**Type I hyperoxaluria** is a metabolic autosomal recessive disease caused by the absence of the hepatic enzyme alanine glyoxalate aminotransferase (AGT).\(^1\)\(^-\)\(^3\) AGT, normally found in peroxisome liver cells, is responsible for the conversion of glyoxalate to glycine.\(^1\)\(^-\)\(^2\)\(^,\)\(^4\) It catalyzes the removal of more than 99% of the circulating glyoxylate, thereby allowing only a small fraction to be oxidized to oxalate, a relatively insoluble end product.\(^1\)\(^-\)\(^2\)\(^,\)\(^4\) This key function becomes quite evident by the overproduction of oxalate that ensues when the enzyme is deficient.\(^4\) At normal physiologic levels, oxalate is easily excreted in the urine. If, however, overproduction exists, the kidneys become overwhelmed and unable to excrete the large amount of crystals. Microcrystallization in the proximal tubules, followed by calcium oxalate precipitation in the renal medulla, leads to interstitial edema and subsequent renal failure.\(^3\) If high levels of oxalate crystals continue to circulate systemically, end-organ deposition can develop, a condition known as oxalosis.\(^1\)

**Case summary**

A 15-year-old boy and his mother were admitted to a large metropolitan medical center operating suite for a living related renal transplant. The recipient was the boy, and his mother was the donor. Three days before surgery, he was admitted to a children’s hospital where he underwent dialysis each day for 5 hours in preparation for surgery. His blood urea nitrogen and creatinine levels before dialysis were 20 mg/dL (7.1 mmol/L) and 14.5 mg/dL (1,282 μmol/L), respectively. On the morning of surgery, he was transported to the preoperative holding area of the adult facility accompanied by his father. His mother was placed on a gurney in the adjacent bed space. The patient was awake, alert, and in good spirits. His medical history was significant for type I hyperoxaluria, hypertension, hyperparathyroidism, hyperphosphatemia, and end-stage renal disease. His surgical history included orthotopic liver transplantation in August 1998 (primary treatment for hyperoxaluria) and repair of a ventricular-septal defect as a child. The preoperative assessment was remarkable only for a grade III systolic ejection murmur. The patient had good left ventricular function as evidenced by an echocardiogram 1 year earlier. Laboratory tests revealed the following findings: sodium, 142 mEq/L (mmol/L); potassium, 3.6 mEq/L mmol/L; chloride, 103 mEq/L mmol/L; blood urea nitrogen, 11 mg/dL (3.9 mmol/L); creatinine, 3.2 mg/dL (283 μmol/L); hemoglobin, 12 g/dL (120 g/L); hematocrit, 37% (0.37); platelets, 164,000 (164 × 10⁹/L); white blood cell count, 2,500/mm³ (2.5 × 10⁹/L); phosphate, 6.2 mg/dL (19 mmol/L); calcium, 9 mg/dL (36 mmol/L); and magnesium, 1.9 mg/dL (0.95 mmol/L). The airway examination results were essentially normal; the patient was described as Mallampati class I with intact dentition.

After obtaining consent for surgery, anesthesia, and blood transfusion from the father, an 18-gauge intravenous catheter was inserted, and the boy was transported to the operating room. Monitors included 2-lead electrocardiogram, noninvasive blood pressure, temperature, and pulse oximetry. General anesthesia was induced with midazolam, 1 mg; fentanyl, 250 μg; sodium thiopental, 225 mg; and cisatracurium, 9 mg, after a priming dose of 1 mg. The
trachea was successfully intubated with a 6.0-cuffed endotracheal tube using a Miller 2 blade. Maintenance anesthetics included an oxygen-air mix (FiO₂ 50%), isoflurane at 1% to 1.5% end tidal, and intermittent boluses of fentanyl, midazolam, and cisatracurium. An orogastric tube was inserted to decompress the stomach, and an esophageal temperature probe also was inserted. A right internal jugular central venous pressure line then was inserted.

Throughout the case, vital signs remained within normal limits, and end-tidal carbon dioxide was maintained between 32 and 40 mm Hg. Before the incision was made, vancomycin, 1 g, was administered over 1 hour for antibiotic prophylaxis, and methylprednisolone, 500 mg intravenously, also was administered. One hour before reperfusion of the donor kidney, mannitol, 50 g, and hydrochlorothiazide, 50 mg, were administered intravenously over 45 minutes to promote renal perfusion. Shortly after the graft kidney was reperfused, dopamine at 3 µg/kg per minute was initiated to promote renal blood flow and was maintained into the postoperative period. The intravenous fluid used was normal saline, and the rate was increased after reperfusion to maintain a state of mild hypervolemia.

Following skin closure, the patient’s muscle relaxant was reversed with 3 mg of neostigmine and 0.6 mg of glycopyrrolate intravenously. After return of normal neuromuscular function and level of consciousness, he was extubated without difficulty and transferred to the postanesthesia care unit receiving 10 L of oxygen. In the postanesthesia care unit, the patient required a total of 5 mg of morphine for analgesia, and he recovered rapidly. He was transferred to a pediatric intensive care unit where he continued to recover. On postoperative day 8, he was evaluated for Achilles tendinitis in his left leg, which was treated with splinting and rest. Twelve days after admission, he was discharged home. His last blood urea nitrogen and creatinine levels were 8.5 mg/dL (3.0 mmol/L) and 0.9 mg/dL (80 µmol/L), respectively.

