THE COURSE AND PROGNOSIS OF COARCTATION OF THE AORTA

BY

MAURICE CAMPBELL AND J. H. BAYLIS

From the Cardiac Department, Guy's Hospital and the Institute of Cardiology

A beautiful description of the collateral circulation was given by Bramwell and Morgan Jones (1941), and Bramwell (1947) discussed the clinical features and prognosis.

The prognosis of patients with coarctation of the aorta is judged mainly from studies of those who have died in hospital, for no large series seemed to have been followed clinically for many years. To advise a patient wisely whether he should undergo operation demands a knowledge of the prognosis, and this will become more difficult to acquire as increasing numbers take advantage of the operation that was shown to be possible by Crafoord and Nylin in 1945. This paper is based on a study of 130 patients, mainly in an attempt to assess the prognosis, about which we have become less satisfied, for an absence of significant symptoms gives no certainty of continuing good health, even for the next few years.

Incidence. Coarctation occurs in from one in a thousand (Fawcett, 1905; Blackford, 1928; and Evans, 1933) to one in four thousand necropsies (Reifenstein *et al.*, 1947). The clinical incidence was 1 in 10,000 in American Army recruits, and 1 in 12,000 in children under 15 years at Toronto (Mustard *et al.*, 1955).

Sex Incidence. In our series there was a ratio of 5 males to 3 females, but earlier there were more men as several were diagnosed during military service. In the necropsy series of Abbott (1928) and Reifenstein *et al.* (1947) it was four times as common in males, and in most surgical series it has been two or three times as common. There was, however, an equal sex distribution in 44 children (Bonham-Carter, 1954) and in 23 consecutive patients in Helsinki (Halonen *et al.*, 1951). As many are without symptoms and as routine medical examinations are more common in men, the true male preponderance may not be as high as has been thought.

Age of Patients when First Seen. Of our patients, 60 per cent were under 20 and only 14 per cent over 30 years of age (Table I): nine have since passed 30, but this only increases the percentage

TABLE I

AGE INCIDENCE (WHEN FIRST SEEN) OF PATIENTS WITH COARCTATION

	0 to 4	5 to 9	10 to 14	15 to 19	20 to 24	25 to 29	30 to 34	35 to 39	40 to 55	Total
Males	6	10	10	21	10	12	9	3	1	82
Females	3	8	13	7	4	8	3	1	1	48
Total	9	18	23	28	14	20	12	4	2	130
With aortic stenosis	—	2	—	—	2	—	2	—	—	6
With aortic regurgitation	1	2	4	7	5	4	3	2	0	28
*Percentage with aortic regurgitation	14		18	22	27		28			22

*In calculating this the patients with aortic stenosis have been excluded: three patients who were under observation when aortic regurgitation developed have been included twice—before and after.

to 21, which is less than we expected. This is partly because adult patients are difficult to follow regularly, and are reluctant to return to hospital when they feel well, but if many were still living it seems that more should have been seen for incidental reasons. We cannot recollect seeing anyone for angina pectoris who was then found to have coarctation of the aorta, though we have seen such with dextrocardia, for example. The age incidence of these cases suggests, but can do no more, that there is a heavy mortality from 20 onwards.

DIAGNOSIS

If the blood pressure is taken whenever there is unusual pulsation in the neck (Bramwell, 1947) and if the femoral pulse is felt whenever there is brachial hypertension or unexplained aortic regurgitation few cases should be missed. Bending forward with the arms hanging by the side (Campbell and Suzman, 1947), has proved useful in demonstrating more clearly the collateral circulation in the back. Rib notching is not often seen before 7 or decisive before 10 years but we have seen it clearly at the age of 6 (Fig. 1): it is not always found especially if there is a good collateral circulation elsewhere, but may become extreme (Fig. 2). Other radiological signs are rarely diagnostic but are often interesting and sometimes important: some examples are shown in Fig. 3.

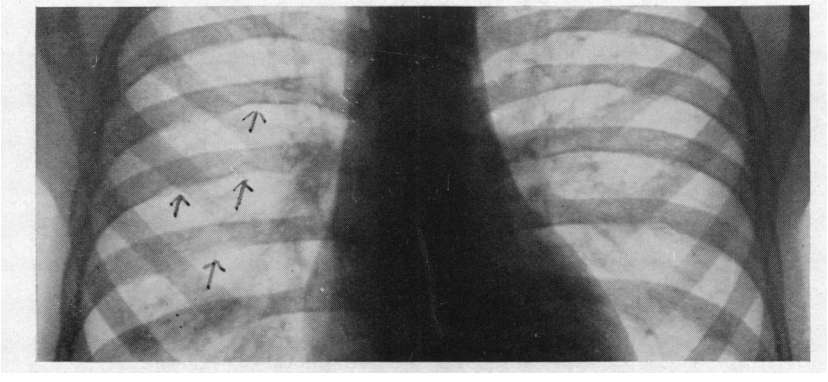


FIG. 1.—Notching of the ribs already clearly visible, especially in the sixth rib, in a boy aged 6 years.

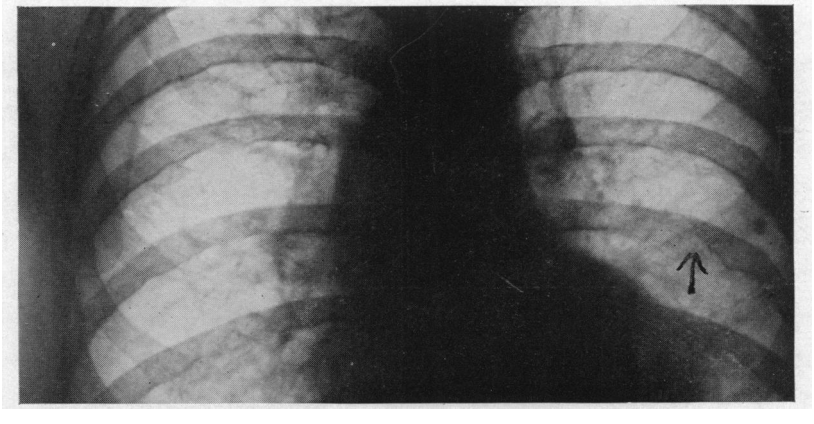


FIG. 2.—Unusually deep rib notching in a woman, aged 52 (Case 21). We do not know of any case where this has led to fracture of a rib, but the tortuous collateral arteries may become aneurysmal and rupture.

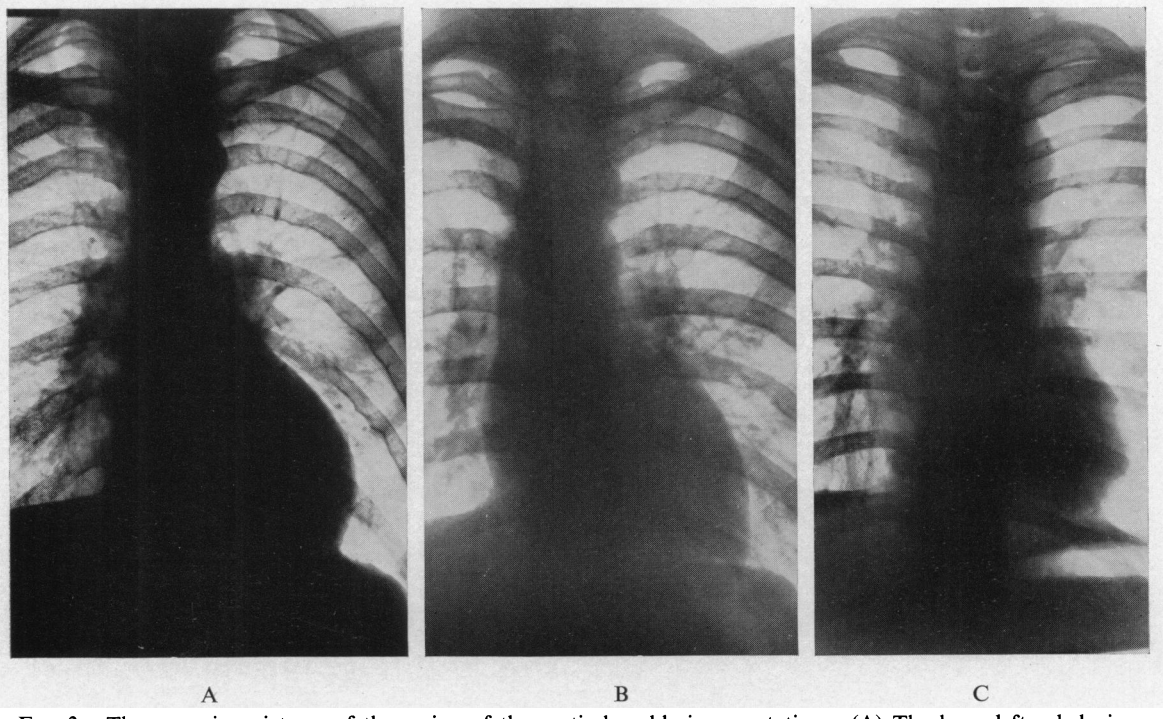


FIG. 3.—Three varying pictures of the region of the aortic knuckle in coarctation. (A) The large left subclavian artery simulates the aortic knuckle. (B) Below the shadow of the left subclavian a second shadow, the descending aorta, is clearly seen. (C) The descending aorta is easily seen and forms a continuous shadow from the site of the aortic knuckle. The heart was not enlarged (c.t.r. 46–49%) in any of these three patients.

