ADDIS ABABA UNIVERISTY

SCHOOL OF MEDICINE

DEPARTMENT OF PEDIATRIC AND CHILD HEALTH

Clinical profile and outcome of repaired cases of Coarctation of Aorta at TASH paediatric cardiology unit Ethiopia from Jan 2009 to Feb 2017G.C

Investigator: Kalkidan Gebremeskel M.D, Senior Resident, College of health science, School of medicine, Department of paediatric and child health

Cellphone - 0911-56-70-58

E Mail - Kalism2@yahoo.com

Advisor: Dr Tamirat Moges, Consultant pediatric cardiologist, chair of department Pediatric and child health, college of health science, Addis Ababa University

Cell phone - 0911-40-52-60

Email - mogest98@yahoo.com

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Abstract

Introduction

Coarctation of aorta (COA) is one of the acyanotic congenital heart disease where the portion of aorta gets narrowed often involving the descending aorta distal to the aortic isthmus. It requires treatment with definitive surgery or balloon dilation and if not treated associated with many complication. But even after the advert of multiple surgical approach it is associated with post-operative complication at times may also need lifelong post op follow up. Little experience is available in Ethiopia regarding post coarctation of aorta repair.

Objectives

To determine clinical profile and post-repair outcome of children with COA who underwent repair for COA.

Methodology: The study was conducted from March 2017 to Sept 30 of 2017 with a retrospective chart review of 39 patients with repair for native COA from Jan 2003 to Feb 2017 both at local and outside of the country. In this study clinical profile, immediate complication and outcome were assessed.

Result : A total of 39 patient with repair of COA of which 27(67.3%) were male and 12(30.8%) female. The age at time of intervention ranges from 3months to 25 years, with mean age of 6.9 ±5.7 yrs. 82.1% of the patients were hypertensive before intervention. 15(38.5%) had complex COA,13(33.3%) discrete COA,7(17.9) simple COA,4(10.3%) with interrupted aorta. Type of intervention was balloon angioplasty with or without stent in 20(51.3%), end to end resection and anastomosis 14(35.9%) and patch arthroplasty in 5(12.8%). Immediate complication seen were rebound hypertension 18(46.15%), post coarctectomy syndrome 3(7.69%). Other complication include Intracranial hemorrage, spinal cord injury ,pneumomedastinium ,chylothorax, Infective endocarditis, pericardial effusion, hemodynamic collapse 2ry to bleeding , lung atelectasis complicate 1(2.56%) each. There was no death following surgery. At 6 month 35 patient were evaluated at follow up clinic and only 40% of patients where hypertensive and echocardiography shows residual COA in 7(20%) and aneurism in 1(2.85%) patient.

Conclusion: the most practiced method of the repair in the study was balloon angioplasty. Rebound hypertension was the most common immediate complication encountered. Mortality rate is very low comparing it from other setting. Surgical repair has significant association with rebound hypertension. Six month follow up echocardiography showed significant number of re-coarctation

ABBREVATIONS AND ACNONYMS

BMI _ Body mass idex

BP – blood pressure

BSA _ body surface area

CV- cardiovascular

CVA – cerebro- vascular accident

CCE – cardiac centre Ethiopia

COA – coarctation of aorta

Diss Ao - descending aorta

JVP – jagular venous pressure

IE _ infective endocarditis

ICH _ intracranial hemorrage

LSB _ left sternal border

LSCA- left sub-clavian artery

Post op – post operative

PR – pulse rate

RR- respiratory rate

SO2 – oxygen saturation

SPSS – software statically package for social science

TASH – Tikur Anbesa Specialized Hospital

TEMP- temperature

1. Introduction

1.1. Background

Coarctation of aorta is constriction of aorta of variable degree. In 98 % of the case the constriction is just below the origin of the subclavian artery at the origin of the ductus arteriosis.**(1**) It is one of the acyanotic heart disease, accounting for 6- 8% of the congenital heart disease**.(2**)

Coactation of aorta may be simple if it occurs alone or with patent ductus arteriosis or can be complex if associated with other intra cardiac congenital lesion. In 85% of the case it is associated with bicuspid aortic valve.(3) And sometimes with left out flow abnormality as shone complex.(4)

COA is associated with extra cardiac vascular anomalies like brochiocephalic artery anatomy variation, collateral artery, aneurism in the circle of willis and anomalous left subcalvian artery and right subclavian artery origin. It may also associated with extra cardiac non vascular anomalies like musculoskeletal, genitourinary gastrointestinal or respiratory anomalies as high as 25% of the cases. (5)

Coarctation of aorta may present as infant with Congestive heart failure, child or young adult with hypertension or inr3cidental murmur. The classic finding of COA is discrepancy in pulse or Blood pressure between the upper and lower extremity.

Echocardiography and Doppler provide non invasive and accurate assessment of COA anatomy and physiology in most of the patients. Occasionally we may need CT or MRI angiography with diagnostic cardiac catheterization for diagnosis.(6)

Surgery remains safe treatment of choice at most center for isolated juxtaductal COA. Through years different surgical techniques have been practice for repair of COA. Which include end to end anastmosis which is the first reported method and still the preferred methods in most centers(7), patch angioplasty second to be inrtoduced(8) which requires less mobilization but with high incidence of aneurism(9-12) , sub clavian flap procedure with less aortic mobilization and avoidance prosthetic material but with un to ward effect on the left sublcalvian artery (13,14) percutaneous balloon angioplasty which gains more acceptance in recurrent repair of COA and remain controversial for primary treatment of native COA,But with some effectiveness on those with native COA(15-21)

The prognosis of repair of COA is excellent. The immediate complication that occurs post repair include rebound hypertension, post coarctectomy syndrome, mesentic arteritis which present with hypertension and acute abdominal pain with symptom and sign of bowel ischemia, spinal cord injury ,residual murmur and residual pressure gradient , recurrent laryngeal nerve with hoarsness of sound and phrenic nerve injury , Chylothorax , infection and bleeding, femoral artery injury and hemmorage especially if it is percutaneous ballon angioplasty in infant.

The late prognosis after COA repair affected by presence of associated intracardiac lesion (22) and the technique of repair. The long term complication includes Recoarctation, aortic aneurism, aortic dissection or rupture, long standing systemic hypertension, left sub clavian steel syndrome, increase risk of coronary artery disease and stroke , and endartritis.

1.1. Statement of the problem

Coarctation of aorta has incidence of 1 in 2500 of life birth, with male to female ratio of 2:1**(1,23)** . Untreated COA has been associated with high mortality 75% die at the age of 46 years because of aortic aneurism dissection or rupture , cerberovascular accident like stroke(**24)**.

