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References

Anesthetic management of a 2-year-old male with propionic acidemia
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Sir—We present a case of a 2-year-old male, with propionic acidemia diagnosed at 3 days of age, scheduled to have percutaneous cystolithotomy, which was complicated with apnea and bronchospasm immediately after extubation.

He was a term but small for gestational age infant, born to a nonconsanguinous parents with no history of genetic disease but sudden infant death syndrome (his brother). He was first evaluated at 3 days of age for lethargy and inability to breast feed and diagnosed with propionic acidemia. At the age of 21 months, he presented with hematuria, and stone protocol computed tomography revealed stones in the collecting system. He was operated by lithotripsy. A month later, stones were seen in the bladder and he was scheduled for cystolithotomy. Since he had bronchitis with fever (39°C), the operation was delayed. During the infection, urine analysis was positive for ketones and his diet was revised.

Preoperatively, his general status was good and social development normal. Weight and height were 8.2 kg and 75 cm, respectively. Vital signs and laboratory data were normal and SpO2 was 98.1% in air, nor was there symptomatology related to the recent infection. Auscultation revealed normal bilateral breath sounds. Chest X-ray demonstrated clear lung fields. He received D10 1/3 NS during the fasting period.

On arrival in the operating room NIBP, ECG, pulse oximetry and endtidal CO2 were used for monitoring. The IV line was dislodged probably during transfer to the operating room, because of this induction was via inhalation of sevoﬂurane (8%) in N2O–O2 (50–50%). During induction, an 22 G cannula was inserted IV and D10 1/3NS restarted. Vecuronium bromide (0.1 mg kg⁻¹) was given for muscle relaxation and tracheal intubation was achieved at the first attempt with a 4 mm uncuffed tube, without complication. The operation lasted 30 min and was uneventful. Muscle relaxant was reversed by neostigmine (0.05 mg kg⁻¹) with atropine (0.01 mg kg⁻¹). As spontaneous respiration began and tone regained, he was extubated. Unfortunately, apnea occurred immediately after extubation with bronchospasm such that he could not be ventilated via face mask. He rapidly desaturatated, became bradycardic and peripheral perfusion deteriorated. He responded well to atropine (0.01 mg kg⁻¹) and was reintubated, then manually ventilated with 100% oxygen. Arterial blood sample was taken and revealed: pH 7.24, PCO2: 6 kPa (47 mmHg), PO2: 35 kPa (271 mmHg), HCO3: 1.7 mmol l⁻¹, SaO2: 100%, BE: −7 mmol l⁻¹ and lactate: 1.7 mmol l⁻¹.

Prednisolone and bicalciron were given. Peripheral perfusion, SpO2 and bradycardia improved. Spontaneous respiration began and he was transferred to the ICU intubated but was later extubated without complication. Blood glucose level was 24 mmol l⁻¹ (432 mg dl⁻¹) but urine ketones were negative. Glucose levels were regulated with an insulin infusion and the following day he was transferred to the ward with normal blood chemistry.

Propionic acidemia, is a rare autosomal recessive, life-threatening, inborn error of metabolism, caused by mutations in the PCCA or PCCB genes, located on chromosome 13 and the long-arm of chromosome 3; encoding two subunits of propionyl Co-A carboxylase enzyme, leading to deficient activity of this mitochondrial enzyme (1,2). The enzyme functions as part of the catabolic pathways of branch-chain amino acids (methionine, valin, isoleucin, and threonin), odd-chain fatty acids and cholesterol (1).
Deficient activity leads to excessive propionate levels in blood and odd-chain fatty acid deposits in liver (3). Propionic acid is also produced by anaerobic fermentation of odd-chain fatty acids in the gastrointestinal tract (1).

This clinically and genetically heterogeneous disease, typically begins shortly after birth with rare cases presenting in young adulthood (4). However, Jacobs et al. presented a case of prenatal diagnosis of propionic acidemia (5). Our patient was diagnosed with propionic acidemia at the age of 3 days.

Most commonly, the disease is characterized by episodic decompensation with dehydration, lethargy, nausea and vomiting as well as a risk for neurologic sequelae (white matter spongiosis or gray matter vacuolization) (4) and has a relapsing course of severe metabolic acidosis, usually precipitated by excessive protein intake, constipation or intercurrent infection (1). Ketoacidosis develops because excessive propionic acid feed-back inhibits the citric acid cycle enzymes (1,3). The disease may present with hypotonia, lethargy, hypoglycemia, seizures, developmental retardation, gastroesophageal reflux, neutropenia and trombocytopenia (bone marrow dysfunction), hyperammonemia (caused by inhibition of acetylglutamate synthetase by propionic acid), hypogammaglobulinemia, osteopenia, pancreatitis and cardiomyopathy. Dietary regulations make the manifestations less likely to occur (1). Our patient's acid-base balance, nutritional state, gastrointestinal function, muscle tone, mental status and social development were all good.

During the fasting period, the patients require IV fluids containing dextrose and sodium bicarbonate to suppress protein catabolism and subsequent acidosis (1). Our patient's fasting period was 4 h and he was given D10 1/3NS during this period. Unfortunately, an initial arterial blood gas sample was not taken, which should have been done as a base-line at the end of this fasting period. We avoided lactic acid containing fluids because these solutions lead to acid load and may be poorly metabolized.

