# 14 A Diagnosis of Exclusion: Demystifying Neurodevelopmental Disorder Associated with Prenatal Alcohol Exposure

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## Case

Jacob is an 8-year-old Russian-born adopted boy presenting for a consultation for mood outbursts and aggressive behavior at home, school, and in the community. From his appearance, Jacob did not fit the textbook criteria of a frail child with "dysmorphic" facial features of prenatal alcohol exposure. Since the features change through development, his parents brought a picture of him as a baby just after being adopted. The picture showed a stark contrast. He appeared to have lonely, distant eyes and was barely fitting into 6-month clothing at 18 months. Since that time, he ate well and gained weight rapidly and was well-developed by the time of his visit.

Perceptive, comfortable, and open yet cautious at times, his favorite activities during psychotherapeutic sessions were toys, art supplies and doll house, moon sand, building blocks, farm animal figures, and army men. He often asked off-topic questions such as "What's your favorite sports hero?" Jacob was concrete, conversant, spontaneous, funny, and light-hearted with 4-year-old bathroom humor scattered throughout the discussions. His face lit up as he asked, "Do you help kids that pee on them self? Or can't sit still?" He skirted around any direct references to himself, changing the subject or distracting himself further in the play activity. When asked directly why he was there, he jumped up and scattered a handful of sand and army men, shoved an oversized chair out from the wall, and climbed behind it. He spent most of the session crouched under a makeshift tent from a quilt draped over the chair, vacillating between sucking his thumb and yelling, "I hate this place! I'm going to break this house! I want to go home!"

Jacob had experienced low frustration tolerance and an easily triggered flash point from an early age. He was in a Russian "baby home" from birth, having been placed there because his alcoholic mother was found unfit by the

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government child welfare agency. When he was adopted at 15 months, he was the size of a 6-month-old baby. He was found to have had a number of medical issues, prematurity, low birth weight, failure to thrive, and developmental delay which was said to be due to lack of stimulation. He was left in the crib for such extended periods of time that he developed torticollis, a chronic muscle spasm of the neck causing his head to tilt to one side. After arriving in the USA, he began to receive occupational therapy though with limited improvement. He attended a private day care and preschool program but had to repeat kindergarten and was failing first grade by the time of his evaluation. Though he struggled on a number of different levels, his school said that he was capable of grade-level work.

Transitions had always been hard for Jacob, especially around loud noises and in crowded settings. Seemingly minor triggers tended to unravel Jacob, leading him to "fight or flee" to get out of the situation. He also had difficulty appreciating the boundaries of personal space, at times leading to others getting hurt. For example, swinging his book bag around at the bus stop may lead to him accidentally hitting another child. Peers and teachers often thought he did it on purpose, and he would be scolded. Jacob would feel so anxious and ashamed that he would have a meltdown when he got home that day.

Since Jacob is a very sensitive and perceptive boy who is eager to please his parents and other adults, their reprimands led to him feeling "bad" and different from other kids his age. On some occasions, his parents would get a glimpse into his internal conflict. He would say, "I like Miss A, she helps me learn…I like reading, science, math, art, music, or gym…I don't like getting in trouble, but when I get mad, I can't control myself…I know it's my fault when I get in trouble…I want to have a good day…I want to be with my sister and go to [aftercare]…"

At school, Jacob had become physically aggressive toward himself, teachers, and classmates, quickly escalating and becoming defiant and disrespectful. Often, this oppositionality would lead to loud, verbal confrontations with the teacher in which Jacob would rip up his work, draw on his desk, and shout "I'm stupid!" or "I'm a baby!" He would begin scratching his arms, face, and forehead. He made frequent, threatening statements – "I will push [item] off your desk!" "You need to shut your mouth and quit talking!" "I am going to scratch my face!" "I am going to the lunchroom and start a food fight."

At home, he would demonstrate physically regressive behaviors, curling up in a ball on the floor in a fetal position, kicking to the point of his shoes coming off, and disregarding others who were speaking to him. Jacob's mood shifted, switching from "attentive to disruptive in a flash." He would also say things like "I want to turn into a giant and crush the school and burn it down!" These periods of upset would require anywhere from 30 to 40 min to de-escalate. He often would complain afterward, "my head hurts" and "I can't control how I act." His behaviors transformed into whining, making crying noises, and even farting in the aftermath of one of these explosive episodes. Despite the intensity of the incident, he rarely would apologize for his behaviors and would move on as though nothing had happened. His family

members and anyone around him would feel terrified during the events and traumatized afterward. He often confabulated to others that his parents would lock him out of the house or physically harm him by kicking or slapping, often acting out the action to whomever was listening.

