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Coarctation of the Aorta:

Experience at the University College Hospital, Ibadan

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Summary

Adegboye VO, Ogunkunle O, Omokhodion SI, Brimmo AI, Adebo OA, Ogunseyinde OO, Obajimi MO. Coarctation of the Aorta: Experience at the University College Hospital, Ibadan. *Nigerian Journal of Paediatrics* **2002;29:27.** Between May 1977 and June 1998, 697 patients with congenital heart diseases were admitted to the cardiothoracic surgical unit (CTSU) at the University College Hospital, Ibadan. Eighteen (2.6 per cent) of the patients with 19 coarctations of the aorta (CoA) were retrospectively studied. The age range of all the patients with CoA was 18 days to 30 years (mean 7.2±8.2 years), but for the 15 patients who underwent surgery, it was one month to 30 years (mean 8.6±8.3 years). Three patients died preoperatively of congenital cardiac anomalies associated with infantile CoA. There were 16 thoracic and two abdominal CoA, while one patient had recurrent CoA. Resection and end-to-end anastomosis was performed in four patients, dacron tube interposition graft in three, and dacron patch graft in four patients. Other procedures were employed in five patients. Operative mortality was 25 per cent. Operative deaths occurred in two infants with isolated CoA, a neonate who had associated pulmonary hypertension and a 17-year old who had surgery for re-coarctation. Complications of surgery included post-operative haemorrhage in two patients, intra-operative hemorrhage in one and hoarseness of the voice in four patients. Paradoxical hypertension occurred in three patients, graft occlusion and wound dehiscence occurred together in one patient and two patients had chylothorax. It is concluded that CoA is a surgically correctable congenital anomaly which is probably less frequently diagnosed locally.

Introduction

COARCTATION of the aorta (CoA) is a congenital cardiovascular defect that shortens life if untreated but can be corrected to render the patient functionally normal.[1] The natural history of untreated CoA depends on the age at presentation and associated cardiac anomalies.[2] Symptomatic infants have a high mortality which depends on the severity of the CoA and the severity of the associated cardiac defects.[3-5] Some symptomatic children with isolated CoA survive for a while on medical treatment alone and even patients with isolated interrupted aortic arch (IAA) have been known to present in later childhood or in adult life.[2] Those that survive untreated until adulthood have a shorter life span compared to normal population.[6,7] The causes of death in CoA include bacterial endocarditis, aortic rupture, intracranial haemorrhage, heart failure or valvar heart disease.[8] Early identification and management of CoA may minimize the incidence of these complications.

The prevalence of CoA at the University College Hospital, Ibadan based on earlier studies, was 6.7 per cent of cases of congenital heart diseases in a population where congenital heart diseases had an incidence of 3.5 per 1000 births.[9] This incidence is similar to that of 6.5 per cent quoted in literature.[8, 10] However, several reports have documented a low incidence of CoA in black communities in predominantly white societies.[11, 12] This report describes our experience with CoA in a black African population.

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Patients and Methods

In the cardiothoracic surgical unit (CTSU) of the University College Hospital, Ibadan, all patients seen and treated between May 1977 and June 1998, were registered. The case notes of the patients who had CoA were obtained and various data were extracted. These included the ages of the patients, their sex, the types of CoA, the associated congenital heart or other diseases and the type of corrective surgery employed. The preoperative evaluations included plain chest X-rays, aortography and 2D echocardiography. Coagulation profiles, liver and renal function tests were determined prior to surgical intervention. No karyotyping was done. Other information extracted from the case notes included cross clamp time, the number of collaterals ligated and the complications of the surgery for CoA.

Results

During the period of study, 697 patients with congenital heart diseases were admitted to the Unit for palliative or curative procedures; 18 (2.6 per cent) of these had CoA. The age range of the patients with CoA in this series was 18 days to 30 years (mean 7.2±8.2 years); for the 15 patients who underwent surgery, it was one month to 30 years (mean 8.6 ± 8.3 years). The male:female ratio was 1.4:1. Table I summarizes the clinical findings, corrective procedures and outcome among these patients.

Diagnosis was made from symptoms and signs in 15 patients while in three patients, the clinical signs of CoA were discovered during hospital visit for unrelated reasons. The major clinical features are shown in Table II. Diagnosis was made in all cases by the difference in arterial pulsation between the upper and lower extremities and cardiomegaly was an invariable finding in all the patients. Congestive cardiac failure was present in three neonates with associated congenital heart defects and three other patients with isolated CoA. Plain chest x-ray film was suggestive of the location of the CoA in five patients (aged nine to 12 years), who had bilateral upper rib notching (Fig. 1), and "3" sign in the left parasternal area. Four patients had aortography (Fig. 2), and four others had only 2D echocardiography. In the latter group, an intrathoracic CoA was not found at surgery in one patient who clinically had radio-femoral arterial pulse dissociation. This patient was one of the two with abdominal CoA (one was suprarenal, while the intraabdominal level of the other was undetermined). There were 17 thoracic CoA. The three patients who died preoperatively had infantile type of CoA; the types of CoA in the 15 patients who underwent surgery are as shown in Table I.

