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# Mood Disorders in Individuals with Genetic Syndromes and Intellectual Disability

Maria Cristina Triguero Veloz Teixeira,  
Maria Luiza Guedes de Mesquita, Marcos Vinícius de Araújo,  
Laís Pereira Khoury and Luiz Renato Rodrigues Carreiro  
*Developmental Disorders Program, Center for Health and Biological Sciences,  
Mackenzie Presbyterian University, São Paulo  
Brazil*

## 1. Introduction

### 1.1 Mood disorders

Mental disorder is a behavioral pattern that causes psychological suffering or incapacity (American Psychiatric Association [APA], 2003). This concept also underlines that whatever the cause of the problem, a mental disorder is the manifestation of a behavioral, psychological or biological dysfunction. The manual offers an organizational plan that groups mental disorders in 16 diagnostic categories. These categories are described according to common traits that allow the specialist to identify the disorder or make differential diagnosis when necessary (APA, 2003). DSM follows the International Statistical Classification of Diseases and Related Health Problems (ICD-10) developed by the World Health Organization (World Health Organization [WHO], 2000).

There are no doubts about the advantages of DSM IV over its prior versions. Criteria have become clearer and more precise and, at the same time, they allow the consideration of much additional information that goes further than the diagnosis *per se*. However, many challenges to the improvement of these criteria have still been discussed in work groups in order to create a new edition in 2013 (Joyce, 2008; APA, 2010).

Mood disorders constitute one of the 16 diagnostic categories. They are a kind of health problem whose main trait is a mood alteration that can follow two distinct directions: a) depressive mood, b) expansive mood. The disorders characterized by depressive mood are: Major Depressive Disorder, Dysthymic Disorder and Depressive Disorder Not Otherwise Specified. Expansive mood comprises Bipolar Disorders Type I and Type II, Cyclothymic Disorder and Bipolar Disorder Not Otherwise Specified (APA, 2003).

The main clinical characteristics of Depressive Disorders are sadness, tearfulness, hopelessness, irritability, lack of interests, diminished appetite, sleep disorders, psychomotor agitation or retardation, diminished energy, tiredness, guilty feelings, attention and concentration problems, memory problems, thoughts of death, suicidal ideation, and/or suicide attempts. Bipolar Disorders also present a persistent, elevated and

abnormal mood alteration, but in two opposite directions, i.e. expansive or irritable and depression. Criteria for a Manic Episode include report inflated self-esteem, euphoria and elevated enthusiasm, grandiosity feelings, diminished need for sleep, flight of ideas, distractibility, psychomotor agitation, excessive involvement in pleasurable activities with high risk potential, pressured speech, among others (APA, 2003). The specificities of typically depressive, manic or mixed frames require an attentive and careful clinical interview to determine characteristics of the disorder, its course and, when necessary, a differential diagnosis.

Mood Disorders due to a General Medical Condition are more likely to present a physical condition of acute beginning. In this case, clinical traits also involve depressive mood, prominently diminished interest or pleasure, and expansive, irritable or elevated mood. These characteristics allow the specialist to classify the disorder according to the main symptomatic frame: a) depressive traits; b) depressive type episode; c) maniac traits, c) mixed traits. The manual specifies that in every mood disorder related to a medical condition it must be verified the presence of different clinical manifestations of depressive, maniac or mixed disorders. Nevertheless, these manifestations do not fulfill all criteria for the disorders and there must be a confirmation of an etiologically related general medical condition.

## 1.2 Intellectual disability

In this chapter, we chose to use the term intellectual disability (ID) as a synonym for mental retardation that has been classically described by DSM IV as a Disorder Usually First Diagnosed in Infancy, Childhood, or Adolescence. The manual describes MR as a disorder marked by significantly subaverage intellectual functioning and an Intellectual Quotient (IQ) of approximately 70 or below, with onset before the age of 18. It can be codified as Mild, Moderate, Severe, Profound Mental Retardation, and Mental Retardation Severity Unspecified (APA, 2003). MR's main characteristic is subaverage intellectual functioning (usually standard deviation 2 below average) and significant impairment in adaptive functioning. Such impairments can be observed through significant limitations in three domains of daily life: conceptual, social and practical abilities. The 11<sup>th</sup> edition of American Association Intellectual and Developmental Disability (2011) define that intellectual disability is characterized by significant limitations both in intellectual functioning and adaptive behavior as expressed in conceptual, social, and practical skills. This disability originates before age 18 (American Association on Intellectual and Developmental Disabilities, 2011).

Genetic syndromes are considered biological risk factors for ID that occurs in pre-natal phase of development. Mood disorders and ID are both complex conditions and involve an interdisciplinary professional work, especially when such conditions are associated. Thus, when such conditions are to be assessed, two diagnostic approaches are needed: an etiological, to identify possible genetic cause for ID, and a functional, to determine the amount of adaptive resource related to environmental needs (Moescheler, 2008).

## 2. Behavioral phenotype and psychiatric phenotype

The development of molecular biology techniques has allowed progress in the study of etiological factors related to genetic evidences in psychiatric disorders. In this context, two

mental disorders stand out: schizophrenia and bipolar disorder (Basset et al., 2003; Craddock et al., 2010; Pun et al., 2011). Nevertheless, to establish a possible causal relation it is necessary to deepen the study of several issues. For instance, why is it that not all individuals with psychiatric disorders present genetic alterations? Why is it that not everyone who has genetic alterations develops psychiatric disorders? (Basset et al., 2003; Leyfer et al., 2006; Mota et al., 2010; Walker et al., 2011). The advances of molecular biology are enormous, however many questions are difficult to be answered. For example, in 1987 mood disorders were studied in several genetic researches that aimed finding the linkage between different genes and bipolar disorder (Egeland et al., 1987). This study was classic in the search for markers in chromosome 11 in a genetically isolated community in Old Order Amish, Pennsylvania. Notwithstanding, this finding could not be confirmed by further research (Kelsoe et al., 1989).

