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A series of diverse cases of aortic anomalies of valve, root, arch, trunk and associated anomalies and their surgical management - A single center study

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Abstract: *Aortic anomalies with associated cardiac and extra cardiac anomalies present a diverse spectrum of diseases that often demand implementation of diverse methods of repair and correction. Among various techniques, encompassing the spectrum from performing Bentall surgery to anatomic and extra-anatomic bypass like ventralization of aorta and using diverse perfusion maneuvers lay the foundation of achieving desirable outcomes in dealing with complex anatomic and physiological challenges offered by these anomalies. Critical and efficient decision making both on table and off table by the operating surgeon/ team is mandatory for balancing the aim of ensuring absolute anatomical and physiological correction that may sometimes prove to be a deterrent to survival against providing graded and adequate correction with a sustainable and non life threatening post op course. Here we present a single center conglomerate of total fifteen cases that primarily had aortic aneurysm/ dissection and co-arctation with associated conditions like LA Myxoma/ Pectus Excavatum/ Affection of Coeliac arteries and cases that otherwise were taken up with a routine treatment strategy but involved exercising mental faculties on table by the surgeon to achieve an acceptable outcome.*

Keywords: *Aortic Aneurysm, Bentall Surgery, Aortic dissection, Extra anatomic bypass, Coarctation of aorta, Ventralization of aorta*

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I. Introduction

Aortic diseases are to a great extent a disease of elderly and are becoming more common as population ages. The incidence of aortic diseases in India is expected to rise with increasing age of population [(1)]. Thoracic aortic aneurysms have an estimated incidence of 5-10 per 100,000 person-years and are classified into aortic root or ascending aortic aneurysms, most common (60%), followed by aneurysms of descending aorta (35%) and aortic arch (<10%). Thoraco-abdominal aortic aneurysm refers to descending thoracic aortic aneurysms that extend distally to involve the abdominal aorta [(2)]. Infrarenal aortic aneurysm is frequent in elderly and causes 1-3% of all deaths among men aged between 65 and 85 years. An infrarenal aortic aneurysm with diameter of 5.0 - 5.5 cm should be treated either surgically or by endovascular therapy [(3)]. As per the International Registry of Acute Aortic Dissection (IRAD), dissection presents with wide range of manifestations, and classical findings are often absent. A high clinical index of suspicion is necessary [(4)]. Acute dissection are classified based on site of main intimal tear : Type A : on ascending aorta; Type B :on transverse arch; Type C : on descending aorta. Extension of process is classified as - antegrade or retrograde [(5)]. Coarctation is found in 0.06% to 0.08% of general polulation, most commonly it involves aortic segment just distal to the origin of Left Subclavian artery [(6)].Profound hypothermia associated with circulatory arrest is the commonest method of cerebral protection during operations on aortic arch [(7),] however presently the approach has shifted to favour Hyothermic Circulatory Arrest with Antegrade Selective Cerebral Perfusion with variable flow [(8)] . Fibrin sealant is safe when applied properly, majority of clinical papers reveal a beneficial effect when used as a hemostatic and sealing agent [(9)]. At our center we use it for securing anastomotic lines.

II. Background

Aortic surgery largely involves management of aneurysms, which have been known as potentially ominous lesions since ancient times [(10)]. Bentall Procedure is considered gold standard in the treatment of patients requiring aortic root replacement. However, late mortality, major bleeding and thromboembolic complications remain a concern [(11)]. Bentall procedure is safe and durable operation, with very good early and long-term results and low rate of re-operation. At univariate analysis, long-term mortality was associated with age, diabetes, Bicuspid Aortic Valve, NYHA class III/IV, emergency treatment, cardiopulmonary bypass time, and coronary artery bypass grafting. Age, Bicuspid Aortic Valve and emergency surgery were important independent predictors of mortality [(12)]. In Marfan patients, the Bentall procedure is associated with excellent mid-term outcome [(13)]. Coarctation of the aorta in adulthood associated with extensive collateralization and also other cardiovascular anomalies which can be addressed simultaneously via an extra anatomic bypass grafting technique with low morbidity and mortality [(14)]. The use of bypass graft for single-staged repair of coarctation of aorta along with treatment of a coexisting cardiac disorder is feasible, because the bypass graft between the aortic arch and descending aorta follows the route of the native aorta [(15)]. Bovine Arch occurs in 15 - 35 % of population in USA [(16)]. A trend towards increased prevalence of bovine arch was seen in patients with dilated aortas (26.2%) compared to controls (20.5%, $p = 0.12$). The association was statistically significant in patients over 70 years old and when dilation involved the aortic arch [(17)]. No cannulation strategy (central aortic/ axillary/ femoral artery) can be completely free from the risk of intraoperative malperfusion during surgery for acute type A dissection [(18)]. Open Thoraco-abdominal Aorta Aneurysm involves surgical removal of a section of aorta and replacing it with an artificial graft. Its a complicated surgery involving large thoracoabdominal incision involving clamping the aorta in stages hence a heart lung machine is required to support circulation [(19)].

