CASE

Frankie is an 8-year-old boy seen in consultation because of school difficulties in 2nd grade and “odd behavior.” His family has been concerned about him for the past 2 years, and he has had psychological testing at school. His parents are seeking another evaluation now because they are concerned that his school performance is declining and his self-confidence is decreasing.

Frankie was born at term weighing 7 lb, 3 oz. Delivery was complicated by the “cord around the neck,” but Apgar scores were 8 and 9, and he had no difficulties in the neonatal period. He went home with his mother. As an infant he was easily soothed and liked to be held. He sat at 6 months, crawled at 8 months, and walked alone at 14 months. He spoke 3 intelligible words at 1 year and was speaking in sentences by 2 years. He was fully toilet-trained by 2 years, 9 months. He had no significant medical illnesses.

His parents began to be concerned about Frankie when his preschool teacher noted a concern about his fine motor skills. She remarked also that, although he had no problems socially, he seemed happy to play alone. From a very early age, he liked ropes and often carried around a length of rope as some children carry a blanket. He was never a difficult child or often carried around a length of rope as some children carry a blanket. He was never a difficult child or disruptive at home or in his preschool. The only problem his parents have had is that he tends to get extremely involved in a particular activity (watching television, building with LEGO blocks), and it is very difficult to turn his attention away from that activity to a family task. At the same time, he is also noted to be easily distracted from tasks in which he has less investment.

Frankie’s father is said to have attention-deficit/hyperactivity disorder (ADHD) and learning disabilities, and he is being treated with an SSRI for depression. He works as a successful carpenter and builder. A maternal uncle had Down syndrome and died at the age of 17 years as a result of heart disease. There is no other contributory family history. A 12-year-old sister is successful academically and socially. Frankie’s mother is a librarian. The grandparents live nearby and share the parents’ concerns about Frankie.

He was evaluated a year earlier by his pediatrician and thought to have attention-deficit disorder, primarily because of distractibility and easy frustration. He is not oppositional but does have emotional outbursts and cries easily when frustrated. He was noted to carry with him a piece of rope and to play with it repetitively. A trial of methylphenidate was considered unsuccessful. An occupational therapy (OT) evaluation revealed low muscle tone and difficulty with visual-motor skills and handwriting. He was said to have “sensory integration problems,” and twice-weekly OT interventions were recommended. His parents requested our opinion about this recommendation.

Frankie’s academic work was considered average by his teacher in all areas, though weakest in spelling and reading. He was particularly strong in oral expression and noted to have excellent vocabulary and knowledge. He hesitated to participate in group activities and frequently needed considerable encouragement. On the other hand, he was noted to be liked by his peers and was not disruptive.

His parents noted that Frankie remembers “everything” in detail. He is very talkative and has a great sense of humor. He loves to speak in groups and is “a ham” when given a microphone. He enjoys crafts of all sorts and playing with LEGO blocks. He likes music. His favorite television programs are educational ones, from which he seems to learn a lot of information easily. He has made several extensive presentations to his class based on what he has learned from such programs.

The Behavioral Assessment System for Children (BASC) checklist was completed by both his teacher and his mother. The teacher’s responses suggested concerns in the subscales of anxiety/depression, attention, adaptability, and atypicality. Social skills were a notable strength. The mother’s responses suggested concerns in attention, adaptability, and social withdrawal.

Frankie’s parents and teacher completed a checklist based on the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV) criteria for ADHD. Both parents noted 3 items among the hyperactivity cluster and 5 among the inattentive cluster to be “often or very true” for Frankie. His teacher noted 4 inattentive items and 2 hyperactivity items to be true.

The Woodcock-Johnson Battery revealed a standard score of 100, with particular strength in short-
term retrieval and relative weaknesses in auditory processing and processing speed. Math skills, reading, and written language were average, whereas knowledge in all areas was in the superior range. On the Wechsler Intelligence Scale for Children (WISC)-III, Frankie’s verbal IQ was 104 and performance was 97. The highest subscales were vocabulary (13) and similarities (12); the lowest were coding (6) and symbol search (6).