Based on school reports and discussions with teachers and administrators, Jacob was performing on grade level and therefore did not qualify for extra services at school. However, he was perceptive enough to recognize that other children finished assignments much faster than he did and were reading and completing math problems accurately. Jacob felt uncomfortable, frustrated, and misunderstood at school and unable to please his parents at home. Daily negative reports from school and his out-of-control behavior at home left his parents feeling inept and hopeless. At a crossroads in understanding his challenges, they began to lower their own expectations for him emotionally and academically, recognizing that they needed different strategies to parent.

## Diagnostic Assessment

Jacob was assessed to have neurodevelopmental disorder associated with prenatal alcohol exposure (ND-PAE) under other specified neurodevelopmental disorder (ICD-10 Code F88). The Diagnostic and Statistical Manual 5th Edition lists ND-PAE as a diagnosis to explain the complex neuropsychiatric, cognitive, behavioral, social, language, communication, and other multisensory deficits associated with maternal alcohol use during pregnancy. Like autistic disorder, symptoms mimic a myriad of neuropsychiatric conditions [1].

Evaluation included a 24-h EEG which showed nonspecific background abnormalities and an MRI which showed thinning of the corpus callosum. He was started on lamotrigine and clonidine to help control impulsivity and hyperactivity. Ultimately, Jacob was hospitalized in a specialty neurobehavioral unit specializing in children and adolescents with neurodevelopmental issues such as ND-PAE. Eventually, after several failed attempts to place Jacob in a variety of public school settings, he was placed in a therapeutic residential school.

Just prior to his admission to the residential school, Jacob quickly began telling about the many things he hoped would be there, a go cart track, a Native American campsite and tool-making center, a tree house, and on and on. Paradoxically, Jacob, who had been so out of control, was able to sit calmly drawing and labeling his vision of the "blueprint" for the school that he was excited to attend.

Adaptive functioning continued to be problematic and was a focus of his comprehensive treatment approach in the residential setting. Activities of daily life including bathing and hygiene were still challenging, and he was socially immature compared to his peers. He had difficulty understanding nonverbal cues and struggled with being gullible and easily influenced by others. Academically, by this point, he was found to be far behind his peers in math, writing, and reading and was found to have an FSIQ rating of 56.

**Clinical Pearl**
Infants who are poorly regulated in their sleep/wake cycle, are difficult to soothe or comfort, or have hyper- or hyposensitivity to light, sound, taste, temperature, textures, or other experiences often have underlying neurodevelopmental issues. It is common for children with undiagnosed ND-PAE to have been expelled from preschool more than once, to be labeled with "behavioral issues" in elementary school, to get into fights with peers and have oppositional behaviors over homework or chores, and to have academic failure or juvenile delinquency. Externalizing, defensive, oppositional, defiant, maladaptive, and acting-out behaviors may occur, particularly when the child is frustrated, conflicted, indecisive, or confronted with a difficult task. For patients presenting with such a complex array of issues, ND-PAE should be high on the differential diagnoses.

## Neurodevelopmental Sequelae of Fetal Alcohol Spectrum Disorder (FASD)

By some estimates, up to 10% of US grade school children have FASD/ND-PAE [2]; clearly many more children are affected than are diagnosed. The main reasons that children like Jacob are not identified with ND-PAE are that the birth history is unknown or that mothers may not admit use during early stages of pregnancy. Like other individuals with ND-PAE, Jacob likely had brain damage from prenatal alcohol exposure given his mother's history of heavy drinking during pregnancy.

The degree of effects of ND-PAE depends upon a mother's nutrition status, genetic predisposition, other lifestyle behaviors (e.g., smoking cigarettes, recreational/illicit substance use), stress level, and medical issues. The timing, duration, frequency, and maternal alcohol concentration also contribute to the range and degree of deficits, with earlier and binge exposures possibly associated with worse outcomes. Extrinsic factors, such as witnessing or experiencing abuse, neglect, loss, and other trauma may further impact neurodevelopment, increasing vulnerability to "fight or flight" reactions.

Prenatal alcohol exposure likely caused Jacob's brain to be "wired differently" as compared to a typically developing child. One way to understand this altered connectivity is to consider renovation of an old 1800 era manor house with electricity installed in the late 1930s. The electric wires will likely need to be routed in different directions because of the placement of support walls that were never intended to be hollowed with cables passing through them. Jacob's brain had connections that were affected by switches not biologically programmed to control those areas. During their migration in early pregnancy, brain cells lost their ability to plot their course and steer in the right direction, leaving them in places they were never intended to develop.

