Anesthesia for a Patient with Williams Syndrome

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(Key words: Williams syndrome, supravalvular aortic stenosis, systolic pressure gradient)

Williams syndrome is a very rare disease associated with characteristic face, cardiac abnormality, and mental retardation[1]. This is a case report of our anesthetic management of a patient with Williams syndrome.

Case Report

A 7-year-old boy weighing 17.2 kg was scheduled for bilateral orchiopexy for repair of retentio testis. Since the age of 5, he had been pointed out to have supravalvular aortic stenosis and was diagnosed as Williams syndrome. The systolic pressure gradient between aorta and left ventricle was 20 mmHg. He had strabismus, epicanthal folds, long philtrum, thick lips, mandibular hypoplasia, and mental retardation with a developmental quotient of 3 years. A grade 2/6 systolic ejection murmur was observed at the area of aortic valve. The size of heart was normal on chest x-ray, but the ECG showed left ventricular hypertrophy.

He was premedicated with atropine 0.17 mg. Anesthesia was induced with 85 mg thiopental. After the administration of 12 mg of d-tubocurarine, endotracheal intubation was performed. Anesthesia was maintained with 66% nitrous oxide in oxygen, intermittent d-tubocurarine, and caudal anesthesia with 0.25% bupivacaine 10 ml and morphine 0.5 mg. The blood pressure in both arms was measured simultaneously with Riva-Rocci method.

The blood pressure before induction of anesthesia was 120/80 mmHg in the right arm and 80/60 mmHg in the left arm, the pressure gradient between both arms being 40 mmHg. After induction of anesthesia, the blood pressure changed to 94/58 mmHg in the right and 90/54 mmHg in the left. The systolic blood pressure gradient between both arms decreased to 0 to 10 mmHg during surgery. Vital signs remained stable throughout the operation. He became fully conscious, and the endotracheal tube was extubated after operation. The blood pressure was 114/70 mmHg in the right and 110/58 mmHg in the left at the end of anesthesia (figure 1). The postoperative course was uneventful.

Discussion

Williams syndrome is an association of supravalvular aortic stenosis, elfin face, and mental retardation (Williams triad). Idiopathic hypercalcemia, pulmonary stenosis, and seizures are also associated[1,2]. Angina and congestive heart failure are common complications because of the myocardial hypertrophy and fixed cardiac output[1]. Occasional sudden death by myocardial ischemia has been reported[1].

The difference of arterial pressure in both arms is referred to as the "Coanda effect," in which the jet of high-velocity blood stream produced by the supravalvular aortic stenosis is directed into the innominate artery, the right arm systolic pressure being greater than that of the left[1]. This arterial pressure

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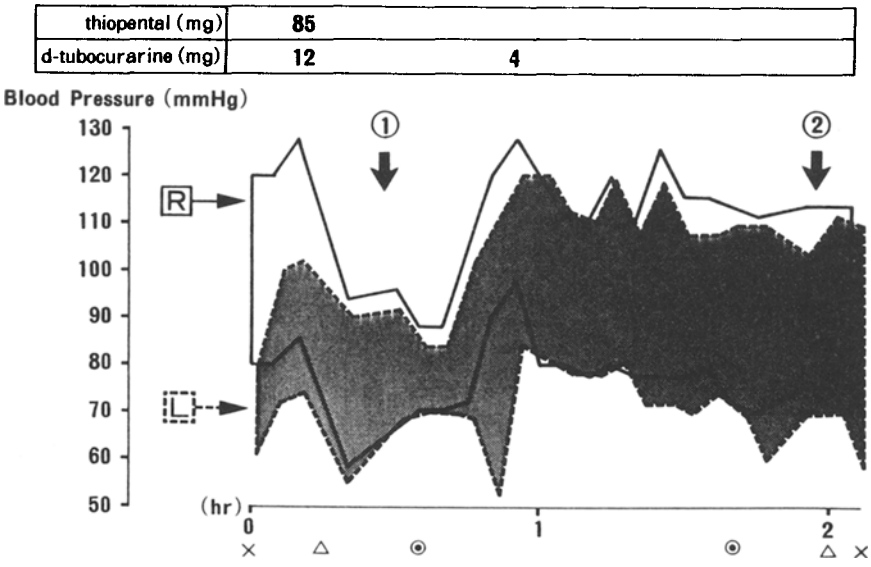

Fig. 1. The blood pressure in both arms of a patient with Williams syndrome
The systolic blood pressure gradient between both arms decreased after induction of anesthesia.
———, R: blood pressure in the right arm
– – – –, L: blood pressure in the left arm
① caudal anesthesia (0.25% bupivacaine 10 ml, and morphine 0.5 mg)
② atropine 0.6mg, and neostigmine 1.7 mg, intravenously

difference decreased from 40 mmHg of pre-induction period to 0 to 10 mmHg following induction of anesthesia, which persists even after emergence from anesthesia. This can probably be exlained by the decrease of high-velocity blood jet due to the decrease of cardiac output by anesthesia and also by the decreased effect of jet stream directed into the innominate artery secondary to the decrease of vascular resistance produced by caudal block[3].

The entire course of anesthesia was uneventful and it is our postulate that the general anesthesia combined with epidural block is the anesthetic technique of choice for patients with Williams syndrome.

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References

1. Rowe RD: Supravalvular aortic stenosis, Heart disease in infancy and childhood. Edited by Keith JD, Rowe RD, Vlad P. New York, Macmillan, 1978, pp.718–724
2. Cramolini GM: Systemic diseases affecting renal parenchyma. Anesthesia and uncommon pediatric diseases. Edited by Katz J, Steward DJ. Philadelphia, WB Saunders, 1987, pp.197–199
3. Payen D, Ecoffey C, Carli P, Dobousset AM: Pulse doppler ascending aortic, carotid, brachial, and femoral artery blood flows during caudal anesthesia in infants. Anesthesiology 67:681–685, 1987