On evaluation, Frankie was a pleasant and cooperative young man who participated in all tasks and related appropriately. His neurological examination was normal. He held a pencil or crayon tensely and wrote slowly and laboriously with tremendous pressure. He did not appear to be anxious during the evaluation. He kept a short piece of rope with him at all times and occasionally fiddled with it. He complied with all requests passively and did not initiate conversation or activities. He had no difficulty making transitions from 1 activity to another even when specifically challenged.

There are several discrete conditions that may be applied in this case. Is it in the child’s best interest to define which diagnosis fits him best? Is it acceptable to provide the parents and teachers with some guidelines for helping Frankie to succeed in school and at home and to “wait and see” how he develops over time? Would some practitioners initiate a trial of a pharmacological intervention?

This Challenging Case is about a school-aged child with an uneven pattern of development characterized by significant strengths and weaknesses in different developmental and behavioral domains. The case was submitted by Dr Ellen Perrin, Professor of Pediatrics at Tufts University School of Medicine in Boston, Massachusetts, and Medical Director of the Center for Children With Special Needs, New England Medical Center. In the first commentary, Dr Perrin sets the stage for the discussion. It is followed by a commentary by Dr Lynn Wegner, a development-behavioral pediatrician who practices in Morrisville, North Carolina.

INDEX TERMS. Asperger syndrome, ADHD, learning disability, transitional object.

Dr Martin T. Stein

The science of medical practice begins with a diagnosis. Before we develop a treatment strategy, we always strive to establish a correct diagnosis. The field of developmental and behavioral pediatrics is based on the same principle. Diagnostic classifications typically cluster a group of observable symptoms and signs that are seen at a particular age and, more or less, follow a known natural history. Although we are usually limited in the diagnostic process because of the absence of a biological marker (as found in most areas of medical practice), a specific diagnosis guides our understanding of a condition, our ability to educate a child and parents about the condition, and our choice of therapeutic interventions.

Pediatricians often see a child with a variety of behavioral symptoms and neurodevelopmental findings that either do not fit neatly into a single diagnostic category or suggest several diagnoses in different domains of development. How important is a unifying diagnosis in clinical practice? Is there a value to listing several diagnoses and then looking for overlapping patterns that may suggest a unifying treatment plan? In some cases, should we feel comfortable without a diagnosis and recommend treatment strategies that target specific behaviors or developmental deficits?

Dr Ellen C. Perrin

This child poses a diagnostic (and, thus, a therapeutic) dilemma. He has some characteristics of Asperger disorder because of his limited social interactions and some repetitive behaviors. On the other hand, he relates well to adults and experiences no significant limitations as a result of his relative lack of interest in social relationships. His repetitive behaviors are not rigid or continuous and cause no difficulties for anyone.

He has some characteristics of ADHD, inattentive type: distractibility, hyperfocusing, difficulty with organization, forgetfulness, and failure to finish activities. On the other hand, he has been able to overcome these difficulties with assistance from parents and teachers. His hyperfocusing appears to have served him well in his particular learning strategies, which he has used effectively to bolster his social relationships and academic achievement.

He also has some characteristics of nonverbal learning disability: he performs far better in oral tasks than in written ones, has excellent verbal memory, and has difficulty with math and social interactions. On the WISC-III, his strengths are in the verbal area and his weaknesses are in the performance area. On the other hand, he has no significant discrepancies in verbal and performance IQ scores and no motoric problems or difficulty with balance or visual-spatial functioning.

Some would argue that he has sensory integration dysfunction. His poor handwriting and difficulty with writing, poor organizational skills, and distractibility are said to be features of this problem, as are his unusual social relationships. On the other hand, he is not hyperactive and neither seeks extreme sensory input nor shuns/fears it.

