International Journal of Medical Anesthesiology

E-ISSN: 2664-3774
P-ISSN: 2664-3766
www.anesthesiologypaper.com
IJMA 2022; 5(3): 07-11
Received: 07-04-2022
Accepted: 09-05-2022

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A rare case of abnormal narrowing and bifurcation of descending thoracic aorta and perioperative anaesthetic management during aortic bypass surgery

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DOI: https://doi.org/10.33545/26643766.2022.v5.i3a.353

Abstract
Hypertension in paediatric population is an emergent global health issue, where the prevalence is varied among nations. Although vascular pathologies are one of the causes for secondary hypertension, mid aortic anomalies are not seen commonly. These patients usually present with uncontrolled hypertension or hypertension emergency or hypertension associated organ dysfunction. Medical management per se will be not enough to achieve a satisfactory control in blood pressure and most of them will have to undergo either surgical repair or an endovascular procedure. Perioperative management of these patients is a challenge to an anaesthetist due to multiple reasons such as haemodynamic instabilities, organ dysfunction, metabolic derangements and the need for invasive monitoring.
Here we present an 8 years and 10 months old boy who was investigated for hypertension and found to have abnormal narrowing and bifurcation of the descending aorta at T10 level and underwent successful aorto-aortic bypass surgery.

Keywords: Hypertension in paediatric population, haemodynamic instabilities, metabolic derangements, abnormal narrowing and bifurcation of descending

Introduction
Multiple vascular pathologies can lead to hypertension in paediatric population. Anomalies in descending aorta beyond the arch is not a commonly seen scenario and most of the time these patients may need to undergo an invasive procedure to achieve satisfactory control in blood pressure and to prevent end organ dysfunction [6]. Thoracic aortic surgeries comprise specific and significant challenges to an anaesthetist in the perioperative period.
Here we present the case of an 8 years and 10 months old boy who underwent an aorto-aortic bypass surgery for an abnormal narrowing and bifurcation of descending aorta at T10 level and discuss about the successful perioperative anaesthetic management.

Case history
Previously healthy, 8 years and 10 months old (body weight 27 Kg) boy was incidentally found to have high blood pressure, while evaluating inward for an episode of shortness of breath 4 months ago. His history and rest of the examination was insignificant.
He had undergone a range of haematological, biochemical and radiological investigations for the evaluation of cause of hypertension. 2D echo revealed moderate left ventricular hypertrophy with long segment narrowing of the lower thoracic to abdominal aorta. Following that, an urgent CT aortogram was performed, which showed abnormal branching of the descending aorta at T10 level (diameter at T9 level is 6mm), right trunk (10.5 mm in diameter) continuing as the distal abdominal aorta and bifurcate at L4 level to common iliac arteries. Left trunk (5 mm in diameter) gives off coeliac axis and terminates as superior mesenteric artery. Bi-lateral kidneys were normal.

Fig 1: CT aortogram

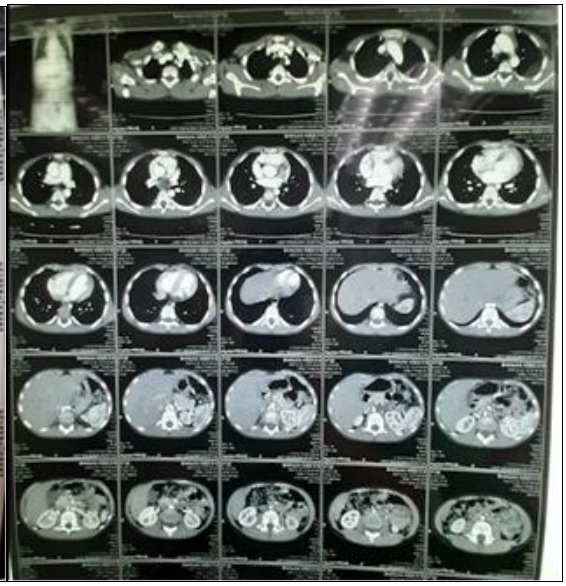

Fig 2: CT aortogram (cross section)

With these findings, he was prepared for the aortic bypass surgery and pre-habilitation was commenced. Blood pressure was optimized with prazocin 1mg 6 hourly and amlodipine 7.5mg mane dose.

