Case Report

Transposition of The Great Arteries, Situs Inversus with Congenital Complete Heart Block: Initial Management and Referring to a Special Unit

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Received: Jan 22, 2021
Revision received: Mar 03, 2021
Accepted after revision: Aug 06, 2021
BKK Med J 2020;17(2): 150-154.
DOI: 10.31524/bkkmedj.2021.22.003
www.bangkokmedjournal.com

Abstract

A case of a term female newborn, 40-week gestational age (by ultrasound (US)) with prenatal diagnosis of situs inversus and levocardia is reported. Transposition of the great arteries (TGA) and irregular fetal bradycardia were prenatally detected. Immediately after birth, electrocardiogram (ECG) monitoring revealed a complete heart block with no sign of hemodynamic compromise. The patient was then transferred to the neonatal intensive care unit (NICU) for ventilatory support. Close observation and further investigation with echocardiogram were undertaken. The patient was transferred by air at the age of 4 days to the Queen Sirikit National Institute of Child Health for definitive cardiovascular treatments. This paper describes how to overcome the challenges of transporting a very young neonate with complex cardiac problems.

Keywords: congenital complete heart block, transposition of the great arteries, air transportation, sinus inversus, levocardia

Congenital complete heart block (CCHB) appears 1 in 20,000 to 1 in 25,000 live births.[1] Half of the infants with congenital complete heart block have associated cardiac malformations.[2,3] Even though neonates frequently tolerate 50-60 beats per minute (bpm) with no hemodynamic compromise, most patients who have CCHB and congenital heart diseases require pacemaker therapy in the early neonatal period.[2,4] We report a newborn with CCHB and complex cardiac diseases who required air transportation from Bangkok Hospital Udon to the National Pediatric Cardiac Center in Bangkok for definitive treatment.

Case report

A term female newborn, 40-week by US was delivered by cesarean section at Bangkok Hospital Udon. The APGAR scores were 8, 9, 9 at 1, 5, 10 minutes respectively with the birth weight of 3,510 grams.

Her mother was a 24-year old Laotian woman, $G_2P_0A_1$, with no underlying disease. The mother had her 1st antenatal care (ANC) at Bangkok Hospital Udon at the gestational age (GA) of 14-week, and due to COVID-19 pandemic, had the rest of her ANC in Lao People Democratic Republic (Lao PDR). She has blood group B with Rh positive. Blood tests for venereal disease research laboratory (VDRL) and anti-HIV were non-reactive and HBsAg test was negative.

The GA of 26-week by US, an examination performed at a Lao PDR medical care clinic, revealed regular heartbeats with a fetal heart rate (FHR) of 138 beats per minute (bpm). The US revealed that the fetal stomach was on the right side while the fetal liver was on the left side of the body, indicating situs inversus.

At the GA of 36-week by US, the examination at a Lao PDR medical care clinic revealed irregular heartbeat with FHR of 58 bpm and situs inversus. The mother requested to be transferred to Bangkok Hospital Udon for further treatments.

At Bangkok Hospital Udon, GA of 38^{+3} week by US, the examination showed situs inversus, levocardia, and TGA. The FHR was 58-62 bpm with irregular fetal bradycardia. There was no sign of hydrops fetalis.

A conference to plan for delivery was set up with a multidisciplinary team consisting of an obstetrician, a neonatologist, and a pediatric cardiologist. The delivery was planned at the GA of 40-week by cesarean section and an emergency management was prepared.

After birth, crying and good tone were noticed. The baby received initial steps in stabilization (keeping warm and maintaining normal temperature in a radiant warmer, positioning, clearing airway, keeping dry, stimulation and ECG monitoring). At the 1st minute assessment, the heart rate was 68-70 bpm, the oxygen saturation (SpO2) was 75% with mild respiratory distress. Oxygen supplement was initiated via a nasal continuous positive airway pressure (CPAP) with pressure of 5 cmH_2O. The baby was transferred to the NICU with cutaneous SpO2 of 88%. At the NICU, the baby was found to have a systolic ejection murmur grade II/VI at left upper parasternal border, her systolic blood pressure was 76 mmHg with the mean arterial pressure of 49 mmHg. The twelve lead ECG (Figure 1) indicated atrioventricular dissociation with atrial rate of 150 bpm and ventricular rate of 70 bpm, this suggested complete atrio-ventricular block.

