

# Anaesthesia And Pulseless Disease: Case Report And Review Of The Literature

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## Abstract

We present a case of Takayasu arteritis and describe the technicalities of administering general anesthesia for the case. Administering general anesthesia in Takayasu arteritis for major vascular procedures is challenging and needs special attention to preoperative optimization of the patient, perioperative monitoring of the cardiovascular system, and maintenance of cerebral and other organ perfusion and normal acid-base status.

## INTRODUCTION

Takayasu arteritis (pulse less disease) is an inflammatory arteriopathy with protean symptomatic manifestations of arterial disease. This is a non-specific granulomatous panarteritis affecting the large and medium sized arteries (1). This disease has a predilection for young Asian women; the etiology is unknown. Long-term relief of symptomatic arterial ischemia results after surgical correction with autogenous or prosthetic bypass grafts. Short and long-term complications of these reconstructions are few.

## CASE REPORT

A 26 year old male patient presented in the Accident and Emergency Department of the Hamad General Hospital, Doha Qatar with a history of dizziness and loss of consciousness when he elevates his left upper limb and inability to work with his left upper limb. He was a painter by occupation. On examination, his temperature was 37° C, pulse 70/ min. regular, with absent pulses in his left upper limb, weight 59kg, B.P.: right upper limb 145/74, left upper limb 92/57, right lower limb 140/64, left lower limb 133/59 mm of Hg. His eyes and vision were normal and there was an ejection systolic murmur all over the precordium and conducted to both the carotid arteries. His chest radiograph showed increased bronchovascular markings and LVH. ECG revealed LVH with strain pattern. A provisional diagnosis of Aortic stenosis and left subclavian steal was made. He was smoking about 5 cigarettes a day. He was not known to have hypertension, diabetes mellitus or hyperlipidemia.

Transthoracic echocardiography revealed moderate LV

dysfunction with LV ejection fraction of 39%, concentric LVH, marked LV and LA enlargement, trivial MR, RV systolic pressure of 34 mm of Hg and severe coarctation of descending thoracic aorta with systolic and mean pressure gradient of 78 and 44 mm Hg. respectively.

MR angiography revealed marked focal stenosis of distal descending thoracic aorta and complete occlusion of left subclavian artery and left common carotid arteries at their origins from the aortic arch - strongly suggestive of Takayasu's disease.

Coronary angiography showed normal coronaries. His C-reactive protein and ANCA reports were negative. Carotid Doppler study showed 50% stenosis of left common carotid artery with reversal of flow in left vertebral artery, confirming the diagnosis of left subclavian steal.

The surgical decision was to do a bypass from the ascending aorta to both left carotid and left subclavian arteries to correct his severe subclavian steal symptoms. The patient underwent ascending aortocarotid-subclavian (left) bypass using Dacron Y graft. Midline sternotomy was done to access to the ascending aorta. The left supra-clavicular incision was done to expose the subclavian artery, while the left carotid artery was exposed through the same neck incision.

## ANESTHETIC MANAGEMENT

All his hematological reports were within normal limits. As premedication, he received 2 mg Ativan at bed time night before surgery and on the morning of surgery. The

monitoring included 5 lead ECG, pulse oximetry, invasive right radial pressure monitoring with a 20 G cannula, arterial blood gas and electrolyte monitoring, CVP monitoring with a triple lumen catheter inserted through right internal jugular vein, nasopharyngeal temperature probe, inspired and expired oxygen, carbon dioxide and anesthetic agent monitoring, and indwelling bladder catheter for urine output monitoring & Bispectral index monitor. Anesthesia was induced with inj.fentanyl 0.2 mg, propofol 100 mg and cisatracurium 10 mg i.v., intubated with 8.5 mm cuffed ET tube and ventilated with Datex-Ohmeda anesthesia ventilator using circle absorber. Anesthesia was maintained with propofol infusion 50 mg/hour, fentanyl 0.12 mg/h using syringe pump, cisatracurium infusion 6 mg/ hour and sevoflurane 0.7-1.5% in 50% oxygen with air. 2 grams of ceftriaxone was given after induction. Titrated infusion of inj. sodium nitroprusside was used to maintain systolic pressure of 90-100 mm of Hg during partial aortic cross-clamping for proximal anastomosis, and inj. Heparin 5000 IU i.v. was given before distal anastomosis on left subclavian and common carotid arteries which was reversed with 50 mg protamine after completion of the anastomosis.

The procedure lasted for 7 hours and all the vital functions monitored remained within normal limits during the procedure. The nasopharyngeal temperature was maintained at 37 ° C using heated water mattress. PaO<sub>2</sub> was 150 to 200 mm Hg. Normocarbica (EtCO<sub>2</sub> of 30-35 mm Hg. and PaCO<sub>2</sub> of 32-39 mmHg.) was maintained since hypercarbia or hypocarbia can alter cerebral blood flow. pH was maintained within the normal range after declamping of the arteries. Adequate depth of anesthesia was confirmed with Bispectral index monitor. The total blood loss was 1200ml. The cell saver was used and 320 ml of autologous blood, 500 ml lactated Ringer's, 1000 ml of 6% hydroxyethyl starch and 1 unit packed red cell were transfused during the procedure. At the end of the procedure the patient was transferred to SICU and electively ventilated for 24 hours while sedated with propofol (70 mg/ hour) and fentanyl (0.05 mg/ hour) infusions.