Intraoperatively, patient was preoxygenated and 18G peripheral canula was inserted. Induction was done with weight-based doses of morphine, propofol and atracurium and intubated with size 6.5mm single lumen ETT. This was followed by the placement of a bronchial blocker into the left main bronchus under fiberoptic bronchoscope guidance. Anaesthesia was maintained with isoflurane and intermittent doses of atracurium were given to maintain the muscle paralysis. Lumbar drain was inserted at L4-L5 level in order to minimize the risk of spinal cord ischaemia. Thoracic epidural catheter was inserted at T8-T9 level for perioperative pain relief. Left internal jugular central venous line and right radial arterial line were inserted and invasive monitoring was established. Nasopharyngeal probe was used to measure core body temperature and precautions were taken to avoid hypothermia. Mean arterial pressure (MAP) was kept at 75-80mmHg range to maintain spinal cord perfusion pressure.

Patient was positioned carefully in right lateral position and pressure points and eyes were well protected. Surgery was carried out via left thoraco-abdominal lateral incision. During surgery, at the level of thoracic aorta, one lung ventilation was carried out. Epidural 0.1% 3- 5ml/hour infusion was commenced, and rate was adjusted according to haemodynamic parameters. Lumbar drain was zeroed at the level of iliac crest and CSF pressure was maintained at 10mmHg. Before initial cross clamping of the aorta, iv heparin 2500U was given.

Initial clamping time at the level of thoracic aorta was 51 minutes. During clamping period there was slight rise in blood pressure, which was managed by deepening the level of anaesthesia with isoflurane. Fluids were optimized and patient was hyperventilated with 100% oxygen prior to clamp release in order to minimize physiological consequences of unclamping.

After completing thoracic aortic graft anastomosis bi lateral lung ventilation was recommenced. 2nd aortic cross clamping was done below the origin of renal arteries and clamping time was 32 minutes. Physiological effects of clamping and unclamping was managed successfully as mentioned above.

Surgery was carried out over 6 hour's duration. Thoracic to abdominal aortic bypass was done with a synthetic graft with about 100ml blood loss. Left side intercostal tube was inserted prior to closure of thoracic cavity. ABG was performed at the end of the surgery showed a pH 7.31, pO2 123mmHg (FiO2 0.5), pCO2 41mmHg, HCO - 26mmol/L, base excess (-) 1.2mmol/L, lactate 2.7mmol/L. Throughout the surgery i.v. normal saline 500ml and ringers lactate 1000ml was infused and urine output was maintained more than 0.5ml/kg/hour.

Fig 3: Thoracic aorta prior to graft anastomosis

Fig 4: Thoracic aorta after graft Anastomosis

Fig 5: Abdominal aorta prior to graft anastomosis

Fig 6: Abdominal aorta after graft Anastomosis

Patient was transferred to paediatric surgical ICU with the ETT, and mechanical ventilation was continued overnight in SIMV pressure control mode. He was sedated with iv morphine 20microgram/hour and iv midazolam 1mg/hour infusion. Epidural 0.1% bupivacaine infusion and regular doses of iv paracetamol was continued for pain management. Haemodynamic parameters were monitored closely, and MAP was maintained at 75-80mmHg range. Lumbar drain was kept in situ and maintained CSF pressure at 10 mmHg while closely monitoring for neurological deficits. Immediate post operative investigations revealed Hb- 10.8g/dl, s. creatinine – 38 micromol/L, Na+ 139mmmol/L, K+ 4.8mmol/L, INR – 1.45, AST - 51U/L and ALT – 68U/L.

