Case report

Anaesthesia techniques and advanced monitoring in CANVAS patients - Implications for postoperative morbidity and patient recovery: A case report

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\textbf{ABSTRACT}

Introduction: Cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) is a rare multisystem neurodegenerative disorder. We describe our perioperative evaluation and care of a patient with CANVAS undergoing a pancreaticoduodenectomy for an ampullary adenocarcinoma, with a focus on perioperative risk stratification and optimisation, intraoperative advanced haemodynamic monitoring and the postoperative care.

Case presentation: A 69-year-old female with CANVAS presented with asymptomatic obstructive jaundice, icterus and abdominal pain. She had limited mobility and deconditioning due to severe generalised neuropathy. Computed tomography confirmed a resectable periampullary tumour. Her Duke Activity Status Index was 8.25 points and Edmonton Frailty Scale score was 11, confirming moderate frailty. However, the Charlson Comorbidity Index was five, indicative of a 21% estimated 10-year survival. Further risk stratification including respiratory function testing, echocardiography and cardiopulmonary exercise testing was conducted. The patient proceeded with surgery after multidisciplinary discussions with her treating medical teams.

Discussion: CANVAS is a rare and challenging condition requiring careful perioperative planning and management. There is no effective treatment for CANVAS. The management approach focuses on mitigating symptoms and improving quality of life. Given that no specific guidelines for managing these patients in the perioperative period have been provided, this report highlights several critical medical issues and implications that should be considered for the successful management of these patients. We demonstrate the role of specific anaesthesia techniques and advanced haemodynamic monitoring in both preventing postoperative morbidity and optimising patient recovery.

Conclusion: CANVAS is a rare and challenging condition in anaesthesia requiring careful perioperative planning and management.

1. Introduction

Cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS) is a rare neurodegenerative disorder. Management of patients with CANVAS is complex and requires careful perioperative planning and management. There are no recommendations outlining the anaesthesia management for patients with CANVAS. Moreover, there is a dearth of information for managing these patients in the perioperative period. We present a patient with CANVAS undergoing a pancreaticoduodenectomy for ampullary adenocarcinoma. We describe our perioperative evaluation and the specific anaesthesia management, with a focus on perioperative risk stratification and optimisation, the anaesthesia techniques and intraoperative advanced haemodynamic monitoring employed, and the postoperative care. This report has been reported in line with the SCARE criteria \cite{1} and reported in line with the PROCESS criteria \cite{2}. 

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2. Case report

A 69-year-old female (51 kg, 157 cm, body mass index 20.7 kg/m²) presented to a university teaching hospital with asymptomatic obstructive jaundice, mild icterus and intermittent upper abdominal pain. She had a history of cerebellar ataxia, neuropathy and vestibular areflexia syndrome (CANVAS). In addition, she had limited mobility and deconditioning due to severe generalised neuropathy. Initially, she had attributed this to abdominal pain caused by spending protracted periods in her wheelchair. She was on no medications and there was no relevant family or genetic history of neurological disorders.

Her presenting laboratory investigations revealed elevated bilirubin (53 μmol/L, normal range 3–17.5 μmol/L), gamma-glutamyl transferase (842 IU/L, normal range 0–118 IU/L), alkaline phosphatase (581 IU/L, normal range 115–460 IU/L), alanine transaminase (175 IU/L, normal range 10–40 IU/L), aspartate transaminase (80 IU/L, normal range 14–35 IU/L) and erythrocyte sedimentation rate (44 mm/h, normal range < 5 mm/h). All other results were within the normal range. Autoimmune hepatitis, infectious hepatitis, primary biliary cholangitis and primary sclerosing cholangitis were excluded by serologic testing. An elevated serum bicarbonate of 29 mmol/L suggested chronic hyperventilation syndrome.

An upper abdominal ultrasound was performed showing common bile duct dilation at 15.4 mm, with no obstructive calculus identified in the biliary tree or the gallbladder. Further investigations included computed tomography (CT), which revealed a double duct sign with a dilated intrahepatic and extrahepatic biliary tree and pancreatic duct. A periampullary adenocarcinoma was confirmed, which was considered resectable and curable with a pancreaticoduodenectomy (Whipple procedure). Staging CT, positron emission tomography and magnetic resonance imaging (MRI) showed no evidence of metastatic disease. The patient underwent preoperative endoscopic retrograde cholangiopancreatography and a biliary stent was placed into the bile duct for drainage. Definitive surgery proceeded six weeks post stent insertion.