Discussion

Type 1 hyperoxaluria is an inherited condition of the liver that results in the overproduction of oxalate. It is relatively rare, affecting only 1:120,000 live births.¹ Onset most often occurs within 5 years and usually will cause end-stage renal disease by the mid-second decade.¹ In the presence of adequate renal function, the mainstay of treatment includes sufficient fluid intake and calcium oxalate crystallization inhibitors.¹ ⁵ Dietary restrictions of foods high in oxalate such as strawberries, nuts, and spinach also should be implemented; however, this has a minimal effect in reducing oxalate levels.¹ Pyridoxine (vitamin B₆) administration also has been shown to be useful for certain patients for reducing oxalate excretion; however, it is not an effective treatment for all.¹ When the glomerular filtration rate falls below a critical threshold, dialysis is required and renal transplantation becomes the preferred treatment option.¹ Aggressive dialysis remains a course of action; however, 6 to 8 hours of treatment are required each day to have any significant effect.¹ Therefore, renal transplantation is considered the best course of treatment when the patient’s glomerular filtration rate drops below 25 mL/min per 1.73 m².¹ Simultaneous hepatorenal transplantation also has been performed to provide a hepatic system that is able to produce adequate levels of AGT.¹ ⁷ This has been reported to increase the graft 1-year survival rate from 30% to 80%.⁶ However, a 1999 report stated that a “cure” for hyperoxaluria might not be necessary if the patient responds to vitamin B₆ and, therefore, disputed the need for more risky hepatic transplantation in these cases.⁷ For the patient described herein, hepatic transplantation took place 1 year before the renal transplantation procedure.

When a patient with type 1 hyperoxaluria enters the operating room, it is important for the anesthesia provider to have both a working knowledge of the disease and an understanding of the treatment options and associated abnormalities that might exist. After longstanding hyperoxaluria, it is common for oxalate crystals to be deposited in other organ systems, a condition known as oxalosis. Although hepatic function is sometimes less efficient, liver damage is not always present.⁴ The most common site of dysfunction is the renal system. When renal excretion of oxalate becomes excessive (>40 mg/d), crystallization in the proximal tubules occurs, followed by calcium oxalate stone formation in the medulla, leading to inflammation in the kidney and subsequent renal failure.³ ⁸ Renal stone formation is increased if urine oxalate concentrations exceed 0.4 mmol/L and urine calcium concentrations exceed 4 mmol/L.¹ During anesthesia, fluid administration should be planned to provide a dilutional effect. Hemodilution must be performed judiciously, however, in the patient with renal insufficiency, cardiovascular disease, or both. If intraoperative diuresis is required, hydrochlorothiazide, which was used in our patient, is the drug of choice. Although hydrochlorothiazide is a less potent diuretic than furosemide, there is a decreased risk of calcuiuria, which can lead to greater stone formation.¹ A combination of both drugs also may be acceptable.¹

Major deposit sites for excessive oxalate crystals are
the bones and joints. Synovitis and other orthopedic complications are also possibilities in this population. In our patient, this may have manifested in the postoperative period as Achilles tendinitis. Careful attention must be given to body positioning to avoid hyperextension or pressure on joints. Calcium oxalate deposits also may be present in the integumentary system; therefore, judicious padding should be used when extended surgical time is anticipated. A careful assessment of peripheral sensation also should be documented, since peripheral neuropathy is common.

The cardiovascular system is another common area affected by oxalate crystal deposition. Although the cause of this patient’s aortic regurgitation could have been related to the earlier repair of the ventricular-septal defect, oxalosis also could have been the cause. Oxalate crystals deposited in the heart can lead to cardiomyopathy and conduction defects. Deposits also can occur in distal arteries, leading to limb gangrene and arterial thrombosis. It is critical that the anesthesia provider be aware of these potential comorbid conditions in the preoperative period. Careful assessments of each patient’s physical activity level, along with preoperative electrocardiogram, chest radiograph, and echocardiogram is appropriate.

Finally, care must be taken in the choice of an anesthetic drug regimen. If muscle relaxation is required for the procedure, cisatracurium is the drug of choice. Cisatracurium undergoes spontaneous degradation at normal pH and temperature (Hofmann elimination), has no active metabolites, and is not dependent on either hepatic or renal function for elimination. Drugs to be avoided include agents with significant active metabolites (eg, meperidine, diazepam). Altered elimination of metabolically active compounds could lead to untoward effects.

Conclusion
Hyperoxaluria is a rare condition in the general population; however, it is likely that anesthesia practitioners will encounter a patient with this disease at least once in their careers, especially if their practice includes transplant recipients. Depending on the extent and progression of disease, these patients can range from healthy and active individuals with limited deficits, to people who are completely incapacitated. In-depth preoperative assessment to detect the range of anomalies, along with an informed approach to the anesthetic, is the key to implementing a safe and effective anesthetic plan individualized for each patient’s physical condition.

REFERENCES

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