

## Introduction

Heterotaxy syndrome, seen in 1 in 10,000 people worldwide, is a condition defined by abnormal arrangement of the heart and intraabdominal organs. It is caused by disruption of left-right axis orientation during early embryonic development. Cardiac malformations are a major component of heterotaxy syndrome and can be associated with considerable morbidity and mortality.

Abnormal cardiac development typically leads to atrial appendage isomerism, resulting in either bilateral paired right atria (right atrial isomerism) or paired left atria (left atrial isomerism). When isomerism of the atria is present, the sinus node may either be duplicated (RAI) or absent/hypoplastic (LAI), often leading to sinus node dysfunction. The following case describes a unique presentation of left atrial isomerism associated with complete heart block, occurring in approximately 10% of patients with left atrial isomerism.

## Case

- Mother with limited prenatal care; 1<sup>st</sup> MFM appt on day of C/S with concern for cardiac anomaly
- Baby boy born at 31 weeks via crash C-section, for concern for fetal decels in MFM office, admitted to NICU for prematurity and persistently low HR (HR in 60s)
- Intubated in delivery room for persistent bradycardia and surfactant given
- Apgars 6/8
- UVC placed and started on Isoproterenol, Epi, and PGE drips
- ECHO on admission: complete AVSD, interrupted IVC, left atrial isomerism, LVOT obstruction, coarctation of the aorta, multiple muscular VSDs
- EKG bradycardia with complete heart block (Fig. 1)
- Pediatric Cardiology with placement of 2 atrial and 2 ventricular wires and paced in VVI at rate of 90 BPM
- Pacing rate increased gradually, infant became hypotensive and acidotic

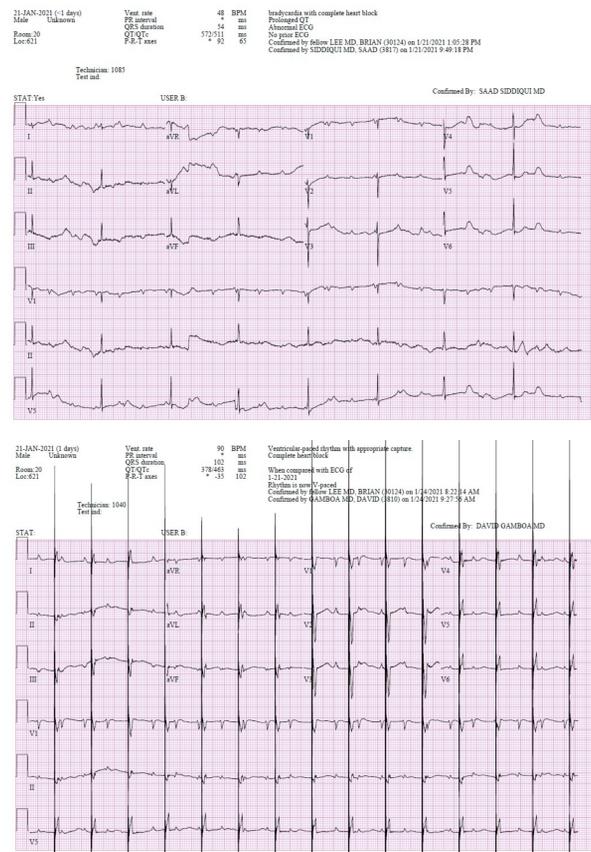


Fig. 1 – Initial EKG revealing complete heart block and bradycardia (top), repeat EKG after temporary pacing wires placed (bottom)

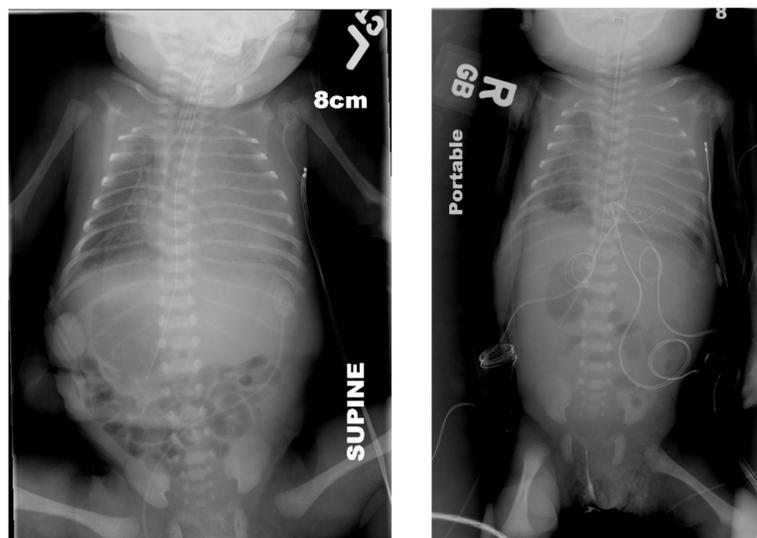


Fig. 2 – Initial XR showing right sided stomach (consistent with heterotaxy syndrome) and lung opacities with possible concern for cardiomegaly (Left). Repeat XR with worsening lung opacities and additional wires seen, including pacing wires and UVC (Right).

## Case (cont.)

- Dopamine started on DOL 2 for hypotension
- Transitioned from conventional vent to HFJV on DOL 2
- On DOL 3 attempted AV synchrony with mode changed from VVI to DDD and infant exhibited 2:1 conduction
- DOL 4 had increasing acidosis with increase in Epi support
- Septic workup completed and patient started on empiric antibiotics
- On DOL 4 evening had decreasing HR, compressions and epinephrine administered, bedside echo with no function after epi administration and time of death called.

## Discussion

This case highlights the severity of heterotaxy syndrome and how timing of delivery factors into prognosis. This infant, in addition to having a complex congenital heart disease, had to battle prematurity and lungs that were underdeveloped (Fig. 2). This case illustrates that not only is continued prenatal care important, but that congenital heart block alone is not a reason for emergent delivery. Infants can be delivered full term as long as no signs of fetal distress, like hydrops fetalis, are present. Additionally, decision to deliver early should not be made on fetal ventricular rate alone, especially given that prematurity is a strong risk factor for postnatal death. Due to increased advances in medicine and cardiac surgery, survival rate for heterotaxy has increased. However, infants born with complete heart block (especially when diagnosed prenatally) is prognostic of a worst outcome and decreased survival. Making prematurity a complication that increases mortality for infants born with complete heart block. The perinatal management is important and includes not only close monitoring, but timing delivery as close to full term as possible to maximize maturity. Additionally, it is important to have a multidisciplinary approach with OB, neonatologist, and cardiologists to best improve outcomes.

## References

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