Twelve years prior, the patient had developed a severe gait disturbance, which was followed by multiple bouts of vertigo and ataxia. CT of the brain showed mild cerebellar atrophy and hydrocephalus, which was treated with a ventriculoperitoneal shunt for a tentative diagnosis of normal pressure hydrocephalus. This failed to relieve any neurological symptoms. Over the following four years, peripheral neuropathy developed. The patient had bilateral carpal tunnel decompressions as a desperate attempt to relieve painful neuropathy and weakness of the hands. Subsequent nerve conduction studies revealed a progressive upper and lower limb sensory neuropathy with absent sensory nerve action potentials. Other common causes of sensory peripheral neuropathy—for example, infections, toxins (such as alcohol), metabolic or hormonal imbalances (including hypothyroidism and vitamin B₁₂ deficiency) as well as hereditary conditions (such as Charcot-Marie-Tooth disease)—were excluded. Neurological assessment confirmed the probable diagnosis of CANVAS. This was based on the clinical course and characteristic MRI findings consistent with cerebellar degeneration. Supportive clinical features and investigations included worsening cerebellar gait and limb ataxia, impaired vestibular function bilaterally, neurophysiologic evidence of sensory ganglionopathy on nerve conduction studies and a significant component of autonomic dysfunction.

Considering the diagnosis and the possibility of a curative resection, further preoperative testing was performed to stratify the patient’s risk of perioperative morbidity and mortality. The Duke Activity Status Index was 8.25 points, reflective of a very poor functional status of approximately 3.08 metabolic equivalents of tasks. The patient’s Edmonton Frailty Scale score was 11, confirming moderate frailty. However, the Charlson Comorbidity Index, which is a validated metric that predicts 10-year survival in patients with multiple comorbidities, was five. This was indicative of a 21% estimated 10-year survival.

Respiratory function tests showed no evidence of obstructive or restrictive lung disease. Mild reduction in gas exchange was apparent, with a carbon monoxide transfer coefficient of 76% (adjusted for haemoglobin and ventilation). Diaphragmatic weakness with normal expiratory muscle function was predicted by maximum inspiratory pressures of 47% and suggested by maximum expiratory pressures of 101%. Transthoracic echocardiography demonstrated normality for biventricular systolic function, valve function and estimated pulmonary artery pressures. It also showed no evidence of left ventricular diastolic impairment.

To further stratify the patient’s perioperative complication risks, cardiopulmonary exercise testing was conducted. During the test, the patient exercised to a maximum load of 63 watts using a 10 W per minute ramp protocol. It was challenging for her to perform the test due to significant pre-existing leg weakness. She could maintain exercise at 35 rpm (recommended 55–60 rpm) with leg fatigue limiting exercise and a maximal oxygen consumption rate (VO₂ peak) of 10 ml/min/kg (48% predicted). The anaerobic threshold was reached within one minute of the ramp protocol commencing, and the lowest achieved ventilatory equivalent for CO₂ (VE/VO₂) was significantly elevated at >40. The ventilatory reserve was normal, and no hypoaxia was observed. Oxygen pulse was reduced to 3.9 ml/beat at maximum exercise. The patient was able to mount a maximum heart rate of 178 bpm (sinus rhythm), with no evidence of cardiac ischaemia on the ECG at any stage of the test. The combination of a low VO₂ peak, low anaerobic threshold and a high VE/VO₂ at anaerobic threshold were indicative of a high perioperative risk of morbidity following major surgery.

Given the benefits and risks of curative surgery, the possibility of tumour progression and the subsequent impact on quality of life, the patient elected to proceed with surgery. This decision was reached after multidisciplinary discussions that included her family and treating neurologist, in addition to discussions with the surgeon, anaesthetist, respiratory physician, intensivist, physiotherapist and dietician. In preparation for surgery, a four-week personalised prehabilitation program was initiated, consisting of daily physiotherapy, incentive spirometry and nutritional optimisation.

