

On-pump coronary artery bypass grafting and postoperative cardiac ICU care in a patient with hereditary spastic paraplegia: a case presentation

Abstract

Background: Hereditary Spastic Paraplegia (HSP) is a rare condition with very little information in the literature regarding its implications for major surgery and perioperative management in critical care. In those cases reported, the authors have avoided general anaesthesia or modified their technique in some way in an attempt to mitigate any potential complications.

Case presentation: A 50 year old gentleman with HSP underwent urgent inpatient on-pump coronary artery bypass grafting. Since the avoidance of general anaesthesia, opioids and muscle relaxation was unavoidable, the anaesthesia and postoperative care proceeded with no modification to the unit's standard practice.

Discussion: This is the first case report of a patient with HSP undergoing major surgery including general anaesthesia, unreversed muscle relaxation, large doses of intraoperative and postoperative opioids and a period of postoperative ventilation.

Conclusion: We have been able to demonstrate that this unit's standard anaesthesia and postoperative care was safe and effective for this patient with HSP.

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Abbreviations: HSP, Hereditary Spastic Paraplegia; CICU, Cardiac Intensive Care Unit; NDMR, Non-depolarising Muscle Relaxant; PCA, Patient Controlled Analgesia; CPAP, Continuous Positive Airway Pressure; HDU, High Dependency Unit; MH, Malignant Hyperthermia; TIVA, Total Intravenous Anaesthesia

Introduction

Hereditary Spastic Paraplegia (HSP) is a relatively rare condition with a prevalence in the order of 2-5/100,000, varying slightly by country.¹ As a result there is very little information in the literature regarding its implications for major surgery and perioperative management in critical care.

Case presentation

This case involves a 50-year-old gentleman presenting for on-pump coronary artery bypass grafting. After a lengthy period of investigation he had been diagnosed with Hereditary Spastic Paraplegia approximately 5 years earlier. He had pre-existing coronary artery disease unrelated to his neurological condition and had undergone coronary artery stenting 12 years earlier.

Approximately one week prior to the day of this surgery he had presented acutely to the Emergency Department with crescendo angina and was listed for urgent inpatient coronary artery bypass grafting. His other comorbidities included treated hypertension and gout. His functional status was limited due to his HSP, which meant that he was able to walk only short distances with the aid of sticks and an external peripheral nerve functional electrical stimulation device. His spasticity was managed with regular oral baclofen.

Prior to surgery, the patient had been premedicated with lorazepam 2mg, one dose the preceding night and one on the morning of surgery. Once in the anaesthetic room full monitoring was established. A wide-bore peripheral cannula and radial arterial line were inserted

while the patient was awake. The patient was then preoxygenated and anaesthesia was induced with 2.5mcg/kg fentanyl, 1mg/kg propofol and 1.2mg/kg rocuronium. Following oral intubation, anaesthesia was maintained with an air/oxygen/isoflurane mixture. An internal jugular central line was then sited with the patient under anaesthesia. Further fentanyl was administered in increments prior to knife-to-skin, bringing the total dose to 10mcg/kg. As per the unit's standard practice, 8mmol magnesium sulphate and 2g tranexamic acid were given once the patient had been moved through to theatre. 300units/kg heparin was administered prior to cardiopulmonary bypass. As cardiopulmonary bypass was established, a further 20mg rocuronium was given.

The cardiopulmonary bypass machine used in this department did not have the capacity to add volatile anaesthetic agents into the circuit. Therefore, while on bypass, anaesthesia was maintained with an infusion of 2% propofol 5mg/kg/hr. Diastolic arrest was achieved with cold blood cardioplegia solution and the patient was kept normothermic during the procedure. Following grafting of the coronary vessels protamine was given to reverse the cumulative heparin dose at 10mg per 100 units. The patient was successfully weaned from cardiopulmonary bypass without the need for any inotropic or vasopressor support. An adequate underlying heart rate and rhythm meant that no pacing wires were sited.

Following completion of surgery the patient was kept sedated with propofol 200mg/hr and was moved to the CICU for ongoing ventilation pending extubation once stable. No further non-depolarising muscle relaxant (NDMR) had been administered, nor was a reversal agent used at any stage during the procedure. Ongoing analgesia was provided in the form of regular paracetamol 1g QDS and a morphine infusion of 1-2mg/hr, which was exchanged for a morphine Patient Controlled Analgesia (PCA) pump (1mg bolus, 5 minute lockout) once the patient was awake enough to use it effectively.