III. Methods

Performing Aortic surgeries at our center is a routine practice. However, here we have listed "unusual" aortic cases not having single anomaly with straightforward treatment protocol. The cases included in our study had a complicated anomaly profile and involved decent brainstorming both pre-operatively and on table to derive on to the final management protocol. Time frame is from September 2017 to March 2020. The age group was between 25 - 65 years and single case of a child of the age 13 years was kept. Out of fifteen patients, twelve were men and three were women. All patients were evaluated thoroughly for their cardiovascular diseases including 2 D Echocardiography and Aortograms and in special circumstances Contrast enhanced Computed Tomographic scans of chest and coronary angiography were done. Interesting associations were bovine aortic arch in two cases, pectus excavatum in one case, Left Atrial Myxoma in one and suspected Marfanoid habitus in one case each of root dilatation in which bentall procedure was performed. Five cases of Coarctation of aorta out of which four were adults out of these four, three had root dilatation and one has severe Aortic Stenosis and Permanent Pacemaker implanted. The issue of massive collateral formation in adults was bothersome and they were referred to the department of cardiology for interventional repair. However, successful outcome couldn't be achieved due to either failure or the endovascular repair not being feasible. Hence we decided to proceed with open surgical repair. Three cases with root dilatation were addressed with bentall procedure followed by extra anatomic bypass/ ventralization of aorta and one case without root dilatation also had VSD as an associated anomaly for which patch closure was done, the case which had severe AS had aortic valve replacement and extra anatomic bypass done. Four thoracic and thoracoabdominal aorta aneurysm with associated anomalies were addressed and lastly one De Bakey type III Aortic Dissection was opened up for graft repair but on table was managed with just apposition of the dissection flap and patient had an unremarkable post op course.

The below table 1 shows patient details with the procedure performed.

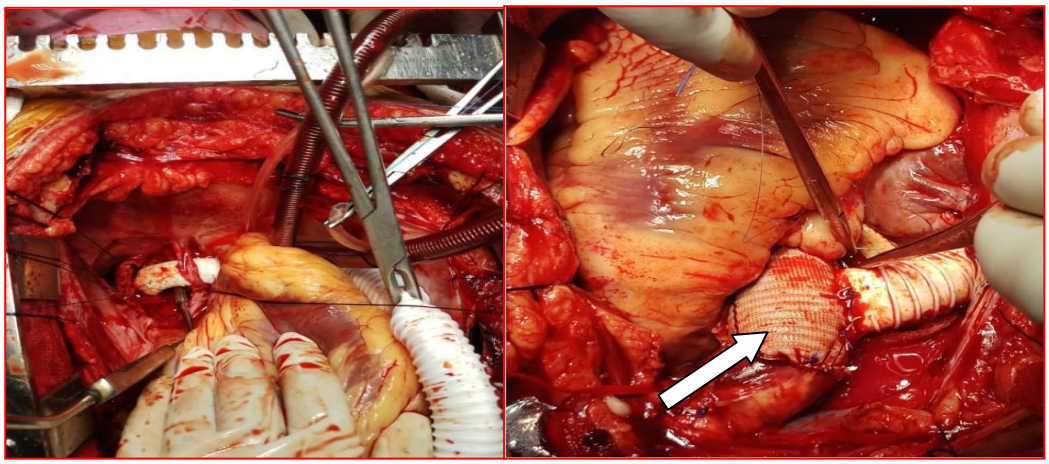
S No	Age (yr)/ Sex	Diagnosis	Primary Procedure	Simultaneous Procedure	Special considerations	Post op
1	45/M	Ascending AA with AS/AR with Small VSD with CoA distal to Lt Subclavian artery	Bentall Procedure with 23 mm Valve Conduit	Extra Anatomic bypass valve conduit to Post Coarct segment using 18 mm polyester graft and patch closure of VSD	FS used liberally to ensure Hemostasis	Continuous ooze. Heavy ionotropic support.
2	45/M	Ascending AA with Severe AR with Bicuspid valve with CoA distal to Lt Subclavian artery	Bentall Procedure with 25 mm Valve Conduit	Extra Anatomic bypass valve conduit to Post Coarct segment using 16 mm ePTFE ring graft	FS used liberally to ensure Hemostasis	Continuous ooze. Heavy ionotropic support.
3	27/M	Ascending AA with Bovine Arch	Bentall Procedure	Nil	FS used	Faint

