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When Asperger's Syndrome and a Nonverbal Learning Disability Look Alike

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When Asperger's Syndrome and a Nonverbal Learning Disability Look Alike*

CASE

Joey was always an active boy. As an infant, he experienced severe colic. As a toddler, his pediatrician described him as "hyperactive and more impulsive than most children his age." Joey walked at 12 months but was unable to successfully drink from a cup until 15 months. His parents reported that he spoke several clear words at 12 months and that his spoken language was always ahead of other children his age. Prior to 3 years old, he had recurrent otitis media, chronic sinusitis, and constipation associated with withholding of stools. At 4 years old, his preschool teacher was concerned with fine motor coordination. A pediatric evaluation revealed intermittent hyperactivity with moderate delay in fine motor skills (eg, difficulty copying a circle and manipulating blocks) and mixed dominance; he was observed to be "extraordinarily engaging with advanced expressive language." The remainder of the neurological examination was normal without focal findings. Growth measurements, including head circumference, were between the 50th and 75th percentiles. A review of adverse events during prenatal and perinatal periods was unrevealing. There was no history of postnatal head trauma or loss of consciousness. Family history did not reveal any neurological or psychological disorders.

At the 6-year health-supervision visit, Joey's parents stated that his kindergarten teacher commented on accelerated reading skills (second- to third-grade level). With occupational therapy, fine motor skills improved moderately; however, he was now described as a "clumsy child." Further questions directed to the parents revealed that Joey's teacher expressed concern about his intermittent hyperactivity and preference for individual play. She told the parents that other children wanted to play with Joey, but he preferred to play and work independently. In the office, the pediatrician observed significant hyperactivity with a limited attention span. He fidgeted throughout the examination. Expressive language was notably advanced. The characteristics of attention-deficit/hyperactivity disorder (ADHD) were discussed with his parents. A plan was made for classroom accommodations and behavioral modification targeted at hyperactivity and social interactions.

The pediatrician, uncomfortable with ADHD as the primary diagnosis, phoned Joey's teacher to learn more about his behavior and learning in school. She

noted that he was "well-behaved and had verbal skills at a superior level." However, she reported, "at times his language was confusing and circuitous." His teacher was also concerned with Joey's unusual social interactions. "He often avoided physical, verbal, or eye contact" with peers and intermittently with his teacher. Although he would sit next to other children, he avoided initiating conversation. In addition, she stated that "he had a hard time focusing on class work, needed constant verbal cues to get back to work," and when she asked him why he was not completing his work, he replied, "I'm thinking."

At 7 years old, a child psychologist evaluated Joey. A Wechsler Intelligence Scale for Children-Revised (WISC-R) revealed a significant difference between verbal and nonverbal abilities: verbal IQ = 136 and performance IQ = 92. The performance subtest scores were all between 7 and 10. The Test for Visual Motor Integration was at the 37th percentile. The Children's Memory Scale revealed verbal memory in the superior range compared to visual memory in the low-average range. The psychologist observed appropriate emotional engagement during the assessment when interacting one-on-one. She concluded that Joey's difficulty integrating social, visual, and emotional information caused him to be easily overwhelmed in social situations and resulted in withdrawal.

The pediatrician reviewed Joey's medical history, the teacher observations, and the recent psychoeducational evaluation. Behavioral observations suggested Asperger's syndrome (AS). However, the neurocognitive test results were striking. She contemplated the next step in diagnosis and treatment.

INDEX TERMS. *Asperger's syndrome, nonverbal learning disability, pragmatic learning disorder.*

Dr Martin T. Stein

AS has received considerable attention during the past decade. Its inclusion in the autistic spectrum disorder (ASD), along with autism and pervasive developmental delay, has informed clinicians, parents, and teachers. Many clinicians have the impression that AS is used as a diagnosis indiscriminately to describe children and adolescents with atypical social behaviors without a careful consideration of alternative behavioral, language, and cognitive conditions. I agree with this observation.

The case of Joey illustrates the numerous diagnostic challenges when considering a diagnosis of AS at an early age. Two experienced clinicians were invited to comment on the case. **Professor Ami Klin** is the Harris Associate Professor of Child Psychology and Psychiatry at Yale University's Child Study Cen-

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ter. Dr Klin's research has focused on the neuropsychology of AS, autism, and nonverbal learning disabilities (NLDs). **Dr Karen Miller** is an Assistant Professor of Pediatrics at Tufts University School of Medicine. She is a developmental-behavioral pediatrician who practices and teaches at the Center for Children With Special Needs at the Tufts-New England Medical Center in Boston, Massachusetts. Dr Miller's special interests include learning and behavior problems in school-aged children.

