

# Behavioural and Emotional Disorders with Onset Usually Occurring in Childhood and Adolescence

*Shoumitro (Shoumi) Deb and Meera Roy*

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### Learning Objectives

- Conduct disorder (CD) in people with intellectual disabilities
  - Oppositional defiant disorder (ODD) in people with intellectual disabilities
  - Intermittent explosive disorder (IED) in people with intellectual disabilities
  - Stereotyped movement disorders in people with intellectual disabilities
  - Other behavioural disorders with onset in childhood and adolescence
  - Assessment of CD, ODD, and IED in people with intellectual disabilities
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- Disruptive, impulse-control, and conduct disorders is a new category under DSM-5 classification system.
  - These disorders originate in childhood and often continue into adulthood.
  - There is a strong association between these disorders and intellectual disabilities (ID).
  - They include conduct disorder, oppositional defiant disorder which is a less severe form of conduct disorder, and intermittent explosive disorder, which was part of impulse control disorder in the previous DSM classifications.
  - By definition many of these disorders have to show an intention behind the behaviour, which is not easy to determine, particularly among those who have severe and profound ID.
  - Other impulse control disorders include pyromania, kleptomania, gambling disorder, compulsive sexual behaviour disorder, trichotillomania, and excoriation disorder.
  - Body-focussed repetitive behaviour, on the other hand, includes stereotyped movement disorder.
  - The aetiology of these disorders is complex and often is the outcome of an interaction between internal factors within the person and external factors such as the environment.
  - Therefore, the management of these disorders requires input from a multitude of professionals with relevant and

specific skills to provide a person-centred assessment and management.

- The ultimate aim should be to improve the quality of life of the person with ID and their family members.

#### Tip

- The DSM-5 criteria are often difficult to apply to people with intellectual disabilities for conduct disorder, intermittent explosive disorder, and oppositional defiant disorder.
- Therefore, instead of making a psychiatric diagnosis as per DSM-5 criteria, it is more important to describe the behaviour which will lead to a better management.

## 19.1 Disruptive, Impulse-Control, and Conduct Disorders

### 19.1.1 Introduction

Disruptive, impulse-control, and conduct disorders (CD) are characterised by impairment in the ability to control emotions and behaviours to an extent that the resulting behaviours have a clinically significant impact on an individual and her or his interactions with others and/or society. The six specified disruptive, impulse-control, and CD are (a) antisocial personality disorder, (b) CD, (c) oppositional defiant disorder (ODD), (d) intermittent explosive disorder (IED), (e) kleptomania, and (f) pyromania. In a way ODD is considered as the less severe form of CD. In order to meet DSM-5 [1] diagnostic criteria for disruptive, impulse-control, and CD, there should be evidence that an individual present with behaviours that violate the rights of others (e.g. aggression, destruction of property) and/or bring the individual into significant conflict with societal norms or authority figures.

In the previous DSM classification such as the DSM-IV-TR [2], CD and ODD were included in the attention deficit hyperactivity

disorder (ADHD) and disruptive behaviour disorder category. In the previous version of DSM, 'Impulse-Control Disorders' included IED, kleptomania, and pyromania. These are now classified within the new DSM-5 classification as disruptive, impulse-control, and CD. Also included in this category are CD, ODD (both previously classed as Disorders Usually First Diagnosed in Infancy, Childhood, or Adolescence) and antisocial personality disorder (see ► Chap. 31).

Previously, trichotillomania and pathological gambling were also included in Impulse-Control Disorders Not Elsewhere Classified. However, in the DSM-5, these disorders are now included within obsessive-compulsive and related disorders and substance-related and addictive disorders respectively. Habit disorders such as finger sucking, body rocking, and bruxism are classed as stereotypic movement disorders and are included among motor disorders under the 'neurodevelopmental disorders' section of DSM-5 [1]. Antisocial personality disorder is included in DSM-5 [1] both under the 'Disruptive, Impulse-control, and CD' category as well as under the 'Personality Disorder' category.

Diagnoses of other specified or unspecified disruptive, impulse-control, and CD may be given when an individual has symptoms which are characteristic of the specific disorders listed above but do not meet the full diagnostic criteria. These diagnoses may be used for individuals with presentations in which there is insufficient information to make a more specific diagnosis.

### 19.1.2 Issues in the Use of Diagnosis in Intellectual Disabilities

There are a number of difficulties when using these terms in relation to people with intellectual disability (ID). These diagnostic categories are hardly used in day-to-day practice, instead a broader term such as problem (challenging) behaviour is used more widely both in clinical practice and in research. The term 'problem behaviour' is used to describe 'socially unacceptable behaviour that causes

distress, harm or disadvantage to the persons themselves or to other people, and usually requires some intervention' [3] and includes behaviours such as verbal or physical aggression to others, physical aggression to property and/or self, screaming, shouting, indiscriminate wandering, objectionable personal habits including smearing of faeces and pica, etc., antisocial behaviour including taking others' possession without permission, etc., unacceptable sexual behaviours, and stereotyped movement not caused by a neurological disorder.

DSM-5 [1] reports that these disorders usually develop in childhood or adolescence. This criterion applies equally to people with ID. However, for individuals with ID there may be a higher likelihood that symptoms remain unrecognised or are misinterpreted until adulthood because of the difficulties encountered when assessing and diagnosing psychiatric and behavioural disorders in this group [4, 5], particularly among children and adolescents. For individuals with ID, before making a diagnosis one has to consider whether the behaviour is both age and developmentally appropriate as in many people with ID, developmental age does not correspond to their chronological age.

One of the reasons that diagnoses such as CD and ODD are rarely used in ID literature is that these disorders assume that the person is behaving with an intent, which is difficult to determine in most people with ID who exhibit problem behaviour. Another problem is with the DSM-5 criterion that stipulates that a diagnosis of IED cannot be made before the age 6 years. As individuals with severe and profound ID are likely to have a mental age below 6 years, this diagnosis cannot be applied to this group of people [6].

Among the general population who do not have ID, disruptive, impulse-control, and CD are known to commonly occur with each other, substance use disorders, and ADHD. This might as well be the case for people with ID. Because of these difficulties with diagnostic categories Deb and colleagues [6] suggested that a diagnosis based on a group of behaviours may be better for an appropriate formulation of management than a syndromic categorisation.

The DSM-5 [1] acknowledges that many of the behaviours associated with disruptive, impulse-control, and CD are present in the general population and that when formulating a diagnosis ‘the frequency, persistence, pervasiveness across situations, and impairment associated with the behaviors...[should] be considered relative to what is normative for a person’s age, gender, and culture’. For individuals with ID, attention should also be paid to what is considered normative relative to an individual’s developmental level.

