NEUROPSYCHOLOGY TODAY

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Pervasive Developmental Disorders: Introduction

It is estimated that one child in 166 has a pervasive developmental disorder (PDD). The prevalence of PDDs has been increasing over the past several decades, which is attributed to new diagnostic categories and broadening of the diagnostic criteria. Given the high prevalence of PDDs and their detrimental effects on children's development and functioning, early diagnosis and referral for behavioral and educational interventions are vital, as they improve the long-term prognosis of PDDs.^{1,2}

PDDs involve dysfunction in such developmental areas as social interaction, communication and language skills, cognitive skills, and behavior (stereotyped, repetitive behaviors, restricted activities and interests). PDDs are highly heritable and most likely involve abnormalities in early development of the central nervous system. As a result, PDDs interfere with the children's normal neurocognitive, emotional, behavioral, and academic functioning.^{1,2}

PDDs, also referred to as Autistic Spectrum Disorders, include autism, Asperger's disorder, Rett's disorder, childhood disintegrative disorder, and PDD not otherwise specified (NOS). Even though these disorders affect the same core developmental domains, they are different in terms of clinical features, age of onset, gender distribution, course, and prognosis.^{1,2}

Inside:

Interventions for PDDs	2
About Dr. Rimma Danov	2
Neurological Abnormalities	
in Children with PDDs	3

Diagnosis and Comorbidity in PDDs

Early identification and intervention in PDDs cannot be overemphasized. Interestingly, research shows that the signs of PDD may be detected even before the children reach their first birthday, since infants who are later diagnosed with a PDD may display abnormal muscle tone, posture, and inactive or disorganized movement patterns. Currently, the early signs of PDDs that are viewed as the "red flags" by pediatric health professionals include lack of social smile, poor eye contact, no babbling, pointing, or gesturing by 12 months of age, no spoken words by 16-18 months of age or two-word phrases by 24 months of age, atypical play behavior, and regression in terms of loss of acquired language or social skills.1,3

These signs are typically first evaluated by the pediatricians, followed by a referral to a multidisciplinary team, which may involve a neuropsychologist, pediatric psychiatrist and psychologist, developmental pediatrician, and neurologist. When there are concerns regarding the child's functioning or positive result on a PDD screening test, pediatricians are advised to refer

(Continued on Page 2)



Types of PDDs

Autism

Autism is the most common and well-researched of the PDDs. The onset of autism is usually between 2 and 3 years of age, although such factors as parental concern, denial, and education, as well as the level of associated mental retardation in the child may mediate how early the child is diagnosed.⁵

Autism affects social, communicative. and cognitive development of children. These children may not like being held or touched, and may exhibit unusual auditory or tactile sensitivity. Autistic children display lack of social reciprocity and eye contact, lack of joint and/or symbolic play, and poor understanding of basic social conventions and expectations, as well as atypical attachment behaviors, emotional expression, and language use (monotonic speech, lack of conceptual words, extreme literalness, pronoun reversal, echolalia, etc.). Some social and communicative skills eventually improve in autistic children, but the vast majority continue to have social difficulties throughout adulthood.5

With regard to cognitive development, autism has been linked to mental retardation, and the severity of social impairment has been found to correlate with IQ in autistic children. During cognitive testing, these children typically perform better when it comes to nonverbal measures (matching items, identifying patterns, solving puzzles, etc.) compared to tests involving language and conceptual reasoning skills.⁵

(Continued on Page 3)

("Diagnosis and Comorbidity in PDDs," continued from p.1)

children for a comprehensive evaluation as early as possible and to closely monitor the child's siblings, since they are at an increased risk of PDDs.¹

To date, there is no single conclusive test or biological marker for PDDs; however, there is a number of screenings and diagnostic tools that the aforementioned health specialists utilize to evaluate children who possibly have a PDD. Primary care physicians usually employ "level 1" measures that are used to determine whether a child may have a PDD. Other specialists utilize "level 2" tests, which help confirm or refute the PDD diagnosis, differentiate from other developmental disabilities, and identify the type and severity of PDD.^{1,2}

Additionally, primary care physicians need to evaluate the child's hearing and vision, since disabilities in these areas may mimic the symptoms of PDDs such as lack of eye contact or response to one's name. Other conditions that should be ruled out during differential diagnosis include lead poisoning, fragile X syndrome, tuberous sclerosis, mental retardation, Down syndrome, Angelman syndrome, etc.^{1,2}

