Schizophrenia Spectrum Disorders

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Introduction

Psychosis is a non-specific term denoting a mental condition in which the sufferer loses contact with reality. Psychosis can have several causes, including organic illness (in which the altered mental state is due to a primary physical cause) and functional disorder (in which a primary abnormality of the mind is considered the cause).

Schizophrenia spectrum disorders (SSDs) comprise a cluster of mental disorders in which the most prominent feature is psychosis. SSDs vary in their duration, intensity, and course. They are more common in people with intellectual disability (ID) than the general population. The assessment, diagnosis and management of these disorders presents challenges to the clinician and the presence of such an illness can have a significant impact on an individual's functioning and quality of life.

Classification of schizophrenia spectrum disorders

SSDs represent a heterogeneous array of clinical syndromes. There is increasing support of a dimensional conceptualisation of psychosis (Esterberg & Compton 2009) but convention dictates categorisation of these disorders based on major signs and symptoms.

The pre-eminent manuals used to classify and diagnose mental illnesses are the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) (American Psychiatric Association 2013) and the International Statistical Classification of Diseases and Related Health Problems (ICD-10) (World Health Organisation 1992).

BOX 1 – schizophrenia spectrum disorders defined in ICD-10

1. **Schizophrenia**

As the archetype psychotic illness, schizophrenia is a disorder of markedly distorted thinking, perception and affect. Main symptoms include thought disorder, delusions, hallucinations, and negative symptoms. Cognitive deficits develop over time. Subtypes have been delineated by course and major features.

- Paranoid schizophrenia

  The most common subtype. Paranoid or persecutory delusions and auditory hallucinations dominate.

- Hebephrenic schizophrenia

  Delusions and hallucinations are less prominent. Behaviour is grossly disorganised and emotional response is lacking or inappropriate.

- Catatonic schizophrenia

  Bizarre motor manifestations are the hallmark of catatonia. These include, but are not limited to, posturing, waxy flexibility, stupor, and mutism.

- Residual (and simple) schizophrenia

  A slow but progressive decline in functioning accompanies social withdrawal and the appearance and deepening of negative symptoms.

2. Schizotypal disorder

Sometimes considered a form of personality disorder, schizotypal disorder is characterised by social and interpersonal deficits. There may be suspiciousness, odd beliefs, eccentric behaviour and unusual perceptual experiences, though no single feature predominates. The disorder runs a chronic course.

3. Persistent delusional disorder

Chronic, frequently lifelong delusions occur in the absence of other psychotic symptoms.

4. Acute and transient psychotic disorder

Psychotic symptoms develop rapidly and are not due to intoxication or an organic condition. There is an equally rapid and usually spontaneous recovery.

5. Induced delusional disorder

Occurs when a delusional belief, and sometimes other psychotic symptoms, are shared by two or more people with close emotional links. The sufferers are typically socially or physically isolated from others. Symptoms resolve in at least one of the sufferers following geographical separation. The illness is also known by the French term folie à deux, literally “madness of two” (Shimizu et al. 2007).

6. Schizoaffective disorders

A disorder with prominent, persistent mood (affective) symptoms and episodic psychotic symptoms, not clearly coinciding with periods of extremes of mood.

DSM-5 recently superseded DSM-IV (American Psychiatric Association 2000). The new edition incorporates some important changes but main categories of SSD still broadly map to ICD-10. Subtypes of schizophrenia have been removed from DSM-5 owing to limited diagnostic stability, low reliability, and poor validity, and there has been an attempt to define disorders along a spectrum with a gradient of symptoms and continuum of severity (Bhati 2013).

Operationalised criteria used to diagnose mental disorders in the general population may not be generalizable to people with ID, particularly in those with the greatest degree of disability (Clarke et al. 1994). The DM-ID (Fletcher et al. 2007) and DC-LD (Royal College of Psychiatrists 2001) have been developed from DSM and ICD criteria respectively, to better serve this population, although there remains debate about their utility in improving ascertainment of SSDs especially in people with more severe ID (Melville 2003).