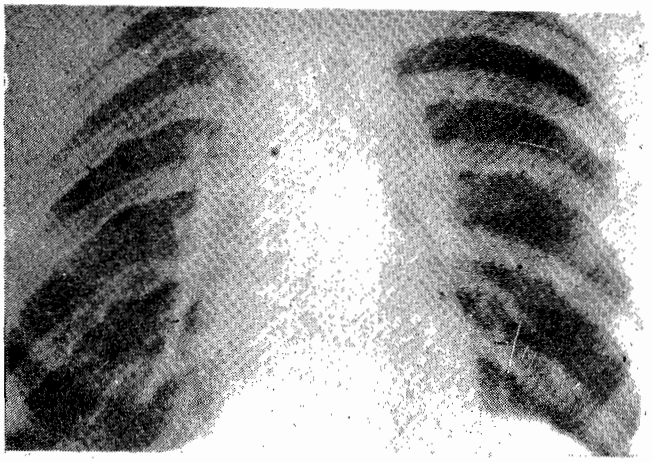

Fig 1. Posteroanterior (PA) view of a chest radiograph showing posterior, bilateral notching of the inferior edges of the ribs.

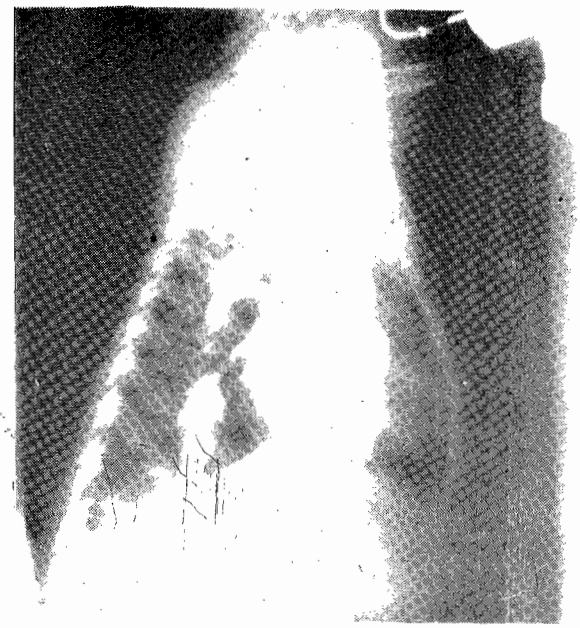

Fig 2. Lateral view of an angiogram showing narrowing of the descending aorta and poststenotic dilatation.

Aortic cross clamp time ranged from 25 to 45 minutes (mean 36.1±5.3 minutes), and one to four collaterals were divided during the corrective procedures. Four patients aged between one month and two years with infantile type of CoA had coarctectomy and end-to-end anastomosis (Fig. 3a and 3b). Three patients had coarctectomy and dacron tube interposition; one of these had IAA. Four other patients had coarctectomy and dacron patch aortoplasty. Other procedures employed in the care of CoA are summarized in Table I. All operative survivors

Table I

Clinical Findings, Corrective Procedures and Outcome among 18 Patients with 19 Coarctations of the Aorta

S/No.	Age	Type of Coarctation	Associated Congenital Heart or other diseases	Procedure	Cross Clam Time (Minutes)	No of Intercostals Divided	Outcome
1	18days	Infantile	Cortriloculare	-	-	-	Died preoperatively
2	21days	Infantile	Ventricular septal defect	-	-	-	Died preoperatively
3	1month	Infantile	Subaortic stenosis	-	-	-	Died preoperatively
4	1month	Infantile	Pulmonary hypertension	Patch aortoplasty and pulmonary banding	25	1	Died on the 3rd post operative day from heart failure
5	1month	Infantile	-	End-to-end anastomosis	32	2	Satisfactory
6	2 months	Infantile	-	End-to-end anastomosis	37	2	Satisfactory
7	1 year	Infantile	-	End-to-end anastomosis	45	3	Died (postoperative haemorrhage)
8	2 years	Infantile	-	End-to-end anastomosis	40	2	Died (postoperative haemorrhage)
9	12 years	Infantile	-	Subclavian aortoplasty	43	-	Satisfactory (paradoxical hypertension)
10	12 years	Infantile	-	Interposition graft	39	3	Satisfactory (paradoxical hypertension)
11	18 years	Infantile	Takayasu aortitis	Interposition graft and PDA ligation	37	4	Satisfactory (paradoxical hypertension)
12	30 years	Aortic interruption	Patent ductus arteriosus (PDA)	Interposition graft and PDA ligation	36	-	Satisfactory
13	*7 years	Adult	-	Patch graft	35	1	Satisfactory
14	9 years	Adult	-	Patch graft	28	1	Satisfactory
15	11 years	Adult	-	Patch graft	32	2	Satisfactory
16	12 years	Adult	-	Patch graft	30	-	Satisfactory (paradoxical hypertension)
17	17 years	Adult {had first operation at age 7yrs (patch graft)}	Re-coarctation	Re-operation	-	-	Died (intraoperative haemorrhage)
18	5 months	Intraabdominal	-	Negative exploratory thoracotomy	40	-	Persistent arm to leg systolic pressure gradient
19	10 years	Intraabdominal suprarenal membrane	-	Excision of abdominal aortic membrane	43	-	Satisfactory