### 19.1.3 Assessment of Behaviour

As a diagnosis of disruptive, impulse-control, and CD cannot be made in most people with ID, particularly for those who have severe and profound ID, Deb and colleagues in DM-ID-2 [6] recommended that in practice, clinicians should make a diagnosis of ‘problem (challenging) behaviour’ outside the DSM-5 [1] framework and describe the frequency and severity of these behaviours along with severity ratings (mild, moderate, severe) and frequency ratings (low, medium, high). They recommend a multidisciplinary, person-centred assessment of the behaviours using a schema based on the assessment of the Behaviour, the Person showing the behaviour, Psychological and psychiatric factors, Medical and organic factors, and Social factors (BMPPS) (see ► Chap. 7 and ► <https://spectrom.wixsite.com/project>).

Assessment of behaviour should include the person’s history of problem behaviour; baseline behaviour prior to the onset of the current behaviour; how the behaviour started, whether they appeared gradually over time or relatively abruptly, perhaps precipitated by an acute event; the frequency, severity, and duration of the behaviour; the nature, content, and context of the behaviour, as some behaviour may occur in certain circumstances/settings but not in others, associated behaviours; and the impact of the behaviour on the person’s life, others’ lives, and the environment. The behaviour may lead to reduced quality of life for the individual and her/his caregiver. The

behaviour may lead to reduced access to services including education, day service, and employment opportunity, and may lead to a threatened or actual loss of placement in a residential setting or day placement. The behaviour may lead to reduced social activities including leisure activities, access to friends, etc.

In severe cases the individual may end up being physically restrained, inappropriately medicated, or taken to a hospital or police station. All these scenarios may subsequently have their impact on the individuals, their behaviour, and their caregivers. For example, some of these outcomes may work as perpetuating factors for the ongoing behaviour. These outcomes may be seen as inappropriate or excessive and the individuals or their caregivers may perceive them as punishments.

A risk assessment should be carried out which should include (a) risk to others, (b) risk to the individual, (c) risk to the environment, and (d) other risks. Where possible use a standardised method of risk assessment but always make a record of the assessment and review them periodically.

#### 19.1.3.1 Assessment of Medical and Organic Factors Should Include

- Chronic physical conditions such as headache, toothache, and pain in other parts of the body.
- Medical conditions such as acid reflux, chest infections that are common in people with ID.
- Epilepsy which is common in people with ID [7].
- Neurological conditions such as cerebral palsy.
- Genetic conditions such as Prader-Willi syndrome, Fragile X syndrome, and Lesch-Nyhan syndrome which are known to be associated with problem behaviour [10].
- Hearing and visual impairments which can lead to problem behaviour form frustration.
- Sensory processing issues in people with ASD.

- Communication problems where the individual may use problem behaviour to communicate or a discrepancy between expression and comprehension.
- Physical disabilities causing frustration.
- Illicit drug and alcohol use, although this may be more of an issue in people with borderline intelligence.
- Adverse effects of prescribed medicines.

### 19.1.3.2 Assessment of Psychological and Psychiatric Factors

Psychiatric disorders such as mood disorders, schizophrenia, other psychoses, and anxiety-related disorders may lead to problem behaviour in people with ID [4]. ASD and ADHD are also common comorbidities resulting in higher rates of problem behaviour. It is important to investigate for these conditions as the management of the problem behaviour will depend on the management of the underlying condition.

Psychological and emotional factors such as bereavement and trauma and abuse (physical, emotional, or sexual), historical, ongoing, recurring or new, or stress and relationship difficulties may lead to loss of self-esteem and isolation, causing and perpetuating problem behaviour.

### 19.1.3.3 Assessment of Social Factors

Environmental factors have an important role to play as some people with ID may present with problem behaviour if they find themselves in an environment which they find too demanding or understimulating. Personalities and attitudes of others towards the person, including care givers can also affect the person with ID. Needless to say, management styles could impact on the individual too, for example confrontational attitude.

## 19.1.4 Conduct Disorder

The essential feature of Conduct Disorder (CD) is a repetitive and persistent pattern of behaviour that violates the basic rights of others

or that breaks major age-appropriate societal norms or rules. Behaviours are grouped into four major categories: (a) aggression to people or animals, (b) destruction of property, (c) deceitfulness or theft, and (d) serious violation of rules. As with all psychiatric conditions, the presentation must cause clinically significant impairment in social, academic, or occupational functioning.

These disorders could be of (a) childhood onset where the behaviour is present before the age of 10 years or (b) adolescent onset where no symptoms are observed before the age of 10 years. Unspecified onset category applies to those for whom there is insufficient information to determine the age of onset of symptoms.

Following are the DSM-5 [1] criteria for diagnosis of CD, none of which could be applied to those who have severe and profound ID as it is not possible to determine the underlying intention behind the behaviour. All three criteria (A, B, C) must be met in order to qualify for the diagnosis.

### ■ DSM-5 Diagnostic Criteria A [1]

Repetitive and persistent pattern of behaviour in which the basic rights of others or major age-appropriate societal norms or rules are violated, as manifested by the presence of three or more of the following criteria in the past 12 months, with at least one criterion present in the past 6 months. The clinician should assess age appropriateness of the behaviour and ascertain that the individual understands societal rules and norms.

### 19.1.4.1 Aggression to People and Animals

- Often bullies, threatens, or intimidates others.
- Often initiates physical fights.
- Has used a weapon that can cause serious physical harm to others (e.g. bat, brick, broken bottle, knife, gun).
- Has been physically cruel to people.
- Has been physically cruel to animals.

In case of people with ID, it is important to ascertain that the individual displaying the

behaviour has understood the significance and impact of her or his actions.

#### 19.1.4.2 Destruction of Property

- Has deliberately engaged in fire setting with the intention of causing serious damage.
- Has deliberately destroyed others' property (other than by fire setting).

In case of people with ID, clinician should assess intent behind the behaviour. Even some people with mild ID may have difficulties with communicating their thoughts and feelings. The intent is even more difficult to determine in people with severe ID.

#### 19.1.4.3 Deceitfulness or Theft

- Has broken into someone else's house, building, or car.
- Often lies to obtain goods or favours or to avoid obligations (i.e. 'cons' others).
- Has stolen items of nontrivial value without confronting a victim (e.g. shoplifting, but without breaking and entering; forgery).

In case of people with ID, it is important to establish that the individual understands the concept of ownership.

#### 19.1.4.4 Serious Violations of Rules

- Often stays out at night despite parental prohibitions, beginning before age 13 years.
- Has run away from home overnight at least twice while living in parental or parental surrogate home (or once without returning for a lengthy period).
- Is often truant from school, beginning before age 13 years.