Mortality COA repair which is isolate is 0% but with complex one with intra cardiac lesion may reach to 2% and may reach to 10% in those with complex cardiac lesion. Morbidity form the procedure may vary from procedure to procedure**.(25-27)**

The long term complication of post coarctation surgery are well establish in studies and includes hypertension, re-coarctation residual gradient in 8-27% in native balloon angioplasty(15,19,21),recurrent stenosis aneurism formation in 5-10% in native balloon angioplasty , Residual murmur, aortic aneurysm, aortic dissection, aortic rapture cerberovascular accident, endarteritis.

The risk of late hypertension may be as high as 10-20% however even if the coarctation repair in infancy (28,29). The occurrence of aortic aneurism may be as high as 24% in those with patch aortoplasty repair of COA(65) and it is 5-10% following balloon angioplasty (19,20,21).Subclavian steal syndrome with growth restriction of left upper exterimity and and intermittent claudication in those with subclavian artery flap.(30,13,14). Cerebrovascular accident is impotant cause of late mortality (31,32)

In Ethiopia the experience of surgery is of recent and started at our cardiac centre which started to be functional at 2009 and only few patients undergo surgery for COA. No written data or research has been done on those patients about the post op profile till now. This research is conducted to close this gap of information and to know what is the post profile and the observed complication in those patient and compare it with other countries experience. And also this research can provide a base line data to further research in this topic as the capacity of the cardiac unit is increasing and the number of patient operated increase over time

2. Literature review

Various literature reviews from developed countries justified that hypertension occurs even after surgery done for coarctation of aorta. Other immediate complication like post coarctectomy sydrome, rebound hypertension and long term complications like recoarctation , long standing hypertension, left subclavian steel syndrome are not un common to be seen in postoperative coarctation patient . But still, surgery significantly decreases hypertension from the preoperative period. This was reflected by the Koller M et'al study which retrospectively analyzed 362 who underwent surgery for coarctation of aortal which showed the incidence of hypertension decrease 82.5% preoperatively to 33.5% at discharge from the hospital and further decrease as futher follow up. (33)

In majority of the studies end to end anastomisis is the most used method. And post op compliacation are not uncommon to see however post op death is very signicicantly lower which emphasis that surgery is the most safe and effective way of treatment.(25-27)

Study done in USA at Mayo clinic , Rochester, Minnesota which include 819 patient form 1946-2005 with isolated coarctation of aorta who underwent primary operative repair , the mean age was 17+_ 13.6 years among them 83 % have pre hypertension. The most freuquently used type of operation in this study was end to end anastmosis (n=632) , followed by, patch angioplasty (n=72), Interposition graft (n=30) subclavian flap and other (n=35). Overall early mortality < 30 days is 2.4%. mean follow up 17.4 $\pm$ 13.9 years. Actuarial survival rate were 93.3 %,86.4% and73.5 % at 10,20 and 30 years respectively. Younger age at time of repair (P< 0.001) and end to end anastomosis technique (P<0.001) were independently associated with low rate o f re-intervention.(34)

In another retrospective Study done in Brno, Czech Republic, centre of cardiovascular surgery and transplantation,376 patient treated at the centre 1978 -2001 the average age at surgery was 7.2+- 8.9 years and 41 patient were younger than 3 months. Most common method used was end to end anastmosis (n =189), followed by patch graft angioplasty (n=136) and subclavian flap angioplasty(n=24). 314 of them has long term follow up with good out come. Of them 24(7.6%) underwent reoperation for recoarctation. The were no relationship found between recoarctation and type of operation they undergo. 15(4.8%) had persistent hypertension although good anatomical repair was done. Best outcome was seen in those with operation done in preschool age where there is reduced late complication of surgery seen.(35)

In another study done in USA a total of 235 adult with coarctation with mean age of 27+- 13 years) were retrospectively reviewed. They are grouped in to three group goup 1 treated with surgery9N=180) group 2 treated with balloon angioplasty or stenting (n=28) and group 3 no previous intervention has been carried out in 26 patient with mild coarctation at diagnosis. Aortic wall complication found in 37 patient(16%).there were no difference among the three groups with respect to total complication (15%,18%, and15%respectively) ascending aortic aneurism (9%,11%,and 12%) or descending aortic aneurisms (4% in all three groups). No significant relationship between previous repair type of repair age at repair, residual Doppler pressure gradient, or systemic hypertension and the occurrence of aortic complications. Only aging and bicuspid aortic valve were signigicantly related to these complications.(36)

A 30 years experience of surgery of coarctation of aorta from Lausanne , Switzerland , review 141 patients who underwent surgery for coarction of aorta form which 30 are neonate ,29 infant , 45 children , and the rest 37 were adult. And the result show no hospital death but late death 1 in the adult and 3 in the pediatric age (2.9%). Re coarctation was seen in 10% of the paitients and end to end anastmosis show lowest incidence of reocoarctation($P< 0.005$).3 patient form late survivors of adult group(8.3%) have medical management of systemic hypertension.(37)

According to another research form Tehran, Iran which is a 10 years retrospective study which analyzed 188 patients who underwent surgical repair for coarctation of aorta.Un like the other studies, in this study the most common method included was patch graft angioplasty (59%) followed by resection and end to end anastomosis(20.7%) , subcalavian flap angioplastySCFA (16.5%) and the result shows over all mortality of 2.6% . and highest evidence of recoactation was found in patch graft angioplasty(12.7%) lowest in SCFA (3.2%). Long segment COA shows Increase chance of recoarctaion. Unlike most other study this study shows no increase incidence of reocarctation in age below 1 year.(38)

A fourteen years Experience form India, Christian medical college hospital, Vellore, India reviewed 51 patient who underwent operation for coarctation of aorta, from which only 3 were asymptomatic unlike most western studies. Operation procedure include resection and graft interposition, resection and end to end anastomosis , bypass grafting and patch angioplasty. Patch angioplasty seemed by far the most satisfactory procedure. Overall mortality was 9.8%. Recoarctation was not seen in this series. The presence of associated defect affect result adversely.(39)

From few experience from African continent , a research done in Ibadan university college hospital cardio- thoracic surgical unit reviewed 694 admission to its unit from 1977 to 1998 form which 19(2.7%) of the admission where for coarctation of aorta and of which 15 patient underwent surgery for coarction of aorta with age ranging 1 month to 30 years. The method of intervention was resection and end to end anastmosis in 4 ,Dacron tube interposition graft in 3, Dacron patch graft in 4 and for the other 5 other method were used. four of the patient (25%) died after operation. Complication of the surgery which were seen include post op haemorrhage in 2 patients, intra operative haemorrhage in 1 , hoarseness of voice in 4 patients, paradoxical hypertension in 3 patient , 2 had chylothrax and 1 had graft occlusion and wound dehiscence. In this morbidty and mortality of coarctation of COA is higher as compare to the developed country and other long term complication is not documented to compare it with the other studies.(40)
In study that compare the prevalence of rebound hypertension in a balloon dilation and those with surgical correction.

