The Diagnosis of Emotional Dysregulation

In People with Developmental Disabilities

3/13/2011

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That they cannot express their subjective feelings, like lack of pleasure, or express their internalized emotions.

According to Michael Rutter's classic population study on the Isle of Wight, 50% of children with mental retardation had a psychiatric disorder (Rutter, Tizard et al. 1976).

Emotional dysregulation (ED) is a term used in the mental health community to refer to an emotional response that is poorly understood, and does not fall within the conventionally accepted range of emotive response. ED may also be referred to as labile mood or mood swings.

Emotional dysregulation can lead to behavioral problems and can interfere with a person's social interactions and relationships at a person's home, school or place of work.

In our line of work, people with developmental disorders (DDs) or intellectual deficiency (ID) manifest ED in accordance to their capacity to express themselves: ranging from raw emotion (in severe DDs) also called challenging behavior, to depression as we know and diagnose it among people with above borderline intellectual quotient.

Challenging Behavior

Challenging Behavior (CB) is the most frequent reason for psychiatric referral and residential placement of individuals with DDs.

A hallmark of CB is self-injurious behavior. This behavior is commonly seen in individuals with DDs, ranging from genetic and metabolic disorders such as Lesch-Nyhan, Rett, and Cornelia de Lange syndromes, to more frequent etiologies, including fragile X, traumatic brain injury, and severe hypoxic ischemic injury. But there are others encountered in the same diagnostic/etiologic categories: behavior that is aggressive, destructive, impulsive, tantrum-like, verbally or physically abusive, and sexually explicit or inappropriate.

Clearly, CBs are multifactorial, and, if treatment is to be effective, the approach to their diagnosis is multidisciplinary. The DSM-IVR conditions that are most closely aligned with these behaviors are conduct disorder and antisocial personality disorder; however, they can also be key aspects of obsessive-compulsive disorder, attention-deficit-hyperactivity disorder, autism spectrum disorders, or bipolar disorder and major depression.

Clinical features of CB

CBs are reported to reach a peak during adolescence and early adulthood, tapering off by age 35 years (Day 1985), (Oliver, Murphy et al. 1987; Borthwick-Duffy 1994). Males tend to exhibit more aggressive behaviors: self-biting, head punching/slapping and head-to-object banging, females, self-cutting (Oliver, Murphy et al. 1987).
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The intensity of the behaviors seems to correlate inversely with IQ (Borthwick-Duffy 1994). Repetitive self-injury has been shown to be the cause of institutionalization because it is a source of major stress to families and caregivers (Bromley and Emerson 1995; Emerson and Bromley 1995)

Behavioral Characteristics

In fragile X and fetal alcohol syndrome have poor self-regulation and impulse control, manifested as decreased attention, hyperactivity, stereotypies and disruptive behavior both at home and at school. (Roebuck, Mattson et al. 1999) In Rett syndrome, it is classic “hand wringing”, while in Lesch-Nyhan there is hand and lip biting. Aggression is found in a mixed population of individuals with cognitive impairment. There can be destruction of property and assault with the head, teeth, fingers, hands, and legs (Harris 1993)

Prevalence studies

There is great variability in studies of prevalence of CBs. This is due to difficulties in types of studies (self-observation versus more rigorous surveys) as well as the populations sampled. There are two large studies in existence, one in England (rates of 7 %) and one in California (rates of 14 %) respectively. When more inclusive criteria are added the rates go up to up to 67 %. This means to say that most individuals exhibit more than one type of challenging behavior, and the bias came from their need for higher level of assistance in daily living inclusive of attendance to psychiatric clinics, hence the zealousness in reporting cases (Emerson, Kiernan et al. 2001).

Etiology and Pathophysiology

The likelihood of a complex interplay between genetic factors, endogenous neurotransmitters, physical discomfort, psychological frustrations, conditioned responses and disruption in social order.

In animal studies, there are consistent findings of structural lesions in the anterior hypothalamus, leading to offensive aggressive behavior. In functional neuroimaging studies there is selective disruption of D1 pathways, with the resultant hypersensitivity in the residual D1 receptors, which may have a role in SIBs. In primate studies, there is the suggestion that a deficit in serotonin activity correlates with impulsive and aggressive behaviors, and that antidepressants, which manipulate the 5HT receptor and the serotonin transporter may have an effect in the levels of aggression (Ferrari, Palanza et al. 2005).