2.1. Intraoperative course

Provision of anaesthesia and surgery were performed by clinicians with expertise in hepatobiliary-pancreatic anaesthesia and surgery respectively. A combined regional–general anaesthetic technique was performed with multimodal analgesia. The patient received a single pre-induction intrathecal injection of 0.5% hyperbaric bupivacaine (15 mg), morphine (200 μg) and clonidine (75 μg). A thoracic epidural was also inserted for both intraoperative and postoperative analgesia. Anaesthesia monitoring included pulse oximetry, five-lead ECG, continuous central venous pressure, intra-arterial pressure, nasal temperature and depth of anaesthesia monitoring. Advanced haemodynamic monitoring (FloTrac® system, Edwards Lifesciences, Irvine USA) was used for real-time monitoring of cardiac index, stroke volume, stroke volume variation and systemic vascular resistance parameters. After the induction of anaesthesia, the trachea was intubated. Anaesthesia was maintained with a propofol target-controlled infusion (Schneider model, effect site concentration of 1.5 μg/ml) and nitrous oxide (66%), maintaining a bispectral index of 40–60. As part of an anaesthesia and opioid sparing multimodal technique, the following intravenous infusions were also administered: lignocaine (1 mg/kg/h, no loading dose), dexmedetomidine (0.3 mcg/kg/h, no loading dose) and magnesium (15 mg/kg load, 15 mg/kg/h). To maximise surgical access and optimise muscle relaxation, a rocuronium infusion was administered at 0.5 mg/kg/h.

Over the 12-hour operation, fluid intervention and the use of vasoactive medication were guided by an individualised, surgery-specific haemodynamic algorithm (see Fig. 1). The patient required a vasopressor infusion (noradrenaline 2–10 μg/min, titrated to keep the mean arterial pressure > 65 mmHg). Surgery was uneventful, with 50 ml of estimated blood loss. In total, 750 ml of crystalloid (plus drug infusion
volumes) was administered. Regular arterial blood gas monitoring was performed with the lowest pH (7.30), standard base excess (−4.5 mmol/l) and haemoglobin (12.9 g/dL) values. Intraoperative blood transfusion was not required. Fig. 2 presents the intraoperative haemodynamic data. One hour prior to the completion of the surgery, the epidural was loaded with ropivacaine 0.2% (8 ml). At the surgery’s completion, the patient was administered sugammadex (200 mg) for reversal of neuromuscular blockade and the trachea was extubated. The patient was alert and pain free when she was transferred to the intensive care unit (ICU) for routine postoperative monitoring.

Postoperatively, free oral fluids were immediately encouraged, and the patient was mobilised to a chair six hours post-surgery. She spent five days in the ICU to optimise her haemodynamic state with twice daily physiotherapy. On postoperative day three, there was a single event of elevated blood lactate to 3.2 mmol/L, which was most likely secondary to hypovolaemia. It resolved rapidly following a 4% albumin bolus. The noradrenaline requirement persisted until postoperative day three; it was then ceased in temporal relation to the removal of the thoracic epidural. The patient received one dose of oral tapentadol (50 mg) during the entire postoperative stay. Paracetamol was charted daily but declined by the patient at every offering. On postoperative day six, the patient was transferred to a general surgical ward. She was discharged on postoperative day 12. Final histopathology of the tumour confirmed a poorly differentiated adenocarcinoma of the ampulla, invading the duodenal mucosa and into the muscularis propria. There was no perineural or lymphovascular invasion. Follow-up at one, three and six months post-discharge revealed no changes in tumour biomarkers (e.g., carbohydrate antigen 19-9, carcinoembryonic antigen, alpha-fetoprotein and chromogranin A), and a return to preoperative physiological functional status.

2.2. Discussion

CANVAS is a rare, progressive, adult onset neurologic disorder characterised by imbalance due to cerebellar gait and limb ataxia, impaired vestibular function bilaterally and non–length dependent sensory neuropathy [3]. The age of onset between 50 and 60 years, with a similar prevalence in males and females [3].

2.2.1. Inheritance

The cause of CANVAS is mainly idiopathic; however, both acquired and familial causes have been identified [4,5]. Autosomal recessive, biallelic repeat expansion of an AAGGG intronic segment in the replication factor C subunit 1 (RFC1) gene on chromosome 4p14 has been attributed as the cause of familial CANVAS and late onset ataxia, particularly if there is coexisting sensory neuropathy and bilateral vestibular areflexia [4,5]. However, autosomal dominant inheritance has also been suggested recently [6].

