Clinical Reports

Anaesthetic management of a patient with familial normokalaemic periodic paralysis

F. Walsh MB MRCPI FFARCSI, D. Kelly MB MRCPI FFARCSI

Purpose: *We describe the anaesthetic management of a patient with the autosomal dominant inherited disease, normokalaemic periodic paralysis. The disease results in intermittent bouts of limb and respiratory muscular weakness in association with hypothermia, stress, prolonged fasting or exercise. Unlike hypokalaemic and hyperkalaemic periodic paralysis, the more common variants of the disease, normokalaemic periodic paralysis is not accompanied by alterations in the plasma potassium concentration.*

Clinical features: *A five-year-old boy presented for emergency scrotal exploration. He had a family history of periodic paralysis and had experienced previous episodes of weakness, two of which had required hospitalization for respiratory distress. On admission there was no evidence of weakness and serum potassium concentration was 4.2 $mMol \cdot L^{-1}$. A spinal anaesthetic was performed and the procedure was uncomplicated by muscle paralysis above the level of the spinal block.*

Conclusion: *Avoidance of known precipitating factors and judicious use of neuromuscular blocking drugs has been advocated in patients with this disorder presenting for surgery. In appropriate circumstances, spinal anaesthesia represents a useful option in patients with normokalaemic periodic paralysis.*

Objectif: *Les auteurs décrivent la gestion anesthésique d'un patient souffrant de paralysie normokaliémique périodique, maladie congénitale à caractère autosomique dominant. Cette affection provoque des poussées intermittentes de faiblesse musculaire périphérique et respiratoire associées à l'hypothermie, au stress, au jeûne prolongé et à l'exercice. Contrairement aux paralysies hypokaliémique et hyperkaliémique, la variante la plus commune de la maladie, la paralysie normokaliémique périodique ne s'accompagne pas de dyskaliémie.*

Caractéristiques cliniques: *Un garçon de cinq ans était programmé en urgence pour une exploration du scrotum. Il présentait déjà une histoire familiale de paralysie périodique et avait déjà subi des épisodes antérieurs de faiblesse musculaire, dont deux avaient nécessité son hospitalisation pour détresse respiratoire. A l'admission, il n'avait pas de faiblesse musculaire évidente et sa kaliémie se situait à 4,3 $mMol \cdot L^{-1}$. Une anesthésie rachidienne a été effectuée sans se compliquer par une paralysie musculaire au-dessus du niveau de l'anesthésie régionale.*

Conclusion: *Si ces patients doivent être opérés, on a préconisé d'éviter les facteurs déclenchants et d'utiliser judicieusement les relaxants musculaires. Lorsque les circonstances le permettent, la rachianesthésie représente une solution de rechange pour les patients qui souffrent de paralysie normokaliémique périodique.*

Key words
ANAESTHESIA: paediatric;
COMPLICATIONS: periodic paralysis;
SYNDROMES: familial periodic paralysis.

From the Department of Anaesthesia, Cork University Hospital, Wilton, Cork, Ireland.

Address correspondence to: Dr. Fergus Walsh, Garden Apartment, 38 Waterloo Rd., Dublin 4, Ireland.

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The familial periodic paralyses are a group of rare, autosomally inherited genetic diseases. They are characterized by intermittent attacks of skeletal muscle weakness, associated with hyper, hypo or normokalaemia. In 1959, Egan and Klein[1] described exacerbation of periodic paralysis in association with anaesthesia. To date, most of the literature on this subject deals with the hypokalaemic and hyperkalaemic variants.[2–7] We describe the anaesthetic management of a five-year-old boy with normokalaemic periodic paralysis undergoing scrotal exploration.

Case report

A five-year-old boy, weighing 25 kg, presented with a six hour history suggestive of testicular torsion.

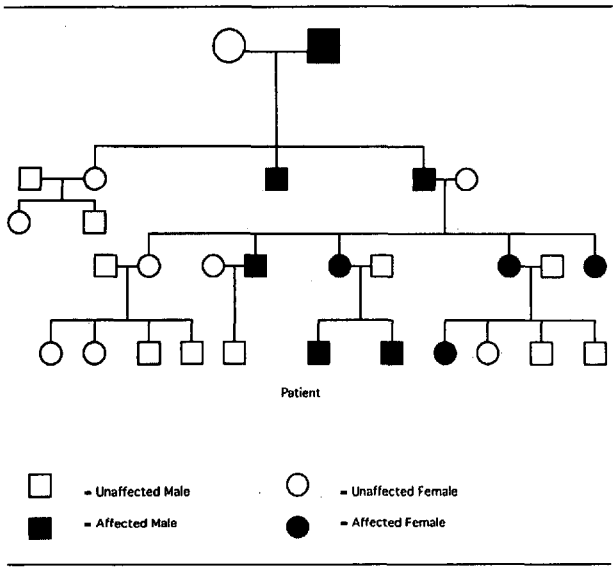

FIGURE Family tree.

Examination, on admission, was unremarkable apart from marked scrotal tenderness. He had no evidence of muscle weakness.

