

# Observations on the Zero to Three Diagnostic Classification

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## OBSERVATIONS ON THE DIAGNOSTIC CLASSIFICATION OF MENTAL HEALTH AND DEVELOPMENTAL DISORDERS OF INFANCY AND EARLY CHILDHOOD

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### SUMMARY

The present article comments on the format and use of the diagnostic classification recently developed by Zero to Three/National Center for Clinical Infant Programs (1994). Our main goal is to stimulate discussion among clinicians about the issues involved in this classification system. First, we put forward some thoughts on the classification system itself and its diagnostic criteria. Second, we address cultural or transcultural factors, particularly regarding the assessment of relationship disturbances in context. Third, some observations are made regarding the diagnostic hierarchy and the place of "organic conditions" in the classification system and their role in differential diagnosis. Fourth, we comment on the problematic issue of diagnosing regulatory disorders. Finally, we offer some observations about attachment disorders and relationship disorders. Brief clinical vignettes illustrate our main points.

### I. DIAGNOSTIC CLASSIFICATION SYSTEM.

This classification system is the first formal one offered for children of this age. It is the product of consensus reached by authors from different theoretical backgrounds and from several infant centers. The diagnostic system addresses the manifestations of emotional, developmental, and relationship disturbances observed in infants and very young children up to age four.

In brief, the classification system models itself after the five axis system followed in the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) (American Psychiatric Association, 1994). The authors point out that the Zero to Three classification is not intended to replace current nosological systems, but instead aims to supplement them for children in this age range.

Axis I diagnoses are intended to reflect the most prominent features of the disorder and are limited to seven major diagnostic categories (traumatic stress disorder, disorders of affect, adjustment disorder, regulatory disorders, sleep behavior disorder, eating behavior disorder and disorders of relating and communicating).

Axis III and IV resemble the DSM-IV system, in that medical conditions and environmental stressors are noted. Axis III is used to describe medical and developmental conditions that affect the child, as well as disorders included in other diagnostic classifications, like DSM IV when such disorder is also evident in the child. Axis IV describes psychosocial stressors according to their severity, duration, and overall impact. Finally, Axis V is used to note the functional and emotional developmental level of the child, according to some specified guidelines

The Zero to Three classification is an enormous step forward. As clinicians and researchers from all over the world use and react to the classification, further contributions will be made to the development of its diagnostic taxonomy. The classification will no doubt be a catalyst for research, facilitating communication between clinicians from different centers via a common nomenclature.

As the first specimen of its kind, the Zero to Three classification system presents some obvious but unavoidable obstacles. Prime among these is that its basis is expert consensus, rather than empirical or scientific evidence. This is also the case, to a large degree, for DSM-IV, as well as for Chapter V of the International Classification of Mental and Behavioral Disorders presented in the tenth edition of the International Classification of Diseases (ICD-10) (World Health Organization, 1992). The boundaries between disorders, and their place in the diagnostic taxonomy, are for the most part based on the consensus of experts, rather than on empirical or scientific evidence.

In the field of infant mental health, there is still relatively scant empirical evidence about the validity of disorders. Several constructions, although clinically quite useful and sound, are not based on empirical evidence (e.g., relationship disturbances and regulatory disorders).

A major step forward -- and to a degree a departure from DSM-IV and ICD-10 -- is the formulation of relationship disorders and disturbances in a way that attempts to diagnose not just one individual or another, but rather the relationship between the two. This approach is theoretically at the opposite pole from DSM-IV, which assumes that all mental disorders exist within the affected individual.

Also, the Zero to Three diagnostic classification encourages clinicians to select one diagnostic category in Axis I that may or may not coexist with a relationship problem (Axis II) between the infant and a caretaker. In general psychiatry, there is a clear tendency to assign multiple diagnoses to an individual patient, in order to capture all the manifestations of difficulties. One individual with a personality disorder, for instance, might also be diagnosed with a substance abuse problem, a substance-related mental disorder, a stress-induced condition, and a preexisting mood disorder. Whether such splintering of problems is really helpful for the individual patient is questionable, as is whether the clinician gains much from accepting the existence of such thin boundaries between some disorders (e.g., anxiety disorders and mood disorders). In the Zero to Three classification, clinicians are encouraged to think in terms of a categorical problem in the infant within the frame of reference of the infant's relationships with caregivers, and taking into account stress factors for the child.

Given that the field of infant mental health is itself in its infancy, it seems more useful not to develop too many diagnostic categories nor to delineate fine distinctions between them. Doing so would lead to multiple diagnoses and a premature separation of conditions, although there is no scientific evidence that such boundaries really exist.

To test the existence of some collections of symptoms, there is clearly a need for much empirical research, which would lead, in turn, to specific diagnostic conceptualizations. This is the main way to validate the existence of definite conditions or disorders as distinct entities with an etiology, a clinical course, and other nosological features. All these features can be ascertained only through clinical research.

Recommendations in the introductory section of the diagnostic classification address the need for the careful evaluation of infants and their caretakers, and emphasize the value of the multidisciplinary nature of the field. That is, on the need to assess infants, their abilities and challenges, and their relationships from several points of view (medical, psychological, occupational therapy, speech and hearing, etc.).

We will make some observations about specific aspects of the diagnostic system, hoping to stimulate further discussion among clinicians and researchers. The classification is undergoing a process of review and critique by clinicians in the U.S. and other countries. To help shape the review process, the members of the diagnostic classification task force have invited reactions from clinicians and researchers.

## **II. CULTURAL/ TRANSCULTURAL FACTORS.**

Clinicians working with infants and young children can clearly see that they are quite dependent on the quality of relationships established with their caretakers. These relationships are determined in large part by the cultural prescriptions and practices within each family and social group. Thus, child care practices and methods of child-rearing are highly specific to cultures. Yet many industrialized countries, such as the United States, Canada, and several European countries, are truly multicultural societies where individuals from very different cultures interact (e.g., a clinician and a family from different cultural backgrounds).

Keeping this diversity in mind is particularly relevant diagnostically when considering conditions such as relationship disturbances. It would seem advisable for the diagnostic criteria for these disorders to clearly indicate that maladaptive patterns of relating or care-taking are distinctly outside those considered adequate within a specific cultural context.