2.2.2. Histopathology

Both the central and peripheral nervous systems are implicated in the histopathology of CANVAS. Centrally, there is widespread depletion of cerebellar Purkinje cells, as well as the loss of neurons in the vestibular, trigeminal and facial gangliaons [4,7]. Peripherally, there is a severe depletion of myelinated fibres in posterior columns, dorsal root ganglia atrophy and axonal loss without active Wallerian degeneration or Schwann cell proliferation [8]. In contrast to axonal neuropathies, where nerves are of normal calibre or even enlarged, axonal atrophy in CANVAS neuropathy results in peripheral nerves that are pathologically smaller [9].

2.2.3. Clinical presentation and diagnosis

The diagnostic criteria for CANVAS have been proposed by
Szmulewicz et al. [5] (see Table 1). The demonstration of cerebellar dysfunction, bilateral vestibulopathy and non-length dependent sensory neuronopathy is essential for the diagnosis. However, the timing of the onset of these components varies, and the progress of symptoms is often slower than other causes of ataxia [8]. Typical findings indicating cerebellar disease include gait ataxia, dysarthria and intermittent gaze-evoked nystagmus [10]. Importantly, the characteristic finding of bilateral vestibulopathy is demonstrated by an abnormal visually enhanced vestibulo-ocular reflex (doll’s head reflex) assessed using high-speed video-oculography [7]. Non-length dependent sensory neuronopathy manifests variably as reduced sensation to vibration, light touch or pinprick, and/or abnormal joint proprioception [7].

Nerve conduction studies demonstrate a reduction in sensory fibre action potential generation, with relative sparing of motor fibres [10,11]. Clinical findings of cerebellar disease can be corroborated through the use of MRI, demonstrating cerebellar atrophy involving vermian lobules (VI, VIIa, VIIb) and Crus I of the cerebellar hemisphere [7]. Although not diagnostic, dysautonomia is an important clinical feature of CANVAS, with variable manifestations including postural dizziness, hypohidrosis, erectile dysfunction and constipation [10].

Surgical phase

| Fluids therapy      | Dissection and resection | Reconstruction | Closure |
|---------------------|--------------------------|----------------|---------|
| 200 mL              | +100 mL                  | +100 mL        | +200 mL |
| Noradrenaline infusion | 2μg/min                  | 5μg/min        | 10 μg/min |
|                     | 4μg/min                  | 9μg/min        |         |

Fig. 2. Intraoperative haemodynamic data (MAP: mean arterial pressure; CI: cardiac index; SVR: systemic vascular resistance; SV: stroke volume; HR: heart rate; SVV: stroke volume variation). Green shaded areas represent haemodynamic “target zones”.

L. Weinberg et al.
evaluating accurate cardiopulmonary reserve and fitness for surgery in patients with significant neuromuscular disease. Patients with severe cerebellar atrophy to CANVAS, vestibulopathy and non-heritage, MRI findings and age of onset (although adult onset FRDA has been reported) [3]. Although MSA-C demonstrates a similar pattern of cerebellar atrophy to CANVAS, vestibulopathy and non–length dependent sensory neuropathy are not present [7]. Other pathologies that may mimic aspects of CANVAS should also be excluded, including diabetic neuropathy, Wernicke's encephalopathy and iatrogenic causes of vestibulopathy (such as aminoglycoside antibiotics) [7].

2.2.4. Management
There is no effective treatment for CANVAS. The management approach focuses on mitigating symptoms and improving quality of life.

2.2.5. Anaesthetic implications
Given the complexity of CANVAS, multiple anaesthetic implications require careful perioperative planning and management. To our knowledge, no case reports have outlined the specific anaesthetic implications for CANVAS patients. Further, no specific guidelines for managing these patients in the perioperative period have been provided.

