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Congenital junctional ectopic tachycardia in the paediatric emergency department

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Brief Report

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Abstract

Congenital junctional ectopic tachycardia is a rare but serious cardiac arrhythmia seen in neonates and young infants. It is frequently resistant and refractory to first-line treatment options such as cardioversion with adenosine and direct current shock, and it carries a high morbidity and mortality rate. The aim of this article is to present the case of congenital junctional ectopic tachycardia observed in a 14-day-old neonate, highlighting the role of ivabradine in the management, followed by a discussion about current approaches to treatment.

Congenital junctional ectopic tachycardia is the least common sustained childhood arrhythmia, with one review of data from major electrophysiology centres revealing only 100 reported cases over the past 40 years.[1]

The classical clinical presentation of congenital junctional ectopic tachycardia is of a neonate or young infant presenting to hospital with a tachycardia and can include a recent history of poor feeding, tachypnoea, or other symptoms of cardiac failure.[1] Electrocardiogram findings show a narrow complex tachycardia with complete atrioventricular discordance or variable retrograde conduction.[1]

Case report

AB was born at 38+5 weeks of gestation by spontaneous vaginal delivery. Her birth weight was 2.51 kg. Antenatal scans were normal. AB had a routine postnatal course with only mild jaundice.

On day 13 of life, AB's parents had noticed that her heart was beating too fast to count and attended their family doctor, where they were advised to seek immediate medical attention in the Emergency Department. On arrival there, the heart rate was 280. AB's parents mentioned that she had been feeding poorly for 1 day. Her mother had felt AB was also breathing fast over this same timeframe. There were no sick contacts at home and no family history of cardiac issues.

AB's respiratory rate was 52 breaths per minute with an oxygen saturation of 98%. Her blood pressure was 83/69, temperature 36.7° centigrade, capillary refill time less than 2 seconds, and blood sugar was 3.9 mmol/L.

AB was immediately brought to the resuscitation bay and commenced on oxygen via nasal prongs. Blood was collected for blood cultures, full blood count, urea and electrolytes, C-reactive protein, liver function tests, group and hold and venous blood gas. Her electrocardiogram (Fig 1) showed a supraventricular tachycardia-like pattern, with a narrow complex tachycardia and AV discordance, and so she was given adenosine. A total of five adenosine doses were administered over the course of AB's resuscitation starting from 150 μg/kg up to a maximum of 500 μg/kg after consultation with a paediatric cardiologist. There were no electrocardiographic changes noticed following any of these doses. AB was also given a bolus of intravenous amiodarone at 2 mg/kg over an hour but there was no change in her heart rate. She received two 10 ml/kg intravenous fluid boluses of 0.9% sodium chloride due to significant hypotension. She was commenced on maintenance intravenous fluid therapy of 0.9% sodium chloride and 5% dextrose at 100 ml/kg/day. She was also covered for sepsis with intravenous cefotaxime, amoxicillin, and gentamicin.

AB's clinical examination was otherwise unremarkable. She remained alert and active throughout the resuscitation. Her chest X-ray and blood results were normal. Her initial venous blood gas displayed a slight metabolic acidosis with a pH of 7.33, pCO_2 6.57, bicarbonate 22.3, base excess 0, lactate 4.6. However, subsequent blood gases showed a gradual worsening of metabolic acidosis, with lactate reaching up to 12. As AB remained tachycardic, she was transferred to the Paediatric ICU in the nearby tertiary hospital for further management.