In individuals with genetic syndromes researchers have found the presence of indicators of psychiatric disorders, especially those associated with ID (Antonell et al., 2006; Bilder et al., 2009; Jarvinen-Pasley et al., 2008; Ridha, 2011). Some of the most studied psychiatric symptoms belong to the clinical frames of psychosis, mood disorders and anxiety disorders (Siegel & Smith, 2010). This has determined an operational definition that helps the recognition of psychiatric alterations according to the type of genetic syndrome. This definition is known as Psychiatric Phenotype (PP). It must be emphasized that PP is different from the concept of Behavioral Phenotype (BP). This difference will be treated in the following paragraphs.

Historically, the first description of behavioral characteristics in genetic syndromes was made by Langdon Down in 1866 in his observations of a group of people that have typical mongoloids that shared physical, mental and behavioral characteristics (Down, 1995). After Langdon Down's original description (1866) the syndrome's typical personality and behavioral traits have become known (Collacott et al., 1998). Almost 100 years have passed until a new study appeared and cast suspicion on the existence of genetically based behavior patterns when, in 1963, Money described the visuospatial difficulties in Turner Syndrome (Artigas-Pallarés, 2002). Despite the introduction of the genotype-phenotype relation, the concept of behavioral phenotype associated with genetic disorders was firstly proposed only in 1976 (Nyhan, 1976). The researcher proposed a relation between the presence of self-injury behavior and the syndrome later known as Lesch-Nyhan (Holland et al., 2003). The children affected by this syndrome present a typical behavioral pattern characterized by stereotypies and self-injury behaviors (Nyhan, 1976). The concept of BP proposed by Nyhan was kept and is still used (Artigas-Pallarés et al., 2006). However, during the 1970's and 1980's few researches showed interest in the study this concept. As from the 1990's new studies were developed in the field of genetics, contributing to the understanding of BP concept and the genotype-phenotype relation. In this relation behavioral problems have been underlined and many of them configure indicators of psychiatric disorders (Artigas-Pallarés, 2002; Artigas-Palláres et al., 2006; Bacheaga et al., 2004; Bilder et al., 2009; Collacot et al., 1998).

The explanation for the origin of different types of BP has two sides. The first appeared with Nyhan's proposal in 1976, which states that traits of behavioral phenotypes are biologically determined, not learned and has genetic origin (Artigas-Palláres, 2002; Holland et al., 2003; Moldavsk et al., 2001; Nyhan, 1972). The other variant suggests that BP patterns in genetic

syndromes are determined by biological factors and influenced by environmental factors, as described in the following definition: "Behavioral, cognitive, linguistic, social and also motor patterns that have a biological basis, without disregarding the influence of the environment in their development (Ruggieri & Arberas, 2003, p. 240).

When a syndrome is described with a certain behavioral, linguistic and cognitive pattern one should not presuppose that all individuals affected by the syndrome will present equivalent typical behaviors, once the expression of BP is determined by biological and environmental factors (Ruggiere & Arberas, 2003; O'Brien, 2000). All of these factors operate in a complex of interactions that contribute to the manifestation at times heterogeneous of some behavioral problems (Gallardo-Pujol, et al., 2009; Jarvinen- Pasley et al., 2008; O'Brien, 2000; Ruggieri & Arberas, 2003). Following this discussion, BP was defined as the higher probability for people with genetic syndromes to present certain behavioral and developmental patterns, when compared to people without the syndrome (Dykens & Hodapp, 2007; Flint, 1998; Tartaglia et al., 2010).

The definition of Psychiatric Phenotype, in its turn, involves that set of psychiatric problems which is more frequently associated with a genetic syndrome and in a larger proportion when compared to people of the same age, gender and level of intellectual disability that do not present other biologically based disease (Clarke, 2003). This definition aims at explaining why some individuals that present genetic alterations develop certain psychiatric disorders. One example is the case of the association between Autistic Disorder (AD) and Fragile X Syndrome. People with this syndrome present in a more consistent way autistic behavioral patterns and, likewise, fragile X mutations rates are raised in children diagnosed with AD (Fombonne et al., 1997). This is one example on how genetic factors can increase the vulnerability to develop certain types of psychiatric disorders.

### **3. Using the classification system of mood disorders of the diagnostic and statistical manual of mental disorders (DSM IV) in populations with intellectual disability**

If we observe in detail the clinical criteria of depressive manifestations on DSM IV, it is possible to verify that the assessment of many of them require a minimum of preserved intellectual abilities. Such abilities include organization, planning, competence in problem solving, abstract thought, working memory, attentional abilities, among others (American Association on Mental Retardation, 2006). Take the example of an adult with intellectual functioning significantly below population average, diagnosed before 18 years old and who presents varied deficits in adaptive functioning with losses in the areas of communication, personal care, social abilities, academic performance, safety and leisure, as well as behavioral indicators of sadness and depression. Suppose this individual has a genetic syndrome. In this case, how is it possible to use depressive disorder clinical criteria that have been developed for normative population? How can a person with moderate or severe ID talk about feelings of emptiness or sadness, lack of interest in activities, diminished appetite, fatigue, difficulties in concentration, feelings of uselessness, death thoughts, suicidal ideation and others (APA, 2003). The raising of these questions has lead to different discussions workgroups. One question is related to the procedures to assess the presence of psychiatric disorder in ID condition (Cooper, 2003;

Hermans & Evenhuis, 2010). The other, seeks new proposals for diagnostic classification of psychiatric disorders in people with learning disabilities and ID (Cooper et al., 2003). Both questionings will be treated next.