Jacob's poorly wired nervous system impaired his ability to function in a world tolerable to most 6- to 8-year-old children. His low frustration tolerance, lack of social skills, limited cognitive abilities, and hypersensitivities to noise, smells, and textures left gaps in his basic life skills. His emotional, cognitive, language, and social skills were more consistent with a 2- to 3-year-old child than an 8-year-old child.

## Reflections from the Family

Until school age, Jacob's parents were unaware of the degree of his disability, having been told when they adopted him that he was growth deficient and had health problems because he had been in an orphanage for the first 15 months of life. The adoption agency said he would "catch up" on his milestones and gain weight once he had gotten proper love and nutrition. They believed that their son was a survivor, possibly more resilient than most other children, and they were determined to help him thrive by giving him a loving, nurturing home environment as they had been told would help him overcome his harsh beginnings.

The family has vivid memories of bringing Jacob home with legs so weak he could not stand at 15 months and of being too small for size 6 months baby clothes. They also remember his anxious feeding habits, gulping down mouthfuls of food as though he would not have another meal.

Jacob's mother vividly remembers the intensity of his violent rages, uncontrollable crying spells, and other significant emotional problems during his early years. Jacob's current living situation in a pastoral environment learning life skills rather than being in a diploma track school setting appears to be much better suited to his neurodevelopmental level. He is proud of working in the cafeteria where he is learning to assist the staff with preparing simple meals, setting the tables, prepping vegetables, and doing minimal chores in lieu of the pressure toward high level math or reading novels.

## Four-Domain Model of ND-PAE

A four-domain model of ND-PAE depicted in Fig. 14.1 provides a structure to understand the complex array of neurodevelopmental issues potentially affected by prenatal alcohol exposure: emotional regulation, social communication, neurocognitive functioning, and motor/coordination/sensory function. The overlapping areas indicate that individuals with prenatal alcohol exposure can have one or more domains of impairment.

Using this approach, individuals suspected of ND-PAE would be referred for the following assessments: neuropsychological testing to rule out neurocognitive issues, speech/language screening to understand their social communication problems, and occupational/physical therapy to identify and treat underlying fine/gross motor

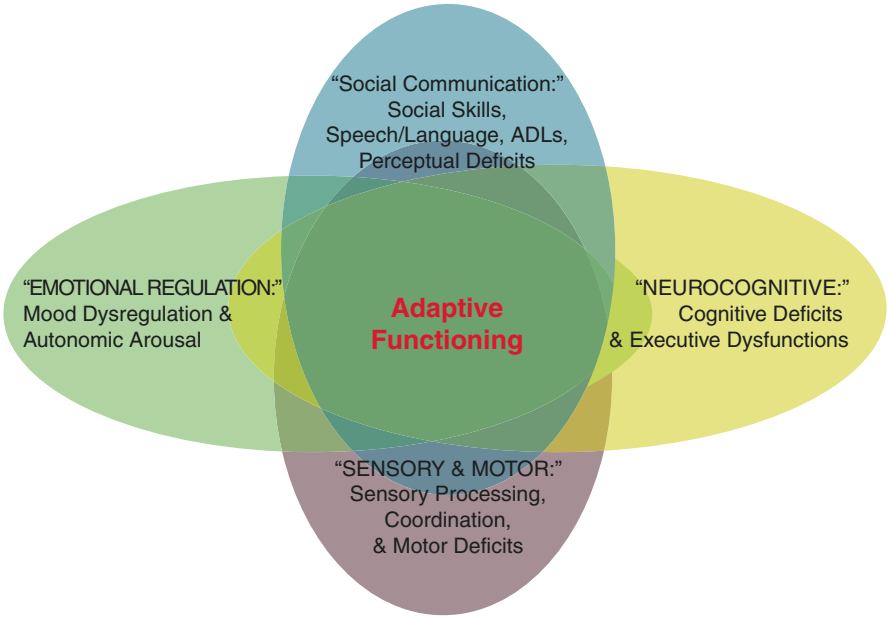


**Fig. 14.1** Domains of ND-PAE for Treatment Planning

deficits or other functional issues. Mood regulation and autonomic arousal would ideally be assessed by a physician. This team of professionals would work with the individual and family to develop a comprehensive treatment plan to enhance strengths and build vocational skills and self-esteem through mentoring, apprenticeships, and experiential learning.

