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# Williams syndrome: anesthetic management for balloon dilatation of supravalvular aortic stenosis

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## Introduction

Williams syndrome is a very rare disease associated with an elfin face, mental retardation, mild growth deficiency, and supravalvular aortic stenosis (SAS). There are only two anesthetic reports on this syndrome. Kato et al. [1] described an anesthetic management for orchiopexy. Patel and Harrison [2] suggested a possible relation between malignant hyperthermia and this disease. Nevertheless, no report has been published on anesthetic considerations for the correction of SAS which can be a fatal aspect of Williams syndrome.

This is a case report of the anesthetic management for balloon dilatation angioplasty of SAS associated with an extremely high pressure gradient and bleeding tendency.

## Case report

An 8-year-old boy who weighed 18.5 kg was scheduled for balloon dilation angioplasty of SAS. He had a characteristic elfin face consisting of a broad forehead, a low nose, pouting lips and a small mandible. He had physical and mental retardation. Anticonvulsants had been administered since he had been diagnosed with epilepsy 5 years previously. Chest X-ray and electrocardiogram showed left ventricular hypertrophy. Nevertheless, no congestive change was noted in the lungs. Echocardiogram revealed moderate aortic valve stenosis and severe SAS. He was evaluated as NYHA class 2. Hematological examinations reported a decreased number of platelets (119 000/mm$^3$) and prolonged bleeding time (10 min). Though prothorombin time and activated partial thromboplastin time were normal, thromboelastography showed an increased K value (8.5 min) and a decreased MA value (42 mm) which suggested slight platelet dysfunction. Other laboratory data were within normal ranges.

The patient was premedicated with 3 mg of bromazepam suppositorium 1 h before induction. Cardiac surgeons and extracorporeal circulation team were standing by in case of emergency. Ten units of concentrated platelets were also in hand besides ordinary blood preparation. After the application of two leads of ECG (II, $V_5$), a noninvasive blood pressure monitor and a pulse oximeter, anesthesia was induced with fentanyl 300 μg, vecuronium 4 mg, and oxygen. Following a 4% lidocaine spray to the larynx, the trachea was intubated with a 6.0-mm non-cuffed Portex oral tube. Intubation was difficult due to the small mandible. Atropine 0.2 mg was administered for bradycardia caused by laryngoscopy. Anesthesia was maintained with total doses of fentanyl 30 μg·kg$^{-1}$, vecuronium 14 mg, midazolam 4 mg, and oxygen during the operation.

Prior to the balloon dilatation, a catheter study was carried out. The left ventricular systolic pressure was 250 mmHg, the poststenotic systolic pressure was 136 mmHg, thus the pressure gradient was 114 mmHg. Two parallel balloons were positioned at the stenosis. They were inflated with contrast media under the pressure of 6 atm for 10 s, then deflated immediately. During this procedure the poststenotic aortic pressure was recorded continuously through a catheter sheath introduced into the left femoral artery. Following every inflation, a transient ST segment depression lasting 15–30 s was observed. Also several premature ventricular systoles were treated with lidocaine 1 mg·kg$^{-1}$. Several prolonged periods of hypotension following deflation of

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the balloons were treated with ephedrine 2 mg. Fourteen consecutive angioplasties were completed in over 3 h. At the end of the procedure, the systolic pressure gradient decreased to 70 mmHg. The procedure was finished without any major complications. Vecuronium was antagonized with a mixture of atropine 0.5 mg and neostigmine 1.0 mg. Fentanyl was antagonized with naloxone 0.12 mg. The patient was closely observed in the ICU for a day, then transferred to the pediatric ward.

## Discussion

Williams et al. [3] reported four cases with broad foreheads, wide mouths, widely set eyes, pouting lips, and pointed chins. All were mentally retarded and had supravalvular aortic stenosis. Beuren et al. [4] added multiple peripheral pulmonary stenosis and identical dental malformations to this syndrome. The incidence of this syndrome is said to be one in 200 000–350 000 births [5]. SAS can induce left ventricular hypertrophy and pump failure by adolescence. Therefore, a surgical treatment of the stenosis is essential. It is common to perform an open angioplasty of the stenotic aorta for children of this age. Nevertheless, because of his coagulopathy, it was decided to employ a balloon catheter dilatation rather than an open angioplasty.

Angina and congestive heart failure could be common complications because of myocardial hypertrophy and fixed cardiac output [3]. During inflation of the balloons, the aorta was completely occluded. Extremely high left ventricular diastolic pressure obstructed coronary perfusion. Thus transient ST depressions and premature ventricular systoles were unavoidable. Anesthetic agents either intravenous or inhalational which have a myocardial depressive effect should be avoided. Due to the thickened myocardium, tachycardia could easily cause an increase in myocardial oxygen demand and ischemic change. We used a relatively high dose of fentanyl with a small dose of midazolam without nitrous oxide for induction and maintenance of anesthesia. This method was thought to be better than the conventional combination of thiopental and halothane or enflurane with nitrous oxide. We successfully kept the heart rate at less than 120 beats·min$^{-1}$ throughout the procedure. To give adequate warning to the operators, it was important to detect cardiovascular events as soon as possible. ECG using two leads (II, $V_5$), blood pressure, and arterial oxygen saturation should be monitored continuously.

Though the balloon catheter dilatation is a popular choice for the treatment of coarctation of the aorta in young infants [6], there has been no report on the balloon dilatation of SAS of Williams syndrome. As far as complications are concerned, the balloon dilatation for both diseases may have similar hazards. It is true that this procedure could cause some fatalities. There is a possibility of a perforation or a rupture of the aorta cuased by either a guide wire or an inflated balloon [7]. To cope with an emergency operation, a cardiovascular surgical team and an extracorporeal circulation team must stand by [8].

A sudden death after balloon dilatation angioplasty was reported [9]. Because of the prolonged effect of general anesthetics, either the patients cardiovascular or respiratory condition could deteriorate. Close monitoring and observation should be carried out postoperatively. The patient must be observed in the ICU for at least 24 h following the procedure [8,9].

In summary, an anesthetic management for balloon dilatation angioplasty of SAS with Williams syndrome was reported. It seemed to be suitable to use a relatively high dose of fentanyl with a small dose of midazolam without nitrous oxide for induction and maintenance of anesthesia. We emphasize the importance of close monitoring including a pulse oximetry during anesthesia and postoperatively.

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