Management of outpatient anesthesia in an unusually case with glutaric aciduria type-1

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Sir—Glutaric aciduria type-1 (GA-1) is an autosomal recessive, uncommon and severe metabolic disorder with a deficiency of glutaryl-CoA dehydrogenase. This disorder mainly detects in early childhood (1). Most children with this disorder have no measurable enzyme activity, but several have had 10–15% residual enzyme activity (2). The population frequency has been estimated at 1 in 30 000 neonates in a Scandinavian study (3). Most patients have a dystonic–dyskinetic syndrome. The patient's symptoms are psychomotor delay, dystonia, spastic quadriparesis and macrocephaly (1). In the literature only two cases with GA-1, who underwent general anesthesia, were reported (4). Our cases are two sisters with GA-1, who were aged 12 (39 kg) and 16 (51 kg) years, respectively, and presented with macrocephaly and psychomotor delay. Laboratory analysis yielded urinary glutaric acid values of 157 and 153 nmol·mol^{-1} creatinine (normal: 0–5.3), respectively. On preanesthetic clinical examination, macrocephaly was observed, with an occipitofrontal circumference of 58 cm and 62 cm (>98%). They were hypotonic, especially in both arms and her reflexes were normal. Their optics discs were pale, and a dystonic posturing of both hands was noted. Father and mother weren't consanguineous. There required sedation for magnetic resonance imaging during routine follow up. The cases weren't premedicated before sedation. They were continuously monitored by electrocardiogram, oxygen saturation (SpO_2), body temperature, and noninvasive blood pressure. A 22 G intravenous cannula was inserted in the arm and a fluid infusion was started with dextrose in normal saline instead. Sedation was achieved by using bolus of propofol 1 mg·kg^{-1}. Sedation was maintained using a bolus of propofol 0.5 mg·kg^{-1} in case of necessity and spontaneous breathing was maintained during this time period. Oxygen was administered via facemask. Procedures time were approximately 10 mins. All the monitored parameters remained within normal ranges during the procedures. The procedures were completed uneventfully. They were being treated with a low-protein diet, riboflavin, and carnitine. The patients were discharged home 1 h after the procedure. GA-1 prevails in communities with a high rate of consanguineous marriages or in groups who are ethnically or culturally isolated. The father and mother of our cases were not relatives. The cerebral damage seen in GA-1 is caused by the direct effect of glutaric acid in the brain and Cerebrospinal fluid (4). Acute neurological crises are typically precipitated by the glutaric acid passage through the blood-brain barrier associated with an infection or some physiological stress. The most common presenting symptoms of GA-1 are macrocephaly, hypotonia or diffuse rigidity, consciousness, seizures, and dystonic limb movements (4). Our patient's symptoms were psychomotor delay, dystonia, spastic quadriparesis and macrocephaly. Metabolic manifestations, such as hypoglycemia and metabolic acidosis, are rarely present except in the acute phases of the disease (5). Our patients didn't have a metabolic problem. The principles of the treatment of GA-1 consist of an initial diet low in lysine and tryptophan to reduce total glutaric acid production, administration of oral L-carnitine supplements as all patients present a secondary carnitine deficiency, and riboflavin, a cofactor of glutaryl-CoA dehydrogenase (4). Our patients were also being treated with a low-protein diet, riboflavin, and carnitine.

An emergency protocol is established for preventing encephalopathic crises that consists of a high-calorie dextrose infusion, rapid correction of fluid deficits, ensuring brisk output of alkaline urine, high-dose intravenous carnitine, and anticonvulsants usage (5). To prevent hypoglycemia and metabolic acidosis in our patients during the sedation, we avoided using Ringer's lactate since it contains lactic acid, and used dextrose in normal saline instead.