More reversible and common causes for SIBs, with potentially reversible causes are medical illnesses that cause pain, like migraine headaches, sinus infection, or gastrointestinal reflux. Poor sleep has also been found to be associated with increased irritability and aggressive behavior.

Some cases with obsessive-compulsive disorder have self-injurious behavior that may respond to SSIRs.
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In clinical depression

Tantrum-like behavior and aggression are also likely to be used as a means of communication in people with language deficits. There are times when operant conditioning and reinforcement of responses form caregivers may serve to habituate and reinforce these behaviors.

Finally, the social and familial disruption encountered in the homes and families of people with DD an IDs are risk factors (home instability, inadequate housing, poverty) are among the correlates in CBs and conduct disorders (Rutter 1985)

Diagnosis of challenging behaviors

The diagnosis of CBs entails a multidisciplinary approach. In the case of a psychiatric evaluation there is the need to identify possible symptoms of depression (here a thorough background history is necessary, yet the frequent unavailability of reliable informants is a hindrance), mania, anxiety, psychosis, or obsessional thoughts need to be investigated.

A symptom-based scale such as the Developmental Behavioral Checklist (Dekker, Nunn et al. 2002) may be necessary. The DBC is a suite of instruments for the assessment of behavioural and emotional problems of children, adolescents and adults with developmental and intellectual disabilities. The Developmental Behaviour Checklist, (DBC), (Einfeld & Tonge, 1992, 1995; 2002) is a questionnaire which is completed by parents or other primary carers or teachers, reporting problems over a six month period. Each behavioural description is scored on a 0, 1, 2 rating where 0 = ‘not true as far as you know’, 1 = ‘somewhat or sometimes true’, and 2 = ‘very true or often true’.

(Einfeld & Tonge, 1992, 1995; 2002) is a questionnaire which is completed by parents or other primary carers or teachers, reporting problems over a six month period. The DBC shares the structure of the Child Behaviour Checklist (Achenbach & Edelbrock, 1983), that is, each behavioural description is scored on a 0, 1, 2 rating where 0 = ‘not true as far as you know’, 1 = ‘somewhat or sometimes true’, and 2 = ‘very true or often true’. The items are completely independently derived from a study of the medical files of 7000 intellectually handicapped children and adolescent seen in a developmental assessment clinic.

Treatment of CBs

In targeting CBs the first line interventions are behavioral.

Pharmacological treatments are used to augment the behavioral intervention. In this sense, modification of the environment (to avoid overstimulation, unpredictability and crowding, and teaching skills to replace maladaptive behaviors with more productive means of communication such as sign language and use of picture communication strategies. The plan must be reassessed as necessary in an already “fluid system”, the challenging patient.

The best approach when using pharmacological agents is to “start low and go slow”. What I usually do is to identify the target symptom and using a symptom rating scale to follow the specific response of the intervention. Then there is the ongoing monitoring of side effects, such as excessive sleepiness, lethargy,
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extrapyramidal symptoms, as well as laboratory assessments of electrolytes, blood sugar levels, liver function tests, and prolactin levels.

The side effect profile of these medications includes tardive dyskinesia, sedation, weight gain and seizures in the case of pre-existing seizure disorder.

**Depression in People with Intellectual Disabilities**

**Epidemiological Issues**

People with all levels of ID have been described as suffering from mood disorders. There are, however, very few systematic, prospective, well-controlled studies with reliable means of assessing the presence of these mood disorders.

The current classification of mental diseases (DSM-IV-TR *Diagnostic and Statistical manual of Mental Disorders*, American Psychiatric Association, 1994; 2000) includes two types of depressive disorders:

a) Major depressive disorders
b) Dysthymic disorders

Major depressive disorder is characterized by one or more major depressive episodes (i.e.: at least two weeks of depressed mood or loss of interest). During the same two-week period, the following 5 or more symptoms of depression must be present, for the diagnosis of MD to be made:

- depressed mood most of the day
- markedly diminished interest or pleasure in all activities
- considerable weight gain or weight loss
- insomnia or hypersomnia
- psychomotor agitation or retardation
- feelings of worthlessness
- diminished ability to think or concentrate
- recurrent thoughts of death

The prevalence of major depressive episodes in the **general population** is estimated to be between 10 and 25% for women, and 5 to 12% for men. Both sexes have been found to have 8% when it comes to dysthymic disorder.
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There is a general consensus however, that in persons with ID, these DSM-IV TR diagnostic criteria apply to people with higher levels of cognitive functioning. For people with severe and profound ID there is the problem of their limited ability to communicate or only non-verbal communication: the effect is that they are unable to express subjective feelings such as lack of pleasure, or "internalized material" (Einfeld and Tonge 1999).