In the tertiary centre, she was intubated and a central venous catheter was inserted. She was given further doses of adenosine up to 500 μg/kg and received 2 days of continuous amiodarone infusion at 20 μg/kg/minute, but again to no avail. She became increasingly acidotic with pH reaching as low as 7, bicarbonate at 12 mmol/L, base excess at minus 8 mmol/L, and lactate at 8 mmol/L. She was prepared to commence treatment with extracorporeal membrane

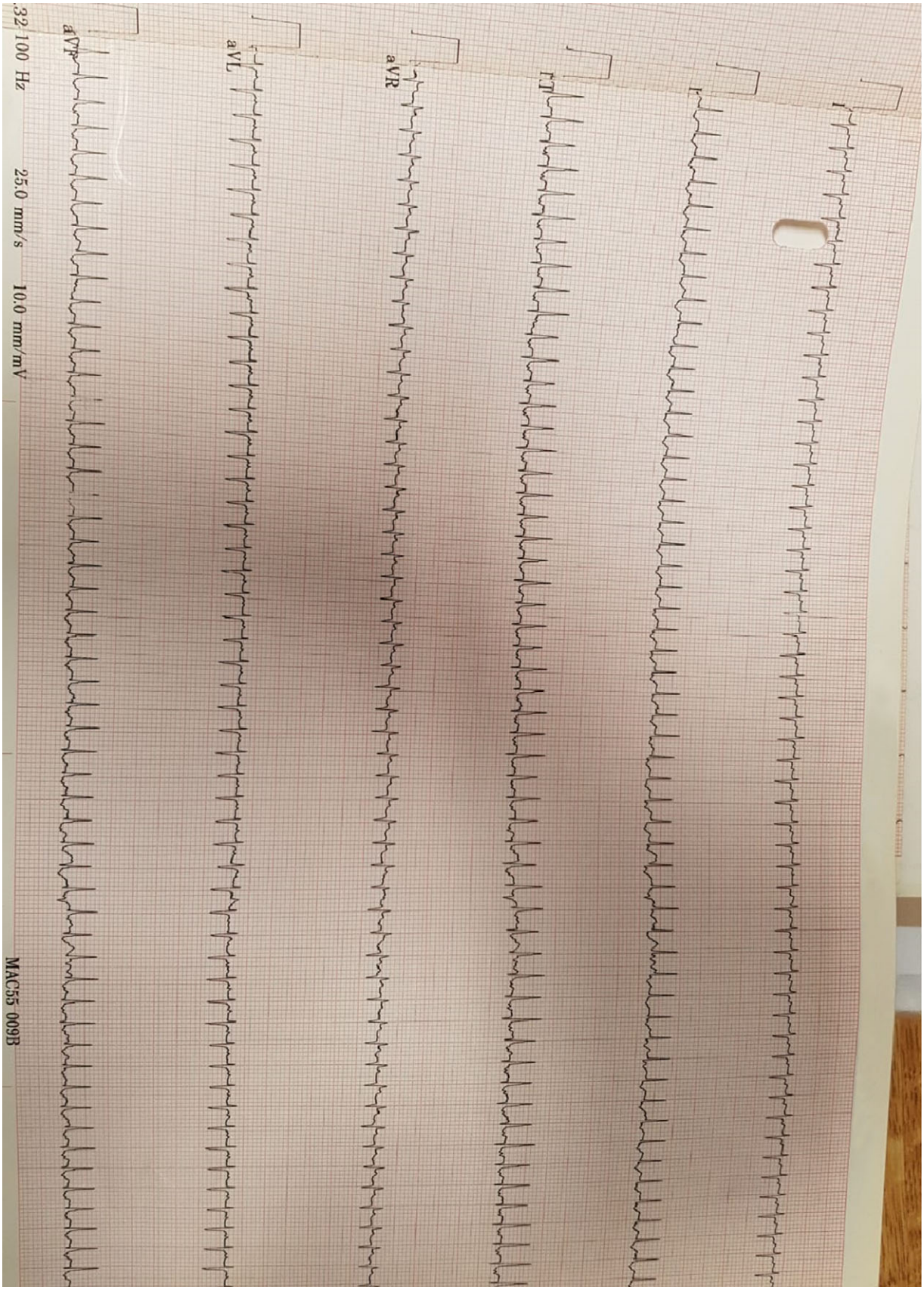

Figure 1. ECG of patient AB showing a narrow complex tachycardia with probable 1:1 retrograde conduction.