Clinical features

The manifestation of SSDs is diverse. People with the condition have one or more of the following core features: delusions, hallucinations, disorganised thinking, motor disturbance, and negative symptoms.

- Delusions are fixed false beliefs that are held despite conflicting information or evidence. The content of such beliefs is often bizarre or implausible and does not reflect an individual's cultural background. Common themes include persecutory delusions (in which an individual believes they are at risk of harm), grandiose delusions (where the belief is of special powers or ability), and delusions of control (where one may believe that their thoughts, feelings or behaviours are under the control of an external force).

- Hallucinations are defined as sensory experiences that are not caused by external stimuli. They can occur in any modality, although auditory hallucinations are the most common type in functional psychoses. Typical content in schizophrenia is of voices that provide a running commentary of a person's actions and refer to the subject in the third person. True hallucinations occur in clear consciousness, the sufferer has no agency over them, and are perceived as intrusive.

- Thought disorder is a clinical sign that reflects disorganised thinking. It is identified by abnormalities in speech and can present in many ways, including speech that

abruptly stops mid-steam (thought blocking) or moves quickly between topics (flight of ideas).

- Catatonia describes a cluster of motor symptoms. The presentation is varied and may include either excessive activity or drastically reduced movements together with a decreased reactivity to the environment (Rajagopal 2007; Francis 2010).

- Negative symptoms cause withdrawal that contributes to impaired psychosocial functioning. Examples include emotional blunting, anhedonia, avolition and poverty of speech (Stahl & Buckley 2007).

Prevalence

Studies investigating prevalence rates of SSDs in people with ID report varying results. Reasons for such variation include, diversity in sampling frames, inconsistent definitions of both ID and psychosis, and differences in case ascertainment and assessment (Buckles et al. 2013). As a general estimate, schizophrenia prevalence is often quoted as 3% in people with ID, compared with an approximately 1% lifetime risk in the general population (Perälä et al. 2007).

Cooper et al. conducted a population-based cohort study of over 1,000 adults with intellectual disability who were followed-up for 2 years (Cooper et al. 2007). The point prevalence of psychotic disorders was reported as 2.6-4.4%, dependent on diagnostic criteria used, and the two year incidence of psychotic episode was 1.4%. Findings of this study are in agreement with earlier work that reported a prevalence rate of schizophrenia of 4.4% in a Welsh cohort of adults with ID (Deb et al. 2001).

Morgan et al report the results of a large study based on a population register in Western Australia. They found that between 3.7 and 5.2% of the study population had an ICD-9 diagnosis of schizophrenia, with the lifetime risk of experiencing at least one episode of non-organic psychosis to be just over 10% (Morgan et al. 2008). These results contrast with a previous Australian study which found a lower point prevalence of 1.3% for the broader diagnosis of ‘psychosis’ (White et al. 2005).

In a study of over 3,000 inmates of British prisons, Hassiotis et al not only found that people with ID were over-represented in the prison system, but also that those with ID were twice as likely as inmates of normal intelligence to have probable psychosis (Hassiotis et al. 2011). Although the time of onset of the psychosis was not recorded, it was speculated that the stressful and complex prison environment might precipitate the development of psychotic illness in those already at risk.

Risk factors

There is no single, clear cause underlying SSDs in people with ID. Work has attempted to identify risk factors that might contribute to the development of SSDs, with the aim of improving the effectiveness of public-health interventions and targeting an 'at risk' group for close monitoring and quick treatment. Owing to its prevalence and societal burden, investigation of risk factors has focused on schizophrenia itself, rather than other types of functional psychosis.

Risk factors for schizophrenia have traditionally been considered as genetic and environmental, but there is increasing evidence in support of interaction of the two in the genesis of the disorder (Van Os et al. 2008). Owing to lack of investigation, not all risk factors have been established in the population with ID but it might be reasonable to assume that exposures that place individuals in the general population at higher risk of developing schizophrenia have similar effects in those with ID. Moreover, the association of certain factors with increased rates of schizophrenia does not prove evidence of causation, and the mechanisms by which risk factors influence the development of such complex psychopathology are generally not well understood. See box 2 for an overview.