With regard to comorbid disorders, PDDs may co-occur with epilepsy and other seizure disorders. Sleep disturbances, gastrointestinal problems, dietary restrictions, allergies, and immunologic abnormalities have been reported in children with PDDs.^{1,2}

Further, children with PDDs are at an increased risk of behavioral and emotional disturbances, including inattention, hyperactivity, obsessivecompulsive disorder, tics, mood lability, oppositional defiant disorder, anxiety, and depression. It has been estimated that up to 78% of children with PDDs have symptoms of Attention Deficit Hyperactivity Disorder (ADHD), although according to the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV-TR), PDD and ADHD cannot be co-diagnosed, since the assumption is that ADHD symptoms are due to the developmental and cognitive disturbances associated with PDDs. Some researchers believe that the evaluation should include

assessment of symptomatology of both disorders when present.¹

As part of the comprehensive evaluation, children may be referred to a neuropsychologist. The main focus of pediatric neuropsychological evaluation is to assess the child's fine motor, sensori-perceptual, and neurocognitive functioning, including attention, memory, reasoning, language, nonverbal and spatial abilities, and learning skills. Additionally, neuropsychologist is qualified to evaluate emotional symptoms of the child and give the appropriate psychiatric diagnosis.⁴

The neuropsychological report includes the results of the objective tests that were used during the exam, as they relate to the child's everyday functioning, and the extent of cognitive and motor deficits in relation to the age norm. Also, the report includes diagnosis(es) and recommendations for treatment and interventions.⁴

Every state guarantees special education and related services for all eligible children. Public schools are federally mandated to provide these services to eligible children in accordance with the Individuals with Disabilities Education Act (IDEA). As for the pre-school children, each state provides early intervention programs for children identified to have developmental disorders up to the age of 3 years. The neuropsychological report can be used to determine the need for preventative and other services (speech therapy, occupational therapy, etc.), as well as special accommodations and individualized educational plan (IEP) at school.2

Interventions for PDDs

To date, there is no single treatment package that can improve the symptoms of PDDs. The intervention approach is developed for each child individually, depending on his or her specific deficits, level of functioning, and needs. It is known, however, that early detection and intervention improves long-term outcomes in these children. Most children with PDDs respond well to highly structured, specialized programs.²

Among the most widely used methods of treating autism is Applied Behavior

Analysis (ABA), which has been found to be effective in reducing inappropriate behavior and in increasing communication and appropriate social behavior. The central concept of ABA is reinforcing desirable behaviors and reducing undesirable ones. It is recommended that behavioral programs are highly structured, build on the child's interests, and teach tasks in a series of simple steps, while actively engaging the child's attention. Parental involvement is also encouraged.²

Other interventions for PDDs include medications (e.g., Olanzapine) that may help alleviate such behavioral symptoms as aggression, self-injury, and tantrums. Dietary restrictions (avoiding casein and gluten) and vitamin B6 taken with magnesium are also utilized in some cases.² A line of homeopatic remedies to decrease some of the emotional and behavioral PDD symptoms has been developed. For more information about this line of products and about local tutoring services go to www.55MyTutor.com.

About Dr. Rimma Danov

Dr. Rimma Danov received her PhD in clinical psychology from Adelphi University in NY. She completed her internship in clinical psychology and neuropsychology at Harvard Medical School and postdoctoral fellowship in pediatric and adult neuropsychology in a private clinic affiliated with NJ Medical School and the Robert Wood Johnson Medical Center. She is an assistant clinical professor at Penn State University, Dept. of Kinesiology, and has served as an assistant clinical professor at NYU School of Medicine, Dept. of Neurology, and Adelphi University, Derner Institute. In the past, she worked as a neuropsychologist for the NJ Devils Hockey Team and was engaged as a coinvestigator of TBI in boxers at the NYS Athletic Commission.

Presently, Dr. Danov maintains a full-time private neuropsychology practice where she examines neurocognitive and neurobehavioral functioning of patients 2-90 years of age with various neurological and neuropsychiatric disorders, such as MS, TBI, CVA, Parkinson's, Alzheimer's, dementia, ADHD, PDD, Autism, learning disabilities, seizures, and many others, using state-of-the-art neuropsychological techniques. Dr. Danov also conducts and publishes research in these areas. She is available for medico-legal consultations and testimony.

