Case Report

Mid aortic syndrome correction: Anaesthetic considerations and management
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Abstract
A twelve years old female patient presented with headache off and on and disparity between upper and lower limb pressures. On the basis of history, physical examination and magnetic resonance angiography, the patient was diagnosed as mid aortic syndrome and planned for elective aorto aortic bypass graft surgery through left thoracotomy.

Double lumen endotracheal tube was placed for lung isolation and episodes of hypertension during proximal and distal cross clamp were controlled with sodium nitroprusside infusion (SNP). Extra pleural catheter was put in at the end of surgery before chest wall closure for postoperative pain control. Both upper and lower limb pressures were monitored in the cardiac intensive care unit (CICU) and over a period of twenty four hours, gradient gradually reduced to 10 mm Hg.

Introduction
Congenital coarctation of the thoracic aorta at the ligamentum arteriosum or in the aortic arch is well recognized, but a much less common variety of aortic coarctation is located in the distal thoracic aorta, abdominal aorta or both and is called "middle aortic syndrome" or "mid-aortic dysplastic syndrome". A high incidence of both visceral and renal artery stenosis is associated with this condition. Untreated patients may present with severe renovascular hypertension, myocardial infarction, heart failure, intracranial haemorrhage and aortic rupture. Most of the patients if left untreated, would die by 40 years of age. Surgery is the treatment of choice with excellent results although angioplasty has been used with variable success in the last two decades.

Case Report
A twelve years old female patient, weighing 43 kg, presented in the clinic with history of headache off and on, high blood pressure (BP) in the upper limb and disparity of pulses between upper and lower limbs. Her blood pressure in the upper limb and lower limb was 150/90 and 70/50 mmHg respectively. On the basis of history, clinical examination and magnetic resonance angiography (MRA), she was diagnosed as a case of mid aortic syndrome. Computed tomography (Figure 1) and MRA showed focal segmental narrowing in mid descending aorta opposite T7 and T8, with good collaterals for splanchnic, spinal cord and lower limb vessels. Trans-thoracic echocardiogram showed a long stenotic segment of descending thoracic aorta with a gradient of 120 mmHg and an incidental finding of supra-valvular aortic stenosis with a 40 mmHg gradient. Her blood pressure was initially controlled with captopril, lasoride and propranolol. She was planned for elective aorto aortic bypass graft surgery through left thoracotomy.

She was admitted in the hospital two days before the surgery to monitor and control her blood pressure. At that time hydralazine was also added for BP control. Patient was premedicated with midazolam 5mg orally one hour before surgery.

In the operating room noninvasive blood pressure, pulse oximeter and electrocardiogram leads were applied. Two 16 guage peripheral intravenous lines were placed. Patient was preoxygenated and induced with fentanyl 100 mg.
microgram, etomidate 10 mg and atracurium 30 mg. She was intubated with double lumen tube (28 French) to facilitate surgery. Arterial lines were inserted in right radial and left femoral artery and Central line was placed in right internal jugular vein. Bispectral index (BIS) was applied to monitor the depth of anaesthesia and was kept between 40 and 60. Nasopharyngeal temperature was used for continuous core body temperature monitoring. Patient was put in right lateral decubitus position, taking care of pressure points and neurovascular compression. One lung ventilation started and ventilator set on pressure control of 20, respiratory rate of 15, inspiratory and expiratory ratio of 1:2 and fraction of inspired oxygen (Fio2) 0.5. Episodes of hypertension during proximal and distal cross clamp were controlled with Sodium Nitropruside (SNP) at rate of 0.5 - 3 microgram /kg/min. Anaesthesia was maintained with propofol 2-4 mg /kg/hour and isoflurane 1-2%. Fentanyl was titrated to the total dose of seven microgram/kg. Perfusion distal to cross clamp was monitored by femoral arterial line and urine output. Intraoperative blood gases showed pH 7.35, PCO2 42, PO2 109, HCO3 22, Na 135, k 3.7 and Hb 7.7 gm/dl. Aorto aortic graft was put in successfully as shown in the Figure 2. Patient remained stable throughout the procedure. Extra pleural catheter was put in at the end of surgery before chest wall closure and continuous infusion of Bupivacaine 0.25% started at the rate of 8-10 ml/hour.

Total blood loss was 550 ml and two units of packed red blood cells transfused during the procedure. Arterial blood gas at the end surgery showed, Ph 7.36, PCO2 41 mmHg, PO2 84 mmHg, HCO3 22 mEq/L, Base excess -2.6, Hb11 gm/dl, Hct 32, K+ 3.97 mEq/L and Na+ 135 mEq/L. Proximal cross clamp time was 50 minutes and distal cross clamp time was 60 minutes. Urine output during clamping was 40 ml and mean lower body pressure was 30-40 mmHg.

Patient was extubated and shifted to cardiac intensive care unit (CICU) with supplemental oxygen and routine monitoring. In the CICU, blood pressure surges were controlled with Sodium Nitropruside (SNP) infusion. Extra pleural catheter and intravenous analgesics were used for pain control. Both upper and lower limb pressures were monitored in CICU and over a period of twenty four hours the gradient reduced to 10 mmHg.