In our clinical work, we have encountered, in particular, the problem of dissonance between different cultural values. For instance, a clinician working with a parent-child dyad from Japan, India, or Latin America might find that the mother is "over involved" with her child, because she does not separate from the child at all during the first few years of life (Guzder, 1992). Furthermore, the mother and father co-sleep with the baby, and they do many things for the infant (e.g., carry him around, spoon feed him, carry his things) for far longer than is typical for Anglo-Saxon children of the same age. The mother may express a preoccupation with her baby and may appear overly solicitous and indulgent. Within the family's original cultural context, this care-taking pattern may be entirely

normal, but it may not appear so outside that specific context. The diagnostic classification should therefore include a cautionary statement along these lines, particularly in regard to evaluating the quality and nature of parent-child relationships.

In many societies, infants are not encouraged to do things for themselves as much as is typical in some Northern European or North American cultural groups. On the contrary, individuation is not encouraged but rather social interdependence and dependency are reinforced and encouraged (Guzder, 1992 ). Assertiveness and self-reliance are not viewed as desirable qualities, in comparison to obedience and compliance.

On first going to a daycare setting or a preschool, a young child from one of these cultures may never have been separated from the mother. The child may never have been looked after by a stranger, such as a baby-sitter. For this child, a separation would be much more stressful than for a child who had "practiced" it all before. In addition, the mother's feelings of anxiety and fear on being separated from her child would be more intense, and might even be considered "pathological" outside their cultural context.

Another area of possible misunderstanding has to do with the role of eye contact and touch in relationships. In several ethnic groups, eye contact is not considered a central part of relationships and might even be avoided (LeVine, 1990; Quinton 1994). As such, it is not essential to the parent-child relationship and may be discouraged. This relationship pattern may apply, for example, to some groups of African-American mother-child dyads and to American Indian families (Quinton, 1994; Canino and Spurlock, 1994) .

In many ethnic groups, physical contact is not considered normative, particularly after a certain age. It is not considered essential as a manifestation of love in several American Indian groups, and it is not practiced in certain other ethnic groups such as India and Japan as the child grows older, for instance to the age of three or four. Failure to consider these two culturally determined patterns of interaction (eye contact and physical touch) in relation to their native cultural context could result in misconstruing them as manifestations of a distant or under involved relationship between an infant and a caretaker.

Another culturally determined aspect of the care-taking relationship is the involvement of multiple and substitute caretakers. In some groups that share care-taking responsibilities, it may not even be clear who the primary caretaker of an infant is. In cases where there is an extended family available, for instance, a grandmother or other relative may assist the biological mother in caring for the infant on a day-to-day basis. There is no indication that this "sharing" of attachment has negative effects for the child (Quinton, 1994).

### **III. DIAGNOSTIC HIERARCHY AND "ORGANIC" CONDITIONS.**

The Zero to Three classification explicitly advises clinicians to consider the role of stress first as the highest order in the hierarchy of diagnoses in an infant's life. Stress can induce manifestations in an infant that mimic other conditions and, therefore, may lead to a misdiagnosis. Before considering other possible diagnostic entities, clinicians are encouraged to rule out the possibility that major stressors may be at play. While this is quite important and clinicians are well advised to keep stress in mind, instead, we believe that clinicians should first rule out another set of conditions even before considering stress as a precipitating factor -- but especially before going on to consider other possible diagnoses. This set of conditions is what used to be called "organic disorders" but in DSM-IV is now referred to as Cognitive Disorders, Substance-Related Disorders, and Conditions Related to a General Medical Condition (which includes any condition diagnosable in the body of knowledge of general medicine).

The reason for this suggestion to modify the hierarchy of diagnosis is that "organic" entities can indeed produce behavioral and emotional symptoms in a child that may mislead the clinician into diagnosing the problem as an emotional disturbance due to stress, or a purely behavioral problem, when instead the situation is due to biological factors. This is an issue of enormous importance that clearly illustrates the necessity of involving physicians (e.g., a pediatrician, child psychiatrist or a child neurologist) in the diagnostic work-up of infants and young children who exhibit behavioral or emotional disturbances. Such involvement can range from direct examination of a child to seeking an outside consultation in cases where there is the possibility of a biologically based underlying condition. It would be inadequate for a clinician to ignore this set of causal factors, and to go on to recommend an approach that did not take into account these etiological factors; as a result, the treatment could be misguided and might delay the recognition and handling of the organic condition.

It would be undesirable, for example, for a child to be given a diagnosis of an anxiety disorder and to begin treatment, under the assumption that the child was reacting to maternal anxiety, when instead he might be suffering from the side effects of prescription medications given for bronchial asthma, such as salbutamol or corticosteroids. Both of these medications could lead to a

presentation of symptoms in a young child that reflect an agitated state, with fears, marked anxiety, and hyperactivity. The initial treatment approach should be to ameliorate the effects of those medications -- ( Celano & Geller, 1993. Bender, Lerner and Poland , 1991) rather than to ascribe them to anxiety in the mother -- and then to attempt to treat the symptoms accordingly.

The diagnostic classification should thus include such diagnostic possibilities, in the highest order of priority. These fall into three main areas: cognitive disorders, substance-related disorders, and medical conditions.

It could be argued that some of these conditions should be addressed only in Axis III (e.g., if an infant has hypothyroidism). However, the central concern is that behavioral and emotional symptoms may be only a manifestation of the medical condition, and so should be characterized as pertaining directly to it : being a part of the condition, or being caused by it. This seems a better option than to diagnose, for instance, a regulatory disorder of hyposensitive type, and then in Axis III to diagnose hypothyroidism as well. It seems more suitable for a diagnosis of a regulatory disorder to be reserved for conditions in which there is no obvious medical basis for the problem. In the case of multi-system developmental disorders , a thorough study of associated medical problems or symptoms is essential for an adequate diagnosis (Gillberg et. al., 1990).

In the next section, we describe some of the main disorders caused by alteration in brain functioning, caused by diseases, as well as by substances or withdrawal from the effects of a substance, including medications. For purposes of reference, we include many conditions and describe them in some detail.

## **A. Cognitive Disorders.**

### **1. DELIRIUM.**

Delirium is a mental disturbance that is characterized by the child's inability to sustain focused attention on one issue longer than a few seconds. Attention then shifts to another stimulus or topic. Because of this difficulty in sustaining attention, the child cannot articulate a stream of logically connected thoughts; instead, the attention focuses and is easily disturbed by irrelevant stimuli.

