Coarctation Repair in an Adult Funambulist (Dombari)

Sushrut Suhas Potwar¹, Uday Eknath Jadhav², Abhishek Rajendra Potnis³, Karan Kumar Shetty³ and Heli Ketan Oza³

Abstract

Five to seven percent of all congenital cardiac disorders are caused by aortic coarctation. Funambulism or tight rope walking is a part of street circus acts earning livelihood for Dombari community, which requires utmost upper and lower limb strength and balance. Coarctation can be repaired surgically or with interventional approach. End-to-end anastomosis, subclavian flap aortoplasty, and extra-anatomic bypass grafts are a few surgical fixes. However, these approaches are not without risks involving the subclavian artery and spinal arteries thus putting limbs in jeopardy. Interventional procedures include balloon angioplasty and use of bare metal or covered stents but are associated with complications like repeated interventions or surgery, migration of stents compromising branches of aorta, thereby causing limb paresis or plegia, and are not very cost effective. The authors hereby discuss a cost-effective surgical management and spinal protection strategy for coarctation in a funambulist patient.

Keywords: Coarctation; Funambulist; Dombari; Spinal protection strategies.

Introduction

Three occurrences of coarctation of the aorta are thought to occur every 10,000 newborns, accounting for 5-7% of all congenital heart disorders [1,2]. Patients with aortic coarctation have a 35-year median life expectancy, and 90% of them pass away without treatment within 50 years. Systemic hypertension and abnormal upper and lower limb pulses are frequently found in adult life together with complaints of exertional headaches, leg tiredness, or claudication. Funambulism refers to tightrope walking that involves the use of arches of feet to grasp the rope between the great and the second toe while pivoting around the ankle joints and maintaining balance with a pole or bilateral extended arms.
This requires substantial lower limb strength and coarctation in such a case can prove to be hazardous to the performer. Crafoord performed the first successful surgical repair of aortic coarctation in 1944 [3].

Adult coarctation is treated using a variety of techniques, including surgery, balloon angioplasty, with or without stents, and pharmaceutical therapy. Several surgical procedures, including resection and end-to-end anastomosis, subclavian flap aortoplasty, and extranatomic repair employing bypass grafts or prosthetic patch aortoplasty, can be used to heal the damage [4].

Balloon angioplasty and covered stents may cause occlusion of local branches of the aorta including spinal arteries causing severe neurological complications like paraplegia or paraparesis (in 10% cases). The following case report presents a valuable management strategy for surgical repair of coarctation in a funambulist belonging to lower economic strata unable to afford interventional therapy.

Case report

A 21-year-old funambulist (Dombari) female, presented with complaints of shortness of breath, headache, weakness in patient’s lower limbs for three years. The patient was diagnosed as having a case of coarctation at an outside center two years back, but the patient refused treatment. After an episode of nasal bleeding, a patient presented at author’s institute. The patient examination revealed 140/90 mmHg pressure in both upper limbs. Bilateral femoral arterial pulses were palpable but weak and delayed compared to the upper limbs.

This requires substantial lower limb strength and coarctation in such a case can prove to be hazardous to the performer. Crafoord performed the first successful surgical repair of aortic coarctation in 1944 [3]. The different methods employed for the treatment of coarctation in adults include surgical, balloon angioplasty, with or without stents and pharmacological therapy. Surgical repair can be accomplished by several techniques like resection and end to end anastomosis, subclavian flap aortoplasty and extranatomic repair using bypass grafts or prosthetic patch aortoplasty [4].

Balloon angioplasty and covered stents may cause occlusion of local branches of the aorta including spinal arteries causing severe neurological complications like paraplegia or paraparesis (in 10% cases). The following case report presents a valuable management strategy for surgical repair of coarctation in a funambulist belonging to lower economic strata unable to afford interventional therapy.

Chest X-ray revealed notching of the inferior border of the 3rd–7th posterior ribs. Echocardiography showed narrowing distal to left subclavian artery with high-velocity color Doppler signal across the narrow segment, gradient of 68 mmHg, post-stenotic dilatation, obliterated large ductus but no other associated cardiac anomalies.