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Dr Ami Klin

From the standpoint of Joey's presentation at the age of 7 years, the following descriptions from the developmental history need to be considered: possible motoric discoordination (later described as "clumsiness"), advanced and possibly precocious expressive language skills, the notion that he was "engaging" early on, and concerns about ADHD. As a 6-year-old, Joey's language could be "confusing and circuitous," and he had "unusual social interaction" but could also be withdrawn or avoidant and self-absorbed in thoughts. As a 7-year-old, intelligence testing revealed superior verbal skills and average or lower-average nonverbal skills, with a similar discrepancy between verbal and visual memory skills. He could interact appropriately with an adult on a one-to-one basis, but there was no report of the quality of social adaptation in more naturalistic environments. The psychologist who tested Joey considered the possibility of an NLD, whereas his pediatrician raised the possible of AS, a variant of autism.

The next step in the diagnostic process should include a developmental disabilities evaluation.¹ The first step would be to obtain a more detailed developmental history with a focus on Joey's social and communicative development. While it is not unusual for a child with good language skills not to come to the attention of professionals specializing in developmental disorders such as autism and other pervasive developmental disorders until school age, it would be highly unusual that, retrospectively, the parents or teachers would identify first concerns when Joey was 4 years old (and not earlier), and that this concern would be primarily in the motor area.

In the case of children with AS, early-onset patterns include unusual social, language, and communication patterns such as a pedantic style (both in choice of words and in tone of voice), reliance on language-mediated interaction (as a "life line") relative to little nonverbal interaction (eg, using facial and bodily gestures) and action-based social engagement (eg, reciprocal play), and awkward social approaches (eg, speaking loudly at a person's face, "talking too much" about unusual, adult-like topics, thus alienating other children). During kindergarten and elementary school years, this unusual style may evolve into attempts to engage others via relentless 1-sided descriptions of facts and information, difficulty in intuiting other people's feelings, intentions,

and motivations, all of which resulting in lack of mutual friendships despite an apparent motivation to engage others.² Self-absorption (eg, in thoughts) or even talking to self or smiling to self may be observed, again indicating a lack of monitoring of other people's reactions to oneself and a greatly reduced awareness of being the object of other people's subjectivity. "Circuitous" (or incoherent) speech may result from a style that is marked by verbal associations and fact-listing without maintaining a single coherent message or otherwise adjusting speech to the conversational partner's needs. Alongside the verbosity, one may see the emergence of all-absorbing circumscribed interests, sometimes involving unusual topics (eg, deep fat fryers, washers and driers, deep-marine biology, the Titanic) or more normative topics (eg, trains, snakes, computer games) but which are pursued in unusual manner (eg, learning of serial numbers, listing of models, exhaustive knowledge that is list-based) and with great intensity that may engulf the entire family's energy. These circumscribed interests typically impact on learning of other things and also interfere with the child's capacity to maintain reciprocal conversations.

In order to explore these various issues, there is a need for sampling the child's social and communicative behaviors directly (eg, using semistructured interviews such as the Autism Diagnostic Observation Schedule)³ as well as obtained reports of the child's behavior in representative environments such as home and school (eg, using the Autism Diagnostic Interview).⁴ Both instruments cover autism-related symptomatology and provide diagnostic algorithms based on *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition* (DSM-IV) definitions.⁵ Of great importance is the measurement of adaptive behavior, or the child's ability to translate cognitive potential into real-life skills, using the Vineland Adaptive Behavior Scales. Almost by definition, individuals with AS exhibit discrepancies of over 3 standard deviations between their verbal IQ and their socialization scores on the Vineland (which one can refer to as "street smarts"), and consideration of these delays is critical for diagnostic considerations and for intervention.⁶ Although children with ADHD may exhibit social and learning vulnerabilities, the early onset, nature, and magnitude of the social disabilities encountered in autism and AS are critical in differential diagnosis. Clearly, however, ADHD can be a comorbid condition; and indeed, individuals with AS may present with other psychiatric disorders, particularly in adolescence, the most common of which are anxiety and depression.

