

CASES THAT TEST YOUR SKILLS

A teen with Down syndrome changes schools and suddenly becomes angry and isolative. His developmental skills regress, and he's talking with imaginary friends. Is this only an adjustment problem?

Regression, depression, and the facts of life

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HISTORY New school, old problems

Mr. E, age 13, was diagnosed with Down syndrome at birth and has mild mental retardation and bilateral sensorineural hearing loss. His pediatrician referred him to our child and adolescent psychiatry clinic for regressed behavior, depression, and apparent psychotic symptoms. He was also having problems sleeping and had begun puberty 8 months earlier.

Five months before referral, Mr. E had graduated from a small elementary school, where he was fully mainstreamed, to a large junior high school, where he spent most of the school day in a functional skills class. About that time, Mr. E began exhibiting nocturnal and daytime enuresis, loss of previously mastered skills, intolerance of novelty and change, and separation difficulty. Although toilet trained at age 7, he started having "accidents" at home, school, and elsewhere. He was reluctant to dress himself, and he resisted going to school.

The youth also talked to himself often and appeared to respond to internal stimuli. He "relived" conversations aloud, described imaginary friends to family and teachers, and spoke to a stuffed dog called Goofy. He would sit and stare into space for up to a half-hour, appearing preoccupied. Family members said he had exhibited these behaviors in grade school but until now appeared to have "outgrown" them.

Once sociable, Mr. E had become increasingly moody, negativistic, and isolative. He spent hours alone in his room. His mother, with whom he was close, reported that he was often angry with her for no apparent reason.

With puberty, his mother noted, Mr. E had begun kissing other developmentally disabled children. He also masturbated, but at his parents' urging he restricted this activity to his room.

On evaluation, Mr. E was pleasant and outgoing. He had the facial dysmorphism and stature typical

of Down syndrome. He smiled often and interacted well, and he attended and adapted to transitions in conversation and activities. His speech was dysarthric (with hyperglossia) and telegraphic; he could speak only four- to five-word sentences.

Was Mr. E exhibiting an adjustment reaction, depression, or a normal developmental response to puberty? Do his psychotic symptoms signal onset of schizophrenia?



Dr. Krassner's and Kraus' observations

Because Down syndrome is the most common genetic cause of mental retardation—seen in approximately 1 in 1,000 live births¹—pediatricians and child psychiatrists see this disorder fairly frequently.

Regression, a form of coping exhibited by many children, is extremely common in youths with Down syndrome² and often has a definite—though sometimes unclear—precipitant. We felt Mr. E's move from a highly responsive, familiar school environment to a far less responsive one that accentuated his differences contributed to many of his symptoms.

Psychosis is less common in Down syndrome than in other developmental disabilities.² Schizophrenia may occur, but diagnosis is complicated by cognition impairments, test-taking skills, and—in Mr. E's case—inability to describe disordered thoughts or hallucinations due to poor language skills.³

Self-talk is common in Down syndrome and might be mistaken for psychosis. Note that despite his chronologic age, Mr. E is develop-

Box 1

Which antidepressants are safe for treating pediatric depression?

The FDA's recent advisory about reports of increased suicidality in youths taking selective serotonin reuptake inhibitors (SSRIs) and other antidepressants for major depressive disorder during clinical trials has raised questions about using these agents in children and adolescents. Until more data become available, however, SSRIs remain the preferred drug therapy for pediatric depression.

- **Based on our experience**, we recommend citalopram, escitalopram, fluoxetine, and sertraline as first-line medications for pediatric depression because their side effects are relatively benign. The reported link between increased risk of suicidal ideation and behavior and use of paroxetine in pediatric patients has not been clearly established, so we cannot extrapolate that possible risk to other SSRIs.
- **Newer antidepressants** should be considered with caution in pediatric patients. Bupropion is contraindicated in patients with a history of seizures, bulimia, or anorexia. Mirtazapine is extremely sedating, with side effects such as weight gain and, in rare cases, agranulocytosis. Nefazodone comes with a "black box" warning for risk of liver toxicity. Trazodone is also sedating and carries a risk of priapism in boys.
- **Older antidepressants**, such as tricyclics, require extreme caution before prescribing to children and adolescents. Tricyclics, with their cardiac side effects, are not recommended for patients with Down syndrome, many of whom have cardiac pathology.

mentally a 6-year-old, and self-talk and imaginary friends are considered normal behaviors for a child that age. What's more, the stress of changing schools may have further compromised his developmental skills.