We avoided drugs metabolized to propionic acid, odd-chain organic acids, odd-chain alcohols, acrylic acid or odd-chain fatty acids (1). Muscle relaxants metabolized by ester hydrolysis (succinylcholine, cisatracurium, and atracurium), including mivacurium should be avoided because their metabolites include odd chain organic molecules. Propofol, should also be avoided because a small portion of the fats included may be metabolized to propionic acid (1). We did not use propofol and we preferred inhalational anesthesia for induction. Muscle relaxation was provided by vecuronium. Lethargic and hypotonic patients may be sensitive to central nervous system (CNS) depressant effects of volatile anesthetics and narcotic analgesics (1). However, our patient had no CNS involvement and was not lethargic or hypotonic. We did not use narcotic analgesics, but a volatile anesthetic agent for both induction and maintenance of anesthesia, although the depressant effects might have been prolonged and this depressant effect may have caused the apnea in our patient. As Harker et al. reported, airway complications can be minimized if tracheal extubation is delayed until muscle strength and vigor is regained (1). Our patient had regained spontaneous respiration and tonus before extubation which was indicated.

Unfortunately, we experienced both apnea and bronchospasm immediately after extubation and it is probable that upper airway irritation from residual effects of the recent bronchitis, despite three asymptomatic weeks and antibiotic therapy were to blame. Delaying elective surgery until at least 4 weeks after infection, when upper airway irritability is known to lessen significantly (6) may be more suitable for these patients.

The apnea and bronchospasm also may have been caused by the exaggerated depressant effects of anesthetic agents related to metabolic acidosis that could have been present at the end of the fasting period, but unfortunately we did not have arterial blood gas analysis.

Supporting Harker et al., we can also say that these patients may be prone to develop respiratory distress secondary to fatigue or upper airway obstruction. Preoperative evaluation should include asking about recent infections and the patients should be followed up for acid-base balance during and at the end of the fasting period. Moreover, postoperative follow up in a postanesthesia care unit is necessary because of possible respiratory complications.

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References
Lumbar sympathetic blockade in a patient with cutis marmorata telangiectatica congenita
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Sir—We present, a 3-year-old girl, with a diagnosis of cutis marmorata telangiectatica congenita (CMTC) from 7 months of age, presenting with limping, pain in all toes of her right foot and ischemic lesions on the skin of the first three toes of the same foot, whose pain, ischemic areas and vascular lesions improved with vasodilator therapy and lumbar sympathetic blockade.

At 7 months of age, she was first evaluated by the Department of Pediatrics. In both thighs, capillaries were prominent and there were hypopigmented lesions coursing from thigh to calf on the medial sides of both legs. Her right foot was hypoplastic. From the superior iliac spine to the medial malleolus; her right leg was 5 cm shorter than the left. From the point 4 cm superior to the proximal pole of the patella, the circumference of the right thigh was 2.5 cm thinner than the left. There was no significant family history and there was no associated anomaly. Punch biopsy specimen revealed dilated venules in the superficial dermis. Arteriovenous colored Doppler USG showed normal main femoral artery and vein, popliteal arterial and venous branches and calf arteries and veins. The dorsalis pedis artery was not observed. At angiography the femoral artery could be followed towards the popliteal artery, but the peroneal and tibial arteries were not clearly observed. She was followed up with these symptoms and signs until 17 months of age without medication.

At 17 months of age, the Department of Cardiovascular Surgery started low-dose indapamide (Fludex, Servier, Neuilly-sur-Seine Cedex, France) and bencyclane hydrogen fumarate (Angiodel, Organon, Oss, The Netherlands). She was followed up until 3 years of age, but the response was limited.

At 3 years of age, limping, pain, and ischemic lesions increased (Figure 1). There was also a temperature difference between the feet; but not measured quantitatively. She was referred to the Departments of Cardiovascular Surgery and Anesthesiology. Indapamide and bencyclane hydrogen fumarate therapy was replaced with femoral intra-arterial ilio prost (Iliomedine, Schering, Berlin, Germany) for 7 days and she was given acetyl salicylic acid orally. Ischemic lesions regressed partially, but temperature difference, pain, and limping persisted. At the end of 7 days, knowing that the pain had become chronic (more than 6 months), we thought that lumbar sympathetic blockade might provide improvement.

We avoided neurolytic blockade, because of the patient’s age. We placed an epidural catheter under general anesthesia and started continuous sympathetic blockade. Unfortunately, the epidural catheter was dislodged on the second day as there had been little change in the symptoms and signs, we decided to administer lumbar sympathetic blockade instead of continuous blockade. We gave 5 ml of 0.5% bupivacaine at levels L2–L4 under general anesthesia with fluoroscopic control, twice a week for each blockade. The limb temperature difference was measured at the beginning and at the end of each blockade with at least a 4–5°C increase in temperature at the lesions. Improvement was maintained with a total of 12 blockades without complication (Figure 2). Our patient was followed up at 3-month intervals for 1 year and total relief was maintained. It has been 2 years since treatment was completed without further complication.

Cutis marmorata telangiectatica congenita, first described by Von Lohuizen (1), in 1922, is an uncommon and distinctive cutaneous vascular malformation composed predominantly of capillary and venous-sized vessels (2). The disorder, affecting predominantly females (3,4), presenting at birth or shortly after (3), is characterized by a fixed reticulated vascular pattern on the skin resembling physiological cutis marmorata, but unlike this, it does not resolve with warming of the skin (3,4). Prominent veins (phlebectasia), telangiectasias, cutaneous atrophy, possible ulceration of the involved skin and hyperkeratosis often accompany the reticulated pattern, usually in a localized distribution, especially affecting the lower limbs (4,5). Even with generalized involvement, it does not occur on