COARCTION OF THE AORTA *

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INTRODUCTION

Systemic Hypertension in the majority of cases affects the arterial tree in all the four extremities more or less uniformly; with pressure in the lower extremities higher than the upper extremities. However there are conditions leading to systemic arterial hypertension with differential involvement of the systemic arteries.

Two common varieties exist. Hypertension in the upper extremities and hypertension in the lower extremities.

The clinical example of the former is coarctation of the aorta and the example of the later is Takayasu's arteritis.

Coarctation of the aorta is a correctable lesion. It occurs in about 2—8% of all cases of Congenital Heart disease in the west.

Clinical diagnosis of coarctation of the aorta requires the examination of the upper and lower limb arterial pulses and pressures.23 cases were diagnosed in the last 7 years in the Haemodynamics labs of NICVD by the authors.

The purpose of this study is to document the clinical presentation, physical findings, haemodynamic angiographic data and surgical results in these patients.

METHODS AND MATERIALS

All patients referred to the authors with the diagnosis of systemic hypertension since May 1975 were included in the study. Twenty three cases were discovered to have delayed or absent femoral pulses and were clinically diagnosed. All patients had detailed history and physical examination. B.P. was checked in both arms and (R) radial and (R) femoral arteries were palpated simultaneously. 12 lead E.C.G., standard chest X-ray P.A. view and RAO LAO views with Barium Swallow were done.

Twenty three cases were discovered to have delayed or absent femoral pulses and were clinically diagnosed as coarctation of the aorta.

Cardiac catheterization was performed in all patients.

Right femoral artery puncture percutaneously in 15 patients; (R) Brachial artery cut down in 5 patients and Lt. brachial artery cut down in 3 patients.

Right Heart Cath. was done in 6 patients. Aortic root injection of 76% urogaphin was made and filming by 35 mm cine-camera was done in the L.A.O. view, Another injection was given just above the coarctation in AP view. Pressures were recorded on Multichannel physiological or E for M VR-12 recorder with the transducer at the mid chest level. All patients were presented for surgery. The results were as detailed in Tables I to V.

DISCUSSION

Coarctation of the aorta forms somewhere between 4-8% of patients with Congenital Heart Disease (1,2,3,) It is also mentioned that it forms less than 0.5% of the hypertensive population (4). Studies on Congenital Heart

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Disease from J.P.M.C. showed only one case among 190 patients with Congenital Heart C.H.D. though hypertension reported from Pakistan is about the same as in the Western Countries (6). The rarity of coarctation may either be due to its true rarity or because of low index of suspicion many cases may be treated in other groups. Table No.1 shows this points very well, only <10% cases were picked up by the refering physicians. The rest were categorized with Rheumatic Heart Disease (43%), essential hypertension (30%) and C.H.D. (9%). All patients having hypertension esp. if they are below 40 years must get their B.P. checked in their lower extremties. Patients with coarctation of the aorta have leg B.P. either equal or less than the upper extremities.

Table I: Distribution of cases according to referral diagnosis.

Coarctation of the aorta	...	2
Essential hypertension	...	8
Mitral insufficency	...	7
Aortic Insufficency/Aortic Stenosis	...	3
P.D.A.	...	2
Renal artery stenosis	...	1
Total	...	23

Simultaneous palpation of (R) radial and (R) femoral pulses also gives a clue. Normally the (R) femoral is felt ahead of the (R) radial but in coarctation the Rt. Radial is felt ahead of the (R) femoral.

Absent femoral pulses coupled with a bruit over the epigastriuum is usually due to abdominal coarctation.

The high ratio of Abdominal coarctation in our patient population has prompted us to study all our cases by Aortography. The abdominal coarct. is very long usually and needs a graft from the thoracic Aorta to the aortic bifurcation as was a one in one of our case.

This patient has been followed for 2 years and his B.P. is well controlled. A one year post-up. Aorto gram showed only 15 mm Hg gradient. Because of the age of our patients all thoracic Coarcts belonged to the post ductal variety. The lumen was usually narrowed eccentrically by a curtain like extension of the media and intima as shown in the aortogram. This is the usual finding as described by Edwards and other (7). However we have also seen concenteric narrowing of the lumen in three of our patients (Aortogram in Fig. 1); large collateral vessels are formed between the branches of the subclavian artery above the coarct with branches of descending aorta below the coarctation. The intercostal arteries posteriorly