The morbidity and mortality of coarctaion of aorta surgery is mainly determined mainly presence of associated cardiac malformation, post operative hypertension and age of patient at time of surgery. According to Koller M et'al study which review 362 patient opereated up on for caortcation of aorta form 1961 – 1980 and analysed retrospectively. Associated cardiovascular malformations were more very common especially in those ages less than 2 years and associated with higher mortality. Early morality was 12.2 % in those age less than 2 years of age and 1.4 % in those age greater than 2 years. Of those 336 followed 6 month to 21 year(8.9 years) and late mortality was 0.8 per patients year and associated cardiac defect and post operative hypertension were responsible for late death. This study also noted that patient operated between 2-9 years showed lowest risk of coarcation residual and post op hypertension.(33)

Depending on the series reviewed and definition used, up to three quarters of all patients with repaired COA will have systemic hypertension by 20±30 years after operation.(41,42,43) However, the mean age of COA repair in these early series was relatively high between 10 years of age and subgroup analysis suggested that earlier repair might be associated with lower prevalence and or less severe late hypertension. In a study of 226 patient undergoing coarctation repair before and after the age of 20 years, Presbitero reported that 30 % and 60% were hypertensive at 20 years follow up, respectively.(42). Similarly, in a study of 571 patients, Cohen and colleagues showed that late hypertension occurred in 7% of those operated on as infants as opposed to 33% who had undergone repair after the age of 14 years.(41) Consistent with these findings, Brouwer and colleagues speculated that the optimal age for elective surgery was 18 months, since this age grouped showed the best results with respect to re stenosis, normal blood pressure, and life expectancy(43)

Study done in Milton S.Hershey medical centre, which review 15 patient with repair of COA, revealed 56% incidence of paradoxical hypertension in immediate post- operative time (46). Comparative Study done 8 patients with balloon dilatation and 7 patient with surgical repair showed that all of the 7 patients with surgical repair showed increase in both SBP and DBP where as in those with balloon angioplasty SBP decrease and that of DBP un affected (47).

2. Objective of the study

General objective: to describe the clinical profile and post-repair outcome of patients who undergo repair for COA.

Specific objectives:

1. To describe the demographic and clinical characteristic of patients
2. Describe the intervention done
3. Determine the outcome of the intervention
4. To describe common complication following intervention

4. Methodology

4.1 study area and study period

Study setting

The study was conducted at pediatric cardiology unit of TASH.

The pediatric cardiology service runs parallaly both in cardiac centre and pediatric department cardiology section of TASH.

Cardiac center Ethiopia was established in 2009 E.C on the ground of Tikur Anbesa specialized hospital Addis Ababa Ethiopia. It is a full operational unit taking care of patients with congenital heart disease and acquired valvular heart disease. It works hand in hand with TASH paediatric cardiac clinic.

Established in September 1974, TASH accommodate more than 720 beds. It is the largest teaching hospital in Ethiopia with more than 1700 medical and non-medical staff. The unit teach both undergraduates and postgraduate students. Pediatric cardiac clinic at TASH works 5 days per week in working hours.

Study period

The study was conducted from March 2017 to September 30, 2017.

4.2 Study design

A Retrospective chart review of cases.

4.3 source population and sampled population

All patient with COA who undergo surgical treatment and catheter intervention both locally and abroad having follow up at TASH paediatric cardiac clinic.

4.4 Inclusion criteria and exclusion criteria

Inclusion criteria: All Coarctaion of aorta patients undergo repair for native COA and with minimum of 6 month follow up post repair

Exclusion criteria

Exclusion criteria include those patients with repair done for re-coarctaion of aorta, patients with incomplete clinical data or lost charts and patients whose intervention done in less than 6 month at the time of the study period.

4.5 Sample size determination and sampling technique

All patients with repair of native COA repair in the study period were included since the number of patient is very low as from the calculated sample size.
Single population proportion formula for estimating sample size was used as follows:

$$n = \frac{z^2 p(1-p)}{w^2}$$

Where: n= is the minimum sample size required

p = is an estimate of the proportion of COA patients who underwent operation for COA.
w= Margin of error for sampling
Z- The standard normal value at (100%−α) confidence level.

Assumptions: Zα1/2= 95 % CI (=1.96), p=50 %(because we don't know the proportion), w=0.05. Therefore, n= $Z^2 * P * (1-p)$, n = $(1.96)^2$ x 0.5 x (1- 0.5)/ $(0.05)^2$, n =384

The estimated sample size based on the above assumption was 384. But because our sample size is less than half of the sample estimate we included all pediatric patients who undergo catheter and operatative intervention for coarctation of aorta in the above specified time.

4.6 Data collection instrument and technique

4.6.1 Questionnaire

A structured questionnaire was prepared data on socio demographic, clinical profile, type of surgical intervention and outcome of the intervention and possible complications was collected by the investigator.

4.6.2 Data collection quality assurance

The data from chart of a patient was collected using a structured format prepared for this purpose.

4.7 study variable

4.7.1 Dependent variable

Re-stenosis, systolic and diastolic blood pressure after surgery, immediate and late post intervention complication, aortic aneurysm, post intervention echocardigrahic gradient across descending aorta, death, lost from follow up.

4.7.2 Independent variable

Age, sex, age at diagnosis, age at initial repair, age at last visit, age at death

Type of COA, type of operation, associated intra-cardiac lesion, weight , length/ height and MUAC before and after repair, chromosomal abnormality, if any aortic valve anomalies, ,cardiomyopathy, moderate to severe left ventricular outflow tract obstruction, moderate to severe aortic valve regurgitation, ascending aortic dilatation.

4.8 Data management and quality assurance

Investigator collected all the data from the patient chart. The principal investigator monitor the data collection and does a quality check ,by counterchecking collected data against information in the chart in randomly selected patient.(manual proof reading)

4.9 Data analysis and interpretation

The data was entered and analysed using statically package for the social science (SPSS) version 20. The data was analysed using descriptive statistics and the statistical significance is tested using chi square. P value less than 0.05 will be considered significant.