By contrast, depression is fairly common in Down syndrome, although it is much less preva-

lent in children than in adults with the developmental disorder.²

Finally, children with Down syndrome often enter puberty early, but without the cognitive or emotional maturity or knowledge to deal with the physiologic changes of adolescence.³ Parents often are reluctant to recognize their developmentally disabled child's sexuality or are uncomfortable providing sexuality education.⁴ Mr. E's parents clearly were unconvinced that his sexual behavior was normal for an adolescent.

TREATMENT Antidepressants lead to improvement

We felt Mr. E regressed secondary to emotional stress caused by switching schools. We viewed his psychotic symptoms as part of an adjustment disorder and attributed most of his other symptoms to depression. We anticipated Mr. E's psychotic symptoms would remit spontaneously and focused on treating his mood and sleep disturbances.

We prescribed sertraline liquid suspension, 10 mg/d titrated across 3 weeks to 40 mg/d. We based our medication choice on clinical experience, mindful of a recent FDA advisory about the use of antidepressants in pediatric patients (*Box 1*). Also, the liquid suspension is easier to titrate than the tablet form, and we feared Mr. E might have trouble swallowing a tablet.

Mr. E's mood and sociability improved after 3 to 4 weeks. Within 6 weeks, he regained some of his previously mastered daily activities. We added zolpidem, 10 mg nightly, to address his sleeping difficulties but discontinued the agent after 2 weeks, when his sleep patterns normalized.

At 2, 4, and 6 weeks, Mr. E was pleasant and cooperative, his thinking less concrete, and his speech more intelligible. His parents reported he was happier and more involved with family activities. At his mother's request, sertraline was changed to 37.5 mg/d in tablet form. The patient remained stable for another month, during which his self-talk, though decreased, continued.

Two weeks later, Mr. E's mother reported that, during a routine dermatologic examination for a chronic, presacral rash, the dermatologist noticed strategic shaving on the boy's thighs, calves, and scrotum. Strategic shaving has been reported among sexually active youths as a means of purportedly increasing their sexual pleasure.

The dermatologist then told Mr. E's mother that her son likely was sexually molested. Based on the boy's differential rates of pubic hair growth, the doctor suspected that the molestation was chronic, dating back at least 3 months and probably continuing until the week before the examination. Upon hearing this, Mr. E's parents were stunned and angry.

What behavioral signs might have suggested sexual abuse? How do the dermatologist's findings alter diagnosis and treatment?



Dr. Krassner's and Kraus' observations

Given the dermatologist's findings, Mr. E's parents asked us whether their son's presenting psychiatric symptoms were manifestations of posttraumatic stress disorder (PTSD).

Until now, explaining Mr. E's symptoms as a reaction to changing schools seemed plausible. His symptoms were improving with treatment, and his sexual behaviors and interest in sexual topics were physiologically normal for his chronologic age. Despite his earlier pubertal experimentations, nothing in his psychosocial history indicated risk for sexual abuse or exploitation.

Still, children with Down syndrome are at higher risk for sexual exploitation than other children,⁴ so the possibility should have been

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explored with the parents. Psychiatrists should watch for physical signs of sexual abuse in these patients during the first examination (*Box 2*).⁴

But how is *sexual abuse* defined in this case? Deficient language skills prevented Mr. E from describing what happened to him, so determining whether he initiated sexual relations and with whom is nearly impossible. The act clearly could be considered *abuse* if Mr. E had been with an adult or older child—even if Mr. E consented. However, if Mr. E had initiated contact with another mentally retarded child, then cause, blame, and semantics become unclear. Either way, the incident could have caused PTSD.⁵

Diagnosing PTSD in non- or semi-verbal or retarded children is extremely difficult.^{6,7} Unlike adults with PTSD, pre-verbal children might not have recurrent, distressing recollections of the trauma, but symbolic displacement may characterize repetitive play, during which themes are expressed.⁸