("Types of PDDs," continued from p.1)

Childhood Disintegrative Disorder

Children with this disorder are characterized by normal early development, followed by a profound regression of acquired communication skills with only minimal subsequent recovery. Once established, childhood disintegrative disorder is behaviorally similar to autism, although the prognosis is worse.⁵

Rett Disorder

Rett disorder is only found in girls, who develop normally during the first few months of life and then demonstrate developmental regression, deceleration of head growth, and loss of purposeful hand movement. During preschool years, Rett disorder resembles autism, with such symptoms as stereotyped motor behaviors and abnormalities in gait or trunk movements, while the associated mental retardation is usually more severe compared to autism. Rett disorder may also involve breath-holding spells and seizures.⁵

Asperger Disorder

This condition mostly occurs in boys, and may or may not be associated with mild mental retardation. It is identified in children somewhat later than autism due to relatively preserved language and cognitive abilities, but like autism, it is also linked to learning problems. The key features of Asperger disorder include naïve, inappropriate, one-sided social interaction; pedantic, adult-like, and monotonic speech; often highly developed verbal reasoning and vocabulary skills, but poor nonverbal communication and poor understanding of abstract language (e.g. sarcasm); difficulty forming friendships and consequent social isolation; intense absorption in specific topics; clumsiness, poor coordination, poor sensory integration, and odd posture. Asperger disorder is associated with better outcomes compared to autism due to higher intellectual skills.5

PDD Not Otherwise Specified (NOS)

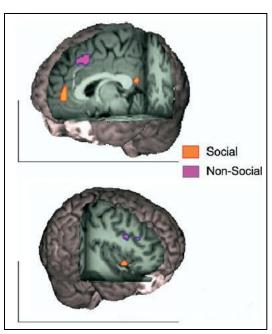
PDD NOS, also referred to atypical PDD, shares some symptoms with autism, since children with this condition exhibit unusual sensitivities and difficulties in social interaction, but

display better cognitive and communicative skills and ability to relate compared to autistic children.⁵

Neurological Abnormalities in Children with PDDs

Children with PDD have been found to have increased prevalence of hypotonia and hyperflexia, as well as epileptoform EEG results, even among the children without seizures. Further, children diagnosed with PDDs may have cerebral and cerebellar abnormalities, as well as atypical basal ganglia, supplementary motor, and anterior cingulate regions which may explain motor deficits seen in PDDs. Specifically, these children may have poor coordination, limb apraxia (inability to perform a voluntary movement), as well as impaired graphesthesis (finger sensory perception), stereognosis (hand object recognition), and alternating and sequential movement. These motor difficulties diminish in many children over time with treatment.3

A recent meta-analysis of numerous functional brain imaging studies revealed that during socially relevant tasks, children with PDDs are



Top: healthy children exhibit greater activation in anterior and posterior cingulate corteces during socially relevant tasks (orange regions) and in presupplementary motor area during other tasks (purple region) compared to children with PDD. Bottom: healthy children's brains are more activated in the right anterior insula during socially relevant tests, and in the middle frontal gyrus during nonsocial tests.⁶

characterized by hypoactivation in the perigenual anterior cortex, which is involved in executive functioning, and hypoactivation in right anterior insula, recently linked to social cognition (see image below). On the other hand, children with PDDs show more brain activation in the rostral anterior cingular cortex region, which is typically suppressed during attentionally demanding nonsocial tasks in normally developing children.⁶

Further, the same study demonstrated that children with PDDs tend to exhibit hypoactivation in amygdala, which plays an important role in evaluating others' facial expressions, affect, and intentions. Importantly, inappropriate recruitment of lower-order processing regions in place of higher-order regions has been noted.⁶

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- 1. Background image (pp.1,4): Jeff Johnson Biological & Medical Visuals
- 2. Girl image (p.1). autism.lovetoknow.
- 3. Brain image (p.3). "Works cited" #6.

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<u>Next Issues</u>- Jan'10: Mild Cognitive Impairment; Feb'10: Premature Birth

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Each insurance carrier determines the medical necessity of every requested neuropsychological exam differently. Our billing staff determines whether the exam will be covered by the insurance before the exam begins and works very hard to obtain an authorization, if needed. If you have questions about a plan that is not listed here, contact our office to find out whether we can obtain an authorization or have recently joined that plan.

Languages

We are very much open to diverse cultures in this practice and value the quality of a bilingual neuropsychological exam performed in the patient's native language. Dr. Danov is a native Russian speaker. Her current clinical staff include native *Russian*, *Spanish* and *Hebrew* speakers.



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