Clearly, the overlaps along these supposedly discrete conditions are many. Is it in the child’s best interest to define which diagnosis fits him best? Is it acceptable (insurance policies aside) to provide the parents and teachers with some guidelines for helping Frankie to succeed in school and at home and to “wait and see” how he develops over time? Would
some try, empirically, some pharmacologic assistance?

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Frankie’s parents have asked very common referral questions: “Our child knows a lot, seems to be liked by his teachers and peers, and yet is not performing as well as he could be and is losing the feelings of competency he had when he first entered school. Why? How can we help him?”

Frankie’s early history provides important diagnostic clues. First, there are signs of possible fine motor delays dating to his preschool years. The subsequent OT referral confirmed “difficulty with visual-motor skills, handwriting, and low muscle tone.” Additional low scores on the WISC-III Coding and Symbol Search confirm continued relative weakness on pencil and paper tasks, and Frankie’s slow handwriting demonstrates the functional impact. A referral for OT assistance could make a real difference to this boy, as the handwriting demands will only increase in school and persistently slow, inexact writing can lead to incredible frustration and even his refusal to attempt classroom assignments.

Second, despite Frankie’s strong oral expression, excellent vocabulary and knowledge, and apparently normal discourse, there are several subtle historical clues suggesting language weakness. Frankie was described as “talkative.” Is this reflective of poor expressive organization? Are the mild reading and spelling weaknesses signs of an underlying phonological processing weakness? Are his lengthy oral reports of favorite educational television programs signs of emerging pragmatic issues? Because the verbal section of the WISC-III is not a measure of language, a referral to a speech/language specialist with expertise in phonological and discourse skills could be very helpful in providing practical suggestions for school and home assistance if real weaknesses are found.

Third, Frankie shows signs of weakness in several areas of executive functions, and these may underlie his “inattentive attention deficit.” His organization skills are weak, he poorly self-monitors and therefore often does not complete activities, and he may have difficulty previewing and planning, all of which may make him reluctant to enter new activities. Explaining this aspect of selective attention weakness to his parents could be very therapeutic as they understand that Frankie will need direct instructions in organization, planning and previewing, keeping things in their correct order, and self-monitoring. Environmental support is the best intervention here. Medication may help improve mental energy, but if Frankie does not tolerate the side effects well, the emphasis at this time should be placed on environmental support. Finally, the affective issues remain. Frankie may be biologically vulnerable to depression, yet it will be very important for the adults around him to not over-rely on that diagnosis for him. Frankie’s temperament may not enhance extroverted behaviors, but this may not reflect withdrawal. As long as Frankie is not ego-dystonic about his lack of social interactions with peers, it may not be a problem. Certainly, a social-emotional assessment can provide the answer to his parents’ concerns. Providing Frankie’s parents with more information about temperament could help them more accurately support their son.

In summary, the best therapeutic recommendations will involve environmental support and minimize diagnostic labels for this boy. Frankie shows signs of mildly uneven skills, but these can be remediated. There are no advantages to labeling him further unless the school requires an attention-deficit disorder, inattentive type, designation to develop a 504 plan for accommodations.

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Web Site Discussion
The case summary for the Challenging Case was posted on the Developmental and Behavioral Pediatrics Web site† (www.dbpeds.org/list) and the Journal’s Web site (www.lww.com/DBP). Comments were solicited.

Robert Wells, PhD, Valley Children’s Hospital, Fresno, California
This is an interesting case describing some common difficulties. There is not much written about children who are overfocused, but this sounds like a piece of the puzzle. Such children tend to become overly involved in their very specific interests and have difficulty moving on. They have subtle obsessive thinking, but rarely show the anxious quality found in obsessive-compulsive disorder. They may also have subtle learning disabilities, and this boy

† 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.dbpeds.org.
sounds like he may have a reasonably significant visual-motor problem. Difficulties with underfocusing are also common. “Underfocusing” is a term describing the symptoms of inattentive ADHD problems initiating, sustaining, and allocating attention in an efficient fashion so tasks are completed. It is not clear how functionally impairing any of these problems are for this young man. Some children with severe overfocusing styles do well on an SSRI, and this may be a part of his father’s history as well. I usually give out some articles explaining the overfocusing attentional style and ways for parents to help them cope with this. This child should receive some OT/physical therapy services and be followed carefully during his school years to make sure he is progressing and is not experiencing any significant functional impairment.