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oxygenation. However, before this was started, a final attempt to reverse her tachycardia was attempted with a combination of ivabradine (0.1 mg/kg) and propranolol (0.3 mg/kg). This proved to be successful almost immediately, bringing her heart rate down to 100 shortly after administration. Extracorporeal membrane oxygenation was abandoned at this stage, and she was managed with ivabradine and propranolol in the Paediatric ICU. A diagnosis of congenital junctional ectopic tachycardia was made. She was extubated on day 4 of her stay there, transferred to the ward on day 5, and subsequently discharged in stable condition on ivabradine 0.05 mg/kg twice daily and propranolol 0.3 mg/kg three times daily, with close follow-up in the cardiology outpatient clinic.

Discussion

The exact mechanism of congenital junctional ectopic tachycardia is not fully understood presently, but it is believed that a region close to the atrioventricular node or bundle of His is responsible by signalling its own independent electrical impulses to the ventricles.[1] This mechanism of enhanced automaticity explains why congenital junctional ectopic tachycardia is not amenable to adenosine or direct current cardioversion.

Mortality from congenital junctional ectopic tachycardia remains high, an earlier case series found a mortality rate as high as 35%,[2] and a more recent series demonstrated a mortality rate of up to 9%.[3] Death frequently results from sudden decompensation of the arrhythmia to ventricular fibrillation or complete heart block or from severe heart failure.[4] Tachycardia-induced cardiomyopathy is also a serious possible sequela of congenital junctional ectopic tachycardia.[2]

There are two main options available for management when confronted with congenital junctional ectopic tachycardia-antiarrhythmic medication and cardiac ablation. The pro-arrhythmic nature of some antiarrhythmic medications can lead to congenital junctional ectopic tachycardia evolving to ventricular fibrillation or complete heart block, and ablative measures may lead to iatrogenic complete heart block.[1]

Congenital junctional ectopic tachycardia often requires two or more antiarrhythmic medications to control.[3–5] Amiodarone is often the first-line medication used to attempt cardioversion in the neonate. Commonly, a second drug agent is added to try reduce the dose of amiodarone used and its associated long-term toxicity.[4] Digoxin has not been found to be very effective in controlling the tachycardia seen in congenital junctional ectopic tachycardia and has been observed to cause ventricular fibrillation in reported cases.[2,4] A combination regimen of flecainide and propranolol has been shown to be effective in a recently reported case.[6] A series of recent cases have demonstrated the efficacy of ivabradine in managing the tachycardia seen in infants with congenital junctional ectopic tachycardia, when other antiarrhythmics have failed.[7–10] Ivabradine is a new-generation antiarrhythmic drug which acts by inhibiting the cardiac pacemaker current I(f) of the sinoatrial node, atrioventricular node, and His-Purkinje system.[7,10] This effectively reduces the spontaneous pacemaker activity of these cells.[7] Ivabradine is a selective negative chronotropic drug without significantly affecting inotropy or other haemodynamics.[7] Our report detailed above adds to the current literature for the potential of ivabradine to be used as a possible new drug agent to manage congenital junctional ectopic tachycardia refractory to first-line antiarrhythmics.

Ablative techniques offer an alternative treatment option for patients with congenital junctional ectopic tachycardia, as well as offering a more definitive treatment option for those already on medications, with associated easing of medication burden. Ablation, however, carries significant risk of complications if performed too early in affected infants, so may be best performed at a deferred stage after initial pharmacological management.[11]

In conclusion, this case report highlights a rare clinical presentation of congenital junctional ectopic tachycardia in an otherwise well 14-day-old neonate, which was highly resistant to initial treatment with adenosine and amiodarone, and responded impressively to a combination of ivabradine and propranolol.

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Conflicts of interest. None.

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