In addition to having problems maintaining focused attention, children affected by delirium may have perceptual disturbances and may suffer oscillations in their state of alertness, going from being hyper-alert to somnolent or with clouded consciousness. Typically, an affected child will be disoriented to his own identity, or to that of familiar people and familiar places, and so will mistake his home for his school, or the hospital for church, etc.

There are also neurovegetative changes, which may be obvious or subtle, and they can be life-threatening (Prugh, Wagonfeld, Metcalf, & Jordan, 1980). There are typical electroencephalographic changes that aid the diagnosis of this condition when it is suspected. Delirium, particularly when it is not severe and lasts for several days (Stoddard & Wilens, 1990), may be mistaken for an adjustment reaction to the stress of hospitalization, to a medical procedure, or to traumatic events. The parents may be unable to comfort the child, who cries inconsolably or is very irritable or scared. The patient may show regressive behavior, such as in speech, self-feeding, and bladder or bowel control.

Delirium is a clinical problem that tends to occur in young children and infants who are hospitalized -- especially those admitted to an intensive care unit. It can be due to stimulus deprivation, a neurological condition, an infection, or a metabolic disturbance, as well as to the adverse effects of medications, or all of the above in different combinations. One further problem faced by mental health workers in medical centers and hospitals is that delirium sometimes has subtle manifestations and may be of long duration (i.e., lasting days without being diagnosed adequately).

Several factors may contribute individually -- or concurrently -- to causing the syndrome of delirium (Stoddard & Wilens, 1990). Chief among these are some antibiotics, aminophylline, chemotherapeutic treatment, anticholinergic medications, cimetidine, prednisone, and opiates that are used to treat pain in some conditions. Other factors may be sleep deprivation, stress, and stimulus deprivation, as well as viral conditions. An electrolytic imbalance, ketoacidosis in diabetic decompensation, and hypoxia, as well as infections of other kinds, may contribute to this condition. The resulting behavioral manifestations may be misjudged as oppositional or uncooperative behavior. The clinician may not think of trying to elicit from the young child any evidence of cognitive disturbance since the parents of the child gave a history that the child is cognitively intact up to this point..

Obviously, to treat these medical conditions, it is essential not to attribute the condition only to emotional factors. Instead, by recognizing existing medical conditions and establishing a treatment plan that takes them into consideration, clinicians can better aid

the child overall.

*Case Example No. 1.* Charles, age three-and-a-half, was admitted to a large pediatric hospital because of marked behavioral change, including rage, extreme fear on being approached (even by his parents), and constant psychomotor agitation. At first, the mental health professional in charge of the consultation suspected, given the child's behavior, that the situation might be due to a stress reaction to possible traumatic events in the home, which the family denied. Further observation and careful questioning revealed that the child's level of consciousness or alertness would fluctuate, tending to become more cloudy at night. He was disoriented to time, place, and person (his own identity and that of his parents), and he couldn't count or name things on request, which he had done before at home. The syndrome of delirium was diagnosed and the child was treated with Valium. Later on, a diagnosis of encephalitis underlying the delirious state was made. Eventually, the boy recovered without sequelae.

## **2. Dementia.**

Most clinicians distinguish between developmental disorders and dementia. In developmental disorders, such as multi-system developmental disorder, or in pervasive developmental disorders (e.g., infantile autism, Asperger's syndrome, etc.), the child fails to acquire certain skills or abilities, or the development is deviated in these areas (e.g., imagination, two-way relatedness, language). With dementia, the child who has achieved normal functioning in these areas and in cognitive skills loses them after a bodily insult or disease. In practice, disintegrative disorder of childhood is an appropriate diagnosis, provided that the child had developed normally during the first two years of life (American Psychiatric Association, 1994).

Dementia can be produced by a number of conditions, including cerebrovascular accident, head trauma, brain tumors, hydrocephalus, multiple sclerosis, infection of the central nervous system (e.g., an acquired herpes virus encephalitis) (Aicardi, 1992; Gillberg, 1995), or a degenerative disease affecting the brain. It can also be caused by endocrine conditions (e.g., hypothyroidism, hypercalcemia, or hypoglycemia), as well as by nutritional deficits (e.g., in thiamine, Vitamin B 12, and niacin). It may be associated with metachromatic leukodystrophy.

The hallmark of dementia is memory loss and other cognitive skills. With very young children, it is necessary for the clinician to ask the parents specifically about difficulties in memory functioning that involve the child's failing to remember incidents from the recent past. There are often personality changes, errors in judgment (taking off from the house, reckless behavior, fearless and daredevil activities) and marked irritability or aggressiveness, with poor impulse control and catastrophic reactions when frustrated. One clue that points to the suspicion of dementia is the progressive nature of the problem and its relatively acute development of symptoms (over the course of months, with a clear onset).

These conditions do occur in very early childhood, and should be considered in the differential diagnosis of young children who have relatively clear and marked onset of behavioral or emotional changes, or who suffer cognitive deterioration. Clinicians who do not take them into account run the risk of missing the early manifestations of these disturbances, and may mistakenly ascribe them to, for instance, stressful circumstances.

*Case Example No. 2:* Recently, Michael, a three year old child was brought for consultation by his mother, who is a nurse. According to his mother, the boy had sequelae of periventricular hemorrhage after his premature birth and resuscitation (hyperactivity, restlessness, and impulsive behaviors). She was concerned that he was developing more marked symptoms of hyperactivity, inattentiveness, and aggression. A physician who had examined the child previously had advised the mother that he was reacting to the fact that his father was a diabetic and had decompensations from time to time, which created much stress for the family. On closer examination, however, it was clear that the child had lost some functioning gains such as language abilities, and that there were some subtle memory changes as well as new episodes of rage. A neurological consultation revealed hydrocephalus, which explained the child's symptoms of initial dementia.

## **B. SUBSTANCE-RELATED DISORDERS.**

These are also "organic conditions" related to the use of substances, or to the organic brain syndromes directly caused by the effects of substances or by withdrawal from them. The substances can include prescription medications, as well as street drugs, such as cocaine or amphetamines.