The child's cognitive profile of strengths and weaknesses can add a great deal to both understanding of the child's social vulnerabilities and to the creation of an individualized program of intervention.⁷ Although AS and NLD are not mutually exclusive diagnoses (because they belong to different nologies or classification systems), they often co-occur.⁸ The NLD profile is known to predispose children to social vulnerabilities because of their typical overreliance on verbally mediated/rote skills and neglect of intuitive, integrative, visual-percep-

tual, and visual-motor skills.⁹ This style results in reduced social intuition, overliteralness, social naivety, and slow social responses. Consideration of this profile in an effort to plan for an educational plan that capitalizes on the child's strengths while addressing the child's weakness becomes, therefore, of great importance in order to maximize the child's outcome and to prevent the emergence of maladaptive behaviors or secondary depression/anxiety.¹⁰

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Dr Karen Miller

The pediatrician is wise to look beyond Joey's hyperactivity and inattention to consider a wider differential diagnosis. In addition to ADHD, she is also considering AS. Another possibility is NLD. Both AS and NLD are present with a history of advanced verbal skills, social-interaction issues, and motor problems. Children with AS or NLD are often initially referred for assessment due to attentional concerns. The DSM-IV¹ notes that essential features of AS are severe and sustain impairment in social interaction with restricted, repetitive patterns of behavior, interests, and activities but without clinically significant delays in cognitive development or language. Although basic language skills are intact in AS, there are delays in the nonverbal communication skills and the social use of language (pragmatics). NLD is a neurological syndrome involving right hemisphere dysfunction and/or significant pertur-

bations of the brain's white matter (long, myelinated fibers).² There are strengths in basic verbal/auditory skills, rote memory, and early literacy skills but deficits in visual-spatial organization, tactile perception, motor functions, social skills, and executive functions such as self-regulation, planning, and problem-solving. Anxiety and difficulty adapting to novel or complex situations are common.

There is considerable overlap between AS and NLD.³ Joey's history of verbosity, precocious early reading skills, and clumsiness could occur with either diagnosis. He shows significant discrepancy on the WISC-R with better developed verbal than nonverbal abilities, a pattern that is characteristic of NLD but can also be seen in AS. ADHD can be a comorbid condition in both disorders. Several points suggest that Joey is better described as having NLD than AS given the information presented. He is emotionally appropriate and engaging in the one-to-one setting, but he has unusual social interactions in peer-group settings. Children with NLD often engage socially with adults but are overwhelmed by the complexities of peer interactions. They have difficulty processing nonverbal social cues, integrating sensory-motor information, organizing their language in a conversation, and adapting to new situations. They frequently become socially anxious and isolated. Although children with AS also participate in social interchange more effectively with adults, their atypical social interactions are still quite evident in that setting. Most importantly, Joey is not described as having repetitive behaviors or restricted interests, a requirement for an AS diagnosis. The use of an Asperger's-specific rating scale such as the Gilliam Asperger's Disorder Scale⁴ may be helpful in surveying for the relevant behaviors that may have been missed on initial interview. A parent, caregiver, or teacher can complete this scale.

Joey would benefit from referral for further assessment. Comprehensive academic testing is needed as children with NLD may be advanced in rote skills (eg, reading words) but have poor applied skills (eg, reading comprehension). Problems with mathematics are characteristic once children with NLD move beyond rote memorization of facts. Written expression is often a major frustration because of difficulties with handwriting and organization. Evaluation should also include assessment of higher-order language and pragmatics skills as well as social-emotional screening for anxiety and adaptive functioning in various settings. ADHD-specific rating scales are needed to assess for attentional dysfunction at home and at school.

Management involves education about NLD, and probably ADHD, with appropriate academic programming, social-skills training, emotional support, and possibly medication. Joey, his family, and his teachers need to understand his strengths and weaknesses.^{5,6} Children with NLD are frequently misunderstood, as their deficits may not cause significant dysfunction until mid-elementary school. This lack of recognition of the specific learning disability also contributes to difficulties obtaining appropriate educational programming. Joey would benefit from ac-

commodations for organization and executive-function deficits, pragmatic language interventions, occupational therapy, and social-skills training. Medication for behaviors associated with ADHD can be beneficial, but the effect may be less robust as the organizational deficits are multifactorial. If there is significant anxiety and depression, counseling and medication may be indicated.⁷ Joey will benefit from ongoing case management and monitoring as children with NLD are at increased risk for academic, social, and emotional problems in secondary school and adulthood.