ID, encoded on DSM IV as MR, is a clinical condition that belongs to the category of Disorders Usually First Diagnosed in Infancy, Childhood, or Adolescence. In the multi-axial system of DSM IV, MR is classified in Axis II together with Personality Disorders (APA, 2003). The manual specifies that, although MR belongs to Axis II, depending on the condition it can be the main diagnosis. However, this classification does not seem appropriate, for it compromises the understanding of what is the ID condition itself. DSM V work group dedicated to childhood and adolescence has already proposed a new category for MR, i.e. Intellectual Developmental Disorder within the condition of Neurodevelopmental Disorders (APA, 2010). According to such discussions, Intellectual Developmental Disorder involves both an intellectual deficit – marked by deficits in mental abilities such as reasoning, problem-solving, planning, abstract thinking, judgment, academic learning and learning from experience – and a deficit in adaptive functioning, which limits participation or restricts performance in one or more aspects of daily activities, such as communication, social participation, functioning at school or work. The onset of these traits must occur during developmental period.

Mood disorders in the ID condition are usually underestimated. There is a tendency to attribute every mood change to the ID condition and this can lead to an obscuration of the diagnosis (American Association on Mental Retardation, 2006). When indicators of mood disorders are attributed to the ID condition, the need for clinical assessment of the patient's mental health and the possibility of intervention are denied. Furthermore, when mental health problems affect people with ID, damages in the adaptive functioning tend to increase (American Association on Mental Retardation, 2006).

Behavioral Phenotype of an individual with a genetic syndrome indicates aspects of the etiological diagnosis which are valuable for professionals that assist him. Unfortunately, not all syndromes have been studied from the psychiatric point of view as they should. Probably, this is associated with the low rates of prevalence of these disorders in general population. In many of them there is consistent data on indicators of mood disorders (Arron et al., 2010; Leyfer et al., 2006; Sinnema et al., 2011; Walker et al., 2011). One of the essential clinical criteria for a health problem to be considered a mental disorder is the persistence of abnormalities that impair several areas of the individual's life (APA, 2003). It is noteworthy that people with genetic syndromes and ID present significant limitations in social, academic and work areas. Hence the use of the manual's diagnostic classifications requires from the specialist expertise in this type of clinical judgement.

Despite the broad acceptance DSM IV and ICD 10, what are their advantages for mood disorder assessment in people with genetic syndromes associated with mental retardation? The multi-axial classification of DSM-IV TR involves an assessment in five axes: Axis I – mental disorders; Axis II – personality disorders and mental retardation; Axis III – medical conditions; Axis IV – psychosocial and environmental problems; and Axis V – global assessment of functioning scale (APA, 2003). As it has been said previously, many of the clinical criteria for depression or bipolar disorder are not easy to be identified in populations with moderate to severe ID. This limitation has lead many groups of specialists to work with them in a different way, considering typical phenotype traits and limitations in intellectual

functioning. The assessment of a depressive or maniac symptom is very important, for it indicates the presence or absence of these symptoms, as well as their severity degree. And, precisely in this aspect, DSM IV clinical criteria can not always be applied in individuals with atypical development (Cooper, 2003).

To respond to the limitations of DSM IV, specialists from London Royal College of Psychiatrists (Cooper, 2003) have stand out for developing a classification system of Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disabilities/Mental Retardation (DC-LD). The group has developed this system based on clinical and research criteria. About the research it is pointed out the need for a diagnostic system different from DSM's for the following reasons: (1) lack of clearness in the diagnostic criteria to help the development of epidemiological studies of incidence and prevalence; (2) need of clinical essay to assess the efficiency of interventions; (3) need of data about the effectiveness of interventions among groups and (4) to identify risk and protection factors for psychiatric disorders in populations with ID. About clinical criteria, the system proposes that the diagnostic process is made in three basic stages: (1) measuring psychopathology; (2) classifying the psychopathology using operationalized diagnostic criteria (descriptive diagnosis) and (3) determining the etiology of the descriptive diagnosis (often multifactorial), using a biological psychological-social-developmental framework (Cooper, et al., 2003, p. 4).

DC-LD has a critical view of DSM IV. The group considers that the classification of disorders in Axis I is ambiguous in case of ID condition. It suggests that, in people with ID, instead of assessing symptoms by language abilities, it should be used their behavioral equivalents. DC-LD recommends avoiding the use of diagnostic subcategories for each disorder. A careful analysis is also made on the behavioral phenotype described for the genetic syndrome. It is not recommended consider in the psychopathology of a disorder some behavioral pattern of the genetic disorder. To illustrate this, it is possible to observe the hyperphagia pattern that marks Prader-Willi Syndrome, the bizarre/aberrant behavior presented by people with Smith-Magenis Syndrome or the impulsiveness described in Cri du Chat Syndrome (Clarke, 2003).

According to DC-LD, for adults with mild intellectual disability it is suggested the use of DSM IV classification (Cooper et al., 2003). Finally, the DC-LD system presents a multi-axial structure that includes a classification guideline for psychiatric disorders associated with genetic syndromes for moderate to severe ID cases. Axis I: severity of intellectual disabilities, Axis II: cause of intellectual disabilities and Axis III: presence of additional psychiatric disorders. It is noteworthy that Axis I and II can adopt diagnostic categories from ICD-10 and DSM IV.