Unfortunately, many babies are exposed *in utero* both to medications and to psychoactive substances, some prescribed and some

abused by the mother. This exposure leads to manifestations of behavioral/regulatory disturbances in the newborn and also in the older infant. It is also well known that alcohol and other street drugs have long-term effects observable in toddlers and preschool children.

To address these problems in the diagnostic classification, we would suggest a system similar to that of *DSM-IV*. There could be a category to address the direct effects of exposure to, and withdrawal from, addictive substances, including their long-term effects (after the newborn period). Substances can affect the infant via three main routes:

(1) Direct transfer from the mother to the fetus via the placental circulation; (2) Direct exposure to the substance via oral, rectal, or transdermal absorption; and (3) Absorption via breast feeding.

In all three of these contexts, intoxication, withdrawal, and long-term effects can occur. In our opinion, it would be best, given the relatively limited knowledge about these effects, to keep the number of conditions to a minimum. Some of the common substance related situations encountered by clinicians are described below. Some detail is provided to aid clinicians in identifying them.

### **IN THE NEWBORN**

**(1) Intoxication** (e.g., alcohol, benzodiazepines) (McElhatton, 1994). Manifestations of this condition are depression of the central nervous system, hyporeactivity to stimuli and poor motor coordination. We have encountered clinical situations in which the mother drank alcohol, or cannabis during labor and the baby was born under the influence of these.

**(2) Withdrawal.** This condition is evident when the mother has used addictive substances up to the time of delivery. Many of the manifestations of Neonatal Withdrawal Syndrome (NWS) occur regardless of the class of drug. Symptoms include irritability, hyperactivity, abnormal sleep pattern, high-pitched persistent crying, tremor, vomiting, diarrhea, weight loss, and failure to gain weight (Levy & Spino, 1993). Withdrawal could be caused by the mother's use of cocaine, amphetamines, alcohol, or opiates.

This problem is commonly encountered in nursery and neonatal intensive care units. It appears to be worse when the baby is delivered at fullterm rather than when the child is premature. The infant manifests behavioral dysregulation, irritability, excessive crying, and disturbances in feeding and sleeping (Doberczak, Kandall, & Wilets, 1991).

Withdrawal from cocaine produces symptoms that have been reported to last from hours to even months after birth (McElhatton, 1994). Amphetamine withdrawal in infants whose mothers were addicted can cause symptoms consisting of shrill cries, irritability, jerking, and sneezing. Also reported were agitation (alternating with lassitude), glassy-eyed stares, and drowsiness lasting up to four days (Neuberg, 1970). The withdrawal symptoms due to opiates may be iatrogenic (i.e., from the continued use of opiates, such as morphine, fentanyl, or methadone) during treatment of a baby in the Neonatal Intensive Care Unit. They may also be precipitated by the acute suppression of exposure to the drugs at birth, in cases where the mother continued her drug use up to the time of delivery (Barr & Jones, 1994; Franck & Vilardi, 1995).

Withdrawal of tricyclic antidepressants (e.g., imipramine or chlorpramine) (Bromiker & Kaplan, 1994) may also produce similar symptoms in newborn infants.

The mother's use of other prescription drugs (e.g., fluoxetine and psychoactive substances) may also cause withdrawal-like symptoms in the infant. *In utero* exposure to marijuana, for example, has been associated (Fried, 1995) with mild autonomic dysfunction and with difficulties in state regulation.

The "addicted" newborn exhibits hyperexcitability, jerky and poorly coordinated movements, excessive crying, intense reactions to visual and auditory stimuli, and poor self-regulation of its physiological state. Other possible symptoms include poor sleep, feeding disturbance, and convulsions.

### **IN THE INFANT (AFTER NEWBORN PERIOD)**

**(1) Acute Behavioral/Emotional Disturbance.** This type of reaction may result from exposure after birth to a wide variety of prescription medications and medical treatments (e.g., corticosteroids, chemotherapy, bronchodilators, psychostimulants, anticonvulsant medications, antihistamines, and sympathomimetics).

**Corticosteroids** are widely used in treating very young children for multiple conditions, particularly allergies and autoimmune

diseases. For example, they are often prescribed for the treatment of asthma. A number of possible behavioral effects, such as agitation, motor hyperactivity, talkativeness, and silliness, can result from their use. Corticosteroids may also cause depressive symptoms, including lethargy, apathy, and tearfulness. In addition, they have been associated with marked irritability and oppositional behavior, particularly when taken by mouth (Bender & Milgrom, 1995).

**Chemotherapy** may cause hyperactivity, hyperreactivity, feeding disturbance, and sleep disturbance, as well as depressive symptoms, apathy, withdrawal, and tearfulness.

**Bronchodilators** may cause agitation, anxiety, tremor, fears, aggressive behavior, and defiance. These types of reactions appear to be particularly noticeable with beta receptor stimulants such as salbutamol (Ventolin), in comparison to theophylline, which is used more rarely as a first-line bronchodilator in young children (Bender & Milgrom, 1995).

**Psychostimulants** (e.g., Methylphenidate, amphetamines) may cause agitation, aggressive and defiant behavior, a sad expression, depressive feelings, and tearfulness.

**Anticonvulsant medications.** To begin with, the child with epilepsy has a higher risk for psychiatric difficulties. However, anticonvulsant medications, in themselves, produce a number of behavioral symptoms. Phenobarbital, for instance, causes irritability, and high degrees of motor activity and restlessness, as well as episodes of rage. It also clearly interferes with learning -- and perhaps even with memory -- by causing a short attention span. Aggression, marked irritability, and compulsive behavior are caused by valproic acid and carbamazepine (Evans, Clay & Gualtieri, 1987; Gillberg, 1995).

**Antihistamines** are sold over the counter and given to young children by parents to treat the symptoms of flu and colds. Of these, diphenhydramine (Benadryl), Astemizole, loratadine, and terfenadine may cause agitation, hallucinations (Bender & Milgrom, 1995) and delirium in some children, particularly when high doses are taken.