BOX 2 – risk factors for schizophrenia

- Genetics – the heritability of schizophrenia in the general population has been estimated at over 80%.A number of gene variants have been implicated in increasing the risk of developing schizophrenia, although individually each genetic variant is associated with only a small effect (Kumar et al. 2014).

- Pregnancy and birth complications– pregnancy and birth complications are more common in people with ID, and may be causative in some instances (Sussmann et al. 2009). Pre- and peri-natal complications have also been associated with the later development of schizophrenia. There is evidence that pregnancy and birth complications are more common in people with ID who develop schizophrenia than in a matched cohort of people with ID who do not (O'Dwyer 1997).

- Urbanicity – there is strong evidence that living in an urban environment increases the chance of developing schizophrenia (Vassos et al. 2012).

- Ethnic minority status – people from an ethnic minority group in the UK who have ID are more likely to develop SSDs than their white counterparts (Tsakanikos et al. 2010).

- Cannabis – use of cannabis increases the incidence of psychosis in the general population, and the response appears to be dose-dependent (Moore et al. 2007). The role of cannabis in the risk of developing SSDs in people with ID has not been thoroughly studied; one study demonstrated that cannabis use is prevalent amongst people with ID presenting to specialist psychiatric services and is particularly associated with those suffering SSDs (Chaplin et al. 2011).

- Negative life events – events such as moving house, death of a relative, and victimisation are common in people with ID and have been associated with the subsequent development of a range of psychiatric problems, including psychotic illness (Hulbert-Williams et al. 2014).

The nature of the link between ID and SSDs

The association between SSDs and intellectual functioning has prompted research in people with ID and SSD with the aim of gaining a broader understanding of the pathophysiology of schizophrenia itself (Moorhead et al. 2009). Such work has challenged the traditional view of schizophrenia as a neurodegenerative condition with onset in early adulthood (indeed, the Kraeplinian term 'dementia praecox' implies a slow progressive decline arising after normal development) and a neurodevelopmental model of schizophrenia is increasingly dominant (Owen et al. 2011; Fatemi & Folsom 2009).

Proponents of a social causative theory of schizophrenia might argue that the increased rates of social and economic disadvantage, exclusion, bullying, and adverse life-events that people with ID suffer cause additional stress which underlies the observation of increased rates of SSDs (and mental illness in general) in this group (Tsakanikos et al. 2007). That is, the effects of a hostile environment are borne out by increased rates of psychopathology in vulnerable individuals. Alternatively, it may be the case that cognitive impairment in itself increases susceptibility to developing an SSD, mediated by 'overload' of an individual's cognitive and adaptive capacity resulting from multiple episodes of only partly-understood stimuli (Doody et al. 1998).

Results from imaging studies, although relatively sparse in people with ID and SSDs, support a neurodevelopmental model, which maintains that SSDs arise as a result of disturbed early development of the nervous system. A number of structural brain abnormalities have been described in individuals with schizophrenia, including enlargement of the ventricles, dilatation of cortical sulci, and a reduction in brain volume with proportionately greater loss in the amygdala and hippocampus, particularly on the left side. Many of the changes in gross brain morphology demonstrated in people with schizophrenia are also seen in people with ID without schizophrenia, however imaging studies have shown the brains of people with co-morbid ID and schizophrenia to show greater similarity to the brains of people of normal intelligence with schizophrenia than to a control group with ID alone (Sanderson et al. 1999). This observation leads to the suggestion that a common pathophysiological process is at work in both comorbid schizophrenia and ID and in schizophrenia alone. Therefore, the association of ID and schizophrenia may be a function of a severe and early-onset form of schizophrenia (of which the global intellectual impairment is part of the natural history of the disease), rather than the pre-existing ID itself acting as a risk factor for the development of schizophrenia (Sanderson et al. 1999; Bonnici et al. 2007). Further advocating a neurodevelopmental theory of schizophrenia is the finding that both functional decline (Fuller et al. 2002) and morphological brain changes are evident at the onset of the disease or may even pre-date the clinical disorder, rather than developing after the disease becomes manifest (Steen et al. 2006). Moreover, people with ID tend to develop schizophrenia at a younger age than those of normal intellectual functioning. Such findings

suggest a biologic underpinning to SSDs, the expression of which may be influenced by later environmental exposures.