Patient was extubated on postoperative day 1 and chest rehabilitation was commenced. Antihypertensives, amlodipine 2.5mg mane and prazocin 0.5mg tds, were recommenced since postoperative day 2 as blood pressure was higher than the upper limit of normal for his age. Epidural catheter was removed on postoperative day 3 and pain relief was achieved with regular doses of paracetamol and oral ibuprofen. Lumbar drain was clamped after 48 hours and observed for the development of neurological deficit for about 8 hours and then removed. Central venous line and arterial line were removed on postoperative day 3.

Postoperative period was complicated with development of left sided lower zone consolidation and effusion which was successfully managed with iv antibiotics, chest physiotherapy, incentive spirometry and breathing exercises. IC drain was removed on postoperative day 6. Patient was transferred to ward on ICU day 5 and discharged from the hospital on postoperative day 13.

Discussion

The global prevalence of hypertension in paediatric population has risen over the last few decades and it is reported as 4%1. It is highly varied among nations, where in America it is 2- 4%2, while in Europe it is ranged between 2-13%3. In South Asia it is recorded as 5.54%4. There are multiple causes for hypertension in this population such as essential hypertension, obesity, familial or due to secondary causes such as renal parenchymal disorders, endocrine disorders or vascular pathologies. Among vascular pathologies, coarctation of aorta and renal artery stenosis or thrombosis are commonly reported [5].

Mid aortic syndrome (MAS) is another rare vascular cause of hypertension in paediatric and adolescent population, where there is a segmental narrowing of the distal thoracic or abdominal aorta and varying involvement of its branches. It can be either idiopathic or acquired [6]. Our case is different from other known vascular pathologies, where there was a narrowing of thoracic aorta associated with abnormal bifurcation at T10 level.

Most of the patients with mid aortic vascular pathologies, like in our case, present with uncontrolled severe hypertension or hypertension associated organ involvement. Apart from that, there can be various other presentations related to vessel involvement such as renal dysfunction and lower limb claudication [7]. Computed tomography angiography (CTA) and magnetic resonance angiography (MRA) can be used to effectively diagnose these conditions [8].

There are few available measures for the management of these patients. It can be pharmacological or an endovascular intervention or a surgical repair. The main aim of management is to control arterial blood pressure and minimize end organ complications. Most of the time pharmacological measures alone will be not enough to achieve a satisfactory control in blood pressure, in which case invasive procedures would be indicated. Aorto-aortic bypass graft surgery is the commonest surgical intervention with largely unknown medium or long term effectiveness. Therefore, multidisciplinary approach is essential to decide on definitive management plan [6]. In our case, surgical intervention was planned early, as he had hypertension related left ventricular hypertrophy at the diagnosis of the condition and his blood pressure was difficult to control with high doses of two oral antihypertensives.

Preoperative optimization is essential and is of paramount importance to achieve a better surgical outcome as they have to undergo a major, high risk vascular surgery. Blood pressure control, optimization of other medical conditions, haemoglobin and nutritional status, commencement of physiotherapy and breathing exercises and psychological support to patient and family are key components in these pre-habilitation programs [9].

Intraoperative management could be a challenging task for an anaesthetist and there are several important anaesthetic considerations. Securing of airway and one lung ventilation is one of the main concerns. Lung isolation can be achieved by using a double lumen tube or a bronchial blocker or an endobronchial intubation with single lumen ETT, with their own pros and cons. Specially in paediatric patients, when selecting lung isolation method and sizes of the airway devices and when performing bronchoscopy, extra precautions should be taken.10 Close monitoring of

haemodynamic, respiratory and ventilatory parameters are essential for the early recognition of complications, such as hypoxia.11 In our patient we used a bronchial blocker as the lung isolation technique due to lack of appropriate size of double lumen tube.