The patient CT angiography revealed a short segment coarctation in the vicinity of a large, obliterated ductus, dilatation of the proximal portion of the left subclavian artery and distal descending aorta (Figure 1).
Surgical intervention

The patient was posted for surgery with a plan for left subclavian artery to descending thoracic aorta bypass with epidural cooling and lumbar cold pack coverage for spinal protection. The chest was opened through a posterolateral thoracotomy via the 4th intercostal space. Short segment coarctation with a large ductus was visualized (Figure 2). In view of the hectic occupation of the patient, a decision was taken to mobilize the coarctation segment and ductus for the direct end-to-end repair of descending thoracic aorta (Figure 3). Authors had to clip and transect approximately three large collaterals for adequate mobilization of the aorta. The ductus was excised with the coarctation segment, and the pulmonary end was closed with a 6-o Prolene suture (Figure 4). The descending aorta was anastomosed end-to-end with 6-o Prolene suture. Post operatively, equalization of pressure was observed in the upper and lower limbs (Figure 5). The patient had an uneventful course post-surgery on biennial follow up in first year followed by annual follow up thereafter.

Figure 1: Pre-op computed tomography scan showing evidence of coarctation distal to the left subclavian artery, dilatation of proximal left subclavian artery and descending thoracic aorta distal to coarctation.

Figure 2: Intra-op image showing coarctation segment.
Figure 3: Intra-op image showing end-to-end anastomosis of descending aorta.

Figure 4: Intra-op image showing completed anastomosis.

Figure 5: Monitor showing equalization of pressures post-surgery.
Discussion

Different surgical treatment options for coarctation include anatomic repair, extra-anatomic repair, and transcatheter interventions depending on the age group of patients. Compared with surgical repair, balloon angioplasty is associated with higher incidence of restenosis and the use of bare metal stents is associated with the formation of aneurysms and paraplegia [5-7]. Surgical treatment in infants is preferred as complications are more common in older patients and can be detrimental as the surgical repair has been associated with extended recovery time, possible phrenic and recurrent laryngeal nerve injury, and lower body paralysis secondary to ischemic spinal cord injury [8].

Paraplegia secondary to spinal cord ischemia or upper limb weakness following subclavian artery flap angioplasty is a potential life-debilitating complication for a funambulist. Hence, after a thorough risk-benefit analysis, a definitive end-to-end repair with spinal protection strategies was planned for the patient. The aortic clamp and sew technique with mild hypothermia was used in this patient. A cross-clamp time of more than 30 minutes is associated with a significantly higher risk of paraplegia [9-11]. Therefore, the procedure was completed within 30 minutes of cross clamping. Hypothermia provides neuroprotection during ischemic events by reducing the cerebral and spinal cord metabolic rate of oxygen at the rate of approximately 5% per degree Celsius and by modulating release of excitatory amino acids [12-14]. Regional hypothermia was employed for cooling spinal cord through epidural space and by applying cold packs on lumbar area of the patient; thus, avoiding complications of systemic hypothermia such as cardiac arrhythmias, coagulopathy, and infections [15,16].

Different pharmacological neuroprotective strategies like use of magnesium sulfate, adenosine and medium molecular weight fraction of hydroxyethyl starch as described previously [17] were also used in perioperative period.

Conclusion

Repair of the coarctation of the aorta can be accomplished in multiple ways; however, the use of the most effective strategy differs on a patient-to-patient basis. Age, occupation, potential complications, and debility affecting daily living and risk-benefit analysis of each technique must be considered to determine the most suitable strategy for a patient. Since no method can alone provide absolute protection against post-operative limb paresis or plegia, an intelligent supplementation with neuroprotective manoeuvres and drugs must be undertaken. The inability to maintain frequent follow ups with cardiologist and re-intervention after endovascular approach due to economic constraint should direct the treatment strategy towards a definitive surgical repair with complete excision of the coarct segment and end to end repair.

This may require adequate mobilization of the aorta and protection of local branches with above mentioned strategies. On a biennial follow up in the first year, neurodeficit or discrepant blood pressure in any limb should be checked; after which annual follow up may be advised.
References