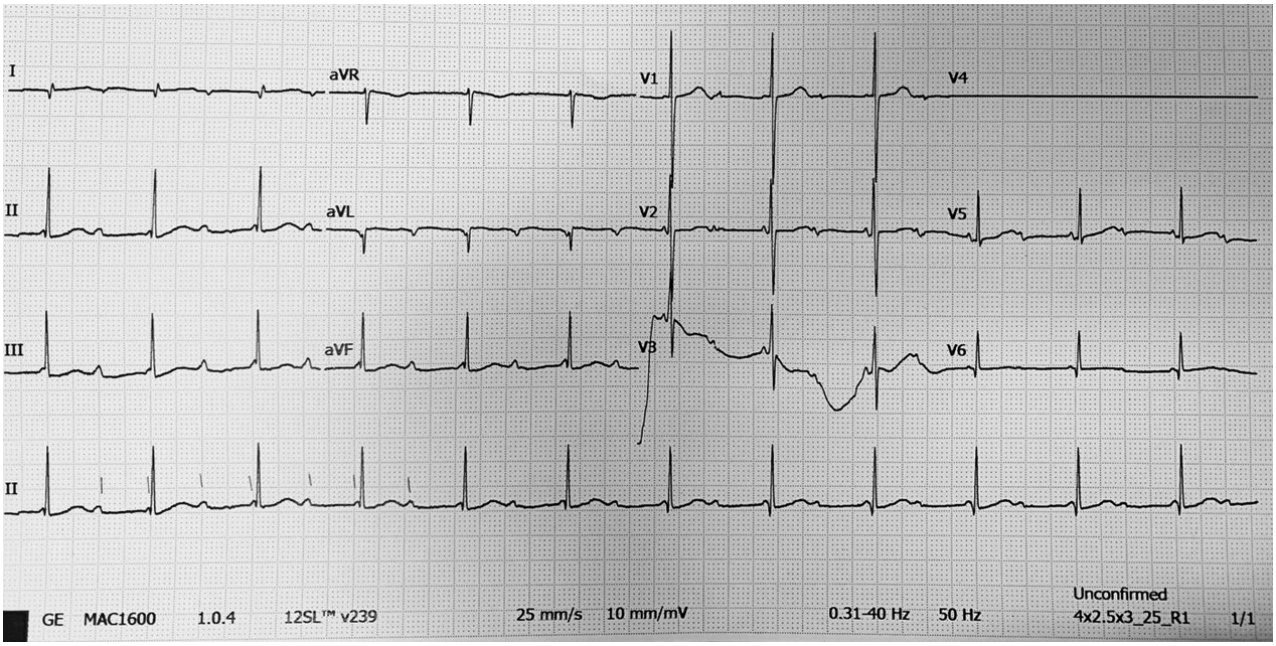

Figure 1: Twelve-lead ECG day 1 after birth

Transthoracic echocardiogram (TTE) showed situs inversus, levocardia, atrial septal defect (ASD) 8 mm with left to right shunt, ventricular septal defect (VSD) 8 mm with left to right shunt, discordance of ventriculo-arterial connection representing a typical dextro-TGA [d-TGA is a potentially life-threatening birth defect in the large arteries of the heart], left aortic arch, and patent ductus arteriosus (PDA) 3.5 mm with left to right shunt.

The chest x-ray at 1 hour (Figure 1) after birth showed mild bilateral perihilar reticular opacities, which were compatible with the transient tachypnea of the newborn (TTNB). The stomach shadow located at the right upper quadrant (RUQ) whilst the liver shadow located at the left upper quadrant (LUQ) of the abdomen. Abdominal ultrasonography (Figure 2) confirmed the position of the liver on the left subphrenic area. The spleen was visualized on the right subphrenic area. Normal appearances of the liver and spleen were also demonstrated. The final diagnosis was a term female newborn 40-week by US with appropriate for GA with situs inversus, TGA, ASD, VSD, PDA, CCHB, and TTNB.

Initial managements included non-invasive respiratory support via heated humidified high flow nasal cannula (HHHFNC) with fraction of inspired oxygen (FiO2) 0.21-0.3, flow 4-6 L/min to maintain pre-ductal SpO2 to more than 85%. Given the hemodynamic stability with d-TGA, ASD, VSD, PDA and congenital complete heart block (the heart rate was 62-78 bpm), the newborn was closely observed in the NICU with ECG monitoring and was prepared to be transferred to a special unit for permanent internal pacemaker placement.

Transportation

After coordinating between hospitals within Bangkok Dusit Medical Services (BDMS) and consulting the specialist opinions (a neonatologist, a pediatric cardiologist, and transportation medicine experts), the newborn was air-transported via a helicopter to shorten the transportation time. The air transport team comprised a neonatologist, a special nurse and an air medical transport physician. The emergency equipment and planning were prepared. Pre-transport transthoracic echocardiogram (TTE) was repeated and it revealed the same