		anomaly with Severe AR	with 23 mm Valve Conduit		liberally to ensure Hemostasis	contractile function of heart. Death 3 hrs post op.
4	33/M	Ascending AA with Root Dilatation with Severe AR with Marfanoid Habitus	Bentall Procedure using 27 mm size valve conduit	Nil	Nil	Unremarkable
5	25/M	Ascending AA with Root Dilatation with Severe AR with Pectus Excavatum	Bentall Procedure using 25 mm size valve conduit	Nil	Difficult and Tedious Sternotomy using oscillating saw.	Unremarkable
6	61/M	Asc AA with Root Dilatation with Severe AR with LA Myxoma with Infra renal Aortic Aneurysm extending to B/L CIA	Bentall Procedure using 23 mm size valve conduit	LA opened Myxoma excised	Nil	Unremarkable
7	13/M	CoA just Proximal to the origin of Lt Subclavian artery with small restrictive VSD & Bicuspid Aortic Valve	Proximal to distal segment bypass using 14 mm polyester Graft with inclusion of Lt Subclavian artery by Lt Thoracotomy	Nil	Intraop assesment of Lt Radial Pressure and LV Pressure No CPB	Unremarkable
8	60/M	Descending TAA with Infra Diaphragmatic Extension	Thoracoabdominal incision with 16 mm Interposition ePTFE graft repair	Nil	FS used liberally to ensure Hemostasis DLT	Unremarkable for 3 days then lung collapse
9	49/M	CoA distal to origin of Lt Subclavian Artery on PPI (implanted previously) with Severe Aortic Valve Stenosis	AVR #21 Cardiamed valve by Midline Sternotomy	Extra Anatomic bypass Ascending Aorta to Post Coarct segment using 16 mm ePTFE graft	DLT FS used liberally to ensure Hemostasis	Poor Cardiac Performance. Heavy ionotropeic support. Death on POD - 1
10	59/M	Severe AR with Asc AA with TR with EF = 30 % with CoA with Massive Cardiomegaly	Bentall Procedure using 21 mm size valve conduit	Conduit to Distal Aorta Extra Anatomic Bypass using 22 mm polyester Graft	DHCA FS used liberally to ensure Hemostasis Packing done intraop due to excessive oozing.	Heavy ionotropic support. Death - POD 1
11	50/F	Asc AA with Severe AR with Bovine Arch	Bentall Procedure with 23 mm valve conduit	Nil	Nil	Unremarkable
12	49/F	Complete Descending Thoracic Aorta Aneurysm with Dissection in entire length with one Large Pseudoaneurysm	Excision with Interposition Graft repair by 16 mm Polyester graft by Lt Thoracotomy	Nil	DLT Femoral Cannulation for Lt Heart Bypass Cell Saver used	Post op lung collapse as evident on reexploration
13	45/M	Supra-Renal Abdominal AA with Extension into Thoracic Aorta and Coeliac Artert origin Aneurysm	Lt Thoraco Abdominal Incision with Interposition Graft repair by 16 mm Polyester graft	Nil Coeliac artery searched vigirously but not delinated	DLT Femoral Cannulation for Lt Heart Bypass	Post Op Lung Collapse and Neurological event leading to death on POD 2
14	48/F	Descending TAA with huge Pseudoaneurysm eroding into vertebral column (T4-T7)	Excision with Interposition Graft repair by 16 mm Polyester graft	Nil	DLT used DHCA used	Neurological event and Lt Lung Collapse
15	64/M	Aortic Dissection (Stanford B/ De Bakey III) upto just proximal to Rt CIA bifurcation with bilateral Renal arteries arising from true lumen	Aorta transected just distal to Lt Subclavian. False lumen closure approximation and End-End Anastomosis done completing repair	Nil	Lt Thoracotomy DLT Lt Femoral Cannulation for partial Bypass False lumen approximation done by Vigorous use of FS and Teflon felt	Unremarkable