**Discussion**

Middle aortic syndrome is a rare entity which is characterized by stenosis in the descending thoracic aorta and comprises of 0.5% to 2.0% of all the Coarctations. The condition is usually associated with some form of vasculitis especially Takayasu's but it may occur in isolation. The condition is usually diagnosed in young adults but may present in childhood as a challenging problem of managing hypertension. Stenting and surgery has been performed to treat this condition.

Anaesthetic plan should include perioperative blood pressure control, maintenance of distal perfusion during cross clamp, spinal cord protection, and anticipation of excessive blood loss and its appropriate replacement, application of one lung ventilation and postoperative pain control. In the present case, preoperatively the blood pressure was controlled with combination of ACE inhibitor, Beta blocker, loop diuretic and a direct acting arterial dilator. Intraoperatively, upper body hypertension was controlled with SNP while lower body hypoperfusion was minimized by adequate volume resuscitation before cross clamping. Fortunately, our patient had adequate collaterals which prevented lower body hypoperfusion. Postoperative pain was addressed with 0.25% Buvacaine infusion through extra pleural catheter.

Recognized complications of surgery are haemorrhage, paradoxical post-operative hypertension and paraplegia. The incidence of paraplegia is about 3 to 9% and prolonged cross lamp time is an important factor in the development of this complication. Long term results after surgery are satisfactory in the absence of arteritis however persistent hypertension and reno-vascular disease appear to affect life expectancy.

**Conclusion**

Appropriate anaesthetic management of mid aortic syndrome correction requires preoperative planning and anticipation of intraoperative adverse outcome. Management should include perioperative blood pressure control, maintenance of distal perfusion during cross clamp, spinal cord protection, anticipation of excessive blood loss and its appropriate replacement, application of one lung ventilation and postoperative pain control.

**References**

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Case Report

Epidural anaesthesia during labour for a patient with congenital complete heart block: A Case Report
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Abstract
We report labour pain management in a full term pregnant patient with Congenital Complete Heart Block. She delivered uneventfully under routine monitoring with facilities for pacing at hand. She previously had an uneventful normal delivery and a D&E, both outside our hospital. Only findings were a low heart rate of 45-50 beats per minute. She never had syncopal attacks. She had a good effort tolerance on ETT. Her ejection fraction was 60% on Echocardiogram. She was given a single shot low dose spinal with fentanyl followed by epidural insertion. She successfully delivered through mid-cavity forceps in about 2.5 hours. The only problem encountered was a transient bradycardia of 40 per minute with a systolic blood pressure of 70 mmHg, which settled with ephedrine.

Introduction
Congenital complete heart block is a rare congenital abnormality in which there is block at the AV node. Its incidence is reported to be 1:20,000 live births. Patients may experience syncpe and dyspnoea due to incomplete ventricular filling. In 53% of patients it may be associated with other cardiac lesions.² It has a strong correlation with positive autoimmune antibodies.³ It tends to worsen over time but rarely may resolve spontaneously. Steroids have also been reported to be effective.⁴ Pregnancy is usually well tolerated with this disorder unless there are symptoms of syncpe and dyspnoea. Pace maker insertion is recommended early in case the patient is symptomatic or has a prolonged Q-T interval or left arterial enlargement on ECG. Regional anaesthesia is recommended to prevent valsalva induced bradycardia or cardiac arrest during expulsive efforts by the patient.

Case
A 29 years old Gravida 2 Para 3, presented with 40 weeks gestation for delivery. She was known to have congenital complete heart block (CCHB). She had no history of attacks of syncop or dizziness. Only transient attacks of shortness of breath were reported, which settled without any treatment. She never had a pacemaker and had a good effort tolerance on ETT. Echocardiogram was within normal limits with an ejection fraction of more than 60%. Her labs were within normal limits. Her base line heart rate was 45-50 beats per minute and a blood pressure of 100/60 mmHg. She had one delivery and one D&E in the past which were both uneventful.

Monitoring was done with ECG, NIBP and SpO₂. Atropine, Swan sheath, Pacemaker and Transcutaneous pacing equipment were all at hand. As she was greatly distressed with pain a single shot spinal with 2.5mg bupivacain and 25µg fentanyl was given in sitting position with a 25G sprout needle followed by epidural catheter insertion with a 16G Tuohye needle. The patient had a transient bradycardia of 35 beats per minute with the systolic blood pressure decreasing to 70 mmHg. This was successfully treated with two boluses of 2.5 mg ephedrine and I/V fluids. A single 10 ml bolus of 0.1% bupivacaine was given after an hour. Infusion was not required as a healthy baby girl was delivered uneventfully with the help of mid cavity forceps.

Discussion
Women with complete heart block cannot augment their cardiac output due to limitation in their heart rate response, but generally they tolerate pregnancy well, unless