Genetic conditions and SSDs

Several genetic conditions that cause intellectual disability are also associated with increased rates of SSDs, although the mechanisms mediating the links have not been defined.

22q11.2 deletion syndrome, also known as velo-cardio-facial or DiGeorge syndrome, is caused by the deletion of a small region of DNA on the long-arm of chromosome 22. It occurs at a population frequency of approximately 1 in 4,000 live births (Botto et al. 2003). Features include facial dysmorphia, cleft palate, structural heart defects and immune disorders in addition to mild-moderate intellectual disability. The syndrome is one of the strongest known risk factors for psychosis – 1% of people with schizophrenia are estimated to have the mutation (Bassett et al. 2010; Horowitz et al. 2005) and up to schizophrenia spectrum disorders develop in up to 41% people with the syndrome (Murphy et al. 1999; Ousley et al. 2013; Pulver et al. 1994; Schneider et al. 2014). A review is provided by Shprintzen (Shprintzen 2008).

Prader-Willi syndrome results from failure of expression of paternally-inherited genes on the long arm of chromosome 15. The characteristic phenotype is of short stature, hyperphagia, morbid obesity, and hypogonadism. The majority have intellectual disability (Dykens et al. 1992). Psychiatric illness is highly prevalent, particularly in the subtype caused by maternal disomy, and usually manifests as an affective psychosis (Boer et al. 2002; Sinnema et al. 2011; Soni et al. 2008).

Usher's syndrome is an autosomal-recessive condition that results from defects in one of several genes. The syndrome involves intellectual disability and varying degrees of deafness, blindness and vestibular dysfunction. Such sensory impairments contribute a further challenge to psychiatric diagnosis. Lifetime incidence of psychosis is increased though the mechanisms underlying this association have yet to be explained (Hess-Röver et al. 1999; Waldeck et al. 2001).

There is evidence that people with ***Down's syndrome*** are less likely to be diagnosed with schizophrenia than people with intellectual disability not due to Down's syndrome (Collacott et al. 1992; Mantry et al. 2008). However, whether this represents a true differential in rates of the illness is not clear.

Differential diagnosis

Several illnesses can present in a similar way to SSDs (see box 3) and must be excluded before diagnosis. A thorough history, physical examination and necessary investigations, such as blood tests and neuroimaging, by trained professionals maybe necessary. BOX 3 – important differentials in the diagnosis of SSDs

Delirium

Delirium is acute cognitive impairment caused by organic illness, such as infection or substance withdrawal. It may be confused with functional psychosis, although there are important differences, including the speed of change in mental state, fluctuating course, and co-incident presentation with physical illness (Gleason 2003).

Autistic spectrum disorder (ASD)

The overlap between core features of autistic spectrum disorders and psychosis, and the challenges this imparts for accurate diagnosis, are considered by Starling and Dossetor and Raja and Azzoni (Starling & Dossetor 2009; Raja & Azzoni 2010). People with ASD have impaired social interactions, commonly talk to themselves, may hold unconventional beliefs or peculiar ideas, have a high likelihood of sensory abnormalities, and often display steretoypies of speech and movement, all of which could be misinterpreted as psychosis. Poor social judgement and theory-of-mind skills may suggest paranoid delusions (Deprey & Ozonoff 2008). However, true delusions and hallucinations are not among the symptoms of ASD. Cochran et al further review the subject and suggest approaches to assessment (Cochran et al. 2013).

Epilepsy

Between 20 and 30% people with ID have epilepsy (Bowley & Kerr 2000). Pre-, post- or inter-ictal changes in behaviour may be mistaken for SSDs. Brief absences may be thought to represent distractibility by unknown stimuli, and hallucinations and abnormal experiences occurring in certain forms of epilepsy may be attributed to mental illness.

Psychotic symptoms may also be seen in affective disorders (as part of a severe depressive or manic episode), post-traumatic stress disorder, and personality disorder.