In case of ID, it is important to carefully examine the home environment and the reasons why the individual is running away. For example, if there is evidence of inadequate care, neglect, or abuse, the relevance of this

behaviour to the diagnosis should be questioned. Careful examination of school environment is necessary to rule out any reasonable cause such as distress due to bullying.

#### ■ DSM-5 Diagnostic Criterion B [1]

The disturbance in behaviour causes clinically significant impairment in social, academic, or occupational functioning.

#### ■ DSM-5 Diagnostic Criterion C [1]

If the individual is 18 years or older, criteria are not met for antisocial personality disorder.

#### ■ Severity:

*Mild:* few if any conduct problems in excess of those required to make the diagnosis *and* conduct problems cause only minor harm to others.

*Moderate:* number of conduct problems and effect on others intermediate between 'mild' and 'severe'.

*Severe:* many conduct problems in excess of those required to make the diagnosis *or* conduct problems cause considerable harm to others.

It cannot be assumed that the person with ID understands the basic rights of others and appropriate social norms. The four categories of behaviours to be rated in CD cannot be viewed as equivalent in the case of people with ID. There are examples of people with ID showing aggression towards people or animals and destroying property, but there are far fewer instances of deceitfulness or serious violation of rules. The difference is in the nature of the intent. Aggression and destruction can occur when the person is frustrated and does not have any other available outlet to express their frustration. Serious violation of rules could also occur in the absence of intent, either because the person does not understand the rules or because the person does not appreciate the consequences of their actions. Deceit, however, is by definition an intended act.

Often people with ID function at a level much lower than their chronological age, which makes it difficult to determine the age appropriateness of the behaviour. When the function



of the behaviour (what the behaviour is trying to achieve) is understood, making a diagnosis of CD becomes difficult, even for those functioning at borderline levels of intelligence.

Clinicians should focus on the interaction between social environment and the individual and not assume that the responsibility lies with the individual for which it is necessary to use multiple sources of information during assessment. People with ID themselves can be essential sources of information.

Assessment should focus on obtaining the most direct information regarding current behaviours. Information on previous problem behaviour if any should be collected to determine onset, predisposing, precipitating, and perpetuating factors for the individual's behaviour. Frequency and severity of each current and past behaviours should be assessed [11] along with repetitiveness and chronicity to indicate the onset and stability over time, and developmental appropriateness.

The determination of age appropriateness depends on an understanding of both normal developmental patterns and the behavioural patterns presented by the person. For example, one might be reluctant to assign the label 'stealing' to a person for whom it is unclear whether she or he understands the concept of personal ownership.

Assessment of environment plays an important part in determining the diagnosis of CD. Behaviours that are inconsistent across settings suggest environmental influence on behaviour. The behaviours may be generated by environmental factors which would weigh against a diagnosis of CD. Importance of making assessments across multiple settings is that multiple informants are likely to be used, thus somewhat mitigating reporting bias.

### 19.1.5 Oppositional Defiant Disorder

The essential feature of Oppositional Defiant Disorder (ODD) is a recurrent pattern of angry, irritable mood with defiant, disobedient, and hostile vindictive behaviour towards at least one individual who is not a sibling.

The behaviour must occur more frequently than would occur typically in individuals of comparable age and development and has to cause significant impairment in social, academic, or occupational functioning. All three DSM-5 [1] criteria (A, B, C) must be met in order to qualify for the diagnosis.

The DSM-5 [1] criteria could not be applied for those who have severe and profound ID and who lack communication skills but could be applied to those who have mild to moderate ID and could communicate with some caveats (see next section).

#### ■ DSM-5 Diagnostic Criteria A [1]

Pattern of angry/irritable mood, argumentative/defiant behaviour, or vindictiveness lasting at least 6 months, during which four or more of the following are present:

1. Often loses temper
2. Often touchy or easily annoyed
3. Often angry and resentful
4. Often argues with authority figures or, for children and adolescents, with adults
5. Often actively defies or refuses to comply with requests from authority figures or with rules
6. Often deliberately annoys others
7. Often blames others for his or her mistakes or misbehaviour
8. Spiteful or vindictive at least twice within the past 6 months

To apply this diagnosis to people with ID, the clinician must rule out any other reasons for the behaviour (see ► Chap. 7). The clinician also needs to carry out a language and cognitive assessment to rule out comprehension and specific cognitive difficulties that might otherwise explain the noncompliance and other associated behaviour. Assessment of both intent and developmental age is also required. Consider a criterion met only if the behaviour occurs more frequently than is typically observed in individuals of comparable age and developmental level.

#### ■ DSM-5 Diagnostic Criterion B [1]

The disturbance in behaviour is associated with distress in the individual or others in the



immediate social context. Further, it causes clinically significant impairment in social, academic, or occupational functioning.

#### ■ **DSM-5 Diagnostic Criterion C [1]**

The behaviours do not occur exclusively during the course of a psychotic disorder, mood disorder or substance use.

Rates of problem behaviour which may or may not include CD and ODD vary widely between 2% and 60% [4, 12, 13] depending on the diagnostic criteria used and the type of population studied. The rates may also vary depending on factors like age, gender, etc. (see ► Chap. 7) [4, 14, 15]. Impairments in impulse control may be associated with aggressive behaviour in people with ID and also borderline intellectual functioning [15, 16].

Given that the locus of control in their lives is often outside their control, people with ID are more likely to feel frustrated and for this to manifest itself as insufficient self-regulation and defiance. For this reason, the clinicians have to be mindful of the danger of possible overdiagnosis of ODD in people with ID.

People with severe ID may not have the necessary language and cognitive abilities to be able to meet some of the criteria, such as argue with others, blame others, or be vindictive. It is thus incumbent on clinicians to conduct a comprehensive assessment of all relevant factors, including physical and mental health status, quality of the environment, possible sources of stress, and possible abuse and come to an accurate formulation which may point away from such a diagnosis. A diagnosis of ODD often lends the impression that the responsibility for the problem lies mainly within the individual themselves, rather than an interaction between the individual and the social environment or with other systemic factors [6].

A thorough assessment is necessary addressing the following issues in order to make the correct diagnosis and formulate an appropriate management plan.

- The nature of the behaviours must be explored in detail, including the circumstances in which they occur, including the predisposing, precipitating, and perpetuating factors.