A difference of blood pressure of more than 30 mm. between the two arms suggests that the left subclavian is involved in the coarctation or, more rarely, arises below it. Palpable pulsation in the abdominal aorta suggests an abnormally low coarctation. We agree with Granstrom (1951) and Walker and Stanfield (1952) that the retinal arteries show striking corkscrew tortuosity without the usual changes seen in hypertensive retinopathy.

Physique. Several observers have commented on the good physique of patients with coarctation (Newman, 1948) and, in general, we agree, though it is not confined to well-built subjects. We have not proved this by measurements, but an unduly large proportion, women as well as men, have a well-developed chest. Often the shoulders are broad and the muscles stand out prominently. The physique was unusually good in rather more than a quarter of these patients, average or good in about half, and poor in less than a quarter. If patients with coarctation and others with atrial septal defect were mixed, 70 per cent could probably be separated correctly without looking at the pulsation in the neck.

COURSE AND PROGNOSIS

Some of these patients have been seen only for a short time or had resection soon after their first visit, but 80 have been followed on an average for five years and a few much longer. We shall describe the state of those seen in each decade to give an idea of the course and prognosis. School children and men liable for military service are seen more often than older patients without any symptoms simply because of the murmur so our picture of the natural course and prognosis may be biased by this, but otherwise we think it reasonably representative. This seems the only way the question can be answered, for patients can rarely be followed for 30 or 40 years.

Two patients have been followed for nearly 25 years. The first still works and feels well but

has deteriorated through the third decade: the second is still well at 49 though her blood pressure was 220/115 when she was first seen.

*Case 2.** This boy was first seen when he was 7 and was a little breathless; when he was 11, the collateral circulation and rib-notching were obvious. The blood pressure was averaged 147/85 at 7, 176/95 at 11, and 210/110 at 16, without any change clinically or in the size of the heart (c.t.r. 47%). Aortic regurgitation developed between 16 and 21, but there was no change in the pressures till he was 26, when they were 240/90 and for the first time there was some left ventricular strain in the cardiogram. He was unwilling to have an operation. He is now 30 and is at regular work with few symptoms. In spite of the aortic regurgitation, the heart shows no more than fulness of the left ventricle (c.t.r. 48%) but the blood pressure is now 260/90 mm.

Case 3. When she was 25 she became a little breathless after a stillbirth in her second pregnancy: her blood pressure was about 210/110 and sometimes 230/120. Thirteen years later her fourth child was born in Guy's Hospital by Cæsarean section: she made a good recovery and the pressure settled from an average of 235/118 to 205/109 resting in the ward. When she was 46 she thought it was time to stop going out to work as well as doing her own work at home. At 49 she is still leading a normal life, but is a little more breathless; the heart is little changed (c.t.r. 56% instead of 54% when 25), the blood pressure is 250/120 and the cardiogram now shows slight inversion of T in V5 and V6 only.

Course during the First Decade. The occurrence of congestive failure in the first months of life is well-known: a mortality of over 50 per cent has been reported in such infants (Nouaille *et al.*, 1954; Mustard *et al.*, 1955) but Gross (1950) thinks that most of them recover with medical treatment as the collateral circulation improves. One patient sent to us with this diagnosis had, in fact, lymphatic obstruction of the type seen in Milroy's disease.

We have seen 28 patients in this decade and three of them have died. The first was a girl, aged 3, with recurrent attacks of failure (Case 22, q.v.). A boy, aged 7, has probably died, for we watched him getting worse and his heart getting larger during six years: his aortic stenosis and incompetence were most likely rheumatic for we saw him with typical rheumatic nodules and pericarditis (*Case 1*). Towards the end of this decade one girl became increasingly short of breath and a year later was admitted with bacterial endocarditis; although she recovered from this she died soon after with congestive failure (Case 18).

One girl, aged 9, had severe migrainous headaches, one boy, aged 3, with a blood pressure of 142/82, had such severe epistaxis that he needed transfusion, and another girl, aged 3, had a very large heart that we attributed to congenital mitral incompetence: all these have had successful operations, but the last too recently to know if the heart will become much smaller.

With these exceptions most children have come to us for routine examination because of their murmur and have seemed normal to their parents. A few were breathless or easily fatigued, and three complained of headaches that may have been due to berry aneurysms. We have no evidence of any others losing ground during this decade. Most of them are developing well and leading a normal life without symptoms, though generally the blood pressure shows a steady increase. In 8 patients followed on an average for 7 years, the mean pressure rose from 147/98 to 170/106, but in six others there was little rise during four years.

Second Decade. We have seen 50 patients in this decade and watched many of them through most of it, with only one death. This girl died in hospital when she was 17: there was no necropsy and the cause of death was said to be " pneumonia and aortic incompetence," but we think she may have had heart failure (*Case 4*).

For most patients this period is uneventful, though the blood pressure continues to rise. Campbell and Suzman (1947) thought that it rose till somewhere between the ages of 15 and 18 and then stabilized and this is supported by our larger series where the steep rise levels off at about 17 or 18 years (see Fig. 4). In 8 cases, followed on the average for eight years, the mean rise was from 168/99 to 193/108; but there were 7 others with no rise during five years—generally older patients followed from about 14 to 19 years of age.

Cerebro-vascular accidents, however, may start in this decade. Four patients came under

* Cases 1–15 are numbered as they were when reported by Campbell and Susman (1947): Cases 16–28 are shown in Table III.

our observation because of such attacks (see p. 483): two made excellent recoveries but two were left with hemiplegia. Cerebral attacks are less often fatal than in the next decade, but may be so or may leave a persistent hemiplegia.

Apart from these major complications symptoms are not common before the third decade: headaches and dyspnœa on exertion are the most usual. Patients rarely complain about their legs, but from this age onwards some coldness or aching on exertion may be elicited by questioning.

Third Decade. This is very different from the two earlier decades. Of the 37 patients, many still regard themselves as symptomless though more have headaches or cannot do as much as others; most of them are leading normal lives—many even active or athletic lives. In general the blood pressure no longer shows any progressive rise and the majority still have hearts of normal size.

This might suggest that the outlook is still just as good, but 5 of the 37 patients have died and most of these died suddenly or rather suddenly within two or three years of a time when they seemed as well as ever. One died from a ruptured aorta, one from a cerebral hæmorrhage, and two from congestive failure (Cases 16, 17, 15, and 19): all these had been in good health two years before. The fifth was cured of bacterial endocarditis, but developed congestive failure three years later and died after operation (Case 23). Few other patients have lost ground seriously, but in two aortic regurgitation was increasing and leading to progressive cardiac enlargement.

The blood pressure is no longer rising at this age and the average figure shows a steady level at 191/105. Individual patients where we have records over several years show the same result, for 12 followed for four years or more showed no change, and only 3, all followed for ten years, showed any rise.

Fourth Decade. Two of the 17 patients died during this decade—a man, aged 32, who had a very large heart and free aortic regurgitation, and a man, aged 31, who had aortic stenosis and was becoming disabled, though the immediate cause of death was resection (Cases 20 and 24). The numbers are large enough to suggest this is a safer period and that most of those with the greatest risk of rupture of the aorta or of cerebral hæmorrhage die during the third decade.

In general, the others are doing well and few of them have increasing symptoms. The only patient we have seen with atrial fibrillation, and it was paroxysmal, was of this age and it has become no worse during four years.

In the early thirties the average blood pressure was no higher than in the twenties, and the slight rise towards the end of this decade is of doubtful significance for the numbers are smaller. Of the 7 individual records available over some years, only one showed much change. His pressure rose from 180/110 to 200/125 between 27 and 31; he was leading a most energetic life loading and driving lorries for long hours, and since he has taken life more easily there has been no further rise in five years.

Fifth and Sixth Decades. There are not enough patients in this period for generalizations. The small number suggests a higher mortality in the previous decade than do our direct observations. Most of the survivors have a high blood pressure and there is a slight increase in the average figure (220/122) after a period of stability, but the numbers are too few to be significant. One woman had a blood pressure of 235/110, a large heart (c.t.r. 56%), and free aortic regurgitation so that we hardly expected her to do well, yet five years later she was very well with no change. Case 3, who has been followed from 25 to 48 is still well though her blood pressure is now 250/120. In a third the blood pressures were lower, 178/92, and had not changed for 23 years.

The oldest patient was first seen when she was 52 and was admitted to hospital with pneumonia: shortly before there had been an episode suggestive of left ventricular failure; her blood pressure remained about 217/128, but these episodes became increasingly troublesome and she died with congestive failure when she was 55 (*Case 21*).