Difficulties in recognising and diagnosing psychotic disorders in people with intellectual disabilities

Accurate diagnosis is essential to direct the correct treatment and supportive interventions.

There are no laboratory, radiological, or psychometric tests that confirm diagnosis which is therefore based on clinical interview and observation. Diagnosis of SSDs in people with ID can be difficult for several reasons.

- Deficits in communication skills and limitations in verbal ability can make it difficult for people with ID to describe their symptoms. This is especially pertinent in psychotic disorders where symptoms include complex and subjective mental phenomena.

- People with ID may express their thoughts in a muddled or disjointed manner resembling thought disorder, or may seem overly-concerned about the motives of others in a way resembling paranoid thinking, but which could be normal when viewed in the context of past experiences of abuse or victimisation.

- Distinguishing developmentally-appropriate behaviours from psychotic experiences can be difficult. Magical thinking and imaginary friends are two such examples that could be misinterpreted as evidence of delusions, or hallucinations (Pickard & Paschos 2005).

- People with ID may lack insight into their illness and thus not report symptoms, quite before barriers to accessing healthcare are considered.

There is some evidence that people with intellectual disability experience a different pattern of psychotic symptoms to people of average intelligence. In people with mild ID, the symptoms and clinical features of schizophrenia are thought to be broadly the same as the population with normal intelligence (Doody et al. 1998). The content of delusions and hallucinations will be commensurate with an individual's developmental level and people with ID are less likely to hold complex systematised delusional beliefs or to report conceptually-complex symptoms such as passivity delusions (Moss et al. 1996; Meadows et al. 1991). Auditory hallucinations have been shown to be the most reliable and consistently-reported psychotic symptom in those with mild ID (Meadows et al. 1991; Moss et al. 1996).

Bouras et al. (2004) compared people diagnosed with schizophrenia spectrum disorders with and without mild ID. The study found that although the groups did not differ in terms of reported psychopathology, the group with ID showed greater levels of observable psychopathology and more negative symptoms (Bouras et al. 2004). This finding was corroborated by a meta-analysis of studies comparing the presentation of schizophrenia in people with mild ID or borderline intellectual functioning with those with average-high IQ which demonstrated those with lower IQ experience substantially greater negative symptoms (Welch et al. 2011). This contrasts with evidence from people with severe-

profound ID in whom negative symptoms seem to be under-represented (Cherry et al. 2000) although there is, of course, inherent difficulty in diagnosing functional decline in people who may never have attained basic adaptive skills, and some authors have questioned the validity of the construct of negative symptoms when applied to people with ID (Hatton et al. 2005; Melville 2003). In recognition of this, the DC-LD gives less significance to negative symptoms than diagnostic criteria developed for use in the general population.

In the absence of detailed self-report, observable signs of psychiatric illness that might manifest as changes in behaviour and functioning are important. Maladaptive behaviours such as unexplained screaming, aggression, and self-injury, can suggest psychotic illness, particularly in those with greater degrees of impairment. Change in presentation is a particularly important indicator of the development of a psychotic illness. Collateral information is vital to establish a baseline pre-morbid state and family and carers should be engaged at all stages, although reliance on third-party information is not without its own complications (Costello & Bouras 2006).

Use of scales and instruments

Given the challenges in diagnosing SSDs described above, several standardised instruments have been developed to assist the clinician in recognising and evaluating psychopathology (Matson et al. 2012).

The Reiss screen (Reiss 1988) and PAS-ADD Checklist (Prosser et al. 1998) are brief carer- or family-rated instruments that can highlight the need for more detailed investigation.

The Mini PAS-ADD Interview requires further training to administer and is used to collate information on psychiatric symptomatology from an informant (Prosser et al. 1997). It has been shown to have good consistency with expert clinical opinion (Prosser et al. 1998).

The PAS-ADD Clinical Interview is a more comprehensive diagnostic interview suitable for direct use with people with ID and separately with informants. Psychotic symptoms are covered in detail and the outcome is a specific diagnosis aligned to diagnostic criteria specified in the ICD or DSM. The Clinical Interview has been validated in the diagnosis of schizophrenia (Moss et al. 1996).

