Perioperative Management of Type A Dissection: focus on Marfan Syndrome

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Learning Objective
To review the anesthetic concerns related to care of the patient with aortic dissection, with a focus on Marfan’s syndrome.

Stem Case and Key Questions
A 34-year-old man with Marfan’s syndrome presents to the emergency room with tearing back pain and a cold right leg. Physical exam is notable for severe hypertension, aortic regurgitation and a cold right lower extremity. His electrocardiogram and myocardial enzymes are within normal limits. His chest X-ray reveals a widened mediastinum.

(1) The patient is currently taking losartan. What is its clinical benefit in Marfan’s syndrome? What is a perioperative concern with this drug?

The patient is admitted directly to the operating room after urgent aortic imaging reveals dissection of the ascending aorta with extension to the aortic bifurcation.

(2) What is the Penn classification of this clinical presentation? How does the clinical presentation of type A dissection affect perioperative risk?

After anesthetic induction, a transesophageal echocardiogram (TEE) is performed.

(3) What clinical questions should the TEE examination address? What findings do you consider particularly important in planning the central aortic repair?

The decision is made to proceed with aortic valve implantation, with replacement of the aortic root, ascending aorta and aortic arch.

(4) Should central aortic repair be performed immediately? Should the ischemic lower extremity be managed first?

(5) Which artery would you recommend should be cannulated for cardiopulmonary bypass? What are the risks and benefits with your choice?

The axillary artery is cannulated via a Dacron graft. The patient is placed on
cardiopulmonary bypass (CPB). Systemic cooling is begun. Deep hypothermic circulatory arrest (DHCA) is planned.

(6) How would you assess high resistance to arterial flow during initiation of CPB? Is this important or not?

(7) Which hematocrit range would you choose during cardiopulmonary bypass? What is your rationale? What are the risks and benefits of your choice?

(8) What type of blood gas management would you prefer in anticipation of DHCA? What are the quoted advantages and disadvantages of alpha-stat versus pH-stat in this clinical setting?

(9) What temperature would you plan for DHCA? Would you prefer an objective end-point such as offered by electroencephalographic or jugular venous saturation monitoring?

The patient has full electroencephalographic monitoring? When the ascending aorta is cross-clamped, the right cerebral hemisphere suddenly becomes isoelectric.

(10) Does this matter? How would you explain this development? What intervention would you recommend? What alternative cerebral monitoring would detect this complication?

The surgeon plans cerebral perfusion during DHCA.

(11) Is this required? What are the possibilities? What is the quality of evidence for a particular choice? Are there any anesthetic concerns?

The surgeon proceeds with anterograde cerebral perfusion (ACP) via the cannulated right axillary artery.

(12) During DHCA/ACP, the left-sided cerebral oximetry declines acutely. What is your diagnosis? What intervention would you recommend?

The aortic arch is repaired. The DHCA time is 55 minutes. Systemic rewarming is commenced. The surgical procedure is completed, as planned. After aortic unclamping, there is persistent ventricular fibrillation.

(13) What are the possible explanations? What interventions would you choose?

Separation from CPB proceeds smoothly. After protamine administration, the surgical field is still wet. The patient develops persistent mediastinal bleeding.

(14) What is your approach to this syndrome?

(15) On the second postoperative day, the patient complains of severe abdominal pain. What is your diagnostic evaluation? What are the therapeutic options?
The following discussion, structured as answers to the 15 case questions, is intended as a starting point for the discussion in the live PBLD session. Further details are also available in the provided references.

**Question (1):** Marfan’s syndrome has an autosomal dominant inheritance, with a prevalence of 1 in 5000 people. The associated aortopathy is ameliorated by antagonism of transforming growth factor-beta with losartan. Angiotensin blockade with losartan is a major risk factor for perioperative vasoplegia that may be catecholamine resistant. Pharmacologic rescue with vasopressin and/or methylene blue can be life-saving.

**Question (2):** The Penn classification stages acute type A aortic dissection by ischemic presentation. Penn staging stratifies perioperative mortality in this disease. The classification is defined as follows: class a (absence of regional or generalized ischemia with a perioperative mortality rate = 3.1%); class b (branch vessel malperfusion causing regional ischemia e.g. cold extremity; stroke, with a perioperative mortality rate = 25.6%); class c (circulatory collapse [systolic blood pressure < 80 mmHg] with or without cardiac involvement, associated with generalized ischemia and a perioperative mortality rate = 17.6%); and, class b + c (branch vessel ischemia and circulatory collapse together with a perioperative mortality rate = 40%).

**Question (3):** Firstly, TEE should confirm the diagnosis of aortic dissection. If dissection is proximal to the left subclavian artery, emergency aortic repair is indicated. Secondly, TEE rule out pericardial tamponade, and myocardial ischemia. Pericardial tamponade requires drainage by pericardiectomy. Coronary dissection with consequent regional wall hypokinesis may require coronary bypass grafting. Thirdly, TEE should evaluate the mechanism and severity of associated aortic insufficiency (AI). The major AI mechanisms are: (i) central AI due to a dilated aortic annulus and/or diastolic intimal prolapse through the aortic valve; and, (ii) eccentric AI due to leaflet restriction from intimal dissection.

