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PANDAS/PANS treatments, awareness evolve, but some experts skeptical

by Jessica Pupillo, Correspondent

One morning, 8-year-old Holland woke up with extreme anxieties, headache and abdominal pain. The previously healthy girl from Scottsdale, Ariz., also had food aversions and light sensitivities, said her mother, Karen Blandini.

For the next two years, the family hunted for a diagnosis and effective treatment, Blandini said. After Holland could no longer talk or get out of bed, she arrived at the Children's Postinfectious Autoimmune Encephalopathy Center of Excellence at the University of Arizona Steele Children's Research Center where she was diagnosed with Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcus (PANDAS).

The term PANDAS was coined in the 1980s by Susan E. Swedo, M.D., FAAP, chief of the Pediatrics and Developmental Neuroscience Branch at the National Institute of Mental Health (NIMH), after she discovered a link between abrupt-onset obsessive-compulsive disorder (OCD) and group A *Streptococcus* (GAS).

The diagnostic criteria for PANDAS include sudden-onset OCD or severely restricted food intake, Dr. Swedo said, and at least two of the following symptoms:

- anxiety;
- emotional lability or depression;
- irritability or aggression;
- behavioral regression;
- attention-deficit/hyperactivity disorder-like symptoms that cause a deterioration in school performance;
- sensory or motor abnormalities; and
- somatic symptoms like sleep disturbances and frequent urination.

PANDAS is a subset of Pediatric Acute-onset Neuropsychiatric Syndrome (PANS), which has the same symptoms but can be caused by infections like varicella or influenza, she said.

While there are no incidence data on PANS yet, Dr. Swedo estimates the disorder impacts about 1% of elementary school-aged children and likely is underdiagnosed.

PANDAS primarily affects children younger than 12 because they are less likely to have streptococcal antibodies.

While more centers like the one in Arizona are being established around the country to treat PANDAS, not all clinicians are convinced that strep infections trigger neuropsychiatric behaviors.

"The main reason PANDAS/PANS is not diagnosed more is because of almost 20 years of controversy," Dr. Swedo said.

Making a diagnosis

PANS is a diagnosis of exclusion, requiring a comprehensive evaluation. The PANDAS Physicians Network (PPN) maintains a diagnostic algorithm on its website (see resources) as well as treatment guidelines.

The PPN draws on the expertise of more than 40 doctors who serve on its scientific and medical advisory boards to help medical professionals better understand the disorders.



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"As literature is published, our expert panel updates our guidelines," said David Brick, M.D., FAAP, PPN president and a pediatric cardiologist. "We want to reflect the standard of care by expert consensus." Much of PPN's diagnostic guidelines are based on a consensus statement from a 2013 PANS Consensus Conference published in the *Journal of Child and Adolescent Psychopharmacology* (<http://bit.ly/2kN2aDh>); updated PPN guidelines are expected to be published in the journal by summer.

Pediatricians should consider PANS any time a child has an abrupt behavior change with obsessive thoughts, said Dr. Swedo, one of the authors of the consensus statement. These changes are so intense and out of character that parents or the child usually can pinpoint the day or hour that conditions began.

Treatment recommendations

It has long been established that in the treatment for rheumatic fever with Sydenham chorea, antibiotics may be recommended despite a negative strep throat culture, Dr. Brick said. Prophylactic levels of antibiotics should be considered for children with severe symptoms of PANDAS, those recovering from immunotherapy or those with multiple GAS-associated neuropsychiatric exacerbations, he said.

Cognitive behavioral therapy can benefit those with mild impairments, according to PPN treatment guidelines. If symptoms persist, nonsteroidal anti-inflammatory drugs, corticosteroids, intravenous immunoglobulin (IVIG) or therapeutic plasma exchange may be necessary, the guidelines state. IVIG and therapeutic plasma exchange, however, can be expensive and difficult to get approved by insurance.

"If (a child is) properly diagnosed, treatment is usually successful," Dr. Brick said. "A pediatrician may see a 7-year-old child who is normal at the beginning of the week and by the end of the week has severely debilitating OCD. These patients can have a significant recovery after IVIG."

IVIG treatment was said to be central to Holland's recovery. "Soon after treatment, she was walking, talking, singing, dancing and playing," her mom said in a video produced by the university. "We were amazed."

Sydney Anne Rice, M.D., M.S., FAAP, a behavioral-developmental pediatrician at the University of Arizona, said her center receives parent and physician referrals from across the country. Many children, like Holland, are very sick when they arrive, requiring support from an immunologist, sleep specialist and behavior analyst. "The quicker these children are identified and treated, the better they do," Dr. Rice said.

Disorder fraught with controversy

While there is no controversy that the children have debilitating OCD, there has been controversy about the cause of the disorder and how to treat it.

The AAP *Red Book*, for example, doesn't recognize a relationship between PANDAS and GAS.

"We know that some children with rheumatic fever had Sydenham chorea, a movement disorder. Many of those children had obsessive-compulsive behaviors as well as the movement disorder," said Margaret C. Fisher, M.D., FAAP, an infectious diseases expert who has served on both the AAP Committee and Section on Infectious Diseases, and has chaired the section. "There is no question that rheumatic fever is associated with GAS infection, so there is a basis for PANDAS."

"While I think some children do have PANDAS, I just don't know how to help them since the information regarding antibiotic therapy is conflicting," Dr. Fisher said. "I am patiently awaiting more studies."

Early studies with a likely selection bias fueled the confusion, Dr. Brick said. In those studies, a few doctors outside of NIMH selected patients with well-established Tourette syndrome or classic OCD symptoms who happened to have a strep infection, rather than patients with sudden-onset severe OCD.



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"These patients didn't respond as expected because they didn't have PANDAS," Dr. Brick said. "Some of the doctors who published the initial studies suggesting PANDAS is controversial are now working with us, realizing that maybe the initial studies were studying the wrong patients. It has taken some time to clear up that initial confusion."

Resources

- [PANDAS Physicians Network](#)
- [PANDAS - Q&A, National Institute of Mental Health](#)