green et al., 2009). In addition, 2 % of service users with schizophrenia exhibit this 22q11 deletion but 6 % of the early onset cases do so (Usiskin et al., 1999). Increased rates of dysmorphic features, cognitive impairment and structural brain abnormalities have led to the proposal of 22q11 deletion as a model for a neurodevelopmental subtype of schizophrenia (Bassett, Chow, & Weksberg, 2000). However many studies of VCFS and psychosis have excluded people with ID.

Obstetric Complications

O'Dwyer (1997) found that in people who develop ID and schizophrenia, the number of obstetric complications increases, compared to those matched for age, sex, severity of ID and epilepsy. These complications included abnormally long labour, dysmaturity, pre-eclamptic toxaemia and maternal episiotomies, all of which may give rise to anoxic cerebral trauma, which the authors concluded was the major aetiological factor predisposing people with ID to develop psychoses. Hucker et al. (1979) studied 24 people with ID and schizophrenia and 40 control subjects. They found higher proportions of people with impaired hearing, gestation below 36 weeks and low birth weight in the group with schizophrenia.

Epilepsy

Deb et al. (2001) and Doody, Johnstone, et al. (1998) found that those with schizophrenia and ID were more likely to have epilepsy compared to those with schizophrenia of typical intelligence. These findings would be expected given that epilepsy is more common in people with ID (Bowley & Kerr, 2000). However, Cowley et al. (2004), Cooper et al. (2007) and Arshad et al. (2011) found in their studies that people with epilepsy and ID had a lower incidence of schizophrenia or non-affective psychosis than in people with ID only. These somewhat conflicting findings from research to date are perhaps unsurprising as epilepsy provides further complications in the diagnosis of SSDs in people with ID. For example, it can be difficult to differentiate temporal lobe epilepsy from schizophrenia. Ring, Zia, Lindeman, and Himlock (2007) found that psychoses were more common in people with ID and epilepsy when there had been no seizures for the preceding 3 months. The phenomenon of 'forced normalisation' has been described in relation to the schizophrenia-like psychoses of epilepsy (Trimble, 1996) whereby a reduction in seizure activity following the introduction of anticonvulsant agents is associated with an exacerbation of psychosis.

Other Risk Factors

Meadows et al. (1991) and Fraser and Nolan (1994) suggested that schizophrenia might have an earlier age of presentation in people with ID. One possible explanation given was that the existence of schizophrenia in the context of ID is more likely to produce a more severe illness

with earlier appearance of psychotic symptoms (Meadows et al., 1991). However, there is also the possibility that the presence of schizophrenia increases the likelihood of an individual being diagnosed as having ID, and vice versa (Sanderson, Doody, Best, Owens, & Johnstone, 2001). There can sometimes also be a delay in presentation of schizophrenia in people with ID due to the difficulties of detection, including the suggestion that the onset of psychosis may sometimes be less noticeable in people with ID (James & Mukherjee, 1996). However, Doody, Johnstone, et al. (1998) found no significant differences in the ages of first symptoms of schizophrenia, consultation, admission or diagnosis between subjects with both mild ID and schizophrenia and matched controls with schizophrenia alone. This was despite the fact that co-morbid subjects had more psychopathology and duration of admissions than the controls.

Doody, Thomson, Miller, and Johnstone (2000) reported that women with ID generally developed schizophrenia later than men with ID, who were also less likely to have a family history of the disorder. They suggested that gender differences in schizophrenia might be more pronounced among the co-morbid population where early cognitive impairment can herald the onset of a more severe form of onset. Cowley et al. (2004) found higher prevalence of SSDs with older age, whereas Cooper (1997) found no differences in the prevalence of schizophrenia between younger and older adults with ID. Ethnicity has also been associated with SSDs in the ID population in the UK, with a higher prevalence in those referred to services being from ethnic minority groups than would have been expected by chance (Chaplin, Thorp, Ismail, Collacott, & Bhaumik, 1996; Cowley et al., 2004; Tsakanikos, Bouras, Costello, & Holt, 2009). Sensory impairments are more common among people with ID (Carvill, 2001). It might be expected that sensory impairments would increase the risk of SSDs in people with ID, but there have been few studies to confirm this. Cooper et al. (2007) found that visual impairment was independently associated with psychosis in adults with ID.