REFERENCES

David M. Snyder, MD, Developmental-Behavioral Pediatrics, Valley Children’s Hospital, Fresno, California

This young man’s pattern of development and behavior suggest the diagnosis of Asperger syndrome. The elements suggesting this include his fine motor deficits, disinterest in social interactions, remarkable accumulation of “facts,” and his always carrying an object—a piece of rope. The latter, I have found, is extremely common in Asperger syndrome and in some other children with autistic spectrum disorders. He certainly does not have the full-blown syndrome, but this possibility raises the question of whether his “attention” problems are secondary to anxiety or obsessive-compulsive disorder elements rather than ADHD per se. If so, an SSRI may be more helpful than a central nervous system stimulant, as Dr Wells suggested. In addition, OT should be arranged for the motor deficits. The case report is equivocal in regards to his social skills. A behavioral observation in a peer group setting by an experienced clinician would be interesting.

By the way, I recently reread Marcel Kinsbourne’s original description of the “overfocused child.” There is no question in my mind that, today, such children would be diagnosed as having Asperger syndrome.

REFERENCE

Frances Page Glascoc, PhD, Adjunct Associate Professor of Pediatrics, Vanderbilt University, Nashville, Tennessee

Is there more information on the breakdown of this child’s Woodcock-Johnson scores? Specifically, I’d like to see the quotients for word attack and passage comprehension. It might also be wise to have a second reading comprehension assessment since the Woodcock-Johnson Psycho-Educational Battery-Revised assesses literal skills. Other measures like the Diagnostic Achievement Battery-3 require children to actually read a passage and answer questions—a far more typical, yet challenging task, on which many students, especially those with emotional issues, do poorly. I’d also like to see the difference between math computations and applied problems for the same reasons described above.

Michael McMillan, PhD, Director of Behavioral Pediatrics, St John Hospital, Detroit, Michigan

This boy obviously has at least mild problems, considering attention and motor skills. What are the expectations of his parents and teachers? Everyone states he is basically happy and does well with peers, although he can play alone. I work in an area where there are 2 extremes, well-educated, wealthy, and often demanding parents to the other end of the continuum. As long as the school makes some accommodations for his poor motor skills and some environmental engineering to maximize his attention, maybe people should leave him alone. Sometimes I wonder if we create problems by expecting perfect kids?

Meg Zweiback, RN, CPNP, Associate Clinical Professor of Nursing, University of California, San Francisco, California

I wonder if teachers, parents, and possibly the clinician are describing a lack of self-initiated, assertive, or even oppositional social interactions. The fact that he is not seen as difficult or disruptive may be because he is not connecting to others. Sometimes an overwhelmed or depressed parent does not notice or is relieved by the absence of demands from a child or sees it as sign of independence.

I think that it would be good to observe this child at school and with family members to see the nature of his social interactions. He may not know how to have a conversation or to play with another child. If his classmates express their liking of him in ways that make him feel included in the social scene—for example, by choosing to sit next to him, joining him in play, or showing appreciation for his oral reports—great. But if he’s being treated like wallpaper, and the children “like” him because they don’t pay any attention to him, I would be concerned. He may be liked simply because there is nothing to dislike.

As children interact, they learn how to manage conflict, when to assert themselves, when to compromise, and how to deal with the complexity of relationships. If this little guy is passing under the radar all the time, he may not be learning how to make friends, even if he is liked.