(DLT - Double Lumen Endotracheal tube for single lung ventilation/ DHCA - Deep Hypothermic Circulatory Arrest/ TAA - Thoracic Aorta Aneurysm/ AR - Aortic Regurgitation/ AS - Aortic Stenosis/ Asc AA - Ascending Aorta Aneurysm/ CoA - Coarctation of Aorta/ TR - Tricuspid Regurgitation/ POD - Post Op Day/ VSD - Ventricular Septal Defect/ CPB - Cardiopulmonary Bypass/ AVR - Aortic Valve Replacement/ ePTFE - expanded Poly Tetra Fluoro Ethylene/ CIA - Common Iliac artery/ LA - Left Atrium/ FS - Fibrin Sealant)

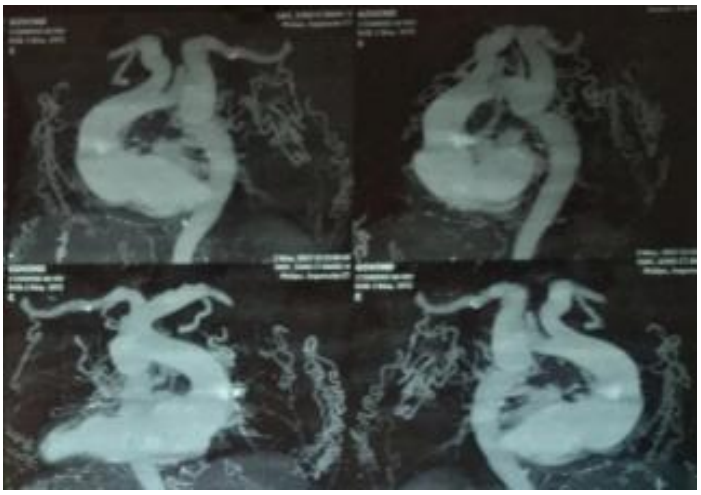
III. (1) Operative Technique

The surgical approach in all cases were standard approaches. Looking at the difficult nature of cases no minimally invasive or short incision techniques were used. Standard incisions - Median sternotomy/ Lt Thoracotomy/ Lt Thoracoabdominal incisions were given. Invasive monitoring of femoral arterial and where ever applicable of radial artery pressures of one/ both extremities were used. Standard cannulation techniques were used and in suitable cases bypass was established by femoral cannulation usually of right and in one case from left side. The Polyester valve conduits were place after appropriate sizing. Polyester/ ePTFE tube grafts were anastomosed to the descending thoracic aorta in end-to-side fashion with the use of a side biting clamp. The graft was then clamped, de-aired carefully and positioned for proximal anastomosis. For thoracoabdominal cases clamps were applied for the duration of anastomosis and then were released. Hemostasis was done vigorously by using Fibrin sealants, Absorbable oxidized regenerated cellulose, electrocautery, monitoring of activated clotting time and post op administration of fresh frozen plasma, platelet concentrates and amino-caproic acid. Post closure if a double lumen tube has been placed was changed to a single lumen tube by the anaesthetic team on table and then the patient was shifted to ICU on mechanical ventilation.