Conclusions about Rise of Blood Pressure with Age. These are shown in Fig. 4 and Table II. Patients with aortic stenosis have been excluded, but not those with aortic regurgitation (see p. 486). and those followed ten years or more have been included twice. The systolic pressure rises much

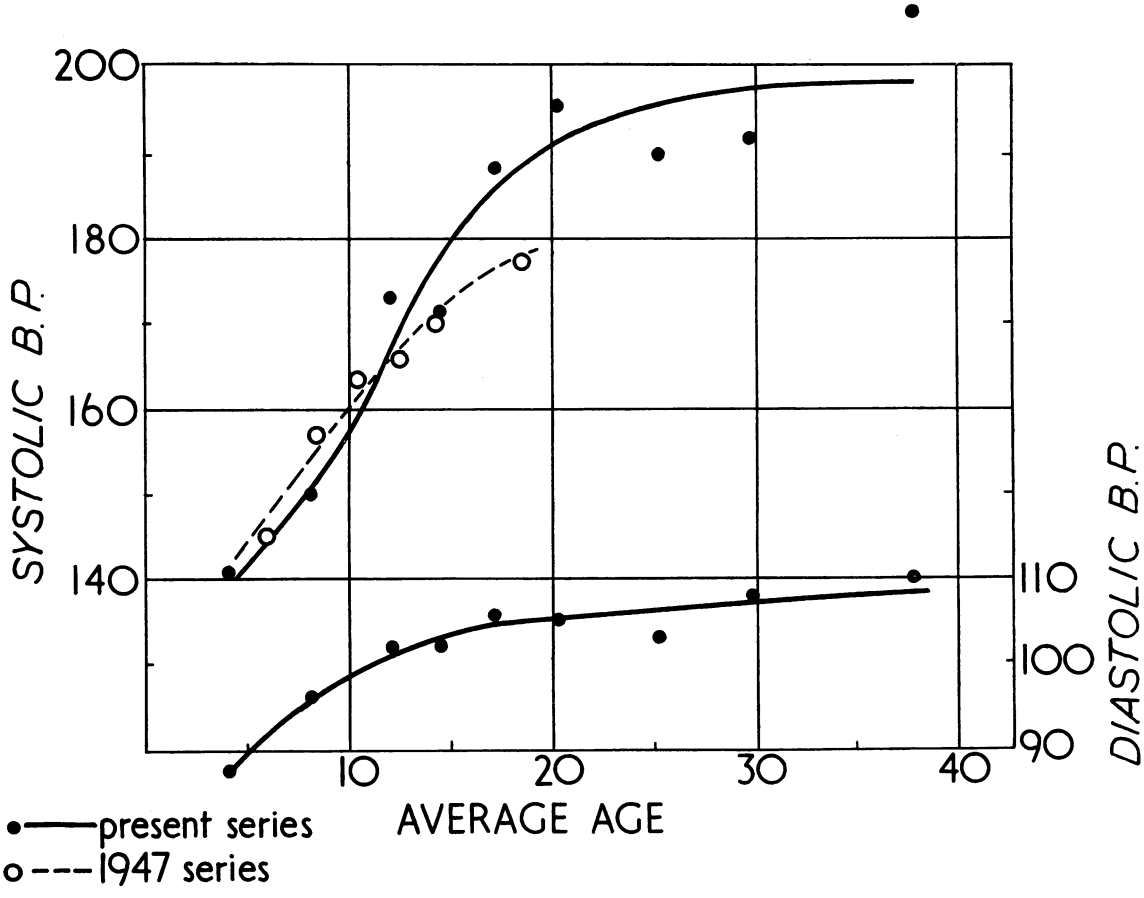


FIG. 4.—The rise of systolic and diastolic blood pressures with age. The curve for the diastolic pressure (below) is of the same shape as the curve in normal subjects, about 30 mm. higher throughout. The curve for the systolic pressure (above) has the same general shape but rises more sharply than the normal curve, so that it is 55 mm. above normal at 5 years and 75 mm. above normal after 20 years of age. The dotted line represents the curve for the systolic pressures reported by Campbell and Suzman (1947) from their own and other reported cases.

TABLE II

BLOOD PRESSURE IN COARCTATION OF THE AORTA RELATED TO AGE

	Average Age	Blood Pressure	
		Systolic	Diastolic
Youngest 15 patients	4·4	141	87
Next 15	8·1	150	96
Next 15	11·5	173	102
Next 15	14·2	171	102
Next 15	17·3	188	106
Next 15	20·7	195	105
Next 15	25·9	190	103
Next 15	29·3	192	108
Oldest 15 patients	37·5	206	110

The patients have been arranged in order of age and grouped in fifteens to minimize individual variations.

more than the diastolic, 55 mm. against 20 mm. from 5 to 35 or 40 years. The rise is not regular: both rise more steeply and fairly regularly until about the age of 17 and then level off and rise very slightly, if at all.

The blood pressure on a smoothed curve rises from 140/87 at the age of 4, to 154/93 at 8, to 169/99 at 12, to 184/104 at 16, and to 193/105 at 20; and only to 197/107 at the age of 30 years.

Conclusions from Patients who have Died. Of the 16 patients who have died (see Table III) 4 give us no information as they were in good health and died unexpectedly after operation. Of the other 12, six were seen because they were already seriously disabled so do not help much in judging the prognosis but the other six were in good health when they were first seen and three were sent only because of a routine medical examination (Case 4, see p. 478; Cases 15–19 see pp. 482–484): there was, therefore, no reason to anticipate their deaths yet they all died within two or three years. The unexpected deaths of these six, mostly in the third decade, has been the main reason for making us think that operation should be advised for most young patients.

Hazards of Coarctation

In cases that have survived infancy, the main causes of death during the first three decades are aortic rupture, bacterial endocarditis or aortitis, and intracranial hæmorrhage. After the age of 30 the incidence of congestive heart failure rises, sometimes without complications, sometimes after bacterial endocarditis, but most often when there is aortic regurgitation. Our patients illustrate all these hazards (Table III). After 40 there is about an even chance of death from an incidental cause.

TABLE III

Some Findings in Patients with Coarctation who have Died

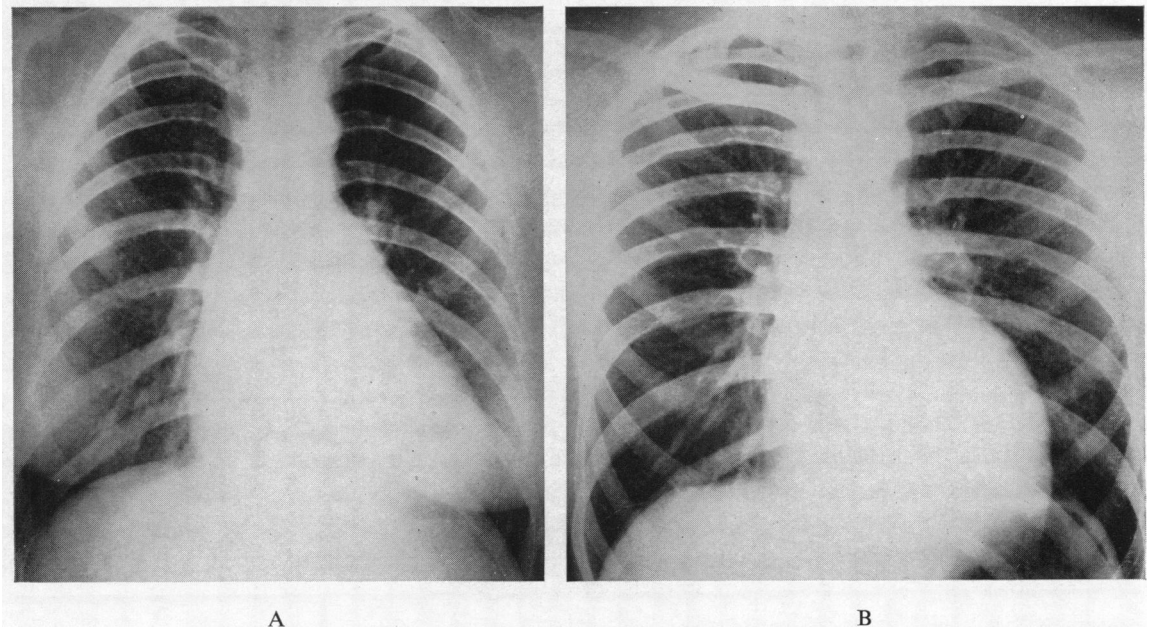
Case No.	Age and sex	B.P.	Aortic valve	C.t.r (%)	Cause of death	Aortic valve cusps	Other findings at necropsy, if any
1	M13	165/60	A.S.A.I.	68	?Rheumatic H.D.	—	No necropsy
4	F 17	200/90	A.I.	56	"Pneumonia and A.I."	—	No necropsy
15	M27	172/109	A.I.	48	C.H.F.	—	No necropsy
16	F 22	210/110	(A.I.)	61	Ruptured aorta	?3	Aortic cusps thickened
17	M27	190/85	(A.I.)	54	Cer. hæmorrhage	2	Aortic cusps thickened and incompetent
18	F 9	170/105	?A.S.	61	Failure after S.B.E.	—	No necropsy
19	F 28	204/106	(A.I.)	50	C.H.F.	—	No necropsy
20	M32	190/101	A.I.	66	?C.H.F. ?S.B.E.	—	No necropsy
21	F 55	217/128	N	52	L.V. failure	—	No necropsy
22	F 3	172/143	N	71	At operation*	3	198 g. Valves thickened: bicuspid pulm. valve
23	M26	200/115	N	58	At operation*	3	L.V. much hypertrophied
24	M31	155/110†	A.S.	60	Aneurysm at site of anastomosis*	3	Aortic valve stenosed (Fig. 8)
25	F 11	190/100	(A.I.)	51	At operation*	3	140 g. Nothing else
26	F 13	180/102	N	53	Cer. hæmorrhage*	—	No necropsy
27	F 14	185/95	N	48	Hge. from graft*	2	240 g. Nothing else
28	M17	210/111	N	48	Acute enteritis*	3	455 g. Small diverticulum in septum only

† Systolic pressure in left ventricle, 198. * Died after operation.

In the 304 cases that came to necropsy in the combined series of Abbott (1928) and Reifenstein *et al.* (1947) the main causes of death were as follows. Over one-quarter (29%) died from incidental causes not connected with the coarctation, but this group is not represented in our series. Just over one-quarter (26%) died from congestive failure which is more prominent in our series. Less than one-quarter (21%) died from aortic rupture. The last quarter of the deaths were equally divided between intracranial hæmorrhage (12%) and bacterial endocarditis (12%).

