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Paediatric Acute-Onset Neurologic Syndrome (PANS) (aka PANDAS): Information for Healthcare Providers



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Sommaire : Paediatric acute neurologic symptoms (PANS) is uncommon, however family physicians should consider PANS when seeing the patient with sudden onset OCD, eating disorder or tic symptoms that appears correlated to a streptococcal infection. As referral to neurologists and psychiatrists may take time, family physicians can intervene quickly by investigating and treating any potential strep infections. Any residual OCD or tic symptoms can be managed as one would manage them from any other cause.

Case

“It was like he was fine one day, and then he woke up the next day, and now he has to wash his hands all the time… Not only that, but he’s regressed at home and school, and he can’t do anything on his own anymore. He’s clingy and anxious all the time… Its just not our child, like he’s been possessed… Its been a week like this now…”

What is Paediatric Acute-Onset Neurologic Syndrome (PANS)?

PANS describes a small group of children/youth that develop a neurologic syndrome with symptoms of Obsessive Compulsive Disorder (OCD) or tic disorders such as Tourette Syndrome, which occurs after a streptococcal infection.

PANS was previously known under the term Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS).

PANS Criteria

1. Abrupt, dramatic onset of obsessive-compulsive disorder or severely restricted food intake
2. Concurrent presence of additional neuropsychiatric symptoms, with similarly severe and acute onset, from at least two of the following seven categories

    - Anxiety
    - Emotional lability and/or depression
    - Irritability, aggression and/or severely oppositional behavoiurs
    - Behavioural (developmental) regression
    - Deterioration in school performance
    - Sensory or motor abnormalities

- Somatic signs and symptoms, including sleep disturbances, enuresis or urinary frequency

1. Symptoms are not better explained by a known neurologic or medical disorder, such as Sydenham chorea, systemic lupus erythematosus, Tourette disorder or others.

Epidemiology

By definition, PANS occurs in children and adolescents and affects a small subset of patients with OCD, but exact prevalence is unknown as there is a lack of population studies.
However, in a study of 136 youth with a lifetime OCD diagnosis, 5% of those with OCD met proposed criteria for PANDAS and/or PANS, of whom two met PANDAS criteria, four met PANS criteria, and one met criteria for both (Jasper-Fayer, 2017).

Pathophysiology

Exactly what happens in PANS is not well understand, however, the following steps are felt to occur:

- Individual develops a strep infection
- The individual's body produces antibodies against the strep bacteria
- The antibodies end up reacting with the person's own brain, specifically areas implicated in OCD, hence leading OCD and other neuropsychiatric symptoms

History

Classic history is the previously well child who presents with sudden onset of various symptoms:

- Primary symptoms such as:
  - Obsessive compulsive disorder (ODC) and/or
  - Tic disorder
- Secondary symptoms include, but are not limited to the following:
  - ADHD symptoms (problems with being inattentive, hyperactive, distractible, fidgety)
  - Separation anxiety, i.e. the child has problems separating from parents
  - Mood changes such as emotional lability, e.g. crying or laughing out loud, which is a marked change from previous
  - Regression
  - Temper tantrums; altered speech, including "baby talk" and selective mutism; limited insight;
  - Auditory and visual hallucinations may occur

Medical history

- Infectious diseases
  - Any symptoms of a pharyngitis?
    - Classic presentation is pharyngitis --> then OCD symptoms
    - However, due to severity of pharyngitis, other presentations are possible (Cooperstock, 2017):
      - OCD without any complaints of pharyngitis
      - OCD symptoms first (i.e. as early as 1-3 days before the onset of group A streptococcal pharyngeal (GAS) symptoms) then pharyngitis
- Autoimmune disease
- Inflammatory disease
- Immunodeficiency

Family history

- Movement disorders
- OCD, hoarding

- Other conditions such as mood / anxiety disorders
- Systemic conditions such as rheumatic disorders

Investigations

If you suspect PANS, then order the following as soon as possible:

- Throat swab
- Anti-streptolysin O titer (ASOT)
- Document when the symptoms started, and when the samples were taken (ideally as close together as possible)
- Other tests
    - Complete blood cell count (CBC) with differential
    - Erythrocyte sedimentation rate (ESR)
    - C-reactive protein (CRP)
    - Metabolic panel, i.e. glucose, calcium, Na, K, Cl, BUN, Cr
    - Is there urinary complaints such as dysuria?
        - Urinalysis (to assess hydration and to rule out inflammation).
    - Is there cloudy urine, i.e. pyuria?
        - Clean-catch urine culture.