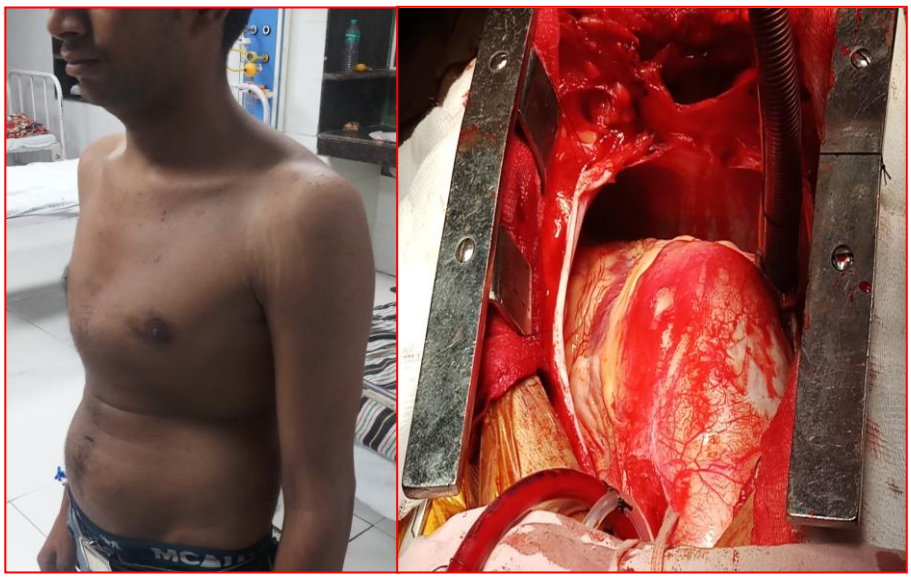
Case No - 01

Bentall procedure with Extra-anatomic bypass (conduit to distal aorta) using ePTFE ring graft. End-side anastomosis of conduit to graft is shown by white arrow.



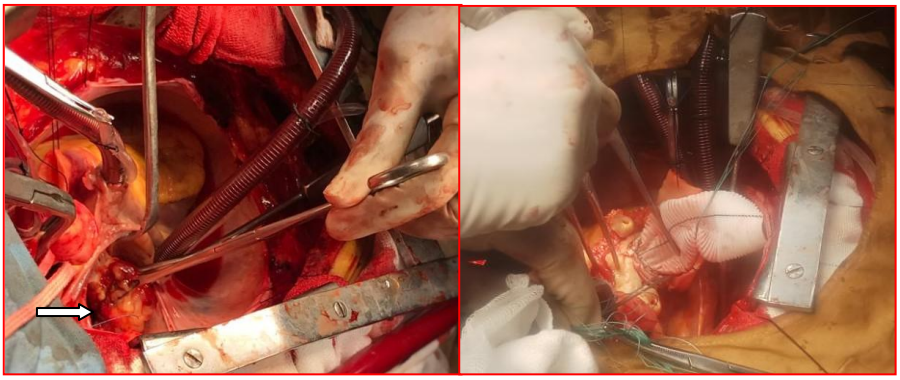
Case No - 02

Adult Coarctation with root dilatation with multiple collaterals seen



Case No - 05

Pectus Excavatum deformity with Aneurysm of Ascending Aorta



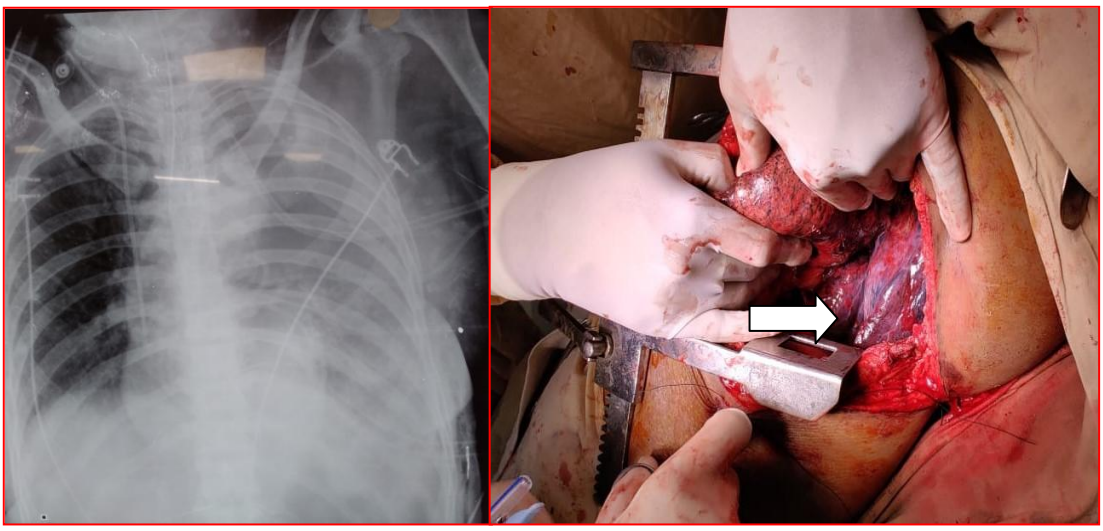
Case No - 06

Excision of Lt Atrial Myxoma (white arrow) followed by placement of valve conduit