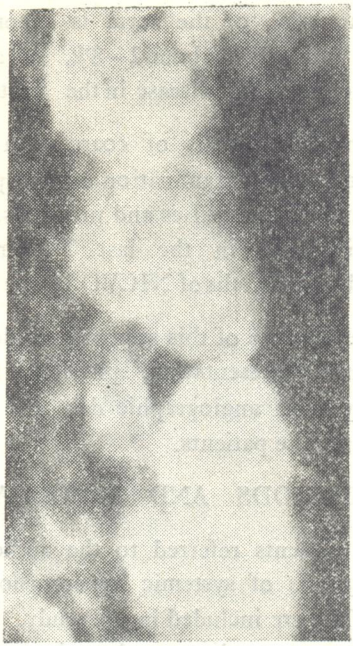

Fig. 1

anastomose with branches of Transverse Cervical artery at these points causing erosion of the rib borders inferiorly resulting in the commonly known rib notching.

The internal mammary artery anastomoses anteriorly with the Intercostal and also with the inferior epigastric artery (see aortogram in Fig. 2).

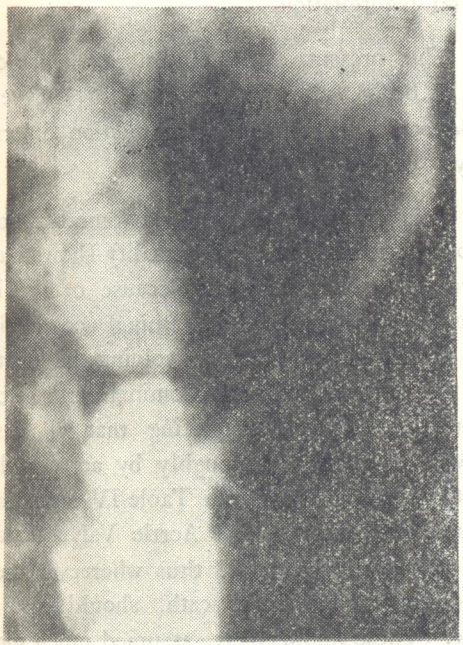

Fig. 2

This anastomosi does not result in rib notching as the intercostal vessel scome out of the groove under the rib and thus are not in close proximity with the rib. The 1st two ribs are also spared because the 1st two inter costal arteries are sub branches of the sub clavian arteries above the coarctatoin. If the subclavian artery of one side arises from below the coarcted segment, rib notching on that side would not develop and thus unilateral rib notching would be seen. Rib notching is usually seen above 5-7 years of age and is also absent in the infantile type (preductal) of coarctation. Other rare causes of rib notching include, Neurofibromotosis, pulmonary atresia, chronic mediastinal fibrosis, A-V shunts of the chest wall. Rib notching is absent in abdominal coorctation or may be present in only the lower few ribs.

Abnormalities of the aortic knuckle has also attracted great attention. In addition to the commonly known sign of "3", high knunckle, low knuckle, or flat or absent knuckle is frequently described.

In the figure of "3" deformity, the central band of figure 3 is due to the constricted part of

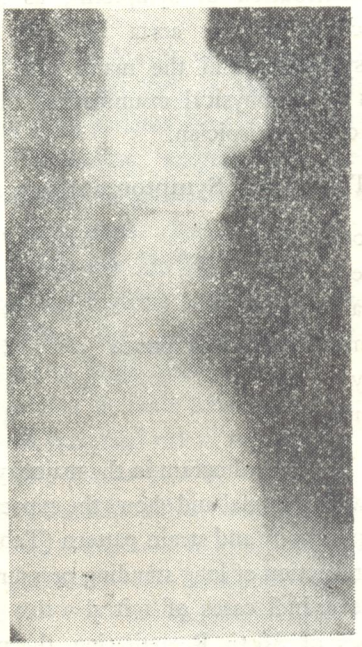

Fig. 3

the coarct, the lower buldge of 3 is due to the post stenotic dilatation and the upper buldge is due to the markedly dilated and tortuous left subclavian artery. The other types of knuckle deformities are produced if there is absence of dilatations in the proximal or distal segment of the coarctation of the aorta. When the subclavian segment is not dilated but post stenotic dilation is present then, low knuckle deformity results, while in the absence of post stenotic dilatation and presence of only subclavian dilatation, high knuckle deformity is seen (Fig.3). The knuckle deformity and rib notching are very useful signs in differentiating the abdominal coarct from the thoracic type. In the chest skiagrams the abdominal coarctation shows marked prominance of Aortic krunckle and absence of rib notching, while the thoracic coarct shows flat knuncle and rib notching. Table No. II shows the frequency distribution of symptoms, as stressed earlier, coarct of the aorta has very little specific symptoms and the major clue in the diagnosis is on physical examination and high index of clinical suspician.

Table II: Symptomatology.