Scheeringa et al have recommended PTSD criteria for preschool children, including:

- social withdrawal
- extreme temper tantrums
- loss of developmental skills
- new separation anxiety
- new onset of aggression
- new fears without obvious links to the trauma.^{5,6}

Treating PTSD in children with developmental disabilities is also difficult. Modalities applicable to adults or mainstream children—such as psychodynamic psychotherapy, cognitive-behavioral therapy (CBT), exposure therapy, and medications—often do not help developmentally disabled children. For example, Mr. E lacks the cognitive apparatus to respond to CBT.

On the other hand, behavioral therapy, reducing risk factors, minimizing dissociative triggers, and educating patients, parents, friends, and teachers about PTSD can help patients such as Mr. E.⁵ Attempting to provide

Box 2

Signs of sexual abuse in pediatric patients

- Aggression
- Anxiety
- Behavior, learning problems at school
- Depression
- Heightened somatic concerns
- Sexualized behavior
- Sleep disturbance
- Withdrawal

structure and maintain routines is a cornerstone of any intervention.

FURTHER TREATMENT A family in turmoil

We addressed Mr. E's symptoms as PTSD-related, though his poor language skills kept us from identifying a trauma. Based on data regarding pediatric PTSD treatment,⁹ we increased sertraline to 50 mg/d and then to 75 mg/d across 2 weeks.

However, an intense legal investigation brought on by the parents, combined with ensuing tumult within the family, worsened Mr. E's symptoms. His self-talk became more pronounced and his isolative behavior reappeared, suggesting that the intrusive, repetitive questioning caused him to re-experience the trauma.


We again increased sertraline, to 100 mg/d, and offered supportive therapy to Mr. E. We tried to educate his parents about understanding his symptoms and managing his behavior and strongly recommended that they undergo crisis therapy to keep their reactions and emotions from hurting Mr. E. The parents declined, however, and alleged that we did not adequately support their pursuit of a diagnosis or legal action, which for them had become synonymous with treatment.

Mr. E's mother brought her son to a psychologist, who engaged him in play therapy. She followed

continued

her son around, noting everything he said. All the while, she failed to resolve her guilt and anger. When we explained to her that these actions were hurting Mr. E's progress, she terminated therapy.

How would you have tried to keep Mr. E's family in therapy?



Dr. Krassner's and Kraus' observations

Treating psychopathology in children carries the risk of strained relations with the patient's family. The risk increases exponentially for developmentally disabled children, as they have little or no input and their parents are exquisitely sensitive to their needs. Further, the revelation that the parents might have somehow failed to avert or anticipate danger to the child complicates their emotional response.

Although the child is the patient, the parent is the consumer. Failure to gain or keep the parents' confidence will hinder or destroy therapy.

Developmentally disabled children who exhibit psychopathology should be screened at the initial examination for sexual abuse. In cases of abuse, an amicable working relationship with traumatized family members—though difficult to maintain—is critical to treatment.

BottomLine

 **Related resources**

- ▶ Ryan RM. Recognition of psychosis in persons who do not use spoken communication. In: Ancill RJ, Holliday S, Higenbottam J (eds). *Schizophrenia: exploring the spectrum of psychosis*. New York: John Wiley & Sons, 1994.

DRUG BRAND NAMES

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|------------------------|-----------------------|
| Bupropion • Wellbutrin | Paroxetine • Paxil |
| Citalopram • Celexa | Sertraline • Zoloft |
| Escitalopram • Lexapro | Trazodone • Desyrel |
| Fluoxetine • Prozac | Venlafaxine • Effexor |
| Mirtazapine • Remeron | Zolpidem • Ambien |
| Nefazodone • Serzone | |

We might have protected our working relationship with Mr. E's parents by recognizing how fragile they were and how intensely they would react to any constructive criticism. Paradoxically, for the short-term we could have tolerated their detrimental behaviors toward Mr. E (such as repeated questioning) in the hopes of protecting a long-term relationship. Spending more time exploring the guilt, anger, and confusion that tormented Mr. E's parents—particularly his mother—also might have helped.

References

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