He was diagnosed as having normokalaemic familial periodic paralysis at the age of four and there was a family history of familial periodic paralysis (Figure). He experienced bouts of severe limb weakness lasting four to six hours every six to eight weeks. On two occasions he had required hospitalisation for episodes of respiratory distress associated with acute attacks of muscle weakness. On each admission, plasma potassium estimations were normal. His only sibling was similarly affected. He was taking no regular medications and had been fasting for two hours. Preoperative plasma potassium (4.2 $mMol \cdot L^{-1}$), sodium (138 $mMol \cdot L^{-1}$), and urea (4.8 $mMol \cdot L^{-1}$) concentrations were within normal limits. In the operating room standard haemodynamic monitoring was applied and a fluid blous, 10 $ml \cdot kg^{-1}$ normal saline, administered. Fentanyl (1 $\mu g \cdot kg^{-1}$) was given for sedation. With the patient in the right lateral position a sub-arachnoid block was performed through the L_{4-5} intervertebral space and 0.75 ml hyperbaric lidocaine 5% were administered through a 26 gauge Quincke tipped spinal needle. This resulted in a satisfactory block on the right side. The scrotum was incised and an ischaemic right testis removed. Throughout surgery haemodynamic variables were stable.

Perioperatively, the patient did not suffer muscle weakness above the level of the block which regressed completely within six hours. Serum electrolyte estimations immediately and at 24 hr after surgery were normal. The patient was ambulant and taking his normal diet six hours after surgery. Analgesia included one dose of morphine (0.2 $mg \cdot kg^{-1}$ *im*) and diclofenac (1.5 $mg \cdot kg^{-1}$ bd *pr*). He was discharged home on the third postoperative day.

Discussion

The familial periodic paralyses are a group of autosomal dominant inherited diseases and are classified into hyperkalaemic, , hypokalaemic and normokalaemic types. The first description of paralysis in association with anaesthesia was in 1959.[1] Episodic attacks of flaccid muscular weakness, mostly affecting the limbs, are usually associated with alteration of the serum potassium concentration and symptoms are ameliorated when the potassium concentration is restored to normal.[7] In the hypokalaemic group, episodes of weakness typically occur following carbohydrate loading, stress, hypothermia or infections. Hyperkalaemic periodic paralysis is classically associated with prolonged fasting and physical exercise.

Normokalaemic (sodium responsive) periodic paralysis is the third variant of this disease process. Whether normokalaemic periodic paralysis is a separate entity or merely a sub-group of the hyperkalaemic form is the subject of debate.[8] Unlike the hyper- and hypokalaemic forms, the onset is often in early childhood and affects the respiratory muscles as well as the limbs. However, similarities between these groups do exist. For example, the paralysis is worsened by the administration of potassium. Since there is no discernable abnormality of plasma potassium concentration treatment is difficult, but includes sodium loading and maintenance of carbohydrate stores. Also kaliuresis induced by acetozolamide or thiazide diuretics has been advocated.[9]

The pathophysiological basis is thought to lie in an abnormality of the voltage gated sodium channel. Isolated muscle fibres from patients with hyperkalaemic periodic paralysis have abnormal sodium conductance, with increased influx of sodium, and an abnormal effect of extracellular potassium on excitability. The sodium channel is genetically disabled and acts in a "leaky" fashion. This results in persistent opening of the sodium channel during depolarization in response to elevated extracellular potassium concentrations.[10]

Miller and Lee[9] have suggested guidelines for the anaesthetic management of patients with periodic paralysis. These include attention to preoperative carbohydrate balance, preoperative correction of electrolyte abnormalities, reduction of stress and maintenance of normothermia. They also recommended that muscle relaxants should be avoided. However, the successful use of atracurium has been reported[11,12] and succinylcholine has been used in standard doses without compli-

cation.[2,12] Postoperative weakness has been reported following pancuronium administration during coronary artery bypass grafting.[4,5] In each of these reported episodes of muscular weakness hypokalaemia was present and train-of-four stimulation using a peripheral nerve stimulator indicated recovery from neuro-muscular blockade. Horton's[3] review of 21 anaesthetics to members of a family with hypokalaemic periodic paralysis uncovered seven patients who suffered mild or severe postoperative paralysis. Muscle relaxation in these patients had been maintained by intravenous infusion of succinyl choline in one and by intermittent boluses in another; d-tubocurarine had been used intermittently in one and the remaining four cases were associated with the use of cyclopropane. There is one report of spinal anaesthesia to provide analgesia for childbirth[3] and another of combined spinal/general anaesthesia[2] in patients with hypokalaemic periodic paralysis. Neither of these cases suffered prolonged post-operative weakness.

In patients with normokalaemic periodic paralysis, normal serum potassium estimations do not preclude the onset of muscle weakness, every effort must be made to avoid known precipitating factors. Therefore, the plasma potassium concentration should be maintained within the normal range, (potassium free) intravenous fluids should be warmed, heating blankets applied, and prophylactic sedation administered. If general anaesthesia is chosen, careful titration of neuromuscular blocking drugs and reversal agents using a peripheral nerve stimulator is recommended.

Our case describes a patient with normokalaemic periodic paralysis undergoing anaesthesia. Despite his young age we chose spinal anaesthesia with a short acting agent (lidocaine) to minimise residual post-operative motor blockade to allow prompt assessment of muscular function. The volume of intra-thecal lidocaine administered provided an adequate level of anaesthesia without compromising respiratory muscle function. Efforts were made to ensure normothermia throughout the procedure. Careful monitoring and analgesia ensured that post-operative pain and stress were minimised and not allowed to precipitate muscle weakness.

As more case reports appear, guidelines for the anaesthetic management of these patients will become clearer. We suggest that, in appropriate circumstances, spinal anaesthesia in patients with normokalaemic periodic paralysis represents a useful option.

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