Dr Martin T. Stein

Frankie is a school-aged child with excellent skills in oral expression, general knowledge, and vocabulary. His “odd behavior” includes a preference to play alone, hypervigilance for some tasks, and a...
chronic attraction to a piece of rope. Although his relative weaknesses (fine motor skills, spelling, reading, and, possibly, slow auditory-processing skills) have been defined during the evaluation, his schoolwork output is consistent with his measured aptitude.

Frankie is a child with an uneven pattern of cognitive, motor, and social development. If his “performance and self-confidence” are compromised as his parents state, then intervention may be appropriate. However, there is limited information to support a compromise in these areas. Rather than reach out for specific diagnostic categories as a means to specific recommendations, it might be more useful to define areas of concern (or “weakness”) where an intervention may be helpful. Before doing this, however, I would arrange an office visit with the parents alone with 3 goals in mind: (a) to describe the idea of “uneven” development in normative terms and guide their understanding that this concept describes the development of most children (and adults), (b) to delineate areas of strengths (for now and the future) and weaknesses that may benefit from an intervention, and (c) to understand how the parents perceive Frankie’s diminishing school performance and self-confidence. I would attempt to guide the interview in a direction that frames symptoms and signs into a format that helps the parents to gain a deeper understanding of the presenting concerns.

Before consideration of Asperger syndrome, mentioned as a possible explanation for some of Frankie’s symptoms by Drs Perrin and Snyder, I would see Frankie in the office for at least a second visit. His “passivity” and absence of “initiating conversation or activities” at the time of the initial office visit may be situational and change with familiarity. It is not uncommon for a child with a slow-to-warm-up temperament to become more animated and engaging at subsequent office visits. To be sure, many of Frankie’s behaviors may be associated with his temperament, characterized by a low-key responsiveness, situational inactivity, and slow adaptability. As suggested by Meg Zweiback, a visit to the school would provide valuable, first-hand information about social interactions. Is he always by himself? Which other kids does he attract during play? Is the hypervigilance significant or intermittent and transient?

It would be helpful to know more about the quality of the trial on methylphenidate. What was the maximum dose? What target behaviors were assessed? In the recent Multimodal Treatment Study of Children With ADHD (MTA), optimal response to core ADHD symptoms was achieved with stimulant medication dosage of 30 to 37 mg per day among children between 7.0 and 9.9 years old in the medication treatment groups. This is higher than the dose of stimulant used in the MTA community treatment group and higher than reported in community surveys of practice patterns among primary care clinicians.

Frankie’s symptoms recorded on a DSM-IV behavior checklist approach the criteria for a diagnosis of the inattentive form of ADHD. His parents endorsed 5 of 9 behaviors, whereas his teacher endorsed 4 of 9 behaviors. Although the DSM-IV standard requires 6 of 9 behavior criteria from both the parent and teacher sources for the inattentive type of ADHD, in the presence of chronicity (>6 months), onset before 7 years old, and a documented functional impairment in educational and social development, Frankie’s assessment approaches diagnostic criteria. I would present this information to the parents and offer another trial of stimulant medication if the initial trial used an inadequate dose of medication or if a second stimulant was not used.

Frankie’s attraction to a short piece of rope is intriguing, but its meaning is uncertain. His parents noted that he has carried the piece of rope from an early age. I would inquire about the home situation and life events around the time he became attracted to the piece of rope.

During the second half of the first year, many infants acquire a “transitional object,” a piece of a rag, a pillow, or a blanket. The child becomes progressively more attached to the object and uses it as a comforter, especially at bedtime, during separations, or following a stressful event. The transitional object is usually given up by age 3 to 4 years as autonomy and individuation encourage psychological separation from the parent. One of the few available long-term studies on transitional objects evaluated 171 healthy children between ages 9 and 13 years. There were no significant differences in psychological or educational outcomes that could be correlated with the acquisition or duration of a transitional object. This was true even among those children who carried an object into their school years.

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The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://pediatrics.aappublications.org/content/114/Supplement_6/1448