Anesthetic Management of Neonates With Congenital Complete Atrioventricular Heart Block Undergoing Pacemaker Implantation

Jan Ammann, MD,* Joachim Winter, MD, PhD,† Ulrich Sunderdiek, MD, PhD,† and Stephan A. Loer, MD, PhD*

CARDIAC OUTPUT OF infants depends on a high heart rate (140-160 beats/min), and decreases in heart rate can only partially be compensated for by increases in stroke volume of the stiff neonatal ventricles. Children with congenital complete heart block, however, have a low heart rate (often <60 beats/min) and are therefore at increased risk of cardiac failure. This applies particularly to the perioperative period when pharmacologically and surgically induced changes in contractility, preload, and afterload occur. Thus, a thorough understanding of the cardiovascular pathophysiology and treatment options of infants with complete heart block is necessary for adequate perioperative care of these children.

The incidence of congenital complete heart block is estimated to range between 0.5 and 1 in 10,000 births.[1,2] Heart block may exist alone or in combination with other congenital malformations.[1,2] It is associated with maternal anti-SS-A/Ro and anti-SS-B/La antibodies, which are transferred via the placenta and which induce an inflammatory tissue injury within the fetal heart.[3] This inflammatory reaction can result in a fibrosis of the atrioventricular node with subsequent conduction abnormalities as well as a progressive endocardial fibrosis.[2-4] In some infants, not only the atrioventricular node but also the sinoatrial node are involved.[5] Mothers of these children often suffer from diseases like lupus erythematosus or Sjögren's syndrome but may also be asymptomatic.[1,2,4,6,7] Prenatal echocardiography allows diagnosis within the 24th to 26th week of gestation so that therapy can be started in utero.[4] Treatment options include corticosteroids to prevent further inflammatory tissue injury, β-sympathomimetics to increase fetal heart rate, plasmapheresis to reduce or eliminate circulating maternal antibodies, and in utero pacing.[1,2,7-9] Often, however, the heart block is irreversible and the affected children require lifelong pacemakers, which are implanted within the first weeks of their lives.[10-12] Because these children are at an increased perioperative risk for cardiac decompensation, 2 cases of neonates requiring pacemaker implantation are reported and possible implications for the perioperative management are discussed.

CASE REPORTS

Patient 1

The female neonate was delivered by cesarean section in the 36th week of gestation. The complete heart block had been diagnosed in the 16th week of gestation, and her ventricular rate during pregnancy, birth, and the time until pacemaker implantation ranged between 54 to 68 beats/min. Her mother suffered from Sjögren's syndrome and had a history of another infant with complete congenital heart block in a previous pregnancy who died after birth. During pregnancy, she received a corticosteroid medication to prevent further inflammatory tissue injury of the fetal heart as well as β-sympathomimetics to increase fetal heart rate during the last weeks of the pregnancy. After cesarean section, the neonate developed respiratory distress syndrome requiring tracheal intubation and mechanical ventilation. On arrival at the pediatric intensive care unit, her skin color was pale, systolic blood pressure was 60 mmHg, and all 4 extremities were cold. Attempts to increase her heart rate by the administration of atropine (0.04 mg) were not successful, and an infusion of isoproterenol (0.17 and 0.36 μg/kg/min) increased her heart rate only by 5 to 10 beats/min without improvement of clinical signs. An echocardiogram and chest x-ray showed an enlarged heart. On the second day of her life, urinary output ceased and she developed a metabolic acidosis (pH 7.23, BE 12.7 mmol/L, HCO_3 concentration 14.2 mmol/L), indicating pending cardiopulmonary decompensation. Her skin color was still pale, and her extremities were still cold. Therefore, she was scheduled for urgent pacemaker implantation.

On arrival in the anesthetic department, her heart rate was 68 beats/min and her blood pressure 60/30 mmHg. Her body weight was 2.3 kg, hemoglobin concentration was 14.3 g/dL, and her metabolic status and electrolytes had been corrected. In addition to a cannula that had been placed before arrival in the operating room in a cephalic vein, a 24-G cannula was placed in a vein of the back of her right hand and isoproterenol was infused at a rate of 0.36 μg/kg/min. An external pacemaker (Oyspka Pace 500 D; Dr Osypka GmbH, Rheinfelden-Herten, Germany) was prepared in case additional external emergency pacing would become necessary before implantation of a permanent pacemaker. During the perioperative period, electrocardiogram, pulse oximetry (probe placed at an earlobe and at a hand), and esophageal body temperature were monitored continuously, and noninvasive arterial blood pressure was monitored intermittently every 3 minutes. To maintain normothermia, the room temperature was increased to 24°C, and the girl was placed on a warm air bed (Bair Hugger; Arizant Healthcare, Eden Prairie, MN).