*Presented at age 17 years (patient No. 17) with re-coarctation

Fig 3a. Intraoperative photograph of coarctation of the aorta showing the left subclavian artery encircled with umbilical tape, and post-stenotic dilatation of the descending aorta.

had satisfactory postoperative status within the four months to 10 years of their follow-up. The operative mortality was 25 per cent; death resulted from intraoperative haemorrhage in one patient, postoperative haemorrhage in two patients and refractory right sided heart failure in one patient. Other complications of surgery included hoarseness of the voice in four patients, paradoxical hypertension in three patients which was appropriately managed with reserpine in one patient and nitroprusside in the others. The patient with aortic interruption developed graft occlusion, which warranted a re-operation; he also had wound infection and dehiscence, which were treated by wound debridement and broad spectrum antibiotics. This patient had a successful outcome. Two patients developed chylothorax, which cleared up without specific treatment. No neurological defects or post-coarctectomy syndromes were observed.

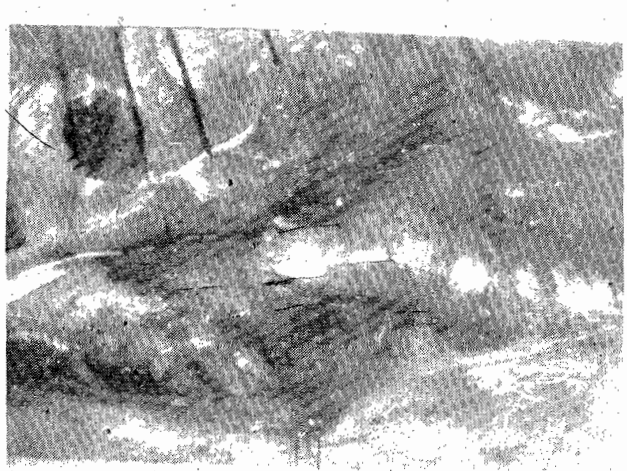

Fig 3b. Intraoperative photograph showing the descending aorta after coarctectomy and end-to-end anastomosis.

Table II

Major Clinical Features in 18 Patients with Coarctation of the Aorta

Clinical Feature	*Number of Patients*
Symptoms	
Throbbing headache	9
Weakness in the lower limbs	8
Claudication in the lower limbs	6
Chest pain	4
Intermittent dizziness	3
Signs	
Radio-femoral pulse dissociation	19
Systolic and/or diastolic murmur	11
Cold lower extremities	9
Palpable arterial chest collaterals	8
Congestive heart failure	6

Discussion

Cardiovascular disease in Africa is dominated by hypertension.[13] Coarctation of the aorta is one of the two common vascular anomalies responsible for hypertension, the other being renal artery stenosis.[14] There are currently three postulated causes of the constriction or the mechanical obstruction in CoA.[15] The concept of ductal tissue spreading into the aorta and subsequently causing constriction after birth.[16-18] Theories of flow as a cause of CoA are supported by the fact that intracardiac anomalies with decreased aortic flow patterns have increased incidence of arch anomalies. Intracardiac anomalies with increased aortic flow, because of decreased pulmonary flow are not associated with CoA.[19-22] The majority of CoA or other arch anomalies which are located preductally may be due to neural crest migratory problems.[15, 23] The pathogenesis of the hypertension in CoA may be more complicated than simple mechanical obstruction.[24] A generalized vasoconstrictor mechanism is likely to be

involved, which may either be related to renin–angiotensin system or to sympathetic nervous activity.[25]