2.3. Preoperative assessment
To stratify the patient's perioperative risk of morbidity and mortality, preoperative assessment needs to include a thorough history, examination and investigations relevant to the patient's condition, comorbidities and risk factors (see Table 2). The evaluation should include the patient's goals for surgery, their functional status, frailty index, cognitive capacity and general fitness for surgery [12]. In our patient's case, the CPET and DASI suggested an elevated risk of postoperative complications. This demonstrates the limited roles and complexity of CPET and DASI in evaluating accurate cardiopulmonary reserve and fitness for surgery in patients with significant neuromuscular disease. Patients with severe neuromuscular disease are poorly represented in cohort studies evaluating functional capacity of patients undergoing major surgery [13]. In this case, multidisciplinary discussions involving the patient, her family, and all other care providers, resulted in a decision to proceed with surgery, weighing the likelihood of cure against the likelihood of complications and the quality of life the patient felt she would be left with if she proceeded with non-surgical treatment options.

2.4. Perioperative management
There are several perioperative considerations for the management of CANVAS dysautonomia. There is an inherent risk of aspiration secondary to gastroparesis. Anaesthetic recommendations for patients with similar ataxic syndromes have suggested rapid sequence induction and endotracheal intubation to mitigate this risk [14,15].

There is a heightened risk of haemodynamic instability due to defective baroreflex function and resistance vessel vasoconstriction, resulting in a less predictable response to anaesthetic and vasoactive agents, as well as to unexpected shifts in blood volume [16,17]. In line with similar literature [14,16,17], we recommend for careful maintenance of euvolaemia by ensuring adequate preoperative hydration and the EV1000 platform as was used with our patient. Atropine-resistant bradycardia is a documented phenomenon secondary to dysautonomia, thus alternatives such as isoprenaline and temporary pacing should be considered in severe cases [18]. We also considered that a dopamine beta-hydroxylase deficiency seen in MSA-C might contribute to poor sympathetic tone [18], and we therefore used noradrenaline as the preferred vasopressor. Importantly, medications for orthostatic hypotension (i.e., fludrocortisone) can increase the risk of supine hypertension perioperatively [17]. Supine hypertension may be managed with transdermal nitrate therapy, although alternative agents (such as short-acting intravenous sodium nitroprusside) may be more suitable given the risk of profound hypotension as well as associated cardio-depressive and neuropathic sequela [17].

Dysautonomia also impairs thermal regulation, increasing the risk of perioperative hypothermia or hyperthermia, which warrants the strict control of theatre temperatures [14].

Significant derangements to respiratory physiology are also a concern for patients with CANVAS. As seen in patients with MSA, there may be an increased risk of respiratory depression from central ventilatory failure and upper airway obstruction from obstructive sleep apnoea or vocal cord abductor paralysis/paresis on induction [14]. Positive pressure ventilation is one approach to managing this; however, it requires cautious monitoring due to the risk of compromising venous return and exacerbating hypotension [16]. The interaction between muscle relaxants and opioids may further compromise airway patency.
Anaesthesia considerations and implications for patients with CANVAS.