After extubation he was kept on intermittent CPAP mask and patient controlled analgesia with fentanyl doses of 10 mcg, lockout time 10 minutes and a 4 hour limit of 180 mcg. The post-operative course was complicated by left lower lobe collapse which resolved gradually with CPAP mask, chest physiotherapy, mucolytic and bronchodilator nebulisations. The lung collapse could be due to the fact that the patient was a heavy smoker or due to lung retraction

during surgery. The total length of SICU stay was 6 days. Post-operatively the patient had normal pulse and blood pressure in his left arm and no neurological deficit.

## DISCUSSION AND REVIEW OF LITERATURE

Takayasu's arteritis (pulse less disease) primarily affects young Asian females, often presenting as upper extremity claudication and minimal to no palpable arterial pulsation in the arms and neck. The lack of peripheral pulses reflects chronic inflammation of the aorta and its major branches. The definitive diagnosis is made on the basis of contrast angiography. Signs and symptoms of Takayasu syndrome manifest on multiple organ system. Decreased perfusion to the brain owing to involvement of the carotid arteries by occlusive inflammatory and thrombotic processes can be manifested as vertigo, visual disturbances, seizures, and cerebrovascular accidents with hemiparesis or hemiplegia. Bruits are often audible over stenosed carotid or subclavian vessels. Hyperextension of the head may decrease carotid blood flow by stretching the arteries. Indeed, patients often hold their heads in a flexed (drooping) position to prevent syncope.

Involvement of the pulmonary arteries by vasculitis occurs in about 50% of patients and can be manifested as pulmonary hypertension. Ventilation-to-perfusion abnormalities owing to occlusion of small pulmonary arteries by the inflammatory process may contribute to unexpected decreases in the PaO<sub>2</sub>. Myocardial ischemia can reflect inflammation of the coronary arteries. There may be involvement of the cardiac valves and cardiac conduction system. Renal artery stenosis can lead to decreased renal function as well as to initiation of events producing renal hypertension. Corticosteroids have been the primary therapy for Takayasu's arteritis in the pulse less phase, since dramatic symptomatic improvement and restoration of absent pulses were reported to follow its use (4). However, the efficacy of corticosteroids in Takayasu's arteritis has not been universally acknowledged, since most patients with symptomatic arterial insufficiency do not manifest dramatic clinical improvement(4,5). Inhibitors of platelet aggregation or oral anticoagulants may be instituted in selected patients. Hypertension may be treated with calcium entry blockers or angiotensin-converting enzyme inhibitors. Life threatening or incapacitating arterial occlusions are sometimes amenable to surgical intervention.

## **ANESTHETIC CONSIDERATIONS**

Takayasu's arteritis may be encountered in patients presenting for obstetric anesthesia, incidental surgery, or such corrective vascular procedures as carotid endarterectomy.

Formulation of a plan for the management of anesthesia must take into account the drugs used for treatment of this syndrome, as well as multiple organ system involvement by vasculitis (2,3). For example, chronic corticosteroid therapy may result in suppression of adrenocortical function, indicating the need for supplemental exogenous corticosteroids during the perioperative period. Regional anesthesia may be a controversial selection in the presence of anticoagulation. Associated musculoskeletal changes can make performance of lumbar epidural or spinal anesthesia difficult.

Nevertheless, lumbar epidural anesthesia has been described for vaginal delivery and tubal ligation in these patients (3). Blood pressure may be difficult to measure noninvasively in the upper extremities. Indeed, blood pressure is predictably decreased in the upper extremities because of narrowing of the arterial lumen. There is a theoretical but undocumented concern regarding cannulation of arteries that may be involved by the inflammatory process characteristic of this syndrome. Nevertheless, a catheter placed in the radial artery is useful for confirming the presence of an adequate perfusion pressure from a catheter placed in the femoral artery is an option, but it should be recognized that systolic blood pressure in the legs will be higher than that present in the central aorta. In addition, constant monitoring of the ECG and of urine output provides an index of the adequacy of perfusion of the heart and kidney. Placement of a pulmonary artery catheter is acceptable if the magnitude of the surgery dictates (2).

In patients with known compromise of carotid blood flow, intraoperative monitoring of the EEG may be useful in detecting cerebral ischemia. It is important to recognize that hyperextension of the head, as during direct laryngoscopy for intubation of the trachea, may compromise blood flow through the carotid arteries shortened as a result of the vascular inflammatory process associated with this disease. Indeed, during the preoperative evaluation of the patient, it is useful to establish the effect of changes in head position on cerebral function. Ankylosing spondylitis and rheumatoid

arthritis may accompany this syndrome. Regardless of the drugs selected to produce anesthesia, the priority must be to maintain an adequate arterial perfusion pressure during the intra-operative period. Therefore, anesthetic - induced decreased blood pressure caused by decreased cardiac output or systemic vascular resistance must be recognized promptly and treated by either adjusting the concentration of anesthetic drugs or expanding the intravascular fluid volume, or both. The administration of a sympathomimetic to maintain perfusion pressure may be helpful until the underlying cause of the decrease in blood pressure can be corrected.

Avoidance of excessive hyperventilation of the lungs, as well as the selection of a volatile anesthetic, perceived to favor maintenance of cerebral blood flow, is a reasonable goal, especially in patients in whom the disease process involves the carotid arteries (2).

## **CONCLUSION**

Administering general anesthesia in Takayasu arteritis for major vascular procedures is challenging and needs special attention to preoperative optimization of the patient, perioperative monitoring of the cardiovascular system, and maintenance of cerebral and other organ perfusion and normal acid-base status.

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