Case Report
pISSN 2383-9309 ▮ eISSN 2383-9317
J Dent Anesth Pain Med 2023;23(2):111-116 ▮ https://doi.org/10.17245/jdapm.2023.23.2.111

General anesthesia with a transcutaneous pacemaker for a Noonan syndrome patient with advanced atrioventricular block discovered in the remote period after open-heart surgery: a case report

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We provided general anesthesia management to a patient with advanced atrioventricular block, which was discovered in the remote period after open-heart surgery. A 21-year-old man with Noonan syndrome was scheduled to undergo excision of a median intramandibular tumor. At 2 months of age, the patient underwent endocardial repair for congenital heart disease. During our preoperative examination, an atrioventricular block was detected, which had not been previously noted. Emergency drugs were administered, and a transcutaneous pacemaker was placed. During anesthesia induction, mask ventilation was easy, and intubation was performed smoothly using a video laryngoscope. The transcutaneous pacemaker was activated in demand mode at a pacing rate of 50 cycles/min approximately throughout the anesthesia time, and the hemodynamic status remained stable. The effect of intraoperatively administered atropine was brief, lasting only a few seconds. Although body movements due to thoracoabdominal muscle spasm were observed during pacemaker activation, they did not interfere with surgery. In postoperative patients with congenital heart disease, an atrioventricular block may be identified in the remote period, and preoperative evaluation should be based on this possibility. In addition, during anesthesia management, it is important to prepare multiple measures to maintain hemodynamic status.

Keywords: Anesthesia, General; Atrioventricular Block; Case Reports; Noonan Syndrome; Pacemaker, Artificial.

INTRODUCTION

Noonan syndrome is a genetic disorder that often presents with facial irregularities, such as ptosis, small mandibles, and short necks, as well as thoracic deformities and intellectual disabilities. Congenital heart defects, such as pulmonary stenosis and ventricular or atrial septal defects, are diagnostic requirements for Noonan syndrome.

Advanced atrioventricular block (AVB), a block with an atrioventricular conduction ratio of 3:1 or more, may lead to complete AVB or cardiac arrest [1]. Permanent pacemaker (PPM) implantation is recommended for patients with AVB who have clinical symptoms, such as dizziness or syncope.

During endocardial repair of congenital heart disease with ventricular septal defects, AVB can occur because

Received: November 7, 2022 • Revised: February 12, 2023 • Accepted: February 21, 2023
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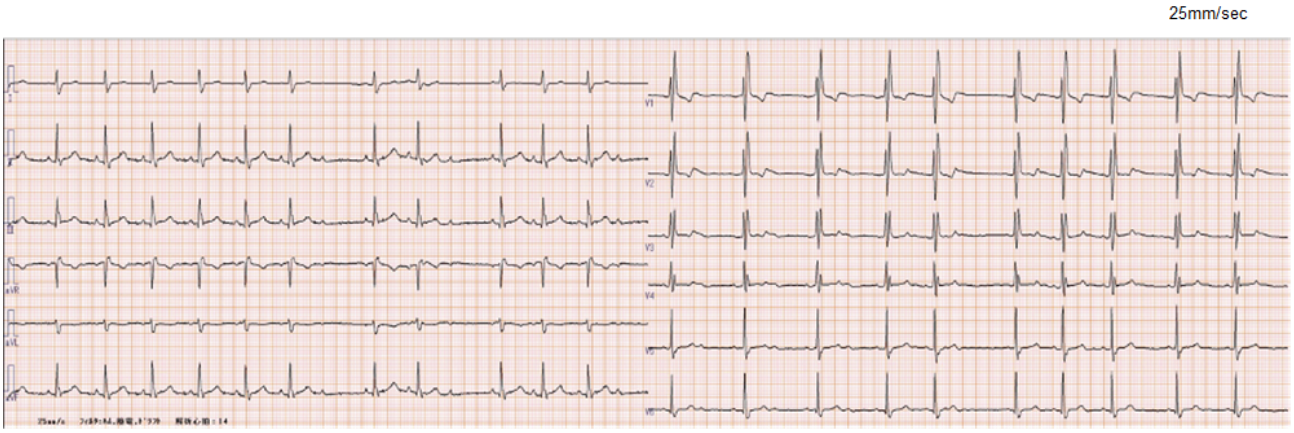


Fig. 1. A 12-lead electrocardiogram taken as our preoperative examination showing second degree Mobitz type II atrioventricular block in addition to complete right bundle branch block