Case No - 11

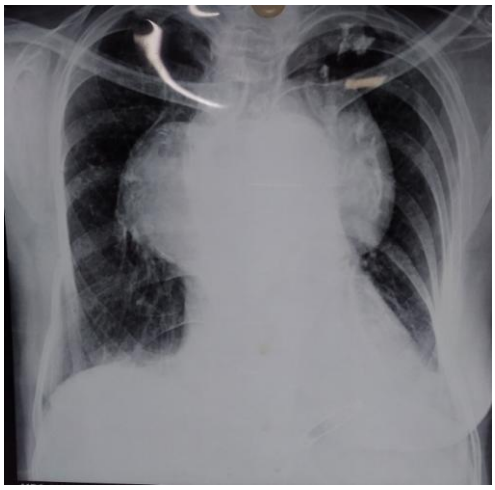
Bentall procedure done with associated Bovine arch.

Unrearkable follow up angiogram.



Case No - 12

Lt Lung collapse post op (on X-ray and on Re-exploration). White arrow indicating lobar collapse.



Case no - 14

Massive Aneurysm of Descending TAA eroding into vertebral column

Case No - 15

Aortic Dissection flap repaired by approximation showing regression of dissection in Abdominal Aorta and Iliac arteries. (No graft used)

IV. Results

In our study, there are total 15 "unusual" aortic cases not having a single anomaly with straightforward treatment protocol. We have achieved remarkable success in performing Bentall procedure in one patient of marfanoid habitus/ one patient of pectus excavatum/ one patient of Lt Atrial myxoma in which myxoma excision was also done. The last patient also had infra-renal aortic aneurysm which was left for second stage repair. Two cases of bovine aortic arch were also subjected to Bentall procedure out of which one had unremarkable post op and one died in the immediate post op due to failure of contractile function of heart. The Debakey type III dissection patient operated only by flap approximation and the 13 year old child in whom CoA repair was done including Lt Subclavian artery in the polyester graft, both had unremarkable post op course owing to on table decision regarding minimalistic intervention.

Three cases of Bentall Procedure combined with extra anatomic ventralization of aorta due to associated Coarctation were done. One had associated VSD which was addressed by patch closure and one had poor LV function (EF = 30%) and this patient suffered mortality on post operative day 1 owing to continuous ooze and poor contractile function requiring heavy ionotropes. One case of CoA with PPI requiring Aortic valve replacement due to associated Severe Aortic Stenosis followed by extra-anatomic bypass suffered mortality on first post op day due to contractile failure.

Two thoracic aorta aneurysm operated showed lung collapse due (double lumen endotracheal tube was used in both) and one also showed confusion in immediate post op, however both were finally extubated.

Two Thoracoabdominal aorta replacement out of which one had associated coeliac artery aneurysm and this patient suffered post op lung collapse and suspected neurological event eventually leading to mortality. The other case after successful extubation suffered lung collapse on third post op, day requiring reventilation.

V. Discussion

As a centre with more than 1000 on bypass cases in a year we have operated on aortic anomalies with remarkable success but, still we need to pursue the lacunae in the decision making, perfusion, post op management of "unusual" aortic cases involving more than one anomaly. Isolated Bentall procedure has given the best outcomes.

Complex adult coarctations associated with arch aneurysm and collaterals pose a challenge. Performing extra anatomic bypass in such scenarios enable surgeon to avoid extensive dissection and mobilization. In our context, extra-anatomic ventralization of aorta for coarctation with heavy collateralization with associated root dilataion were attempted as interventional dilatation by cardiologist were not feasible or successful. . Exposure of descending thoracic aorta through midline sternotomy by posterior pericardial approach was first described by Vijayanagar et al. in 1980 [(20)]. He placed the extra anatomical graft from ascending aorta to the descending aorta along the left heart border. Powell et al. described the same technique with extra anatomical graft placed along the right heart border in 1983 [(21)]. These cases are difficult to manage due to the issues of neurological events and hemostasis.

The problem of post op lung collapse was encountered in cases of Thoracic/ Thoracoabdominal aortic surgery worth mentioning the use of DLT in these cases.

An integrated team approach with anaesthesist and perfusionist need to be developed with diligence to further improve the outcomes in difficult and diverse aortic surgeries.

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