Rupture of the Aorta

One of our patients died from this cause. The first part of the aorta was prominent on X-ray examination (Fig. 5A) and this may perhaps give some warning of the risk; otherwise, it cannot be foreseen for as a rule the blood pressure is not specially high (Reifenstein *et al.*, 1947).



A B

FIG. 5.—(A) A large heart (c.t.r. 61%) from a woman, aged 20, with coarctation. The first part of the aorta was unusually prominent and she died with a ruptured aorta two years later. (B) A large heart with a rounded left ventricle (c.t.r. 54%) from a man, aged 21, with coarctation and aortic stenosis.

Case 16. A woman, aged 20, had always been a little breathless and had aortic regurgitation with a blood pressure of 210/110. She remained well for two years but suddenly became unconscious and died shortly afterwards. At necropsy, the arch of the aorta was thin and aneurysmal and there was a rupture at this site, presumably due to a dissection as the pericardial sac contained 600 ml. of blood. The aortic cusps and the anterior cusp of the mitral valve were somewhat thickened.

In one girl, aged 11, the aorta was difficult to sew up at operation, probably because of the poor quality of the media, so there may have been a risk of aortic rupture (Case 25). Rupture often occurs through a dissecting aneurysm but sometimes through a pre-existing saccular aneurysm. Two of our patients had such aneurysms. One had a large aneurysm proximal to the coarctation, both successfully resected (Brock and Graham, 1952), and the other had a calcified aneurysm just below it (Case 23). In the series of Clagett *et al.* (1954) there were 11 instances of aneurysms distal to, and only 2 proximal to the coarctation: in general, they found the aortic medial changes greater in men.

Aortic rupture occurs at two sites, proximal to the coarctation but generally in the ascending aorta rather than just proximal, and less often distal to it. Among the 304 cases in the series of Abbott and of Reifenstein there were 56 instances of the former and 16 of the latter.

Proximal rupture starts as a dissecting aneurysm: it is associated with medial degenerative changes or rarely with infective aortitis, e.g. in 2 of the 56 quoted. It may occur at any age but mainly in the second and third decades. Among 141 patients under the age of 40 with dissecting aneurysms, 99 of whom died from aortic rupture, Schnitker and Bayer (1944) found coarctation of some degree recorded in 45—in 19 of the 27 aged between 11 and 20, and in 17 of the 53 aged between 21 and 30 years.

Spontaneous rupture of the ascending aorta is the main hazard of pregnancy in patients with coarctation, and was responsible for 6 of 11 cases where death was directly associated with pregnancy (Rosenthal, 1955), in one during labour and in the rest during the third trimester. In Schnitker and Bayer's series there were 92 males and 49 females, and 24 of these 49 were or just had been pregnant and 15 were in the third trimester: the blood pressure was not as a rule known to be specially high and they suggest that metabolic factors during pregnancy predispose to cystic medio-necrotic changes in the aorta.

Distal rupture occurs spontaneously and from endo-aortitis. The latter occurs mainly in the first two decades, and in the 8 cases reviewed by France *et al.* (1950) 6 were under 18, the average age at death being 17 years. In Abbott and Reifenstein's 16 cases, aortitis was present in 6 at least. Spontaneous rupture occurs from rupture of aneurysms, sometimes traction aneurysms at the site of the ligamentum arteriosum (Monckeberg, 1907), or through a jet lesion (Bellet and Gelfand, 1952) or from mucinous medial degeneration (Zaslow and Krasnoff, 1943). In contrast with the aortitis group, the average age at death in ten cases was 37 (France, 1950), only one being under 18 years. No instance of distal rupture has been recorded with complete aortic atresia.

Intracranial Hæmorrhage

This is responsible for about one death in eight. Two of our patients died from this cause, one the day after operation when there was a temporary rise of blood pressure (Case 26).

Case 17. A man, aged 27, had coarctation (B.P. 190/85), aortic regurgitation, and familial polyposis of the colon; in view of the risk of this becoming malignant, he had colectomy, which was well borne. But 18 days after his discharge, he had a sudden agonizing headache and died a few hours later. Necropsy confirmed a large hæmorrhage that had disrupted the right internal capsule but had not broken through into the ventricle. The aortic valve was bicuspid and incompetent.

Apart from the danger of death there is the risk of residual cerebral damage, sometimes with hemiplegia. Among our 80 patients followed for an average period of five years, only one, a girl aged 13, has had a cerebral episode. When sitting at tea she dropped a cup and said she had a severe headache, and stumbled on the way to bed. The headache persisted for a day, and the left arm and leg felt numb. She had quite recovered in three days except for numbness and tingling in the leg which was more sensitive to pain. When she was seen the blood pressure was no higher than usual, 145/90: four years later it was lower, 138/87, and she has continued to lead a normal life.

Four other patients had episodes of this type before we saw them—two with a less satisfactory outcome. A girl, aged 15, had signs of meningeal irritation and the cerebro-spinal fluid was blood-stained and under increased pressure: her blood pressure was always labile and rose to 190/110 with very gentle exercise. A man, aged 28, had a subarachnoid hæmorrhage soon after his recovery from bacterial endocarditis. Both made good recoveries and have had successful resections since.

A boy, aged 19, was admitted with a blood pressure of 260/120 instead of the usual 197/116 and with hemiplegia which persisted: four years after resection he is getting on well with a pressure of 157/95. A girl, aged 13, had frequent headaches and one severe attack was followed by loss of consciousness and a left hemiplegia; coarctation was diagnosed and she had a successful resection: six years later she is well with a blood pressure of 133/92 instead of 185/120: her leg has recovered but her left arm is of little use. Earlier diagnosis and treatment could probably have prevented these two tragedies.

Intracranial hæmorrhage is due in about half the cases to berry aneurysms that can be demonstrated pathologically, and rarely to mycotic aneurysms. Unlike aortic rupture, it shows some correlation with the level of the blood pressure (Reifenstein *et al.*, 1947). Death from intracranial hæmorrhage is most common in patients between 10 and 30 years of age; there was only one earlier than this among the 31 cases of Abbott and of Reifenstein, and in 11 proven ruptured berry aneurysms in association with coarctation, the youngest was 13 years old (Wright, 1949).

Bacterial Endocarditis and Aortitis

These were responsible for another one-eighth of the deaths in the combined series, a proportion that has now been greatly reduced though not abolished. They still, however, cause death indirectly, from congestive failure after the infection has been cured.

Case 18. A girl, aged 9, had not been able to do as much as usual for three years. Additional aortic stenosis was suspected as there was a striking murmur and thrill in the aortic area and a large heart (c.t.r. 61%) with S–T depression from V2–V6, but the blood pressure was 174/98. Operation was advised but there was some delay and she was admitted elsewhere with bacterial endocarditis (blood culture positive, hæmoglobin 33%, an added aortic diastolic murmur, and B.P. down to 140/70). She seemed to have recovered after prolonged treatment with penicillin when she went home five months later, but was back in three weeks with congestive failure from which she died a month later. There was no necropsy.

Two other patients developed bacterial endocarditis during the period of follow-up, one year and six years after they were first seen; both made good recoveries. Two others were sent to us just after treatment for bacterial endocarditis because this led to the recognition of their coarctation. Another (Case 23) had bacterial endocarditis three years before he came to us with congestive heart failure. In three cases the causative organism was *Streptococcus viridans* but in the other three blood cultures were repeatedly negative.

Endocarditis is mainly a hazard of bicuspid aortic valves, and may be followed by free aortic regurgitation and cardiac hypertrophy (Reifenstein *et al.*, 1947). No follow-up study of successfully treated cases has been found, and there was only one instance of coarctation in 442 cases of treated bacterial endocarditis (Cates and Christie, 1951).

Aortitis occurring alone was the cause of death in 6 per cent of Reifenstein's series, the predominant sites of infection being the aorta just distal to the coarctation and the ascending aorta: it was frequently found with endocarditis.

Congestive Heart Failure

This was the commonest cause of death in our series—in 8 patients, if we include two already ill, who died after operation. The girl, aged 3, had an unusual coarctation (Case 22). Two, aged 9 and 24, had failure after recovering from bacterial endocarditis (Cases 18 and 23). The man aged 32 had aortic stenosis also (Case 24). Of the other four, aged 17, 27, 29, and 55, the three younger ones all had aortic regurgitation. Notes of the two who were in good health when they were first seen and of the unusual infant follow.

Case 15. A man, aged 25, had no symptoms and was diagnosed on his discharge from the Navy. He had aortic regurgitation, though the diastolic pressure was high (B.P. 172/109) and the heart was not enlarged. Two years later he became ill and died within three months, probably from congestive failure, although when seen he had been leading a normal life.

Case 19. A woman, aged 26, seen during her first pregnancy, remained well for two years but then developed congestive failure. She was very anæmic but bacterial endocarditis was not confirmed. A loud aortic diastolic murmur, without much change in the pulse pressure (200/100 instead of 203/109), was heard for the first time. Her anæmia improved very slowly and some congestive failure persisted, but after three months in hospital she was allowed to go home. She died soon afterwards and there was no necropsy.