Neuropathology

In the classification systems of DSM-IV (American Psychiatric Association, 1994) and ICD-10 (World Health Organisation, 1992), it is stipulated that schizophrenia should not be diagnosed in the presence of 'overt' brain disease, which is more common in people with ID (Hagberg & Kyllerman, 1983). This complicates the study of neuropathology in individuals with both schizophrenia and ID. A recent case-controlled study of volumetric cerebral magnetic resonance reported that in terms of brain structure, people with ID and schizophrenia resemble those with schizophrenia more closely than those with ID (Sanderson, Best, Doody, Owens, & Johnstone, 1999). These findings were confirmed by further studies (Moorhead et al., 2004; Sanderson et al., 2001). Sanderson et al. (1999) suggested that the higher frequency of schizophrenia in ID was due to the greater tendency of people with schizophrenia to develop cognitive deficits and that within the ID population there may be people whose deficits result from undiagnosed schizophrenia. These authors support the view that co-morbid schizophrenia and ID may represent an early onset and severe form of schizophrenia, rather than ID complicated by psychosis (Moorhead et al., 2005; Spencer et al., 2007). It could be then that a subset of adolescents with ID have a form of schizophrenia that has not yet made itself manifest. Hence Johnstone et al. (2007) looked at schizotypal traits in adolescents with ID and used these to screen for those likely to develop schizophrenia at a later date. Welch et al. (2010) found that within a sample of people with ID those with schizotypal traits showed reduced amygdala volume on magnetic resonance imaging scanning compared to those with ID alone.

Psychopathology

Presentation

Disagreement remains as to what extent the presentation of SSDs is different in people with and

without ID. Most of the literature suggests that there are no *substantial* differences in the symptoms of schizophrenia between people with ID and people of normal intelligence (Hucker et al., 1979; Moss, Prosser, & Goldberg, 1996; Reid, 1972). Meadows et al. (1991) suggested that there was no difference in the presentations of symptoms of schizophrenia between those with and without ID although there was still a trend for less overt psychopathology in 13 of the 16 symptoms. Bassett et al. (2003) also found no significant differences in the schizophrenia phenotype between service users with and without ID. However other studies have found differences in presentation, especially a tendency for less florid positive symptoms of schizophrenia in those with ID. For example, Linnaker and Helle (1994) found that those diagnosed with schizophrenia with the Psychopathology Instrument for Mentally Retarded Adults (PIMRA) had fewer delusions, flatter affect and more incoherent speech than people with schizophrenia of normal intelligence, especially persecutory delusions and formal thought disorder. Bouras et al. (2004) showed that those with ID and SSDs showed more observable and fewer self-reported symptoms than people with SSDs alone.

Turner (1989) described the symptoms of schizophrenia in people with ID as 'often shallow or banal'. Because of their reduced opportunity to engage in normal life experiences and social opportunities, the delusions of people with ID, when they occur, may be relatively simple. For example, there is little attempt to interpret strange subjective phenomena and secondary elaboration of abnormal perceptions or odd beliefs producing complex delusional systems rarely occurs (Reid, 1989). Complex hallucinations such as voices giving as running commentary in the third person or discussing the person between themselves are also uncommon. Sovner (1986) described this phenomenon as an example of 'psychosocial masking' of symptoms in people with ID. James and Mukherjee (1996) found that the presenting complaints of schizophrenia in people with ID were very often a decline in functioning, deterioration of skills and social withdrawal, rather than hallucinations and delusions. Reid (1993) suggested that hebephrenic, paranoid and catatonic subtypes of schizophrenia could all be seen in people with ID. However, most clinicians believe that it is less often possible to sub-classify schizophrenia in people with ID than in those with more typical IQ.

Positive and Negative Symptoms

Although researchers in the field of psychosis in ID have used the categorisation of psychotic symptoms into 'positive' and 'negative' (Andreasen, 1982), the validity of these in the ID population remains unclear (Melville, 2003). There have again been some apparently conflicting findings; for example, Bannerjee, Morgan, Lewis, Rowe, and White (2001) and Cherry, Penn, Matson, and Bamburg (2000) emphasised positive symptoms in the presentation of schizophrenia in people with ID. Doody, Johnstone, et al. (1998) and Bouras et al. (2004) found 'negative' symptoms to be more common in people with ID and SSDs than in SSDs alone. Moss et al. (1996) showed negative symptoms to have low specificity for schizophrenia in adults with ID. They found that auditory hallucinations seemed easiest to elicit in people with ID and schizophrenia, followed by thought disorder, and then by delusions relating to replacement of will.