#### **4. Assessment of mood disorders in people with mental retardation**

Both DSM IV and DC-LD's new system are supported by clinical judgments to raise diagnostic hypothesis for mood disorders. This judgment is a special type of assessment that is based on empirical evidence. However, there are other procedures that help this clinical judgment. The observational method and psychological instruments (scales, inventories or semi-structured interviews) are some examples. In any case, they must not be used as the only source of information for the diagnosis.

The observational method helps professional practices for modifying and management behavioral problems (Martin & Pear, 2009; Silvaes, 2000). This type of assessment procedure requires that the observer has been previously trained in three areas: clinical traits of the mood disorder he wishes to observe the ID condition and the Behavioral Phenotype described for the genetic syndrome. To observe mood indicators of an individual with ID requires the specialist to focus on the way the person behaves, the frequency of the behavior's occurrence, intensity and also environmental factors possibly related to mood change. The main advantage of the observational method is that it allows the control of variables directly related to the observed behavior, whether it happens in a natural environment or in laboratories. Otherwise, behavioral observation is not always sensitive for the assessment of mood states and feelings (Martin & Pear, 2009).

The indirect assessment procedures use standardized tools that rely on information provided by informers/caregivers. For people with ID it is necessary to consider the vast number of standardized tools available to assess general behavioral alterations in children and adults with atypical development. These instruments track only behavioral problems associated with the behavioral phenotype of genetic syndromes, i. e. aggression, self-injury, behavioral stereotypy, repetitive responses, tantrums, irritability, agitation, lethargy, among others (Aman et al., 1985a; Brinkley et al., 2007; Brown et al., 2002; Hill et al., 2008; Kaland et al., 2008; Karabekiroglu & Aman, 2009; Keil et al., 2009; Narita et al., 2004; Polk et al., 2008; Rojahn et al., 1997). Some tools for this assessment are: Behavior Problems Inventory (BPI-01) (Rojahn, J. et al., 2001), Aberrant Behavior Checklist (ABC) (Aman et al., 1985b), Communication and Symbolic Behavior Scales Developmental Profile (CSBS) (Wetherby et al., 2002), The Repetitive Behavior Scale Revised (RBS-R) (Bodfish, et al., 2000), Questions about Behavior Function Scale (QABF) (Matson et al., 1999). These instruments offer discerning indicators of behaviors and are of relatively fast application and easy correction.

However, other tools are specifically designed to assess behavioral indicators of psychiatric disorders, such as mood disorders. In childhood and adolescence some of the most used instruments for this end are the behavioral assessment inventories of the Achenbach System of Empirically Based Assessment - ASEBA (Achenbach & Rescorla, 2001). Its application involves asking parents or caregivers about the child's competence in different areas, as well as the assessment of the existence of diseases and disabilities, parental worries about the child and positive aspects they recognize in them. Another set of questions assesses emotional and behavioral problems encoded in the scales of internalization, externalization and total problems. These inventories have been translated, adapted and internationally validated (Achenbach & Rescorla, 2001; Dykens et al., 2002, 2007; Graham et al., 2005). Some of the most commonly used instruments in childhood and adolescence are: Child Behavior Checklist (CBCL), Youth Self-Report (YSR) and Teacher's Report Form (TRF). All three inventories provide specific DSM-oriented scales. In the affective problems scale, for example, statements about the behavior of the assessed child are consistent with DSM's clinical criteria described for Major Depressive Disorder and Dysthymic Disorder. The inventory requires the informant to say if the statement is: not true of the child, somewhat or sometimes true, very true or often true of the child. Some of the statements found in the inventory are: there is very little he/she enjoys, he/she cries a lot, he/she deliberately harms him/herself or attempts suicide, he/she doesn't eat well, he/she feels worthless or inferior, he/she feels too guilty or overtired without good reason (Achenbach System of Empirically Based Assessment [ASEBA], 2011).

In the form of a semi-structured psychiatric interview, the Schedule for Affective Disorders and Schizophrenia for School-Age Children – Present and Lifetime Version (K-SADS-PL) is another tool designed to assess the symptomatology of psychiatric disorders in the present and throughout the life of children between 6 and 18 years old. The questions from the first version of this interview were based on DSM III and DSM III R's clinical criteria for psychiatric disorders in childhood (APA, 1980; APA, 1987; Chambers et al., 1985). After the publication of the manual's fourth edition, Kaufman and colleagues developed a new version of the interview that comprised the assessment of the child's symptoms in the present and throughout his/her lifetime (Present and Lifetime Version), according to clinical criteria of Disorders Usually First Diagnosed in Infancy, Childhood or Adolescence (Kaufman et al., 1997). K-SADS-PL provides objective questions and criteria to assess each symptom. Some of the assessed diagnoses are: Major Depression, Dysthymia, Mania, Hypomania, Cyclothymia, Bipolar Disorders, Schizoaffective Disorder, Attention Deficit Hyperactivity Disorder, Conduct Disorder, among others. This interview collects data from parents/informants and also from the child. A conclusive score about the symptomatology is based on the summary of all this information. For example, on the field of depressive disorders the interview assesses if the child has ever felt very sad and depressed, and how often it has happened; subjective feelings of irritability; bad mood; anger; lack of interest; little motivation; thoughts of death; suicidal ideation, among others. Concerning manic affective states, the following problems are assessed: exalted mood, excessively optimistic attitude, excessive happiness, diminished need for sleep, among others.