Case 22. A girl, aged 3, had recurrent congestive failure for 18 months. The striking pulsation in the neck seemed to exclude aortic stenosis, which was suspected because the pressure was not high enough to explain the very large heart and recurrent failure. At operation the pressure was 168/79 in the left ventricle and 172/143 in the first part of the aorta, though it had been 130/95 in the brachial arteries. The first part of the aorta was dilated but became narrowed and obstructed before the origin of any aortic branches. At necropsy, the heart weighed 198 g. and the left ventricle was greatly hypertrophied. The aortic valve was thickened but not stenosed: the pulmonary valve was bicuspid.

Congestive failure is uncommon in young patients in the absence of some additional lesion. Among the 19 patients dying in failure in Reifenstein's series, 18 had coincident valvular, coronary arterial, or some cardiorenal disease; the average age at death was 39 years and only one was under

20; and all six in whom the duration of failure was a year or less had advanced deformity, usually stenosis, of the aortic valve. Our patients show that coarctation with no complication except aortic regurgitation may lead to congestive failure by the third decade.

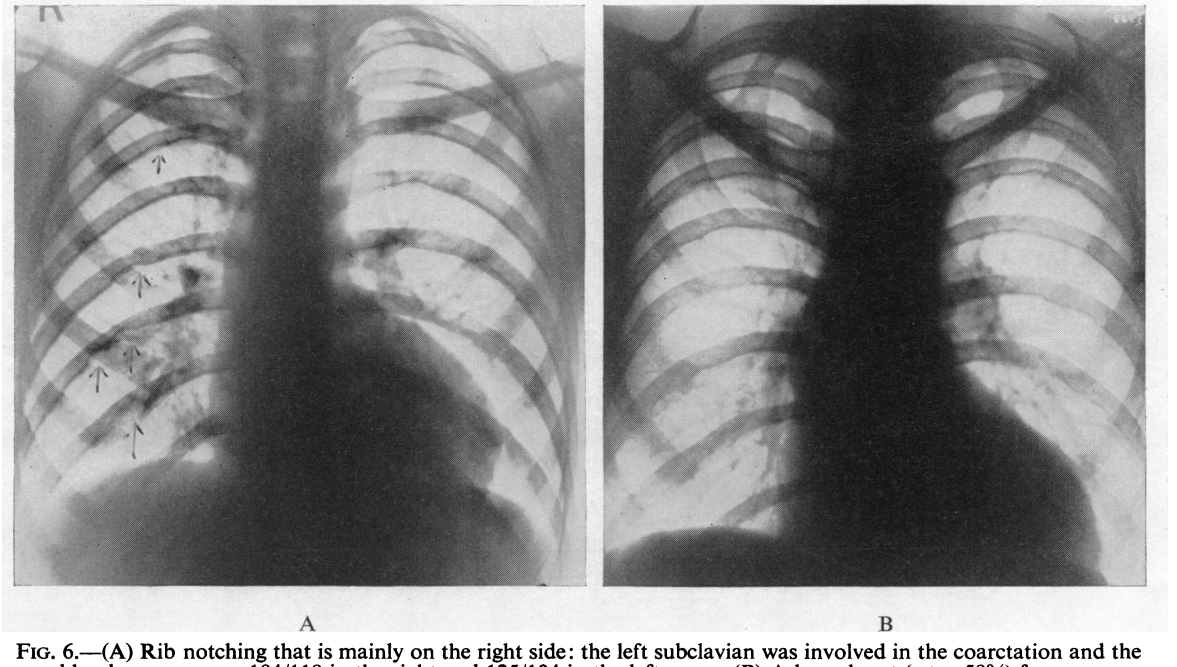
Coarctation at or Above the Origin of the Left Subclavian Artery

Brown (1950) states that this is rare as only 20 cases had been recorded, but many more have been found in recent series. When the origin of the left subclavian artery is involved in the coarctation its recognition is of surgical importance, and is suggested by *consistent* differences of pressures in the right and left brachial arteries.

Differences of 20 mm. are not significant. We have, for example, found consistent differences in several patients, e.g. 210/110 and 190/110, 210/130 and 195/125, 235/118 and 220/112—the highest reading in each of these being in the right arm. Such differences do not militate against successful operation, e.g. readings of 215/106 and 194/105 became 182/93 and 159/90 after a resection.

Large differences are, however, of great significance. In one patient in whom no operation was possible owing to the relationship of the aortic branches to the coarctation, no difference was recorded between the two arms and the *right* subclavian was the last branch before the coarctation; but in the other, the readings in the right arm were 184/118 and in the left 125/104, and this warned us of the involvement of the left subclavian in the coarctation and of the difficulty of operation which, in fact, proved insuperable in 1950, though it may be overcome by grafting. In another boy, the pressure averaged 162/107 in the right, and 130/97 in the left arm; he had few symptoms and after an aortogram in 1949 operation was not advised as the left subclavian seemed involved in the coarctation: six years later his condition is unchanged.

The last two were good examples of unilateral rib notching, though in the former there was slight notching of one rib on the left side as well as the major notching on the right side (Fig. 6). Valdoni (1955) finds that the pressure is higher than usual when the collateral circulation depends on one side only.



A B

FIG. 6.—(A) Rib notching that is mainly on the right side: the left subclavian was involved in the coarctation and the blood pressure was 184/118 in the right and 125/104 in the left arm. (B) A large heart (c.t.r. 58%) from a man, aged 33, with coarctation and calcification of the stenosed aortic valves. The dilatation of the first part of the aorta is characteristic of congenital aortic stenosis but is not seen in Fig. 5B.

Exceptionally, the difference may be the other way round or both subclavians may be obstructed. The readings averaged 162/100 in the right and 177/110 in the left arm in one case, and 140/110 in the right and 190/100 in the left arm in Case 25: the right subclavian (3 mm. in diameter) arose below and behind the large and dilated left subclavian from a very small projection of the aorta above the coarctation, which was 1 cm. long.

The origins of both subclavians were obstructed, partly by thrombosis, near the coarctation and the blood pressure was 200/115 in the temporal artery at a time when it was only 130/90 in the brachial arteries in Case 23 who died after operation.

Aortic Regurgitation and Coarctation

Some degree of aortic regurgitation was indicated by a diastolic murmur in nearly a quarter of these patients. It is, we think, most often due to a bicuspid aortic valve that has become atherosclerotic. Generally, it is slight and the murmur is a soft one, there is no water-hammer pulse, and the pulse pressure is no wider than usual. We soon found that the diastolic pressure was higher than in other forms of aortic regurgitation, and that it was rarely below 100, though in the course of years the usual picture with a wide pulse pressure and a low diastolic pressure may develop. This is well shown in Fig. 7, relating the systolic and diastolic pressures, for most of those with aortic regurgitation (shown by the white circles) fall among the others, and are absent only from the narrow upper margin showing patients with relatively high diastolic pressures: there are only 4 patients with regurgitation standing out below the lower margin of the general scatter, where the diastolic pressure was low because the regurgitation was more significant.

Excluding the six patients with aortic stenosis, some of whom had regurgitation also, 28 of 124 patients had aortic regurgitation. This incidence of 23 per cent is not much below that of bicuspid

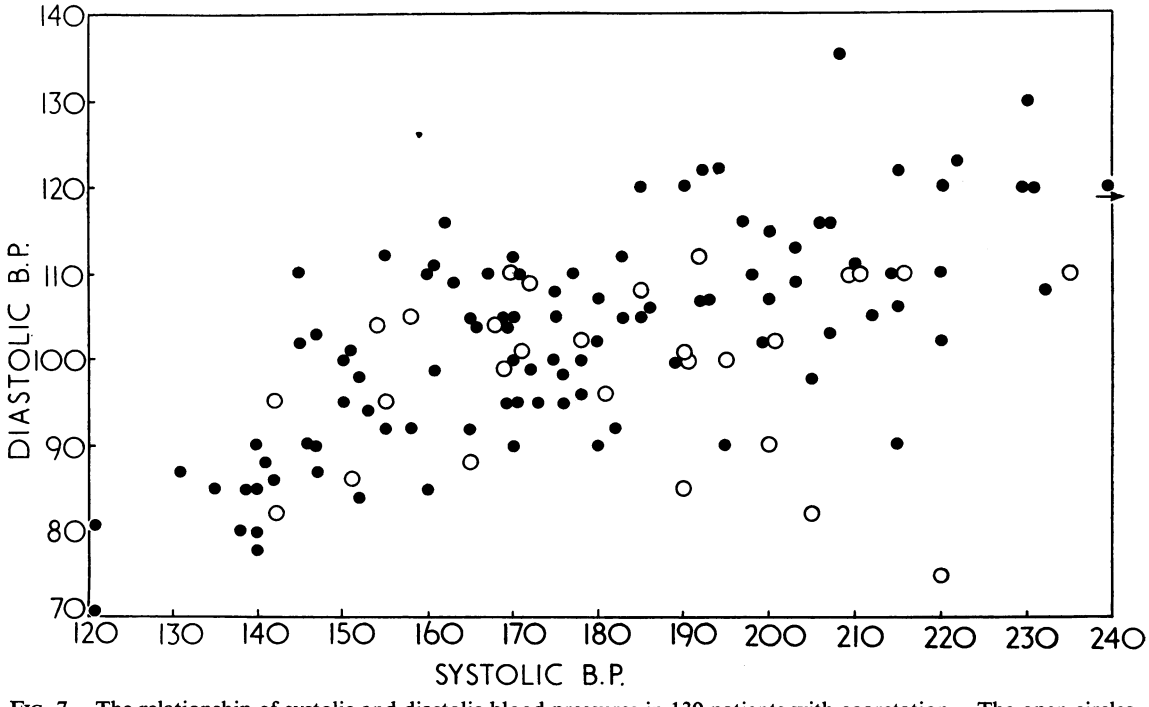


Fig. 7.—The relationship of systolic and diastolic blood pressures in 130 patients with coarctation. The open circles represent those with aortic regurgitation and, with four exceptions where the diastolic pressure was lower than usual they fall within the normal scatter, showing that aortic regurgitation in cases of coarctation generally has little influence on the level of the diastolic pressure.

aortic valves, from 25 (Abbott, 1928) to 42 per cent (Reifenstein *et al.*, 1947). It increases with increasing age; being present in 16 per cent of those under 15, in 22 per cent of those between 15 and 19, and in 27 per cent of those over 20 (see Table I). Sometimes we know when the regurgitation developed. In Case 2, it was between 16 and 21 without any immediate change in the blood pressure though it widened from 210/110 to 260/90 during the next eight years; in Case 19 between 26 and 28, the pressure changing only from 203/109 to 200/100; and in another man between 18 and 21 years of age, the pressure changing from 200/100 to 218/100.