'Behavioural' Symptoms

Some have argued that there may be possible behavioural manifestations or 'behavioural equivalents' of mental disorders such as psychosis in people with ID (Dosen, 2005). Associations have been found between psychotic symptoms and problem behaviours in people with ID, but their relationship remains unclear (Myrbakk & von Tetzchner, 2008). Certainly some behavioural problems respond to antipsychotic medication suggesting that they might have been caused or exacerbated by underlying psychosis that is difficult (if not impossible) to diagnose clearly using standard diagnostic criteria. It has been argued that people with ID and psychosis may

present with other atypical features. For example, hysterical type symptoms such as over-breathing, pseudo-fits, gait disturbance and Ganser states may be more frequent in people with ID and psychosis (Dosen & Day, 2001; Hucker et al., 1979; Reid, 1972). Some schizophreniform psychoses also present acutely with apparent changes in consciousness (Reid, 1993). Turner (1989) reported that disturbed and aggressive behaviour, bizarre rituals and 'hysterical' behaviours are relatively more common in ID and may complicate the presentation of psychosis in ID. People with ID and schizophrenia may also present with social withdrawal, fearfulness and sleep disturbance (Eaton & Menolascino, 1982; Myers, 1999). The 'DC-LD' guide (Royal College of Psychiatrists, 2001) suggested that early signs of a psychotic illness could be new problem behaviours (especially when odd or bizarre or uncharacteristic for the person), or an increase in frequency or severity of pre-existing behaviours.

Behaviour that might suggest auditory hallucinations could be the person with ID shouting back apparently at people not present when this has not been their previous behaviour. Similarly, suspiciousness, blunted or incongruent affect and social withdrawal not previously part of the person's personality and behaviour could also be suggestive of a schizophreniform-type psychosis. Non-verbal evidence for possible psychosis by necessity becomes of greater diagnostic significance in the more severely intellectually disabled. It has been claimed that that it is possible using observations and non-verbal communication to diagnose schizophrenia in people with more severe ID. Both Eaton and Menolascino (1982) and Cherry et al. (2000) claimed behaviour characteristic of schizophrenia could be identified in people with severe ID. Heaton-Ward (1977) also argued that for some people with more severe ID, their emotional lability, noisy outbursts and disorganised, purposeless activity, including aggression, destructiveness and self-mutilation, could be considered 'psychotic'. However, he also recognised that it would be impossible to determine whether such individuals were demonstrating lack of insight and loss of sense of reality, which are fundamental to psychosis. Turner (1989) noted the persistent comment on catatonia in the literature. Hucker et al. (1979), Eaton and Menolascino (1982) and Heaton-Ward (1977) all remarked upon the high proportion of their study samples of subjects with ID and schizophrenia who showed catatonic features. The relationship between catatonia and schizophrenia in people with ID is complex and still needs further clarification.

Cognitive Functioning

An episodic psychotic illness may be associated with a reduction in transient functioning to the mild or even moderate level of ID (Russell & Tanguay, 1981). A chronic psychosis may, however, lead to a prolonged or even permanent reduction in intellectual and social functioning. Sometimes those with chronic schizophrenia are thus mistakenly diagnosed as also having ID because of deterioration in intellectual functioning. There has been little specifically published though in the literature of neuropsychological findings in people with ID and schizophrenia. Heaton-Ward (1977) found a 20 % fall in IQ in some of his service users with ID and schizophrenia over time. Doody, Johnstone, et al. (1998) found that their co-morbid subjects were more likely to have impairment of episodic memory, which may affect their compliance with treatment. They also had greater impairment of theory of mind on second-order tests than subjects with schizophrenia and normal pre-morbid IQ (Doody, Gotz, Johnstone, Frith, & Cunningham Owens, 1998). Rowe, Rudkin, and Crawford (2000) found that there was an increased rate of mixed handedness among people with co-morbid ID and schizophrenia compared to controls. Mixed handedness is taken as an index of diminished cerebral dominance or laterality, and thus these findings also supported the idea of a neurodevelopmental hypothesis of schizophrenia. Weinberger (1987) has suggested that people with ID and schizophrenia form a subgroup of schizophrenia with a neurodevelopmental aetiology. Doody, Johnstone, et al. (1998) found evidence consistent with the neurodevelopmental

hypothesis in their study of co-morbid ID and schizophrenia. For example, they found that their co-morbid subjects had more 'soft' neurological signs than controls. They suggested that there is a form of schizophrenia that manifests in childhood with cognitive impairment prior to the onset of psychotic symptoms.