For adulthood there are also several instruments to track mood disorders indicators in people with ID. In 2010, Research in Developmental Disabilities published a review article that aimed at obtaining information on psychometric properties of screening instruments for depression in adults with ID (Hermans & Evenhuis, 2010). The period of time covered by the review was of 28 years, and the databases revised were PUBMED, PSYCHINFO and EMBASE. Only articles in English were included. A quality grade was granted to instruments according to their psychometric properties. Among the instruments that reached the best grades were Glasgow Depression Scale for people with a Learning Disability (GDS-LD), Reiss Screen for Maladaptive Behavior (RSMB), Diagnostic Assessment for the Severely Handicapped-II (DASH-II), Mini Psychiatric Assessment Schedule for Adults with Intellectual Disabilities (Mini PAS-ADD), Assessment of Dual Diagnosis (ADD), Psychiatric Assessment Schedule for Adults with a Developmental Disability (PAS-ADD), Glasgow Depression Scale – Carer Supplement (GDS-CS), Self-Report Depression Questionnaire (SRDQ) and Psychopathology Instrument for Mentally Retarded Adults (PIMRA). The authors draw attention to the need for many instruments to be assessed from the point of view of their sensibility and specificity to identify affective problems in people with ID, since not all of the studies fulfilled properly these psychometric properties.

The previous theoretical considerations allow us to present next a brief synthesis of the main mood disorders identified in people with ID and genetic syndromes, such as Prader Willi syndrome, Down syndrome and Williams syndrome.

## **5. Mood disorders indicators in genetic syndromes**

The proportion of mental health problems in individuals with intellectual disabilities associated with genetic syndromes is twice to three times greater than general in the general

population. Despite that, the identification of emotional problems is complex due to the expression of behavior phenotype variability (Sturmey et al., 2010; Clarke, 2003). However, in individuals with genetic syndromes and ID the acknowledgement of psychopathological conditions such as depression and bipolar disorder is relatively recent compared to typically developing people (Adams & Oliver, 2011; Hayes et al., 2011; Matson & Laud, 2007; Mutkins et al., 2011; Tsiouris et al., 2011).

In ID condition, mood disorders have received special attention due to the impairments that this kind of problem causes in the accomplishment of daily life activities. People with intellectual disabilities may exhibit significant limitations in adaptive behavior which can impair their social life (Denis et al., 2011; Koskentausta et al., 2007). These limitations can be caused by difficult management of psychiatric and emotional problems, such as shyness, phobias, mood changes, nervousness, somatic complaints, sadness, withdrawal, worries (Bolsoni-Silva et al., 2010; Walker et al., 2011).

Within the category of mood disorders, depressive disorders have received special attention in genetic syndromes accompanied by ID. Due to the interference in language expressive abilities that can be caused by the intellectual impairment, it is recommended, whenever it is possible, the assessment of these individuals according to behavioral equivalents for depression. Previous studies have described some of these behavioral equivalents which usually have acute beginning and do not have medical causes. Some examples are: aggressive behavior, tearfulness, self-injury, diminished daily life abilities, social isolation, somatic complaints, hypochondria, appetite alterations, weight changes, psychomotor agitation or retardation (Benson & Brooks, 2008; Cooper, et al., 2007; Smiley & Cooper, 2003). It will be presented next a brief characterization of genetic syndromes and their main associated mood disorders.

### **5.1 Prader Willi syndrome**

Prader Willi Syndrome (PWS) is a genetic disorder caused by the lack of expression of paternally inherited imprinted genes on chromosomal region 15q11-q13. Genes from this region have differential expression according to the paternal origin so that the maternal and paternal copies must be present for normal genetic expression. Criteria of PWS are well established and laboratory diagnosis is made in 95% of the cases: in 70% of the patients paternal deletion is found in chromosome region 15q11-q13 and 25% have uniparental disomy. The incidence ranges from 1:10.000 to 15.000 births (Cassidy & Driscoll, 2009; Jin, 2011). The syndrome is characterized by two clinical phases. In the first cardinal symptoms are neonatal hypotonia, difficulties to be fed, lethargy, weak cry and hyporeflexia. In the second phase, from six months on, there is an improvement of hypotonia, weight gain, progressive development of hyperphagia and obesity. Genital alterations such as cryptorchidism, micropenis and scrotal hypoplasia are found in male children and hypoplasia of female external genitals in female children (Holm et al., 1993).

The Behavioral Phenotype described in people with this disease indicates many behavioral problems in childhood, adolescence and adulthood such as: obsessive-compulsive spectrum behaviors, anger and aggressive bursts, tendency to rigidity, oppositional behavior, lying and stealing, mild to moderate ID, neuropsychological alterations associated with prefrontal lobe activity in attentional abilities, executive functions (for instance, inhibitory control and

selective attention) and visuospatial organization. The most frequent psychological complaints are of the emotional-affective type, psychotic disorders spectrum complaints, withdrawal, mood changes and depression (Benarroch et al., 2007; Dykens & Roof, 2008; Goldstone et al., 2004; Goos & Ragsdale, 2008; Gunay-Aygun et al., 2001; Hinton et al., 2006; Jauregi et al., 2007; Koenig et al., 2004; Semenza et al., 2008.)

Although the interest for the study of mood disorders in people with PWS dates back to the 1990's (Dykens & Cassidy, 1995), recent research have been trying to assess the association between certain types of mood disorders and two of the main genetic subtypes of the disease: deletion and maternal uniparental disomy (Hartley et al., 2005). For instance, a recent study has put forward the hypothesis that deletion subtype is more associated with depressive disorders without psychotic symptoms, as opposed to maternal uniparental disomy that presented more association with bipolar disorder and psychotic symptoms (Soni, et al., 2008). Notwithstanding such findings are still contradictory.