After a period of two hours of positive pressure ventilation on the CICU there was sufficient spontaneous respiratory effort to change the mode of ventilation to CPAP/Pressure Support. After a further two hours of assisted spontaneous ventilation, the patient was cardiovascularly stable, warm, generating adequate tidal volumes with minimal support, awake, following commands and had good motor power, so was extubated. Supplemental oxygen continued to be provided via a facemask. The patient remained on the CICU until his second postoperative day before being stepped down to the High Dependency Unit (HDU). From there he was moved to the post-op ward before his eventual discharge home.

Discussion

None of the clinicians involved in this gentleman's perioperative care had any prior experience of his condition. A literature search was carried out to attempt to best plan his care but there are very few case reports of patients with HSP undergoing general anaesthesia, and none for cardiac or indeed any other form of similarly invasive surgery. HSP (also known as Familial Spastic Paraplegia or Strumpell-Lorrain syndrome) is a rare genetic condition. It is not a single disease entity, more the final common pathway for a number of different genetic abnormalities.² The result is progressive lower limb weakness, spasticity, gait disturbance, hyperreflexia and often features sphincter and mild dorsal column disturbance. The disease predominantly affects the longer neurones within the spinothalamic tract and there is no involvement of speech, swallowing or respiratory function. While it is disabling, the patient's lifespan is not usually shortened.^{2,3} There are no specific drug treatments, instead disease management centres around physical therapy, assistive walking devices and orthotics to maintain function. Benzodiazepines, baclofen or injections of botulinum toxin are used to reduce spasticity and clonus.

In those case reports we were able to find, the authors had adjusted their technique due to concerns over postoperative respiratory distress due to muscle weakness or the fear of causing a lasting impact in the patient's functional status. The techniques employed included avoiding general anaesthesia and instead opting for a regional anaesthetic^{4,5} in order to keep the patient awake and spontaneously ventilating; avoiding the use of NDMRs and opioids⁶ to minimise the risk of postoperative respiratory depression and muscle weakness; or through the planned use of sugammadex for the reversal of rocuronium in cases where a NDMR was used.⁷

This case is unique amongst those in the published literature in that no specific modifications were made to the anaesthetic technique with consideration to the HSP. Avoidance of intraoperative nitrous oxide (due to the potential for subacute combined degeneration of the spinal cord with prolonged exposure) and suxamethonium (due to the potential for significant hyperkalaemia causing cardiac arrest) seemed to be reasonable precautions in general, but neither of those were relevant to the routine practice for this procedure.

Unlike certain muscular dystrophies and myotonias, there was no suggestion of a predisposition towards Malignant Hyperpyrexia (MH) with HSP. While a TIVA technique was used during the period of cardiopulmonary bypass, and would have been possible as the sole technique throughout, it was felt that the beneficial ischaemic preconditioning effects of isoflurane merited its use. Rocuronium is the standard NDMR used in this department. Reversal agents are not routinely given to patients undergoing cardiopulmonary bypass as they are all taken back to CICU ventilated and are only extubated once they have been rewarmed, are cardiovascularly stable and breathing

adequately. However, sugammadex was available should it have been necessary, which meant that rocuronium was felt to have been a reasonable choice.

The period of weaning from the ventilator and waking in CICU prior to extubation was uneventful and followed a time course that would have been expected for a patient in a similar state of health without any coexisting neurological condition. There was no evidence of any residual muscle weakness over and above the patient's baseline. We were therefore able to safely anaesthetise and care for this patient in CICU with no specific modification to the usual technique practiced within that unit, and with no residual effects or extended period of ventilation.

Several aspects of our management may be unique to the setting of cardiac anaesthesia rather than applicable to all major surgery for patients with HSP. Firstly, we knew from the start that we had the luxury of a CICU bed for the immediate postoperative period and this was a factor in the decision-making regarding the anaesthetic technique. Secondly, the effects of cardiopulmonary bypass on the pharmacokinetics and pharmacodynamics of the opioid and NDMR could have favourably altered the outcome.

Conclusion

HSP is an uncommon disease and there was no experience within our unit nor in the literature to guide out practice. There were theoretical concerns relating to potential complications, but in the absence of any specific contraindications it was felt that the safest course of action was to deviate as little as possible from the standard practice to which that the staff in theatres and CICU would be familiar.

The procedure was necessary and a general anaesthetic and postoperative ventilation on CICU were unavoidable. The conscious use of short acting drugs or those with readily available antagonists and the knowledge that an extended period of ventilation could have been accommodated if necessary meant that we were confident to proceed. We would recommend that general anaesthesia, including the use of opioids and NDMRs, is safe for on-pump cardiac surgery in patients with HSP. The same could also be said for similarly major forms of surgery requiring postoperative critical care management, with the caveat that the usual care and attention is paid to the reversal of NDMRs at the end of surgery where necessary.

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Conflicts of interests

Authors declare that there is no conflict of interest.

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