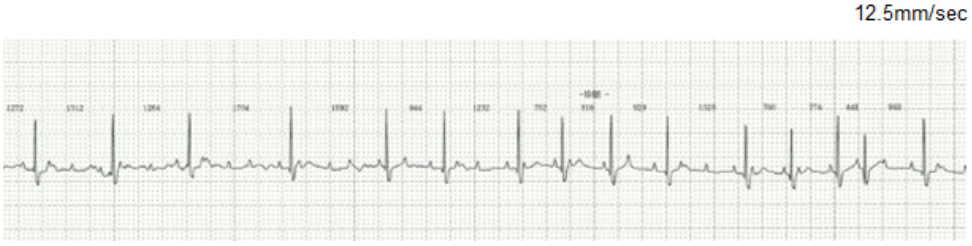


Fig. 2. Excerpt from the 24-h Holter electrocardiogram showing 2:1 and 3:1 atrioventricular block

of damage to the atrioventricular node or bundle of His, with an incidence of 1.4–7% [2,3].

In this report, we describe the general anesthesia management of a patient with Noonan syndrome who had an advanced AVB first revealed on preoperative examination more than 20 years after open heart surgery and who underwent dental surgery before PPM implantation was determined to be necessary or not.

CASE REPORT

A 21-year-old man, 153 cm in height and 58.3 kg in weight, complained of pain in the chin for several months prior to his visit to an oral surgeon at our hospital. He had a gingival fistula with oral infection, and intramandibular tumor resection was planned under general anesthesia.

At 2 months of age, the patient underwent closure of a ventricular septal defect and patent ductus arteriosus. Postoperatively, the patient developed neonatal ischemic renal failure due to left renal artery infarction. At 18 months, the patient underwent vesicoureteral neostomy for vesicoureteral reflux. Around the same time, he was diagnosed with Noonan syndrome due to congenital heart defects, short neck, and intellectual disability.

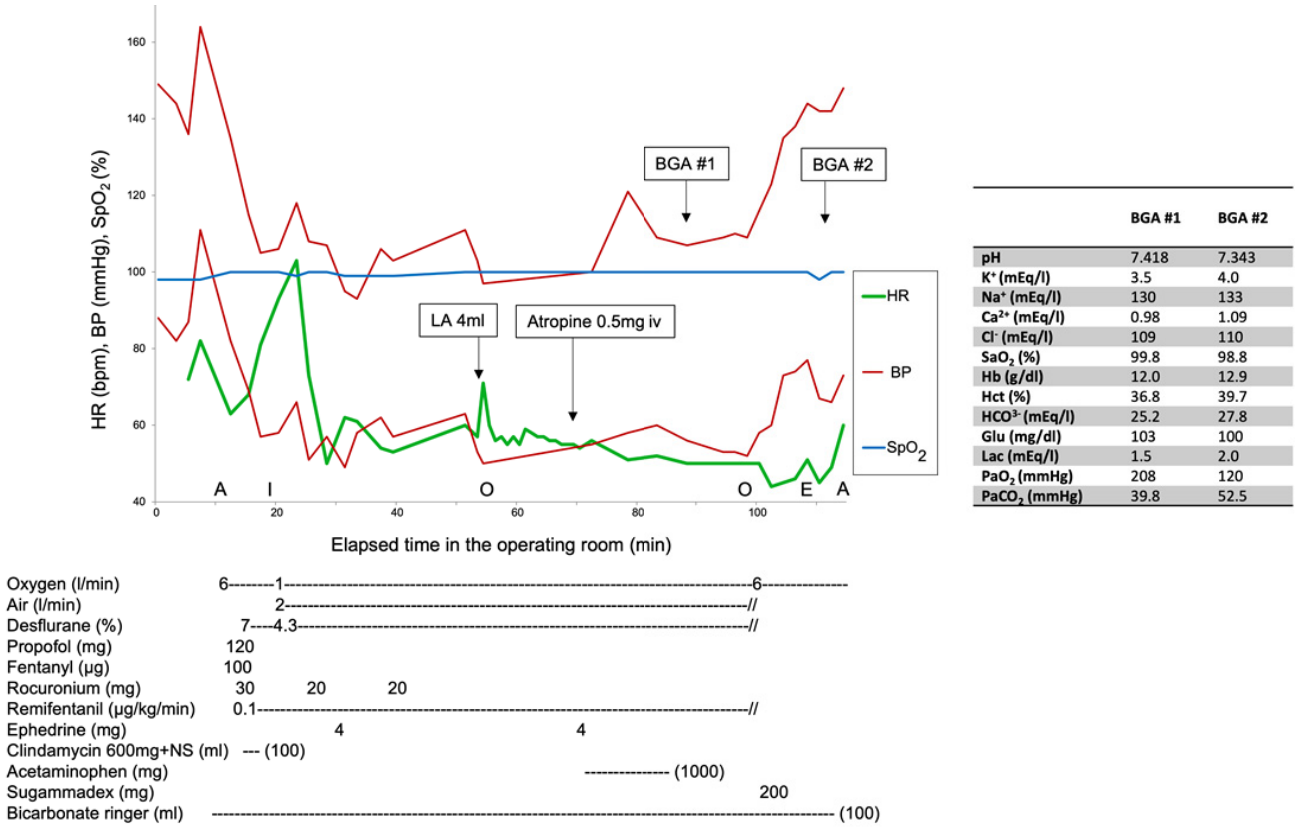
Blood tests showed no abnormal data, including normal renal function (serum creatinine, 1.03 mg/dL; estimated glomerular filtration rate, 78.4 mL/min/1.73 m^2). He was prescribed angiotensin-converting enzyme inhibitors because of persistent proteinuria.