**Table 2**

| Parameter                  | Concern                                                                 | Management considerations |
|----------------------------|-------------------------------------------------------------------------|----------------------------|
| **Preoperative**           |                                                                         |                            |
| for surgery                | Goals of surgery                                                       | History, physical examination and investigations |
|                           | Fitness for surgery                                                    | Frailty scoring            |
|                           | Patient comorbidities                                                  | Duke Activity Status Index |
|                           | Functional status                                                      | Nutritional and body composition assessment |
|                           | Frailty                                                                | Multidisciplinary discussion including surgeon, anaesthetist, neurologist, respiratory physician, intensivist, physiotherapist, dietician |
| **Preoperative assessment**| Dyautonomia                                                            | Autonomic function assessment (i.e., postural blood pressure test) |
|                           | Sleep-disordered breathing                                             | Screening tool (i.e., STOP-BANG questionnaire) |
|                           | Cardiac status                                                         | Polysomnography            |
|                           | Upper airway obstruction                                               | Cardiac function testing (i.e., echocardiography, dynamic stress evaluation) |
|                           | Respiratory dysfunction                                                 | Cardiopulmonary exercise stress test |
|                           | Polypharmacy                                                           | Airway assessment (e.g., Mallampati score) |
|                           | Thermal dysregulation                                                  | Respiratory function testing |
|                           | Nutritional risk assessment                                             | Blood gas analysis         |
|                           |                                                                        | Medication review          |
| **Nutritional assessment** | Prone to severe nutritional risk defined as:                           | Nutritional intervention plan |
|                           | - Weight loss >10–15% within six months                                | Avoid prolonged preoperative fasting |
|                           | - BMI < 18.5 kg/m²                                                     | Consider carbohydrate drink with 800 ml the night before and 400 ml 2 h before surgery |
|                           | - Nutritional risk assessment score > 5                                | Allow unlimited clear fluids until 2 h before surgery |
| **Perioperative**          | Prone to pressure injuries                                             | Additional gel supports for all joints, cottonwood padding for sacrum and buttocks |
| Positioning                |                                                                         |                            |
| **Anaesthetic technique**  | Patient non-cooperation, involuntary movements, dystonia, rigidity      | Consider combined regional–general anaesthetic technique |
|                           | Cardiopulmonary complications of general anaesthesia                   |                            |
| **Anaesthetic agent**      | Increased sensitivity to depolarising and non-depolarising agents      | Avoid depolarising agents (i.e., succinylcholine) |
|                           | Perioperative opioid toxicity                                           | Use non-depolarising agents with caution |
| **Cardiovascular**         | Intraoperative hypotension                                             | Avoid opioids and consider multimodal or regional techniques |
|                           | Labile haemodynamic response to fluid shifts and anaesthesia           | Preoperative hydration      |
|                           | Systolic hypertension                                                  | Advanced perioperative haemodynamic monitoring to guide appropriate fluid and vasopressor support |
| **Respiratory**            | Upper airway obstruction                                               | Endotracheal intubation     |
|                           | Central respiratory failure                                            | Tracheostomy kit on standby |

**Table 2 (continued)**

| Parameter                  | Concern                                                                 | Management considerations |
|----------------------------|-------------------------------------------------------------------------|----------------------------|
| Gastrointestinal           | Risk of aspiration                                                     | Rapid sequence induction   |
|                           |                                                                         | Endotracheal intubation     |
| Neurological               | Cognitive impairment                                                   | Appropriate and targeted communication, support and interaction |
|                           |                                                                         | Strict control of theatre temperature |
| Thermoregulation           | Hyperthermia                                                           | Patient temperature monitoring |
|                           | Hypothermia                                                            |                            |
| Postoperative              | Complex multisystem management                                         | Individualised postoperative care and support |
| Intensive care support     |                                                                         |                            |
| Analgesia                  | Postoperative opioid toxicity                                          | Cautious use of opioids    |
|                           |                                                                         |                            |
| Haemodynamic management    | Postoperative hypotension and autonomic dysregulation                  | Advanced perioperative haemodynamic monitoring to guide appropriate fluid and vasopressor support |
|                           | Supine hypertension                                                    |                            |
|                           | Labile haemodynamic response to fluid shifts                           |                            |
|                           | Immobility                                                             |                            |
|                           | Haemodynamic instability                                               |                            |
| Compliations               |                                                                         |                            |
| Nutritional support        | Exacerbated stress-related catabolism                                  | Integration of nutrition into the overall management of the patient |
|                           | Impaired metabolic regulation                                          | Re-establishment of enteral feeding as early as possible |
|                           |                                                                         | Metabolic control (e.g., of blood glucose) |
|                           |                                                                         | Reduction of factors that exacerbate stress-related catabolism or impair gastrointestinal function |
|                           |                                                                         | Minimise paralytic agents for ventilator-dependent patients |
|                           |                                                                         | Early mobilisation to facilitate protein synthesis and muscle function |
| Psychosocial support       | Risk of postoperative depression and anxiety                            | Early identification and involvement of family, carers and relatives |
|                           |                                                                         | Early involvement of social worker, psychologist or psychiatrist |

and respiratory drive [19], which is why we opted for an opioid sparing technique.

Choice of anaesthetic technique should be directed towards maintaining haemodynamic stability, muscle relaxation, adequate respiratory function and effective pain relief both perioperatively and postoperatively. Our use of a combined regional–general anaesthesia allowed for both an opioid and anaesthesia sparing technique that facilitated a rapid emergence of anaesthesia and successful extubation at the end of the operation prior to ICU transfer. The advantage of a regional approach is that the anaesthetist can mitigate the risks of respiratory and cardiovascular complications without compromising efficacy, which has been demonstrated safely with both spinal and epidural techniques in MSA, FRDA and SCA patients [19–22]. However, achieving successful regional anaesthesia may be technically difficult due to patient non-cooperation, involuntary movements, dystonia and muscular rigidity [15]. In such cases, general anaesthesia may be the preferred modality, with institution of regional techniques post-induction of anaesthesia [16].