Management

The principles of treatment of schizophrenia in people with ID are essentially similar to those without ID. There have been a few studies that have suggested that treatment with antipsychotics in ID is broadly similar in efficacy with no significant risk in side effects (e.g. Craft & Schiff, 1980; Reid, 1972). Shedlack, Hennen, Magee, and Cheron (2005) found that in those with ID and SSDs, there was substantial improvement in social withdrawal following treatment with both typical and atypical antipsychotic medication. Some studies have reported beneficial effects (and sometimes fewer side effects) with atypical antipsychotics (e.g. Advokat, Mayville, & Matson, 2000; Shastri, Alla, & Sabaratnam, 2006; Williams, Clarke, Bouras, Martin, & Holt, 2000). However Matson and Mahan (2010) cautioned that a long-term perspective on atypical antipsychotic use in people with ID is still not yet possible in the same way as it is with typical antipsychotic medications. It is still not clear whether people with ID and SSDs, or at least a subgroup of them, are more likely to be treatment-resistant. Clozapine is efficacious and well tolerated in people with ID and co-morbid mental illness including schizophrenia (Antonacci & de Groot, 2000). Varghese and Banerjee (2010) reported in their audit of their community teams that of seven of their treatment-resistant service users, only two had been considered for clozapine. They noted the additional difficulties in prescribing clozapine for clinicians treating people with ID.

Clarke (2001) argued that people with brain pathology are more likely to develop tardive dyskinesia than the general population. Sachdev (1992) found that 34 % of those people with ID receiving antipsychotic medication in a long-stay institution had tardive dyskinesia. Gingell and Nadararajah (1994) also suggested that people with ID might be at greater risk of developing movement disorders when taking antipsychotic medication. However, the evidence for increased risk of tardive dyskinesia due to antipsychotic medication in people with ID and SSDs relative to those with SSDs alone is not yet conclusive. For example, Gualtieri, Shroeder, Hicks, and Quade (1986) found no evidence that ID increased the risk. One possible reason for this discrepancy in findings is that differentiating between movement disorders and medication side effects is again more difficult in service users with ID. Rogers, Karki, Bartlett, and Pocock (1991) showed that people with more severe ID also have high rates of motor disorders not attributable to antipsychotic medication. The presence of muscle tone abnormalities and stereotypies including tics, mannerism and self-stimulatory behaviours can all mask the presence of antipsychotic-induced movement disorders (Shedlack et al., 2005). One reason is the increased propensity towards tardive dyskinesia in people with ID relative to those with normal IQ (Cohen, Khan, Zheng, & Chiles, 1991; Fodstad et al., 2010; Rao, Cowie, & Matthew, 1987). However none of these studies have conclusively answered the complex question of whether people with ID and SSDs are even more likely to develop tardive dyskinesia as a result of being treated with antipsychotic medication than people with SSDs but without ID would be. It has also been suggested that the risk of neuroleptic malignant syndrome may also be possibly increased in people with ID on antipsychotic medication (Boyd, 1992).

Many clinicians have reported that the optimal dosage levels of antipsychotic medications in clinical practice appear to be lower in service users with ID than in those with normal IQ (Menolascino, Ruedrich, Golden, & Wilson, 1985). Problems such as possible exacerbation of pre-existing cognitive impairment, the lowering of seizure thresholds and the reduced ability to communicate symptoms and side effects can make the prescribing of antipsychotics for people with ID and SSDs even more difficult than

in people with SSDs without ID. There is a consensus that antipsychotic prescribing often needs to be instigated and increased more cautiously than in service users without ID. Duggan and Brylewski (1999) argued that there was still insufficient evidence to judge the efficacy of antipsychotics when used specifically in people with ID. The use of antipsychotics to treat SSDs in this service user group therefore is still largely based on extrapolation from the evidence base of general adult psychiatry. There remains a need for randomised controlled trials of the efficacy of antipsychotic medication in people with ID and SSDs. Electroconvulsive therapy (ECT) is sometimes used in the treatment of SSDs (e.g. in catatonic states), but there has been little published regarding its use as treatment of SSDs specifically in people with ID (Chanpattana, 1999).

There have often been criticisms of prescribing antipsychotic medication for people with ID and SSDs given the dearth of specific evidence for 'biological' treatments, but there is also not a great deal of evidence regarding the use of psychosocial (including family) interventions. Crowley, Rose, Smith, Hobster, and Ansell (2008) conducted a preliminary study of using two psycho-educational groups for eight people with the dual diagnoses of psychosis and ID. Participants were reported to have had their knowledge increased about psychosis, medication, early signs of relapse and the role of stress. Behavioural techniques have been used to improve the social skills of individuals with psychosis and ID (Hatton, 2002). The application of either individual or family cognitive-behaviour therapy (CBT) approaches has not been systematically evaluated adequately as yet in people ID who have psychosis. A case series of CBT for five service users with psychosis and mild ID has been reported (Haddock, Lobban, Hatton, & Carson, 2004). This included two cases in which family interventions were also integrated into the individual CBT. Taylor, Lindsay, and Willner (2008) have argued that it is unjustified to not make available psychotherapy based on cognitive-behavioural approaches for people with ID and psychopathology.