4.10 Ethical consideration

The study proposal was approved by the department research review committee, of department of paediatrics and child Health College of health sciences, Addis Ababa University.

Personal identifiers like name of participants were not used to maintain confidentiality.

OPERATIONAL DEFINATION

Pressure gradient - difference between upper limb and lower limb blood pressure greater than 10 mmHg. (Between the brachial and the popliteal artery.)

Differential cyanosis – difference in oxygen saturation between the upper and the lower extremity

Hypertension – systolic or diastolic pressure above 95^{th} percentile for age and sex value

Pre-hypertension – systolic or diastolic blood pressure between 90^{th} and 95^{th} percentile for age sex value

Post repair residual coarctation of aorta - presence of aortic gradient immediately after repair

Re -coarctation - development of re-stenosis after an initially successful repair.

Intervention - refer to treatment of the lesion either by balloon angioplasty or surgical correction.

Lost from follow up- patient who is alive but not attaining follow up

Unknown status – patient current condition unknown

Outcome – outcome of the patient was measured with presence of immediate complication of repair, the post intervention blood pressure and the presence of late complication of repair like re-coarcation of the aorta.

Congestive heart failure: impairment of cardiac pumping or filling function evident by Tachypenia (respiratory rate higher than for age), sign of respiratory distress, presence of hepatomegaly, Increase JVP and peripheral edema.

Discrete Coarctation of aorta – Coarctation of aorta with no associated cardiac lesion

Simple coarctation of aorta – Coarcation of aorta with Patent ductus arteriosis

Complex coarctation – coarctation of aorta associated with congenital heart lesion other than patent ductus arteriosis.

4.11 Result disseminations

The result of this study will be presented in the department's research defence day and also submitted. As well as it will be presented to the scientific conferences in the college of health science, Addis Ababa University and Possibility through publication in scientific journal.

Result

A total of 39 patient with native COA repair reviewed 27(69.3%) were male and 12(30.8%) female. Majority of the patient were from Addis Ababa 24(61.5 %) followed by Oromiya 8(20.5%) and South nation and nationality 5(12.8) there rest 2(5.2%) form other nation.

The age at the time of diagnosis ranges from 12 days to 25 years, with mean of 4.68 ± 5.68 years. Age at the time of intervention was 3 months to 25 years, with mean of 6.89 ± 5.33 years. The latest intervention was done 6 months back and the earliest 13.5 years back. (see Table 1 and Table 2)

The most commonest presentation before intervention as shown in Figure 1 was high BP 22(56.4%) followed by exertional dysnea and incidental finding of murmur.

TABLE 1: Age distribution at time of intervention

Age at time of intervention	Number	Percentage
< l year	1	2.56
1-5 years	19	48.71
5- 18 years	17	43.59
>18 years	2	5.12
Total	39	100

Table 2: time between time of data collection and time of intervention

Time between survey and the time of intervetion	Number	Percentage
6 to 24 months	13	33.33%
25 to 60 months	15	38.46%
>25 months	11	28.2%
Total	39	100%

Three (7.69%) of the patient has family history cardiac disease. None of the patient has documented or diagnosed genetic abnormality on their chart.

The systolic BP of those patient ranges from 76 mmHg to 173 mmHg with mean of 128.2 ±19.25mmHg and the diastolic BP range from 33 mmHg to 108 mmHg with median73.5 ±19.33mmHg. Stage of BP of patients before repair of COA is shown in Table 3.

Figure 1: presenting compliant of patents

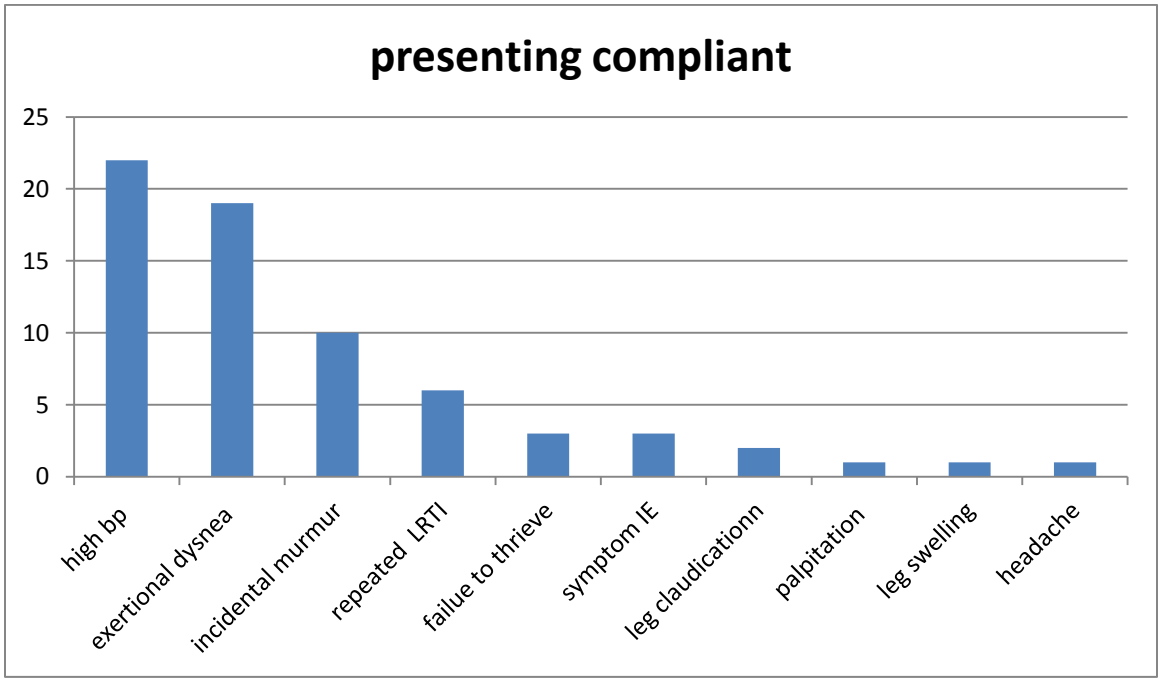


Table 3 : Blood pressure of patient before COA repair

Blood pressure	number	Percent
Stage II hypertension	22	56.41
Stage I hypertension	9	23.1
Pre-hypertensive	2	5.1
Normal blood pressure	5	12.8
No BP record	1	2.56
Total	39	100

Regarding the femoral pulse 13 (33.3%) have palpable femoral pulse and 18(46.2%) have weak femoral pulse and the rest 8(20.5%) have absent femoral pulse .21 (53.8%) had radio-femoral delay and 10(25.6%) had no radio –femoral delay.