A recently published cohort study with an expressive sample of 68 individuals with clinical and genetic diagnosis of PWS has allowed researchers to share the sample in four age groups: < 25 years old, 25-34 years old, 35-44 years old, ≥ 45 years old. They were also able to make comparative analysis for age and genetic subtype according to the prevalence of psychiatric disorders, among them mood disorders and psychotic disorders. The instrument used to verify psychopathological indicators was the Developmental Behavior Checklist for Adults, which was answered by the main caregiver of each participant (Sinnema et al., 2011). This study compared mood disorders distribution of the diagnoses according to the genetic subtype of deletion and maternal uniparental disomy. The findings indicate that, whatever the genetic subtype is, it was observed a relatively equivalent distribution of 16 to 20% of the sample in the fulfilling of typical bipolar disorder symptoms with psychotic symptoms and psychotic disorder isolatedly. The depressive symptoms observed were isolation, diminished interest or pleasure in making activities, impairment of self-care abilities, depressive mood or sadness. In the scale of the inventory that assesses clinical indicators of depression it was observed that the number of affective problems increased with the age of the participants. Among the main symptoms verified in the frame of bipolar disorder with psychotic symptoms were: mood fluctuations that involve hypomania and depression, paranoid ideas and hallucination (Sinnema et al., 2011).

So far, one Brazilian study was published about behavioral phenotype of PWS (Mesquita et al., 2010). The sample of this research was composed of 11 children and adolescents with clinical and molecular cytogenetic diagnosis of PWS and it explored indicators of mood disorders through the use of Achenbach & Rescorla's Child Behavior Checklist 6-18 - CBCL/6-18 (Achenbach & Rescorla, 2001). It was verified that out of CBCL/6-18's four DSM-oriented scales, the affective problems scale indicated the presence isolation, depression, sadness, excessive dependence of adults and affective problems. It was also observed positive and statistically significant correlation between social problems and the scales of isolation and depression (Mesquita et al., 2010). Despite the restricted sample and convenience criteria for sample selection, this was the first Brazilian research to describe the behavioral phenotype of children and adolescents with the syndrome, and some of the findings were consistent with previous studies (Lindgren et al., 2000; Semenza et al., 2008).

## 5.2 Williams syndrome

Another target genetic disorder in the study of depressive disorders is Williams syndrome (WS). WS is a genetic disorder caused the deletion of multiple genes of the long arm of chromosome 7 (region 7q11-23) (Sugayama et al., 2007). Previous studies registered incidence rates of 1:20.000 to 1:50.000 live births and prevalence between 1:7.500 and 1:20.000 live births (Meyer-Lindenberg et al., 2006; Sugayama et al., 2007). Initially, the diagnosis of the syndrome is based on clinical criteria such as typical facial dysmorphism during childhood (flat mid-face, periorbital fullness or swelling, upturned nose, prominent cheeks); cardiovascular diseases (supravalvar aortic stenosis); connective tissue and calcium level alterations (Cardoso-Martins & Silva, 2008; Herreros et al., 2007; Jarvinen-Pasley et al., 2008; Martens et al., 2008; Rossi, et al., 2006, 2009). However, diagnostic confirmation can only happen by means of cytogenetic examination (Fluorescence *in situ* Hybridization Test - FISH) (Sugayama et al., 2007).

The syndrome is characterized by different cognitive and behavioral alterations (Porter & Dodd, 2011). Typical cognitive patterns of the syndrome's phenotype are: intellectual disability in various degrees, neuropsychomotor development delay in the first years of life, good performance in tasks that require the use of expressive language, impairment in receptive language abilities, syntactic-pragmatic, structural and functional alterations of language, use of clichés, sound effects, intonation resources, echolalia, deficits in visuospatial abilities, attention deficit, alterations in executive functions that involve work memory, inhibitory control and planning of task accomplishment (Mervis & John, 2010; Teixeira et al., 2010).

About the Behavioral Phenotype, studies with children from 5 years old, report significant contrast between deficits and excess of abilities that interfere considerably in their adaptation in familiar, social and school environment (Artigas-Pallarés, 2002; Bellugi et al., 2000; Cardoso-Martins & Silva, 2008; Davies et al., 1998; Jarvinen-Pasley et al., 2008). Among the excesses it is found empathy, hypersociability, agitation, various behavioral stereotypies and anxiety. Among behavioral deficits some traits are difficulties in establishing and maintaining relationships, establishing limits for physical proximity, maintaining cooperative games, sharing interests and waiting for his/her turn in conversations.

Although the most prevalent mental disability in the syndrome is Attention Deficit Hyperactivity Disorder, the behavioral and emotional phenotype of individuals with Williams Syndrome is characterized by other affective problems, such as Generalized Anxiety Disorder, phobias, fears and social withdrawal (Martens et al., 2008; Mervis & Becerra, 2007; Leyfer et al., 2006; Martens et al., 2008). Previous studies that used behavior problems assessment scales verified the prevalence of behavioral indicators of depression, almost always associated with anxiety and somatic complaints (Pérez-García, et al., 2011). Also about the contrasts between excesses and deficits in abilities, in a systematic review study on WBS (Mervis & John, 2010), underscore the high rate of social interactions that they establish and social abilities deficits observed during these interactions. Some of these contrasts are likely to contribute to the development of affective problems, especially concerning hypersociability and, at the same time, lack of social abilities that can end up with the individual's withdrawal.