It is either that coarctation of the aorta is uncommon among blacks or it is underdiagnosed in our environment. The fact that 18 cases of coarctation of the aorta were seen over a twenty-one year period in a tertiary institution requires further investigation. CoA constituted about five per cent of all cases of congenital heart diseases operated upon at the University College Hospital between 1968 and 1970.[26] A similar frequency (four per cent) was obtained for the period between 1975 and 1977 at the same institution.[27,28] In a recent collection of patients with congenital heart disease (CHD) who had corrective surgery under "Save a child's heart" programme, only one patient out of 91 (1.1 per cent) had CoA (personal communication). It does seem as if fewer patients get diagnosed and fewer patients get to surgery in our catchment area. However, workers in white communities have suggested a low incidence of CoA among blacks in their communities.[11]

The hallmarks of early diagnosis of CoA include a high index of suspicion and alertness to symptoms and signs. Various degree of severity of CoA starts to manifest at the time the pulmonary vascular resistance starts to decrease which also coincides with the closure of the ductus arteriosus.[2, 24] Because of the older age of our patients, more than 70 per cent were diagnosed on the basis of clinical features, supplemented by a simple investigation such as plain chest radiograph. However, symptomatic infants pose the greatest challenges to diagnosis and management in most peripheral hospitals. Because of lack of widespread use of echocardiography with doppler studies, evaluation of these symptomatic infants have been incomplete. This may be the major reason why there were few cases. The mortality among these patients depends on the severity of the coarctation and the nature of any associated congenital cardiac defects.[3-5] Many of such patients probably die before referral and without evaluation. However, the salvage rate in the presence of congenital cardiac defect is poor universally,[2,21] hence the preoperative mortality of 16.7 per cent in our series is not unique.

When there are clinical features suggestive of CoA, the actual localization of site and type of obstruction should be mapped out by angiocardiography or aortography.[2] Newer, non-invasive methods of imaging such as two dimensional and doppler echocardiography demonstrate the site of obstruction and suggest or exclude associated anomalies; they also provide an estimate of the arterial pressure gradient.[11, 29,30] However, difficulty in localizing CoA using these non-invasive techniques could still occur,[29] as we experienced in one of our patients. The current thinking is that echocardiography is adequate for definitive diagnosis of CoA in neonates and infants, but other techniques such as cardiac catheterization[18] or magnetic resonance imaging may clarify issues in older children[32] when echocardiographic findings are inconclusive.

The presence of coarctation is generally sufficient indication for surgical correction.[6, 33] The major questions are the timing and method of repair.[1, 2,6,11] The timing for elective repair of coarctation of the aorta is perhaps the most important determinant of surgical results.[1-6,11] Repairs done in late childhood or adulthood, and repairs in infancy using the classical method of resection and end-to-end anastomosis have specific attendant complications. The former provides relief of some symptoms, but there is an increased incidence of persistent hypertension and its associated morbidity.[34, 35] Three out of the four patients who had paradoxical hypertension were 12 years old and the fourth patient was 18 years old. The repair in infancy has a high incidence (up to 60 per cent) of residual or recurrent stenosis.[2,35] However, re-coarctation was less frequent in our series because most repairs were carried out in the older age group. The current trend is for elective repair between ages one and six years in order to decrease the incidence of re-coarctation and minimize complications of late repair.[2,4] Some workers however, prefer repair at the time of diagnosis in symptomatic and asymptomatic patients.[3,35] This latter approach was the only option we had; the mean age of our cases who underwent surgery was about eight years.

Surgical options depend on age at surgery, length of coarctation and surgical techniques that are considered appropriate.[1,36,37] Coarctectomy and end-to-end anastomosis though easier in younger children is more permanently beneficial if the aorta is allowed to approach its adult size. With increasing age however, the aorta is more sclerotic, less elastic, and more difficult to approximate and suture. Besides, aneurysms of the intercostal arteries are usually found in older patients.[1] Longer segments of coarctation need either aortic replacement or subclavian flap aortoplasty. Our options depended on the most feasible technique at surgery based on these principles. For children, some points must be borne in mind: (i) all excess tissue around the coarctation site must be carefully dissected from the aorta; (ii) as much of the aorta as possible should be resected for the anastomosis to be well away from any residual constrictive material associated with the ductus; and very careful approximation must be done with fine interrupted sutures.[11,36] When re-coarctation occurs, balloon angioplasty for recurrent coarctation has given the most favourable results and seems equally successful regardless of prior procedure.[38] The operation is associated with absence of mortality, low morbidity, effective reduction of the gradient, restenosis of only 10–13 per cent and aneurysm formation of only 2–4 per cent.[38-40] However, balloon angioplasty does not seem appropriate for neonates and infants, and remains questionable for

children with unoperated CoA.[38] Re-operations for re-coarctation have a high mortality rate.[39] The operative mortality in our series of 25 per cent comes close to the range of 0-24 per cent of a collected series totalling 1,189 patients.[38]

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