- Assess the amount of control the individual seems to have over these behaviours.
- Make a list of all current and previous behaviours being displayed.
- Clarify the referral question.
- Determine who are affected by the behaviour, where and when do they occur?
- Define them in each area of the individual's life.
- Determine whether these behaviours are developmentally appropriate or not?
- Obtain a careful biopsychosocial history, to chart onset, course, and contributing risk factors.
- Assess the strengths and weaknesses of the individual that may have an impact on the behaviour.
- Assess the individual's cognitive, communication, occupational, and social function.
- Exclude comorbidities such as anxiety disorder, ASD, ADHD, PTSD, history of abuse that may have a bearing on the behaviour.
- How has the individual's behaviour affected the social environment and vice versa?
- What systematic factors contribute to or maintain the disruptive behaviour?
- What has the individual been referred for and by whom?
- Evaluate the reliability of the referral and informant sources.
- Gather information from as many sources as possible including the individual with ID and from direct observation of behaviour in different settings where possible.
- Consider the fact that transient oppositional behaviour may be common among preschool children and adolescents.

Emerson and Hatton [13] reported ODD among 13.3% of youth with ID compared with 2.3% in typically developing youths. In contrast Christensen and colleagues [17] reported among 5–9-year-old children, a point prevalence of ODD between 21% and 29% among the typically developing children compared with 35–44% among those who had borderline intellectual functioning and 39–48% among children who had ID. They noticed comorbidities with other disorders,

particularly ADHD, among children with ID. However, they did not find any inter-group differences in the rate of ODD according to gender, age of onset, or stability of the behaviour.

### 19.1.6 Intermittent Explosive Disorder

Intermittent Explosive Disorder (IED) is characterised by recurrent impulsive aggressive outbursts that are grossly out of proportion to any provocation or psychosocial stressor. Explosive episodes may be frequent and less severe (non-injurious) and less frequent but involving severe aggressive behaviour, such as destruction of property or physical assault with injury against an animal or a person. An important qualifier is that the aggressive outbursts are impulsive in nature and not premeditated or instrumental.

The recurrent aggressive outbursts cause marked distress or impairment in occupational or interpersonal functioning or are associated with social, financial, or legal consequences. The aggression in IED is characteristically more intense than what is observed in ADHD, CD, ODD, and antisocial and borderline personality disorders. IED should not be diagnosed in children younger than 6 years of age or in children aged 6–18 years if aggression occurs in the context of an adjustment disorder. This criterion makes this diagnosis invalid for people who have severe and profound ID, whose developmental age is likely to be lower than 6 years. This diagnosis may also be difficult to apply to those with less severe degree of ID (see notes in the next section).

IED should not be diagnosed in people with disruptive mood dysregulation disorder (DMDD), or whose impulsive aggressive outbursts are attributable to another medical condition or to the physiological effects of a substance or another mental disorder. All three DSM-5 [1] criteria (A, B, C) must be met in order to qualify for the diagnosis.

#### ■ DSM-5 Diagnostic Criteria A [1]

Pattern of angry/irritable mood, argumentative/defiant behaviour, or vindictiveness lasting at least 6 months, during which four or more of the following are present:

1. Often loses temper
2. Often touchy or easily annoyed
3. Often angry and resentful
4. Often argues with authority figures or, for children and adolescents, with adults
5. Often actively defies or refuses to comply with requests from authority figures or with rules
6. Often deliberately annoys others
7. Often blames others for his or her mistakes or misbehaviour
8. Spiteful or vindictive at least twice within the past 6 months

To apply this diagnosis to people with ID, the clinician must rule out any other reasons for the behaviour (see ► Chap. 7). The clinician also needs to carry out a language and cognitive assessment to rule out comprehension and specific cognitive difficulties that might otherwise explain the noncompliance and other associated behaviour. Assessment of both intent and developmental age is required. Consider a criterion met only if the behaviour occurs more frequently than is typically observed in individuals of comparable age and developmental level.

#### ■ DSM-5 Diagnostic Criterion B [1]

The disturbance in behaviour is associated with distress in the individual or others in the immediate social context. Further it causes clinically significant impairment in social, academic, or occupational functioning.

#### ■ DSM-5 Diagnostic Criterion C [1]

The behaviours do not occur exclusively during the course of a psychotic disorder, mood disorder, or substance use.

The onset of recurrent, problematic impulsive aggressive behaviour is most common in late childhood or adolescence [18]. The core

features of IED typically are persistent and continue for many years. The course of the disorder may be episodic, with recurring periods of impulsive aggressive outbursts. Coccaro and colleagues [19] reported a lifetime prevalence rate of 11% and a 1-month prevalence of 2.3% in a community sample of adults. These prevalence rates depend on the definition used for IED.

Certain factors make people with ID vulnerable to IED. For example, they may find it difficult to control their impulse compared with those who possess better psychological defence mechanism and stress management skills to deal with their inner frustration and anxiety. It may be difficult to be sure of the instrumental nature of behaviour in people with significant intellectual and communication impairment because the underlying intention could not be assumed. Other factors such as comorbid seizure disorders, developmental brain abnormalities, and environmental factors may all contribute to the behaviour (see ► Chap. 7). Some highlighted a role of life events as possible contributing factor to IED [20] among adults with ID, which may suggest a connection between explosive aggression and hyperarousal provoked by specific conditioned fear experiences in these individuals.

A multidisciplinary assessment is needed to confirm a diagnosis of IED in an individual with ID. The multidisciplinary team may include a psychiatrist, a psychologist or behaviour specialist, a speech and language therapist, a nurse, an occupational therapist, a social worker, and a neurologist where necessary. Caregivers including family members and other close informants such as teachers should also contribute to the discussion. Individual with ID should also take part in the discussion where necessary and appropriate.

A thorough physical and mental state examination is necessary to exclude any medical and psychiatric cause of aggressive behaviour. A neurologist may assist in the diagnosis and management of epilepsy and other neurological disorders which may be contributing to the aggressive behaviour [7–9] (see ► Chap. 7). The psychologist or behaviour specialist/

nurse may help in gathering data on the nature, frequency, target, and impact (damage or injury) of aggression and carrying out a functional assessment of the behaviour. Finally, any history of abuse or trauma should be discussed in the context of identifying stimuli that may trigger an aggressive fear response.

Biological including genetic, developmental, psychosocial including environmental and family factors should be carefully assessed. The following conditions could be coexistent with or should be differentiated from IED.

- Disruptive mood dysregulation disorders (DMDD)
- Antisocial personality disorder or borderline personality disorder
- Delirium
- Major neurocognitive disorders
- Aggressive behaviour due to another medical condition
- Substance intoxication or substance withdrawal
- ADHD
- ODD
- CD
- ASD

### 19.1.7 Disruptive Mood Dysregulation Disorders

In DSM-5 [1] a new diagnostic category called ‘Disruptive Mood Dysregulation Disorders (DMDD)’ has been created under the overall ‘Depressive disorder’ category. This was done to categorise youths who display severe irritability and aggression chronically. DMDD is defined primarily by two features: (a) frequent severe temper outburst, and (b) persistent irritability evident every day for most of the day. Unlike the previous diagnosis of ‘severe mood dysregulation’, DMDD excludes hyperarousal (e.g. insomnia, agitation, distractibility, racing thoughts) from the essential criteria. Although theoretically DMDD is distinguishable from CD and ODD on the basis of severity, it is not so easy in practice to apply this distinction.

