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Anesthesia Management in an 11-Month Old Infant with Pompe Disease

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ABSTRACT

Pompe disease is a glycogen storage disease (GSD) type II. Infantile-onset Pompe disease is fatal presenting with cardiac and skeletal myopathies and has an autosomal recessive pattern of inheritance with the prevalence rate of 1 in 40,000 live births (1). Its common symptoms include cardiomegaly, hypotonia, failure to thrive (FTT) and hepatomegaly (1).

The patient was a 4 kg, 11-month-old infant with the history of jaundice and recurrent seizures under treatment with phenytoin (15 mg/day) and phenobarbital (15 mg/day). He was hypotonic, cachectic and pale (Hb=9.5) when presented to the anesthesia clinic of Labbafi Nejad Hospital for bilateral lensectomy.

Induction and maintenance of anesthesia were carried out via the inhalation anesthesia method (N_2O/O_2 and sevoflurane). Laryngeal mask airway (LMA) was placed when achieving the appropriate depth of anesthesia. Bilateral lensectomy took 2 hours. After completion of the operation, the patient regained consciousness. His vital signs were stable and he was transferred to the recovery room and then to the ward. He was discharged from the hospital the day after the operation with no complications. ***(Tanaffos 2009; 8(1): 79-83)***

Key words*: Glycogen Storage Disease, Lensectomy, Anesthesia, Pompe disease*

INTRODUCTION

Pompe disease is a glycogen storage disease (GSD) type II (1) accompanying by a series of organic disorders and has an autosomal recessive (AR) pattern of inheritance (2). It presents in two forms including infantile or late onset (2, 3) and has a wide range of manifestations regarding onset of the disease, organs involvements and life expectancy (3). Its infantile-onset type is very dangerous and fatal (2, 3).

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Pompe disease is caused by acid alpha-glucosidase deficiency. This enzyme is responsible for degradation of glycogen polymers into glucose in lysosomes which are found within cardiac and skeletal muscles known as two major glycogen storage tissues (4). It is noteworthy to mention that cardiomyopathy and skeletal myopathy are observed in patients with complete enzyme deficiency resulting in death during first year of life. Skeletal myopathy eventually develops respiratory failure which is an obvious manifestation in patients with partial enzyme deficiency (4). Metabolic myopathy is due to a defect in muscular metabolism presenting as exercise intolerance or a progressive muscle

weakness (5). Infantile-onset Pompe disease is very severe and manifested with obvious cardiomypathy, hypotonia, hepatomegaly, failure to thrive (FTT) and death before 1 year of age.

Its late-onset form may initiate at any age and there is no severe cardiac involvement but skeletal muscles dysfunction is progressive. Other organs like liver, lung, spleen, endothelium, brain, peripheral nerves and anterior horn of spinal cord are involved as well (3).

Treatment mostly consists of enzyme replacement therapy (ERT) including acid alpha-glucosidase (myozyme) in late-onset form of the disease. Although it is not well-known which of these symptom resolves with ERT, most of them are preventable with continuing this treatment (6). The best treatment method in partial enzyme deficiency patients with respiratory symptoms is ERT (4). In case of no treatment, aggravation of the disease leads to progressive hypertrophic cardiomyopathty, arrhythmia, heart failure (systolic and diastolic), chronic respiratory failure and eventually death (2).

CASE SUMMARIES

The patient was an 11-month-old male infant delivered at full term by a vaginal delivery. He weighed 2,100 gram at birth (low birth weight) and was the 2nd offspring and his parents were not relatives. He became icteric after birth and underwent phototherapy. He also had a history of recurrent tonic-clonic seizures and was being treated with phenytoin (15 mg/day) and phenobarbital (15 mg/day). He was hypotonic, cachectic, breastfed and weighed 4 kg when admitted to the anesthesia clinic for bilateral lensectomy. Heart, lungs and abdomen were normal on physical examination. Cell blood count (CBC) and pediatric consultation were requested for him. The results demonstrated Hb=9.5 and an unidentified metabolic disorder was diagnosed. Biotin, carnitine syrup, folic acid, vitamin B6 and omeprazole were started for him and continued until the operation day.

On the operation day he was anemic, pale and hypotonic and had FTT. He was NPO 5 hours before operation. His chest x-ray and laboratory tests were normal except for Hct=32 and Hb=9.5. His temperature was 36.8˚C, SPO_2=95%, and heart rate= 126/min. To prevent hypothermia, his extremities, chest and head were covered with sheet wadding. An IV line was taken using catheter (22 G) and 40 cc of 1/3-2/3 IV fluid (10 cc/kg) was administered before the induction of anesthesia. Temperature of the operating room was set at 25˚C. The patient was monitored by precordial stethoscope, ECG, pulse oximeter and BIS (bispectral index) monitor. Considering the patient's health condition, we decided not to use any drug injection. Therefore, anesthesia was induced by using Mapleson F system and inhalation anesthesia method with 50% N_2O/O_2 and very low concentrations of sevoflurane. Sevoflurane was started with 0.5% concentration and it was increased by every 3-4 respirations. The patient was anesthetized after about a minute when the sevoflurane concentration reached 3%. Considering the HR= 115/min and BIS=62, the patient was hyperventilated for a short period of time and when the pupils were in the midline and BIS was reduced to 48, size 1 LMA was inserted. It fit perfectly by inflating cuff. It had a good compliance with no air leakage. At this time the permission for starting the operation was given considering the above mentioned conditions.