Spinal cord protection is another major intraoperative consideration, where paraplegia has been reported in 4-16% of thoracic aortic surgeries. Spinal cord perfusion pressure (SCPP) is determined by the MAP and cerebro-spinal fluid pressure (CSFP).

$$SCPP = MAP - CSFP^{12}$$

Following aortic cross clamping spinal cord blood supply will be compromised due to rise in CSFP and reduction in arterial blood flow. To minimize this devastating complication, CSF drainage has a major role [12]. Apart from that, maintaining of MAP around 80mmHg, alleviating other physiological insults such as hypoxia, hypercarbia, acidosis, hypothermia and ischaemia - reperfusion injury have significant impact on spinal cord protection. Intraoperative neurophysiological monitoring has a great value in early diagnosis of spinal cord injury and act promptly to reverse any correctable insult [13]. Some surgical techniques are also described to minimize spinal cord ischaemia such as sequential clamping of the aorta and reimplantation of intercostal and lumbar segmental vessels [12]. We inserted a lumbar drain at L4-L5 level after induction and CSFP was maintained at 10mmHg. Apart from that, precautions were taken to minimize other physiological insults.

Moreover, exaggerated physiological effects of clamping and de-clamping of aorta should be anticipated during thoracic aortic surgeries. During clamping, the risk of myocardial insufficiency due to increased afterload and preload and distal organ ischaemia are possible insults while sudden hypotension and ischaemia-reperfusion injury are major concerns during de-clamping. Fluid optimization and use of vasodilators will help to alleviate clamping related haemodynamic insults. Preparing for de-clamping is essential to minimize sudden haemodynamic instabilities. Vasopressors, 10% calcium gluconate and NaHCO - should be available inside the theatre and a period of hyperventilation will be beneficial to minimize acidosis [12, 14]. In our patient, we did not experience major haemodynamic or metabolic instabilities during clamping or unclamping of the aorta and we were able to manage without vasodilators or vasopressors.

Establishment of invasive monitoring, avoidance of hypothermia, haemostasis and patient blood management and careful positioning of the patient are some other important aspects to be considered during the intraoperative period. Pain management is paramount to achieve a good postoperative outcome. It should be multimodal with intravenous drugs, such as opioids and paracetamol, central neuraxial techniques and other regional thechniques [12, 14]. We were able to provide effective analgesia with epidural 0.1% bupivacaine infusion, i.v. morphine and i.v. paracetamol 2 doses 6hours apart.

Postoperatively, patients are monitored closely in an intensive care unit and most of these patients will require a short period of sedation and ventilation until haemodynamic and metabolic stability is achieved. Invasive monitoring needs to be continued in initial postoperative period. If lumbar drain is inserted intraoperatively, it should be kept in situ for 48-72 hours and prior to removal, development of neurological deficit after clamping it should be excluded. If not inserted, should be ready to be insert a lumbar drain at any point where spinal cord ischaemia is suspected. Pain management and monitoring for organ dysfunction such as acute kidney injury, paralytic ileus or mesenteric ischaemia, myocardial dysfunction and coagulopathy are other key components in postoperative care. Chest and limb physiotherapy are similarly important other aspects in postoperative period to improve perioperative morbidity and mortality and should be initiated from early postoperative period [12]. Our patient was closely monitored for haemodynamic instabilities, metabolic derangements and organ dysfunctions, especially spinal cord deficits in a paediatric ICU for 6 days.

Conclusion

Various vascular pathologies can lead to uncontrolled hypertension in paediatric population [5], while mid thoracic aortic anomalies are a rare cause of hypertension [6]. Most of the patients will require an invasive procedure at one point of their disease trajectory, as it is difficult to control hypertension with pharmacological methods alone [7]. Anesthetics management of these patients in perioperative period include many specific considerations and challenges to an anaesthetist14. Proper preoperative optimization, intraoperative meticulous management of haemodynamic and metabolic instabilities and excellent postoperative care will improve perioperative morbidity and mortality in these patients.

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