The DASH-II (diagnostic assessment for the severely handicapped-revised) is an alternative scale for use in people with severe and profound ID, and has been demonstrated to be useful in screening for schizophrenia in this group (Bamburg et al. 2001).

Management

Treatment approaches can be broadly divided into pharmacological and non-pharmacological.

The evidence base for such interventions in people with intellectual disability is generally lacking and management is guided by research in the non-ID population and clinical experience.

Pharmacological treatment

The mainstay of pharmacological management in schizophrenia spectrum disorders is anti-psychotic medication. However, there is a lack of high quality evidence for their use in people with intellectual disability (Duggan & Brylewski 2004) and studies suggest that a significant proportion of prescribed anti-psychotics are used to treat behavioural, rather than psychotic symptoms (De Kuijper et al. 2010).

Atypical anti-psychotics have largely replaced older anti-psychotics as first-line treatment and seem to be better tolerated (Connor & Posever 1998; Advokat et al. 2000). Choice is determined by individual factors and presence of co-morbid conditions, and should be made jointly by practitioners and patients where possible (NICE, 2014). An important side-effect of atypical antipsychotics is the 'metabolic syndrome' comprising obesity, insulin resistance, impaired glucose tolerance, and dyslipidaemia (Newcomer 2007). However, an observational study conducted by Frighi et al found that there were no clinical or statistically significant differences in metabolic indices between people with ID treated with anti-psychotics and those who were anti-psychotic naïve, although there was a trend towards increased rates of type 2 diabetes in the treated group (Frighi et al. 2011) Guidelines recommend regular monitoring of blood glucose, lipids and weight for people taking anti-psychotic medication (American Diabetes Association; et al. 2004), although there are indications that people with ID frequently do not receive such investigations (Devapriam et al. 2009; Teeluckdharry et al. 2013)

Extra-pyramidal side-effects (EPSEs) of anti-psychotic medication comprise drug-induced parkinsonism, akathisia, acute dystonic reactions, and tardive dyskinesia. Such side-effects can be persistent, impair quality of life and may be mistaken for core symptoms of the ID. Although the newer, atypical anti-psychotics have been promoted as having less propensity to cause EPSEs than older drugs, there is evidence that those with ID who take atypical anti-psychotic drugs remain at increased risk of developing abnormal movement disorders (Fodstad et al. 2010). The Matson Evaluation of Drug Side-Effects (MEDS) has been developed as a comprehensive informant-based measure that can be used to assess side-effects of psychotropic medication in people with ID (Matson et al. 1998).

The atypical anti-psychotic clozapine is effective in improving symptoms in treatment-resistant schizophrenia (Kane 1992). It is licensed where two adequate trials of alternative anti-psychotics have failed. Studies suggest that clozapine is safe and efficacious in people with intellectual disability (Antonacci & De Groot 2000; Thalayasingam et al. 2004). Clozapine has also been used to treat aggression in people with ID with some success in small trials (Cohen & Underwood 1994) but a more recent review concluded that "research on the use of clozapine to manage behaviour among individuals with ID is inconclusive at best" (Singh et al. 2010). In addition, the risk of serious side-effects, including potentially fatal agranulocytosis, and necessity of regular blood tests may deter clinicians from using the drug.

Non-pharmacological treatment

Psychosocial interventions are often used in combination with medication and require input from the wider multi-disciplinary team. A broad range of psychosocial interventions has been used, including cognitive behavioural therapy (CBT), psychoeducation programmes, and family-based interventions (Chien et al. 2013). The focus may be on managing or reducing symptoms, psycho-education and improving insight, or maximising functional ability. Delivery of therapies needs to be adapted to reflect a person's developmental level which may involve incorporating flexibility in the location and structure of sessions, and supporting the individual with prompts or accessible written information.