Joaquin *et al*., reported that there were no reports on the superiority of one anesthetic drug over another in patients with GA-1 in the literature (4). The following should be taken into consideration in anesthesia management: the possibility of pulmonary aspiration, prolonged responses to nondepolarizing muscle relaxants, and hyperkalemic responses to succinylcholine. The patients with severe dystonia may be at a greater risk for aspiration of gastric contents during general anesthesia. Appropriate measures to avoid aspiration or its complications must be undertaken, including fasting, use of H2 blockers and rapid-sequence anesthesia induction with cricoid pressure (4). Before sedation, 25 mg of ranitidine was given intravenous to patients having fasting for 8 h in order to prevent aspiration of gastric content. We used a bolus of propofol to maintain a sedation as protected spontanaous breathing.

In concluded that we suggested performing sedation with propofol without any difficulty in these cases.

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Late eosinophilic pleural effusion after cardiac surgery in a neonate – prompt response to corticosteroid therapy

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Sir—Eosinophilic pleural effusion (EPE) defined as the presence of 10% or more eosinophils of the nucleated cells in the pleural fluid is well documented in adults (1) and may explain about 25% of postsurgical effusions (2). It is accompanied by blood eosinophilia in 25–75% of the cases. Mechanical or inflammatory pleural injury is assumed to be the triggering factor (1,3). Very few cases of EPE have been reported in children. In two cases of EPE following pediatric cardiac surgery, corticosteroids resulted in resolution of the pleural effusion within 48 h after the start of treatment (4).

We report a case of long-lasting pleural effusion in a girl with transposition of the great arteries and a ventricular septal defect, who underwent corrective surgery at 9 days of age. She was extubated and drains removed on the second postoperative day. Two weeks later, pulmonary function was deteriorating, requiring reinstitution of ventilator treatment. Bilateral pleural effusions were diagnosed and chest drains inserted draining opalescent exudates of 500–600 ml daily. Several surgical attempts (one resternotomy, bilateral thoracotomies) using sutures, clips, and compressions were performed during the third and fourth postoperative weeks aiming at occluding lymphatic vessels. The girl fasted and received total parenteral nutrition and octreotide (a somatostatin analog) (5). The effusions changed character from opalescent to straw-colored 4 weeks after the initial surgery. There were no signs of infection, and cardiac catheterization was unremarkable. Blood eosinophilia (34% of the leucocytes) was now detected (total number of leucocytes $6.7 \times 10^9\,l^{-1}$). 25–30% of the pleural exudates leucocytes were eosinophiles. Two weeks later, methylprednisolone 4 $mg{\cdot}kg^{-1}{\cdot}day^{-1}$ was started as the situation remained unchanged, and the blood eosinophil count was now 63% (total amount of leucocytes $8.6 \times 10^9\,l^{-1}$). Within 24 h, blood eosinophils were normalized at $0.04 \times 10^9\,l^{-1}$. Pleural drainage decreased within 3 days, and the chest drains could be removed, and the girl extubated a week after the start of corticosteroids. Methylprednisolone doses were slowly reduced during the following 2 months. Repeated blood eosinophil counts during this step-down were within normal range ($0.04–0.4 \times 10^9\,l^{-1}$), and the pleural effusion did not return.

Blood eosinophilia absolute eosinophil count greater than $0.7 \times 10^9\,l^{-1}$ is not uncommon in neonates. Possible causes are allergic reaction to antibiotics or blood transfusions, heart surgery or infection (6). Because we did not measure pleural effusion eosinophil count until after the blood eosinophilia was discovered, we cannot determine whether the pleural fluid eosinophilia was the primary reaction or not. The prompt reaction to corticosteroid therapy with diminishing pleural effusion and blood eosinophils is a strong indication of an inflammatory trigger causing an eosinophilic local and systemic response.

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Airway rescue with an esophageal airway in a child with a huge venous malformation in oral and maxillofacial region — a case report

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Sir—A 14-year-old male child with a huge venous malformation in oral and maxillofacial region was hospitalized and scheduled for copper needle plus sclerotherapy under general anesthesia. The venous malformation was noticed as a deep blue discoloration since birth and