This causes confusion among the DMDD diagnosis and CD and ODD diagnoses. Therefore, some suggested that DMDD should be used as a specifier of ODD rather than a separate diagnosis [21]. Lochman and colleagues [21] also highlighted the danger of potential misdiagnosis of severe chronic irritability as a bipolar disorder which may lead to inappropriate use of medication.

### 19.1.8 Pyromania

A diagnosis of pyromania is made when a person repeatedly sets fires deliberately to relieve arousal or obtain pleasure from or relief of unpleasant tension upon watching the fire. The act is repetitive, deliberate, and for the purpose of relief of tension. This is not done for any other purpose or caused by any other mental disorder.

Only a small proportion of people with ID who set fires can be diagnosed with pyromania since most cases of fire setting occur for reasons that would exclude the diagnosis. The presence of ID particularly when severe and profound is likely to cause difficulty in assessing motivation and the presence of tension and relief of tension. In the case of severe or profound ID such assessments would be very difficult, if not impossible to carry out. Similarly, the observer of this act would have difficulty assessing whether there is a motive behind the act. DSM-5 [1] criteria also exclude a diagnosis of pyromania in the presence of impaired judgement which may be the case for many people with ID. Therefore, we do not think that this diagnosis could be applied to those who have severe and profound ID and who are unable to communicate their thoughts, feelings, and emotions.

#### ■ DSM-5 Diagnostic Criteria [1]

- A. Deliberate and purposeful fire setting on more than one occasion.
- B. Tension or affective arousal before the act.
- C. Fascination with, interest in, curiosity about, or attraction to fire and its situa-

tional contexts (e.g. paraphernalia, uses, consequences).

- D. Pleasure, gratification, or relief when setting fires or when witnessing or participating in their aftermath.
- E. The fire setting is not done for monetary gain, as an expression of sociopolitical ideology, to conceal criminal activity, to express anger or vengeance, to improve one's living circumstances, in response to a delusion or hallucination, or as a result of impaired judgement (e.g. in major neurocognitive disorders, intellectual disabilities, substance intoxication).
- F. The fire setting is not better explained by CD, a manic episode, or anti-social personality disorder.

This diagnosis has to be distinguished from abnormal rituals and interests that may be seen in a person with ASD. The clinician has to consider the difficulty in ascertaining the exact inner feelings of a person with ID.

Whereas the acts of fire setting and arson are committed by people with ID, it is not clear how many of these acts will qualify for a diagnosis of pyromania which is likely to be made of a very small subgroup of fire-setters [22–27]. Published reports identify individuals as fire setters or arsonists in a forensic setting but not in the community. It is not clear whether fire-setting is more or less prevalent among people with ID than in the general population who do not have ID. However, Jacobson [28] reported an association between the rate of fire-settings and an increasing severity of ID. Taylor and colleagues [29, 30] showed that group intervention could be effective for women with ID who set fires.

### 19.1.9 Kleptomania

The diagnosis requires repetitive stealing not due to the need for personal use, monetary value, anger, vengeance, or in response to delusions, hallucinations, or other mental disorders. The stealing is done to alleviate tension. It may be difficult to determine

whether these conditions are met in mild and moderate ID and it is not possible to make this diagnosis in people with severe and profound ID.

The diagnosis of kleptomania requires ascertainment of motives and the presence of tension and gratification upon stealing. Some people with ID will have difficulty describing these measures due to poor verbal skills, lack of understanding of what these measures mean, and even, inability to understand the concept of ownership and personal property.

■ **A.DSM-5 Diagnostic Criteria [1]**

- B. Recurrent failure to resist impulses to steal objects that are not needed for personal use or for their monetary value.
- C. Increasing sense of tension immediately before committing the theft.
- D. Pleasure, gratification, or relief at the time of committing the theft.
- E. The stealing is not committed to express anger or vengeance and is not in response to a delusion or a hallucination.
- F. The stealing is not better explained by CD, a manic episode, or antisocial personality disorder.

In case of people with ID, it is important to assess that the individual understands the sense of ownership and normal societal rules regarding possession. The difficulty of assessing inner feelings of a person with ID should be considered.

No information is available on the development and course of this disorder. The prevalence is unknown, although stealing without meeting the criteria for kleptomania is not unusual among people with ID. This may be seen as part of mania, CD, or antisocial personality disorder. Kleptomania would severely interfere with social adaptation. No research literature concerning kleptomania in people with ID could be found except a case report in the context of naltrexone treatment. Jacobson [28] reported an increased rate of property theft associated with increased severity of ID.

### 19.1.10 Trichotillomania

Also known as ‘hair pulling disorder’, involves recurrent, irresistible urge to pull hair from scalp, eyebrows, eyelids, and other areas of the body, despite repeated attempts to stop or decrease hair pulling. In DSM-5 [1], this is classified under Obsessive Compulsive and Related Disorders.

■ **DSM-5 Diagnostic Criteria [1]**

- A. Recurrent pulling out of one’s hair, resulting in hair loss.
- B. Repeated attempts to decrease or stop hair pulling.
- C. The hair pulling causes clinically significant distress or impairment in social, occupational, or other important area of functioning.
- D. The hair pulling or hair loss is not attributable to another medical condition (e.g. a dermatological condition).
- E. The hair pulling is not better explained by the symptoms of another mental disorder (e.g. attempts to improve a perceived defect or flaw in appearance in body dysmorphic disorder).

Hair pulling can happen in people with ID, as part of a syndrome such as Down syndrome, Prader-Willi syndrome as part of the behavioural phenotype or in people with autism and sensory issues. While they would meet Criterion A, the rest of the criteria would not be met. Pulling out hair and consuming it is a cause of bowel obstruction in people with ID [31].

### 19.1.11 Excoriation (Skin-Picking) Disorder

■ **DSM-5 Diagnostic Criteria [1]**

- A. Recurrent skin picking resulting in skin lesions.
- B. Repeated attempts to decrease or stop skin picking.
- C. The skin picking causes clinically significant distress or impairment in social,



- occupational, or other important areas of functioning.
- D. The skin picking is not attributable to the physiological effects of a substance (e.g. cocaine) or another medical condition (e.g. scabies).
  - E. The skin picking is not better explained by symptoms of another medical condition (e.g. tactile hallucinations or delusions in a psychotic disorder, attempts to improve a perceived defect or flaw in appearance in body dysmorphic syndrome, stereotypies in stereotypic movement disorder, or intention to harm oneself in non-suicidal self-injury).

