A 18-year-old female presented to an adolescent clinic with a 3-week history of fatigue, feeling cold, “bone pain,” and sharp, intermittent abdominal pain. She also reported a 9-kg unintentional weight loss over the past month, accompanied by decreased appetite and increased thirst and urinary frequency. She denied nausea, vomiting, diarrhoea, fever, or recent sick contacts. Her medical history was only notable for a history of attention-deficit/hyperactivity disorder. Her social history was significant for current marijuana use (4-5 joints per day), a past history of voluntary inpatient rehabilitation for treatment of ecstasy use, and a past history of sexual abuse. Family history was significant for diabetes, hypothyroidism, rheumatoid arthritis, and substance abuse. A metabolic panel revealed significant hypokalemia (potassium 2.2 mmol/L) and metabolic acidosis (CO₂ of 16 mmol/L) with a normal anion gap. When hypokalemia was noted, the patient was referred to the emergency department. Potassium repletion was initiated and an electrocardiogram was performed; the results were normal. She was then admitted to the hospital for further work-up and management of her weight loss and electrolyte abnormalities.

On hospital admission, her height was 161 cm and weight was 46.8 kg (body mass index of 18 kg/m² [5th-10th percentile]). Pulse was 66 beats per minute and blood pressure was 113/72 mm Hg. Phosphorus was initially normal in the emergency room, but dropped to 2.3 mg/dL after admission. Urine pH was 7.0 and specific gravity was 1.008. Urine toxicology was positive for cannabis. Potassium supplementation was continued during her hospitalization. Nephrology, psychiatry, and adolescent medicine services were consulted. Nephrology suggested the possibility of renal tubular disease, with inappropriate potassium reabsorption by the renal tubules. However, a nutritional deficiency secondary to disordered eating was thought to be the more likely etiology of her hypokalemia. A renal ultrasound was normal. Despite the patient’s denial of depressed mood or suicidal ideation, inpatient psychiatric hospitalization was recommended for the treatment of the patient’s neurovegetative symptoms, which were thought to be consistent with depression, and for weight monitoring. However, the patient and her family refused psychiatric hospitalization and she was discharged home on oral potassium and phosphorus supplementation, with outpatient primary care and nephrology follow-up. Serum potassium on discharge was 3.4 mmol/L.

When seen in follow-up as an outpatient 2 months later, the patient’s pain and fatigue had improved and she had gained 2 kg since hospital discharge. She was no longer taking potassium or phosphorus supplementation. Despite improvement in her symptoms, her serum potassium remained low (3.2 mmol/L), and her electrolytes showed a persistent normal anion gap.

**Case Challenge**

For diagnosis, see page 62

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.

**An 18-Year-Old Female with Weight Loss and Hypokalemia**

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continued on page 62
TABLE 1.
American-European Consensus Group Classification Criteria for Sjogren’s Syndromea

Ocular symptoms (at least one of the following)
- Daily, persistent, troublesome dry eyes for more than 3 months
- A recurrent sensation of sand or gravel in the eyes
- Using a tear substitute more than three times a day

Oral symptoms (at least one of the following)
- Dry mouth for more than 3 months
- Recurrently or persistently swollen salivary glands
- Frequently drinks liquids to aid in swallowing dry food

Ocular signs (at least one of the following)
- Positive Schirmer’s testb of both eyes without anesthesia
- Elevated Rose Bengalc score

Histopathology: focal lymphocytic sialoadenitis

Salivary gland involvement (at least one of the following)
- Low unstimulated whole salivary flow
- Parotid sialography showing the presence of diffuse sialectasias without evidence of obstruction in the major ducts
- Salivary scintigraphy showing delayed uptake, reduced concentration, and/or delayed excretion of tracer

Autoantibodies
- Antibodies to Ro (SSA) and/or La (SSB) antigens

aData from Vitali et al.7
bObjective test to assess for tear production and ocular dryness.
dData from Vitali et al.7

Diagnosis:
Sjogren’s Syndrome

DISCUSSION
Sjogren’s syndrome (SS) is a chronic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands, including the salivary and lacrimal glands. Although the principal targets of the disease are glandular, extra-glandular and/or systemic features can also be seen. SS can be defined as either primary or secondary, depending on whether there is another autoimmune disease present (most commonly systemic lupus erythematosus or rheumatoid arthritis). The classification criteria by the American-European Consensus Group were revised in 2002 and have been validated in adults with SS with a sensitivity and specificity of 89.5% and 95.2%, respectively (Table 1).1 Currently, there are no validated criteria for the diagnosis of pediatric SS.

RTA is a clinical phenomenon frequently associated with adult SS, but less often seen in childhood SS. Distal renal tubular acidosis is the most commonly occurring presentation of renal disease in SS and results in the inability to excrete hydrogen ions at the level of the distal tubule. It presents with a normal anion gap metabolic acidosis, hypokalemia, and poorly acidified urine. Clinically significant renal manifestations of SS tend to appear in younger patients and precede the more classic symptoms of xerostomia (dry mouth) and xerophthalmia (dry eyes).2 The renal insult is thought to be primarily interstitial disease, with interstitial infiltration by lymphocytes on renal biopsy.3 In addition to distal RTA (type 1), renal complications may also present as proximal RTA (type 2), nephrogenic diabetes insipidus, nephrocalcinosis, inability to concentrate urine, and Fanconi syndrome. Less commonly, and typically later in the disease, patients with SS may develop immune complex glomerulonephritis.3

The natural course of renal disease in SS is typically reassuring, with most patients demonstrating latent disease and a good prognosis. Potassium supplementation and correction of acid-base disequilibrium is the mainstay of therapy for most patients. However, 3%-9% of patients demonstrate clinically significant renal disease.4 Although rare, presentation of severe renal disease may include hypokalemic paralysis, cardiac arrest, and renal failure.4 The presence of distal RTA confers a significantly increased incidence of B-cell lymphoma in adults with SS. This association has not been studied in the pediatric population.5

CONCLUSION
In an adolescent presenting with hypokalemia due to RTA, SS should be included in the differential diagno-
sis. Although renal failure due to SS is rare, SS is likely underdiagnosed in adolescence, due to atypical presentation of disease in pediatric patients. Cases of preventable renal failure in addition to other systemic complications due to this autoimmune process may therefore be missed if the diagnosis is not considered. Symptoms consistent with distal RTA overlap with other clinical entities, such as disordered eating, which can contribute to the difficulty in making the diagnosis. Substance abuse may also provoke RTA, making the correct diagnosis challenging in a patient with this comorbid psychiatric condition. Once diagnosed, appropriate monitoring of renal function and initiation of appropriate supplementation and immunosuppressive therapy, when indicated, may improve prognosis.

REFERENCES