The weight of patent range from 5 kg to 54 kg with mean weight 18.36 ±10.05 kg . Height range from 60 to 169 Cm with mean of 107.7 ± 27.18 Cm and BMI range from 12 to 19.40 with mean 14.9 ±1.9/m^2. The BMI for age and height for age of the patient is shown in table 4.

Coming to the auscultatory finding before intervention the most common cardiac finding in those patient was systolic click or murmur best heard at the LSB which was finding of 29 (74.3%)followed by murmur of MR heard in 7, murmur of AR in 4 patient and murmur of AS in 2 patient . P2 accentuation was documented in 3(7.69%). There were no auscultatory cardiac finding in 3(7.69%) of the patients.

Table 4: BMI for age and Height for age of patient before intervention.

	BMI	Height
Above +3 Z score	0	1(2.56%)
Between +2 and +3 Z score	1(2.56%)	1(2.56%)
Between +2 and – 2 Z score	26(66.7%)	20(51.3%)
Between – 2 and -3 Z score	6(15.4%)	8(20.5%)
Below – 3 Z score	2(5.1%)	5(12.8%)
Not available	4(10.3%)	4(10.3%)

None of the patient at time of evaluation before intervention fulfils the criteria for CHF. Based on the creatine level just before intervention only 1(2.7%) patient had raised creatine and 3 patients data about renal status was missing and the rest 35(97.2%) patient out 36 had normal creatine level. Coming to nervous system manifestation before intervention 1 (2.6%) had seizure disorder and 1 (2.6%) had history of focal neurologic deficit. Chest x ray finding of patient Is shown in table5.

Table5 :chest X-ray finding of patients

Chest X- ray finding	Number	Percent
Normal	6	15.3%
Cardiomegaly	30	76.9%
Increase pulmonary blood flow	14	35.9%
Rib notching	2	5.12%
Indentation of aora	1	2.6%

Electrocardiography of patient before the intervention shows Left ventricular hypertrophy in 26(66.67%) and Biventricular Hypertrophy In 3(7.6%) ,other abnormality in 6(15.4%) of patients and was normal in 6(15.4%) of those patients.

Coming to the type of the COA as shown in Figure 2, thirteen (33.3%) had discrete and 7(17.9 %) had simple COA with PDA, 15(38.5%) had complex COA, 2(5.1%) had isolate interrupted aorta and 2 (5.1%) had complex interrupted aorta.

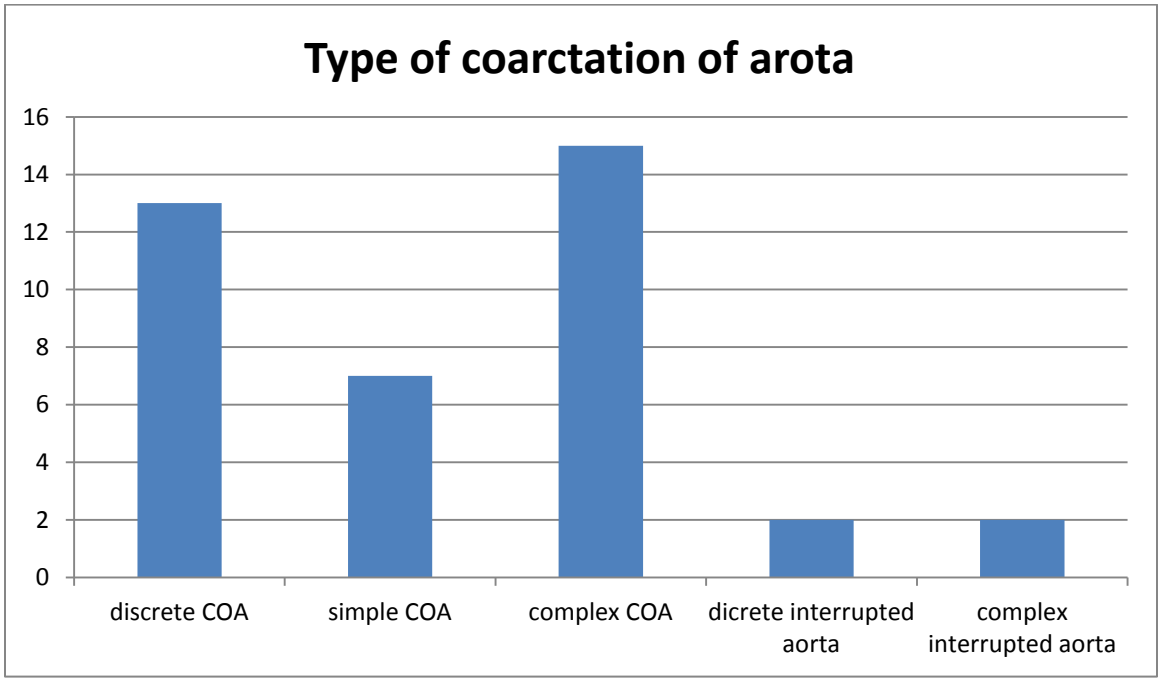
The most common associated lesion found was Bicuspid AV seen in 15 (38.5%) Followed by PDA 11(28.2%) see Table 6

Echocardiograhic evidence of pulmonary hypertension in 2 (5.1%) of the 39 patient, aortic aneurism 2(5.1%) of patients, evidence of dilated cardiomyopathy was seen in 4 (10.25%) of the patient.

Table 6: frequency of associated cardiac lesion

Associated cardiac lesion	Number	Percent
PDA	11	28.2%
Shone complex	9	23.07%
VSD	8	20.5%
Hypoplastic aortic arch and Ismuth	2	5.1%

Figure 2: type of coarctation of aorta



Cardiac catheterization for diagnosis purpose was done for only 1 patient but for purpose of intervention it was done for 22(56.4%) of 39 patients. After cardiac catheterization and echocardiography 7(17.9%) was diagnosed to have pulmonary hypertension.

Thirty (76.9%) of the 39 patient was on antihypertensive medication before surgery. ACE inhibitor was given to 24 out of 30 (80%) patients. ACE alone was given for 10 and combined with other drug in 14 of them. Following ACE inhibitor Loop diutetic given in 10 (33.3 %) of those on treatment followed by Beta blocker 3(10%). Other infrequently used drugs were calcium channel blocker , spirinolactone , digoxine.