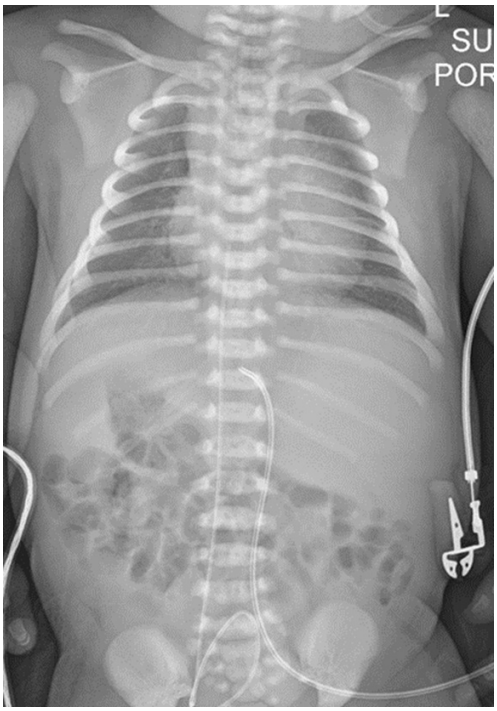

Figure 2: Portable chest x-ray at 1 hour after birth.

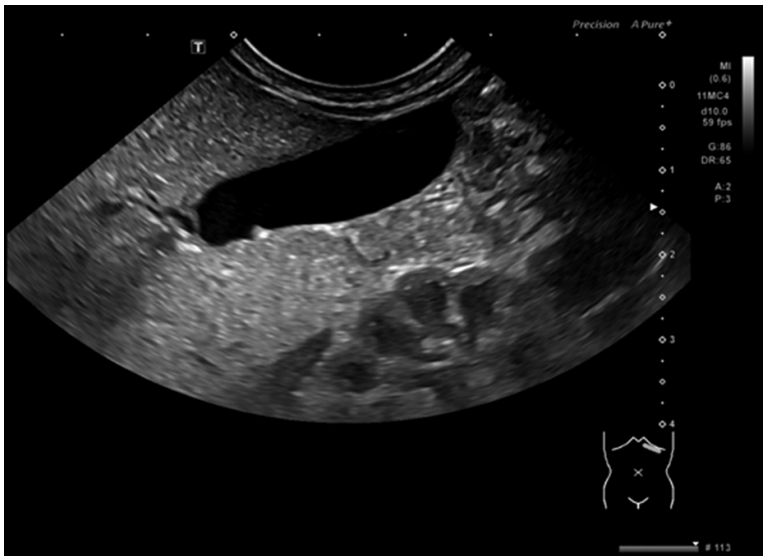

Figure 3: Abdominal ultrasound demonstrating the liver located at left subphrenic region

results as earlier. Usual resuscitation kit including airway management, ventilatory support, and circulatory support were made available. Emergency drugs and a temporary transcutaneous pacing were also prepared and made ready to use including a PGE1. These are a precaution because the patient did not have cyanosis and the presence of large ASD would allow mixing of oxygenated and de-oxygenated blood thus the patient was expected to be stable even if PDA were closed during the flight. The transportation was done when the patient was 4 days old. A nasal CPAP with pressure of 5 cmH_2O was implemented for respiratory support during the transportation and oxygen saturation of 85-90% was maintained because too much oxygen would cause pulmonary vasodilation and decreased systemic blood flow. The transportation was made during day time due to the availability of management. Transportation via a helicopter was chosen because it shortened the transportation time to only a few hours. And instead of fixed-wing aircrafts, helicopters fly at much lower altitude (4,000 - 5,000 feet compared with 20,000 - 30,000 feet of those fixed wings) where partial pressure of oxygen is not so different from ground level pressure.[5] At sea level, effective oxygen percentage is 20.9%. At 4,000 feet above sea level, partial pressure of oxygen is equivalent to 17.9% O_2 at sea level compared with 9.7% at 20,000 feet above sea level.[5] The team anticipated that flying at this lower altitude would have minimal effects for the patient because of the presence of a large ASD and the lower partial pressure of oxygen during the flight could be corrected by oxygen supplementation. Remaining issues that might have occurred included bradycardia, hypotension, and heart failure. The team was careful and vigilant on monitoring heart rate, blood pressure, respiratory rate, and oxygen saturation during the journey.

Discussion

Congenital complete heart block with complex structural heart disease (d-TGA, ASD, VSD, PDA in this case) is an emergency medical condition, which should be managed by a team of specialized physicians and highly-trained hospital personnel. In this case, the managements were divided into 4 phases including prenatal, during delivery, post-natal, and transportation period.

Prenatal phase

A conference that was set up with an obstetrician, a neonatologist, a pediatric cardiologist, and a cardiothoracic surgeon to plan for managing the first golden hour thoroughly and in a timely manner was very helpful. Intrauterine transfer was the most secure transportation. However, during the COVID-19 epidemic, according to the instructions issued by the Center for COVID-19 Situation Administration of Thailand, it would have been too complicated to transfer a Laotian pregnant woman to the Queen Sirikit National Institute of Child Health in Bangkok, Thailand. As a result, the team decided to deliver the baby at Bangkok Hospital Udon then transfer the newborn to the tertiary medical center. Appropriate staffing and equipment were prepared. In case of symptomatic bradycardia, an external cardiac pacemaker was ready to be used and the coordination with the hospitals capable of inserting an internal pacemaker was made.

