症 例 (Case report)

Anesthetic Consideration in a Patient with Noonan's Syndrome

Ryu Okutani*, Ayumi Fujita**, Taeko Fukuda**

Noonan's syndrome exhibits Turner's syndrome-like symptoms, namely a short truncus, webbed neck, ocular hypertelorism, and ptosis. But, Noonan's syndrome has normal chromosomes[1]. For associated cardiac anomalies, pulmonary stenosis, atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus (PDA), and hypertrophic cardiomyopathy (HCM) have been reported[2]. This syndrome is often associate with a risk of developing malignant hyperthermia. Recently we experienced anesthetic management in a patient with Noonan's syndrome with HCM who underwent orchiopexy. The details are described in the following paragraphs.

Case Presentation

A 6-year-old boy, height 110cm and weight 18kg, was scheduled for the surgery of bilateral-orchiopexy.

The patient was delivered at a gestational age of 36 weeks by forceps in an asphyctic state. A birth weight was 3,500 g. An electroencephalographic examination revealed an abnormal spike. He had been under medication with anti-convulsant until the age of 3, but he has not had any obvious convulsive attacks. Developmental delays were evident; his neck became steady at 3 months and he learned to walk at 3 years. No chromosomal anomalies were recorded at a chromosomal analysis performed immediately after birth. Family history was unremarkable. Characteristic findings of Noonan's syndrome, such as ptosis, wide-set eyes, antlmongolold slant of the eyes, nystagmus, low-set ears, and cryinkly hair, were noted. A delay in intellectual development was recognized; language comprehension is 1 year 9 months, and social age is 1 year 10 months at age of 6.

No abnormalities were found in the blood chemical analysis. CTR was 56% in chest X-ray film. Left axial deviation was found in electrocardiography. Echocardiography revealed asymmetrical hypertrophy of the ventricular septum and narrowing of the left ventricular chamber. No obstruction was noted in the left ventricular outflow tract. HCM with no out flow obstruction was diagnosed. Left ventricular ejection fraction was 90% and cardiac output was 1.5 $\ell \cdot min^{-1}$. A cerebral CT examination showed slight atrophy of the bilateral frontal lobes and temporal lobe, but electroencephalographic examination revealed no obvious abnormalies.

Two hours prior to entering the operating room, 9mg of diazepam was orally administered followed by intramuscular injection of 0.25mg atropine 45 minutes before surgery. After monitoring devices, such as electrocardiographic electrodes and pulse oximeter, were attached to the patient, anesthesia was induced by inhalation of 4 $\ell \cdot min.^{-1}$nitrous oxide, 2 $\ell \cdot min.^{-1}$oxygen, and sevoflurane up to 5% through mask under spontaneous breathing. After the patient was anesthetized, avenous line was secured and 2mg of vecuronium, a muscle relaxant,

*Department of Anesthesiology, Hyogo Prefectural Nishinomiya Hospital
**Department of Anesthesiology, Hyogo College of Medicine

was injected to facilitate intubation. The endotracheal tube size 5Fr was intubated without difficulty. Anesthesia was maintained with 66% nitrous oxide and 0.5-2% sevoflurane in oxygen, and intermittent 1mg vecuronium iv. During the surgical procedure, the circulatory system was monitored by transthoracic echocardiography to detect obstruction of the left ventricular outflow tract and to evaluate the left ventricular volume.

Except for a slight rise in temperature (from 36.4 to 37.1℃), the surgical procedure was completed normally. After the effect of the muscle relaxant was reversed by atropine 0.4mg and neostigmine 1mg at the end of surgery, the tracheal tube was removed. The total time required for surgery was 2 hours and 35 minutes and for anesthesia was 3 hours and 25 minutes. The volume of lacted Ringer's fluid infused, urinary volume, and blood loss were 480mℓ, 100mℓ, and 18g, respectively. Recovery from anesthesia was uneventful, and after 2 hours observation in the recovery room, the patient was returned to his ward. Throughout the perioperative period, there were no signs suggesting the development of malignant hyperthermia.

Discussion

The incidence of Noonan's syndrome is reported to be 1/1,000 to 1/2,500 in all birth[3]being higher than that of Turner's syndrome. It is frequency complicated with abnormalities of the urogenital and cardiac systems[4]. The problems in anesthetic management of patients with Noonan's syndrome are following;

1. Difficulty in airway management due to micrognathia and a webbed neck.
2. Presence of cardiac complications such as pulmonary stenosis, ASD VSD, and HCM
3. Possibility of developing malignant hyperthermia.

Since the present patient was free of airway anomalies such as micrognathia, webbed neck, and macroglossia, the trachea was intubated without any problems. However, it is wise to prepare for such a difficulty in airway management, as we prepared a laryngeal mask and a bronchofiber apparatus.

Noonan's syndrome is very frequently complicated with congenital heart disease (26～30%)[3] so an echocardiographic examination is absolutely necessary as a part of the preoperative cardiac evaluation. Pulmonary stenosis most frequently occurs, and ASD, PDA, VSD, and HCM have been reported[2]as other cardiac anomalies of this syndrome. The frequency of incidence of HCA is said to range from 26% to 32%[3] in cardiac anomalies.

The present patient suffered from HCM. Although the preoperative echocardiography did not show stenosis of the left ventricular outflow tract at rest, there is a risk of develping an obstruction at the outflow tract during anesthesia and surgery. Fortunately, the patient exhibited satisfactory myocardial contraction and his left ventricular wall motions were normal. Therefore use of inhalation anesthetics which possessed a pronounced cardiac depressive effect was acceptable[5]. During the surgical procedure, transthoracic echocardiography (Color Doppler) showed no obstruction of the left ventricular outflow tract, and that the left ventricular contents evaluated by tomography of the four chambers maintained an appropriate preloading.

The relationship between malignant hypertherrmia and Noonan's syndrome has not been elucidated, but it has been reported that the former occurs frequently in chidren with Turner phenotype[6]. Therefore particular attention is required in patients with this syndrome who had an exaggerated serum creatinine phosphokinase level prior to surgery. Intra-operative monitoring of body temperature is essential.

We successfully managed a patient with Noonan's syndrome operated for orchiopexy under general anesthesia using sevoflurane, nitrous oxide in oxygen and iv. vecuronium. Preand intraoperative monitoring using echocardiography contributed to the successful management.

References

1) Summitt RL. : Turner syndrome and Noonan's syndrome. J Pediat 74 : 155-156, 1969
2) Matsuzaki Y, Akiba T, Yoshikawa M, et al : Evaluation of heart disease associated with Noonan's syndrome. Shonika Rinsyo 44 : 1991-1995, 1991
3) Nora JJ, Lortscher RH, Spangler RD : Echocardiographic studies of left ventricular disease in Ullrich-Noonan syndrome. Am J Dis Child 129 : 1417-1420, 1975
4) Hayakawa F, Tsukidate K, Mori O, et al : Clinical evaluation of 30 cases of Turner syndrome and 16 cases of Noonan's syndrome (in Japanese). Shonika Rinsyo 38 : 2625-2631, 1985
5) Campbell AM, Bousfield JD : Anaesthesia in a patient with Noonan's syndrome and cardiomyopathy. Anaesthesia 47 : 131-133, 1992
6) Pinsky L. : The XX-XY Turner phenotype and malignant hypertherrmia. Lancet 19 : 383, 1972

(Circ Cont 15 : 453～455, 1994)