Prognosis

It has long been established that in service users with schizophrenia there is a relationship between pre-psychotic IQ and disease prognosis (Offord & Cross, 1971). There has been very little published, however, into the long-term prognosis in people with ID and schizophrenia. There have not been either any published studies of the latency period or prodrome before presentation of psychotic symptoms in this service user group. Most clinicians believe that the course of schizophrenia in people with ID will tend to be more severe than in people with normal IQ. However, again the literature is scant and findings have not always been consistent. For example, Reid (1972) reported that the psychoses ran a more benign course in people with ID, especially in those with more severe ID.

Bouras et al. (2004) showed that in a group of people with ID and SSDs, matched for duration of illness, there were greater functional disabilities compared to a group with SSDs attending a general mental health outpatient clinic. The authors suggested that the increased prevalence of observable psychopathology might lead to increased risk of stigma and social isolation for those with SSDs who also had ID. The higher rate of negative symptoms seen also might contribute to social withdrawal and isolation and raised the question of whether those with ID as well as SSDs might have progressed more rapidly to the chronic deficit state of SSDs. The authors suggested that people with ID are more debilitated by SSDs than those without ID and may thus need additional input. Doody, Johnstone, et al. (1998) found that service users with dual diagnosis of schizophrenia and ID had fewer psychiatric admissions but for longer periods of time, and that at point of discharge they needed more support than service users with schizophrenia alone. With some outcome parameters, such as total time in hospital and offending behaviour, there was no evidence that the comorbid group were more impaired than the schizophrenia control group. However they also found that those with both conditions received

more community supports than control subjects with schizophrenia alone. Doody et al. (2000) showed that in people with co-morbid ID and schizophrenia, males with an early age of onset and no known family history of either ID or schizophrenia were more likely to require care and treatment in a high-security hospital. Chaplin et al. (2006) also found that people in their study with lower IQ (including those with borderline low IQ as well as those with ID) and schizophrenia had a lower quality of life, more severe psychotic symptoms and reduced functioning compared to those with schizophrenia and higher IQ.

SSDs in people with ID cause a great deal of human suffering, carer burden and also significant service and care costs. Hemmings, O'Hara, et al. (2009) found that the SSDs were overall the largest group of diagnoses for people with ID admitted to both a specialist and generic mental health units in their study in South London. In Taiwan, Lai, Hung, Lin, Chien, and Lin (2011) have reported that a large proportion of inpatient admissions and associated financial costs were accounted for by a coexisting diagnosis of schizophrenia in people with ID. For community services, focus group studies (Hemmings, Underwood, & Bouras, 2009a) and a Delphi study (Hemmings, Underwood, & Bouras, 2009b) have helped to identify a consensus of what needs to be provided for this disadvantaged dually diagnosed service user group. There is some evidence that outcomes may be better when people with both ID and psychosis are treated by specialist rather than generic services (Raitasuo, Taiminen, & Salokangas, 1999).

Conclusion

The evidence base regarding SSDs in people with ID remains limited. There is some evidence though to suggest that people with ID and SSDs may have even more complex needs than service users with SSDs of typical intelligence and may be harder to treat. We are not likely to see any clear-cut relationships between ID and SSDs when ID is itself on a spectrum with the general population. Although in some individuals with ID the development of SSDs may be by chance, it seems that for others there may be direct and indirect links. The relationship between IQ and schizophrenia is complex, as lower IQ may cause vulnerability to the development of schizophrenia or be an early manifestation of the disorder (Offord & Cross, 1971). Many believe that co-morbid ID and schizophrenia may often represent a severe form of schizophrenia with poorer outcomes (Doody, Johnstone, et al., 1998) and both arising from the same genetic aetiology. The majority of studies of SSDs have excluded people with premorbid ID but is important that further research does not unless there are compelling reasons to do so. Studying SSDs in people with ID might not only improve diagnosis and treatment but also help to identify those with ID who will go on to develop these severe mental illnesses. It may also provide one important way forward in the understanding of the SSDs in the general population. Research across the biopsychosocial spectrum is needed and this must include developing our knowledge on the best service provision and outcomes for this dually disadvantaged group of service users and their careers.

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