It is, therefore, possible for atherosclerotic bicuspid valves to become incompetent early in life. This is more important than a specially high blood pressure or the effects of rheumatic infection. The way in which the diastolic murmur may at first be faint, heard at one visit and not at the next, and the age incidence are not suggestive of rheumatic disease and fit in better with bicuspid valves gradually becoming incompetent.

When patients with coarctation have aortic valve disease, it has often been attributed to a rheumatic infection both by clinicians and by pathologists. In our view, however, this is uncommon. Among the 130 patients there are only 8 who gave a history of rheumatic fever. It is true that four of these had aortic regurgitation and a fifth had pure aortic stenosis but this was thought to be congenital at necropsy, and there was no valvular disease in the others. Even more important there was no clinical or pathological evidence of mitral stenosis in these or in any of the patients with aortic valve disease, and it was present in only 4 per cent of the necropsies in Reifenstein's series. We cannot be certain that all our patients were asked about a past rheumatic history, but if some were omitted, we doubt the correctness of the rheumatic history in others, who were kept in bed because of a systolic murmur and not because of joint pains; and it may well be that hearing this murmur and failing to diagnose coarctation was the reason for thinking them rheumatic.

Aortic Stenosis and Coarctation

This combination has been emphasized by Taussig (1947) and by Smith and Matthews (1955) who collected 24 reported cases with necropsies; it was present in 6 of our patients, more often than would be expected by chance.

Case 24. A man, aged 31, had been getting worse for two years. The heart was large (c.t.r. 60%) and the blood pressure was 155/110 in both arms but 198/36 in the left ventricle. His coarctation was resected and his aortic valve divided, but he died four weeks later. The mitral valve was normal and the aortic valve had two large cusps and one small cusp, all much thickened and fibrosed with a large fibrous calcified mass between two of the cusps (Fig. 8).

A man, aged 21, has coarctation (B.P. 158/109; c.t.r. 54%; Fig. 5B), signs of aortic stenosis, but few symptoms. A man, aged 23, has coarctation (B.P. 153/102; c.t.r. 53%) and fairly severe aortic stenosis with some anginal pain. A man, aged 33, is still getting on well, though he has a blood pressure of 175/100, a large heart (c.t.r. 58%), and aortic stenosis with calcification of the valve (Fig. 6B). A girl, aged 9, has been described (Case 18). In the sixth (Case 1) we thought that the stenosis was rheumatic. The last five all had some aortic regurgitation as well.

Association with Other Congenital Abnormalities

With Other Cardiac Abnormalities. We have not seen many other additional lesions except in the arrangement of the aortic branches. One cyanotic girl, aged 9, had Eisenmenger's complex also (Campbell and Cardell, 1953). A girl, aged 5, was thought to have transposition of the great vessels as well. A boy, aged 16, had coarctation (B.P. 180/110), an enormous heart (c.t.r. 75%), loud systolic and diastolic murmurs at the apex, and atrial fibrillation: the diagnosis of congenital mitral incompetence (and stenosis) was supported by catheterization. A boy, aged 7, had pulmonary stenosis with a systolic gradient of 28 mm. and slight coarctation (B.P. 130/84). A girl, aged 16, with recent atrial fibrillation was found to have slight mitral stenosis, bilateral S.V.C., a pulmonary vein that drained into the right S.V.C., and slight coarctation (B.P. 130/85). Two patients, one with Fallot's tetralogy (Case 15, Brinton and Campbell, 1953), and one with a patent ductus (Case 23, Campbell, 1955), were found at necropsy to have a trivial coarctation. These seven have not been included.

The following have been included as coarctation was the main lesion. One man, aged 23, who had a successful resection, had a loud systolic murmur and thrill in the pulmonary area and a pressure gradient

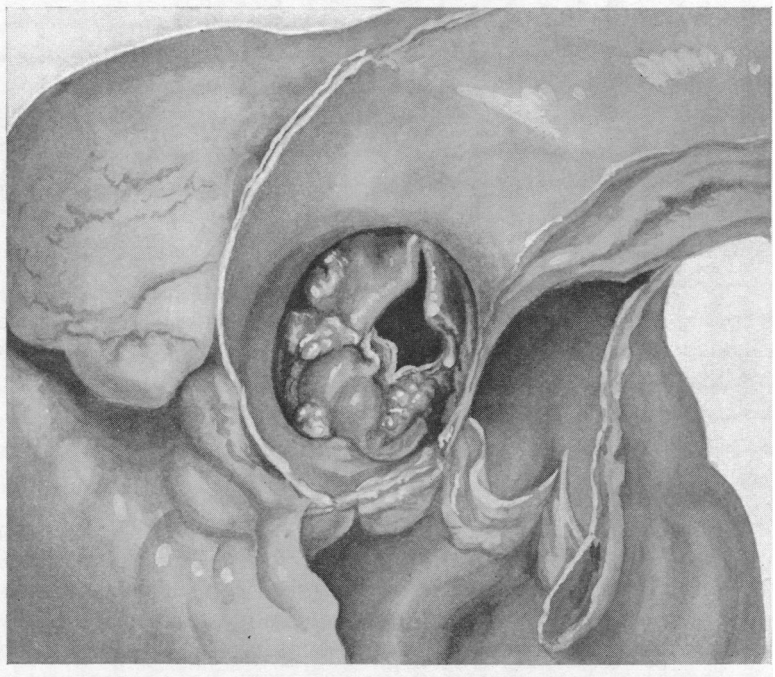


FIG. 8.—A heavily calcified and stenotic aortic valve that had been divided three weeks before his death, from a man, aged 31, with coarctation. Case 24.

of 11 mm. across the pulmonary valve—hardly enough by itself to prove stenosis, but probably enough with the physical signs. Wood (1950) has reported one patient with coarctation and a ventricular septal defect. We think this combination is uncommon: it was suspected in three of our patients, but catheterization showed no abnormality in two, while in the third a pulmonary vein drained into the superior vena cava.

We have seen only three patients with a patent ductus in addition, although this is found in 10 per cent of some series. In one boy, aged 2, a blood pressure of 140/70 was accepted as due to a large patent ductus, which from the site of the murmur was thought to be on the right side: when he was 7 it was heard best on the left, the pressure had risen to 175/105, and coarctation was recognized.

With Turner's Syndrome. Turner (1938) described the syndrome of a webbed neck, cubitus valgus, and infantilism—a sub-variety of ovarian agenesis, now more commonly known as gonadal dysplasia (Polani *et al.*, 1956). Albright *et al.* (1942) first drew attention to the association of coarctation with this syndrome. Three of our four apparently female patients with Turner's syndrome are included in a series of gonadal dysplasia where the chromatin sex has been determined by Polani *et al.* (1953) on skin, blood, or both, and all three are chromosomal males. Of the 54 patients with gonadal dysplasia investigated by Polani (personal communication) or recorded by Grumbach *et al.* (1955) 49 have male and 5 female chromosomal sex: webbing of the neck was present in 31 and coarctation in 12 of these. All those with coarctation and gonadal dysplasia were chromosomal males and had also a third abnormality, notably webbing of the neck.

There were not many with *other non-cardiac congenital defects.* One girl had bilateral cervical ribs, and one man had had a successful operation for hypospadias. One girl had lymphatic swelling of the legs and she had ptsosis of the left eye-lid also, and another had bilateral nerve deafness. Nor have we often found a family history of other defects. The mother of one patient had a patent ductus and the daughter of another had pulmonary valve stenosis, and since writing this we have seen the sister of one of our patients also with coarctation. Bonham-Carter informs us that he has seen two siblings, both with coarctation, a patent ductus, and an aberrant subclavian artery.

THE SIZE OF THE HEART

Bonnet (1903) pointed out that the heart might be of normal size: it may remain so over many years even when the blood pressure is very high (Lewis, 1933). Brown (1950) stated that it might

be normal in size or slightly enlarged, or sometimes when there was aortic regurgitation greatly enlarged.

It is surprising how many patients have small vertical hearts, though generally there is some fulness of the left ventricle on radioscopy. The large well-developed chest is only part of the explanation. Some increase is quite common but large hearts are uncommon. In considering size we have omitted 8 cases with complications: six had aortic stenosis and in a seventh this was suggested by the left ventricular strain but other evidence supported mitral incompetence, while the eighth had recurrent heart failure in infancy (Case 22). The heart was only moderately large in two of these (c.t.r. 53–54%) but large in three (c.t.r. 58–61%) and enormous in three (c.t.r. 68–71%).