Neuraxial anaesthesia has also been associated with worsening dysautonomia, resulting in unpredictable cardiopulmonary responses and neurological symptom progression [23,24]. However, evidence
supporting the latter is scarce [14]. Our patient’s case was further complicated by a pre-existing ventriculoperitoneal shunt in situ. With the exception of lumbo-peritoneal shunts [25,26] (where spinal anaesthesia may have rapid wash-out due to peritoneal spread), no contraindications to neuraxial anaesthesia appear in patients with pre-existing cerebrospinal fluid (CSF) shunts. This supported our decision to use a central neuraxial approach. During surgery, the CSF shunt was carefully protected in a sterile endopouch retrieval bag (Ethicon Endo-Surgery, LLC, Puerto Rico, USA). Regarding anaesthetic agent selection, there was concern that patients with SCA and FRDA have heightened sensitivity to depolarising and non-depolarising muscle relaxants [15,23]. While non-depolarising agents can be used safely with caution [15] (as with our patient), studies have tended to avoid using depolarising agents due to the increased risk of hyperkalaemia, excessive neuromuscular paralysis and the need for prolonged mechanical ventilation [13,24].

2.5. Postoperative management

Postoperative management should aim to restore patients to their baseline level of function as soon as possible by anticipating complications, optimising pain relief, recommencing timely oral intake and employing aggressive physiotherapy to facilitate early mobilisation [27,28]. Given the baseline autonomic dysfunction and elevated peri-operative risks associated with CANVAS, the recovery period may be complicated by haemodynamic changes, which should be anticipated early. Our patient spent the first six postoperative days in the ICU to optimise her haemodynamic state with fluid therapy and vasoactive medications (Table 3).

3. Conclusion

CANVAS is a rare and challenging condition in anaesthesia requiring careful perioperative planning and management. Our study highlights several issues and implications that should be considered for the successful management of patients with this condition. We also demonstrate that using specific anaesthetic techniques and advanced monitoring may play a role in both preventing postoperative morbidity and optimising patient recovery.

Ethics approval

Austin Health Research Ethics Committee waived the requirement for ethical approval as written consent has been obtained by the patient.

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None declared.

CRediT authorship contribution statement

A/Prof Laurence Weinberg, A/Prof Vijayaragavan Muralidharan were responsible for study concept, data collection, data interpretation, collation of all images and writing of the paper. Dr. Akshay Hungenahally, Dr. Joshua Meyerov, Dr. Daniel RA Cox assisted with the literature review, data collection and interpretation, and writing of the paper. Dr. Lachlan F Miles assisted with the interpretation of the risk assessment and cardiopulmonary exercise test results and drafting of the article.

Table 3

| Parameter                  | Presenting bloods | Day 14 post-stenting | Intraoperatively | Intensive care | Discharge |
|----------------------------|-------------------|----------------------|------------------|---------------|-----------|
| Arterial blood gas         |                   |                      | Baseline         | Tumour removal | Wound closure | Arrival | POD 1 | POD 3 | POD 6 | POD 12 |
| FiO₂ (− / −)               | − −               | − −                  | 0.6              | 0.3           | 0.7        | 0.7      | 0.4   | 0.4   | 0.28  | Room air |
| pH (− / −)                 | − −               | − −                  | 7.35             | 7.30          | 7.34       | 7.27    | 7.35  | 7.46  | 7.43  | −        |
| PaO₂ (mmHg) (− / −)        | 221               | 115                  | 46               | 51            | 38         | 49      | 41.9  | 37.3  | 35.1  | −        |
| PaCO₂ (mmHg) (− / −)       | 144               | 142                  | 139              | 145           | 146        | 147     | 148   | 144   | 142   | 145     |
| Sodium (mmol/L) (− / −)    | 4.6               | 4.3                  | 3.7              | 4.5           | 4.3        | 4.2     | 3.8   | 4.8   | 4.3   | 3.9     |
| Potassium (mmol/L) (− / −) | 105               | 105                  | 107              | 108           | 109        | 110     | 107   | 108   | 106   | 104     |
| Chloride (mmol/L) (− / −)  | 28                | 29                   | 25.2             | 21.6          | 22.5       | 23.1    | 26.9  | 25.8  | 25.7  | −        |
| Base excess (mEq/L) (− / −)| −                 | −                    | 0.3              | −0.8          | −4.5       | −2.9    | −2.1  | 2.8   | 1.6   | 2.7     |
| Lactate (mmol/L) (− / −)   | −                 | −                    | 0.8              | 0.8           | 1.5        | 1.8     | 1.5   | 3.2   | 1.9   | 1.4     |
| Haemoglobin (g/dL) (− / −) | 136               | 134                  | 118              | 124           | 129        | 115     | 116   | 110   | 92    | 93      |
| Glucose (mmol/L) (− / −)   | 6.2               | 5.9                  | 5.8              | 7.3           | 8.1        | 6.4     | 9.1   | 8.6   | 7.7   | 8.1     |