*Recently, we treated Jonathan, a two and a half year old child whose mother brought him to consultation due to restlessness and aggressive behavior. On questioning, after several sessions, the mother admitted that she would give him spoonfuls of diphenhydramine (Benadryl) to "calm him down"; she had thought of this approach herself. She had repeatedly requested that the psychiatrist treat her child with dextroamphetamine, as she had a nephew (older) that she described as "hyper" who took this medicine. She worked in the night shift and was the sole caretaker of her son in the mornings. She needed that the child remain very quiet or preferably sleep in the morning, so that she could sleep, or alternatively, that he play silently by himself for several hours at a time.*

*The aggressive behavior had to do with these expectations, but clearly worsened with the side effects of the antihistaminics.*

**Sympathomimetics** (e.g., pseudoephedrine, amphetamines) may cause symptoms of confusion, agitation, and anxiety, particularly in very young children. In less severe cases, sympathomimetics may cause restlessness, irritability, and disturbed sleep (Bender & Milgrom, 1995).

**(2) Long-term Effects. These effects have been linked to in utero exposure to alcohol, cocaine, amphetamines, cannabis or marijuana, and tobacco.**

**Fetal Alcohol Syndrome** is characterized by well-known dysmorphic features, such as coarse facial features, diminished cephalic perimeter, and intellectual handicap. In addition, affected infants and very young children frequently suffer from short attention span, hyperactivity, impulsivity, and aggressive behavior (Steinhausen, Gobel, & Nestler, 1984; Steinhausen, Willms, & Spohr, 1994). These effects are often accompanied by sensory hypersensitivity to the surrounding environment (Weinner & Morse, 1994). Language development may also be delayed in affected children.

**Exposure to cocaine in utero** causes short attention span, hyperactivity, aggressive behavior, and learning difficulties (Fox, 1994) in the long term.

**Exposure to amphetamines in utero** produces manifestations similar to those resulting from the mother's cocaine use (e.g., short attention span, hyperactivity, aggressiveness, and learning difficulties) noted years later.

**Exposure to cannabis in utero** also can result in a short attention span. Other deleterious effects include concentration difficulties

and learning problems (Fried, 1995).

### C. MEDICAL CONDITIONS.

It is essential for clinicians to have several possible diagnoses in mind so as to not overlook a medical condition that might manifest itself via behavioral or emotional symptoms. Taking this approach is helpful in working not only with newborns but also in dealing with any young children under four years of age.

Clinicians should keep in mind that many different medical conditions produce signs and symptoms of behavioral/emotional disturbance that may derail the diagnosis and result in inappropriate or delayed treatment. These illnesses may range from more serious, lifelong conditions such as epilepsy and hypothyroidism to those that surface primarily in the early developmental stages (e.g., infantile spasms, fragile X syndrome).

**Epilepsy.** In the young child, petit mal seizures, in which the child appears "absent" or "gone" for periods of several seconds at a time, and "disconnected" from the surroundings, can be mistaken for attempts in the child to avoid the painful feeling of a memory or for dissociation. In some children, this can occur many times a day and can be mistaken as a reaction to anxiety-producing situations or stressful circumstances. The EEG reveals a typical brain wave pattern of spike and wave (three cycles per second) that is indicative of the underlying problem.

*Case Example No. 3: Recently, Mary, a nine-month-old girl, was referred for consultation at the request of the staff of her daycare center. The initial concern was that her adolescent mother appeared rather indifferent toward the baby. The mother complained also that her own sleep was disrupted because the baby had an extremely "restless sleep," which was initially considered as a manifestation of an overall tendency to be restless and dys-regulated. A videotape of a nap the child took at the daycare center illustrated her restless sleep patterns. The consultants could see that the child exhibited myoclonic rhythmic discharges that occurred only during sleep. A subsequent neurological consultation confirmed the presence of a seizure disorder.*

**Infantile Spasms** (e.g., West Syndrome, Salaam seizures). It is well known that the condition of infantile spasms is often misdiagnosed as colic in the very young infant (Holmes, 1992). When the convulsions are infrequent – usually in the beginning -- the child with this form of seizure disorder (i.e., myoclonia in which there is sudden flexion of the trunk, arms, and legs) appears to curve its body in response to some sudden stimulus, interpreted by the parent as colic. In our experience, this condition is not thought of frequently, and the child is misdiagnosed as having colic. This disorder appears typically in the first two years of life, with its peak of onset between four and six months (Holmes, 1992). The treatment generally includes the use of medication (i.e., adrenocorticotropic hormone).

**Hypothyroidism** is a possibility that should be considered in children who are excessively passive, under-reactive, or "floppy." Screening tests in the newborn are done in some countries, but not many. Prevalence of this disorder is approximately one in every 3,000-4,000 births (Ilicky & Larsson, 1991) with it occurring twice as frequently in boys as in girls. A child with this condition exhibits some of the features of regulatory disorders of the under-reactive type, such as being very placid and non-reactive. The child may appear socially withdrawn or depressed. In less severe cases, there are problems with motor coordination, attention span, and perceptual peculiarities that can lead to learning problems. A diagnostic work-up can determine whether an infant has the condition, which is treatable, thus preventing negative effects in intellect and overall development. Despite prompt treatment, there are learning difficulties later on in about 25% of all cases. Hypothyroidism has also been associated with autism (Gillberg, 1995).

**Fragile X Syndrome.** This disorder is more frequent than previously thought and can easily be overlooked in infants. It has been estimated that one in every 400-500 people may have a carrier status (Goldson & Hagerman, 1992). After Down's syndrome, it is the second most common chromosomal cause of mental handicap in children. In infants, it is characterized by hypotonia, with the affected child being very difficult temperamentally. The infant is very sensitive to stimuli, easily overwhelmed, and quite irritable. Episodes of intense crying may alternate with temper tantrums. There may also be failure to thrive because of unexplainable problems or feeding difficulties. Sleep difficulties are also common. When the child is older, difficulties with attention span and hyperactivity may surface. When fragile X syndrome is diagnosed in the preschool years, folate treatment might help alleviate the hyperactivity (Hagerman, 1991). In more serious cases, there may exist a multi-system developmental disorder or even autistic disorder. This syndrome can also occur in girls, and it can be present in children with normal cognitive functioning (Mazzocco, Hagerman, Cronister-Silverman, & Pennington, 1992).