During the operation, the appropriate dosage of sevoflurane (2-3%) was repeatedly adjusted based on the heart rate, brachial pulse rate and BIS monitoring and use of precordial stethoscope.

One hour after the initiation of surgery, patient's heart rate started to decline incrementally and dropped from 120 to 110. At this time BIS was 35; therefore, sevoflurane dosage was reduced to 0.5%

and 0.1 mg atropine (0.025 mg/kg) was administered intravenously. After waiting for 30 seconds since no change was noticed, the second dose of 0.1 mg atropine was repeated with no response. Therefore, 0.2 mg IV ephedrine was administered and heart rate started to rise and reached 118/min quite rapidly. The heart rate stayed stable between 114-120/min during the remaining operation period. Patient's respiration was assisted throughout the operation (respiration rate= 30/min).

Since patient's condition was stable, the 2nd eye operation was started which took about 45 minutes. During this time, about 20 cc of 1/3-2/3 IV fluid was administered to the patient (the total amount of administered fluid was 15 cc/kg). Five minutes before completion of surgery, sevoflurane dosage was decreased and 2 minutes before ending the surgery administration of gases was discontinued and the patient was ventilated with 100% oxygen. Three to four minutes after completion of surgery when the patient had spontaneous breathing LMA was extracted and 7-8 minutes later, the patient regained consciousness and was transferred to the recovery room where he was monitored (ECG, SPO_2) and received oxygen via face mask. After about 40 minutes, the patient received 20 cc of 1/3-2/3 IV fluid and was breastfed after 20 minutes. He was transferred to the ward 20 minutes later with no complication. On the post-operative visit in the same evening, he had no complication. Electrolytes were tested which were within the normal limit. He was discharged the next day in good and stable general condition.

DISCUSSION

Metabolic disorder usually involves several important organs (heart, liver, lungs, etc.) (3) resulting in conduction disorders of the heart, cardiomyopathy, hepatomegaly, etc (2,3). In a case series study in 2007, 9 patients with infantile-onset Pompe disease were evaluated. These patients had developed cardiovascular arrest or fatal arrhythmia immediately after the induction of anesthesia (1). Based on report on metabolic disorders, patients in whom propofol had been used for induction of anesthesia had developed cardiac disorders and ventricular fibrillation (VF) (2).

Halothane causes significant myocardial depression through decreasing the heart rate and its effect on the heart rate is much more significant than sevoflurane. Therefore, we chose sevoflurane as the sole anesthetic agent for our case (7-11). On the other hand, since sevoflurane had much more significant effect on neuromuscular blockade compared to halothane, there was no need to administer a muscle relaxant (12).

Since our patient had a metabolic disorder, it was necessary to start feeding him as soon as possible after the completion of surgery in order to prevent further metabolic disturbances. That was another reason for choosing sevoflurane which is the drug of choice for such cases and can be revered very quickly resulting in patient awakening faster at the end of surgery (8, 13-17).

Considering the patient's special condition, we tried our best to maintain his hemodynamic status stable and avoided using any IV drugs (5). In several studies despite using sevoflurane for the induction of anesthesia, the patients developed hypotension, significant decrease of oxygen saturation and VF leading to death due to the infusion of propofol (18). We only used sevoflurane for our patient. We used LMA to provide our patient with the right dosage and at the same time minimum required amount of drug and to prevent overdose as well. By using BIS monitor for the patient, adequate dosage of sevoflurane was adjusted helping to maintain the anesthesia at an optimal level (not getting too much or too little anesthesia because they are both dangerous to the patient (19,20).

During the operation at one point the patient developed a conduction disorder of the heart resulting in decreased HR (110/min.) and cardiac output which was reversed by ephedrine (18).

Eye is a very sensitive organ and is innervated by several nerves. Therefore, eye surgeries are accompanied by a severe hemodynamic response. The risk is several times greater if the depth of anesthesia is not matched with the trauma/type of surgery endangering patient's life or vision. Therefore, it is essential to achieve an adequate depth of anesthesia. We met this requirement by adding a BIS monitor to the preexisting monitors which helped preventing an insufficient depth of anesthesia resulting in patient awakening during the operation or over dosage (19,20).

CONCLUSION

1- Using BIS monitor for controlling the depth of anesthesia is very helpful in such severely-ill patients.
2- Sevoflurane is the drug of choice for anesthesia induction in such cases because it does not cause bradycardia (like halothane does), does not drop cardiac output and does not dangerously affect patient's hemodynamic status (7). It also results in a faster recovery from anesthesia at the end of surgery (8, 13-16) and an adequate depth of anesthesia with sevoflurane can be maintained by the help of BIS monitoring (19,20) and adequate muscle relaxation can be achieved by this drug (12).
3- Since propofol causes myocardial depression and severe hypotension (18), it would be better if not used in similar cases with metabolic disorders because it may result in severe exacerbation of the preexisting condition.
4- It is crucial to stabilize the heart rate and maintain the hemodynamic stability in such cases (7,10)
5- To maintain the patients' hemodynamic status stable, it is necessary to preserve the preload. Therefore, fluid therapy should be performed with extreme precision in these patients (2).
6- Considering skeletal muscles weakness and probability of respiratory depression, we should pay a special attention to respiratory symptoms which, fortunately, were not detected in our patient (4, 5).

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