Intrapartum Management of Hyperkalaemic Periodic Paralysis

Sarah Winter-Irving[1], Hayley Anderson[1], Emma Chesterman[1]

[1]Women's and Childrens Health, St George Hospital, Kogarah

BACKGROUND

Hyperkalaemic periodic paralysis (HPP) is a rare autosomal dominant genetic condition caused by a mutation in the SCN4A gene resulting in malfunction of skeletal muscle sodium channels and characterised by transient hyperkalaemia-induced muscle weakness or paralysis (1). Common triggers include physical exertion, inactivity, cold-exposure and fasting. Frequency of episodes increases in pregnancy (2) however literature describing management of HPP in pregnancy is scarce.

AIMS

We describe intrapartum management of a woman with HPP who achieved a vacuum-assisted vaginal birth.

CASE

A 34-year-old nulliparous woman booked into our maternity service at fourteen weeks' gestation. Her medical history included HPP diagnosed at age 14 and managed with low-potassium diet, acetazolamide 250mg orally daily and avoidance of triggers. She was referred antenatally to neurology and anaesthetics and a plan was made for monitoring and managing HPP intrapartum.

She presented in spontaneous labour at 40+3 weeks' gestation, prior to a planned induction of labour. An intravenous cannula was inserted and baseline bloods collected. Continuous fetal monitoring was undertaken and an epidural block (EDB) was sited at the patient's request. Given immobility was a known trigger, following the EDB the patient's partner regularly moved her legs in a pedalling motion. Prior to the EDB, an intravenous fluid bolus of Hartmann's was inadvertently commenced however this was quickly recognised and changed to 0.9% sodium chloride. The multidisciplinary intrapartum plan was instituted including maintenance of normothermia and regular carbohydrate ingestion. Despite these measures, she developed bilateral upper limb weakness and paraesthesias. ECG and serum potassium were monitored serially and worsening hyperkalaemia was observed (peak of 7.0mmol/L) however without ECGs features of hyperkalaemia. Hyperkalaemia was initially managed with inhaled salbutamol however serum potassium remained elevated despite this. A multidisciplinary team approach among obstetric, neurology, anaesthetic and intensive care teams was maintained and a joint decision made to transfer our patient to the ICU for telemetry and hyperkalaemia management including intravenous calcium gluconate, and continuous intravenous insulin and dextrose.

Labour progressed spontaneously to full cervical dilatation. Given concerns for fetal distress, delivery was expedited via vacuum-assisted delivery with birth of a live female infant in good condition.

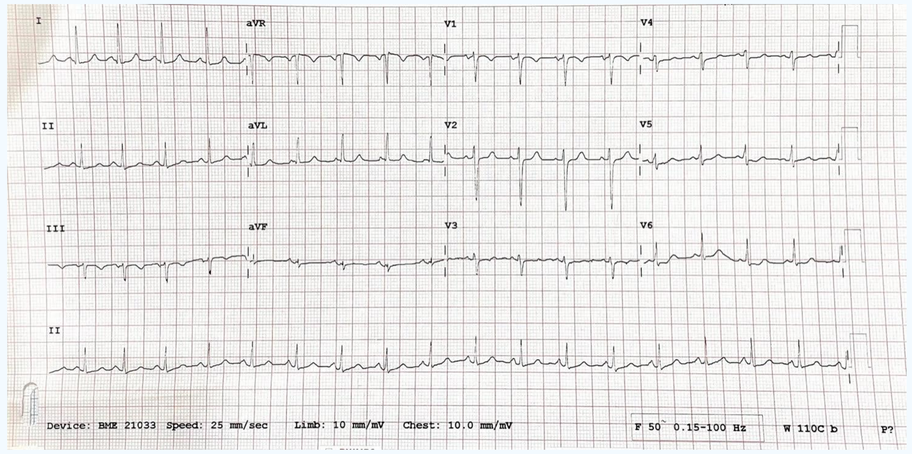

Figure 1. ECG taken at time of peak serum potassium, 7.0mmol/L, with no features of hyperkalaemia demonstrated

RESULTS

Maternal potassium levels were monitored post-partum and remained normal. The neonate was admitted to the special care nursery for observation and cardiac monitoring. Electrolytes including potassium were normal at one hour and one day of life and cardiac monitoring was unremarkable. Our patient and baby remained well and were discharged home two days post-partum with outpatient neurology follow up.

DISCUSSION

This case illustrates successful intrapartum management of HPP. It highlights the importance of a multidisciplinary team approach to managing patients with rare conditions including HPP, to minimise risk of harm to mother and baby (3).

References

1. Horga A, Raja Rayan DL, Matthews E, Sud R, Fialho D, Durran SC, et al. Prevalence study of genetically defined skeletal muscle channelopathies in England. Neurology. 2013;80(16):1472-5.
2. Charles G, Zheng C, Lehmann-Horn F, Jurkat-Rott K, Levitt J. Characterization of hyperkalemic periodic paralysis: a survey of genetically diagnosed individuals. J Neurol. 2013;260(10):2606-13.
3. Mackenzie MJ, Pickering E, Yentis SM. Anaesthetic management of labour and caesarean delivery of a patient with hyperkalaemic periodic paralysis. Int. 2006;15(4):329-31.