As with hair pulling, skin picking is not uncommon, particularly in people with ID and autism and people with behavioural phenotypes such as Smith Magenis syndrome.

### 19.1.12 Gambling Disorder

#### ■ DSM-5 Criteria

- A. Persistent and recurring problematic gambling behaviour leading to clinically significant impairment or distress as indicated by individual exhibiting four or more of the following in a 12-month period:
  1. Needs to gamble with increasing amounts of money in order to achieve the desired excitement.
  2. Is restless or irritable when attempting to cut down or stop gambling.
  3. Has made unsuccessful efforts to control, cut back, or stop gambling.
  4. Is often preoccupied with gambling (e.g. having persistent thoughts of reliving past gambling experiences, handicapping or planning the next venture, thinking of ways to get money with which to gamble).
  5. Often gambles when feeling distressed (e.g. helpless, guilty, anxious, depressed).
  6. After losing money gambling, often returns another day to get even (chasing one's losses)

7. Lies to conceal the extent of involvement with gambling.
  8. Has jeopardised or lost a significant relationship, job or educational or career opportunity because of gambling.
  9. Relies on others to provide money to relieve desperate financial situations caused by gambling.
- B. The gambling behaviour is not better explained by a manic episode.

Gambling disorder can be episodic or persistent and can be mild (4–5 criteria met), moderate [7, 10], or severe [11, 12].

The prevalence of gambling disorder in people with ID is not known. People with more severe degrees of ID will be supervised and may not have access to finances to gamble. It may be fair to say that there is a complex interplay of factors in people with ID. Tromans and colleagues [32] describe the association of temporal lobe epilepsy, ASD, and gambling disorder in a man with mild ID.

## 19.2 Secondary Impulse Control Disorders

### 19.2.1 Hoarding Disorder

Hoarding disorder affects both men and women. Epidemiological studies report significantly higher prevalence among men which contrasts clinical samples where women predominate. Community surveys estimate point prevalence of clinically significant hoarding to be approximately 2–6% [1]. Hoarding symptoms are three times more common in older adults, aged 55–94 years compared with younger adults.

Hoarding begins early in life, starts interfering with everyday functioning by mid-twenties and significant impairment by mid-thirties. Individuals have persistent difficulties discarding or parting with possessions, regardless of their actual value. The main reasons given are the perceived utility or the aes-



thetic value of the item or the strong sentimental attachment to the possessions. Commonly saved items include newspapers, magazines, old clothing, books, bags, mail, and paperwork, but virtually any item can be saved. Individuals experience distress when faced with the prospect of discarding them which emphasises the intentional aspect of hoarding. They accumulate large number of items which fill up and clutter living areas so that they are no longer habitable so much so that the individual may not be able to sleep on their bed or cook in their kitchen. Third parties such as family members, local authorities may have to intervene to remove clutter. Symptoms such as discarding items must cause clinically significant distress. People with hoarding disorder may show features of indecisiveness, perfectionism, avoidance, procrastination, difficulty in planning and organisation, and distractibility. Half of these people have a relative with the condition.

#### ■ DSM-5 Diagnostic Criteria [1]

- A. Persistent difficulty discarding or parting with possessions, regardless of their actual value.
- B. This difficulty is due to a perceived need to save the items and to distress associated with discarding them.
- C. The difficulty discarding possessions results in the accumulation of possessions that congest and clutter active living areas and subsequently compromises their intended use. If living areas are uncluttered, it is because of the interventions of third parties (e.g. family members, cleaners, authorities).
- D. The hoarding causes clinically significant distress or impairment in social, occupational, or other important areas of functioning (including maintaining a safe environment for self and others).
- E. The hoarding is not attributable to another medical condition (e.g. brain injury, cerebrovascular disease, Prader-Willi syndrome).
- F. The hoarding is not better explained by the symptoms of another mental disorder (e.g. obsessions in obsessive compulsive disorder, decreased energy in major depressive disorder, delusions in schizo-

phrenia or another psychotic disorder, cognitive deficits in major neurocognitive disorder, restricted interests in ASD).

Specify if: *with excessive acquisition*: if difficulty discarding possession is accompanied by excessive acquisition of items that are not needed or for which there is no available space.

#### ■ Specify if:

*with good or fair insight*: The individual recognises that hoarding related beliefs and behaviours (pertaining to difficulty discarding items, clutter, or excessive acquisition) are problematic.

*with poor insight*: The individual is mostly convinced that hoarding-related beliefs and behaviours (pertaining to difficulty discarding items, clutter, or excessive acquisition) are not problematic despite evidence to the contrary.

*with absent insight/delusional beliefs*: The individual is completely convinced that hoarding related beliefs and behaviours (pertaining to difficulty discarding items, clutter, or excessive acquisition) are not problematic despite evidence to the contrary.

Although ID itself is an exclusion in the diagnosis of hoarding disorder, this behaviour occurs in people with ID. Testa and colleagues [33] found that among 61 children with ID, 16.4% exhibited hoarding as a major clinical issue. When the children with ID and hoarding were compared with age-, sex-, and IQ-matched ID participants without hoarding, hoarders exhibited a slower learning curve on word list-learning task.

Diogenes syndrome is characterised by marked self-neglect, domestic squalor, social withdrawal, and hoarding of rubbish and is named after Greek philosopher Diogenes, known for his pursuit of self-sufficiency, freedom from material possessions, and disregard for social niceties and creature comforts. Williams and colleagues [34] describe two women with mild ID who fit the criteria for Diogenes syndrome and discuss the possible differential diagnoses. Hoarding was a significant feature in both of them. One of them made a significant improvement when she returned to the care of her mother and the hoarding stopped completely but the situation continued with the second one.

Hoarding is part of the behavioural phenotype of Prader-Willi syndrome. Dykens and colleagues [35] examined the nature, severity, and correlates of non-food obsessions and compulsions in 91 people with Prader-Willi syndrome and found that 47% showed hoarding behaviour. Participants hoarded a variety of objects such as old batteries, trash, papers, pens, and toiletries. Respondents often noted that some objects were collected and stored in a specific manner, for example books or toys in a series had to be purchased and then arranged in sequential order. Many care givers noted the need for subjects to sort and arrange toys, clothing, or papers according to specific rules based on size, shape, colour, or simply until they were 'just right'. As repetitive behaviours occur in 86% of people with Williams syndrome, Royston and colleagues [36] examined the profile of behaviours, comparing them with those with Prader-Willi and Down syndromes. They administered the Repetitive Behaviour Questionnaire to care givers of adults with Williams ( $n = 96$ ), Prader-Willi ( $n = 103$ ), and Down ( $n = 78$ ) syndromes, respectively. There were few group differences, although participants with Williams syndrome were more likely to show body stereotypies. Individuals with Williams syndrome also showed more hoarding and less tidying behaviours than those with Down syndrome.