The presented patient requires prosthetic replacement of the aortic root, ascending aorta and aortic arch. If possible, the native aortic valve should be preserved for optimal valve function and freedom from anticoagulation. Aortic valve reimplantation in a prosthetic aortic graft with fashioned sinuses of Valsalva will minimize native aortic leaflet stress.

**Question (4):** Emergent central aortic repair is recommended in acute type A dissection. In the majority of cases, branch vessel malperfusion resolves with central aortic repair. However, Penn presentation type b is still associated with a high mortality. Consequently, there are expert centers that first restore perfusion with intimal fenestration and/or endovascular stenting followed by delayed central aortic repair. There is currently a divergence of expert opinion as to which management approach is superior.

**Question (5):** This patient requires a total arch replacement due to his aortopathy. Since the ascending aorta is dissected and friable, it is often not the first choice for arterial cannulation. Typically, a major non-dissected aortic branch is preferred. Cannulation of the axillary artery via a Dacron graft permits anterograde cerebral arterial perfusion both during CPB and DHCA. If the axillary artery is cannulated, an ipsilateral radial artery catheter will often not yield accurate readings.

**Question (6):** High arterial resistance suggests cannulation of the aortic false lumen. The arterial cannula may have to be revised. TEE can visualize a guidewire in the true lumen of the thoracic aorta, and thus facilitate cannulation of the true arterial lumen. It is vital
that the anesthesiologist monitor aortic flow on initiation of CPB to detect malperfusion syndromes related to cannulation.

**Question (7)**: Aortic arch repair still induces significant perioperative organ injury. A hematocrit of about 25% during CPB may reduce this organ dysfunction, balancing the risks of anemia from a lower hematocrit and the risk of increased blood viscosity from a higher hematocrit.

**Question (8)**: Alpha-stat pH blood gas management is currently the standard for adult CPB/DHCA. A concern with pH-stat in adults is an increased cerebral embolic risk due to cerebral vasodilatation and enhanced cerebral perfusion. In pediatrics, however, pH-stat may be preferred because it promotes neuroprotection due to uniform cerebral cooling from the associated vasodilatation.

**Question (9)**: Expert centers differ significantly in their conduct of DHCA for aortic arch repair. Three cooling standards for DHCA have evolved: cooling to a goal temperature such as 18 degrees Celsius; cooling to an isoelectric EEG; and, cooling to a jugular venous saturation of 95%. Each PBLD participant is encouraged to research his/her institutional practice. Practice differences in DHCA will then be explored and discussed during the PBLD session.

**Question (10)**: This scenario is typical of brachiocephalic malperfusion. In this example, the surgeon was immediately advised to fenestrate the intimal flap above the aortic cross-clamp, whereupon the electroencephalographic signals briskly returned to normal. In our practice, there is about a 3% incidence of this intraoperative complication.

Brachiocephalic perfusion can be monitored with ultrasound (transesophageal; transpharyngeal; transcutaneous), regional oximetry and/or transcranial Doppler. Choice of monitoring is currently based on expert opinion and limited case series.

**Question (11)**: Expert centers differ significantly in their conduct of cerebral perfusion. There are currently 3 options: none; retrograde cerebral perfusion [RCP]; and, anterograde cerebral perfusion [ACP].

RCP is easy, cools the brain, and provides partial metabolic substrate and cerebral emboli washout. Although ACP may be technically more demanding, it is more physiological and lowers stroke rate when DHCA time > 45 minutes.

**Question (12)**: This scenario is typical of left cerebral ischemia. Assuming adequate ACP flow via the right axillary artery, there is inadequate collateral flow via the Circle of Willis. Based on autopsy studies, the Circle of Willis in this scenario is insufficient in up to 20% of cases. The solution is to perfuse the left brain via selective cannulation of the left common carotid artery and restore the left-sided cerebral oximetry to baseline.

**Question (13)**: Persistent ventricular fibrillation after aortic unclamping is from myocardial ischemia. It typically resolves with time during reperfusion. However, it may require lidocaine or amiodarone administration. In aortic root replacement, the left main coronary artery button may be dissected. This coronary ischemia may require coronary artery bypass grafting.

**Question (14)**: The patient requires mediastinal exploration if surgical bleeding is suspected. Factor VIIa is also indicated to augment hemostasis. Although not validated in randomized controlled trials, case series suggests adequate safety and efficacy in this setting.

**Question (15)**: This scenario is typical of splanchnic malperfusion, and may require surgical bypass, intimal fenestration, and/or endovascular stenting. Aortic stenting is
contraindicated Marfan’s syndrome, since the weak aorta will not anchor a stent graft.

References