**Brain Tumors.** Although these are relatively rare in infancy, their initial presentation may be only changes in the child's mood or behavior, only later leading to suspicion of a neurological condition. A recent analysis of 74 cases of children with tumors( Edgeworth, Bullock, Bailey, Gallagher and Crouchman, 1996 ) revealed that in 47% of cases the initial manifestation for periods of weeks or longer were these behavioral changes. These included the child becoming irritable, disrupted sleep pattern, excessive crying, needing more attention in the child before two years of age. Between two and three years of age, they had disruption in their eating habits, temper tantrums, and irritable behavior. The clinician should keep these possibilities in mind when assessing such behavioral changes in a young child.

**Central Nervous System Infection** (e.g., encephalitis ortoxoplasmosis). The child has marked behavioral changes, may appear irritable, aggressive, out of control, or markedly passive and apathetic. Later on, the condition may lead to intellectual deficit, learning disturbance, language disorder, or autistic disorder (Rutter & Bartak, 1971). The problem may be manifested initially only by language difficulty, leading to echolalia, social withdrawal, and changes in behavior (e.g., hyperactivity, episodes of rage, and poor judgment) (Aicardi 1992; Gillberg 1995). The relatively acute onset of these symptoms in a previously normal child should raise suspicion about the possibility of encephalitis. Adequate treatment could prevent sequelae that include autistic disorder (DeLong, Bean, & Brown, 1981) or dementia.

**HIV Infection.** The encephalopathy caused by HIV infection is considered the most common cause of dementia in childhood (Goodman, 1994). Neuronal damage seems to occur through the "innocent bystander" phenomenon(Thompson, Westwell, & Viney, 1994), that is, the negative reactions of neighboring neurons to the destruction of immune cells in the brain. The child may have a congenital infection or acquire one later. Manifestations may include failure to develop skills or the actual loss of skills, as in true dementia, in which the child loses previously acquired abilities (Goodman, 1994). This condition is of great epidemiological importance. The World Health Organization has estimated that in 1992 there were 600,000 children worldwide infected with the AIDS virus(Chung & Grinker, 1992 ). The manifestations of HIV infection affecting the central nervous system include developmental delays that initially may be subtle. The child may also have problems in motor functioning and in sensory abilities, such as visual spatial perception problems (Aylward, Butz, Hutton, Joyner, & Vogelhut 1992). As early as two months, the signs of delay may be present that can lead to apathy, social withdrawal, feeding and sleeping disturbance, and intellectual deterioration. The initial manifestation may be failure to thrive (Thompson, Westwell, & Viney, 1994).

**Diencephalic Syndrome.** This symptom complex may be caused by tumors of the thalamus, hypothalamus, and pineal region. They are fortunately rare. Symptoms start very early in life, but the disorder is usually diagnosed after the first year. In the case of mid-line gliomas of childhood (tumors that occupy the third ventricle), the manifestations are of diencephalic syndrome. This consists of failure to thrive and feeding difficulties, such as food refusal, leading to the "concentration camp syndrome" of emaciation (Cohen & Duffner, 1992).

**Phenilketonuria.** This condition is relatively rare, but its early diagnosis can lead to treatment that may prevent a number of subsequent dysfunctions. Its prevalence in the United States is one in 15,000 births, but in some countries it is much more frequent (Aicardi, 1992). Infants are not routinely screened for this condition worldwide. Initial manifestations may include irritability and refusal of feedings, which can lead to poor weight gain, as well as repeated vomiting. The baby may appear hypertonic, exhibiting rhythmic rocking and tremor of the hands. Or there may be considerable hyperactivity and uncontrollable temper associated with multiple behavioral problems (Nyhan, 1992). The child may manifest slow psychomotor development and social withdrawal. In some cases, the only manifestation is a regulatory disturbance, with high levels of motor activity and a short attention span.

**Duchenne muscular dystrophy.** Like other congenital muscular dystrophies present at birth, this condition may be manifested only later by muscle weakness, body floppiness, and difficulties in sucking and feeding (Dick, Bertorini, & Igarashi, 1992).

This condition may not be recognized in the infant or very young child, nor even suspected at all until much later. There are abnormalities in motor development, but these may be only mildly delayed. The disorder may manifest itself by the second or third year of life via retarded social development, social isolation, and depressive symptoms.

**Neurofibromatosis.** This condition may go undiagnosed in the young child for a long time. It consists of a tendency to form tumors in the central nervous system, skin, and internal organs. Its prevalence in the general population is one in 3,000 (Riccardi & Eichner, 1986), but milder forms are thought to be much more frequent, often going undiagnosed. Two subtypes, I and II, are linked to alterations in chromosomes 17 and 22, respectively. The characteristic *cafe au lait* spots on the surface of the skin may not be evident in the young child. Direct DNA diagnosis is available for the type I variety. It may be characterized by disorders of motor

control or perception, and hyperactivity. Also, a multi-system developmental disorder may also characterize the type I variety (Gillberg, 1995).

**Williams Syndrome.** This relatively rare congenital disorder is identifiable by a characteristic facies, cognitive deficits, and supravalvular aortic stenosis, sometimes with hypercalcemia. The facial features are thick and include large lips, a long philtrum, and an upturned nose with depressed nasal bridge. There may be epicanthic folds, ocular defects, strabismus, and a preponderance of blue eyes with a stellate iris pattern. Affected children are often born with below average weight and commonly experience early feeding problems (Martin, Snodgrass & Cohen, 1984), such as sucking difficulties, frequent regurgitation, or even vomiting (Meyerson & Frank, 1987). These feeding problems may lead to a failure to thrive. Older children with this disorder may engage in cheek biting and cheek sucking. Later on, there may be difficulties in chewing and swallowing food. The voice is described as characteristically of low pitch and hoarse, metallic, or peculiar. Older children may be hypersensitive to sounds or may exhibit hyperacusis, crying and screaming in response to noises, or attempting to cover their ears (Arnold, Yule & Martin, 1985). In the preschool years, there is frequently loquacity and the child has very long utterances (ten - 13 words per utterance, on average). There is a characteristic "cocktail party syndrome" to these expressions, in which the child just talks in a disconnected way, using irrelevant words. Later on in life, there may be noticeable hyperactivity, attention deficit, and autistic-like features of symptoms of multi-system developmental problems.