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Web Site Discussion

The case summary for this Challenging Case was posted on the Developmental and Behavioral Pediatrics Web site† (www.dppeds.org.list) and the *Journal's* Web site (www.lww.com/DBP). Selected responses are included.

Keith Goulden, MD

We are at a crossroads with respect to the diagnosis of AS. On one hand, there is an increased prevalence of ASD from improved recognition of early and mild cases; on the other hand, there are many children with social-interaction problems that do not involve a "lack of social and emotional reciprocity" (the critical DSM-IV criterion for autism).

One choice is to continue trying to differentiate

children with atypical cognitive profiles. Joey has hyperlexia, which may be associated with superior verbal intelligence or a pragmatic language disorder (or both). He may also meet criteria for NLD, although achievement testing is not reported. Hyperactivity and inattentiveness suggest ADHD. Who are Joey's peers, and how much interaction with classmates is expected? The current gold-standard assessment for autism/AS includes a structured interview such as the Autism Diagnostic Interview and a structured observation such as the Autism Diagnostic Observation Scale. The purpose of these assessments is to establish whether Joey has a "lack of social and emotional reciprocity." Formal assessment of pragmatics and fine motor/sensory function would also be useful.

The other choice is to continue progressively widening the definition of AS to include all such children and use the label of AS or pervasive developmental delay to promote access to social-skills training groups, further medical or mental health specialty care, and parent support groups. The effort to distinguish AS, associated with deficits in visual-perceptual functioning and attention and strengths in verbal cognitive abilities, from a verbally gifted child with NLD and ADHD is considerable. With limited support resources, labels continue to be used as "gatekeepers" for services.

Richard Coolman, MD

I am cautious about labeling. AS is a life-long disability that impacts not only learning, but it also impacts independence, employment, and adult relationships. When cases are unclear in clinic or when parents aren't ready for the label, I provide information and community contacts. Parents are justifiably leery of self-fulfilling prophecies, particularly if schools have little in the way of experience with or supports for children with AS. I would explain that if Joey has AS, even with appropriate interventions including stimulant treatment of ADHD, occupational therapy, social pragmatics groups, and classroom accommodations around organizational deficits, the diagnosis will become clearer as educational demands become more abstract and peers become more sarcastic.

I think that one step at a time is often the best way to proceed. In addition to the interventions mentioned, Joey should qualify for 504 plan accommodations for ADHD and/or NLD, and his parents should be connected with an advocacy support group. Diagnosis of AS is sometimes driven by eligibility requirements, and if additional services are available only through an individualized education plan (IEP), then parents are often willing to take that step.

I am concerned with efforts to identify historical and ultrasuccessful persons like Thomas Jefferson and Bill Gates as having AS. While it may give kids role models, it also minimizes the disability. If they haven't required the supports, they probably have the phenotype, not the syndrome.

I often urge investment in a supportive, noncompetitive martial arts program. I recently sat behind a

† A bimonthly discussion of an upcoming Challenging Case takes place at the Developmental and Behavioral Pediatrics Web site. This Web site is sponsored by the Maternal and Child Health Bureau and the American Academy of Pediatrics section on Developmental and Behavioral Pediatrics. Henry L. Shapiro, MD, is the editor of the Web site. Martin Stein, MD, the Challenging Case editor, incorporates comments from the Web discussion into the published Challenging Case. To become part of the discussion at the Developmental and Behavioral Pediatrics home page, go to www.dppeds.org.

proud couple whose 13-year-old son was on stage at our Asian Celebration performing Kata successfully with his peers; for several years they doubted he would ever fully participate. A 22-year-old who attained a black belt was profiled in our newspaper. Both exemplify a big impact for kids who often need help finding niches.

David M. Snyder, MD

This case raises several diagnostic questions as well as the question of what the primary care pediatrician should do next.

The diagnostic questions pertain to the following possible diagnoses:

1. ADHD
2. NLD
3. Pragmatic language disorder
4. AS

Joey's high activity level and fidgetiness seems to be long standing. However, these motor symptoms are insufficient to make the diagnosis of ADHD. I would ask about the degree to which his attention span is "selective." Is he inattentive only when engaged in tasks requiring nonverbal processing, his area of cognitive weakness? Is his "daydreaming" the typical internal distractibility of an ADHD-affected, inattentive type, or is he either preoccupied with some anxiety or tuning out when he doesn't comprehend the task? I find that a bright 7-year-old can usually describe the content of daydreams. I ask: "Do you daydream about the same thing for a long time, or does your mind skip from one thought to another to another?" The latter pattern is more characteristic of ADHD. He should be assessed for anxiety. A trial of stimulants might well be contraindicated.