The co-arcted segment range from complete obliteration to 6 mm with mean of 3.01 ± 1.80 mm and the peak pressure gradient range from 30 mmHg to 97mmHg with mean 63.4 ± 15.89 mmHg before intervention

Regarding the type of intervention 20 out of 39(51.3%) has non-surgical intervention out of which.4 with balloon angioplasty only (10.3%) and 16 (41%) balloon angioplasty with stent.

Surgical intervention was done for 19 of the patient (48.7%). End to end resection and anastomosis was done for 14(35.9%) patients followed by the patch arthroplasty in 5(12.8%) of the patients.

The immediate peak pressure gradient after repair ranges from nill to 60 mmHg with mean of 21.37 mmHg and SD 12.77.

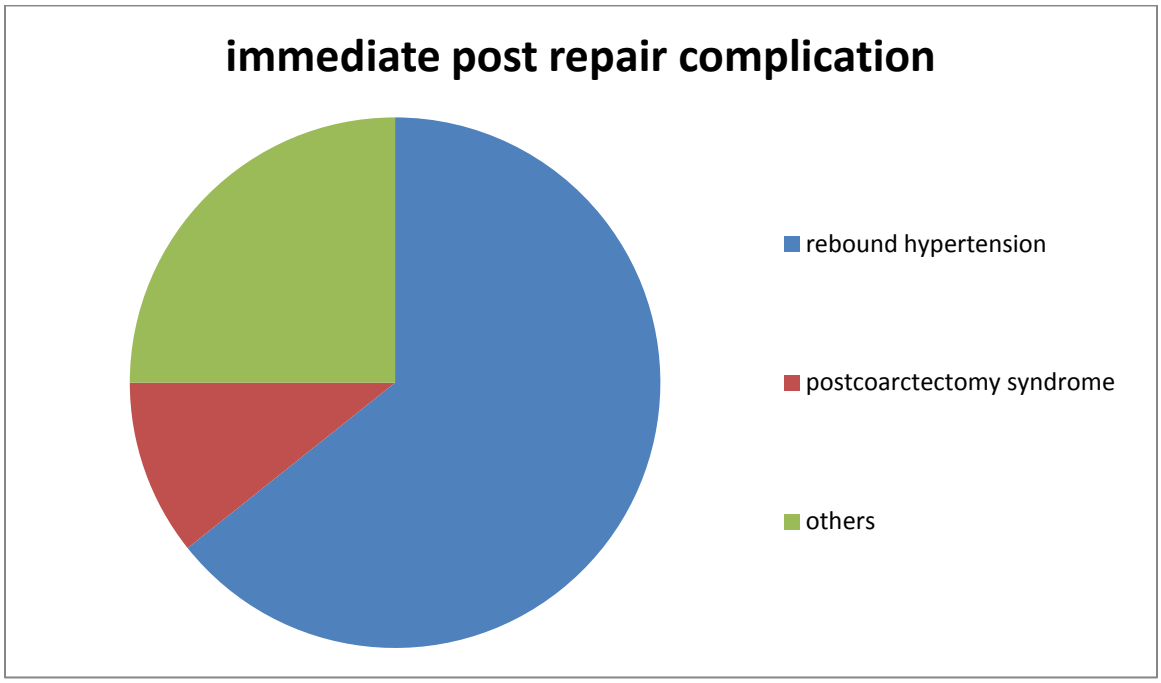
Coming to the immediate post intervention complication following repair 18(46.15%) of the 39 patients had no complication . 18 (46.15%) had rebound hypertension,3(7.69%) had postcoarctectomy syndrome,1(2.56%) had sign of ICH,1(2.56%) had pneumomedastinium,1(2.56%)had pericardial effusion,1(2.56%) had spinal cord injury with paralysis,1(2.56 %) had chylothorax,1 (2.56%) had lung atelectasis and 1(2.56%) had IE following repair. See Figure 3.
Blood pressure profile at 6 months after repair of COA is shown in table 7.

Regarding post intervention medication 17(43.6%) were off medication and the rest were on prescribed medication of which the frequent medication was ACE inhibitor taken 17(43.6%) of the patient ACE alone was taken by 11(28.6%) and in combination with other drug 6(15.38%). Following ACE beta blocker and diuretic were the frequent drug each with 4(15.38%) of the 39 patients.

Table 7 : Blood pressure 6 month after the repair of COA.

BP after intervention	Number	Percent
Stage II hypertension	2	5.1
Stage I hypertension	12	30.8
Pre- hypertension	5	12.8
Normal BP	17	43.6
No record	3	7.7
Total	39	100

Graph 3: Frequency of immediate post intervention complication of COA repair.



Thirty five of the patient had echocardiography result at 6 month after intervention with the finding of normal echocardiographic finding in 15(42.8%), residual COA 7(20%), evidence of LVH in 4(11.43%) and Aneurism in 1(2.85%) . Finding associated with the associated cardiac problem was seen in 7(20%) of the patients.

The mean SBP decreased 128.2± 19.2mmHg to 111.5 ±12.13 mmHg , DBP form 73.5 ± 19.3 to 69.2 ± 12.13 and that of peak pressure gradient across desending aorta form 63.4 ±15.89 to 21.37 to 12.7 following the repair. Out of the 4 who had balloon angioplasty 2 had recoarctation on the 6 month echocardiography and only 1 out 14 patient with balloon angioplasty with stent and in 3 of the 13 with end to end anastomosis and 2 of the 5 with patch arthroplasty .

Rebound hypertension shows stastically significant association with the type of intervention with P value of (0.0104) associated with surgical intervention specially with patch arthroplasty.

At time of the study only 26(66.7%) were on follow up ,7(17.9%) lost from follow up but were alive,1 (2.6%) died and for 5 (12.8%) current status of the patient was unknown. Three of the patient had one time re intervention for diagnosis of re coarctation.

Table 8 : cross tabulation between type of intervention and rebound hypertension

	Rebound hypertension		Total	P- value = 0.0104
	Yes	no		
Balloon angioplasty	5	15	20	
Surgical intervention	13	6	19	
Total	18	21	39	

Table 9 : cross tabulation between type of intervention and re- coarctation

		Residual COA		Total	P value = 0.05
		Yes	NO		
type of intervention done	balloon angioplasty	2	2	4	
	balloon angioplasty with stent	1	13	14	
	resection and end to end anastomosis	3	10	13	
	patch arthroplasty	2	3	5	
Total		8	28	36	

Discussion

Coarcation of aorta repair has been shown to be safe and definitive treatment modality in those patient with COA. Though morbidity and mortality of the procedure is not as such prounced as its benefit it is not un common to see immediate and long term complication following the repair of COA. This warrant the need of life long follow up in those patient even after repair.