**Angelman Syndrome.** This disorder was previously considered very rare, but it is increasingly being reported in the literature, as it is recognized earlier in life, especially in infancy (Van Lierde, Atza, Giardino, & Viani, 1990). The earlier the diagnosis, the more rehabilitative interventions can be implemented (Yamada & Volpe, 1990). Although infants may have a normal physical examination, the disorder may be suspected when there are prominent jerky movements, marked instability even when sitting, and feeding difficulties. The abnormal facial features (e.g., wide mouth, prominent mandible, and micro-brachycephalia) may become prominent only much later. The feeding problems consist of frequent spitting or food refusal, as well as chewing and swallowing difficulties, that may lead to a failure to thrive. The affected child may also need little sleep. Later on, mental retardation and seizure activity become evident. Striking and characteristic episodes of unprovoked laughter (which resulted in a nickname of the "happy puppet syndrome") tend to appear between 16 months and three years of age (Van Lierde et al., 1990).

**Sotos Disorder.** This rare disorder, associated with developmental delay, involves early maturation of bones and accelerated growth in early childhood, as well as distinctive facial features (i.e., gargoyle features). The behavioral profile of these children comprises language abnormalities, frequent temper tantrums, attention deficit, destructiveness, difficulties in social relatedness, and marked eating and sleeping difficulties (Dodge, Holmes, & Sotos, 1983; Rutter & Cole, 1991).

#### **Agenesis and Dysgenesis of the Corpus Callosum (e.g., Aicardi Syndrome).**

Agenesis of the corpus callosum may be found through more than 2% of all CT scans, which are usually indicated to diagnose neurological or behavioral difficulties (Gillberg, 1995). There are two forms, symptomatic and asymptomatic. The symptomatic type affects girls only and leads to Aicardi syndrome, which includes infantile spasms, mental retardation, complex partial seizures, and ocular malformations. The "asymptomatic form" is manifested via behavioral disturbances. In the three- to four-year-old child, these behavioral problems may appear as difficulties in relating and communicating, and as explosions of anger or rage; in the older child, there may be obsessive-compulsive behavior. These behaviors are often accompanied by seizures, macrocephaly, and hypertelorism (eyes more separated than normal).

**Menkes' Kinky Hair Syndrome (trichopoliodystrophy).** In our experience, this condition is often misdiagnosed early on. The child may be admitted to a pediatric hospital due to failure to thrive; the pale skin, puffy appearance, variations in temperature, and strikingly peculiar hair may make clinicians suspect that the child is being neglected and maltreated. Instead, the problem is a neurodegenerative disorder that eventually causes psychomotor deterioration and seizures. At the beginning of the disease, the child may exhibit early feeding difficulties and irritability. Phenomena-like seizures, and quadriplegia may appear later (Maertens & Dyken, 1992).

#### **Summary**

These conditions are only some of those that could easily be overlooked, and whose symptoms might be mistakenly assumed to have an emotional or relational basis. Of course, in many cases, there is the possibility that the symptoms may be a superimposition of underlying substance-related or medically caused problems, intertwined with emotional and behavioral reactions to stressful

circumstances or relational problems.

## V. REGULATORY DISORDERS

The regulatory disorders have received much attention recently, especially from clinicians. Occupational therapists have contributed greatly to the clinical description of disturbances in sensory abilities, processing difficulties, motor and coordination problems.

An obvious problem is that the diagnostic criteria included in the Zero to Three Classification only identify "hyper or hypo" sensitivity or reactivity in the various sensory channels as being problematic, and then only when it is coupled with a behavioral disturbance. One problem is that there are no thresholds to define when there is hyper or hyposensitivity in a given sensory channel. For instance, when should a clinician decide that a child is too sensitive to noise or to visual stimuli without this being a too individualized decision? To ascertain whether the child has a problem in these sensory modalities, we can rely on a number of symptom checklists used by occupational therapists and other clinicians (Anzalone, 1993; DeGangi & Poisson, 1995; DeGangi, Wietlisbach, Goodin, & Scheiner, 1993).

Clinicians should be advised to not make a diagnosis solely on the basis of a parental report obtained on a symptom checklist. Many parents are unfamiliar with what is "normal" in terms of a child's sensitivities. For instance, a very stressed mother might believe that her normally exploratory toddler is "hyperactive" or is "on the go all the time," because she (the mother) is exhausted and perceives a normal level of activity as being pathological. Clearly, it would be advisable to obtain specific examples of what the parent means, and also to conduct a firsthand examination of the child.

Regarding the symptom guidelines, it is our impression that many behaviors manifested by normal children might easily be assigned a pathological quality. Chief among these are calming difficulties, special sleep requirements, demanding adult company, inability to wait, cravings, overreacting to loud sounds or strong visual images, not wanting to have eyes covered, using smell to explore surroundings, pica, coordination difficulties, not wanting diaper to be changed, and sensitivity to temperature.

**Difficulty calming when talked to from a distance.** Many infants will not calm down when talked to from a distance, so this should not be considered a symptom. If the item read, "never calms when talked to," or "always requires approaches other than talking," it would narrow the identification process and be more appropriate.

**Need for special conditions to fall asleep** (e.g., rocking, stroking hair, audio tapes). We teach parents that they are well advised to create rituals to facilitate the transition of their child to sleep. It, therefore, seems puzzling that such conditions might be considered indicative of a problem. The implication also is that the child should not need such conditions and should be able to go to sleep by himself or herself, which does not take into account other factors.

**Demanding adult company constantly.** This item is somewhat confusing. Infants, who admittedly should be under constant supervision rather than left to themselves, normally seek proximity to the care-taking figure most of the time.

**Difficulty in waiting for what she or he wants.** This criterion appears too broad. Even very normal toddlers have difficulty waiting for what they want. It might be more useful to state that the child has unusually limited patience or is extremely demanding, or that he or she almost always "falls apart" immediately.

**Craving certain foods or activities.** Children (and adults, too, for that matter) prefer certain foods and activities. It might be best to specify a very restricted range of food preferences, such as fad foods, or a grossly restricted range of activities.

**Reactions to loud sounds.** This type of reaction seems typical for young children. Perhaps the item should be restated to qualify (e.g., an abnormally intense or unusual reaction to loud sounds).

**Objection to having vision occluded.** In our experience, infants normally do not like to have their eyes occluded, other than very briefly within the context of play.

**Gets excited by complex visual images.** This would seem to be a normal response in young children or in anyone, for that matter.