There were 128 other patients, a few who had been followed for ten years being included in two decades. Most have normal or even small vertical hearts, nearly two-thirds having cardiothoracic ratios of 50 per cent or less, nearly half of 48 per cent or less, and nearly one-third of 46 per cent or less (Table IV). Age alone did not have a great influence though the smaller hearts were less common in those over thirty: they were found in patients with very high blood pressure and in others with aortic regurgitation but not when this was severe.

TABLE IV

THE SIZE OF THE HEART IN COARCTATION OF THE AORTA

Age in years	No. of cases	Percentages with cardiothoracic ratios of			
		48 and less	49–52	53–56	57 and over
0–19	73	50	34	16	0
20–29..	36	52	22	17	9
30 and over	19	26	32	32	10
Total	128	47	30	19	4

At the other extreme there were only 14 (11%) with ratios of 55 per cent or more, and in 9 of these it was only 55 or 56 per cent: 3 of these 9 were over 30 and 4 of the 6 under 30 had aortic regurgitation. Of the 5 with larger hearts (c.t.r. 57% or more), 4 had aortic regurgitation: one developed a larger heart (c.t.r. 51% at 22 to 57% at 29 years, with a blood pressure of 220/75): one woman, aged 39, with a large heart (c.t.r. 57%) was just as well after five years with no further increase; the third (c.t.r. 61%) died two years later with a ruptured aorta (Case 16); and the fourth, aged 32, with the largest heart (c.t.r. 66%) and free aortic regurgitation died a year later (Case 20). In the fifth his congestive failure and large heart (c.t.r. 58%) had followed bacterial endocarditis (Case 23).

Few patients showed any increase while under observation, but this happened in two, both with aortic regurgitation. One has just been mentioned and in the other it increased between 15 and 20 years of age (c.t.r. 49 to 53%), although he seemed as well with no change in the blood pressure, 165/89: he showed unusual dilatation of his descending aorta (Fig. 3C). Both these patients should have had an operation when they were first seen, but at that time we regarded aortic regurgitation as a contra-indication.

It is clear that most patients continue to have normal hearts for many years in spite of the high blood pressure, sometimes even with the lesser degrees of aortic regurgitation. Sometimes, after thirty, the heart may become large even without any regurgitation.

ELECTROCARDIOGRAPHIC CHANGES

Lewis (1933) stated that there was generally left axis deviation but 4 of his 6 cases were over 40: Campbell and Suzman (1947) were surprised to find a normal axis as common as left axis deviation. Brown (1950) says " There are no characteristic changes in the electrocardiogram which is often

physiological. Ultimately there is the picture of left ventricular hypertrophy." We thought these views were due to the use of standard leads only but they are confirmed by chest leads. We have omitted the same 8 patients, mostly with aortic stenosis, all of whom had some, often severe, left ventricular strain.

Among the other 120 patients, more than one-quarter had normal electrocardiograms, often in spite of a high blood pressure for a long time, but this was uncommon when there was aortic regurgitation as well. Half had some left ventricular preponderance, and nearly one-quarter had some strain, but generally no more than a little S–T depression or minimal T inversion in leads V5, V6, or V7 (Fig. 9).

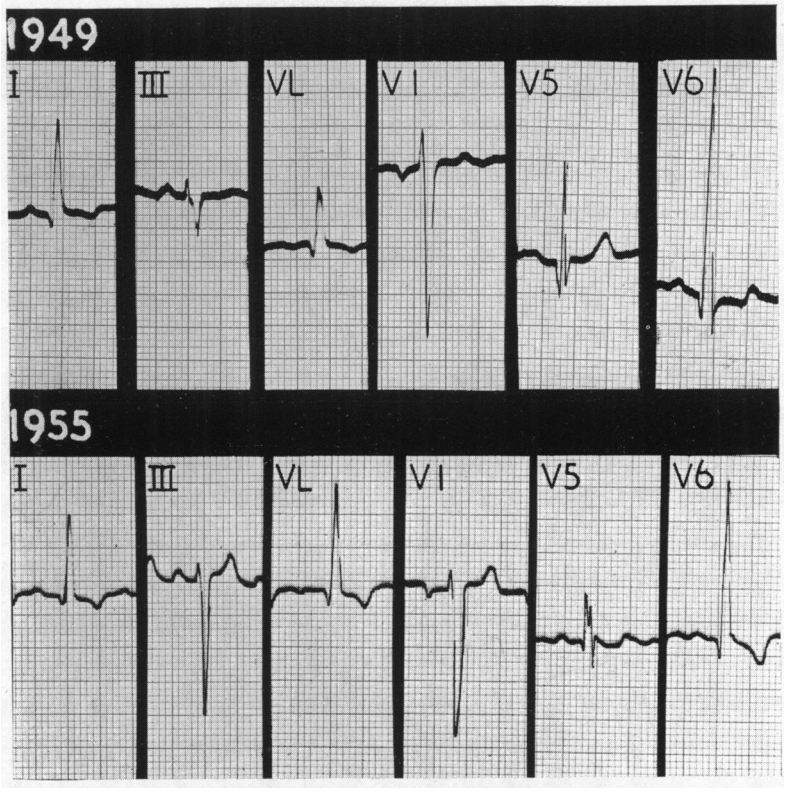


FIG. 9.—Increasing left ventricular strain in a patient aged 22, where the heart had also become larger. (A) In 1949 with slight S–T depression and a late-rising T wave in V6. (B) In 1955, with a biphasic instead of a large upright T in V5 and with moderately deep T inversion in V6.

We had diagnosed preponderance on a prominent S in V1 and a prominent R in V6 without exact measurements, but this is a lower criterion than is generally accepted. If S in V1+R in V6 must be 35 mm. for the diagnosis of left ventricular preponderance the number would be much less, though S in V2 was often larger than in V1 and would bring more cases up to the required standard.

Slight left ventricular strain may occur from the second decade onwards, but is much more common in patients over thirty, when normal cardiograms are less common. Slight aortic regurgitation did not have much influence, but as it became more severe it was more often associated with left ventricular strain (Fig. 9). The youngest patient with the strain pattern was 18; he was almost without symptoms but the flat T waves in V5 and V6 were one reason for advising resection and became upright after it: ventricular strain in young patients is, we think, a special indication for resection.

The Results of Surgical Treatment

Most patients with coarctation are enjoying good health with few symptoms, so an operation that carries a considerable risk is not easy to advise; yet disaster may arrive with little warning and a successful resection improves the outlook for the future. It is easier when there are serious symptoms or an unusually high blood pressure, or when the patient is young and the risk less. But the diagnosis is still made late in many patients, sometimes only when symptoms are severe or even when there is heart failure. We have come to think that operation should be advised in most children, that after the age of 8 if not sooner there should be no delay, and that early aortic regurgitation is an added reason, for the natural prognosis seems less favourable than we had supposed. Crafoord (1955) rarely recommends surgery before 5 or 10 years, but thinks it should be urged for all patients under 20, advised for most between 20 and 30, but needs careful consideration after that age. He, too, looks on aortic regurgitation as an indication, particularly when the heart is not greatly enlarged.

During the last eight years 46 patients with coarctation have been operated on, all except two by Sir Russell Brock at Guy's Hospital. Ten were between 10 and 14, nine between 15 and 19, and nine between 20 and 24 years. Most of them (60%) were, therefore, between 10 and 24 years of age, but eleven were older and seven were younger. Clearly they were not all of the ideal age and this will be a problem for some time. Most of them had been under observation before operation, sometimes for several years.

Operative Mortality. Seven of the 46 patients died but 3 of these were among the 9 admitted directly under Sir Russell. It seems fair to exclude these three since two had recurrent congestive failure (Cases 22 and 23) and in the third the coarctation could not be resected at a previous thoracotomy (Case 25), and if so, the mortality was 9 per cent. In 648 resections that we have reviewed the mortality was 7 per cent, but many authors, including ourselves, think that it is now about 5 per cent in cases without complications: Gross (1953) has reported only two deaths in his last 100 cases.

Four of the deaths were particularly unfortunate as the operation seemed successful, but one died from fulminating enteritis soon after it (Case 28), one from hæmorrhage through a branch of the graft (Case 27), one from subarachnoid hæmorrhage following a rise of blood pressure to 200 the day after it (Case 26), and one from an aneurysm at the anastomosis associated with a staphylococcal septicæmia (Case 24). The last two occurrences have been reported in other series and suggest the need for prophylactic antibiotics and for hypotensive drugs after operation. Three of these were young patients, aged 17, 14, and 13, so the risks are not limited to older patients. It is disappointing, for there were no deaths among the first 18 patients operated on from 1948–53.

Among the 45 deaths in the 648 resections reviewed, 35 seemed to be in cases without complications. The cause of death was given in 23 of these; in 9 it was hæmorrhage or cardiac arrest at or just after operation, in 10 late hæmorrhage from the suture line (infection being proved in four of these), in 2 cerebral hæmorrhage, in 1 cerebral damage, and in 1 mesenteric necrotizing arteritis. Intracranial hæmorrhage that was not fatal complicated three cases and paralysis of the legs three others. One late death from bacterial endocarditis has been recorded and aortic regurgitation has progressed in one boy of 15 in spite of a successful resection (O'Sullivan and Steinberg, 1953).

Results in this Series. In two early cases, no resection was possible but one of these may prove operable now that the use of grafts is more developed. In fact, grafts were used in 10 of the 46, but this is a higher proportion than usual.