A previous study with an expressive number of participants (n=190) has allowed the author to share the sample in three age groups: 6-9, 10-13 and 14-18. The instruments used were

Child Behavior Checklist (CBCL/6-18), Behavior Assessment System for Children (BASC), Revised Children's Manifest Anxiety Scale (RCMAS) and Yale-Brown Obsessive-Compulsive Symptom Checklist (Y-BOCS). Concerning depressive disorders indicators it was found that the group of adolescents between 14-18 years old reached the highest scores in the anxiety/depression scale of CBCL when compared to the remaining groups (Switaj, 2000).

Other studies have compared behavioral indicators of mood disorders among people with different genetic syndromes. One example is a research that involved three groups of syndromes: Fragile X syndrome, Down syndrome and Williams syndrome. This study shows CBCL clinical profile for the total WBS cohort. The scales with highest percentage of individuals within subclinical range (T-scores between 65 and 69) and clinical range (T-scores higher than 70) were anxiety/depression and attention problems. Global internalizing and total scales also included a significant percentage of patients in either subthreshold or clinical range. Although this research aimed at contributing to the description of WBS behavioral phenotype, as in previous studies, anxiety and depression appeared with the highest mean scores in WBS with many individuals within clinical range (Pérez-García, 2011).

In Brazil there are only two published studies on the identification of behavior problems and mood disorders indicators in children and adolescents with WS. The first aimed at describing behavioral, cognitive and language profiles and identifying autistic behavior in a group of 10 children and adolescents with WBS, age range 5-16 years old, and 10 typically developing children and adolescents. The instruments used were: Test of Nonverbal Intelligence (Leiter-R); Child Behavior Checklist (CBCL/1½-5; CBCL/6-18); Language Exam (TIPITI); and Autism Screening Questionnaire (ASQ). One-way ANOVAs were applied to compare the mean of compatible scales of CBCL/1½-5 and CBCL/6-18 inventories between groups with WBS and CG. There were statistically significant differences between groups on the scales of internalizing problems, total emotional/behavioral problems and affective problems (Teixeira et al., 2010). The second study was made with 22 children and adolescents with WS, age range 7 years 5 months old to 18 years 3 months old, with clinical and molecular cytogenetic diagnosis for WS confirmed by Fluorescence *in situ* Hybridization (FISH) techniques. All participants were members of the Brazilian Association of Williams Syndrome. The sample's intellectual performance was assessed through block design and vocabulary subtests of Wechsler Intelligence Scale for Children (WISC-III). The result was an estimate intelligence quotient compatible with inferior classifications that characterize mild to moderate intellectual disability. The results concerning behavior problems and mood disorders were assessed through the use of Child Behavior Checklist (CBCL/6-18) and indicated a bordering classification in the scales of anxiety and depression, affective problems and in the internalizing scale of the inventory, which is formed by the subscales of anxiety and depression, isolation and depression, and somatic complaints. The scale of total emotional/behavioral problems indicated clinical range compatible scores (Segin, 2010).

### 5.3 Down syndrome

Down syndrome (DS) is another genetic disorder that is associated with psychiatric disorders with difficult social and familiar handling. The disease is caused by

chromosome 21 trisomy and is the most common genetic cause for ID. It is also one of the few aneuploidy conditions compatible with post-natal surviving chances. The syndrome has a prevalence of approximately 1:700 births and is associated with more than 80 clinical conditions that include congenital cardiac malformations, duodenal stenosis, hypotonia, immune system deficiency, raised risk for dementia of the Alzheimer type in adulthood, among other conditions of higher or lower severity according to the variability of each case (Carr, 1994; Harris et al., 1996; Sherman et al., 2007; Sommer & Henrique-Silva, 2008). Some typical patterns of DS's behavioral phenotype in children and adolescents are increased sociability, mood oscillations and stubbornness. Among cognitive alterations it is described ID, joint attention problems, deficits in verbal processing of information, deficits in working memory and several executive functions, inattention and expressive language deficits (Adams et al., 2008; Balci & Ahmet, 2007; Fidler, 2005; Ruggieri & Arberas, 2003).

In 2007 it was published a systematic review study that identified from other researches the main behavioral alterations and psychiatric disorders in individuals with DS in three development phases: childhood, adolescence and adulthood (Dykens, 2007). It is noteworthy the fact that children with DS do not present high rates of behavioral problems, emotional and psychiatric alterations when compared to other groups of children with ID. However, when compared to typically developing children, a larger incidence of externalizing behaviors is reported for the DS group (manipulation, oppositionism, concentration problems, impulsiveness, attention deficit hyperactivity disorder, oppositional defiant disorder, low rates of co-occurrence of autistic spectrum disorders). In adolescence, the author warns about improvements that can happen in the course of some cognitive symptoms which are identified in childhood. For instance: hyperactivity, concentration problems and inattention. Nevertheless, the beginning or aggravation of affective problems that indicate mood disorders (such as internalizing symptoms, especially withdrawal, being more secretive and quiet, preference to be alone) happen in this phase of development. In adulthood, although there is less literature material, studies point to the increase of neurological diseases such as Dementia of the Alzheimer Type with aggressiveness symptoms, high rates of depression, isolation, negative mood, passiveness, insomnia, diminished appetite, schizophrenia and obsessive-compulsive disorder.