A 12-lead electrocardiogram obtained preoperatively revealed a second-degree Mobitz type II AVB (Fig. 1). The patient had interrupted his visits to the pediatric cardiology department for approximately 3 years before the preoperative examination. We requested closer examination by the attending pediatric cardiologist. Echocardiography revealed no problems with cardiac function and no residual defects. A 24-h Holter electrocardiogram revealed 2:1 and 3:1 AVB (Fig. 2,

Table 1. The summary of the 24-hour Holter electrocardiogram analysis

HR	A minimum of 32 bpm, a maximum of 85 bpm and a mean of 53 bpm.
ST events	Nothing.
Arrhythmia	· The occasional multifocal PVCs occurred. · 2:1 and severe AVB were detected.
R–R interval prolongations of > 2.0 seconds	· 248 times/24-hour. · A maximum R–R interval was 3 seconds.

AVB, atrioventricular block; bpm, beats per minute; HR, heart rate; PVC, paroxysmal ventricular contraction.



	BGA #1	BGA #2
pH	7.418	7.343
K^+ (mEq/l)	3.5	4.0
Na^+ (mEq/l)	130	133
Ca^{2+} (mEq/l)	0.98	1.09
Cl^- (mEq/l)	109	110
SaO_2 (%)	99.8	98.8
Hb (g/dl)	12.0	12.9
Hct (%)	36.8	39.7
HCO^{3-} (mEq/l)	25.2	27.8
Glu (mg/dl)	103	100
Lac (mEq/l)	1.5	2.0
PaO_2 (mmHg)	208	120
$PaCO_2$ (mmHg)	39.8	52.5

Fig. 3. Anesthesia record. A, start and end of anesthesia; BP, blood pressure; BGA, blood gas analysis; E, extubation; HR, heart rate; I, intubation; LA, local anesthesia (2% lidocaine + 1/80,000 adrenaline); NS, normal saline; O, start and end of operation; SpO_2, saturation of percutaneous oxygen.

Table 1). However, the pediatric cardiologist decided that there was no urgent need for PPM implantation, and that the patient's future indications should be carefully considered because the patient had no subjective symptoms or history of AVB-induced syncope. The site of atrioventricular conduction defects remained unknown.

After discussion with the pediatric cardiologist, we decided to prioritize dental surgery to reduce the risk of infection in cases of PPM implantation. The pediatric cardiologist's opinion was that there would be no perioperative circulatory crisis. In addition to emergency medications, a transcutaneous pacemaker (TCP) was prepared and in-hospital support was confirmed in the event of a critical situation.

Upon entering the operating room, the patient's heart rate was 70 beats/min (bpm), and the Mobitz type II AVB was scattered. The patient was cooperative while securing the venous route and applied a pacing pad. General anesthesia was induced with fentanyl, propofol, and rocuronium (Fig. 3). Mask ventilation was easy, and intubation was performed using a video laryngoscope (McGRATH MAC™). General anesthesia was

maintained with desflurane and remifentanil in a mixture of air and oxygen (Fig. 3).

After anesthesia induction, pacing was confirmed to be effective at a pacing output of 125 mA in collaboration with a clinical engineer. When the pacing rate was set at 50 cycles/min in the demand mode, TCP was activated soon after the surgery started.