Subsequent discussion is based on our first 30 patients who survived after resection. Four have been followed for from five to seven years, 9 for three or four years, 9 for two years, 6 for one year, and 2 for less than this. The blood pressure was always greatly reduced but not always to normal levels. The result during the first year was generally a reliable indication of the subsequent course and only in one patient has the pressure tended to rise during six years though not nearly to the original level. Only one patient failed to get a good result: his blood pressure fell but two months later rose to near the previous level and he then returned to Africa and has not been traced.

The final results will need a long follow-up for there must still be some risk of endocarditis on bicuspid aortic valves and of rupture of the aorta or of a berry aneurysm even if these are much reduced by the lower blood pressure. Berry aneurysms occur at sites of congenital weakness of the arterial wall and it is reasonable to expect that both their development and their rupture will be reduced by resection at an early age. The site of anastomosis and the graft itself, when this has been used, may not always remain satisfactory. So far, however, there have been no untoward episodes in these patients.

Sometimes the good result can be assessed by relief of symptoms but many patients felt so well and were so active that improvement in symptoms could not be expected. Several of them, however, felt better and some were obviously so. Two have returned to work that they had been forced to give up, and another is working much harder and yet is less aware of symptoms even though he had and has aortic regurgitation.

One example of an excellent result follows. A woman, aged 28, had been well till she was 25 but since then had found her work in a fruit shop as well as housework increasingly difficult because of headaches. Five years later she is doing these easily and looking after her small son, all without symptoms. Her blood pressure has fallen from 207/103 to 130/81 and her cardiogram is more normal. Her renal function tests, done two years after operation because there had been some albuminuria, were completely normal.

Changes in Blood Pressure. Apart from a long follow-up the success of the operation in safeguarding the patient's future can be judged best by the changes in the pressure. These are difficult to assess because of their variability but we have taken the average of many readings often over several years, and think that they give an accurate picture (Table V). Generally the systolic blood pressure stabilizes about 40 mm. lower and the diastolic about 20 mm. lower than it was, but there is, of course, great variation. One man still has a diastolic pressure of 101 mm. but as his systolic

TABLE V

LEVEL OF BLOOD PRESSURE BEFORE AND AFTER OPERATION FOR COARCTATION

	Before		After		Reduction	
	Systolic	Diastolic	Systolic	Diastolic	Systolic	Diastolic
5 patients with the highest systolic pressures	207	115	148	90	59	25
Next 5 patients	197	109	144	87	53	22
Next 5 patients	185	112	142	87	43	25
Next 5 patients	173	104	143	84	30	20
Next 5 patients	162	102	127	78	35	24
5 children aged 3–13 with the lowest systolic pressures ..	146	88	117	75	29	13
Average of all patients ..	178	105	137	83	41	21

has fallen from 207 to 156 the result was counted as good. No other ptaient had a diastolic pressure over 95 mm. though two were at this level 6 and 4 years after. There were ten others where it was about 90 mm.

Most studies of patients after resection have reported the blood pressure as normal or reduced without much detail, though Hallenbeck *et al.* (1951) says that it was below 130/90 in 49 per cent, Gross (1953) says that it was below 140, with corresponding reductions for children, in 88 per cent, and Haxton and Milnes Walker (1954) report an average fall of 47/24 mm. in 25 cases. A more detailed study by Counihan (1956) finds that although the brachial blood pressure is much reduced it rarely falls to normal. By chance, the average pressure before operation was 178/105 in our series and 177/99 in his, but after it our figure was 137/83 against his 157/87. It is, of course, easy

to record the lower readings and neglect the higher ones, but we took them under the same conditions before and after operation, and they are about 10 mm. higher than the average figures obtained in the ward after the operation.

There has been a tendency to assume that when the pressure does not fall to normal, the surgeon has not relieved the obstruction completely, but Counihan doubts this and found no precise relationship between the fall of blood pressure and the apparent increase in the aortic diameter at the anastomosis.

Aortic Regurgitation. This was present in 8 of the 46 patients and did not seem to affect the result. At one time we thought aortic regurgitation a contra-indication but have come to look on it as an indication, for successful operation may delay its progress unless it is too far advanced. In one the diastolic murmur has become more difficult to hear.

The Size of the Heart after Operation. The heart is often of normal size and rarely very large, and those who have had operation have been within the usual range. In three of those who died, however, the heart was very large (c.t.r. 58–71%). In 21 of the 30 who survived with a completed resection it was not enlarged (c.t.r. 40–50) and remained just the same, and in 9 with larger hearts (c.t.r. 52–56%) the average cardiothoracic ratio fell from 53·4 to 52·4 per cent. Most of the larger hearts, however, were in adults where reduction in size—absolute as opposed to relative size in a growing child—must be hard to obtain and if obtainable must take time.

Electrocardiographic Changes. Of the 30 patients who had a completed resection, 11 had some left ventricular strain, generally no more than T waves that were flat or biphasic or slightly inverted in leads V5 and V6, and sometimes this was one of the reasons for advising operation. All these and another 14 had left ventricular preponderance, but 5 had not even this.

Generally the signs of left ventricular strain were lessened by successful operation, and in 6 they were almost or completely abolished (Fig. 10). Sometimes but less constantly the left ventricular preponderance also diminished.

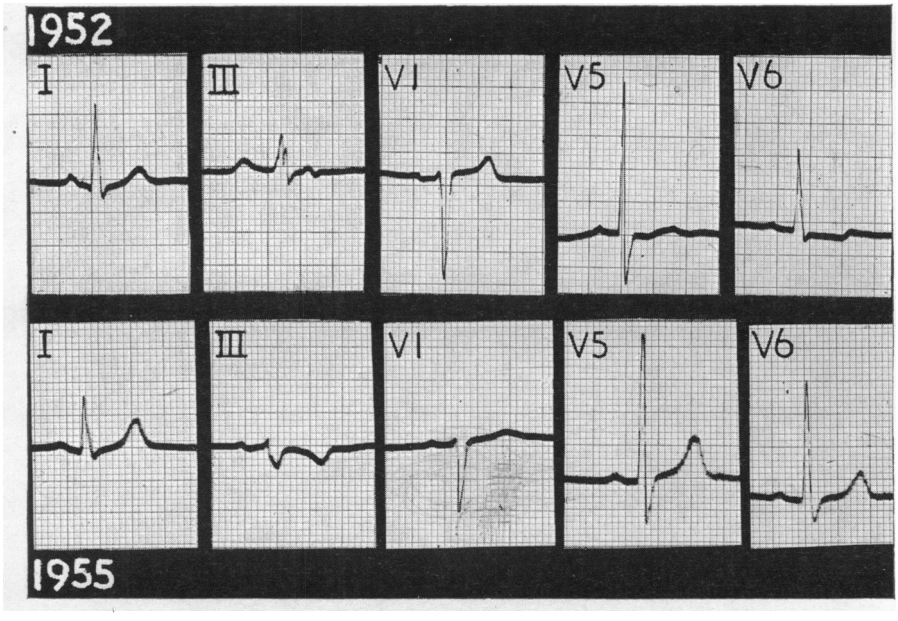


FIG. 10.—The disappearance of slight left ventricular strain after a successful operation. The slight S–T depression and small T waves in V5 and V6 have been replaced by large upright T waves. (A) Before operation when he was 18. (B) Three years after.

Summary and Conclusions

We have tried to summarize the progress of 130 patients with coarctation of the aorta, 80 of whom have been followed for an average period of five years. One-quarter of them have aortic regurgitation, generally from atherosclerotic changes on bicuspid valves, the percentage increasing as they get older. Generally the regurgitation remains slight for many years with the diastolic pressure still over 100, but becomes more serious as time goes on.

About 5 per cent have aortic stenosis, more often we think congenital than atherosclerotic. Patent ductus arteriosus was not common in our series and simple ventricular septal defect was not seen. Other congenital abnormalities are not common but occur.

Most patients get on well for the first two decades, though the blood pressure rises from an average of 145/90 at 5 years to 190/105 at 17 years of age or so, and then remains steady. The heart is often of normal size and rarely becomes very large unless there is free aortic regurgitation. Left ventricular preponderance is present in less than half the electrocardiograms and much left ventricular strain is rare in the absence of aortic stenosis, though slight degrees of this may suggest that a patient is not doing well.

In spite of the satisfactory progress of most patients, six who were in good health when they were first seen died suddenly or relatively so, most often in the third decade. We think, therefore, that most children should have surgical resection unless the evidence suggests the coarctation is trivial. Early aortic regurgitation is a further indication for it is likely to get worse unless arrested.

The results of surgical treatment are generally good. In our first 30 patients surviving with a completed resection all but one had much lower blood pressures and the average fall was 41 mm. for the systolic and 21 mm. for the diastolic pressure. Those who had disabling symptoms were much improved.

We are greatly indebted to Sir Russell Brock for asking us to see several of these patients, and still more for his work in making possible the last section on surgical treatment. We should like to thank Dr. Charles Baker and Dr. Ralph Kauntze for allowing us to include some of their patients: also Miss Waldron for Fig. 4, 7, and 8, and Mr. Engel of the Photographic Dept., Guy's Hospital, for the photographs.

One of us was Medical Registrar to Dr. John Fawcett for several years and does not remember that he ever mentioned coarctation though he had written an excellent paper on the subject in 1905: evidently he thought of it as a condition that was diagnosed at necropsy. It is interesting that 4 of his 18 cases had bicuspid valves ("two valves had coalesced") and 2 (including one with bicuspid values) had aortic stenosis.

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* The 648 resections that are reviewed are taken from these papers.