Fluid management and vasoactive infusions

| Parameter                  | Presenting bloods | Day 14 post-stenting | Intraoperatively | Intensive care | Discharge |
|----------------------------|-------------------|----------------------|------------------|---------------|-----------|
| Fluid balance (mL) (− / −) | − −               | − −                  | +350             | −             | +1249     | +1100   | −1285 | +540  | −        |
| Balanced crystalloid (mL)  | − −               | − −                  | 650              | −             | 1300      | 1250    | −     | −     | −        |
| Colloid (mL)               | − −               | − −                  | Albumin 20% 200 mL | −            | −         | −       | −     | −     | −        |
| Oral fluids (mL) (− / −)   | − −               | − −                  | −                | −             | 260       | 350     | −     | −     | −        |
| Noradrenaline (μg/min) (− / −)| − −                  | − −                  | 2                | 7             | 8         | 8       | 7     | 3     | −        |
| Weight (kg) (− / −)        | 50.1              | 50.2                 | −                | −             | −         | −       | −     | −     | −        |
| Frusemide (mg) (− / −)     | − −               | − −                  | −                | −             | 53.5      | 55.9    | 54.5  | 52.7  | −        |
| Thoracic epidural infusion | − −               | − −                  | −                | −             | 20        | −       | −     | −     | −        |

Renal function

| Parameter                  | Presenting bloods | Day 14 post-stenting | Intraoperatively | Intensive care | Discharge |
|----------------------------|-------------------|----------------------|------------------|---------------|-----------|
| Urea (mmol/L) (− / −)      | 4.9               | 5.0                  | −                | −             | −         | −       | −     | −     | −        |
| Creatinine (μmol/L) (− / −)| 75                | 66                   | −                | −             | −         | −       | 6.7   | 5.4   | 5.1    | 4.1     |
| Estimated glomerular filtration rate (mL/min) (− / −) | 89 | 82 | − | − | − | − | >90 | >90 | >90 | >90 |

Liver function tests

| Parameter                  | Presenting bloods | Day 14 post-stenting | Intraoperatively | Intensive care | Discharge |
|----------------------------|-------------------|----------------------|------------------|---------------|-----------|
| Total bilirubin (μmol/L) (− / −)| 66                | 7                    | −                | −             | −         | −       | 8     | 8     | 7      | 6       |
| Albumin (g/L) (− / −)      | 38                | 37                   | −                | −             | −         | −       | 23    | 22    | 24     | 29      |
| Alanine aminotransferase (IU/L) (− / −) | 236 | 19 | − | − | − | − | 29 | 28 | 24 | 20 |
| Aspartate aminotransferase (IU/L) (− / −) | 98 | 65 | − | − | − | − | 145 | 129 | 112 | 76 |
| Alkaline phosphatase (IU/L) (− / −) | 551 | 91 | − | − | − | − | 103 | 95 | 93 | 86 |
| Gamma-glutamyl transferase (IU/L) (− / −) | 1046 | 65 | − | − | − | − | 145 | 129 | 112 | 97 |
Guarantor

A/Prof Laurence is the guarantor.

Research registration

N/A.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

None.

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