6-8 weeks later

- Repeat ASOT

Diagnosis

Presumptive diagnosis can be made with

1) Throat swab (+) plus

2) ASOT (+) whereby:

- High ASOT associated with OCD or tic-like symptoms
- Low, falling ASOT associated with the convalescent phase

Note: Combination of Throat swab (+) plus ASOT (+) is required. Having only throat swab (+) or ASOT (+) alone is insufficient to make the diagnosis.

Differential Diagnosis

Consider the following differential diagnosis for an individual with PANS:

Neuropsychiatric / Neurologic	
• Transient tic disorder (aka provisional tic disorder)	Are there tics? Have they been present less than 12-months?
• Tourette’s syndrome	Are there multiple tics, e.g. vocal plus motor, etc.? Have tics been present at least a year?
• Sydenham chorea	Is there rapid, irregular, aimless involuntary movements of arms, legs, trunk or facial muscles?

• Autoimmune encephalitis	Any impaired memory and understanding? Any troubles with balance, speech or vision? Any weakness/numbness? Any changes of consciousness? Mental health symptoms such as anxiety, loss of inhibition, hallucinations?
Systemic	
• Systemic autoimmune disease	Fatigue? Achy muscles? Swelling and redness? Low grade fever? Troubles with focus? Numbness/tingling of hands and feet? Hair loss? Skin rashes?
• Wilson's disease	Any troubles with fatigue? Jaundice symptoms such as yellowing of skin and whites of eyes? Problems with speech, swallowing or coordination? Uncontrolled movements or muscle stiffness?
Psychiatric	
• Obsessive compulsive disorder (OCD)	Any compulsions? Any obsessions?
• Anorexia nervosa	Any loss of appetite with intent to lose weight?
• Avoidant/restrictive food intake disorder	Does the person have problems with eating (e.g. due to physical issues such as nausea, getting full too quickly, sensory issues)? Do they deny having worries about gaining weight?
• Bipolar disorder	Are there manic episodes, with increased mood and decreased need for sleep?

Management in Primary Care

Are there OCD symptoms felt due to PANS (i.e. abrupt onset of neuropsychiatric symptoms and evidence of recent GAS infection, e.g. positive throat swab)?

- Antibiotic course for 7-10 days
    - Even if the episode of GAS was already treated, it is recommended to treat again due to failure rates for penicillin and amoxicillin therapy.
    - Antistreptococcal therapy is associated with prompt resolution of PANS symptoms.
- Consider referral to neurology or mental health clinic.

Are there OCD symptoms that are persistent?

- When a child has symptoms of OCD with PANS, the symptoms of OCD are treated the same way that one would treat a child with OCD symptoms unrelated to PANS.
- First line for mild to moderate OCD
    - Non-medication strategies such as cognitive behavioural therapy (CBT).
    - Consider referral to a mental health professional that provides CBT (e.g. psychologist, mental health clinic, etc.)

Are non-medication strategies ineffective? Consider a trial of medication (WFSBP Treatment Guidelines for Anxiety, Obsessive-Compulsive and Post-Traumatic Stress Disorders).

- First-line for OCD
    - Fluvoxamine (Luvox)
    - Fluoxetine (Prozac)
    - Sertraline (Zoloft)
    - Citalopram (Celexa)
- Second-line for OCD
    - Clomipramine

Experimental treatments include intravenous immunoglobulin (IVIG) however there is insufficient evidence to recommend IVIG in most individuals, nor is it even available in most centres.

Websites for Professionals

PANDAS Physicians Network is a rich source of information about PANS/PANDAS, including clinical guidelines, and other practical information.
https://www.pandasppn.org

Patient Information

PANDAS: Frequently Asked Questions about Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections
https://www.nimh.nih.gov/health/publications/pandas/index.shtml

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About this Document

Written by members of the eMentalHealth.ca/PrimaryCare team which includes members of the Department of Psychiatry and Family Medicine at the University of Ottawa. Special acknowledgements to Dr. Asif Doja, neurologist. Reviewed by members of the Family Medicine Program at the University of Ottawa, including Dr's Farad Motamedi; Mireille St-Jean; Eric Wooltorton.

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