

PEDIATRIC ACUTE-ONSET NEUROPSYCHIATRIC SYNDROME (PANS)

KEYWORDS

PANS, OCD, Depression

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ABSTRACT Pediatric Acute-onset Neuropsychiatric Syndrome (PANS) is a clinical diagnosis given to children who have a dramatic – sometimes overnight – onset of neuropsychiatric symptoms including obsessions/compulsions or food restriction. They are often diagnosed with obsessive-compulsive disorder (OCD) or an eating disorder, but the sudden onset of symptoms separates PANS from these other disorders. In addition, they may have symptoms of depression, irritability, anxiety, and have difficulty with schoolwork. The cause of PANS is unknown in most cases but is thought to be triggered by infections, metabolic disturbances, and other inflammatory reactions.



INTRODUCTION

Pediatric Acute-onset Neuropsychiatric Syndrome "PANS" is a clinically defined disorder characterized by the sudden onset of obsessive-compulsive symptoms "OCD" or eating restrictions, concomitant with acute behavioral deterioration and/or motor/sensory changes in at least two of eight domains.[1][2] Additional psychiatric symptoms include anxiety (typically separation anxiety), attention deficit, hyperkinesis, emotional lability and/or depression, irritability, aggressiveness or oppositional behavior, and academic as well as cognitive decline. Associated neurologic findings include thought disorders, mental impairments, motor or vocal tics, increased sensory sensitivities, choreiform finger movements, deteriorating penmanship, and urinary frequency and/or enuresis. Sleep disruptions (disturbed REM sleep) are also characteristic. The definitive diagnosis also necessitates a course that follows a relapsing-remitting pattern. In case of postpuberty chronicity residual symptoms and their severity increases.[3][4]

CAUSES

The initial onset and subsequent exacerbations are usually incited by a variety of recognizable infections. Alternatively, these conditions may be associated with life stresses. In other cases, there is no clear inciting factor.[1]

Infections

A variety of inciting infections have been observed. The most common infection sites are in the upper respiratory tract: including rhinitis, sinusitis, and pharyngitis. The specific microbe most commonly recognized has been group A Streptococcus. Mycoplasma pneumonia, influenza, and other common viruses have also been noted. Influenza has often been well-documented anecdotally at both initial onset and exacerbations of PANS.[4]

A number of additional infections, including gastrointestinal infections, dental infection, herpes simplex, varicella, Epstein-Barr virus, enterovirus, Kawasaki disease, and anaphylactoid purpura, have been mentioned to be associated with the onset or exacerbation of PANS symptoms in a small number of cases.[1]

Environment

At the time of first PANS presentation, note any family history of

pharyngitis, impetigo, perianal dermatitis, and GAS infections. If possible, family members should have a throat swab cultured for GAS. Ongoing vigilance against GAS infections in any of the patient's close contacts is important, as symptom exacerbations have been reported following exposure to a sibling with GAS (even when the PANDAS patient had no evidence of infection). Prompt medical attention to symptoms suggestive of streptococcal infection is important not only to protect the patient, but also siblings, who may be at an increased genetic risk for PANS.[7]

Inflammation

Additionally, studies have shown a high rate of concurrent autoimmunity in patients with PANS and their first degree family members further supporting a role for inflammation in PANS

SYMPTOMS

The cardinal symptom of PANS is the abrupt onset of obsessive-compulsive disorder that causes significant enough distress to disrupt the child's normal home, school, and social activities. The child may manifest overwhelming fears, compulsive rituals and obsessive thoughts. These thoughts and behaviors arise suddenly, seemingly out **of nowhere.**

DIAGNOSIS

There are three criteria for diagnosing PANS:

- An abrupt and dramatic onset of obsessive-compulsive disorder or anorexia.
- The concurrent presence of at least two additional neuropsychiatric symptoms with severe and acute onset.
- Symptoms that are unexplainable by a known neurologic or medical disorder.

SUBTYPES

PANDAS ("Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections") is a subset of PANS and was first reported by Dr. Swedo at the National Institute of Mental Health in 1998. PANDAS has 5 distinct criteria for diagnosis, including:

- · abrupt "overnight" OCD or dramatic, disabling tics;
- $\bullet \quad \hbox{a relapsing-remitting, episodic symptom course;} \\$
- $\bullet \quad young\,age\,at\,onset\,(average\,of\,6-7\,years);\\$
- · presence of neurologic abnormalities;
- and temporal association between symptom onset and Group A streptococcal (GAS) infection.

The 5 criteria usually are accompanied by similar comorbid symptoms as found in PANS.[8]

MANAGEMENT

ORIGINAL RESEARCH PAPER

Mainstays of treatment include psychoeducational, psychotherapeutic, behavioral, family, school-based and pharmacological interventions, which reduce suffering and improve functioning until the immunologic and infectious processes are addressed.[5]

Accommodations

The majority of children with PANS/PANDAS require some type of school accommodation. Anxieties, OCD symptoms, tics, frequent urination, attention deficit disorder symptoms, handwriting difficulties, poor cognitive/physical stamina, difficulties with processing speed, memory issues, pain, and frequent absences create great challenges to the children and their teachers.[5]

Many schools have scarce resources, so persistence and creativity in the school and family will yield better outcomes. The IEP should provide specific, individually-tailored accommodations that will help the child with PANS/PANDAS attend and benefit from school. These might include:

- General—Excusing the child's absences, and not requiring make-up assignments or tests,
- Separation anxiety—allowing a parent to be in or near the classroom (perhaps helping out),
- OC symptoms excusing the child from certain activities, allowing him to complete assignments using alternate methods (e.g., typing homework, rather than erasing and rewriting repeatedly; listening to audio books, rather than reading and rereading),
- Urinary urgency/frequency leaving the class without asking permission
- Dysgraphia/handwriting difficulties—having a note-taker in class, dictating tests and homework, enlarging worksheets, writing on large grid paper, using a keyboard, voice-recognition software or audio recorder,
- Dyscalculia/math difficulties using a calculator or times table and working with a resource teacher or tutor,
- Slowed processing speed decreasing the number and length of assignments, allowing extra time for tests and in-class assignments and giving directions in written and oral form,
- Poor physical and cognitive stamina/pain shorter school day with reduced academic load, less homework, rest periods during the day (perhaps in the nurse's office), omitting or adapting physical education requirements.

When acute exacerbations of PANS/PANDAS symptoms have abated, these accommodations should be re-evaluated. Attending school and doing school work may be part of an appropriate psychotherapeutic plan.[5]

TREATMENTS

For treatment guidelines, refer to the PANDAS Physicians Network. PPN's goal is to help medical professionals understand, diagnose and treat PANS and PANDAS. The network provides research, diagnostic, and treatment tools. PPN Guidelines for Diagnostics and Therapeutics are developed by PPN committees and advisors from the top academic medical institutions in the United States. The members have worked with, treated, and studied the patients and the disorder. PANS and PANDAS are interdisciplinary disorders, so the relevant disciplines are represented on the PPN committees and special advisory council. Some of the disciplines include: Psychiatrists, Pediatric Neurologists, Immunologists, Microbiologists, Rheumatologists, Geneticists, Otolaryngologists, etc.

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