During the TCP activation, body movements due to thoracoabdominal muscle spasms were observed. Although 40 mg of additional rocuronium was administered, body movements could not be completely controlled. Further muscle relaxants were not added, and the pacing settings were not changed because the surgeon commented that body movements would not interfere with the surgery. When atropine (0.5 mg) was administered, a brief increase in the heart rate was observed up to 60 bpm, lasting only a few seconds. Arterial blood gas analysis revealed no abnormal values, and TCP was used for 84 min until his heart rate increased just before awaking from anesthesia and was activated 22 times for a total duration of approximately 20 min.

After discontinuation of the anesthetic agents, the patient recovered rapidly. After extubation, the heart rate was 44–60 bpm, and the blood pressure was stable at 130–140/60–70 mmHg. The operation time was 42 min, and the anesthesia time was 104 min. He did not complain of palpitations or discomfort after returning to the ward and was discharged the following day.

DISCUSSION

In this case, we safely managed a patient with Noonan syndrome, first identified preoperatively as having advanced AVB while maintaining his circulatory dynamics.

AVB that are seen after endocardial repair of congenital heart disease do not always occur immediately after surgery but can occur even in the remote period, although infrequent (0.3–0.7%) [2,3]. This patient had no history of AVB for approximately 18 years after undergoing ventricular septal defect and patent ductus arteriosus closure and had stopped regular visits to the hospital. Thus, it is important to keep in mind that AVB may be revealed in postoperative patients with congenital heart disease during the perioperative period, even if it was not previously detected.

According to the guidelines, “decisions about the need for a pacemaker are necessarily influenced by the presence or absence of symptoms directly attributable to bradycardia” [4]. With regard to the recommendations for permanent pacing, asymptomatic third-degree AVB with an average ventricular rate of 40 bpm or faster when awake would be considered class II (there is conflicting evidence and/or a divergence of opinion), whereas bradycardia with symptoms presumed to be due to AVB would be class I (there is evidence and/or general agreement). As observed in the present case, even in cases with relatively high recommendations in the guidelines, PPM may not be immediately indicated in all patients. Careful perioperative planning is essential for anesthetic management in such cases.

Generally, atropine is the first choice of treatment for bradycardia [5]. However, if the site of atrioventricular conduction disturbance is peripheral to the bundle of His, not only is atropine ineffective, but atrioventricular conduction may be exacerbated by an abnormally prolonged refractory period and increased atrial rate [1, 6,7]. The guidelines [5] state that percutaneous pacing is the first choice for bradycardia in patients with second-degree or more severe AVB, and atropine should be a temporary measure before pacing. In this patient, the site of the atrioventricular conduction defect was not identified, and it was not possible to preoperatively determine how atropine would work. We subsequently administered atropine in situations that were effective in controlling the heart rate with TCP to determine whether atropine would be effective when used after awakening. However, this approach was largely ineffective. Therefore, when managing general anesthesia in patients with AVB in which the site of atrioventricular conduction disturbance has not been identified, as in this case, it is important to administer atropine with caution and to

prepare for other drug therapies, such as isoproterenol, dopamine, dobutamine, epinephrine, [8] and other nonpharmacologic therapies.

Nonpharmacological therapies include transvenous pacing and the use of TCP. Some case reports suggest that a catheter should be inserted before surgery if an advanced AVB is known preoperatively [7,9,10]. However, in this case, the catheter was not inserted preoperatively because the pediatric cardiologist evaluated that perioperative circulatory crisis would not occur. In addition, we wanted to minimize invasiveness, emotional stress, and length of hospital stay with consideration of intellectual disability. Problems with the use of TCP include inadequate pacing, body movement, and burns. It has been reported that body movement during TCP operation causes problems during the operation [11]. In such cases, preoperative insertion of a transvenous catheter may be required. Fortunately, in this case, the body movement caused by the TCP did not interfere with the surgery. Nevertheless, it is important to discuss the surgical technique and body's tolerance to movement with the surgeon in advance.

In summary, we managed general anesthesia in a patient with Noonan syndrome who was preoperatively diagnosed with advanced AVB before PPM implantation, which was determined to be necessary. In postoperative patients with congenital heart disease, AVB may be identified in the remote period, even if it was not previously reported. Thus, preoperative evaluation should consider the possibility of an AVB being present. In addition, for perioperative anesthesia management, it is important to prepare for multiple measures, including nonpharmacological therapies, such as TCP, along with drugs, to maintain hemodynamic status.

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CONSENT: Written consent was obtained from the patient's parents for this report. We received a Certificate of Exemption from the Ethical Review Board for Life Science and Medical Research at Hokkaido University Hospital regarding the ethical issues of this case report.

DECLARATION OF INTERESTS: All authors have no conflicts of interest to disclose.

FUNDING: None.

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