The "V-P" spread of 136-92 on the WISC-R represents nearly 3 standard deviations. This is very suggestive of an NLD. We are told he is reading well above grade level. How does he do in math? My guess is that it is slightly below grade level—not low enough for the teacher to identify a math learning disorder but clearly lower than predicted by the verbal IQ. He really should have an academic assessment, although the school might need some persuading to do this.

Joey's communication difficulties, as described, are more characteristic of a pragmatic language disorder than of AS. He has difficulty initiating conversation and, I would guess, with nonverbal communication—reading body language, turn-taking, maintaining eye contact, etc. The "pedantic" speech characteristic of AS is not described. He should have a language evaluation by a speech-language pathologist who is familiar with pragmatic language disorders. A language-based therapy approach should be considered as an alternative to social-skills training.

Does Joey have AS? He may well meet criteria for impaired social interaction, but there are no suggestions of "restricted repetitive and stereotyped patterns of behavior, interests, and activities." Without these, the diagnosis of AS is not appropriate. If the

pediatrician has difficulty establishing these criteria, a referral to a clinician experienced in diagnosing ASDs should be made.

The diagnosis of an ASD presents a significant emotional burden for parents, and this label can distract school personnel from assessing a child's learning problems and providing needed special education services. It should not be made without a complete, valid diagnostic assessment.

The differential diagnosis of AS, NLD, and pragmatic language disorders is often challenging. I find the DSM-IV quite helpful in this regard. Unfortunately, too many children do get labeled without reference to the consensually validated diagnostic criteria.

Dr Martin T. Stein

The process of differentiating the characteristics of AS, an NLD, and a pragmatic language disorder arguably may be the most challenging diagnostic tasks in developmental-behavioral pediatrics. Overlapping behaviors and cognitive processes and coexisting conditions challenge even the most experienced clinicians. That there are differences in these conditions is not an issue. How important a precise diagnosis is as a guide to treatment is perhaps the most important question for a clinician who treats these patients.

Joey exhibits social behaviors and speech patterns consistent with either AS or NLD as described by both Drs Klin and Miller. The absence in the case summary of restricted repetitive behavior and stereotypic behavior or interests leads one away from AS. However, as Dr Klin emphasized, a formal assessment of observed behaviors and standardized parent/teacher behavior reports should be performed and may reveal behaviors not documented in a clinical setting.

Although Joey's pediatrician recognized the significant difference between verbal and performance scores on the WISC-R, the only other available objective data on the psychoeducational assessment were below-average scores in visual-motor integration and visual memory. In addition, an assessment of academic achievement will guide Joey's educational interventions. Dr Klin pointed out the value of an objective measurement of adaptive behavior. It defines "the child's ability to translate cognitive potential into real-life skills...[and assess] street smarts."

The diagnostic formulation is not complete with the distinction between AS and NLD. There are clues in Joey's history of behaviors consistent with ADHD (hyperactivity, impulsivity, and inattention in the classroom) and social anxiety (social avoidance). Although the case summary does not include enough information to substantiate a DSM-IV diagnosis with either of these conditions, there is sufficient information to place them at the "problem level" in the *Diagnostic and Statistical Manual for Primary Care*¹ diagnostic format. Behavioral management, a social-skills group-training program, and medication should be considered with these coexisting conditions; they may be as important to Joey's treatment as a better understanding of his atypical cognitive pro-

file. Several commentators recommended a learning environment that accounts for Joey's cognitive strengths and weaknesses; it can be achieved through an IEP in his school.

What is the ongoing role of the pediatrician in assisting Joey and his parents?

Use a chronic disease model when developing management strategies.^{2,3}

At the time of the initial assessment and at follow-up visits, inform Joey and his parents about test results and what new treatments they might suggest.

Recognize that the educational and behavioral challenges will change as Joey's development progresses through the school-age period and into adolescence.

Assist parents in their advocacy role especially with Joey's school. Inform them about the 504 plan and IEP process and the responsibility of the school to develop an appropriate and effective educational plan.⁴

Recognize the potential for coexisting conditions; continuously reassess and refer when appropriate.

Consider connecting Joey's parents with another family in your practice who has a child with a similar condition. The pediatrician is in a good position to decide if such a meeting will be helpful for a particular family.

Discuss Joey's strengths and praise his accomplishment at each visit.

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