

Pediatric Acute-Onset Neuropsychiatric Syndrome



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KEYWORDS

- PANDAS • Pediatric autoimmune neuropsychiatric disorder
- Streptococcal infections

KEY POINTS

- A subtype of obsessive-compulsive disorder (OCD) that consists of an abrupt and severe onset has been described.
- The clinical presentation of pediatric acute-onset neuropsychiatric syndrome (PANS) can be differentiated from classic pediatric OCD by its course and its more global neuropsychiatric dysfunction.
- The causal mechanism is unsettled, and it is possible that multiple triggers initiate a neuro-immune process that converges to a common pathway leading to the clinical presentation.
- Research is under way regarding potential treatments, but is currently limited.
- Empirical studies and case reports suggest that potential PANS treatments should consist mostly of therapies that target immune and infectious causes.

OVERVIEW: NATURE OF PROBLEM

Pediatric autoimmune neuropsychiatric disorder associated with *Streptococcus* (PANDAS) is characterized by an abrupt (24–48 hours) onset of obsessive-compulsive disorder (OCD) and/or tics. Associated symptoms include emotional lability, separation anxiety, deterioration in handwriting, poor attention, and attention deficit/hyperactivity disorder (ADHD)-like impulsivity, deteriorating visual-spatial abilities, math and reading deficits, and enuresis (**Box 1**).¹ Neuropsychiatric symptoms tend to emerge 7 to 14 days after a group A streptococcal (GAS) infection. The course

Disclosures: See last page of article.

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Abbreviations	
ADHD	Attention deficit/hyperactivity disorder
ASO	Antistreptolysin
CaM kinase II	Calcium-calmodulin-dependent protein kinase II
CANS	Childhood acute neuropsychiatric symptoms
CBT	Cognitive-behavioral therapy
ELISA	Enzyme-linked immunosorbent assay
GAS	Group A streptococcal
IgG	Immunoglobulin G
IgM	Immunoglobulin M
IVIG	Intravenous immunoglobulin
MP	<i>Mycoplasma pneumoniae</i>
NMDAR	<i>N</i> -methyl-D-aspartate receptor
NSAIDs	Nonsteroidal anti-inflammatory drugs
OCD	Obsessive-compulsive disorder
PANDAS	Pediatric autoimmune neuropsychiatric disorder associated with streptococcal infections
PANS	Pediatric acute-onset neuropsychiatric syndrome
PITANDS	Pediatric infection-triggered autoimmune neuropsychiatric disorders
RF	Rheumatic fever
SC	Sydenham chorea
TPE	Therapeutic plasma exchange
TS	Tourette syndrome
URI	Upper respiratory infection

of the illness is classically or primarily relapsing/remitting, with symptom flares occurring months to years after the initial onset. Many PANDAS youth also display a saw-tooth pattern characterized by dramatic flares followed by considerable improvement, but not fully remitting. A few will display a progressively deteriorative course with each relapse, and a few children will remain remitted after recovering from the initial episode.

Recently, a group of researchers and clinicians familiar with PANDAS met and established a reiteration of this subtype termed pediatric acute-onset neuropsychiatric syndrome (PANS).² PANS is characterized by a clinical presentation similar to that of PANDAS; however, the cause is not defined as being exclusively due to a GAS infection (Box 2). In addition, PANS does not account for patients presenting with tics without OCD.² In this iteration, food refusal and anorexia presentations are

Box 1 Criteria for PANDAS
1. Presence of obsessive-compulsive disorder (OCD) and/or a tic disorder
2. Prepubertal symptom onset
3. Episodic course characterized by acute, severe onset and dramatic symptom exacerbations
4. Neurological abnormalities (eg, choreiform movements) present during symptom exacerbations
5. Temporal relationship between Group A streptococcal (GAS) infections and symptom exacerbations
<i>Adapted from Ahmad G, Duffy JM, Farquhar C, et al. Barrier agents for adhesion prevention after gynaecological surgery. Cochrane Database Syst Rev 2008;(2):CD000475.</i>

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