Another recent literature review study analyzed 30 articles on depression in DS (Walker et al., 2011). The work draws attention to precautions for the diagnosis of depressive disorders in people with DS, once there are two frequently associated medical conditions: dementia and hypothyroidism. However, people with DS present several compatible symptoms with mood disorders of Major Depression Disorder type, such as reduced interest and pleasure, depressed affect, psychomotor retardation, tearfulness, reduced energy, loss of appetite, sleep disorders, hypochondria, aggression or tantrums and reduced speech. As in PWS, it can be identified an association of depressive symptoms with psychotic ones, such as hallucinations, obsessive ideas and compulsion. This makes a proper diagnosis even more difficult. The study concluded that there are many "blind spots" in our current clinical knowledge on depression in DS. Further studies should include systematic assessment of treatment strategies for depression in DS and incorporate the investigation of the role of social support, psychotherapy and biological treatments (Walker et al., 2011).

## 6. Conclusion

There are several specificities in the assessment of mood disorders in genetic syndromes accompanied by ID, especially due to the difficulty in having access to reliable verbal reports on the degree of psychological suffering of these individuals. Seeing that, it should be taken into account the use of standardized instruments in the assessment process to allow access of this information through informant's reports. Thus, it is suggested that the diagnosis of mood disorders in people with ID to be done mainly with the use of behavioral and/or psychiatric assessment instruments that allow adaptations for using DSM-IV's clinical criteria. Besides, in ID populations it is necessary to assess adaptive behaviors and risk and protection factors involved in the etiology and in the course of a mood disorder.

The chapter has pointed the need of interdisciplinary approaches in the assessment and intervention of mood disorders in populations with genetic syndromes associated with ID. Such approach allows the identification of impaired or preserved areas of cognitive, affective, behavioral and social functioning and, therefore, planning interventions that incorporate more appropriate repertoire to their functioning.

Although prevalence rates of genetic syndromes are low, especially WS and PWS, it is imperative to develop epidemiologic studies that can contribute to more accurate knowledge on the incidence and prevalence of mood disorders in this population.

The table 1 summarizes the contents of this chapter. There are reported the main problems and recommendations in the assessment of mood disorders in genetic syndromes associated with intellectual disability.

Problems	Recomendations
Mood disorders and ID are both complex conditions.	The assessment process ought to involve an interdisciplinary professional team.
Mood disorders in the ID condition are usually underestimated. There is a tendency to attribute every mood change to the ID condition and this can lead to an obscuration of the diagnosis.	The clinical judgments may be established by special type of assessments that have to be based on empirical evidence.
How to access mood disorders indicators in individuals with ID?	There are some procedures that can help clinical judgment. The observational method and use of psychological instruments (scales, inventories or semi-structured interviews) are some examples. In any case, they must not be used as the only source of information for the diagnosis.
How to assess depression disorders indicators in individuals with expressive language impairment?	It is recommended to assess these individuals according to behavioral equivalents for depression.

Problems	Recomendations
Some times, it is common to confound typical behavioral patterns of a syndrome with signs of mood disorders.	Improve knowledge about behavioral phenotype of the syndrome to stablish differences between typical behavioral patterns and mood disorders indicators.
High incidence of mental health problems in Prader Willi syndrome.	It is necessary to consider that the most frequent mental health and psychological complaints in PWS are the emotional-affective problems as withdrawal, mood changes and depression, in addition to psychotic disorders spectrum.
High incidence of language alterations and depression in the Williams syndrome.	During assessment of the Williams syndrome, it is necessary to consider that the language domain is characterized by strength in concrete vocabulary and grammatical abilities but weakness in relational/conceptual and pragmatic language. Thus, it is necessary to be careful about assessing verbal report of these individuals.
High incidence of attention deficit hyperactivity disorder in the Williams syndrome.	During assessment of the Williams syndrome, it is necessary to consider that performance on cognitive tests may be influenced by the signs of this disorder.
High incidence of mental health problems in Down syndrome.	It is necessary to consider that people with DS may present several compatible symptoms with mood disorders, such as reduced interest and pleasure, depressed affect, psychomotor retardation, tearfulness, reduced energy, loss of appetite, sleep disorders, hypochondria, aggression or tantrums and reduced speech.

Table 1. Main problems and recommendations in the assessment of mood disorders in genetic syndromes associated with intellectual disability.

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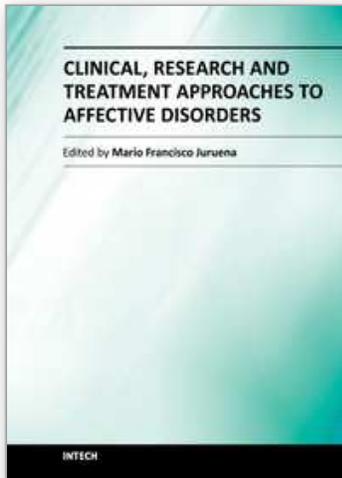
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## **Clinical, Research and Treatment Approaches to Affective Disorders**

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The causes, development and outcomes of disorders are determined by the relationship of psychological, social and cultural factors with biochemistry and physiology. Biochemistry and physiology are not disconnected and different from the rest of our experiences and life events. This system is based on current studies that report that the brain and its cognitive processes show a fantastic synchronization. Written by the foremost experts on Affective Disorders worldwide, this book is characterized by its innovative, refreshing, and highly sensitive perspective on current knowledge of diagnostic, neurobiology, early life stress and treatment of Mood Disorders. The authors share a deep understanding of unique challenges and difficulties involved in Affective Disorders, and have achieved a balance among clinical, research and new treatment approaches to Affective Disorders. The chapters are written in a comprehensive, easily readable, and highly accessible style, stimulating readers, clinicians and researchers.

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Phone: +86-21-62489820  
Fax: +86-21-62489821

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