Palpitation	...	17 (74%)
Headache	...	13 (56%)
Dyspnoea on exertion	...	8 (35%)
Paroxysmal Nocturnal dyspnoea	...	2 (9%)
Infertility	...	1 (4%)
Asymptomatic	...	4 (17%)

The Electrocardiogram in the majority of our patients was abnormal and shows the expected left vent. hypertrophy and strain pattern (Table III). This is a reflection of long standing pressure overload however in 3 cases of mitral valve disease and 2 cases of Aortic Regugitation, the preload may have contributed to the hypertrophy.

Table III: E.C.G. Findings in Coarctation.

Left ventricular hypertrophy by Voltage Criteria	...	...	11
L.V.H. with Strain	...	...	3
Left atrial hypertrophy	...	...	5
L.B.B.B.	...	...	1
Normal	...	...	4

Left atrial enlargement was present in six patients, only 3 cases had apparent, significant Mitral regurgitation.

The presence of Left anterior hemiblock and L.B.B.B. may be indication of left vent. hypertrophy or independent conduction abnormality.

Contrary to the previous reports, none of our cases with ECG abnormality showed any increased risk at surgery. Because of the increased prevalence of abdominal coarctation and other types of arteritis involving the lengths of the aorta as well as abdominal aorta in our population, we are suggesting that all cases showed be studied thoroughly by aortography. Furthermore as seen from Table IV significant Mitral Valve disease and Aortic Valve disease was present in 5 cases/23 thus whereever there is suspicion, Rt. heart cath. should also be done. These findings have assumed much more importance in the light of recent findings of high cardiovascular related mortality and morbidity in long term follow-up of operated cases of coarctation of the aorta. The angiographic findings in our cases showed localized coarcts with eccentric lumen in the majority of our cases. This is due to the extension of the intima and media into the lumen in the form of a curtain as described by Edwards et al. The actual lumen may be only few millimeter. Only 18%

cases showed concentric localized luminal narrowing. Out of the thoracic coarcts only two cases had elongated segments requiring a graft for repair. In contrast, the abdominal coarcts had long narrow segments in the majority of cases.

Biscupid aortic valve was seen in 8 cases. Only two had some degree of Aortic regurgitation. Two other patients had less than 50 mm Hg gradient across the Aortic Valve.

Hypertension in coarctation of the aorta is still ill understood. Our study shows that distribution of systolic and diastolic blood pressure in 4 extremities is different in the upper and lower limbs in thoracic and abdominal coarction.

Table IV: Angiographic Findings

Location	Post ductal (PD)	...	17 (74%)
	Abdominal (Abd.)	...	6 (26%)
Morphology			
A) PD —	Eccentric Lumen	...	12 (70%)
	Concentric Lumen	...	3 (18%)
	Elongated Segment	...	2 (12%)
B) Abd.	Localized	...	2 (33%)
	Elongated	...	4 (67%)
Associated Anomalies			
	Biscuspid A.V.	...	8
	Mild A.I.	...	2
	Aneurysm in mesentric vessel.	...	3
	Severe M.I.	...	3

In thoracic coarcts usually the systolic B.P. is higher but diastolic pressures are only mildly or moderately elevated in the upper limbs while in abdominal coarctation both pressures are severely elevated.

The origin of (Lt.) subclavian artery from below the coarct and sometime anomalous origin of (Rt.) subclavian artery from below the coarct also causes lower pressures in the respective arm Regarding the aetiology of hypertension in coarctation of the aorta, the common thinking is that it is due to simple haemodynamic effect of obstruction in the aortic lumen. However, intensive investigations has been carried out on the role of Renin-Angiotensin-Aldosterone system (R.A.A.) in coarctation of the aorta both before and after surgery, but the results have been inconclusive so far. More recently, R.A.A. system has been studied after volume depletion but no conclusive results have been reached even in these studies.

Excellent studies are now available on the immediate long term effect of corrective surgery on B.P. in coarct of the aorta. 30-40% patients were normotensive at the time of discharge in the series of March et. al. but this proportion increases to 60-70% in the next four months. About 40% patients remained hypertensive 11-25 years following surgery. In the series of Simon et.al., 18 patients had residual coarct, seven were reoperated. Residual coarctation is even more common (25%) in patients operated at earlier age (infant) with end to end anastomosis. Thus the subclavian flap technique is utilized in these cases now-a-days. Maron and Humphries in their long term follow up study showed increased risk of premature cardiovascular mortality in the post op. cases of coarct. of the aorta. Mitral valve disease, Aortic Valve Disease, S.B.E. and C.V.A. were among some of the persistant risks to the life of these patients. The risk was none in patients who were operated after 25 years of age. It is also interesting to note that patients who were normotensives on

Table V: Comparison of Thoracic and Abdominal Coarcts.