**Uses smell to explore objects or environments.** In itself, this is not an abnormality. Everybody explores things through smell and other sensory channels. One would suspect that it would be considered abnormal for children to always approach every new thing first by smelling it, or just to be intensely sensitive to smells. This item could be made more specific by identifying the extent of

abnormality more specifically.

**Chewing on non-edible objects.** A qualifying word is needed in this item to indicate when this behavior crosses over into the abnormal. Practically all children, particularly young infants, mouth or chew on objects from time to time. This criterion should be more restrictive by referring to children who are constantly chewing on non-edible materials.

**Difficulty in riding bicycle or using gym equipment.** This activity does not seem pertinent to toddlers, as many children learn to ride a three-wheeler (tricycle) or a two-wheeler (bicycle) at a much later age.

**Dislikes having diaper changed or being wiped.** This criterion would be more appropriate if it included restrictive language such as an "intense negative reaction." Such terminology would indicate that the infant *always* cries or protests when the diaper is changed. In reality, many infants do not "like" to be wiped.

**Is sensitive to temperature of food or water.** Restricting this criterion to include an unusual sensitivity to temperature, rather than simple dislike, would be preferable. In our opinion, a dislike of mild, hot, or cold items does not, in itself, constitute an abnormality. A marked avoidance of, or preference for, a certain temperature would be more indicative of a problematic situation.

The diagnostic guidelines in these areas lead to the danger of identifying children as "too sensitive," which would include most children. Some further qualifying statements would no doubt help clinicians determine if a child is clearly abnormal in a particular area. Also, there are no guidelines as to how many excessive or deficit features a child must have to qualify for certain diagnoses.

It would be most desirable if agreement could be reached about how many abnormalities or symptoms should be present, and in how many areas, in order for the diagnosis to be made. Some clinicians might be content with being able to identify symptoms in two areas, while others would require more.

## VI. COMORBIDITY.

Having praised the general recommendation that clinicians should try to encompass the child's or the dyad's problem with one diagnosis only, there should be room for exceptional circumstances.

*Case Example No 4: Recently we consulted on a 20 month old infant, Joseph, who had several of these problems. He had had a near-death experience after drowning in the swimming pool at home. Shortly after this experience, he developed nightmares, a number of fears, temper tantrums, and aggressive behavior. He was diagnosed as having a traumatic stress disorder. However, on closer examination, it appeared obvious that even before the accident, the child had been quite hyperactive, had sought out intense stimuli (e.g., loud noises), had had an extremely short attention span, had cried excessively, and had been very sensitive to tactile stimuli, hot temperatures, and visual stimuli. Thus, the clinical picture was one of a regulatory disorder that predated the drowning accident. Afterward, the manifestations of the stress disturbance were superimposed on the child's preexisting difficulties. To capture the complexity of this child's problems, it would have been more useful to consider both diagnostic entities as coexisting rather than to designate one over the other.*

## VII. ATTACHMENT DISORDERS.

The diagnostic classification includes a chapter on Reactive Attachment Deprivation /Maltreatment Disorder of Infancy (pp. 29-30). The description in this area leaves the impression that a history of poor parental care or a change in caretakers is almost sufficient alone to warrant this diagnosis. There is no phenomenological description of what symptoms should be present in the child in order to diagnose an attachment disorder. This is unfortunate, because it would be difficult to distinguish this type of disorder from the relationship disorders described elsewhere in the system as under-involved and abusive. A more detailed description of the symptoms that clinicians should look for might be developed from an examination of the work of Lieberman and Zeanah (1995) and Zeanah and Emde (1994). The presence of an Axis II diagnosis that describes current abnormalities in relationships is not sufficient, because children might have an attachment disorder as a consequence of a previous (rather than a current) abnormal relationship with a caretaker.

Finally, a somewhat surprising point is the placing of gender identity disturbances in the section of mood disorders. In the traditional nosology mood or affective disturbances do not include aspects like gender identity disorder.

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## **APPENDIX**

We offer some suggestions on a number of conditions that might be included in the classification, in order to help clinicians rule out some of the conditions mentioned above, before considering a purely stress-related, behavioral or emotional condition in an infant.

### **COGNITIVE DISORDERS**

1. **Delirium**, caused by: Medication or substance, stimulus deprivation, medical condition, or by several etiologic factors concurrently.
2. **Dementia**, caused by: Infection, HIV infection, cerebrovascular accident, or other medical condition.

### **SUBSTANCE-RELATED DISORDERS IN NEWBORN**

#### **(1) Intoxication**

- Alcohol
- Benzodiazepine
- Cocaine

#### **(2) Withdrawal**

- Cocaine.
- Amphetamines
- Alcohol
- Opiates
- Antidepressants
- Other (may include fluoxetine, other psychoactive substances)

### **SUBSTANCE-RELATED DISORDERS IN INFANT**

#### **(1) Acute Behavioral/ Emotional Disturbance**

- Corticosteroids
- Chemotherapy
- Bronchodilators
- Psychostimulants
- Other

#### **(2) Long-Term Effects**

- Fetal Alcohol Syndrome.
- Exposure to cocaine in utero.

- Exposure to amphetamines in utero .

- Exposure to cannabis in utero

- Other

**Behavioral/Emotional Disturbance Due to a General Medical Condition (i.e., a condition of General Medicine): Name of the condition, e.g.,**

**Hypothyroidism**

**Fragile X syndrome**

**Epilepsy**

**Central Nervous System Infection**

**HIV Infection**

**Phenilketonuria**

**Duchenne muscular dystrophy**

**Williams Syndrome**

**Angelman Syndrome**

**De Sotos Syndrome**

**Aicardi Syndrome**

**Menke's Kinky Hair Syndrome**

**Neurofibromatosis**

**Other medical conditions**

**SLEEP DISORDER DUE TO EXPOSURE TO SUBSTANCE**

**SLEEP DISORDER DUE TO MEDICAL CONDITION**

**FEEDING OR EATING DISORDER DUE TO EFFECTS OF SUBSTANCE**

**FEEDING OR EATING DISORDER DUE TO MEDICAL CONDITION**

**REGULATORY DISORDER DUE TO EFFECTS OF SUBSTANCE**

**REGULATORY DISORDER DUE TO MEDICAL CONDITION**