Case reports detailing successful group and individual interventions for people with ID and SSDs have been published (Allott et al. 2013; Hurley 2012; Crowley et al. 2008) but interventions have not been systematically evaluated and robust evidence of their effectiveness and acceptability is lacking

Psychosis co-occurring with intellectual disability confers additional carer burden (Irazábal et al. 2012) and efforts should be made to involve and support family carers (National Institute for Health and Care Excellence 2014). Research in the mainstream population has consistently demonstrated the effectiveness of family therapy in improving several aspects of the condition including frequency of relapse and number of admissions to hospital (Pharoah et al. 2010). Although further research is required, adapted family therapy shows signs of being a promising treatment in people with ID (Marshall & Ferris 2012).

Service provision

The majority of people with intellectual disability and psychosis will be managed in the community and their usual home. People with ID and SSDs are likely to be heavy consumers of psychiatric resources (Spiller et al. 2007). Amongst people with ID suffering mental illness, those with schizophrenia have been shown to use psychiatric services more than those with

any other diagnosis (Morgan et al. 2008). Despite this, there has been little research to inform the most effective models of community care (Balogh et al. 2008; Hemmings 2008). A consultation exercise involving a large multi-disciplinary group identified the "need for a focused approach on the service user and their illness" and "working within the wider context of the service user" as essential components of services managing people with ID and psychosis (Hemmings et al. 2009).

Assertive community treatment (ACT) teams are a feature of many mainstream psychiatric services and work with people with severe and chronic mental illness who engage poorly with services (Stein & Test 1980). Evaluation of such services for people with intellectual disabilities have failed to show evidence of improvement in any outcome measure (Martin et al. 2005; Oliver et al. 2002).

At times of symptom exacerbation or where the risk associated with the illness is too great, facilities for hospital admission must be available. Being diagnosed with a schizophrenia spectrum condition increases the likelihood of having a psychiatric admission over people with other psychiatric diagnoses (Cowley et al. 2005). Moreover, a study conducted in Taiwan found that hospital admissions for people with ID and co-occurring schizophrenia cost more than admissions for people with ID and other mental illnesses (Lai et al. 2011).

Whether psychiatric services for people with ID who develop serious mental illness, including SSDs, are provided within specialist ID teams or by generic mental health services is the focus of an, as yet unanswered debate (Hemmings et al. 2014).

Outcome / prognosis

The 'recovery model' of rehabilitation from serious mental illness has gained currency in the general population. Advocates from within both professional and service-user and carer populations uphold that principles of hope, healing and empowerment should drive the development of positive, person-centred services that work collaboratively with, and are acceptable to, users of those services (Warner 2009). Under this model, recovery is not merely remission from symptoms but implies broader gains in social and functional outcome. Respect for individual's experience and their priorities are at the heart of the model. Although the concept of 'recovery' from SSDs in this sense has sparsely been covered in people with ID, certain aspects of the model such as supported employment, social skills training, and promotion of community inclusion, will be familiar to those working in ID services.

Despite the optimism inherent in the recovery model, SSDs tend to run a chronic course and can impair relationships, functioning and quality of life (Morgan et al. 2008; Jääskeläinen et

al. 2012). A study by Cooper et al reported a full remission rate of just under 15% over a 2 year period (Cooper et al. 2009).

When comparing people with schizophrenia spectrum disorders with and without co-occurring intellectual disability, studies have shown people with borderline intellectual functioning or diagnosed intellectual disability have a more severe illness, greater functional disability and lower quality of life (Bouras et al. 2004; Chaplin et al. 2006).

Conclusion

SSDs are not uncommon in people with ID and confer an additional degree of impairment. The presentation of these disorders in people with ID is often atypical and as such they can be difficult to recognise and might be overlooked. Treatment guidelines are extrapolated from those used in the general population, as research addressing interventions specifically tailored to people with ID is relatively sparse.

Key summary points

- Schizophrenia spectrum disorders are more common in people with ID than the general population.
- The nature of the link between SSDs and ID is not clear and the development of pathology is likely to be multifactorial. There is increasing evidence in support of aberrant early development of the nervous system in predisposing individuals to developing SSDs.
- The clinical manifestations of SSDs are diverse and are likely to be different to those in the general population. The disorders can be difficult to recognise in people with ID, leading to under-diagnosis and treatment, and continuing distress and disturbance.
- Treatment methods are extrapolated from research in the general population, but there are issues unique to people with ID that must also be considered.

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