Anesthesia was induced and maintained with sevoflurane (end-tidal concentration 2.0-2.5 vol%). The lungs were mechanically ventilated with oxygen in air (F_IO_2 0.5) with a tidal volume of 10 mL/kg of body weight at a frequency of 30 to 40 breaths/min. End-tidal CO_2 concentration was maintained between 36 and 40 mmHg. Peripheral oxygen saturation measured at the left hand was above 95% throughout the procedure. She received an infusion of one third glucose 5% and two thirds Ringer's solution (9 mL/kg/h). Intraoperative electrocardiogram showed a third-degree atrioventricular block with an atrial rate of 140 beats/min and a ventricular rate of 68 beats/min. Systolic blood pressure during the course of anesthesia ranged between 60 to 66 mmHg.

After implantation of a permanent VVI pacemaker via the right subclavian vein (St. Jude Medical, Sylmar, CA; bipolar screwing lead Medtronic 5076, Minneapolis, MN), her heart was paced with a rate of 100 beats/min (Figs 1 and 2), and she was transferred to a pediatric intensive care unit (ICU). She was extubated 1 day after the implantation without any complications. The child was discharged from the pediatric ICU on day 3 after pacemaker implantation.

*From the Departments of *Anesthesiology and †Thoracic and Cardiovascular Surgery, University Hospital Düsseldorf, Düsseldorf, Germany.*

Address reprint requests to Stephan Alexander Loer, MD, PhD, Klinik für Anaesthesiologie, Universitätsklinikum Düsseldorf, Moorenstr 5, 40225 Düsseldorf, Germany. E-mail: loer@med.uni-duesseldorf.de

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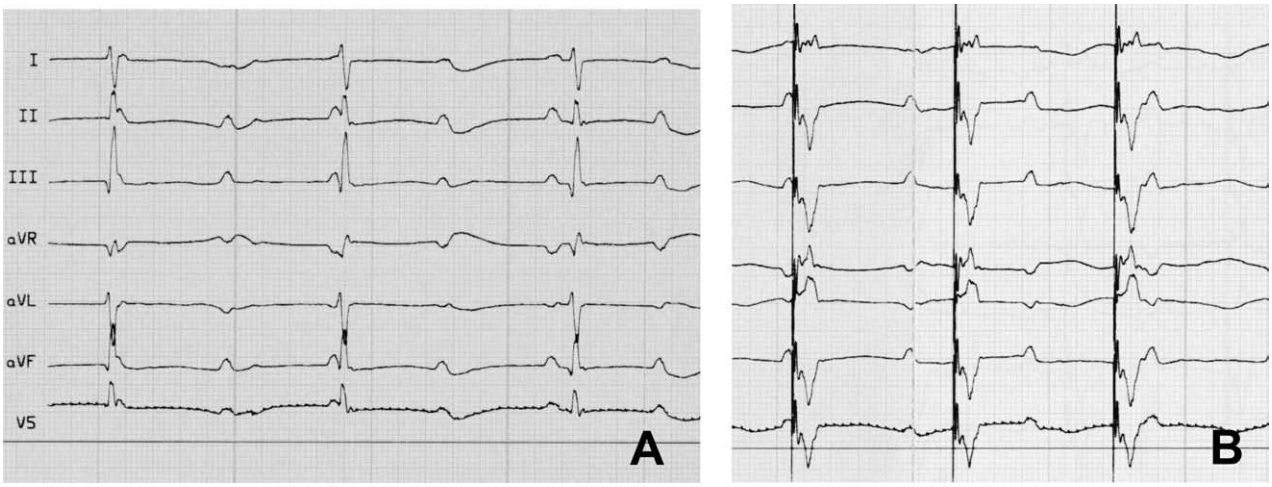

Fig 1. (A) Intraoperative electrocardiogram of a 2-day-old anesthetized girl with congenital complete heart block showing an atrial rate of 140 beats/min and a ventricular rate of 68 beats/min before lead implantation. (B) After pacemaker implantation, the girl was paced with a rate of 100 beats/min by the implanted VVI pacemaker.