Another factor is the great variation in existence in the levels of social cognitive and social skills among people within the same level of functioning, as well as the different cognitive levels (mild to profound). Therefore there has been concern that depressive disorders are not properly recognized in this population, therefore, under-diagnosed, even though studies by King (1994), Gitta (1995) have emphasized the high incidence of these disorders among people with ID.

Another methodological issue found when studying mental health problems in general among adults with IDs is that both conditions (ID and mental illness) can confound the detection of either. There are many people with serious mental health problems who perform poorly in IQ tests and may be misclassified as having ID. The opposite may happen, with low functioning ID patients whose low IQ masks the effect of mental illness, thus missing the effect of the ID. Finally there is the overemphasis of symptoms of mental illness mistakenly assumed to be due to the ID rather than to mental illness (Smiley 2005). In subpopulations, like Down syndrome (Cooper and Collacott 1994) or pervasive developmental disorders single case studies (Ghaziuddin and Tsai 1991), (Stavrakaki, Antochi et al. 2004) exist.

Mood symptoms (depression) have been identified in more recent studies (Clarke and Gomez 1999; Tsiouris, Cohen et al. 2003). There are several recent studies of depression in adults with ID, and they are probably underestimated numbers. Point prevalence rates of depressive disorders in adults with ID point to 4 % (Meins 1995), (Deb, Thomas et al. 2001).

On the other hand, a biased rate (when studying adults with ID attending a psychiatric out-patient clinic may have inflated numbers, such as 26 or 9.1 %, of a total sample of 285 found to have depressive or dysthymic disorder (Stavrakaki 1997).

Etiology

The causes of depressive disorders in people with ID are similar to the causes of depressive disorders in the general population.

There is no clarity as yet whether depressive disorders are due to biological or psychosocial influences. (Lunsky, Bradley et al. 2006). In subpopulations, such as Down syndrome, there are genetic and biological factors (Tu and Zellweger 1965) (Ghaziuddin and Tsai 1991). Genetic disorders with mood disorders are also linked with fragile-X syndrome, Down syndrome, fetal alcohol syndrome and Williams syndrome (Einfeld, Tonge et al. 2001).

The relationship between stressful life events and depressive disorders is particularly important for people with ID, whose lives are characterized by frequent major adverse life events, such as separation form caregivers, and other traumatic events (Stavrakaki 1997). Major depression was also associated
That they cannot express their subjective feelings, like lack of pleasure, or express their internalized emotions with sexual assault, physical assault, parental loss and parental separation. Important risk factors include lack of social support, daily troubles and negative social interactions (Lunsky and Benson 2001; Lunsky 2003)

Symptom Presentation

It appears that depressive symptoms in adults with ID vary according to the degree of their disability the higher their intellectual ability, the closer their symptoms of depression are to those of the general population. In people with severe disability, depression seems to be presented with atypical symptoms or “behavioral equivalents” : atypical symptoms such as irritability, psychomotor agitation, increased behavioral problems and loss of adaptive behavior are found amongst patients identified as having severe ID (Meins 1995).

One classification bases the existence of symptoms (common or distinct) on the degree of disability a patient has (Marston, Perry et al. 1997).

**Common symptoms**, (based on degree of disability) were:

1. Depressed affect
2. Sleep disturbance

The **distinct symptoms**, (based on the degree of disability) were:

**For mild ID**

1. Tearfulness
2. Diurnal mood variation
3. Loss of energy
4. Loss of interest
5. Low self-esteem
6. For moderate ID
7. Social Isolation
8. Self-injurious behavior
9. Weight loss

**For severe/ profound ID**

10. Screaming
11. Aggression
12. Self-injurious behavior

In order to develop a clear impression of what are the **real prevalence rates**, one must have strict and well-developed criteria for the diagnosis of depressive disorders in people with ID, as without these, there may be a potential under-representation of the problem. Therefore, one must cover issues such as:
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- Defining what constitutes ID
- Defining what constitutes Mental Health problems
- A method of identification and assessment of a case
- Use of appropriate diagnostic criteria
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References


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