A 32-year-old woman with a history of severe intellectual disability was brought to the hospital by emergency medical services after being found unconscious on the bathroom floor by her parents. In the weeks prior to her admission, the patient had experienced numerous falls. Her Glasgow Coma Scale score in the emergency department was 9. She had a temperature of 87°F and a heart rate of approximately 50 beats per minute, indicative of severe hypothermia and bradycardia, respectively. Abnormal laboratory findings upon admission included a thyroid-stimulating hormone level of 68.3 uIU/mL and a potassium level of 3.1 mmol/L, as well as evidence of a urinary tract infection (UTI). Computed tomography without contrast of the head showed no acute intracranial or structural abnormalities.

The patient was admitted to the intensive care unit for further care and monitoring. Endocrinology was consulted, and upon examination they found the patient in a state of stupor with periorbital edema and with decreased bilateral Achilles’ reflexes. She was given 275 mcg of intravenous (IV) levothyroxine once daily in the morning and 100 mg of IV hydrocortisone every 8 hours. An adrenocorticotropic hormone stimulation test the next morning showed borderline results for adrenal function, so IV hydrocortisone was continued; it was later decreased to twice daily as she improved.

The patient’s parents were present in the room during the initial evaluation. Mental status examination showed a woman with blunted affect with a prominent nasal bridge and narrow forehead. During the initial evaluation she looked around the room and appeared to be responding to internal stimuli. Her voice was high pitched and she muttered responses to questions posed by the doctor during the interview. Her parents confirmed that the patient was not at baseline, namely due to their daughter’s new-onset paranoia, her talking to people who were not there, and reportedly seeing snakes on the wall. At baseline, the parents denied any history of paranoia, auditory/visual hallucinations, or suicidal thoughts or intent. However, the parents did note constant behavioral issues, specifically temper tantrums, impulsivity, and self-injurious behaviors when the patient did not get her way, as well as an insatiable appetite (in contrast to poor feeding as an infant).
The patient was prescribed 0.5 mg of risperidone twice daily orally and she quickly improved. Within 3 days she demonstrated alertness during interviews, resolution of visual hallucinations and paranoia, and reinstatement of her circadian rhythm. Her family confirmed that she had essentially returned to her baseline state of mind with resolved behavioral disturbances. She was successfully discharged to a skilled nursing facility for physical rehabilitation now that her behavior was controlled and her cognition had improved. Her follow-up care was coordinated at the facility.

DISCUSSION

Prader-Willi Syndrome

Prader-Willi syndrome (PWS) was first described in 1956 by Prader et al.\(^1\) in a report of several cases of “a syndrome characterized by obesity, small stature, cryptorchidism, and oligophrenia” after an initial hypotonic presentation in infancy. Patients with PWS also have increased rates of endocrine abnormalities such as hypothyroidism, adrenal insufficiency, diabetes mellitus type 2, and low levels of growth hormone.\(^2\)

PWS is the result of a microdeletion on part of paternal chromosome 15, with an estimated incidence of 1 in 16,000.\(^3,4\) Current consensus diagnosis has been formalized in a list of major and minor criteria with a scoring scale for clinical diagnosis.\(^5\) Major criteria include central hypotonia accompanied by poor suck at birth that gradually improves, resultant failure to thrive, subsequent hyperphagia and obesity after age 1 year and before age 6 years, dolichocephaly, hypogonadism, and microdeletion by testing.\(^5\) The patient in this case met criteria by prior testing per collateral history and by current presentation.

Myxedema Coma

Myxedema coma is a clinical diagnosis, and signs include hypothermia, hypoglycemia, hypotension, and altered mental status. It is an endocrine emergency and requires immediate supplementation of thyroid hormone in a hospital setting.\(^6\) In addition, patients with PWS with an acute high-stress illness are at much higher risk of developing adrenal insufficiency.\(^2\) In the case discussed here, risk plus the patient’s borderline adrenal function test both indicated the need for treatment with hydrocortisone, which was started by the endocrinology team prior to our consultation.

Delirium

Delirium may be difficult to diagnose in someone with premorbid intellectual disability; moreover, the condition is poorly studied due to the transient nature of the condition, limited assessment of premorbid condition, and ethical concerns.\(^7\) To best understand altered mental status, a baseline must be established, and fortunately the patient’s caregivers were present to provide collateral information. They confirmed a baseline of severe intellectual disability with no prior episodes of paranoia, visual hallucinations, or disturbances of consciousness and sleep.

Numerous possible etiologies for the delirium included the recent myxedema coma, adrenal insufficiency, steroid therapy, and UTI at presentation. The patient had been receiving treatment for all these conditions as indicated; therefore, the main concern at the time of consultation for both the primary team and the psychiatry team was symptomatic management, as the patient was responding to internal stimuli with altered circadian rhythms and worsening overnight physical aggression.

The psychiatry team analyzed both the risks and benefits of treatment options, both for the delirium itself and its contributing, potentially reversible factors. Levothyroxine treatment for the myxedema coma was necessary, as well as antibiotics for the UTI and fluid replenishment for hypotension. Glucocorticoid therapy for adrenal insufficiency was of particular importance here, as adrenal insufficiency is known to cause psychosis.\(^8\) However, given the recommendations of the endocrinology department, the severity of the patient’s illness, and subsequent improvement on an antipsychotic medication, the patient continued and eventually completed her steroid regimen without further issue.

For the symptoms of the delirium itself, we recommended non-
pharmacologic measures such as maintaining day/night cycles, consideration of a patient sitter, and use of physical restraints if necessary but only as a last resort. We felt the patient’s aggression and paranoia required pharmacotherapy to avoid unnecessary restraint and harm to self or others. Atypical antipsychotics are first-line treatment due to concerns of extrapyramidal side effects that can occur with typical antipsychotics. The best studied of the atypical antipsychotics for delirium are risperidone, quetiapine, and olanzapine, the last of which was given to the patient with little noted effect. Given the patient’s comorbid obesity and presenting myxedema coma leading to falls, concerns for metabolic syndrome and hypotension led us to discontinue the previously started olanzapine and to avoid quetiapine altogether. Instead, we chose the high-potency dopamine D2 receptor antagonist risperidone, which has also been shown to help with severe behavioral disturbances in PWS.

CONCLUSION
The primary reason for the psychiatric consultation in this case was not to diagnose PWS or myxedema coma, but rather to advise in the management of delirium. However, consideration of the underlying syndrome and its comorbidities contributing to the delirium was necessary in selecting an appropriate treatment. As shown in this case, psychiatrists can use their expertise to aid in selection of appropriate, indicated treatments for altered mental status while balancing risks and benefits to avoid unwanted sequelae, which in this case led to a positive outcome.

REFERENCES