ISSN 2287-0237 (online)/ 2228-9674 (print)

During delivery phase

Having a neonatologist serving as the leader of the transfer team was an excellent decision. Special equipment was prepared including an ECG and a defibrillator for external (transcutaneous) pacing in the case of emergency. The treatments were planned in accordance with the neonatal resuscitation 2020 and bradycardia guidelines in the Pediatric Advanced Life Support (PALS).

Post-natal phase

To ensure harmony, three principal goals were set. They were:

1. To maintain hemodynamic stability
2. To provide ventilator support
3. To correct electrolyte imbalances

In this case, the newborn was not cyanotic as other TGA cases because of high left to right shunt ratio that emerged from large ASD and VSD. The PDA was still open but neither the open or closed status affected hemodynamics and oxygen transportation. The newborn was able to tolerate a heart rate of 60-70 bpm without hemodynamic instability. However, the hemodynamics could be compromised due to decreasing of pulmonary pressure resulting in increasing left to right shunt and slow heart rate leading to heart failure.[6] From the condition, it was necessary to transport the newborn to the special care unit for internal pacemaker placement. And according to ACC/AHA/HRS 2008 guidelines, permanent pacemaker implantation is indicated for congenital third-degree AV block in the infant with congenital heart disease and a ventricular rate less than 70 bpm.[7]

Transportation phase

The team compared air and ground transportation (by an ambulance) and focused on the significant difference of transportation time; it would take almost 8 to 10 hours for the ground transportation while the air transportation would take only a few hours. Time is a key factor and can determine the difference between life and death in severe disease. Eventually, air transportation was chosen with a neonatologist onboard to support the patient during the flight. According to Hirakawa E.[8] neonatal transportation via air is as safe as ground transportation, the time taken to provide first aid and intensive care is significantly reduced compared to ground transportation. Air transportation could also contribute to prevention of intraventricular hemorrhage (IVH) in neonatal transportation. Air medical transportation can be by airplane or helicopter. Airplane is usually used when patient is transported for a long distance, for example, between countries.[9] A helicopter was used in this case, and since it is a low flying aircraft, the effects of high altitude on the baby's oxygen saturation is minimal and should not interfere with the baby's hemodynamic status even in a newborn with complex congenital heart disease. The co-ordination was set up with the destination hospital to be prepared for the transfer. Schedule, clinical information, medical history and previous managements were communicated. The emergency managements and equipment were prepared including resuscitation drugs (especially atropine and epinephrine). In case of bad weather or when the flight is unavailable, ground transportation may be used. In the latter scenario, more preparation would be needed because of longer transportation time. An external pacemaker, a defibrillator, and emergency drugs should be available as well as co-ordination with hospitals along the route to the destination should be made to prepare for any emergency conditions.

The first priority during transfer is the safety of the patient. Prevention of hypothermia, hypoxemia, hypovolemia, hypoglycemia, and acidosis by applying the **S.T.A.B.L.E** program.

S: **sugar and safe care.** Maintain the balancing of fluid and blood sugar. Assessment of volume status and blood sugar level before referring were needed. Prepare lines (umbilical arterial catheter and umbilical vein catheter) for drug or intravenous fluid infusion.

T: **temperature.** Controlling the suitable temperature by setting the patient in the transport incubator. Set the temperature according to the neutral thermal environment (NTE). In this case, the transport incubator was set temperature at 30 degrees Celsius and 50% humidity.

A: **artificial breathing.** Support ventilation by nasal CPAP with suitable pressure support and FiO2

B: **blood pressure.** Monitoring blood pressure and circulation. Observing and treating heart failure that might occur.

L: **lab work.** Evaluation of laboratory results to guide the treatments.

E: **emotional support.** Explain to the family the necessity and risk of the transportation. It would be better if the parents could accompany their baby. However, due to limited capacity in the aircraft, the parents took a commercial aircraft afterwards.

Conclusion

A term female newborn, 40-week gestational age (by US) with prenatal diagnosis of situs inversus and levocardia was transferred four days after birth from a province in northeast Thailand. The TGA and irregular fetal bradycardia indicated special arrangements. With a comprehensive preparation and an excellent multi-disciplinary teamwork, transportation with a helicopter went smoothly and the patient with complex cardiac disease and complete heart block received proper care at a national institute in Bangkok.

Declaration of conflict of interest

The authors declare no conflict of interest.

Acknowledgements

Special thanks to Chaiyos Kunanusont, MD, PhD (BHRC) for advising on this report and to Somarch Wongkhomthong CEO, BDMS group 4 for the opportunity to make this report.

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