Patient 2

The second neonate was also delivered by cesarean section with a complete heart block but no other heart malformation in the 38th week of gestation. The heart block had been diagnosed in the 24th week of gestation, and the heart rate during pregnancy, birth, and the time until pacemaker implantation ranged between 48 to 54 beats/min. Her mother suffered from an autoimmune disease without any clinical manifestations. In a previous pregnancy, she had also delivered an infant with a complete congenital heart block who died within the first weeks of his life. To prevent further inflammatory tissue injury of the fetal heart, she had received a corticosteroid medication during the last weeks of the pregnancy. After the cesarean section, the girl showed no signs of congestive heart failure; however, attempts to increase her heart rate pharmacologically failed so she was scheduled for elective pacemaker implantation.

At the age of 26 days and with a body weight of 3 kg, she arrived in the operating room without any signs of heart failure. Her heart rate was 50 beats/min, and her blood pressure was 60/28 mmHg. Hemoglobin concentration (17.5 g/dL) and other values were within the normal range. Before induction of general anesthesia, a 24-G cannula was placed in a vein of her right foot and an external pacemaker (Oyspka Pace 500 D) was prepared in case external pacing became necessary. Electrocardiogram, pulse oximetry, and body temperature were monitored continuously, and arterial blood pressure was monitored intermittently every 3 minutes. After preoxygenation, anesthesia was induced with 4 mg/kg of thiopental and maintained with sevoflurane (end-tidal concentration 2.5-2.8 vol%). Thereafter, her trachea was intubated with a cuffed endotracheal tube (internal diameter 3.0 mm), and the lungs were mechanically ventilated with a tidal volume of 10 mL/kg body weight at a frequency of 30 to 40 breaths/min maintaining end-tidal CO_2 between 38 and 43 mmHg. Peripheral oxygen saturation was above 94% during mechanical ventilation with 50% oxygen in air. A 20-G catheter of 8-cm length was introduced into the inferior vena cava via the right femoral vein using a guidewire. This catheter was placed to allow infusion of vasoactive drugs and to attempt transvenous emergency pacing if required. During the operation, she received an infusion of one third glucose 5% and two thirds Ringer's solution (7 mL/kg/h). Before skin incision, 1 μg/kg of fentanyl was administered intravenously. Intraoperative electrocardiogram recording showed a third-degree atrioventricular block with an atrial rate of 120 beats/min, a ventricular rate of 50 beats/min, and a long QT-interval (above 600 milliseconds) (Fig 3). Systolic blood pressure during the implantation of a pacemaker lead ranged between 60 to 70 mmHg.

After implantation of the pacemaker lead, her heart was paced with a rate of 70 beats/min and she was transferred to a pediatric ICU. In the ICU, her heart rate was increased stepwise to 100 beats/min. This procedure was chosen to allow adaptation of her cardiovascular system to the increased heart rate. She was extubated in the afternoon of the same day without any complications.

DISCUSSION

Neonates with complete congenital heart block undergoing general anesthesia may have an increased risk of cardiac complications. Their heart rate is fixed at low rates (often below 60 beats/min) so that any changes in volume loading, contractility, or systemic vascular resistance may predispose to cardiac failure. Although neonates with complete congenital heart block show some degree of compensatory adaptation to the persisting slow ventricular rate in the form of an increase in fractional shortening and ventricular size already in utero,[13,14] anesthetics and the operative procedure may predispose to cardiac decompensation. Because the incidence of this abnormality is low, recommendations on the perioperative management of these infants are mainly based on case reports.[15-18]

Neonates in general have an increased sensitivity to volume loading and a poor tolerance to changes in systemic vascular resistance. This applies particularly to neonates with a low and fixed heart rate because of congenital heart block. Thus, in addition to the principles applying to all neonates requiring general anesthesia (strict control and maintenance of body temperature, normovolemia, normocapnia, and normoxia), specific effects of the administered anesthetics on the cardiovascular system should be considered and pharmacologic as well as electrical measures to increase heart rate should be prepared before induction of general anesthesia.

Most volatile and intravenous anesthetics influence myocardial contractility, heart rate, and systemic flow resistance so