In this study where 39 primary repair of coarctation of aorta review over a period of 14 years , the age at the time of intervention range from 3 months to 25 years with the mean age being 6.9 ± 5.3 years . The mean age is lower to that of the USA study which is 17 ±13.6 years and is comparable with that of the Czech republic study which was 7.2 ±8.9 years .(34,35)

The incidence of hypertension before the repair in our study was 82.1% which is comparable with the USA study which was 82.5%. But post repair rate of hypertension was 40% in our study which is higher when we compare it with the same study in USA study, which shows a drop of hypertension to 33.5 % after repair of coarctation of aorta . This may be explain that though the total number of patient with hypertension was not significantly drop as in the USA study, the stage of hypertension was decrease from the pre intervention time which shows 56.4% of stage 2 hypertension Compare to 6.25% in those with post intervention time . And also the mean SBP and DBP significantly decrease from the the pre intervention value.(33)

Type of coarctation in our study was complex COA in 38.5 % ,simple COA 17.9%,discret in 33.3% interrupted in 10.3% of the patients. The most associated cardiac lesion were PDA followed by shone comlex and VSD. There was no significant association with the type of coarctation and the post op complication in this study.

The type of intervention in our study is mostly non- surgical (balloon angioplasty with or with out stent) which account to 51.3%. The most commonest intervention in our study was balloon angioplasty with stent(41%) followed by end to end anastomosis 35.9%, patch arthorplasty 12.8% and lastly balloon angioplasty without stent 10.3%. According to this result non operative repair was the most practice method in our study which is unlike all the other studies. In USA, Czech republic , Switzerland ,Indian and Nigerian studies where the most practice method of a repair was end to end anastomosis and with the Iranian study where patch artroplasty was commonest in that study. The reason for this couldn't be explained since majority of the intervention were mission based and based on the practice of different countries. (35,37,38,39,40)

In this study the early mortality form the intervention was 0% which comparable to the swittzerland which is also 0%(37). But very low when compare to Indian study which mortality rate of 9.8% , Nigerian experience which is 25% and the USA study (12.2 % -1.4%). This result could be explained that all the repair done in our study are elective and no emergency procedure was done as such intervention are mission based and are not all year round which exclude very critical patient who would have less chance of survival after intervention. And none of our patent where having sign of CHF and where on cardiorespiratory support before intervention.(35,39,40)

None of the immediate complication was seen in the 46.5% of the patients. And out of the immediate complication rebound hypertension take the lion share which complicate 46.5% of patient's immediate course. This rebound hypertension was seen mostly with that of surgical intervention which shows significant association with P value of 0.0104. Rebound hypertension complicate 5 of the 20 patient with non -operative management (25%) ,13 of the 19 of the operative intervention(68.4%) and all of the patient with patch arthroplasty. This is result is In consistent with the comparative study between the operative VS non- operative which shows all of the operative repair complicate with rebound hypertension as compare to the catheter intervention with P value <0.05 (47).

The incidence of rebound hypertension which is 46.5 % is high when compare to the Nigerian study which occurs in only 3 of 16 patient (18.75 %) . This incidence is comparable to the study in Milton S. Hershey medical center which showed that 56% of 25 patents complicate with paradoxical hypertension following repair of COA (46). None of our patient has hoarseness of sound ,comparing it form the Nigerian study which 4(25%) had hoarseness of sound this might be explained by majority of our patient had non operative type of intervention which is associated with lesser risk of recurrent laryngeal nerve injury. Post- operative haemorrhage, chylothorax graft occlusion and wound dehiscence occurs more in the Nigerian study than our study. Even the mortality which is 4(25%) is very high compare to our study even if the number of patient of the study is too small to generalize. The decreased morbidity and mortality in our study may be explained by the above mentioned reason that our intervention is for those elective patient with no emergency condition. In this Nigerian study it was not possible to compare the long term complication with our set up. (40)

Though the long term complication of the patient in our study was not further study , the 6 month post intervention profile shows residual coarctation in 7 of 35 patient with echocardiography at that time which account for 20% which is a very significant. Even though this study is not a cohort study to compare it with those of USA study and those patient profile was taken only for the first 6 month of post intervention period, this percentage of patient which is 20% is higher than the USA figure which was 7.6%(35). But further study is needed to know the exact figure as time of follow up increases.

There was no significant stastical association between the type of coarctation of aorta and type of intervention and reocarction in the follow up Echocardiography. This result is consistent with the USA study.(35) Only one patient echocardiography show aortic aneurism at 6 month post intervention and type of intervention for that patient was balloon angioplasty with stent. This result is difficult to compare to that of other research with aortic complication of repair COA. This shows if subsequent Echocardiography done for those patient long term aortic complication of repair of COA are not Uncommon to see.

In our study, though not the scope of the study, only 26 (66.7%) are in follow up , 7(17.9%) lost from follow up but alive , 5(12.8%) status unknown and one late death at the time of the study .This follow up rate is very low and many drop out from the follow up clinic making it difficult to study long term complication in those patients further.

Conclusion

Repair of COA significantly decrease BP and pressure cross the Dis Ao. The most commonest type of COA in this study was complex COA followed by discrete COA. The most common mode of intervention was ballon agioplaty with stent followed by resection and end to end anastomosis. Rebound hypertension followed by postcoarctectomy syndrome, were the most common immediate complication following repair of COA. There was no early death following intervention. There was no significant association between type of intervention and risk of coartaiton as well as the age of intervention. Significant association was found between surgical intervention of COA repair and that of incidence of rebound hypertension. Recoarction of COA complicates significant number of post intervention patient form the long complication. Only 66.7% of patient was still on follow up at the time of the study. Significant number patients drop out form the follow up.

Limitation of the study

The number of patient on which the study was conducted was very small to have strong recommendation
Incomplete data of patients make analysis more difficult.
Significant drop out from the follow up clinic makes it difficult to study on long term out come of the patients.
Intervention at CCE are mission based with professionals came from different countries. So practice depends on where they came .

Recommendation

Scheduled follow up and echocardiographic evaluation at flow up clinic is needed
Strong advice should be given to those patients with COA repair about the need of follow up and the importance of follow up should be explained so significant drop out from follow up decrease.
Why this significant number of patient drop out from follow up need further study.