## Emotional Regulation: Mood Dysregulation and Autonomic Arousal

Children with ND-PAE, compared with neurotypical children, may be less able to adapt to their surroundings, accept responsibility, identify social cues, demonstrate appropriate behavior, and bond with peers. Given the nature of their impairments, they may lack self-discipline, be short-sighted and impulsive, easily be swayed by peers, and overreact to stressors. Described by their parents and caretakers as "moral chameleons," it is no wonder that 60% of children with FASD ultimately encounter legal trouble [3].

Often, the effects of alcohol on the central nervous system (CNS) produce a highly mood-dysregulated child, having seemingly random or easily provoked episodes of frustration, irritability, aggression, and anger. Infants and toddlers with ND-PAE can present with Regulatory Disorder Type I, II, or III [4]. This may lead to infants and toddlers seeming to be easily agitated, overstimulated, and hyperaroused. This leads to impulsive aggression and physically lashing out during stressful or emotionally charged situations.

Many are prone to mood dysregulation that, on the face of it, may appear to mimic bipolar disorder and be characterized by intermittent explosive outbursts and episodes of rage triggered by the slightest insult, sideways glance, or annoyance. As a result, they are often more vulnerable to emotional overreaction, poor frustration tolerance, hypersensitivity to criticism, and suspicion about both positive and negative stimuli and may be overwhelmed by apparently minimal life stressors.

Such altered "arousal patterns" are associated with difficulties settling to sleep. Altered or disrupted sleep has been described by many parents and caregivers of children with ND-PAE. At the same time, few studies have been conducted to tease out the differences between typically developing children's sleep disorders and those of children with ND-PAE [5]. In animal models, prenatal alcohol affects the circadian rhythm in the hypothalamus by changing the metabolism in pro-opiomelanocortin (POMC)-producing neurons as well as the expression of "clock regulatory genes" [6].

## Neurocognitive: Cognitive and Executive Dysfunctions

Individuals with ND-PAE may have deficits in a variety of cognitive domains, including but not limited to fronto-executive dysfunction (organization, concentration, processing speed, working memory, problem solving, attention, impulse control, etc.), intellectual disability, or specific learning disabilities. These disruptions in cognitive functioning often lead to a failure to understand consequences and limited insight.

## Social Communication: Language, Social Skills, and Perceptual Deficits

A variety of speech and language and related socialization disabilities can also be seen in individuals with ND-PAE. The misuse of language integral to social cognition and communication are quite common problems in adolescents or young adults with ND-PAE. At times, these patients are misdiagnosed with autistic spectrum disorder or Asperger's syndrome [7]. Individuals with ND-PAE may have indiscriminate or immature behaviors. Behavior problems range from silly or irritating socially inappropriate behaviors to overtly aggressive and sometimes risky behaviors.

## Sensory and Motor Processing and Coordination Issues

Many patients with ND-PAE have sensory integration (also known as "sensory processing") issues, including hypo- or hypersensitivities to noise, touch, proprioceptive stimuli, smells, tastes, and/or visual stimuli. As infants, they are often difficult to soothe, may not seem to enjoy their caregivers, and can suffer from a range of

other regulatory problems. As toddlers and young children, they frequently are sensitive to environmental sounds, lights, and fans and may be easily irritated by loud voices or music. Older children, adolescents, and adults may cope by avoiding situations or environments which provoke their sensitivities.

## Lessons Learned About Neuropsychiatry

Because effects of prenatal alcohol exposure can mimic a variety of psychiatric disorders, it is important to accurately diagnose the condition in order to develop the most appropriate treatment plan. Moderate to heavy prenatal alcohol exposure can cause a wide range of deficits in children that are somewhat resistant to standard techniques of treatment. Children may have underlying cardiac defects, conduction anomalies, or arrhythmias associated with prenatal alcohol exposure, so it is important to rule out underlying heart conditions prior to beginning treatment with stimulants. Additionally, leptomeningeal heterotopias and other brain anomalies caused by prenatal alcohol exposure can be linked with seizure disorders. Medications that lower the seizure threshold can sometimes unmask such conditions (e.g., bupropion).

Additionally, traditional psychotherapy, cognitive and behavioral approaches, and other forms of non-pharmacological treatment should take into account subtle receptive and/or expressive language deficits, nonverbal learning disorders, social pragmatic challenges, or other communication issues. Other children may have auditory processing issues, cognitive or executive functioning problems, or difficulties interfering with their ability to benefit from traditional behavioral management.

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