Lane and colleagues [37] suggest that hoarding can be complicated for individuals with developmental disabilities living in congregate settings because it can interfere with staff and peer relationships and affect potential community placement. Berry and Schell [38] evaluated the effects of individualised reinforcement and item return procedures on hoarding behaviour in a multiple baseline across three persons with severe ID. Systematic preference assessment procedures identified items used in the individualised reinforcement procedures. There was a reduction in hoarding behaviour for each person when individualised treatment and where item return procedures were applied, and these reductions were maintained when direct support staff were trained to provide treatment. Kellet and colleagues [39] provided 12 cognitive behav-

iour therapy (CBT) sessions to 14 people with mild ID on domiciliary visits at their residential facilities in the community and followed them up 6 months later. They found no evidence of relapse or adverse events and suggest from this open label trial that CBT may be a safe intervention for people with ID who exhibit hoarding behaviour.

### 19.2.2 Compulsive Sexual Behaviour Disorders

Compulsive sexual behaviour disorder has been proposed for inclusion as an impulse control disorder in the ICD-11 [40]. It is characterised by a persistent pattern of failure to control intense, repetitive sexual impulses or urges, resulting in repetitive sexual behaviour over an extended period (e.g. six months or more) that causes marked distress or impairment in personal, family, social, educational, occupational, or other important areas of functioning [41].

The pattern is manifested in one or more of the following:

- (a) Engaging in repetitive sexual activities has become a central focus of the person's life to the point of neglecting health and personal care or other interests, activities, and responsibilities.
- (b) The person has made numerous unsuccessful efforts to control or significantly reduce repetitive sexual behaviour.
- (c) The person continues to engage in repetitive sexual behaviour despite adverse consequences (e.g. repeated relationship disruption, occupational consequences, negative impact on health).
- (d) The person continues to engage in repetitive sexual behaviour even when she or he derives little or no satisfaction from it [41].

Compulsive sexual disorder or sexual addiction is not recognised in the latest DSM-5 [1] classification system. It was previously included in the DSM-III-TR [42] as a descriptor under the general diagnosis of 'Sexual Disorder Not Otherwise Specified'.

Pang and Masiran [43] described a 22-year-old man referred from the genitourinary clinic where he had been treated for gonococcal urethritis. He experienced high libido from the age of 19 years, admitted to having sexual intercourse with more than one hundred different commercial sex workers in the last three years, with increase in frequency to gain satisfaction, having to widen his search of area to find sex workers, having to spend huge amounts of money on this activity which he financed by working as a shop assistant and by borrowing from relatives. There were withdrawal symptoms from abstinence and the sexual activity was carried out to the exclusion of all other activities. He has been physically abused by his father as a child. His academic functioning at school was poor, having failed examinations. He could not handle money or carry out age appropriate calculations. He is reported as meeting the criteria for compulsive sexual behaviour disorder in the context of mild ID and was treated by addiction services. Psychological work reduced his craving significantly.

Singh and Coffey [44] reported a case of a young man with mild ID, autism, OCD, and relapse of bipolar affective disorder who had developed ulcers on his penis from excessive masturbation. While an inpatient, he was treated with lithium and as his mood stabilised, so did the sexual behaviour. Nishimura and colleagues [45] also described a young woman with ID who started masturbating in public, with no other features of a mood disorder who did not respond to antipsychotics but improved with lithium treatment. Lamy and colleagues [46] describe a woman with ID who had herself been experienced sexual and physical abuse. She then perpetrated sexual abuse on her children.

Thom and colleagues [47] in their review suggested that the prevalence of inappropriate sexual behaviour in people with ID can be as high as 15–33%, but the nature tends to be inappropriate than violative. It may be associated with ID syndromes. For example, Fisher and colleagues [48] found a complex association between Klinefelter syndrome and hypersexuality, paraphilic behaviours, and gender dysphoria, which were mediated by obsessive

compulsive and autistic traits. Börjeson-Forssman-Lehmann syndrome (BFLs) is an X-linked inherited disorder characterised by unusual facial features, abnormal fat distribution, and ID. Behaviour in persons with BFLs is in general friendly but can be challenging with externalising and thrill-seeking features but their social skills are good. de Winter and colleagues [49] described a group of four BFLs patients with inappropriate sexual behaviour as part of their behavioural phenotype.

Finally, as Sajith and colleague [50] point out, men with ID sometimes exhibit inappropriate sexual behaviours, which may be termed challenging behaviours or offending behaviours depending on the circumstances and whether criminal justice system is involved. Hawk and colleagues [51] have shown that the rate of sex offence charges was nearly twice as high among defendants with ID as among defendants without ID.

In conclusion, compulsive sexual behaviours can happen in people with ID for various reasons; as part of affective disorder, behavioural phenotype, due to environmental reasons, or as an entity in itself. However, it is important to explore the knowledge of the person with ID of sexual function, sexual relationship, and social norms before assuming that any socially inappropriate sexual acts on their part including sex offences are intentional.

## 19.3 Body-Focussed Repetitive Behaviour Disorder

### 19.3.1 Stereotypic Movement Disorder

#### ■ DSM-5 Diagnostic Criteria [1]

- A. Repetitive, seemingly driven, and apparently purposeless motor behaviour (e.g. hand shaking or waving, body rocking, head banging, self-biting, hitting own body).
- B. The repetitive motor behaviour interferes with social, academic, or other activities and may result in self-injury.

- C. Onset is in the early developmental period.
- D. The repetitive motor behaviour is not attributable to the physiological effects of a substance or neurological condition and is not better explained by another neurodevelopmental or mental disorder (e.g. trichotillomania, OCD).

■ **Specify if:**

With self-injurious behaviour or behaviours that would result in an injury if preventive measures were not used.

Without self-injurious behaviour.

■ **Specify if:**

Associated with a known medical or genetic condition, neurodevelopmental disorder, or environmental factor (e.g. Lesch-Nyhan syndrome, ID, intrauterine alcohol exposure).

Mild, if symptoms can be suppressed by sensory stimulus or distraction; moderate, if explicit protective measures and behaviour modifications are required; and severe if continuous monitoring and protective measures are required to prevent self-injury.