Batching and Cohorting of patients for future survival and morbidity study is needed

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Annex I

QUESTIONNARI

1 .Demographic Data

Name: _____ Age _____

Sex: M □ F □ Card no_____ Tel no_____

Address _____ Woreda_____ Kebele_____ house no_____

2. Age at time of diagnosis: _____

3. Age at time of surgery _____

4. Duration of follow up after surgery _____

5. Presenting symptom

- Headache yes ☐ No ☐
- Palpitation Yes ☐ No ☐
- Pain or weakness of lower extremity up on walking Yes ☐ No ☐
- Exertional dysnea Yes ☐ No ☐
- Leg swelling Yes ☐ No ☐
- Decrease in mental status Yes ☐ No ☐
- Hem paresis Yes ☐ No ☐
- Fever and symptom suggestive of endarteritis Yes ☐ No ☐
- Chest pain Yes ☐ No ☐
- No symptom incidental finding during evaluation for other compliant ☐
- Family history of CHD Yes ☐ No ☐
- Any known genetic syndrome(Turner syndrome) Yes ☐ No ☐
- Other _____

6 .Pre op physical examination

Vital sign: PR=_____

Peripheral artery pulses

- Radial: Absent ☐ present ☐ Not documented ☐

- Cubital: Absent ☐ present ☐ Not documented ☐
- Femoral Absent ☐ present ☐ Not documented ☐
- Popliteal Absent ☐ present ☐ Not documented ☐
- Pedis Dorsalis Absent ☐ present ☐ Not documented ☐

RR = _____ Temperature= _____

O2 saturation Upper extremity = _____ Lower extremity = _____

Blood pressure:

UL= _____ UR= _____ LL= _____ LR= _____

Blood pressure gradient = _____ BP percentile_____ systolic ____ Diastolic_____

Anthropometry :

Weight in kg _____ Height in cm:_____ MUAC _____

BMA: _____ BSA:_____

HEENT- dysmoriphic feature Absent ☐ present ☐

Respiratory system

- On respiratory distress yes ☐ No ☐
- Sign of pulmonary congestion(basal crackles) Yes ☐ No ☐
- Continuous murmur and thrill over the lung field of the co-lateral Yes ☐ NO ☐

Cardiovascular system

- Differential cyanosis Yes ☐ No ☐ Not documented ☐
- Pulse: bounding pulse over the upper body yes ☐ No ☐not documented ☐
  - Pulse discrepancy b/n upper left and right extremity Yes☐ NO ☐ not documented ☐
  - Pulse discrepancy b/n upper and lower extremity Yes ☐No☐not documented ☐
  - Radio-femoral delay: Yes☐ NO ☐ Not documented ☐
  - JVP raised: Yes☐ NO ☐ Not documented ☐

Precordial examination

- Apical heave Yes ☐ No ☐ Not documented ☐
- Left sternal border heave Yes ☐ No ☐ Not documented ☐
- P2 accentuation Yes ☐ No ☐
- Systolic thrill over the sternal notch Yes ☐ No ☐
- Murmur of AS Yes ☐ No ☐
- Systolic ejection click over the left middle sternal border Yes ☐ No ☐
- Continuous murmur of collateral Yes ☐ No ☐

Abdomen – Hepatomegally Yes ☐ No ☐

Positive sign of fluids collection: Yes ☐ No ☐

Msk and intugumentary system: Oedema yes ☐ No ☐

Nervous system

- Decrease in mental status: Yes ☐ NO ☐
- Seizure: Yes ☐ No ☐
- Focal neurologic deficit: Yes ☐ No ☐

7. Investigation

Haematology HCT_____ Hgb _____ PLT _____

RFT: BUN _____ Creatine: _____

LFT: SGOT_____ SGPT _____ AKP _____

Chest x ray: Normal ☐ Cardiomegally ☐

Increase pulmonary flow ☐ Indentation of aorta ☐

Rib notching ☐

ECG: LV hypertrophy ☐ RV hypertrophy ☐

Echocardiography:

Associated intra cardiac lesion ☐ If yes specify_____

Ascd AO diameter (mm)_____

Desc AO diameter (mm)_____

Coarcted segment diameter (mm) _____

DSAoPressure gradient before angioplasty by echo/Catheter(mmHg) _____

DSAo pressure gradient after surgical repair echo/ Catheter(mmHg) _____

Cardiac catheterization: Done ☐ **not done** ☐

If done, finding _____

8. Pre op medical treatment

Antihypertensive treatment Yes ☐ No ☐

If yes, B blocker ☐ Diuretics ☐ ACE inhibitor ☐

Other specify _____

Treatment for Congestive heart failure Yes ☐ No ☐

9. Type of operation

Operation Note:

Site of operation-

Type of surgery: Resection and end to end ananstomosis ☐

Prostetic patch arthroplasty ☐

Subclavian flap arthroplasty ☐

Bypass graft ☐

Percutanious ballon angioplasty ☐

9. Anticipated complication:

Paradoxical hypertension yes ☐ No ☐

If yes, treated with- ACE Inhibitor ☐ B Blocker ☐

Other specify_____

Post coarctectomy syndrome yes ☐ No ☐

Hoarseness of sound Yes ☐ No ☐

Ipsilateral diaphragmatic paralysis(pherenic nerve injury) Yes ☐ No ☐

Paralysis of lower extremity (Spinal cord injury) **Yes** ☐ **No** ☐

Hemodynamic collapse 2ry to hemmoragge **Yes** ☐ **No** ☐

Residual hypertension: Resting hypertension ☐

Exercise hypertension ☐

BP: UL =_____ UR = _____ LU= _____ LU= _____

Pressure gradient = _____

If, hypertensive treatment received

B Blocker ☐ ACE ☐ Other specify _____

Symptom and sign suggestive of intracranial haemorrhage Yes ☐ NO ☐

Symptom and sign suggestive of endarteritis yes ☐ No ☐

Proven endarteritis: yes ☐ No ☐

LSCA Steal syndrome Yes ☐ No ☐

Aortic aneurism: yes ☐ No ☐

Others _____

10. Follow up profile at 1 yr of post operation

BP= UR _____ UL _____ LL _____ LU_____

Pressure gradient:_____ persentile of BP _____ systolic _____ diastolic

If still hypertensive, treatment received

ACE ☐ B Blocker ☐ other Specify _____

Post op echocardiography

Residual coarctation of aorta ☐ Left ventricular hypertrophy ☐

Aortic aneurism ☐ Discend OA diameter r _____

Peak pressure across the Diss aorta _____