		Thoracic Coarctation		Abdominal Coarctation
		Pre-Ductal	Post-Ductal	
AGE		Infants	Adolescents, children, adults	Adults
Sex		M ≻ F	M ≻ F (16:1)	F ≻ M (5:1)
L.V.F.		Early	Late	?
HYPERTENSION	Type	—	Systolic mainly	Systolic & Diastolic
Pressure in Extremities	Systolic	—	UL ≻ LL	UL ≻ LL
	Diastolic	—	UL=LL	UL ≻ LL
Aortic Valve Deformity		—	Bicuspid	None
Mitral Valve Deformity		—	+	—
V.S.D.		+	—	—
P.D.A.		++	±	—
Renal Artery Stenosis		—	+	+
Associated Aneurysm of medium size art.		—	+	++
Rib notching		—	+	only lower few ribs.
Aortic knuckle		—	Sign of 3	markedly promine.

long term follow up had experienced a longer preop. hypertensive period, than the ones who were hypertensives (24 vs. 17 years).

Patients having residual hypertension on long term follow up should be studied for recurrence of coarctation and renal artery stenosis. Fowler et.al. described one case of residual hypertension who had operation of recurrence of coarct. of the aorta developing a pheochromocytoma post-operatively. Normotensive children may become hypertensive only during exercise and may be candidates for recurrence.

The optimal age for post-ductal coarct repair is not well settled. The goal is that it should be done at a time when the aortic lumen has achieved enough diameter to prevent subsequent recurrence without delaying so much as to cause secondary changes in the Cardiovascular system as a result of hypertension and collaterals. 8-12 years is the general age recommended. Post-ductal coarcts should be operated before age 5 years only if complications exist. The preductal coarctation is repaired early in life by the subclavian technique and other associated anomalies must also be corrected. In summary, our findings show that all patients having hypertesion below the age of 40 years should also have examination of the femoral pulses and B.P. checked in the lower extremities to detect coarctation of the aorta. Because of the associated Aortic and Mitral valve disease and more so the high frequency of abdominal coarctation in

our population, all suspected patients should be studied carefully. These patients need continued medical supervision and if need be treatment following surgery.

SUMMARY

Twenty three cases of coarctation of the aorta were studied in the haemodynamic laboratory of N.I.C.V.D., from May 1975 to Nov. 1982. Six were females, and seventeen were males. Age ranged between 12-35 years. Six had abdominal coarctation, 17 were of the post ductal variety.

All post ductal coarcts had successful surgical repair. Two of the abdominal coarcts had successful surgery.

Two patients had associated Mitral Valve disease and 3 had small aneurysms in the mesenteric vessel.

Mild A.I. was present in 5 patients. Biscupid Aortic Valve was present in 9 patients. Our study shows that abdominal coarctation is more common in females. Femoral pulses are usually absent, there is a continous bruit in the epigastrium or below and rib notching is absent.

Post ductal coarctation is more common in males (16:1), femoral pulses are present but delayed, ejection systolic or continuous murmur is usually present over the frontal chest or inter-scapular area and rib notching is present.

REFERENCES

1. Sinka S.N.., Coarctation of the Aorta in Infancy Circ. 40:385, 1969.
2. Najafi. H. Julian D.C., Coarctation of the Aorta in Adult Patients. J. Thoracic Cardiovascular Surg. 61:125, 1971.
3. Syed, S.A. et. al., N.I.C.V.D monograph Published May 47, Karachi.
4. Razia J. Rahimtoola et. al., Congenital Heart Disease in Children. Pakistan Heart Journal 13:21, 1980.
5. Sharif M., et. al., Hypertension below age 40 years. Pak. Heart J. 2:49, 1969.
6. Barry J. Maron and Humphries, J., Prognosis in Surgically Corrected Coarctation of the Aorta. Circulation 47:119, 1973.
7. Arthur B. Simon., Coarctation of Aorta. Am. J. Cardiology. 50:456, 1974.
8. March H.J. and Gerbode J., Immediate and remote effects of Resection on HBP in Coarct of the Aorta, Brit. Heart J., 22:361, 1960.
9. Freed D. Michael et. al., Exercise Induced Hypertension after Surgical Repair of Coarct. of Aorta. Am. J. Cardiology, 43:253, 1979.
10. Parker B.F. et. al., Preoperative and Post Operative Renin Levels in Coarct of the Aorta. Circulation 66:513, 1982.

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