Essential features of stereotypic movement disorder are that they are repetitive, seemingly driven, and apparently purposeless [1]. They are often rhythmical movements of head, hand, or body without obvious adaptive function and may not respond to effort to stop them. These may be seen in typically developing children, but they can be stopped when attention is drawn to them or when the child is distracted. The child may also demonstrate self-restraining behaviour, that is sit on their hands.

In typically developing infants, there is a predictable sequence of rhythmic motor development, which appears stereotypic and declines by 24 months of age [52]. This stereotypic, rhythmic motor behaviour may appear at later ages in children with Down syndrome and those with significant motor impairment [53, 54]. Moreover, children with significant developmental disabilities engage in relatively high rates of stereotypes as compared with typically developing infants and toddlers [54, 55]. Hoch and colleagues [56] compared two groups of young children, one with develop-

mental delay and one without, and asked parents to complete standardised measures on the presence of stereotypes and self-injury. They found that stereotyped behaviour was reported significantly more frequently by parents of children with developmental delay than by parents of typically developing children (85.7% vs. 42.8%). Over half of the children with delay (53%) were described as having at least one stereotype that was considered a moderate or severe problem in comparison with only 2% of the children without developmental delay on a standardised measure of repetitive behaviour.

Stereotypic behaviour is variable as each individual presents with her or his own signature behaviour. Examples of non-self-injurious stereotyped behaviour include body rocking, hands flapping, finger flicking, and head nodding. Examples of self-injurious stereotypic behaviour can be repeated head banging, face slapping, eye poking, and biting parts of the body. Frequency of occurrence of these behaviours can vary and be related to stress, excitement, boredom, fatigue, and multitude of factors specific to the person. In both humans and nonhuman primates, aberrant stereotyped behaviour including self-injury has been associated with adverse environmental circumstances and early CNS damage [57].

Stereotypic behaviours occur in a variety of neurodevelopmental conditions, particularly in people with ID and ASD. Bodfish and colleagues [58] found a prevalence of stereotypy of 60.9% and self-injury 46.6%; and compulsion when they surveyed people with severe and profound ID. Rojahn and colleagues [59] found 54% of 432 randomly selected residents from a developmental centre had stereotyped behaviour and 43%, self-injury. The determination of the functional profile of a targeted behaviour has important implications for the design of customised behavioural interventions. Medeiros and colleagues [60] investigated the relationship between the level of ID and the functional profile of aggression, stereotypy, and self-injurious behaviour. They found that individuals with mild ID tended to use self-injurious

behaviour less often for tangible rewards or to escape social demands than individuals with severe ID who tended to use them for these same purposes significantly more often. This could be helpful in devising interventions, together with language and communication training which may help people with severe ID express themselves about when they wanted a break from social demands. Research also showed that aggressive and stereotypic behaviour problems can function, regardless of the severity of ID, for the purposes of attention, sensory stimulation, pain reduction, social escape, and tangible reinforcement. A functional assessment is, therefore, an essential part of the assessment process (see Assessment section in the ► Chap. 7).

Despite their core significance in ASD, it is not clear whether there are distinct groups of stereotypic behaviours with different specificity to autism. Carcani-Rathwell and colleagues [61] found that subjects with a Pervasive Developmental Disorder (PDD) diagnosis were more likely to have repetitive stereotyped behaviours than those with only ID. Within the PDD group, those with ID had higher rates of all repetitive stereotyped behaviours. The study suggests that there may be two distinct sub-groups of repetitive behaviours whereby the sensory and motor repetitive behaviours are generally associated with lower developmental age and less specific to the autistic syndrome whereas the ‘higher-order’ behaviours, that is cognitive rigidity symptoms, may be a more ‘autism specific’ feature. Bodfish and colleagues [62] compared adults with autism with adults with ID and found that both groups had significant patterns of repetitive behaviour co-occurrence. Autistic subjects had significantly greater severity ratings for compulsions, stereotypy, and self-injury. Repetitive behaviour severity also predicted severity of autism. Pedersen and colleagues [63] found that restricted interests or repetitive behaviours significantly differentiated between children with autism and ID.

Behavioural features of children with severe ID remain difficult to disentangle from those of children with autism. Polyak and colleagues [64] suggest that current estimates of autism prevalence fail to take into account the

impact of comorbidity of ID and epilepsy on autism diagnosis.

Self-injury also co-occurs in a number of syndromes associated with ID including Lesch-Nyhan, Smith Magenis, Rett, Cornelia de-Lange, Prader-Willi, and fragile X syndrome [10]. Disruption to the activity dependent neuroprotector homeobox (ADNP) gene is among the most common heterozygous gene mutation seen associated with ASD. Affected individuals have a constellation of features including autism, ID, dysmorphic features, and hypotonia. Arnett and colleagues [65] compared individuals with ADNP syndrome with those with autism due to CHD8 mutation and idiopathic autism and found that ADNP syndrome group’s profile of social communication and restricted and repetitive behaviours deficits to be notably different from those of other autism subtypes. The youths with ADNP syndrome had relatively mild social communication deficits (despite impaired verbal intelligence and low expressive language abilities) coupled with stereotyped motor restricted repetitive behaviours. The social communication deficits were mild enough as to not to warrant a diagnosis of DSM-5 [1] ASD in about 30% of cases, although all of them had a diagnosis of ID. The aetiology of social communication and repetitive stereotyped behaviours in autism is not well understood. Arnett and colleagues [65] suggest that genetic subtypes of ASD may each be associated with a unique aetiology of autism trait covariance. Within the idiopathic group, it could be polygenic. However, within single gene disorder, individual differences in the location and effect of the genetic variant could have vastly different effects on protein functioning. Yet, these disparate events frequently produce a similar behavioural, physical, and/or medical phenotype, implying convergence to a common neurobiological endophenotype.

Haw and colleagues [66] carried out a study on the movement disorders seen in a cohort of 145 people with Down syndrome living both inpatient and community facilities in Surrey, UK. They found that over a third of these people experienced stereotypies and no association was found between stereotypies



and age, hospital or community placement, exposure to neuroleptics, behavioural disorder, or dementia.

For treatment of all the conditions described in ► Chaps. 11 and 12.

### Key Points

- The DSM-5 criteria are often difficult to apply to people with intellectual disabilities for conduct disorder, intermittent explosive disorder, and oppositional defiant disorder.
- Therefore, instead of making a psychiatric diagnosis as per DSM-5 criteria, it is more important to describe the behaviour for a better formulation leading to a better intervention.
- Both pharmacological and non-pharmacological interventions are used but the evidence in their support is poor.
- It is, therefore, important to make a very thorough multidisciplinary assessment of these behaviours in order to achieve a successful treatment outcome.
- The ultimate goal for the intervention is to improve the quality of life of people with intellectual disabilities and their care givers.

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