Management of a Parturient with Gordon’s Syndrome

Abstract Type: Case Report/Case Series
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Introduction: Gordon’s syndrome is characterised by hypertension and hyperkalemia with normal glomerular function. Hyperkalemia can be severe with serum potassium levels exceeding 7.0 mmol/l and is invariably associated with an acidaemia. This explains growth failure, intellectual impairment and muscle weakness seen in some severely affected individuals. The pathophysiology is the inability of the kidney to excrete an excessive dietary sodium load. The treatment involves dietary sodium restriction and thiazide diuretics.

Case Report: A 20 yr old primigravida woman presented at term, in labour, requesting epidural analgesia. She was recently diagnosed with minor form of Gordon’s syndrome featuring hyperkalemia and hypertension. She had been on thiazide diuretic which was discontinued during pregnancy. Epidural was sited and patient was administered drugs as per local protocol and effective analgesia was achieved. As labour was slow to progress, a decision was made to deliver her by caesarean section. Epidural was topped up with 14mls of rapid top up mix (Lidocaine 2% 10 ml + Bupivacaine 0.5% with Epinephrine 1:200,000 10ml and 2ml of 8.4% sodium bicarbonate) in 5mls increments. A sensory level to cold to thoracic 2 dermatome was achieved within 15 min with a motor block Bromage score 4. After the delivery of the baby, patient complained of difficulty in breathing, was only able to whisper and was unable to lift her head against gravity. The decision to induce general anaesthetic was taken and patient had a rapid sequence induction with thiopentone and suxamethonium. Insulin-glucose infusion was commenced to bring down the potassium levels. General anaesthetic was maintained with sevoflurane and remifentanil. The patient was kept asleep for 3 hours, after which she was extubated with good cough reflex and normal muscle power.

Discussion: There were multiple issues in this case. Firstly it was decided not to lower the potassium levels due to the chronicity of the disease. Secondly the issue of weakness was thought to be due to either a high epidural block or hyperkalemic paralysis. Gordon syndrome is regarded as a separate entity from hyperkalemic periodic paralysis, because hyperkalemia is chronic. The third problem involved the use of muscle relaxants. The patient required rapid sequence intubation of the trachea. It would seem logical to avoid suxamethonium but the chronicity of hyperkalemia, familiarity with suxamethonium and a probable difficult and emergent airway prompted the use of suxamethonium with insulin-glucose infusion to reduce any surges in potassium.

References:
2. GE power et al; Emergency anaesthesia in a patient with gordon syndrome Anaesth Intensive care 2004;32:275-277